



# Cardiology in the Young

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## Cardiac Dysrhythmias and Electrophysiology

### O-01

#### Appropriate therapy for implantable cardioverter defibrillators used as primary versus secondary prophylaxis: A danish pediatric population

Jani Thuraiayah, Annette Schophuus Jensen, Berit Philbert, Lars Sondergaard, Christian Jons

Department of Cardiology, The Heart Centre, Rigshospitalet, Copenhagen University Hospital, Blegdamsvej 9, Copenhagen 2100, Denmark

**Background and Aim:** Implantable cardioverter defibrillators (ICD) are used for primary prevention in children highly suspicious of life-threatening arrhythmias due to family history and malignant syncope. This practice might potentially result in a lower incidence of appropriate therapy compared with ICDs implanted for secondary prevention following documented ventricular arrhythmias. We aimed to compare the rate of appropriate therapy after ICD implantation for primary versus secondary prevention in young children.

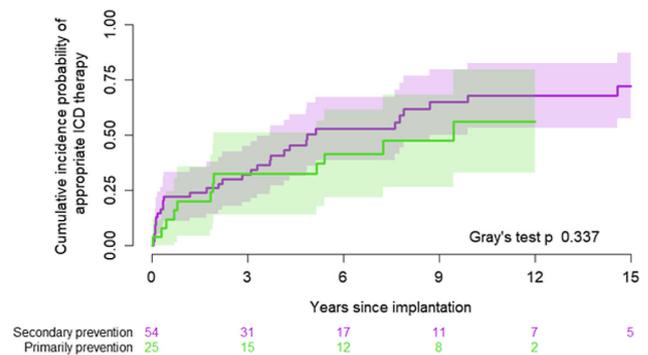
**Method:** Using Danish ICD registry, this retrospective nationwide cohort study included children aged  $\leq 15$  years who received their first ICD between 1988 and 2020. Patient characteristics, medical history including mortality, device indication, therapy and complications were retrieved from electronic medical files. The primary endpoint was time to appropriate therapy, defined as shock or antitachycardia pacing for ventricular tachycardia or fibrillation.

**Results:** During the study period, a total of 81 ICDs (39 ICDs per million live births) were implanted. Among these, 79 were included in the outcome analysis due to retrievable follow-up data. At baseline, the majority had channelopathies (44%) or structural heart diseases (42%), whereas 25 (32%) and 54 (68%) devices were implanted as primary and secondary prophylaxis, respectively. The median age at primary implantation was 13.9 and 11.6 years ( $p < 0.05$ ), respectively. During a median follow-up of 9.0 (IQR: 4.8–13.9) years, 44 patients experienced appropriate device therapy and 6 died, with no difference for primary and secondary prevention recipients ( $p = 0.34$  and  $p = 0.83$ ). The 10-year cumulative incidence of appropriate therapy was 56% for primary prevention recipients and 68% for secondary prevention recipients, whereas 10-year survival probability was 90% (95% CI: 76–100%) and 81% (95% CI: 63%–99%), respectively. All deaths

were of cardiovascular cause. Bleeding occurred in 0 versus 2, infection in 5 versus 4, and inappropriate shock therapy in 2 versus 11 patients, with an overall combined trend towards more events for ICDs implanted as secondary prophylaxis ( $p = 0.08$ ).

**Conclusions:** ICD implantation in young children was relatively rare. The rate of appropriate therapy was similar among primary and secondary ICD recipients, indicating too restrictive primary prophylaxis implantation, which should be investigated in future studies. The incidence of complications was highest among secondary ICD recipients.

**Keywords:** Appropriate ICD therapy, Pediatrics, Complications, Implantable cardioverter-defibrillator



Time to appropriate device therapy (appropriate antitachycardia pacing or appropriate shock therapy) in the cohort based on indication

### O-02

#### Myocardial work in ventricular preexcitation

Tereza Zmatlíková, Olena Iurchenko, Jan Kovanda, Viktor Tomek, Peter Kubuš, Terezia Tavačová, Jan Janoušek  
Children's Heart Centre, 2nd Faculty of Medicine, Charles University in Prague and Motol University Hospital, Prague, Czech Republic

**Background and Aim:** Ventricular preexcitation alters the electromechanical activation sequence and may induce left ventricular (LV) contractile discoordination and pathologic remodelling in selected cases.

**Aim:** To evaluate LV myocardial work before and after accessory pathway (AP) ablation in patients with WPW preexcitation and to compare them to normal controls.

**Method:** Mechanical activation mapping and myocardial work calculation was performed in 11 paediatric patients with the WPW syndrome/pattern and left sided or paraseptal APs undergoing catheter ablation (successful in 10/11) and 8 healthy age-matched controls using speckle tracking echocardiography and the GE ECHOPAC version 204.57.0.745 software with the integrated AFI module. An 18-segmental echocardiographic LV model was used. Global LV myocardial work efficiency (GWE) was calculated as the ratio of global constructive work to the sum of global constructive and global wasted work (GWW). The time from delta wave onset to R wave peak in lead V6 (delta to R) was used to quantify the degree of preexcitation.

**Results:** Before ablation GWW was higher (164 vs 102 mmHg%,  $P = 0.015$ ) and GWE lower (median 91 vs 94%,  $P = 0.010$ ) in patients vs controls. After successful ablation GWW decreased from 164 to 93 mmHg% ( $P = 0.006$ ) and GWE increased from 91.1 to 94.0 % ( $P = 0.006$ ). GWE tended to correlate negatively with the delta to R interval ( $R = 0.59$ ,  $P = 0.056$ ). Before ablation, segmental WE increased with distance from ventricular AP insertion from median 69.5 % at AP location to 98.0 % in distant segments ( $P < 0.001$ ). After ablation, WE was equally distributed among LV segments. Mechanical activation mapping localized APs with a median (IQR) segmental distance of 0 (0–1) segments from the EP mapped location.

**Conclusions:** Ventricular preexcitation induces significant LV myocardial work inefficiency in segments adjacent to AP which correlates with the degree of preexcitation and is normalized after ablation. The amount of wasted work in asymptomatic preexcitation may be considered when assessing the potential for pathologic LV remodelling or discussing the indication for prophylactic ablation. Mechanical activation mapping may be used to localize manifest APs.

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**Keywords:** myocardial work, preexcitation, wolff-parkinson-white syndrome, speckle tracking echocardiography, accessory pathway, ablation

### O-03

#### Ventricular arrhythmias in primary benign cardiac tumors in children: anatomical and histological predictors in a long term follow up

Silvia Placidi, Giulio Calcagni, Camilla Calvieri, Stella Maiolo, Corrado Di Mambro, Massimo Stefano Silveti, Sonia Albanese, Lorenzo Galletti, Fabrizio Drago

Department of Pediatric Cardiology and Cardiac Surgery, Bambino Gesù Children's Hospital and Research Institute, Rome, Italy

**Background and Aim:** Cardiac primary tumors in children are extremely rare and incidence of arrhythmias depends mostly by histological type. Predisposing factors are still widely debated. Our aim was to assess predictors of ventricular tachycardia occurrence in a large cohort of children.

**Method:** We retrospectively enrolled 102 children <18 years old with primary benign cardiac tumor referred to Bambino Gesù Children Hospital between 1980 and 2014.

Data about the type, location and dimensions of the cardiac tumor were obtained. Clinical data, electrocardiograms (ECG), Holter monitoring and medical therapy for each patient were collected from hospital records.

**Results:** Among the 102 cardiac tumors with a median age of the arrhythmic presentation of 52 months (IQR 7–132), there were 78 rhabdomyoma, 13 fibroma, 5 mixoma and 6 others (1 lipoma,

2 angioma and 3 teratoma). During a mean follow up of 15 years (median 14, IQR 8–19 years), 44 of the 102 (43%) children presented clinically significant arrhythmias. Fourteen (14%) had supraventricular arrhythmias, which were sustained in 8. Twenty-seven patients (27%) had ventricular arrhythmias, which were sustained in 16. Two patients showed ventricular pre-excitation and one (1%) a complete AV block.

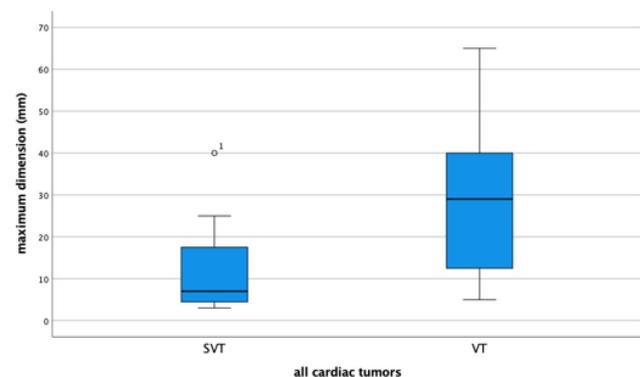
Two patients underwent cardiac transplant (2%) and 9 PMK or ICD implantation (8, 8%).

Six patients died (5, 9%), in 3 cases due to arrhythmias, in other 3 cases, death occurred without any cardiac cause documented.

Biventricular sided primary cardiac tumors showed an increased occurrence of sustained arrhythmias if compared to those only left or right sided ( $p = 0.019$ ). Moreover, cardiac tumors with dimension >15 mm were significantly associated with occurrence of ventricular arrhythmias if compared to those <15 mm ( $p = 0.033$ ). Among rhabdomyomas, those presenting ventricular arrhythmias had greater dimensions if compared with those showing supraventricular arrhythmias ( $p = 0.006$ ). At Cox regression analysis, adjusted for antiarrhythmic therapy, fibroma remained independently associated with ventricular sustained tachycardia occurrence during follow up. (H.R. 4.238 95% CI 1.386– 12.959,  $p = 0.011$ ).

**Conclusions:** Anatomic characteristics, biventricular side and dimensions above 15 mm seem to be associated with ventricular arrhythmias occurrence. Among histological types, cardiac fibroma seems to be independently associated to ventricular sustained tachycardia occurrence during follow up.

**Keywords:** Cardiac primary tumors, ventricular arrhythmias, risk stratification



Type of arrhythmias according to maximum dimension of cardiac tumors

### O-04

#### The clinical course of asymptomatic children with RYR2-mediated catecholaminergic polymorphic ventricular tachycardia

Puck J. Peltenburg<sup>1</sup>, Sing Yap<sup>20</sup>, Isabelle Denjoy<sup>16</sup>, Guillermo J. Perez<sup>15</sup>, Martijn Bos<sup>14</sup>, Vincent Probst<sup>2</sup>, Heikki Swan<sup>3</sup>, Esther Zorio<sup>4</sup>, Georgia Sarquella Brugada<sup>5</sup>, Kristina Haugaa<sup>6</sup>, Moniek Cox<sup>7</sup>, Jonathan R Skinner<sup>8</sup>, Peter Schwartz<sup>9</sup>, Minoru Horie<sup>10</sup>, Annika Rydberg<sup>11</sup>, Shubhayan Sanatani<sup>12</sup>, Jan Till<sup>13</sup>, Michael J Ackerman<sup>14</sup>, Ramon Brugada<sup>15</sup>, Antoine E Leenhardt<sup>16</sup>, Janneke A. E. Kammeraad<sup>17</sup>, Arthur A. M. Wilde<sup>18</sup>, Nico A. Blom<sup>19</sup>, Sally Ann B. Clur<sup>1</sup>, Christian Van Der Werf<sup>18</sup>

<sup>1</sup>Department of Pediatric Cardiology, AmsterdamUMC, Amsterdam, the Netherlands; <sup>2</sup>Université de Nantes, CHU Nantes, CNRS, INSERM, l'institut du thorax, France; <sup>3</sup>Heart and Lung Centre, Helsinki University

Hospital and Helsinki University, Finland; <sup>4</sup>Department of Cardiology, Hospital Universitario y Politécnico La Fe, Valencia, Spain; <sup>5</sup>Pediatric Arrhythmias, Inherited Cardiac Diseases and Sudden Death Unit, Hospital Sant Joan de Déu, Spain; Medical Science Department, School of Medicine, Universitat de Girona, Spain; <sup>6</sup>Department of Cardiology, ProCardio Center for Innovation, Oslo University Hospital, Rikshospitalet, Norway; <sup>7</sup>Department of Cardiology, University of Groningen, University Medical Centre Groningen, The Netherlands; <sup>8</sup>Cardiac Inherited Disease Group New Zealand, Green Lane Paediatric and Congenital Cardiac Services, Starship Children's Hospital, Auckland, New Zealand; <sup>9</sup>Istituto Auxologico Italiano, IRCCS, Center for Cardiac Arrhythmias of Genetic Origin, Milan, Italy; <sup>10</sup>Department of Cardiovascular Medicine, Shiga University of Medical Science, Otsu, Japan; <sup>11</sup>Department of Pediatric Cardiology, Umea University, Umea, Sweden; <sup>12</sup>BC Children's Hospital, Vancouver, Canada. Department of Pediatrics, University of British Columbia, Vancouver, Canada.; <sup>13</sup>Department of Cardiology, Royal Brompton Hospital, London, United Kingdom; <sup>14</sup>Departments of Cardiovascular Medicine, Pediatric and Adolescent Medicine, and Molecular Pharmacology & Experimental Therapeutics, Division of Heart Rhythm Services and Pediatric Cardiology, Windland Smith Rice Sudden Death Genomics Laboratory, Mayo Clinic, Rochester, United States; <sup>15</sup>Pediatric Arrhythmias, Inherited Cardiac Diseases and Sudden Death Unit, Hospital Sant Joan de Déu, Spain, Medical Science Department, School of Medicine, Universitat de Girona, Spain, Spain; <sup>16</sup>Service de cardiologie - Rythmologie CHU Paris Nord-Val de Seine - Hôpital Xavier Bichat-Claude Bernard, Paris, France; <sup>17</sup>Department of Pediatric Cardiology, Erasmus MC - Sophia, Rotterdam, The Netherlands; <sup>18</sup>Amsterdam UMC, University of Amsterdam, Heart Centre, Department of Clinical and Experimental Cardiology, Amsterdam Cardiovascular Sciences, The Netherlands; <sup>19</sup>Department of Pediatric Cardiology, Willem-Alexander Children's Hospital, Leiden University Medical Centre, the Netherlands; <sup>20</sup>Department of Cardiology, Erasmus MC, Rotterdam, The Netherlands

**Background and Aim:** Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rare, potentially lethal inherited cardiac arrhythmia, associated with *RYR2* variants. Data on asymptomatic children with a *RYR2* variant, a growing population due to cascade screening, are lacking. We aimed to assess the age at CPVT phenotype development and the incidence of symptoms in these children.

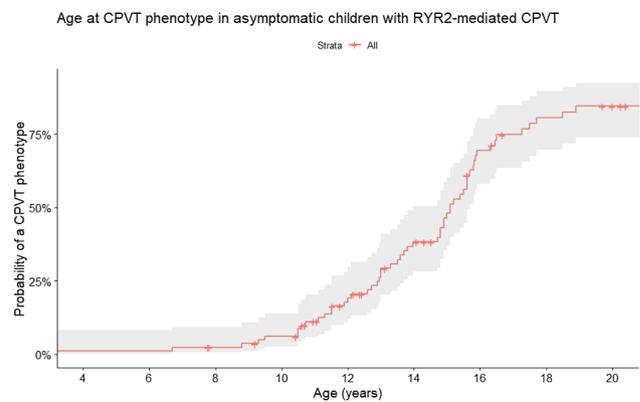
**Method:** All children with a *RYR2* (likely) pathogenic variant from the International CPVT Registry who were asymptomatic (defined as absence of syncope or sudden cardiac arrest) were included if an exercise-stress test (EST) or Holter off anti-arrhythmic drugs at baseline was available. A CPVT phenotype was defined as the presence of any ventricular ectopy on EST or couplet or worse on Holter. Data on symptoms were collected and all ESTs during follow-up of phenotype-negative children at baseline were assessed.

**Results:** One-hundred-twenty-one children (median age at baseline 10.6 [IQR: 6.9-13.8] years, 61 (50.4%) female, 91 (75.2%) ESTs at baseline) from 51 families with one of 44 different *RYR2* variants were included and followed-up for a median duration of 7.0 [IQR: 4.7-11.4] years. At baseline, 42 (34.7%) children were phenotype-positive. Subsequently 64 (52.9%) children developed a phenotype, including 34 (53.1%) on beta-blocker therapy. The median age at start of beta-blocker was 12 [IQR: 9-15] years and 27 children (22.3%) never received an anti-arrhythmic drug. When censored at the start of beta-blockers, 48% (95% CI: 37-61%) of the children developed a phenotype by 15 years of age (Figure). Five children had a syncope, of whom three were phenotype-positive, four used a  $\beta$ 1-selective beta-blocker, and in three non-adherence was confirmed. All three phenotype-positive children had recurrent symptoms, partly

related to non-adherence; syncope in two and an appropriate implantable cardioverter-defibrillator shock in one. Two children (1.6%) had an aborted cardiac arrest. Both were phenotype-positive and used a  $\beta$ 1-selective beta-blocker and flecainide, one confirmed non-adherence.

**Conclusions:** The majority of asymptomatic children with *RYR2*-mediated CPVT developed a phenotype around 15 years of age. The incidence of symptoms was relatively low and symptoms were mostly preceded by the development of a phenotype and related to non-adherence. This information could guide when to initiate therapy in this population.

**Keywords:** catecholaminergic polymorphic ventricular tachycardia, ryanodine, phenotype, exercise-stress-test



CPVT phenotype development over age

## O-05

### Initial experience of left bundle branch pacing in paediatric patients. the future is here

Ferran Roses Noguer<sup>1</sup>, Oscar Cano<sup>4</sup>, Jaume Francisco<sup>2</sup>, Alba Santos<sup>2</sup>, Andrea Bueno<sup>1</sup>, Joaquin Osca<sup>4</sup>, Ferran Gran<sup>1</sup>, Bea Insa<sup>3</sup>, Begoña Benito<sup>2</sup>, Jordi Perez<sup>2</sup>, Núria Rivas<sup>2</sup>

<sup>1</sup>Department of Paediatric Cardiology, Vall d'Hebron University Hospital, Barcelona, Spain; Department of Paediatric Cardiology, Royal Brompton Hospital, London, UK; GuardHeart European Reference Network; <sup>2</sup>Department of Cardiology, Vall d'Hebron University Hospital, Barcelona, Spain; <sup>3</sup>Department of Pediatric Cardiology, Hospital Universitario La Fe, Valencia, Spain; <sup>4</sup>Department of Cardiology, Hospital Universitario La Fe, Valencia, Spain

**Background and Aim:** Left bundle branch pacing is a new pacing technique that prevents RV pacing-induced left dysfunction, and it has been proven helpful in adult patients.

**Method:** Prospective multicentric study to describe the initial experience of the first 36 consecutive paediatric patients in Spain treated with left bundle branch pacing since December 2019 at the Hospital de la Vall d'Hebron and at the Hospital de la Fe in Valencia.

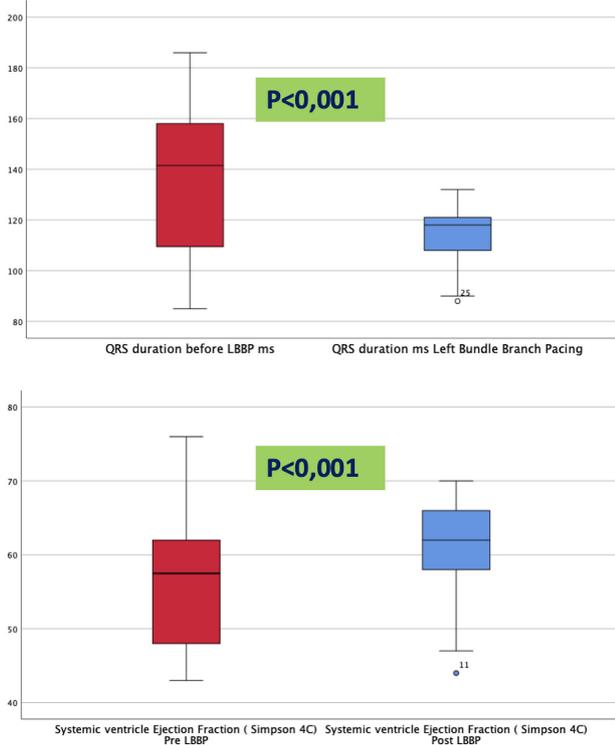
**Results:** Thirty-six patients (26 females) were included, with a median age of 11, 9 years (3, 5 years – 17, 6 years) and a median weight of 41 kg (minimum 16 – maximum 70). The diagnosis was congenital atrioventricular block (AVB) in 18 patients, AVB post-surgery of a CHD in 14 patients and 4 patients with sinus node dysfunction. 33 had a systemic Left Ventricle, and 3 had a systemic RV. 21 patients had a previous pacemaker (16 epicardial and 5 TV). The procedure was performed under general anaesthesia, guided by fluoroscopy and 3D transesophageal ultrasound. In

34 patients, the implant was through the left axillary vein, and only 2 cases were implanted through the right side. There were no complications during implantation or after a median follow-up time of 1, 6 years (1 month – 4, 17 years). The duration of the QRS was reduced from 142 ms (IQR 158–111) to 118 ( IQR 121–116)  $p < 0.001$ , and the systemic ventricle ejection fraction increased from 56% (IQR 48–62) to 63 ( IQR 58–66)  $p < 0.001$

**Conclusions:** Left bundle branch pacing is a safe and feasible technique in paediatric patients when performed in expert centres. The technique significantly reduces the duration of the QRS and improves the systemic ventricle EF and should be considered the implant technique of choice in paediatrics when choosing a transvenous implant.

**Keywords:** Left Bundle Branch pacing, Paediatric cardiology

Figure 1. QRS duration and SV Ejection Fraction before and after LBBP.



## O-06

### Molecular genetic testing in children and adolescents with inherited arrhythmia syndromes – results of the german multicenter cogia study

Lea Lippert<sup>1</sup>, Tobias Burkard<sup>1</sup>, Franziska Markel<sup>2</sup>, Volker Debus<sup>3</sup>, Fridrike Stute<sup>5</sup>, Anna Kamphues<sup>6</sup>, Robert Dalla Pozza<sup>6</sup>, Stefan Rupp<sup>9</sup>, Maria Gonzalez Y Gonzalez<sup>9</sup>, Claudia Junge<sup>10</sup>, Majed Kanaan<sup>11</sup>, Kirstin Hoff<sup>12</sup>, Anja Hanser<sup>13</sup>, Sven Dittmann<sup>4</sup>, Dominik Sebastian Westphal<sup>15</sup>, Sebastian Clauß<sup>7</sup>, Marc Phillip Hitz<sup>12</sup>, Kolja Becker<sup>12</sup>, Stefan Kääb<sup>8</sup>, Peter Ewert<sup>1</sup>, Michael Hofbeck<sup>13</sup>, Wolfgang Wällisch<sup>14</sup>, Sven Dittrich<sup>14</sup>, Anselm Uebing<sup>12</sup>, Jan-Hendrik Nürnberg<sup>16</sup>, Joachim Hebe<sup>16</sup>, Rainer Kozlik-Feldmann<sup>5</sup>, Philipp Beerbaum<sup>10</sup>, Gunter Kerst<sup>11</sup>, Hans-Gerd Kehl<sup>3</sup>, Eric Schulze-Bahr<sup>4</sup>, Roman Gebauer<sup>2</sup>, Gabriele Hessling<sup>1</sup>, Cordula Maria Wolf<sup>1</sup>

<sup>1</sup>German Heart Center Munich, Technical University Munich, Germany; <sup>2</sup>German Heart Center Leipzig, Leipzig University, Leipzig, Germany; <sup>3</sup>University Hospital Münster, Pediatric cardiology, Münster, Germany; <sup>4</sup>University Hospital Münster, Institute for Genetics of Heart Diseases, Münster, Germany; <sup>5</sup>University Hospital Hamburg-Eppendorf, Hamburg, Germany; <sup>6</sup>Division of Pediatric Cardiology and Intensive Care, University Hospital, LMU Munich, Munich, Germany; <sup>7</sup>Hospital Ludwig-Maximilians-University Munich, Munich, Germany; <sup>8</sup>Institute of Surgical Research at the Walter-Brendel-Centre of Experimental Medicine, University Hospital, LMU Munich, Munich, Germany; <sup>9</sup>German Center for Cardiovascular Research (DZHK), Partner Site Munich, Munich Heart Alliance (MHA), Munich, Germany; <sup>10</sup>Interfaculty Center for Endocrine and Cardiovascular Disease Network Modelling and Clinical Transfer (ICONLMU), LMU Munich, Munich, Germany; <sup>11</sup>Hospital Ludwig-Maximilians-University Munich, Munich, Germany; <sup>12</sup>German Center for Cardiovascular Research (DZHK), Partner Site Munich, Munich Heart Alliance (MHA), Munich, Germany; <sup>13</sup>Interfaculty Center for Endocrine and Cardiovascular Disease Network Modelling and Clinical Transfer (ICONLMU), LMU Munich, Munich, Germany; <sup>14</sup>University Hospital Giessen and Marburg, Giessen and Marburg, Germany; <sup>15</sup>University Hospital Hannover, Hannover, Germany; <sup>16</sup>University Hospital Aachen, Aachen, Germany; <sup>17</sup>University Hospital Schleswig-Holstein, Kiel, Germany; <sup>18</sup>University Hospital Tübingen, Tübingen, Germany; <sup>19</sup>University Hospital Erlangen, Erlangen, Germany; <sup>20</sup>Institute of Human Genetics, Klinikum rechts der Isar, School of Medicine, Technical University of Munich, Munich, Germany; <sup>21</sup>Department of Internal Medicine I, Klinikum rechts der Isar, School of Medicine, Technical University of Munich, Munich, Germany; <sup>22</sup>Electrophysiology Bremen for Nürnberg and Hebe

**Background and Aim:** Molecular genetic testing is often performed as part of the clinical work-up in children with inherited arrhythmia syndromes, such as Long QT syndrome (LQTS), Brugada syndrome (BrS) and catecholaminergic polymorphic ventricular tachycardia (CPVT).

To describe distribution of genetic variants and association with the occurrence of malignant arrhythmias and (aborted) sudden cardiac death (SCD) in children and adolescents with inherited arrhythmia syndromes.

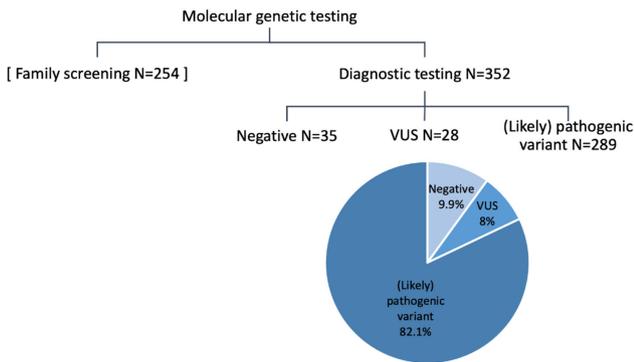
**Method:** Retrospective multicenter (12 tertiary care pediatric cardiology centers) data collection from patients with a clinical and/or molecular genetic diagnosis of an inherited arrhythmia syndrome  $\leq 18$  years. Major arrhythmic events (MAE) were defined as SCD, aborted SCD, appropriate discharge of implantable cardioverter-defibrillator (ICD) or documented sustained ventricular tachycardia (susVT). The patient group with family screening was excluded for parts of the analysis because of asymptomatic courses.

**Results:** Testing was performed as part of family cascade screening in 254 asymptomatic patients (median age 3.6 years (IQR 0.3;9.9)) and as diagnostic testing within clinical work-up in 352 patients (median age 10.8 years (IQR 5.0;14.6)). Within the latter group a (likely) pathogenic variant was identified in 82.1% (289/352), a variant of uncertain significance (VUS) in 8.0% (28/352) and a negative genetic result in 9.9% (35/352), respectively. Affected genes with (likely) pathogenic variants included KCNQ1 in 44.6% (129/289), KCNH2 in 23.5% (68/289), SCN5A in 12.8% (37/289), RYR2 in 12.5% (36/289) and other genes in 6.6% (19/289). In patients with LQT1 genetic testing was positive in 96.6% (114/118).

SCD (N = 2), aborted SCD (N = 64), appropriate ICD discharge (N = 12) and susVT (N = 15) occurred in 28.0% (81/289) of patients with a (likely) pathogenic variant, in 25.0% (7/28) of patients with a VUS and in 22.9% (8/35) of patients with a negative molecular genetic test result, respectively. The highest MAE-rate was found in patients with a RYR2-mutation (63.9%, 23/36).

**Conclusions:** Molecular genetic testing performed in children and adolescents with a clinical diagnosis of an inherited arrhythmia syndrome was positive in 82.1% and highest in LQT1, therefore adding a significant diagnostic yield within this entity of rare diseases. MAE had an almost similar distribution between patients with or without a positive genetic finding. MAE prevailed if the RYR2 gene was affected.

**Keywords:** inherited arrhythmia syndrome, sudden cardiac death, malignant arrhythmia, molecular genetic testing



Molecular genetic testing in children and adolescents with inherited arrhythmia syndromes

#### O-07

### 3D-electroanatomic mapping-guided conduction system pacing preserves electromechanical function in paediatric patients with atrioventricular block

Massimo Stefano Silvetti, Fabrizio Gimigliano, Chiara Mizzon, Fabio Anselmo Saputo, Ilaria Tamburri, Marta Campisi, Virginia Battista, Ilaria Cazzoli, Cristina Raimondo, Giacomo Silvetti, Fabrizio Drago Paediatric Cardiology and Cardiac Arrhythmia/Syncope Unit, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy - European Reference Network for Rare and Low Prevalence Complex Disease of the Heart (ERN GUARD-Heart)

**Background and Aim:** Transvenous paediatric pacing from alternative ventricular sites including conduction system pacing may prevent ventricular dyssynchrony and systolic dysfunction. These sites may be difficult to reach and require higher fluoroscopic exposures. The use of three-dimensional-electroanatomic mapping system (3D-EAM) may guide lead implantation and reduce fluoroscopy. Purpose of this study is the outcome of 3D-EAM-guided alternative sites pacing in paediatric patients.

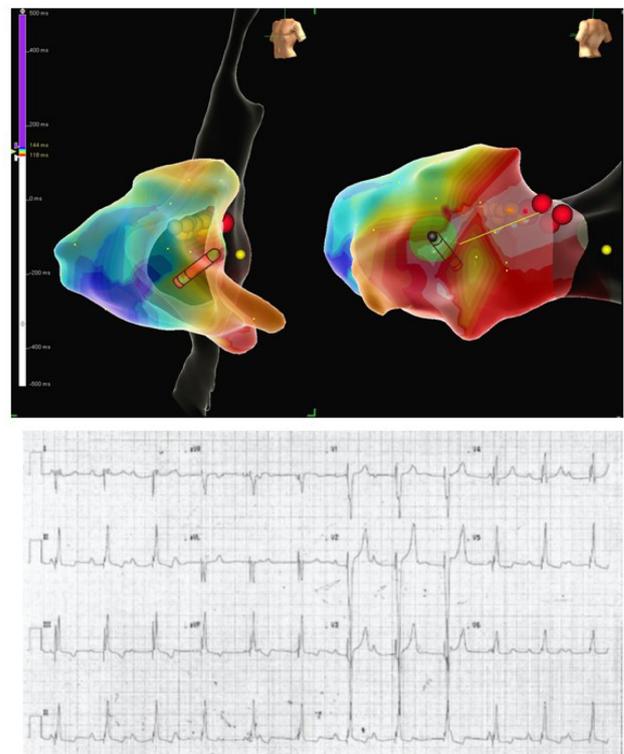
**Method:** Retrospective analysis of children and young patients with congenital or acquired complete atrioventricular block (CAVB) with or without other congenital heart defects (CHD) who underwent 3D-EAM-guided transvenous pacing in alternative sites of the subpulmonary ventricle, to perform non-selective His bundle pacing (NSHBP), pacing of ventricular septum close to conduction system (VS-CSP) or right ventricular outflow tract pacing (RVOT). 3D-pacing map guided stylet-directed screw-in lead implantation toward septal sites with narrower paced QRS. Procedure and follow-up data were recorded. Parameters of

ECG (QRS duration, systemic ventricular activation time, VAT) and of the echocardiogram of the systemic ventricle (3D ejection fraction, EF, global longitudinal strain, GLS) were registered and compared at baseline (pre-implantation) and at 3-year follow-up. Data are reported as median (25th-75th centiles).  $P < 0.05$  was significant.

**Results:** 64 patients (47 females) with CAVB, of whom 11 with CHD, underwent 3D EAM-guided pacing (31 VVIR, 33 DDD; 10 NSHBP, 5 RVOT, 49 VS-CSP) (Figure 1) at age 12 (8-15) years, weight 44 (27-57) kg. Prior pacing (RV apex, RV and LV free wall, also biventricular) was present in 27 patients. Fluoroscopy exposure was: 3.0 (1.2-5.0) mGy and 90 (33-146) microGy/m<sup>2</sup>. Baseline QRS was 85 (80-130) ms, post-implantation QRS 115 (100-120) ms ( $P = 0.002$ ), with VAT 80 (70-81) ms. In patients with prior pacing, QRS shortened: baseline QRS 130 (120-160) vs. 120 (110-125) ms post-implantation ( $P = 0.004$ ). Echographic parameters at baseline and at 3-year follow-up showed non-significant differences: EF 63 (58-69)% vs. 59 (56-63)%; GLS -22 (-28-14)% vs. -21 (-26-20)%.

**Conclusions:** 3D-EAM-guided alternative site pacing including conduction system pacing was accomplished with low fluoroscopic exposure in paediatric patients with CAVB with or without CHD and preserved electromechanical ventricular function at mid-term follow-up.

**Keywords:** cardiac pacing, paediatric age, conduction system pacing, systolic function



**Figure 1** 3d-eam right ventricular pacing map. red dots show His bundle and proximal right bundle branch. The white area shows the area with the narrowest paced QRS. The lead tip is shown. Panel below: ECG during VVIR pacing

## O-08

**Initial data from the international prospective “observational study on catheter ablation in pediatric patients” (OSCA-PED)**

Ulrich Krause<sup>1</sup>, Jan Janousek<sup>2</sup>, Peter Kubus<sup>2</sup>, Nico Blom<sup>3</sup>, Robin A. Bertels<sup>3</sup>, Rami Fogelman<sup>4</sup>, Tom Einbinder<sup>4</sup>, Janneke Kammeraad<sup>5</sup>, Sip Wijchers<sup>6</sup>, Sergio Laranjo<sup>7</sup>, Stefan Kurath Koller<sup>8</sup>, Radoslaw Pietrzak<sup>9</sup>, Thomas Paul<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology, Intensive Care Medicine and Neonatology, University Medical Center, Georg-August-University Göttingen, Germany; <sup>2</sup>Children's Heart Centre, 2nd Faculty of Medicine, Charles University in Prague and Motol University Hospital, Prague, Czech Republic; <sup>3</sup>Department of Pediatric Cardiology, Leiden University Medical Center/Amsterdam University Medical Center, The Netherlands; <sup>4</sup>The Schneider Children's Medical Center of Israel, Cardiology Institute, Petach Tiqva, Sackler School of Medicine, Tel Aviv University, Israel; <sup>5</sup>Department of Pediatric Cardiology, Erasmus MC – Sophia Children's Hospital, Rotterdam, The Netherlands; <sup>6</sup>Department of Cardiology, Erasmus MC, Rotterdam, The Netherlands; <sup>7</sup>Serviço de Cardiologia Pediátrica do Hospital de Santa Marta, Centro de Referência Nacional de Cardiopatias Congénitas, Centro Hospitalar Universitário de Lisboa Central, Lisbon, Portugal, member of the European Reference Network (ERN) GUARD-HEART; <sup>8</sup>Pediatric Cardiology, Medical University Graz, Austria; <sup>9</sup>Department of Pediatric Cardiology and General Pediatrics, Medical University of Warsaw, Poland

**Background and Aim:** Large-scale data on catheter ablation in pediatric patients is scarce. The present study was projected to generate current benchmark data on catheter ablation as technical improvements significantly impacted this treatment measure over the last decade.

**Method:** Prospective multicenter study on catheter ablation of supraventricular and ventricular substrates in pediatric patients <18 years of age.

**Results:** A total of 930 subjects from 8 European pediatric EP centers were enrolled. Mean age was 12.8 ± 3.8 years, mean body weight was 50.9 ± 19.6 kg. Congenital heart disease was present in 44/930 (4.7%). Most common indication was patients'/parents' preference (n = 686, 74%), followed by prophylactic accessory pathway ablation (n = 110, 12%). Substrates targeted were atrio-ventricular reentrant tachycardia (AVRT; n = 547, 59%), atrio-ventricular nodal reentrant tachycardia (AVNRT; n = 296, 32%), focal atrial tachycardia (FAT; n = 31, 3%), intraatrial reentrant tachycardia (IART; n = 18, 2%) and ventricular tachycardia/premature ventricular contractions (VT/PVC; n = 38, 4%). Procedural success rate was 93% for AVRT, 99.7% for AVNRT, 84% for FAT, 89% for IART and 93% for VT/PVC. Cryoenergy was used in n = 54/547 (10%) subjects with AVRT, 22/296 (7%) with AVNRT and 3/31 (10%) with FAT. Procedural success using cryoenergy was lower compared to radiofrequency ablation (RFA) in patients with AVRT (74% cryoenergy vs. 95% RFA, p<0.001). Procedural success for cryoenergy ablation of AVNRT and FAT did not differ from results obtained with RFA. Complications reported were complete AV block requiring permanent pacing in 3/930 (0.3%), transient AV block with complete recovery in 4/930 (0.4%), coronary artery injury in 2/930 (0.2%) and groin vessel injury 1/930 (0.1%). **Conclusions:** Catheter ablation of supraventricular and ventricular substrates was highly effective with lower acute procedural success for FAT and IART. Safety profile of pediatric catheter ablation was

high. Complete AV block requiring permanent pacing and injury of the coronary arteries were rare but significant. Cryoenergy was a safe alternative for ablation of substrates in close proximity to the cardiac conduction system and the coronary arteries. This higher safety profile was, at least for ablation of accessory pathways, counterbalanced by a lower success rate.

**Keywords:** catheter ablation, supra ventricular tachycardia, ventricular tachycardia, cryo-energy, multicenter

**Fetal Cardiology**

## O-09

**Use of pressure wires during fetal aortic valvuloplasty in fetuses with critical aortic stenosis**

Andreas Tulzer<sup>1</sup>, Julian Hochpöchlner<sup>1</sup>, Iris Schamreiter<sup>2</sup>, Kathrin Holzer<sup>1</sup>, Gerald Tulzer<sup>1</sup>

<sup>1</sup>Pediatric Cardiology, Children's Heart Center Linz, Linz, Austria; <sup>2</sup>Department of Prenatal Medicine, Kepler University Hospital, Linz, Austria

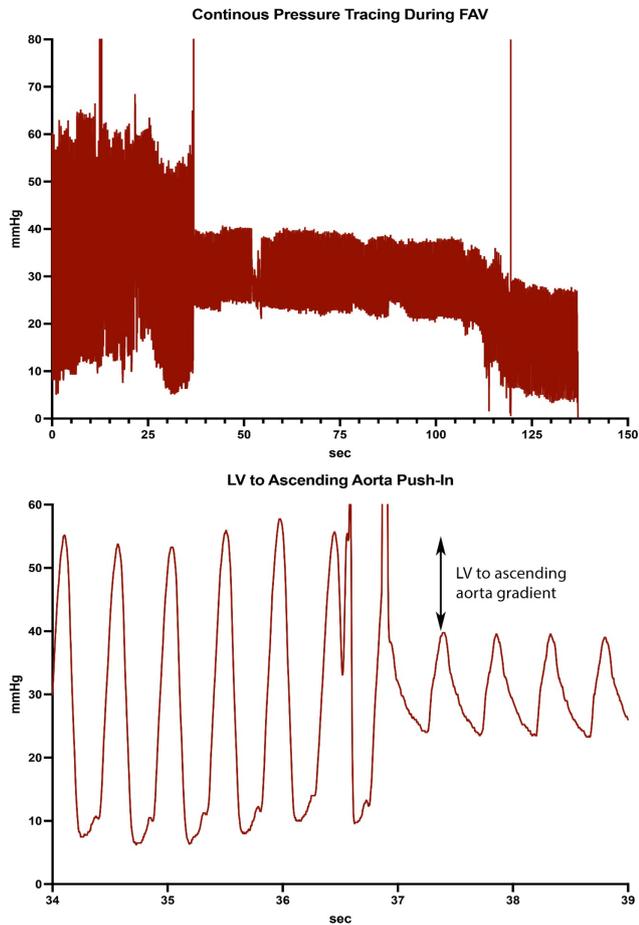
**Background and Aim:** Selection criteria of fetuses with critical aortic stenosis (CAS) for fetal aortic valvuloplasty (FAV) are still not perfect. Some ventricles show good remodeling after a successful FAV, some not. FAV still carries a 3–4% risk of procedure related mortality. The aim of this study was to characterize pre-intervention fetal left ventricular function using pressure guide-wires and to assess their utility.

**Method:** Instead of standard 0.014 inch guide wires we used 0.014 inch guide wires with a pressure transducer at its tip (Philips Volcano Verrata Plus). Patient's charts were analyzed with regard to echocardiographic parameters (Vmax of mitral regurgitation and aortic valve) and invasively acquired parameters (dp/dt, end-diastolic (EDP) and peak pressure of the LV (LVP), systolic blood pressure in the ascending aorta).

**Results:** Between September 2021 and October 2022, 12 FAV were performed in 11 fetuses using pressure guide-wires at a median gestational age of 26+4/7 weeks (22+4/7 to 30+3/7). Pressures could not be acquired in two patients (one broken wire and one technical problem). In fetuses with adequate LV pressure tracing before valve dilation, median EDP was 12.5mmHg (8–21mmHg) with a median LVP of 46 mmHg (27–53mmHg). LV dp/dt was low in all patients with a median of 366mmHg/sec (250–467mmHg/sec). LV to ascending aorta gradients were low in all patients due to poor LV function (median 9mmHg, range 3–19mmHg). Median systolic arterial blood pressure was 40mmHg (25–43mmHg). Continuous pressure monitoring allowed good information about the guidewire position even if imaging was not good.

**Conclusions:** Measuring invasive pressures during FAV using a pressure guide-wire is feasible and provides valuable information on fetal left ventricular performance. The use of pressure guide wires improves safety and may reduce procedure time due to better information about the wire position. Current technical equipment is still suboptimal, error-prone and needs improvement.

**Keywords:** Critical aortic stenosis, Fetal cardiac intervention, Invasive pressure measurements, eHLHS  
Pressure tracing in a fetus with CAS at 25+6 weeks



The upper image shows entire continuous pressure measurement during fetal aortic valvuloplasty in a fetus with critical aortic stenosis. The bottom image shows the change of pressure curves as the pressure guidewire is moved from the LV to the ascending aorta.

#### O-10 Haemodynamic effects of acute maternal hyperoxygenation in fetuses with suspected coarctation of the aorta

Milou PM van Poppel<sup>1</sup>, Johannes K Steinweg<sup>1</sup>, Reza Razavi<sup>2</sup>, John M Simpson<sup>2</sup>, Kuberan Pushparajah<sup>2</sup>, Trisha V Vigneswaran<sup>3</sup>, David FA Lloyd<sup>2</sup>

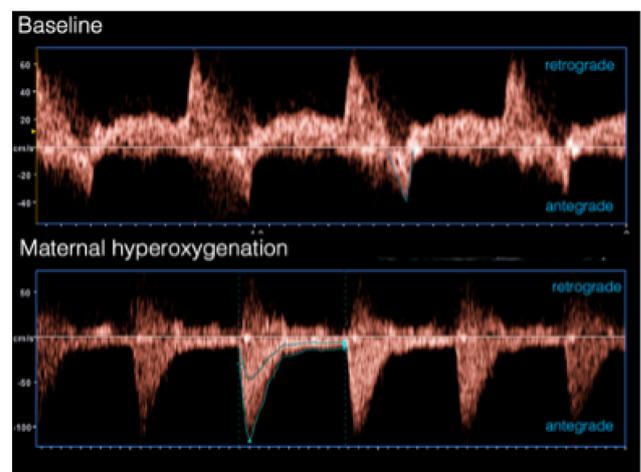
<sup>1</sup>School of Biomedical Engineering & Imaging Sciences, King's College London, London, United Kingdom; <sup>2</sup>School of Biomedical Engineering & Imaging Sciences, King's College London, London, United Kingdom; Department of Congenital heart disease, Evelina London Children's Hospital, London, United Kingdom; <sup>3</sup>Department of Congenital heart disease, Evelina London Children's Hospital, London, United Kingdom

**Background and Aim:** Acute maternal hyperoxygenation (AMH) is known to modulate the fetal circulation by reducing pulmonary vascular resistance and increasing pulmonary venous return to

the left atrium. We report the haemodynamic response to AMH in fetuses with suspected coarctation of the aorta (CoA). **Method:** Pregnant women carrying a fetus with suspected CoA were prospectively recruited for AMH with fetal cardiac MRI and echocardiography at baseline and after 15 minutes 10L/min 100% oxygen via non-rebreather face mask. Phase-contrast MRI (PC-MRI) data was acquired in the ascending aorta (AAO), superior vena cava (SVC), descending aorta, main pulmonary artery, arterial duct, and umbilical vein. Aortic isthmus (AoI) flow was derived (AAO – SVC). PC-MRI data was retrospectively gated and indexed to fetal MRI weight (ml/min/kg). Ultrasound metrics of pulmonary vasoreactivity, preload, afterload and cardiac haemodynamics were studied. Outcome was defined as neonatal CoA requiring surgical repair or false positive (FP-CoA). **Results:** Twenty-five cases were recruited at gestational age range 30–35 weeks (mean  $32.9 \pm 1.3$ ). Baseline and AMH PC-MRI measurements were obtained in 19/25 (76%). Neonatal CoA was confirmed in 5/19 (26%). Paired echocardiography was available for 13/19, pulmonary vasoreactivity to AMH was observed in all these cases. Left ventricular output was not significantly increased during AMH in either CoA or FP-CoA on MRI or ultrasound; a trend towards increased left to right flow at the foramen ovale was noted on MRI. Mean SVC flow decreased significantly during AMH in FP-CoA cases ( $138 \pm 30.9$  vs  $105.6 \pm 18.9$ ,  $p = 0.003$ ). FP-CoA cases showed a non-significant trend to increased antegrade flow in the distal aortic arch with AMH on echocardiography compared to CoA (see example Figure 1). Mean AoI flow increased significantly in both CoA and FP-CoA, from  $-14.4 \pm 47.6$  to  $7.2 \pm 43.4$  ( $p = 0.042$ ) and  $-19.4 \pm 49.2$  to  $30.2 \pm 31.6$  ( $p = 0.002$ ) respectively. No significant change in right ventricular or combined cardiac output; middle cerebral artery PI z-score or umbilical artery PI z-score with AMH were seen.

**Conclusions:** A significant increase in antegrade AoI flow was seen with AMH which was of greater magnitude in the FP-CoA group. Response of the cerebrovascular circulation may be important in explaining appearances of FP-CoA.

**Keywords:** Congenital heart disease, Coarctation, Fetal, Magnetic Resonance Imaging



**Figure 1** Haemodynamic changes to AMH as seen with fetal ultrasound in the distal aortic arch.

## O-11

**Mri nutmeg lung pattern as the useful diagnostic tool of the restrictive pulmonary venous return in fetuses with various congenital heart defects**

Monika Bekiesinska Figatowska<sup>1</sup>, Agnieszka Grzyb<sup>2</sup>, Joanna Dangel<sup>2</sup>  
<sup>1</sup>Diagnostic Imaging Department, Institute of Mother and Child, Warsaw, Poland; <sup>2</sup>Department of Perinatal Cardiology and Congenital Anomalies, Centre of Postgraduate Medical Education, Warsaw, Poland

**Background and Aim:** Usefulness of magnetic resonance imaging (MRI) in diagnosis of pulmonary lymphangiectasia in fetuses with congenital heart defects (CHD) and obstructive pulmonary venous return is quite new diagnostic tool. Doppler pulmonary venous flow is routinely checked during fetal echocardiography. It seemed to be that this method is not as effective as MRI. The aim of this study was to check usefulness of MRI in the diagnosis of severity of pulmonary venous obstruction known as nutmeg lung pattern (NLP) in fetuses with diagnosed critical CHD.

**Method:** 30 MRI were performed in 29 fetuses with CHD and Doppler signs of the obstruction of the pulmonary venous return. MRI was performed between 22 and 36 gestational weeks (GW; median 33). The first 6 fetuses underwent MRI using an old scanner (GE Signa HDxt), the remaining 23 examinations were performed with GE Signa Artist. Detailed diagnosis and findings are presented in the Table. One fetus was examined twice: at 22 and 28 GW.

**Results:** In the first 6 fetuses the lung images were inconclusive, with an indication of possible nutmeg pattern in 5 of them. In one it was false positive, whereas in the other four the image of NLP was compatible with the biphasic pulmonary venous Doppler. In a new scanner the quality of lung images in SSFSE/T2-weighted was very good and enabled diagnosis of NLP in 10 out of 30 fetuses (33.3%) and showed possible NLP in further 3 fetuses. In the fetus scanned twice, MRI showed the progression of the disease with development of NLP in the second examination after 6 weeks. In 5 fetuses with preserved diastolic flow and deep reverse A flow in the pulmonary veins MRI showed evident NLP. All cases with NLP died in utero or shortly after birth.

**Conclusions:** Our preliminary results proved that the ability to detect the NLP is dependent on instrumentation issues (hardware and sequence/protocol settings). In modern MRI scanners, it is unambiguously detected in most cases and is an important element of parental counseling and perinatal care planning.

**Keywords:** fetal MRI, nutmeg lung pattern, fetal echocardiography, obstructive pulmonary venous flow

## O-12

**Prenatal diagnosis, pregnancy and postnatal outcomes in fetuses having redundant foramen ovale flap aneurysm associate with/without heart defects**

Balaganesh Karmegaraj<sup>1</sup>, Balaji Srimurugan<sup>2</sup>, Balu Vaidyanathan<sup>3</sup>  
<sup>1</sup>Sowmi Fetal and Pediatric Heart Centre, Tirunelveli, Tamilnadu, India; Department of Paediatric Cardiology, Amrita Institute of Medical Sciences and Research Centre, Kochi, Kerala, India; <sup>2</sup>Department of Paediatric Cardiovascular Surgery, Amrita Institute of Medical Sciences And Research Centre, Kochi, Kerala, India; <sup>3</sup>Department of Paediatric Cardiology, Amrita Institute of Medical Sciences and Research Centre, Kochi, Kerala, India

**Background and Aim:** Isolated redundant foramen ovale flap aneurysm in the absence of restrictive foramen ovale is believed to be a cause for pseudocoarctation of aorta in fetus since the impediment of blood flow to the left heart can be severe, resembling the picture

of left ventricular hypoplasia with retrograde aortic flow. We present here our experience on fetuses with redundant foramen ovale flap aneurysm associated with and without congenital heart disease and its outcomes.

**Method:** Prospective study (January 2020 to August 2022). All fetuses with foramen ovale flap aneurysm (foramen ovale flap diameter to left atrial diameter > 0.65) with and without associated heart defects were included. Imaging techniques like 2D with color and 4D spatio-temporal image correlation fetal imaging were used to evaluate anomalies and its hemodynamics. The imaging, in-utero hemodynamics, pregnancy, and postnatal outcomes were presented.

**Results:** During the study period, a total of 1312 fetal echocardiography were performed. Twenty-one fetuses with redundant foramen ovale flap aneurysm were included. Out of which fourteen fetuses had isolated redundant foramen ovale flap aneurysm and seven fetuses had associated heart defects (systemic venous anomalies in four; tetralogy of Fallot in two; ventricular septal defect in one). A fetus with isolated left superior vena cava progressed to borderline left ventricle with arch hypoplasia after birth. Prenatal borderline left ventricle in fetuses with tetralogy recovered to normal left ventricle postnatally resulted in diagnostic confusion prenatally. A fetus with multiple muscular ventricular septal defects associated with absent ductus venosus developed coarctation of aorta after birth. All fetuses with isolated foramen ovale flap aneurysm were delivered with no dysmorphism and cardiac defects. Genetic evaluation performed in fetuses with associated heart defects was normal. Three out four fetuses with intracardiac defects underwent biventricular repair with good outcomes avoiding prenatal termination of pregnancy.

**Conclusions:** Though isolated redundant foramen ovale flap aneurysm carried a benign prognosis, fetuses with associated intracardiac defects resulted in diagnostic confusion specifically in decision making on biventricular repair.

**Keywords:** fetus, redundant foramen ovale flap aneurysm, congenital heart disease

## O-13

**Single ventricle journey from fetus to first procedure. A united kingdom multicentre experience**

Peter John Lillitos<sup>1</sup>, Oscar Nolan<sup>2</sup>, Daniel Cave<sup>1</sup>, Catherine Lomax<sup>2</sup>, Shuba Barwick<sup>1</sup>, James Bentham<sup>1</sup>, Anna Seale<sup>2</sup>

<sup>1</sup>Department of Congenital Cardiology, Leeds Teaching Hospitals NHS Trust, United Kingdom; <sup>2</sup>Department of Cardiology, Birmingham Women's and Children's NHS Foundation Trust, United Kingdom

**Background and Aim:** United Kingdom single ventricle (SV) palliation outcomes after first procedure are well documented. However, publication of survival from fetal diagnosis to first procedure is lacking. With view to better informing parental-fetal counselling, we aimed to define survival and examine risk-factors for non-survival, for fetal SV diagnoses to their first postnatal procedure at two large UK centres.

**Method:** Retrospective review of all SV fetal diagnoses over a 7-year period (2015 to 2021) in two large UK congenital cardiac centres: Leeds and Birmingham. SV diagnoses were defined by our group: aortic-atresia, HLHS, mitral-atresia, double-inlet-left-ventricle (DILV), pulmonary-atresia-intact-ventricular-septum (PA-IVS), tricuspid-atresia (TA), double-outlet-right-ventricle counselled as SV (DORV-SV), Ebstein's counselled as SV, unbalanced AVSD (uAVSD), other-SV and indeterminate-SV. Survival from fetal diagnosis with intention-to-treat (ITT) and attrition rate to birth and then first procedure including

biventricular pathway are reported. Multivariable data was collected to examine maternal/fetal factors predicting non-survival. **Results:** There were 668 fetal SV diagnoses with 417 (62%) electing for ITT. Of ITT, 386 (93%) were live births and 336 (81%) underwent a first procedure. Biventricular pathway was deemed possible in 5 (1%): Ebstein's 4, uAVSD 1. Fetal survival (with ITT) to first procedure or biventricular pathway for individual lesions: aortic-atresia 2/2 (100%), HLHS 129/157 (82%), mitral-atresia 8/10 (80%), DILV 34/38 (89%), PA-IVS 23/24 (96%), TA 38/43 (88%), DORV-SV 30/35 (86%), Ebstein's 10/22 (45%), uAVSD 31/45 (69%), other-SV 33/37 (89%) and indeterminate-SV 3/4 (75%). Risk factors for mortality were decreasing gestation at birth (OR 1.5, 95% CI 1.3–1.7,  $p < 0.001$ ), decreasing birthweight (OR 3.4, 95% CI 1.6–7.1,  $p = 0.001$ ), increasing maternal age (OR 1.1, 95% CI 1.0–1.1,  $p = 0.001$ ), extracardiac anomaly (OR 2.6, 95% CI 1.1–6.1,  $p = 0.02$ ) and genetic diagnosis (OR 2.6, 95% CI 1.2–5.7,  $p = 0.02$ ).

**Conclusions:** Twenty percent of ITT fetuses diagnosed with SV circulation will not reach the first interventional procedure. Risk varies according to morphological subtype. Prematurity, decreasing birthweight, increasing maternal age, and concurrent extracardiac/genetic diagnoses decreased the prospect of reaching cardiac intervention. This data will help inform parents faced with a new fetal diagnosis of SV and highlights surgical outcome data alone should not be used in fetal counselling.

**Keywords:** Single ventricle, fetal diagnosis, survival, surgery, intervention

#### O-14

##### Maternal ANTI-RO antibody levels are strongly associated with the development of immune-mediated fetal heart disease

Edgar Jaeggi<sup>1</sup>, Kulasingam Kulasingam<sup>2</sup>, Jack Chen<sup>1</sup>, Steve Fan<sup>2</sup>, Laskin Laskin<sup>3</sup>, Robert Hamilton<sup>1</sup>, Linda Hiraki<sup>1</sup>, Earl Silverman<sup>1</sup>, Lusía Sepiashvili<sup>1</sup>

<sup>1</sup>The Hospital for Sick Children, Toronto, Canada; <sup>2</sup>Toronto General Hospital, University Health Network, Toronto, Canada; <sup>3</sup>Mount Sinai Hospital, Toronto, Canada

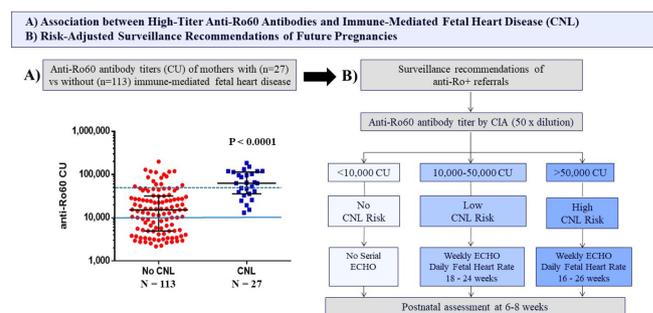
**Background and Aim:** Anti-Ro antibody-positive mothers are frequently referred for serial echocardiography due to the fetal risk of developing heart block and endocardial fibroelastosis. Little is known why only some and not all offspring develop the cardiac manifestations of neonatal lupus (CNL). This prospective study examined associations between anti-Ro antibody titers and CNL. **Method:** Antibody-positive mothers referred since 2018 for fetal echocardiography prior to CNL (group 1;  $n = 240$ ) or with CNL (group 2;  $n = 18$ ) were included. At the baseline fetal echocardiogram, maternal anti-Ro antibody titers were measured with a chemiluminescence immuno-assay (CIA; BIO-FLASH). Additional testing on diluted serum samples was used to quantify antibody titers above the analytical measuring range (AMR) of the assay (Ro60:  $\geq 1375$  chemiluminescent units (CU); Ro52:  $\geq 1685$  CU).

**Results:** In group 1, CNL occurred in 0% (0/107 referrals) with anti-Ro60 antibody titers  $< 1375$  CU and 7% (9/133) with anti-Ro60 titers  $\geq 1375$  CU. Of 122 group 1 mothers with anti-Ro60 antibody titers  $\geq 1375$  CU, event rates of CNL ( $n = 9$ ) were 0% (0/45) with titers from 1375–10,000 CU, 5% (3/56) with titers from 10,000–50,000 CU, and 29% (6/21) with titers  $> 50,000$  CU (odds ratio (OR): 13.1;  $p = 0.0008$ ). If anti-Ro52 antibody titers were also elevated  $> 10,000$  CU, the event rate of CNL increased to 55% (5/9 cases; OR 34.1;  $p < 0.0001$ ). Of group 2 referrals with CNL, 0% (0/18) had anti-Ro60 antibody titers

$< 10,000$  CU, 44% (8/18) had titers from 10,000–50,000 CU and 56% (10/18) had titers  $> 50,000$  CU.

**Conclusions:** CNL is associated with substantially higher anti-Ro antibody titers than are obtained with a standard immunoassay. By performing end-point dilutions of patient sera, we were able to significantly extend the measuring range of antibody assays and determine a cutoff (Ro60  $< 10,000$  CU) that is predictive of a pregnancy outcome without CNL and no need of serial fetal echocardiograms (Figure). For cases with higher Ro60 titers, we found significant correlations between event rates of fetal CNL and the amount of maternal anti-Ro antibodies. Serum dilution to quantitate anti-Ro antibody titers should therefore be included in any risk-based surveillance and management strategies of pregnant women with autoantibodies.

**Keywords:** Antibodies, Autoimmune Disease, Immunoassay, Fetal, Neonatal Lupus, Heart Block



A) shows associations between immune-mediated fetal heart disease (CNL) with maternal anti-Ro antibody titers by CIA. CNL is associated with significantly higher anti-Ro60 antibody titers when compared with unaffected pregnancies. Straight line shows cutoff (Ro60 CIA:  $< 10,000$  CU) below which no CNL was observed while dotted line shows threshold associated a high CNL risk (Ro60 CIA:  $> 50,000$  CU). B) Serum dilution to quantitate anti-Ro60 titers should be included in any risk-based surveillance and management strategies of pregnant women with autoantibodies

#### O-15

##### Prenatal diagnosis, associated findings, and early postnatal outcomes in fetuses with congenitally corrected transposition of the great arteries.

Maria Kavga, Adeolu Banjoko, Esther Poole, John Stickley, Tariq Desai, Paul Miller, Michael Harris, Natasha Khan, Anna Seale  
 Heart Unit, Birmingham Women's and Children's Hospital, NHS foundation Trust, Birmingham, United Kingdom

**Background and Aim:** Congenitally corrected transposition of the great arteries (ccTGA) is a rare malformation with diverse morphology. There is limited evidence about the morphology and outcomes of prenatally diagnosed patients with ccTGA. We assessed features of fetuses with ccTGA and evaluated neonatal outcomes.

**Method:** Retrospective review of fetuses with ccTGA at Birmingham Women's and Children's Hospital born from 2005 to 2020. Pre- and postnatal imaging was reviewed, morphology and outcomes were assessed.

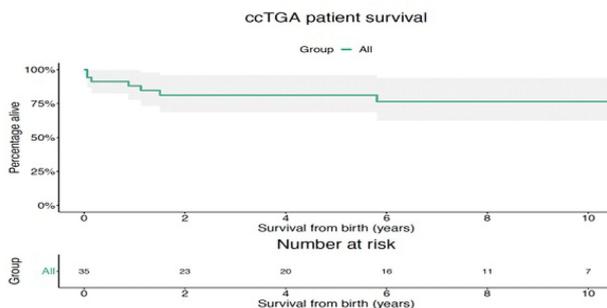
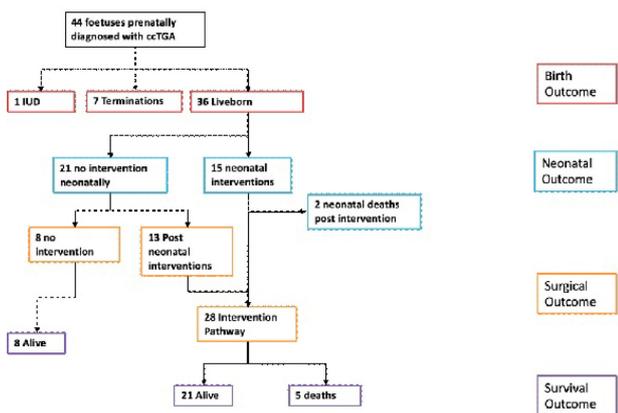
**Results:** Of forty-four fetuses identified, ten had unavailable prenatal data and one was postnatally diagnosed with isomerism. Isolated

ccTGA without associated cardiac anomalies (Group I) was found in 24.2% (8/33), ccTGA with large VSD (Group II) in 30.3% (10/33), ccTGA with pulmonary obstruction (Group III) in 18.1% (6/33) and ccTGA with complex anomalies (Group IV, including right ventricular hypoplasia, straddling tricuspid valve or aortic coarctation) in 27.2% (9/33). Tricuspid regurgitation (TR) was observed in 10/33 (30.3%) and heart block in 4/33 (12.1%) prenatally. Six (13.6%) underwent genetic testing with only one abnormality detected (duplication of chromosome 20). Six extra-cardiac anomalies were reported prenatally and postnatally. Pregnancy was discontinued in seven, of which four were in Group IV and three had severe TR. There was one intrauterine death (Group IV with TR). Therefore, there were thirty-six live-born. Coarctation of the aorta was found in five postnatally but not suspected prenatally, in one pulmonary stenosis was underestimated; otherwise prenatally morphology was confirmed. Cardiac intervention was performed in 28/36 liveborn (77.7%) with 15/36 (41.7%) undergoing neonatal intervention. (Figure 1a).

Overall, seven died of which four were in groups III and IV. Five of the seven had heart block, four diagnosed prenatally. Four had prenatal severe TR. Estimated survival at 1 and 5 years was 87.8% and 80.5% respectively. (Figure 1b)

**Conclusions:** Accurate prenatal diagnosis of ccTGA is critical for appropriate counselling. Careful evaluation of the aortic arch is challenging and recommended. Early outcomes of our study are favourable with 75% undergoing surgery in early life. Fetuses with prenatal diagnosis of complex associated abnormalities, heart block and tricuspid regurgitation appear to do less well.

**Keywords:** prenatal, diagnosis, morphology, outcome



**Figure 1.** 1a. Outcomes of 44 Fetuses Prenatally Diagnosed with ccTGA - 1b. Kaplan Meier Survival Curve for liveborn cohort. Survival at 6 years is 75%

**O-16**

**Early diagnosis of tuberous sclerosis complex is a prenatal diagnosis**

Monika Bekiesinska Figatowska<sup>1</sup>, Paulina Sobieraj<sup>1</sup>, Monika Pasieczna<sup>2</sup>, Joanna Dangel<sup>3</sup>

<sup>1</sup>Diagnostic Imaging Department, Institute of Mother and Child, Warsaw, Poland; <sup>2</sup>Second Department of Obstetrics and Gynecology, Medical University of Warsaw, Warsaw, Poland; <sup>3</sup>Department of Perinatal Cardiology and Congenital Anomalies, Centre of Postgraduate Medical Education, Warsaw, Poland

**Background and Aim:** Strong emphasis is placed on early (first 4 post-natal months) detection of tuberous sclerosis complex (TSC) and introduction of antiepileptic treatment before seizure onset in order to improve neurodevelopmental outcome of affected children. Up to 90% of children with cardiac rhabdomyomas have TSC, at least 50% of children with TSC have cardiac rhabdomyomas, and almost 100% of fetuses with multiple rhabdomyomas have TSC. Rhabdomyomas are detected during fetal ultrasound and constitute major diagnostic TSC sign. Prenatal magnetic resonance imaging (MRI) allows detection of cerebral major TSC manifestations: cortical tubers (CT), subependymal nodules (SEN), subependymal giant cell astrocytomas (SEGA). The aim of this study was to check the importance of fetal MRI in diagnosis of TSC in fetuses with cardiac tumors.

**Method:** We retrospectively reviewed 48 fetuses with cardiac tumors detected during fetal echocardiography between 19 and 36 GW (median 31) and referred for MRI to confirm/rule out TSC: 4 with one cardiac tumor, 28 with 2 or more cardiac tumors, in 16 number of cardiac tumors was unclear. MRI was performed at 24–37 GW (median: 34).

**Results:** In 11 fetuses (22.9%) cardiac tumor(s) remained the only TSC criterion. In remaining 37 cases (77.1%) TSC was diagnosed based on at least 2 major criteria, first of which was cardiac tumor(s) detected during fetal echocardiography. SENs were detected on MRI in all cases (37/37 = 100.0%). In 6 (6/37 = 16.2%) SEGAs were diagnosed based on their location near foramen of Monro and size larger than 10 mm. Cortical tubers/transmantle sign were found in 24 (24/37 = 64.9%).

**Conclusions:** Proper care of children with TSC should be established during perinatal period. It is necessary to emphasize education of women to report on mandatory sonographic examinations during pregnancy, quality of US and need to refer pregnant patients to MRI if cardiac tumor is depicted on fetal ECHO. All specialists who are involved in prenatal diagnosis – obstetricians, pediatric cardiologists and radiologists performing prenatal MRI are clue for early diagnosis of TSC – before seizures occurred and though many patients can win race against time thanks to prenatal imaging.

**Keywords:** fetal MRI, tuberous sclerosis complex, fetal cardiac tumors, fetal echocardiography, subependymal nodules, subependymal giant cell astrocytomas

**Neurodevelopment and psychosocial care**

**O-17**

**Impact of electric seizures and aeg background pattern during cardiac surgery.**

Debora Cañizo Vazquez<sup>1</sup>, Marta Perez<sup>2</sup>, Alba Rivas Piorno<sup>1</sup>, Lucia Rossi<sup>2</sup>, Joan Sanchez De Toledo<sup>2</sup>, Marta Camprubi Camprubi<sup>1</sup>

<sup>1</sup>BCNatal, Barcelona Center for Maternal Fetal and Neonatal Medicine, Hospital Sant Joan de Déu and Hospital Clinic; <sup>2</sup>Pediatric Cardiology Department. Sant Joan de Déu Hospital, Barcelona, Spain

**Background and Aim:** Perioperative brain injury is common in young infants undergoing cardiac surgery (CS). We aimed to determine the relationship between intra-surgical electrical seizures and the background pattern of amplitude-integrated electroencephalography (aEEG) with brain damage biomarkers and 2-year neurodevelopmental outcome in infants undergoing CS.

**Method:** A total of 120 newborn infants undergoing CS underwent aEEG monitoring during surgery. Seizure activity and aEEG background pattern were recorded and analyzed. Brain damage biomarkers (protein s100b and neuronal specific enolase (NSE)) were analyzed immediately after surgery and 72 hours post. Survivors underwent neurodevelopmental outcome assessment using the Bayley Scales at 2 years.

**Results:** Sixty-one per cent of the patients were operated with less than one month. Cardiopulmonary bypass was used in 58.47%. Intraoperative electrical seizures occurred in 22.87%. Seizures were more frequent in surgeries with higher Aristoteles score ( $p = 0.001$ ). Those patients with seizures presented higher levels of s100b immediately after surgery ( $p = 0.0019$ ). Mean time of seizure burden was 30 min [9.6–71.4]. Seizure burden was correlated with time of aortic clamps ( $\rho = 0.60$ ;  $p < 0.05$ ) and temperature ( $\rho = -0.32$ ;  $p = 0.02$ ). Those surgeries that require Aortic clamp presented worse aEEG patterns. There is no correlation between seizures or aEEG pattern and Bayley scores at 2 years.

**Conclusions:** Patients with seizures presented higher levels of S100B. Aortic Clamp and hypothermia are the main risks for intra-operative seizures and abnormal background patterns. Intraoperative seizures were common in this cohort of infants but did not impact on 2-year neurodevelopmental outcome. Post-surgical monitorization will be important to determine the impact of the recovery of the aEEG patterns on the long-term outcome.

**Keywords:** neuromonitoring, brain injury, electroencephalogram

## Nursing and Allied Health Professionals

### O-18

#### My transition, my care: providing individualised congenital heart disease transition care.

Danielle Poole

Department of Paediatric Cardiology, Royal Brompton and Harefield Hospitals Part of Guys and St Thomas' NHS Foundation Trust, London

**Background and Aim:** Congenital Heart Disease (CHD) is the most common birth defect and 90% of children born with CHD will survive into adulthood. Transition describes the practical process of supporting young people in the move from children's to adult's healthcare services. The published standards and guidelines for transition care outline the core themes of transitional care for young people with CHD. Collectively there is a strong emphasis on providing individualised and developmentally appropriate transition care. This quality improvement (QI) project aimed to identify the transition priorities of young people aged 12–21 and explore the potential differences in priorities according to stage of adolescence (early, middle, late) and classification of CHD (mild, moderate, severe).

**Method:** Through PDSA (plan, do, study, act) cycles a clinic proforma was developed to identify the transition priorities of young people. This was utilised in all CHD transition clinics between April and July 2022.

**Results:** 88% ( $n = 206$ ) of young people attending clinic completed the proforma. Young people prioritised 0–12 transition themes

(mean 3.39, SD 2.1). 'Exercise' was the most popular transition theme for discussion and was identified by 38% of young people. Followed by 'diagnosis and previous surgeries' which was identified by 31% and 'differences between paediatric adult care' which was identified by 29%. Chi-square analysis identified a statistically significant difference between stages of adolescence and prioritising; 'my future healthcare plan' ( $\chi^2 = 6.2227$   $p = 0.044541$ ), 'symptoms' ( $\chi^2 = 11.7074$   $p = 0.002869$ ), 'education and employment' ( $\chi^2 = 5.998$   $p = 0.049837$ ), 'reproductive health' ( $\chi^2 = 14.5661$   $p = 0.000687$ ), 'alcohol, smoking and drugs' ( $\chi^2 = 17.6659$   $p = 0.000146$ ) and 'extreme sports and rollercoasters' ( $\chi^2 = 8.1851$   $p = 0.016696$ ). Interestingly, disease classification did not influence transition priorities.

**Conclusions:** This QI project has transformed patient care within the CHD transition clinic. Care has progressed from an integrated care pathway to a patient-led model of transition where young people identify their priorities for their consultation by using the clinic proforma. The results offer novel findings to inform patient care, which places importance on stage of adolescence.

**Keywords:** congenital heart disease, adult congenital heart disease, transition, quality improvement

### O-19

#### Psychological quality of life in mothers of children with congenital heart disease: A longitudinal study from hospital discharge to adolescence

Melanie Ehrler<sup>1</sup>, Corina Wettach<sup>1</sup>, Ingrid Beck<sup>2</sup>, Emanuela R.

Valsangiacomo Buechel<sup>3</sup>, Markus A. Landolt<sup>4</sup>

<sup>1</sup>Child Development Center, University Children's Hospital Zurich, Zurich, Switzerland; <sup>2</sup>Child Development Center, University Children's Hospital Zurich, Zurich, Switzerland and Department of Pediatrics, Ostschweizer Kinderspital, St. Gallen, Switzerland; <sup>3</sup>Division of Cardiology, Pediatric Heart Center, University Children's Hospital Zurich, Zurich, Switzerland; <sup>4</sup>Department of Psychosomatics and Psychiatry, University Children's Hospital Zurich, Zurich, Switzerland and Division of Child and Adolescent Psychology, Department of Psychology, University of Zurich, Zurich, Switzerland

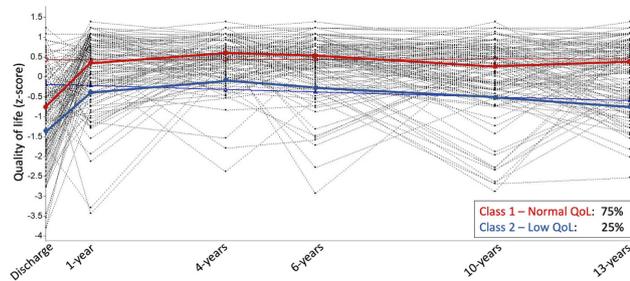
**Background and Aim:** Having a child with complex congenital heart disease (cCHD) may affect parental mental health. We investigated the long-term trajectories of psychological quality of life (QoL) in mothers of a child with cCHD and examined risk factors for persistent low QoL.

**Method:** 125 mothers of children with cCHD filled in a standardized questionnaire on QoL after the child's open-heart surgery and when the child was 1, 4, 6, 10, and 13 years old. A z-score for QoL was calculated using Swiss norms. Latent class growth analysis (LCGA) identified subgroups of mothers with regards to their QoL-trajectories over time. Regression analysis investigated predictors for chronically low QoL.

**Results:** Compared to Swiss norms, mothers of children with cCHD had significantly lower QoL immediately after open-heart surgery ( $\beta = -0.30$ ). Subsequently, QoL improved to a normal level ( $\beta = 0.05$  to  $0.27$ ). LCGA revealed two distinct groups of QoL trajectories: A group with normal QoL (75% of mothers, mean  $z = -0.76$  to  $0.62$ ) and a group with chronically low QoL (25% of mothers, mean  $z = -1.32$  to  $-0.10$ ). Chronically low QoL was associated with poorer social support (OR = 3.39,  $p = 0.008$ ) but not with parental education (OR = 1.08,  $p = 0.533$ ), migration status (OR = 0.55,  $p = 0.257$ ), number of open-heart surgeries (OR = 0.78,  $p = 0.432$ ) having a child with an univentricular CHD (OR = 0.39,  $p = 0.258$ ) or with low IQ (OR = 0.98,  $p = 0.260$ ).

**Conclusions:** Mothers of a child with cCHD are at risk for low QoL when their child undergoes open-heart surgery. One in four mothers continues to have chronically low QoL throughout their child's development, especially those mothers with poor social support. Family-based approaches are needed to support these mothers in order to reinforce both, parental well-being and child development in the long-term.

**Keywords:** Quality of life, mental health, parents, mothers, longitudinal study, well-being  
Trajectories of QoL in mothers of a child with cCHD. Groups identified by LCGA.



Latent class growth analysis revealed two classes of QoL in mothers of children with cCHD. Black dashed lines = individual trajectories. Bold red line = average trajectory of mothers of the "normal QoL" class. Thin red line = linear regression line of mothers of the "normal QoL" class. Bold blue line = average trajectory of mothers of the "low QoL" class. Thin blue line = linear regression line of mothers of the "low QoL" class. Missing data was imputed by chained equations.

## O-20

### Impact of postoperative necrotizing enterocolitis on neurodevelopmental outcome – the swiss neurodevelopmental outcome registry for children with chd

Walter Knirsch<sup>1</sup>, Alexandra De Silvestro<sup>1</sup>, Verena Rathke<sup>1</sup>, Christelle L'ebrel<sup>2</sup>, Julia Natterer<sup>3</sup>, Juliane Schneider<sup>4</sup>, Nicole Sekarski<sup>5</sup>, Beatrice Latal<sup>6</sup>, Cristina Borradori Tolsa<sup>7</sup>, Maya Bouhabib<sup>8</sup>, Katharina Fuhrer Kradolfer<sup>9</sup>, Martin Glöckler<sup>10</sup>, Damian Hutter<sup>10</sup>, Marc R. Pfluger<sup>10</sup>, Lena Kaiser<sup>10</sup>, Angelo Polito<sup>11</sup>, Janet Kelly<sup>12</sup>, Michael Von Rhein<sup>6</sup>, Swiss Neurodevelopmental Outcome Registry For Children With Congenital Heart Disease (orchid)<sup>13</sup>

<sup>1</sup>Pediatric Cardiology, Pediatric Heart Center; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland; <sup>2</sup>Pediatric Cardiology, Woman-Mother-Child Department, University Hospital Lausanne, Switzerland; <sup>3</sup>Pediatric Cardiology, Woman-Child-Adolescent Department, University Hospitals and Faculty of Medicine, University of Geneva, Switzerland; <sup>4</sup>Pediatric Intensive Care Unit, Woman-Mother-Child Department, University Hospital Lausanne, Switzerland; <sup>5</sup>Neonatology, Woman-Mother-Child Department, University Hospital Lausanne, Switzerland; <sup>6</sup>Pediatric Cardiology, Woman-Mother-Child Department, University Hospital Lausanne, Switzerland; <sup>7</sup>Child Development Center; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland; <sup>8</sup>Development and Growth, Department of Pediatrics, University Hospitals and Faculty of Medicine, University of Geneva, Switzerland; <sup>9</sup>Pediatric Cardiology, Woman-Child-Adolescent Department, University Hospitals and Faculty of Medicine, University of

Geneva, Switzerland; <sup>9</sup>Pediatric Neurology, Bern, Switzerland; <sup>10</sup>Pediatric Cardiology, Center for Congenital Heart Disease, Department of Cardiology and Cardiac Surgery, University Children's Hospital, University of Bern, Switzerland; <sup>11</sup>Pediatric and Neonatal Intensive Care Unit, Department of Pediatrics, Gynecology and Obstetrics, University Hospitals and Faculty of Medicine, University of Geneva, Switzerland; <sup>12</sup>Neonatology and Pediatric Intensive Care; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland; <sup>13</sup>on behalf of Swiss Neurodevelopmental Outcome Registry for Children with Congenital Heart Disease

**Background and Aim:** Necrotizing enterocolitis (NEC) is a major extra-cardiac complication in congenital heart disease (CHD). While length of ICU and hospital stay are associated with worse neurodevelopmental outcomes, the impact of postoperative NEC for neurodevelopmental (ND) outcome is unknown, which we analysed in a prospective clinical data set.

**Method:** The analysis based on data of the Swiss neurodevelopmental Outcome Registry for Children with complex Congenital Heart Disease (ORCHID), a nation-wide registry (founded in 2018) on the ND outcome of patients undergoing early neonatal cardiac surgery or hybrid palliation at < 6 weeks of age. Between 2019 and 2021 we analysed clinical characteristics, rate of complicating postoperative cardiogenic NEC ( $\geq$  Bell stage II), and their one year ND outcome (Bayley III).

**Results:** 101 patients (n = 63 female) were included. Cardiac diagnoses were biventricular (n = 75) univentricular (n = 21), or borderline types of CHD (n = 4). Surgery was conducted with (n = 86) or without (n = 15) cardiopulmonary bypass (CPB) at a median age (IQR) of 8 (6) days, most patients within RACHS category 3 or 4 (n = 73). CPB time was 213 $\pm$ 85 min, cross clamping time 125 $\pm$ 51 min, including selective cerebral perfusion (n = 53) at moderate lowest core temperature of 31.2 $\pm$ 4.0 °C. After surgery, cardiogenic NEC occurred in 16 patients, representing the second most frequent complications after respiratory complications (n = 35). NEC was treated by antibiotics/parenteral nutrition for at least 5 days (n = 15) resp. abdominal surgery (n = 1). On average, the Bayley III scores at 11.5 $\pm$ 1.5 months of age showed cognitive (102.2 $\pm$ 14.9), language (93.8 $\pm$ 13.0), and motor composite scores (88.7 $\pm$ 15.8) in the normal range, with a trend towards lower values in patients after NEC. Length of ICU (13.5 with vs. 8 days without NEC, p<0.05) resp. hospital stay (49 with vs. 31.5 days without NEC, p<0.05) were longer in patients after complicating NEC.

**Conclusions:** Postoperative cardiogenic NEC is associated with longer ICU resp. hospital stay, but not affecting ND outcome at one year of age as a solely covariate. Systematic nation-wide registries such as Swiss ORCHID with ever-growing patient populations serve as an excellent research platform to better understand the impact of perioperative risk factors on the long-term ND outcome.

**Keywords:** Neurodevelopmental outcome, registry, neonatal cardiac surgery, complication

## O-21

### Hippocampal subfield alterations and working memory in children with congenital heart disease

Ruth O'gorman Tuura<sup>1</sup>, Melanie Ehrler<sup>2</sup>, Nadja Naef<sup>2</sup>, Alenka Schmid<sup>2</sup>, Felicitas Koch<sup>2</sup>, Fraser Callaghan<sup>1</sup>, Oliver Kretschmar<sup>3</sup>, Beatrice Latal<sup>2</sup>

<sup>1</sup>Center for MR Research, University Children's Hospital Zurich, Switzerland; <sup>2</sup>Child Development Center, University Children's Hospital Zurich, Switzerland; <sup>3</sup>Department of Cardiology, University Children's Hospital Zurich, Switzerland

**Background and Aim:** Children and adults with congenital heart disease (CHD) are at risk of structural brain abnormalities including reduced brain volumes.<sup>1</sup> One of the most consistent findings has been an association between reduced hippocampal volumes and memory impairment,<sup>2</sup> but the volumes of individual hippocampal subfields are largely unexplored in the context of different working memory domains in CHD. This study aimed to examine the

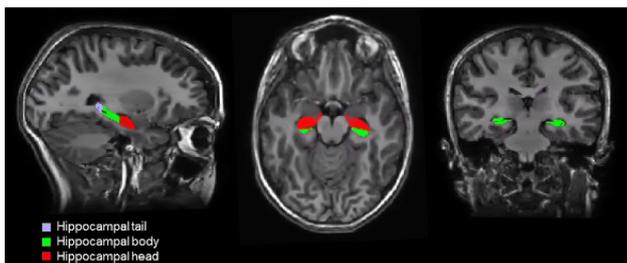
association between hippocampal subfield volumes and verbal and spatial working memory, in children with complex CHD and age-matched control children.

**Method:** The participant group included 57 children with complex CHD without genetic comorbidities, who had undergone cardiopulmonary bypass surgery before the age of 6, and 82 healthy control children. Verbal working memory was assessed with the working memory index from the Wechsler Intelligence Scale for children (WISC-IV), and visuo-spatial working memory was assessed from the CORSI block-tapping test. Hippocampal subvolumes were assessed from segmentation of a high resolution 3D T1-weighted MRI volume in Freesurfer 7.1. The volume of the left and right hippocampal head, body, and tail were extracted, and groupwise differences were tested with a univariate ANOVA. Post-hoc analyses were performed to assess the effect of cyanotic vs acyanotic CHD on the hippocampal subvolumes, and associations between working memory measures and hippocampal volumes were tested with partial Spearman correlations. The total brain volume was included as a covariate for all analyses.

**Results:** The CHD group showed poorer verbal and spatial working memory scores and smaller hippocampal subvolumes, particularly for the hippocampal tail bilaterally ( $p < 0.003$ ). Verbal and spatial working memory were differentially related to the hippocampal subvolumes, and hippocampal volumes in cyanotic CHD were smaller than in acyanotic CHD.

**Conclusions:** The hippocampus is a core region for memory function and is sensitive to hypoxic injury. Here we observed that hippocampal volumes were smaller in cyanotic CHD than in acyanotic CHD, supporting the link between hypoxia, hippocampal damage, and memory impairment. Children with CHD also showed poorer working memory scores, which were differentially related to the hippocampal subvolumes. Early detection of hippocampal changes may help to identify children with complex CHD at risk for memory impairments.

**Keywords:** congenital heart, brain, working memory, hippocampus, MRI



3D T1-weighted SPGR volume from one participant, with the Freesurfer hippocampal subfield segmentation for the head, body, and tail of the hippocampus superimposed.

## Nursing and Allied Health Professionals

### O-22

#### Facilitators and barriers for continued follow-up care after transfer – from a healthcare provider's perspective

Sandra Skogby<sup>1</sup>, Eva Goossens<sup>2</sup>, Bengt Johansson<sup>3</sup>, Philip Moons<sup>4</sup>, Ewa Lena Bratt<sup>5</sup>

<sup>1</sup>Institute of Health and Care Sciences, University of Gothenburg, Gothenburg, Sweden; <sup>2</sup>Department of Paediatric Cardiology, Queen Silvia Children's Hospital, Gothenburg, Sweden; <sup>3</sup>KU Leuven Department of Public Health and Primary Care, Leuven, Belgium; <sup>4</sup>KU Leuven Department of Public Health and Primary Care, Leuven, Belgium; <sup>5</sup>Centre for Research and Innovation in Care, Department of Nursing and Midwifery, Faculty of Medicine and Health Sciences, University of Antwerp, Antwerp, Belgium; <sup>3</sup>Heart Centre and Department of Surgery and Perioperative Sciences, Umeå University, Umeå, Sweden; <sup>4</sup>Institute of Health and Care Sciences, University of Gothenburg, Gothenburg, Sweden; <sup>5</sup>KU Leuven Department of Public Health and Primary Care, Leuven, Belgium; <sup>6</sup>Department of Paediatrics and Child Health, University of Cape Town, Cape Town, South Africa; <sup>7</sup>Institute of Health and Care Sciences, University of Gothenburg, Gothenburg, Sweden; <sup>8</sup>Department of Paediatric Cardiology, Queen Silvia Children's Hospital, Gothenburg, Sweden

**Background and Aim:** Young people with congenital heart disease (CHD) are frequently affected by discontinued follow-up when transferring from paediatric to adult care. Identified predictors for discontinuation include mostly patient-related factors, and further knowledge of hospital and healthcare system factors is needed. The overall purpose of this study is to provide broader perspectives and complement previous descriptions of barriers and facilitators for continued follow-up after transfer. The study explores patient-, hospital-, and healthcare system-related factors affecting continued follow-up care after transfer, as perceived by paediatric cardiology and adult congenital heart disease (ACHD) healthcare providers (HCPs) in Sweden and Belgium.

**Method:** Methods include individual interviews with cardiologists, nurses, and administrative staff at eight university hospitals in Sweden and Belgium, subjected to qualitative content analysis.

**Results:** Three main categories of factors perceived by HCPs to affect continued follow-up care after transfer emerged from the qualitative content analysis, including: "Care structure", "Care processes", and "Patient characteristics and circumstances". Individual characteristics and circumstances of patients were perceived to affect continued follow-up, including challenges of transition to adulthood, personal motivation for follow-up, and individual resources. Processes of care were perceived to affect continued follow-up, here quality of care delivery and care relationships were highlighted as well as personal responsibility for follow-up, collaboration across clinics, and transitional care interventions, such as patient education. Factors on a hospital and healthcare system level, including care access, out-of-pocket costs, organization of care and clinic resources, were highlighted as affecting continued follow-up. Overall, particularly highlighted success factors included: preparations for transfer, structured transfer routines, the collaboration between paediatric and adult care, transitional care interventions, personal care relationships, full-time dedicated staff, sufficient administrative resources, and active

measures to keep patients in care, such as calling non-attending patients. Few differences appeared between paediatric and ACHD HCP, as well as between Swedish and Belgian HCPs.

**Conclusions:** Factors regarding patient-, hospital- and healthcare system levels were perceived by HCPs to influence continued follow-up. Process and structure-related aspects of care were perceived as more influential than individual patient characteristics. The identified factors most probably interact, creating synergistic effects, and therefore should not only be considered separately but also jointly.

**Keywords:** Adolescent; Continuity of patient care; Heart Defects, Congenital; Patient transfer; Young Adult

### O-23

#### Optimizing transfer to adult care for young people with congenital heart disease

Birgitte Lykkeberg

Cardiology Clinic, The Heart Centre, Copenhagen University Hospital – Rigshospitalet, Denmark

**Background and Aim:** When the healthcare of people with congenital heart disease (CHD) is transferred from pediatric to adult care, there is a risk of discontinuity of care (3.6 - 62.7%) which has been associated with increased morbidity. Moreover, studies emphasize that young people with CHD have a significant lack of knowledge about their disease. To develop self-management skills and autonomy international guidelines recommend engaging in an educational transition program before the actual event of transfer to adult care. Our transition program has been established since 2008, however, a structured approach and new components were needed in order secure continuity of high-quality care when young people transfer from pediatric to adult care.

**AIM:** To optimize a transition program in adult care for young people with CHD.

**Method:** Patient participation, reviewing the literature, and organization of pathways were applied. To get insight into opinions from young people and to identify areas for development, workshops were held with the participation of experts in the field together with a young person. Observational notes were taken with an observational study approach and analyzed thematically. International guidelines were reviewed for practices and issues that could be adapted to the transition program.

**Results:** Young people benefit from enhanced collaboration between pediatric and adult clinics when they are offered a transfer visit ahead of the actual transfer to adult care. This visit is important as it helps young people to gain confidence in the new healthcare team in adult care. Hence, the provision of transition visits in the adult clinic should occur until young people are in their mid-twenties. A transition coordinator care model was implemented as it comprehensively addresses many challenges associated with transition.

**Conclusions:** With inspiration from research, collaboration with a pediatric ward, inviting experts and young people, several new components have been implemented in our transition program. The clinical pathways have been developed to ensure continuity of care for young people with CHD. As a result of this, the number of transition consultations has increased from 63 in 2020 to 90 in 2021.

**Keywords:** Optimizing, transfer, continuity of care, congenital heart disease

### O-24

#### Sustained effects of cardiac rehabilitation on the exercise function, quality of life and self-esteem of patients with complex congenital heart diseases

Ana Clara Félix<sup>1</sup>, Sérgio Laranjo<sup>1</sup>, Inês Perez<sup>2</sup>, Jorge Soares<sup>2</sup>, Mário Duque<sup>1</sup>, Fátima F Pinto<sup>1</sup>

<sup>1</sup>Serviço de Cardiologia Pediátrica, Centro de Referência de Cardiopatias Congénitas do CHULC, Hospital de Santa Marta, Centro Hospitalar e Universitário de Lisboa Central, Lisboa, Portugal; <sup>2</sup>Serviço de Medicina Física e Reabilitação, Hospital de Santa Marta, Centro Hospitalar e Universitário de Lisboa Central, Lisboa, Portugal

**Background and Aim:** The exercise capacity of patients with congenital heart disease (CHD) is often depressed. Past studies have documented the acute benefits of cardiac rehabilitation in children with CHD. The objective of this study was to characterise the acute and long-term effects of a cardiac rehabilitation program on the exercise function, quality of life and self-esteem of children with CHD.

**Method:** Eighteen patients, ages 13 to 21 years, with complex CHD who were referred for the cardiac rehabilitation programme and found to have a peak oxygen consumption (VO<sub>2</sub>) < 80% of predicted were enrolled in this study. The programme consisted of three sessions of exercise training/week for 12 weeks, focusing on improving functional capacity, motor skills and health literacy. Each session comprised endurance, resistance, strength, and flexibility exercises. A gamification approach was followed to motivate the younger patients. Patients had post-rehabilitation CPET at the end of the programme and the twelve months' follow-up. Changes in exercise function relative to baseline, pre-cardiac rehabilitation exercise tests were compared with changes observed in a group of control subjects with similar diagnoses, who also had 2 exercise tests separated by a year but did not undergo cardiac rehabilitation. Health-related quality of life (HR-QoL) and self-esteem questionnaires were applied at the three assessment moments.

**Results:** At the end of the programme, Improvements were found in 16 patients (89%). Peak VO<sub>2</sub> rose from 21.5+/-7 to 32.1+/-6mL/kg/min, and the ventilatory anaerobic threshold from 15.6+/-4 to 18.2+/-4mL/kg/min. The peak heart rate and peak respiratory exchange ratio did not change. No patient experienced rehabilitation-related complications. The exercise function did not change significantly at the twelve months of follow-up; the predicted peak oxygen consumption percentage remained superior to the baseline. These changes were also associated with HR-QoL and self-esteem improvements. In contrast, among the control subjects, statistically insignificant decreases in peak VO<sub>2</sub> were observed on the final exercise test compared with values obtained at baseline.

**Conclusions:** Cardiac rehabilitation can improve the exercise performance of children with CHD. Additionally, cardiac rehabilitation produces significant, sustained improvements in exercise function, HR-QoL and self-esteem. Routine use of formal cardiac rehabilitation may significantly reduce the morbidity of complex CHD.

**Keywords:** cardiac rehabilitation, congenital heart disease, HR-QoL, self esteem

## Genetics, Basic Science and Myocardial Disease

## O-25

**Left ventricular non-compaction in childhood: echocardiographic follow-up and prevalence in first-degree relatives**

Thilde Olivia Kock<sup>1</sup>, Marie Fagerlund Boerresen<sup>1</sup>, Anne Sophie Sillesen<sup>2</sup>, Ruth Ottilia Birgitta Vøgg<sup>2</sup>, Jakob Boesgaard Norsk<sup>2</sup>, Maria Munk Paerregaard<sup>2</sup>, Niels Grove Vejstrup<sup>3</sup>, Alex Hoerby Christensen<sup>1</sup>, Kasper Karmark Iversen<sup>2</sup>, Henning Bundgaard<sup>3</sup>, Anna Axelsson Raja<sup>3</sup>  
<sup>1</sup>Department of Cardiology, Copenhagen University Hospital Herlev-Gentofte, Copenhagen, Denmark; <sup>2</sup>Department of Cardiology, Copenhagen University Hospital Rigshospitalet, Copenhagen, Denmark; <sup>3</sup>Department of Cardiology, Copenhagen University Hospital Rigshospitalet, Copenhagen, Denmark

**Background and Aim:** Left ventricular non-compaction (LVNC) is characterized by excessive trabeculations of the left ventricular wall. The clinical importance and heredity of LVNC with normal systolic function is unclear. We aimed to describe the development of the left ventricular function and pattern in 2–4-year-old children with LVNC, diagnosed at birth, compared to matched controls. Additionally, we aimed to describe the prevalence of LVNC in first-degree relatives.

**Method:** A follow-up transthoracic echocardiography was performed at 2–4 years of age. Cases were matched 1:4 to controls. First-degree relatives were also offered inclusion. LVNC was defined as a ratio of non-compact to compact myocardium of  $\geq 2$  in at least one left ventricular segment measured in end-diastole.

**Results:** Of the 16 children diagnosed with LVNC at birth, 14 (median age 3 (interquartile range (IQR) 3–4) years, 71% male) were reevaluated together with 56 children without LVNC at birth (age 4 (IQR 3–4) years, 71% male). Also, 37 first-degree relatives of children with LVNC (age 31 (IQR 4–38) years, 46% male) and 146 first-degree relatives of children without LVNC (age 33 (IQR 11–40) years, 50% male) were included. In cases, the proportion of trabeculated segments (8% vs. 13%,  $p = 0.81$ ) and left ventricular ejection fraction (LVEF) (50% vs. 49%,  $p = 0.91$ ), were unchanged from birth to follow-up. LVEF was significantly lower in cases compared with controls at follow-up (49% vs. 60%,  $p < 0.001$ ). Criteria for LVNC was fulfilled in 11 out of 37 (30%) first-degree relatives to cases, whereas none of the first-degree relatives of controls fulfilled criteria ( $p < 0.001$ ). LVEF was significantly lower in first-degree relatives of cases fulfilling criteria of LVNC compared to first-degree relatives of controls (51% vs. 60%,  $p < 0.001$ ).

**Conclusions:** Children with LVNC diagnosed at birth as part of a population study still had a reduced systolic function when compared to controls but showed no further progression of left ventricular dysfunction or extent of trabeculation at the age of 2–4 years. Of the first-degree relatives to children with LVNC, 30% (95%CI 15–44%) fulfilled the criteria for LVNC and had reduced systolic function compared to controls. These findings strongly support family-screening and clinical follow-up of children with LVNC.

**Keywords:** left ventricular non-compaction, cardiomyopathy

## O-26

**Hypoxia-inducible factor-1 $\alpha$  signaling contributes to disease pathogenesis in sarcomeric hypertrophic cardiomyopathy**

Sarala Raj Murthi<sup>1</sup>, Andreas Petry<sup>2</sup>, Bachuki Shashikadze<sup>5</sup>, Damir Kračun<sup>2</sup>, Mathieu Klop<sup>2</sup>, Luisa Lehmann<sup>2</sup>, Joachim Schmitt<sup>3</sup>, Karin Klingel<sup>4</sup>, Florian Flenkenthaler<sup>5</sup>, Jan B. Stöckl<sup>5</sup>, Josh Gorham<sup>6</sup>, Gianluca Santamaria<sup>10</sup>, Christopher N. Toepfer<sup>12</sup>, Xinpei Chen<sup>9</sup>, Manuel Schmid<sup>12</sup>, Zsuzsanna Mayer<sup>7</sup>, Sabine Bauer<sup>8</sup>, Alessandra Moretti<sup>14</sup>, Thomas Fröhlich<sup>5</sup>, Peter Ewert<sup>11</sup>, Stefan Holdenrieder<sup>7</sup>, Agnes Görlach<sup>13</sup>, Jonathan G. Seidman<sup>6</sup>, Christine E. Seidman<sup>6</sup>, Cordula M. Wolf<sup>11</sup>

<sup>1</sup>Department of Pediatric Cardiology and Congenital Heart Defects, German Heart Center Munich, Technical University of Munich, School of Medicine & Health, Munich, Germany; <sup>2</sup>Experimental and Molecular Pediatric Cardiology, German Heart Center Munich at the Technical University Munich, Munich, Germany; <sup>3</sup>Institute of Pharmacology and Clinical Pharmacology, University Hospital Düsseldorf and Cardiovascular Research Institute Düsseldorf (CARID), Heinrich-Heine-University, Düsseldorf, Germany; <sup>4</sup>Institute for Pathology and Neuropathology, University Hospital of Tübingen, Cardiopathology, Tübingen, Germany; <sup>5</sup>Laboratory for Functional Genome Analysis LAFUGA Gene Center, LMU Munich, Munich, Germany; <sup>6</sup>Department of Genetics, Harvard Medical School, Boston, USA; <sup>7</sup>Institute for Laboratory Medicine, German Heart Center Munich, Munich, Germany; <sup>8</sup>Department of Cardiology, German Heart Center Munich, Technical University of Munich, School of Medicine & Health, Munich, Germany; <sup>9</sup>Experimental and Molecular Pediatric Cardiology, German Heart Center Munich at the Technical University Munich, Munich, Germany & School of Medicine, Technical University of Munich, Munich, Germany; <sup>10</sup>First Department of Medicine, Cardiology, Klinikum Rechts der Isar, School of Medicine & Health, Technical University of Munich, Germany & Department of Experimental and Clinical Medicine, “Magna Graecia” University of Catanzaro, Italy; <sup>11</sup>Department of Pediatric Cardiology and Congenital Heart Defects, German Heart Center Munich, Technical University of Munich, School of Medicine & Health, Munich, Germany & DZHK (German Centre for Cardiovascular Research), partner site Munich Heart Alliance, Munich, Germany; <sup>12</sup>Department of Genetics, Harvard Medical School, Boston, USA & Cardiovascular Medicine, Radcliffe Department of Medicine, University of Oxford, United Kingdom & Wellcome Centre for Human Genetics, University of Oxford, United Kingdom; <sup>13</sup>Experimental and Molecular Pediatric Cardiology, German Heart Center Munich at the Technical University Munich, Munich, Germany & DZHK (German Centre for Cardiovascular Research), partner site Munich Heart Alliance, Munich, Germany & School of Medicine, Technical University of Munich, Munich, Germany; <sup>14</sup>First Department of Medicine, Cardiology, Klinikum Rechts der Isar, School of Medicine & Health, Technical University of Munich, Germany & DZHK (German Centre for Cardiovascular Research), partner site Munich Heart Alliance, Munich, Germany

**Background and Aim:** Hypertrophic cardiomyopathy (HCM) caused by autosomal-dominant mutations in genes that code for the structural proteins of the sarcomere, is the most common inherited heart disease. HCM is associated with progressive myocardial hypertrophy and fibrosis, ventricular dysfunction, and arrhythmias. Disease onset during childhood and adolescence carries the risk of morbidity and sudden cardiac death. Hypoxia and

the main regulator of the cellular hypoxic response hypoxia-inducible transcription factor-1 $\alpha$  (HIF-1 $\alpha$ ) have been associated with HCM, however their exact role are not elucidated yet. The aim of the study is to determine the role of HIF-1 $\alpha$  in HCM disease pathogenesis.

**Method:** The effect of cardiomyocyte-specific HIF-1 $\alpha$  knockout (cHIF1 $\alpha$ KO) was studied in the established MHCR719W+/KO HCM mouse model that exhibits classical features of human HCM. Disease phenotype was analyzed by echocardiography, histopathology, immunohistochemistry, and determination of oxidative stress. Whole transcriptome and proteome analysis and subsequently pathway analysis were performed. Furthermore, serum proteomic analysis was carried out in patients affected by HCM to validate the contribution of HIF-1 $\alpha$  signaling in human HCM.

**Results:** HIF-1 $\alpha$  protein and HIF downstream targets, such as Prkaa2, are upregulated in left ventricular tissue of adult MHCR719W+/KO HCM mice. Cardiomyocyte-specific abolishment of HIF-1 $\alpha$  did not cause any cardiac abnormalities in cHIF1 $\alpha$ KO mice, but caused amelioration of disease phenotype in MYH6R719W+/KO - cHIF1 $\alpha$ KO mice, as evidenced by decreased left ventricular wall thickness on echocardiography, reduced myocardial fibrosis on histopathology, reduced disordered SRX/DRX state, diminished ROS production, and downregulation of HCM markers Actg1, Des, Lmna, Myh7 and Tpm4. In addition, cardiomyocyte-specific HIF-1 $\alpha$  knockout induced normalization of pathologic left ventricular remodeling signaling evidenced on whole transcriptome bulk mRNA sequencing and proteomics analysis in MYH6R719W+/KO - cHIF1 $\alpha$ KO mice. Proteomics performed of serum samples from patients with early onset HCM (N = 16) compared to age- and gender-matched healthy controls (N = 15) revealed HIF targets LTBP1, RECK, WDR1 and IGF2 as significantly dysregulated.

**Conclusions:** Cardiomyocyte-specific knockout of the HIF-1 $\alpha$  attenuates disease phenotype in the hypertrophic cardiomyopathy MYH6R719W+/KO mouse model by the regulation of HIF-1 $\alpha$  and its associated adaptive pathways. Targeting HIF-1 $\alpha$  might serve as therapeutic option to prevent disease progression in HCM.

**Keywords:** Hypertrophic cardiomyopathy, hypoxia, HIF-1 $\alpha$ , hypertrophy, myocardial fibrosis.

## O-27

### Cardiovascular screening in cutis laxa syndromes

Ilse Meerschaut<sup>1</sup>, Aude Beyens<sup>2</sup>, Katya De Groot<sup>3</sup>, Kristof Vandekerckhove<sup>3</sup>, Joseph Panzer<sup>3</sup>, Sofie Symoens<sup>2</sup>, Marie Claude Boiteux<sup>4</sup>, Karolien Aelbrecht<sup>5</sup>, Laura Muiño Mosquera<sup>1</sup>, Bert Callewaert<sup>2</sup>

<sup>1</sup>Center for Medical Genetics, Ghent University Hospital, Ghent, Belgium; <sup>2</sup>Department of Pediatric Cardiology, Ghent University Hospital, Ghent, Belgium; <sup>3</sup>Center for Medical Genetics, Ghent University Hospital, Ghent, Belgium; <sup>4</sup>Department of Biomolecular Medicine, Ghent University, Ghent, Belgium; <sup>5</sup>Department of Pediatric Cardiology, Ghent University Hospital, Ghent, Belgium; <sup>6</sup>Cutis laxa internationale, patient organization for cutis laxa, Bons-en-Chablais, France; <sup>7</sup>Center for Medical Genetics, Ghent University Hospital, Ghent, Belgium

**Background and Aim:** Cutis laxa (CL) syndromes comprise a heterogeneous group of rare inherited or acquired connective tissue disorders characterized by a loose, redundant skin. Inherited CL results from impaired synthesis and/or assembly of elastic fibers. Clinical subgroups are determined on the mode of inheritance and the type and extend of other organ involvement, including lung emphysema (ELN, FBLN5, LTBP4) and/or cardiovascular

manifestations (ELN, FBLN4) versus growth retardation (PYCR1, ALDH18A1) and skeletal anomalies (ATP6V0A2), with or without neurodevelopmental impairment or corneal clouding or cataract (PYCR1, ALDH18A1). Acquired CL results from elastic fiber degradation, often preceded by inflammation. Arterial aneurysms and stenoses, and/or tortuosity have been described in ELN, FBLN4, FBLN5, and LTBP4-related CL, but cardiovascular involvement has not been studied in detail in most CL subtypes. This study describes the cardiovascular features in individuals with CL presenting at the 6th CL days at Ghent University Hospital (September 2022).

**Method:** We evaluated 17 individuals (13 females) with a molecular or clinical diagnosis of ELN (6, one analysis pending), ALDH18A1 (1), FBLN5 (1), LTBP4 (2), ATP6V0A2 (4), PYCR1 (1)-related CL, unknown type (1), or acquired CL (1) with transthoracic echocardiography (TTE) and blood pressure measurement.

**Results:** The mean age of all individuals was 21.4  $\pm$  17.1 years, the range 2 – 52 years. Overall, TTE visualization was poor (likely due to lung emphysema). Abnormalities were found in 11 patients (64.7%). Five had tricuspid and/or mitral valve prolapse and/or insufficiency (FBLN5 (1/1), LTBP4 (2/2), ATP6V0A2 (1/4), unknown (1/1)), one had a bicuspid aortic valve (ATP6V0A2 (1/4)) and 7 had aortic insufficiency (ELN (3/6), FBLN5 (1/1), LTBP4 (2/2), unknown (1/1)). Three patients had dilatation of the aortic sinus (ATP6V0A2 (1/4), ELN (1/6)) or the ascending aorta (ALDH18A1 (1/1)). Myocardial mass index was increased in 6 individuals (ELN (2/6), ALDH18A1 (1/1), LTBP4 (1/2), ATP6V0A2 (1/4), unknown (1/1)). Two adults had arterial hypertension (ELN (2/6)).

**Conclusions:** Our findings underscore the importance of cardiovascular screening in CL, with special attention for arterial hypertension, valvular anomalies and aortic dilatation. Due to limited imaging quality of TTE in CL, an MRI head-to-pelvis could be of added value.

**Keywords:** cutis laxa, cardiovascular screening, arterial hypertension, valvular anomalies, aortic dilatation

## O-28

### Natural history and predictors of mortality and arrhythmic events in children with rasopathy-associated hypertrophic cardiomyopathy

Olga Dimitra Boleti<sup>1</sup>, Gabrielle Norrish<sup>1</sup>, Ella Field<sup>1</sup>, Kathleen Dady<sup>2</sup>, Kim Summers<sup>1</sup>, Gauri Nepali<sup>3</sup>, Vinay Bhole<sup>3</sup>, Orhan Uzun<sup>4</sup>, Piers Edward Francis Daubney<sup>5</sup>, Graham A Stuart<sup>6</sup>, Precyia Fernandes<sup>7</sup>, Karen Mcleod<sup>7</sup>, Maria Iliina<sup>7</sup>, Muhammad Najih Ali Liaqath<sup>8</sup>, Tara Bharucha<sup>8</sup>, Grazia Delle Donne<sup>9</sup>, Katie Linter<sup>10</sup>, Caroline Jones<sup>11</sup>, Jonathan Searle<sup>12</sup>, Sujeev Mathur<sup>13</sup>, Nicola Boyd<sup>14</sup>, Zdenka Reinhardt<sup>14</sup>, Satish Adwani<sup>12</sup>, Terence Prendiville<sup>15</sup>, Juan Pablo Kaski<sup>1</sup>

<sup>1</sup>Institute of Cardiovascular Sciences, University College London, London, United Kingdom; <sup>2</sup>Centre for Inherited Cardiovascular Diseases, Great Ormond Street Hospital, London, United Kingdom; <sup>3</sup>The Heart Unit, Birmingham Children's Hospital, Birmingham, United Kingdom; <sup>4</sup>Children's Heart Unit, University Hospital of Wales, Cardiff, United Kingdom; <sup>5</sup>Department of Paediatric Cardiology, Royal Brompton and Harefield NHS Trust, London, United Kingdom; <sup>6</sup>Department of Paediatric Cardiology, Bristol Royal Hospital for Children, Bristol, United Kingdom; <sup>7</sup>Department of Paediatric Cardiology, Royal Hospital for Children, Glasgow, United Kingdom; <sup>8</sup>Department of Paediatric Cardiology, Southampton General Hospital, Southampton, United Kingdom; <sup>9</sup>Department of Paediatric Cardiology, Leeds General Infirmary, Leeds, United Kingdom; <sup>10</sup>Department of Paediatric Cardiology, Glenfield Hospital, Leicester, United Kingdom;

<sup>11</sup>Department of Paediatric Cardiology, Alder Hey Children's Hospital,

Liverpool, United Kingdom; <sup>12</sup>Department of Paediatric Cardiology, John Radcliffe Hospital, Oxford, United Kingdom; <sup>13</sup>Children's Heart Service, Evelina Children's Hospital, London, United Kingdom; <sup>14</sup>Department of Paediatric Cardiology, The Freeman Hospital, Newcastle, United Kingdom; <sup>15</sup>The Children's Heart Centre, Our Lady's Children's Hospital, Dublin, Republic of Ireland

**Background and Aim:** The RASopathies are a group of developmental disorders caused by germline variants in components of the RAS-MAPK pathway, commonly associated with hypertrophic cardiomyopathy (HCM). The aim of this study was to describe the natural history and predictors of all-cause mortality and major arrhythmic cardiac events (MACE) in a large, multi-centre cohort of paediatric patients with a RASopathy syndrome and HCM.

**Method:** The study cohort consisted of patients  $\leq 18$  years with HCM and a clinical and/or genetic diagnosis of a RASopathy syndrome (Noonan syndrome (NS), NS with multiple lentiginos (NSML), Costello syndrome (CS), cardiofaciocutaneous syndrome (CFCS), Noonan-like syndrome), evaluated between January 1, 1985, and December 31, 2020, in 14 paediatric cardiology centres in the United Kingdom and the Republic of Ireland.

**Results:** One-hundred-and-forty-nine patients were included (111 (74.5%) Noonan Syndrome (NS); 12 (8.05%) Noonan Syndrome with Multiple Lentiginos (NSML); 6 (4.03%) Costello Syndrome (CS); 6 (4.03%) Cardiofaciocutaneous Syndrome (CFCS); and 14 (9.4%) with Noonan-like syndrome patients). NSML patients had higher maximal wall thickness (MWT) (z score 17.02 (10.57–32.78),  $p = 0.004$ ) and higher left ventricular outflow tract (LVOT) gradient values (60mmHg, 36–80,  $p = 0.019$ ), but there were no other significant differences in cardiac phenotype between the different RASopathy syndromes. Over a median follow up of 197.5 months (93.58–370), 23 patients (15.43%) died, at a median age of 48.94 months (3.29–175.84). Overall survival was 96.45% (91.69–98.51), 90.42% (84.04–94.33) and 84.12 (75.42–89.94) at 1, 5 and 10 years, respectively, but this varied by RASopathy syndrome. RASopathy syndrome, specifically Noonan-like syndrome (Hazard ratio 4.18,  $p = 0.032$ ) and congestive cardiac failure admission (Hazard ratio 7.35,  $p = 0.037$ ) were independent predictors of all-cause mortality on multivariate analysis and LVOT gradient was an independent predictor for MACE (Hazard ratio 1.02,  $p = 0.025$ ).

**Conclusions:** These findings highlight a distinct category of patients with Noonan-like syndrome with a milder HCM phenotype but significantly worse survival, and identify predictors of adverse outcome in patients with RASopathy-related HCM.

**Keywords:** paediatric, hypertrophic cardiomyopathy, RASopathies, genetics, mortality, MACE

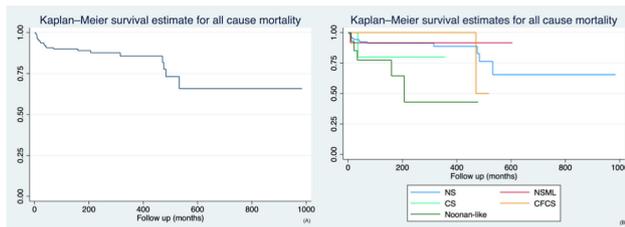


Figure 2: Kaplan-Meier survival analysis for all-cause mortality for whole cohort (A) and for RASopathy syndrome (B) for Follow up (months)

**Figure 1** Kaplan-Meier analysis for all cause mortality for whole cohort (A) and for RASopathy syndrome (B) for Follow up (months)

## O-29

### Clinical course of rhabdomyomas in tuberous sclerosis complex – 16-years single centre experience

Ivana Cerovic<sup>1</sup>, Vladislav Vukomanovic<sup>2</sup>, Ruzica Kraljanc<sup>3</sup>, Sanja Ninic<sup>1</sup>, Sasa Popovic<sup>1</sup>, Stasa Krasic<sup>1</sup>, Sergej Prijic<sup>2</sup>

<sup>1</sup>Department of Cardiology, Mother and Child Health Institute of Serbia, Belgrade, Serbia; <sup>2</sup>School of Medicine, University of Belgrade; Department of Cardiology, Mother and Child Health Institute of Serbia, Belgrade, Serbia; <sup>3</sup>School of Medicine, University of Belgrade; Department of Neurology, Mother and Child Health Institute of Serbia, Belgrade, Serbia

**Background and Aim:** cardiac rhabdomyomas occur in over 50% of patients with tuberous sclerosis complex (TSC), with variable clinical significance.

**Method:** retrospective analysis included 54/88 patients with TSC who underwent cardiac evaluation in period from June 2006 until September 2022.

**Results:** In total, 36 (66.7%) patients were diagnosed with cardiac rhabdomyoma (CR) – 44.4% had solitary, 11.1% two and 44.4% had multiple tumors, localised predominantly in ventricles (92%). Initial echocardiography was performed at the age of  $4.1 \pm 4.9$  years. Two patients were diagnosed prenatally. Largest diameter and cross-sectional area (CSA) of CR was  $9.8 \pm 6.4$  mm and  $57.7 \pm 67.9$  mm<sup>2</sup>, respectively. Subgroup of infants included 17/54 (31.5%) of patients, evaluated at the average age of  $4.2 \pm 3.5$  months. CRs were found in 76.5% of these patients (solitary and two tumors in 11.8% and 17.6%, respectively, multiple in 47.1%), with largest diameter of  $12.2 \pm 7.7$  mm and CSA of  $72.9 \pm 88.4$  mm<sup>2</sup>. Average follow-up of all patients was  $4.6 \pm 2.9$  years. CRs showed significant reduction in both diameter and CSA,  $6.4 \pm 6.0$  mm ( $p < 0.05$ ) and  $34.2 \pm 46.6$  mm<sup>2</sup> ( $p < 0.05$ ), respectively, while in 13.8% they resolved completely. Patients diagnosed in infancy initially showed tendency towards CR size reduction, achieving statistical significance after  $3.7 \pm 2.9$  years of follow up, with diameter of  $7.1 \pm 8.4$  mm ( $p < 0.05$ ) and CSA of  $39.9 \pm 61.8$  mm<sup>2</sup> ( $p < 0.05$ ). In 25% of them CRs disappeared. Four patients received mTOR inhibitors therapy and average regression of their CRs was  $6.8 \pm 4.8$  mm compared to  $1.3 \pm 2.9$  mm ( $p < 0.01$ ) in the rest of patients without specific treatment. Three patients (8.3%) had hemodynamically significant obstruction of blood flow. Five patients (12.2%) manifested with ECG abnormalities, two of them had atrial ectopic tachycardia, two had Wolf-Parkinson-White syndrome and one had salvos of premature atrial contractions.

**Conclusions:** cardiac rhabdomyomas in patients with tuberous sclerosis complex have tendency of spontaneous regression, but can occasionally cause life-threatening conditions. mTOR inhibitors potentiate their natural course and might be treatment of choice for symptomatic patients.

**Keywords:** cardiac rhabdomyoma, tuberous sclerosis complex, manifestations, clinical course

## O-30

### Impact of genotype on aortic disease progression in patients with marfan syndrome and loeys-dietz syndrome

Aitor Guitarte Vidaurre, Remi Vincent, Thomas Edouard, Caroline Munzer, Fernanda Bajanca, Maud Langeois, Bertrand Chesneau, Yves Dulac

Centre de Référence du syndrome de Marfan et des syndromes apparentés, Hôpital des Enfants, CHU de Toulouse, Toulouse, France.

**Background and Aim:** Marfan and Loays-Dietz syndromes are associated connective tissue disorders progressing from childhood.

Major morbidity and mortality in both syndromes relate to aorta dilation predisposing to aortic tear.

We investigated a putative relationship between genotype and aortic disease progression in young patients that can potentially lead to severe cardiovascular events.

**Method:** 100 patients (51 male, 49 female) with pathogenic variants in FBN1 (N = 84), TGFβR1 / R2 (N = 8) and SMAD3 (N = 8) genes were subject to retrospective analysis. The average age at study onset was 13.8 years and data were collected over 5 years. The FBN1 variants were classified as premature termination codon (PTC, N = 33) or dominant negative (DN, N = 51). Among DN variants 14 cases of cysteine loss (Cys-), severely affecting protein conformation, were analysed separately. Aortic diameters and aortic dilation at the level of the Valsalva sinuses were assessed by echocardiography.

**Results:** A link was found between gender and aortic diameter evolution (median females 3.8mm vs males 6.3mm; p = 0.0089) and growth rate (1.0mm/year females vs 1.4 mm/year males; p = 0.0061). No overall differences were found between Marfan and Loeys-Dietz syndrome neither between PTC, DN and Loeys-Dietz patients.

However, the aortic diameter evolution over 5 years was significantly higher in Cys- patients, with median 8.70mm against 4.00mm for other genotypes (p = 0.0007); OR adjusted for gender was 1.3[1.1-1.5], p = 0.003. The median growth rate was 1.53mm/year in Cys- patients against 1.04mm/year for other patients (p = 0.0127), adjusted on gender.

No significant difference was found between gender distribution in Cys- and the remaining patients (p = 0.15). Interestingly, mitral valve surgery was only reported in PTC patients (3/33, p = 0.03).

**Conclusions:** Our study shows that gender differences in aortic disease progression are significant since childhood. Male patients should be monitored more closely, with emphasis on FBN1 variants with loss of cysteine.

**Keywords:** marfan syndrome, loeys dietz syndrome, genetics

**O-31**

**Childhood-onset lamin a/c cardiomyopathy**

Marit Kristine Smedsrud<sup>1</sup>, Monica Chivulescu<sup>2</sup>, Marianne Inngjerdigen Forså<sup>2</sup>, Nina Eide Hasselberg<sup>2</sup>, Andreas Früh<sup>3</sup>, Kristina Hermann Haugaa<sup>2</sup>

<sup>1</sup>ProCardio Center for Innovation, Department of Paediatric Cardiology, Oslo University Hospital, Rikshospitalet, Norway;

<sup>2</sup>ProCardio Center for Innovation, Department of Cardiology, Oslo University Hospital, Rikshospitalet, Norway; <sup>3</sup>Department of Paediatric Cardiology, Oslo University Hospital, Rikshospitalet, Norway

**Background and Aim:** Lamin A/C gene (LMNA) mutations cause familial dilated cardiomyopathy (DCM) with a malignant cardiac phenotype characterized by atrioventricular (AV) block and supraventricular and ventricular arrhythmias, often preceding cardiac dilatation and dysfunction. Current guidelines recommend genetic and clinical screening of family members, starting at 10-12 years of age. However, children with LMNA mutations are underrepresented in research publications and there is limited data on the onset of a cardiac phenotype in this population.

We aimed to investigate the penetrance of LMNA phenotype during childhood and specifically the occurrence of cardiac events in LMNA genotype positive children.

**Method:** We conducted a single-centre, longitudinal cohort study including genotype-positive LMNA patients and genotype positive relatives, all ≤18 years of age, followed between 2009 and 2022. The patients were examined by electrocardiography, Holter monitoring, cardiac magnetic resonance imaging, and echocardiography. A cardiac phenotype was defined as the presence of atrioventricular (AV) block, atrial fibrillation/ flutter (AF), ventricular tachycardia (VT), and/or echocardiographic DCM. Cardiac events were defined as AF, VT, sudden cardiac arrest (SCA) or heart transplantation (Htx).

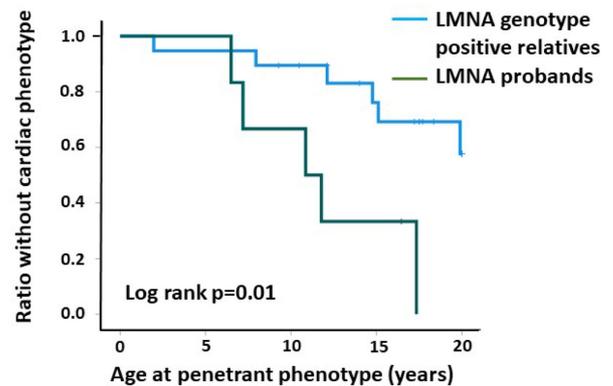
**Results:** We clinically followed 25 LMNA genotype-positive children (5 probands with cardiac phenotype, 1 proband with neuromuscular phenotype only, 19 relatives, age 9.7 [IQR 6.8-13.1] years) of which 11 (44%) were phenotype-positive at end of 9.7 years follow-up at a median age of 11.7 [IQR 7.2-15.1] years. Among the LMNA genotype-positive relatives, 32% (6/19) became phenotype-positive at a median age of 13.4 [IQR 6.5-16.3] years (Figure).

New onset cardiac events occurred in 8 children (32%) during follow-up, of which half of them in patients ≤12 years of age. Fifty% (4/8) of the cardiac events occurred in relatives. Two children had AF, 3 had non-sustained VT, and 1 underwent SCA. Htx was performed in 2 unrelated children, at 6 and 8 years of age, respectively.

**Conclusions:** In a paediatric cohort of LMNA genotype positive probands and relatives, we found a high occurrence of AF, VT and HTx. Half of the events occurred in children ≤12 years of age. One third of the paediatric LMNA genotype-positive relatives had a cardiac phenotype during follow-up, highlighting the importance of early family screening and cardiological follow-up.

**Keywords:** Lamin A/C cardiomyopathy, family screening, disease penetrance

Kaplan-Meier plot



<b>Probands</b>	<b>6</b>	<b>6</b>	<b>4</b>	<b>2</b>	<b>1</b>
<b>Relatives</b>	<b>19</b>	<b>18</b>	<b>17</b>	<b>14</b>	<b>13</b>

Comparison of ages of documented cardiac phenotype between 6 LMNA genotype-positive paediatric probands and 19 LMNA genotype-positive paediatric relatives

## O-32

**Analysis of the mutational spectrum and diagnostic yield in a cohort of patients referred for study of hypertrophic cardiomyopathy**

Ivonne Johana Cárdenas Reyes<sup>1</sup>, Luis De La Higuera Romero<sup>3</sup>, Almudena Amor Salamanca<sup>1</sup>, Soledad García Hernández<sup>1</sup>, María Valverde Gómez<sup>1</sup>, Diego Cabrera Argaña<sup>1</sup>, Laura Cazón Varela<sup>2</sup>, Marlene Perez Barbeito<sup>2</sup>, Iria Gómez Díaz<sup>2</sup>, Rosalia Peteiro Debén<sup>2</sup>, María Sánchez Flores<sup>2</sup>, Guillermo Smith Ramos<sup>1</sup>, Anahi Sanluis Verdes<sup>2</sup>, Xusto Fernández Fernández<sup>3</sup>, Martín Ortiz Genga<sup>1</sup>, Juan Pablo Ochoa<sup>1</sup>

<sup>1</sup>Scientific Committee, Health in Code. A Coruña, Spain; <sup>2</sup>Genetic Diagnosis Area, Health in Code. A Coruña, Spain; <sup>3</sup>Data Scientist, Health in Code. A Coruña, Spain

**Background and Aim:** Advances in genetic in recent decades have made it possible to identify in a significant percentage of hypertrophic cardiomyopathy (HCM) cases the molecular cause of the disease. Knowing the genetic etiology allows a more precise diagnosis, guiding both treatment and follow-up in carriers. The aim of this study is to evaluate the mutational spectrum and diagnostic yield of the genetic study in a cohort of pediatric patients with HCM.

**Method:** Retrospective study in which the presence of disease-causing genetic variants (P/LP) was evaluated in a cohort of patients diagnosed with HCM under 18 years of age and studied by next-generation-sequencing (NGS).

**Results:** 471 individuals diagnosed with HCM before the age of 18 years were studied in our center, identifying a disease-causing variant in 53% (n = 244) of the cases. In the group aged 0 to 1 years (n = 28), half of the cases were explained by variants in sarcomeric genes, and the other half by genes associated with HCM in the context of syndromic disease (mainly RASopathies). In the cohort of patients aged 1-17 years (n = 216), 88% of cases had pathogenic variants in sarcomeric genes (mainly MYH7, MYBPC3, and TNNT3), 6% in non-sarcomeric genes, and 5% in genes associated with RASopathies, mitochondrial or metabolic diseases. Complex genotypes in sarcomeric genes (homozygous, compound heterozygous or digenic carriers) were identified in 7% (n = 33) of cases.

**Conclusions:** Pathogenic variants in sarcomeric genes were the most frequent cause of disease, although in the 0-1 years group, variants associated with HCM in the context of syndromic disease are also relevant. The diagnostic yield of the genetic study in our cohort of pediatric patients with this phenotype was greater than 50%. The systematic performance of a comprehensive genetic study by NGS should be considered in pediatric patients with HCM.

**Keywords:** Hypertrophic cardiomyopathy, genetic study, pediatrics

## O-33

**Human cardiophere-derived cells with activated mitochondria for better myocardial regenerative therapy**

Masahiro Shiraishi<sup>1</sup>, Daisuke Sasaki<sup>2</sup>, Atsuhito Takeda<sup>2</sup>, Mitsue Hibino<sup>3</sup>, Hideyoshi Harashima<sup>4</sup>, Yuma Yamada<sup>5</sup>

<sup>1</sup>Department of Pediatrics, Graduate School of Medicine, Hokkaido University, Sapporo, Japan; Faculty of Pharmaceutical Sciences, Hokkaido University, Sapporo, Japan; <sup>2</sup>Department of Pediatrics, Graduate School of Medicine, Hokkaido University, Sapporo, Japan; <sup>3</sup>Faculty of Pharmaceutical Sciences, Hokkaido University, Sapporo, Japan; Faculty of Engineering, Hokkaido University, Sapporo, Japan; <sup>4</sup>Faculty of Pharmaceutical Sciences, Hokkaido University, Sapporo, Japan; <sup>5</sup>Faculty of Pharmaceutical Sciences, Hokkaido University, Sapporo, Japan; Fusion Oriented Research for disruptive Science and

Technology (FOREST) Program, Japan Science and Technology Agency (JST) Japan, Saitama, Japan

**Background and Aim:** Heart failure is one of the most common causes of death in the world, and treatment of patients with severe heart failure still has limited benefits. Cell transplantation is a promising therapeutic strategy for myocardial regeneration therapy. To improve therapeutic effects, we are developing a culture medium additive to enhance the mitochondrial function of cardiomyocytes for transplantation. We have succeeded in delivering mitochondrial activation molecules to mouse-derived cardiac progenitor cells using mitochondrial target drug delivery system (MITO-Porter system). In this study, we investigated whether the mitochondrial function of human-derived myocardial precursor cells could be enhanced with a view to clinical application of MITO-Porter.

**Method:** Human cardiophere-derived cells (CDCs) were isolated from excised myocardium during surgery for congenital heart disease, and coenzyme Q10 (CoQ), an antioxidant and mitochondrial electron transport coenzyme, was selected as the mitochondrial activating molecule. MITO-Porter encapsulating CoQ can be prepared in large quantities using microfluidic device and added to serum contained medium to deliver CoQ to the mitochondria of human CDCs for transplantation (referred to herein as human MITO cells). We optimized the protocol for preparing human MITO cells based on the oxygen consumption rate (OCR), which is an index of mitochondrial function. Oxidative stress in human MITO cells under ischemic conditions was evaluated in vitro by exposing human MITO cells to hydrogen peroxide (H<sub>2</sub>O<sub>2</sub>). We then verified the therapeutic effect of cell transplantation therapy using human MITO cells on a rat myocardial ischemia-reperfusion injury.

**Results:** Intracellular reactive oxygen species (ROS) levels upon H<sub>2</sub>O<sub>2</sub> exposure were significantly lower in human MITO cells than in non-treated CDCs. Furthermore, human MITO cells transplantation showed improvement in cardiac function and suppression of myocardial fibrosis compared with non-treated CDCs transplantation. These effects were observed not only by myocardial administration but also by intravenous administration of human MITO cells.

**Conclusions:** Transplantation of human MITO cells into ischemic myocardium showed a stronger therapeutic effect compared with non-treated CDCs transplantation. This study is the first attempt to verify that mitochondrial delivery of functional compound would contribute to improving the outcome of human-derived CDCs transplantation therapy.

**Keywords:** heart failure, myocardial regenerative therapy, cell therapy, mitochondria

**General Cardiology**

## O-34

**Utility of the single ventricular stroke work index for predicting the prognosis of patients with functional single-ventricle physiology**

Takashi Honda<sup>1</sup>, Rumi Watanabe<sup>1</sup>, Manabu Takanashi<sup>1</sup>, Atsushi Kitagawa<sup>1</sup>, Fumiaki Shikata<sup>2</sup>, Yoichiro Hirata<sup>1</sup>, Kagami Miyaji<sup>2</sup>, Kenji Ishikura<sup>1</sup>

<sup>1</sup>Department of Pediatrics, Kitasato University School of Medicine, Sagami-hara, Japan; <sup>2</sup>Department of Cardiovascular Surgery, Kitasato University School of Medicine, Sagami-hara, Japan

**Background and Aim:** The stroke work index (SWI) has been shown to reflect ventricular workload and considers both ventricular systolic and diastolic function. However, this index has never been

applied for patients with functional single-ventricle physiology. In this study, we evaluated the utility of single ventricular SVWI (SVSWI) for predicting the prognosis of single-ventricle patients. **Method:** This was a retrospective cohort study including 36 single-ventricle patients who underwent Blalock-Taussig shunt or pulmonary artery banding. Based on the results of cardiac catheterization before the Glenn procedure, we calculated SVSWI using the following equation:  $SVSWI = \text{stroke volume of the single ventricle/body surface area} \times (\text{mean aortic pressure} - \text{atrial pressure}) \times 0.0136$ . Stroke volume was calculated based on the sum of systemic and pulmonary blood flow. We evaluated the utility of SVSWI for predicting the rate of reaching Fontan completion within 2 years after catheterization and survival. Pearson's chi-square test or the Kaplan–Meier method was used for statistical analysis.

**Results:** The mean age at catheterization was  $0.53 \pm 0.46$  years. The patients were divided into a high SVSWI group (Group H,  $SVSWI > 60$ ,  $N = 19$ ) and a low SVSWI group (Group L,  $SVSWI < 60$ ,  $N = 17$ ). The rate of reaching Fontan completion was significantly higher in Group H than in Group L (100% vs. 76%,  $p = 0.0242$ ). The survival rate was also significantly higher in Group H ( $p = 0.0081$ ). The 5-year survival rates for Group H and L were 100% and 81%, and the 10-year survival rates were 100% and 65%, respectively. However, neither the rate of reaching Fontan completion nor mortality was predicted based on cardiac index, pulmonary artery index or end diastolic pressure of the main ventricle.

**Conclusions:** SVSWI is theoretically proportional to the area enclosed by the pressure–volume loop, reflecting the stroke work of the single ventricle. Our results revealed that we can simply predict the prognosis of single-ventricle patients based on SVSWI, as early as before the Glenn procedure, regardless of cardiac morphology. It has also been demonstrated that the prognosis of single-ventricle patients is determined by cardiac function before the Glenn procedure and that the first palliation and preservation of ventricular function are of critical for their prognosis.

**Keywords:** Stroke work index, single-ventricle physiology, Prognosis, Stroke work

## Pulmonary hypertension, heart failure and transplantation

### O-35

#### The impact of the obesity on the fontan hemodynamics during transition to the adult

Makoto Kudo, Takuya Takahashi, Kanchi Saito, Akira Sato, Satoshi Nakano, Yumi Sato, Hirofumi Saiki

Department of Pediatrics and Pediatric Cardiology, Iwate Medical University, Shiwa, Japan

**Background and Aim:** Obesity is a risk for developing heart failure in the adults, where increased circulatory volume as well as augmented ventricular afterload exacerbates cardiovascular remodeling. Since the characteristic feature of Fontan circulation including augmented afterload and compromised preload reserve attributes Fontan related end-organ dysfunction, obesity during adolescence may further accelerate organ dysfunction. We tested our hypothesis that obesity in the Fontan circulation additively increase the cardiovascular load and compromise organ function.

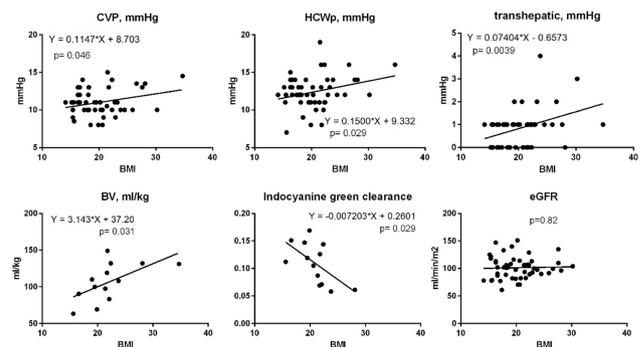
**Method:** The consecutive 54 Fontan patients older than 13 years old were enrolled in this study. The patients were categorized into 3 groups based on the Japanese obesity criteria using body mass index (BMI); obesity ( $>25$ ,  $n = 7$ ), lean ( $<18.5$ ,  $n = 21$ ), and

moderate ( $18.5\text{--}25$ ,  $n = 26$ ). The hemodynamic data and its interaction with markers for end-organ dysfunction were analyzed.

**Results:** The age for each group was similar ( $17 \pm 3$ ,  $16 \pm 5$ ,  $18 \pm 5$  years old). While cardiac index, systemic vascular resistance, and blood pressure (BP) were similar, central venous pressure (CVP  $12.7 \pm 2.1^*$ ,  $11.8 \pm 1.7$ ,  $10.8 \pm 2.0$  mmHg), pulmonary resistance ( $R_p$   $2.2 \pm 1.0^*$ ,  $2.0 \pm 0.8$ ,  $1.5 \pm 0.5$  mmHg), hepatic wedge pressure (HCWp  $14.0 \pm 1.6^*$ ,  $12.2 \pm 1.6$ ,  $11.7 \pm 2.1$  mmHg) were markedly high in the obese patients ( $*$ :  $p < 0.05$  as compared with moderate). Indeed, the BMI was positively correlated with CVP, HCWp and trans-hepatic pressure. Interestingly, blood volume (BV, ml/kg) measured by dye dilution and CVP augmentation by contrast load (mmHg/ml) were positively correlated with BMI ( $p = 0.031$ ,  $0.023$ , respectively), suggesting saturated vascular bed in the patients with high BMI. In addition, BP suppression with general anesthesia induction were closely correlated with higher BMI, but independent of BV, suggesting reactive vascular constriction as the contributor of BP augmentation in the obese patients ( $p = 0.0079$ , ANCOVA). Although BMI was independent of eGFR, serum hepatic fibrosis markers (procollagen type III peptide, type IV collagen 7s, hyaluronic acid) and hepatic elasticity markers (Shear wave elasticity, Dispersion slope), it was closely correlated with suppressed hepatic function as represented by ICG clearance rate ( $p = 0.029$ ).

**Conclusions:** The Fontan patients with obesity had unfavorable vascular hemodynamics including reactive BP augmentation and CVP elevation with saturated vascular bed. Although hepatic fibrosis was unaffected in adolescence, additive hemodynamic load was associated with hepatic dysfunction.

**Keywords:** Fontan, obesity, end-organ dysfunction, FALD,



The impact of BMI on the Fontan hemodynamics and organ function

## Interventional Cardiology

### O-36

#### Retrospective review of m3c-neck experience with transcatheter management of coronary artery fistulas

Raymond N Haddad, Damien Bonnet, Sophie Malekzadeh Milani  
M3C-Necker, Hôpital Universitaire Necker-Enfants malades, Assistance  
Publique-Hôpitaux de Paris (AP-HP), Paris, France.

**Background and Aim:** Coronary artery fistulas (CAFs) are rare coronary anomalies and transcatheter closure remains debatable. We aim to evaluate our experience with transcatheter management of CAFs.

**Method:** Retrospective institutional data review of all children in whom echocardiographically suspected CAFs were confirmed during cardiac catheterization from 2000 to 2022. Procedural considerations, techniques, standard safety, and outcomes were assessed.

**Results:** 92 CAFs were identified in 76 patients (44% males) with a median age of 3.8 years (IQR, 0.8–7) and weight of 15 Kg (IQR, 8.5–25). 25 (32.9%) patients had concomitant congenital anomalies and 9 (11.8%) had coronary artery anomalies. 39/51 (76.5%) patients with isolated CAFs were clinically asymptomatic at diagnosis. 27 (35.5%) patients had pre-procedural CT angiography. The CAFs originated from the left main coronary artery left (42.4%), right coronary artery (38.1%), left anterior descending (14.1%), and circumflex coronary artery (5.4%). The drainage sites were the right ventricle (57.6%), right atrium (22.8%), pulmonary arteries (14.1%), left atrium (2.2%), coronary sinus (2.2%), and left ventricle (1.1%). 23/76 (30.3%) patients with 35/92 (38%) small CAFs had no intervention with a benign clinical long-term follow-up. 8/76 (10.5%) patients with 9/92 (9.8%) CAFs not amenable to percutaneous closure were directly sent for surgery. 45/76 (59.2%) patients had percutaneous closure of 48/92 (52.2%) CAFs using microcoils (31.3%), device occluders (58.3%), or a combination of both (10.4%). Occlusion material was exchanged before release in 4 (8.9%) patients. Devices were deployed transvenously using a track wire loop in 19/48 (39.6%) CAFs. The closure approach was modified per-operatively in 4 (8.9%) patients. Percutaneous closure was unsuccessful in 3 (6.7%) patients of which 2 had surgical ligation. Twelve complications occurred including 7 transient ST–T wave changes, 2 asymptomatic coronary pseudo-stenosis, one coronary dissection, and one pulmonary edema. Repeat closure was needed in 3 (6.7%) patients for residual leak and was unsuccessful in 2 of them. One patient had trivial CAF recanalization with an asymptomatic 12-year-follow-up.

**Conclusions:** Transcatheter closure of CAFs is feasible and effective in carefully selected patients. Complications are frequent but not permanent. Surgery is a valuable upfront option in large and technically complex CAFs or a bailout of failed percutaneous attempts.

**Keywords:** coronary artery fistula, coil, percutaneous closure, plugs, children.

### O-37

#### Atrial flow regulator device in children and in patients with congenital heart disease: an international registry

Enrico Piccinelli<sup>1</sup>, Mara Pilati<sup>2</sup>, Micol Rebonato<sup>2</sup>, Kostantin Averin<sup>3</sup>, Grazyna Brzezinska Rajszys<sup>4</sup>, Adam Kolesnik<sup>4</sup>, Alban Elouen Baruteau<sup>5</sup>, Andreas Eicken<sup>6</sup>, James Kuo<sup>7</sup>, Matthew Jones<sup>8</sup>, Carles Bautista<sup>9</sup>, Alain Fraisse<sup>9</sup>, Biagio Castaldi<sup>10</sup>, Lisa Bianco<sup>11</sup>, Pedro Betrian Blasco<sup>11</sup>, Roberto Formigari<sup>2</sup>, Gianfranco Butera<sup>2</sup>  
<sup>1</sup>Ospedale Pediatrico Bambino Gesù, Rome, Italy; <sup>2</sup>PolitoBIOMed Lab, Politecnico di Torino, Italy; <sup>3</sup>Ospedale Pediatrico Bambino Gesù, Rome, Italy; <sup>4</sup>Stollery Children's Hospital, Edmonton, Canada; <sup>5</sup>The Children's Memorial Health Institute, Varsavia, Poland; <sup>6</sup>Centre hospitalier universitaire de Nantes, Nantes, France; <sup>7</sup>Deutsches Herzzentrum München, München, Germany; <sup>8</sup>Children's Healthcare of Atlanta, Atlanta, US; <sup>9</sup>Evelina Children's hospital, London, UK; <sup>9</sup>Royal Brompton Hospital, London, UK; <sup>10</sup>Azienda ospedaliera di Padova, Padova, Italy; <sup>11</sup>Vall d'Hebron University Hospital, Barcellona, Spain

**Background and Aim:** The Occlutech Atrial Flow Regulator (AFR®) is a self-expandable double-disc nitinol device with a

central fenestration. Once deployed percutaneously, the central portion of the device stents the atrial septum leaving a pre-selected calibrated atrial communication. Its use has been approved in the adult population with heart failure and described for pulmonary hypertension. Only case reports and small series have been published about its use in the paediatric population and congenital heart disease (CHD).

**Method:** This is a multicenter retrospective registry involving ten centres worldwide. Patients of any age with CHD or patients aged < 18 years with pulmonary hypertension or cardiomyopathy needing AFR implantation were included.

**Results:** From 2017 to 2022, 40 patients underwent AFR implantation. The median age of the population was 58, 5 (IQR 31.5–142.5) months and the median weight was 17 (IQR 10–46) Kg. 26 patients had CHD, 9 children had cardiomyopathy and 5 a structurally normal heart. Indications for implantation were left heart failure in 16 patients (40%), right heart hypertension in 11 patients (27, 5%), severe desaturation in fenestrated Fontan in 5 patients (12, 5%), Fontan failure in 4 patients (10%) and ECMO in 4 patients (10%). AFR implantation required balloon pre-dilatation in 65% of patients. The implantation success rate was 100%. There was a 5% rate of intra-procedural complications, consisting of two immediate AFR occlusions. Two patients with dilated cardiomyopathy on ECMO died during the hospital stay, while another was complicated by cardiac arrest. All the other patients were discharged without complications. At a median follow-up of 330 days (IQR 125–593), 92, 5% of patients were alive. A child died of sepsis 3 months after the procedure. All but one device were patent at follow-up. At follow-up 20 patients improved their NYHA class, 12 didn't change it and only one with idiopathic pulmonary hypertension worsened.

**Conclusions:** AFR implantation in patients with CHD and children with severe pulmonary hypertension or cardiomyopathy is promising and seems to have beneficial effects at short-term follow-up. The AFR has the potential to provide benefits in terms of symptoms and survival to a variety of patients with limited treatment options and indeterminate prognosis.

**Keywords:** Atrial Flow Regulator, congenital heart disease, cardiomyopathy, pulmonary hypertension, ECMO

AFR device



## O-38

**Transcatheter balloon dilation of pulmonary artery band outcomes: A multi-centre study**

Dr. Precyia Philo Fernandes<sup>1</sup>, Dr. Patrick Noonan<sup>1</sup>, Dr. Ryan Abumehdi<sup>2</sup>, Dr. Oliver Stumper<sup>2</sup>, Dr. Matthew Jones<sup>3</sup>, Dr. Sok Leng Kang<sup>4</sup>, Dr. Ines Hribernik<sup>5</sup>, Dr. Jamie Bentham<sup>5</sup>, Dr. Najih Muhammad Liaqath Ali<sup>6</sup>, Dr. Nicholas Hayes<sup>6</sup>, Dr. Abhishek Oswal<sup>7</sup>, Dr. Demetris Taliotis<sup>7</sup>

<sup>1</sup>Royal Hospital for Children Glasgow UK; <sup>2</sup>Birmingham Childrens Hospital UK; <sup>3</sup>Evelina Childrens Hospital London UK; <sup>4</sup>Alder Hey Childrens Hospital Liverpool UK; <sup>5</sup>Leeds Childrens Hospital UK; <sup>6</sup>Southampton General Hospital UK; <sup>7</sup>Bristol Childrens Hospital UK

**Background and Aim:** Pulmonary Artery banding (PAB) is an operation carried out for congenital heart defects, essentially to control increased pulmonary blood flow. This is performed to protect pulmonary vasculature from long term irreversible changes and damage leading to pulmonary hypertension while awaiting complete repair or also as destination therapy. There are very few studies and case reports in the literature that review the long-term outcomes of the patients who underwent percutaneous de-banding of pulmonary artery bands (dPAB).

We aimed to assess the safety and efficacy of balloon dilatation of PAB and review the outcomes.

**Method:** Data was collected from various Cardiothoracic centres within UK according a proposed excel spread sheet after ethical approval. PAB safety was determined from short term outcomes such as immediate post intervention clinical status, length of stay in hospital, survival to discharge, complication rate and long term outcome included survival at 1 year. Efficacy of the procedure was determined by assessing the Invasive and non-invasive gradient and oxygen saturations pre and post procedure.

**Results:** 7 UK wide cardiothoracic centres data from Jan 2011 to Jan 2021 was analysed. A total of 65 patients underwent 69 dPAB procedures. Median age was 12 months and median weight was 9kg at intervention. M:F ratio was 1.4. Average post operative length of stay was 1-2 days and follow up duration was up to 10 years. There was statistically significant reduction in median invasive gradient from 74mmHg to 45mmHg (p value of <0.001) and median transthoracic echo gradient from 54mmHg to 24mmHg (p value of <0.001) post intervention. Post operative complications included 1 extravasation injury and 2 hemopericardium. There was one death associated with concomitant VSD closure. Overall complication rate of 4.3%

**Conclusions:** dPAB appears to be safe and effective to achieve desired loosening of the band. There is low intraoperative and post operative complication rate. Reintervention rate is low in those where the dPAB was performed as single procedure. Limitations of this study was that this was a retrospective study and does not compare results with classic PA bands or surgical outcomes and number of patients was relatively small.

**Keywords:** PAB, pulmonary artery banding dilatation, pulmonary artery debanding

## Cardiac Imaging

## O-39

**Ventricular performance in patients with transposition of the great arteries after arterial switch operation at mid-term follow-up**

Covadonga Terol Espinosa De Los Monteros<sup>1</sup>, Roel L. F. Van Der Palen<sup>1</sup>, Jef Van Den Eynde<sup>2</sup>, Lukas Rammeloo<sup>3</sup>, Mark G. Hazekamp<sup>4</sup>, Nico A. Blom<sup>1</sup>, Irene M. Kuipers<sup>3</sup>, Arend D. J. Ten Harkel<sup>1</sup>

<sup>1</sup>Department of Pediatrics, Division of Pediatric Cardiology, Leiden University Medical Center, Leiden, The Netherlands; <sup>2</sup>Department of Cardiovascular Sciences, KU Leuven, Leuven, Belgium.; <sup>3</sup>Department of Paediatrics, Division of Paediatric Cardiology, Amsterdam University Medical Center, Amsterdam, the Netherlands; <sup>4</sup>Department of Pediatric Cardiac Surgery, Leiden University Medical Center, Leiden, The Netherlands

**Background and Aim:** Ventricular performance after arterial switch operation (ASO) for transposition of the great arteries (TGA) may be impaired at mid- to long-term follow-up, especially in more complex TGA patients such as those with ventricular septal defect (VSD), Taussig Bing anomaly (TBA), aortic arch obstruction, and/or unusual coronary arteries anatomy. With issue Doppler imaging (TDI) and speckle tracking imaging (STI) we can detect more subtle changes in ventricular performance than with conventional echocardiographic parameters. We aimed to investigate ventricular performance with these imaging techniques in a large cohort of TGA patients with simple and more complex anatomy, and compare it to a group of healthy controls (HC).

**Method:** Conventional, TDI and STI echocardiographic parameters were obtained from 144 TGA patients (64.6% male, age 10.7±5.04 years) and 128 HC (52.3% male, age 9.71±5.17 years). The TGA cohort included patients with intact ventricular septum (n = 94), VSD (n = 30) and TBA (n = 20).

**Results:** Whereas conventional echocardiographic parameters suggested preserved function at 10 years post-ASO, TDI and STI parameter demonstrated that TGA patients had impaired biventricular systolic performance compared to HC (LVs' 9±2cm/s vs 10±3cm/s, p<0.001; RVs' 9±cm/s vs 12±2cm/s, p<0.001; Septal s' 6±1cm/s vs 7±1cm/s, p<0.001 (figure A-C); mean 4-chamber longitudinal strain: 16±3% vs 20±2%, p<0.001) (figure F). Similarly, diastolic performance was impaired (LV E/e' 6.4±2 vs 5.9±2, p = 0.004; RV E/e' 6.3±2 vs 4.4±1, p<0.001) (figure D-E). These differences were more evident at younger ages, especially at the apical segments (figure G-H). The TBA subgroup showed more pronounced ventricular impairment, especially at the septal levels, compared to other TGA subgroups. Finally, multivariate linear regression identified older age at ASO, delayed sternal closure and any reoperations/reinterventions post-ASO as predictors worse biventricular performance.

**Conclusions:** Assessment of ventricular performance with TDI and STI showed that TGA patients 10 years after ASO, in particular the

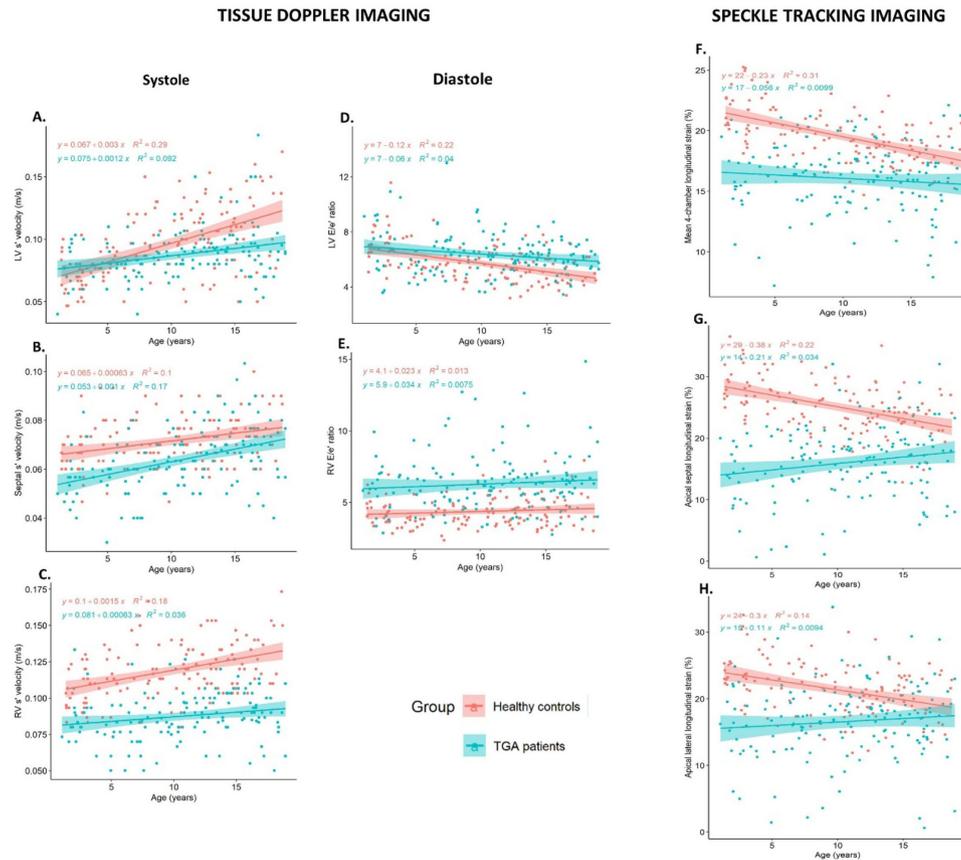
TBA subgroup, have decreased biventricular systolic and diastolic function as compared with healthy controls, especially at the septal regions.

**Keywords:** Transposition of the great arteries, arterial switch operation, cardiac function, tissue Doppler imaging, speckle tracking strain imaging.

Ventricular performance parameters

**Method:** All consecutive patients who were referred for stress perfusion at the Paediatric Cardiac MRI department at the Royal Brompton Hospital from November 2016 to August 2022 were included. The indications, impact on the management and complications were assessed.

**Results:** A total of 70 stress perfusion studies were performed in 65 patients (39 male, 26 female). Thirty-three scans were done in patients with structurally normal heart, 26 in congenital heart dis-



Dot plots and regression lines representing the difference in tissue doppler imaging and speckle tracking imaging parameters between healthy controls (red) and TGA patients (blue), according to age.

## O-40

### Role of stress perfusion in cardiac magnetic resonance imaging in paediatric patients

Palak Gupta<sup>1</sup>, Inga Voges<sup>3</sup>, Raad Mohiaddin<sup>2</sup>, Cemil Izgi<sup>2</sup>, Dudley Pennell<sup>2</sup>, Sylvia Krupickova<sup>2</sup>

<sup>1</sup>Paediatric Cardiology, Royal Brompton Hospital and Harefield NHS Foundation Trust, Guy's and St Thomas' NHS Foundation Trust; <sup>2</sup>Cardiovascular Magnetic Resonance Department, Royal Brompton Hospital, Guy's and St Thomas' NHS Foundation Trust; <sup>3</sup>Department of Congenital Heart Disease and Paediatric Cardiology, University Hospital Schleswig-Holstein, Campus Kiel, Germany

**Background and Aim:** Perfusion imaging enables us to detect and quantify myocardial ischaemia in patients with congenital and acquired heart lesions.

The aim of this study was to evaluate the safety, feasibility and benefit of stress perfusion imaging in children

ease, 6 in cardiomyopathy and 5 in acquired heart disease. General anaesthesia was required in 4 patients (6 studies). Adenosine was used in 34 and Regadenoson in 36 studies to stress the myocardium. Aminophylline was used to reverse the effect of Regadenoson before rest perfusion. Five patients were 0-5 years old, 5 patients 6-10, 33 11-15 and 27 patients 16-18 years of age at the time of the scan. Complications were observed in 1 patient who experienced excessive sneezing, coughing, itching and facial pallor suspicious of allergic reaction to gadolinium contrast or adverse reaction to Aminophylline.

Inducible ischemia was found in 8 patients (11 studies) – 4 with congenital heart defect (2 post-arterial switch operation for transposition of great arteries, 1 post-repair for anomalous origin of left coronary artery (LCA) from pulmonary artery, one post-repair for abnormal origin of LCA from right coronary sinus and intramural course). All these were managed conservatively, one of them underwent cardiac catheterisation. Out of 3 patients with hypertrophic cardiomyopathy, one underwent extended septal myectomy.

One patient with LCA stenosis presumably due to Kawasaki disease presented with out of hospital arrest. He underwent emergency stenting of LCA. He had circumferential perfusion defect on 3 occasions during the follow-up, which helped to guide the interventions. He has been recently referred for cardiac transplantation

**Conclusions:** Stress perfusion imaging is a safe tool for estimating myocardial perfusion in children and has an added value for the management of these patients

**Keywords:** Stress perfusion, ischaemia, children, regadenoson

**O-41**

**More than 25 years experience in diagnosis and treatment of newborns with pulmonary atresia with intact ventricular septum.**

Giulia Tuo, Martino Cheli, Andrea Moscatelli, Dario Paladini, Francesco Santoro, Maurizio Marasini  
G. Gaslini Institute

**Background and Aim:** Pulmonary atresia with intact ventricular septum (PAIVS) is characterized by varying degrees of right ventricle underdevelopment and great morphologic variability of tricuspid valve (TV), pulmonary valve (PV) and coronary arteries. To describe the long-term outcome of newborn patients managed for PAIVS or critical pulmonary valve stenosis (CPS) and to identify pre and postnatal echocardiographic predictors of biventricular repair for those receiving radiofrequency pulmonary valve perforation (PVP) at birth.

**Method:** All patients born between January 1991 and June 2022 with or without a prenatal diagnosis of PAIVS were retrospectively

included. Monopartite (n 16), bipartite (n 39), and tripartite (n 46) right ventricle (RV) subgroups were created on morphologic characteristics at birth. TV and PV annulus z score, tricuspid valve/mitral valve ratio, RV/left ventricular length ratio and preprocedural RV pressure, presence of coronary fistulas, were analyzed in all patients who underwent PVP. Retrospective analysis of fetal population included also degree of tricuspid regurgitation (TR) at last evaluation.

**Results:** 101 patients were diagnosed with PAIVS or CPS of whom 55 during the prenatal period (Figure 1). Eighty-three patients underwent PVP. In table 1 clinical and echocardiographic data of patients submitted to PVP. 33/83 (40%) needed further percutaneous or surgical procedures during the neonatal period. The overall mortality was 6%. 10 patients were lost at follow up. At a median follow up of 8 years (range, 0–32 years), 8 patients underwent a bidirectional Glenn procedure whereas all the other patients achieved a biventricular circulation without any further intervention in 39 of them. Patients who died or had biventricular circulation failure showed a higher incidence of bipartite RV and a lower median tricuspid Z value [-2.75 (range from -4.8 to -1) vs -1.5 (range from -4 to 1); p=0, 003] Table 2. In our experience, fetal TV/MV 0.7, tricuspid z score > -3 and presence of tricuspid regurgitation correlated with a favourable outcome.

**Conclusions:** Tricuspid valve size and RV morphology are the main predictors of outcome in patients with PAIVS or CPS. PVP is an effective first-stage procedure. The right heart appeared to be adequate to maintain a long-term biventricular circulation in the large majority of cases

**Keywords:** Pulmonary atresia, Echocardiography, Percutaneous valve perforation

Table 1. Patients submitted to PVP

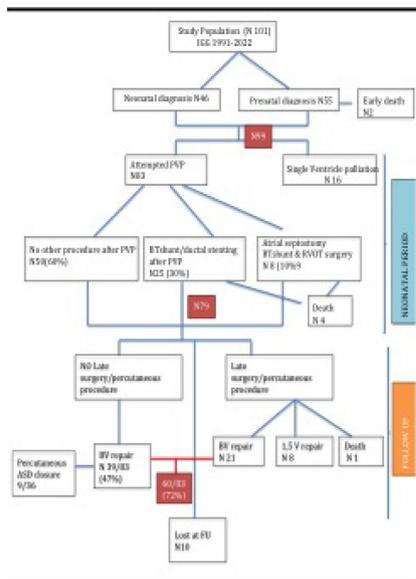
Characteristics of patients	N = 83
GA at birth, (week)	Median (range) 38 (34;41)
Weight, (g)	Median (range) 3000 (1800;4400)
Tricuspid Z value	Median (range) -1.7 (-4.5 ; 1)
TV, (mm)	Median (range) 9 (5 ; 13)
TV/MV ratio	Median (range) 0.75 (0.4;1)
PV, (mm)	Median (range) 7(4.5;10)
Pulmonary valve Z value	Median (range) -1.4 (-4.5 ; 1)
RV/LV ratio	Median (range) 0.76(0.5;1)
preprocedural RVP mmHg	110 (80;200)
Age at PVP,(hours)	Median (range) 48 (1; 147)
Follow up, (yrs)	Median (range) 8.11 (0 ; 31.6)

RV right ventricle; GA, gestational age; PVP pulmonary valve perforation

Table 2. Patients submitted to PVP. Success (BV repair)- Failure (1,5 V repair/death)

Characteristics of patients	Success N=71	Failure N=28	p value
	n (%)	n (%)	
<b>RV morphology</b>			<b>0.001*</b>
Tripartite	46 (64)	1 (3.5)	
Bipartite	25 (35)	13 (46)	
Monopartite	0	14(50)	
	Median (range)	Median (range)	
GE at birth (week)	38 (32 ; 41)	39 (32 ; 40)	0.92
Weight (g)	3000 (2150 ; 4100)	2950 (1800 ; 4400)	0.62
Z value Tricuspid Valve	-1.5 (-4 ; 1)	-2.75 (-4.8 ; -1)	<b>0.003</b>
Tricuspid Valve (mm)	9 (5.5 ; 14)	6 (4 ; 11)	<b>0.001</b>
TV/MV ratio	0.8 (0.5;1)	0.50.4(0.7;)	<b>0.001</b>
PV, (mm)	7 (4.5;10)	4(2;6.5)	<b>0.001</b>
PV Z value	-1.1(-4.5)	-2.5(-1;-9)	<b>0.008</b>
RV/LV ratio	0.8	0.6	<b>0.001</b>
preprocedural RVP mmHg	115	115	<b>0.13</b>
Age at PVP (h)	38 (2; 147)	18.5 (1 ; 62)	0.19
Follow up (yrs)	5.7 (1.2 ; 12.7)	7.1 (1.2 ; 14.1)	0.35

RV right ventricle; GA, gestational age; PVP pulmonary valve perforation. \* test x<sup>2</sup>



Flowchart of study population and data related to patients that underwent PVP

## O-42

**Ebstein's valve rotation angle: A quantitative index of anatomical severity and clinical risk in Ebstein's anomaly**

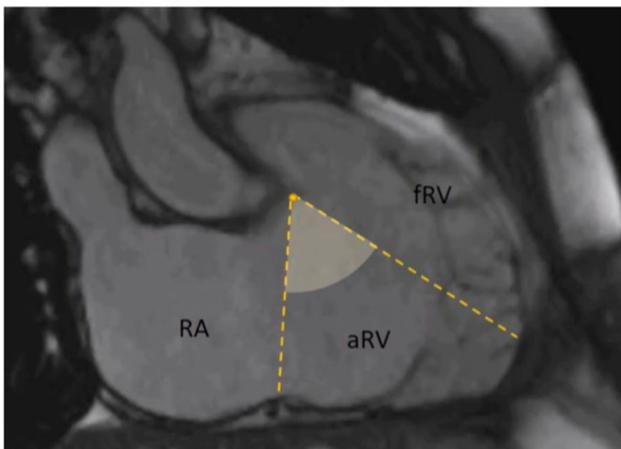
Giulia Pasqualin<sup>1</sup>, Beatrice Cavazza<sup>1</sup>, Francesca Bevilacqua<sup>1</sup>, Andreea Andronache<sup>1</sup>, Martina Evangelista<sup>1</sup>, Teresa Concetti<sup>1</sup>, Luciane Piazza<sup>1</sup>, Luca Giugno<sup>1</sup>, Paolo Ferrero<sup>1</sup>, Massimo Lombardi<sup>2</sup>, Antonio Saracino<sup>1</sup>, Massimo Chessa<sup>1</sup>, Alessandro Giamberti<sup>1</sup>

<sup>1</sup>Pediatric and Adult Cardiology Heart Centre, IRCCS Policlinico San Donato, Milan, Italy; <sup>2</sup>Multimodality Cardiac Imaging Section, IRCCS Policlinico San Donato, Milan, Italy

**Background and Aim:** The study aimed to identify cardiovascular magnetic resonance (CMR) imaging parameters representative of Ebstein's Anomaly (EA) anatomical and clinical severity.

**Method:** Consecutive CMR examinations of patients with EA referred to our Center were evaluated retrospectively. Displacement index (DI) was defined as the distance between the atrio-ventricular groove and the septal leaflet hinge point over the body surface area. The Ebstein's valve rotation angle (EVRA) was quantified as in Figure 1. The Celermajer index was calculated using area (Cel-a) and volumes (Cel-v) as the ratio between the atrialized right ventricle (aRV) plus the right atrium over the sum of the functional right ventricle (fRV) and left chambers. The occurrence of unfavorable outcome was defined as the presence of sustained brady-tachyarrhythmia or New York Heart Association class  $\geq$  II for unoperated patients and the presence of an at least moderate tricuspid regurgitation or a sustained brady-tachyarrhythmia during the post-operative hospital stay for patients undergone surgical correction.

**Results:** The study included 45 patients (age  $28 \pm 15$  years, 64% males). DI, Cel-a, Cel-v and EVRA tended to increase with worsening Carpentier classification ( $p = 0.001$ ,  $p = 0.02$ ,  $p = 0.02$ ,  $p < 0.0001$  respectively). EVRA was the only parameter that showed to discriminate patients with unfavorable outcome both in the group of unoperated ( $58 \pm 27^\circ$  vs.  $30 \pm 27^\circ$ ,  $p = 0.02$ ) and operated patients ( $68 \pm 13.9^\circ$  vs.  $43.5 \pm 26.9^\circ$ ,  $p = 0.03$ ). Among the other analyzed parameters, DI ( $48 \pm 18$  mm/m<sup>2</sup> vs.  $28 \pm 14$  mm/m<sup>2</sup>,



**Figure 1** Cine steady-state free precession sequence showing Ebstein's Valve Rotation angle (EVRA) calculation from a right ventricular 3 chamber view. EVRA is enclosed between a line passing through the anatomical tricuspid anulus and one through the functional valve orifice at end-systole. RA: right atrium; aRV atrialized right ventricle; fRV functional right ventricle

$p = 0.01$ ), aRV indexed end-diastolic-volume (EDVi) ( $31 \pm 22$  ml/m<sup>2</sup> vs.  $68 \pm 45$  ml/m<sup>2</sup>,  $p = 0.03$ ) and anatomical RV (ARV) EDVi ( $128 \pm 32$  ml/m<sup>2</sup> vs.  $192 \pm 81$  ml/m<sup>2</sup>,  $p = 0.03$ ) were significantly higher in unoperated patients with unfavorable follow-up, while both fRV/ARV long axis ( $0.66 \pm 0.2$  vs.  $0.52 \pm 0.14$ ,  $p = 0.04$ ) and volumes ( $p = 0.87 \pm 0.29$  vs.  $0.67 \pm 0.11$ ,  $p = 0.03$ ) were significantly lower. For patients with an unfavorable post-operative stay, FRV-EDVi was significantly higher ( $145 \pm 31$  ml/m<sup>2</sup> vs.  $110 \pm 32$  ml/m<sup>2</sup>,  $p = 0.03$ ). The EVRA value that showed to best discriminate patients who had an unfavorable follow-up event was  $44.5^\circ$  with a sensitivity of 0.57 (0.37–0.76, 95%CI) and a specificity of 0.88 (0.69–0.96, 95%CI).

**Conclusions:** EVRA is an emerging parameter that may help to provide a quantitative assessment of EA anatomical severity and may be integrated in the risk stratification for clinical follow-up and surgical planning.

**Keywords:** Ebstein's, Anomaly, Cardiovascular, Magnetic, Resonance

## O-43

**Double aortic arch: A comparison of fetal cmr, postnatal ct and surgical findings**

Milou PM van Poppel<sup>1</sup>, David FA Lloyd<sup>2</sup>, Johannes K Steinweg<sup>1</sup>, Sujeev Mathur<sup>3</sup>, James Wong<sup>3</sup>, Simone Speggorin<sup>3</sup>, Vita Zidere<sup>3</sup>, Reza Razavi<sup>2</sup>, John M Simpson<sup>2</sup>, Kuberan Pushparajah<sup>2</sup>, Trisha V Vigneswaran<sup>3</sup>

<sup>1</sup>School of Biomedical Engineering & Imaging Sciences, King's College London, London, United Kingdom; <sup>2</sup>School of Biomedical Engineering & Imaging Sciences, King's College London, London, United Kingdom; Department of Congenital Heart Disease, Evelina London Children's Hospital, London, United Kingdom; <sup>3</sup>Department of Congenital Heart Disease, Evelina London Children's Hospital, London, United Kingdom

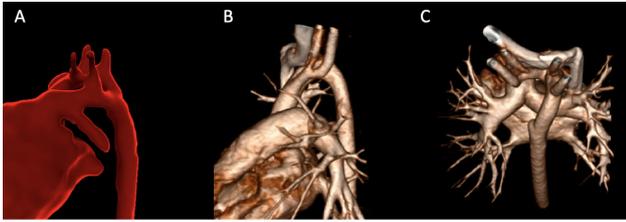
**Background and Aim:** A double aortic arch (DAA) can demonstrate variations in morphology after birth and can be mislabelled as right aortic arch (RAA) if a region of one of the arches becomes atretic. This study aims to highlight the prenatal to postnatal evolution to different morphological DAA types using 3D fetal cardiac magnetic resonance imaging (CMR) and postnatal CT imaging.

**Method:** Fetuses with DAA diagnosis were studied using fetal CMR, and arch morphology was compared to postnatal CT and surgical findings. Three parameters were obtained using fetal CMR: 'isthmus:ductal ratio' (ratio of cross-sectional diameter of LAA isthmus to that of arterial duct), 'Z-angle' (posterior course of LAA quantified by measuring angle between RAA and LAA) and 'isthmal displacement ratio' (distance between LAA isthmus and descending aorta indexed to size of descending aorta).

**Results:** Between 2016–2022, 33 fetuses with DAA underwent fetal CMR, demonstrating in all cases a complete DAA with left sided arterial duct. The RAA was dominant in 31/33(94%). Postnatal CT was undertaken at mean age of 3.1 months (range 0.1–6.9) demonstrating a complete DAA with patency of both arches in 10/33(30%), LAA coarctation was present in seven of these. Atresia of LAA isthmus was present in 22/33(67%), and atresia of the transverse arch between left carotid and left subclavian artery was present in 1/33. Surgery confirmed DAA in all 33 patients (mean age 6.2 months, range 0.1–17.5). Compared to patients with postnatal complete DAA, patients with postnatal atresia of LAA isthmus had a significantly larger mean Z-angle ( $43.7 \pm 9.9$  vs.  $35.6 \pm 4.6$ ,  $p = 0.005$ ), a lower mean isthmus:ductal ratio ( $0.49 \pm 0.1$  vs.  $0.63 \pm 0.15$ ,  $p = 0.033$ ) and higher mean isthmal displacement ratio ( $1.1 \pm 0.3$  vs.  $0.8 \pm 0.3$ ,  $p = 0.016$ ).

**Conclusions:** Fetal CMR provides novel insights into perinatal evolution of DAA. In DAA, the smaller LAA can develop coarctation or atresia following constriction of the arterial duct postnatally, making diagnosis of DAA challenging even with contrast-enhanced CT. Fetal arch morphology in patients with postnatal atresia of LAA isthmus indicates a small and more proximally located LAA isthmus, similar as seen in coarctation of the aorta. There is a potentially important role for prenatal 3D vascular imaging in cases where DAA may be suspected.

**Keywords:** Congenital heart disease, Vascular ring, Magnetic Resonance Imaging, Fetal



**Figure 1** Left lateral view of fetal CMR at gestational age of 29 +4 weeks (Fig1A) and left lateral and superior view of postnatal CT (Fig1 B,C) at 3.5 months showing distal atresia of left aortic arch posterior to left subclavian artery. Surgery was undertaken at age of 4 months where atretic left aortic arch and ligamentum arteriosum were divided.

#### O-44

##### **Cardiac magnetic resonance imaging 6 months after the onset of pims-ts – a retrospective follow-up study**

Thomas Karas<sup>1</sup>, Mario Scala<sup>2</sup>, Ariane Biebl<sup>3</sup>, Jürgen Steiner<sup>1</sup>, Gerald Tulzer<sup>1</sup>

<sup>1</sup>Division of Cardiology, Department of Paediatrics, Kepler University Hospital, Linz, Austria; <sup>2</sup>Division of Radiology, Department of Paediatrics, Kepler University Hospital, Linz, Austria; <sup>3</sup>Division of Infectiology, Department of Paediatrics, Kepler University Hospital, Linz, Austria

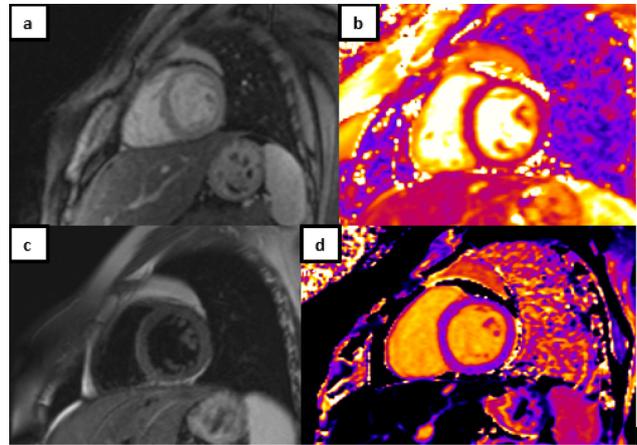
**Background and Aim:** In 2020, reports revealed paediatric cases of significant systemic hyperinflammation and shock with multiorgan involvement called paediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2 (PIMS-TS). A small proportion of children seem to suffer from persistent left ventricular (LV) dysfunction at discharge, but there is uncertainty as to predictive factors toward these patients. The primary aim of this retrospective descriptive single centre study was to investigate if the cardiac damage in PIMS-TS persists during follow-up in these patients.

**Method:** Between December 2020 and February 2022 we included 15 consecutive children hospitalized at the paediatric department of the Kepler University Hospital Linz fulfilling the WHO criteria for PIMS-TS with cardiac involvement. Cardiac magnetic resonance imaging (CMRI) was performed six months after the onset of symptoms to evaluate a possible persistent myocardial damage. **Results:** A total of 15 patients (80% male) with a median age of 9, 1 years were included. MRI-analysis was performed in 12 patients a median of 202 days after the onset of symptoms leading to the diagnosis of PIMS-TS. During admission a majority of our small cohort presented with elevated cardio-specific markers like NT-proBNP

(median 9693 ng/L) and Trop T-hs (median 201, 7 ng/L). A third of patients presented with reduced left ventricular function in first transthoracic echocardiography. In CMRI a majority of patients showed normal values of left ventricular volume. Seventy-three percent (n = 8/11) of individuals had regular LV function (median LVEF 62%) while the rest of the study group showed mild to moderately reduced LVEF-values. No signs of late gadolinium enhancement (LGE) were found, therefore no patient had evidence of persistent myocardial damage.

**Conclusions:** No long-term changes of cardiac function were found in CMRI six months after the onset of PIMS-TS. Regarding our patient cohort this study contributes to the thesis that cardiac impairment during PIMS-TS is not persistent. Nevertheless CMRI could be useful to determine a small group of children, within the population suffering from PIMS-TS, with persistent cardiac involvement during follow up.

**Keywords:** PIMS-TS, Cardiac magnetic resonance imaging, SARS-CoV-2



**Figure 1** Examples of typical cardiac MRI findings in the studied population (a—normal T1W, b—normal T2-TRUE FISP map, c—normal T2W, d—normal T2 map; short-axis views).

#### O-45

##### **Advanced echocardiographic and cmr features of patients with repaired tetralogy of fallot in the long-term follow-up**

Anna Gozzi, Jolanda Sabatino, Jennifer Fumanelli, Angela Di Candia, Martina Avesani, Domenico Sirico, Biagio Castaldi, Elena Reffo, Alessia Cerutti, Roberta Biffanti, Giovanni Di Salvo

Department for Women's and Children's Health, University Hospital of Padova, Pediatric and Congenital Cardiology Unit, Via Nicolò Giustiniani, 2, 35128 Padova, Italy

**Background and Aim:** Tetralogy of Fallot is the most common cyanotic congenital heart disease, spToF survivors are growing rapidly and develop late complications (including sudden cardiac death and life-threatening ventricular arrhythmia). The aim of this study was to focus on advanced echocardiographic and CMR features of patients spToF in a long-term follow-up, finding non-invasive predictors of adverse prognosis.

**Method:** This is a retrospective cohort study including more than 200 patients with repaired ToF, not undergoing pulmonary valve

replacement. EKG, cardiopulmonary exercise testing, echocardiographic and cardiac magnetic resonance data were reviewed, calculating Right Myocardial Work Index (area of the right ventricular pressure strain loops), estimating Right Constructive Work, Wasted Work and Work Efficiency. Patients were screened according to age at surgery (< and >12 months) and time since the repair (< and  $\geq$  25 years), using a composite end-point of major adverse cardiovascular events (sudden cardiac death, sustained/non-sustained ventricular tachycardia, resuscitated sudden cardiac death, or syncope).

**Results:** CPET values weren't significantly different in patients > 25 years since surgery. LV EF measured either by CMR or echocardiography were slightly significantly reduced in patients > 25 years since corrective surgery. Older patients at surgery (>12 months) have significantly reduced CPET VO<sub>2</sub>max, larger RA areas, LA areas and RV basal diameter. During the follow-up 1 patient died suddenly, 5 shew ventricular tachycardia, other 4 syncope. On univariate analysis, echocardiographic LA area/BSA and RV longitudinal strain were significantly related to the combined end point. Patients with reported Holter ECG VT presented significantly reduced RV function (CMR-EF, echocardiographic FAC, RWI). Borderline/impaired predicted VO<sub>2</sub> max at CPET in related to significantly reduced RV FW longitudinal strain and MWI.

**Conclusions:** Long-term survival and clinical conditions after surgery in infancy are good, the late post-surgery functional status could be excellent up to 25 years after repair. Older patients at surgery show reduced functional status compared with earlier repair. The echocardiographic/CMR RV function is related to worse prognosis, recurrent VT, reduced functional status. RV MWI is feasible in patients spTOF; disadvantageous right ventricular work may be a more sensitive indicator of right ventricular impairment compared with standard echocardiographic parameters, and is able to predict exercise capacity.

**Keywords:** Tetralogy of Fallot, Right Ventricular Function, Advanced Echocardiography, Myocardial Work, CMR

## O-46

### Predictive factors of left ventricular dysfunction after pulmonary valve balloon dilation in neonates with critical pulmonary stenosis

Patrizio Moras<sup>1</sup>, Luciano Pasquini<sup>1</sup>, Cosimo Marco Campanale<sup>1</sup>, Marco Masci<sup>1</sup>, Yeva Guk<sup>1</sup>, Valeria Donato<sup>1</sup>, Maria Carolina Colucci<sup>2</sup>, Luca Di Chiara<sup>3</sup>, Alessandra Toscano<sup>1</sup>

<sup>1</sup>Perinatal Cardiology Unit, Department of Medical and Surgical for Fetus-Neonate, Bambino Gesù Children's Hospital IRCCS, Rome, Italy; <sup>2</sup>Unit of Pediatrics, NESMOS Department, Sapienza University c/o Sant'Andrea University Hospital, Rome, Italy; <sup>3</sup>Pediatric Cardiac Intensive Care Unit, Department of Pediatric Cardiology and Cardiac Surgery, Bambino Gesù Children's Hospital, Rome, Italy

**Background and Aim:** Critical pulmonary valve stenosis (PS) is a ductal-dependent cyanotic congenital heart disease that requires pulmonary valve balloon dilation (PVBD) soon after birth. Left ventricular dysfunction following relief of right ventricular compression is a major complication following transcatheter valvuloplasty. Pathophysiological mechanisms are not entirely clear. Aims of this study were to assess global and regional left ventricular (LV) and right ventricular (RV) systolic function before and after PVBD, to identify echocardiographic predictors of post-PVBD LV dysfunction and to provide possible pathophysiological explanations.

**Method:** Neonates who required urgent PVBD from January 2019 to September 2022 at our institution were retrospectively

analyzed. Patients were divided into 2 groups depending on whether they developed post-PVBD LV dysfunction (group 1) or not (group 2). Full echocardiographic assessment was performed at birth and within 14 days after the procedure.

**Results:** Of the 16 neonates identified, 9 (56%) were prenatally diagnosed. Eight patients developed post-PVBD LV systolic dysfunction (ejection fraction  $46.0 \pm 5.6\%$ ) and 7 received inotropes administration. In group 1, peak systolic longitudinal strain of LV septal segments was significantly reduced compared to LV lateral segments ( $-13.92 \pm 2.7\%$  vs.  $-18.47 \pm 2.1\%$ ,  $p < 0.001$ ). At regression analysis, statistically significant relationships were found between TV annulus Z-score ( $p = 0.029$ , OR 5.1, CI 95% 1.0-16.3), pre-PVBD LV GLS ( $p = 0.005$ , OR 3.4, CI 95% 1.7-5.8), pulmonary artery systolic pressure ( $p = 0.040$ , OR 1.2, CI 95% 1.1-1.6) and post-PVBD LV dysfunction.

**Conclusions:** LV dysfunction is a common complication following PVBD. It occurs mainly in the first days after the procedure and often requires inotropes administration. Predictive factors include high TV annulus Z-score, low pre-catheterization LV GLS and high PASP. PVBD LV dysfunction may involve negative ventricular-ventricular interaction, as large the interventricular septum shows the most significant impairments in strain.

**Keywords:** Critical pulmonary valve stenosis, congenital heart disease, strain, speckle-tracking, systolic function

### Predictive factors of LV dysfunction



Predictive factors of post-balloon PV valvuloplasty LV dysfunction are: TV annulus Z-score, pre-valvuloplasty GLS and pulmonary artery systolic pressure. Abbreviations: LV, left ventricular; PV, pulmonary valve; TV, tricuspid valve; GLS, global longitudinal strain

## Congenital Heart Surgery

### O-47

#### Impact of cardiopulmonary bypass on cerebrovascular autoregulation assessed by ultrafast ultrasound imaging

Julien Aguet<sup>1</sup>, Nikan Fakhari<sup>2</sup>, Minh Nguyen<sup>2</sup>, Luc Mertens<sup>2</sup>, Elod Szabo<sup>3</sup>, Birgit Ertl Wagner<sup>2</sup>, Lynn Crawford<sup>3</sup>, Christoph Haller<sup>4</sup>, David Baron<sup>4</sup>, Jerome Baranger<sup>2</sup>, Olivier Villemain<sup>2</sup>

<sup>1</sup>Department of Diagnostic Imaging, The Hospital for Sick Children, Toronto, Canada; <sup>2</sup>Department of Pediatrics, Division of Cardiology, The Hospital for Sick Children, Toronto, Canada; <sup>3</sup>Department of Anesthesia and Pain Medicine, The Hospital for Sick Children, Toronto, Canada; <sup>4</sup>Department of Surgery, Division of Cardiovascular Surgery, The Hospital for Sick Children, Toronto, Canada

**Background and Aim:** Newborns with congenital heart disease (CHD) requiring cardiac surgery with intraoperative cardio-pulmonary bypass (CPB) are at risk of neurodevelopmental impairment. The impact of intraoperative cardiopulmonary bypass (CPB), deep hypothermia and selective cerebral perfusion on the brain is not well understood. Ultrafast power Doppler (UPD) is a novel ultrasound

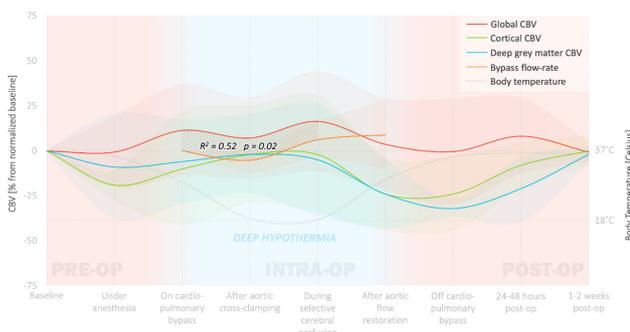
technique able to assess cerebral blood volume (CBV), used as a surrogate of cerebral perfusion. This study aimed to demonstrate the use of UPD to monitor cerebral blood volume (CBV) during cardio-pulmonary bypass in neonates.

**Method:** Freehand transfontanelar ultrafast ultrasound using a 5.7 MHz phased array ultrasound probe (6SD, General Electric) connected to a programmable and portable ultrafast ultrasound scanner (Vantage 256, Verasonics Inc.). Newborns with hypoplastic left heart syndrome (HLHS,  $n=5$ ) selected to undergo a Norwood procedure, were scanned within the first days of life (considered as baseline), intra-operatively at different circulatory conditions (for example under cardio-pulmonary bypass, during selective cerebral perfusion) and post-operatively. Variation in average CBV was expressed in percentage to normalized baseline on mid-sagittal, mid-coronal and parasagittal planes. Associations between CBV and temperature, blood pressure, blood gases and CPB flowrate (CPB-FR) were studied. Maximum CBV variation was compared with brain injury on post-operative MRI.

**Results:** Global CBV was not significantly different between HLHS and controls ( $p=0.98$ ) and between pre- and post-operative ( $p=0.62$ ). UPD was able to monitor changes in CBV throughout cardiac surgery, revealing regional differences in CBV during hypothermia suggesting decreased flow to the basal ganglia despite increased global CBV. During deep hypothermia, only CBV correlated with CPB-FR ( $R^2=0.52$ ,  $p=0.02$ ) (see Figure). Good intra-observer agreement was shown (mean difference:  $-0.01$ ; limits of agreement:  $-0.29$ - $0.27$ ). Maximum CBV variation was  $\leq 15\%$  in one HLHS patient, the only without any brain injury on MRI. All other patients had higher CBV variation (mean:  $35\%$ , range:  $15$ - $47\%$ ) and showed brain injury on MRI.

**Conclusions:** UPD can assess global and regional brain perfusion variation during neonatal cardiac surgery. This first intra-operative application demonstrated an association between CBV and CPB-FR, suggesting loss of cerebrovascular autoregulation. Measurement of CBV by UPD could enable optimization of cerebral perfusion during cardiac surgery in neonates.

**Keywords:** ultrafast power Doppler, brain perfusion, deep hypothermia, cardio-pulmonary bypass



**Figure** Global and regional cerebral blood volume in neonates ( $n=5$ ) undergoing cardiac surgery with cardio-pulmonary bypass. Variation of global and regional average cerebral blood volume (CBV) compared to bypass flow-rate over time from pre-operative baseline to 1-2 weeks after surgery with significant association between global CBV and bypass flow-rate during deep hypothermia ( $R^2=0.52$ ,  $p=0.02$ ). CBV is expressed in % to normalized pre-operative baseline and represented with  $\pm 1$  SD. Bypass flow-rate is expressed in % to normalized baseline flow at initiation.

## O-48

### A new method of aortic root harvesting in complex transposition of the great artery with pulmonary stenosis and coronary arteries anomaly.

Tomasz Nalecz<sup>1</sup>, Alexandre Pelouze<sup>1</sup>, Maurice Beghetti<sup>2</sup>, Julie Wacker<sup>2</sup>, Tornike Sologashvili<sup>1</sup>

<sup>1</sup>Cardiovascular Surgery Unit, University Hospitals of Geneva, Geneva, Switzerland; <sup>2</sup>Pediatric Cardiology Unit, University Hospitals of Geneva, Geneva, Switzerland

**Background and Aim:** In complex cases of transposition of the great arteries (TGA) with pulmonary stenosis, when Rastelli procedure cannot be performed (because of inlet ventricular septal defect, or atrioventricular valve straddling), surgical options are Nikaidoh or double root rotation. However, associated coronary arteries anomaly, such as LAD originating from the right coronary artery and crossing the right ventricular infundibulum, often disqualify this group of patients for a biventricular reparation. We recently developed a new surgical technique allowing for aortic root harvesting in the setting of coronary arteries anomaly.

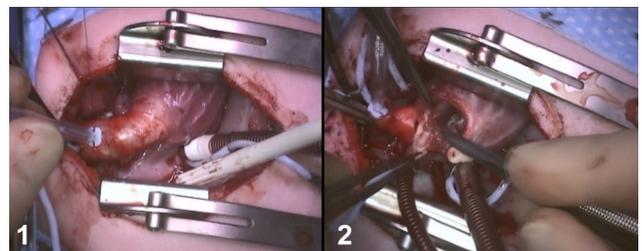
**Method:** We present our small experience with this technique. The aortic root is harvested with minimal muscle margins, of less than 2 millimeters. (Surgical technique video/photo).

**Results:** We used this aortic root harvesting technique in two patients undergoing double root rotation for complex TGA, pulmonary stenosis and coronary arteries anomaly. One patient was 5 days old and weighed 3 kg, she is doing well without aortic regurgitation (AR) or stenosis at 2 years follow up. The other patient was 5 weeks old, and 4 kg. He has minimal AR at one month follow up. We did not notice aortic valve destabilization, and no left ventricular outflow tract obstruction in our two patients.

**Conclusions:** Coronary arteries anomaly should not be a contraindication for Nikaidoh or double root rotation providing that the aortic root is harvested with our technique, using the smallest muscular margins possible. Application of this technique to double root rotation without coronary artery anomalies may be of benefit, minimizing the trauma on the right ventricle, but warrants further research.

**Keywords:** Aortic root harvesting, Nikaidoh, Double root rotation, coronary anomalies, complex transposition of the great arteries

### Root harvesting technique



1. LAD originating from the right coronary artery and crossing the right ventricular infundibulum 2. Aortic root harvesting

## O-49

**Ukrainian 25 years experience of ross procedure**

Oleksandr Romaniuk<sup>1</sup>, Yuliia Klymyshyn<sup>2</sup>, Ihor Lebid<sup>2</sup>, Nadiia Rudenko<sup>1</sup>, Ilya Yemets<sup>1</sup>

<sup>1</sup>Ukrainian Children's Cardiac Center, Kyiv, Ukraine. Shupyk National Medical Academy of Postgraduate Education, Kyiv, Ukraine; <sup>2</sup>Ukrainian Children's Cardiac Center, Kyiv, Ukraine.

**Background and Aim:** To examine long-term outcomes after Ross procedure (RP) for twenty-five year period in UCCC, Ukraine. **Method:** Ross procedure was performed in 349 consecutive patients from 1996 to 2021. The mean age at the time of operation was 132±101 months (from 0, 3 to 648 months). 264 patients were male - 76% and 85 (24%) - female. Newborn patients were 16 (4, 5%), age less than 1 year was 33 (9, 5%), 1 to 18 years 254 (73%) and more than 18 years - 46 (13%). Aortic stenosis was identified in 177 (51%) patients, aortic insufficiency in 132 (38%) and concomitant lesion in 40 (11%) patients.

**Results:** Overall mortality was 25 (7, 2%) patients, in hospital mortality was in 16 (4, 5%) patients, 9 (2, 7%) died during the long-term follow-up. Follow-up was 96% complete. Regression and correlation statistical analysis were used to assess the results.

NeoAoV reoperation required in 13 (3, 9%) patients (in 5 cases - repair of AoV and in 8 cases - AVR). Indications for reoperation were severe neoAV insufficiency and AscAo dilatation, n = 12 (5, 1%), reSubAo stenosis n = 1 (0, 5%). 56 (17%) patients had the RV-PA conduit procedure - conduit replacement. Independent predictors of autograft reoperation were older age (more than 11y) at the time of surgery (r = 0, 01, p = 0, 023) and preoperative aortic regurgitation (r = 0, 1, p = 0, 04). Factors which decrease risks of autograft disfunctions and reoperations are aortic root reinforcement (r = -0, 165, p = 0, 043), congenital aortic valve anomaly (r = -0, 249, p = 0, 002), bicuspid aortic valve (r = -0, 285, p = 0, 001). Independent predictors of RV-PA connection reoperation were younger age at the time of surgery (r = -0, 174, p = 0, 03) and small conduit size (20 mm and less) (r = -0, 38, p = 0, 001).

**Conclusions:** UCCC have the highest experience of RP in Ukraine. RP is an operation with low mortality and good long-term results. Reoperations on neoAoV was rare 3.9%. Conduits of RV-PA require more reoperations (17%) than neoAoV and it is important to modified further study.

**Keywords:** Ross procedure, aortic valve, noeaortic valve, pulmonary artery, conduit.

## O-50

**Truncus arteriosus repair at the red cross war memorial hospital, cape town, south africa: A 20-year review of surgical practice and outcomes**

Allen Moodley<sup>1</sup>, Heidi Meyer<sup>2</sup>, Paul Human<sup>1</sup>, Liesl Joanna Zühlke<sup>3</sup>, Liesl Joanna Zühlke<sup>4</sup>, Andre' Brooks<sup>1</sup>

<sup>1</sup>Department of Cardiothoracic Surgery, University of Cape Town, South Africa; <sup>2</sup>Department of Anesthesia and Perioperative Medicine, University of Cape Town, South Africa; <sup>3</sup>Medical Research Council Francie Van Zyl Drive Tygerberg, Cape Town, South Africa; <sup>4</sup>Division of Paediatric Cardiology, Department of Paediatrics Faculty of Health Sciences, University of Cape Town, South Africa

**Background and Aim:** A description of the post-operative outcomes following Truncus Arteriosus (TA) repair over 20 years before and following the transition to non-conduit repair. Primary outcomes were 30-day and overall, in-hospital mortality in paediatric patients who underwent TA repair at Red Cross War Memorial

Children's Hospital (RCWMCH). Secondary outcomes encompassed (a) Incidence of postoperative complications and (b) medium-term outcomes, including reinterventions, late deaths, and loss to follow-up.

**Method:** A single-centre retrospective study of all consecutive patients who undertook the repair of TA from January 1999 to December 2018 at RCWMH. Patients with an interrupted aortic arch or previous pulmonary artery banding were excluded.

**Results:** Fifty-four patients had TA repair during the study period. Thirty-four (63.0%) patients had a conduit repair, and 20 (37.0%) patients had a non-conduit repair. There were 2 intraoperative deaths. Thirty-day in-hospital mortality was 22.2%. Overall, in-hospital mortality was 29.6%.

Twenty-nine (55.8%) of fifty-two patients suffered a postoperative complication. No major disparity in the number of postoperative complications amongst the cases who underwent a conduit versus a non-conduit repair (p = 0.675). The most common overall complications were cardiovascular and infective. Overall, 10/54 (18.5%) developed low cardiac output syndrome (LCOS) in the post-operative period. LCOS was similar between the conduit group (6/34, 17.6%) and the non-conduit group (4/20, 20%). Thirteen of fifty-two (24.1%) patients had an infective complication. The number of infective complications was similar between the two groups (p = 1.0). Of the eighteen patients with pre-operative moderate truncal valve regurgitation, nine (50%) patients had residual moderate postoperative truncal valve regurgitation.

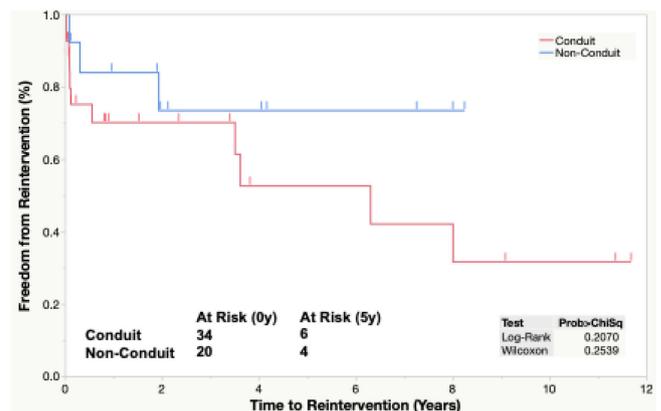
A total of 38 patients were followed up post-hospital discharge with 11 patients (28.9%) lost to follow-up and 8 (21.1%) late mortalities observed.

The actuarial survival for the conduit group was 77.5%, 53.4% and 44.5% at 6, 12 and 27 months respectively and the non-conduit group was 58.6% at 6 months. The overall freedom from re-operation between the conduit group and the non-conduit group was 66.2% vs 86.5%, 66.2% vs 76.9% and 29.8% vs 64.1% at 1, 2 and 8 years respectively.

**Conclusions:** There was no difference in postoperative mortality between the conduit and non-conduit repair. Reintervention rates were lower in the non-conduit group.

**Keywords:** Truncus Arteriosus Surgery, Aortopulmonary Septal Defect, Persistent Truncus Arteriosus, Cardiac Septal Defects, Congenital Heart Defect

Kaplan Meier: Reinterventions



Reinterventions on Truncus Arteriosus Repair comparing conduit to non-conduit repair

## O-51

**Arterial switch for transposition of the great arteries with intact ventricular septum: findings from linked national datasets on 17 years of practice**

Ferran Espuny Pujol<sup>1</sup>, Dan Mihai Dorobantu<sup>2</sup>, Martin Kostolny<sup>3</sup>, Katherine L Brown<sup>3</sup>, Rodney C Franklin<sup>4</sup>, Sonya Crowe<sup>1</sup>, Christina Pagel<sup>1</sup>, Serban C Stoica<sup>5</sup>

<sup>1</sup>Clinical Operational Research Unit (CORU), Department of Mathematics, University College London, UK; <sup>2</sup>Children's Health and Exercise Research Centre (CHERC), University of Exeter, Exeter, UK; <sup>3</sup>Population Health Sciences, University of Bristol, Bristol, UK; <sup>4</sup>University Hospitals Bristol and Weston National Health Service Foundation Trust, Bristol, UK; <sup>5</sup>Heart and Lung Division, Great Ormond Street Hospital NIHR Biomedical Research Centre, London, UK; <sup>6</sup>Department of Paediatric Cardiology, Royal Brompton and Harefield National Health Service Foundation Trust, London, UK; <sup>7</sup>University Hospitals Bristol and Weston National Health Service Foundation Trust, Bristol, UK

**Background and Aim:** Long term mortality and reintervention after surgery to correct transposition of the great arteries with intact septum (TGA-IVS) are favourable according to single centre studies. Less is known about hospital resource utilization and the impact of treatment choices and timing on these outcomes. We aim at describing survival, reintervention and hospital resource utilization in the years after arterial switch operation (ASO); exploring predictors for late mortality and reintervention; investigating the impact of balloon atrial septostomy (BAS) as procedure preceding ASO, as opposed to ASO only.

**Method:** Clinical follow-up data and life status for all patients undergoing ASO between April 2000 and March 2017 in England and Wales were collected from five linked national (registry and administrative) datasets and explored using multivariable regressions (Cox, cause-specific hazards) and matching.

**Results:** A total of 1772 patients (12 centres) were included, with median ASO age of 9.5 days (IQR 6.5, 14.5) which slightly decreased over time. Mortality and cardiac reintervention at 30 days after ASO were 1.7% and 2.0% respectively, and at 10 years after ASO were 3.2% (95% CI 2.5%, 4.2%) and 10.7% (9.1%, 12.2%). Patients stayed a median of 7 days (IQR 5, 11) in intensive care during ASO spell: all of them received invasive ventilation and inotrope support, while 3.5% required ECMO and 14.0% renal support. Median time spent in hospital during the ASO spell was 19 days (IQR 14, 24), and over the following year 7 days

(IQR 4, 11), decreasing to 1 outpatient day/year beyond the 5th year. ASO age and BAS use were not associated with late mortality and reintervention, but cardiac or congenital comorbidities, low weight, and need for more intensive care at ASO were. Matching for patient characteristics, BAS followed by ASO and ASO as first procedure, performed within the first 3 weeks of life, had comparable early and late outcomes, including hospital resource utilization.

**Conclusions:** Mortality and hospital resource utilization are low, while the risk of reintervention remains elevated. The current approach of early ASO and individualised use of BAS allows for flexibility in treatment timing and choices, and a focus on reducing mortality for subgroups at clinical risk.

**Keywords:** transposition of the great arteries, arterial switch, balloon atrial septostomy, multicentre study, reintervention, hospital resource utilization

## O-52

**Two year clinical, functional and histological outcome of a tissue engineered valvular pulmonary conduit evaluated in a chronic growing animal model**

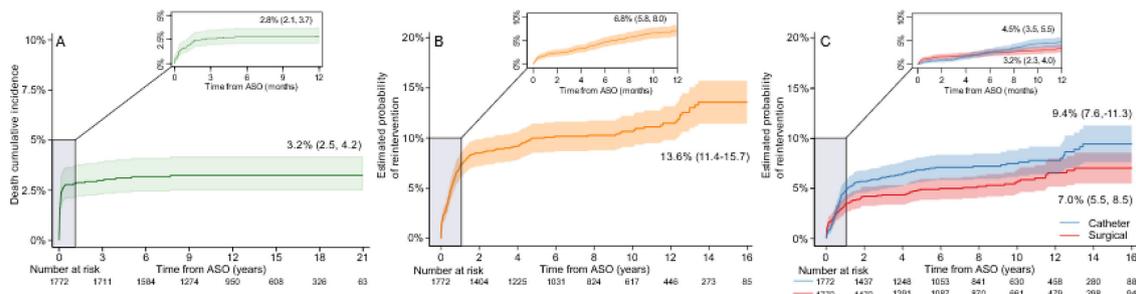
Walter Knirsch<sup>1</sup>, Bernard Krüger<sup>2</sup>, Alexandra Malbon<sup>3</sup>, Thea Fleischmann<sup>4</sup>, Miriam Weisskopf<sup>4</sup>, Thomas Frauenfelder<sup>5</sup>, Hitendu Dave<sup>6</sup>, Frithjof Lemme<sup>6</sup>, Michael Hübler<sup>6</sup>, Martin Schweiger<sup>6</sup>

<sup>1</sup>Pediatric Cardiology, Pediatric Heart Center; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland; <sup>2</sup>Cardiac Anesthesia, Institute of Anesthesiology, University Hospital; Anesthesiology, University Children's Hospital Zurich, Switzerland; <sup>3</sup>Institute for Veterinary Pathology Vetsuisse Faculty, University of Zurich, Switzerland; <sup>4</sup>Center for Surgical Research and Central Biological Laboratory, University Hospital Zurich, Switzerland; <sup>5</sup>Institute of Diagnostic and Interventional Radiology, University Hospital Zurich, Switzerland; <sup>6</sup>Congenital Cardiovascular Surgery, Pediatric Heart Center; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland

**Background and Aim:** Previously, we established a chronic growing lamb model to test a self-constructed valvular pulmonary conduit, using a biologic (porcine) decellularized small intestinal submucosal extracellular matrix scaffold.

**Method:** The tissue engineered valved pulmonary conduit was implanted in orthotopic position in 19 lambs. Here, the two-year

## Mid and late outcomes after ASO in simple TGA-IVS



Panel A: Probability of death (Kaplan-Meier) over 21 years. Panel B: Probability of cardiac reintervention conditional on survival (Conditional Probability Function) over 16 years. Panel C: Probability of cardiac reintervention conditional on survival (Conditional Probability Function) by reintervention type (surgical or transcatheter). Survival median (IQR; max) follow-up is 12.4 (8.6,16.7;21.8) years; reintervention follow-up is 5.4 (4.4,12.8;16.9) years. All inserts are enlarged for the 1st year of follow-up. ASO, arterial switch operation; TGA-IVS, transposition of the great arteries with intact septum.

follow-up results of surviving animals are reported: Clinical outcome, laboratory markers, in-vivo conduit functionality assessed by transoesophageal echocardiography (TEE) and CT-scan, and post-mortem histopathological analysis. Each pulmonary valve leaflet along with the conduit wall was histologically assessed by using the following stains: hematoxylin-eosin, Elastic van Gieson, CD31,  $\alpha$ -smooth muscle actin, Alcian blue, Von Kossa, and Masson's trichrome. Data are presented as number (%) or median (range).

**Results:** Seven (36.8%) lambs died within the first two days after surgery due to technical reasons (learning curve). During short-term follow-up three lambs died unexpectedly. Follow-up was available in nine sheep undergoing planned euthanasia at 9, 12, 18 and 24 months. Clinically, up to 24 months follow-up body weight increased from 33 (27–38) kg to 59 (44–72) kg, ( $p = 0.002$ ). No clinical or laboratory signs of haemolysis were present. The TEE examination of the valved conduit at last follow up showed peak and mean systolic Doppler pressure gradients of 18.8 (7–21) and 7.8 (3–13) mmHg, respectively. Valvular regurgitation was trivial ( $n = 4$ ), mild ( $n = 3$ ), moderate ( $n = 1$ ) or severe ( $n = 1$ ). Premortem CT scan revealed no conduit or leaflet calcification. Histopathological examination was performed in all sheep euthanised from 12 months onwards. Within each animal variability existed between the leaflets. Overall, a low (<25%) to moderate (25%–75%) stromal re-population was found with a low rate of endothelialisation but a relatively high rate of chondroid and osseous metaplasia and an ongoing mild-to-moderate inflammatory process. The conduit wall generally showed a much greater degree of incorporation into the native vessel and fewer detrimental reactive changes (metaplasia and inflammation).

**Conclusions:** While functional aspects of tissue engineered valvular pulmonary conduits seem satisfactory up to 24 months follow-up, histologic findings show strong disadvantages regarding the concept of cell repopulation and regeneration of the valve leaflets, which consecutively limits its clinical use.

**Keywords:** Valve, animal, tissue engineering, outcome

## O-53

### Delayed sternal closure following complex cardiac surgery in neonates

Amr Ashry<sup>1</sup>, Shubhi Gupta<sup>2</sup>, Hesham Ahmed<sup>3</sup>, Ramesh Kutty<sup>4</sup>, Ram Dhannapumeni<sup>4</sup>, Rafael Guerrero<sup>5</sup>, Attilio A. Lotto<sup>6</sup>

<sup>1</sup>Department of Paediatric Cardiac Surgery, Alder Hey Children's Hospital, Liverpool, United Kingdom; <sup>2</sup>Department of Cardiothoracic Surgery, Assiut University Hospital, Faculty of Medicine, Assiut University, Assiut, Egypt.; <sup>3</sup>School of Medicine, University of Liverpool, Liverpool, United Kingdom.; <sup>4</sup>Department of Cardiothoracic Surgery, Faculty of Medicine, Menoufia University, Menoufia, Egypt.; <sup>5</sup>Department of Paediatric Cardiac Surgery, Alder Hey Children's Hospital, Liverpool, United Kingdom.; <sup>6</sup>Department of Paediatric Cardiac Surgery, Alder Hey Children's Hospital, Liverpool, United Kingdom.; <sup>7</sup>Department of Paediatric Cardiac Surgery, Assiut University Hospital, Assiut, Egypt.; <sup>8</sup>School of Medicine, University of Liverpool, Liverpool, United Kingdom.; <sup>9</sup>Department of Paediatric Cardiac Surgery, Assiut University Hospital, Assiut, Egypt.; <sup>10</sup>School of Medicine, University of Liverpool, Liverpool, United Kingdom.; <sup>11</sup>Department of Paediatric Cardiac Surgery, Assiut University Hospital, Assiut, Egypt.; <sup>12</sup>School of Medicine, University of Liverpool, Liverpool, United Kingdom.; <sup>13</sup>Department of Paediatric Cardiac Surgery, Assiut University Hospital, Assiut, Egypt.; <sup>14</sup>School of Medicine, University of Liverpool, Liverpool, United Kingdom.; <sup>15</sup>Department of Paediatric Cardiac Surgery, Assiut University Hospital, Assiut, Egypt.; <sup>16</sup>School of Medicine, University of Liverpool, Liverpool, United Kingdom.

**Background and Aim:** Delayed sternal closure (DSC) is a well-known strategy for management of neonates following complex cardiac procedures requiring prolonged cardiopulmonary bypass (CPB) times and/or deep hypothermic circulatory arrest, which predispose to myocardial oedema, low cardiac output and haemodynamic instability. The purpose of this study is to evaluate clinical

outcomes and morbidity associated with DSC in this group of patients.

**Method:** Retrospective review of neonates who underwent DSC after complex congenital cardiac surgery in a single centre from 2015 to 2021. Out of 357 neonates who had cardiac surgery via median sternotomy, we identified 187 cases who had DSC.

**Results:** Mean age and weight were  $12.8 \pm 6.8$  days and  $3.3 \pm 0.5$  Kg, respectively. Mean RACHS-1 Score was  $4 \pm 1$ . Arterial Switch operation for either simple or complex TGA, hypoplastic aortic arch repair associated or not with VSD and Norwood Operation were the most frequent performed procedures. Mean aortic cross clamp time and CPB time were  $102.8 \pm 52.5$  and  $183 \pm 81$  minutes. Mean days of opened chest were  $3.8 \pm 5.8$  days. Mean ICU and hospital stay were  $12.8 \pm 16.6$  and  $25.9 \pm 36.9$  days. Intra-operative ECMO was required in 6 cases (3.2%), while 21 patients (11.2%) needed post-operative ECMO for ECPR. 30-day hospital mortality was 4.8% ( $N = 9$ ). 8 out of 27 patients (29.6%) who required ECMO died. Only 2 patients (1.07%) needed sternal wound debridement for deep wound infection, while 19 patients (10.2%) had superficial wound infection that was managed conservatively. Univariate analysis showed that DSC days ( $p = 0.011$ ), ECMO ( $p = 0.0001$ ), aortic cross clamp time ( $p = 0.007$ ) and CPB time ( $p = 0.006$ ) associated with 30-day mortality, while in multivariate analysis, only ECMO was significant ( $p = 0.002$ ) for mortality. However, only RACHS-1 score was an independent risk factor for sternal wound infection in both univariate ( $p = 0.019$ ) and multivariate analysis ( $p = 0.052$ ).

**Conclusions:** Delayed sternal closure (DSC) is a safe and effective therapeutic option following complex cardiac surgery in neonates. DSC might also speed up establishing ECMO during ECPR which nonetheless remains a risk factor for mortality. Moreover, higher RACHS-1 score is associated with sternal wound infection.

**Keywords:** Delayed sternal closure, Neonates, ECMO, Congenital Cardiac Surgery

## O-54

### Management of congenitally corrected transposition of the great arteries: A 25-year single center experience.

Irene M. Kuipers<sup>1</sup>, Roel L.f. Van Der Palen<sup>2</sup>, Dave R. Koolbergen<sup>4</sup>, Timofey Nevvazhay<sup>4</sup>, Nico A. Blom<sup>3</sup>, Mark G. Hazekamp<sup>4</sup>

<sup>1</sup>Department of Pediatrics, Division of Pediatric Cardiology, Amsterdam University Medical Centre, Amsterdam, the Netherlands; <sup>2</sup>Department of Pediatrics, Division of Pediatric Cardiology, Leiden University Medical Centre, Leiden, The Netherlands; <sup>3</sup>Department of Pediatrics, Division of Pediatric Cardiology, Leiden University Medical Centre, Leiden & Department of Pediatrics, Division of Pediatric Cardiology, Amsterdam University Medical Centre, Amsterdam, The Netherlands; <sup>4</sup>Department of Cardiothoracic Surgery, University Hospital Leiden, Leiden & Department of Cardiothoracic Surgery, Amsterdam University Medical Centre, Amsterdam, The Netherlands

**Background and Aim:** To assess outcomes of individualized management strategies for congenitally corrected transposition of the great arteries (ccTGA) based on a 25-year single-center experience.

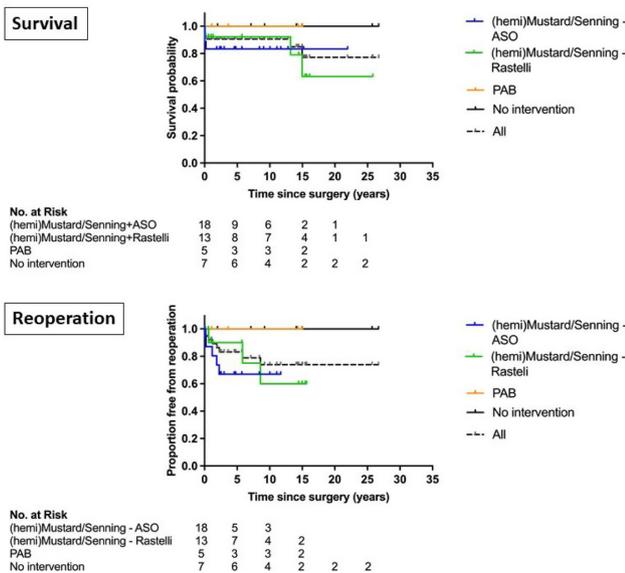
**Method:** From 1998 to 2022, 43 patients underwent different treatment strategies: Senning/Mustard with arterial switch (ASO) (group A, 18), Senning/Mustard with Rastelli (group B, 13) [24 had bidirectional cavopulmonary anastomosis with hemi-Mustard], pulmonary artery banding (PAB) (group C, 5), no intervention (group D, 7). Patient records, operative reports and follow-up echocardiography images were reviewed.

**Results:** Median age at corrective surgery was 2.5 (IQR 1.5–3.5), 3.3 (IQR 1.4–5.7) and 3.2 (IQR 0.7–9.3) years (groups A, B, C).

In group A and B, 17 and 10 patients had previous surgical interventions, respectively. In-hospital mortality occurred in 4 patients (group A: 3; group B: 1); late death in 2 (group B, one non-cardiac related death, sepsis)(Figure). Two patients were lost-to-follow-up. Median follow-up of hospital survivors was 8.4 (IQR 3.0-11.7), 13.8 (IQR 1.1-15.6), 14.3 (IQR 2.3-15.0) and 14.1 (IQR 7.1-25.7) years (group A, B, C, D). Eight patients were reoperated: RV-PA conduit replacement (2), aortic valve replacement (1), valve sparing root replacement (1), baffle obstruction (3) and residual VSD closure (1) (Figure). Three patients needed percutaneous interventions for pulmonary artery stenosis. Four patients received PAB as destination therapy, one patient is waiting for hemi-Mustard-Glenn-ASO. A 'no intervention' strategy was followed in 2 patients with VSD and pulmonary stenosis and in 6 without associated lesions. The ejection fraction of the systemic (LV) ventricle in group A, B is median 48.0 (IQR 43.3-54.5) %, the ejection fraction of the systemic (RV) ventricle in group C, D is median 51.0 (IQR 44.0-52.0)%. Tricuspid valve regurgitation improved in all except one of 9 patients with (moderate or more) tricuspid valve regurgitation in group A, B, C. All patients are in NYHA class I, except one patient (group A) who is in NYHA class III.

**Conclusions:** Individualized treatment is important. No surgery for isolated ccTGA seems reasonable. In patients with late diagnosis palliative PAB can replace anatomical repair while preserving systemic RV function and preventing important tricuspid insufficiency. After early learning curve anatomical repair of ccTGA results in durable good outcomes.

**Keywords:** ccTGA, surgical management, banding, anatomical repair



Survival and Reoperation Kaplan Meier Curve

O-55

**Transcatheter interventions in children with functionally univentricular heart disease: A 21 year single centre experience**

Richard Parry<sup>1</sup>, Adrian Crucean<sup>2</sup>, Vinay Bhole<sup>1</sup>, Paul Miller<sup>1</sup>, Rami Dhillon<sup>1</sup>, Phil Botha<sup>2</sup>, Natasha Khan<sup>2</sup>, Tim Jones<sup>2</sup>, Chetan Mehta<sup>1</sup>, Milind Chaudhari<sup>1</sup>, Oliver Stumper<sup>1</sup>

<sup>1</sup>Department of Cardiology, Heart Unit, Birmingham Women's and Children's NHS Foundation Trust, Birmingham, United Kingdom;

<sup>2</sup>Department of Cardiac Surgery, Heart Unit, Birmingham Women's and Children's NHS Foundation Trust, Birmingham, United Kingdom

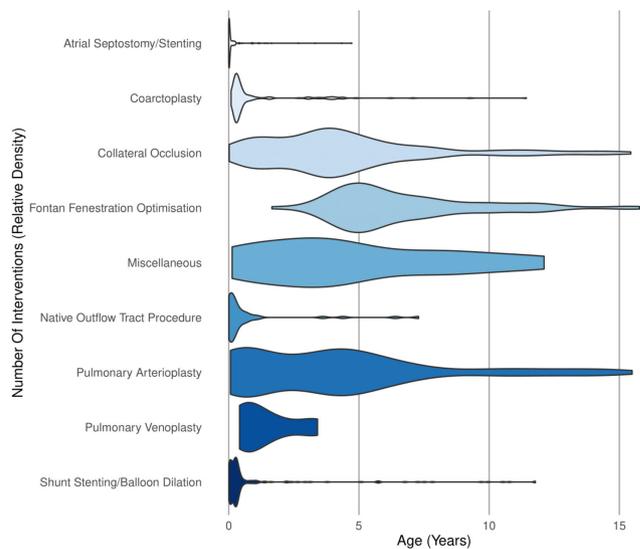
**Background and Aim:** Functionally Univentricular Heart disease (FUVH) comprises markedly heterogeneous anatomical substrates with additional myocardial, vascular, conduction and lymphatic abnormalities. We characterise the common transcatheter interventions required during staged palliation for preparation and maintenance of an adequately functioning Fontan circulation.

**Method:** A retrospective analysis of all patients with FUVH treated at Birmingham Children's Hospital between 01/01/2000 and 31/12/2021. Details of catheter interventions and outcome were recorded and analysed.

**Results:** 982 patients were included, of whom 526 (53.6%) had HLHS. 1,355 interventions were carried out in 577 patients (58.8% of patients). The most common interventions were pulmonary angioplasty (289, 21.3%), collateral occlusion (229, 16.9%), Fontan fenestration optimisation (211, 15.6%), and shunt stenting/balloon dilation (204, 15.1%). The most common interventions at each stage were atrial septostomy (76.5%) before stage I palliation (the majority in non HLHS patients), shunt stenting / balloon dilation (32.9%) between I and II, collateral occlusion (42.0%) between II and III, and modulating Fontan fenestration size (57.3%) post III. There were four transcatheter Fontan takedowns.

Patients with HLHS were more likely to receive transcatheter intervention between stage I and II of palliation (p<0.001) and early (<=30 days) after stage III palliation (p = 0.025), but less likely before stage I (p<0.001). Risk factors for late (>30 days) post stage III transcatheter intervention included HLHS (p = 0.028) and early post stage III transcatheter intervention (p<0.001). Transcatheter intervention before stage III palliation was not associated with late post stage III transcatheter intervention (p = 0.200), nor freedom from death or cardiac transplant after stage III palliation (p = 0.490). Freedom from transcatheter reintervention late after stage III palliation at 16 years of age was 47.2%.

Safety was excellent, with 5 deaths on table after failed bail out surgery (0.4%), 14 urgent transfers to surgery (1.0%), and 20 unplanned admissions to PICU (1.5%). Of the 1,327 interventions with sufficient data, 1,234 (93.0%) of attempted interventions were a technical success.



**Figure 1** A violin plot showing the relative density of common transcatheter interventions at given ages.

**Conclusions:** In a contemporary cohort of FUVH patients, catheter interventions provide a safe and effective treatment option for various anatomical, physiological, conduction and lymphatic complications throughout childhood. A highly specialised catheter interventional service is integral to the overall success of a FUVH management programme.

**Keywords:** Fontan, Interventional Cardiology

## O-56

### Right ventricular remodelling after covered stent correction of sinus venosus asds

San Fui Yong, Saleha Kabir, Matthew Jones, Shakeel Qureshi, Eric Rosenthal  
Department of Paediatric and Adult Congenital Heart Disease, Evelina London Children's Hospital, Guy's and St Thomas' NHS Foundation Trust, London, United Kingdom

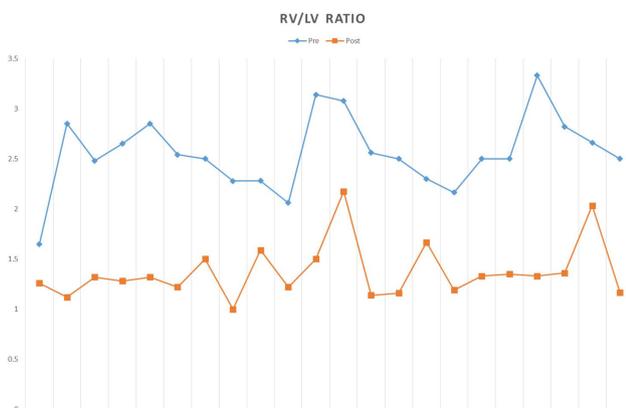
**Background and Aim:** Transcatheter correction of sinus venosus ASDs using 10 zig covered Cheatham-Platinum stents has been undertaken in our unit since 2016. Whilst there is increasing experience with this approach in other units, medium-term results are sparse. We now report on follow up and the effect on right ventricular remodelling.

**Method:** Ongoing prospective clinical review and acquisition of MRI data at least one year after the procedure.

**Results:** Stent implantation was performed successfully in 46/47 patients. In one patient, the stent embolised and was retrieved at the time of surgical repair. MRI scans were available in 28 patients following the procedure (unavailable in claustrophobic patients, those living abroad, and those awaiting 1 year scans). On average, the QP:QS decreased from 2.57 to 1.18, the RVEDVi from 154 to 95 ml/m<sup>2</sup> and the RV/LV ratio from 2.67 to 1.37. In 3 patients, the QP:QS was > 1.5:1. Of these, two patients had an anomalous pulmonary vein draining high into the SVC that had intentionally been left draining into the SVC. The third patient had a residual QP:QS of 1.7:1 and only a modest decrease in the RV volume (RV/LV 2.18); an additional stent was placed at a second procedure and abolished the shunt. No patients developed pulmonary vein obstruction or sinus node dysfunction. In 2 patients with a pre-existing need for pacing, the pacemaker leads were passed through the covered stent.

**Conclusions:** Transcatheter correction of sinus venosus ASDs is achievable with low morbidity. The reduction in shunt is accompanied by remodelling of the RV without evidence of sinus node dysfunction or pulmonary vein obstruction. Whilst further experience is required, this approach may become the standard of care in carefully selected patients.

**Keywords:** congenital heart disease, sinus venosus atrial septal defect, stent



## O-57

### Device closure of ventricular septal defect using multifunctional vsd occluder- medium-term results with special reference to complications

Masood Sadiq, Ahmad Usaid Qureshi, Uzma Kazmi, Najam Hyder  
The Children's Hospital, University of Child Health Sciences, Lahore, Pakistan

**Background and Aim:** Device closure of ventricular septal defect is a complex procedure with potential complications like complete heart block and aortic regurgitation. LifeTech™ multifunctional (KONAR-MFTM) VSD Occluder (MFO) is a promising new device with good safety in short term.

**Aim:** To assess the safety, efficacy and medium term outcome of ventricular septal defect closure, using MFO.

**Method:** 74 patients with haemodynamically significant, restrictive ventricular septal defect underwent closure with KONAR-MFTM device from April 2019 to March 2022. Clinical, echocardiographic and angiographic data were collected and reviewed. Patients were followed up at 1, 3, 6, 12, 24 and 36 months.

**Results:** RESULTS: The median age was 7(1.7-36) years and median weight was 18(11-79) kg. Of 74 patients, 20(27%) had a high muscular and 54(73%) a perimembranous defect, of which 7 had mild prolapse of right coronary cusp. The left ventricular end median ventricular septal defect size was 8.6(3.9-14.4) mm. Retrograde approach was adopted in 66(89.1%) patients. Fourteen patients (18.9%) had a small residual leak and there was a slight increase in aortic regurgitation in two patients. Two devices embolized to pulmonary artery, which were retrieved, and defect closed with a larger device. At a median follow up of 30(5-41) months the residual leak persisted in 2(2.7%) patients. Mild aortic regurgitation in 2 patients remained unchanged. There were no major complications.

**Conclusions:** Device closure of ventricular septal defect using KONAR-MFTM device is safe and effective in medium-term follow-up. Small delivery system allows majority of defects to be closed retrograde with a very low complications rate.

**Keywords:** Ventricular septal defect, Congenital heart disease, interventions, follow-up

## O-58

### Prediction model to estimate the predicted probability of LPA stenting after bdcpc in in single ventricle patients: our first experience

Alessia Callegari<sup>1</sup>, Jana Logoteta<sup>1</sup>, Walter Knirsch<sup>1</sup>, Robert Cesnjevar<sup>2</sup>, Hitendu Dave<sup>2</sup>, Oliver Kretschmar<sup>1</sup>, Daniel Quandt<sup>1</sup>

<sup>1</sup>University Children's Hospital Zurich, Pediatric Heart Centre – Division of Pediatric Cardiology, University of Zurich; Switzerland;

<sup>2</sup>University Children's Hospital Zurich, Pediatric Heart Centre – Division of Congenital Cardiothoracic Surgery, University of Zurich; Switzerland

**Background and Aim:** A comparison of single ventricle (SV) patients with and without PA-stent implantation post-BDCPC regarding associated risk-factors was performed to create a prediction model to estimate the predicted probability of LPA-stent implantation post-BDCPC. We aim to test the efficacy of this model for SV patients pre-BDCPC at our institution.

**Method:** The comparison was a single centre, retrospective, longitudinal study (2004-2021) on 144 SV patients. Baseline characteristics, clinical-, surgical-course from birth to TCPC, angiographic PA and (neo-)ascending dimensions at the cardiac catheter pre-BDCPC were assessed. The prediction model was built on the results of a logistic regression (p<0.001) performed to ascertain the factors related to the likelihood of LPA-stent implantation

post-BDCPC. Once the model was defined, we tested its prospective sensitivity/specificity on a follow-up group of 13 patients that received a BDCPC (2021–2022).

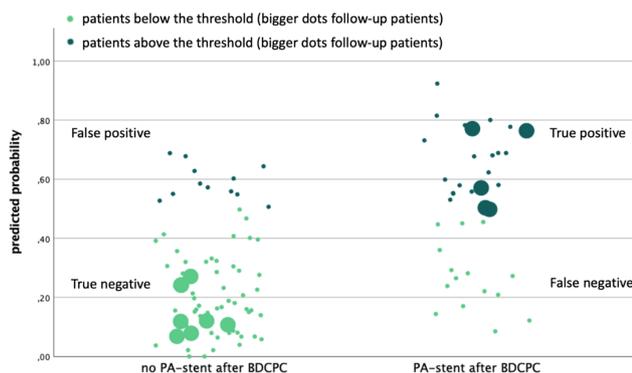
**Results:** A total of 44/144 (30%) patients received a LPA-stent at median (IQR) 12 (1–136) days post-BDCPC. The logistic regression identified that smaller diameter of LPA (mm/m<sup>2</sup>) and larger diameter of the (neo-)ascending aorta pre-BDCPC were associated with an increased likelihood of LPA-stent implantation post-BDCPC.

Based on these results, we built a prediction model to estimate the predicted probability of LPA-stent implantation post-BDCPC. The equation of the model is: predicted logit =  $-3.010911 + (-0.130738 * (\text{LPA in mm/m}^2)) + (0.053830 * (\text{neo-ascending aorta in mm}))$  // predicted probability =  $(e^{\text{predicted logit}}) / (1 + e^{\text{predicted logit}})$ . We tested the specificity/sensitivity of the model on a ROC-curve (AUC = 0.78) and defined the threshold with the highest specificity/sensitivity. This threshold correctly classified 29/44 (65%) of the patients with LPA-stent (true positives) and incorrectly included 12/100 (12%) of the patients without LPA-stent (false positive).

A test phase of the model on the following 13 patients correctly classified 5/5 (100%) of the patients with LPA-stent and incorrectly included 0/8 (0%) without LPA-stent post-BDCPC (Figure 1).

**Conclusions:** Our results suggest that a correct pre-BDCPC risk-stratification for LPA-stent implantation is theoretically feasible with this prediction model at our institution. For patients at risk an exit-angiography / hybrid-procedure on the LPA should be discussed. A surgical relieve at BDCPC of the compression of the (new-)ascending aorta on the LPA could also be considered.

**Keywords:** LPA-Stent, single ventricle patients, pulmonary arteries



**Figure 1** The threshold with the highest sensitivity and specificity was 0.45. Jittered scatter plot showing how this threshold correctly classified 100% of the follow-up patients with a PA-stent (true positives) and incorrectly included 0% of the follow-up patients without a PA-stent (false positive).

### O-59

#### Mid-term outcomes of secundum asd closure with the gore® cardioform asd occluder - results for the combined cohorts in the gore assured clinical trial

Athar M Qureshi<sup>1</sup>, Robert J. J. Sommer<sup>2</sup>, Gareth Morgan<sup>3</sup>, Robert G. Gray<sup>4</sup>, Barry Love<sup>5</sup>, Bryan H. Goldstein<sup>6</sup>, Lissa Sugeng<sup>7</sup>, Matthew J. Gillespie<sup>8</sup>

<sup>1</sup>Texas Children's Hospital/Baylor College of Medicine; <sup>2</sup>Columbia University of New York; <sup>3</sup>The Children's Hospital of Colorado; <sup>4</sup>University of Utah; <sup>5</sup>Mt. Sinai Medical Center; <sup>6</sup>Cincinnati Children's Hospital/University of Pittsburgh Medical Center; <sup>7</sup>Yale University School of Medicine; <sup>8</sup>Children's Hospital of Philadelphia

**Background and Aim:** The GORE® CARDIOFORM ASD Occluder (W. L. GORE & Associates, Flagstaff, AZ) was approved in the US on May 28, 2019 for closure of ostium secundum atrial septal defects (ASD). Here we report the mid-term results of the COMBINED Training, Pivotal, and Continued Access cohorts of the GORE ASSURED clinical trial.

**Method:** This was a prospective, multicenter, single-arm clinical trial comparing the GORE® CARDIOFORM ASD Occluder with existing benchmark data from other U.S. FDA approved devices. This clinical investigation was designed to enroll up to 704 subjects (including 2 training cases per site) from a maximum of 22 clinical investigational sites in the U.S. Enrollment occurred sequentially in two phases – Pivotal (125 pivotal subjects, in addition to 2 training cases per site) and Continued Access (up to 535 subjects).

**Results:** A total of 613 patients with a mean age of 19.0 years (range 1 to 84 years) with a mean weight of  $44.7 \pm 29.7$  kg underwent attempted ostium secundum ASD closure. The mean stop flow ASD diameter was  $17.7 \pm 5.3$  mm. The primary endpoint of 6-month clinical closure success was met in 100% of patients (516 patients with complete data). The primary endpoint of 6-month composite clinical success (577 patients with complete data) was met in 87.2% of patients (technical failure in 8.1%, 30-day serious adverse events in 4.3% of patients, and device events in 2.9% of patients). Clinically significant new arrhythmias were observed in 25/613 (4.1%) of patients at 6 months. WFF was noted in 152/468 (32.5%) of patients completing a 6-month fluoroscopy, (19.8%, 18.9%, 28.6%, 74.2% and 63.2% of patients with 27 mm, 32 mm, 37 mm, 44 mm and 48 mm devices, respectively). No clinical sequelae were observed in any patient with WFF. Device-related serious adverse events were similar in patients with and without WFF [1/152 (0.7%) versus 7/316 (2.2%).

**Conclusions:** In this largest congenital ASD device trial to date, the GORE ASSURED Clinical Trial study results met the primary endpoints. Due to its unique features, size range and safety profile, the GORE® CARDIOFORM ASD Occluder expands the options for transcatheter ostium secundum ASD closure.

**Keywords:** ASD, device

### O-60

#### Safety and efficacy of transcatheter closure of persistent ductus arteriosus in 2 to 6 kg infants: an international study

Paul Padovani<sup>1</sup>, Caroline Ovaert<sup>2</sup>, Gianfranco Butera<sup>3</sup>, Marc Gewillig<sup>4</sup>, Hugues Lucron<sup>5</sup>, Beatrice Kahl<sup>6</sup>, Sebastien Hascoet<sup>7</sup>, Bruno Lefort<sup>8</sup>, Zakia Saliba<sup>9</sup>, Hasri Samion<sup>10</sup>, Sonia El Saiedi<sup>11</sup>, Henrik Holmstrom<sup>12</sup>, Jean Bernard Selly<sup>13</sup>, Hélène Bouwaist<sup>14</sup>, Alain Fraisse<sup>15</sup>, Domenico Sirico<sup>16</sup>, Ali Houeijeh<sup>17</sup>, Clément Karsenty<sup>18</sup>, Céline Grunenvald Gronier<sup>1</sup>, Mathilde Méor<sup>19</sup>, Nadir Benbrik<sup>1</sup>, Damien Bonnet<sup>19</sup>, Sophie Malekzadeh Milani<sup>19</sup>, Alban Elouen Baruteau<sup>1</sup>

<sup>1</sup>Chu Nantes, Nantes, France; <sup>2</sup>Chu Marseille, Marseille, France; <sup>3</sup>IRCCS Ospedale Pediatrico Bambino Gesù, Rome, Italy; <sup>4</sup>University Hospital Leuven, Louvain, Belgique; <sup>5</sup>Chu Martinique, Fort-de-France, France; <sup>6</sup>Pediatric Heart Center, Division of Pediatric Cardiology, Medical University Vienna, Austria; <sup>7</sup>Hôpital Marie-Lannelongue, Le Plessis-Robinson, France; <sup>8</sup>Chu Tours, Tours, France; <sup>9</sup>CHU Hôtel-Dieu de France, Beirut, Lebanon; <sup>10</sup>Institut Jantung Negara, Kuala Lumpur, Malaysia; <sup>11</sup>Cairo University Children's Hospital, Le Caire, Egypt; <sup>12</sup>Department of Paediatric Cardiology, University of Oslo, Norway; <sup>13</sup>CHU La Réunion, Saint-Denis, France; <sup>14</sup>CHU Grenoble, Grenoble, France; <sup>15</sup>Royal Brompton Hospital, London, United Kingdom; <sup>16</sup>University Hospital of Padova, Padoue, Italy; <sup>17</sup>Chu Lille, Lille, France; <sup>18</sup>Chu Toulouse, Toulouse, France; <sup>19</sup>M3c-Necker, Paris, France

**Background and Aim:** Transcatheter patent ductus arteriosus (PDA) closure is increasingly used in smaller patients. Percutaneous approach has proved to be safe and efficient in <2 kg preterm infants. However, data are limited and major safety concerns remain when applied to the 2 to 6 kg body weight range. We aim to assess efficacy and safety of transcatheter PDA occlusion in low-body-weight infants.

**Method:** Patients weighing 2 to 6 kg in whom transcatheter closure of an isolated PDA was attempted in 19 tertiary pediatric cardiology units (9 countries, 2000 to 2022) were retrospectively analyzed to review the outcome.

**Results:** A total of 658 infants (m/fsex ratio: 0.50; 46% ex-preterm) underwent attempted transcatheter PDA closure at a median age of 5.3 month (IQR 3.2;6.5) and a median weight of 4675 gr (min 2000; max 6000). Patients were all symptomatic (respiratory support: 36%, hospitalization for heart failure: 66.5%, nutrition care: 49.4%) with a hemodynamically significant ductus. PDA was large (minimal ductal diameter:  $4.1 \pm 1.7$  mm) and mainly conical (Krichenko type A: 50%) or tubular (Krichenko type C: 28%). Procedure was performed under both fluoroscopy (fluoro time:  $8.7 \pm 8.1$  min) and transthoracic echocardiography guidance in all patients, mainly under general anesthesia (87%) with both arterial and venous femoral access (90%). Successful device implantation and complete PDA occlusion was achieved in 607 (92%) patients using various devices in 93.3% (Amplatzer Duct Occluder: 76%, Amplatzer Vascular Plug: 7.6%, Occlutech Duct Occluder: 2.9%) and coils in 6.5%. Major complications occurred in 4.6% cases, including device embolization requiring surgery in 21 (3.4%). In one patient device embolized to the descending aorta 1 hour after successful device delivery, leading to death (mortality: 0.1%). The incidence of device-related obstruction of the aorta, left pulmonary artery were low (1.1% and 2.4%, respectively) as was access-related peripheral vascular injury (1.3%).

**Conclusions:** Although challenging, transcatheter PDA closure can be safely achieved in 2 to 6 kg infants with both good success rate and reasonable complication rate. These are preliminary results from a wider international study that will allow better delineation of risk stratification in this challenging body weight group.

**Keywords:** patent ductus arteriosus, percutaneous, transcatheter closure, adverse event

## O-61

### Early and mid-term results of transcatheter pda closure in extremely-low-birth-weight patient: A nationwide prospective series.

Sophie Guiti Malekzadeh Milani<sup>1</sup>, Bruno Lefort<sup>2</sup>, Claire Dauphin<sup>3</sup>, Stéphanie Douchin<sup>4</sup>, Ahmad Akhavi<sup>5</sup>, Mohamed Bakhloul<sup>6</sup>, Nadir Benbrick<sup>7</sup>, Mathilde Meot<sup>1</sup>, Damien Bonnet<sup>1</sup>, Alban Baruteau<sup>7</sup>

<sup>1</sup>M3C Pediatric cardiology, Hôpital Necker-Enfants Malades, Paris, France; <sup>2</sup>CHU Tours, Tours, France; <sup>3</sup>CHU Clermont-Ferrand, Clermont-Ferrand, France; <sup>4</sup>CHU Grenoble, Grenoble, France; <sup>5</sup>CHU Reims, Reims, France; <sup>6</sup>CHU Lyon, Lyon, France; <sup>7</sup>CHU Nantes, Nantes, France

**Background and Aim:** Transcatheter persistent ductus arteriosus (PDA) in extremely-low-birth-weight infants has proved to be feasible and safe but midterm outcomes data are scarce. We report herein results from the French nationwide Premiclose registry.

**Method:** All infants who attempted device PDA closure at a procedural weight <2 kg were prospectively included from 7 tertiary institutions between October 2017 and June 2022. Demographic and procedural data were reviewed as well as respiratory and cardiac follow-up.

**Results:** 274 patients (mean birth weight: 812 +/-213 grams, mean gestational age: 25.8 +/- 1.9 weeks) were included. Mean procedural age and weight were 31 +/-12 days and 1250 +/-330 grams

respectively. Patients were all symptomatic with a hemodynamically significant PDA. Procedure was performed under both fluoroscopy (fluoro time: 4.5 +/-4 min) and transthoracic echocardiography guidance in all patients; 240 (87.6%) procedures were contrast free. Devices implanted were Pico occluder in all cases except 2 who received Amplatzer Vascular Plug. Successful PDA closure was achieved in 97%. Complications occurred in 10 patients, with major adverse events in 3 (1.1%) consisting in device embolization in 2 and tamponade in 1. There was no procedural death. Post ligation cardiac syndrome was noted in 4% and escalation of respiratory support in 20% of the cases. During follow-up, 8 late complications were reported with 3 patients requiring surgical repair of aortic coarctation. Over time, significantly smaller patients were treated with significantly lower complications. Neurologic assessment is ongoing and will be presented.

**Conclusions:** Our large series confirms favorable results of transcatheter PDA closure in extremely-low-birth-weight infants, along with demonstrating learning curve, high procedural success, low procedural complications and low rate of post ligation cardiac syndrome but escalation in respiratory support. Late cardiac complications were few but long term follow-up remains mandatory.

**Keywords:** Preterm, transcatheter closure, PDA

## O-62

### Transcatheter pda closure in preterm infants weighing less than 1.5kg

Conall Thomas Morgan<sup>1</sup>, Laurent Desjardins<sup>1</sup>, Abdulaziz Alosaimi<sup>1</sup>, Bonny Jasani<sup>2</sup>, Linh Ly<sup>2</sup>, Luc Mertens<sup>1</sup>, Audrey Marshall<sup>1</sup>, Rajiv Chaturvedi<sup>1</sup>, Lee Benson<sup>1</sup>

<sup>1</sup>Division of Cardiology, The Labatt Family Heart Centre, The Hospital for Sick Children, Toronto, Canada; <sup>2</sup>Division of Neonatology, The Hospital for Sick Children, Toronto, Canada

**Background and Aim:** Transcatheter PDA closure (TCPC) in preterm infants has emerged as an alternative to surgical ligation. The objectives to this study were to describe our experience of TCPC in preterm infants under 1.5kg at our centre.

**Method:** A retrospective review of premature infants weighing under 1.5kg undergoing TCPC between 2019 and 2021 at The Hospital for Sick Children was performed.

**Results:** The cohort consisted of 42 neonates born at a gestational age between 22 and 26 weeks. The mean age and weight at the time of the procedure was  $29.9 \pm 1.6$  days (range 26 to 34 days) and  $1080 \pm 184$  g (769g to 1486g). Eighteen babies were less than 1000g. Venous only access was used in all 42 patients. The Amplatzer Pico Occluder Device (Abbott) was used in 93% of the babies (n = 39). Procedural success was achieved in 98% (n = 41). There were 2 major adverse events due to device positioning; left pulmonary artery and descending aorta obstruction, requiring eventual surgical intervention in each. Two babies developed severe tricuspid regurgitation which improved to mild at latest follow up. There were no reports of infective endocarditis or procedure related deaths. Moderate to severe LV dysfunction developed in 8 patients (19%) within 24 hours of the procedure, requiring milrinone initiation in 6. The LV function recovered in all within 1 week. An occlusive venous thrombosis was identified in 7% of patients (n = 3). Follow up was complete to  $269 \pm 164$  days. On long term follow up, three patients had mild flow acceleration in the LPA (maximum velocity less than 2.5m/s) and no patient developed arch obstruction. There was one late death attributed to complications of prematurity.

**Conclusions:** TCPC is a safe alternative to surgical PDA ligation in preterm infants <1500g. Acute LV dysfunction is common but resolves. Late LPA or arch obstruction was not seen in our cohort.

**Keywords:** transcatheter, PDA, preterm

## O-63

**Remote ischaemic preconditioning in paediatric cancer patients receiving anthracycline therapy: A double-blinded randomized trial**

Y F Cheung<sup>1</sup>, Vivian W Y Li<sup>1</sup>, Edwina K F So<sup>1</sup>, Frankie W T Cheng<sup>2</sup>, Jeffery P W Yau<sup>2</sup>, S Y Chiu<sup>2</sup>, Wilfred H S Wong<sup>1</sup>, Daniel K L Cheuk<sup>2</sup>  
<sup>1</sup>Department of Paediatrics & Adolescent Medicine, The University of Hong Kong, Hong Kong; <sup>2</sup>Department of Paediatrics & Adolescent Medicine, Hong Kong Children's Hospital, Hong Kong

**Background and Aim:** Anthracycline cardiotoxicity is a concern in survivors of childhood cancers. Recent evidence suggests that remote ischaemic preconditioning (RIPC) may offer non-pharmacological myocardial protection.

**Method:** We performed a double-blind randomized controlled trial to determine the impact of remote ischaemic preconditioning (RIPC) on myocardial injury in paediatric cancer patients receiving anthracycline-based chemotherapy. Eligible patients were randomized to receive RIPC (three cycles of five-minute inflation of a blood pressure cuff placed over one limb to 15 mmHg above systolic pressure) or sham intervention. The intervention was applied within 60 minutes before initiation of the first dose and up to four cycles of anthracycline therapy. Primary outcome was plasma high sensitivity troponin T level. Secondary outcome measures included echocardiographic indices of left ventricular (LV) systolic and diastolic function and occurrence of cardiovascular events.

**Results:** A total of 68 children aged 10.9±3.9 years were randomized to receive RIPC (n = 34) or sham (n = 34) intervention. There were no significant differences in baseline demographic parameters and laboratory results. Plasma levels of hs-cTnT showed progressive increase across time points in both the RIPC (p<0.001) and the sham group (p<0.001). However, at each of the time points, there were no significant differences between the two groups (all p>0.05). Left ventricular tissue Doppler and strain parameters were also similar between the two groups at different time points. None of the patients developed heart failure or cardiac arrhythmias.

**Conclusions:** RIPC did not exhibit cardioprotective effects in childhood cancer patients receiving anthracycline-based chemotherapy.

**Keywords:** remote ischaemic preconditioning, paediatric cancer anthracycline

## O-64

**The utility of speckle tracking imaging to predict short and mid-term outcomes after reverse potts shunt in patients with idiopathic pah**

Shatabdi Giri<sup>1</sup>, Prashant Bobhate<sup>1</sup>, Santosh Wadile<sup>2</sup>

<sup>1</sup>Children Heart Centre, Kokilaben Dhirubhai Ambani Hospital and Medical Research Institute, Mumbai, India; <sup>2</sup>Sri Sathya Sai Sanjeevani Hospital, Mumbai, India

**Background and Aim:** Potts shunt has been suggested as effective palliative therapy and a bridge to heart-lung transplant for patients with Idiopathic pulmonary artery hypertension (PAH) not associated with congenital heart disease. However, not all patients may benefit from this high-risk procedure

**Method:** This is a prospective single-center study performed to correlate right ventricular functional parameters derived from ECHO with invasive hemodynamic and laboratory parameters and analyze the factors associated with poor prognosis after the Potts shunt.

**Results:** Our study included 19 patients (14 females and 5 males) with PAH and no intra- or extra-cardiac shunt undergoing Potts shunt (16 surgical Potts shunt and 3 PDA Stent) at a single tertiary care center.

The Median follow-up was 38 (range 5-53 months). Right atrial (RA) strain correlated negatively with mean RA pressure (r2 = -0.78, p value = 0.001) and NT pro-BNP (r2 = -0.62, p value = 0.01) while right ventricular global longitudinal strain (RVGLS) correlated with cardiac index (r2 = -0.52, p value = 0.03), Pulmonary vascular resistance index (PVRi) (r2 = 0.6, p value = 0.01), and NT Pro-BNP (r2 = 0.65, p value = 0.003). RV free wall strain (RVFWS) correlated with cardiac index (r2 = -0.53, p value = 0.03), PVRi (r2 = 0.6, p value = 0.01) and NT pro-BNP (r2 = 0.57, p value = 0.01). Eight patients died and eleven patients showed sustained clinical and echocardiographic improvement. The patients showing benefit after Potts shunt had significantly lower preoperative RV GLS (-11.35% versus -2.85%, p value:0.002) and pooled RV FWS (-14.8% versus -2.2, p value:0.004) and significantly higher right ventricular fractional area change (FAC) (30.45% versus 7.39%, p value:0.001) than the patients who did not benefit from the procedure.

**Conclusions:** Functional ECHO parameters and speckle tracking imaging correlates well with invasive hemodynamic and laboratory parameters and can be helpful in predicting short and midterm outcomes after Potts shunt in patients with Idiopathic pulmonary hypertension.

**Keywords:** Pulmonary hypertension, Potts shunt, Right ventricular function, Global longitudinal strain, Speckle tracking Echocardiography

Comparison of both the groups based on various demographic, ECHO and Cardiac Cathparameters

Demographic Parameters	Group 1	Group 2	p value
Age (yrs)	9.5 (4.25, 16.5)	13.5 (1.35, 26)	
Weight (kg)	23.85 (12.87, 55.13)	32.15 (9.7, 50.28)	0.947
Height (cm)	124.4±30.98	124.17±40.06	0.990
BSA (m2)	0.93±0.45	1.02±0.51	0.729
NT pro-BNP (pg/ml)	3264 (1987, 5432)	7791 (2328, 9867)	0.238
<b>ECHO parameters</b>			
RVGLS (%)	-11.35 (-13.5, -10.8)	-2.85 (-3.2, -2.15)	<b>0.002*</b>
RVFWS (%)	-14.8 (-16.6, -11.0)	-2.185 (-5.93, -1.09)	<b>0.004*</b>
RA strain (%)	23 (18.55, 30.65)	11.1 (3.98, 13.53)	<b>0.001*</b>
FAC (%)	30.45 (22.55, 36.7)	7.39 (2.1, 14.14)	<b>0.001*</b>
RI	1.485 (1.39, 1.54)	1.68 (1.46, 1.85)	<b>0.043*</b>
TAPSE	13.4 (10, 17.7)	11.5 (8, 13.7)	0.25
<b>Cath parameters</b>			
mPAP (mmHg)	74.27±16.9	79.29±14.68	0.515
mABP (mmHg)	71.73±9.49	70±12	0.767
Mean RA pressure (mmHg)	10±3.5	13.6±3	<b>0.04*</b>
PVRI (Woodu.m2)	20.71±5.97	27.43±9.89	0.089
CI (L/m2)	3.3±0.99	2.61±1.08	<b>0.03*</b>
SVRI (Woodu.m2)	20.43±5.4	26.87±10.55	0.105
Rp/Rs	1.04±0.21	1.04±0.09	0.955
Follow up duration (months)	43.5 (30.2, 53.2)	6.33 (5.12, )	0.038

## O-65

**Rejection surveillance with cell-free dna using droplet digital pcr: A swedish national study**

Jens Böhmer<sup>1</sup>, Jens Böhmer<sup>2</sup>, Marianne Jonsson<sup>3</sup>, Håkan Wåhlander<sup>1</sup>, Håkan Wåhlander<sup>2</sup>, Katarina Hansens<sup>4</sup>, Maria Sjöborg Alpman<sup>5</sup>, Julia Asp<sup>3</sup>, Julia Asp<sup>6</sup>, Carina Wasslavik<sup>3</sup>, Jan Sunnegårdh<sup>1</sup>, Anne Ricksten<sup>3</sup>, Göran Dellgren<sup>7</sup>, Göran Dellgren<sup>8</sup>

<sup>1</sup>Pediatric Heart Center, Sahlgrenska University Hospital; <sup>2</sup>Department of Pediatrics, Institute of Clinical Sciences, Sahlgrenska Academy, University of Gothenburg; <sup>3</sup>Department of Laboratory Medicine, Institute of Biochemistry, Sahlgrenska Academy, University of Gothenburg; <sup>4</sup>Pediatric Heart Center, Skane University Hospital;

<sup>5</sup>Pediatric Heart Center, Karolinska University Hospital; <sup>6</sup>Department of Clinical Chemistry, Sahlgrenska University Hospital; <sup>7</sup>Department of Cardiothoracic Surgery, Sahlgrenska University Hospital; <sup>8</sup>Transplant Institute, Sahlgrenska University Hospital

**Background and Aim:** Heart transplantation (HTx) comes with a higher risk for rejection compared with other solid organs. Graft surveillance largely depends on scheduled endomyocardial biopsies (EMB) as the gold standard due to the lack of reliable biomarkers. Donor-derived cell-free DNA (dd-cfDNA) has gained attention as a potential marker to assess organ function after HTx.

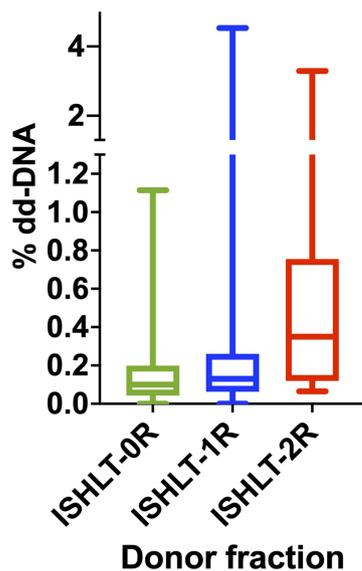
**Method:** In a collaboration between the two pediatric heart centers performing heart transplantations in Sweden, a prospective cohort study was conducted 2016–2021. Patients were followed with parallel measurements of dd-cfDNA and EMB during one year after HTx. Single-nucleotide polymorphisms (SNP) were chosen to distinguish between recipient and donor. After a targeted preamplification-step, droplet digital PCR was conducted.

**Results:** A total of 29 consecutive patients were included, 193 samples from 24 patients have so far been analyzed. Donor fraction (DF) levels showed a median of 0.13 %, with a highly significant increase on samples during the first 14 days after HTx compared to later. When comparing different grades of rejection, there were highly significant differences between ISHLT-0R, 1R and 2R.

**Conclusions:** We constructed a technically robust method to measure cell-free DNA after HTx. Results for total levels of DF are well in concordance with the available literature in the field. High initial levels probably reflect ischemia-induced and surgical damage. There are significant differences between levels of rejection, making cfDNA a promising marker for organ surveillance.

**Keywords:** cell-free DNA, rejection, pediatric heart transplantation, droplet digital PCR

Rejection



Significantly different levels of dd-cfDNA between rejection grades as compared by one-way ANOVA ( $p < 0.0001$ ).

## O-66

### Survival of children and adolescents with pulmonary arterial hypertension. data from the bnp-pl registry – PAH children

Joanna Kwiatkowska<sup>1</sup>, Rafal Surmacz<sup>2</sup>, Malgorzata Zuk<sup>3</sup>, Marcin Waligora<sup>4</sup>, Anna Migdal<sup>5</sup>, Jacek Kusa<sup>5</sup>, Elzbieta Skiba<sup>5</sup>, Pawel Banaszak<sup>6</sup>, Agata Michalczyk<sup>6</sup>, Katarzyna Ostrowska<sup>8</sup>, Joanna Pluzanska<sup>8</sup>, Magdalena Bazgier<sup>1</sup>, Halszka Kamińska<sup>7</sup>, Barbara Wojcicka Urbanska<sup>7</sup>, Aldona Lesko<sup>10</sup>, Sebastian Goreczny<sup>10</sup>, Grzegorz Kopec<sup>9</sup>

<sup>1</sup>Department of Pediatric Cardiology and Congenital Heart Defect, Medical University of Gdansk, Gdansk, Poland; <sup>2</sup>Department of Pediatric Cardiology, Poznan University of Medical Sciences, Poznan, Poland; <sup>3</sup>The Children's Memorial Health Institute, Warsaw, Poland.; <sup>4</sup>Pulmonary Circulation Center, Department of Cardiac and Vascular Diseases, Faculty of Medicine, Jagiellonian University Medical College, John Paul II Hospital in Krakow, Krakow, Poland.; <sup>5</sup>Center for Innovative Medical Education, Department of Medical Education, Faculty of Medicine, Jagiellonian University Medical College, Kraków, Poland.; <sup>6</sup>Pediatric Cardiology Department, Regional Specialist Hospital- Research and Development Center in Wroclaw, Poland; <sup>7</sup>Department of Congenital Heart Defects and Pediatric Cardiology, Silesian Centre for Heart Diseases, Zabrze, Poland; <sup>8</sup>Department of Pediatric Cardiology and General Pediatrics, Medical University of Warsaw, Warsaw, Poland; <sup>9</sup>Institute of Polish Mother's Health Center, Lodz, Poland; <sup>10</sup>Pulmonary Circulation Center, Department of Cardiac and Vascular Diseases, Faculty of Medicine, Jagiellonian University Medical College, John Paul II Hospital in Krakow, Krakow, Poland; <sup>10</sup>Department of Pediatric Cardiology, University Children's Hospital, Jagiellonian University Medical College, Kraków, Poland

**Background and Aim:** In Poland the care of children suspected of pulmonary arterial hypertension (PAH) is centralized in 8 medical paediatric centres. The Working Group on Pulmonary Circulation of the Polish Cardiac Society established a Data Base of pulmonary hypertension in the Polish population (BNP-PL) in 2018. We present the survival of children with PAH based on the registry results.

**Method:** From 01.03.2018 to 30.08.2022 we prospectively enrolled all PAH patients from 3 months to 18 years of age. The PAH diagnosis was based on the right heart catheterization. The functional class was assessed based on the WHO-FC. Anthropometric measurements were evaluated on the basis of percentile charts of height and BMI. Data is presented as median (interquartile range).

**Results:** 138 PAH children (females,  $n = 70$ , 50.7%) were enrolled in the present analysis. The median percentile of height was 10 (3–50) and median weight percentile was 17 (3–50). Most frequent cause of PAH was CHD ( $n = 92$ ; 66.7%), followed by non-reactive IPAH ( $n = 33$ ; 23.9%), reactive IPAH ( $n = 9$ , 6.5%), portopulmonary PAH ( $n = 2$ ; 1.4%) and one patient each with PAH associated with connective tissue disease and hereditary PAH. At enrolment 25.4% of patients were still in WHO-FC III. The majority of children (95.7%) were treated with PAH specific medications but only a half of them with either double combination therapy or triple combination therapy. During prospective observation (up to 54 months), 16 (11.6%) patients died and those had higher NT-proBNP [1592 (665–4519.5) vs 314 (120–680),  $p = 0.0002$ ], worse 6-minute walking distance [101 (0–383) vs 346 (202–450);  $p = 0.01$ ] and were more often diagnosed with IPAH than with other aetiologies of PAH [9 (19.6%) vs 7 (7.6%),  $p = 0.03$ ]. During follow-up, 21 patients reached adult

age and were transferred for a further treatment in the PAH centre dedicated for adults.

**Conclusions:** Almost all PAH cases were associated with CHD or IPAH without significant representation of other PAH subclasses. Most children had growth retardation, suggesting the need for active dietary interventions. Patients diagnosed with IPAH, high NT-proBNP and with poor exercise capacity have worse survival

**Keywords:** pulmonary arterial hypertension; pediatric registry, children's survival

## O-67

### **Risk factors for pulmonary hypertension in preterm infants with bronchopulmonary dysplasia**

Anne C Kesting<sup>1</sup>, Sven C Weber<sup>1</sup>, Martin Koestenberger<sup>2</sup>, Felix Berger<sup>1</sup>, Christoph Czernik<sup>3</sup>, Hannes Sallmon<sup>1</sup>

<sup>1</sup>Department of Congenital Heart Disease/Pediatric Cardiology, Deutsches Herzzentrum Berlin (DHZB) and Charité - Universitätsmedizin Berlin, Berlin, Germany; <sup>2</sup>Division of Pediatric Cardiology, Graz Medical University, Graz, Austria; <sup>3</sup>Department of Neonatology, Charité - Universitätsmedizin Berlin, Berlin, Germany

**Background and Aim:** Pulmonary hypertension (PH) in preterm infants with bronchopulmonary dysplasia (BPD) considerably contributes to morbidity and mortality. However, risk factors associated with BPD-PH development are still poorly defined. Herein, we aimed to determine demographic and clinical factors associated with BPD-PH.

**Method:** We retrospectively analyzed a single center cohort of 63 very low birth weight (VLBW) infants with BPD born between 2015 and 2020 (BPD no PH, n=33; BPD-PH, n=30). Demographic and clinical data were obtained by chart review. Differences between groups were analyzed in order to identify risk factors associated with BPD-PH using SPSS statistics software (Chicago, IL, USA).

**Results:** BPD infants with and without PH did not differ in regard to sex and gestational age (p<0.05). Infants with BPD-PH exhibited significantly lower birthweights (p = 0.035) and a higher incidence of intrauterine growth restriction (IUGR; p=0.002). Patients with BPD-PH also required a longer duration of invasive ventilation (p = 0.012) and oxygen supplementation (p = 0.000), while the duration for non-invasive ventilation was similar between groups (p<0.05). Furthermore, infants with BPD-PH received significantly more red blood cell (RBC) transfusions (p = 0.002). Interestingly, there were no significant differences in regard to the incidence of patent ductus arteriosus (PDA) or the occurrence of hemodynamically significant PDA (hsPDA) in BPD infants with and without BPD-PH (both p>0.05). Intraventricular hemorrhage (grade > 2), late onset sepsis, necrotizing enterocolitis and retinopathy of prematurity were similar between groups. However, infants with BPD-PH had a significantly longer inpatient stay (p = 0.034) and in-hospital death occurred only in infants with BPD-PH (n = 7).

**Conclusions:** Development of BPD-PH was associated with several potential risk factors, including lower birth weight, IUGR, RBC transfusions, longer durations of invasive ventilation and oxygen supplementation. However, PDA or hsPDA were not associated with BPD-PH. The occurrence of in-hospital death only in BPD infants with PH underlines the severity of the condition. Further prospective studies are required to delineate the possible relationships between potential risk factors and BPD-PH development in the most vulnerable cohort of VLBW infants with BPD.

**Keywords:** pulmonary hypertension, preterm infants, bronchopulmonary dysplasia

## O-68

### **Immunological risks for post-transplant lymphoproliferative disease: the immunology of thymectomy and childhood cardiac transplant (ithaca) study**

Ugonna T Offor<sup>1</sup>, Heather M Long<sup>2</sup>, Andrew Fliby<sup>3</sup>, Milos Ognjanovi<sup>4</sup>, Jacob Simmonds<sup>5</sup>, Gareth Parry<sup>6</sup>, Abbas Khushnood<sup>6</sup>, Andrew Gemmerly<sup>7</sup>, Chris M Bacon<sup>8</sup>, Simon Bomken<sup>1</sup>, Zdenka Reinhardt<sup>6</sup>

<sup>1</sup>Wolfson Childhood Cancer Research Centre, Translational and Clinical Research Institute, Newcastle University, Newcastle upon Tyne, UK; <sup>2</sup>Department of Paediatric Haematology and Oncology, The Great North Children's Hospital, Newcastle upon Tyne Hospitals NHS Foundation Trust., Newcastle upon Tyne, UK.; <sup>3</sup>Institute of Immunology and Immunotherapy, College of Medical and Dental Sciences, University of Birmingham, Birmingham, UK.; <sup>4</sup>Flow Cytometry Core Facility, Biosciences Institute, Faculty of Medical Sciences, Newcastle University, Newcastle Upon Tyne, UK.; <sup>5</sup>Department of Paediatric Nephrology, Great North Children's Hospital, Newcastle Upon Tyne, UK.; <sup>6</sup>Department of Cardiology, Great Ormond Street Hospital for Children NHS Foundation Trust, London, UK.; <sup>7</sup>Department of Cardiopulmonary Transplantation, Newcastle upon Tyne Hospitals NHS Foundation Trust, Newcastle upon Tyne, UK.; <sup>8</sup>Translational and Clinical Research Institute, Newcastle University, Newcastle upon Tyne, UK; <sup>9</sup>Department of Paediatric Immunology and Haematopoietic Stem Cell Transplantation, The Great North Children's Hospital, Newcastle upon Tyne Hospitals NHS Foundation Trust, Newcastle upon Tyne, UK; <sup>10</sup>Department of Cellular Pathology, Newcastle upon Tyne Hospitals NHS Foundation Trust, Newcastle upon Tyne, UK.

**Background and Aim:** Post-transplant lymphoproliferative disease (PTLD) is a rare but potentially fatal cancer that can occur in children after a solid organ transplant. Most childhood PTLD is caused by Epstein-Barr Virus (EBV). The incidence and adverse outcomes of EBV-PTLD are of particular concern in paediatric heart transplant (PHT) patients. We have previously shown that PHT patients with underlying congenital heart disease and early thymectomy at < 1 year old are at significantly higher risk of developing EBV-PTLD. This study aims to examine the role of early thymectomy in the development of EBV-PTLD by monitoring the temporal trajectory of immune reconstitution in PHT patients and their response(s) to EBV exposure.

**Method:** Prospective and sequentially immune monitoring will be performed for 34 PHT and 6 renal transplant patients (aged 0-18 years) over a 2-year period, and stratified into early (<1-year) vs late (>1-year)/non-thymectomy groups. Blood samples will be taken before transplant and at 3-, 6-, 12- and 24 months post-transplant. Single cell analysis of circulating immune cells and functional stimulation of EBV-specific T-lymphocytes will be performed using high dimensional spectral flow cytometry and MHC I/II tetramer assay respectively. EBV antibodies and viral load will be monitored at each of the defined study timepoints. Clinical data will also be collected on induction and maintenance immunosuppression regimens, sociodemographic characteristics and post-transplant sequelae.

**Results:** The ITHACA study opened to patient recruitment in March 2022 and has recruited 8 participants as of November 2022. Mononuclear cells have been isolated and cryopreserved from patient blood samples along with serum for quantification of EBV antibodies. A 30-colour full spectrum flow cytometry panel has been developed for deep immunophenotyping of circulating T-/B-lymphocytes, NK cells, and antigen presenting cells (monocytes and dendritic cells). This panel includes multiple markers for cellular exhaustion and senescence (PD-1, CTLA4, LAG-3, TIM-3) and has been validated using blood samples from healthy volunteers.

**Conclusions:** Study analyses will provide key insight to the complex mechanisms involved in EBV-PTLD. The study will help identify immune signatures that are risk predictive for PTLD. Potential biomarkers will be explored to support future patient risk-stratification and facilitate the development of clinical pathways that mitigate PTLD risk.

**Keywords:** EBV, PTLD, Paediatrics, Transplantation, Immunology

## O-69

### Pharmacokinetics and safety of selexipag in children with pulmonary arterial hypertension: A prospective open-label, single-arm, ongoing phase 2 study

Maurice Beghetti<sup>1</sup>, Shahin Moledina<sup>2</sup>, Lene Nygaard Axelsen<sup>3</sup>, Julian I. Borissoff<sup>3</sup>, Mahdi Farhan<sup>3</sup>, Simon Grill<sup>3</sup>, Sining Leng<sup>3</sup>, Alberto Russo<sup>4</sup>, Chong Xu<sup>3</sup>, Catherine Lesage<sup>3</sup>

<sup>1</sup>Paediatric Cardiology Unit, University Hospitals of Geneva, Geneva, Switzerland; <sup>2</sup>Paediatric Cardiology, Great Ormond Street Hospital, London, UK; <sup>3</sup>Actelion Pharmaceuticals Ltd, a Janssen Pharmaceutical Company of Johnson & Johnson; <sup>4</sup>Janssen-Cilag SpA, Cologno Monzese, Italy

**Background and Aim:** Selexipag, an oral selective prostacyclin receptor agonist, is approved for the treatment of pulmonary arterial hypertension (PAH) in adults. This study assesses the pharmacokinetics, safety, and efficacy of selexipag in children aged  $\geq 2$  years with PAH, with the primary objective to determine a paediatric dosing regimen providing similar exposure to that observed in adults. Pharmacokinetics results and interim safety and exploratory efficacy data are reported here.

**Method:** This prospective, multicenter, open-label, single-arm, Phase 2 study (NCT03492177) evaluates selexipag in children with PAH by age cohorts ( $\geq 2$ – $<6$ ;  $\geq 6$ – $<12$ ;  $\geq 12$ – $<18$  years) and weight groups ( $\geq 9$ – $<25$ ;  $\geq 25$ – $<50$ ;  $\geq 50$  kg). Selexipag was up-titrated for 12 weeks, followed by an ongoing long-term maintenance period. The combined steady-state exposure to selexipag and its metabolite corrected for potency (AUC $\tau$ , combined, ss) was determined during the up-titration period. Safety, tolerability, efficacy, and acceptability were monitored throughout the study. **Results:** Sixty-three children were enrolled and 59 completed  $\geq 16$  weeks of treatment (median treatment duration: 144.4 weeks). The dosing regimen ensured similar exposure in children to that observed in adults; the difference in AUC $\tau$ , combined, ss from that expected based on adult-extrapolated data was  $<25\%$  for all weight groups (Table) and age cohorts. Sixty patients (95.2%) had  $\geq 1$  adverse event (AE). During the up-titration period, 54 patients (85.7%) reported AEs; most common were headache (31.7%), vomiting (30.2%) and diarrhoea (27.0%). During long-term maintenance period, 44 patients (69.8%) reported AEs; vomiting (19.0%), COVID-19 (14.3%), and diarrhoea (11.1%) were most common. Overall, 23 patients (36.5%) had serious AEs (selexipag-related in 3 patients). Seven patients (11.1%) had AEs leading to discontinuation, including 6 deaths (9.5%), all unrelated to selexipag. Trends towards improvement in 6-minute walk distance (mean [SD] change: 21.6 [72.5] meters), NT-proBNP ( $-78.5$  [383.1] pmol/L) and echocardiography parameters were observed at Week 16. Selexipag tablets were well accepted in all age cohorts.

**Conclusions:** In children aged  $\geq 2$  years, the data support the weight-based paediatric dosing regimen being safe, well tolerated, and appropriate for further clinical evaluation of selexipag in paediatric PAH. The nature and frequency of AEs were similar to those observed in adults with PAH.

**Keywords:** Selexipag, paediatric, PAH, safety, PK

**Table. Dosing regimen and pharmacokinetic results**

Body weight group	Dosing regimen		Pharmacokinetic results	
	Starting dose ( $\mu\text{g}$ , b.i.d.)	Maximum dose allowed ( $\mu\text{g}$ , b.i.d.)	N	Difference from expected AUC $\tau$ ,ss* [90% CI], %
$\geq 9$ to $<25$ kg	100	800	28	17.7 [1.13; 37.0]
$\geq 25$ to $<50$ kg	150	1200	19	5.9 [-13.3; 29.2]
$\geq 50$ kg	200	1600	12	6.1 [-19.3; 39.4]
Overall			59	11.37 [-0.2; 24.3]

Dosing regimen was selected based on pharmacokinetic extrapolation from adults. Selexipag was up-titrated weekly in increments equal to the starting dose to the individual maximally tolerated dose or to the maximum dose allowed.

\*Combined sum of selexipag and ACT-333679 (metabolite) exposures (area under the plasma concentration-time curve over a dose interval at steady state, AUC $\tau$ ,combined,ss) weighted by their potency, and scaled to the respective starting doses. A difference in the point estimates up to  $\pm 30\%$  was considered acceptable.

## O-70

### Pediatric heart transplantation in Spain after circulatory determination of death and in situ normothermic regional perfusion

Luis García Guereta<sup>1</sup>, Alvaro Gonzalez Rocafort<sup>2</sup>, Juan Miguel Gil Jaurena<sup>3</sup>, Manuela Camino<sup>4</sup>, Ferran Gran<sup>5</sup>, Raul Abella<sup>6</sup>, Paula Burgos<sup>2</sup>, Juan Jose Menendez<sup>7</sup>, Belen Estebanez<sup>10</sup>, Jose Zamorano<sup>3</sup>, Laura Butragueño<sup>8</sup>, Braulio De La Calle<sup>11</sup>, Vicente Sanchez Salmeron<sup>6</sup>, Joan Balcells<sup>9</sup>, Christopher Mazo<sup>12</sup>, Edume Lorence<sup>13</sup>, Alicia Perez Blanco<sup>14</sup>

<sup>1</sup>Department of Pediatric Cardiology, Hospital Universitario La Paz, Madrid, Spain; <sup>2</sup>Department of Pediatric Cardiac Surgery, Hospital Universitario La Paz, Madrid, Spain; <sup>3</sup>Department of Pediatric Cardiac Surgery, Hospital Universitario Gregorio Marañón, Madrid, Spain; <sup>4</sup>Department of Pediatric Cardiology, Hospital Universitario Gregorio Marañón, Madrid, Spain; <sup>5</sup>Department of Pediatric Cardiology, Hospital Universitario de la Vall d'Hebron, Barcelona, Spain; <sup>6</sup>Department of Pediatric Cardiac Surgery, Hospital Universitario de la Vall d'Hebron, Barcelona, Spain; <sup>7</sup>Department of Pediatric Intensive Care, Hospital Universitario La Paz, Madrid, Spain; <sup>8</sup>Department of Pediatric Intensive Care, Hospital Universitario Gregorio Marañón, Madrid, Spain; <sup>9</sup>Department of Pediatric Intensive Care, Hospital Universitario de la Vall d'Hebron, Barcelona, Spain; <sup>10</sup>Transplantation Coordination, Hospital Universitario La Paz, Madrid, Spain; <sup>11</sup>Transplantation Coordination, Hospital Universitario Gregorio Marañón, Madrid, Spain; <sup>12</sup>Transplantation Coordination, Hospital Universitario de la Vall d'Hebron, Barcelona, Spain; <sup>13</sup>Transplantation Coordination, Hospital Universitario Donostia; <sup>14</sup>Organización Nacional de Trasplantes

**Background and Aim:** Organ transplantation programs following controlled donation after circulatory death (cDCD) have been developed in only 17 countries and include mainly kidney and liver transplantation and only recently pediatric heart transplantation (PHT). In 2021 a national protocol was developed by pediatric heart transplantation teams and Organización Nacional de Trasplantes (ONT). We report our initial national experience with PHT after cDCD

**Method:** Hearts donors after cDCD were allocated to pediatric patients ( $<18$ y) who had consented at listing. Procedures were performed from March 14th 2021 to Nov 2022. Withdrawal of life support was undertaken at theatre. Functional warm ischemic time (FWIT) was the period between hypoperfusion and normothermic regional perfusion (NRP) establishment. Circulatory cessation was defined by asystole or absence of flow by invasive arterial monitoring or echo absence of aortic flow. Hands off period (5 min) was followed by median sternotomy, clamping of supraaortic arteries, cannulation of aorta and RA and in situ normothermic regional perfusion (NRP) with cardiopulmonary bypass or ECMO. 5 min were allowed for heart beat to be recovered and 30 min to wean from ECMO. In situ assessment of the heart was performed before explant. Retrieval of heart and other organs

was performed following the national protocol adapted by each center and ice stored for transportation

**Results:** Table show details of donors and recipients. One of the hearts showed LVEF<50% and was discarded. 4 recipients had CHD, 3 were on LVAD and 1 on ECMO at transplant. fWIT was 18-30 min except for the first case and cold ischemic time 132-298 min. Two patients had primary heart failure one of whom recovered after ECMO for 3 days. There were 5/8 survivors. 2/3 deaths occurred in children in a critical situation at transplant. During 2022 6 of 28 donors for PHT were cDCD (21% of total PHT). 8 other organs (kidney, Liver, Lung, Multivisceral) were implanted from same donors.

**Conclusions:** cDCD constitute an additional source of heart donors thus increasing the probability of transplantation for pediatric listed patients. Detailed assessment of the donor must be undertaken in order to assure adequate transplant outcomes.

**Keywords:** Heart transplantation, Donation after circulatory death

Hospital	Date	Rec.age/weight	Rec Diag	ECMO/VAD	Don age/weight	ABO/ABOI	H/Morgan	fWIT	cold IT	Local/distant	outcome
1	11/14/21	14 y/40 Kg	OH		12a/52 Kg	ABOc	Morgan	30	174	Local	Alive
1	6/16/2022	2m/3.4 Kg	DCM	ECMO/VAD	3m/4 kg	ABOc	Morgan	29	176	Local	D graft failure
1	10/22/2022	3m/5.5 Kg	DCM	VAD	6m/8 Kg	ABOc	Morgan	24	298	D 350 km	Alive
1	10/29/2022	8y/24 Kg	chd(HLHS)		14a/23 Kg	ABOc	Heart	23	280	D 350 km	Alive
2	3/11/21	2m/3.1 Kg	DCM		3 d/3.4 Kg	ABOI	Heart	32	245	D 300 Km	D ARI at 6 mo
2	3/12/22	2y/12 kg	DCM		2 y/12 Kg	ND	not done	ND	ND	ND	ND FEV1 < 50%
2	3/17/2022	1y/15 Kg	OH		6a/21 kg	ABOc	Morgan	22	235	1 km	Alive
3	6/22/2022	8m/6 Kg	DCM	VAD	18 m/12 Kg	ABOI	Morgan	18	202	D 400 Km	Alive
3	5/12/22	3m/3.6 Kg	DCM	ECMO	2m/2.9 Kg	ABO c	Heart	20	132	L	D MO failure

**Table Legend:** Rec: recipient; Don: Donor; Morgan: Multiorgan donation; fWIT: functional warm ischemic time; cold IT: cold ischemic time; MO: multiorgan

**Image 1** Table with donor and recipients data

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## Adult Congenital Heart Disease

### O-71

#### Gut dysbiosis in patients with fontan circulation

Hideo Ohuchi<sup>1</sup>, Ryotaro Asano<sup>2</sup>, Aki Mori Mori<sup>1</sup>, Tomohiko Ishibashi<sup>3</sup>, Kenichi Kurosaki<sup>1</sup>, Yoshikazu Nakaoka<sup>3</sup>

<sup>1</sup>Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Suita, Japan; <sup>2</sup>Department of Cardiovascular Medicine, National Cerebral and Cardiovascular Center, Suita, Japan;

<sup>3</sup>Department of Vascular Physiology, National Cerebral and Cardiovascular Center, Suita, Japan

**Background and Aim:** Gut dysbiosis causes systemic inflammation through the leaky intestine due to low perfusion and/or venous congestion and reflects disease severity in patients with chronic heart failure (HF). Gut dysbiosis may play a significant role of failing pathophysiology in patients with Fontan circulation (FC). This study was to explore a presence of gut dysbiosis and to clarify its association with Fontan pathophysiology.

**Method:** We prospectively assessed gut microbiomes with 16SrRNA analysis of bacterial DNA extracted from fecal sample in 108 consecutive patients with FC (24 ± 8 years) and 44 healthy controls (22 ± 11 years) between 2019 and 2022. α diversity of the microbiome was assessed by observed number of operational taxonomic unit (OTU) and Faith's phylogenetic diversity (FPD) and the β diversity was assessed by UniFrac analysis and these diversities were compared with Fontan pathophysiology, including hemodynamics, von Willebrand factor (VWF) as an indicator of bacterial

translocation, M2BPGi as an index of liver fibrosis, peak oxygen uptake (PVO<sub>2</sub>) and HF re-hospitalization.

**Results:** Compared to controls, observed OTU (221 ± 59 vs. 267 ± 104) and FPD (22 ± 5 vs. 25 ± 8) were lower (p ≤ 0.01 for both) and β diversity was significantly different in FC patients (p ≤ 0.01). In FC patients, observed OTU and FPD were correlated inversely with central venous pressure and positively with PVO<sub>2</sub> (p ≤ 0.05 for both). In addition, both α diversity indices were inversely correlated with plasma levels of VWF and M2BPGi (p ≤ 0.05 for both). During a mean follow-up of 18 months after the assessment, 16 patients required unscheduled HF re-hospitalization and the decrease in both α diversity indices were associated with the re-hospitalization (p ≤ 0.05 for both).

**Conclusions:** Patients with FC exhibited gut dysbiosis with low α and β diversities when compared to healthy subjects. The gut dysbiosis was associated with reduced exercise capacity and a high risk of HF re-hospitalization. Furthermore, this dysbiosis may be involved in the mechanisms responsible for the progression of Fontan associated liver disease through the leaky gut.

**Keywords:** Fontan, gut dysbiosis, heart failure, Fontan associated liver disease

### O-72

#### Liver life - liver stiffness affected through food intake by fontan-patients, teens and adults, determined with elastography

Richard Mühlberg, Annabell Braun, Nikolaus Haas, Marcus Fischer, Zora Meyer

Division of Pediatric Cardiology and Intensive Care, University Hospital, Ludwig Maximilian University of Munich, München, Deutschland

**Background and Aim:** For several years, patients with single ventricle hearts have been palliated according to the Fontan-Principle. It is known that due to the increased post hepatic pressure, chronic liver congestion develops, which can lead to liver fibrosis, cirrhosis and carcinoma. Liver elastography is available as a non-invasive sonographic method to determine the stage of fibrosis. The measured values depend on many influencing factors, such as food intake, age and respiration. These possible confounders have not been investigated in Fontan-Patients so far. This study validates the influence of food intake on liver stiffness in Fontan-Patients and healthy subjects in order to optimize the examination conditions in the future. The impact of respiration on liver stiffness is discussed in a separate article.

**Method:** A total of 25 patients with Fontan circulation (group 1) and a healthy control group, which consist of 50 volunteers (group 2), were recruited for the study. Liver stiffness was evaluated using liver elastography once before (T0, after 6 hours of fasting) and at specific time intervals (T:15, 30, 45, 60, 90, 120, 150, 180) after standardized food intake. Each time measurements were performed in inspiration and expiration. A standardized meal from comparable studies, a 500 ml chocolate drink was used to simulate ingestion. The nutrient distribution (600kcal) corresponds approximately to an average meal.

**Results:** There was a significant increase in liver stiffness at T15 and T30 after food intake in the control group (group2: T15 = 0, 78 kPa = 19, 0%, T30 = 0, 64 kPa = 16, 7%), whereas liver stiffness did not differ significantly at any time in Fontan-Patients between fasting (T0) and postprandial values. Because of age influence on liver stiffness, we divided each group into two equally sized subgroups (1a, 1b and 2a, 2b) of different ages as part of the data analysis. No significant difference was found between the age groups.

**Conclusions:** With these results it is shown for the first time that in Fontan-Patients the time of food intake has no clinical significance

for the values obtained in liver elastography. In future, this will be of particular importance for ambulant controls of Fontan-Patients, as fasting is not required for the examination.

**Keywords:** sonography, congenital heart disease, follow-up

### O-73

#### Should veno-venous collaterals be closed in fontan patients?

Shin Ono, Kazuma Tsuno, Takeshi Ikegawa, Takuya Wakamiya, Ki Sung Kim, Sadamitsu Yanagi, Hideaki Ueda

Department of Cardiology, Kanagawa Children's Medical Center

**Background and Aim:** Veno-venous collaterals (VVC) occur in some patients after Fontan operation, which may decrease SpO<sub>2</sub> but may also alleviate elevated central venous pressure. It is unclear whether VVC has an adverse or positive effect on the Fontan circulation. We have previously reported an inverse relationship between peripheral venous pressure during peak exercise (peakVP: mmHg) and oxygen uptake during peak exercise (peakVO<sub>2</sub>: l/min/kg) in Fontan patients. In this study, we investigated whether there is a difference in the correlations between peakVP and peakVO<sub>2</sub> in Fontan patients and compared hemodynamics with and without VVC.

**Method:** In 47 postoperative Fontan patients (19 with VVC), peakVP and peakVO<sub>2</sub> were measured by cardiopulmonary exercise testing using a Treadmill while peripheral venous pressure was measured. peakVP was used as the dependent variable, and

peakVO<sub>2</sub> and presence of VVC were used as the explanatory variables. Multiple regression analysis with interaction was performed to compare the correlation between peakVP and peakVO<sub>2</sub> with and without VVC.

**Results:** Two regression equations were created as follows, with and without VVC. Two regression equations showed statistically significant difference ( $P < 0.0001$ ). Two regression lines intersected at peakVO<sub>2</sub>: 27.8.

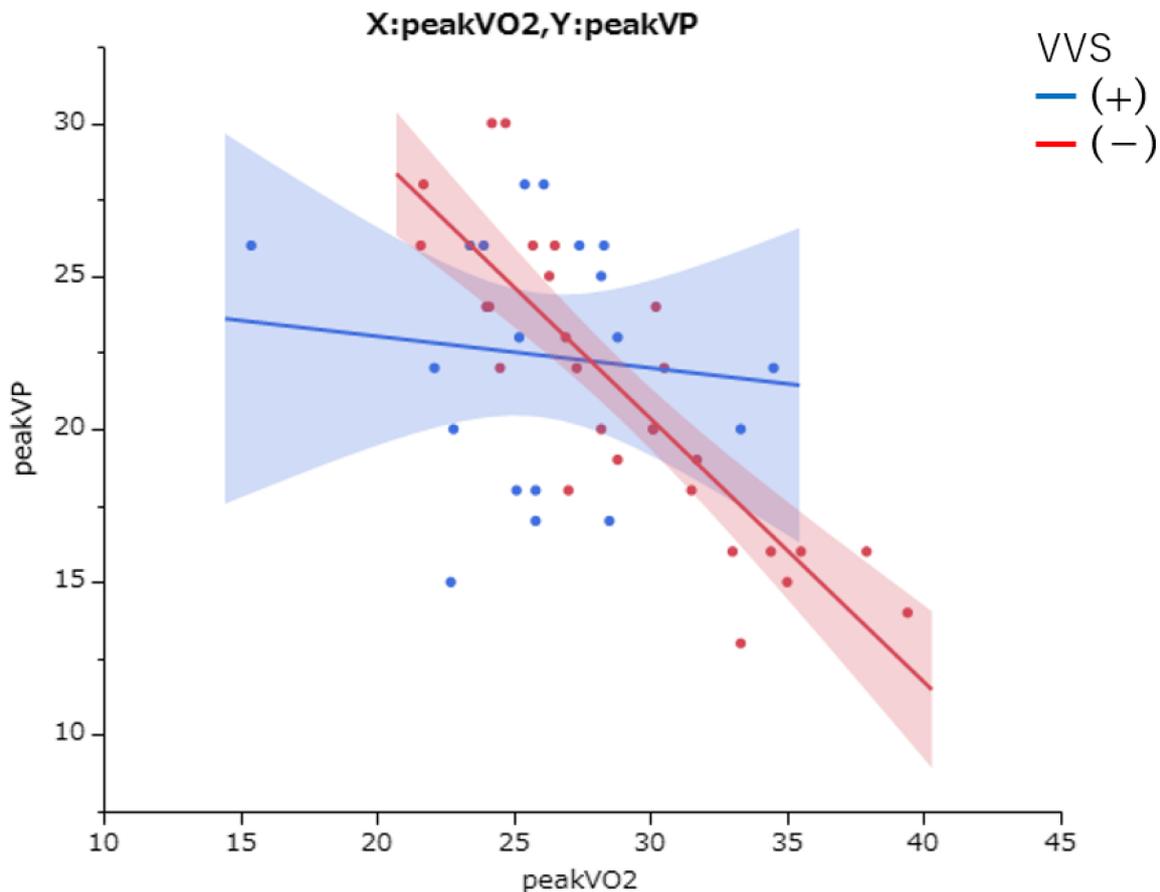
VVC (+): PeakVP =  $35.6 - (0.48 \times \text{peakVO}_2) - 0.005 + [0.38 \times (\text{peakVO}_2 - 27.8)]$

VVC (-): PeakVP =  $35.6 - (0.48 \times \text{peakVO}_2) + 0.005 + [-0.38 \times (\text{peakVO}_2 - 27.8)]$ .

**Conclusions:** The presence of VVC buffers the increase in PeakVP against the decrease in PeakVO<sub>2</sub>. The cutoff value for PeakVO<sub>2</sub> is 27.8, with PeakVO<sub>2</sub> < 27.8 resulting in lower PeakVP in patients with VVC and PeakVO<sub>2</sub> ≥ 27.8 resulting in higher PeakVP. In other words, embolization of the VVC in patients with PeakVO<sub>2</sub> < 27.8 may exacerbate elevated venous pressure during exercise, while PeakVO<sub>2</sub> ≥ 27.8 may reduce venous pressure during exercise. PeakVO<sub>2</sub> can be a criterion for determining who should be treated for embolization of the VVC in postoperative Fontan patients.

**Keywords:** Fontan, Veno-venous collaterals, Cardiopulmonary Exercise Test, venous pressure

Correlation between PeakVP and PeakVO<sub>2</sub> with and without VVC



Two regression equations showed statistically significant difference ( $P < 0.0001$ ). Two regression lines intersected at peakVO<sub>2</sub>: 27.8.

O-74

**Development and validation of a deep learning algorithm for cardiac MR image quantification in patients with tetralogy of fallot**

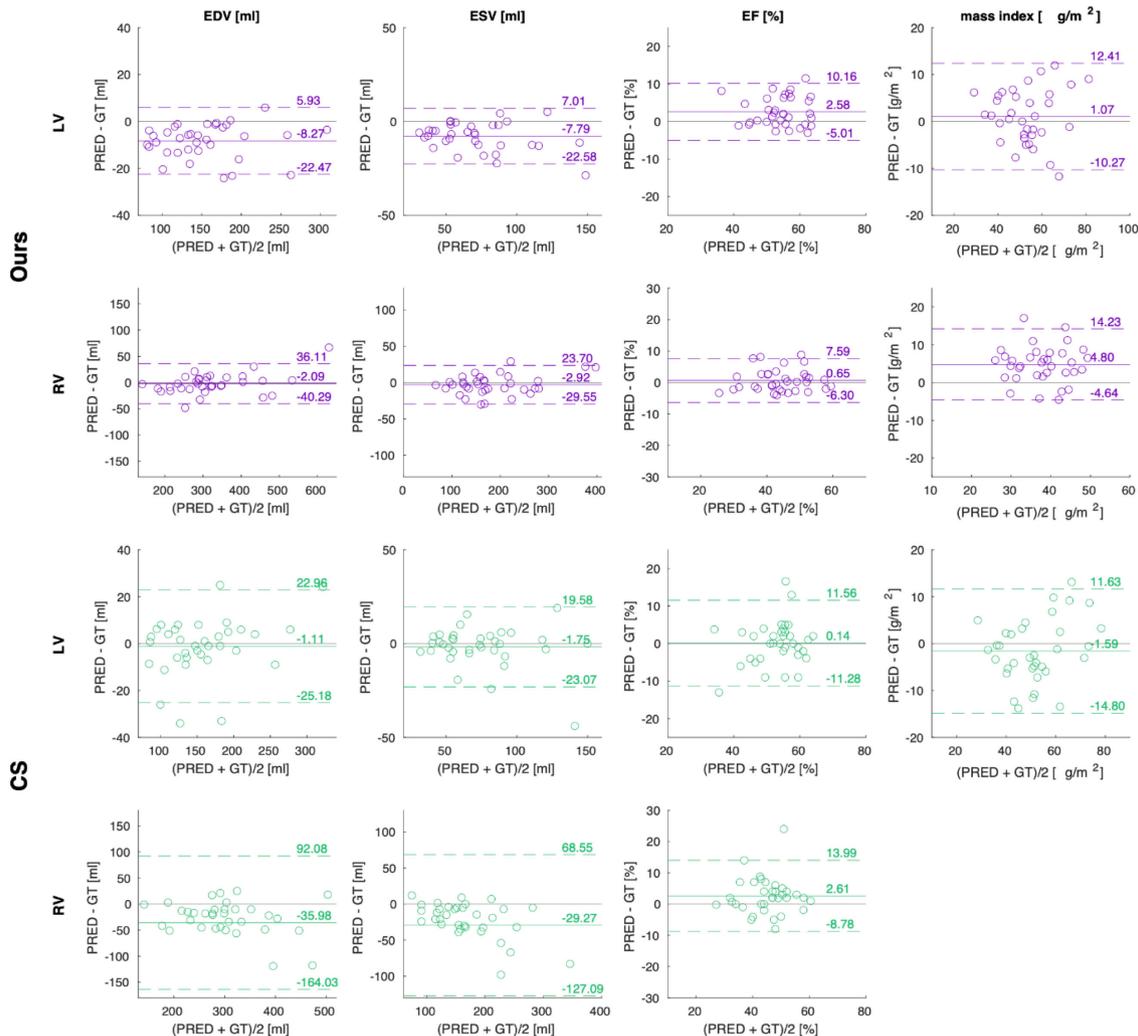
Sofie Tilborg<sup>1</sup>, Tiffany Liang<sup>3</sup>, Stavroula Raptis<sup>3</sup>, Ayako Ishikita<sup>3</sup>, Werner Budts<sup>4</sup>, Tom Dresselaers<sup>5</sup>, Jan Bogaert<sup>5</sup>, Frederik Maes<sup>1</sup>, Rachel M. Wald<sup>2</sup>, Alexander Van De Bruaene<sup>4</sup>

<sup>1</sup>Department of Electrical Engineering, Division of Processing Speech and Images, KU Leuven, Leuven, Belgium; <sup>2</sup>Medical Imaging Research Center, UZ Leuven, Leuven, Belgium; <sup>3</sup>Department of Medical Imaging, Toronto General Hospital, University of Toronto, Toronto, Canada; <sup>4</sup>Division of Cardiology, Peter Munk Cardiac Centre, University of Toronto, Canada; <sup>5</sup>Division of Cardiology, Peter Munk Cardiac Centre, University of Toronto, Canada; <sup>6</sup>Department of Cardiovascular Sciences, KU Leuven, Leuven, Belgium; <sup>7</sup>Department of Imaging and Pathology, Division of Radiology, KU Leuven, Leuven, Belgium

**Background and Aim:** Deep learning is a state-of-the-art technique for automatic segmentation of LV and RV in cardiac MRI. However, these methods are typically developed and validated

in structurally normal hearts or in acquired heart disease, and as such are not applicable to congenital cardiac disease lesions such as tetralogy of Fallot (TOF). This study aimed to (1) develop and validate a fully automated approach for biventricular segmentation and quantification in patients with repaired TOF and (2) compare this approach to a commercially available software (CS). **Method:** We used an institutional dataset with 131 TOF subjects, divided into n = 96 subjects for model development and n = 35 for validation, and the public ACDC dataset (Bernard, 2018) containing 100 subjects with no or acquired cardiac disease. LV and RV endocardium at end-diastole (ED) and end-systole (ES), along with the epicardium at ED, were manually delineated by a single expert radiologist in the TOF dataset. We trained a 3D convolutional neural network (CNN) on three different data splits: 1) ACDC dataset, 2) TOF dataset, 3) both datasets combined. We compared their performances in a 5-fold cross-validation on the TOF development dataset using the Dice similarity coefficient (DSC) which calculates the percent of overlap between manual and automatic segmentations. Additionally, we calculated EDV, ESV, EF and myocardial mass index for LV and RV on the

Bland-Altman plot comparing our approach with commercial software



Bland-Altman plots for our approach (Ours) and for the commercial software (CS) evaluated on the test split of the TOF dataset. Statistically significant differences as assessed by the Wilcoxon signed rank test with a significance level of 5% were only found for RV EDV and RV ESV. For CS, one outlier for RV EDV (PRED-GT = -364ml) and one outlier for RV ESV (PRED-GT = -280ml) were not shown.

TOF validation dataset by applying our best performing CNN to all frames, selecting ED and ES automatically as minimum and maximum volumes. This was compared to an automatic commercial software.

**Results:** The significantly best DSCs were obtained when training on ACDC+TOF (DSC = 88.6±9.9%), and the worst when training on ACDC alone (DSC = 80.7±14.0%). TOF alone achieved a DSC of 88.2±10.0%. Our LV quantification showed no significant difference with CS. However, RV EDV and ESV were significantly underestimated by CS, especially for larger volumes, with a bias of -35.5ml and -29.3ml respectively, which was only -2.1ml and -2.9ml for our approach (see figure).

**Conclusions:** We developed and validated a deep learning approach for biventricular quantification in TOF patients. Compared to the commercial solution, our approach is superior for RV quantification, indicating its potential value in clinical practice.

**Keywords:** tetralogy of Fallot, left and right ventricle, deep learning, segmentation, quantification

### O-75

#### **Percutaneous edge-to-edge tricuspid repair in patients with systemic right ventricle: A french multicenter cohort study**

Xavier Iriart<sup>1</sup>, Alexandre Silini<sup>1</sup>, Corina Vasile<sup>1</sup>, Zakaria Jalal<sup>1</sup>, Pascal Amedro<sup>1</sup>, Valentin Femenia<sup>3</sup>, Robin Le Ruz<sup>2</sup>, Charlene Bredy<sup>3</sup>, Helene Bouvaist<sup>4</sup>, Jean Benoit Thambo<sup>1</sup>, Patrice Guerin<sup>2</sup>

<sup>1</sup>Department of Pediatric and Adult Congenital Cardiology, University Hospital of Bordeaux, France; <sup>2</sup>Department of Cardiology, University Hospital of Nantes, France; <sup>3</sup>Department of Cardiology, University Hospital of Montpellier, France; <sup>4</sup>Department of Cardiology, University Hospital of Grenoble, France

**Background and Aim:** Systemic tricuspid regurgitation (STR) is a crucial predictor of reduced functional capacity, heart failure, and mortality in patients with systemic right ventricle (SRV) and a common therapeutic dilemma because medical treatment has not proven to be effective, while valvular surgery is associated with a high rate of complications. Transcatheter edge-to-edge repair (TEER) may be a safe and effective alternative to surgery in high-risk surgical patients.

**Method:** The TEER SRV cohort is a multicenter, longitudinal, descriptive, prospective cohort study of patients with SRV undergoing TEER for severe STR. Eight high-risk surgical patients with severe systemic tricuspid regurgitation (TR) undergoing percutaneous repair with the MitraClip or TriClip system (Abbott) were enrolled in the study between May 2019 and October 2021. We collected and compared clinical (symptoms, quality of life questionnaires), biological (renal function, BNP/NT-proBNP levels), VO<sub>2</sub> max, echocardiographic, and cross-sectional imaging (TR severity, RV dilatation, and dysfunction) data at baseline and six months after the procedure.

**Results:** Early complications included two successfully ablated common flutters in acTGA. Clinically, we observed an improvement in functional status after six months and in the median physical (+9.4 points) and mental (+11 points) quality of life scores. None had been hospitalized for heart failure at six months (vs. 71% within the 6 months before TEER). The median BNP and NT-proBNP blood levels decreased by 105 and 178 pg/mL, respectively, whereas cardiopulmonary exercise testing data did not substantially change. The effective regurgitant orifice area and regurgitant volume assessed by transthoracic

echocardiography improved by 21.5 mm<sup>2</sup> and 14.9 mL, respectively, whereas the median SRV volumes (measured by magnetic resonance imaging or computed tomography) decreased by 38.5 mL/m<sup>2</sup> and 33 mL/m<sup>2</sup> (end-diastolic and end-systolic volumes, respectively). The SRV ejection fraction increased by 11.5%.

**Conclusions:** Percutaneous TEER for STR appears feasible, safe, and effective. It improves patient symptoms, quality of life, heart failure biomarkers, STR grade, and SRV dilation and function at six months. Additional dedicated studies and registries are necessary to predict the outcomes and to define the optimal indications for TEER.

**Keywords:** systemic right ventricle, tricuspid regurgitation, percutaneous edge-to-edge repair.

### O-76

#### **Prognostic value of blood biomarker - hs-TnT, NT-probnp and CRP - for event free survival in adults with congenital heart disease**

Laura Willinger<sup>1</sup>, Alfred Hager<sup>1</sup>, Renate Oberhoffer Fritz<sup>2</sup>, Peter Ewert<sup>1</sup>, Jan Müller<sup>2</sup>

<sup>1</sup>Department of Congenital Heart Disease and Pediatric Cardiology, Deutsches Herzzentrum München, Technische Universität München, Germany; <sup>2</sup>Institute of Preventive Pediatrics, Technische Universität München, Germany

**Background and Aim:** High-sensitive-troponin-T (hs-TnT), N-terminal pro B-type natriuretic peptide (NT-proBNP), and C-reactive protein (CRP) are frequently used as prognostic biomarkers in symptomatic and/or hospitalized adults with congenital heart disease (ACHD). However, their prognostic value and clinical utility for risk-stratification in clinically stable ACHD is not yet well established. The aim of this study was to examine the predictive value of hs-TnT, NT-proBNP and CRP for survival and cardiovascular events in clinically stable ACHD.

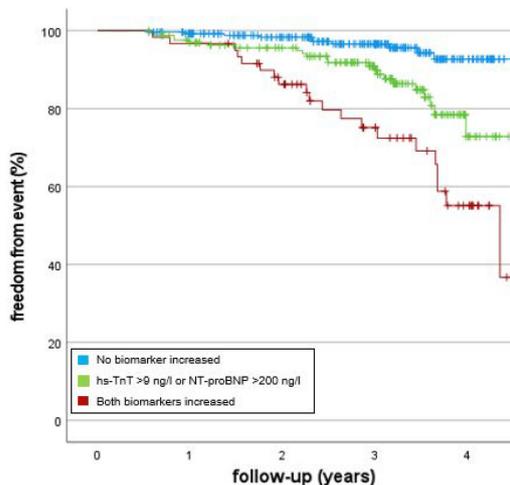
**Method:** This is a prospective cohort study with 495 outpatient ACHD (43.9 ± 10.0 years, 49.1% female) who underwent venous blood sampling including hs-TnT, NT-proBNP and CRP. According to ACC criteria 79 patients had a simple, 146 a moderate and 267 a complex severity class of heart defects, while 3 participants could not be defined. All ACHD were in NYHA class I or II at inclusion. Patients were followed for survival and the occurrence of cardiovascular events including sustained ventricular tachycardia, hospitalization with cardiac decompensation, electrophysiological procedure (ablation), interventional catheterization, pacemaker implantation and cardiac surgery. Survival analyses was performed via Cox proportional hazards regression analysis and Kaplan-Meier curves.

**Results:** After a mean follow-up of 2.8 ± 1.0 years, 53 patients (10.7%) reached a cardiac-related endpoint. Hs-TnT (p = .005) and NT-proBNP (p = .018) were independent predictors of death or cardiac-related events in stable ACHD, whilst the prognostic value of CRP vanished after multivariable adjustment (p = .057). Cut-off values with best sensitivity and specificity for event-free survival were hs-TnT ≤ 9 ng/l and NT-proBNP ≤ 200 ng/l according to ROC curve analysis. Patients with increased hs-TnT and NT-proBNP had a 7.7-fold (CI 3.57 – 16.40, p < 0.001) increased risk for death and cardiac-related events compared to patients without elevated blood values.

**Conclusions:** In stable outpatient ACHD subclinical values of hs-TnT and NT-proBNP are a useful, simple, and independent prognostic tool for adverse cardiac events and survival. These blood

biomarkers provide valuable prognostic information for regular clinical aftercare in specialized centers already at subclinical level and may contribute to improved risk stratification in ACHD.

**Keywords:** blood biomarker, hs-TnT, CRP, NT-proBNP, congenital heart disease



Patients at risk (n)	0	1	2	3	4
No biomarker increased	255	240	198	128	30
One biomarker increased	167	153	133	99	12
Both biomarkers increased	60	58	47	30	12

Kaplan-Meier survival curves according to the presence or absence of risk factors based on the cut-off values displayed in the ROC curve (hs-TnT >9 ng/l; NT-proBNP >200 ng/l).

### O-77

#### Outcomes and prognostic factors of congenitally corrected transposition of great vessels in children

Sylvie Di Filippo<sup>1</sup>, Oriane Besset<sup>1</sup>, Marine Nouhaud<sup>1</sup>, Olivier Metton<sup>1</sup>, Julia Mitchell<sup>1</sup>, Sylvie Di Filippo<sup>2</sup>

<sup>1</sup>Department of Congenital Heart Disease, Claude Bernard University, Lyon, France; <sup>2</sup>Congenital Cardiology, Cardiothoracic Center of Monaco

**Background and Aim:** Double discordance (DD) is a rare congenital heart disease, associated or not with other various and complex cardiac lesions. Isolated DD is a well-tolerated cardiopathy but long-term prognosis is impaired by heart failure from dysfunction of systemic right ventricle (RV).

The aims of this study are: 1) to compare isolated DD with DD associated to complex lesions, 2) to evaluate the clinical outcome of patients with isolated DD, and 3) to determine if early operation on asymptomatic children in infancy has long-term functional benefit.

**Method:** This is a retrospective single-center descriptive analysis of patients who reached the inclusion criteria. The patients were divided into two groups: DDI (double discordance isolated) and CCC (complex DD). Demographics, clinical data, imaging measurements, major events and outcomes were collected.

**Results:** 151 patients (91 males) were included: 63 in CCC (41.8%) and 88 in DDI (58.2%). Age at diagnosis was  $6.9 \pm 15.9$  years:  $1 \pm 3.5$  years in CCC and  $10.4 \pm 19$  years in DDI ( $p = 0.0068$ ). Atrioventricular block occurred in 38.5%, pacemaker implantation in 32.4% at age  $20.4 \pm 20.9$  years, and first episode of heart failure at  $33.8 \pm 25$  years. All patients in CCC underwent one or more surgeries, and 35 (40.2%) in DDI had at least one surgery. At end follow-up, patients were in NYHA class 1, 2, 3 or 4 in

respectively 13%, 45%, 29% and 13% of cases. Heart failure, pacemaker and death or transplantation were similar between CCC and DDI. Tricuspid regurgitation in DDI was  $\geq 3$  in 7 cases in DDI,  $< 3$  in 52, and RV ejection fraction was  $47\% \pm 11.7\%$ . Surgery was performed in 35 of DDI patients (40%): first surgery at  $4.7 \pm 8.9$  years, second at  $8.2 \pm 10$  years. Survival was higher in unoperated DDI ( $p = 0.006$ ) and operated patients had worse survival after double switch operation, compared to non-operated ( $p < 0.0001$ ).

**Conclusions:** Our results showed better prognosis and survival rates in unoperated patients with isolated double discordance anatomy. The study could not conclude on the long-term advantage of the double switch program compared to the natural history of isolated double discordance

**Keywords:** Congenital heart disease, Congenitally corrected transposition, Double discordance, Double switch program

### O-78

#### The risk of premature atherosclerosis in survivors of childhood cancer: A nested case-control study from the PCS2 cohort

Jonathan P Wong<sup>1</sup>, Cameron Slorach<sup>1</sup>, Wei Hui<sup>1</sup>, Mark Friedberg<sup>1</sup>, Emily Lam<sup>1</sup>, Kyle Runeckles<sup>2</sup>, Steve Fan<sup>2</sup>, Paul Nathan<sup>1</sup>, Luc Mertens<sup>1</sup>  
<sup>1</sup>Department of Pediatrics, The Hospital for Sick Children, University of Toronto, Toronto, Canada; <sup>2</sup>Ted Rogers Computational Program, Ted Rogers Centre for Heart Research, Peter Munk Cardiac Centre, University Health Network, Toronto, Canada

**Background and Aim:** Cardiac disease is the most common cause of non-cancer related mortality in childhood cancer survivors (CCS). There is growing evidence that coronary artery disease (CAD) causes significant morbidity in these survivors. Few studies have adequately assessed the prevalence of atherosclerosis in pediatric CCS, which have largely reported positive signs of premature atherosclerosis. These studies were limited by small numbers and a single modality of assessment used. We aimed to determine the presence and extent of non-invasive markers of early atherosclerosis among a cohort of CCS compared to matched healthy controls using a comprehensive non-invasive assessment protocol.

**Method:** We conducted a nested case-control study among CCS who had received anthracycline therapy from the PCS2 (Preventing Cardiac Sequelae in Pediatric Cancer Survivors) prospective study at the Hospital for Sick Children, Toronto, and Princess Margaret Cancer Centre, Toronto. Each participant underwent non-invasive ventricular and vascular assessment using advanced functional echocardiography, carotid intima media thickness (cIMT), pulse wave velocity (PWV) and flow mediated dilation (FMD). This study received institutional research ethics approval and the participants provided informed consent.

**Results:** 206 CCS were recruited and matched to 150 healthy controls. The mean age was  $17.38 \pm 8.11$  and 94 (45.6%) were female. Contrary to prior reports, we found no difference in non-invasive markers of premature atherosclerosis between CCS and controls, including cIMT ( $0.046 \text{ cm} \pm 0.005$  vs  $0.046 \text{ cm} \pm 0.005$ ,  $p = 0.91$ ), carotid to femoral PWV ( $5.5 \pm 0.9$  vs  $5.6 \pm 0.9$ ,  $p = 0.33$ ), and FMD ( $6.36\% \pm 3.11$  vs  $6.25\% \pm 3.16$ ,  $p = 0.60$ ). Similar to previous reports, there were a significant difference in mitral valve E/E' ratio ( $5.48 \pm 1.40$  vs  $5.09 \pm 1.04$ ,  $p = 0.003$ ) as a marker of diastolic function. Additionally, there was a statistically significant, but clinically insignificant, difference in systolic function measured by M-mode EF between the two groups ( $65\% \pm 6$  vs  $69\% \pm 6$ ,  $p < 0.001$ ) and average global longitudinal strain ( $-20.85 \pm 2.25$  vs  $-21.64 \pm 2.07$ ,  $p = 0.001$ ).

**Conclusions:** This study reports no difference in presence of non-invasive markers of premature atherosclerosis between CCS and healthy controls using the largest reported cohort of CCS and utilizing a comprehensive non-invasive assessment protocol. Additional study is needed to determine what factors place this population at increased risk of CAD.

**Keywords:** Coronary artery disease, Childhood Cancer Survivor, Carotid intima media thickness, Flow mediated dilation, Pulse wave velocity

## Cardiovascular Morphology

### O-79

#### A loss-of-function variant in INVS in hypoplastic left heart syndrome

Maarab Mohammed Al Korashy<sup>1</sup>, Amal Mohammed Al Hashem<sup>2</sup>, Faten Mohammed Al Hadheq<sup>1</sup>, Saud Abdrabouh Takroni<sup>1</sup>, Nadiyah Salem Al Ruwaili<sup>3</sup>, Zuhair Nasser Al Hassan<sup>1</sup>

<sup>1</sup>Center of Genomic Medicine, King Faisal Specialist Hospital & Research Centre (KFSH&RC), Riyadh, Saudi Arabia.; <sup>2</sup>Division of Medical Genetics, Department of Pediatrics, Prince Sultan Medical Military City, Riyadh, Saudi Arabia.; <sup>3</sup>Heart center Department, King Faisal Specialist Hospital & Research Centre (KFSH&RC), Riyadh, Saudi Arabia

**Background and Aim:** Hypoplastic left heart syndrome (HLHS), is a serious complex congenital heart disease that affects the left ventricle, aorta, and mitral valve. While the genetic basis of HLHS remains largely undetermined, variants in several genes have been identified. Recessive mutations in INVS are known to cause infantile nephronophthisis which can be associated with heart valve and septal defects. To our knowledge, mutated INVS has not been reported in association with HLHS.

**Method:** In a consanguineous family with three affected children with HLHS and renal malformations, we performed a combined approach of homozygosity mapping and whole exome sequencing. Family segregation was conducted on filtered variants that were validated by Sanger sequencing.

**Results:** A homozygous loss-of-function pathogenic variant (c.1807C>T, p.Arg603Ter) in INVS was detected. Homozygosity mapping and family segregation were consistent with a recessive inheritance of the heart defect.

**Conclusions:** Our findings suggest that loss-of-function variants in INVS are associated with HLHS. We suggest adopting next generation sequencing, that covers INVS, in the diagnostic evaluation of familial forms of HLHS

**Keywords:** Hypoplastic Left Heart Syndrome, Congenital Heart Disease, Whole Exome Sequencing.

### O-80

#### Virtual reality volume rendering in assessment of ductus arteriosus anatomy in duct-dependent pulmonary circulation

Nikola Topuzov<sup>1</sup>, Antoni Ludew<sup>1</sup>, Maksym Helman<sup>1</sup>, Anna Migda<sup>2</sup>, Grażyna Brzezińska Rajszyz<sup>2</sup>, Maria Zubrzycka<sup>2</sup>, Bożena Rewers<sup>2</sup>, Andrzej Kościeszka<sup>3</sup>, Adam Koleśnik<sup>4</sup>

<sup>1</sup>Department of Descriptive and Clinical Anatomy, Medical University of Warsaw, Warsaw, Poland; <sup>2</sup>Cardiovascular Interventions Laboratory, The Children's Memorial Health Institute, Warsaw, Poland;

<sup>3</sup>Department of Pediatric Radiology, The Children's Memorial Health Institute, Warsaw, Poland; <sup>4</sup>Department of Descriptive and Clinical Anatomy, Medical University of Warsaw, Warsaw, Poland;

Cardiovascular Interventions Laboratory, The Children's Memorial Health Institute, Warsaw, Poland

**Background and Aim:** Congenital heart defects with duct-dependent pulmonary circulation comprise a considerable amount of patients requiring early surgical or interventional palliative treatment. The increasing popularity of transcatheter ductus arteriosus (DA) stenting raises a question – what patterns of anatomy can be found in this group of patients? The aim of our study was to assess the value of direct virtual reality volume rendering in assessment of DA anatomy in patients with duct-dependent pulmonary circulation.

**Method:** 19 anonymised CT scans of patients aged from 1 to 43 days (median = 15) were obtained and analysed in virtual reality using VMersive software (VR-Learning, Warsaw, Poland). The group consisted of 11 patients with tetralogy of Fallot, 6 with double outlet right ventricle and 2 with tricuspid atresia.

The morphology of DA and aortic arch was analysed and categorised. Following measurements were performed: DA diameter and length, presence and diameter of DA and PA stenosis, number of DA segments and their internal angles. McGoon ratio was calculated as a summation of left and right PA divided by descending aorta diameter.

**Results:** DA shape was categorised into 3 categories: straight (5.2%), single-bend (47.4%, divided into L-shaped and U-shaped) and tortuous (47.4%, divided into S-shaped and spiral), depending on the number of segments and their spatial relations. Mean DA length was 16.4 mm and diameter 4.1 mm. Stenosis in DA was identified in 3 cases (1 in proximal part, 2 in distal). Stenosis in left PA was identified in 8 cases (2 proximal, 3 proximal and distal, 3 whole length) and 1 case in proximal right PA.

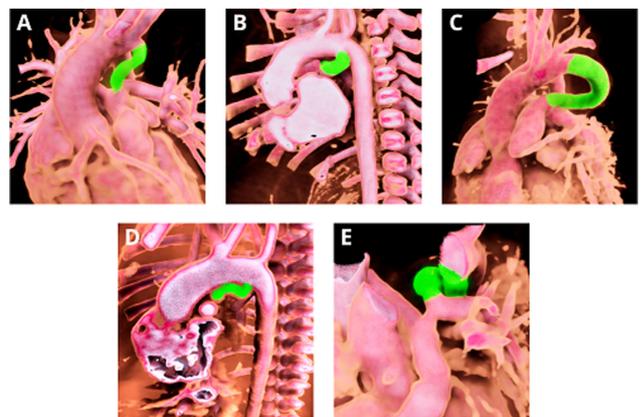
The morphology of the aortic arch and DA was categorised into 3 types: left-sided arch and DA (63.2%), right-sided arch and DA (10.5%) and right-sided arch with left sided DA (26.3%).

McGoon ratio ranged from 1.08 to 1.77.

**Conclusions:** Virtual reality volume rendering allows for spatial recognition of shape and topographic relations of DA and aortic arch, as well as performing various measurements. The method allows for precise assessment of DA ostium, its distal connection site and presence of stenoses in DA and PAs, which could prove valuable for planning of transcatheter DA stenting.

**Keywords:** cardiovascular morphology, ductus arteriosus, virtual reality volume rendering, duct-dependent pulmonary circulation

#### Types of ductus arteriosus morphology



Straight (A); Single-bended: L-shaped (B) and U-shaped (C); tortuous: S-shaped (D) and spiral (E)

## O-81

**Creation of a neonatal congenital heart digital twin to assist with parent recognition of cardiac compromise following congenital heart surgery**

Michael E Kim, Nadine A Kasparian, Meghan M Chlebowski, Michelle Rios, Ian Anderson, Matthew Necamp, William Burke, David Davis, David Ls Morales, Ryan A Moore  
The Heart Institute, Cincinnati Children's Hospital Medical Center, Department of Pediatrics, Cincinnati, OH, USA

**Background and Aim:** Neonates with critical congenital heart disease (CHD) remain high risk following cardiac surgery, even under the care of a skilled healthcare team. This risk can increase following discharge with any level of cardiac compromise. Recently, our team conducted a study which independently identified two high risk groups following congenital heart surgery (CHS): 1) families with lower socioeconomic status and 2) families of neonates. Currently, there is no formal training for families to recognize potential cardiac compromise (i.e., cyanosis, tachypnea, poor perfusion). Strategies to address this gap in parent training are needed.

**Method:** We developed a realistic digital twin of a neonate s/p CHS to be utilized in a virtual reality (VR) environment to assist parents with recognition of signs of cardiac compromise. Employing clinician and simulation educator feedback, the neonate was created to have easily differentiated levels of cyanosis, tachypnea, and poor perfusion. We tested the CHD neonate in the VR environment for feasibility and acceptability amongst a diverse user group. The neonate was coded with various skin tones to address diversity, equity, and inclusion criteria of the potential end-user.

**Results:** A small pilot user group (n = 10) of clinicians, nurses, trainees, simulation educators, and parents ran through a prototype VR scenario that included the neonatal CHD digital twin. The neonatal avatar was noted to be highly realistic by all. The various levels of cardiac compromise (e.g. mild, moderate, and severe) were able to be easily differentiated by all users for cyanosis, tachypnea, and poor perfusion. The neonate interactivity and VR environment were noted to be moderately difficult to navigate. Additional feedback was elicited from the pilot group to improve user interface (UI) and user experience (UX) for the next phase focused on a larger pilot group of parent end-users.

**Conclusions:** This is the first digital twin of a CHD neonate to be developed specifically for VR training of parents to improve recognition of cardiac compromise. A select group of users provided



Example of neonatal digital twin in the VR environment demonstrating cyanosis

critical feedback to enhance the UI/UX for the next development phase. Future plans include a parent VR simulation study to determine the psychosocial and clinical benefits of the training.

**Keywords:** virtual reality, digital twin, simulation, discharge planning

Neonatal Digital Twin (VR)

## O-82

**Exploring the GUT microbiota of infants with complex congenital heart disease undergoing cardiopulmonary bypass (the gumibear study)**

Claire Magnier<sup>1</sup>, Kiera Murphy<sup>2</sup>, Fatma Koc<sup>2</sup>, Dominic Jenkins<sup>3</sup>, Mong Hoi Tan<sup>3</sup>, Molly O' Toole<sup>3</sup>, Jordan Boyle<sup>3</sup>, Niamh Maguire<sup>3</sup>, Adam James<sup>4</sup>, Sophie Duignan<sup>4</sup>, R. Paul Ross<sup>2</sup>, Catherine Stanton<sup>2</sup>, Colin J. Mc Mahon<sup>4</sup>

<sup>1</sup>School of Nursing, Midwifery & Health Systems, University College Dublin, Ireland; <sup>2</sup>APC Microbiome Ireland, University College Cork, Co Cork, Ireland; <sup>3</sup>Children's Health Ireland at Crumlin, Dublin, Ireland; <sup>4</sup>Department Paediatric Cardiology, Children's Health Ireland at Crumlin, Dublin, Ireland

**Background and Aim:** The gut microbiota develops from birth, playing a major role in infant health and development. The composition of the gut microbiota is influenced by several factors including mode of delivery, gestational age, feed type and treatment with antibiotics. Infants diagnosed with 'Congenital Heart Disease' (CHD) often require surgery involving cardiopulmonary bypass (CPB) early in life. The impact of this type of surgery on the integrity of the gut microbiome is poorly understood.

**Method:** In this pilot study we investigated the gut microbiota composition of 13 infants with CHD undergoing surgery involving CPB and matched healthy controls. Stool samples were collected from all participants pre-surgery and post-surgery, and were analysed using 16S ribosomal DNA sequencing.

**Results:** The gut microbiota of pre-surgery CHD subjects separate significantly from control subjects using Principal Component Analysis (Adonis, p <0.001). Significantly lower abundances of Actinobacteria (p <0.001) and higher abundances of Proteobacteria (p = 0.009) were observed in CHD patients at phylum level. At genus level, abundances of Bifidobacterium were significantly lower in CHD group (p <0.001). Post-surgery CHD subjects also separately significantly from control (Adonis, p <0.001). Similarly, lower abundances of Actinobacteria (p = 0.004) and higher abundances of Proteobacteria (p = 0.002) were observed post-surgery. At genus level, increased abundances of Escherichia Shigella (p <0.001), Enterococcus (p = 0.001) and Eisenbergiella (p = 0.003), in addition to reduced Bifidobacterium (p = 0.002) were detected in post-surgery CHD subjects.

**Conclusions:** Alterations in gut microbiota composition characterised by increased Proteobacteria and reduced abundance of beneficial Bifidobacterium appears to be a hallmark of patients with CHD. Gut microbiota alterations appeared to be exacerbated by CPB surgery leading to increases in pathogenic microbes Escherichia Shigella, Enterococcus and Eisenbergiella. Further studies are warranted to investigate the microbiota and its role in CPB and the potential for microbiota modulation in optimising outcomes for CPB patients.

**Keywords:** Congenital heart disease; microbiome, cardiopulmonary bypass

## O-83

**Cardiac rhabdomyomas presenting with critical cardiac obstruction in neonates and infants**

Li Yen Ng<sup>1</sup>, Terence Prendiville<sup>1</sup>, Orla Franklin<sup>1</sup>, Jonathan McGuinness<sup>2</sup>, Damien Kenny<sup>1</sup>, Colin J McMahon<sup>1</sup>

<sup>1</sup>Department of Paediatric Cardiology at Children's Health Ireland at Crumlin, Dublin, Ireland; <sup>2</sup>Department of Paediatric Cardiothoracic Surgery at Children's Health Ireland at Crumlin, Dublin, Ireland.

**Background and Aim:** Cardiac rhabdomyoma is a benign tumour commonly associated with tuberous sclerosis, a multisystem neurocutaneous genetic disorder, which results from a mutation in the TSC1 or TSC2 genes. The clinical presentation varies from asymptomatic to sudden cardiac death due to severe inflow or outflow tract obstruction. Treatment strategies for haemodynamically significant lesions include medical therapy with mTOR inhibitors (mTORi) or surgical resection.

We present our experience of children with cardiac rhabdomyoma and significant cardiac obstruction who required intervention over the last decade.

**Method:** Retrospective review of patients with obstructive cardiac rhabdomyoma who received mTORi therapy, surgical resection or hybrid procedure [bilateral pulmonary artery (BPA) bands and ductus arteriosus (PDA) stenting]. Medical records were reviewed for each patient.

**Results:** See Table 1 for summary of the patients' demographic, treatment strategies and outcome.

Five patients presented with haemodynamically significant lesions. Three patients were antenatally diagnosed: 2 of 3 patients with critical left ventricular outflow tract obstruction (LVOTO) and left ventricular failure (LVF), and 1 patient with critical right ventricular outflow tract obstruction (RVOTO). Two patients presented postnatally: 1 patient presented with poor feeding and murmur at 6 days old, echocardiography confirmed a solitary tumour causing severe RVOTO; 1 patient with multiple tumours causing right inflow and LVOTO presented with aborted cardiac arrest at 4 months of age.

Two patients with critical LVOTO and LVF underwent LVOT tumour resection, hybrid BPA bands PDA stenting and mTORi therapy. Of the 2 patients with RVOTO, 1 underwent solitary tumour resection and 1 patient with polycystic kidney

Table 1: Patient demographic, treatment strategies and clinical outcome.

	Case 1	Case 2	Case 3	Case 4	Case 5
Sex	M	M	F	M	F
Antenatal diagnosis (Y/N)	Y	N	Y	Y	N
Presentation	Critical LVOTO, LVF	Aborted cardiac arrest. LVOTO and RV inflow obstruction	Critical RVOTO	Critical LVOTO, LVF	Cardiac murmur. Severe RVOTO
Age of diagnosis	At birth	4 months old	At birth	At birth	6 days old
Tumour burden	Multiple	Multiple	Multiple	Multiple	Single
Weight at diagnosis (kg)	3.09	6	2.86	3.2	3.16
mTORi	Sirolimus	Sirolimus x 7 months, switched to Everolimus x 1 month.	Sirolimus	Sirolimus	No
mTORi dose	0.3mg OD	Sirolimus 0.3mg OD;	0.2mg OD	0.5mg OD	N/A
Peak mTORi level (ng/ml)	92	Level low despite dose adjustment	47.8	108	N/A
Surgical resection (Y/N)	Y. LVOT tumour.	N	N	Y. LVOT tumour.	Y. RVOT tumour.
Hybrid BPA bands + PDA stent	Y	N	N	Y	N
Comorbidities	Factor VII deficiency	Ventricular arrhythmia on amiodarone and sotalol.	PKD with preserved renal function. Hypertension.	AKI, seizure	N/A
Age at latest follow up	RIP	2 years 1 month old	3 years old	4 years old	6 years old
Outcome	Died on day 17 of life due to postop bleeding disorder	Multiple LV tumour, 1 RA tumour, no outflow/ inflow obstruction; preserved biventricular systolic function. Frequent NSVT asymptomatic.	Small residual RV tumour. No inflow/ outflow obstruction. Preserved ventricular systolic function.	Small residual RV tumour. No inflow/ outflow obstruction. Preserved ventricular systolic function.	Alive. No residual cardiac rhabdomyoma.

Abbreviations: M: Male; F: Female; Y: Yes; N: No; LVOTO: left ventricular outflow tract obstruction; RVOTO: right ventricular outflow tract obstruction; LVF: left ventricle failure; RV: right ventricle; mTORi: mTOR inhibitor; LVOT: left ventricular outflow tract; RVOT: right ventricular outflow tract; BPA: bilateral pulmonary arteries; PDA: patent ductus arteriosus; PKD: polycystic kidney disease; AKI: acute kidney injury; RIP: rest in peace; RA: right atrium; NSVT: non-sustained ventricular tachycardia.

disease (PKD) underwent PDA stenting with mTORi therapy. mTORi resulted in rapid tumour resolution in 3 patients, however 1 gene negative patient responded minimally. One patient with factor VII deficiency died postoperatively due to failure to achieve haemostasis. Of the remaining 4 live patients, 1 has epilepsy, 1 has polycystic kidney disease and hypertension, and 1 is asymptomatic with non-sustained ventricular tachycardia.

**Conclusions:** mTOR inhibition may prove effective in alleviating critical outflow or inflow tract obstruction. However, a bespoke approach may be required including mTOR inhibition, surgical debulking and/or a hybrid procedure to alleviate severe outflow tract obstruction and maintain systemic cardiac output.

**Keywords:** Obstructive Cardiac rhabdomyoma, Tuberous Sclerosis, mTOR inhibitor, surgical debulking of tumours, hybrid procedure

## O-84

### Trying to define ascending aorta dilatation in paediatric patients with bicuspid aortic valve. comparison of known nomograms and biometric parameters.

Andrea Freixa Benavente<sup>1</sup>, Diana Carolina Juzga Corrales<sup>1</sup>, Ferran Rosés Noguer<sup>1</sup>, Ariadna Ayerza Casas<sup>2</sup>, Marc Figueras Coll<sup>3</sup>, Sílvia Escribà Bon<sup>4</sup>, Beatriz Plata Izquierdo<sup>5</sup>, Rosa Collell<sup>6</sup>, María Arantzazu González Marín<sup>7</sup>, José Manuel Siu<sup>8</sup>, Moisés Sorí<sup>9</sup>, Leticia Albert<sup>10</sup>, Sílvia Teodoro<sup>11</sup>, Monica Rodríguez<sup>12</sup>, Olga Domínguez García<sup>13</sup>, Sara Rellán<sup>14</sup>, Begoña Manso<sup>15</sup>, Bernardo López Abel<sup>16</sup>, Rosa Álvarez Pérez<sup>17</sup>, Manuel Portillo Marquez<sup>18</sup>, Erika Rezola<sup>19</sup>, Fernando Centeno Malfaz<sup>20</sup>, Ruth Solana Gracia<sup>21</sup>, Henar Rojo Sombrero<sup>22</sup>, Anna Sabaté Rotés<sup>1</sup>

<sup>1</sup>Paediatric Cardiology, Vall Hebron University Hospital, Barcelona, Spain; <sup>2</sup>Paediatric Cardiology, Lozano Blesa Hospital, Zaragoza, Spain; <sup>3</sup>Paediatric Cardiology, Josep Trueta Hospital, Girona, Spain; <sup>4</sup>Paediatric Cardiology, Son Espases Hospital, Palma de Mallorca, Spain; <sup>5</sup>Paediatric Cardiology, Salamanca Hospital, Salamanca, Spain; <sup>6</sup>Paediatric Cardiology, St Joan de Reus, Reus, Spain; <sup>7</sup>Paediatric Cardiology, Ciudad Real General Hospital, Ciudad Real, Spain; <sup>8</sup>Paediatric Cardiology, Hospital de Nens, Barcelona, Spain; <sup>9</sup>Paediatric Cardiology, Cartagena Hospital, Cartagena, Spain; <sup>10</sup>Paediatric Cardiology, 12 de Octubre Hospital, Madrid, Spain; <sup>11</sup>Paediatric Cardiology, Parc Taulí Hospital, Sabadell, Spain; <sup>12</sup>Paediatric Cardiology, Montepíncipe Hospital, Madrid, Spain; <sup>13</sup>Paediatric Cardiology, Virgen de la Salud Hospital, Toledo, Spain; <sup>14</sup>Paediatric Cardiology, Hospital Clínico de Valladolid, Spain; <sup>15</sup>Paediatric Cardiology, Virgen del Rocío Hospital, Sevilla, Spain; <sup>16</sup>Paediatric Cardiology, Complejo Hospitalario Universitario Santiago de Compostela, Spain; <sup>17</sup>Paediatric Cardiology, Sant Pau Hospital, Barcelona, Spain; <sup>18</sup>Paediatric Cardiology, Hospital de Mérida, Spain; <sup>19</sup>Paediatric Cardiology, Donosti Hospital, Spain;

<sup>20</sup>Paediatric Cardiology, Río Ortega Hospital, Valladolid, Spain;

<sup>21</sup>Paediatric Cardiology, Intanta Leonor Hospital, Madrid, Spain;

<sup>22</sup>Paediatric Cardiology, Príncipe Asturias Hospital, Madrid, Spain

**Background and Aim:** Ascending aorta (AA) dilatation is a common finding in paediatric patients with bicuspid aortic valve (BAV). However, there are discrepancies on the degree of dilatation depending on the nomogram used. This study sought to determine differences amongst aorta dilatation definitions comparing the classical Halifax z-score (HZ), the newer z-score of Lopez (LZ) and biometric parameters indexing methods,

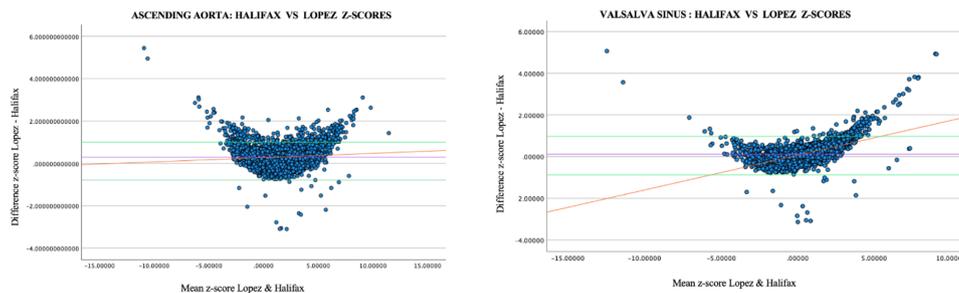
**Method:** It was a retrospective review of children affected by BAV included in the Spanish registry. Bland-Altman plots were used for agreement between both nomograms, and Pearson rank coefficient for correlation. Normality was checked by Kolmogorov-Smirnov test. A second analysis was performed correlating these nomograms against the definition of dilatation indexed by body surface area (BSA), considering dilated when >21 mm per square-meter. Threshold at z-scores +2 and +3 were set for comparison. The nomograms and indexed values were also submitted to concordance analysis by kappa index, considering >0, 4 a positive result.

**Results:** A total of 4013 examinations were included. Mean age at time of examination was 8, 02 years (standard deviation  $\pm$  5, 06). Correlation between HZ and LZ was  $\rho = 0, 98$ , both at AA and at Valsalva sinus (VS). HZ z-scores were significantly lower than LZ, but clinically non-relevant: mean z-score at VS was  $0, 26 \pm 1, 58$  for Halifax and  $0, 38 \pm 1, 86$  for López, and at AA was  $1, 4 \pm 2, 15$  by Halifax and  $1, 70 \pm 2, 19$  by López. Women and younger patients showed significantly higher differences between both nomograms at AA level. Bland Altman plots showed maximum agreement of both nomograms nearby z-score 0 but poorer at extreme values at VS level. For ascending aorta agreement was higher at z-score +2. Z-score threshold >2 in LZ nomogram correlated best with BSA-indexed values, both at AA ( $\rho = 0, 400$ ) and VS ( $\rho = 0, 328$ ). However, none of the threshold showed concordance by kappa index. For patients older  $\geq 10$  years or  $BSA \geq 1.5$  square-meters there was a higher number of patients classified as dilated when using LZ compared to BSA-indexing (41.05% versus 13.45% and 40, 10 versus 5, 95%).

**Conclusions:** Lopez nomogram correlates with the already established Halifax z-score despite resulting in non-clinically relevant higher z-scores. If  $\geq 10$  years or  $BSA \geq 1.5m^2$  Lopez z-score classified more patients as dilated compared to BSA-indexing.

**Keywords:** Bicuspid aortic valve, aorta dilatation, nomograms, z-score, body surface area.

BLAND ALTMAN PLOTS: COMPARISON OF BOTH Z-SCORES



The graphs show how both z-score correlate, It allows the reader to understand when maximum agreement is achieved and when the z-scores differ.

## O-85

**The cardiovascular phenotype of teenagers after preterm birth**

Thomas Salaets<sup>1</sup>, Art Schuermans<sup>2</sup>, Anke Raaijmakers<sup>3</sup>, Zenyu Zhang<sup>4</sup>, Jan Staessen<sup>4</sup>, Karel Allegaert<sup>5</sup>

<sup>1</sup>Division of Pediatric Cardiology, University Hospitals Leuven, Belgium & Echocardiography Lab, Hospital for Sick Children Toronto, Canada; <sup>2</sup>KULeuven, Belgium; <sup>3</sup>Department of Pediatrics, Hospitals ZNA Jan Palfijn, Antwerp, Belgium; <sup>4</sup>Department of Cardiovascular Sciences, KULeuven, Belgium; <sup>5</sup>Department of Development and Regeneration & Department of Pharmaceutical and Pharmacological Sciences, KULeuven, Belgium

**Background and Aim:** Survivors of preterm birth are at increased risk for cardiovascular disease. Several authors have also shown altered cardiac structure and function in former preterms but it remains unclear whether this is due to decreased cardiomyocyte endowment or due to remodeling in the context of vascular disease.

**Method:** The PREMATCH-study recruited 93 extremely low birth weight (ELBW) children (born <1000g) and 87 age-matched healthy controls for a prospective assessment of cardiovascular health at a median age of 11.0 years. Comprehensive echocardiograms were performed to evaluate standard cardiac structure and function parameters. Blood pressure was recorded and vascular stiffness assessed using applanation tonometry.

**Results:** The ELBW children were smaller in both length and weight than the controls. Markers of cardiac size indicated smaller hearts in ELBW-cases (mean LVEDD 4.06cm vs 4.22cm;  $p < 0.001$ ), but not if indexed for BSA (mean LVEDD z-score -0.51 vs -0.32;  $p = 0.119$ ). There was no difference in sphericity of the LV. LV mass was lower in ELBW than in controls (median LVM 75.3g vs 79.7g;  $p = 0.003$ ), however proportionally to LV size (mean mass to volume ratio 1.10g/ml vs 1.08g/ml;  $p = 0.436$ ).

Function-wise, there is no difference in LV ejection fraction but a decreased LV  $s'$  in ELBW (mean  $s'$  z-score -1.22 vs -0.92;  $p = 0.002$ ) might suggest a lower longitudinal function.

Blood pressure is higher in the ELBW-group (median SBPpercentile 84 vs 58;  $p < 0.001$ ; median DBPpercentile 69 vs 55;  $p < 0.001$ ). Carotid-to-femoral pulse wave velocity (PWV) was not significantly different between ELBW-cases and controls (mean PWV 4.13 vs 4.37;  $p = 0.20$ ).

**Conclusions:** In our study the cardiac phenotype of children born with an extremely low birth weight was relatively mild. Cardiac size is decreased, but in line with the decreased body size. There are no arguments for hypertrophy or systolic dysfunction. Additionally there are no arguments for increased vascular stiffness. These findings contrast several other studies. Nevertheless blood pressure is higher in ELBW-cases. Possibly, at this relatively young age, more sensitive methods are needed to assess cardiac remodelling after preterm birth. The possibility to evaluate the relationship between vascular and cardiac manifestations is a major strength of this study.

**Keywords:** preterm birth, cardiovascular health

## O-86

**Hypoplastic left heart syndrome: experience in our center after the implementation of a multidisciplinary care program**

Maria Lucia Rossi<sup>1</sup>, Emma Grau Sio<sup>1</sup>, Bosco Alejandro Moscoso Garrido<sup>2</sup>, Marta Campubí Campubí<sup>3</sup>, Fredy Prada<sup>1</sup>, Jose Maria Cafarena<sup>2</sup>, Joan Sanchez De Toledo<sup>4</sup>, Maria Clara Escobar Diaz<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain; Cardiovascular Research Group, Sant Joan de Déu Research Institute, Barcelona, Spain; <sup>2</sup>Department of Pediatric Cardiac

Surgery, Hospital Sant Joan de Déu, Barcelona, Spain; <sup>3</sup>Neonatal Intensive Care Unit, Hospital Sant Joan de Déu, Barcelona, Spain; <sup>4</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain; Department of Critical Care Medicine, University of Pittsburgh, Pittsburgh PA, US.

**Background and Aim:** The hypoplastic left heart syndrome (HLHS) is a complex heart disease that presents many challenges for post-natal management. Prenatal diagnosis and protocolized pre and post-surgical management influence the prognosis.

We aimed to compare our outcomes before and after the implementation of a protocolized multidisciplinary program.

**Method:** In 2017, we implemented a protocolized management that included training, and the creation of a multidisciplinary working group that included medical doctors, specialized nurse practitioners, psychologists, and social workers. There are also specific pre and post-surgical protocols and guides that ensure continuous intra-hospital monitoring and a rigorous outpatient follow-up.

This retrospective study included patients with HLHS born between 2010 and 2022, and results were analyzed dividing the cohort into two groups: from 2010 to 2016 (group one), and 2017 to 2022 (group two).

**Results:** Thirty-one patients were included, 14/31(45%) in group one and 17/31(55%) in group two. There were no differences between groups regarding prenatal diagnosis (85% vs. 88%,  $p = 0.6$ ), gestational age (38 [37-39] vs. 39 [38-39] weeks,  $p = 0.4$ ), and birth-weight (2870 [2810-2930] vs. 3215 [2695-3547] grams,  $p = 0.6$ ). Six patients (43%) in group one received comfort care vs. 2/17 (12%) in group two ( $p = 0.04$ ).

Of the patients with active management, 8/8(100%) vs. 3/17(17%) received mechanical ventilation and only those in the second group received non-invasive ventilation and a hypoxic mixture ( $p < 0.001$ ) before the first intervention.

Patients in both groups underwent surgery after 7 days [6-14]. In the second period, we performed more Norwood/Sano or Damus-Kaye-Stansel surgeries (86% vs. 62%) and fewer hybrids (13% vs. 37%). Postoperative ECMO was used mostly in the second period [1/8(12%) vs. 6/15(40%)]. Hospital stay was 55 [47-75] vs. 83 [52-129] days.

Hospital mortality in patients with active management [(3/8(37%) vs. 2/15(13%)] and overall mortality [10/14(71%) vs. 7/17(41%)] decreased after implementation of the protocolized program. Interstage mortality did not show significant differences [1/8(13%) vs. 3/15(20%)].

**Conclusions:** In our cohort, survival in patients with HLHS has improved thanks to the implementation of the protocolized management and the cumulated experience. The complexity of managing these patients makes it necessary for centers of excellence to achieve better survival and quality of life.

**Keywords:** Hypoplastic left heart syndrome, multidisciplinary program

**Sports Cardiology, Physical Activity and Prevention**

## O-87

**Exercise capacity remains stable in fontan patients entering adulthood**

Maren Emilie Hebeltoft Ravndal<sup>1</sup>, Lars Idorn<sup>2</sup>, Anne Kathrine Møller Nielsen<sup>1</sup>, Benjamin Kelly<sup>3</sup>, Kim Gjerum Nielsen<sup>2</sup>, Dorte Guldbrand Nielsen<sup>4</sup>, Vibeke Hjortdal<sup>1</sup>

<sup>1</sup>Department of Cardiothoracic Surgery, Copenhagen University Hospital, Copenhagen, Denmark; <sup>2</sup>Department of Pediatrics and Adolescent

Medicine, Copenhagen University Hospital, Copenhagen, Denmark; <sup>3</sup>Department of Cardiothoracic and Vascular Surgery, Aarhus University Hospital, Aarhus, Denmark; <sup>4</sup>Department of Cardiology, Aarhus University Hospital, Aarhus, Denmark

**Background and Aim:** Impaired exercise capacity is a frequent complication in Fontan patients and the functional capacity has been described to progressively decline with age. However, there are few longitudinal studies, and the pediatric age group dominates. We aimed to describe the natural course of exercise capacity in a population-based Fontan cohort, as these patients enter adulthood.

**Method:** Two serial cardiorespiratory exercise tests (CPX) were performed in Danish Fontan patients. Thirty patients (15 males) reached a respiratory exchange rate of minimum 1.0 in both CPX-tests and were included in the analyses. The time interval between the two tests was ten years, with a mean age of 17 years in study 1 and 27 years in study 2. Peak oxygen consumption (VO<sub>2</sub>) and oxygen pulse were measured. Results from the two assessments were compared for each patient using paired t-tests.

**Results:** 60% had a predominantly left ventricle and 40% a predominantly right ventricle. Types of total cavopulmonary connections included extracardiac conduit (47%), lateral tunnel (50%) and classical Fontan (3%). Mean age at Fontan completion was 5.8 years (range 1.5–25.7). There was no significant change in exercise capacity in these 30 patients during the study period. VO<sub>2</sub> remained stable with peak VO<sub>2</sub> 24.9 ml/min/kg (range 14.1–47.2) in study 1 and 23.9 ml/min/kg (range 11.4–41.1) in study 2 ( $p = 0.34$ ), and mean percent predicted peak VO<sub>2</sub> at 67.2% (range 30.0–108.0) in study 1 and 68.5% (range 40.0–97.0) in study 2 ( $p = 0.68$ ). Percent predicted O<sub>2</sub>-pulse was also stable with 88.9% (range 31.5–157.0) in study 1 and 87.8% (range 52.3–155.4) in study 2 ( $p = 0.81$ ).

**Conclusions:** Exercise capacity remained stable during transition from childhood to adulthood in this cohort of Danish Fontan patients.

**Keywords:** TCPC, Fontan, CPX

## O-88

### Leg-focused high-weight resistance training in children with a fontan circulation

Linda Elisabeth Scheffers<sup>1</sup>, Willem A Helbing<sup>2</sup>, Thomas Pereira<sup>3</sup>, Elisabeth Muji Utens<sup>4</sup>, Karolijn Dulfer<sup>5</sup>, Sylvia Walet<sup>6</sup>, Laurens Koopman<sup>2</sup>, Linda E Van Den Berg<sup>7</sup>

<sup>1</sup>Department of Pediatrics, division of Pediatric Cardiology, Erasmus MC - Sophia Children's Hospital, Rotterdam, Netherlands; Department of Pediatrics, Center for Lysosomal and Metabolic Diseases, Erasmus MC, Rotterdam, Netherlands; Department of Pediatric Gastroenterology, division of Pediatric Cardiology, Erasmus MC - Sophia Children's Hospital, Rotterdam, Netherlands; 3. Respiratory Medicine and Allergology, Erasmus MC - Sophia Children's Hospital, Rotterdam, Netherlands; <sup>2</sup>Department of Pediatrics, division of Pediatric Cardiology, Erasmus MC - Sophia Children's Hospital, Rotterdam, Netherlands; Department of Radiology, Erasmus MC - Sophia Children's Hospital, Rotterdam, Netherlands; <sup>3</sup>Department of Pediatrics, division of Pediatric Cardiology, Erasmus MC - Sophia Children's Hospital, Rotterdam, Netherlands; <sup>4</sup>Department of Child and Adolescent Psychiatry/Psychology, Erasmus MC - Sophia Children's Hospital, Rotterdam, Netherlands; Department of Child and Adolescent Psychiatry, Amsterdam University Medical Center/Levvel, Amsterdam, Netherlands Research Institute of Child Development and Education, University of Amsterdam, Amsterdam, Netherlands; <sup>5</sup>Intensive Care Unit, Department of Pediatrics and Pediatric Surgery, Erasmus MC - Sophia Children's Hospital, Rotterdam, Netherlands; <sup>6</sup>Division of Dietetics, Department of Internal

Medicine, Erasmus MC - Sophia Children's Hospital, Rotterdam, Netherlands; <sup>7</sup>Department of Orthopedics and Sportsmedicine, Erasmus MC - Sophia Children's Hospital, Rotterdam, Netherlands

**Background and Aim:** Sufficiently effective therapy to improve reduced exercise capacity in Fontan patients is currently lacking. Resistance training, focused on the lower leg muscles, has been suggested to augment the peripheral muscle pump and thereby enhance exercise capacity. Therefore, we aimed to augment the peripheral muscle pump with high-weight resistance training, to improve exercise capacity in Fontan patients.

**Method:** This study was a randomized semi-cross-over controlled trial. The effects the 12-week leg focused high-weight resistance training (3 supervised training sessions a week) supported by a high-protein diet were measured on (sub) maximal exercise capacity, cardiac function (measured using MR), muscle strength and quality of life in pediatric Fontan patients.

**Results:** Twenty-eight pediatric Fontan patients were included, 27 patients, (median age 12.9 years [IQR: 10.5–16.2]), successfully completed the program. At baseline patients had a reduced exercise capacity of 33.3 ml/kg/min [27.1–37.4] (73% [62–79] of predicted). Peak VO<sub>2</sub>/kg improved significantly with +6.2 ml/kg/min [95% CI: 3.4–9] (+18%)  $p < 0.001$ , compared to the control period. Peak load improved with 22 watts [95% CI: 12–32]  $p < 0.001$ , compared to the control period. Indexed single ventricle stroke volume increased significantly (43 ml/m<sup>2</sup> [40–49] vs 46 [41–53],  $p = 0.014$ ), as did inferior caval vein flow (28 ml/m<sup>2</sup> [20–33] vs 31 [27–39],  $p = 0.015$ ), while superior caval vein flow and EDV/Mass ratio remained unchanged. Maximum walked distance during the 6-minute walking test and average heart rate during submaximal exercise testing improved significantly after training (+28 meters,  $p = 0.021$  versus controls, average heartrate -8 beats  $p = 0.042$  versus controls). Seven out of nine measured muscle groups, including all leg muscles, increased significantly compared to the control period. Children improved significantly on the physical functioning and change in health domains of quality of life (child health questionnaire). Parents reported quality of life also improved significantly on the general health, change in health, bodily pain and general health perception domains compared to the control period.

**Conclusions:** In a relatively large group of 27 older Fontan children, 12-weeks of leg focused high-weight resistance training improved exercise capacity, cardiac output, muscle strength, and quality of life.

**Keywords:** Fontan procedure; Exercise capacity, cardiac function, cardiac rehabilitation; resistance training

## O-89

### Energy drinks: do they give the pediatric cardiovascular system wings? - results of the educate-study

Felix Sebastian Oberhoffer, Pengzhu Li, André Jakob, Robert Dalla Pozza, Nikolaus Alexander Haas, Guido Mandilaras  
Division of Pediatric Cardiology and Intensive Care, University Hospital, LMU Munich, Munich, Germany

**Background and Aim:** Energy Drinks (ED) are sweetened beverages that contain stimulants such as caffeine, guarana or taurine. EDs are popular among children and teenagers. Multiple adverse cardiovascular events associated with ED consumption were reported in minors. Despite EDs' increasing popularity, their acute effects on the pediatric cardiovascular system had not been investigated. Therefore, the EDUCATE-Study (Energy Drinks - Unexplored Cardiovascular Alterations in TEens and TwEens) was initiated by our institution. This abstract shortly summarizes published data

concerning the ED-induced effects on pediatric blood pressure, arterial stiffness, heart rhythm and electrocardiographic time intervals demonstrated in the EDUCATE-Study.

**Method:** The EDUCATE-study was a randomized, single-blind, placebo-controlled, crossover clinical trial. Study participants were asked to consume a weight-adjusted ED amount (3 mg caffeine per kg bodyweight) or a placebo beverage on two consecutive days. Systolic (SBP, mmHg) and diastolic blood pressure (DBP, mmHg) were measured by an oscillometric device. Peak circumferential strain of the common carotid arteries (CCA CS, %), a marker of arterial stiffness, was assessed sonographically by 2D speckle tracking. SBP, DBP and CCA CS were evaluated at the following time points: baseline as well as 30, 60, 120 and 240 minutes after beverage consumption. Heart rhythm and electrocardiographic time intervals were recorded via a 3-lead Holter monitor and analysed up to 240 minutes after beverage consumption.

**Results:** In total, 27 children and adolescents were included (14.53 ±2.40 years, 14 male). The ED consumption led to a significantly higher SBP (5.23 mmHg,  $p < 0.0001$ ) and DBP (3.29 mmHg,  $p < 0.001$ ). CCA CS was significantly lower ( $11.78 \pm 2.70$  % vs.  $12.29 \pm 2.68$  %,  $p = 0.043$ ) after the ED intake indicating an increased arterial stiffness. Moreover, the ED consumption resulted in a significantly higher number of supraventricular extrasystoles (incidence rate ratio: 1.700 (1.058, 2.732),  $p = 0.0276$ ) and a significantly lower heart rate ( $79.54 \pm 8.85$  bpm vs.  $82.65 \pm 8.81$  bpm,  $p = 0.012$ ).

**Conclusions:** The EDUCATE-Study suggests that acute and single dose ED intake is associated with negative impacts on the pediatric cardiovascular system. Minors, particularly those presenting with cardiovascular risk factors (e.g., arterial hypertension, arrhythmia), should be discouraged from consuming EDs. The chronic effects of ED consumption on the pediatric cardiovascular system need to be addressed in the future.

**Keywords:** Energy Drinks, Cardiovascular, Prevention, Pediatrics

## O-90

### Early life predictors for blood pressure in healthy adolescents are not associated with enhanced carotid intima-media thickness (cimt)

Isabelle Adrienne Van Der Linden<sup>1</sup>, Rozan Roodenburg<sup>1</sup>, Sanne Laura Nijhof<sup>2</sup>, Cornelis Korstiaan Van Der Ent<sup>3</sup>, Sabine Elena Ineke Van Der Laan<sup>3</sup>, Henk Simon Schipper<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology, Wilhelmina Children's Hospital, University Medical Center Utrecht, Utrecht, The Netherlands;

<sup>2</sup>Department of Pediatrics, Wilhelmina Children's Hospital, University Medical Center Utrecht, Utrecht University, Utrecht, The Netherlands;

<sup>3</sup>Department of Pediatric Pulmonology, Wilhelmina Children's Hospital, University Medical Center Utrecht, Utrecht University, Utrecht, The Netherlands

**Background and Aim:** Atherosclerosis development starts as early as childhood. Early life determinants including birth weight, body mass index (BMI) increase during childhood, physical activity, and fruit intake can modulate the development of hypertension and other risk factors for atherosclerosis. Birth cohort studies such as the Wheezing Illnesses Study Leidsche Rijn (WHISTLER) offer a unique opportunity to study early life determinants of blood pressure and preclinical atherosclerosis later in life. The aim of the current study is to investigate whether early life determinants predict blood pressure in adolescence, and preclinical atherosclerosis as assessed by carotid intima-media thickness (cIMT) measurements.

**Method:** The WHISTLER birth cohort study enrolled healthy children at birth, and followed them until 16 years of age ( $n = 232$  children). Physical measurements and questionnaires were performed at 5 years of age, and at 12-16 years. Data included birth weight, BMI, waist/hip ratio, physical activity, fruit intake, family history of atherosclerosis, blood pressure, and cIMT measurements using ultrasonography. We used multivariable linear regression analyses to evaluate cross-sectional determinants of blood pressure and cIMT at 12-16 years, and early life predictors for blood pressure and cIMT at 12-16 years.

**Results:** The cross-sectional analysis in healthy adolescents 12-16 years of age showed BMI as the main determinant of blood pressure. Early life predictors for blood pressure included the increase in BMI between 5 and 12-16 years of age, and blood pressure at 5 years. For cIMT measurements in healthy adolescents, we did not identify any cross-sectional determinants. Early life predictors for cIMT included only cIMT at 5 years.

**Conclusions:** Early life predictors for blood pressure in healthy adolescents include the increase in BMI between 5 and 12-16 years of age, and blood pressure at 5 years. These early life determinants were not associated with a higher cIMT in healthy adolescents. Only cIMT at 5 years was associated with cIMT in healthy adolescents. Our results show that blood pressure and its early life determinants are not associated with cIMT in healthy adolescents. This suggests that cIMT measurements in healthy adolescents may not fully capture their atherosclerotic risk, in line with other recent studies.

**Keywords:** adolescents, atherosclerosis, cIMT, children, cardiovascular risk factors

## O-91

### Cutaneous microcirculation in patients with congenital heart disease or marfan syndrome

Constance Gesina Weismann (1, 2)<sup>1</sup>, Michal Odarnarsky<sup>2</sup>, Petru Liuba<sup>2</sup>

<sup>1</sup>Department of Pediatric Cardiology, Skåne University Hospital, Department of Clinical Sciences, Lund University, Sweden; Department of Pediatric Cardiology and Pediatric Intensive Care, Ludwig-Maximilian University, Munich, Germany; <sup>2</sup>Department of Pediatric Cardiology, Skåne University Hospital, Department of Clinical Sciences, Lund University, Sweden

**Background and Aim:** Microcirculation plays an important role in the regulation of cardiovascular function and end-organ perfusion. Dysfunction of the microcirculation is an early marker of acquired cardiovascular diseases. However, data on patients with congenital heart disease is scarce. It was the aim of this study to evaluate endothelium-dependent and endothelium-independent microcirculatory function in patients with congenital cardiac disease.

**Method:** Prospective study of children and adults with history of repaired aortic coarctation (CoA), bicuspid aortic valve (BAV), Marfan syndrome (MFS) or single ventricle following Fontan palliation (SV). Laser Doppler (Periflux5000, Perimed AB, Sweden) was used to assess

cutaneous microcirculation non-invasively. The change of cutaneous microcirculatory flow in response to iontophoresis with acetylcholine (ACh) as an endothelium-dependent vasodilator and sodium nitroprusside (SNP) as endothelium-independent vasodilator was assessed quantitatively using a standardized protocol. The area under the curve over 2 minute cycles at baseline and maximal response were selected for analyses. Logarithmic transformation was performed. Multiple regression analyses correcting for age and sex as well as partial correlations were carried out.

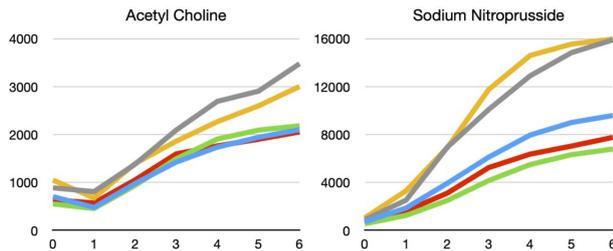
**Results:** 211 subjects were included (56 CoA, 46 BAV, 20 MFS, 30 SV, 59 controls). Mean age 29 (range 8-65) years. 13% of

participants were on vasodilatory medications, 12% were exposed to nicotine. At baseline, the SV group had a significantly lower microcirculatory perfusion than controls, while MFS patients had relatively increased perfusion. The response to transcutaneous iontophoresis of ACh was not significantly different between the groups, though there was a trend towards a diminished maximal response in the SV group. However, following iontophoresis of SNP, we saw an increased response in the BAV and MFS groups, while vasodilation was diminished in the SV group. The CoA group had a trend towards lower baseline perfusion and response to SNP. Laser Doppler outcome variables did not correlate with taking a vasodilatory medication or being exposed to nicotine.

**Conclusions:** While patients with SV physiology have impaired microcirculation at baseline and in response to iontophoresis of vasodilatory medications, the response to endothelium-independent vasodilatory stimuli appears increased in both MFS and BAV patients. The regulation of cutaneous microcirculation in SV patients will be the subject of future studies.

**Keywords:** microcirculation, endothelium, single ventricle, Marfan syndrome, bicuspid aortic valve, coarctation

Laser Doppler Figure and Table



**Figure:** Microcirculatory flow at baseline and in response to iontophoresis boluses of Acetyl Choline (left) or Sodium Nitroprusside (right) per group. Number of boluses is indicated on the timeline (x-axis). Flow is quantified as area under the curve over 2 minute cycles each. Groups: Control (blue), Coarctation (green), Bicuspid Aortic Valve (grey), Marfan syndrome (yellow), Single Ventricle with Fontan circulation (red).

**Table:** Cutaneous microcirculation at baseline and its maximal response to iontophoresis of Acetyl Choline (ACh) or Sodium Nitroprusside (SNP).

	CoA		BAV		MFS		SV	
	B	p	B	p	B	p	B	p
Base	-0.081	0.065	0.055	0.235	0.148	0.015*	-0.119	0.032*
ACh max	-0.026	0.73	0.101	0.213	0.079	0.467	-0.143	0.134
SNP max	-0.132	0.075	0.206	0.009*	0.207	0.042*	-0.192	0.041*

B-values (corrected for age and sex) and the respective p values are presented per patient group compared to healthy controls. P-values <0.05 were considered significant (\*). A trend was defined as a p<0.2 and marked in *italic*.

O-92

**Stairway to heaven - evaluation of cardiorespiratory fitness with a simple standardized stair climbing test in comparison to standard cardiopulmonary**

Simone Katrin Dold, Melina Kristina Uhl, Katharina Veronika Huber, Maurice Pablo Mall, Anne Lentz, Lara Emmrich, Johanna Wander, Nikolaus Alexander Haas

Department of Paediatric Cardiology and Paediatric Intensive Care, Ludwig-Maximilians-University, Munich, Germany

**Background and Aim:** According to the WHO, cardiovascular diseases are the leading cause of death worldwide. Intensive research

has shown that the cardiorespiratory exercise capacity of our organism is one of the decisive, if not the main factor in prevention of cardiovascular events or to curb existing risk factors and influence morbidity and mortality. Nevertheless, this parameter is only rarely recorded in routine clinical examinations. Currently, after individual indication, spiroergometry is considered the gold standard for assessing individual maximal exercise capacity. Alternatively, only the submaximal six-minute walking test (6MWT) is available so far. The aim of this study is to develop a standardized stair climbing test (SCT) with a reliable correlation to spiroergometry and 6MWT.

**Method:** In our pilot studies 25 healthy children (10-18 years), 52 healthy young adults (18-30 years), 27 Fontan patients (9-29 years) and 10 patients after heart transplantation (10-33 years) were included.

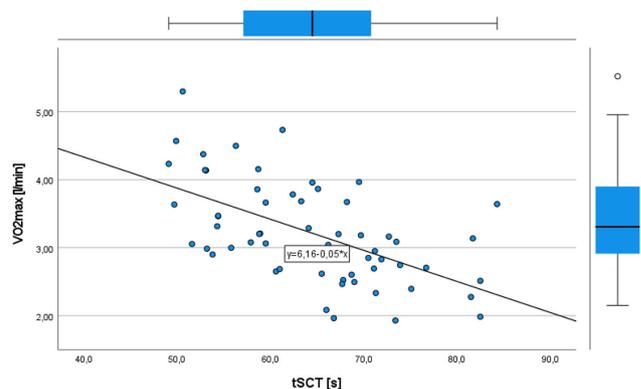
We tested the individuals' CRF by a treadmill exercise test and 6MWT (DGPK protocols) and a defined SCT according to our standardized protocol. In our SCT the individuals had to climb four floors (13, 14 m height) up and down as fast as possible; the time (tSCT) was stopped, and the vital signs were measured during this test. The correlation of the results from the simple exercise tests to CPET was evaluated.

**Results:** We could demonstrate a significant correlation between the measured time during the SCT and VO2max for instance in healthy adults:  $r = -0.783$ ;  $p = <0.001$ . There was also a good correlation between the distance achieved during the 6MWT and the passage time of our stair climbing test ( $r = -0.709$ ;  $p = <0.001$ ). Finally, the calculated oxygen pulse (VO2max/HFmax) showed a reliable correlation with the values of the stair climbing test ( $r = -0.779$ ;  $p = <0.001$ ).

**Conclusions:** We could demonstrate a significant correlation of the results of our SCT and the standard CPET VO2max and oxygen pulse in healthy subjects and various defined group of patients. Therefore, the SCT can be used as a simple and cheap standardized exercise test to assess CRF in healthy and chronically ill patients. In addition it seems useful to detect early cardiorespiratory changes as a screening test.

**Keywords:** cardiorespiratory fitness, exercise testing, stair climbing test

Correlation VO2max(l/min) to tSCT(s)



## O-93

**Aerobic fitness change with time in children with congenital heart disease: A controlled cohort study**

Arthur Gavotto<sup>1</sup>, Martina Avesani<sup>2</sup>, Sophie Guillaumont<sup>3</sup>, Helena Huguet<sup>4</sup>, Marie Christine Picot<sup>4</sup>, Stephan Matecki<sup>1</sup>, Pascal Amedro<sup>2</sup>

<sup>1</sup>Pediatric and Congenital Cardiology Department, M3C Regional Reference CHD Centre, Montpellier University Hospital, Montpellier, France; <sup>2</sup>Pediatric and Congenital Cardiology Department, M3C National Reference CHD Centre, Bordeaux University Hospital, IHU Liryc, INSERM 1045, University of Bordeaux, Bordeaux, France; <sup>3</sup>Paediatric Cardiology and Rehabilitation Unit, St-Pierre Institute, Palavas-Les-Flots, France; <sup>4</sup>Epidemiology and Clinical Research Department, University Hospital, Montpellier, France.

**Background and Aim:** To evaluate the change in aerobic fitness (VO<sub>2</sub>max), measured by cardio-pulmonary exercise test (CPET), in children with congenital heart disease (CHD), compared to matched healthy controls, and identify predictors of VO<sub>2</sub>max change with time in this specific population.

**Method:** This longitudinal multicentre cohort study was carried out from 2010 to 2020. We included CHD paediatric patients from the cohort of a previous cross-sectional study, who had a second CPET at least 1 year after the first one, during their follow-up. Trial registration: NCT04815577.

**Results:** We included 936 children, 296 in the CHD group and 640 controls. Mean time between baseline and final CPET was 4.4 ± 1.7 years. After matching on age and gender and adjustment for age and BMI, the mean VO<sub>2</sub>max group difference was 10.5% ± 1.0% of percent-predict VO<sub>2</sub>max at baseline and increased to 19.1% ±

1.3% at final assessment. In the CHD group, the proportion of children with impaired aerobic fitness was significantly higher at final than at baseline CPET assessment (51.4% vs 20.3%; P < 0.01). The mean annual VO<sub>2</sub>max decrease was significantly worse in the CHD group than in controls (-1.88% ± 0.19% of percent-predict VO<sub>2</sub>max/year vs. -0.44% ± 0.27% of percent-predict VO<sub>2</sub>max/year, P < 0.01, respectively). In multivariate analyse, male gender, a high initial VO<sub>2</sub>max, a high BMI, and the number of cardiac surgical procedures ≥2, were predictors of the VO<sub>2</sub>max decrease with time.

**Conclusions:** The VO<sub>2</sub>max decrease with time is more pronounced in children with CHD compared to healthy matched controls. This study highlighted the importance of serial CPET assessment in children with CHD.

**Keywords:** Congenital heart disease, VO<sub>2</sub>, Cardio-pulmonary exercise test, Paediatrics, Physical capacity

## O-94

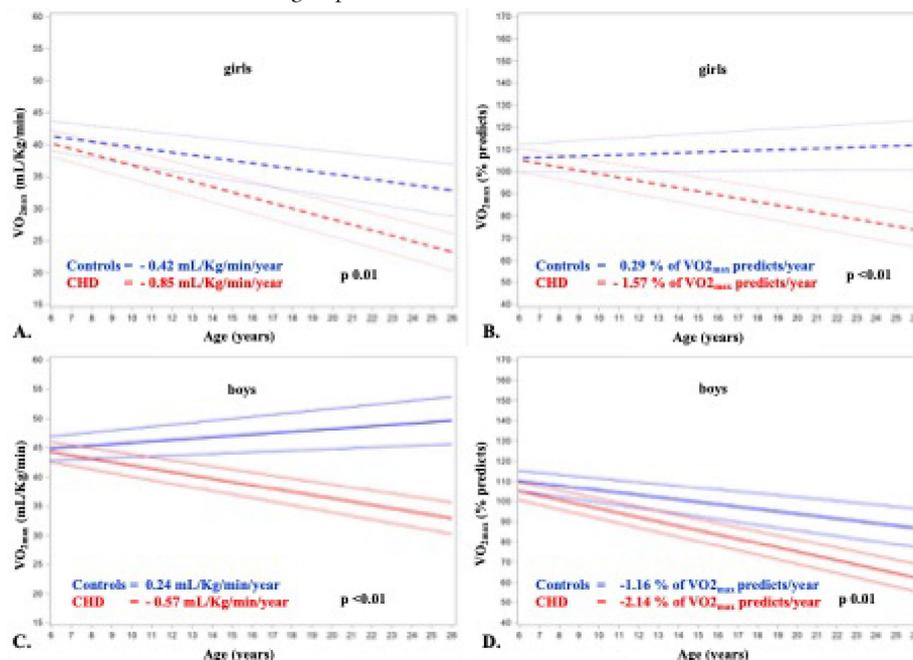
**Anti-coagulation with warfarin for paediatric cardiac indications: A national review**

Stephen T. O'Brien<sup>1</sup>, Sophie Duignan<sup>1</sup>, Kathleen Crumlish<sup>1</sup>, Jacqueline O'Donoghue<sup>1</sup>, Orla Franklin<sup>1</sup>, Phillip Roberts<sup>1</sup>, Colin J. Mahon<sup>2</sup>

<sup>1</sup>Department of Paediatric Cardiology, Children's Health Ireland at Crumlin, D12 N512 Dublin, Ireland; <sup>2</sup>School of Medicine, University College Dublin, Belfield, D04 V1W8 Dublin, Ireland

**Background and Aim:** Warfarin is used as anti-coagulation for paediatric patients for a wide range of cardiac indications but carries the disadvantage of requiring INR monitoring and dose adjustment.

VO<sub>2</sub>max change with time in CHD and control groups.



The blue lines represent the VO<sub>2</sub>max change with time in the control group, and the red lines in the CHD group. The density lines represent the median of VO<sub>2</sub>max change with time and the dashed lines represent the standard deviation of this value. VO<sub>2</sub>max change with time is expressed in mL/Kg/min (A, girls; C, boys) and percent-predicts (B, girls; D, boys).

Management of warfarin therapy is challenging due to its narrow therapeutic window and is further complicated in children by dietary changes, frequent illnesses and developing systems of metabolism and haemostasis. We aimed to retrospectively review the indications, INR levels, percentage time in range, and dose adjustments of the national cohort.

**Method:** The warfarin database was used to identify patients actively being prescribed warfarin by the cardiology team during a four-week study period, May 2022. A retrospective review was then performed of identified patients' medical records to assess the indication, time in INR target range and frequency of phlebotomy. We looked at all INR results and the percentage time in range (ITR%) and calculated the median ITR% by indication. The first 4 weeks of treatment were excluded to ensure patients were established on warfarin.

**Results:** Twenty-six patients were identified. The most common indication for warfarin use was in patients post-TCPC (total cavo-pulmonary connection) (n = 18, 69%). We demonstrated a variability in duration of warfarin therapy following TCPC, (median of 9 months, range 6-19 months). Nineteen (73%) of patients had used the coagucheck machine for home measurement of INR. The median frequency of phlebotomy for all indications was one test every 1.47 weeks and the median %ITR was 55% (29% – 86%) and this was also broken down by indication (see table). Of note, the percentage under target range in the patients with mechanical mitral (2) and aortic valves (1) was found to be 23% and 33% respectively.

**Conclusions:** Warfarin is commonly used for anticoagulation in paediatric patients with a wide range of cardiac conditions. These data demonstrate a high frequency of INR values outside of the target range. This necessitates frequent phlebotomy and dose changes, resulting in a significant effect on quality of life of these patients and their families. Emerging evidence for direct oral anticoagulants in paediatrics may represent an alternative to warfarin in the future although more research is required for their use in certain cardiac indications.

**Keywords:** Warfarin, Paediatric, Cardiology

Number of patients	Indication	Target INR	%ITR	Frequency of phlebotomy
26	All Indications	Variable (see below)	55.4% (29.7 – 86.4)	Every 1.47 weeks
18	Post-TCPC	1.8-2.5 (13) 2.0-3.0 (5)	58.6% (29.7 – 74.2)	Every 1.56 weeks
2	Mechanical Mitral Valve	3.0-4.0 (1) 3.5-4.5 (1)	45.6%	Every 1.06 weeks
1	Mechanical Aortic Valve	2.5-3.5	49.8%	Every 1.7 weeks
1	PIMS-TS with coronary aneurysm	2.0 – 3.0	37.5%	Every 1.4 weeks
1	Kawasaki Disease with large coronary aneurysms	1.8 – 2.2	46.9%	Every 1.3 weeks
1	Post trans-conduit puncture closure of mitral valve	1.8 – 2.5	86.4%	Every 2.2 weeks
1	CVA post BD Glenn	2.0 – 3.0	74.1%	Every 2 weeks
1	Early valve dysfunction post PPVR	2 – 2.5	45.2%	Every 1.1 weeks

Summary of Results

## Paediatric Cardiovascular Intensive Care

### O-95

#### Predicting cardiogenic shock in multisystem inflammatory syndrome in children

Said Bichali<sup>1</sup>, Mathilde Bonnet<sup>1</sup>, Marie Emilie Lampin<sup>2</sup>, Jean Benoit Baudelet<sup>1</sup>, Héloïse Reumaux<sup>3</sup>, Olivia Domanski<sup>1</sup>, Thameur Rakza<sup>1</sup>, Alexandre Delarue<sup>1</sup>, Morgan Recher<sup>2</sup>, Jérôme Soquet<sup>4</sup>, Francois Dubos<sup>5</sup>, Stéphane Leteurre<sup>2</sup>, Ali Houeijeh<sup>1</sup>, Francois Godart<sup>1</sup>

<sup>1</sup>Univ. Lille, CHU Lille, Pediatric Cardiology, F-59000 Lille, France;

<sup>2</sup>Univ. Lille, CHU Lille, Pediatric Intensive Care, F-59000 Lille,

France; Univ. Lille, Ulr 2694 - metrics: évaluation des technologies de santé et des pratiques médicales, F-59000 Lille, France;

<sup>3</sup>Univ. Lille, CHU Lille, Pediatric Rheumatology, F-59000 Lille, France;

<sup>4</sup>Univ. Lille, CHU Lille, Cardiac Surgery, F-59000 Lille, France;

<sup>5</sup>Univ. Lille, CHU Lille, Pediatric Emergency, F-59000 Lille, France.

**Background and Aim:** NT-proBNP has been shown to be the key cardiac marker to differentiate between serious and non-serious conditions related to SARS-CoV-2. This study aims to predict cardiogenic shock at diagnosis of multisystem inflammatory syndrome in children (MIS-C or PIMS-TS) using NT-proBNP.

**Method:** A single-center prospective cohort observational study was conducted between May 2020 and July 2022 at a tertiary care hospital. Children meeting the World Health Organization MIS-C criteria were included. The primary outcome was the occurrence of cardiogenic shock, assessed using logistic regression and receiver operating characteristic curve analysis.

**Results:** Sixty children were assessed for inclusion and 31 children were finally analysed (52% males, median age 8.8 (5.7-10.7) years). Nine children (29%) had cardiogenic shock. Median NT-proBNP level at diagnosis was 10870 (2328-16836) ng/L. At the time of diagnosis, NT-proBNP was associated with cardiogenic shock (OR 3.11 (1.23-7.91), p = 0.02). NT-proBNP at diagnosis  $\geq$  11254 ng/L had a sensitivity of 89%, and a specificity of 68% in predicting cardiogenic shock. The addition of age > 8 years and time to diagnosis  $\geq$  6 days had a sensitivity of 89%, a specificity of 91%, a positive predictive value of 80%, and a negative predictive value of 95% for cardiogenic shock.

**Conclusions:** NT-proBNP at diagnosis  $\geq$  11254 ng/L predicted cardiogenic shock in MIS-C. This biological marker, associated with an older age and a long time to diagnosis, could help to improve triage and intensify treatment.

**Keywords:** COVID 19, heart failure, biomarker, MIS-C, PIMS, diagnostic delay

### O-96

#### Prevalence and patterns of antibiotic treatment in neonates with critical congenital heart defects

Jessica A. Davis<sup>1</sup>, Tracy Baust<sup>2</sup>, Michael J. Morowitz<sup>3</sup>

<sup>1</sup>Department of Health Promotion and Development, University of Pittsburgh, Pittsburgh, Pennsylvania, United States of America;

<sup>2</sup>Department of Critical Care Medicine, UPMC Children's Hospital of Pittsburgh, Pittsburgh, Pennsylvania, United States of America;

<sup>3</sup>Department of Surgery, University of Pittsburgh, Pittsburgh, Pennsylvania, United States of America

**Background and Aim:** Antibiotic treatment in the setting of negative blood cultures has been associated with adverse outcomes in pre-term and full-term neonates without congenital heart defects.

Although data exists demonstrating widespread antibiotic treatment across other critically ill neonatal populations, there are no available data regarding the prevalence and potential consequences of antibiotic treatment among neonates with critical congenital heart defects (CCHD) beyond perioperative prophylaxis. The purpose of this study is to describe prevalence and patterns of antibiotic treatment in the first 28 days of life in a cohort of neonates with CCHD.

**Method:** A secondary analysis was performed using electronic health record data of full-term neonates with CCHD who required cardiac surgery in the first 28 days of life and were admitted to a children's hospital in the Northeastern United States of America between April 2016 and April 2020. Neonates in this cohort had no other defects or chromosomal abnormalities, and all received prostaglandins preoperatively. Descriptive statistics were generated to show cohort characteristics, antibiotic treatment days, and indications for use. Pearson Chi-Square, binary logistic regression, independent samples t-test were used to analyze potential predictors.

**Results:** A total of 74 neonates with CCHD were included, with the majority being prenatally diagnosed (57/74, 77%) and male (48/74, 64.9%). Thirty-eight (51%) received antibiotics in the first 48 hours after birth and 65 (88%) in the first seven days. Antibiotic treatment ranged from three to 27 days (mean 10 days, SD 6 days). Two neonates had positive blood cultures (2.7%). Prenatal diagnosis was not a significant predictor of antibiotic treatment in the first 48 hours after birth and there were no significant predictors for antibiotic treatment in the first seven days. Biventricular repair (41/74, 55.4%) was associated with fewer antibiotic treatment days ( $P = .016$ ) in the first 28 days of life.

**Conclusions:** These are the first available data on antibiotic treatment beyond perioperative prophylaxis in neonates with CCHD. Antibiotic treatment may be highly prevalent among CCHD and more common in those with univentricular defects. Larger, multi-center studies are needed to corroborate these findings and enhance understanding of potential consequences of antibiotic treatment in the setting of negative blood cultures.

**Keywords:** antibiotics, sepsis, neonate, critical congenital heart defects

#### O-97

##### **Clinical profile and pre-operative critical care needs of children with TAPVC: A retrospective study from a pediatric cardiac critical care unit**

Arun Kumar Baranwal, Priyanka Goyal, Rakesh Kumar Pilonia, Navpreet Kaur  
Postgraduate Institute of Medical Education and Research (PGIMER), Chandigarh, India

**Background and Aim:** Managing Total anomalous pulmonary venous connection (TAPVC) is highly challenging. There is scarcity of data on their pre-operative clinical profile and critical care needs from lower-middle income countries. Current study is conducted to identify clinical profile, pre-operative critical care needs and outcome among patients with TAPVC in a newly commissioned pediatric cardiac intensive care unit in a lower-middle income country.

**Method:** Medical records of all admitted children with diagnosis of TAPVC during its first 4 years of commissioning (Nov'18-Oct'22) were reviewed. Data regarding demographic and clinical details, their critical care needs and final outcome were collected and analyzed.

**Results:** Out of 920 patients, 43 had TAPVC (Male:Female; 5:1). They were diagnosed at postnatal age (IQR) of 67 (28-150) days, and presented to our hospital at 67 (32-180) days. Respiratory distress ( $n = 40$ , 81.39%) and cyanosis ( $n = 14$ , 32.5%) were common presenting symptoms. Supracardiac type was most common ( $n = 22$ , 51.16%). Unobstructed and obstructed TAPVC accounted for 35 (81.39%) and 8 (18.60%) patients respectively. Critical care requirements included mechanical ventilation [invasive;  $n = 16$ , 37.20%; Obstructed,  $n = 4$ , duration, 5.5 (1.25-14.25) days; Unobstructed,  $n = 12$ , 3.5 (3-6.5) days] and non-invasive;  $n = 12$ , 27.90%), inotropic support ( $n = 17$ , 39.53%), peritoneal dialysis ( $n = 2$ , 4.65%) and chest tube drainage for pneumothorax ( $n = 2$ , 4.65%). One patient with coarctation needed prostaglandin E1. Twenty-three patients (53.5%) had pulmonary hypertension. Common associated cardiac defects were ASD ( $n = 40$ , 93%) and TR ( $n = 18$ , 41.8%). Twenty one (48.8%) patients underwent surgery. Five (23.80%) of them died post-operatively; all were obstructive in type. Median admission-surgery interval was 10 (8-22) days. 10 (23.3%) patients were sick and died before surgery could be done. Rest were discharged, post-stabilization, for surgery on follow up.

**Conclusions:** Identification and thus presentation to tertiary care facility is delayed in TAPVC patients. A significant number of patients need mechanical ventilation and inotropes prior to surgery. Obstructed TAPVC is associated with poor outcome. Appropriate pre-operative critical care is a boon to children with late presentation.

**Keywords:** Total Anomalous Pulmonary Venous Connection, Clinical profile, Critical Care needs, Outcome, Lower-Middle Income country

#### O-98

##### **Left atrial strain and duration of intensive care in children with congenital diaphragmatic hernia**

Katarina Övermo Tydén, Balvín Jonsson, Carmen Mesas Burgos, Felicia Nordenstam  
Department of Women's and Children's Healths, Karolinska Institutet, Sweden

**Background and Aim:** Congenital diaphragmatic hernia (CDH) is a severe malformation syndrome that affects approximately 2, 5/10000 live births. The newborn child presents with different degrees of pulmonary hypoplasia and abnormal pulmonary vascular development. Children born with CDH present with various degrees of pulmonary hypertension, left heart hypoplasia and left and right heart dysfunction. Today, there is increased concern about the important role of left ventricle (LV) dysfunction in children with CDH. If there is LV dysfunction, traditional treatment can worsen the situation and drive the child into cardiorespiratory failure. Hence it is of importance to evaluate left ventricle function, both systolic and diastolic. Left atrial strain is an established method

to evaluate diastolic dysfunction in adults. Today, only a few studies have been published on LA strain in neonates and there are only a handful publications on reference values.

**Method:** This was a retrospective observational cohort study, including children born between 2018–2020 with CDH and treated at Karolinska University Hospital. A retrospective examination of echocardiograms was performed.

LA strain was correlated with days spent at intensive care unit (ICU) and need for extra corporal membrane oxygenation (ECMO). LA strain analyzes were performed in TomTec version TTA2 (TOMTEC imaging Systems GmbH, Unterschleissheim, Germany).

**Results:** A total of 41 children born with CDH were included. 58 % (n = 27) were male. Gestational age was between 32+0 – 42+1 and average birthweight was 2.9 kg (range 1.5–4.3 kg)

Children with LA strain less than 35 (n = 27) had a significantly longer stay in ICU (mean 18.7 days, 95% CI 13.9–23.5 days) compared to children with LA strain more than 35 (n = 14) (mean 7, 79 days, 95% CI 2.2–13.4 days) (p = 0.001).

**Conclusions:** LA strain is a feasible ECHO measurement to assess diastolic ventricular function in children born with CDH. Ventricular diastolic dysfunction assessed with LA strain is related to longer stay in the intensive care unit.

**Keywords:** Left atrial strain, cardiac dysfunction, Congenital diaphragmatic hernia, intensive care

## O-99

### **Cardiac dysfunction requiring intensive care management in pims-ts and covid 19 infection; ireland's experience through the pandemic**

*Nicola Mccay, Niamh Beirne, Erike Brereton, Martina Healy, Orla Franklin*

*Children's Health Ireland, Crumlin, Dublin, Ireland*

**Background and Aim:** Coronavirus disease 2019 (Covid-19) is an infectious disease caused by the severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) Although primary infection is usually mild in children, paediatric Inflammatory Multi-system syndrome; temporally associated with SARS-CoV-2 (PIMS-TS), can result in significant cardiac manifestations.

**Method:** This is a retrospective multi-centre observational study investigating cardiac disease and short-term outcomes in paediatric

patients with Covid-19 infection, PIMS-TS and covid vaccine-related myocarditis requiring intensive care (PICU) management in Ireland between March 2020 and August 2022.

Data collection was coordinated through retrospective chart, echocardiography database and electronic record review.

**Results:** • 126 children were admitted to PICU in Ireland with a Covid-19 related illness between March 2020 and August 2022.

- 68.2 % of patients had acute Covid-19 infection, 30.9% PIMS-TS and 0.7% post-covid vaccine-related myocarditis.

- 60.5% of patients in ICU with PIMS-TS had evidence of cardiac involvement:

- o 36.8% had reduced left ventricular function; 21.1% mild and 15.8% moderate.

- o 31.6% of PIMS-TS patients had significant atrioventricular valve regurgitation, 7.9% had coronary artery changes, 10.5% had pericardial effusion.

- 13.9% of patients in PICU with primary covid-19 infection had cardiac dysfunction, 80% mild. There were no cases with coronary artery involvement or AV valve regurgitation.

- In PIMS-TS patients 53.8% required inotropic support compared to 11.5% of Covid 19 infection

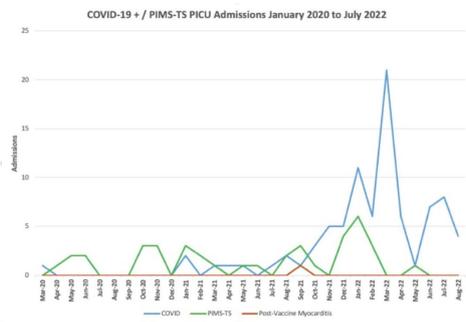
- 53.9% of patients with acute Covid19 infection required invasive or non-invasive ventilation compared to 23.2% with PIMS-TS.

- 93.7% of patients with PIMS-TS related cardiac disease and 100% with Covid-19 had a normal echo at the time of discharge from hospital.

**Conclusions:** The burden of cardiac disease in children requiring PICU care for Covid-19 was high in the acute phase of the illness, reassuringly all cardiac investigations had normalised at the time of short-term follow-up.

**Keywords:** Paediatric intensive care, cardiac dysfunction, Covid-19, PIMS-TS

Data; patients in PICU in Ireland with Covid-19 related illness 2020–2022



Ventilation Status	COVID-19	PIMS-TS	Post-Vaccine Myocarditis	Grand Total (n)	%
Invasively Ventilated	36	8		44	35%
NIV or No Ventilation Required	51	31	1	83	65%
<b>Total</b>	<b>87</b>	<b>39</b>	<b>1</b>	<b>127</b>	

PIMS-TS LV systolic dysfunction on inital ECHO		Patients
None		24
Mild		8
Moderate		6
Severe		
<b>Grand Total</b>		<b>38</b>

Covid-19 LV systolic dysfunction initial ECHO		Patients
None		31
Mild		5
Severe		1
Moderate		
<b>Grand Total</b>		<b>37</b>

Medications	COVID-19	PIMS-TS	Post-Vaccine Myocarditis	Grand Total
Vaccinated	2 *(18 Unknown)	2	1	5
Inotropes	10	21		31
Milrinone	8	22	1	31
Steroids	37	37		74
IVIG	11	31	1	43

Key data /results collated following our multi-centre observational study

**O-100**

**Thrombogenic myocardial infarction in newborn**

Franziska Markel<sup>1</sup>, Roman Antonin Gebauer<sup>1</sup>, Marcel Vollroth<sup>2</sup>, Frank Thomas Riede<sup>1</sup>

<sup>1</sup>Heart Center Leipzig, Department for Pediatric Cardiology, Struempellstrasse 39, 04289 Leipzig, Germany; <sup>2</sup>Heart Center Leipzig, Department for Pediatric Cardiosurgery, Struempellstrasse 39, 04289 Leipzig, Germany

**Background and Aim:** Myocardial infarctions in newborns are rare and have a high mortality rate. 1, 2 We present a seldom case of a newborn with a left coronary artery thrombus, who presented in cardiogenic shock.

**Method:** Our patient was a term-hypertrophic newborn that showed relevant hypoglycemia in the regular controls of the birth clinic. 24 hours after birth, she was transmitted to the NICU due to cyanosis, dyspnoea, and hypoxemia, with a maximum lactate of 20, and pH of 6, 9. Sonographically a left ventricular obstruction was suspected; an infusion with prostaglandin was initiated and the patient was transmitted to our heart center. She presented in cardiogenic shock with ventricular tachycardia and minimal output. A first defibrillation attempt of the VT was unsuccessful, finally, the rhythm converted into sinus rhythm after treating the hyperkalemia and the lactate acidosis. The ECG in SR revealed a left anterior hemiblock and relevant ischemia of the left heart. Echocardiographically we diagnosed a thrombus in the left

ventricle and above the aortic valve, impeding blood flow to the left coronary artery. As the catheter lab showed a total occlusion of the LCA, an emergency surgical thrombectomy was initiated and extracorporeal membrane oxygenation was started.

**Results:** As hyperkalemia and acidosis can aggravate ventricular tachycardia these causes have to be treated beforehand to sufficiently be able to convert the patient into sinus rhythm. In our case, the ventricular rhythm disorder was suspicious with an unusual QRS morphology, but it masked the relevant ischemia in the ECG and made the finding of the diagnosis even harder. The etiology of the thrombus can be varying from infection, autoimmune disorders, oncologic diseases or inherited thrombophilias such as Faktor-V-Leiden mutation, or Protein C- or S-deficiency.

**Conclusions:** Although a myocardial infarction due to thrombosis on the second day of life is a very rare diagnosis, it has to be kept in mind as one possible factor for a cardiogenic shock.

**Keywords:** Newborn, cardiogenic shock, thrombus, myocardial ischemia

## General Cardiology

### O-101

#### Clinical features and echocardiography of multisystem inflammatory syndrome in children in the republic of Ireland, April 2020 to october 2022.

Bryony Treston<sup>1</sup>, Bazlin Ramly<sup>1</sup>, Eoin Fitzgerald<sup>1</sup>, Karina Forde<sup>1</sup>, Niall Linnane<sup>2</sup>, Orla Franklin<sup>2</sup>, Colin McMahon<sup>2</sup>, Timothy R Leahy<sup>3</sup>, Patrick Gavin<sup>1</sup>

<sup>1</sup>Department of Infectious Diseases and Immunology, CHI Crumlin and Temple Street, Dublin, Ireland; <sup>2</sup>Department of Paediatric Cardiology, CHI Crumlin and Temple Street, Dublin, Ireland; <sup>3</sup>Department of Infectious Diseases and Immunology, CHI Crumlin and Temple Street, Department of Paediatrics, University of Dublin, Trinity College, Dublin, Ireland

**Background and Aim:** Our aim was to describe the clinical presentation of multisystem inflammatory syndrome – in children (MIS-C) in the Republic of Ireland, since the onset of the SARS-CoV-2 pandemic.

**Method:** Through prospective surveillance all paediatric patients treated for MIS-C in the Republic of Ireland from April 2020 to October 2022 were included.

**Results:** 118 children were diagnosed with MIS-C during the study period, (median age 6.6 years; males 57%). Of these, 78 were deemed inflammatory type MIS-C and 40 Kawasaki disease type MIS-C. Almost half had a bodyweight >91st centile for age. Gastrointestinal and mucocutaneous symptoms were the most common presenting complaint (both 67%), followed by respiratory symptoms (61%) and lymphadenopathy (36%). ICU admission was required for 27% of patients. Almost half of patients had abnormal echocardiography at presentation. Impaired myocardial contractility and/or myocarditis were more common among inflammatory-type MIS-C cases, whereas coronary artery abnormalities were more common among KD-type. In analysis of MIS-C cases over the first year of the pandemic (n = 54), all but three cases had normal echocardiograms at follow-up (median 2 months) with the majority returning to normal within a few days of presentation. Lymphocyte and platelet counts were significantly lower among inflammatory-type MIS-C cases compared to KD-type cases on admission (0.8 vs 2.8, p = 0.003 and 187 vs 437, p = 0.0004). Twenty-nine (54%) MIS-C patients had evidence of previous SARS-CoV-2 infection, with a higher rate in the inflammatory type (72%) compared to the

KD type (8%). There were no fatalities among our cohort and all patients were discharged home.

**Conclusions:** MIS-C patients in Ireland had similar clinical features, echocardiography, and laboratory results to international case series. However, disease severity was less in Ireland with lower ICU admission rates, minimal morbidity, and no fatalities. While half of children had abnormal echocardiograms at presentation, resolution of these abnormalities mirrored the clinical picture of a prompt full recovery in the majority of cases.

**Keywords:** MIS-C, PIMS-TS, Kawasaki Disease, COVID-19, Echocardiography

### O-102

#### Aspirin resistance in a cohort of paediatric patients with in-situ bio-prosthetic heart valves

Sean T Kelleher<sup>1</sup>, Irene E Regan<sup>2</sup>, Damien Kenny<sup>1</sup>, Kevin P Walsh<sup>1</sup>, Jonathon G McGuinness<sup>3</sup>, Philip A Roberts<sup>1</sup>, Dermot Cox<sup>4</sup>, Colin J McMahon<sup>5</sup>

<sup>1</sup>Department of Paediatric Cardiology, CHI at Crumlin, Dublin, Ireland; <sup>2</sup>Department of Haematology and Coagulation, CHI at Crumlin, Dublin, Ireland; <sup>3</sup>Department of Cardiothoracic Surgery, CHI at Crumlin, Dublin, Ireland; <sup>4</sup>School of Pharmacology, Royal College of Surgeons in Ireland, Dublin, Ireland; <sup>5</sup>School of Medicine, University College Dublin, Ireland & Department of Paediatric Cardiology, CHI at Crumlin, Dublin, Ireland

**Background and Aim:** Aspirin is widely prescribed to reduce the risk of thromboembolic events in at-risk paediatric patients with congenital heart disease (CHD). Patients with bio-prosthetic heart valves are at an additional risk of sub-clinical thrombus formation with postulated impact on valve-longevity and infective endocarditis risk, in whom aspirin may attenuate this risk. The incidence of aspirin resistance amongst patient with CHD has a wide range of reported incidence. The aim of this prospective observational study was to determine the rate of aspirin resistance in a cohort of paediatric patients with in-situ bio-prosthetic heart valves.

**Method:** Patients were recruited prospectively from ward and out-patient settings. Inclusion criteria; in-situ bio-prosthetic bovine or porcine heart valves, and current aspirin therapy. Exclusion criteria; age >18 years, known coagulation disorder or decreased platelet function, thrombocytopenia <150 x10<sup>9</sup>/L, haemoglobin <100g/L, medications interfering with platelet function, active endocarditis. All patients underwent recent echocardiography. Aspirin resistance was defined as platelet inhibition below 50% by thrombo-elastography (TEGPM) and light transmission platelet aggregation (LTA) below 20% using arachidonic acid agonist.

**Results:** There were 21 patients with an average age of 8.9 years (range: 0.3–18.5). Eight were female (38%) and 13 were male (62%). Most patients (90%, n = 19/21) had complex abnormalities of the right ventricular outflow tract with pulmonary atresia (n = 8) being the most common single diagnosis. In 9 patients (43%), the valve was situated within a conduit, in whom 44% (n = 4/9) had a valve sited percutaneously into an existing conduit. Patients received standard therapeutic doses of aspirin of 3–5mg/kg/day up to a maximum of 75mg/day. One patient received a lower dose (0.5mg/kg) due to bleeding. Five patients (24%) were aspirin non-responsive (TEGPM 0–26.5%, LTA 24.1–35%).

**Conclusions:** Twenty-four percent of patients with in-situ bio-prosthetic heart valves in our cohort demonstrated aspirin resistance. Given this relatively high rate in this at-risk group, we advocate testing for aspirin resistance be performed in all such patients. Further studies are needed to demonstrate whether aspirin resistance is associated with adverse outcomes in this group.

**Keywords:** Aspirin, Resistance, Bio-prosthetic valve

## Adult Congenital Heart Disease

### P-001

#### Outcome of down patients with repaired versus unrepaired atrioventricular septal defect

Mathies Daene<sup>1</sup>, Lore De Pauw<sup>1</sup>, Pieter De Meester<sup>2</sup>, Els Troost<sup>2</sup>, Philip Moons<sup>3</sup>, Marc Gewillig<sup>4</sup>, Filip Rega<sup>5</sup>, Alexander Van De Bruaene<sup>2</sup>, Werner Budts<sup>2</sup>

<sup>1</sup>Faculty of Medicine, Catholic University Leuven, Belgium; <sup>2</sup>Department of Cardiovascular Sciences, Catholic University of Leuven, Belgium; Congenital and Structural Cardiology, University Hospitals Leuven, Belgium; <sup>3</sup>Department of Public Health and Primary Care, Catholic University of Leuven, Belgium; <sup>4</sup>Institute of Health and Care Sciences, University of Gothenburg, Sweden; <sup>5</sup>Department of Paediatrics and Child Health, University of Cape Town, Cape Town, South Africa; <sup>6</sup>Pediatric Cardiology, University Hospitals Leuven, Belgium; <sup>7</sup>Department of Cardiovascular Sciences, Catholic University of Leuven, Belgium; <sup>8</sup>Cardiac Surgery, University Hospitals Leuven, Belgium; <sup>9</sup>Department of Cardiovascular Sciences, Catholic University of Leuven, Belgium

**Background and Aim:** Patients with Down Syndrome (DS) are frequently born with an atrioventricular septal defect (AVSD). Surgical repair of the defect aims to minimize mortality and morbidity. However, a surgical intervention, specifically in DS patients, is not without risk and a subgroup of patients underwent only conservative non-surgical treatment. Outcome data of these different approaches are scarce. The aim of this retrospective study was to compare the long-term outcome of DS patients with and without surgery for AVSD.

**Method:** DS patients registered with AVSD in the hospital's database from January 1980 till December 2020 were selected. Patient characteristics, peri-operative if appropriate, and follow-up data were obtained from the medical files.

**Results:** In total, 72 unrepaired (36 male, 50%) and 134 repaired patients (61 male, 46%) were included. After a maximum of 60 years of follow-up, the all-cause mortality was 45.8% and 17.1%, respectively. Thirty-six percent and 13%, respectively, were labeled as non-cardiovascular death. Mean survival time for unrepaired AVSD was 40.7 years (95% CI 36.1 – 45.2) and for repaired AVSD 38.5 years (95% CI 35.3 – 41.6) (Log rank  $p = 0.465$ ). However, the survival rate 35 years after birth was 62.1% for unrepaired patients versus 81.7% for repaired patients. Mortality rates were the highest the first months after surgical repair.

**Conclusions:** The mean survival rate of Down patients, born with an AVSD, did not differ between repair or not. However, long-term survival rate was higher in patients who underwent surgical repair. Mortality was highest the first months after surgery.

**Keywords:** Down syndrome, atrioventricular septal defect, atrioventricular canal, surgery, Eisenmenger syndrome, outcome

### P-002

#### Decreased cardiac reserve in asymptomatic patients after ASO for transposition of the great arteries

Beatrice Santens<sup>1</sup>, Alexander Van De Bruaene<sup>1</sup>, Pieter De Meester<sup>1</sup>, Guido Claessen<sup>2</sup>, Philip Moons<sup>3</sup>, Piet Claus<sup>2</sup>, Kaatje Goetschalckx<sup>2</sup>, Jan Bogaert<sup>4</sup>, Werner Budts<sup>1</sup>

<sup>1</sup>Congenital and Structural Cardiology, University Hospitals Leuven, Belgium; <sup>2</sup>Department of Cardiovascular Sciences, Catholic University Leuven, Belgium; <sup>3</sup>Cardiology, University Hospitals Leuven, Belgium; <sup>4</sup>Department of Cardiovascular Sciences, Catholic University, Belgium; <sup>5</sup>Department of Public Health and Primary Care, Catholic University Leuven, Leuven, Belgium; <sup>6</sup>Institute of Health and Care Sciences, University of Gothenburg, Gothenburg, Sweden; <sup>7</sup>Department of

Paediatrics and Child Health, University of Cape Town, Cape Town, South Africa; <sup>8</sup>Department of Radiology, University Hospitals Leuven, Leuven, Belgium; <sup>9</sup>Department of Imaging and Pathology, Catholic University Leuven, Leuven, Belgium

**Background and Aim:** Exercise capacity is impaired in patients after arterial switch operation (ASO) for complete transposition of the great arteries. Maximal oxygen consumption is related with outcome. This study assessed ventricular function by advanced echocardiography and cardiac magnetic resonance (CMR) imaging at rest and during exercise, to determine exercise capacity in ASO patients, and to correlate ventricular function with exercise capacity to identify early markers of deterioration.

**Method:** Forty-four patients (71% male, mean age 25 years - range 18-40 years) were included during routine clinical follow-up. Assessment involved physical examination, 12-lead ECG, echocardiography, and cardiopulmonary exercise test (CPET) (day 1). On day 2 CMR imaging at rest and during exercise was performed. Blood was sampled for biomarkers.

**Results:** All patients reported New York Heart Association class I, the overall cohort had an impaired exercise capacity. Fragmented QRS was present in 27%. Exercise CMR showed that 20% of patients had abnormal cardiac reserve (CR) of the left ventricle (LV) and 25% had reduced CR of the right ventricle (RV). CR LV and CR RV were significantly associated with impaired exercise capacity. Pathological patterns on myocardial delayed enhancement and hinge point fibrosis were detected. Biomarkers were normal.

**Conclusions:** This study found that in some asymptomatic ASO patients electrical, LV and RV changes at rest, and signs of fibrosis are present. Maximal exercise capacity is impaired and linearly related to the CR of the LV and the RV. Therefore, exercise CMR might play a role in detecting subclinical deterioration of ASO patients.

**Keywords:** transposition of the great arteries, arterial switch operation, impaired exercise capacity, contractile reserve

### P-003

#### Factors that may influence symptoms during pregnancy in women with congenital heart disease - data from the swedish birth registry

Annika Bay<sup>1</sup>, Åsa Burström<sup>2</sup>, Malin Berghammer<sup>3</sup>

<sup>1</sup>Department of Nursing, Umea University, Umea, Sweden;

<sup>2</sup>Department of Neurobiology, Care Sciences and Society, Karolinska Institutet, Stockholm, Sweden; <sup>3</sup>Institute of Health Sciences, University West, Trollhättan, Sweden; <sup>4</sup>Queen Silvia Childrens Hospital, Sahlgrenska University Hospital, Gothenburg, Sweden

**Background and Aim:** Women with a congenital heart defect (CHD) contains the largest group of women with heart disease during pregnancy. Women with complex heart defects count as a high-risk pregnancy, which is associated with an increased risk of miscarriage, premature labor as well as other complications during pregnancy. For women with complications, the risk for complications after pregnancy and labor is also higher. Since pregnancy-related symptoms easily could be mixed up with signs of heart failure, there is a need to further investigate the prevalence of symptoms and compare whether certain factors are more common in women who experience symptoms during pregnancy. **AIM:** To investigate what factors influence symptoms during pregnancy in women with congenital heart disease.

**Method:** Women in the national register of CHD, with a previous pregnancy beyond 22 weeks of gestation, were identified in the national birth register. Descriptive statistical analysis was performed

in order to compare women with symptoms and those without symptoms.

**Results:** Twelve hundred and seventy-seven women (mean age  $29.7 \pm 5.0$ ) with CHD were identified in the birth registry and 307 of them experienced symptoms. The most common symptom was fatigue, followed by dyspnea, palpitations, edema, syncope and chest pain. Symptoms were more common among women who lived alone (44% vs. 24%,  $p = 0.02$ ), had a lower education level  $\leq 12$  years (27% vs. 18%,  $p \leq 0.001$ ), had more than 3 children (51% vs. 23%,  $p \leq 0.001$ ), were not so physically active (28% vs. 21%,  $p = 0.01$ ), had an impaired left ventricular function (36% vs. 23%,  $p = 0.01$ ) and had an ongoing cardiovascular medication (33% vs. 23%,  $p = 0.004$ ).

**Conclusions:** The vast majority of women with CHD undergo pregnancy without symptoms from their heart defect. However, it appears that factors other than those related to the heart defect also affect the burden of symptoms and may be noted early in pregnancy.

**Keywords:** ACHD, symptoms, reproduction, pregnancy

**Table 1.** Overview of variables.

	Total n=1277	Symptoms n=307	No symptoms n=970	p-value
Age, years, mean $\pm$ SD	29.7 (5.0)	29.9 (5.1)	29.7 (5.0)	0.40
Family situation, n(%)				0.02
Living alone	27	12 (44%)	15 (56%)	
Living together	1193	269 (24%)	864 (76.3%)	
Education level n(%)				0.001
$\leq 12$ years	557	149 (27%)	408 (73%)	
$> 12$ years	471	86 (18%)	385 (82%)	
Complexity of the heart disease n(%)				0.15
Simple lesions	492	129 (42%)	363 (37%)	
Moderate/severe lesions	785	178 (58%)	607 (62%)	
No. of children, mean $\pm$ SD				0.001
0-3	1227	283 (23%)	944 (77%)	
$> 3$	47	24 (51%)	23 (49%)	
Physical active (n(%)				0.01
$\leq 3$ hours/week	709	201 (28%)	508 (72%)	
$> 3$ hours/week	379	81 (21%)	298 (79%)	
Left ventricle function, n(%)				0.01
Impaired	75	27 (36%)	48 (64%)	
Not impaired	1141	258 (23%)	883 (77%)	
Cardiovascular medications, n(%)				0.004
Yes	162	54 (33%)	108 (67%)	
No	1095	249 (23%)	846 (77%)	

n, number; SD, Standard deviations; Bold figures denotes  $p < 0.05$

**P-005**

**Assessment of microvascular dysfunction in adults with congenital heart disease**

Inne Vanreusel<sup>1</sup>, Wendy Hens<sup>2</sup>, Ervin De Bliëk<sup>3</sup>, Hilde Heuten<sup>1</sup>, Bernard Paelinck<sup>1</sup>, Emeline Van Craenenbroeck<sup>1</sup>, Hein Heidebuchel<sup>1</sup>, Vincent Segers<sup>1</sup>, An Van Berendonck<sup>1</sup>

<sup>1</sup>Department of Cardiology, Antwerp University Hospital, Edegem, Belgium; Research Group Cardiovascular Diseases, GENCOR, University of Antwerp, Antwerp, Belgium; <sup>2</sup>Department of Cardiology,

Antwerp University Hospital, Edegem, Belgium; Cardiac Rehabilitation Centre, Antwerp University Hospital, Edegem, Belgium; Department of Rehabilitation Sciences and Physiotherapy, Faculty of Medicine and Health Sciences, MOVANT Research Group, University of Antwerp, Antwerp, Belgium; <sup>3</sup>Department of Cardiology, Antwerp University Hospital, Edegem, Belgium; Research Group Cardiovascular Diseases, GENCOR, University of Antwerp, Antwerp, Belgium; Cardiac Rehabilitation Centre, Antwerp University Hospital, Edegem, Belgium

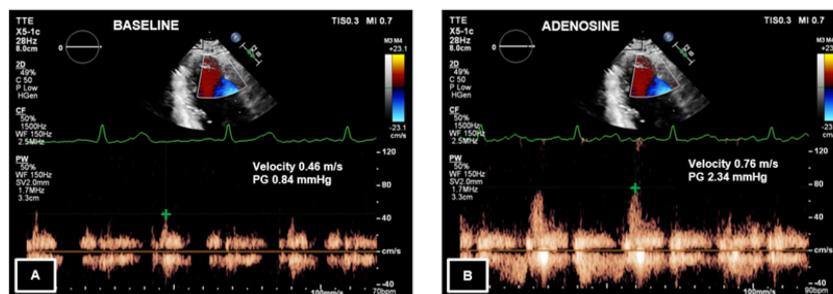
**Background and Aim:** The role of coronary (CMD) and peripheral microvascular dysfunction (PMD) in adults with congenital heart disease (ACHD) is incompletely understood. Adenosine stress transthoracic echocardiography (TTE) is a validated method to assess coronary flow reserve (CFR), but has not been used to assess CMD in ACHD. Here, we aim to determine the presence of CMD and PMD in ACHD non-invasively and to relate these to exercise capacity.

**Method:** We conducted a pilot study in 13 ACHD patients (5 males, median age 31 years), including patients with cyanotic (n = 7) and acyanotic (n = 6) defects and patients with (n = 5) and without (n = 8) history of cardiac surgery. CMD was assessed by CFR of the LAD with cardiac ultrasound. CFR was measured by pulsed-wave Doppler at rest and after adenosine administration intravenously and calculated as the ratio between hyperemic and baseline average peak coronary flow velocity values (Figure 1). A cut-off value of  $< 2.5$  was used to define CMD. PMD was measured by arterial tonometry at the fingertip. A reactive hyperemia index (RHI) of  $< 2.1$  was defined as reduced. Lab tests were performed and patients underwent cardiopulmonary exercise testing. We examined associations of CFR and RHI with patient characteristics (e.g., age, sex, BMI), common biochemical parameters (e.g., hemoglobin, NT-proBNP) and percent-predicted peak oxygen consumption (% ppVO<sub>2</sub>; reference values following Wasserman et al.) as well as between CFR and RHI.

**Results:** CFR could successfully be measured in 9 out of 13 patients, and CMD was observed in 9 out of these 9. RHI was reduced in 9 out of 13 patients and in 8 out of 9 with CMD. CFR was higher in patients with LDL-c  $< 115$  mg/dl compared to patients with LDL-c values  $> 115$  mg/dl ( $p = 0.03$ ). We found significant negative correlations between CFR and systolic blood pressure ( $\rho = -0.75$ ;  $p = 0.02$ ), hemoglobin ( $\rho = -0.67$ ;  $p = 0.049$ ), fasting glucose ( $\rho = -0.82$ ;  $p = 0.007$ ); and a positive correlation with %ppVO<sub>2</sub> ( $\rho = 0.74$ ;  $p = 0.02$ ).

**Conclusions:** This pilot study indicates that microvascular dysfunction is a common finding in ACHD patients, supporting more extensive studies to determine its prevalence, extent and pathological contribution to prognosis.

**Keywords:** Microvascular (dys)function, Coronary flow reserve, "Microcirculation"[Mesh], Adults with congenital heart disease, "Heart Defects, Congenital"[Mesh]



**Figure 1** Measuring coronary flow velocity by transthoracic cardiac ultrasound; pulsed-wave Doppler in the distal left anterior descending artery at rest (A) and after adenosine administration intravenously (B). The traces were characteristically biphasic with a prominent diastolic component. At baseline and during hyperemia, three optimal profiles of peak diastolic Doppler flow velocities were measured, and the results were averaged. CFR was calculated as the ratio between hyperemic and basal average peak velocities.

**P-006****Retinal vessel analysis in adult patients with congenital heart disease.**

Jan Müller<sup>1</sup>, Daniel Göder<sup>1</sup>, Julia Hock<sup>2</sup>, Leon Brudy<sup>1</sup>, Peter Ewert<sup>2</sup>, Renate Oberhoffer Fritz<sup>1</sup>

<sup>1</sup>Institute of Preventive Pediatrics, Technische Universität München, Munich, Germany; <sup>2</sup>Department of Pediatric Cardiology and Congenital Heart Disease, Deutsches Herzzentrum München, Technische Universität München, Munich, Germany

**Background and Aim:** Retinal vessel analysis gives important information on the damage to microvascular structures and enables first initial conclusions about cerebro- and cardiovascular risk.

**Method:** 91 Adults with Congenital Heart Disease (ACHD) (38.0 ± 9.6 years, 40 female) and 68 healthy controls (RC) (30.2 ± 12.4 years, 34 female) were analyzed via SVAdrs ocular fundus camera (Imedos Systems GmbH, Jena, Germany) and analyzed for »Arteriolar-to-venular ratio« (AVR), »Central retinal arteriolar equivalent« (CRAE), and »Central retinal venular equivalent« (CRVE)

**Results:** Mean AVR was 0.786 ± 0.100 on the left eye retina, and 0.816 ± 0.102 on the right eye retina for ACHD. In the healthy control group AVR was 0.840 ± 0.083 on the left eye retina, and 0.830 ± 0.082 on the right eye. After adjusting for age, sex, BMI and systolic blood pressure ACHD showed significantly reduced values on both retinas for AVR (left: ACHD: 0.785 ± 0.101 vs. RC: 0.841 ± 0.084, p = .002; right: ACHD: 0.814 ± 0.099 vs. RC: 0.830 ± 0.082, p = 0.022). Furthermore, CRAE (left: ACHD: 175 ± 29 vs. RC: 204 ± 20, p < .001; right: ACHD: 181 ± 25 vs. RC: 203 ± 21, p < .001) and CRVE (left: ACHD: 224 ± 31 vs. RC: 244 ± 19, p < .001; right: ACHD: 225 ± 31 vs. RC: 245 ± 21, p < .001) were narrower.

**Conclusions:** ACHD showed significantly worse values in several retinal vessel parameters in comparison to healthy controls. The clinical significance of these findings and further subgroup analysis needs to be evaluated in future studies on microcirculation of ACHD.

**Keywords:** Microcirculation, Retinal vessel analysis, Congenital Heart Disease

**P-008****Safety of transcatheter ASD closure in elderly patients**

Petya Stoyanova Parashkevova<sup>1</sup>, Lyubomir Georgiev Dimitrov<sup>1</sup>, Ivan Getov Velkovski<sup>1</sup>, Valeri Lyubenov Gelev<sup>2</sup>

<sup>1</sup>Department of Pediatric Cardiology, National Hospital of Cardiology, Sofia, Bulgaria; <sup>2</sup>Department of Cardiology, Acibadem City Clinic Tokuda Hospital, Sofia, Bulgaria

**Background and Aim:** Atrial septal defect (ASD) is the most common type of congenital heart defect diagnosed in adults but closure in elderly patients is controversial. We aimed to evaluate the outcomes and safety of transcatheter closure of secundum atrial septal defects (ASDs) in two elderly patients.

**Method:** The first patient is a 74-year-old man with a history of ST-elevation myocardial infarction (STEMI) in 2010, left ventricular dysfunction, chronic atrial fibrillation, heart failure NYHA class III, and arterial hypertension grade III. The second patient is a 70-year-old woman with newly diagnosed ASD due to an attack of atrial fibrillation. The patient has long-standing grade 3 arterial hypertension. In addition to routine examinations, both underwent computer tomography (CT) and 3D reconstruction of the

defect to assess its size and morphology. Patients were considered suitable for transcatheter closure of the ASD.

**Results:** Transcatheter atrial septal defect device implantation is a safe procedure. However, in elderly patients there may be some complications like acute pulmonary edema when the increase in left atrial pressure after the closure of ASD impairs left ventricular diastolic function. Though left atrial pressure is often monitored during test occlusion of a defect, it is not clear at what cut-off value device implantation is contraindicated. To prevent this complication the diuretic therapy in our patients was escalated two days before the procedure. The ratio of pulmonary blood flow to systemic blood flow (Qp/Qs) were 3.11 and 2.5 respectively. Both had elevated pulmonary artery pressure, mPAP 35 and 30 respectively. During the procedure, both patients underwent balloon sizing of the defects and balloon occlusion for 10 min. Pulmonary capillary wedge pressure (PCWP) increased from 18 to 21, and from 19 to 20 respectively during the balloon occlusion. The defects were closed with Amplatzer septal occluders. Patients were discharged from the hospital on postprocedure day 3.

**Conclusions:** Closure of ASDs in elderly patients can be a real challenge to cardiologists. The procedure can cause significant complications like acute pulmonary edema. Careful patient selection and preemptive measures like balloon occlusion and measurement of PCWP can make the procedure safer and the periprocedural period smoother.

**Keywords:** ASD, transcatheter closure, balloon occlusion, elderly

**P-009****10-year single-center experience of surgical pulmonary valve replacement in guch patients**

Armin Darius Peivandi, Sven Martens, Andreas Rukosujew, Sabrina Martens

Department of Cardiothoracic Surgery, University Hospital Muenster, Muenster, Germany

**Background and Aim:** Surgical pulmonary valve replacement (sPVR) after initial repair of various congenital cardiac malformations is standard therapy in GUCH patients. In order to examine our center's results, we analyzed our patient cohort of the last 10 years.

**Method:** Between January 2013 and August 2022 all GUCH patients who underwent sPVR were included in the study. Primary diagnoses as well as peri-operative data and in-hospital mortality were analyzed. Normality of continuous variables was tested using Shapiro-Wilk test. Either median (IQR3-1) or n (%) were calculated.

**Results:** 80 GUCH patients (60% male, age 42 (52.7-29) years) underwent surgical pulmonary valve replacement. Main underlying disease was tetralogy of Fallot (n = 48, 60%). Most patients received a stented bioprosthesis (Edwards Perimount) 93.75% (n = 75) of large diameter (25 mm; n = 63, 78.75%) to provide the basis for future valve interventions. X-clamp time was 69 (105.25-0) minutes. In-hospital mortality of our cohort was low (6.25% (n = 5)).

**Conclusions:** sPVR is a safe therapeutic option in GUCH patients with low mortality, providing an opportunity for further interventions in the future.

**Keywords:** surgical pulmonary valve replacement, GUCH patients, Fallot tetralogy

## P-011

**Long-term home-based physical endurance and inspiratory muscle training for fontan patients**

Lena Walzer<sup>1</sup>, Peter Kramer<sup>1</sup>, Anastasia Schleiger<sup>1</sup>, Stefan Dirks<sup>1</sup>, Bernd Wolfarth<sup>2</sup>, Thomas Thouet<sup>2</sup>, Felix Berger<sup>1</sup>, Hannes Sallmon<sup>1</sup>, Marcus Kelm<sup>1</sup>, Stanislav Ovrutsky<sup>1</sup>

<sup>1</sup>Department of Congenital Heart Disease—Pediatric Cardiology, Deutsches Herzzentrum Berlin, Berlin, Germany; <sup>2</sup>Department of Sports Medicine, Charité—Universitätsmedizin Berlin, Berlin, Germany

**Background and Aim:** Lifestyle adaptations such as regular physical activity are recommended for patients with heart disease and are also of increasing interest for patients after Fontan palliation. Our aim was to investigate the effects of individualized home endurance training (IHET) on a bicycle ergometer in combination with inspiratory muscle training (IMT) in pediatric and adult Fontan patients.

**Method:** A total of 25 patients were initially enrolled to a prospective intervention study, out of which 16 patients aged 18.25±8.68 years (7 female, 9 male) completed follow-up at 22 months, including cardiopulmonary exercise testing (CPET). A Magbike® AM-5i/3i (DKN Technology) bicycle ergometer was used for the home exercise program and a POWERbreathe Medic plus handheld device (HaB GmbH) was used for the IMT. Training was increased over the course of the study to 80 minutes of basic endurance training per week, 25 minutes of interval training per week, and IMT with 30 breaths per day for 6–7 days per week.

**Results:** Compared to baseline, at 22 month follow-up exercise capacity increased from 1.93±0.63 to 2.15±0.6W/kg,  $p = 0.040$ ). Oxygen pulse increased from 8.55±3.12 to 9.58±2.73ml/beat ( $p = 0.068$ ). Vital capacity increased from 80 to 85% of reference values ( $p = 0.047$ ). Mean inspiratory pressure increased from 6.7±2.3 to 8.6±4kPa ( $p = 0.013$ ) and expiratory pressure from 6.6±2.3 to 8±3.7kPa ( $p = 0.04$ ), respectively. Peak oxygen uptake (VO<sub>2</sub>peak) increased from 1.25±0.34 to 1.49±0.4l/min ( $p = 0.028$ ). No adverse events or unplanned interventions occurred during the study.

**Conclusions:** In Fontan patients, IHET in combination with IMT lead to improvements in exercise capacity, oxygen pulse, VO<sub>2</sub>peak, mean inspiratory and expiratory airway pressure. IHET/IMT interventions may introduce an additional safe treatment strategy for improving physical fitness and respiratory muscle function in Fontan patients, which might help to delay the inevitable deterioration of physical capacity and hemodynamics. Further studies are needed to understand the long-term impact and compare long-term outcomes to age and sex matched controls.

**Keywords:** Fontan patients, Physical and Inspiratory Training

## P-012

**A case of successful lymphatic embolization for refractory PLE after fontan surgery**

Atsuhito Takeda<sup>1</sup>, Daisuke Abo<sup>2</sup>, Hirokuni Yamazawa<sup>1</sup>, Gaku Izumi<sup>1</sup>, Daisuke Sasaki<sup>1</sup>, Ayako Nagai<sup>1</sup>, Takao Tsujioka<sup>1</sup>, Yuji Manuo<sup>1</sup>

<sup>1</sup>Department of Pediatrics, Hokkaido University Graduate School of Medicine, Sapporo, JAPAN; <sup>2</sup>Department of Diagnostics and Interventional Radiology, Hokkaido University Graduate School of Medicine, Sapporo, JAPAN

**Background and Aim:** Prolonged protein-losing enteropathy (PLE) after Fontan surgery is one of the poor prognostic factors. Recently, lymphatic embolization has been tried for refractory PLE, but there are still few reports. Here we report a case of

successful lymphatic embolization for refractory PLE after Fontan surgery.

**Method:** (Case presentation)

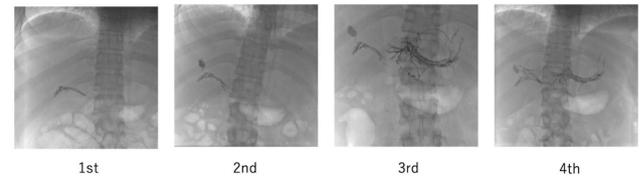
**Results:** The patient was diagnosed with a concordant criss-cross heart, straddling tricuspid valve, and small right ventricle by echocardiography at birth. Following surgical operation was performed: pulmonary artery banding at 2 months, palliative arterial switch at 2 years, bidirectional Glenn at 5 years, and TCPC completion with extracardiac conduit at 6 years. At the age of 15, he developed PLE with generalized edema, which temporarily improved with steroids, but later became prolonged. At this time, aortic root enlargement and left pulmonary artery stenosis were observed. At the age of 19 years, he was referred to our hospital and underwent David's surgery and Fontan root reconstruction, but PLE re-worsened after the surgery. Despite hemodynamical improvement and previously reported medical therapy for PLE, PLE became refractory over the 7 years with a serum albumin level of less than 2.0 g/dL. Therefore, transhepatic lymphatic embolization was performed. n-butyl-2-cyanoacrylate (NBCA)-Lipiodol was injected into the lymphatic system around the portal umbilicus, near the hepatic sickle mesentery, and in the ventral region of the pancreatic body. After the fourth embolization, gastrointestinal bleeding from the duodenum was observed, requiring endoscopic hemostasis several times, but the serum albumin level reached 3.0 g/dL without albumin infusion.

Lymphatic embolization is indicated for patients with refractory PLE. Multiple embolization procedures would be necessary in cases of extensive lymphatic leakage. The main complication was gastrointestinal bleeding, which is closely associated with the therapeutic efficacy of PLE.

**Conclusions:** Lymphatic embolization was effective when performed multiple times while taking into consideration adverse events such as gastrointestinal bleeding.

**Keywords:** Fontan, PLE, Lymphatic embolization

## Transhepatic lymphatic embolization for PLE



NBCA(20%)-Lipiodol was injected into the lymphatic system

## P-013

**Long term pacing mode effect on echocardiographic strain analysis in fontan palliated children**

Sonia Albanese, Mario Panebianco, Carolina D'anna, Laura Asta, Camilla Calvieri, Corrado Di Mambro, Massimo Silveti, Fabrizio Drago

Department of Cardiac Surgery, Cardiology, Heart and Lung Transplantation Bambino Gesù' Children's Hospital IRCCS, Roma, Italy

**Background and Aim:** Fontan circulation leads frequently to arrhythmias that sometimes require pacemaker (PMK) implantation. Despite a large number of Fontan PMK patients, there are few studies characterizing their population. Our aim was to compare echocardiographic characteristics in these patients according to pacing mode stimulation.

**Method:** During a period of 13 yrs, we prospectively enrolled 55 children undergoing Fontan operation and subsequent PMK implantation. Data on surgical intervention, PMK implantation, PMK complications and reoperations, pacing modality and echocardiographic measures were collected. Echocardiographic speckle tracking strain analysis and 3D ejection fraction evaluation were obtained by two independent pediatric cardiologists.

**Results:** 55 children (27 female, 48%), 35 (64%) of whom with single ventricle, underwent Fontan operation with extracardiac conduit in 50 (89%) and fenestration in 13 (23%) at a median age of 3 yrs (IQR 2–6 yrs) and subsequent surgical epicardial PMK implantation after median 4 yrs (IQR 0–10) from the operation. Indications for PMK implantation were sinus node dysfunction (SND) in 38 (68%) and atrioventricular block (AVB) in 18 (32%). Surgical access was a sternotomy in 42 cases (76%) and a left thoracotomy in 13 (24%). Twenty-six patients (47%) underwent a second PMK procedure 12 yrs (IQR 5–17 yrs) later for atrial electrode fracture in 9 (35%), ventricular electrode fracture in 7 (27%), both electrodes fracture in 6 (23%), electrodes dysfunction in 2 (7.6%) and high electrodes thresholds in 2 (7.7%). Among those, 18 (69.2%) underwent re-implantation, 4 (15.4%) electrode repair, and 4 (15.4%) system upgrading to CRT. During the observation period, 4 patients underwent cardiac transplant and one died.

Echocardiographic analysis comparison between patients with AV monopolar (n = 10, 18%) and AV bipolar (n = 13, 24%) stimulation showed no difference in 3D LVEF, whereas circumferential strain ( $-20 \pm 6, 3\%$  vs  $-26, 5 \pm 3\%$ ,  $p = 0.013$ ) and left atrial volume ( $44 \pm 23$  ml vs  $23 \pm 11$  ml,  $p = 0.003$ ) were significantly better in those receiving bipolar stimulation.

**Conclusions:** During long-term follow up, PMK atrioventricular bipolar pacing seems to positively affect circumferential strain and left atrium dimension more than monopolar modality in Fontan palliated children.

**Keywords:** Fontan, PMK, echocardiography, strain, bipolar stimulation

#### P-014

##### **Efficacy and safety of direct-acting oral anticoagulants (doac) versus vitamin k antagonist (VKA) for patients with congenital heart disease (CHD)**

Sedra Al Yousef

Department of Cardiology, Rigshospitalet, Copenhagen, Denmark

**Background and Aim:** Patients with congenital heart disease (CHD) have an increased risk of arrhythmia and thrombo-embolic events. Therefore, prophylactic anticoagulation therapy is common in CHD, but it is debatable whether direct-acting oral anti-coagulants (DOAC) is as efficient and safe vitamin K antagonist (VKA) in this patient population. The aims of this study were to investigate the temporal trends in the use of DOAC and VKA, as well as the thrombo-embolic and bleeding events of these anti-coagulants in Danish CHD patients.

**Method:** A retrospective descriptive cohort study including all CHD patients identified from The Danish National Patient Register (DNPR). With an individual linkage to Danish nationwide registries, oral anti-coagulants, as well as bleeding and thromboembolic events from 1995 to 2018 were obtained. Outcome was calculated as events per patient years at risk defined as the total years in which patients were exposed to the medication.

**Results:** During the study period, 3,501 patients with CHD were treated with either DOAC or VKA. DOACs constituted of 0.3% of all anti-coagulants among CHD patients in 2009 but increased to 38.0% in 2018. Compared to the VKA, patients on DOACs had both a higher thromboembolic rate (94 vs. 74 events per 100 patient year) as well as a higher bleeding rate (58 vs. 18 events per 100 patient year)

**Conclusions:** DOAC has increasingly been replacing VKA in treating CHD patients in Denmark. Nevertheless, the findings in this study suggest that DOAC is both less effective and safe than VKA in patients with CHD.

**Keywords:** CHD, DOAC, VKA, anticoagulation, arrhythmia

#### P-015

##### **A case of multiple late complications of mustard operation for transposition of great arteries**

Alkestis Eleni Kalesi<sup>1</sup>, Maria Karakosta<sup>1</sup>, Nikolaos Miaris<sup>1</sup>, Dimitrios Boukis<sup>1</sup>, Evangelia Grigoraki<sup>1</sup>, Nearchos Kasinos<sup>1</sup>, Antonios Destounis<sup>1</sup>, Anastasios Theodosios Georgilas<sup>1</sup>, Evangelia Nyktari<sup>2</sup>, Panagiotis Zachos<sup>3</sup>

<sup>1</sup>Echocardiography Training Center, Cardiology Department, "Tzanio" General Hospital, Piraeus, Greece; <sup>2</sup>CMR Unit, Onassis Cardiac Surgery Center, Athens, Greece; <sup>3</sup>Pediatric Cardiology and Adult Congenital Heart Disease Department, G.H. of Karditsa, Karditsa Greece

**Background and Aim:** Complete transposition of the great arteries (D-TGA) stands for 5% of congenital heart diseases (CHD). In this entity, there is ventriculo-arterial discordance and arterial-ventricular concordance. If not surgically corrected, survival into adulthood is very poor. Surgical approaches prior to the 1980s were Mustard and Senning procedures that tried to reverse the systemic and pulmonary venous inflow, by placing baffles within the atria. Common complications of these operations are baffle leaks or stenosis and heart failure.

**Method:** We report a case of multiple late complications of Mustard operation for D-TGA. The diagnostic imaging techniques that were used, included echocardiography and cardiac magnetic resonance (CMR).

**Results:** The case concerns a 51-year-old male who presented to our CHD department complaining about shortness of breath for the past 3 months. From his medical history, he was operated at the age of 5 because of D-TGA with a Mustard technique. His electrocardiogram was in sinus rhythm with first degree atrioventricular block and right bundle branch block. The heart ultrasound revealed systemic right ventricular dilatation with impaired systolic contractility. The subpulmonary left ventricle was also dilated with preserved ejection fraction and concomitant significant pulmonary valve regurgitation. At least two leaks were observed at the baffle of systemic veins. A CMR followed, demonstrated the presence of two leaks between the pulmonary veins' baffle and the inferior vena cava (IVC) limb. There was no systemic and pulmonary venous pathway stenosis, while Qp/Qs was 1, 6. Cardiac catheterization confirmed the above findings.

**Conclusions:** Baffle leakage in patients undergoing Mustard surgery due to D-TGA is a common late complication (25%). Nevertheless, its relationship with the worsening of systemic ventricular function and the possible reverse remodeling after correction of leaks is of great interest.

**Keywords:** d-TGA, Mustard operation, baffle leak, late complications

## Mustard operation

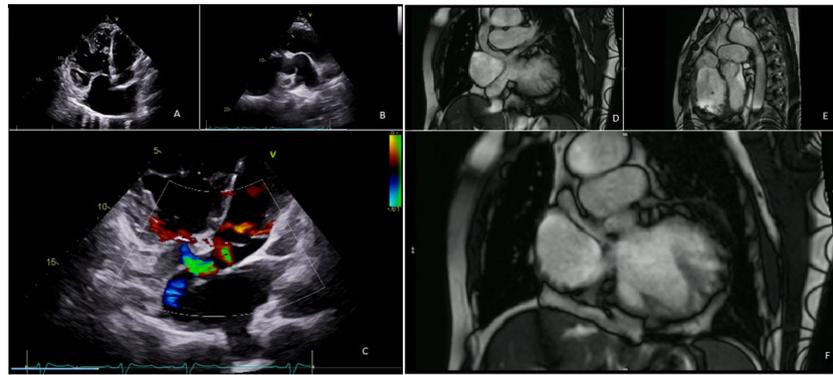


FIGURE: A: Echo. Systemic right ventricular dilatation and hypertrophy. B: Echo. Parallel course of aortic and pulmonary artery, indicative of D-TGA. C: Echo. Presence of at least two leaks in the systemic venous baffles. D: CMR. Inferior and superior vena cava limbs without stenotic phenomena. E: CMR. Severe pulmonary valve regurgitation. F: CMR. Leakage to the inferior vena cava limb.

## P-016

### Effects of exercise training on lymphatic function in fontan patients – a controlled trial

Lene Thorup<sup>1</sup>, Aida Luiza Ribeiro Turquetto<sup>2</sup>, Luciana Patrick Amato<sup>2</sup>, Benjamin Kelly<sup>3</sup>, Vibeke Hjortdal<sup>1</sup>

<sup>1</sup>Department of Cardiothoracic Surgery, Copenhagen University Hospital (Rigshospitalet), Copenhagen, Denmark; <sup>2</sup>Heart Institute (InCor), University of Sao Paulo Medical School, São Paulo, Brazil; <sup>3</sup>Department of Cardiothoracic and Vascular Surgery, Aarhus University Hospital, Aarhus, Denmark

**Background and Aim:** Every day, the lymphatic system returns about 8 L of lymphatic fluid to the systemic circulation. Thus, a compromised lymphatic system can cause severe symptoms such as protein losing enteropathy and plastic bronchitis. Patients living with a Fontan circulation are especially prone to this, possibly due to their elevated central venous pressure.

Exercise training in Fontan patients has gained recognition for improving exercise capacity and perhaps also Fontan physiology. Furthermore, regression of PLE symptoms after starting exercise programs have been described and animal studies have demonstrated an improvement of lymphatic dysfunction when upon exercise. This has not been investigated in humans.

The aim of this study was to investigate the effect of exercise training on lymphatic function in Fontan patients and increase knowledge about the lymphatic physiology in this patient group.

**Method:** This was conducted as a controlled study of 24 Fontan patients. Lymphatic function was investigated using Near Infrared fluorescence Imaging of the lower limb with intradermal Indocyanine Green injection, to determine lymphatic contraction frequency, velocity, and pressure at relaxed state and during lymphatic stress (gravitation). A baseline investigation was conducted in October 2022 followed by convenience sampling of 10 patients to receive 36 sessions of supervised exercise training and 14 patients to no intervention. After two months a follow-up investigation will be conducted on all patients and results compared between the two groups.

**Results:** Results are pending.

**Conclusions:** This study will contribute with knowledge of both baseline lymphatic function in a patient population at risk of lymphatic complications as well as insights on possible new management strategies for Fontan patients.

**Keywords:** Lymphatics, Fontan, Exercise Training

## P-017

### Operated tetralogy of fallot: clinical and echocardiographic approach in adult life

Mahdi Walha, Rania Gargouri, Yesmine Keskes, Hassen Gargouri, Niez Laaribi, Yecine Abdelmoula, Leila Abid

Cardiology Department, Hedi Chaker university hospital, Sfax, Tunisia

**Background and Aim:** The evolution of open heart surgery has radically improved the prognosis of these patients. However, the evolution is marked by a significant percentage of morbidity. We studied in our work the different complications as well as their predictive factors to improve the management of patients with tetralogy of Fallot.

**Method:** 30 consecutive patients followed in our department for operated tetralogy of fallot aged over 18 years were included. We performed a collection of the patients' symptoms as well as an echocardiographic study including a detailed study of the right ventricle and the right ventricle outflow tract. Exercise capacity was studied using the 6-minute walk test

**Results:** This is a series of 30 patients with a median age of 26.5 years with extremes between 18 and 69 years. 56.67% were female. 21 patients had a regular form of fallot, 7 patients had hypoplasia of the PAs and 2 patients had a birth defect of the coronary arteries. Age at surgery ranged from 1 to 47 years with a median of 4 years.

3 patients had a residual ventricular septal defect, 5 patients had a dilated aorta, 17 patients had a dilated RV which was associated with significant PI ( $p = 0.001$ ), older surgery ( $p = 0.034$ ).

RV systolic function was impaired in 11 patients. Male sex ( $p = 0.002$ ), increased body surface area, wide QRS, and the presence of pulmonary stenosis were associated with impaired RV systolic function.

The strain of the RV was studied and was impaired in case of pulmonary stenosis ( $r = 0.437$ ,  $p = 0.018$ )

A significant PI was noted in 40% of cases, it was associated with an older age at the time of surgery ( $p = 0.05$ ) and an irregular form of fallot ( $p = 0.023$ )

A significant pulmonary stenosis was noted in 36.7% of cases. It was associated with male sex and caused an alteration of the systolic function of the RV

The exercise capacity was measured by 6min Walk-test.

Decreased functional capacity was noted with either systolic or diastolic RV dysfunction.

**Conclusions:** male gender, increased body surface area, advanced age at the time of surgery are parameters to consider for patients followed for tetralogy of fallot surgery.

**Keywords:** Tetralogy of Fallot, Prognosis, RV strain, 6-min walk test

## P-018

### Predictive factors of mortality and heart failure in children and adults with ebstein anomaly

Dimitri Herreira<sup>1</sup>, Olivier Metton<sup>1</sup>, Julia Mitchell<sup>1</sup>, Francis Bessiere<sup>1</sup>, Sylvie Di Filippo<sup>2</sup>

<sup>1</sup>Department of Congenital Heart Disease, Claude Bernard University, Lyon, France; <sup>2</sup>Congenital Cardiology, Cardiothoracic Center of Monaco

**Background and Aim:** The objective of the study was to analyze outcomes of patients with Ebstein anomaly and assess risk factors for death and/or heart failure over follow-up

**Method:** All patients diagnosed with Ebstein anomaly were included in this retrospective single-center study. Demographics, clinical data, transthoracic echocardiographic data, outcomes and events were assessed.

**Results:** Seventy-six patients were included, 34 males (45%), diagnosed at mean age 21.9years, median 13.9 (neonates 27%, children 27%, adult 46%). NYHA class at time of diagnosis was I, II, III and IV in respectively 55%, 35%, 7% and 3%. ASD was present in 71% of cases, WPW in 16%, obstructive RV outflow tract lesion in 12%, VSD in 10%. Follow-up was 2.8 to 66 years, mean 28 and median 25 years. Supraventricular tachycardia occurred in 43.5%. Pacemaker implantation occurred in 12%. Three deaths occurred (4%), due to uncontrolled heart failure (HF). HF was observed in 26% of cases, thromboembolic events in 9%, stroke in 10.5%. Mean age at event was 39 to 41 years. Freedom from HF was 90% at 40years, 80% at 50years and 50% at 70years. Freedom from procedure was 85%, 80%, 70%, 50%, 20% at respectively 10, 20, 50 and 70 years of age. Death correlated with HF, and was not associated with Ebstein anatomy type, age at diagnostic, arrhythmias, RV volumes, pacemaker implantation, tricuspid valve replacement, number of procedures, ASD closure or ablation. HF occurrence was associated with major type of Ebstein anomaly ( $p = 0.03$ ), tricuspid regurgitation grade III-IV ( $p = 0.03$ ), neonatal age at diagnosis ( $p = 0.09$ ), arrhythmias ( $p = 0.003$ ) and more specifically IART, and with the number of procedures over follow-up (3 or more,  $p = 0.006$ ).

**Conclusions:** Long-term survival rates of Ebstein anomaly patients were favorable. Main risk factor for death was heart failure. Predictive factors for HF were major Ebstein and TR severity, arrhythmias and iterative procedures.

**Keywords:** Ebstein anomaly, mortality, heart failure, risk factor

## P-021

### Shared decision-making in decisionally-impaired adults considerations for the congenital heart provider

David Vinas Fernandez<sup>1</sup>, Ibett Colina Garcia<sup>2</sup>, Rafael Alonso Gonzalez<sup>1</sup>, Craig S Broberg<sup>2</sup>

<sup>1</sup>Toronto ACHD Program, Peter Munk Cardiac Centre, University Health Network, University of Toronto, Toronto, Ontario, Canada; <sup>2</sup>Knight Cardiovascular Institute, Oregon Health & Science University, Portland, Oregon, USA.

**Background and Aim:** In recent years, due to increased life expectancy of people with disabilities and a profound cultural

transformation, there has been a change in the trend towards patient empowerment, leaving behind a more paternalistic approach. As providers who care for patients with impaired decision-making capacity, whether or not they are legally incompetent, to the emphasis is usually on the question of: “how do I determine what is or would be the patient’s own decision?” We aimed to develop visual material as guidance and discussion for the congenital heart provider facing these challenging situations.

**Method:** After review of available literature from sources including scientific studies, patient advocacy groups, ethical boards, and government agencies, agreement was reached on techniques to approach the decisionally-impaired. All contemporary documents advocate a shared decision process as currently accepted best-practice. Visual material was developed in a collaboration with all the authors. This was adapted from the three-talk model for shared decision-making (G. Elwyn et al. BMJ 2017;359:j4891)

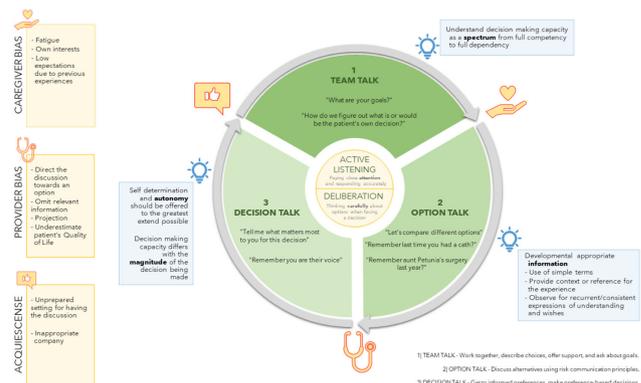
**Results:** A user-friendly and visual algorithm for the shared decision process adapted to assisting decisionally-impaired adults was developed emphasizing autonomy and informed shared decision making. Following the three-talk model for shared decision-making, the graph also includes usual biases for providers to be mindful of and helpful tips. This is attached as supplemental material.

**Conclusions:** For health care providers, skills for assisting an appropriate shared decision-making process include developmentally appropriate disclosure of information and ongoing education about options in simple language. Autonomy, and so decision-making capacity, is not binomial but a spectrum from full competency to full dependency. Offering visual information with tips and tricks and usual pitfalls to watch for and awareness of certain biases, will improve the care provided to this special population and their caregivers and families.

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**Keywords:** shared decision-making, autonomy, mental incompetence, disability, congenital heart disease, ethics.

Three-talk model for shared decision-making in decisionally-impaired adults with congenital heart disease



Adapted from the three-talk model for shared decision-making (G. Elwyn et al. BMJ 2017;359:j4891)

## P-023

**Challenges and outcomes of establishing a department of adult congenital heart surgery in a private hospital in Greece**

Andreas C. Petropoulos<sup>1</sup>, Panagiotis Sfridis<sup>2</sup>, Evangelia Ntalarizou<sup>2</sup>, Angelos Katsianis<sup>5</sup>, Vasilios Kyriakopoulos<sup>3</sup>, Chrisovalantis Pothitakis<sup>3</sup>, Zois Panagopoulos<sup>4</sup>, Prodomos Azariathis<sup>5</sup>, Afksendiyos Kalangos<sup>6</sup>

<sup>1</sup>Department of Adult Congenital Heart Disease Surgery, Euroclinic Hospital, Athens Greece, The "Aziz Aliyen" National Postgraduate and CME Medical Training Center, Baku-Azerbaijan; <sup>2</sup>Department of Cardiothoracic Surgery, "St.Sophia" Children's Hospital", Athens Greece; <sup>3</sup>Department of Anaesthesiology, Euroclinic Hospital, Athens, Greece; <sup>4</sup>Intensive Care Unit, Euroclinic Hospital, Athens, Greece; <sup>5</sup>Department of Adult Congenital Heart Disease Surgery, Euroclinic Hospital, Athens Greece; <sup>6</sup>Department of Adult Congenital Heart Disease Surgery, Euroclinic Hospital, Athens Greece, Department of Pediatric and Congenital Cardiac Surgery, Koç University, Istanbul, Turkey

**Background and Aim:** Incidence of adults suffering unknown – simple or surgically palliated congenital heart diseases (ACHD) is rising worldwide. Since 2000 overcome the pediatric population (60/40%). Studies predict further increase (75/25%) by 2050. This exceptional population suffering from vast morbidity, facing early mortality rate; have many social-economic challengers. They need multidisciplinary medical teams. Aiming to present Challenges and Outcomes during two years from establishment ACHD surgery service in Greek private hospital.

**Method:** Since, November 2020 a department of ACHD Surgery was established in Athens. We evaluated and operated on 30 patients suffering from CHD. 19/30(63%) men, 11/30(37%) women. Age ranged 19 - 82 years. Mean age 40, 7 years. Patients suffered: 1. MvR+6/30(20%), 2. Left AVvR+ in c-AVSD 1/30(3.3%), 3. MvR+ & AovS 1/30(3.3%), 4.ToF s/p PAvR+7/30(23%), 5. Tv atresia – TCPC, MvR+ 1/30(3.3%), 6. Tv atresia+ mBT-shunt 1/30(3.3%), 7. C-c -TGA + TvR+ 1/30(3.3%), 8. p-AVSD+ LVOT obstruction+ LAVvR+1/30(3.3%), 9. AovS 7/30( 23%), 10. AAo Aneurism-Marfan syndrome 1/30(3.3%), 11.AAo-Aneurism + BAovS 1/30(3.3%), Ebstein type C 2/30(6.6%). NYHA stages: II: 9/30(29.7%), III: 14/30( 46.2%), III<IV: 7/30(23.3%)

**Results:** Patients had sternotomies, previously 1-5 mean 1.9/ patient. We delivered 32 surgeries. Two reoperations (1. Tamponade, 2. Perivalvular leakage). Operative mortality was 1/30 (3.3%), c-c-TGA with sever TvR+ patient. Perioperative mortality 2/30 (7th s/p: PH crises, RV failure in p-AVSD case & 21th s/p: infection, sever MvR+ & sever AovS). 1/30 (3.3%) late mortality 37th s/p infection, Fontan patient with sever MvR+. Seriou's complications 2/30(6.7%), 1. Need permanent pacemaker 2. 53th s/p VT with arrest, followed, (L) hemiplegia. Intubation period: 2-16 (mean 11 days). Length of ICU stay 3-16 (mean 11 days). Length of hospitalization 8 -37 (mean 15 days). **Conclusions:** Establishing GUCH surgical service faces many Challengers: high morbidity, need sophisticated imaging, preoperative, perioperative management of HF/ PH, excessive bleeding due to additional coagulopathy and previous sternotomies. Basic socioeconomic issues were expensive hospital fees due to longer ICU and IPC stay and use of homographs as well as pour insurance coverage of many patients. Total mortality 13.2% is accepted for this sick and complex population.

**Keywords:** Adult Congenital Heart Surgery Service, Challengers, Outcomes

## P-024

**Respiratory function of adult congenital heart disease**

Yasumi Nakashima, Nao Inoue, Hisashi Sugiyama  
Department of pediatric cardiology, Seirei Hamamatsu General Hospital, Hamamatsu, Japan

**Background and Aim:** Although respiratory impairment has been reported to play a role in prognosis in patients with chronic heart failure, there are few reports on respiratory function in adult congenital heart disease (ACHD).

**OBJECTIVES:** To determine the prevalence and severity of respiratory impairment in ACHD.

**Method:** ACHD Patients over 15 years old who had respiratory function tests performed at our hospital were included in the study. We reviewed % vital capacity (%VC) and % predicted Forced Expiratory Volume in one second (%FEV1.0). We examined the relationship between respiratory function and cardiac surgery, surgical procedure (intracardiac repair (ICR) or Fontan), number of thoracotomies, and prognosis.

**Results:** 105 patients were enrolled in this study (46 unoperated and 59 operated (42 ICR and 17 Fontan). The mean age was older in the unoperated group than in the operated group (45.3±18.0 vs. 29.6 ±14.1, respectively, p<0.05). The number of thoracotomies was 1.8±0.9 for ICR and 2.9±1.1 for Fontan, with Fontan patients having significantly more thoracotomies (p<0.01). The mean %VC in the subject was 92.2 ± 20.3% and mean FEV 1.0% was 84.9 ± 20.0%. The %VC was lower in operated group (unoperated vs. operated group: 102.7 ± 19.6% vs. 84.5 ± 17.0%, respectively, p<0.05). % FEV1.0 was not significantly different (unoperated vs. operated group 81.9 ± 9.6% vs. 84.8 ± 8.6%, respectively). The number of thoracotomies and %VC were negatively correlated (p<0.001, r = -0.52), but not with %FEV 1.0. The mean %VC in ICR and Fontan patients were 86.3±15.7% vs. 78.2±19.1%, and %FEV1.0 was 84.6 ±9.5% vs. 85.2±6.4%, respectively, with no significant difference in %FEV1.0 or %VC. The percentage of cases with %VC <70% was significantly higher in Fontan (ICR vs. Fontan 7.1 vs. 35.3%). There were 6 deaths (2 unoperated, 4 operated). The %VC of deaths was significantly lower than that of survivors (75.9±16.3% vs. 93.5 ±20.1%, p<0.001). The FEV1.0% of the deaths (73.5±12.2%) was in the normal range, but lower than that of the survivors (84.1 ±8.6%) (p<0.001).

**Conclusions:** %VC decreases in operated ACHD. Fontan Patients often have severe respiratory dysfunction. Impaired respiratory function may also be associated with prognosis in patients with ACHD.

**Keywords:** respiratory function, adult congenital heart disease

## P-025

**Severity of aortic coarctation impact to renal perfusion**

Sigitas Cesna<sup>1</sup>, Augustinas Bielinis<sup>2</sup>, Kamile Cerlinskaite Bajore<sup>1</sup>, Virgilijus Tarutis<sup>3</sup>

<sup>1</sup>Clinic of Cardiac and Vascular Diseases, Institute of Clinical Medicine, Faculty of Medicine, Vilnius University, Vilnius, Lithuania; <sup>2</sup>Center of Radiology and Nuclear Medicine, Department of Nuclear Medicine, Vilnius University Hospital Santaros Clinics, Vilnius, Lithuania; <sup>3</sup>Center of Cardiothoracic Surgery, Clinic of Cardiac and Vascular Diseases, Institute of Clinical Medicine, Faculty of Medicine, Vilnius University, Vilnius, Lithuania

**Background and Aim:** Longterm outcome data suggest that up to 50% of patients with repaired aortic coarctation (CoA) have

persistent hypertension. Eventually significant CoA may lead to long standing renal hypoperfusion and may impact residual hypertension. In this pilot study we try to evaluate renal perfusion changes and relation to the severity of CoA.

**Method:** 20 patients with CoA underwent CT scan and were distributed in 2 groups (10 patients in each group). Patients with significant reCoA or native CoA had isthmus/AoD index <0.7 and Group 2 had isthmus/AoD index >0.7. The <sup>99m</sup>Tc-MAG3 captopril scintigraphy was performed for both patients groups. Time to peak, time to half-peak, peak to half-peak, 30 min/peak and 20 min/3 min count ratios were determined for whole-kidney ROIs. **Results:** Time to peak after captopril test for Group 1 was 3.01 [2.78, 3.75] and 3.46 [3.23, 3.63], for Group 2 was 3.18 [2.73, 4.13] and 4.23 [3.39, 4.93] for left and right kidneys, respectively ( $p > 0.05$ ). The median time to ½ peak was significantly lower in Group 2 (0.64 [0.44, 0.68] and 0.62 [0.47, 0.72] versus 0.85 [0.61, 1.02] and 0.86 [0.70, 1.00] for left and right kidneys, respectively), which show faster parenchymal perfusion in Group 2. Prolongation of time to ½ peak after captopril test was significantly higher in Group 2 ( $p = 0.009$  for left kidney and  $p = 0.008$  for right kidney), which can suggest signs of renal hypoperfusion. There were no differences between groups by other parameters obtained by renogram curves.

**Conclusions:** For patients with clinically significant CoA faster parenchymal perfusion and prolongation of time to ½ peak after captopril test aware about possibility to have renal hypoperfusion. Notably, this gives additional data for CoA treatment indications. However, further studies with larger number of patients needed to gain stronger clinical data.

**Keywords:** aortic coarctation, renal scintigraphy, ACHD, renal hypoperfusion, residual hypertension

## P-028

### Adult congenital heart disease (ACHD) training: A pan european survey and joint ESC and AEPC collaboration

Colin J. Mc Mahon<sup>1</sup>, Margarita Brida<sup>2</sup>, Petra Jenkins<sup>3</sup>, Annemien Van Den Bosch<sup>4</sup>, Joerg Stein<sup>5</sup>, Inga Voges<sup>6</sup>, Magalie Ladouceur<sup>7</sup>, Senka Meshihovic Dinarevic<sup>8</sup>, Gabriela Doros<sup>9</sup>, Kevin Walsh<sup>10</sup>, Ruth Heying<sup>11</sup>, Ornella Milanesi<sup>12</sup>, Massimo Chessa<sup>13</sup>, Werner Budts<sup>14</sup>

<sup>1</sup>Department Paediatric Cardiology, Children's Health Ireland, Crumlin, Dublin, Ireland; <sup>2</sup>Division of Adult Congenital Heart Disease, Department of Cardiovascular Medicine, University Hospital Centre Zagreb, Kispaticeva ul. 12, Zagreb 10000, Croatia.; <sup>3</sup>ACHD Department, Liverpool Heart and Chest NHS Foundation Trust; <sup>4</sup>Department of Cardiology, Erasmus University Medical Center, Rotterdam, The Netherlands; <sup>5</sup>Department of Paediatrics III, Cardiology, Pulmology, Allergology and Cystic Fibrosis, Medical University, Innsbruck, Austria; <sup>6</sup>Department of Congenital Heart Disease and Pediatric Cardiology, University Hospital Schleswig-Holstein, Kiel, Germany; <sup>7</sup>Hôpital Européen Georges Pompidou, Université de Paris, Paris, France; <sup>8</sup>Eurofarm Polyclinic, University of Mostar, Sarajevo, Bosnia and Herzegovina; <sup>9</sup>"Victor Babes" UMF, Ilird Pediatric Clinic, Louis Turcanu Emergency Children Hospital, Timisoara, Romania; <sup>10</sup>Department of Cardiology, Mater Misericordiae University Hospital, Dublin, Ireland; <sup>11</sup>Department of Paediatric Cardiology, UZ Leuven, Leuven, Belgium; <sup>12</sup>Paediatric Cardiac Unit, Department of Paediatrics, University of Padova, School of Medicine, Padova, Italy; <sup>13</sup>ACHD Unit - Pediatric and Adult Congenital Heart Centre, IRCCS-Policlinico San Donato, Piazza Edmondo Malan, 2, Milan 20097, Italy; <sup>14</sup>Congenital and Structural Cardiology, University Hospitals Leuven, and Department of Cardiovascular Sciences, Catholic University Leuven, Leuven, Belgium.

**Background and Aim:** Limited data exist on training of European adult congenital heart disease (ACHD) cardiologists.

**Method:** A structured and approved questionnaire was circulated to the lead ACHD specialist or where not available the national delegate of the Association for European Paediatric and Congenital Cardiology in 34 European countries.

**Results:** Delegates from 29 countries (85%) responded. ACHD cardiology was reported as recognised as a distinct speciality by the respective ministry of Health in one country (3%). Five countries (17%) have formally accredited ACHD training programmes, seventeen (55%) have informal (not accredited or certified) training, and seven (26%) have very limited or no programme. Twenty-three countries (79%) described training ACHD doctors on the job. Six countries (21%) have a curriculum. Four countries (14%) reported having a national training director. The median number of ACHD centres was 4 (range 0-28), median number of ACHD surgical centres was 3 (0-26) and the median number of ACHD training centres was 2 (range 0-28). There was one ACHD cardiology centre per 2.69 million population (range 0.66-8.63 million), one ACHD training centre per 3.43 million population (range 0.66-21.8 million), and one ACHD surgical centre per 3.57 million population (range 1.25-9.34 million population). The median number of ACHD cardiology fellows per training programme was 1 (range 0-3), and median duration of training was 2 years (range 1-5 years). Only two countries (6%) match training posts with postgraduate consultant cardiology posts. An established exit examination in ACHD was conducted in only 2 countries (6%) and formal certification provided by 1 country (3%). ACHD cardiologist number is affected by gross domestic product ( $R^2 = 0.32$ ).

**Conclusions:** Formal accredited training in ACHD is quite limited and poorly structured across the majority of European countries. Although formal fellowship programmes exist in a minority of countries, several countries have informal training or 'train people on the job'. A tiny minority of countries ( $n = 2$ ) provide either exit examination or certification. Many ACHD cardiologists still migrate from paediatric or adult cardiology. Harmonisation of ACHD training and standardisation of an exit examination and certification could reduce variation in training thereby promoting high-quality patient care by European ACHD cardiologists.

**Keywords:** Adult congenital heart disease, education, training

## Cardiac Dysrhythmias and Electrophysiology

### P-031

#### Successful treatment of frequent premature ventricular contractions and ventricular tachycardia with flecainide in a case of RYR1-related myopathy

Yuji Maruo<sup>1</sup>, Atsuhito Takeda<sup>2</sup>

<sup>1</sup>Department of Pediatrics, Hokkaido University Graduate School of Medicine, Sapporo, Japan; Department of Pediatrics, Japanese Red Cross Kitami Hospital, Kitami, Japan; <sup>2</sup>Department of Pediatrics, Hokkaido University Graduate School of Medicine, Sapporo, Japan

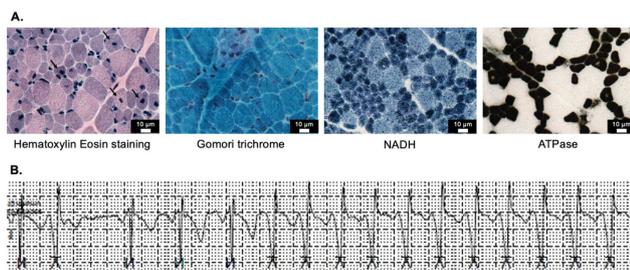
**Background and Aim:** Ryanodine Receptor 1 (RYR1)-related myopathies are a group of congenital muscle diseases caused by RYR1 mutations. These mutations may cause centronuclear myopathy, a congenital neuromuscular disorder characterized by clinical muscle weakness and pathological presence of centrally-placed nuclei on muscle biopsy. Mutations in RYR2 cause ventricular arrhythmias that are treatable with flecainide; however, reports of ventricular arrhythmias in RYR1-related myopathies are rare.

**Method:** A 15-year-old man presented with centronuclear myopathy with RYR1 mutations, and exhibited frequent premature ventricular contractions and non-sustained ventricular tachycardia that was successfully treated with flecainide.

**Results:** Neurological disease manifested at 7 months with hypotonia and delayed motor development. Skeletal muscle biopsy performed at 4 years of age revealed: type 1 fiber atrophy (5 to 10  $\mu\text{m}$  in diameter), type 1 fiber predominance (accounting for 85%), myofibers with centrally-placed nuclei (accounting for 10%), and absence of nemaline bodies or core structures; leading to the diagnosis of centronuclear myopathy (Figure A). At 15 years of age, premature ventricular contractions were identified on the electrocardiogram, and 24-hour Holter monitoring revealed 38, 065 premature ventricular contractions per day (27% of total QRS complexes) and non-sustained ventricular tachycardia (Figure B). Treatment with verapamil was initiated; however, it was not beneficial. Therefore, flecainide was added as a treatment, which decreased premature ventricular contractions frequency. At the age of 21 years, non-sustained ventricular tachycardia disappeared, and premature ventricular contractions almost disappeared at age of 22 years. Mutation screening for known causative genes for congenital myopathy using a custom-made panel revealed novel pathogenic variants, c.13216delG (p.E4406fs) and c.14874G>C (p.K4958N), in *RYR1*. Ryanodine receptors are calcium-release channels embedded in the endoplasmic reticulum membrane, an intracellular calcium storage site. Flecainide is reported to prevent catecholaminergic polymorphic ventricular tachycardia by inhibiting RYR2 receptor-mediated  $\text{Ca}^{2+}$  release. A similar mechanism may exist in the case of our patient by inhibiting RYR1 receptor, thereby ameliorating ventricular arrhythmias. In the most recent follow-up at the age of 23 years, no life-threatening cardiac complications were observed.

**Conclusions:** Patients with RYR1-related myopathies can present with potentially fatal ventricular arrhythmia in adulthood, which may be effectively treated using flecainide.

**Keywords:** RYR1-related myopathy, centronuclear myopathy, premature ventricular contraction, ventricular tachycardia, flecainide



Pathological findings of biopsied skeletal muscle (Figure A) and findings of 24-hour Holter monitoring (Figure B)

### P-032

#### Impact of maternal connective tissue disease on conduction abnormalities in the offspring

Sarah Sofie Andersen<sup>1</sup>, Terese Frellesen Neumann<sup>1</sup>, Sofie Dannesbo<sup>1</sup>, Otilia Birgitta Vøgg<sup>2</sup>, Anna Axelsson Raja<sup>1</sup>, Karen Schreiber<sup>3</sup>, Alex Christensen<sup>2</sup>, Kasper Iversen<sup>2</sup>, Henning Bundgaard<sup>1</sup>, Anne Sophie Sillesen<sup>2</sup>

<sup>1</sup>Department of Cardiology, Copenhagen University Hospital, Rigshospitalet, Copenhagen, Denmark; <sup>2</sup>Department of Cardiology, Herlev-Gentofte Hospital, Copenhagen University Hospital,

Copenhagen, Denmark.; <sup>3</sup>Department of Rheumatology, Sønderborg Hospital, Denmark

**Background and Aim:** Maternal systemic connective tissue diseases (CTD) and their antibodies may be associated with adverse fetal outcome, including increased risk of congenital heart disease (CHD) and complete heart block in the offspring. Whether this increased risk also applies to less severe cardiac abnormalities in the offspring, including other conduction abnormalities than complete heart block, is currently unknown.

Based on the to date largest cohort of newborns of mothers with CTD, we aim to assess the association between CTD and electrocardiographic/conductive abnormalities in the newborn.

**Method:** The study is a part of the Copenhagen Baby Heart Study (CBHS), a population study including more than 25, 000 newborns between April 2016 and October 2018. Included newborns had a transthoracic echocardiogram and an electrocardiogram (ECG) obtained during the first 30 days after birth (median 12 days).

The present study will include all children from the CBHS cohort born to mothers with CTD. Mothers with CTD will be identified using the Danish registries. Assuming an incidence of CTDs at 2%, we expect to include approximately 500 newborns to mothers with CTD. Cases will be matched 1:1 to controls matched on sex, gestational age, age and weight, and maternal age. Maternal medical charts will be reviewed to validate the CTD diagnosis and to obtain information about disease severity, treatment, etc. Based on the ECGs obtained in the newborns, the primary end-point is a composite of conduction abnormalities (heart rate, p-wave duration, PQ distance second, and third degree atrioventricular block, QRS duration and the QTc interval).

**Results:** Preliminary results from this study are expected in spring 2023.

**Conclusions:** It is unclear whether children born to mothers with CTD should have routine systematic cardiac evaluation and whether children born to mothers with subgroups of CTD are at a higher risk of conduction abnormalities compared to children born after uncomplicated pregnancies. The cohort size of CBHS and the thorough cardiac examination of included newborns provide a unique opportunity for new insights in this field, including whether a routine neonatal ECG should be recommended in this risk group.

**Keywords:** Conduction abnormalities, connective tissue disease, fetal outcome

### P-033

#### Cardiac resynchronization therapy in congenital heart disease

Yannic Wilberg<sup>1</sup>, Philine S. Warrnicke<sup>1</sup>, Theodor Tirilomis<sup>2</sup>, Matthias J. Müller<sup>1</sup>, Thomas Paul<sup>1</sup>, Ulrich Krause<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology, Intensive Care Medicine and Neonatology, University Medical Center, Georg-August-University Göttingen, Germany; <sup>2</sup>Department of Heart, Thoracic and Cardiovascular Surgery, University Medical Center, Georg-August-University Göttingen, Germany

**Background and Aim:** Cardiac resynchronization therapy (CRT) is well-established for the treatment of chronic heart failure (CHF) in adults with left bundle branch block. CRT is increasingly used in patients with congenital heart disease (CHD), however, data on outcome is still sparse. Aim of this study is to describe the use and significance of CRT in CHD.

**Method:** Single center, retrospective study; 41 subjects with CHD and implantation of a CRT device between 2003 and 2022 were enrolled. Outcome with respect to QRS duration and systemic ventricular ejection fraction (EF) and NYHA classification was

analyzed. Follow-up (FUP) was divided as follows: FUP1: < 1 year, FUP2: 1-5 years, FUP3: > 5 years. Non-responders were defined as subjects with deterioration of EF by > 5% or one NYHA classification during FUP. Survival rates and complications were assessed.

Median age at implantation was 38 years (IQR 23.5 years), median EF 43% (IQR 12.5%), median NYHA classification 2, median QRS duration 160 ms (IQR 60 ms). 24 patients (58.5%) had conventional pacing before CRT. Most common diagnoses were Tetralogy of Fallot (13/41), ccTGA (5/41), AVSD (4/41) and VSD (4/41). A systemic right ventricle (sRV) was present in 19.5% of the patients who were more likely to have non-transvenous leads implanted (5/8 vs. 4/33,  $p = 0.003^*$ ).

**Results:** Patients were followed for a mean of  $6.5 \pm 4.1$  years. Mean QRS duration shortened during CRT in all FUP periods ( $p = 0.002^*$  FUP1,  $p = 0.029^*$  FUP2,  $p = 0.016^*$  FUP3). Median EF increased in FUP2 and FUP3 ( $p < 0.001^*$ ,  $p = 0.013^*$ ). Median NYHA classification decreased significantly in FUP2 ( $p = 0.004^*$ ). Non-responder rates were 17.1%/6.7%/21.7% for NYHA status and 14.3%/0%/6.7% for EF with respect to each FUP period (FUP1/FUP2/FUP3). Non-transvenous leads were not inferior regarding perioperative complications ( $p = 0.197$ ) or lead failure ( $p = 0.56$ ).

**Conclusions:** CRT improved the course of CHD patients by preserving or improving EF and heart failure symptoms. Therefore, CHD patients with CHF should be timely evaluated for CRT. As eligibility criteria for CRT in the CHD population are not well established, individual factors need to be taken into consideration.

**Keywords:** cardiac resynchronization, chronic heart failure, congenital heart disease, pacing

#### P-034

##### The impact of maternal and perinatal factors on the neonatal electrocardiogram

Caroline Boye Thygesen<sup>1</sup>, Maria Munk Pærregaard<sup>1</sup>, Julie Molin<sup>1</sup>, Lene Friis Eskildsen<sup>2</sup>, Anne Sophie Sillesen<sup>1</sup>, Ruth Ottilia Birgitta Vøgg<sup>1</sup>, Anna Axelsson Rajá<sup>3</sup>, Kasper Karmark Iversen<sup>4</sup>, Henning Bundgaard<sup>5</sup>, Alex Hørby Christensen<sup>6</sup>

<sup>1</sup>Department of Cardiology, Herlev-Gentofte Hospital, Copenhagen University Hospital, Copenhagen, Denmark; <sup>2</sup>Department of Obstetrics and Gynaecology, Amager-Hvidovre Hospital, Copenhagen University Hospital, Copenhagen, Denmark; <sup>3</sup>Department of Cardiology, Herlev-Gentofte Hospital, Copenhagen University Hospital, Copenhagen, Denmark; <sup>4</sup>The Capital Regions Unit for Inherited Cardiac Diseases, Department of Cardiology, The Heart Center, Rigshospitalet, Copenhagen University Hospital, Copenhagen, Denmark; <sup>5</sup>Department of Clinical Medicine, University of Copenhagen, Denmark; <sup>6</sup>The Capital Regions Unit for Inherited Cardiac Diseases, Department of Cardiology, The Heart Center, Rigshospitalet, Copenhagen University Hospital, Copenhagen, Denmark; <sup>7</sup>Department of Clinical Medicine, University of Copenhagen, Denmark; <sup>8</sup>Department of Cardiology, Herlev-Gentofte Hospital, Copenhagen University Hospital, Copenhagen, Denmark; <sup>9</sup>The Capital Regions Unit for Inherited Cardiac Diseases, Department of Cardiology, The Heart Center, Rigshospitalet, Copenhagen University Hospital, Copenhagen, Denmark; <sup>10</sup>Department of Clinical Medicine, University of Copenhagen, Denmark

**Background and Aim:** Myocardial development is ongoing at the time of birth, and knowledge on the potential effects of maternal and perinatal factors on the neonatal cardiac conduction system, is

sparse. The aim of this study was to investigate the impact of maternal and perinatal factors on the neonatal electrocardiogram in a large, unselected cohort of neonates.

**Method:** In a multicentre, prospective, population-based cohort study, neonates underwent cardiac evaluation with electrocardiograms and echocardiograms during the first month of life. Associations between medical and demographic data, pregnancy, and birth-related factors and electrocardiographic parameters were assessed.

**Results:** A total of 15,928 singletons with normal echocardiograms were included (52% boys; median age at examination 11 days). The neonates were divided into groups by accumulated number of maternal and perinatal factors: 0 ( $n = 1, 412$ ), 1 ( $n = 3, 199$ ), 2 ( $n = 3, 319$ ), 3 ( $n = 3, 044$ ), 4 ( $n = 2, 462$ ), and  $\geq 5$  ( $n = 2, 492$ ), and differences in ECG parameters between groups were analysed. We observed an additive effect with leftward shift in the QRS axis and prolongation of the QT interval (all  $p < 0.01$ ) depending on the number of maternal and perinatal factors. Comparing extreme groups (0 vs.  $\geq 5$  maternal/perinatal factors) we found a 4.3% more left-shifted QRS axis ( $117$  vs.  $112^\circ$ ,  $p < 0.001$ ) and a 0.8% prolonged QTcFridericia (QTcF;  $363$  vs.  $366$  ms,  $p < 0.001$ ); the effect on QTcF was more pronounced in neonates examined in the first week of life ( $360$  vs.  $368$  ms,  $p < 0.0001$ ).

**Conclusions:** We observed a cumulative effect of maternal and perinatal factors on neonatal electrocardiographic parameters, including a significantly more left-shifted QRS axis and increased duration of the QT interval. The effects were most pronounced during the first week of life. Our findings add to the knowledge on the neonatal cardiac transition and may guide clinical management in the neonatal phase.

**Keywords:** Neonates, electrocardiography, maternal factors, perinatal factors, transition, Copenhagen Baby Heart Study

#### P-035

##### Long-term outcomes of patients with implantable cardioverter-defibrillators implanted in childhood: transvenous vs. nontransvenous system

Terezia Tavačová<sup>1</sup>, Peter Kubuš<sup>1</sup>, Alice Krebsová<sup>2</sup>, Jan Janoušek<sup>1</sup>

<sup>1</sup>Children's Heart Centre, 2nd Faculty of Medicine, Charles University and Motol University Hospital, Prague, Czech Republic; <sup>2</sup>Cardiology Department, Institute for Clinical and Experimental Medicine, Prague, Czech Republic

**Background and Aim:** Implantable cardioverter-defibrillators (ICD) have been established as an effective and safe therapy for prevention of sudden cardiac death in children. Controversy regarding ICD type (transvenous, T vs. non-transvenous, NT) led us to perform long-term analysis of outcomes in these two groups.

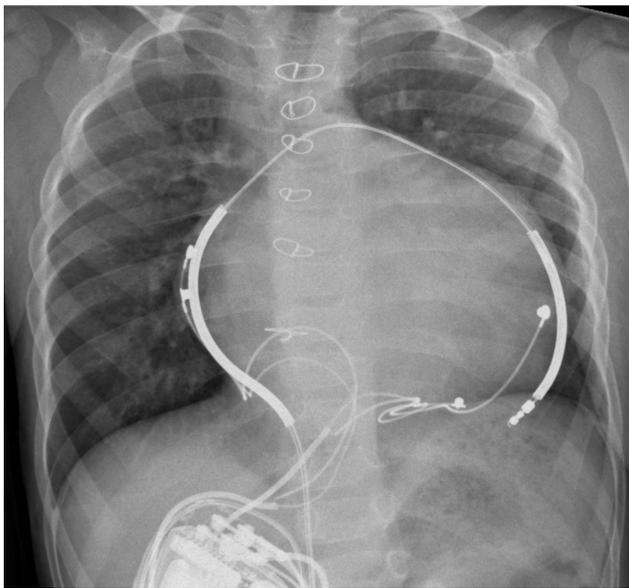
**Method:** A nationwide cohort of all patients ( $N = 109$ , male 73, 67.0%) with ICDs implanted during childhood from 1993-2022 at median (IQR) age 14.3(10.7-16.6) years was retrospectively studied. Data were retrieved from the institutional databases and medical records and cross-mapped with the National Death Registry. Patients were followed-up for a median (IQR) of 62.2(27.1-127.3) months.

**Results:** 94 patients received T and 15 NT systems (pericardial coil in 11, see Figure, pleural coil in 1 and subcutaneous coil in 3 patients). Totally subcutaneous ICD systems were not included. Patients with NT ICDs were significantly younger (median age 4.6 vs. 15.3 years,  $p < 0.001$ ) and smaller (median weight 17.0 vs. 58.6 kg,  $p < 0.001$ ) at implant. Median follow-up was comparable with 67.4 months in NT and 61.0 months in T group,  $p = 0.843$ . There were 6 deaths (5.5%), all in patients with T systems,

yielding a 5/10 years survival probability of 93.8/91.5%. Five years after ICD implantation freedom from appropriate therapy was 56.3/60.3% ( $p = 0.886$ ) and from inappropriate therapy 85.7/88.8% ( $p = 0.751$ ) in NT vs. T systems, respectively. A total of 37 surgical revisions for ICD related complications (except ERI enforced generator replacement) had to be performed 32 patients (29.4%). None of the revisions in the NT group was associated with shock coil malfunction or strangulation of the heart in pericardial coil systems. Five years after implantation freedom from surgical revision in NT vs. T group was 72.7/73.7%, resp. ( $p = 0.961$ ).

**Conclusions:** NT ICDs are as effective and safe as T ICDs in treatment of malignant arrhythmias in children. The burden of surgical revisions is comparable.

**Keywords:** implantable cardioverter-defibrillator, nontransvenous, transvenous, pediatric, arrhythmia



**Figure** Nontransvenous pericardial ICD system

**P-036**

**Perinatal malignant arrhythmias in timothy syndrome - updates from an international cohort**

Alexandra Matthews<sup>1</sup>, Katherine Timothy<sup>2</sup>, Andy Golden<sup>3</sup>, Cecilia Gonzalez Corcia<sup>4</sup>

<sup>1</sup>Department of Paediatric Cardiology, Bristol Royal Hospital for Children, UK; <sup>2</sup>Timothy Syndrome Foundation, Ellicott City, USA; <sup>3</sup>National Institute of Diabetes and Digestive and Kidney Diseases, NIH, Bethesda, USA; <sup>4</sup>Department of Paediatric Cardiology, CHU Sainte-Justine, Montreal, Canada

**Background and Aim:** Timothy syndrome (TS) is an extremely rare, multisystem disorder. It is classically associated with long QT (LQT), syndactyly, and life-threatening ventricular arrhythmias. TS arises from a de novo mutation in exon 8A (type 1) and exon 8 (type 2) of the CACNA1C gene. Rhythm abnormalities in TS may appear as early as fetal life. Diagnosis in the neonatal period allows maximal medical and device therapy to be implemented early to avoid malignant arrhythmias and sudden cardiac death.

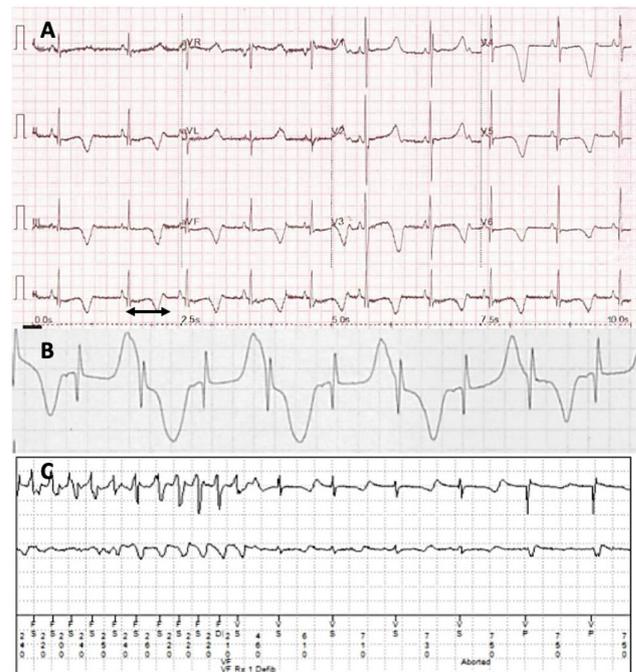
**Method:** Retrospective cohort study using data from the Timothy Syndrome Foundation, a foundation for TS patients around the world. All patients included have confirmed type 1 Timothy syndrome (TS1) (CACNA1C, G406R in exon 8A).

**Results:** Forty-four cases of TS1 were identified (26 male; 60%) over the past 28 years. The mean gestational age was 35.7 weeks (range 28 weeks - term). Eighteen patients (41%) were born prematurely (<37 weeks gestation). Fetal bradycardia secondary to atrioventricular block (AVB) presented in 17 patients (39%) and prompted the premature delivery in 12 patients (27%). In the fetal bradycardic patients, only 8 (47%) were appropriately diagnosed with AVB and had a mean gestation age of 34.4 weeks compared to 37 weeks in patients without fetal bradycardia ( $p < 0.05$ ). At birth LQT was identified in 25 patients (57%). From this group, 12 patients (48%) were diagnosed with TS1 in the neonatal period, compared with 3 patients (7%) from the group in which LQT was not identified at birth ( $p < 0.05$ ). Of the 18 deceased patients, 7 patients (39%) died in the first year of life, with an average age at death of 2.5 months. Eight patients died aged between 23 months and 6 years. The age of death was unknown for 3 patients.

**Conclusions:** Fetal bradycardia is an unusual but high-risk manifestation of the extremely prolonged QT in TS1. Fetal AVB is infrequently diagnosed as the cause of bradycardia in the TS population, resulting in a high incidence of premature delivery. TS1 is associated with a high early mortality yet most patients are diagnosed beyond the neonatal period. Neonatal detection of TS1 allows early intervention to prevent life-threatening arrhythmias.

**Keywords:** Fetal arrhythmia, Long QT, Bradycardia

**ECG recordings of arrhythmias in Timothy Syndrome**



**Figure 2:** ECG recordings of arrhythmias seen in TS. Panel A: 12-lead ECG showing extreme QT and QTc prolongation (black arrow: 600ms and 577 ms, respectively) in a TS1 patient treated with Nadolol and Mexilitine. Panel B: Holter monitoring showing a biphasic T wave alternans pattern in a patient with TS1. Panel C: ICD download of an episode of self-terminating VF in a patient with TS1.

ECG recordings demonstrating long QT, T wave alternans and ventricular fibrillation in patients with Timothy Syndrome.

## P-037

**Real world prevalence of cardiac channelopathies in a paediatric tertiary care center**

Manfred Marx<sup>1</sup>, Sulaima Albinni<sup>1</sup>, Julian Heno<sup>1</sup>, Marianne Guechenberger<sup>2</sup>, Ina Michel Behnke<sup>1</sup>

<sup>1</sup>Medical University of Vienna, Department of Paediatrics and Adolescent medicine, Paediatric Heart Centre; <sup>2</sup>Medical University of Vienna, Department of Cardiology, AKH-Vienna

**Background and Aim:** Inherited arrhythmogenic diseases (IADs) such as Long QT Syndrome (LQTS), Brugada Syndrome (BrS), Arrhythmogenic Rightventricular Cardiomyopathy (ARVC) Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) and cardiac Laminopathies are rare genetic conditions with the risk of arrhythmogenic sudden cardiac death as the first symptom. Early diagnosis followed by appropriate treatment can considerably decrease mortality rate. However, we speculate that the “Real World” numbers of paediatric patients with IADs are significantly lower than previously published prevalence data.

**Method:** In this single centre observational study, we reviewed all patients with IADs at our outpatient clinic for cardiac arrhythmias for the year 2021. Assuming that our paediatric outpatient clinic covers all patients aged from 0-18 years and that every individual patient is checked at least once a year, one single year of observation should include all patients with the above mentioned IADs within all age groups between zero and 18 years. Prevalence for each IAD were calculated for the catchment area.

**Results:** Among 676418 persons aged 0 – 18y in the region of Eastern Austria, a total of 86 (45m, 41f) patients with IADs were followed in the year 2021. The overall prevalence for IADs was 1: 7865. “Real World” prevalence in comparison to the literature is listed in figure 1.

**Conclusions:** There is a significant gap between the number of patients with IADs being treated at our paediatric outpatient clinic and the expected number of cases in the population of the catchment area according to published literature data. Active multidisciplinary approaches with intensive collaboration of paediatric and adult cardiologists combined with elected screening programs are warranted to detect patients and screen family members.

**Keywords:** epidemiology, arrhythmias, sudden cardiac death, children

Observation period 1.1. 2021 - 1.1. 2022 Eastern Austria			
		“Real world” prevalence	Literature prevalence
LQTS 1-13	44	1: 15373	1: 2000
LQTS 1	24	1: 28184	--
LQTS 2	13	1: 52032	
LQTS 3	4	1: 169104	
BrS	24	1: 28184	1: 5000 - 2000
ARVC	11	1: 61493	1: 5000 - 2000
CPVT	4	1: 169104	1: 10000
Lamin A/C	3	1: 225472	Extremely rare

**Figure 1** Real World prevalence versus published data

## P-039

**Transvenous lead advancement in paediatric pacing to overcome growth stretching**

Luìgina Porco<sup>1</sup>, Massimo Stefano Silveti<sup>1</sup>, Marianna Cicienia<sup>1</sup>, Ilaria Tamburri<sup>1</sup>, Giacomo Silveti<sup>2</sup>, Marta Campisi<sup>1</sup>, Fabio Anselmo Saputo<sup>1</sup>, Virginia Battista<sup>1</sup>, Fabrizio Drago<sup>1</sup>

<sup>1</sup>Pediatric Cardiology and Cardiac Arrhythmias Unit, Department of Pediatric Cardiology and Cardiac Surgery, Bambino Gesù Children’s Hospital, IRCCS, Piazza Sant’Onofrio 4 – 00165, Rome, Italy. European Reference Network for Rare and Low Prevalence Complex Disease of the Heart (ERN GUARD-Heart); <sup>2</sup>Sapienza University of Rome, Piazzale Aldo Moro, 5, 00185 Rome, Italy

**Background and Aim:** one of the main complications of transvenous leads implanted in paediatric patients is the stretching of the lead caused by somatic growth. It may cause pacing and sensing defects and lead dislodgement or even fracture. Absorbable lead ligature and atrial loop have been proposed to prevent this risk. However, the loop may induce traction, dislodge, and may unroll too early and impair lead or cardiac function. Lead advancement through pushing it from the pocket may solve growth-induced traction and spare the electrode throughout childhood until post-puberty. Objective of the study is the retrospective analysis of the outcome of the transvenous lead advancement in children with a pacemaker (PM) in a single tertiary paediatric center.

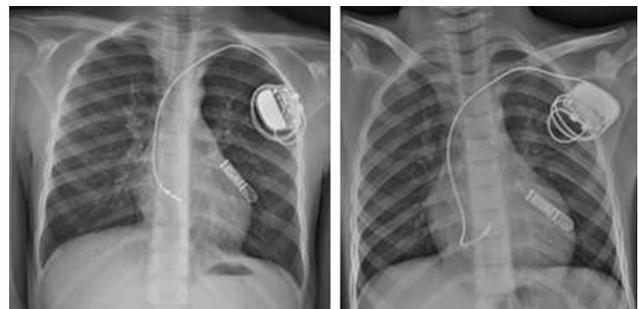
**Method:** consecutive patients with a VVIR PM implanted for isolated congenital complete atrioventricular block (no structural heart disease) in alternative right ventricular (RV) pacing sites, with lead stretching underwent a trial of lead advancement during general anaesthesia, cefuroxime antibiotic prophylaxis, from 2014 to 2021. After venous angiography showed venous patency, the PM pocket was opened, the lead was released from subcutaneous adhesences and with a stylet was gently advanced to create a semi-loop in the atrium without dislodging the tip. Lead data (threshold, sensing, impedance) were compared before and after the procedure. Data are expressed as median (25th-75th centiles).

**Results:** 12 patients underwent PM implantation at 6.8 (5.8-8.0) years of age, 20 (18-21) kg, 116 (106-120) cm, with the lead positioned at parahisian(3) or mid-septum (5 patients) sites. During a follow-up of 3 (1-5) years, advancement procedures were 1.5 (1-4) per patient. Delta between procedures was: age 16 (12-20) months, height 8 (7-11) cm and weight 5 (3-7) kg. All leads were successfully advanced without any procedural complications. Procedure time (skin to skin) was 96 (73-108) minutes, fluoroscopy was 0.5 (0.2-1.4) mGy, 16 (10-46) microGy/m2. Electrical lead parameters did not showed significant differences between consecutive control times.

**Conclusions:** the advancement of transvenous leads in children is a safe and effective procedure, with low fluoroscopy exposure, without significant acute and chronic complications. This procedure seems to preserve transvenous leads until growth has completed.

**Keywords:** Cardiac Pacing, Congenital Atrioventricular Block, Endocardial Lead, Pacing Complication, Pacemaker, Pediatrics

## A case of lead advancement



On the left, lead stretched due to growth (compared to the time of implantation patient 10 cm higher); on the right, the result of the lead advancement

## P-040

**Icd outcome in pediatric arrhythmogenic cardiomyopathy: A single center experience**

Marianna Cicien<sup>1</sup>, Massimo Stefano Silveti<sup>1</sup>, Fabio Anselmo Saputo<sup>1</sup>, Fabrizio Gimigliano<sup>1</sup>, Nicoletta Cantarutti<sup>1</sup>, Rachele Adoriso<sup>2</sup>, Anwar Baban<sup>1</sup>, Fabrizio Drago<sup>1</sup>

<sup>1</sup>Pediatric Cardiology and Arrhythmia/Syncope Complex Unit, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy European Reference Network for Rare and Low Prevalence Complex Disease of the Heart (ERN GUARD-Heart).; <sup>2</sup>Heart Failure and Transplant, Mechanical Circulatory Support Complex Unit, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy

**Background and Aim:** Arrhythmogenic cardiomyopathy (ACM) is a very rare condition among pediatric patients. Once diagnosed, the risk stratification is mandatory in order to prevent sudden cardiac death (SCD) which can sometimes be the first manifestation of the disease as well. Moreover, it must be periodically reassessed due to the progression of the disease.

**Method:** In this is a single center retrospective observational study, all pediatric patients (<18 y.o.) diagnosed with ACM since 2009 in Our Institution were collected. Only the patients with a confirmed diagnosis according to the "Padua Criteria" were enrolled. Implantable cardioverter defibrillator (ICD) implantation was decided according to the current recommendations/clinical studies, and outcome was recorded during follow-up.

**Results:** Twenty-two patients (age range 10 -17 y.o.), had a "definite" (21) or "borderline (1) ACM diagnosis (13 females) according to the "2020 Padua Criteria". Out of them, 16 patients (72.7%) received an ICD: 3 (18.8%) a transvenous device and 13 (81.2%) a subcutaneous one (S-ICD). In 4 (25%) patients, ICD was implanted for secondary prevention (2 hemodynamically unstable sustained ventricular tachycardia (VT) and 2 hemodynamically stable sustained VT). In the other 12 (75%) patients, it was implanted for primary prevention: in 2 with class I indication (severe right ventricular dysfunction), in 8 with class IIa indication (syncope, non-sustained VT, moderate ventricular dysfunction) and in 2 with class IIb due to several "minor" risk factors.

During the follow-up ( $5.59 \pm 3.4$  years), appropriate ICD therapies occurred in 3 (18.8%) patients for sustained VTs, without defibrillation failures. Inappropriate shocks occurred in 2 cases: one for T-oversensing and one for high-rate sinus tachycardia exceeding the VT limit.

**Conclusions:** ICD is indicated and effective in the majority of pediatric patients due to the high risk of SCD. Despite that, appropriate ICD therapies occurred in only a minority of patients. The rate of inappropriate shocks is even more rare. This reassuring follow-up may be due to the efficacy of medical antiarrhythmic and anti-remodeling therapy, promptly started once the diagnosed is made. Certainly, other risk and protective factors and protective ones remain still unknown and a specific pediatric risk stratification model is advised in the future.

**Keywords:** arrhythmogenic cardiomyopathy, sudden cardiac death, implanted cardioverter defibrillator, pediatric patients

## P-041

**A rare complication of paediatric pm and icd: intracardiac lead thrombosis**

Marie Laure Yammine, Massimo Stefano Silveti, Fabrizio Gimigliano, Gabriele Rinelli, Marta Campisi, Virginia Battista, Iliaria Tamburri, Giacomo Silveti, Fabio Anselmo Saputo, Fabrizio Drago  
Pediatric Cardiology and Cardiac Arrhythmias Unit, Department of Pediatric Cardiology and Cardiac Surgery, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy

**Background and Aim:** Implantable electronic devices (IEDs) are increasingly used in children and young adults with specific cardiac rhythm disorders. However, both pacemakers (PMs) and implantable cardioverter defibrillators (ICDs) may have several downsides. Literature lacks of data concerning leads thrombosis, thus we analyzed pacing lead thrombosis' occurrence in our PMK/ICD patients.

**Method:** In this single center study, we retrospectively collected clinical details of children or young adults with IEDs and evidence of pacing lead thrombosis. Data are reported as median (interquartile range).

**Results:** Since 2015, five patients (2 males) aged 16 (12-22) years showed transthoracic echocardiographic evidence of lead vegetations during routine follow-up after 5 (1-16) years from lead's initial implantation. Age at device implantation was 9 (5-12) years. Two patients had dual-chamber ICD, 3 had PMs (2 DDD, 1 VVI). No patient had any early wound complication after implantation. At the time of thrombosis detection, no patient reported any recent fever or symptoms of clinical illness. Devices and leads were normally functioning in all patients. Blood examination revealed absence of infectious or inflammatory signs, and all patients had negative hemocultures for aerobic, anaerobic bacteria and fungi. Therefore, lead thrombosis was diagnosed. Transesophageal echocardiogram (TEE) confirmed the echo finding. Thrombophilia testing revealed that 4 out of 5 patients had methylenetetrahydrofolate reductase (MTHFR) mutation, homozygous and heterozygous (2 each). Patients received oral anticoagulation therapy (OAT) and TEE showed complete recovery in all patients after 6-12 months.

**Conclusions:** Pacing lead thrombosis is a rare complication in paediatric/young adult patients with PMK/ICD, well treated with OAT. MTHFR mutation is a risk factor for this complication.

**Keywords:** Lead thrombosis, pacemakers, implantable cardioverter defibrillators

## P-043

**Case report of smart wearable technology in paediatric cardiology**

Arveen Devindaran, Victor Ofje, Amos Wong  
Department of Paediatric Cardiology, University Hospital of Wales, Cardiff, Wales, UK

**Background and Aim:** In the past decade many new wearable devices have emerged by big consumer technologies,

including several that have been widely adapted by the general public and clinicians. Commercially available devices are able to collect multitude of data including heart rate, heart rhythm, saturations, BP, activity and, much more. We published a case report documenting the usage of such device as a diagnostic tool which lead to appropriate management.

**Method:** Retrospective data interpretation of the electrophysiology report from the smart wearable was done and significant data was collected and sufficient evidence was gathered to proceed with the treatment for the patient.

**Results:** 9-year-old male presented with chest pains and palpitations for several years. All investigations including multiple ECGs, prolonged Holter monitoring, exercise stress test and echocardiogram were normal. The inconclusive investigations did not justify medical/surgical treatment despite ongoing symptoms. Patient was given a Withings Move ECG watch to monitor cardiac rhythm during palpitations. Patient managed to document a wide complex rhythm at a rate of 300bpm. Therefore, a diagnosis of ventricular fibrillation was made and the patient has now been listed for an electrophysiology study and ablation.

**Conclusions:** There are benefits and disadvantages of non-medical grade smart wearable devices. Patient selection criteria is important for using such devices. Typically, its useful for patients with intermittent symptoms over a long time. However, there are minimal studies to compare them against medical grade devices. Therefore, clinical studies must be done to validate the data and accuracy of such technology. One must also be careful in interpreting results from non-medical grade devices. As more advancement happens in this area, we strongly believe smart wearable technology has an important role to play in the future.

**Keywords:** smart wearables, palpitations, remote monitoring

Rhythm strip from smart wearable device



#### P-044

##### Phenotypic heterogeneity in a family with *teclr*-related cpvt type 3: 1st case in kuwait

Mohammad Ebrahim<sup>1</sup>, Ali A M A A M Alkhabbaz<sup>2</sup>, Mohammad Metwally<sup>3</sup>, Buthaina Bash<sup>4</sup>, Ali Al Sayegh<sup>5</sup>, Jerome Philip Saul<sup>6</sup>  
<sup>1</sup>Department of Pediatrics, Kuwait University Faculty of Medicine, Kuwait; <sup>2</sup>Affiliated with Chest Diseases Hospital, Kuwait; <sup>3</sup>Kuwait University Faculty of Medicine, Kuwait; <sup>4</sup>Department of Pediatric Cardiology, Chest Diseases Hospital, Kuwait; <sup>5</sup>Department of Genetics, Ganima Al Ganem Center, Kuwait; <sup>6</sup>Department of Cardiology, Chest Diseases Hospital, Kuwait; <sup>6</sup>Department of Pediatrics, West Virginia University, West Virginia, USA

**Background and Aim:** This is not applicable.

**Method:** An 11-year-old, previously healthy boy, collapsed suddenly while walking casually with family. He was found pulseless, requiring CPR and defibrillation therapy to convert from presumed polymorphic ventricular tachycardia (PMVT) into sinus rhythm with prolonged QT interval (~600 msec) and unstable repolarization. He was briefly started on dobutamine before developing 2 episodes of PMVT despite being on Lidocaine infusion. Thereafter, he was switched to non-selective short-acting beta blockers (due to resting bradycardia) without further VT. He eventually was transitioned to Nadolol (~0.8 mg/kg). He had frequent polymorphic/bidirectional ventricular complex ectopy mainly during stressful periods (e.g., MRI). He has no apparent neurological sequelae.

During his hospital stay, he had a normal echocardiogram and cardiac MRI. Moreover, he underwent electrophysiology study (EPS) with easily inducible unstable PMVT requiring defibrillation therapy. Furthermore, he underwent arrhythmia panel testing through commercial laboratory (Invitae, San Francisco, USA), which was negative. A transvenous dual chamber ICD, was placed prior to discharge. Whole Exome Sequencing (WES) eventually confirmed the high clinical suspicion for homozygous *TECL1* (c.331+1G>A) through another commercial laboratory (Centogene, Rostock, Germany).

He is a product of consanguineous marriage with no family history of concern. Both parents and the rest of the three siblings' electrocardiograms (ECG) were normal. His oldest brother's phenotype

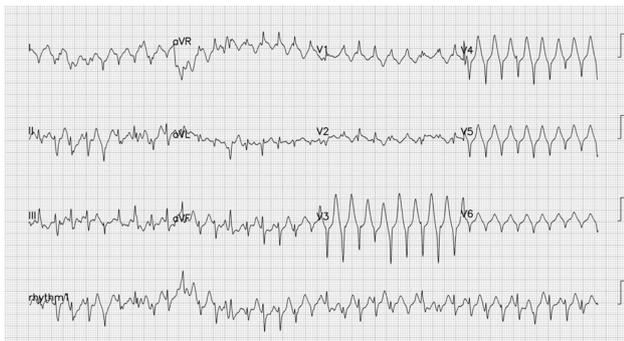
was highly suggestive of CPVT during his screening sprint exercise stress test (EST), however, without QT prolongation. He was confirmed for the same homozygous mutation. Otherwise, segregation studies through Sanger sequencing confirmed the heterozygosity status for both parents and one sibling.

After discharge, Flecainide was added to his regimen with great response in subsequent EST.

**Results:** This part (the discussion) will be submitted in another submission as the total word count of the Case Report is nearly 550 words. The organizing committee member Ms. Lara Volkan instructed as such.

**Conclusions:** I hereby confirm that the consent of the relevant patients has been obtained to submit this Case Report abstract.

**Keywords:** TECRL, CPVT, Kuwait



**Figure 1** Ventricular tachycardia (narrow)

#### P-046

##### **Conduction system pacing guided by 3D mapping in paediatric patients with congenitally corrected transposition of the great arteries.**

Carla Favocchia, Massimo Stefano Silvetti, Fabio Anselmo Saputo, Ilaria Tamburri, Chiara Mizzon, Marta Campisi, Fabrizio Gimigliano, Gabriele Rinelli, Giacomo Silvetti, Lucilla Ravà, Fabrizio Drago  
Paediatric Cardiology and Cardiac Arrhythmia/Syncope Unit and Epidemiology Institute, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy

**Background and Aim:** In congenitally corrected transposition of the great arteries (CCTGA) the right ventricle (RV) is systemic. Atrioventricular block (AVB) and systolic dysfunction are frequently observed. Permanent pacing of the subpulmonary left ventricle (LV) may worsen RV dysfunction. Aim of this study was to seek out if LV conduction system pacing (LVCSP) guided

by three-dimensional-electroanatomic mapping systems (3D-EAM) can preserve RV systolic function in CCTGA patients with AVB.

**Method:** Retrospective analysis of paediatric patients with CCTGA who underwent 3D-EAM-guided LVCSP. 3D-pacing map guided lead implantation toward septal sites with narrower paced QRS close to conduction system. Electrocardiograms, echocardiograms, and lead parameters were compared at baseline (pre-implantation) and at 1-year follow-up. RV function was evaluated by 3D ejection fraction (EF), fractional area change (FAC), RV global longitudinal strain (GLS). Data are reported as median (25th-75th centiles).

**Results:** Seven CCTGA patients aged 15 (9-17) years, with complete/advanced AVB (4 with prior epicardial pacing including 2 biventricular pacing), underwent 3D-guided LVCSP (5 DDD, 2 VVIR). Baseline echocardiographic parameters were impaired in most patients. No acute/chronic complications occurred. Ventricular pacing was >90%. At 1-year follow-up QRS duration showed no significant changes compared to baseline: 120 (80-130) ms vs. 120 (100-125) ms ( $p = 0.9$ ). However, QRS duration shortened in comparison with prior epicardial pacing. Lead parameters remained good despite ventricular threshold increased. Systemic RV function was preserved: FAC and GLS improved significantly [33 (30-43)% and -13 (-14 -11)% vs. 40 (40-45)%,  $p = 0.04$ , and -17 (-20-15)%,  $p = 0.03$ , respectively], and all patients showed normal RVEF (>45%).

**Conclusions:** 3D-EAM-guided CSP preserved RV systolic function in paediatric patients with CCTGA and AV block after short-term follow-up.

**Keywords:** Congenitally corrected transposition of the great arteries, cardiac pacing, ventricular systolic function, paediatric age, radiation exposure, non-fluoroscopic mapping system.

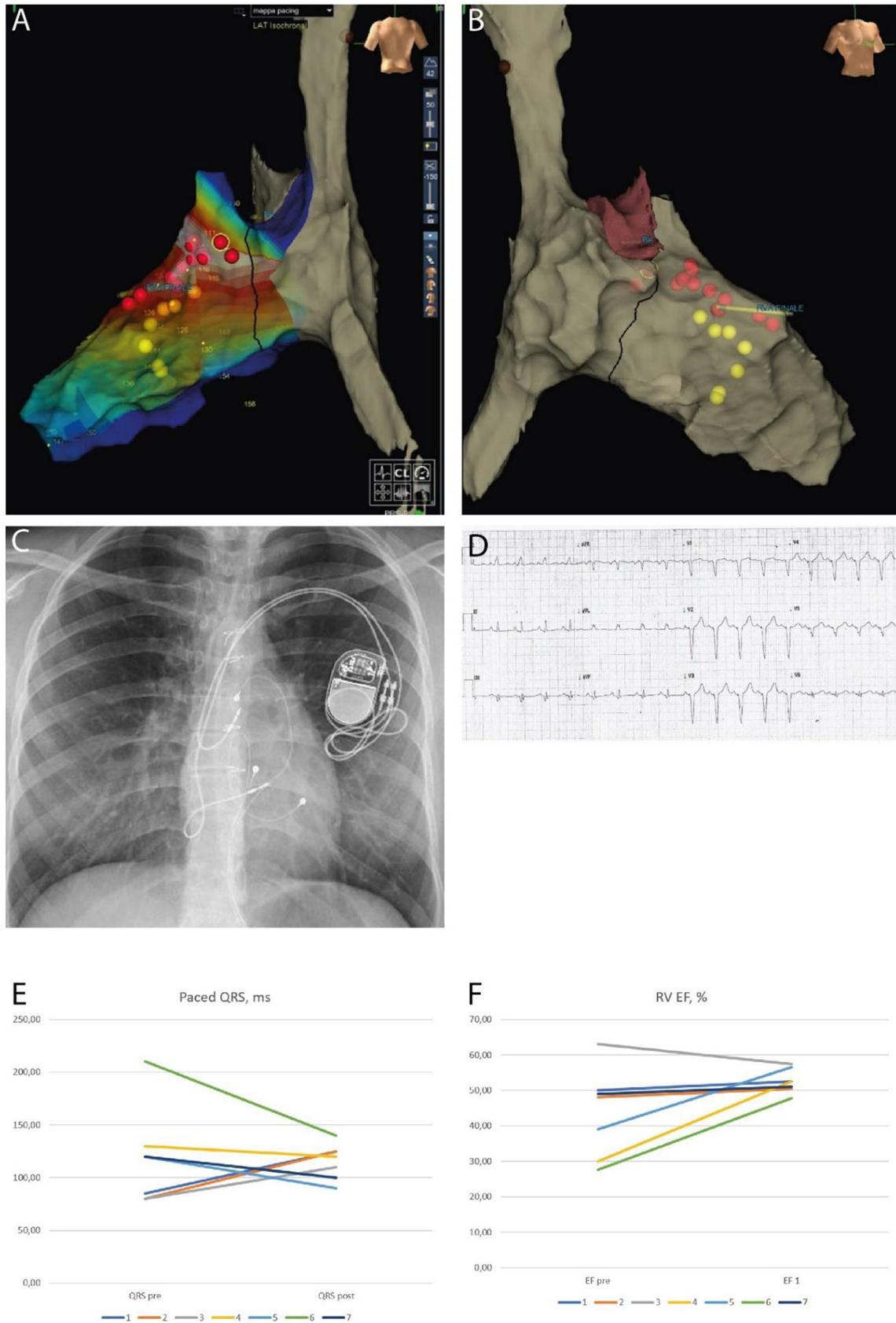
#### P-047

##### **Prevalence, characteristics and natural history of wolff-parkinson-white syndrome in neonates**

Maria Munk Perregaard<sup>1</sup>, Joachim Haartmann<sup>1</sup>, Christian Pihl<sup>1</sup>, Jesper Kock<sup>1</sup>, Adrian Pietersen<sup>1</sup>, Kasper Karmark Iversen<sup>1</sup>, Henning Bundgaard<sup>2</sup>, Alex Hørbj Christensen<sup>1</sup>

<sup>1</sup>Department of Cardiology, Herlev-Gentofte Hospital, Copenhagen University Hospital, Copenhagen, Denmark.; <sup>2</sup>The Capital Regions Unit for Inherited Cardiac Diseases, Department of Cardiology, The Heart Center, Rigshospitalet, Copenhagen University Hospital, Copenhagen, Denmark.

**Background and Aim:** Wolff-Parkinson-White (WPW) syndrome is characterized by an accessory electrical pathway between the atria and ventricles. Clinically, the condition is associated with congenital heart disease, supraventricular tachycardia and sudden cardiac death. Our aims were to determine the prevalence of WPW syndrome in an unselected neonatal population, describe



**Figure 1** 1A-B: LV conduction system in CCTGA and 3D-EAM pacing map. C: Chest X-Ray. D: ECG post-3D-EAM guided LV pacing E-F: Graphs of ECG and echo changes.

electrocardiographic and echocardiographic characteristics, and the natural history during childhood.

**Method:** Electrocardiograms (ECGs) and echocardiograms of neonates (aged 0-30 days) from a large, prospective, general population study were included. WPW cases were identified through manual evaluation of outliers in PR-interval, QRS-duration, and QRS axis. Neonates with confirmed pre-excitation on their initial ECG were offered cardiac follow-up. Localization of the accessory pathway was assessed by different algorithms. Cases were matched 1:4 to controls by age, sex, weight, and gestational age. **Results:** Among the 17, 489 ECGs, we identified 17 (76% boys) neonates with WPW syndrome consistent with a prevalence of 0.1%. The neonatal heart rate, PR-interval, QRS-duration, QTc(Bazett), maximum amplitudes in R-V1 and S-V6 were significantly different between cases and controls (all  $p < 0.05$ ), whereas the QRS axis, max amplitude in S-V1 and R-V6 did not differ (all  $p > 0.05$ ). The accessory pathway was primarily predicted to the left side of the heart (59-100%, depending on algorithm applied). Echocardiographic measurements of the neonates' left ventricular diameter and function, wall thicknesses, and doppler measurements of trans-mitral- and main pulmonary artery blood flow were similar for cases and controls (all  $p > 0.05$ ). One newborn had significant mitral regurgitation while all other newborns had structurally normal hearts; there were no cases of Ebstein's anomaly. At follow-up (available in 14/17 children) the WPW pattern persisted in four children, while the ECG had normalized in the ten remaining children.

**Conclusions:** The prevalence of WPW syndrome in our cohort of asymptomatic neonates was 0.1%. The syndrome was more frequent in boys, generally not associated with structural heart disease, and the accessory pathway was mostly left-sided. A striking

observation was, that the WPW pattern in most children could not be reproduced on follow-up ECGs, suggesting either that the ECG pattern primarily is intermittent, or that normalization occurs.

**Keywords:** Wolff-Parkinson-White, electrocardiography, neonates, supraventricular tachycardia

**P-048**

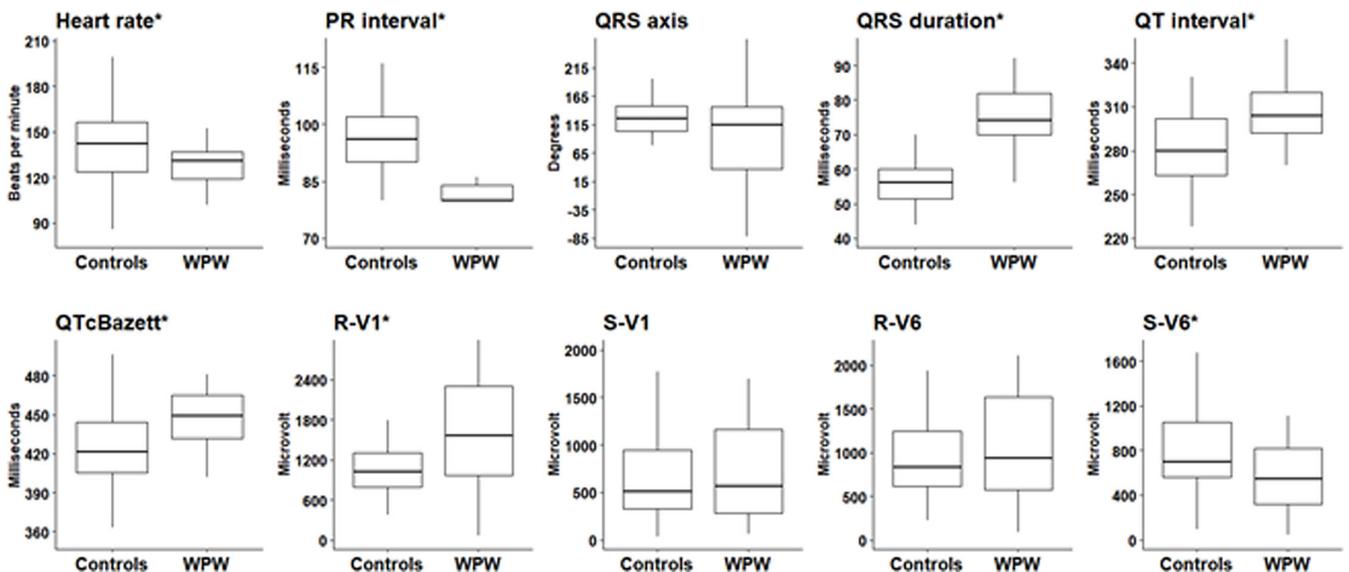
**Diagnostic approach to the paediatric brugada syndrome: A survey of international common practice**

*Christine Stratmann, Maria Cecilia Gonzalez Corcia  
Department of Paediatric Cardiology, Bristol Royal Hospital for Children, Bristol, United Kingdom*

**Background and Aim:** The Brugada syndrome (BrS) is a rare inherited disease that predisposes to cardiac arrhythmias and sudden cardiac death. Up to this date diagnostic criteria, clinical markers of risk and the prognostic value of different underlying or associated genetic variants remain heavily discussed in the scientific community. Due to a highly variable genotype-phenotype correlation, individual risk stratification is challenging, and the diagnostic and therapeutic approach has varied significantly in the past.

**Method:** At present, there is no universal recommendation for screening and diagnosis of the largely asymptomatic paediatric population. With the aim to create an up-to-date picture of the international common practice regarding the diagnostic approach to the BrS in the young, we have invited the 36 centres participating in the International Paediatric Brugada Syndrome Registry in 2021 to complete a questionnaire.

Boxplot - WPW vs. Controls



Box plots of ECG parameters for neonates with WPW syndrome and matched controls. ECG parameters with significant differences between the groups are marked with \*. The horizontal line in the middle is the median value, box depicts the interquartile range (IQR), and the whiskers 1.5 times the IQR.

**Results:** Twenty-one centers from 12 countries and 4 continents responded to the survey. The results show that there is a more uniform picture of diagnostic tools employed than in previous surveys, with all centres screening with a 12-lead ECG, including optional high V1 and V2 placement (n = 21), alongside with a precise clinical history and physical examination. 62% of sites (n = 13) also perform a cardiac ultrasound and an ambulatory electrocardiographic monitor as part of the initial work-up. The use of drug challenges has increased to 85% (n = 7 of 20). Surprisingly, 100% of the centres use genetic testing as a diagnostic tool during childhood and adolescence. The interpretation and clinical consequences of these result, however, still vary significantly.

**Conclusions:** This review of the present clinical practice around the world in managing BrS in the young reflects the efforts of our community to bring light to the understanding of this disease. Scientific advances have made the diagnostic value of many available examinations increasingly clear, and we see a more homogenous diagnostic strategy than in the past. However, genetic testing has introduced an additional layer of complexity and, whilst widely employed, it is yet not completely understood. Further investigations are needed, especially in view of dealing with the large group of asymptomatic family members and silent carriers of pathogenic mutations.

**Keywords:** arrhythmia, brugada syndrome, sudden cardiac death, screening

#### P-049

##### **Premature ventricular contractions affect the mechanics of the left ventricle during the sinus rhythm**

Radosław Pietrzak, Tomasz Marcin Książczyk, Magda Franke, Bożena Werner

Department of Pediatric Cardiology and General Pediatrics, Medical University of Warsaw

**Background and Aim:** Premature ventricular contractions (PVCs) are frequently observed in children and are considered to be benign. However, this hypothesis is uncertain as observations indicate some associations between PVCs and left ventricular (LV) systolic dysfunction. The aim of this study was to investigate regional and global myocardial mechanics during the sinus rhythm, in patients with frequent PVCs.

**Method:** The study group consisted of 33 PVC children aged 13–17 years with normal systolic LV function (EF Teicholz above 60%). The control group consisted of 30 healthy volunteers. 24-hour-Holter-ECG and Echocardiography were performed. Frequent PVCs were defined as 10% of the arrhythmia burden. The echocardiographical speckle tracking analysis was made from an apical 4-chamber view in 6 segments: basic, medial, and apical in the septum and lateral wall of LV. LV Global longitudinal strain (GLS) and dyssynchrony (measured as the SD of time to peak (TTP) of the regional strains) and dyssynchrony index (DI) defined as a ratio of TTP to aortic valve closure time (ACT) were analyzed. ACT was defined as the time from the beginning of QRS to the begin

of aortic valve closure. DI was calculated for every segment of the LV and dyssynchrony was diagnosed when DI was above 1. GLS, and dyssynchrony, were measured in sinus beat directly after PVC and compared between the study and control group, and patients with and without dyssynchrony.

**Results:** In PVC children vs. controls GLS was  $-18.8\pm 4.8\%$  vs.  $-22.1\pm 3.3\%$ ,  $p < 0.01$  and dyssynchrony was  $61.7\pm 14$  ms vs.  $46.7\pm 7$  ms,  $p < 0.01$ , respectively. In basal septal segment (BSS) dyssynchrony was diagnosed in 12 patients. After comparing GLS and dyssynchrony in PVC patients with and without dyssynchrony in BSS, GLS was  $-17.8\pm 5\%$  vs.  $-20.1\pm 4.3\%$ ,  $p = 0.1$ , and dyssynchrony was  $71.8\pm 18$ ms. vs.  $50.4\pm 5.4$ ,  $p = 0.1$ , respectively. In the remaining segments, the number of patients with dyssynchrony was too low for the analysis (below 6).

**Conclusions:** In patients with frequent PVCs, the sinus beat directly after PVC demonstrates significant mechanical abnormalities. This finding suggests that not only dyssynchronous contraction, but intrinsic perturbations are responsible for mechanical abnormalities observed in PVC patients. Dyssynchrony in BSS does not influence the mechanical function of myocardium in PVC patients.

**Keywords:** premature ventricular contractions, left ventricular mechanics, children

#### P-051

##### **An atypical kawasaki presentation in a two-month-old**

Natalie Abbassi, Ramona Onita

Department of Paediatrics, Colchester General Hospital, Colchester, UK

**Background and Aim:** we present a 2-month-old boy who presented with high temperatures, lethargy and poor feeding during Covid lockdown. He was treated for PIMS-TS/ Kawasaki disease.

**Method:** upon admission he had a full septic screen including a lumbar puncture in view of a rising CRP, and was discharged after 7 days on ambulatory IV ceftriaxone to complete a 10-day course. He was re-admitted only 24 hours later with further temperature spikes and in presumed supraventricular tachycardia with a heart rate of 200–220b/min. He received 3 doses of adenosine up to 400mcg/kg with little sustained effect. He was then loaded with Amiodarone to which he responded and was transferred to PICU in a tertiary cardiology centre. He received a further dose of adenosine and CPR in view of profound bradycardia afterwards. Retrospectively, he had sinus tachycardia with first degree heart block, mimicking SVT. Following PIMS-TS treatment with immunoglobulin and steroids, the 1st degree heart block improved and PR interval normalised.

**Results:** further investigations were completed. A routine echo showed dilated coronary arteries, along with a positive PIMS-TS panel but with negative Covid antibodies, PCR and antibody and negative microbiology in blood and CSF cultures.

**Conclusions:** he was treated with intravenous immunoglobulins, aspirin, and methylprednisolone in view of a PIMS-TS/ Kawasaki picture. He has since been discharged from follow-up. This case showed the unusual presentation of Kawasaki disease/

PIMS-TS in the context of negative Covid screen and the importance of its differential diagnosis.

**Keywords:** Kawasaki, PIMS-TS, tachycardia, Covid

#### P-052

##### **Cardiopulmonary exercise testing in children with structurally normal heart and premature ventricular complexes: the first results of prospective study**

Rita Kunigeliene<sup>1</sup>, Odeta Kinciniene<sup>3</sup>, Vytautas Usonis<sup>1</sup>, Germanas Marinskis<sup>2</sup>

<sup>1</sup>Clinics of Children Diseases, Institute of Clinical Medicine, Vilnius University Faculty of Medicine, Vilnius, Lithuania; <sup>2</sup>Department of Cardiology, Vilnius University Hospital Santaros Clinics, Vilnius Lithuania; <sup>3</sup>Department of Children Diseases, Vilnius University Hospital Santaros Clinics, Vilnius Lithuania

**Background and Aim:** Premature ventricular complexes (PVCs) are frequent in the pediatric age group. Although PVCs are usually considered benign, frequent PVCs can cause ventricular dysfunction and might be the first sign of cardiomyopathy. The aim of the study: to evaluate the role of cardiopulmonary exercise testing factors as a prognostic tool in children with structurally normal hearts and various amounts of PVCs.

**Method:** This is a prospective analysis of 40 children with structurally normal hearts and ventricular PVCs. Patients underwent 24 hours electrocardiogram (24ECG) and cardiopulmonary exercise testing (CPET) on treadmill (BTL Cardiopoint CPET) using modified ramp protocol. Maximal oxygen uptake (VO<sub>2</sub> max), carbon dioxide elimination (VCO<sub>2</sub>), anaerobic threshold (AT), respiratory exchange rate (RER), VE/VO<sub>2</sub> were measured during the CPET. The exercise test was terminated if patients get exhausted, develop symptoms or have abnormal blood pressure results during exercise. According to the amount of PVCs in 24ECG, we divided patients into two groups: ≤10 000 and >10 000 PVCs per 24 hours.

Statistical analysis performed with R. Nominal variables tested for normal distribution with Shapiro-Francia test. Nominal values were presented with average and standart deviation. Welch test used to compare means between normally distributed nominal variables. Fisher exact test used to compare categorical variables. The p value <0, 05 was considered statistically significant.

**Results:** 22 boys and 18 girls 12, 4±3, 3 years old with 12 743±10 000 PVCs per 24 hours. CPET duration was 7, 46±0, 08 minutes. Patients reached VO<sub>2</sub> max 1712±777 ml/min; VCO<sub>2</sub> 1963 ±925ml/min, AT 952, 5±317, 3 ml/min, VE/VCO<sub>2</sub> 44, 3±12, RER 1, 17±0, 21. There were no difference by age, body surface area, maximal load, VCO<sub>2</sub>; AT; RER; VE/VCO<sub>2</sub> between the groups. VO<sub>2</sub> max was significantly higher in ≤10 000 PVCs group (p = 0, 03).

**Conclusions:** VO<sub>2</sub> max seems to be the promising factor for evaluation children with PVCs.

**Keywords:** cardiopulmonary exercise test, children, premature ventricular complexes

#### P-053

##### **WPW syndrome due to an left atrial appendage-to-left ventricular connection: successful ablation in a boy presenting with aborted sudden cardiac death**

Mustafa Gülgün<sup>1</sup>, Stefan Ostermayer<sup>1</sup>, Eric Tietz<sup>2</sup>, Timm Dirrichs<sup>2</sup>, Sevinc Özcan<sup>2</sup>, Christiane Katharina Kuhl<sup>2</sup>, Gunter Kerst<sup>3</sup>

<sup>1</sup>Department of Pediatric Cardiology, RWTH University Hospital, Aachen, Germany; <sup>2</sup>Department of Diagnostic and Interventional

Radiology, RWTH University Hospital, Aachen, Germany;

<sup>3</sup>Department of Pediatric Cardiology, Klinikum Stuttgart, Stuttgart, Germany

**Background and Aim:** Sudden cardiac death (SCD) may be the first clinical manifestation of patients with a Wolff-Parkinson-White (WPW) ECG pattern. Here, we report the electrophysiological findings and successful endocardial catheter ablation of an epicardial accessory pathway (AP) connecting the left atrial appendage (LAA) with the left ventricle in a 12-year-old boy with WPW syndrome after aborted sudden cardiac death.

**Method:** A 12-year-old boy was referred to our hospital after aborted SCD while exercising. He was resuscitated by using an external automated cardiac defibrillator. He had a history of surgical closure of an apical and inlet ventricular septal defect (VSD) during infancy as well as symptomatic manifest WPW syndrome for the last 5 months. During endocardial electrophysiological study, the shortest pre-excited R-R interval during induced atrial fibrillation was 200 ms, indicating a high risk AP. Neither ventricular tachycardia nor fibrillation could be induced by programmed ventricular stimulation. Surface ECG and inducible atrioventricular reciprocating tachycardia (AVRT, cycle length of 315 ms) suggested a left-sided AP. After transseptal puncture, the left sided AP was mapped at the base of the LAA. Using conventional RF energy, the AP could not be eliminated. Computed tomography of the LAA revealed a trilobular morphology (Figure 1A). During the second endocardial catheter ablation procedure using irrigated contact-force controlled RF energy, 4 distinct fibers leading to subtle changes of the endocardial electrograms and the surface ECG were ablated in a stepwise manner leading finally to an abolition of the delta wave (Figure 1B). No ST-segment changes were observed during catheter ablation. At the end of the procedure, coronary artery stenosis was excluded by coronary angiography. In the light of aborted SCD history in the context of WPW syndrome and surgical repair of two VSDs, a subcutaneous ICD was implanted.

**Results:** During follow-up period of 8 months, there was no reappearance of the delta wave in the ECG and the patient remained asymptomatic.

**Conclusions:** In this 12-year old boy, endocardial catheter ablation of an epicardial left atrial appendage-to-left ventricular accessory pathway was feasible by meticulously ablating each of the multiple connecting fibers from within the left atrial appendage after delineating the LAA morphology by CT.

**Keywords:** wpw, accessory pathway, appendage, SCD, children

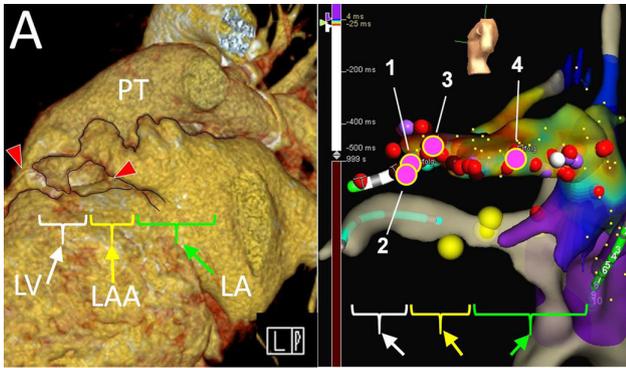


Figure 1 Figure 1A,1B

#### P-054

##### Results of the introduction of the decapolar navigational diagnostic catheter on safety, x-ray exposure and success rate of AVNRT ablation in children

Tomasz Marcin Książczyk, Radosław Pietrzak, Klaudia Obszajczyk, Magda Franke, Bożena Werner  
Department of Pediatric Cardiology and General Pediatrics, Medical University of Warsaw, Warsaw, Poland

**Background and Aim:** The use of the fluoroscopy during electrophysiology procedures in children is directed by the ALARA (As-Low-As-Reasonably-Achievable) rule. The use of non-fluoroscopic mapping systems allowed to significantly reduce the X-ray exposure during ablation procedures. With CARTO 3D-mapping system diagnostic catheters are not visualized until the mapping catheter is introduced, hence the use of the fluoroscopy is required for the positioning of the diagnostic catheters. DECANAV is a navigational decapolar diagnostic catheter that allows to use 3D mapping system and avoid fluoroscopy from the beginning of the procedure. It has been made available in our center from the February 2022 and is now routinely used for SVT ablations.

The aim of this study was to analyze how the use of the DECANV catheter has influenced the X-Ray exposure, safety, success rate and total procedural time of the pediatric atrioventricular nodal reentry tachycardia (AVNRT) ablations.

**Method:** We performed a retrospective analysis of all AVNRT ablations performed in our center using DECANAV catheter (February to October 2022) compared to all AVNRT ablations that were performed using CARTO mapping system but with traditional non-navigational diagnostic catheters (January 2020 to January 2022). In both groups anthropometrical data, fluoroscopy time, radiation dose, total procedure time, acute success and complication rates were evaluated.

**Results:** The studied groups consisted of 14 patients (DECANAV) and 24 (controls). Both groups did not differ significantly with regard to mean age and body weight: 15.1 years vs. 15.2 years and 60.5 kg vs. 60.9 kg in DECANAV and controls, respectively. In the DECANAV, fluoroscopy time was significantly shorter and radiation dose smaller than in the controls: median of 11sec vs. 151sec ( $p < 0.001$ ) and median of 0, 09 mGy/kg vs. 1, 32 mGy/kg ( $p < 0, 001$ ), respectively. Total procedure time: mean of

86min DECANAV group vs 73min in the controls ( $p = 0, 058$ ) and acute procedural success rate: 93% DECANAV vs 92% controls ( $p = 1, 0$ ) did not differ between the groups. There were no complications reported in neither of groups.

**Conclusions:** The use of the navigational decapolar catheter with CARTO mapping system allows to greatly limit the X-Ray exposure without affecting safety, total procedure time, and acute success rate in AVNRT ablation in children.

**Keywords:** AVNRT, ablation, non-fluoroscopic mapping

#### P-056

##### Phenotypic heterogeneity in a family with TECRL-related CPVT type 3: 1st case in kuwait

Mohammad Ebrahim<sup>1</sup>, Ali A M A M Alkhabbaz<sup>2</sup>, Mohammad Metwally<sup>3</sup>, Buthaina Albash<sup>4</sup>, Ali Al Sayegh<sup>5</sup>, Jerome Philip Saul<sup>6</sup>  
<sup>1</sup>Department of Pediatrics, Kuwait University Faculty of Medicine, Kuwait; Affiliated with Chest Diseases Hospital, Kuwait; <sup>2</sup>Kuwait University Faculty of Medicine, Kuwait; <sup>3</sup>Department of Pediatric Cardiology, Chest Diseases Hospital, Kuwait; <sup>4</sup>Department of Genetics, Ganima Al Ganem Center, Kuwait; <sup>5</sup>Department of Cardiology, Chest Diseases Hospital, Kuwait; <sup>6</sup>Department of Pediatrics, West Virginia University, West Virginia, USA

**Background and Aim:** Not applicable

**Method:** Not applicable

**Results:** We report additional evidence of the pathogenicity of the previously described highly conserved TECRL (c.331+1G>A) splice site mutation, at chromosome 7p14-p22 in one other Sudanese family. Similarly, segregation analysis indicated an autosomal recessive form of inheritance. This family had extensive history of sudden cardiac arrest (SCA), mainly during activity and at a very young age. Additionally, the ECGs of the affected members showed mild QTc prolongation and PVCs. This same mutation has been also reported in the setting of oligogenic inheritance in another Tunisian family.

Furthermore, TECRL (c.331+1 >A) variant was not reported as single nucleotide polymorphisms in the general population including in Saudi Arabian individuals. Moreover, functional studies from a symptomatic patient of the same Sudanese family demonstrated increased susceptibility to triggered activity, exaggerated with noradrenaline, mainly due to elevated diastolic intracellular calcium levels.

Luckily, Flecainide therapy had been shown to reduce the triggered activity specifically for TECRL (c.331+1G>A). Additionally, we have noted significant improvement of EST after the addition of Flecainide in the proband. Combination therapy had been instituted for both of our homozygous patients.

Our patient's initial QT intervals and repolarization had been very prolonged and unstable. While this may be related to the initial prolonged cardiac arrest, it took a while for the QT interval to approach upper normal value. This may explain the more severe phenotype of the proband. Also, atrial pacing did seem to stabilize repolarization markedly, similar to previous data for long QT types 1-3. It should be considered for patients with severe long QT phenotype.

Based on our experience, and on what has been reported in the literature, we call for including TECRL in the arrhythmia panels that are commercially available worldwide.

**Conclusions:** Not applicable

**Keywords:** TECRL, CPVT, Kuwait



**Figure 1** The proband experienced this ventricular tachycardia (narrow) a few hours after presentation

### P-057

#### ECG screening: is time to routinely introduce it in apparently healthy schoolchildren?

Stella Maiolo<sup>1</sup>, Same As Above Same As Above<sup>2</sup>, Daniela Righi<sup>1</sup>, Federica Cali<sup>1</sup>, Simone Paglia<sup>1</sup>, Irma Battipaglia<sup>1</sup>, Cristina Raimondo<sup>1</sup>, Elisa Del Vecchio<sup>1</sup>, Fabrizio Drago<sup>1</sup>

<sup>1</sup>Paediatric Cardiology and Cardiac Arrhythmias Unit, Department of Paediatric Cardiology and Cardiac Surgery, Bambino Gesù Children's Hospital and Research Institute, Piazza Sant'Onofrio 4, Rome 00165, Italy.; <sup>2</sup>Department of Pediatrics, Sapienza University of Rome, 00165 Rome, Italy.

**Background and Aim:** BACKGROUND: Studies report SCD in healthy children between 1.3–8.5 cases per 100 000 person-year, but such a low prevalence carries nonetheless a high burden for society. Aetiology is symptomatic or pre-symptomatic cardiomyopathies (CMP), channelopathies, congenital heart diseases (CHD), myocarditis and coronary anomalies.

In Italy, ECG-screening is performed in young athletes > 7 years. Furthermore, there are no Italian studies using ECG as a screening tool for heart anomalies in apparently normal pediatric population. The aim of our study is to evaluate the yield of ECG-screening in identifying any heart anomalies including those potentially at risk of SCD.

**Method:** METHODS: We prospectively analysed school-based ECGs, complemented by informed consent and a cardiovascular questionnaire from 2013 to 2022 in healthy schoolchildren between 3 and 13 years.

ECG anomalies were divided into major (short/long QTc, Brugada pattern, depolarization/repolarization anomaly suggestive of CMP, ventricular preexcitation, >II-degree AVB, bundle branch blocks, ectopic beats) and minor abnormalities (intraventricular and atrioventricular disturbances, short-PR, early repolarization, minor axis deviations, sinus bradycardia and tachycardia). **Results:** RESULTS: 18032 patients underwent ECG. Among them: 9016 (52%) were males. The 42% of the study population never had a prior ECG.

ECG anomalies were detected in 1267 (7.03%) cases. In 1082 (6%) children the anomalies were minor and in 185 (1.03%) were major and confirmed in 171 (0.95% of the entire study population at II-level evaluation.

**Conclusions:** CONCLUSION: Our study confirms that the ECG screening in children has a low cost and a very good efficacy and should be mandatory for the early detection of heart anomalies at potential risk of SCD.

**Keywords:** ECG-screening, sudden cardiac death prevention, low-cost prevention, children arrhythmias

### P-058

#### Autosomal recessive long QT syndrome: clinical aspects and therapy

Daniela Righi<sup>1</sup>, Luigina Porco<sup>1</sup>, Corrado Di Mambro<sup>1</sup>, Anwar Baban<sup>1</sup>, Maria Gnazzo<sup>2</sup>, Francesca Romana Lepri<sup>2</sup>, Antonio Novelli<sup>2</sup>, Alberto Eugenio Tozzi<sup>3</sup>, Fabrizio Drago<sup>1</sup>

<sup>1</sup>Pediatric Cardiology and Cardiac Arrhythmias Unit, Department of Pediatric Cardiology and Cardiac Surgery, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy; <sup>2</sup>Medical Genetics Unit, Medical Genetics Laboratory, Bambino Gesù Children's Hospital IRCCS, Rome, Italy; <sup>3</sup>Multifactorial and Complex Diseases Research Area, Bambino Gesù Children's Hospital IRCCS, Rome, Italy

**Background and Aim:** The recessive form of LQTS without an auditory phenotype represents patients with a severe clinical course, as the Jervell and Lange-Nielsen syndrome (J-LNS). Indeed, as in J-LNS, these patients have a very early onset and major QTc prolongation, and  $\beta$ -blockers have limited efficacy.

The aim of the study is to investigate the clinical characteristics and to evaluate the therapy effectiveness in children with recessive form of LQTS.

**Method:** From September 2012 to September 2021 data about 10 children diagnosed with recessive autosomal form of LQTS at the Bambino Gesù Children's Hospital were collected.

**Results:** The genetic analysis showed compound heterozygous variants of the gene in 3 patients (30%) and homozygous variants of the KCNQ1 gene in the other 7 (70%). All the patients with homozygous KCNQ1 variants presented deafness.

Mean follow-up was 2.9 years (0.52 – 9.79 years).

TWA was observed even on therapy with nadolol in 5/10 patients. A QTc value  $\geq 550$ ms was recorded in 5/10 and 2/10 showed values higher than 600ms. Moreover, 4/10 (40%) patients experienced one or more episodes of syncope, and 4/10 patients experimented a cardiorespiratory arrest (ACR): 2 with homozygous KCNQ1 variants and 2 with compound heterozygous.

Three of 4 (75%) patients with syncope showed also a ACR, while only 1/6 (16.6%) patients without syncope experienced ACR. Furthermore, 2/2 (100%) children with a QTc value more than 600ms presented an ACR, whilst it occurred in only 2/8 (25%) patients with a QTc value under 600ms.

All patients are treated with Nadolol, in 5 Mexiletine was added, due to suboptimal modulation of the QT value or persistence of TWA. In these, Mexiletine allowed a shortening of QTc in a medium value of 77ms (range 25–106ms) and in 4 patients with TWA obtained the disappearance of TWA.

**Conclusions:** In our study, recessive form of LQTS with homozygous variants seems associated with deafness. Patients with episodes of syncope and with QTc value >600ms even in beta-blocker therapy have an increased risk of ACR.

Mexiletine added to beta-blocker was proven effective in these patients to reduce QTc value and to remove TWA

**Keywords:** LQTS, recessive form, Mexiletine

## P-059

**The arrhythmic “threat” of non-compaction cardiomyopathy in children**

Irma Battipaglia, Nicoletta Cantanutti, Marianna Ciconia, Virginia Battista, Elena Pelliccione, Fabrizio Drago  
Paediatric Cardiology and Cardiac Arrhythmias/Syncope Unit,  
Department of Paediatric Cardiology, Bambino Gesù Children’s Hospital  
IRCCS, Rome, Italy

**Background and Aim:** Non-compaction cardiomyopathy (NCM) is defined by the appearance of ventricular trabeculae and deep inter-trabecular recesses on cardiac imaging. Clinical manifestations and outcomes vary greatly: it can be associated to cardiac dilation or hypertrophy, systolic dysfunction, arrhythmic events with sudden cardiac death (SCD). At present, few data are available on NCM in children. The aim of this work is identifying clinical characteristics and outcomes in children with NCM.

**Method:** We retrospectively analysed data from consecutive patients arriving at our hospital with diagnosis of NCM from 2013 to 2022. Data regarding familiarity, clinical history, imaging, cardiac biomarkers, arrhythmias and outcomes were collected.

**Results:** One-hundred forty-six children with imaging diagnosis of NCM were enrolled (84 males, mean age  $13 \pm 6$ ). At time of first presentation 133 (91%) patients had a diagnosis of NCM, 8 (5, 4%) patients had a mixed phenotype of NCM/dilated cardiomyopathy; 2 (1, 3%) patients had NCM and hypertrophy, in 3 (2%) cases a congenital heart defect was present. One patient was in NYHA class IV (NCM associated to mitral cleft and a genetic mutation on SCN5A gene); all others were in NYHA class I or II. Twenty-two (15%) patients had left ventricle dysfunction (mean EF was 48%); cardiac biomarkers were positive in 11 patients. In 52 (35%) children arrhythmic manifestations were documented: 4 cases of complete AV block; 1 sinus node dysfunction; 4 supraventricular tachyarrhythmias (1 ectopic atrial tachycardia, 2 AV re-entrant tachycardias, 1 case of atrial fibrillation); 9 patients had ventricular tachyarrhythmias (premature ventricular complexes in 2, sustained or not sustained ventricular tachycardias in 6, ventricular fibrillation in 1 case). Two patients had ICD implanted in primary or secondary prevention according to current guidelines. Mean follow-up was  $39 \pm 19$  months. No SCD occurred. Patients with ICD received appropriate shocks and ATP therapies from the devices.

**Conclusions:** NCM in children has different clinical manifestations. Most patients show class NYHA I functional status and good outcomes at follow-up (no heart failure nor arrhythmic or thromboembolic complications). Arrhythmias are not so rare (35%); 6% of patients had ventricular arrhythmias and 2 of them (22%) had ICD implanted. No SCD occurred in our cohort.

**Keywords:** non-compaction cardiomyopathy, arrhythmias, sudden cardiac death

## P-060

**Utility of implantable loop recorders in the diagnosis and management of cardiac symptoms in children and adolescents**

Pedro Agudo Montore<sup>1</sup>, Theano Theofanous<sup>2</sup>, María Cecilia González Corcia<sup>2</sup>

<sup>1</sup>Department of Paediatric Cardiology, Virgen del Rocío University Hospital, Seville, Spain; <sup>2</sup>Department of Paediatric Cardiology, Bristol Royal Hospital for Children, Bristol, United Kingdom

**Background and Aim:** Implantable loop recorders (ILR) have emerged as long-term effective monitoring tools for severe or recurrent rhythm abnormalities or unexplained cardiac symptoms. Scarce information is available on the use and results of this technology in the paediatric age. The aim of the study was to report the results of a cohort of paediatric patients that benefited from ILR implantation.

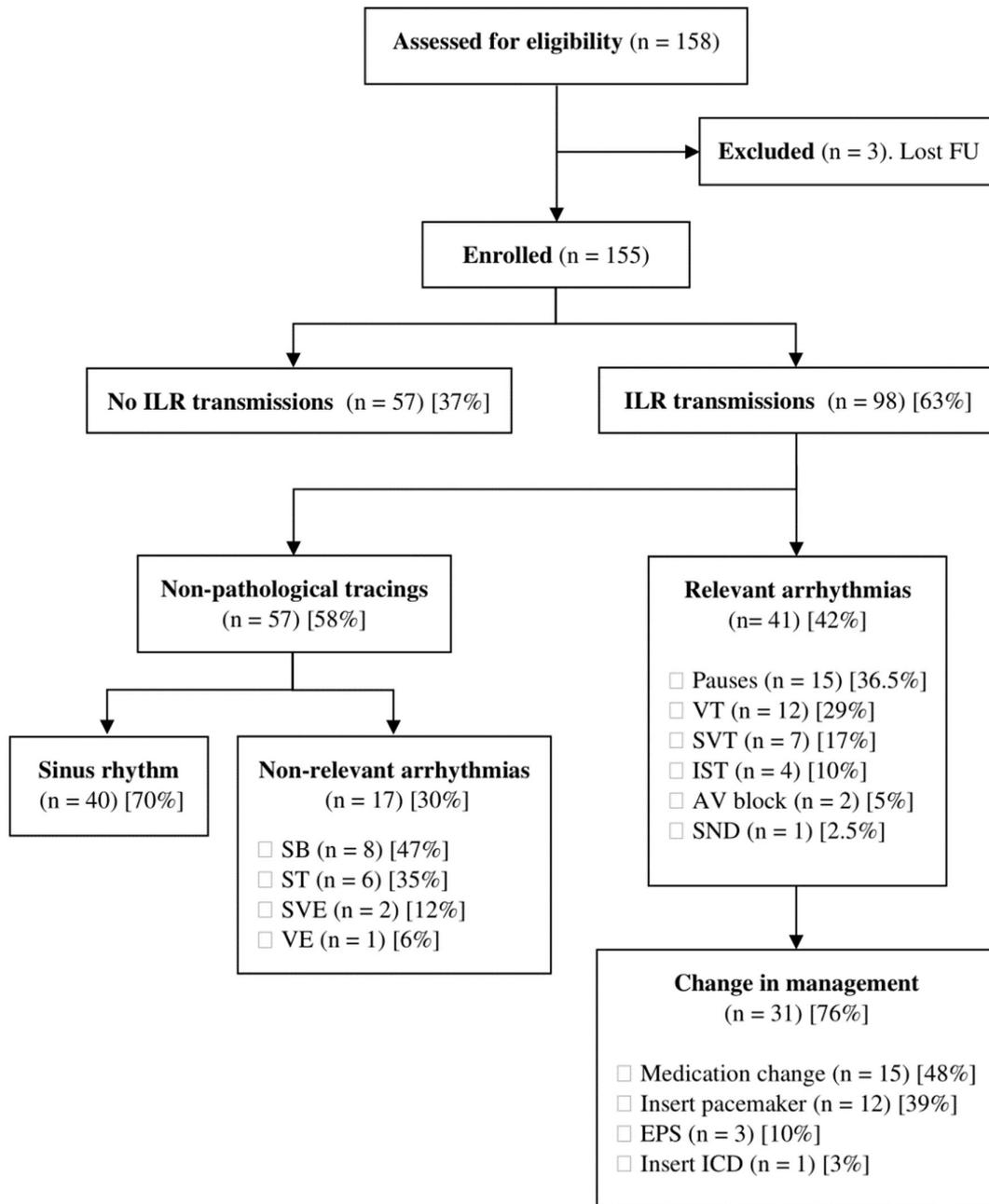
**Method:** Retrospective chart review of clinical data, implantation indications, diagnostic recordings and changes in patient management in paediatric patients who underwent Reveal LINQ ILR implantation from January 2016 to February 2022 at a single paediatric cardiology department in Bristol Royal Hospital for Children.

**Results:** In a paediatric cohort of 155 patients (mean age at implantation 10.3 years), 98 patients (63%) had ILR transmissions, 41 (42%) of which demonstrated a diagnostic arrhythmia recording. The median time to a diagnostic transmission was 175 days. The most common abnormality was sinus pauses (37%), followed by ventricular tachycardia (30%). Of the total transmissions, 78 (80%) were patient-activated and 21% of them had pathological traces. From the device-activated transmission ( $n = 33$ , 34%), 79% showed pathological tracings. Some of the transmissions were simultaneously patient and device-activated. In the arrhythmia-negative group ( $n = 57$ ), 85% of patients activated transmissions during periods of symptoms, with 70% of transmissions showing normal sinus rhythm. In 80% of patients with an ILR-related diagnosis, there was an arrhythmia management plan change, including medication change, a pacemaker or implantable cardioverter defibrillator implantation, and an indication to perform an electrophysiology study. No major complications occurred; 4 (2.4%) patients had minor complications.

**Conclusions:** ILRs have emerged as a new technology proven to be effective and safe in diagnosing and managing paediatric arrhythmias.

**Keywords:** Implantable loop recorder, paediatric, arrhythmias, inherited cardiac conditions, congenital heart diseases.

**Figure 2. ILR outcomes flowchart.**



AV stands for atrioventricular; EPS, electrophysiology study; FU, follow-up; ICD, implantable cardioverter-defibrillator; ILR, implantable loop recorder; IST, inappropriate sinus tachycardia; SB, sinus bradycardia; SND, sinus node dysfunction; ST, sinus tachycardia; SVE, supraventricular ectopy; SVT, supraventricular tachycardia; VE, ventricular ectopy; VT, ventricular tachycardia.

## P-061

**Myocardial and arrhythmic involvement with neuromuscular disorders: clinical and etiological heterogeneity in a cohort of pediatric population.**

Anwar Baban<sup>1</sup>, Giovanni Parlapiano<sup>1</sup>, Fabiana Fattori<sup>2</sup>, Viola Alesi<sup>3</sup>, Federica Cali<sup>1</sup>, Lorenzo De Luca<sup>1</sup>, Marianna Cicienia<sup>1</sup>, Giulia Pascolini<sup>1</sup>, Enrico Silvio Bertini<sup>2</sup>, Carlo Dionisi Vici<sup>4</sup>, Antonio Novelli<sup>3</sup>, Diego Martinelli<sup>4</sup>, Fabrizio Drago<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology and Cardiac Surgery, Heart and Lung Transplantation, Bambino Gesù Children Hospital and Research Institute, IRCCS, 00165 Rome, Italy, The European Reference Network for Rare, Low Prevalence and Complex Diseases of the Heart-ERN GUARD-Heart; <sup>2</sup>Unit of Neuromuscular and Neurodegenerative Disorders, Bambino Gesù Children Hospital and Research Institute, IRCCS, 00165 Rome, Italy; <sup>3</sup>Laboratory of Medical Genetics, Bambino Gesù Children Hospital and Research Institute, IRCCS, 00165 Rome, Italy; <sup>4</sup>Division of Metabolism, Bambino Gesù Children Hospital and Research Institute, IRCCS, 00165 Rome, Italy

**Background and Aim:** Neuromuscular disorders (NMDs) in children are heterogenous condition with often multisystemic signs and frequent cardiac involvement. Childhood onset forms can predispose to various progressive cardiac abnormalities including cardiomyopathies (CMPs), valvulopathies, atrioventricular conduction defects (AVCD), supraventricular tachycardia (SVT) and ventricular arrhythmias (VA).

**Method:** Our tertiary academic pediatric center represent the referral point for the center and south of Italy for the multidisciplinary management of patients with NMDs. In this study we include data

from patients with NMDs and cardiac involvement who had multisystemic clinical assessment, multiorgan screening and genetic study through NGS sequencing, microarray analysis and short tandem repeat expansion analysis.

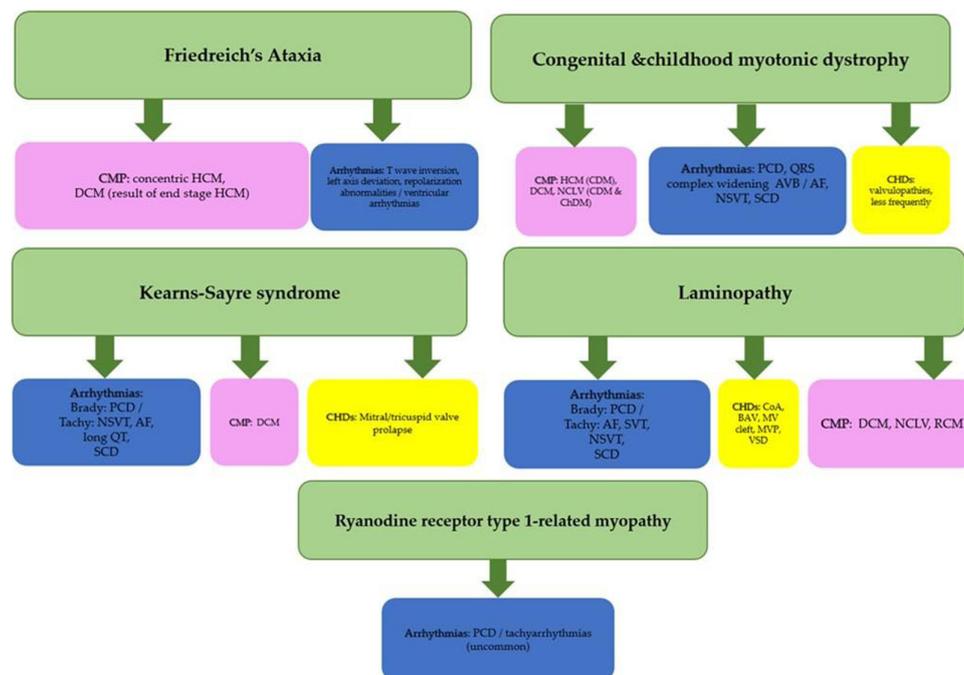
**Results:** We identified and described the possible cardiac spectrum in term of myocardial, structural, and arrhythmic involvement in five specific NMDs: Friedreich's Ataxia (FRDA), congenital and childhood forms of Myotonic Dystrophy type 1 (DM1), Kearns Sayre Syndrome (KSS), Ryanodine receptor type 1-related myopathies (RYR1-RM) and Laminopathies.

**Conclusions:** The diagnosis of certain NMDs is often overlooked and the cardiac aspect can provide signs of their presence even prior to overt neurological diagnosis. In this study we underline the importance of detecting the early onset rare photocopies cardiac manifestations in children' NMDs including its structural (valvular), myocardial (CMPs) and arrhythmic (brady-tachyarrhythmias) aspects. The diagnostic approach must be multidisciplinary and the management must be personalized in nature especially in the rare case of severe early onset forms. These patients need for periodic surveillance by a dedicated heart team.

Part of this topic is reviewed in: PMID: 34827576, PMCID: PMC8615674, DOI: 10.3390/biom11111578

This study is part of the multidisciplinary management project for patients with Pearson and Kearns-Sayre syndrome "Bando Finalizzato GR-2018-12368395".

**Keywords:** Cardiac involvement in NMDs, multisystemic conditions, genetic heterogeneity, multidisciplinary personalized approach.



**Figure 1** Major myocardial and arrhythmic aspects in neuromuscular disorders.

## P-062

**Heart and beyond the heart: clinical spectrum of cacna1c variants, literature revision**

Anwar Baban<sup>1</sup>, Maria Gnazzo<sup>2</sup>, Bernadette Corica<sup>1</sup>, Federica Cali<sup>1</sup>, Nicoletta Cantarutti<sup>1</sup>, Giovanni Parlapiano<sup>1</sup>, Daniela Righi<sup>1</sup>, Corrado Di Mambro<sup>1</sup>, Massimo Stefano Silvetti<sup>1</sup>, Antonio Novelli<sup>2</sup>, Fabrizio Drago<sup>1</sup>  
<sup>1</sup>Department of Pediatric Cardiology and Cardiac Surgery, Heart and Lung Transplantation, Bambino Gesù Children Hospital and Research Institute, IRCCS, 00165 Rome, Italy, The European Reference Network for Rare, Low Prevalence and Complex Diseases of the Heart-ERN GUARD-Heart; <sup>2</sup>Laboratory of Medical Genetics, Bambino Gesù Children Hospital and Research Institute, IRCCS, 00165 Rome, Italy

**Background and Aim:** CACNA1C is a gene encoding the CaV1.2 calcium channel and several cardiac conditions are potentially associated with pathogenic variants of this gene. The aim of this study is to explore genotype-phenotype correlations related to CACNA1C ever described variants and vast phenotypic spectrum both at cardiac and extracardiac level.

**Method:** We analyzed the phenotypic spectrum of 88 patients with CACNA1C variants (CACNA1Cv) from literature. We studied the association between CACNA1Cv and clinical parameters: arrhythmias, structural heart defects, cardiomyopathy, and survival or aborted cardiac arrest. We followed the American College Medical Genetics (ACMG) scoring system to grade variants' pathogenicity and their localization domains. Wide phenotypic spectrum in terms of upper limb anomalies, neurocognitive, and immunological involvement is analysed.

**Results:** CACNA1Cv with high ACMG scores were associated with higher mortality than variants with lower scores ( $p = 0.035$ ). CACNA1Cv in Cytoplasmic vs other Domain did not find significance. ACMG score 4-5 are associated to higher mortality rate 21.3% and LQTS (95.2%) compared to ACMG score 3 in which mortality rate was 11.1% and LQTS was 75%.

**Conclusions:** CACNA1Cv show wide phenotypic expressivity. It can range from "unisystemic" manifestations: only neurological or cardiac or multisystemic. In the present paper an attempt is undertaken to realize potential genotype phenotype correlation on the basis of literature data.

**Keywords:** CACNA1C variants, Arrhythmia, genotype-phenotype correlations, mortality

## P-063

**Refractory fetal and neonatal supraventricular tachycardia associated with mitral valve mass: A case report**

Marisa Pereira<sup>1</sup>, Catarina Almeida<sup>1</sup>, João Sarmento<sup>1</sup>, João Sarmento<sup>2</sup>, Joana Miranda<sup>1</sup>, Joana Miranda<sup>3</sup>

<sup>1</sup>Pediatric Cardiology Department, Centro Hospitalar Universitário de São João, Porto, Portugal; <sup>2</sup>Department of Gynecology-Obstetrics and Pediatrics, Faculty of Medicine of the University of Porto, Porto, Portugal; <sup>3</sup>UnIC@RISE, Department of Surgery and Physiology, Faculty of Medicine of the University of Porto, Portugal

**Background and Aim:** Primary cardiac tumors in children are rare and mostly benign, however they can be associated with serious cardiovascular complications, including clinically significant arrhythmias. They produce a wide variety of arrhythmias, including low-grade ectopic, pre-excitation and sustained supraventricular tachycardia (SVT), non-sustained and sustained ventricular tachycardia and sudden cardiac arrest.

The optimal approach for pediatric patients with cardiac tumors remains unclear, particularly when severe arrhythmias are present. **Method:** We present a case of prenatal diagnosis (28 weeks of gestation) of SVT associated with a mitral valve mass and fetal

hydrops. Transplacental treatment was performed with digoxin and flecainide for 14 days, followed by amiodarone. However, due to persistent fetal arrhythmia and maternal pharmacological toxicity, delivery by cesarean-section was performed at 30 weeks. The female newborn, weighing 2000g, presented SVT resistant to chemical cardioversion with adenosine and amiodarone.

On day-2 of life, a wide complex tachycardia episode was recorded, with no sustained reversal after synchronized electrical cardioversion, so lidocaine was associated. Persistent sinus rhythm was restored on day 9, at which time oral propranolol was started after amiodarone and lidocaine discontinuation.

On day-42, after 5 days with frequent premature atrial contractions, she restarted incessant SVT, requiring new chemical cardioversion and subsequent management with propranolol and flecainide. She has maintained sinus rhythm since day-53.

The echocardiogram presented a mass in the anterior leaflet of the mitral valve, without flow obstruction nor regurgitation. A computed tomography was performed (day-54) and detected both cardiac and liver mass, with similar characteristics and compatible with hemangiomas. Both showed involution during hospitalization, coinciding with improvement in the rhythm.

**Results:** The infant was discharged on day-67 with propranolol and flecainide. She remained asymptomatic with no documented episodes of SVT on regular ECG nor on 24-hour Holter monitoring.

**Conclusions:** We report a rare case of fetal and neonatal sustained SVT associated with a mitral hemangioma, which was medically controlled. Yet, it was associated with neonatal complications (prematurity, anemia, bronchopulmonary dysplasia and probably iatrogenic hypothyroidism) and maternal complications (pharmacological toxicity by digoxin and amiodarone).

Further follow-up is required to evaluate the duration of therapy as well as the characteristics and evolution of the cardiac hemangioma

**Keywords:** primary cardiac tumor, arrhythmias, supraventricular tachycardia, fetal, neonatal, infant

## P-064

**Clinical profile of paediatric atrial flutter – a tertiary care centre experience over 30 years**

Muthukumaran Ravichandran, Amos Wong, Orhan Uzun, Arveen Devindaran

Department of Paediatric Cardiology, University Hospital of Wales, Cardiff, United Kingdom

**Background and Aim:** Atrial Flutter (AFL) is rare and usually noted in neonatal age group or in children with an underlying heart disease. In this retrospective analysis, we intend to report the clinical presentation, management and follow up of all paediatric cases presented to the University Hospital of Wales over the past 30 years.

**Method:** A retrospective analysis of all paediatric AFL cases within South Wales presented to the University Hospital of Wales.

**Results:** There were 35 patients with paediatric AFL. 28 presented in perinatal period (20 prenatal and eight within 28 days of life). Remaining seven were diagnosed between one month and 16 years. Eight patients had structural heart disease. Tetralogy of Fallot (ToF), atrial septal defect (ASD) and Ebstein anomaly constituted approximately 25% each of these anomalies.

28 children (including all 20 prenatal) were asymptomatic at presentation apart from incidental tachycardia. Two presented with suspected sepsis. Other presentations included respiratory distress, vomiting / diarrhoea, lethargy, cough / coryza.

18 patients were treated with flecainide and digoxin combination antenatally. Five of them required postnatal medications owing to atrioventricular re-entry tachycardia (AVRT), of which four exhibited underlying Wolff Parkinson white syndrome (WPW).

21 children needed DC cardioversion to revert to sinus rhythm. Four postnatally diagnosed children had spontaneous resolution of their arrhythmia while all other children reverted with antenatal medication.

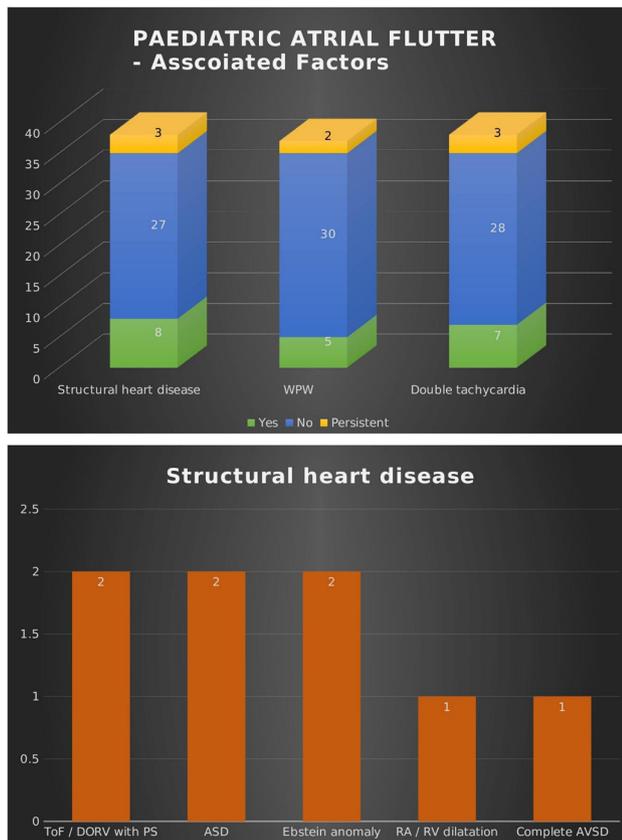
Six children had refractory AFL of which two had WPW, three with structural heart disease and one developed heart failure due to persistent AFL from the fetal period. Three patients had AVRT with AFL, two of whom are still receiving antiarrhythmic treatment. Only one patient is continuing to have persistent AFL despite treatment.

In total, five children had WPW (one successfully ablated, one failed ablation and three awaiting electrophysiology study). There was no mortality in this group.

**Conclusions:** Paediatric AFL seen in prenatal period is usually not associated with a structural heart disease, unlike the ones present later in infancy and childhood. This study shows that structural heart disease or pre-excitation are more likely to produce refractory AFL with or without AVRT with associated complications requiring long term follow up.

**Keywords:** Atrial flutter, pre-excitation, atrioventricular re-entrant tachycardia, cardioversion

AFL associated factors and structural heart disease profile



Top picture: Among the total population (35 children), 22% (8 children) had structural heart disease among whom three had refractory flutter. 14% (5 children) had WPW among whom two had refractory disease. 20% (7 children) had double tachycardia (AFL + AVRT) among whom 3 had refractory AFL. Bottom picture: Among the structural heart diseases, ToF/DORV, ASD and Ebstein anomaly each contributed to 25% cases (2 children) each with Complete AVSD & RA/RV dilation forming the rest (1 each).

## P-065

### Wolf parkinson white syndrome or asymptomatic ventricular pre-excitation in children: electrophysiological properties changes in two different timing

Cristina Raimondo, Silvia Garibaldi, Antonino Maria Quintilio Alberio, Ilaria Cazzoli, Massimo Stefano Silveti, Vincenzo Pazzano, Ilaria Tamburri, Fabrizio Drago

Paediatric Cardiology and Cardiac Arrhythmias Unit, Department of Paediatric Cardiology and Cardiac Surgery, Bambino Gesù Children's Hospital and Research Institute, Piazza Sant'Onofrio 4, Rome 00165, Italy.

**Background and Aim:** In patients with Wolf Parkinson White (WPW) syndrome or asymptomatic ventricular pre-excitation (VP), electrophysiological study (EPS) is recommended to assess and to stratify the risk of life-threatening arrhythmias and SCD. However, little information exist on the timing of EPS study. In particular, the electrophysiological properties variations of accessory pathway (AP) in patients with asymptomatic or symptomatic VP remain to be defined.

The aim of our study was to investigate the evolution of clinical and electrophysiological datas of the manifest PV in symptomatic or asymptomatic children, examined on two separate timing, at least 2 years apart from one EPS to the next.

**Method:** Between January 2011 and July 2022, we enrolled forty-four children and young adults (32 male, mean age 10 years  $\pm$  2, 42) with manifest ventricular preexcitation underwent, as our standard management, at two electrophysiological studies (EPS, transesophageal and/or endocavitary), both at rest and during adrenergic stress (exercise testing or isoproterenol infusion) in two different timing (T0 and T1) within a minimal interval of 2 year. No transcatheter ablation was performed between two EPS. Clinical and electrophysiological data were collected and compared.

**Results:** We observed a significant modification in atrioventricular accessory pathway conductive properties from T0 to T1 in basal study. In particular, at the baseline, preAVA value and 1/1 conduction VA time were respectively 306, 59  $\pm$  43 and 292, 61  $\pm$  61, 7 and significantly decreased on occasion of second EPS evaluation (T1) (respectively to 279, 51  $\pm$  41 with p 0, 004 and 267, 13  $\pm$  51, 6, with p 0, 003). Furthermore, an ARVT were induced in 14 patients in T1 respect 9 patient in T0 (p < 0, 03) and AF in 16 patients in T1 respect 7 patients in T0 (p 0, 24). This results were not confirmed on adrenergic test during EPS evaluation.

We also reported a clinically but not statistically relevant reduction of SPERR1 time evaluated during EPS with stress test from T0 to T1 (from 251, 44  $\pm$  47, 9 to 206, 9  $\pm$  36, 7; p = 0, 18).

**Conclusions:** There were significant and clinically relevant changes of electrophysiological datas in young patients with symptomatic and asymptomatic VP underwent two EPS in two different timing. Therefore a systematic evaluation with repeated EPS should be considered in this population.

**Keywords:** Wolf Parkinson White, electrophysiological study, risk stratification

## P-066

### Voltage abnormalities in 12-lead resting ECG in pediatric duchenne muscular dystrophy patients

Magdalena Bazgier<sup>1</sup>, Jarosław Meyer Szary<sup>1</sup>, Suramya Chandra<sup>1</sup>, Katarzyna Lisowska<sup>1</sup>, Jolanta Wierzbza<sup>2</sup>, Joanna Kwiatkowska<sup>1</sup>

<sup>1</sup>Department of Paediatric Cardiology and Congenital Heart Defects, Medical University of Gdańsk, Gdańsk, Poland; <sup>2</sup>Department of Paediatrics, Haematology and Oncology, Department of General Nursery, Medical University of Gdańsk, Poland

**Background and Aim:** Electrocardiographic changes are one of the earliest and leading manifestations of cardiac involvement in Duchenne muscular dystrophy (DMD). In our study we sought to determine the prevalence of ECG abnormalities, their severity and age-dependency.

**Method:** Heart rhythm(HR), heart rate, conduction time and voltages were assessed in resting ECG recordings acquired between 2015 and 2021 in patients with genetically confirmed DMD in a single-centre observational study.

Reference data by Rijenbeek et al.(2001) based on n = 944 boys were used as the control group. For t-test calculations following assumptions were made: (1)normal distribution of the reference sample analysed parameters after transformation based on the calculated regressions, (2)z-score mean 0 and SD 1 of normalised parameters, (3)the published 98 centile minus the mean representing 2 SD. For P, PQ, QRS duration assessment mean 12-lead values were used. Statistical analysis was performed using Wizard Pro 1.9.38(Evan Miller, Chicago, IL). All data are reported as mean  $\pm$  standard deviation.

**Results:** Ninety-seven ECG recordings, all with sinus rhythm, in 66 male and 1 female patients aged  $9.3\pm 4.2$  years were analysed. HR of  $103\pm 17$ bpm (z-score  $0.39\pm 0.31$ ) was found to be increased ( $p<0.001$ ), severity age-dependent ( $p<0.001$ , Fig 1A).

Whereas the P and PR duration were decreased (z-scores of  $-0.72\pm 0.24$ ,  $p<0.001$  and  $-0.42\pm 0.17$ ,  $p<0.001$  respectively), the QRS duration was increased (z-score  $0.23\pm 0.25$ ,  $p<0.001$ ). P wave shortening severity was age-dependent ( $p<0.001$ , Fig.1B), with no correlation to HR. Based on z-score calculation increased voltages of R waves were found in leads: I ( $0.30\pm 0.47$ ), aVL ( $0.46\pm 0.58$ ), V1 ( $0.19\pm 0.37$ , Fig. 1C), V2 ( $0.33\pm 0.48$ ); Q waves in II ( $0.21\pm 0.47$ ), aVF ( $0.20\pm 0.26$ , Fig.1D), V6 ( $0.19\pm 0.26$ ) and of the S waves in III ( $0.38\pm 0.75$ ). Decreased voltages of S waves were found in leads V1 ( $-0.15\pm 0.27$ ), V2 ( $-0.37\pm 0.35$ ), V4 ( $-0.20\pm 0.40$ ), V6 ( $-0.36\pm 0.29$ ); of R waves in V4 ( $-0.24\pm 0.18$ ), V6 ( $-0.16\pm 0.13$ );  $p<0.001$  for all calculations. QRS fragmentations were found in n = 54(55, 7%) studies, most frequently affecting R waves in leads III, aVF, aVL and S waves in V1 (16%, 15%, 13% and 19% respectively).

**Conclusions:** While electrocardiographic changes in Duchenne muscular dystrophy, especially voltage abnormalities, are extensive, their role in heart failure development, long-term prognostic value and clinical utility remain unclear.

**Keywords:** Duchenne Muscular Dystrophy, Resting ECG, Voltage abnormalities

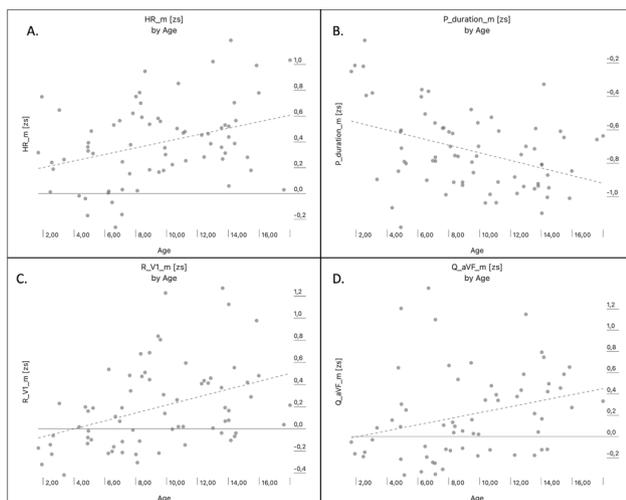


Fig. 1. Age dependency of selected parameters: A. Heart rate [z-score], B. P wave duration [z-score], C. R wave voltage in V1 lead [z-score], D. Q wave voltage in aVF lead [z-score]

## P-067

### Subcutaneous implantable cardioverter-defibrillators in children and adults with complex congenital heart diseases: single centre experience.

Linda Litwin<sup>1</sup>, Agata Grochowina Major<sup>1</sup>, Dominika Musioł<sup>1</sup>, Sławomir Pluta<sup>2</sup>, Adam Sokal<sup>2</sup>, Oskar Kowalski<sup>3</sup>, Roland Fiszer<sup>4</sup>

<sup>1</sup>Department of Congenital Heart Defects and Pediatric Cardiology, Silesian Center for Heart Diseases, Zabrze, Poland; <sup>2</sup>Department of Cardiology, Congenital Heart Diseases and Electrotherapy, Silesian Center for Heart Diseases, Zabrze, Poland; <sup>3</sup>Department of Dietetics, FOHS in Bytom, Medical University of Silesia, Katowice, Poland; <sup>4</sup>Department of Congenital Heart Defects and Pediatric Cardiology, FMS in Zabrze, Medical University of Silesia, Poland

**Background and Aim:** Subcutaneous implantable cardioverter-defibrillators (S-ICD) are increasingly used in children and adults with complex congenital heart defects, however the evidence on safety and effectiveness of S-ICD is yet to be established in this populations. We report early follow-up data from single centre S-ICD registry.

**Method:** Observational follow-up study includes patients with S-ICD implanted up to 18 years of age or in case of complex congenital heart defect. We assessed the class of recommendation at the time of procedure according to 2021 PACES Expert Consensus Statement (patients < 18 years of age) and the 2015 ESC Guidelines. We analyzed early and late procedural complications, inappropriate shocks, appropriate therapy, and patients' outcome.

**Results:** From 2017 to 2022 we implanted S-ICD in 8 patients using standard 3 incisions technique, defibrillator threshold testing was abandoned in 2 patients. There was 1 minor procedural complication (subcutaneous emphysema) and no late complications. One patient received the appropriate shock (effective) and another one inappropriate shocks (reprogramming avoided further episodes). No patient required transition to transvenous device. DCM - dilated cardiomyopathy; BS - Brugada syndrome; HCM - hypertrophic cardiomyopathy; cc-TGA - congenitally corrected transposition of the great arteries; PA - pulmonary atresia; MAPCAs - major aortopulmonary collateral arteries

**Conclusions:** These short-term results suggest that S-ICD implantation in populations of adolescents and adults with complex congenital heart defects is safe and effective. Clinical decision on ICD implantation can be challenging due to significant discrepancies between current guidelines, especially in middle to late adolescence.

**Keywords:** implantable cardioverter-defibrillator, sudden cardiac death, cardiomyopathy, Brugada Syndrome, congenital heart defect, child

## P-068

### Subclinical cardiac autonomic dysfunctions in cancer patients: role of heart rate variability?

Ekta Khandelwal<sup>1</sup>, Sumeet Tripathi<sup>2</sup>

<sup>1</sup>Ekta Khandelwal, AIIMS Raipur, Raipur, INDIA; <sup>2</sup>Sumeet Tripathi, Pt. JNMCC, Raipur, INDIA

**Background and Aim:** Cardiac Autonomic dysfunction is common and among one of the least recognized impairment in cancer patients like lymphoma, leukemia, carcinomas of lung, breast, ovary, prostate, testicles and pancreas. It is one of the significant causes of morbidity, mortality and affecting the compromised quality of life. Despite several recent therapeutic advances, these cancer patients face an increased risk of developing autonomic neuropathy along with treatment-related cardiovascular

autonomic dysfunctions. Aim of the study is assessment of cardiovascular autonomic functions to monitor the prognosis of the disease and side effects of chemotherapy on cardiovascular autonomic dysfunctions.

**Method:** Total 46 patients of various cancers on cardiotoxic chemotherapy were assessed for Heart rate variability (HRV) and Pulse wave analysis (PWA). Data obtained were compared with aged matched controls by using statistical software for Time domain, Frequency Domain & Non linear analysis for HRV. Pulse transit Time, Crest time, decay time for PWA.

**Results:** HRV Parameters of sympathetic and parasympathetic limb like SDDSD, RMSSD, SDNN, pN50, LF/HF ratio, Total Power, SD1 & SD2 were significantly lower in Cancer patients then controls ( $p > 0.005$ ). PWA showed increase in PPT in patients then controls with altered other parameters depicting vascular dysfunctions.

**Conclusions:** Thorough detection of subclinical cardiac autonomic dysfunction in cancer patients is of vital importance for risk stratification and subsequent management. As the cardiac regulation is dependent on non-linear deterministic system, the non-linear dynamics measures should be preferred.

**Keywords:** Cardiac autonomic dysfunction, Heart rate Variability, Pulse wave analysis

#### P-070

##### When you hear hoofbeats, think of horses – not always true: case of a newborn with a rare cause of bradycardia

Sascha Bapistella, Andrea Martig, Clemens von Kalkreuth, Birgit Donner

Department of Paediatric Cardiology, University Children's Hospital Basel, Basel, Switzerland

**Background and Aim:** Bradycardia is a potentially serious symptom in newborns. Knowing the age-appropriate heart rate values may save lives. In the present case we demonstrate how a thorough diagnostic work-up identified the cause of life-threatening arrhythmias.

**Method:** A neonate was admitted to the hospital on its 7th day of life after sleeping for 8.5 hours without crying or drinking. The mother had thyrostatic therapy in pregnancy due to Grave's disease and fetal hypothyroidism had been suggested in second trimester, which could be ruled out postpartum. Therefore, the family had been discharged on 5th day of life.

On admission, the heart rate was 74–78 bpm, the further clinical investigation was unremarkable.

ECG showed a prolongation of QTc interval of 527 ms. Presuming the clinical diagnosis of long-QT syndrome (Schwartz-Score 3.5), therapy with propranolol and magnesium was started. Parents were counselled in respect of strict avoidance of QT-prolonging drugs. Genetic testing excluded LQTS1–3. The parents were discharged home after training in infant resuscitation and AED usage with home ECG monitoring.

On the 28th day of life, the patient went into ventricular fibrillation, which was successfully terminated with 15 J.

**Results:** In the meantime, expanded genetic testing revealed a Calmodulin 3 (CALM3; LRG\_1082t1) sequence variant (c.422A>G, a heterozygous de novo mutation). Until now this mutation is described in two other symptomatic patients (P. Schwartz pers. communication). Therefore, the diagnosis of an autosomal dominant hereditary CALM3-associated long QT-syndrome was made.

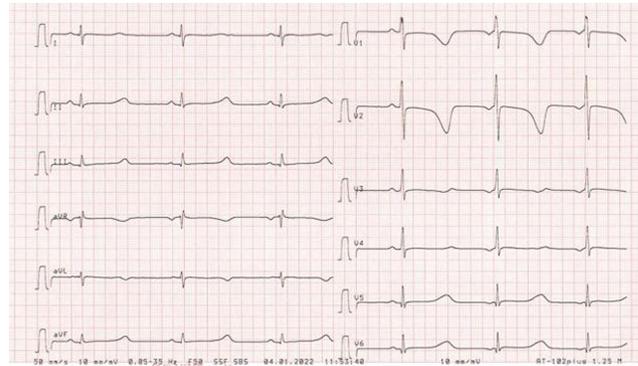
**Conclusions:** Medical therapy was modified by increasing the dosage of beta-blockers (propranolol up to 5 mg/kg/d) and adding

mexilitine (class IB antiarrhythmic). An AICD was implanted. In the last 10 months no further cardiac events occurred.

Calmodulinopathies are an extremely rare cause of long-QT syndrome (<100 cases have been reported worldwide, Crotti et al. 2019). Typically, these are associated with a severe course despite antiarrhythmic therapy. The median age at the first event is 1.5 years. Life-threatening arrhythmias appear in 78% of patients throughout live.

In the present case, extending the genetic analysis was helpful in risk stratification, medical therapy and counselling and might therefore be considered in patients with evidence of elevated QTc and unknown underlying cause.

**Keywords:** Bradycardia, long-QT Syndrome



ECG on admission (7th Day of Life)

#### P-072

##### Arrhythmias and outcomes of ebstein anomaly in children and adults

Dimitri Herrera Nativi<sup>1</sup>, Francis Bessiere<sup>1</sup>, Fanny Deville<sup>1</sup>, Julia Mitchell<sup>1</sup>, Olivier Metton<sup>1</sup>, Sylvie Di Filippo<sup>2</sup>

<sup>1</sup>Pediatric and Congenital Cardiology, Claude Bernard University, Lyon, France; <sup>2</sup>Congenital Cardiology, Cardiothoracic Center of Monaco

**Background and Aim:** Ebstein anomaly outcomes often complicate with arrhythmias. The objectives of the study were to analyze the occurrence and characteristics of arrhythmic events in children and adults with Ebstein anomaly.

**Method:** Single-center study including patients diagnosed with Ebstein anomaly over a 20-year period of time.

**Results:** 76 patients were analyzed, 34 males (45%), aged at diagnosis  $21.9 \pm 21.4$  years, median 13.9: 21 neonates (27.5%), 20 children (26.5%) and 35 adults (46%). Fourty-seven had symptoms at diagnosis (62%): 23 cyanosis, 35 dyspnea and heart failure, 13 palpitations, while 29 (38%) had no symptom. NYHA class was I, II, III and IV in respectively 55%, 35.5%, 6.5% and 3% of the cases. Ebstein anomaly was major type in 19% (14 cases), minor in 35% and intermediate in 46%. ASD was present in 54 cases (71%), VSD in 8 (10%), pulmonary hypoplasia in 5 (6.6%), an accessory pathway in 12 (16%). Interatrial shunt was left to right in 8%, right to left in 36% and bidirectional in 15%. Supraventricular tachycardia occurred in 33 cases (43.4%): supraventricular reentrant tachycardia (SVRT) in 14 (18.4%) and atrial fibrillation (AF) in 19 (25%). SVRT was associated with accessory pathway (41% versus 14%,  $p = 0.06$ ). Mean age at first arrhythmia was 39 years: 25.3 years for SVRT compared to 45.3 years for AF ( $p = 0.0003$ ). Right bundle branch was present in 48 cases (63%). ECG accessory

pathway was associated with no right bundle branch,  $p = 0.09$ . Atrioventricular block was diagnosed in 16 cases (21%) and pacemaker was implanted in 9 (12%). Ablation was performed in 12 (15.8%). Freedom from pacemaker implantation was 90%, 80%, and 60% at respectively 55, 65, and 75 years of age. Freedom from arrhythmia was 90%, 80%, 60%, 50% and 25% at respectively 10, 40, 50, 55 and 70 years of age. Arrhythmias episodes and specifically AF, were associated with HF (43.7% vs 13.6%,  $p = 0.03$ ) but not with death.

**Conclusions:** SVRT and AF complicate the outcomes of patients with Ebstein anomaly, earlier for SVRT and cause of HF for AF, but do not correlate with mortality.

**Keywords:** Ebstein anomaly, arrhythmias, outcomes, prognosis

### P-073

#### “I have a Dream”: Zero or near-zero x-ray pacemaker implantation in paediatric patients.

Massimo Stefano Silveti, Vincenzo Pazzano, Irma Battipaglia, Fabio Anselmo Saputo, Iliara Tamburri, Marta Campisi, Virginia Battista, Giacomo Silveti, Fabrizio Drago

Paediatric Cardiology and Cardiac Arrhythmia/Syncope Unit, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy - European Reference Network for Rare and Low Prevalence Complex Disease of the Heart (ERN GUARD-Heart).

**Background and Aim:** Transvenous implantation of pacemaker (PM) is performed with fluoroscopy for leads' insertion and implantation in the heart chambers. This implies radiation exposure for patients and operators. Complex implant procedures as alternative right ventricular (RV) pacing sites, may prevent pacing-induced ventricular dysfunction, but often requires higher radiation doses. A quite low radiation exposure was reported in very few and small sized paediatric experiences. The use of three-dimensional-electroanatomic mapping system (3D-EAM) may reduce fluoroscopy and guide lead implantation. Purpose of this study is to seek out if a 3D-EAM-guided transvenous implantation into RV alternative pacing sites in paediatric patients can be accomplished with zero or near-zero X-rays.

**Method:** Retrospective analysis of children and adolescents with congenital or acquired (idiopathic) complete atrioventricular block (CAVB) without other congenital heart defects who underwent 3D-EAM-guided pacing in alternative RV sites. The implant procedure was divided in 4 steps: 1-contrast venography; 2- 3D-mapping: with a steerable catheter (femoral vein), the 3D-EAM acquired geometric reconstruction of the right heart and a pacing map identified RV sites with narrower paced QRS; 3-axillary vein puncture; 4-lead and pacemaker implantation: 3D-EAM guided stylet-directed screw-in lead implantation toward desired RV sites. Data are reported as median (25th-75th centiles).

**Results:** 53 CAVB patients (41 females), underwent 3D EAM-guided pacing (27 VVIR, 26 DDD PM) at age 11.5 (7.7-14) years, weight 42 (26-54) kg. Pacing sites were: RV outflow tract (4 patients), conduction system area pacing (49 patients). Paced QRS was 115 (100-120) ms. Procedure time was 170 (143-193) min, total fluoroscopy exposure was: 2.0 (1.0-4.6) mGy and 58 (30-130) microGy/m<sup>2</sup>. The lowest exposures were: 0.2 mGy, 8 microGy/m<sup>2</sup> (VVIR) and 0.6 mGy, 15 microGy/m<sup>2</sup> (DDD). Six patients showed exposures  $\leq 0.5$  mGy and 4  $\leq 10$  microGy/m<sup>2</sup>. The radiation doses of the 4 steps are reported in Table 1.

**Conclusions:** 3D-EAM-guided alternative RV pacing sites was accomplished in a large paediatric cohort with very low fluoroscopic exposure, close to zero in some cases. Therefore, the use

of 3D-EAM demonstrated a relevant lowering of radiation doses also in difficult and complex pacing procedures in paediatric patients, thus reducing radiological risks for patients and operators. The dream may become reality.

**Keywords:** cardiac pacing, radiation exposure, alternative pacing sites, paediatric patients.

### Fluoroscopic Exposure times

	mGy	µGy/m <sup>2</sup>
1. Venography	0	3.7 (1.9-9.3)
2. 3D mapping	0	1.5 (0.2-4.1)
3. Vein puncture	0	5.4 (2.0-15.0)
4. Lead-PM implant	1.7 (0.8-3.2)	35.1 (16.5-94.3)
Total	2.0 (1.0-4.6)	58.0 (30.3-129.8)

**Table 1** Radiation exposures of the 4 steps of pacemaker implantation and total doses in the study cohort. Data are reported as median (25th-75th centiles)

### P-074

#### Neurologic manifestations in children with congenital arrhythmia syndromes subanalysis of the german cogia study

Fridrike Stute<sup>1</sup>, Lea Lippert<sup>2</sup>, Volker Debus<sup>14</sup>, Robert Dalla Pozza<sup>5</sup>, Stefan Rupp<sup>7</sup>, Maria Gonzales Y Gonzales<sup>7</sup>, Sven Dittmann<sup>4</sup>, Dominik Sebastian Westphal<sup>6</sup>, Peter Ewert<sup>2</sup>, Michael Hofbek<sup>11</sup>, Sven Dittich<sup>13</sup>, Jan Hendrik Nuernberg<sup>12</sup>, Joachim Hebe<sup>12</sup>, Anselm Uebing<sup>10</sup>, Philipp Beerbaum<sup>8</sup>, Majed Kanaan<sup>9</sup>, Gunter Kerst<sup>15</sup>, Hans Gerd Kehl<sup>14</sup>, Eric Schulze Bahr<sup>4</sup>, Roman Gebauer<sup>3</sup>, Joerg Sachweh<sup>1</sup>, Thomas S Mir<sup>1</sup>, Rainer Kozlik Feldmann<sup>1</sup>, Gabriele Hessling<sup>2</sup>, Cordula Wolf<sup>2</sup>

<sup>1</sup>Department of Pediatric Cardiology, University Heart & Vascular Center Hamburg - UKE, Germany; <sup>2</sup>Department of Congenital Heart Diseases / Pediatric Cardiology, German Heart Center Munich, Technical University Munich, Germany; <sup>3</sup>Department of Pediatric Cardiology, Leipzig Heart Center, Leipzig University, Leipzig, Germany; <sup>4</sup>Institute for Genetics of Heart Disease, University Hospital Munster, Germany; <sup>5</sup>Division of Pediatric Cardiology and Intensive Care, University Hospital, LMU Munich, Germany; <sup>6</sup>Institute of Human Genetics, Klinikum rechts der Isar, School of Medicine, Technical University of Munich, Munich, Germany. Department of Internal Medicine I, Klinikum rechts der Isar, School of Medicine, Technical University of Munich, Munich, Germany; <sup>7</sup>Department of Pediatric Cardiology, University Hospital Giessen and Marburg, Germany; <sup>8</sup>Department of Pediatric Cardiology and Intensive Care, University Hospital Hannover, Germany; <sup>9</sup>Department of Pediatric Cardiology, University Hospital Aachen, Germany; <sup>10</sup>Department of Congenital Heart Defects and Pediatric Cardiology, University Hospital Schleswig-Holstein, Kiel, Germany; <sup>11</sup>Department of Pediatric Cardiology, Pulmonology and Intensive Care, University Hospital Tubingen, Germany; <sup>12</sup>Elektrophysiologie Bremen, Bremen, Germany; <sup>13</sup>Department of Pediatric Cardiology, University Hospital Erlangen, Erlangen, Germany; <sup>14</sup>Department of Pediatric Cardiology, University Hospital Munster, Munster, Germany; <sup>15</sup>Devision of Pediatric Cardiology, Klinikum Stuttgart, Stuttgart, Germany

**Background and Aim:** Channelopathies are rare diseases with cardiologic, neurologic and musculoskeletal manifestations related to location, gain or loss of function of the altered ion channel.

Particularly challenging is the management of children with overlapping symptoms. The objective is to describe the clinical course in relation to molecular genetics in children and adolescents with congenital arrhythmia syndromes with additional neurologic manifestations.

**Method:** Based on the retrospective multicenter data of the COGIA study (Clinical course, outcome and genetics of inherited arrhythmias in children) pediatric patients with a clinical and/or molecular genetic diagnosis of an inherited arrhythmia syndrome (Long QT, Brugada, Short QT, CPVT) and additional neurologic diagnosis were subjected to a subanalysis.

**Results:** Among 632 patients with childhood onset inherited arrhythmia syndrome (median age at time of diagnosis 6.1 [IQR 0.4;11.3] years; follow-up 4.5 [IQR 1.9;9.4] years), LQTS was present in 73, 6% [LQTS1 in 32, 8 (207), LQTS2 in 22, 6 (143), LQTS3 in 6, 6 (42), other LQTS in 11, 6%, BrS in 9, 8%, CPVT in 9% and other in 7, 6% of patients. A pathogenic genetic variant was identified in 502/583 (86.1%) of patients tested.

In 27 patients (4.3%) an additional neurological diagnosis was present. This included epilepsy/seizures not explained by hypoxic events in 15 patients [2.4%; 7 KCNQ1, 4 KCNH2, 1 SCN5A, 1 RYR2, 1 KCNJ4, 1 unknown) or relevant neurologic impairment i.e. paralysis, developmental disorder in 12 patients [1.9%; 4 KCNQ1, 3 KCNH2, 1 SCN5A, 4 KCNJ2]). In none of the cases a second mutation explaining the neurologic findings was found.

**Conclusions:** About 5% of pediatric patients with congenital arrhythmia syndromes show additional severe neurologic impairment. An accurate genetic characterization of the altered ion channel might help to predict prognosis and to adjust therapy strategies.

**Keywords:** Congenital arrhythmia syndrome, Long QT, Brugada, Epilepsy

#### P-075

##### Predicting reflex syncope based on plethysmography: A new wearable device development and preliminary results

Sérgio Matoso Laranjo<sup>1</sup>, Guilherme Carvalho Lourenço<sup>1</sup>, Helena Fonseca<sup>2</sup>, Lourenço Rodrigues<sup>4</sup>, Andre Lourenço<sup>3</sup>, Fátima F. Pinto<sup>1</sup>, Mário Martins Oliveira<sup>2</sup>

<sup>1</sup>Serviço de Cardiologia Pediátrica, Centro de Referência de Cardiopatias Congénitas do CHULC, Hospital de Santa Marta, Centro Hospitalar e Universitário de Lisboa Central, Lisboa, Portugal; <sup>2</sup>Serviço de Cardiologia, Hospital de Santa Marta, Centro Hospitalar e Universitário de Lisboa Central, Lisboa, Portugal; <sup>3</sup>Instituto Superior de Engenharia de Lisboa; <sup>4</sup>Cardioid

**Background and Aim:** Recurrent reflex syncope (RSync) is a common clinical condition that significantly impacts a patient's life. The underlying mechanism is a transient global cerebral hypoperfusion accompanied by a rapid drop in blood pressure (BP). Isometric counterpressure manoeuvres (legs/arms) induce BP to increase during impending syncope, avoiding or delaying losing consciousness. Therefore, detecting the onset of the syncope mechanism as early as possible is of major importance, particularly in patients (P) without prodromes.

**AIM:** to develop an innovative wearable, efficient and reliable device that can anticipate the RSync.

**Method:** P with recurrent syncope referred for head-up tilt testing (HUT) were monitored using a continuous non-invasive arterial BP system (TaskForce Monitor, CNSystems, Graz, Austria), complemented with the synchronised acquisition of plethysmography (PPG) signals using an innovative wearable device. The device used in this experiment is based on the Maxim platform - a wristband PPG sensor with three integrated wavelengths. It allows the

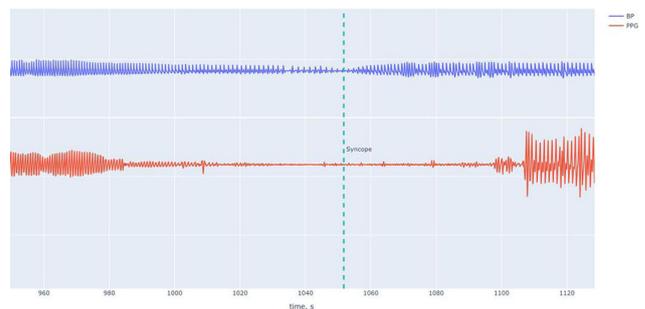
instantaneous heart rate and oxygen saturation monitoring and the extraction of the raw sensor data, enabling the analysis conducted in this study. The continuous BP signal was synchronised with the PPG by extracting the intervals between systolic peaks and finding the delay on PPG that minimised the quadratic difference between the tachogram series. The human verification of the initial estimate further refined synchronisation. From the aligned signals, a segment of 120 sec was extracted from the basal period and the 120 sec prior to the syncopal event.

**Results:** Ten patients (age <18 years) with HUT-induced RSync were enrolled in this proof-of-concept study. Patients from different VASIS classifications were enrolled: one cardioinhibitory, five mixed-type and four vasodepressor. By computing the areas in each segment, normalised by the amplitude of the basal pulses in each signal, a consistent and significant reduction of the PPG amplitude and wave patterns were found between pre-syncopal and basal periods. Furthermore, these changes started, at an average, 60 sec before syncope, preceding the systolic BP, stroke volume and cardiac output changes.

**Conclusions:** Predicting RSync is feasible by monitoring PPG amplitude and morphology changes along the time. This new approach may have a relevant impact on the future management of RSync.

**Keywords:** Syncope, PPG, prediction

#### PPG tracing



#### P-076

##### Surpassing the complex substrate of accessory pathways ablation in Ebstein anomaly

Sérgio Matoso Laranjo<sup>1</sup>, Guilherme Lourenço<sup>1</sup>, Guilherme Portugal<sup>2</sup>, Pedro Cunha<sup>2</sup>, Lidia Sousa<sup>2</sup>, Conceição Trigo<sup>1</sup>, Fátima Pinto<sup>1</sup>, Rui Cruz Ferreira<sup>2</sup>, Mário Martins Oliveira<sup>2</sup>

<sup>1</sup>Serviço de Cardiologia Pediátrica, Centro de Referência de Cardiopatias Congénitas do CHULC, Hospital de Santa Marta, Centro Hospitalar e Universitário de Lisboa Central, Lisboa, Portugal; <sup>2</sup>Serviço de Cardiologia, Hospital de Santa Marta, Centro Hospitalar e Universitário de Lisboa Central, Lisboa, Portugal

**Background and Aim:** Data regarding long-term follow-up of radiofrequency catheter ablation (RFCA) of accessory pathways (APs) in patients (P) with Ebstein's anomaly are limited. This type of procedure is considered challenging due to multiple and broad APs. The present study aimed to describe the electrophysiological features of APs in P with Ebstein's anomaly, and report our RFCA experience with an open-window electroanatomic 3D mapping using high-density mapping catheters in these patients.

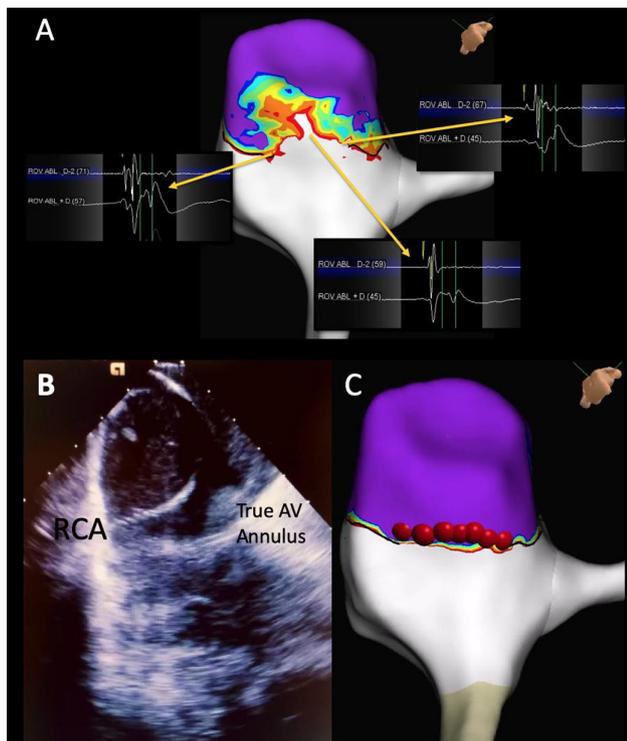
**Method:** A retrospective study of 15 consecutive Ebstein anomaly P with APs who underwent an electrophysiologic study and RFCA from 2013 to 2022.

**Results:** There were a total of 21 manifested non-decremental APs. APs were mainly located on the posterior, posteroseptal, and posterolateral tricuspid annulus. The index procedure was unsuccessful in six P, with the need for a redo procedure. This redo procedure was performed with a high-density catheter (Pentaray, Biosense or HD-Grid, Abbott), using the open-window annotation algorithm (Abbott's NavX Precision or BiosenseWebster Carto3), guided by CT integration and intracardiac echo. Broad APs were documented in all these six patients (width range 2–15cm) and successfully ablated. In one P, the AP encompassed  $\frac{3}{4}$  of the TA, resulting in a complete AV block after the procedure, having fitted a pacemaker. All P remained free from tachycardias during  $15 \pm 8$  months of follow-up, with the majority ( $n = 12$ ) having sinus rhythm with morphology of right bundle branch block, while three patients showed a narrow QRS.

**Conclusions:** RFCA in P with Ebstein anomaly is challenging, but safe, and has a high long-term success rate. APs are predominantly right-sided, manifest and localized to the lower half of the anatomic tricuspid annulus. Some APs have broad widths. In this population, the new high-resolution mapping catheters, using the open-window annotation, produce an improved anatomical resolution of the APs, increasing the odds of success.

**Keywords:** Ebstein, WPW, RF ablation

Open window mapping AP in Ebstein patient



P-077

#### Pathophysiology of reflex syncope response: role of the autonomic nervous system and baroreflex function

Sérgio Matoso Laranjo<sup>1</sup>, Guilherme Lourenço<sup>1</sup>, Teresa Mateus<sup>1</sup>, Helena Fonseca<sup>2</sup>, Isabel Rocha<sup>3</sup>, Conceição Trigo<sup>1</sup>, Fátima F. Pinto<sup>1</sup>, Mário Oliveira<sup>2</sup>  
<sup>1</sup>Serviço de Cardiologia Pediátrica, Centro de Referência de Cardiopatias Congénitas do CHULC, Hospital de Santa Marta, Centro Hospitalar e

Universitário de Lisboa Central, Lisboa, Portugal; <sup>2</sup>Serviço de Cardiologia, Hospital de Santa Marta, Centro Hospitalar e Universitário de Lisboa Central, Lisboa, Portugal; <sup>3</sup>Instituto de Fisiologia, Faculdade de Medicina da Universidade de Lisboa, Universidade de Lisboa, Lisboa, Portugal

**Background and Aim:** Syncope is a common medical problem during the lifetime, with a recurrence rate of 35%. Excluding cardiac disease, most syncopes are of reflex origin, and despite their frequent occurrence, their mechanisms are not yet well defined. Furthermore, while studies in the adult population with vasovagal syncope are abounding, few studies (with conflicting results) have investigated the vasovagal syncope mechanisms in paediatric patients.

**Method:** 238 patients were enrolled (range 12–18 years-old, mean age  $13.4 \pm 4.1$  years, 64.3% females), having experienced, on average,  $3.7 \pm 2.9$  syncope episodes before the HUT. The population was divided according to the HUT response: tilt-positive patients (fainters) and tilt-negative patients (non-fainters). Heart rate (HR), blood pressure (BP), and cardiac haemodynamics were continuously monitored using a Task Force Monitor. In addition, HR variability (HRV), BP variability, baroreflex sensitivity (BEI), cross-spectral wavelet coherence, and phase were evaluated.

**Results:** The test was positive in 99 (41.8%) patients, representing the fainters' group. 38 patients (38%) were defined as cardioinhibitory (type 2), 35 patients (35.4%) mixed type (Type 1) syncope and 26 (26.6%) as vasodepressor (Type 3). Fainters showed significantly higher HR, lower stroke volume and total peripheral resistance values during HUT. Four phases of cardiovascular responses leading to syncope could be described. Additionally, a significant rise in sympathetic activity characterised Fainters' HRV response to HUT. In brief, the core dynamic changes to LF included sudden and initial rise of sympathetic tone immediately after tilting up (Phase 1), followed by a significant decrease of sympathetic activity (Phase 2), the second overshoot of activity (Phase 3), and, then, a steady fall-off 1–2 minutes before syncope (Phase 4). Despite similar BEI in the supine position, the fainters' group showed less systolic BP ramps and a higher lag of the baroreflex response. After HUT, the fainters' group showed a progressive but significant BEI decrease.

**Conclusions:** These results strengthen the hypothesis that impaired baroreflex function and an imbalance between the two branches of the autonomic nervous system may represent a pathophysiological marker of altered response to orthostatic stress and play a role in the pathophysiology of reflex syncope. The findings can stratify reflex syncope patients to define an integrated and personalised therapeutic approach.

**Keywords:** Syncope, heart rate variability,

P-078

#### The impact of frequent premature ventricular complexes morphology to cardiopulmonary exercise testing performance in children

Rita Kunigeliene<sup>1</sup>, Odeta Kinciniene<sup>1</sup>, Vytautas Usonis<sup>1</sup>, Germanas Marinskis<sup>2</sup>

<sup>1</sup>Clinics of Children Diseases, Institute of Clinical Medicine, Vilnius University Faculty of Medicine, Vilnius, Lithuania; <sup>2</sup>Department of Cardiology, Vilnius University Hospital Santaros Clinics, Vilnius Lithuania

**Background and Aim:** Although premature ventricular complexes (PVCs) are considered benign in children, frequent and particular morphology PVCs can cause ventricular dysfunction and might be the first sign of cardiomyopathy and sometimes indicate limitations

of physical activity. However, evidence-based guidelines have documented that regular physical activity is beneficial for cardiovascular disease prevention and exercise testing is one of the tools in risk stratification for patients with PVCs.

The aim is to evaluate the role of cardiopulmonary exercise testing (CPET) factors as a prognostic tool in children with structurally normal hearts and various morphology of PVCs.

**Method:** A prospective analysis of 19 children with structurally normal heart and frequent ventricular PVCs (>10 % per 24 hours). Patients underwent electrocardiogram and CPET on treadmill (BTL Cardiopoint CPET) using modified ramp protocol. Peak oxygen uptake per kilogram (VO<sub>2</sub>/kg), carbon dioxide elimination (VCO<sub>2</sub>), anaerobic threshold (AT), respiratory exchange rate (RER), VE/VO<sub>2</sub> were measured. According to the morphology of PVCs in ECG, patients divided into two groups: LBBB morphology and RBBB morphology PVCs. Statistical analysis performed with R. Nominal variables for normal distribution tested with Shapiro-Francia test. Nominal values were presented with minimum, maximum and median. Welch test used to compare means between normally distributed nominal variables. Fisher exact test used to compare categorical variables. The p value <0, 05 was considered statistically significant.

**Results:** 6 boys and 13 girls 6–17 (median 12, 5) years old with 11–37 (median 21, 98) % of PVC per 24 hours. LBBB morphology PVCs were in 4 and RBBB morphology PVCs – in 15 cases. CPET duration was 4:59–10:36 (median 6:55) minutes. Patients reached maximal load of 50 – 285 (median 150) watts; RER 0, 93 – 2, 05 (median 1, 1); VO<sub>2</sub> peak 18, 9 – 36, 3 (median 31, 2) ml/kg/min; VCO<sub>2</sub> 676–2674 (median 1631) ml/min, AT 390–1377 (median 901) ml/min, VE/VO<sub>2</sub> 36, 1–49, 7 (median 41, 4). We didn't find statistically significant differences between CPET factors and PVCs morphology groups, but anaerobic threshold (AT) seems to be the promising factor.

**Conclusions:** Anaerobic threshold (AT) seems to be the promising factor for evaluation of PVCs during CPET. It is necessary to include more patients into the study for more significant and clear results.

**Keywords:** CPET, children, frequent PVC, normal heart

#### P-079

##### **Implantable cardioverter-defibrillator in a patient with life-threatening inherited type 3 CPVT gen (TECRL) after sudden cardiac arrest.**

Elhadi Husein Aburawi<sup>1</sup>, Mohamed Mattar Al Mehairi<sup>2</sup>, Zahurul B Bhuiyan<sup>3</sup>

<sup>1</sup>CMHS, UAE University P. O. Box 15551, Al Ain, Abu Dhabi, UAE; <sup>2</sup>Department of Cardiology, Tawam Hospital, SEHA, UAE;

<sup>3</sup>Unit of Cardiogenetic Research, Division of Genetic Medicine, BT.02.251, Beaumont 29, CH-1011 Lausanne, Switzerland.

**Background and Aim:** Patients with ventricular tachycardia (VT) are amenable to sudden cardiac arrest (SCA) and death. The standard treatment is antiarrhythmic medications plus implantable cardioverter defibrillator (ICD). It has been reported that in patients with catecholaminergic polymorphic ventricular tachycardia (CPVT), ICD shocks are sometimes futile and may even trigger fatal electrical storms. Furthermore, ICD is not associated with improved survival and instead, it was associated with both a high rate of appropriate and inappropriate ICD shocks along with other device-related complications. The routine ICD insertion is not recommended according to the present guideline-directed therapy to prevent all the potential disadvantages of an ICD. The aim is to demonstrate that ICD could prevent inappropriate shocks and

related complications in homozygous TECRL truncating mutation, CPVT Type 3

**Method:** AA is 16-year-old boy, who was diagnosed as homozygous for a TECRL truncating mutation, CPVT Type 3 at the age of 8 years. He presented 3 years of age to pediatric ED with SCA, while he was running at nearby Park. He was resuscitated for VT, admitted to PICU and started on Nadolol and Magnesium sulphate. He had MRI brain, which was normal. He recovered quite well, though he had another survival SCA in the word. Family history indicated 3 deaths after SCA during exercise in his first-degree cousins plus his brother.

**Results:** He underwent ICD insertion at 3.5 years old, VVI/ 40 BPM, ®Medtronic Single Chamber ICD VR EVERA XT, and had no more symptoms since then. During his last 12 years follow up, the most recent interrogation revealed satisfactory testing of all pacing parameters without documented malfunctions. For the last 12 years duration of the ICD implantation, the patient remained stable in terms of CPVT recurrences and the pacing parameters functions without documentation of appropriate or inappropriate shocks.

**Conclusions:** Given the fact that CPVT is a cardiac channelopathy, that has known association with SCD, lower threshold for ICD implantation may need further clinical trials. These studies are particularly done in the presence of concurrent technology of ICDs that showed convincing evidence of minimizing inappropriate shocks and related complications.

**Keywords:** CPVT, ICD, Secondary prevention, Sudden cardiac arrest

#### P-082

##### **Arrhythmias in ebstein's anomaly: 15 – years' single center experience of catheter ablations**

Valentyna Khanenova<sup>1</sup>, Maryna Meshkova<sup>1</sup>, Nadiia Rudenko<sup>2</sup>, Illya Yemets<sup>1</sup>

<sup>1</sup>Ukrainian Children's Cardiac Center; <sup>2</sup>Shupyk National Healthcare University of Ukraine

**Background and Aim:** Arrhythmias are common in patients with Ebstein's anomaly (EA). Although most arrhythmias can be corrected with catheter ablation, certain problems make this procedure more difficult in EA. This study examines the mechanisms of various EA associated arrhythmias and catheter ablation outcomes as a single center over 15 years' experience.

**Method:** The clinical and procedural data of catheter ablations in patients with EA in Ukrainian Children's Cardiac Center were analyzed.

**Results:** Twenty-two patients (mean age 14 ± 6 years) had 26 ablations from 2007 to 2022. There were 27 accessory pathways (AP) in 19 procedures, 2 cases of cavotricuspid isthmus-dependent atrial flutter (CTI-AFL), 3 focal atrial tachycardias (AT), 1 ventricular tachycardia (VT) and 1 atrioventricular nodal reciprocal tachycardia (AVNRT). In 6 patients (35%) multiple AP were present. Four patients (15, 3%) required multiple procedures to repeat ablation of the same arrhythmia. The procedure success rate after the first ablation was 85%.

**Conclusions:** Most EA related arrhythmias are amenable to catheter ablation. However, ablation procedures are complex and the need for repeat procedures is particularly high because some patients have multiple ablation targets and due to technical issues associated with a dysplastic tricuspid annulus and enlarged right atrium.

**Keywords:** Ebstein's anomaly, arrhythmias, catheter ablation, accessory pathway.

## P-083

**Celebrating 25th anniversary of a paediatric arrhythmia unit in barcelona. what have we learned?**

Georgia Sarquella Brugada, Josep Brugada Tarradellas  
 Unidad de Arritmias Pediátricas, Cardiopatías Familiares y Muerte Súbita, Hospital Sant Joan de Déu, 08950 Barcelona, España; European Reference Network for Rare, Low Prevalence and Complex Diseases of the Heart (ERN GUARD-Heart), 1105 AZ Amsterdam, The Netherlands.

**Background and Aim:** In 1998, the first paediatric ablation was performed in our institution. After 25 years, more than 3500 procedures have been performed in children from birth to 18 years of age. Much has been learned, much has been changed. Nevertheless, the initial vision of the Unit keeps leading our everyday work: to save lives, by preventing sudden death in children. **Method:** We present the 25-year experience of a paediatric arrhythmia unit. 3500 procedures have been performed in children from 5 days to 18 years of life. Including ablations (67%), EP studies (11%), drug challenges (9%), device implantation (10%), and others. **Results:** Technology has changed overtime. From conventional radiofrequency-based cases at the beginning (100% before 2016) to the inclusion of new technology (cryoablation (less than 1%)), to mapping systems (since 2020, 50% of cases). Device implantation techniques have been personalized in this small population.

**Conclusions:** Socially, several differences have been seen compared to adult centres: humanization techniques during procedures, patient journey focused on young patient experience, training programmes specialized in paediatric electrophysiology. All these make particularly different the focus to explore and treat paediatric arrhythmias.

**Keywords:** paediatric ablation, arrhythmia, cryoablation, mapping systems

## P-084

**Management of cardiac arrhythmias in paediatric patients with TANGO2-deficiency disorder**

Georgia Sarquella Brugada<sup>1</sup>, Jose Cruzalegui Gomez<sup>1</sup>, Sergi Cesar Diaz<sup>1</sup>, Fredy Chipa Casani<sup>2</sup>, Estefania Martínez Barrios<sup>3</sup>, Victoria Fiol<sup>2</sup>, Patricia Cerralbo<sup>3</sup>, Andrea Greco<sup>2</sup>, Josep Brugada Tarradellas<sup>1</sup>, Oscar Campuzano Larrea<sup>4</sup>

<sup>1</sup>Unidad de Arritmias Pediátricas, Cardiopatías Familiares y Muerte Súbita, Hospital Sant Joan de Déu, 08950 Barcelona, España; European Reference Network for Rare, Low Prevalence and Complex Diseases of the Heart (ERN GUARD-Heart), 1105 AZ Amsterdam, The Netherlands.; <sup>2</sup>Unidad de Arritmias Pediátricas, Cardiopatías Familiares y Muerte Súbita, Hospital Sant Joan de Déu, 08950 Barcelona, España; <sup>3</sup>Arritmies Pediàtriques, Cardiologia Genètica i Mort Sobtada, Malalties Cardiovasculars en el Desenvolupament, Institut de Recerca Sant Joan de Déu, Esplugues de Llobregat, 08950 Barcelona, Spain.; <sup>4</sup>Medical Science Department, School of Medicine, Universitat de Girona, 17003 Girona, Spain; Cardiovascular Genetics Center, University of Girona-IDIBGI, 17190 Girona, Spain

**Background and Aim:** TANGO2-deficiency disorder (TDD) is a rare paediatric condition caused by bi-allelic mutations in the TANGO2 gene. TDD is characterised by susceptibility to metabolic crises with rhabdomyolysis, encephalopathy, lactic acidosis, hypothyroidism and difficult to manage tachyarrhythmias.

**Method:** Retrospective, single-centre, case series study of patients with TDD (<18 years), with ECG and subcutaneous Holter monitoring, both during metabolic crises and compensated phases. We

describe the electrocardiographic (ECG) findings at baseline and during metabolic crises in patients with TDD.

**Results:** During the period 2013-2021, 12 patients with TDD were found (median age 6.8 years, IQR 2-11.5 years). In the inter-crisis period, bradyarrhythmias were evidenced: 2/12 patients presented sinus bradycardia, 2/12 pauses of 4 and 10 seconds, 1/12 Wenckebach type block; and tachyarrhythmias: 1/12 presented paroxysmal supraventricular tachycardia and 1/12 flutter. 25% had long QT during non-acute phases (median QTc max 476 ms, IQR 456ms-530ms). There were 14 hospitalisations for metabolic crises. 57.1% had QTc interval prolongation, 35.7% ventricular tachycardias (4/14 monomorphic VT, 1/14 torsade de pointes) and 1/14 Brugada type 3 pattern. Three patients died in the context of severe metabolic acidosis and multi-organ involvement (2/12 due to VT and 1/12 due to cardiogenic shock, bradycardia associated with QTc of 635ms and severe ventricular dysfunction).

**Conclusions:** In paediatric patients with TDD, the most frequent ECG abnormality was QTc interval prolongation with or without metabolic crises. There is a high risk of tachyarrhythmias, mainly VT, being the main cause of death in children with TDD. Subcutaneous Holter monitoring is recommended. During metabolic crises, there should be close ECG monitoring.

**Keywords:** TANGO2 deficiency disorder, arrhythmia management, Holter monitoring

## P-089

**Ventricular tachycardia substrates in young patients with repaired tetralogy of fallot**

Justin Wallet<sup>1</sup>, Yoshitaka Kimura<sup>1</sup>, Nico Blom<sup>2</sup>, Monique Jongbloed<sup>3</sup>, Robin Bertels<sup>2</sup>, Mark Hazekamp<sup>4</sup>, Katja Zeppenfeld<sup>1</sup>

<sup>1</sup>Department of Cardiology, Heart Lung Centre, Leiden University Medical Centre, Leiden, The Netherlands & Willem Einthoven Centre of Arrhythmia Research and Management & Centre for Congenital Heart Disease Amsterdam-Leiden (CAHAL), The Netherlands; <sup>2</sup>Department of Paediatric Cardiology, Leiden University Medical Centre, Leiden, The Netherlands & Centre for Congenital Heart Disease Amsterdam-Leiden (CAHAL), The Netherlands; <sup>3</sup>Department of Cardiology, Heart Lung Centre, and department of Anatomy & Embryology, Leiden University Medical Centre, Leiden, The Netherlands & Centre for Congenital Heart Disease Amsterdam-Leiden (CAHAL), The Netherlands; <sup>4</sup>Department of Cardiothoracic Surgery, Leiden University Medical Centre, Leiden, The Netherlands & Centre for Congenital Heart Disease Amsterdam-Leiden (CAHAL), The Netherlands

**Background and Aim:** Patients with repaired tetralogy of Fallot (rTOF) have an increased risk of ventricular tachycardia (VT) after the age of 30 years (Y), with slow conducting anatomical isthmus (SCAI) 3, between the pulmonary annulus and the ventricular septal defect (VSD) patch, as dominant VT substrate. However, we aim to assess the prevalence of SCAI, the associated factors in SCAI development, and the occurrence of clinical VT, in rTOF patients <30Y.

**Method:** From a database of 146 patients with rTOF or related lesions who underwent electroanatomical mapping (EAM) and programmed electrical stimulation (PES) between 2007 and 2022 for treatment of ventricular arrhythmias (VA), risk stratification or before pulmonary valve replacement (PVR), those aged <30Y at time of procedure were analysed.

**Results:** Of 146 rTOF patients, 55 were <30Y (16Y[IQR 14-22]); 33(60%) had TOF, 9(16%) TOF with double outlet right ventricle, and 13(24%) complex TOF variants, including 8 patients with pulmonary atresia with VSD. Initial intracardiac repair (age 0.8Y[0.4-1.4]) was performed via a ventriculotomy in 10(18%).

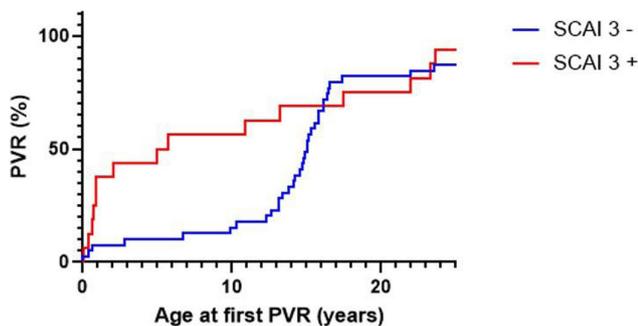
An RV to pulmonary artery (RV-PA) conduit was inserted during repair or early after (<1Y) repair in 12(22%). In 14(25%) PVR was performed later in life (age 10.1Y[2.7-14.3]). Five (9%) underwent EAM/PES after spontaneous VA, 40(73%) before PVR, and 10(18%) for risk stratification. Monomorphic VT was inducible in 8(15%), including 4/5 patients with spontaneous VA, and was proven related to SCAI 3 in 7/8(88%).

In 16(29%) SCAI 3 could be identified; no other SCAI (1, 2, 4) was present. In univariable analysis complex TOF (OR 6.8[95%CI 1.8-26.4]), ventriculotomy (OR 5.3[1.2-22.3]), any prior PVR (OR 4.8[1.4-16.7]), and initial RV-PA conduit/early PVR (OR 15.4[3.3-71.8]) were significantly associated with SCAI 3. In multivariable analysis initial RV-PA conduit/early PVR remained the only independent predictor of SCAI 3 (OR 13.0[1.2-142.4]).

**Conclusions:** SCAI 3 is present in 29% of rTOF patients <30Y referred for EAM/PES and the dominant substrate for spontaneous and inducible VTs. RV-PA conduit placement during initial repair or early thereafter (<1Y) is associated with SCAI 3 in young rTOF. Whether the early conduit/valve placement causes scarring at the infundibulum with consecutive slow conduction and/or whether the complex variants contribute to SCAI requires additional studies.

**Keywords:** Congenital heart disease, ventricular tachycardia, slow conducting anatomical isthmus, electroanatomical mapping, catheter or surgical ablation

Age at first PVR in relation to presence of SCAI 3



#### P-090

##### Congestive heart failure in infants diagnosed with supraventricular arrhythmias

Minna Mecklin<sup>1</sup>, Anniina Linnanmäki<sup>2</sup>, Anita Hiippala<sup>3</sup>, Topias Leino<sup>4</sup>, Anita Arola<sup>4</sup>, Markku Leskinen<sup>5</sup>, Hanna Ruotsalainen<sup>6</sup>, Juha Matti Happonen<sup>3</sup>, Tuija Poutanen<sup>1</sup>

<sup>1</sup>Tampere University Hospital, Department of Paediatrics, Paediatric Cardiology; <sup>2</sup>Tampere University, Faculty of Medicine and Health Technology; <sup>3</sup>Helsinki University Hospital, Department of Paediatrics, Paediatric Cardiology; <sup>4</sup>Turku University Hospital, Department of Paediatrics and Adolescent Medicine, Paediatric Cardiology; <sup>5</sup>Oulu University Hospital, Department of Paediatrics, Paediatric Cardiology; <sup>6</sup>Kuopio University Hospital, Department of Paediatrics, Paediatric Cardiology

**Background and Aim:** Supraventricular arrhythmias (SA) in infants are related to morbidity and may lead to congestive heart failure (CHF). We describe clinical findings of infants diagnosed with

SA and sought to find factors related to CHF without congenital heart disease (CHD).

**Method:** This is a descriptive retrospective cohort study. The medical records of infants diagnosed with SA before age of 12 months between 2005 and 2017 were collected from five university hospitals. Infants diagnosed with CHD were excluded. We compared patients with CHF to those without CHF at time of the diagnosis of SA.

**Results:** A total 350 infants were diagnosed with SA of which 59 infants with CHD were excluded. Out of 291 infants, 71 (24%) had CHF. Subtypes of SA were atrioventricular re-entrant tachycardia (n = 236), atrial tachycardia (n = 26), atrial flutter (n = 16), multifocal atrial tachycardia (n = 6), permanent junctional reciprocating tachycardia (n = 6) and junctional ectopic tachycardia (n = 1). The subtype of SA was not associated with CHF. In infants with CHF, the typical findings were left ventricular (LV) dysfunction (59%), mitral valve regurgitation (49%) and increased LV size (24%). The infants with CHF were older at admission (median, 15 vs 5 days, p < 0.001), the mean duration of symptoms was longer (21 vs 5 hours, p < 0.001) and median rate of arrhythmia was higher (273/min vs 250/min, p 0.029) compared to infants without CHF.

**Conclusions:** The SA in infants without CHD may lead to CHF. The risk factors for CHF were older age at admission, longer duration of symptoms and higher ventricular rate during SA.

**Keywords:** Supraventricular tachycardia, supraventricular arrhythmias, congestive heart failure, infants

#### P-091

##### Comparing predictive power of primacy, hcmrisk-kids and ESC HCM risk-calculator in assessing risk for sudden cardiac death

Ingegerd Östman Smith<sup>1</sup>, Gunnar Sjöberg<sup>2</sup>, Jenny Alenius Dahlqvist<sup>3</sup>, Per Larsson<sup>4</sup>, Eva Fernlund<sup>5</sup>

<sup>1</sup>Institute of Clinical Specialties, Sahlgrenska Academy, Gothenburg University; Children's Heart Center, Queen Silvia Children's Hospital, Gothenburg, Sweden; <sup>2</sup>Department of Women's and Children's Health, Karolinska Institute, Stockholm, Sweden; <sup>3</sup>Department of Clinical Sciences, Umeå University, Umeå, Sweden; <sup>4</sup>Department of Pediatric Cardiology, Uppsala University, Children's Hospital, Uppsala, Sweden; <sup>5</sup>Department of Clinical and Experimental Medicine, Linköping University, Division of Pediatrics, Crown Princess Victoria Children's Hospital, Linköping University Hospital, Linköping; Department of Clinical Sciences Lund, Lund University, Skåne University Hospital, Pediatric Cardiology, Lund, Sweden

**Background and Aim:** Hypertrophic cardiomyopathy (HCM) is the commonest cause of medical sudden death in children after infancy and recently two different paediatric risk calculators to aid in patient selection for implantation of a primary prophylactic internal cardiac defibrillator (ICD) have been published. The ESC adult HCM risk-calculator can be used from age 16y. This study aims to compare their performance in non-syndrome associated HCM-cases.

**Method:** 177 HCM-patients with diagnosis below age 19y, 64% male, were identified in Swedish and English geographical cohorts. Median age at diagnosis was 10.6y [IQR 4.1-14.2]. All survivors had ≥5y of follow-up to assess the end-point of sudden death, cardiac arrest or appropriate ICD-discharge (SCD/CA) within 5y; median follow-up was 10.8 [7.7-15.1]. PRIMACY, HCMRisk-Kids and ESC HCM-Risk-SCD values were calculated using web-based calculators.

**Results:** A total of 34 SCD/CA occurred at median age of 17y [12.1-23.3]. The median calculated percentage risk in the cohort

at diagnosis were quite different: HCMRisk-Kids 3.92% [2.75–6.77] versus PRIMACY 6.66% [4.39–10.42], both being overestimates as only 14 SCD/CA occurred in first five years (annual risk 1.58%), with 22 SCD/CA within 10y of diagnosis. Comparing both with ROC-curves as continuous functions gives a C-statistic of 0.81 [0.69–0.93],  $p < 0.001$  for HCM Risk-Kids versus 0.71 [0.54–0.89],  $p = 0.008$  for PRIMACY. HCMRisk-Kids recommends a cut-off of  $\geq 6\%$ , which gives a C-statistic of 0.75 [0.62–0.88],  $p = 0.002$ ; PRIMACY does not advocate a specific cut-off, but a distribution-derived cut-off of  $\geq 9\%$  has a C-statistic of 0.69 [0.54–0.84],  $p = 0.019$ . For 80 patients values at 16–18 yrs of age for ESC HCM Risk-SCD could be compared with the paediatric algorithms: 2.71% [2.01–4.38] (ESCHCM-Risk) versus 4.24% [2.78–8.26] (HCMRisk-Kids) and 12.7% [9.22–16.7] (PRIMACY). As the majority of SCD/CA occurred before 23 years of age there was poor statistical power to compare the three, but ESCHCM-Risk  $\geq 6\%$  had a C-statistic of 0.63 [0.33–0.94],  $p = 0.38$ , the lowest C-statistic of the three algorithms in this age-range.

**Conclusions:** HCMRisk-Kids appears to have the best predictive power of the two paediatric algorithms, and appears to work well up to 18y of age. The ESCHCM-Risk performs less well for patients between 16 and 18 years of age.

**Keywords:** sudden cardiac death, hypertrophic cardiomyopathy, primary prevention, internal cardiac defibrillators

#### P-092

##### **Incessant post-operative arrhythmia in a toddler – when the usual therapy is not enough**

Guilherme Lourenço<sup>1</sup>, Sérgio Laranjo<sup>1</sup>, Mário Oliveira<sup>2</sup>, Conceição Trigo<sup>1</sup>, Fátima F. Pinto<sup>1</sup>

<sup>1</sup>Paediatric Cardiology Department, Reference Centre for Congenital Heart Disease, Centro Hospitalar Universitário de Lisboa Central, Lisboa, Portugal; <sup>2</sup>Cardiology Department, Reference Centre for Congenital Heart Disease, Centro Hospitalar Universitário de Lisboa Central, Lisboa, Portugal

**Background and Aim:** Early post-operative atrial tachycardias in congenital heart disease are rare, can lead to significant morbidity and mortality, and are notoriously hard to treat.

**Method:** case report

**Results:** An 8-months-old toddler (8.350kg) was admitted to our hospital for uncontrolled arrhythmia. He had a previous history of Berry Syndrome – aortopulmonary window with interrupted aortic arch and aortic origin of right pulmonary artery – having already been subjected to corrective cardiac surgery during the neonatal period, and interventional catheterization and reoperation due to residual lesions (re-coarctation of the aorta and severe stenosis of the branch pulmonary arteries).

In a routine Holter, multiple sustained episodes of narrow and broad complex tachycardia were identified, with almost incessant arrhythmia and maximum heart rate (HR) of 301bpm. He was admitted and focal atrial tachycardia (FAT) with HR-dependent conduction aberrancy was diagnosed with 12-lead ECG. The arrhythmia was uncontrollable with a combination of propranolol, amiodarone and flecainide at maximum doses. Residual lesions were excluded by magnetic resonance and diagnostic catheterization, and an electrophysiological study was performed. Under general anaesthesia the arrhythmia subsided, but FAT was easily and reproducibly induced by means of isoprenaline bolus. The tachycardia cycle was ~370ms, with irregular AV conduction (1:1, 2:1; heart-rate dependent left bundle branch block). An electroanatomical map was performed, with earliest activation on the base of right atrial appendix. Radiofrequency energy was applied at

this location, and tachycardia was no longer inducible. The procedure had no complications. He was discharged on propranolol, with permanent sinus rhythm for 7 days.

One month later he was again readmitted due to arrhythmia recurrence, which was easily controlled with low-dose oral verapamil. At three years follow-up he remains in normal sinus rhythm.

**Conclusions:** In our patient, due to incessant and poorly tolerated FAT, it was necessary to use high risk, seldomly-used therapies for this age group – EPS with ablation and calcium channel blockers. These were accomplished with success and without complications.

**Keywords:** Atrial, tachycardia, toddler, ablation, verapamil

ECG / electroanatomical map



12-lead ECG and electroanatomical activation time map of the arrhythmia

#### P-093

##### **Practice variability, treatment efficacy and long-term outcomes in fetal and neonatal atrial flutter**

Magdalena Bazgier, Jarosław Meyer Szary, Julia Przeniosło, Joanna Kwiatkowska

Department of Paediatric Cardiology and Congenital Heart Defects, Medical University of Gdańsk, Gdańsk, Poland

**Background and Aim:** Atrial flutter (AFL) in fetuses and neonates is a rare, yet possibly life-threatening arrhythmia, with direct current (DCC) and pharmacological cardioversion as therapeutic options. The aim of the study was to evaluate the practice variability, treatment efficacy and long-term outcomes in fetal and neonatal AFL. **Method:** Fetal and neonatal cases of AFL diagnosed in years 2015–2022 were analyzed in a single-centre, retrospective observational study. Complex congenital heart defects were considered an exclusion criterium. All data are reported as median(range).

**Results:** In the studied period 11 cases of AFL were identified, 2 excluded based on the criterium above. Nine patients, 6 females, were analyzed. Prenatal tachycardia diagnosis was made in 6 cases. In 2 cases (diagnosed at 30th and 36th weeks of gestation) treatment was initiated with maternally administered amiodarone or amiodarone and digoxine leading to conversion to sinus rhythm (SR) prenatally in 1 case and spontaneous conversion to SR in first hours of life in the other case. Fetal tachycardia was an indication for C-section in 2 cases and labor induction in 1 case followed by postnatal AFL diagnosis based on resting ECG. Latest diagnosis was made in 11th day of life. All patients were born in good general condition at median gestational age of 39(36–41) weeks, birth weight of 3525 (2580–4160)g. In 8 patients AFL was registered postnatally with maximum heart rate of 225 (190–250)bpm. Six patients were treated with intravenous amiodarone with starting

dose of 5 mg/kg every 8 hours resulting in conversion to SR in all but 1 case, in which DCC was performed. In 3 cases conversion to SR was achieved with DCC with 1, 7 (0, 6–2)J/kg as primary mode of treatment (and no pharmacotherapy). All patients were discharged with normal ejection fraction (median of 69%, 61–74%) and sinus rhythm. In 7 patients follow-up of 1, 9 (1, 0–59, 6) months was available with no recurrence of arrhythmia.

**Conclusions:** In neonatal AFL conversion to SR can be safely achieved with both pharmacotherapy and DCC with good arrhythmia-free long-term prognosis. Due to limited data, lack of unequivocal guidelines resulting in inhomogeneous management, further multicenter studies are crucial for determination of optimal mode of treatment.

**Keywords:** Atrial flutter, neonatal arrhythmias, fetal arrhythmias

#### P-094

##### **Congenital diverticulum of the left ventricle, as an accidental finding in a healthy child. clinical case**

Olha Zborovska<sup>1</sup>, Andriana Malska<sup>2</sup>

<sup>1</sup>Department of Pediatrics №2, National Pirogov Memorial Medical University, Vinnytsya, Ukraine; <sup>2</sup>Department of Propedeutics of Pediatrics and Medical Genetics, Danylo Halytsky Lviv National Medical University

**Background and Aim:** Left ventricular diverticulum (LVD) is a rare congenital cardiac abnormality that occurs with the incidence of 0.05%. Embryologically, LVD is formed due to a close location of the structures from which the anterior part of the diaphragm and part of the primary pericardium are formed. The fusion between these structures leads to the formation of a diverticulum. This anomaly often becomes a part of the Pentalogy of Cantrell. **Aim:** to present a clinical case of a 6-month-old child with LVD, which was routinely diagnosed during 2D Echo-cardiographic examination.

**Method:** Clinically asymptomatic child underwent a 2D Echocardiographic examination, prior to the beginning of treatment of a juvenile hemangioma with  $\beta$ -blockers. The EKG showed sinus rhythm and ST segment elevation in the standard and left thoracic leads. 2D Echo revealed aneurysmal protrusion of the left ventricle of 26x17 mm, with thinned ventricular walls. MRI of the heart confirmed the muscular type of LVD in the apex. CT scan showed a pericardial and an anterior diaphragm defect, which allowed us to combine these findings into a partial Pentalogy of Cantrell.

**Results:** According to the literature surgical correction of LVD is not indicated for asymptomatic patients, such children require long-term observation. However, the presence of LVD is a risk factor for the development of the life-threatening ventricular arrhythmias, systemic thromboemboli in the presence of blood clots in the diverticulum cavity and its spontaneous rupture. The other dangerous factor for our patient is the presence of a defect in the diaphragm, which can be complicated by the hernia pinching, reflux esophagitis, gastrointestinal bleeding, etc. Therefore the decision of the surgical correction of the LVD was undertaken.

**Conclusions:** LVD is often associated with other cardiac and extracardiac abnormalities and is dangerous for its complications even in asymptomatic patients. The results of longitudinal observations of such patients show a significant increase in mortality among such patients with age due to the high risk of spontaneous diverticulum

rupture. Surgical correction is an important treatment of such patients with LVD in order to prevent complications.

**Keywords:** congenital heart defects, left ventricle diverticulum, children, surgical correction

#### P-096

##### **3D MRA with image-based navigation for highly-efficient imaging in patients with congenital heart disease- single center study**

Anastasia Fotaki<sup>1</sup>, Kuberan Pushparajah<sup>1</sup>, Karl Kunze<sup>2</sup>, Radhouene Neji<sup>2</sup>, Christopher Rush<sup>3</sup>, Rene Botnar<sup>1</sup>, Claudia Prieto<sup>1</sup>

<sup>1</sup>School of Biomedical Engineering and Imaging Sciences, King's College London, London, United Kingdom; <sup>2</sup>MR Research Collaborations, Siemens Healthcare Limited, Camberley, United Kingdom; <sup>3</sup>Guy's and St Thomas's Hospital, Adult Congenital Heart Disease Department

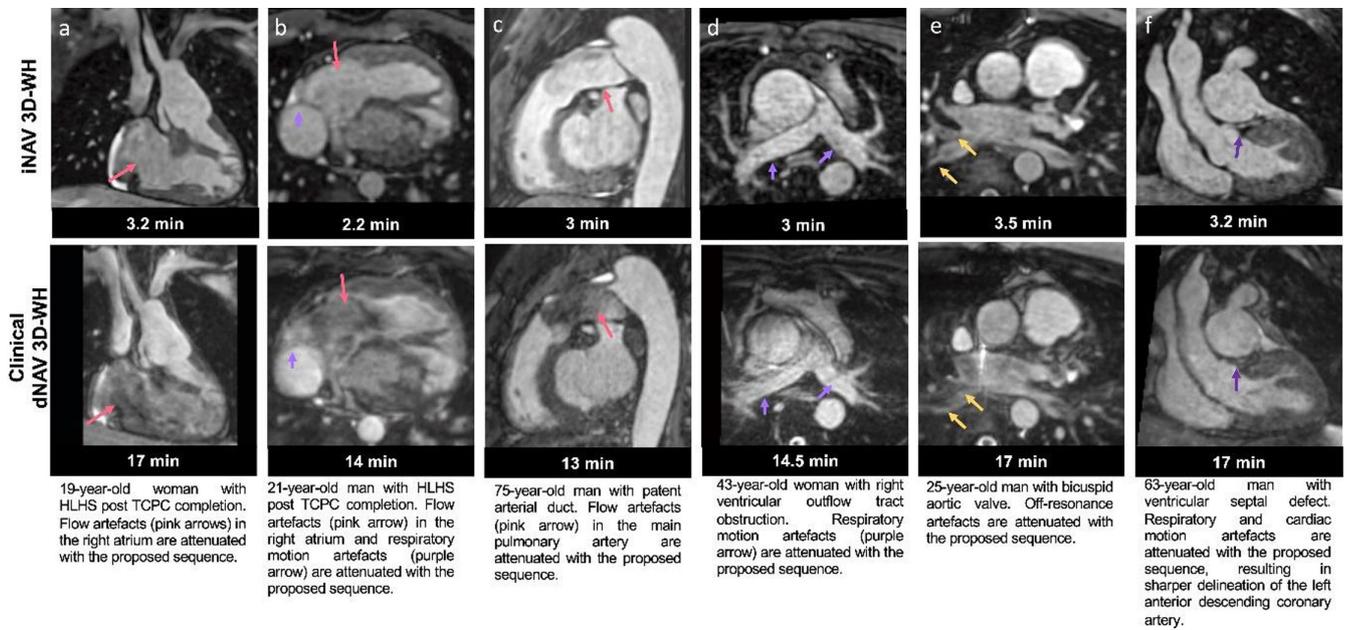
**Background and Aim:** 3D-whole heart(3D-WH) imaging sequences are utilised for the morphological evaluation of patients with Congenital Heart Disease(CHD)1, 2. Current approaches are limited by long scan times due to diaphragmatic navigators(dNAV) as well as frequent image artefacts2, 3, 4 We have proposed a framework that applies image-based navigation(iNAV) for efficient 3D-WH imaging5. The aim of this study is to validate the proposed framework against the clinical dNAV 3D-WH sequence.

**Method:** The iNAV 3D-WH research sequence was evaluated in 40 patients (32  $\pm$ 11 years old, 22 women) with CHD on a 1.5T system (MAGNETOM Aera, Siemens). The proposed approach included iNAV6 for respiratory and cardiac motion correction, 4-fold undersampled acquisition and an enhanced T2 preparation pulse for the attenuation of artefacts5. The clinical framework comprised of diaphragmatic gating(dNAV) and 2-fold undersampling. Resolution and acquisition window duration were same for both2. Two blinded experts, recorded their diagnostic confidence for sequential segmental analysis (Likert scale:1–4,  $\geq$  3: diagnostic dataset) and were asked to identify the presence of branch pulmonary arteries, aortic arch, coronary and pulmonary venous anatomical abnormalities. Each abnormality was scored on a 5-point Likert scale (1: Definitely not present, 2: Probably not present, 3: Unclear, 4: Probably present, 5: Definitely present). Scores of 1 and 2 were coded as abnormality is absent, and 4 and 5 were coded as abnormality is present. A score of 3 was coded as a misdiagnosis. Co-axial vascular dimensions at the level of the aortic root, mid right and left pulmonary arteries were compared using Bland-Altman analysis.

**Results:** Good quality depiction of all intrapericardiac structures(Fig. 1) was achieved with the proposed iNAV-3D-WH in shorter scan time than the conventional [3.2 $\pm$ 0.7min vs 15 $\pm$ 3.2 min,  $p < 0.0001$ ]. Diagnostic confidence was higher for the proposed than for the clinical sequence [4(4, 4) vs 4(3, 4)  $p = 0.009$ ]. The overall diagnostic accuracy for the diagnosis of the aforementioned abnormalities was higher for the research dequence(0.99 vs 0.94,  $P < 0.01$ ). Bland-Altman analysis demonstrated very good agreement in the co-axial vascular measurements between the two datasets, with negligible bias ( $< 0.7$  mm).

**Conclusions:** The iNAV T2prep-bSSFP sequence offers superior quality imaging to the conventional dNAV T2-prep bSSFP sequence in shorter acquisition time. Multi-center studies are warranted for forthcoming clinical adoption.

**Keywords:** 3D whole-heart MRI, scan efficiency, image-based navigation



**Figure 1** Comparison of the proposed image-based navigation 3D-WH images to the diaphragmatic-based navigation 3D-WH sequence. Flow related artefacts in areas of slow (a,b) and turbulent flow (c), respiratory and cardiac motion artefacts (b, d, f) along with off-resonance artefacts (e) are reduced with the proposed iNAV 3D-WH sequence.

**P-097**

**Echocardiographic findings in MIS-C patients**

*Ikrām Chamtouri<sup>1</sup>, Farah Thabet<sup>2</sup>*

<sup>1</sup>Cardiology B department, Fattouma Bourguiba University Hospital, Monastir, Tunisia; <sup>2</sup>Pediatrics department, Fattouma Bourguiba University Hospital, Monastir, Tunisia

**Background and Aim:** Background. Cardiovascular lesions are frequent MIS-C children. Even no clear myocardial damage has revealed at the acute phase of MIS-C in some children, cardiac involvement could be subtle. The aim of the study was to evaluate infra clinical myocardial involvement MIS-C patients by speckle tracking echocardiography at the early phase of MIS-C and six months later.

**Method:** Methods

This is a prospective study performed between March 2022 and september 2022. All consecutive children aged <16 years and hospitalized for MIS-C. MIS-C diagnosis was obtained by positive immunoglobulin M or G detection in serological sample. Consent of all children’s parents was obtained. Exclusion criteria were Children with already reduced left ventricle ejection fraction (LVEF)<50%, pulmonary embolism, myocarditis, cardiogenic shock, persistent arrhythmia and coronary artery aneurysm.

**Results:** Results. 27 children were enrolled with mean age was 5.4 ± 3 years. Compared to the early phase of MIS-C, there was no significant difference in conventional echocardiography parameters at 6-month follow-up. Both left and right ventricles global longitudinal strain (LVGLS and RVGLS) were reduced at the acute phase of MIS-C (> -22) in 45.6% and 56% of children respectively. Segmental strain was more damaged in the anterior wall. After 6 months, LVGLS and RVGLS returned to the normal values.

**Conclusions:** In MIS-C patients subclinical myocardial injury is common. However, during a short follow-up, there is a myocardial recovery.

**Keywords:** MIS-C, cardiac involvement, strain

**P-098**

**Molecular imaging of microcalcification in thoracic aortopathy: associations with high-risk features**

*Alexander James Fletcher<sup>1</sup>, Jennifer Nash<sup>2</sup>, Maaz Syed<sup>2</sup>, Gillian Macnaught<sup>3</sup>, Edwin van Beek<sup>3</sup>, Piotr Slomka<sup>5</sup>, Michelle Williams<sup>2</sup>, Marc Dweck<sup>2</sup>, Niki Walker<sup>4</sup>, David Newby<sup>2</sup>*

<sup>1</sup>Child Health, School of Medicine Dentistry and Nursing, University of Glasgow, Glasgow, United Kingdom; <sup>2</sup>Centre for Cardiovascular Science, University of Edinburgh, Edinburgh, United Kingdom; <sup>3</sup>Centre for Cardiovascular Science, University of Edinburgh, Edinburgh, United Kingdom; <sup>4</sup>Edinburgh Imaging Facility, Queens Medical Research Institute, University of Edinburgh, Edinburgh, United Kingdom; <sup>5</sup>Scottish Adult Congenital Cardiology Service, Golden Jubilee National Hospital, Clydebank, Glasgow, United Kingdom; <sup>5</sup>Cedars-Sinai Medical Centre, Department of Imaging (Division of Nuclear Cardiology), Los Angeles, USA

**Background and Aim:** In patients with thoracic aortopathy, vessel wall microcalcification has recently emerged as an early marker of aortic wall pathology, and has the potential to identify those with high risk disease that may be suitable for early intervention. In a prospective molecular imaging study (NCT04083118), we aim to explore, for the first time, the relationship between thoracic aortic wall microcalcification and biological features of high risk thoracic aortopathy.

**Method:** Patients with bicuspid aortic valve and age/sex matched controls without aortopathy underwent baseline demographic and haemodynamic assessment (blood pressure and applanation tonometry) as well as same-day hybrid  $^{18}\text{F}$ -sodium fluoride positron emission tomography-computed tomography ( $^{18}\text{F}$ -NaF PET-CT, a radiotracer which binds to microcalcification) and magnetic resonance imaging. The relationship between baseline ascending aortic  $^{18}\text{F}$ -NaF activity and demographic, aortic stiffness (stiffness index) and MRI-assessed aortic dilatation (indexed aortic diameter) was assessed (multivariable linear regression and Spearman's rho). The relationship to progression of aortic stiffness and aortic diameter to historical MRI data was also explored (Spearman's rho).

**Results:** Seventy-five patients with bicuspid aortic valve (age  $52.5 \pm 7.5$  years, 24% female) and 18 age/sex matched control subjects (age  $50.6 \pm 6.2$  years, 28% female) were included in the analysis. Those with bicuspid aortic valve and aortic dilatation had increased ascending aortic  $^{18}\text{F}$ -NaF activity compared with control subjects ( $1.11 \pm 0.06$  versus  $1.06 \pm 0.08$ ,  $p = 0.029$ , Figure) which remained significant after adjustment for age. An overall weak association was found between ascending aortic  $^{18}\text{F}$ -NaF activity and aortic diameter (Spearman's rho 0.24,  $p = 0.018$ ). There was a moderate

association between  $^{18}\text{F}$ -NaF activity and localised stiffness index (Spearman's rho,  $r = 0.3$ ,  $p = 0.01$ , Figure), which was present after adjustment for age and diameter ( $\beta = 11.03$ ,  $p = 0.019$ ). Of the 29 bicuspid aortic valve patients with historical MRI  $7.2 \pm 2.5$  years previously,  $^{18}\text{F}$ -NaF correlated with progression of aortic strain ( $r = -0.47$ ,  $p = 0.013$ ) but not diameter ( $r = 0.14$ ,  $p = 0.47$ )

**Conclusions:** We report higher ascending aortic  $^{18}\text{F}$ -NaF activity in patients with bicuspid aortic valve and aortic dilatation than control subjects. Further, ascending aortic  $^{18}\text{F}$ -NaF activity and was associated with baseline ascending stiffness index and progression of ascending aortic strain; hallmarks of biological remodelling. Whether  $^{18}\text{F}$ -NaF PET can prospectively predict dissection events requires further research.

**Keywords:** Aortopathy, bicuspid aortic valve, dissection, calcification, positron emission tomography, magnetic resonance imaging

### P-099

#### Hypoplastic left heart syndrome has asymmetric leaflet expansion and increased tenting volume of the tricuspid valve: A prospective 3DE study

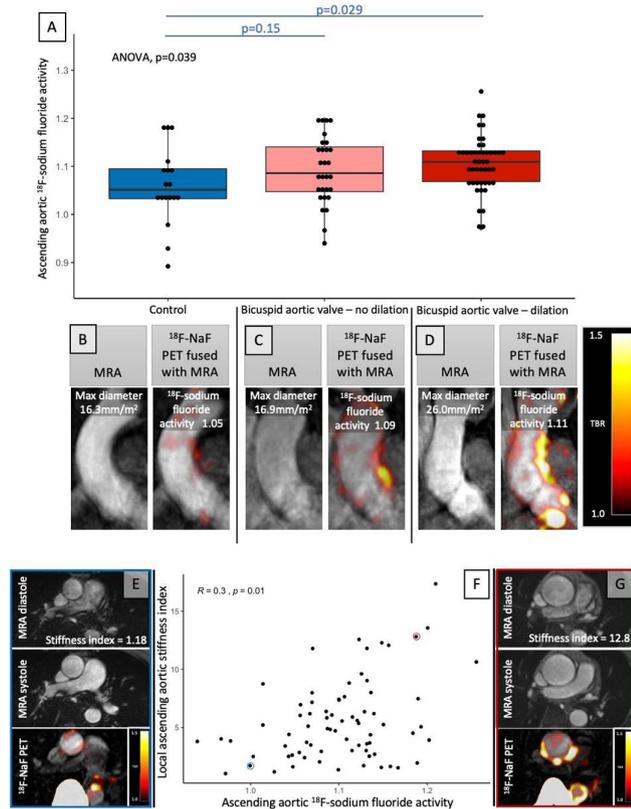
Martha Maria Moran, Sachie Shigemitsu, Tuqa Al Lawati, Nee Szcze Khoo, Timothy Colen, Richard Thompson, Justin Grenier, Daren Freed, Devilliers Jonker, Mohammed Alaklabi, Lily Lin  
Department of Pediatric Cardiology, Stollery Children's Hospital, University of Alberta, Edmonton, Canada

**Background and Aim:** Tricuspid regurgitation is associated with increased mortality in children with hypoplastic left heart syndrome (HLHS) while knowledge on its mechanisms is limited. We recently presented normal TV maturation in children which shows symmetrical leaflet size and expansion through childhood with slight progressive increase of tenting volume when indexed to body surface area (BSA). This study aims to compare differences between normal tricuspid valves (TV), HLHS TVs with no tricuspid regurgitation (TR) and those requiring repair (TVR) to further understand the pathophysiology of TV failure in HLHS.

**Method:** This is a cross-sectional study with 96 prospectively acquired three-dimensional echocardiograms of TV (33 healthy children, 33 HLHS no TR, 30 HLHS TVR) analyzed using a proprietary software for leaflet and annular quantification. TV leaflets were segmented into anterior (AL), septal (SL) and posterior (PL) analyzed in mid-systole to measure areas, prolapse and tenting volumes as well as TV annular size, eccentricity index (ratio of major and minor axis) and bending angle. Variables, where applicable, were indexed to BSA. Between group comparison the Kruskal-Wallis test was used. When significantly different was detected, post hoc comparisons between age groups using the Dunn's test were performed.

**Results:** When compared to healthy children, HLHS with no TR had greater annuli dimensions, similar eccentricity index with preserved annular bending angle. HLHS leaflets had proportionally larger AL, smaller SL and overall increased tenting volume. In addition, HLHS requiring TVR had further annular dilation, AL expansion and greater prolapse (see Table).

**Conclusions:** HLHS TV are larger and has greater tenting volume with an asymmetric expansion of the AL followed by PL when compared to normal TV. Given differences in loading conditions, these changes may reflect "normal" adaptive mechanisms to maintain competency as TV annulus expands. The finding of further TV annular dilation, AL expansion and greater prolapse in HLHS requiring TVR, suggests a maladaptive process impacting



(A) Patients with bicuspid aortic valve and proximal dilatation of the aorta had significantly increased ascending aortic  $^{18}\text{F}$ -sodium fluoride activity than controls. (B-D) Visual representation of the median case associated with each group. (E) A patient with low stiffness index and  $^{18}\text{F}$ -sodium fluoride content. (F) A moderate association was found between aortic stiffness index and ascending aortic  $^{18}\text{F}$ -sodium fluoride activity. (G) An example of a patient with high stiffness index and high  $^{18}\text{F}$ -sodium fluoride activity in the ascending aorta. MRA = magnetic resonance angiogram,  $^{18}\text{F}$ -NaF PET =  $^{18}\text{F}$ -sodium fluoride positron emission tomography

disproportionally the AL, may play an important role in the development of TR in HLHS.

**Keywords:** 3DE, HLHS, Tricuspid valve

midazolam in a large cohort of children and adolescents undergoing cardiovascular magnetic resonance imaging (CMR).

*Method:* CMR sedation protocols from 2010 to 2020 were retrospectively analysed. Sedation incidents, peripheral oxygen

**Table:** Comparison of 3DE parameters between normal TV, HLHS TV that did not undergo TVR and HLHS TV that required TVR. Values expressed as median (25th, 75th percentile). NS denotes non-significance.

Variables	Normal TV (N=33)	HLHS TV no TVR (N=33)	HLHS TV requiring TVR (N=30)	Kruskal-Wallis p-value	Post-hoc comparison of mean rank between groups Dunn's test p-value		
					Normal vs HLHS no TVR	Normal vs HLHS TVR	HLHS no TVR vs HLHS TVR
Age (months)	11.8 (3.5, 28.6)	24.7 (3.5, 36.0)	23.2 (3.9, 39.9)	NS			
Body surface area (BSA) (m2)	0.47 (0.35, 0.60)	0.36 (0.32, 0.56)	0.49 (0.31, 0.58)	NS			
Male gender (%)	0.60	0.7	0.67	NS			
TR grade (1-4)	1.0 (1.0, 1.0)	1.0 (1.0, 2.0)	3.0 (3.0, 3.0)	<0.0001	NS	<0.0001	<0.0001
<b>Annular parameters</b>							
TV total annular area (cm <sup>2</sup> )	1.8 (1.3, 2.6)	3.3 (3.0, 4.4)	5.1 (4.0, 6.6)	<0.0001	<0.0001	<0.0001	0.01
TV annular circumference (cm)	4.9 (4.1, 5.8)	6.8 (6.3, 7.6)	8.2 (7.3, 9.3)	<0.0001	<0.0001	<0.0001	0.012
TV annulus Anteroposterior dimension (cm)	1.4 (1.2, 1.6)	1.9 (1.5, 2.1)	2.2 (2.0, 2.7)	<0.0001	0.0011	<0.0001	0.0025
TV annulus lateral dimension (cm)	1.6 (1.3, 1.9)	2.3 (2.0, 2.6)	2.7 (2.5, 3.1)	<0.0001	<0.0001	<0.0001	0.0113
TV annular eccentricity index	1.1 (1.1, 1.2)	1.2 (1.2, 1.3)	1.2 (1.1, 1.3)	0.0005	0.0003	NS	NS
Bending angle (degrees)	156.1 (150.0, 161.7)	152.5 (144.8, 159.4)	158.2 (151.5, 162.4)	NS (0.06)			
<b>Leaflet parameters</b>							
Total leaflet area (cm <sup>2</sup> )	1.8 (1.3, 2.7)	3.7 (3.3, 4.8)	5.5 (4.3, 7.3)	<0.0001	<0.0001	<0.0001	0.0237
AL leaflet area (cm <sup>2</sup> )	0.6 (0.5, 0.8)	1.8 (1.4, 2.6)	3.1 (1.9, 3.8)	<0.0001	<0.0001	<0.0001	0.0393
PL leaflet area (cm <sup>2</sup> )	0.5 (0.4, 1.0)	1.2 (0.9, 1.5)	1.4 (1.0, 2.2)	<0.0001	0.0006	<0.0001	NS
SL leaflet area (cm <sup>2</sup> )	0.6 (0.4, 0.9)	0.8 (0.4, 1.3)	1.2 (0.8, 1.6)	0.0043	NS	0.0031	NS
AL/total leaflet area ratio	0.35 (0.27, 0.42)	0.46 (0.41, 0.57)	0.51 (0.42, 0.62)	<0.0001	<0.0001	<0.0001	NS
PL/total leaflet area ratio	0.34 (0.26, 0.39)	0.31 (0.24, 0.37)	0.23 (0.18, 0.34)	0.0092	NS	0.0067	NS
SL/total leaflet area ratio	0.32 (0.26, 0.36)	0.22 (0.14, 0.26)	0.21 (0.16, 0.29)	<0.0001	<0.0001	0.0001	NS
Total leaflet prolapse indexed volume (µl/cm <sup>2</sup> )	0.0003 (0.0001, 0.003)	0.0006 (0.000005, 0.004)	0.02 (0.002, 0.04)	0.0009	NS	0.0021	0.005
AL prolapse indexed vol	0.0002 (0, 0.002)	0.0004 (0.000004, 0.006)	0.009 (0.0006, 0.04)	0.0005	NS	0.0004	0.0214
PL prolapse indexed vol	0.0001 (0, 0.001)	0.000008 (0, 0.0005)	0.0002 (0, 0.03)	NS			
SL prolapse indexed vol	0.0003 (0, 0.001)	0.000003 (0, 0.002)	0.0007 (0.0000006, 0.01)	NS			
Total leaflet tethering indexed volume (µl/cm <sup>2</sup> )	0.05 (0.02, 0.08)	0.1 (0.09, 0.2)	0.08 (0.04, 0.2)	<0.0001	<0.0001	0.0208	NS
AL tethering vol (µl)	0.05 (0.02, 0.08)	0.1 (0.07, 0.2)	0.08 (0.03, 0.2)	0.0001	<0.0001	NS	NS
PL tethering vol (µl)	0.06 (0.02, 0.08)	0.1 (0.08, 0.2)	0.1 (0.04, 0.2)	<0.0001	<0.0001	0.0201	NS
SL tethering vol (µl)	0.05 (0.03, 0.08)	0.1 (0.09, 0.2)	0.1 (0.05, 0.2)	<0.0001	<0.0001	0.0008	NS

**P-100**

**Retrospective study of the safety of deep sedation during cardiovascular MRI in children and adolescents with congenital and acquired heart disease**

Marcel Schmidt<sup>1</sup>, Sylvia Krupickova<sup>2</sup>, Anselm Uebing<sup>1</sup>, Philipp Wegner<sup>1</sup>, Dominik Gabbert<sup>1</sup>, Inga Voges<sup>1</sup>

<sup>1</sup>Department of Congenital Heart Disease and Paediatric Cardiology, University Hospital Schleswig-Holstein, Campus Kiel; <sup>2</sup>Department of Pediatric Cardiology, Royal Brompton Hospital, London, UK

*Background and Aim:* Deep sedation with propofol in young patients with cardiovascular disease still confronts physicians with uncertainties. In this study, we assessed the safety and efficacy of a standardised deep sedation protocol including propofol and

saturation, heart rate, doses of propofol and midazolam and the Richmond Agitation-Sedation Scale (RASS) were documented. Furthermore, cardiac diagnoses as well as the severity of congenital cardiac defects defined as simple, moderate and severe (Warnes et al. 2001) were noted.

*Results:* Protocols of 523 patients (5.9 ± 3.7 years) were evaluated. 374 (71.5%) patients had a severe, 121 (23.1%) had a moderate, 4 (0.8%) had a mild cardiac defect and 24 (4.6%) patients had acquired or non-structural cardiac disease. Out of the group with severe cardiac defects, 283 (75.7%) patients had a single ventricle with 200 (53.5%) being hypoplastic left heart syndrome patients. To achieve a sedation level on the RASS of 2, sedation was performed using a combination of propofol and midazolam in 516 (98.7%) patients. On average, 154.3±90.9 mg of propofol and

1.5±0.7 milligrams of midazolam were given to achieve a RAS of 2 throughout the scan.

Eight sedation incidents (1,5%) occurred. The cases were limited to anaphylactic reactions, coughing fits, unexpected oxygen saturation drops and a questionable aspiration in one patient. None of those patients needed intubation and/or intensive care treatment.

31 (5.9%) studies were aborted due to patient agitation, extravasating venous pathways or coughing attacks, but for most patients relevant CMR data were already acquired. Mild cardiac arrhythmias such as bradycardia and extrasystoles occurred in three patients (0.6%).

**Conclusions:** Deep sedation with propofol and midazolam for CMR seems to be a safe and feasible way even in children and adolescents with severe congenital cardiac defects.

**Keywords:** Cardiovascular magnetic resonance imaging, sedation

## P-101

### Patency of neonatally identified interatrial communications in young children

Sofie Dannesbo<sup>1</sup>, Anna Maria Dehn<sup>2</sup>, Ruth Ottilia Birgitta Vøgg<sup>3</sup>, Hæarter A Boyd<sup>4</sup>, Finn Stener Jørgensen<sup>5</sup>, Vibeke Hjortdal<sup>2</sup>, Anne Sophie Sillesen<sup>1</sup>, Anna Axelsson Raja<sup>1</sup>, Niels Vejstrup<sup>1</sup>, Kasper Iversen<sup>3</sup>, Henning Bundgaard<sup>1</sup>

<sup>1</sup>Department of Cardiology, Copenhagen University Hospital Rigshospitalet, Denmark; <sup>2</sup>Department of Cardiothoracic Surgery, Copenhagen University Hospital Rigshospitalet, Copenhagen, Denmark; <sup>3</sup>Department of Cardiology, Copenhagen University Hospital Herlev, Copenhagen, Denmark; <sup>4</sup>Department of Epidemiology Research, Statens Serum Institut, Copenhagen, Denmark; <sup>5</sup>Fetal Medicine Unit, Department of Obstetrics and Gynecology, Copenhagen University Hospital Hvidovre, Hvidovre, Denmark

**Background and Aim:** Atrial septal defect (ASD) of the secundum type is one of the most common congenital heart defects. In newborns, it is challenging to distinguish ASDs from normal physiological interatrial communications (IACs) within the oval fossa, i.e. patency of the oval foramen (PFO). On this basis, we recently developed an algorithm for classification of neonatal IACs identified by transthoracic echocardiography. According to the algorithm, an IAC are defined as an ASD if the defect are ≥4 mm, has a location in the inferior part of the septum or if multiple communications are present. In this study, we investigate the patency of neonatally identified IACs categorized by the algorithm as ASDs or PFOs in young children.

**Method:** Children participating in the Copenhagen Baby Heart Study, who had an IAC detected during the first 30 days after birth, are included for a follow-up echocardiogram at 4–5 years of age. We aim to include 600 children with a neonatally identified IAC categorized as PFO and 600 categorized as ASD according to the algorithm. We exclude children with other types of congenital heart disease, twins, children born prematurely or small for gestational age, and children exposed to maternal diabetes or hypertensive disorders of pregnancy. The follow-up echocardiograms are assessed for patency of an IAC by a single operator blinded to the neonatal IAC subtype. A patent IAC are defined as the presence of a color Doppler flow crossing the atrial septum in combination with either a visual defect on the transectional image or flow acceleration in the color Doppler signal.

**Results:** Data collection is ongoing. Currently, 152 children (median age 4.7 [interquartile range 4.5–5.0] years, 63% female) have been examined. Neonatally, the IACs were classified as ASDs in 57 newborns and PFOs in 95 newborns. The follow-up echocardiograms showed patency of the IAC in 17 of the children with an ASD (30%) and 13 of the children with PFO (14%), ( $p = 0.015$  for difference).

**Conclusions:** These preliminary findings show a significantly higher patency of neonatal ASDs versus neonatal PFOs in children examined at 4–5 years of age. This supports the value of the novel diagnostic algorithm.

**Keywords:** Interatrial communication, Atrial septal defect, Patent foramen ovale, Echocardiography, The Copenhagen Baby Heart Study

## P-102

### Management of incidentally discovered huge right atrial appendage aneurysm

Mohamed Metwally Abdelal, Mustafa Hassan Alqabandi  
Chest Disease Hospital Kuwait

**Background and Aim:** Aneurysms of the right atrial appendage are extremely rare, particularly in pediatric patients. Most of the RAA aneurysms are congenital but genetic predisposition has not been confirmed. It is important because of its complications such as arrhythmia, life-threatening systemic and pulmonary thromboembolism and rupture

**Method:** An 8-year-old boy was admitted to general pediatric ward with short febrile illness.

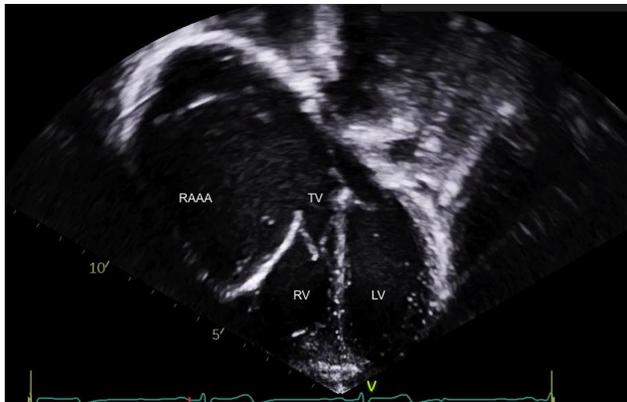
Referred to cardiologist due to significant cardiomegaly in chest X-ray. There was no past medical history of note. Cardiac examination was unremarkable. Transthoracic and transesophageal echocardiography revealed the presence of a large, thin-walled aneurysmal structure in continuity with the free wall of the right atrium, discrete from the caval veins, At the anatomical site of the right atrial appendage measuring 6.2 x 6.6 cm.

CMR showed Aneurysmal dilatation of the right atrial appendage, measuring 10 cm x 8.5 cm x 6.5 cm

**Results:** Even though the condition was accidentally discovered, and the child was asymptomatic, there was increasing risk of paradoxical embolism due to the presence of patent foramen ovale. In addition to the compression over the tricuspid valve which prevents its proper growth. Additionally, the possibility of other complications like arrhythmias or rupture were adding more risks. Thus, after a detailed discussion between the cardiologists and the surgeons, the decision was for surgical resection of the aneurysm.

**Conclusions:** The management of right atrial appendage aneurysms is a matter of debate, because the long-term outcomes of conservative versus surgical treatments have not yet been studied. In some studies, surgical treatment was effective in preventing thromboembolisms and lowering the risk of atrial arrhythmia, which is one of the most common complications of these aneurysms. In other studies, progressive increasing in size of the aneurysm and tricuspid valve compression was an indication for surgery. In cases in which the right appendage is affected, there are no other associated defects, and the patient is asymptomatic, management is more controversial

**Keywords:** Right atrial appendage aneurysm, Congenital heart disease



**Figure 1** 2D echocardiography four chamber view. Showing the large RAA Compressing RV AND TV.

**P-103**

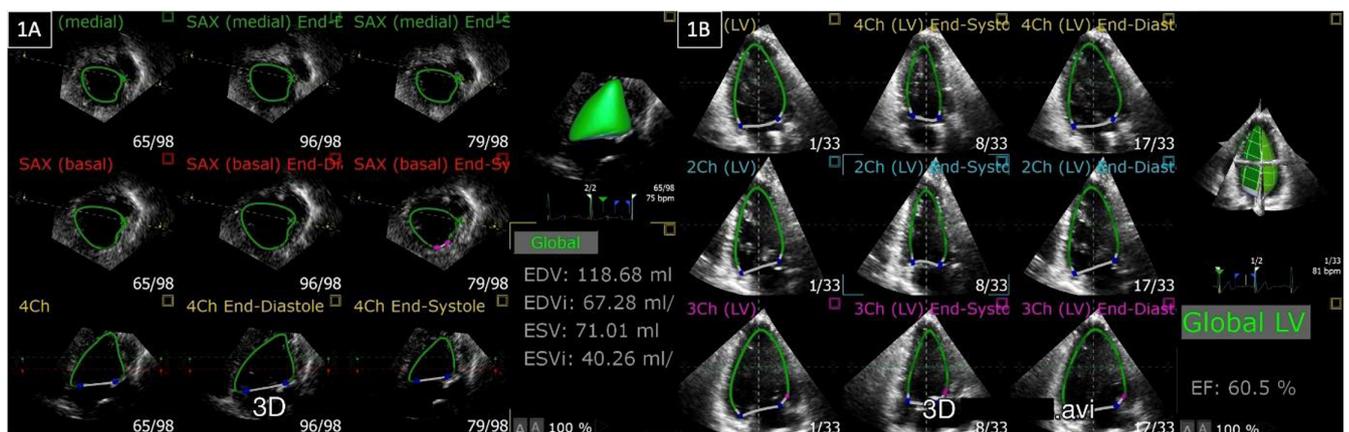
**Correlation between 3D-echocardiography and cardiopulmonary fitness in patients with univentricular heart: A multicentre prospective study**

Pascal Amedro<sup>1</sup>, Valentin Femenia<sup>1</sup>, Sophie Guillaumont<sup>2</sup>, Charlene Bredy<sup>3</sup>, Martina Avesani<sup>1</sup>, Xavier Iriart<sup>1</sup>, François Roubertie<sup>1</sup>, Bertrand Leobon<sup>1</sup>, Marie Christine Picot<sup>4</sup>, Jean Benoit Thambo<sup>1</sup>

<sup>1</sup>Pediatric and Congenital Cardiology Department, M3C National Reference CHD Centre, Bordeaux University Hospital, IHU Liryc, INSERM 1045, University of Bordeaux, Bordeaux, France; <sup>2</sup>Pediatric Cardiology and Rehabilitation Unit, St-Pierre Institute, Palavas-Les-Flots, France.; <sup>3</sup>Pediatric and Congenital Cardiology Department, M3C Regional Reference CHD Centre, Montpellier University Hospital, Montpellier, France; <sup>4</sup>Epidemiology and Clinical Research Department, University Hospital, Montpellier, France.

**Background and Aim:** Prognosis of patients with a functional single ventricle (SV) has improved, with better cardiopulmonary fitness, health-related quality of life (HRQoL), and survival. Conventional echocardiography remains the first-line technique in SV follow-up. Three-dimensional (3D) echocardiography

3D-echocardiography tracking for single ventricle



**Legend:** The green lines represent the endocardial tracking for morphological right SV (1A) and left SV (1B). Abbreviations: SV, single ventricle; SAX, short axis view; 4Ch, 4 chamber view; 2Ch, 2 chamber view; 3Ch, 3 chamber view; EF, ejection fraction; EDV, end-diastolic volume; EDVi, indexed end-diastolic volume; ESV, end-systolic volume; ESVi, indexed end-systolic volume; LV, left ventricle.

has shown recent interest in congenital cardiology, but its ability to predict functional status in SV remains unknown.

In this study, we aimed to evaluate, in patients with SV, the association between 3D-echocardiography parameters and functional status determined by cardiopulmonary fitness.

**Method:** Children and adults with functional SV were prospectively enrolled in this multicentre study. Cardiopulmonary fitness was assessed by cardiopulmonary exercise test (CPET) with measures of maximum oxygen uptake (VO<sub>2</sub>max) and ventilatory efficiency (VE/VCO<sub>2</sub> slope). 3D-echocardiography was performed with off-line reproducibility analyses using TomTec Arena software. HRQoL was assessed using the SF36 questionnaire.

**Results:** A total of 33 patients were screened, and 3D-echocardiography analyses were feasible in 22 subjects (mean age 28±9 years). 3D-echocardiography ejection fraction correlated with percent-predicted VO<sub>2</sub>max (r = 0.64, P<0.01), VE/VCO<sub>2</sub> slope (r = -0.41, P = 0.05), 2D- echocardiography ejection fraction (r = 0.55, P<0.01), and HRQoL physical functioning dimension (r = 0.56, P = 0.04). 3D-echocardiography indexed end-systolic volume correlated with percent-predicted VO<sub>2</sub>max (r = -0.45, P = 0.03), and VE/VCO<sub>2</sub> slope (r = 0.65, P<0.01). 3D- echocardiography reproducibility was good.

**Conclusions:** SV ejection fraction and volumes measured by 3D-echocardiography correlated with cardiopulmonary fitness, as determined by two main prognostic CPET parameters, e.g., the VO<sub>2</sub>max and the VE/VCO<sub>2</sub> slope. Despite a good reproducibility, the feasibility of 3D- echocardiography remained limited. 3D-echocardiography may be of interest in SV follow-up, provided the technique and analysis software are improved.

**Keywords:** single ventricle, three-dimensional echocardiography, aerobic fitness, maximum oxygen uptake, congenital heart disease.

**P-104**

**Prevalence and subtypes of interatrial communications in 12, 718 newborns**

Elisabeth Maria Blixenkroner Møller<sup>1</sup>, Sofie Dannesbo<sup>2</sup>, Anna Maria Dehn<sup>3</sup>, Christian Alexander Pihl<sup>1</sup>, Anne Sophie Sillesen<sup>1</sup>, Ruth Otilia Birgitta Voegg<sup>1</sup>, Anna Axelsson Raja<sup>3</sup>, Steven Colan<sup>4</sup>, Luc Mertens<sup>5</sup>, Niels Vejstrup<sup>3</sup>, Henning Bundgaard<sup>3</sup>, Kasper Karmark Iversen<sup>1</sup>

<sup>1</sup>Copenhagen University Hospital, Herlev-Gentofte, Copenhagen;  
<sup>2</sup>Copenhagen University Hospital, Herlev-Gentofte, Copenhagen and  
 Copenhagen University Hospital, Rigshospitalet, Copenhagen;  
<sup>3</sup>Copenhagen University Hospital, Rigshospitalet, Copenhagen; <sup>4</sup>Boston  
 Children's Hospital, Boston, USA; <sup>5</sup>The Hospital for Sick Children,  
 Toronto, Canada

**Background and Aim:** The prevalence of interatrial communications (IACs) (i.e. patent foramen ovale (PFO) or atrial septal defect (ASD)) in newborns has previously been reported to be between 24% and 92%, but the topic is lacking a universal classification of IACs. We have previously proposed an echocardiographic diagnostic algorithm for the classification of IACs with inter- and intra-observer agreements superior to standard assessment by experts in pediatric echocardiography. The aim of this study is to determine the prevalence of subtypes of IACs in newborns in a large unselected cohort.  
**Method:** Echocardiograms of newborns (age 0–30 days) included in The Copenhagen Baby Heart Study, a large, prospective population study (n>25, 000), were analyzed according to our new diagnostic IAC algorithm, classifying IACs into three subtypes of PFOs and three subtypes of ASDs. An IAC was defined according to the algorithm as color Doppler flow crossing the atrial septum with either a visible communication on the 2D image or flow acceleration in the color Doppler signal.  
**Results:** Of the 16, 276 echocardiograms analyzed, 3, 616 (22.2%) were excluded due to suboptimal visualization of the atrial septum and 9 (<0.1%) due to severe heart disease, leaving 12, 718 newborns (median age 12 [IQR 8;15] days, 48.1% female) included in the study. An IAC was present in 10, 033 (78.9%) newborns. According to the algorithm, 9, 274 (72.9%) of the newborns had a PFO, while 759 (6.0%) fulfilled criteria for having an

ASD. Of PFOs, 4, 451 (47, 9%) was only detectable with color Doppler (flow-defined PFO), 2, 391 (25, 8%) had a visible channel-like structure on 2D (channel-like PFO), and 2, 442 (26, 3%) had a size of <4 mm (size-defined PFO). Of ASDs, 368 (48.5%) had a defect size ≥4 mm (size-defined ASD), 364 (48.0%) had more than one interatrial communication (fenestrated ASD), and 27 (3.6%) had the defect located in the lower third part of the septum (location-defined ASD).

**Conclusions:** An IAC was present in almost 80% of newborns aged 0–30 days. ASDs were present in 6%. Future follow-up studies of this cohort of children are expected to provide clinically useful information on the natural history and hemodynamic impact of these thoroughly categorized ASD and PFO subtypes.

**Keywords:** Echocardiography, interatrial communication, atrial septal defect, patent foramen ovale, Copenhagen Baby Heart Study

**P-105**

**Local arterial stiffness assessment on humans: comparison between the bramwell-hill equation and direct pulse wave imaging by ultrafast ultrasound**

Rahma Rasouli<sup>1</sup>, Jerome Baranger<sup>2</sup>, Cameron Slorach<sup>2</sup>, Minh Nguyen<sup>2</sup>, Patrick Segers<sup>3</sup>, Vitor Guerra<sup>2</sup>, Mathieu Pernot<sup>4</sup>, Wei Hui<sup>2</sup>, Luc Mertens<sup>2</sup>, Olivier Villemain<sup>2</sup>

<sup>1</sup>Faculty of Medicine, University of Toronto, Toronto, Canada;  
<sup>2</sup>Department of Paediatrics, Hospital for Sick Children, Toronto, Canada;  
<sup>3</sup>Institute Biomedical Technology, Ghent University, Ghent, Belgium;  
<sup>4</sup>Physics for Medicine Paris, PSL Research University, Paris, France

Algorithm used in Methods, and graph showing Results

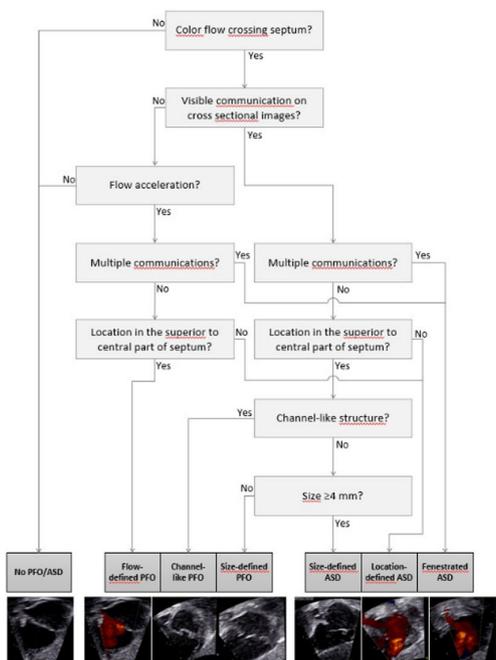


Figure 1: The new diagnostic algorithm for classification of interatrial communications in newborns. Here displayed with image examples.

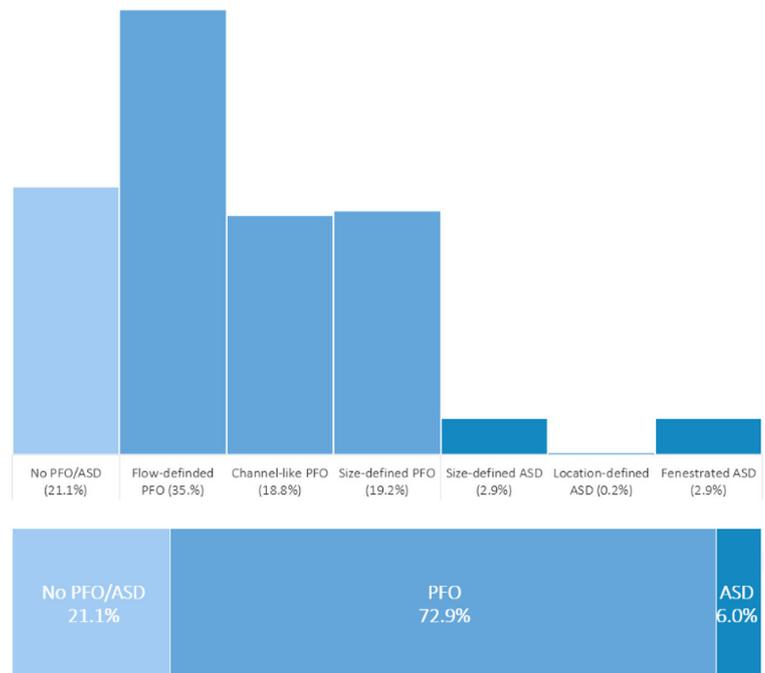


Figure 2: Distribution of subtypes.

Figure 1: The new algorithm used in determining the type of IAC found on TTE image.

**Background and Aim:** Pulse wave velocity (PWV), which is the propagation speed of a pressure wave along an arterial wall, is the clinical surrogate to assess arterial stiffness non-invasively. Currently, the most common clinical tool for locally measuring PWV is the theoretical Bramwell-Hill (B-H) equation. Ultrafast ultrasound imaging (UUI) is able to directly measure local PWV through imaging with high temporal resolution. This study compares local carotid PWV measured by the B-H equation and UUI at rest and during exercise.

**Method:** 21 healthy volunteers aged between 13 and 49 years (mean 25.9 +/- 8.9 years) were enrolled in an exercise trial at the Hospital for Sick Children. Demographic and clinical data were collected from the participants, and carotid properties (intima-media thickness, distensibility, diameter,  $\beta$ -index) were obtained by ultrasound. PWV assessment by B-H equation and by UUI were performed at rest and at submaximal exercise. PWVs assessed by UUI were measured in early systole (systolic foot, PWV-SF) and in end-systole (dicrotic notch, PWV-DN). Only one PWV is obtained by B-H (PWV-BH). Data were analysed using Students t-tests, Bland-Altman plots, and Pearson correlation analysis.

**Results:** On average, the B-H equation significantly underestimated the PWV compared to PWV-SF measurement in both rest (4.25 vs 6.65 m/s) and exercise (4.48 vs. 8.13 m/s) conditions ( $p < 0.001$ ). On Bland-Altman plot analysis, the underestimation of the B-H method is more pronounced at higher velocities ( $R^2 = 0.58$ ). At rest and during exercise, PWV-SF was positively correlated to multiple parameters including age ( $p < 0.05$ ), weight ( $p < 0.01$ ), and diastolic parameters [minimum carotid diameter ( $p < 0.01$ ) and diastolic blood pressure ( $p < 0.05$ )], but PWV-BH was not ( $p > 0.05$  for each of these parameters). PWV-DN were significantly higher than PWV-SF, at rest ( $p < 0.01$ ) and during exercise ( $p < 0.01$ ).

**Conclusions:** We observed significant differences between PWV-SF, PWV-DN, and PWV-BH. Due to the physiological variation of the PWV during the cardiac cycle, these parameters appear to estimate different stiffness states of the arterial wall. PWV-SF assessed by UUI is correlated with diastolic blood pressure and passive stiffness properties of the arterial wall.

**Keywords:** Ultrafast ultrasound imaging, Arterial stiffness, Pulse wave velocity, Bramwell-hill equation

#### P-106

##### **Echocardiographic normative reference intervals for left ventricular parameters in 13, 454 infants – a copenhagen baby heart study**

R Ottilia Birgitta Voegg<sup>1</sup>, Anne Sophie Sillesen<sup>1</sup>, Jan Wohlfahrt<sup>2</sup>, Christian Alexander Pihl<sup>1</sup>, Jakob Boesgaard Norsk<sup>1</sup>, Anna Axelsson Raja<sup>3</sup>, Niels Vejstrup<sup>3</sup>, Eleni Elia<sup>4</sup>, Lynn Sleeper<sup>4</sup>, Steven Colan<sup>4</sup>, Kasper Karmark Iversen<sup>1</sup>, Heather Allison Boyd<sup>2</sup>, Henning Bundgaard<sup>4</sup>  
<sup>1</sup>Department of Cardiology, Herlev Hospital, Copenhagen University Hospital, Copenhagen, Denmark; <sup>2</sup>Department of Epidemiology Research, Statens Serum Institut, Copenhagen, Denmark; <sup>3</sup>The Heart Centre, Rigshospitalet, Copenhagen University Hospital, Copenhagen, Denmark; <sup>4</sup>Department of Cardiology, Boston Children's Hospital, Boston, United States of America

**Background and Aim:** Reference intervals are required to distinguish between normal and pathological structural and functional cardiac variation. Left ventricular parameters are particularly important in the diagnostic work-up of infants suspected of congenital heart diseases and are also predictors of clinical outcome. However, data from healthy infants are limited, and current reference intervals provide an inadequate approximation of normal reference ranges.

The aim of the present study was to develop neonatal normative reference intervals and Z-scores for echocardiographic measurements of left ventricular structure and function based on a large group of healthy newborns.

**Method:** The study population included 13, 454 healthy infants from the Copenhagen Baby Heart Study (CBHS) who were born at term (37 to 42 completed weeks' gestation) to healthy mothers (excluding mothers with maternal pre-gestational or gestational diabetes, hypertensive disorders of pregnancy, systemic connective tissue disorders, neurological disorders, psychiatric disorders, kidney disease, congenital heart disease), had an echocardiogram performed within 30 days of birth, and did not have congenital heart disease at CBHS examination (excluding children with septal defects, valve disease, conotruncal defect, other).

We developed nomograms for ten left ventricular parameters by modelling these measures as a function of body surface area through joint modelling of the mean, standard deviation, skewness and kurtosis. We then evaluated the impact of infant sex and age at echocardiography on model fit.

**Results:** Infants in the study population (48.5% females) had a median body surface area of 0.23 m<sup>2</sup> (interquartile range 0.22–0.25 m<sup>2</sup>) and a median age of 12.0 days (interquartile range 8.0–15.0 days) at examination. All normative reference intervals performed well in both sexes without stratification on infant sex. In contrast, creation of separate reference models for infants examined <7 days of age and those examined at 7–30 days of age was necessary to optimize reference intervals performance.

**Conclusions:** This study provides normative reference intervals and Z-scores for ten clinically widely used echocardiographic measures of left ventricular structure and function based on a large cohort of newborns. These results provide highly needed reference material for clinical application by paediatric cardiologists serving newborn populations.

**Keywords:** Echocardiography, paediatric, normative reference intervals, Z-scores, cardiovascular growth

#### P-108

##### **Hypoplastic left heart syndrome and congenital heart block, a rare association in a newborn.**

Andriana Malska<sup>1</sup>, Olha Kuryliak<sup>2</sup>

<sup>1</sup>Department of Propedeutics of Pediatrics, Danylo Halytsky Lviv National Medical University; <sup>2</sup>Lviv Regional Children's Hospital OHMATDYT

**Background and Aim:** Hypoplastic left heart syndrome (HLHS) and congenital complete heart block (CCHB) are a rare association and very limited amount of literature data is available on this combined pathology.

CCHB is a rare condition with an incidence of 1 of 20, 000 live births, whereas HLHS occurs more frequently in - 1 of 5000 live births. We present a clinical case of unusual association of CCHB and a HLHS in a newborn.

**Method:** A newborn boy was urgently admitted to the neonatal ICU directly from the maternity hospital with the heart rate of 40 beats per minute. An urgent EKG and Echo-cardiography were performed. ECG revealed – III-rd stage complete AV block.

On Echocardiography a Hypoplastic left heart syndrome with mitral and aortic atresia, aortic arch hypoplasia and unique atrium and pulmonary artery dilatation were diagnosed.

Child was born on the 37-th week of gestation, from 11- th pregnancy, by physiological delivery, with APGAR score of 8\8. All prenatal ultrasounds did not reveal any pathology.

**Results:** Child was started on infusion of prostaglandin E and inotropic therapy.

After consultation by cardiac surgeons, surgical correction was denied due to the presence of complete AV block. Patient was intubated due to the progressive respiratory and heart failure, which eventually led to his death on the third day of life.

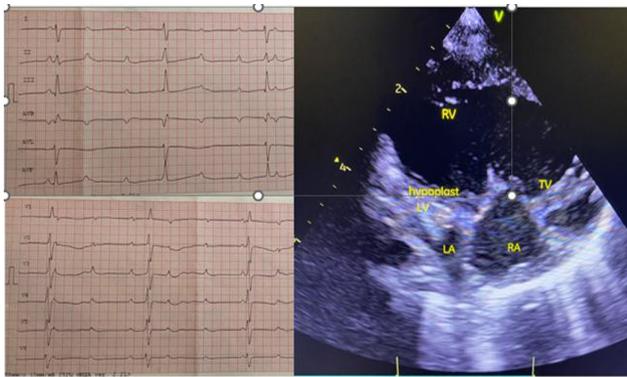
**Conclusions:** The etiologic relation of these two pathologies remains unclear. According to the literature data 70–90% of cases of isolated CCHB are caused by maternal anti-Ro and anti-La antibodies, which cross the placenta and lead to fibrosis of the AV node or by genetic defects, such as mutations in the SCN5A gene. Other theories suggest that compromised coronary blood flow in late fetal life could be a cause of CCHB.

In our case, maternal auto-antibody titers were negative. It is possible that the heart block could be linked to the structural heart disease – HLHS.

Only two similar cases of such combination are described in the literature.

Autopsy was denied by the family; therefore, the cause of these pathologies remains unknown.

**Keywords:** Hypoplastic left heart syndrome, complete congenital atrio-ventricular block, neonate.



**Figure 2:** Graph showing the distribution of the subtypes of IACs.

#### P-109

##### **A rare case of pulmonary atresia intact ventricular septum with a right ventricle-dependent coronary circulation through a large coronary fistula**

Mohamed Metwally Abdelaal, Sumaya Mahmoud Ibrahim, Ahmed Nabil Soliman, Manal Farouk, Marwa Adel, Mustafa Abdullah Al Qbandi  
chest Disease Hospital Kuwait

**Background and Aim:** Right ventricle (RV)-dependent coronary circulation is known to be a poor prognostic indicator in pulmonary atresia with intact ventricular septum. The RV-dependent coronary circulation is defined as the presence of coronary-cameral fistulae with proximal coronary atresia or stenosis, resulting in regions of myocardium being primarily perfused from the RV.

**Method:** A male newborn who was presented with cyanosis immediately after birth. Started on prostaglandin. Was referred to the pediatric cardiology center, his echocardiogram showed: Small right ventricular cavity (bipartite) with mild tricuspid regurgitation which showed a supra systemic right ventricular pressure. The interventricular septum was bowing towards the left. The tricuspid valve Z score was -5, there was no RVOT, the pulmonary

artery was atretic with a confluent pulmonary artery branch. There was a large patent ductus arteriosus from the under surface of the aortic arch supplying the pulmonary artery branches. CT scan revealed mildly tortuous and dilated fistulous tract between left main coronary artery and right ventricle. The rest of the coronary arteries was not seen.

A cardiac catheterization was done which showed a large coronary fistula from the right ventricular sinusoids to the root of the aorta. The left main coronary artery was arising from this fistula. The right coronary artery was absent.

RV dependent coronary circulation was diagnosed. The decision was for a right BT shunt palliation and a univentricular repair pathway latter to avoid decompressing the RV (Figure 1).

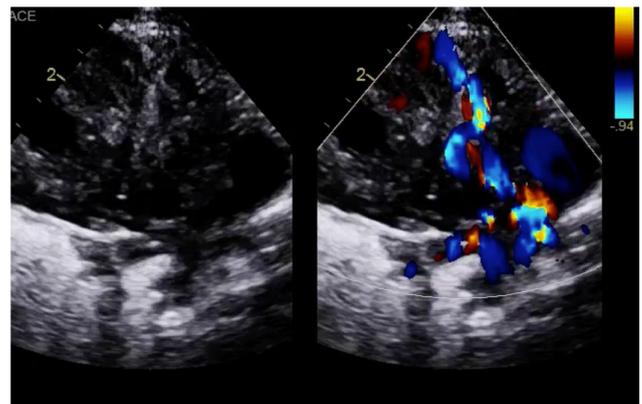
**Results:** The baby had a right BT shunt surgery and PDA ligation on day 6 of life. With satisfactory saturation after surgery.

On day 15th of life the baby had sudden cardiac arrest which was not responding to resuscitation.

**Conclusions:** RV dependent coronary circulation with a large coronary fistula in pulmonary atresia intact septum, makes the patient at risk to a sudden death even without attempts to decompress the right ventricle.

**Keywords:** Coronary fistula, Pulmonary atresia intact septum

Long axis view with anterior tilt



**Figure 1** Large coronary fistula from right ventricle to left coronary system

#### P-110

##### **Unusual mitral valve malformation which mimic ebstein anomaly of the mitral valve**

Mohamed Metwally Abdelaal, Sumaya Mahmoud Ibrahim, Mustafa Abdullah Al Qbandi  
chest Disease Hospital Kuwait

**Background and Aim:** Congenital malformations of the mitral valve are often complex and affect multiple segments of the valve apparatus. We are reporting a case of abnormal mitral valve associated with coarctation of the aorta. The abnormality mimic Ebstein anomaly of the mitral valve. The so-called “Ebstein-like” anomaly of the mitral valve was recently described and consists of a deformity in the mural leaflet, which can be attached to the ventricular wall to varying degrees. The milder forms are asymptomatic and are detected in echocardiographic findings, 2 whereas more severe forms give rise to mitral regurgitation.

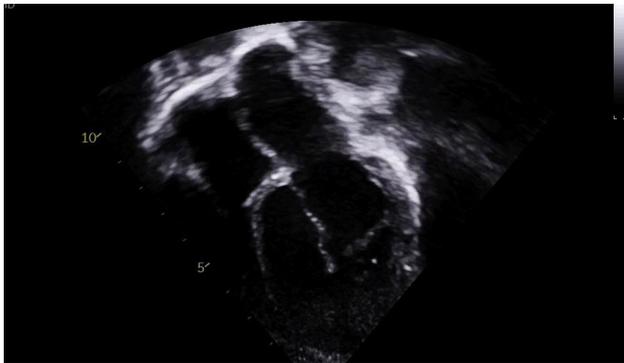
**Method:** Five days old neonate was referred to the pediatric cardiology center due to weak femoral pulsations. Echo cardiograph revealed a tight coarctation and abnormal mitral valve. The anterior mitral valve leaflet was long and attached to the papillary muscle with short chordae. While, the posterior leaflet was tethered to the left ventricle free wall in addition to short chordal attachment to the papillary muscle. (Figure 1). The level of coarctation of both leaflets was downward displaced. There was mild to moderate mitral valve regurgitation and there was no significant stenosis, mean inflow gradient was 3 mmHg. Since the mitral valve was satisfactory functioning the decision was not to operate on the mitral valve and to do coarctation repair only, which was done at seven days of age with a good result.

**Results:** After coarctation surgery the child remain stable on ACE inhibitors with no progress of mitral valve regurgitation. The child is now 3 years of age with no important changes of his mitral valve function.

**Conclusions:** Ebstein's anomaly generally affects the tricuspid valve, though a similar injury can affect the mitral valve. The functional orifice is shifted downwards into the inlet; the most affected leaflet is the mural. Since in our patient the mitral valve function was not levelly affected it was decided not to operate and continue follow up.

**Keywords:** Ebstein, Mitral valve

Four chamber view



**Figure 1** Down ward displacement of mitral valve and the posterior leaflet is adherent to the free wall

#### P-111

##### **Impact of maternal connective tissue disease on functional and structural abnormalities of the heart in the offspring**

*Terese Frellesen Neumann<sup>1</sup>, Sarah Sofie Andersen<sup>1</sup>, Sofie Dannesbo<sup>1</sup>, Anne Sophie Sillesen<sup>2</sup>, Ottilia Birgitta Vøgg<sup>2</sup>, Karen Schreiber<sup>3</sup>, Kasper Iversen<sup>2</sup>, Henning Bundgaard<sup>2</sup>, Anna Axelsson Raja<sup>1</sup>*

<sup>1</sup>Department of Cardiology, Copenhagen University Hospital, Rigshospitalet, Copenhagen, Denmark; <sup>2</sup>Department of Cardiology, Herlev-Gentofte Hospital, Copenhagen University Hospital, Copenhagen, Denmark; <sup>3</sup>Department of Rheumatology, Danish Hospital for Rheumatic Diseases, Sønderborg, Denmark

**Background and Aim:** Some maternal systemic connective tissue diseases (CTD) and their antibodies are associated with adverse fetal outcomes, including increased risk of congenital heart diseases (CHD). Whether this increased risk also applies for less severe structural or functional cardiac abnormalities in the fetus is currently unknown.

Based on a large cohort of newborns born to mothers with CTD, we aim to assess the association between maternal CTD and structural or functional cardiac abnormalities in the newborns.

**Method:** In the Copenhagen Baby Heart Study (CBHS) we included more than 25, 000 newborns between April 2016 and October 2018. All participants underwent systematic transthoracic echocardiography (TTE) within the first 60 days after birth.

The cohort selected for the present study include newborns in the CBHS born to mothers diagnosed with CTD, as identified through the Danish national health registries. Assuming an overall incidence of CTDs at 2% we expect to include approximately 500 newborns to mothers with CTD. Cases will be matched 1:1 to controls, based on child sex; gestational age at birth; age and weight at time of TTE; and maternal age at delivery. Maternal medical charts will be reviewed to validate the CTD diagnosis and obtain information about disease severity, treatment, etc. Based on the TTEs obtained in the newborns the primary endpoint is a composite of functional and structural abnormalities in the heart of the newborns, including left ventricular structural and functional abnormalities, septum defects, and valve anomalies.

**Results:** Preliminary results are expected in spring 2023.

**Conclusions:** It is unclear whether children of mothers with CTD should have routine cardiac evaluation at birth, and whether certain subgroups of maternal CTD have a higher risk of CHD or less severe cardiac abnormalities in the offspring than others. The size of CBHS and the comprehensive assessment of the included children provide a unique opportunity for obtaining new knowledge to this field, including better insights into whether routine TTE after birth should be considered in certain subgroups.

**Keywords:** systemic tissue diseases, echocardiography, congenital heart

#### P-112

##### **Maternal pre-existing diabetes and gestational diabetes and the risk of bicuspid aortic valve in the offspring.**

*Magdalena Hansson<sup>1</sup>, Jakob Norsk<sup>1</sup>, Anne Sophie Sillesen<sup>1</sup>, Line Stormly<sup>1</sup>, Sofie Dannesbo<sup>2</sup>, Ruth Ottilia Birgitta Vøgg<sup>1</sup>, Anna Axelsson Raja<sup>2</sup>, Niels Vejstrup<sup>2</sup>, Kasper Iversen<sup>1</sup>, Henning Bundgaard<sup>2</sup>*  
<sup>1</sup>Herlev Hospital - Copenhagen University Hospital, Department of Cardiology, Herlev, Denmark; <sup>2</sup>Rigshospitalet - Copenhagen University Hospital, Department of Cardiology, Copenhagen, Denmark

**Background and Aim:** Approximately 6% of all pregnancies in Denmark are affected by maternal pre-existing or gestational diabetes, well-known risk factors for congenital heart defects. Bicuspid aortic valve is one of the most common congenital heart defects. Most bicuspid aortic valve cases are sporadic. While previous studies have demonstrated a strong genetic component, a monogenic inheritance does not explain all cases and a multifactorial aetiology has been proposed. Maternal diabetes increases the risk of structural heart disease in the offspring but the association between maternal diabetes and the risk of bicuspid aortic valve in the offspring has been sparsely investigated.

The purpose of the present study is to assess the prevalence of bicuspid aortic valve in children born to mothers with pre-existing or gestational diabetes, with systematic transthoracic echocardiography (TTE), in a large, population-based cohort of neonates, and compare with children born to mothers without diabetes.

**Method:** Systematic, standardized TTE was performed in over 25.000 neonates included in a population-based cohort study between 2016 and 2018, the Copenhagen Baby Heart Study. Currently a continued inclusion of neonates of mothers with

pre-existing or gestational diabetes is ongoing to increase the exposed group of neonates.

Information about maternal pre-existing and gestational diabetes is collected from review of the mother's medical records and from an Obstetrical Database maintained by the participating hospitals.

The exposed neonates will be compared with controls, neonates in the cohort not exposed to maternal diabetes.

**Results:** The collection of data is still ongoing, but preliminary results are expected during the spring of 2023.

**Conclusions:** We expect the results of this study to increase our knowledge of the effects of maternal pre-existing and gestational diabetes on the offspring's risk of bicuspid aortic valve.

Both negative and positive results will be of importance since it can help clinicians in their guidance of women and their pregnancies. Depending on the results of this study, echocardiographic screening of infants born to mothers with diabetes may be indicated.

**Keywords:** Congenital Heart Defects, Bicuspid Aortic Valve, Maternal Diabetes, Echocardiography

### P-113

#### Screening for rheumatic heart disease in sierra leone: A pilot study

Francisco Javier Díez<sup>1</sup>, Ruth Solana<sup>2</sup>

<sup>1</sup>Department of Artificial Intelligence, Universidad Nacional de Educación a Distancia (UNED), Madrid, Spain; <sup>2</sup>Department of Paediatric Cardiology, Hospital Infanta Leonor, Madrid, Spain

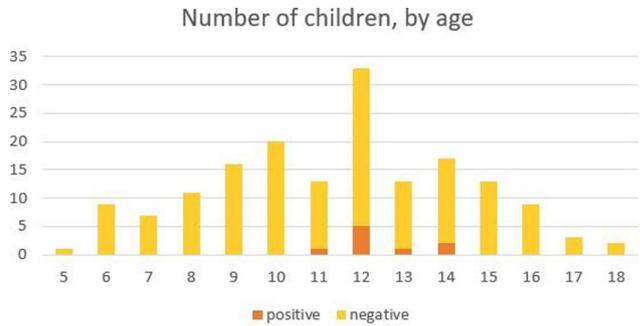
**Background and Aim:** Rheumatic heart disease (RHD) affects about 30 million people worldwide, causing about 300,000 deaths per year and many cases of disability in young people. Echocardiographic screening allows secondary prevention with penicillin prophylaxis. The aim of our study was to detect RHD in children in Makeni, the fifth largest city in Sierra Leone. **Method:** Around 200 children participating in three simultaneous summer camps organised by "Viva Makeni", a Spanish non-government organisation (NGO), were given an informed consent form. Those who returned it signed by their parents or guardians were examined on-site by a paediatric cardiologist (R.S.) using a handheld Philips Lumify S4-1 probe connected to a tablet. The study protocol prescribed that children with suspected RHD be subsequently examined with a standard sonograph at a hospital in Makeni, but this was not possible due to unexpected logistical impediments (lost luggage).

**Results:** 167 children aged between 5 and 18 years (mean: 11.45 years) were examined. Of these, 61, 7% were female. The study found 9 cases of regurgitation (5.4% of the children; 95% CI: 2.5 to 10.0): 5 trivial mitral regurgitations, 2 mild mitral regurgitations and 2 trivial aortic regurgitations, all in children between 11 and 14 years. The prevalence in the age group 12-14 years was 12.7% (95% CI: 5.6 to 23.5).

**Conclusions:** Given that the prevalence of definite RHD in other African countries lies between 1-3%, it is surprising that our study found no positive cases. The fact that rheumatic heart damage accumulates with age and the high prevalence of regurgitations in the age group 12-14 years make it even more surprising that no regurgitations were found above that age. These results suggest that there is a strong selection bias; it might be explained because the children were selected from several schools in a country that suffers from severe school dropout, which worsens with age and mainly affects socioeconomically disadvantaged children, who are at the same time the most vulnerable to RHD. In addition, the prevalence of RHD in Sierra Leone might be low due to

the ease of obtaining antibiotics, even without a prescription. Further studies are necessary to test these explanatory hypotheses.

**Keywords:** rheumatic heart disease, echocardiographic screening, secondary prophylaxis, Sierra Leone



Nine regurgitations were found in 167 children, all of them mild or trivial. The prevalence in the age group 12-14 years was 12.7%.

### P-115

#### Virtual treatment planning for fontan-palliated patients: prediction of hepatic blood flow distribution

Marie Schafstedde<sup>1</sup>, Sarah Nordmeyer<sup>1</sup>, Peter Kramer<sup>2</sup>, Anastasia

Schleiger<sup>2</sup>, Felix Berger<sup>3</sup>, Stanislav Ovroutski<sup>2</sup>, Jan Brüning<sup>4</sup>

<sup>1</sup>German Heart Center Berlin (DHZB), Berlin, Germany & Institute of Computer-assisted Cardiovascular Medicine, Berlin, Germany & German Center for Cardiovascular Research (DZHK), Berlin, Germany;

<sup>2</sup>German Heart Center Berlin (DHZB), Berlin, Germany; <sup>3</sup>German Heart Center Berlin (DHZB), Berlin, Germany & German Center for Cardiovascular Research (DZHK), Berlin, Germany; <sup>4</sup>Institute of Computer-assisted Cardiovascular Medicine, Berlin, Germany

**Background and Aim:** Formation of pulmonary arteriovenous malformations (PAVM) in Fontan-palliated patients is hypothesized to be caused or at least affected by the distribution of hepatic blood (HFD) towards the left and right pulmonary artery. In patients with PAVM and uneven HFD, surgical or interventional treatment facilitating an even HFD is a promising approach to achieve remodeling of PAVM. However, while the pre-interventional hemodynamics can be assessed via angiography or velocity encoded MRI, prediction of the post-intervention hemodynamics is difficult due to the complex and heterogeneous anatomy of Fontan-palliated patients. Here, computer-based approaches might allow outcome prediction of different treatment strategies for a given patient.

**Method:** The patient-specific anatomy of the total cavopulmonary connection (TCPC) of three Fontan-palliated patients with PAVM was reconstructed using CT or a combination of CT and MRI images. Due to severe cyanosis, these patients were considered for surgery or catheter-based intervention aiming at hepatic blood flow re-routing. First, numerical simulations were performed. Based on the patient-specific anatomy and the pre-interventional hemodynamics, different treatment strategies were identified during heart team meetings including pediatric cardiologists and congenital heart surgeons. The patient-specific models were altered virtually to mimic these treatment strategies. Finally, for each virtual treatment the resulting HFD was calculated via numerical simulations.

**Results:** Reconstruction of the complex anatomy of the TCPC was possible in all three patients despite presence of metallic stents. To facilitate this, combined approaches using both CT as well as MRI data were required in two patients. For each patient at least one virtual treatment strategy that was considered viable and simultaneously resulted in a more even HFD was identified.

**Conclusions:** Virtual treatment planning is a promising approach for patients with complex anatomies, such as the TCPC, which do not allow intuitive assessment of the post-operative changes. Especially as treatment strategies vary widely with respect to their respective risk, an objective outcome assessment might help to identify the ideal treatment strategy for a specific patient.

**Keywords:** Fontan, hepatic blood flow distribution, pulmonary arteriovenous malformations, virtual treatment planning

#### P-116

##### Does magnetic resonance feature tracking help to decide operation timing in patients with severe aortic regurgitation and combined stenosis?

Nerejda Shehu<sup>1</sup>, Julika Sieker<sup>1</sup>, Bettina Reich<sup>1</sup>, Heiko Stern<sup>1</sup>, Stefan Martinoff<sup>2</sup>, Peter Ewert<sup>1</sup>, Christian Meierhofer<sup>1</sup>

<sup>1</sup>Department of Congenital Heart Disease and Paediatric Cardiology, German Heart Center, TUM, Munich, Germany; <sup>2</sup>Department of Radiology, German Heart Center, TUM, Munich, Germany

**Background and Aim:** Cardiovascular magnetic resonance feature tracking (CMR-FT) is increasingly used in cardiology to detect early morphological alterations of cardiac function. Patients with bicuspid aortic valve (BAV) and high degree of aortic regurgitation (AR), do not display marked left ventricular dilation or ventricular dysfunction if coexisting valvular stenosis is present. As a consequence, the optimal timing for surgery remains unclear in asymptomatic patients with severe AR combined with AS.

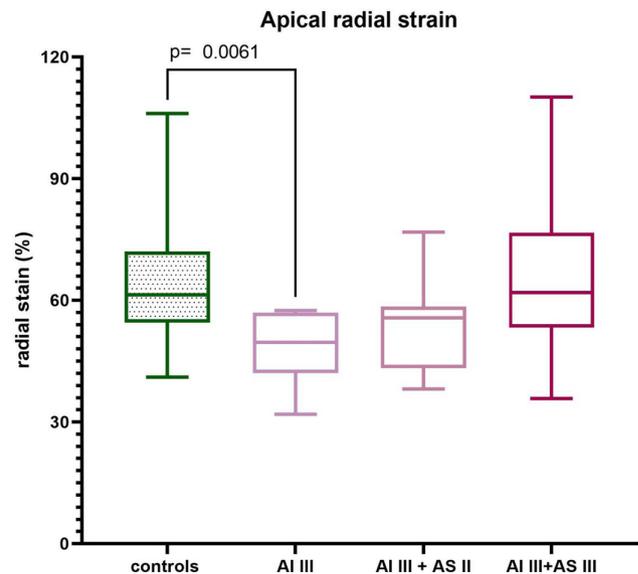
**Method:** We retrospectively analysed 26 asymptomatic patients (median age 25 years; range 9–65 years) with BAV and severe AR with more than 40% regurgitation fraction. Left ventricular (LV) longitudinal strain (LS) was evaluated in the four chamber view, circumferential (CS) and radial (RS) strain in short axis slices. The patients were divided into 3 groups depending on the severity of AS associated; 8 patients had mild AS (velocity encoding (VENC) < 300 cm/s), 7 patients had moderate AS (300 ≤ VENC < 400 cm/s) and 11 patients had severe AS (VENC ≥ 400 cm/s). CMR-FT data of 22 healthy volunteers (median age 21.5 years; range 14–48 years) were used for comparison.

**Results:** RS and CS of the apical segments were pronounced lower in patients with AR and a mild AS compared to controls only. The median apical RS was 50%, range 32 to 57% vs. 61%, range 41% to 106%,  $p < 0.0061$ ; the median apical CS was -25%, range -19% to -26% vs. -27%, range -22% to -32%;  $p = 0.0097$ . No other significant differences of global and regional RS, CS and LS were present, neither between each of the three groups of combined valvular vitium and healthy controls, nor comparing the aforementioned three patients groups to each other.

**Conclusions:** There was no difference in most of all parameters of CMR-FT between patients and controls. Most of CMR-FT parameters do not get worse if the aortic stenotic component increase. Only the apical RS and the apical CS of patients with severe AR and mild AS show reduced values compared to healthy controls. This finding hardly supports the use of CMR-FT in deciding optimal surgery timing in asymptomatic patients with combined aortic regurgitation and stenosis.

**Keywords:** magnetic resonance feature tracking, bicuspid aortic valve, aortic regurgitation, aortic stenosis

apical radial strain



#### P-117

##### Cardiac findings in newborn twins

Julie Molin<sup>1</sup>, Maria Munk Pærregaard<sup>1</sup>, Christian Alexander Pihl<sup>1</sup>, Caroline Boye Thygesen<sup>1</sup>, Adrian Pietersen<sup>1</sup>, Sofie Dannesbo<sup>1</sup>, Jakob Boesgaard Norsk<sup>1</sup>, Anna Axelsson Raja<sup>2</sup>, Ruth Otilia B Vøgg<sup>1</sup>, Anne Sophie Sillesen<sup>1</sup>, Kasper Karmark Iversen<sup>1</sup>, Henning Bundgaard<sup>2</sup>, Alex Hørby Christensen<sup>1</sup>

<sup>1</sup>Department of Cardiology, Herlev-Gentofte Hospital, Copenhagen University Hospital, Copenhagen, Denmark.; <sup>2</sup>The Capital Regions Unit for Inherited Cardiac Diseases, Department of Cardiology, The Heart Center, Rigshospitalet, Copenhagen University Hospital, Copenhagen, Denmark.

**Background and Aim:** The prevalence of twin pregnancies has increased in the last decades and is a well-established risk factor for developing congenital heart disease. However, whether this also applies to an unselected, general population-based cohort with contemporary surveillance is unknown. The study aimed to evaluate cardiac findings in newborn twins from the general population and investigate if newborn twins may require systematic evaluation of cardiac parameters.

**Method:** Prospective cohort study of newborns with echocardiography and electrocardiography (ECG) performed during the first month of life. Cardiac findings were compared between twins and singletons matched 1:3 on sex, age, and weight.

**Results:** We included 412 newborn twins (16% monozygotic; 50% boys; median age at examination 13 days) and 1,236 singletons. We found an increased prevalence of non-severe structural heart disease in twins compared to singletons (6.6 vs. 4.0%,  $p = 0.04$ ). The most common abnormality was ventricular septal defects in both groups. Comparing cardiac parameters showed that twins had a thinner left ventricular posterior wall in diastole (LVPWd; median 1.82 vs. 1.87 mm,  $p = 0.02$ ), a smaller diameter of the left atrium (median 10.6 vs. 11.1 mm,  $p = 0.04$ ), higher heart rate (median 148 vs. 144 bpm,  $p = 0.04$ ), a more left-shifted QRS axis (median 106 vs. 111°,  $p < 0.001$ ), and a lower maximum R-wave amplitude in V1 (median 927 vs. 1,015  $\mu$ V,  $p = 0.02$ ) compared to singletons. After multifactorial adjustment for

potential confounders, the effect of twinning on echocardiographic and ECG parameters persisted only for LVPWd ( $p < 0.05$ ). **Conclusions:** Despite contemporary surveillance, we found an increased prevalence of non-severe structural heart disease in a population-based cohort of newborn twins. However, the effect of twinning on cardiac parameters was modest and generally did not persist after correction for likely confounding factors

**Keywords:** Multiple pregnancy, congenital heart disease, echocardiography, ECG

**Background and Aim:** The electrocardiogram (ECG) is a valuable tool for detecting left ventricular hypertrophy (LVH) in adults. Previous studies investigated whether ECG also is a useful tool for detecting LVH in children have produced mixed results. We therefore aimed at describing the evolution of the QRS complex in the first month of life and investigate the association between QRS complex features and echocardiographic measurements of left ventricular mass in a large population-based study of neonates

Echocardiographic and electrocardiographic findings in twins and singletons.						
	All twins (n=412)	Singletons (n=1,236)	P-value*	Twin A (n=175)	Twin B (n=175)	P-value†
FS, %	31 (26-42)	31 (26-41)	0.80	31 (25-41)	31 (27-42)	0.95
LVEF, %	62 (53-76)	62 (54-75)	0.64	62 (52-75)	62 (55-76)	0.45
IVSd, mm	2.33 (1.54-3.16)	2.32 (1.53-3.37)	0.34	2.33 (1.54-3.17)	2.29 (1.51-3.06)	0.46
LVPWd, mm	1.82 (1.12-2.90)	1.87 (1.18-3.11)	<b>0.02</b>	1.81 (1.12-2.88)	1.83 (1.11-2.76)	0.84
LVIDd, mm	18.7 (14.9-22.1)	18.7 (15.6-22.6)	0.28	18.8 (14.7-22.1)	18.8 (14.8-22.0)	0.28
LVIDs, mm	12.8 (10.1-15.4)	12.8 (10.4-16.0)	0.40	13.0 (10.2-16.0)	12.8 (10.2-15.2)	0.35
PW-LVOT Vmax, m/s	0.78 (0.51-1.11)	0.77 (0.49-1.10)	0.12	0.80 (0.51-1.11)	0.78 (0.52-1.12)	0.20
LA diameter, mm <sup>‡</sup>	10.6 (7.4-14.3)	11.1 (7.0-14.5)	<b>0.04</b>	11.0 (8.3-13.5)	10.4 (7.4-14.7)	0.27
Heart rate, bpm	148 (105-188)	144 (102-190)	<b>0.04</b>	148 (106-190)	149 (104-185)	0.97
PR interval, ms	94 (80-116)	94 (80-120)	0.14	94 (80-122)	94 (80-114)	0.38
QRS axis, degrees	106 (71-198)	111 (72-205)	<b>&lt;0.001</b>	105 (70-188)	106 (73-188)	0.73
QRS duration, ms	52 (42-66)	54 (42-66)	0.17	52 (43-65)	52 (44-66)	0.65
QT interval, ms	270 (229-331)	274 (224-330)	0.37	272 (219-322)	270 (230-338)	0.49
QTcBazett, ms	419 (379-468)	420 (373-474)	0.43	420 (379-480)	417 (379-464)	0.17
QTcFridericia, ms	363 (323-411)	363 (317-411)	0.94	365 (321-415)	364 (324-413)	0.23
R-V1, $\mu$ V	927 (310-2,227)	1015 (281-2,350)	<b>0.02</b>	947 (300-2,076)	957 (298-2,320)	0.85
S-V1, $\mu$ V	581 (107-1,633)	639 (89-1,946)	0.21	556 (99-1,643)	659 (170-1,617)	<b>&lt;0.05</b>
R-V6, $\mu$ V <sup>§</sup>	964 (237-1,955)	874 (244-2,070)	0.07	978 (338-1,908)	959 (219-1,935)	0.15
S-V6, $\mu$ V	576 (131-1,488)	551 (107-1,697)	0.91	612 (131-1,537)	566 (146-1,420)	0.68

Variables are displayed as medians (2-98%iles). Abbreviations: FS: fractional shortening; LVEF: left ventricular ejection fraction; LVIDd/LVIDs: left ventricular internal end-diastole/end-systole diameter; IVSd: interventricular end-diastolic septal thickness; LVPWd: left ventricular end-diastolic posterior wall thickness; PW-LVOT Vmax: pulse-wave left ventricular outflow tract maximum velocity; LA: left atrial diameter in systole; bpm: beats per minute; R-V1: Maximum R-wave amplitude in V1; S-V1: maximum S-wave amplitude in V1; R-V6: maximum R-wave amplitude in V6; S-V6: maximum S-wave amplitude in V6. \*Comparison between twins and singletons. †Comparison between twin A and B. ‡Data available in 199 twins and 564 singletons. §Data from V6 available on 292 twins and 873 singletons. Significant values are marked with bold.

## P-118

### The neonatal QRS complex and its association with left ventricular mass

Julie Molin<sup>1</sup>, Joachim Hartmann<sup>1</sup>, Caroline Boye Thygesen<sup>1</sup>, Maria Munk Perregaard<sup>1</sup>, Anne Sophie Sillesen<sup>1</sup>, Anna Raja Axelsson<sup>2</sup>, Ruth Otilia B Vøgg<sup>1</sup>, Kasper Kammark Iversen<sup>1</sup>, Henning Bundgaard<sup>2</sup>, Alex Horby Christensen<sup>1</sup>

<sup>1</sup>Department of Cardiology, Copenhagen University Hospital - Herlev-Gentofte Hospital, Copenhagen, Denmark and Department of Clinical Medicine, University of Copenhagen, Copenhagen, Denmark.; <sup>2</sup>The Capital Regions Unit for Inherited Cardiac Diseases, Department of Cardiology, The Heart Center, Rigshospitalet, Copenhagen University Hospital, Copenhagen, Denmark.

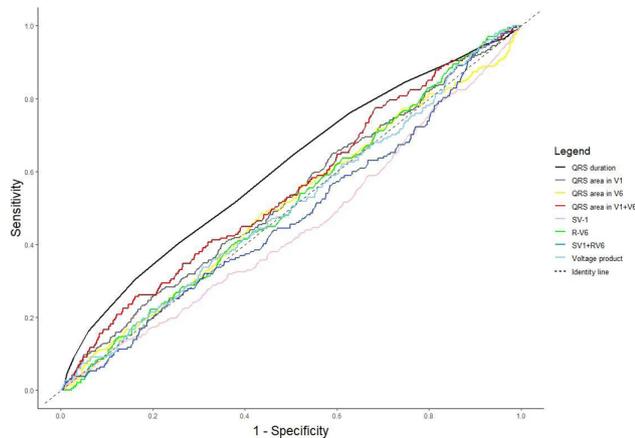
**Method:** Prospective cohort study of neonates with electrocardiogram (ECG) and echocardiography performed during the first month of life. Echocardiographically determined left ventricular mass index (LVMI) was investigated and the correlation with electrocardiographic markers of LVH (defined as LVMI  $\geq$ 98% percentile) was analyzed.

**Results:** We included 16, 450 neonates (52% boys; median post-natal age 11 days) with a median weight of 3.6 kg at the time of examination. For the entire cohort, the median QRS duration was 56 ms and the median LVMI was 26.5 g/cm<sup>2</sup>, which both increased during the first month of life (54 vs. 56 ms, and 24.7 vs. 28.6 g/cm<sup>2</sup>; both  $p < 0.001$ ). Receiver operator characteristics and area under the curve (AUC) analyses for all the investigated

ECG features (QRS duration, QRS areas in V1 and V6, the absolute sum of QRS area in V1+V6, maximum amplitudes of S-waves in V1 and R-waves in V6, the sum of maximum amplitudes in S-V1+R-V6, and the Sokolow-Lyon voltage product  $QRS_{duration} \cdot (S-V1+R-V6)$ ) showed poor results for identification of increased LVMI. The AUC values were between 0.494–0.607 with the highest value for QRS duration. We found low sensitivity (ranging from 0 to 9%), but high specificity (ranging from 97.2 to 98.1 %) to detect LVH in neonates for all the investigated ECG features.

**Conclusions:** We present reference values for QRS complex features and their association with LVMI in neonates from a large, unselected, population-based cohort. The QRS complex gradually evolved during the first month of life but had a low correlation with LVMI. Our results indicate a poor diagnostic value of using ECG features as a diagnostic tool to determine LVMI in neonates.

**Keywords:** QRS complex, electrocardiography, left ventricular mass, neonates, reference values



Receiver operator characteristics curve for each ECG feature ( $\geq 98$  percentile) to detect left ventricular hypertrophy (left ventricular mass index  $\geq 98$  percentile). Abbreviations: S-V1: maximum S-wave amplitude in V1; R-V6: maximum R-wave amplitude in V6. Sokolow-Lyon voltage product:  $QRS_{duration} \cdot (S-V1+R-V6)$ .

**P-119**

**MRI quantification of collateral burden under general anaesthetic and physiological sleep conditions in the single ventricle circulation post norwood**

Malenka Mona Bissell<sup>1</sup>, Peter Lillitos<sup>1</sup>, Hannah Panayiotou<sup>1</sup>, Rawan Abuzinadah<sup>2</sup>, Vitor Ramos<sup>1</sup>, Jutta Scheffczik<sup>3</sup>, Jamie Bentham<sup>1</sup>, Helen Michael<sup>1</sup>

<sup>1</sup>Department of Congenital Cardiology, Leeds Teaching Hospitals NHS Trust, Leeds, United Kingdom; <sup>2</sup>Leeds Institute of Cardiovascular and Metabolic Medicine, University of Leeds, Leeds, United Kingdom;

<sup>3</sup>Department of Anaesthesia, Leeds Teaching Hospitals NHS Trust, Leeds, United Kingdom

**Background and Aim:** In hypoplastic left heart syndrome(HLHS) cardiovascular magnetic resonance imaging(CMR) is often used to quantify collateral burden. General anaesthetic(GA) is known to alter physiology.

**AIM:**

Pilot study to assess the impact of GA conditions on collateral assessment in HLHS post Norwood stage1.

**Method:** 30 HLHS infants post Norwood 1 surgery were included in the study –25 under GA and 5 underwent non-sedated feed and wrap(F&W) CMR in a dedicated neonatal incubator under natural sleep conditions. All patients had flow assessments in: native&neo-aortic valve, descending aorta(DAo), superior vena cava(SVC), Sano or BT-shunt and branch pulmonary arteries(BPA). 16 patients also underwent pulmonary vein(PV) assessment. Collateral flow was calculated as: effective aortic flow–(SVC+DAo) and if available as: PV flow–BPA flow.

In addition, cardiac catheterisation data was available in all babies to invasively categorise the degree of collateral burden as: none (green), mild (amber), excessive (red).

**Results:** 9/25 in the GA group had undergone BT shunt, all remaining infants had a Sano shunt. The F&W group were younger (mean age 2.9+–0.2months vs GA 3.7+–0.9months;  $p < 0.01$ ). Weight was similar between groups (mean F&W 4.7+–0.9kg vs GA 5.1+–0.7kg;  $p = 0.44$ ).

11 GA patients had at least some collateralisation at catheterisation. In 6/11(55%) of these GA MRI had suggested no significant collateral flow. Only 3/11 (27%) had agreement between assessment of collateral burden by catheter(1 some degree and 2 excessive) versus CMR– in others, GA-CMR underestimated collateralisation.

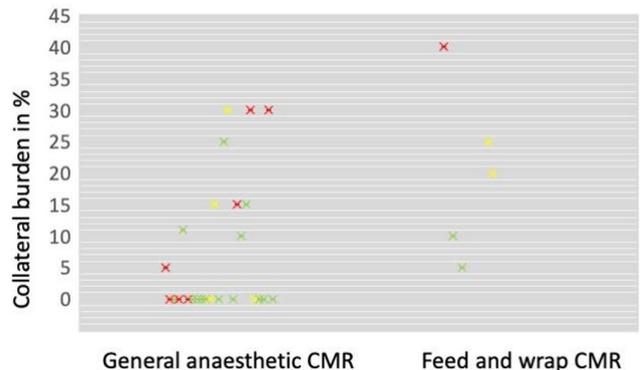
All 3/3(100%) F&W babies had some degree of collateralisation on MRI– and a corresponding degree of collateral burden assessed by catheter(Fisher’s exact test  $p = 0.05$ ).

Only 1 patient(GA group) had a visible collateral on contrast angiogram.

**Conclusions:** Compared to catheterisation, CMR assessment of collaterals under GA conditions underestimated the degree of collateralisation in the majority of cases(70%). Whilst catheter assessment is also undertaken under GA, GA CMR also involves paralysis and breath-holding which may contribute to more artificial conditions. This pilot study suggests that CMR under more physiological conditions(natural sleep) may be beneficial in collateral assessment in the HLHS group pre Glenn surgery. Additional CMR angiography with contrast has a low yield for collateral assessment, possibly due to the low resolution in this age group.

**Keywords:** HLHS, Norwood, flow, collateral, CMR, MRI

**Collateral flow quantification by CMR**



Percentage of collateral burden quantified by general anaesthetic CMR and feed and wrap CMR. Collateral burden by cardiac catheterisation categorised as: none (green), mild (amber), excessive (red)

## P-120

**Altered haemodynamics in the reconstructed neo-aorta after the norwood operation in hypoplastic left heart syndrome**

Rawan Abuzinadah<sup>1</sup>, Hannah Panayiotou<sup>2</sup>, David Shelley<sup>3</sup>, David Broadbent<sup>4</sup>, Ning Jin<sup>5</sup>, Helen Michael<sup>2</sup>, Julio Sotelo<sup>6</sup>, Malenka Mona Bissell<sup>1</sup>

<sup>1</sup>Leeds Institute of Cardiovascular and Metabolic Medicine, University of Leeds, United Kingdom; <sup>2</sup>Department of Paediatric Cardiology, Leeds Teaching Hospitals Trust, Leeds, United Kingdom; <sup>3</sup>Department of Cardiology, Leeds Teaching Hospitals Trust, Leeds, United Kingdom; <sup>4</sup>Department of Medical Physics, Leeds Teaching Hospitals Trust, Leeds, United Kingdom; <sup>5</sup>Siemens Medical Solutions USA, Inc., Chicago, United States of America; <sup>6</sup>School of Biomedical Engineering, Universidad de Valparaíso, Valparaíso, Chile. Millennium Institute for Intelligent Healthcare Engineering, iHEALTH, Santiago, Chile

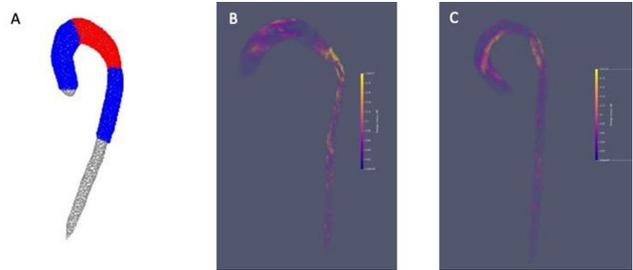
**Background and Aim:** Hypoplastic left heart syndrome (HLHS) is considered one of the most complex congenital cardiac conditions that requires staged surgical palliation. During the first stage surgery (Norwood) the ascending aorta and the arch are reconstructed to create a neo-aorta that is capable of providing blood to the body. **AIMS:** To assess how flow haemodynamics differ in reconstructed aortas in HLHS post Norwood 1 versus healthy babies

**Method:** 14 babies were prospectively recruited into the study (5 Neonates with HLHS who had undergone stage 1 Norwood operation with a sano shunt and 9 healthy control babies[HV]). All babies underwent neonatal feed and wrap cardiac magnetic resonance imaging (CMR), non-sedated, non-contrast, and free breathing in a dedicated CMR compatible incubator. A prototype compressed sensing accelerated 4D Flow CMR sequence was used for aortic assessment. A research software was used for analysis. The aorta was divided into 3 aortic sections (ascending aorta; transverse arch; native descending aorta) and the following data was extracted: the maximum velocity, and the mean values of: wall shear stress, viscous dissipation, energy loss, kinetic energy, curvature, diameter, area and helicity density. To compare each parameter in each aortic section between both groups, a Mann-Whitney U test was used.  $P < 0.05$  was considered as significant.

**Results:** The HLHS cohort was older than the HV (HLHS median 60days, range: 60-90days vs HV 3days, range: 1-9days;  $p = 0.001$ ). Maximum velocity in the ascending aorta was higher in HLHS than HV (HLHS median = 0.89 [0.29] m/s vs HV median = 0.72 [0.20] m/s;  $p = 0.019$ ). In the descending aorta energy loss was higher in HLHS (HLHS median = 0.08 [0.06] uW vs HV median = 0.05[0.06] uW);  $p = 0.042$ ). The relative Helicity Density in the transverse arch was lower in HLHS (HLHS median = -0.05[0.11] vs HV median = 0.02[0.26];  $p = 0.029$ ). The curvature of the transverse arch was also significantly lower in HLHS (median = 54.49[19.46] 1/m vs 79.80 [28.11] 1/m;  $p = 0.004$ ).

**Conclusions:** The neo-aorta after reconstruction during the Norwood operation has an abnormal geometry (decreased curvature) and altered flow (relative helicity density) in the transverse arch, with increased energy loss in the native descending aorta. These imaging markers might become useful in further research of predicting re-coarctation in HLHS.

**Keywords:** HLHS, 4D flow MRI, CMR, Norwood, Aorta



A) Analysis sections of the aorta: Ascending aorta (Section 1); Transverse arch (Section 2); Descending aorta (Section 3) B) Energy loss in a HLHS baby - yellow areas of high energy loss; c) Energy loss in a healthy volunteer baby

## P-121

**Speckle tracking echocardiography in patients with multisystem inflammatory syndrome in children (MIS-C): A retrospective cohort study**

Stejara Augusta Netea<sup>1</sup>, Nico A Blom<sup>2</sup>, Taco W Kuijpers<sup>1</sup>, Irene M Kuipers<sup>2</sup>

<sup>1</sup>Amsterdam UMC, University of Amsterdam, Pediatric Immunology, Rheumatology and Infectious Disease, Meibergdreef 9, Amsterdam, Netherlands; <sup>2</sup>Amsterdam UMC, University of Amsterdam, Pediatric Cardiology, Meibergdreef 9, Amsterdam, Netherlands

**Background and Aim:** Multisystem Inflammatory Syndrome in Children (MIS-C) is a SARS-CoV-2 related hyperinflammatory syndrome with a high risk of cardiovascular complications, including cardiac dysfunction in about half of the cases. We aimed to investigate the short- and long-term cardiac outcomes in MIS-C in combination with laboratory outcomes to guide future studies and clinical recommendations.

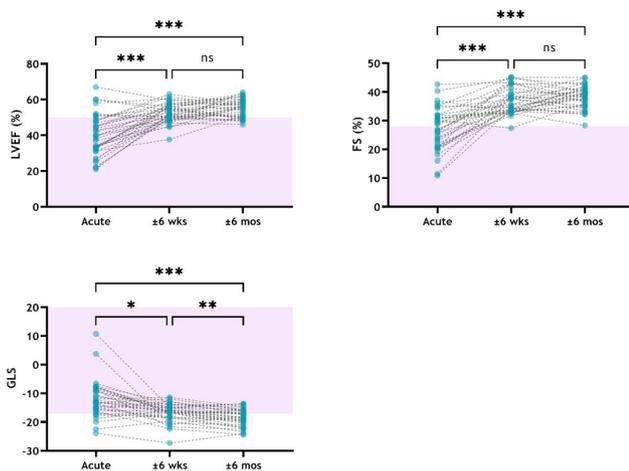
**Method:** We conducted a cohort study including children diagnosed with MIS-C admitted in the Amsterdam UMC. Data concerning clinical characteristics, conventional echocardiography (i.e., left ventricle ejection fraction [LVEF], shortening fraction [SF]) and speckle tracking were collected during the acute (nadir value during admission), early convalescent ( $\pm 6$  weeks) and late convalescence ( $\pm 6$  months). A paired analysis was performed to assess over-time patterns (Friedman test). Spearman correlations and a multivariate logistic regression analysis were performed to identify predictors for late speckle tracking impairment.

**Results:** We included 48 MIS-C patients (median age 11.9 [IQR 6.9-14.5] years old, 58.3% male). 83.0% Of patients presented with cardiac dysfunction in the acute phase based on conventional echocardiographic parameters (LVEF  $< 50\%$  and/or FS  $< 28\%$ ). 8.5% patients had a preserved LVEF and FS, while the GLS was elevated ( $> [-17\%]$ ). In the longitudinal analysis (Figure 1), LVEF and FS reached a plateau level within the first six weeks of follow-up, while the GLS continued to significantly decrease from the acute (median -13.1% [IQR [-15.9]-[-8.7]%) to early convalescent phase (median -16.4% [IQR [-18.4]-[-14.8]%),

$P = 0.02$  [vs. acute]) into the late convalescent phase (median -18.2% [IQR [-20.6]-[-15.9]%,  $P = 0.005$  [vs. acute]). In a multivariate analysis, soluble troponin ( $>62.0$  ng/L) was associated with GLS dysfunction ( $>[-17]$ %) in the late convalescence (OR = 8.2 [95% CI 1.1-60.4]). Acute troponin levels significantly correlated with the GLS during late convalescence ( $r = 0.5$ ,  $P = 0.002$ ).

**Conclusions:** Speckle tracking echocardiography more sensitively detects (persisting) subclinical myocardial impairment than conventional echocardiography. GLS in combination with other markers, such as troponin, may be useful in predicting delayed cardiac recovery.

**Keywords:** MIS-C, speckle tracking, myocardial strain imaging



**Figure 1.** Over-time patterns of LVEF, FS and GLS

#### P-122

##### **Speckle tracking echocardiography in patients with multisystem inflammatory syndrome in children (MIS-C): A systematic review**

Stejara A Netea<sup>1</sup>, Nico A Blom<sup>1</sup>, Taco W Kuijpers<sup>2</sup>, Irene M Kuipers<sup>1</sup>  
<sup>1</sup>Amsterdam UMC, University of Amsterdam, Pediatric Cardiology, Meibergdreef 9, Amsterdam, Netherlands; <sup>2</sup>Amsterdam UMC, University of Amsterdam, Pediatric Immunology, Rheumatology and Infectious Disease, Meibergdreef 9, Amsterdam, Netherlands

**Background and Aim:** Multisystem Inflammatory Syndrome in Children (MIS-C) is a SARS-CoV-2 related hyperinflammatory syndrome with a high risk of cardiovascular complications, including cardiac dysfunction in about half of the cases. A vast number of studies have been published in a short time-frame investigating the value of speckle tracking to follow up subclinical cardiac damage, in particular when conventional parameters (left ventricle ejection fraction [LVEF], fractional shortening [FS]) remain normal. Therefore, we did a systematic review to identify all studies and performed a meta-analysis to evaluate the value of speckle tracking as a marker for subclinical cardiac damage.

**Method:** We performed a systematic search in Pubmed/Medline to identify all available data on speckle tracking in MIS-C up until

November 9th 2022. We excluded case reports and case series ( $n < 10$ ) with a description of non-consecutive participants. Data on study design (e.g., definitions for decreased LVEF, FS and speckle tracking parameters), the study population and results at various time points during follow-up were extracted.

**Results:** We identified fifteen eligible studies. Definitions for a decreased speckle tracking values varied widely and patient populations were limited. Although findings were heterogeneous, a majority of the studies reported a delayed recovery of speckle tracking parameters when compared to conventional parameters (i.e., LVEF, FS).

**Conclusions:** Speckle tracking echocardiography may be useful in detecting (persisting) subclinical myocardial impairment. However, data is still limited and larger studies with a more in-depth approach - including the combined use of soluble blood markers, multiple imaging modalities and stress tests - are required to draw clear conclusions on clinical relevance and follow-up recommendations.

**Keywords:** MIS-C, speckle tracking, myocardial strain imaging

#### P-123

##### **Fragmented QRS efficiency to predict right cardiac function in patients with repaired tetralogy of fallot: comparison with cardiac magnetic resonance**

Bita Zargaran, Hassan Mottaghi Moghaddam, Mohammadreza Naghibi  
 Pediatric Department, Faculty of Medicine, Mashhad University of Medical Science, Mashhad, Iran

**Background and Aim:** The incidence of Tetralogy of Fallot (TOF) as the most common cyanotic congenital heart disease (CHD) is 7-10%. During this time, according to the patient situation pulmonary valve replacement (PVR) should be considered. cardiac magnetic resonance (CMR) is the gold standard but expensive modality. the electrocardiogram (ECG) is accessible in every clinic. The parameters of ECG can reveal the patient's prognosis. We aim to assess the correlation between electrocardiogram findings with cardiac parameters in CMR of the patients with repaired TOF.

**Method:** In this cross-sectional study, we collected the medical report of the patients who underwent TOF repair. According to echocardiography findings, CMR to decide PVR was done. The ECG was magnified by Photoshop and interpretation was done. The parameters in the CMR report and ECG were analysed by SPSS. we considered  $P < 0.05$  as the statistically significant  
**Results:** Of 20 patients (age  $14.26 \pm 9.17$ ) in this study, 75% underwent PVR. Patients with fQRS had significantly higher right ventricle diastolic (RVED) and systolic (RVES) volumes. We found no significant correlation between ventricles function with QT duration and QT correction (QTc). Also, low right ventricle ejection fraction (RVEF) significantly correlated with fragmented QRS (fQRS).

**Conclusions:** Increased RVED and RVES volume were shown in patients with fQRS. Early detection of lower RVEF could be based on the existence of fQRS. According to the parameters of ECG, we could assess the RV function to prove the decision of the PVR.

**Keywords:** Fragmented QRS, Cardiac magnetic resonance, tetralogy of fallot

## P-124

**BMI affects ventricular function recovery in children after PIMS-TS**

Halszka Kamińska<sup>1</sup>, Anna Rożnowska Wójtowicz<sup>1</sup>, Andrzej Cacko<sup>2</sup>, Bożena Werner<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology and General Pediatrics, Medical University of Warsaw, Poland; <sup>2</sup>Department of Medical Informatics and Telemedicine, Medical University of Warsaw, Poland

**Background and Aim:** Pediatric Inflammatory Multisystem Syndrome Temporally Associated with SARS-CoV-2 (PIMS-TS) affects cardiac function with usually fast improvement, but potential long-term effects are still unknown. Our aim was to assess body mass index's (BMI) impact on the pace of cardiac muscle recovery.

**Method:** Prospective study enclosed 172 children aged 1–17 years (mean 8.2 years) diagnosed with PIMS-TS between June 2020 and February 2022 and controlled 6 weeks and 6 months after the disease. In all patients at control points three-dimensional echocardiography (3D-ECHO) and global longitudinal strain (GLS) were used to assess left and right ventricular function. The results were compared between the subgroups of patients with BMI > 75 percentile and thinner children.

**Results:** In all patients the means of left and right ventricular ejection fraction (LVEF and RVEF) in 3D-ECHO and average GLS were within normal limits at the time of all check-up points. In all children the function of left ventricle improved further between 6 weeks and 6 months according to both GLS and LVEF ( $p < 0.05$ ) while right ventricular function remained unchanged. In patients with BMI > 75 percentile the mean of left ventricular GLS after 6 weeks was  $-20.22 \pm 3.07\%$  and after 6 months  $-22.01 \pm 2.26\%$ ; both significantly lower than in thinner children (respectively:  $-22.23 \pm 3.54\%$ ,  $p = 0.008$  and  $-24.01 \pm 3.59\%$ ,  $p = 0.034$ ). Right ventricular GLS was lower in children with higher BMI after 6 months ( $-23.12 \pm 3.01\%$  vs  $-25.83 \pm 3.88\%$ ,  $p = 0.028$ ), no difference was noted after 6 weeks. The comparison of 3D-ECHO LVEF and RVEF between children with higher and lower BMI showed no significant difference after 6 weeks (LVEF  $61.19 \pm 4.29\%$  vs  $60.67 \pm 2.63\%$ ,  $p = 0.496$  and RVEF  $63.55 \pm 3.61\%$  vs  $64.83 \pm 3.03\%$ ,  $p = 0.119$ ) and 6 months (LVEF  $62.92 \pm 2.95\%$  vs  $63.87 \pm 3.58\%$ ,  $p = 0.297$  and RVEF  $64.11 \pm 3.39\%$  vs  $64.87 \pm 2.59\%$ ,  $p = 0.231$ ).

**Conclusions:** Patients with BMI > 75 percentile have lower left ventricular GLS than thinner children during follow-up after PIMS-TS and lower right ventricular GLS 6 months after the disease.

Lower left ventricular GLS in children with BMI > 75 percentile may indicate poorer left ventricular performance in children with higher BMI.

GLS is applicable and sensitive test for functional assessment of cardiac muscle recovery after PIMS-TS.

**Keywords:** PIMS-TS, BMI, global longitudinal strain, follow-up, cardiac function recovery

## P-125

**The prognostic role of systolic longitudinal strain in congenital heart disease with systemic right ventricle: A systematic review and meta-analysis**

Dan M. Dorobantu<sup>1</sup>, Nurul H. Amir<sup>2</sup>, Curtis A. Wade<sup>3</sup>, Chetanya Sharma<sup>4</sup>, A. Graham Stuart<sup>5</sup>, Guido E. Pieleś<sup>6</sup>, Craig A. Williams<sup>3</sup>

<sup>1</sup>Children's Health and Exercise Research Center (CHERC), University of Exeter, Exeter, UK; Congenital Heart Unit, Bristol Royal Hospital for Children and Heart Institute, Bristol, UK; <sup>2</sup>Department of Translational Health Sciences, Bristol Medical School, University of Bristol, Bristol, UK; Faculty of Sports Science and Recreation, Universiti Teknologi MARA Cawangan Perlis, Kampus Arau, Arau, Perlis, Malaysia; <sup>3</sup>Children's Health and Exercise Research Center (CHERC), University of Exeter, Exeter, UK; <sup>4</sup>Congenital Heart Unit, Bristol Royal Hospital for Children and Heart Institute, Bristol, UK; <sup>5</sup>Congenital Heart Unit, Bristol Royal Hospital for Children and Heart Institute, Bristol, UK; National Institute for Health Research Cardiovascular Biomedical Research Centre, University of Bristol, UK; <sup>6</sup>Sports Medicine Department and the Athlete Health and Performance Research Centre, ASPETAR Qatar Orthopaedic and Sports Medicine Hospital, Doha, Qatar; Institute of Sport Exercise and Health, University College London, London, UK; National Institute for Health Research Cardiovascular Biomedical Research Centre, University of Bristol, UK

**Background and Aim:** Congenital heart disease (CHD) with systemic right ventricle (RV) are associated with high mortality and morbidity. There is need for better tools for risk stratification in this population, such as evaluating the RV using speckle tracking echocardiography (STE). This study aims to evaluate the current literature on the prognostic role of STE derived RV longitudinal strain (RV-SI) in CHD with systemic RV.

**Method:** EMBASE, Medline, Web of Science, Scopus and CENTRAL databases were searched from inception to February 2022, for terms related to CHD, STE and prognosis. Patients with transposition of the great arteries treated by atrial switch techniques (asTGA), congenitally corrected TGA (ccTGA) and hypoplastic left heart syndrome (HLHS) were selected. Outcomes of interest were classified as major adverse cardiac events (MACE) and included death, heart failure, heart transplant and/or ventricular arrhythmia. For asTGA and ccTGA a meta-analysis was possible, while for HLHS a synthesis approach was used.

**Results:** A total of  $n = 1254$  abstracts were screened,  $n = 114$  full-text papers assessed for eligibility and  $n = 11$  cohorts met the inclusion and exclusion criteria:  $n = 4$  on asTGA ( $n = 255$  patients),  $n = 4$  on ccTGA ( $n = 317$  patients),  $n = 4$  on pre-Norwood/Stage 1 HLHS ( $n = 113$  patients),  $n = 4$  on interstage HLHS ( $n = 155$  patients) and  $n = 3$  post-Glenn/Stage 2 HLHS ( $n = 69$  patients).

In pooled analysis of asTGA and ccTGA, RV-SI was found to be predictive of MACE both in univariable analyses (HR = 1.12/%, [1.03; 1.22]) and multivariable analyses (HR = 1.17/%, [1.05; 1.31]). This association was positive in single disease cohorts (asTGA or ccTGA) and mixed cohorts alike. In HLHS, RV-SI was not found to be predictive of MACE when measured before Norwood/Stage 1 (3/4 found no associations). Interstage RV-SI was associated with MACE in 4/4 cohorts, after Norwood/Stage and before Glenn/Stage 2. All 3 studies reporting RV-SI after Glenn/Stage 2 reported associations with MACE. (Table)

**Conclusions:** RV-SI should be used in risk stratification for patients with TGA and systemic RV (asTGA and ccTGA), being predictive of MACE. In HLHS, there is limited prognostic value of RV-SI measured before Norwood/Stage 1, but there is evidence to support its role in risk stratification during the interstage period, and after the Glenn/Stage 2 procedure.

**Keywords:** speckle tracking echocardiography, congenital heart disease, systemic right ventricle, systematic review, transposition of the great arteries, hypoplastic left heart syndrome

Associations of RV strain to outcomes in HLHS – synthesis of findings		
Study	Outcome	Measure of effect
Lin et al. 2018	Death or cardiac transplant	<i>Before Glenn/Stage 2</i> AUC: RV-S <sub>i</sub> : 0.71
Altit et al. 2019	Death or cardiac transplant	<i>Before Norwood/Stage 1</i> RV-S <sub>i</sub> : HR=1.07%, p=0.44
Rosner et al. 2019	Death, cardiac transplant, or heart failure	<i>Before or after Glenn/Stage 2</i> 9/15 death/transplant had RV-S <sub>i</sub> >-8% <i>Before Norwood/Stage 1</i> RV-S <sub>i</sub> : HR=1.33%, p=0.17
Colquitt et al. 2019	Death, cardiac transplant or more than moderate RV systolic dysfunction	<i>After Norwood/Stage 1</i> RV-S <sub>i</sub> : HR=1.36%, p=0.02
		<i>Before Glenn/Stage 2</i> RV-S <sub>i</sub> : HR=1.4%, p<0.01
		<i>After Glenn/Stage 2</i> RV-S <sub>i</sub> : HR=1.09%, p=0.032
		<i>Before Norwood/Stage 1</i> RV-S <sub>i</sub> : -15.8% vs -16.5%, p=0.7*
Borrelli et al. 2020 and Michielon et al. 2020	Death or cardiac transplant	<i>After Norwood/Stage 1</i> RV-S <sub>i</sub> : -16.3% vs -11.7%, p=0.02*
		RV-S <sub>i</sub> : multivariable coefficient 0.09%, p=0.003
		<i>Before Glenn/Stage 2</i> RV-S <sub>i</sub> : -16.6% vs -13.2%, p=0.03*
		<i>After Glenn/Stage 2</i> RV-S <sub>i</sub> : -15.5% vs -8%, p=0.0003*
Forsha et al. 2020	Death or cardiac transplant	<i>Before Norwood/Stage 1</i> All 6 deaths had basal septal RV-S <sub>i</sub> >-4.6%

\* Survivors vs events; RV-S<sub>i</sub>: right ventricle (RV) longitudinal strain; AUC: area under the receiver operating characteristic curve; HR, hazard ratio; HLHS, hypoplastic left heart syndrome.

**P-126 Hemodynamic analysis of pulmonary artery for congenital right-side heart disease in adulthood by 4D-Flow MRI**

Akio Inage<sup>1</sup>, Kanako Kishiki<sup>2</sup>, Tadahiro Yoshikawa<sup>2</sup>  
<sup>1</sup>Department of Pediatrics, Japanese Red Cross Medical Center, Tokyo, Japan; <sup>2</sup>Division of Pediatric Cardiology, Sakakibara Heart Institute, Tokyo, Japan; <sup>2</sup>Division of Pediatric Cardiology, Sakakibara Heart Institute, Tokyo, Japan

**Background and Aim:** 4D-flow MRI has been attracting attention as a tool that can visually and quantitatively evaluate and analyze blood flow dynamics. We focused on energy loss and helicity and evaluated hemodynamics in the pulmonary artery of congenital right-side heart disease in adulthood using 4D-flow MRI.

**Method:** In 20 adults who underwent cardiovascular MRI at Sakakibara heart institute, we measured energy loss (EL) and helicity of the main pulmonary artery (MPA). The maximum value (peak EL) in one heartbeat and the total value were obtained for EL. Helicity was defined as a vector quantity, with clockwise rotation being positive and counterclockwise rotation being negative, and the total value was calculated.

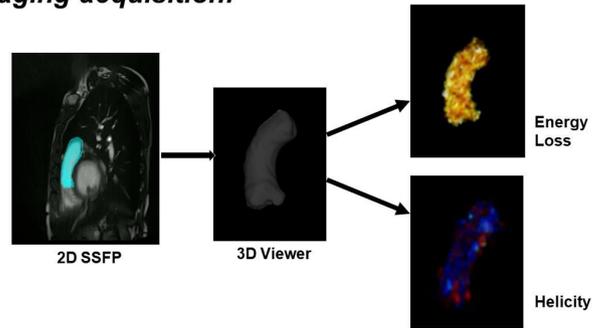
**Results:** The mean age was 27.9 years. The cases included 15 cases of postoperative tetralogy of Fallot (TOF) and 5 cases of congenital heart diseases with pulmonary arterial hypertension (CHD-PAH). The diagnosis of the CHD-PAH group was as follows: 2 postoperative TOF, 1 postoperative double outlet right ventricle, 1 postoperative atrial septal defect (ASD), and 1 unrepaired ASD. Peak EL, time to peak EL, and total helicity were higher in the CHD-PAH group than TOF group. Peak EL values were significantly higher during systole but were observed during diastole in 8 patients (40%). The combined mean value of helicity was positive, with clockwise predominance in 12 cases (60%). A strong correlation was found between peak EL and clockwise helicity (r = 0.68) and counterclockwise helicity (r = -0.73). In addition, a correlation was observed between diastolic EL/BSA and diastolic clockwise helicity (r = 0.51) and diastolic counterclockwise

helicity (r = -0.52). A correlation was also observed with the pulmonary regurgitant fraction (r = 0.49).

**Conclusions:** In this study, helicity in the MPA was dominant clockwise in congenital right-side heart disease in adulthood. It was suggested that EL was involved in helicity throughout the entire cardiac cycle and that peak EL had a particularly large effect.

**Keywords:** 4D-Flow MRI, adult congenital right-side heart disease, pulmonary artery, energy loss, helicity

**Imaging acquisition:**



While looking at both the 2D SSFP and phase images, which are cine images, we traced from the pulmonary valve to before the right and left pulmonary artery branches, and calculated energy loss and helicity.

**P-127 Non-invasive myocardial work indices in patients with fontan circulation**

Alessia Basso, Jolanda Sabatino, Roberta Biffanti, Giovanni Di Salvo, Domenico Sirico  
 Paediatric Congenital Cardiology Unit, Women and Child Health Department, Padova University Hospital, Padova, Italy

**Background and Aim:** There have been no quantitative analyses of the effects of Fontan circulation on myocardial work. Recently, a novel non-invasive method for calculating MW has been introduced on the basis of speckle tracking analysis with the estimation of LV pressure from brachial artery cuff pressure. The aim of this study was to evaluate the diagnostic performance of the non-invasive myocardial work indices in predicting subclinical myocardial work impairment in Fontan patients.

**Method:** A total of 69 patients were included and compared with healthy age- and sex- matched controls (CTRL). Ventricular systolic function and global longitudinal strain (GLS) were assessed. Cardiopulmonary exercise test was performed. Global myocardial work index (MWI) was calculated as the area of the LV pressure strain loops. From MWI, global Constructive Work (MCW), Wasted Work (MWW) and Work Efficiency (MWE) were estimated.

**Results:** The two groups were comparable for blood pressure, weight end height. Mean age of Fontan patients was 21.0±9.2 years. MWI (1162 ± 364 mmHg% vs 1777 ± 240 mmHg%, p < 0.001), MCW (1554 ± 450 mmHg% vs 2102 ± 221 mmHg%, p = 0.001) and MWE (90 ± 6% vs 96 ± 2% p = 0.001) were significantly reduced in Fontan patients compared with healthy CTRL. Moreover, GLS (-13, 9± 3, 1 % vs -21, 2 ± 1.5%, p < 0.001) and the ejection fraction (EF) (58, 9 ± 4.5% vs 63.3 ± 3.9%, p < 0.002) were significantly lower in Fontan patients. Fontan patients with normal EF showed, however, significantly reduced values of MWI compared with CTRL

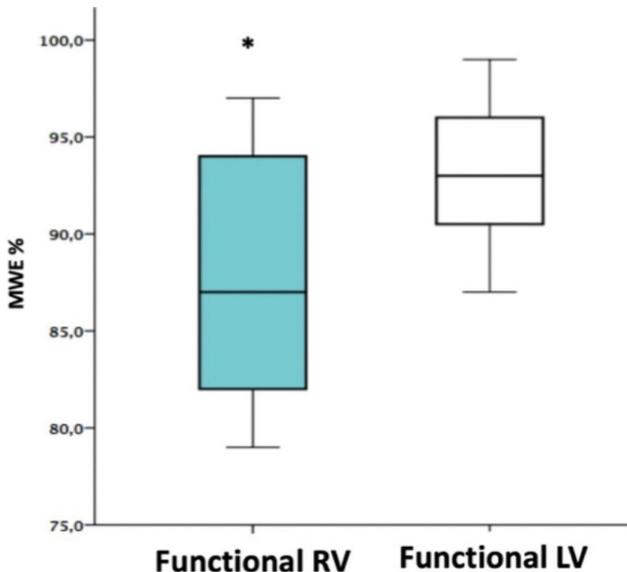
( $p < 0.05$ ). Fontan patients with functional right ventricle showed significantly reduced MWE compared with patients with functional left ventricle ( $p = 0.030$ ).

In univariate analysis, peak  $VO_2$  was significantly associated with age, SatO<sub>2</sub>, MWI. In multivariate regression, lower peak  $VO_2$  was associated with older age ( $p = 0.003$ ) and lower MWI ( $p = 0.026$ ).

**Conclusions:** Fontan physiology is associated with disadvantageous ventricular work. In Fontan patients estimation of MWI may be a more sensitive indicator of myocardial work impairment compared with EF, and is able to predict exercise capacity.

**Keywords:** Fontan, echocardiography, myocardial work

MWE in Fontan circulation



Fontan patients with functional right ventricle showed significantly reduced MWE compared with patients with functional left ventricle ( $p = 0.030$ )

#### P-128

##### How not to misdiagnose anomalous origin of the right coronary artery from the pulmonary artery in the pediatric population

Hiyam Mahmoud<sup>1</sup>, Eliza Cinteza<sup>2</sup>, Cristiana Voicu<sup>1</sup>, Cristina Filip<sup>1</sup>, Georgiana Nicolae<sup>1</sup>, Gabriela Duica<sup>2</sup>, Amelia Aria<sup>1</sup>, Iulian Rotaru<sup>1</sup>, Irina Margarint<sup>1</sup>, Tammam Youssef<sup>1</sup>, Alin Nicolescu<sup>1</sup>

<sup>1</sup>"Marie Curie" Emergency Children's Hospital, Bucharest, Romania;

<sup>2</sup>"Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania

**Background and Aim:** Coronary artery disease in the pediatric population gathers different aspects than in adults. The most frequent pathologies include anomalies of origination, course, or intrinsic anatomy of the coronaries. Anomalous origin of the right coronary artery from the pulmonary artery, ARCAPA, is an extremely rare congenital heart disease (CHD). First described in 1885, this anomaly encountered only 200 cases until nowadays. We report two cases of ARCAPA diagnosed over the last year at the "Marie Curie" Emergency Children's Hospital in Bucharest, Romania, the latter being diagnosed at distance from open-heart surgery for another CHD.

**Method:** Screening echocardiography for a 2-month-old girl raised the suspicion of coronary artery fistula and, in evolution, she presented progressive dilation of the left coronary artery. Therefore, she

was admitted for complete evaluation at the age of 3 years old. Coronarography visualized extensive collateral circulation, increased diameter of the left main coronary artery, and retrograde filling of the right coronary towards the pulmonary artery trunk, confirming the diagnosis of ARCAPA. The patient went through surgical correction with reimplantation of the right coronary to the ascending aorta.

Our second case is of a 2-year-10-month-old male child diagnosed with aortopulmonary window surgically corrected at birth. Routine echocardiographic reevaluation raised the suspicion of a coronary artery anomaly and coronary angiography set the diagnosis of ARCAPA. Surgical correction was performed. Both patients had a full postoperative recovery and good evolution at 6 months follow-up.

**Results:** Although asymptomatic, this pathology can lead to cardiac syncope, congestive heart failure, and sudden cardiac death caused by "steal syndrome" and exercise-induced myocardial ischemia. The diagnostic algorithm is based on paraclinical investigations. The electrocardiogram can be normal or may show indirect signs of ischemia. Routine echocardiography can raise the suspicion of coronary artery anomaly, but invasive coronarography, angioCT, and angioMRI are mandatory to confirm the diagnosis.

**Conclusions:** Diagnostic tools in ARCAPA are centered on imagistic investigations. Because echocardiography can reveal indirect signs of anomalous origin of the right coronary artery, the diagnosis of ARCAPA should be considered until invasive imagistic investigations prove otherwise. Early surgical corrective intervention is recommended to prevent cardiac complications, even if the patient is asymptomatic.

**Keywords:** coronary artery disease, ARCAPA, congenital heart disease, reimplantation, ischemia, coronary angiography

#### P-130

##### Early cardiac involvement in multisystem inflammatory syndrome in children secondary to different SARS-CoV-2 virus variants infection

Alessia Basso<sup>1</sup>, Domenico Sirico<sup>1</sup>, Giulia Bragantini<sup>2</sup>, Jolanda Sabatino<sup>1</sup>, Elena Reffo<sup>1</sup>, Giovanni Di Salvo<sup>1</sup>

<sup>1</sup>Paediatric Congenital Cardiology Unit, Women and Child Health Department, Padova University Hospital, Padova, Italy; <sup>2</sup>Cardiology Unit, San Bortolo Hospital, Vicenza, Italy

**Background and Aim:** Multisystem Inflammatory Syndrome in Children (MIS-C) is a known severe acute condition affecting children previously exposed to SARS-CoV-2. Cardiovascular involvement in MIS-C is frequent. The aim of our study was to describe the early cardiac manifestations in patients with MIS-C, before and after the spread of the Omicron variant during the last months of 2021 in Veneto region in Italy.

**Method:** This is a retrospective observational two centers study. 41 patients with confirmed MIS-C diagnosis were enrolled. Children were divided into two temporal groups: 32 subject in the "pre-omicron era" (Apr 2020– Oct 2021) or group A and 9 in the "Omicron era" (Jan 2022– Aug 2022) or group B. Clinical, laboratory and microbiological data were measured. All children underwent standard transthoracic echocardiography with speckle-tracking analysis of left ventricle global longitudinal strain (LVGLS). 23 (72%) patients of the group A and 6 (67%) of the group B underwent CMR during the acute/subacute phase.

**Results:** Age of presentation was comparable between groups. There were no significant differences in clinical presentation. Cardiovascular symptoms were similar between the groups. The need for inotropic support was significantly higher for the group B (X<sup>2</sup> 4.56,  $p = 0.03$ ). Overall early survival was 100%. All patients showed an hyperinflammatory state, however PCR and D-Dimer

were significantly higher in the Omicron patients as well as NT-proBNP. No differences in LVEF and LVGLS were detected between the two groups (LVEF 58±10.6% vs. 54±8.3%, p = 0.22; LVGLS -17±4.3% vs -18.4±3.2, p 0.52). Coronary dilatation was infrequent and similar between groups. LGE evidence on CMR with non-ischemic pattern was comparable between the two groups (35.2% and 50%, X2 0.46, p = 0.49). During the “Omicron era” patients with MIS-C showed higher rate of pericardial effusion regardless of the diagnostic imaging technique, echocardiography or CMR scan.

**Conclusions:** MIS-C can occur in a small but not negligible proportion of children previously affected by Sars-Cov-2 infection. Despite mild differences in clinical, laboratory and instrumental findings, the extent of subclinical cardiac damage in MIS-C did not differ before and after Omicron variant spread, highlighting the etiologic pivotal role of immuno-mediate response over the infection by different SARS-CoV-2 virus variants.

**Keywords:** Multisystem Inflammatory Syndrome in Children, COVID19, speckle-tracking, echocardiography, cardiac MRI,

**P-132**

**Contrast-free 3D MRA for efficient sequential segmental analysis in patients with congenital heart disease- single center study**

Anastasia Fotaki<sup>1</sup>, Kuberan Pushparajah<sup>1</sup>, Karl Kunze<sup>2</sup>, Radhouene Neji<sup>2</sup>, Alessandra Frigiola<sup>3</sup>, Rene Botnar<sup>1</sup>, Claudia Prieto<sup>1</sup>  
<sup>1</sup>School of Biomedical Engineering and Imaging Sciences, King’s College London, London, United Kingdom; <sup>2</sup>MR Research Collaborations, Siemens Healthcare Limited, Camberley, United Kingdom; <sup>3</sup>Guy’s and St Thomas’s Hospital, Adult Congenital Heart Disease Department

**Background and Aim:** 3D-MRA sequences are utilised for the morphological evaluation of patients with Congenital Heart Disease (CHD)1, 2. Limitations of the current approaches include long scan times due to diaphragmatic navigators, artefacts in areas of turbulent or slow flow (Fontan pathway) and off-resonance artefacts, primarily

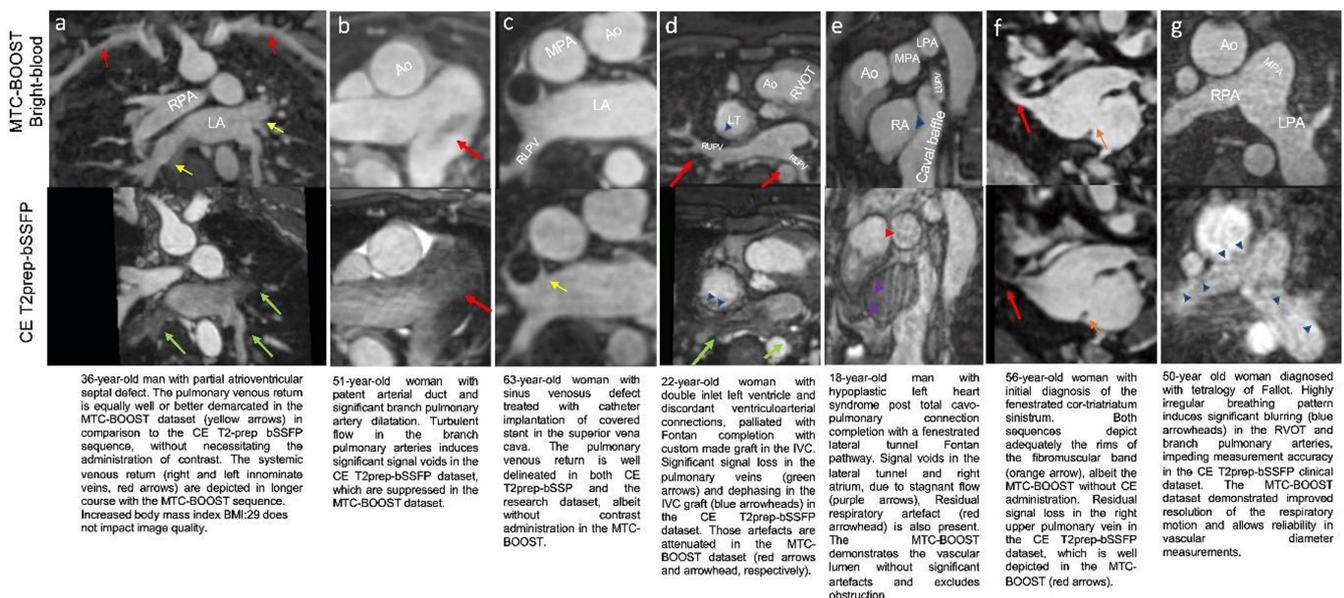
in the pulmonary veins2, 3, 4. Gadolinium based contrast agents are often employed to enhance the anatomical visualisation, albeit with potential side effects, increased preparation time and costs5. We have proposed a sequence that exploits a Magnetisation Transfer preparation pulse (MTC-BOOST)6, that can mitigate the aforementioned shortcomings. The aim of this study is to validate the MTC-BOOST against the clinical contrast-enhanced (CE) sequence.

**Method:** An MTC-BOOST research sequence was evaluated in 15 patients (40 ±14 years old, 10 Female) with CHD on a 1.5T system (Aera, Siemens). 2D image-based navigation7 was utilised to enable respiratory and cardiac motion correction. The clinical sequence was the contrast-enhanced 3D T2-prep bSSFP (Gadobutrol 0.15mmol/kg) with same mage resolution (1.5mm3) and similar acquisition window parameters. Acquisition time for the clinical and the MTC-BOOST dataset was recorded. Two experts blinded to the technique used, recorded their diagnostic confidence to make full sequential segmental analysis (≥ 3: full diagnostic confidence) and assessed image quality scores for 14 intrapericardiac structures using 5-point Likert scale (3≥ diagnostic).

**Results:** Good quality depiction of all intrapericardiac structures was achieved with MTC-BOOST in short scan time, 10 ±2 min vs 14 ±7min (clinical), p<0.0001. Diagnostic confidence was similar to CE clinical sequence [4(4, 4) vs 4(4, 4) p>0.99]. Image quality scores for the assessed vascular structures were consistently high in the MTC-BOOST bright-blood dataset with marked superiority of the MTC-BOOST sequence in the delineation of pulmonary and systemic veins and main pulmonary artery (p<0.01). MTC-BOOST resulted in improved luminal signal and was robust towards rapid, turbulent flow and off-resonance effects (Fig. 1).

**Conclusions:** The MTC-BOOST sequence offers superior quality imaging to the conventional CE T2-prep bSSFP sequence and is equivalent to the diagnostic performance of the CE T2-prep bSSFP in shorter acquisition time. It has potential to enhance the time- and cost-effectiveness of bright-blood 3D-WH imaging in patients with CHD.

**Keywords:** contrast-free MRA, sequential segmental analysis



**Figure 1:** Comparison of bright blood MTC-BOOST images against the clinical contrast-enhanced T2-prepared balanced steady state free precession (CE T2-prep bSSFP) sequence. Off-resonance artefacts (a,d,e), flow related artefacts in areas of turbulent flow (b) and slow flow (e) along with respiratory motion artefacts (g) are attenuated with the proposed MTC-BOOST sequence. Abbreviations: CE: contrast-enhanced, IVC: inferior vena cava, MTC-BOOST: magnetisation transfer contrast bright- and black-blood phase sensitive, T2prep-bSSFP: T2prepare-balanced steady state free precession

## P-133

**Left atrial function and strain in pediatric hypertrophic cardiomyopathy with preserved ejection fraction**Iolanda Muntean<sup>1</sup>, Cristina Blesneac<sup>2</sup>, Amalia Fagarasan<sup>1</sup>, Liliana Gozar<sup>1</sup>, Carmen Suteu<sup>2</sup>, Ana Sorina Pasc<sup>2</sup>, Laura Beligan<sup>2</sup><sup>1</sup>University of Medicine, Pharmacy, Science and Technology „George Emil Palade“ of Târgu Mureş, Romania & Emergency Institute for Cardiovascular Diseases and Transplantation of Targu Mures, Romania; <sup>2</sup>Emergency Institute for Cardiovascular Diseases and Transplantation of Targu Mures, Romania

**Background and Aim:** Left atrial dysfunction and strain are considered early markers of diastolic dysfunction in adults with hypertrophic cardiomyopathy. Assessment of left atrial function and strain in children is not well studied yet. The aim of the present study was to assess left atrial mechanics in pediatric patients with hypertrophic cardiomyopathy with preserved ejection fraction compared with healthy controls.

**Method:** Left atrial two-dimensional volumetric and speckle-tracking parameters were analyzed in fifteen children with hypertrophic cardiomyopathy with preserved ejection fraction and fifteen age and sex matched controls. Maximum and minimum indexed left atrial volume and left atrial strain parameters (left atrial reservoir strain, conduit strain and contractile strain) were measured offline, using special software. Also, conventional echocardiographic and tissue Doppler indices were performed. Brain natriuretic peptide level was determined in all studied children.

**Results:** Maximum and minimum indexed left atrial volumes were significantly higher in children with hypertrophic cardiomyopathy than in controls (23, 59+/-8, 05 vs 16, 59+/-5, 17, p = 0, 013 for maximum, 7, 03+/-2, 93 vs 3, 14+/-1, 38, p = 0, 0001 for minimum, respectively). Compared to controls, children with hypertrophic cardiomyopathy had lower left atrial reservoir strain (36% vs 55%, p = 0, 0001) and left atrial conduit strain (absolute value 19, 49 vs 41, 96, p = 0, 0001). The maximum septal thickness (z score) has correlated with the maximum and minimum indexed left atrial volumes (r = 0, 735, p = 0, 0001 and r = 0, 746, p = 0, 0001, respectively), left atrial reservoir strain (r = 0, 538, p = 0, 005), left atrial conduit strain (r = 0, 565, p = 0, 003), E'/A' (r = 0, 418, p = 0, 034), E/E' (r = 0, 485, p = 0, 012) and also with the brain natriuretic peptide level (r = 0, 633, p = 0, 001).

**Conclusions:** Children with hypertrophic cardiomyopathy with preserved ejection fraction have reduced left atrial mechanics measurable by volumetric and strain analysis. The septal thickness may determine the degree of impairment of the left atrial function. Acknowledgements: "This work was supported by the University of Medicine, Pharmacy, Science and Technology „George Emil Palade“ of Târgu Mureş, Research Grant number 511/5/17.01.2022".

**Keywords:** Left atrial function, strain, hypertrophic cardiomyopathy, children

## P-134

**The exceptional value of t1 native mapping during cardiac magnetic resonance, in follow up of patients with surgically corrected fallot tetralogy**Niki Lama<sup>1</sup>, Panagiotis Zachos<sup>2</sup>, Efstiratos Karavasilis<sup>1</sup>, Georgios Brouboulis<sup>1</sup>, Ioannis Papadopoulos<sup>1</sup>, Alkistis Kalesi<sup>5</sup>, Maria Karakosta<sup>5</sup>, Sotiria Apostolopoulou<sup>4</sup>, Nearchos Kasinos<sup>5</sup>, Antonios Destounis<sup>5</sup>, Anastasios Theodosis Georgilas<sup>5</sup>, Nikolaos L. Kelekis<sup>3</sup><sup>1</sup>Research Unit of Radiology and Medical Imaging, <sup>2</sup>2nd Department of Radiology, School of Medicine, National and Kapodistrian University ofAthens, Athens, Greece; <sup>2</sup>Department of Health Economics, School of Medicine, National and Kapodistrian University of Athens, Athens, Greece; <sup>3</sup>2nd Department of Radiology, School of Medicine, National and Kapodistrian University of Athens, Athens, Greece; <sup>4</sup>Pediatric Cardiology and Adult Congenital Heart Disease Department, Onassis Cardiac Surgery Center, Athens, Greece; <sup>5</sup>Echocardiography Training Center, Cardiology Department, General Hospital Tzaneio, Athens, Greece

**Background and Aim:** Imaging has a key role in follow up of patients with surgically corrected tetralogy of Fallot (TOF), and especially MRI can quantitatively evaluate the pulmonary valve insufficiency and right ventricular function, providing valuable prognostic information's. T1 mapping is a pixel-wise quantification method, of myocardial T1 values. Our goal is to highlight the benefit of T1 native, in detailed characterization of dynamic changes in heart and evaluation of these patients.

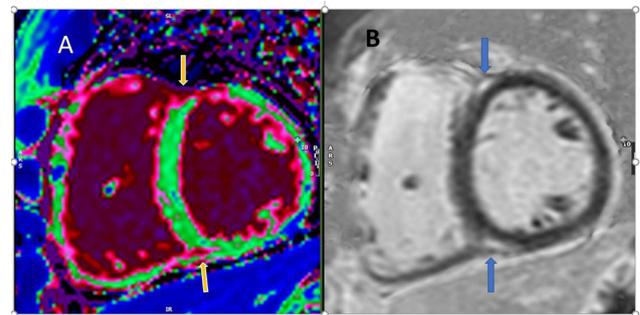
**Method:** All patients underwent a CMR study with our 3T MRI scanner. A simple linear regression analysis was conducted to investigate possible associations between the global native T1 relaxation time and volumetric ventricular data such as end-diastolic volume, end-systolic volume, ejection fraction and stroke volume. All analyses were performed using IBM SPSS v.20 software.

**Results:** We studied 11 patients who underwent CMR. The mean values were: native global T1 relaxation time = 1265, 45 ± 40.73ms; right/left ventricular stroke volume = 56.77±22.71/43.14±7.65ml; right/left ejection fraction = 49.09±8.79/55.82 ± 7.65%; right/left EDVi = 117.60±44.10/79.08±22.32ml; right/left ESVi = 60.75±26.42/35.93±15.67 ml. The regression analyses results indicated that there is strong correlation between the global native T1 relaxation time and the function of the right ventricle. In particular, global native T1 relaxation time was a significant predictor for stroke volume [F(1, 9) = 13.019; p = 0.006], end-diastolic volume [(F(1, 9) = 20.427; p = 0, 001] and for end-systolic volume [F(1, 9) = 10.145; p = 0, 011]. Almost all patients had not elevated LV Native T1.

**Conclusions:** In our study we highlight the benefit of T1 native mapping in detailed characterization of myocardial dynamic changes and evaluation of patients with surgically corrected TOF. T1 Native had significant correlation with RV volumes. Native T1 mapping also showed an added value, revealing focal fibrosis at the right ventricle insertion points, but was unable to detect areas of fibrosis in the right ventricle.

**Keywords:** T1 native, CMR, Fallot

## T1 NATIVE vs LGE



T1 NATIVE SA image (left side) of a patient with surgically corrected Fallot tetralogy in correlation with LGE SA image (right side).

**P-136****Experience with the left ventricular training for congenitally corrected transposition of the great arteries**Olena Stohova<sup>1</sup>, Nadia Rudenko<sup>2</sup>, Illya Yemets<sup>1</sup><sup>1</sup>Ukrainian Children's Cardiac Center, Kyiv; <sup>2</sup>Shupyk National Medical Academy of Postgraduate Education, Kyiv, Ukraine

**Background and Aim:** Assessment of left ventricular training in patients with congenitally corrected transposition of the great arteries (CCTGA).

**Method:** Pulmonary artery banding (PAB) was performed in 11 patients with CCTGA and intact interventricular septum or restrictive interventricular defect from 2005 to 2021. Mean age was 29 (7–82) mo, and mean weight was 11, 5 (5, 8–22) kg. Patients were assessed by intraoperative pressure measurement, intraoperative transesophageal echocardiography (ITEE) and transthoracic echocardiography (TEE) after the operation.

**Results:** Indications for operation were: left ventricular training for restricted ventricular septal defect in 2 patients; left ventricular training for intact interventricular septum in 9 patients; severe tricuspid valve insufficiency with intact ventricular septum in 4 patients; moderate tricuspid valve insufficiency with intact ventricular septum in 3 patients. Intraoperative left ventricular pressure consisted in average of 64% (53%–82%) from systemic pressure. Peak pressure gradient across the pulmonary artery measured by the TTE was in average 37 (18–60) mmHg in early postoperative period. There was no operative mortality. One patient required reducing the band length due to hemodynamic changes on the second postoperative day. In the follow up period two patients died after 8 months and 5 years after PAB. The next operation stage, double switch operation, was performed in 5 patients in average of 48, 6 (39–57) mo after PAB. Peak pressure gradient across the pulmonary artery measured by the TTE was in average of 75, 4 (50–120) mmHg, LV myocardial mass index was 66, 8(54, 5–78, 9) g/m<sup>2</sup>. Left ventricular pressure consisted in average of 90, 2% (77%–111%) from systemic pressure by angiography.

**Conclusions:** PAB can be recommended as a stage operation for patients with CCTGA with intact ventricular septum or restricted ventricular septal defect. On average, it took about 4 years to LV training in order to carry out the next stage – the double switching operation.

**Keywords:** congenitally corrected transposition of the great arteries, left ventricular training

**P-137****The impact of maternal covid-19 during pregnancy on the fetal heart: does it cause subclinical postnatal echocardiographic cardiac dysfunction?**Liat Gelemter Yaniv<sup>1</sup>, Arieh Riskin<sup>2</sup>, Livnat Sharkansky<sup>3</sup><sup>1</sup>Department of Pediatrics, Bnai Zion Medical Center, Haifa, Israel, The Ruth and Bruce Rappaport Faculty of Medicine, Technion, Haifa, Israel;<sup>2</sup>Department of Neonatology, Bnai Zion Medical Center, Haifa, Israel, The Ruth and Bruce Rappaport Faculty of Medicine, Technion, Haifa, Israel; <sup>3</sup>Department of Pediatrics, Bnai Zion Medical Center, Haifa, Israel

**Background and Aim:** COVID-19 infection during pregnancy may be associated with increased risks of preeclampsia, preterm birth and other adverse pregnancy outcomes and poses a higher risk for fetal distress. A potential effect on the fetal heart may result in myocarditis, cardiomyopathy, or valves malformation, as was suggested for other viruses. We looked for subclinical echocardiographic findings (valvular, pulmonary hypertension and

myocardial function), in asymptomatic neonates born to mothers who had COVID-19 during pregnancy.

**Method:** All neonates born to mothers with COVID-19 during pregnancy underwent echocardiography between days of life 1–4. Time of infection, severity, immunization status, gestational and perinatal history were collected. All mothers recovered from COVID-19 at the time of delivery and were otherwise healthy.

**Results:** 55 asymptomatic neonates underwent echocardiography. 85.4% were born at term. 96.3% had normal weight. Valvular findings occurred in 3 newborns (5.4%): 2 mild mitral regurgitation and one had severe pulmonary stenosis (PS) requiring intervention. Three neonates had ventricular septal defects and one had arch hypoplasia. All neonates had normal myocardial contractility and TAPSE. 14.5% of mothers had COVID-19 during the first trimester, 27.2% had COVID-19 during the second trimester and 45.4% had COVID-19 during the third trimester. 34.5% of mothers were asymptomatic and 51% had mild symptoms. 36.8% of mothers were not immunized against COVID-19 at all.

**Conclusions:** Of the asymptomatic newborns, one had severe valvular malformation (PS). None developed cardiac dysfunction, (myocarditis, cardiomyopathy). Structural abnormalities (VSD, arch hypoplasia) were not considered to be related to COVID-19. None had pulmonary hypertension. Most women had asymptomatic/ mild COVID-19. It might be attributed to Omicron being the dominant variant during the study period and vaccine protection in some. At this point, additional cardiac (valvular or myocardial) morbidity attributed to maternal COVID-19 during pregnancy was not detected among asymptomatic newborns compared to the known baseline prevalence. Further data is needed to elaborate our knowledge of the possible cardiac effect of maternal illness. Also, it cannot be determined whether maternal disease severity influences fetal cardiac manifestations since all mothers had a mild disease.

**Keywords:** maternal COVID-19, neonatal COVID-19, neonatal echocardiography

**P-138****A case of multiple life-threatening cardiac rhabdomyomas in a newborn baby**Sally Hall, Teresa Brooks, Jan Forster, Sarah Whiting, Sara Moore, Grazia Delle Donne, Antigoni Deri  
Leeds Congenital Heart Unit, Leeds, UK

**Background and Aim:** Intracardiac rhabdomyomas are rare, affecting just 0.14% of fetuses. Although histologically benign, the position of the muscular tumours within the heart can cause life threatening complications. Here we describe the presentation, management and progress of a newborn baby with multiple, life-threatening rhabdomyomas.

**Method:** The patient initially presented to our service in late fetal life at 39+4 weeks gestation when a growth scan revealed several muscular swellings within the heart. The mother was transferred to our tertiary cardiac centre where fetal echo showed extensive rhabdomyomas. Owing to biventricular outflow tract obstruction, the family was counselled that prognosis was poor. However, following induction of labour, the baby was born in good condition. Echo showed multiple well defined rhabdomyomas within both ventricles and the right atrium. Although the ventricular rhabdomyomas impeded on both ventricular outflow tracts, obstruction was mild with a dynamic Doppler trace in the right ventricular outflow tract of 2.6m/s and a maximum velocity in the left ventricular outflow tract of 2.1m/s.

Over the first few days of life the baby developed significant arrhythmias with variable AV block and episodes of atrial

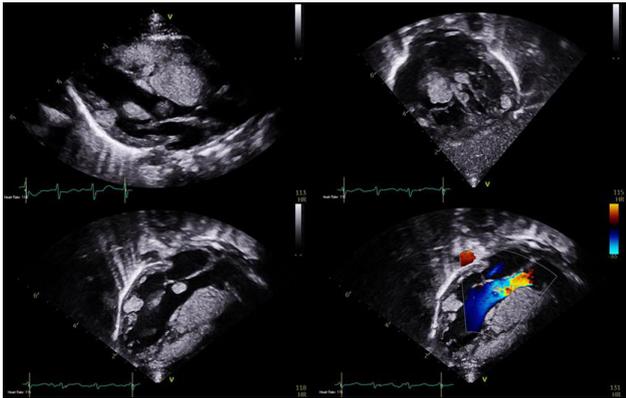
tachycardia. The multidisciplinary team was of the opinion that the child was at high risk of sudden death either by occlusion of the outflow tracts by one or more rhabdomyoma or as a result of a catastrophic arrhythmia. Surgery was not an option due to the quantity and distribution of the rhabdomyomas and so a medical treatment was sought. Following a literature review, the baby was commenced on sirolimus, an mTOR inhibitor immunosuppressant more commonly used as an anti-rejection medication post renal transplant.

**Results:** Early evidence of improvement was seen on echo at 4 and 8 weeks. Sirolimus treatment continued due to persistent arrhythmias. By 8 months all the ventricular rhabdomyomas had vanished except for a small residual right atrial tumour. In addition, an ambulatory ECG was clear of arrhythmias.

**Conclusions:** Our experience in using sirolimus to treat cardiac rhabdomyomas supports the ever increasing body of evidence that the immunosuppressant drug promotes rhabdomyoma regression. In addition, the drug led to resolution of arrhythmias, dramatically reducing the risk of sudden death.

**Keywords:** rhabdomyoma, sirolimus, arrhythmia

#### Multiple intracardiac rhabdomyomas



Although extensive rhabdomyomas could be seen within the heart on an echo shortly after birth, there was no significant obstruction to flow

#### P-139

##### Utility of longitudinal strain in depicting risk of major adverse cardiac events in children with diagnosed myocarditis

Nele Rolfs<sup>1</sup>, Bernd Opgen Rhein<sup>2</sup>, Felix Berger<sup>3</sup>, Thomas Pickardt<sup>4</sup>, Daniel Messroghli<sup>5</sup>, Stephan Schubert<sup>6</sup>, Franziska Seidel<sup>3</sup>, Fatima I Lunze<sup>7</sup>

<sup>1</sup>Department of Congenital Heart Disease - Pediatric Cardiology, German Heart Center Berlin, Berlin, Germany; <sup>2</sup>Department for Pediatric Cardiology, Charité – Universitätsmedizin Berlin, corporate member of Freie Universität Berlin and Humboldt-Universität zu Berlin, Berlin, Germany; <sup>3</sup>Department of Congenital Heart Disease - Pediatric Cardiology, German Heart Center Berlin, Berlin, Germany; <sup>4</sup>Department for Pediatric Cardiology, Charité – Universitätsmedizin Berlin, corporate member of Freie Universität Berlin and Humboldt-Universität zu Berlin, Berlin, Germany; <sup>5</sup>Department of Congenital Heart Disease - Pediatric Cardiology, German Heart Center Berlin, Berlin, Germany; <sup>6</sup>Department for Pediatric Cardiology, Charité – Universitätsmedizin Berlin, corporate member of Freie Universität Berlin and Humboldt-Universität zu Berlin, Berlin, Germany; <sup>7</sup>Department of Congenital Heart Disease - Pediatric Cardiology, German Heart Center Berlin, Berlin, Germany

member of Freie Universität Berlin and Humboldt-Universität zu Berlin, Berlin, Germany; DZHK (German Centre for Cardiovascular Research), partner site Berlin, Germany; <sup>4</sup>Competence Network for Congenital Heart Defects, Berlin, Germany; <sup>5</sup>Internal Medicine - Cardiology, German Heart Center Berlin, Germany; Internal Medicine and Cardiology, Charité – Universitätsmedizin Berlin, corporate member of Freie Universität Berlin and Humboldt-Universität zu Berlin, Berlin, Germany; DZHK (German Centre for Cardiovascular Research), partner site Berlin, Germany; <sup>6</sup>Center for Congenital Heart Disease/Pediatric Cardiology, Heart- and Diabetescenter NRW & University Clinic of Ruhr-University Bochum, Bad Oeynhausen, Germany; Department of Congenital Heart Disease - Pediatric Cardiology, German Heart Center Berlin, Berlin, Germany; DZHK (German Centre for Cardiovascular Research), partner site Berlin, Germany; <sup>7</sup>Department of Congenital Heart Disease - Pediatric Cardiology, German Heart Center Berlin, Berlin, Germany; DZHK (German Centre for Cardiovascular Research), partner site Berlin, Germany

**Background and Aim:** Myocarditis has various clinical manifestation and may lead to heart failure with left ventricular (LV) dysfunction and major adverse cardiac events (MACE). Conventional echocardiography has limited diagnostic value in these patients. Therefore, we aim to evaluate the diagnostic and prognostic value of speckle tracking echocardiography derived global longitudinal strain (GLS) in children with diagnosed myocarditis.

**Method:** We enrolled patients admitted at the pediatric cardiology department of the German Heart Center Berlin and Charité Universitätsmedizin Berlin between 2015 and 2021. All patients had confirmed diagnosis of myocarditis, either by endomyocardial biopsy (EMB) and/or cardiac magnetic resonance imaging (CMR). We assessed echocardiographic parameters at initial admission including functional and myocardial performance parameters. Comparison was made between patients with and without MACE including severe arrhythmias, heart transplantation (HTX), mechanical circulatory support (MCS), resuscitation and death. We aim to provide a pilot-study within the multicentered, prospective myocarditis registry “MYKKE”.

**Results:** We included 20 patients with a median age (IQR) of 3, 8 (0, 6–15, 0) years, 60% were male. Diagnosis of myocarditis was confirmed in all patients by EMB (n = 18) and/or CMR (n = 6). Median LV ejection fraction (LVEF) was 25 (19–54) %, end-diastolic dimension (EDD) Z-score 4, 6 (2, 3–7, 1) and median GLS epicardial -7, 8 (-4, 5; -12, 2) %, mid-wall -8, 0 (-5, 6; -14, 5) % and endocardial -9, 7 (-6, 3; -14, 7) %. Fifteen patients (75 %) experienced at least one MACE (MCS n = 13, resuscitation n = 8, severe arrhythmias n = 7, HTX n = 6, death n = 3). Patients with MACE had significantly lower LVEF (21 vs. 62 %, p = 0, 001) and GLS (epicardial -5, 9 vs. -17, 2 %; mid-wall -6, 8 vs. -17, 8 %; endocardial -8, 0 vs. -20, 5 %, for all p ≤ 0, 002) compared to those without MACE. Five Patients had preserved LVEF > 50 % with median GLS epicardial -17, 2 %, mid-wall -17, 8 % and endocardial -20, 5 %. Of those, only one presented with MACE (atrioventricular block III°). Echocardiographic evaluation in that patient revealed discrepancy between preserved LVEF and reduced GLS.

**Conclusions:** GLS is a promising parameter for assessing myocardial performance and predicting MACE in children with diagnosed myocarditis. In patients with preserved LVEF GLS might also be a reliable parameter. Therefore, an evaluation of the entire MYKKE cohort is planned.

**Keywords:** myocarditis, echocardiography, global longitudinal strain, MACE

## P-140

**Giant coronary aneurysm after kawasaki disease: A case report**

Marisa Pereira<sup>1</sup>, André Carvalho<sup>2</sup>, Ana Reis E Melo<sup>3</sup>, Ana Reis E Melo<sup>4</sup>, Ana Correia Costa<sup>1</sup>, Ana Correia Costa<sup>5</sup>

<sup>1</sup>Pediatric Cardiology Department, Centro Hospitalar Universitário de São João, Porto, Portugal; <sup>2</sup>Radiology Department, Centro Hospitalar Universitário de São João, Porto, Portugal; <sup>3</sup>Pediatric Department, Centro Hospitalar Universitário de São João, Porto, Portugal; <sup>4</sup>Biomedicine Department, Faculty of Medicine, University of Porto, Porto, Portugal; <sup>5</sup>Department of Gynecology-Obstetrics and Pediatrics, Faculty of Medicine, University of Porto, Porto, Portugal

**Background and Aim:** Kawasaki disease is one of the most common childhood vasculitis. It is typically self-limiting but may progress to the development of coronary artery aneurysms, which occurs in approximately 25% of untreated cases; with adequate treatment reducing the risk to 3-5%.

**Method:** We present the case of a previously healthy 3-year-old boy, evaluated in the emergency department because of 6 days of fever,odynophagia and asthenia. Physical examination revealed cheilitis, raspberry tongue, bilateral non-exudative conjunctivitis, trunk rash, rash and perineal desquamation, edema and erythema of palms, bilateral cervical adenopathies, gallop rhythm and hypotension. The analytical study demonstrated anemia, leukocytosis, thrombocytosis; hyponatremia, hypocalcemia; hypoalbuminemia; elevation of TGP, GGT and DHL; hyperbilirubinemia; hypertriglyceridemia; elevation of ferritin and C-reactive protein; elevation of troponin I and BNP; and sterile pyuria. The electrocardiogram was normal. The echocardiogram showed mild depression of left ventricular systolic function and normal coronary arteries. Considering these findings, the diagnostic hypothesis of complete Kawasaki disease was made, and timely and adequate treatment was initiated. Nevertheless, on the fourteenth day of the disease, he evolved into the development of giant coronary artery aneurysms, initially identified on echocardiography (giant left coronary artery aneurysm, z-score +38.7; dilation of right coronary artery, z-score +2.78) and later confirmed by CT angiography (giant fusiform aneurysm of left anterior descending artery, z-score +43.18; focal dilation of left circumflex artery, z-score +11.79; diffuse fusiform aneurysm of right coronary artery, z-score +18.01).

**Results:** About 9 months later, coronary angiography was performed and revealed the persistence of a giant aneurysm of the common trunk of the left coronary artery. He is under hypocoagulation, antiplatelet aggregation and beta-blocker, besides control of cardiovascular risk factors. He remains asymptomatic and without changes in the electrocardiogram.

**Conclusions:** In this case of Kawasaki disease, the initial cardiac involvement consisted of depression of ventricular systolic function, from which he recovered completely. Subsequently, despite adequate and timely treatment, he evolved into the development of giant coronary artery aneurysms that still persist after 1 year of follow-up. The possibility of progression to coronary aneurysms should always be considered and we should maintain regular imaging reevaluation of these patients.

**Keywords:** Kawasaki disease, vasculitis, coronary aneurysm

Giant aneurysm of the common trunk of the left coronary artery.



Coronary angiography showing giant aneurysm of the common trunk of the left coronary artery.

## P-145

**Correlation of cardiopulmonary exercise testing (CPET) parameters, in adults with repaired fallot tetralogy, and cardiac magnetic resonance (CMR).**

Niki Lama<sup>1</sup>, Panagiotis Zachos<sup>2</sup>, Efstratios Karavasilis<sup>1</sup>, Alkistis Kalesi<sup>4</sup>, Maria Karakosta<sup>4</sup>, Georgios Velonakis<sup>1</sup>, Nearchos Kasinos<sup>4</sup>, Antonios Destounis<sup>4</sup>, Sotiria Apostolopoulou<sup>3</sup>, Anastasios Theodosios Georgilas<sup>4</sup>, Nikolaos L. Kelekis<sup>5</sup>

<sup>1</sup>Research Unit of Radiology and Medical Imaging, 2nd Department of Radiology, School of Medicine, National and Kapodistrian University of Athens, Athens, Greece; <sup>2</sup>Department of Health Economics, School of Medicine, National and Kapodistrian University of Athens, Athens, Greece; <sup>3</sup>Pediatric Cardiology and Adult Congenital Heart Disease Department, Onassis Cardiac Surgery Center, Athens, Greece; <sup>4</sup>Echocardiography Training Center, Cardiology Department, General Hospital Tzaneio, Athens, Greece; <sup>5</sup>2nd Department of Radiology, School of Medicine, National and Kapodistrian University of Athens, Athens, Greece

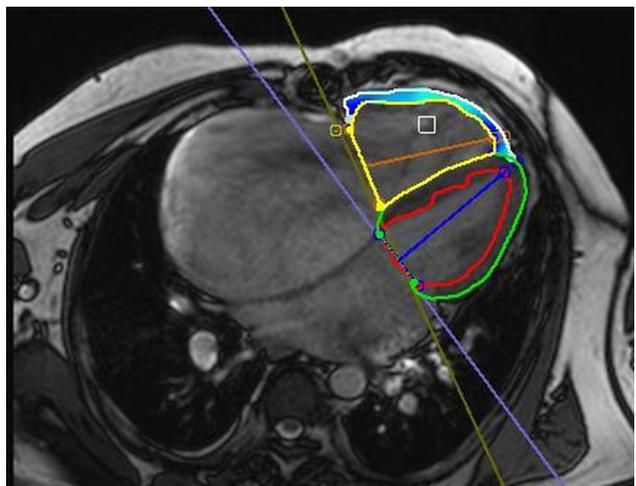
**Background and Aim:** Tetralogy of Fallot is the most common complex congenital heart anomaly. Over the past years, advantages in diagnosis and management of these patients resulted in progressively increase survival rates. The goal of this study is to assess the possible correlation between cardiopulmonary exercise testing and CMR in TOF patients' follow up.

**Method:** CMR is a non-invasive method, able to quantify right and left ventricular function and volumes without radiation exposure and without the need of contrast. CPET is a dynamic test, providing non-invasive assessment of the cardiopulmonary system at rest and during exercise. A simple linear regression analysis was conducted to investigate possible associations between the volumetric ventricular data, (LVEDVi, LVESVi, LVE, RVEDVi, RVESVi) and ventilation metrics (VO<sub>2</sub>max, VE/VCO<sub>2</sub>, AT). All analyses were performed using IBM SPSS v.20 software. **Results:** We studied 12 patients whose mean values were calculated as: LVEDVi = 78.17±18.11 ml/m<sup>2</sup>; LVESVi = 31.33±8.89 ml/m<sup>2</sup>; LVEF = 59.83±6.91 %; RVEDVi = 101.00±29.57ml/m<sup>2</sup>; RVESVi = 50.08±19.44ml/m<sup>2</sup>; RVEF = 51.58±8.2%; VO<sub>2</sub>max = 24.95 ±6.23 (ml/kg/min); VE/VCO<sub>2</sub> = 30.50±5.42; AT = 24.67±13.58. Regression analyses revealed the correlation between LVEDVi and AT [F(1, 10) = 7.992, p = 0.018]. Our study included patients with all degrees of pulmonary regurgitation.

**Conclusions:** AT represents the key moment at which, cardiopulmonary O<sub>2</sub> supply is not sufficient considering the muscles O<sub>2</sub> demand. Our study between different CMR and CPET parameters, revealed a stronger correlation between AT and LVEDVi.

**Keywords:** Tetralogy of Fallot, cardiopulmonary exercise testing, CMR

Assessment of ventricular function and volumes



Cine 4chamber view

## P-147

**Newly high-frame rate blood speckle tracking echocardiography for new insights of aortic flow pattern in children**

Massimiliano Cantinotti, Pietro Marchese, Eliana Franchi, Cecilia Viacava, Nadia Assanta  
Fondazione CNR-Regione Toscana G. Monasterio Massa

**Background and Aim:** High frame rate blood speckle tracking (BST) is a newly echocardiographic technique that allow the study of blood flow and direct visualization of flow patterns into cardiac chambers and vessels walls. The present investigation aims to investigate flow patterns in the aortic root and ascending aorta by BST echocardiography in healthy children.

**Method:** Echocardiographic BST examinations were performed from a focused and zoomed view of the aortic root and the ascending aorta. Flow was classified as laminar, helical, or vortical. Laminar flow was defined as the predominance of particles movement parallel to the major axis of the vessel. Helical flow: particles moving in a spiral producing helical streamlines. Vortical flow: circular or swirling motion

**Results:** From the 100 subjects recruited for the study, 18 were excluded for suboptimal image quality, yielding 82 healthy children (47 males and 35 females) were included for final analysis. Median (IQR) age was 8.2 (5.6-11.0) years, mand median (IQR) body surface area (BSA) was 1.01 (0.79-1.16) m<sup>2</sup>. In all the healthy children the systolic aortic flow hemodynamic was divided into three main phases. Initially, in early systole, immediately after the aortic valve opening, flow pattern in the aortic root was laminar. When the flow reached the tip of the aortic valve cusp in mid-systole, however, the flow split into two helical branches in opposite directions (one to the right ascending aortic wall and one to the left aortic wall) and formed systolic sinus vortices behind the opened valve leaflets. These two branches occupied part of the aortic root and the beginning of the ascending aorta. Finally, in the late systole, the flow returned to be mainly laminar within the ascending aorta. Vortices were visualized in only a single phase of the cardiac cycle in 25 subjects—14 (56.0%) were evident in early diastole and 11 (44.0%) in late systole.

**Conclusions:** Here, we demonstrate the feasibility and reproducibility for the evaluation of aortic flow patterns by BST. We found that flow in the root and ascending aorta was characterized by three main phases and two principal flow patterns: laminar and double helical motions.

**Keywords:** Echocardiography, blood speckle tracking, , aortic flow, children

## P-148

**Organization of pediatric echocardiography laboratories: impact of sonographers on clinical, academic, and financial performance**

Maelys Vener<sup>1</sup>, Nick Arbic<sup>1</sup>, Xavier Iriart<sup>2</sup>, Andreea Dragulescu<sup>1</sup>, Jean Benoit Thambo<sup>2</sup>, Mark K Friedberg<sup>1</sup>, Vitor Guerra<sup>1</sup>, Conall Thomas Morgan<sup>1</sup>, Luc Mertens<sup>1</sup>, Olivier Villemain<sup>1</sup>

<sup>1</sup>Division of Cardiology, Department of Pediatrics, The Hospital for Sick Children, University of Toronto, Toronto, ON, Canada; <sup>2</sup>Department of Pediatric and Adult Congenital Cardiology, Hôpital Cardiologique du Haut-Lévêque, CHU de Bordeaux, Bordeaux, France

**Background and Aim:** Echocardiography has evolved the first-line imaging for diagnosis and management of pediatric and congenital heart disease all over the world. While it recognized as essential component of pediatric cardiac care delivery, organization of pediatric echocardiography services is very heterogeneous across the

world, mainly related to significant differences in material and human resources in heterogeneous health care systems. While in some services sonographers (defined as expert technicians in pediatric echocardiography) are an essential part of the organizational structure, other laboratories operate only with physicians trained in echocardiography.

**Method:** The aims of this work were: 1/ to describe the role of pediatric sonographers and their impact on clinical, academic and financial performance; 2/ to compare two different organizational models (with and without sonographers) with their advantages and disadvantages. Our description was mainly based on two sample hospitals and the authors personal experiences moving between centers. This includes the University Hospital of Bordeaux, Aquitaine, France (physician model) and the Hospital for Sick Children, Toronto, Ontario, Canada (sonographer model).

**Results:** To our knowledge, 11 countries use a sonographer-based structure in pediatric cardiology (including North America, Australia, New Zealand, most countries in Arabian peninsula, Israel, Singapore and a few rare European countries). Sonographer system leads to time-consuming studies but guarantees standardization in data collection, image quality, completeness and storage while physician system is more flexible, providing time-effective studies, reducing intermediaries in patient's journey and allowing reactivity in imaging-based decision-making. On the other hand, the physician system sacrifices medical time and does not guarantee as high a reproducibility and promotion of research as in the sonographer system. Regarding the training of young physicians, sonographer system provides progressive technical teaching from experienced technicians, which remains relatively disconnected from the clinical training, while learning pediatric echocardiography is a daily on-the-job training in the physician system.

**Conclusions:** Different models of care provision are possible and decisions on organizational models need to be adjusted to the demands and available resources.

**Keywords:** pediatric echocardiography, laboratory management, sonographers, pediatric cardiology, congenital heart disease

## P-149

**Left atrial strain by speckle-tracking: incremental role in diastolic assessment of pediatric patients with chronic kidney disease**

Flora Maciel Penachio, Rosana Prado Laurino, Andreia Watanabe, Maria De Fátima Rodrigues Diniz, Karen Saori Shiraishi Sawamura, Alessandro Cavalcanti Lianza, Carolina Rocha Brito Menezes, Isabela Sousa Lobo Silva, Gabriela Nunes Leal  
Instituto da Criança e do Adolescente do Hospital das Clínicas da Universidade de São Paulo

**Background and Aim:** Cardiovascular complications are the leading mortality cause among children with chronic kidney diseases (CKD). Left ventricle (LV) diastolic dysfunction predicts poor cardiovascular outcomes. Diastolic function evaluation is usually limited to spectral and tissue Doppler imaging, known to be less reliable in pediatrics. We aimed to investigate left atrial (LA) strain in diastolic assessment of pediatric CKD.

**Method:** From February 2019 to July 2022, 55 pediatric CKD patients without cardiovascular symptoms and 55 controls were evaluated.

**Results:** Patients and controls had similar age [9.78 (0.89 – 17.54) vs. 10.72 (1.03 – 18, 44) years; p = 0.41] and gender (36M:19F vs. 34M:21F; p = 0.84). LV EF was preserved ( $\geq 55\%$ ) in all patients. E/e' was higher in CKD [6.99 (4.75 – 14.20) vs. 6.38

(3.88 – 11.11);  $p = 0.009$ ], although it was above normal limits in only one individual. Comparing CKD and controls, LA reservoir strain was lower ( $48.22 \pm 10.62\%$  vs.  $58.52 \pm 10.70\%$ ;  $p < 0.0001$ ) and LA stiffness index was higher [ $0.14 (0.08 - 0.48) \%$ -1 vs.  $0.11 (0.06 - 0.23) \%$ -1;  $p < 0.0001$ ]. LV hypertrophy was associated with lower LA reservoir strain ( $42.05 \pm 8.74\%$  vs.  $52.99 \pm 9.52\%$ ;  $p < 0.0001$ ), higher LA stiffness index [ $0.23 (0.11 - 0.48) \%$ -1 vs.  $0.13 (0.08 - 0.23) \%$ -1;  $p < 0.0001$ ] and filling index ( $2.39 \pm 0.63 \text{ cm/s} \times \%$ -1 vs.  $1.74 \pm 0.47 \text{ cm/s} \times \%$ -1;  $p = 0.0001$ ). Uncontrolled hypertension was associated with lower LA reservoir strain ( $41.9 \pm 10.6\%$  vs.  $35.6 \pm 9.7$ ;  $p = 0.005$ ). CKD stage showed negative correlation with LA reservoir strain ( $r = -0.37$ ;  $p = 0.006$ ) and positive correlation with LA stiffness index ( $r = 0.48$ ;  $p = 0.0002$ ). E/e' showed inferior accuracy in differentiating CKD patients from controls (AUC = 0.64), when compared with LA reservoir strain (AUC = 0.75) and LA stiffness index (AUC = 0.73). LA reservoir strain showed the best accuracy in differentiating dialysis from non-dialysis patients (AUC = 0.77). **Conclusions:** LA strain showed better accuracy than conventional parameters, concerning diastolic evaluation in pediatric CKD. Our findings favor LA strain incorporation to routine evaluation in this group.

**Keywords:** pediatric chronic kidney disease, speckle-tracking echocardiography, left atrial strain

#### P-150

##### **Real-time remote-mentored echocardiography in management of neonates with critical congenital heart defects**

Håvard Solvin<sup>1</sup>, Simone Diab<sup>4</sup>, Henrik Holmström<sup>4</sup>, Ole Jakob Elle<sup>3</sup>, Henrik Brun<sup>2</sup>

<sup>1</sup>Institute of Clinical Medicine, University of Oslo. Kirkeveien 166, 0450 Oslo, Oslo Norway; <sup>2</sup>The Intervention Centre, Technology and Innovation Clinic, Oslo University Hospital. Sognsvannsveien 20, 0372 Oslo, Oslo Norway; <sup>3</sup>Department of Informatics, University of Oslo. Gaustadalleen 23 b, 0373 Oslo, Oslo Norway; <sup>4</sup>Department of Paediatric Cardiology, Oslo University Hospital. Sognsvannsveien 20, 0372 Oslo, Oslo Norway

**Background and Aim:** Around one third of neonates with critical congenital heart defects (CCHD) in Norway are born without a prenatal diagnosis and a plan for postnatal care. Management of suspected CCHD relies on early echocardiographic diagnosis and availability of competent echocardiographers is limited in local hospitals. The present study evaluates the use of remote-mentoring echocardiography to identify neonates with CCHD in need of immediate transfer to a pediatric cardiac surgery center.

**Method:** The setup included a pediatric cardiologist in another room, guiding an examining physician with little experience in echocardiographic imaging of congenital heart defects (CHD). The examining physician was in most cases an anesthesiologist or a resident doctor of adult cardiology. This remote-mentoring pair was blinded to the condition of the neonate and presented with a simplified patient history. The echocardiographic images were streamed to the laptop of the mentor, along with a webcam feed showing the position of the probe. Voice communication took place by cellphone. The task was to decide whether the neonate had a CCHD in need of immediate transfer to a pediatric cardiac surgery center if the neonate was at a local hospital. The result was compared to the previously completed echocardiographic report and clinical decision of the patient-responsible pediatric cardiologist. The neonates and their parents were recruited from, and the remote-mentored echocardiography took place in a neonatal intensive care unit at a tertiary healthcare center.

**Results:** Fifteen neonates were recruited. All six neonates with CCHD who needed immediate transfer were correctly labeled by the remote-guidance pair. One neonate with Tetralogy of Fallot (TOF) was erroneously labeled as needing immediate transfer. Eight neonates without need for immediate transfer were correctly labeled. All parents answered they would accept remote mentoring if their child needed echocardiography at a local hospital.

**Conclusions:** In the present study, remote-mentored echocardiography performed by echocardiographers with little experience in imaging of CHD identified all neonates with CCHD in need of immediate transfer for specialist care. One unnecessary transport of a neonate with stable TOF was proposed. Remote-mentored echocardiography shows promising results for improving neonatal diagnostics of CCHD in local hospitals.

**Keywords:** remote, guidance, mentoring, echocardiography, neonate, congenital heart defect

#### P-151

##### **Non-contrast 4-dimensional flow angiography – accuracy of vessel areas**

Barbara Elisabeth Ursula Burkhardt<sup>1</sup>, Anke Meinhold<sup>2</sup>, Julia Geiger<sup>3</sup>, Emanuela Regina Valsangiacomo Büchel<sup>1</sup>, Christian Johannes Kellenberger<sup>3</sup>, Fraser Maurice Callaghan<sup>3</sup>

<sup>1</sup>Pediatric Cardiology, Pediatric Heart Center, Department of Surgery, University Children's Hospital Zurich, Switzerland; Children's Research Center, University Children's Hospital Zurich, Switzerland;

<sup>2</sup>Department of Intensive Care and Neonatology, University Children's Hospital Zurich, Switzerland; Children's Research Center, University Children's Hospital Zurich, Switzerland; <sup>3</sup>Department of Diagnostic Imaging, University Children's Hospital Zurich, Switzerland; Children's Research Center, University Children's Hospital Zurich, Switzerland

**Background and Aim:** 4-dimensional (4D) cardiovascular magnetic resonance (CMR) flow acquisitions are usually performed after injection of contrast agents and cover a similar volume as contrast enhanced MR angiography. There are, however, regulatory restrictions for the use of contrast. We aimed to test if 4D flow derived images are suitable for measuring vessel dimensions, applicable for cases when contrast injection is not possible.

**Method:** Whole heart 4D flow and 2-dimensional (2D) phase contrast sequences were acquired on a 1.5 Tesla GE scanner. 2D planes at the same location and orientation as the 2D data for the ascending aorta (AAO) and the main (MPA), right (RPA), and left (LPA) pulmonary arteries were extracted from 4D flow at all cardiac phases, and vessel boundaries were marked by two readers, one with repeated measurements. Vessel cross sectional areas (CSA) were compared between 4D flow and the systolic phase of 2D flow modulus images using Wilcoxon test and relative differences. Inter- and intraobserver agreement was assessed with Bland-Altman analysis.

**Results:** Images of 16 consecutive patients undergoing CMR without contrast agents (age  $19.4 \pm 14.2$  years) were analyzed. 4D flow in-plane resolution was  $1.4 \pm 0.03 \text{ mm}$ , slice thickness  $2.1 \pm 0.23 \text{ mm}$ ; 2D in-plane resolution was  $1.2 \pm 0.2 \text{ mm}$ , slice thickness:  $4.1 \pm 0.7 \text{ mm}$ .

4D flow derived CSA were (median (interquartile range)) 538 (378-616) mm<sup>2</sup> for AAO, 542 (441-863) mm<sup>2</sup> for MPA, 205 (174-253) mm<sup>2</sup> for RPA, and 219 (191-283) mm<sup>2</sup> for LPA.

Measurements were significantly greater on 4D than 2D (AAO  $p = 0.001$ ; MPA  $p = 0.01$ ; RPA  $p = 0.003$ ; LPA  $p = 0.011$ ). Relative differences between 4D and 2D flow were 12 (4-18)% for AAO, 6 (0-24)% for MPA, 14 (1-20)% for RPA, and 27 (0-52)% for LPA.

Interobserver bias (limits of agreement; LOA) (mm<sup>2</sup>) were 41.4 (-26.1 – 108.8) for AAO, 43.4 (-113.6 – 200.5) for MPA, 35.1 (1.7 – 68.5) for RPA, and 29.4 (-96.9 – 155.6) for LPA.

Intraobserver differences (LOA) (mm<sup>2</sup>) were -8.9 (-53.8 – 36) for AAO, 3.7 (-68.8 – 76.1) for MPA, 8.7 (-42.4 – 59.9) for RPA, and -2.4 (-79 – 74.2) for LPA.

**Conclusions:** Vascular areas are greater on 4D compared to 2D flow images. Non-contrast 4D flow reconstructed images are insufficient for vessel dimension quantification.

**Keywords:** cardiac magnetic resonance, flow imaging, angiography, non-contrast imaging, great vessels

## P-152

### When the valve stops clicking

Precyia Philo Fernandes, Christie Brockway, Nikki Walker, Andrew Mclean, Lorna Swan, Hamish Walker  
Scottish Adult Congenital Heart Services, Golden Jubilee National Hospital Glasgow, Scotland UK

**Background and Aim:** 19year ♂ diagnosed at birth with absent left sided atrio-ventricular connection, congenitally corrected transposed great arteries, dominant morphological left ventricle, hypoplastic morphological right ventricle, small superior atrial septal defect and a restrictive muscular outlet ventricular septal defect. He initially underwent atrial septectomy and pulmonary artery banding at 3months of age. The outflow tract obstruction introduced by the VSD was treated by a Damus-Kaye-Stansel procedure combined with a bidirectional right superior cavopulmonary anastomosis at 2 years. He subsequently had total cavopulmonary connection using 18mm, fenestrated extracardiac conduit at 5years. He developed progressive neo-aortic valve regurgitation and underwent a mechanical valve replacement (MVR) at 14years. The mechanical valve was not well seen at follow-up echocardiograms. The patient reported he could no longer hear the valve click 5 months postsurgery.

**Method:** Cardiac MRI was performed

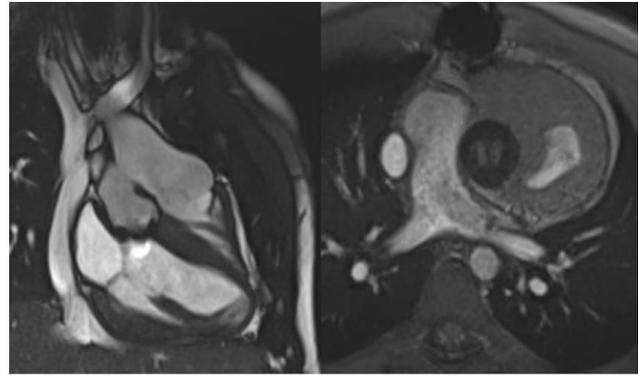
**Results:** Cardiac MRI demonstrated unobstructed cavopulmonary pathways and Damus-Kaye-Stansel anastomosis. Concentric dominant left ventricular hypertrophy was apparent with function. The mechanical neo-aortic valve prosthesis was fixed in closed position with no anterograde flow. A restrictive muscular VSD represented the only outflow from the dominant LV with peak velocity upto 4m/s. Following discussion at ACHD joint conference, the patient was offered redo neo-aortic MVR with tissue valve and is current awaiting surgery.

**Conclusions:** Good valve function is an important factor in the outcome of patients with single ventricle physiology. Valve replacement in this group is uncommon with limited data on long term outcomes. Atrio-ventricular valve replacement is known to have worse outcomes in comparison to semilunar valve replacement. Undertaking a redo replacement of semilunar valve imposes significant higher risk but was felt to be indicated in this patient. The aetiology of mechanical valve failure in this patient was thought to reflect the dual outflow, with a reduction in flow across the prosthetic valve. Redo valve replacement with a tissue valve was considered to be the best option to avoid further valve thrombosis.

This case emphasises the importance of routine cardiac MRI in the assessment of congenital heart disease.

Although this patient was clinically well, a major surgical complication had remained unnoticed due to the assumption that incomplete echo imaging was sufficient.

**Keywords:** Mechanical valve, Fontan, TCPC



Neo-Aortic valve prosthesis fixed in closed position

## P-153

### Echocardiographic indices of LV dysfunction in duchenne muscular dystrophy

Jaroslav Meyer Szary<sup>1</sup>, Magdalena Bazgier<sup>1</sup>, Karolina Śledzińska<sup>2</sup>, Jolanta Wierzba<sup>2</sup>, Joanna Kwiatkowska<sup>1</sup>  
<sup>1</sup>Department of Paediatric Cardiology and Congenital Heart Defects, Medical University of Gdansk, Gdansk, Poland; <sup>2</sup>Department of Internal and Pediatric Nursing, Faculty of Health Sciences with Institute of Maritime and Tropical Medicine, Medical University of Gdansk, Gdansk, Poland

**Background and Aim:** Duchenne muscular dystrophy (DMD) is a rare genetic disease causing dystrophin deficiency and muscle dysfunction resulting in loss of ambulation, respiratory failure and cardiomyopathy. Commonly the onset of cardiomyopathy and heart failure is observed in early adulthood and less commonly already in teenagers. Annual echocardiography is recommended since the diagnosis but its role as well as prognostic value is unknown. The aim was to characterize echocardiographic findings in the childhood period.

**Method:** Standard echocardiographic protocol using B-mode imaging and M-mode (Teichholtz) as well as Doppler was used to acquire measurements. Since it was shown that most echocardiographic indices best correlate with BSA, BSA matched controls were selected from healthy peers. Statistical calculations were done using Wizard 2.0.10 using parametric and non-parametric tests according to data distribution. Multiple studies in the same patient were considered separate data points. Data are given as mean ±SD (p-value).

**Results:** Ninety-three DMD patients having 324 echocardiographic studies and 278 controls with 278 studies were included. Patients were 9.7 ±4.1 and controls 9.4 ±4.2 years old, p = 0.030 and BSA 1.13 ±0.36 vs 0.95 ±0.24 (p = 0.884), Height was 132.1 ±21.8 vs 125.6 ±18.6 (p = 0.076) as expected by matching. DMD patients were found to be of higher Weight 36.0 ±17.5 vs 26.1 ±9.3 (p<0.001) and BMI 19.4 ±4.5 vs 16.0 ±2.2 (p<0.001). IVSd 7.0 ±1.4 vs 6.4 ±1.3 (p = 0.825), IVSs 10.0 ±1.7 vs 9.4 ±1.7 (p = 0.166), LVDd 40.6 ±5.3 vs 37.7 ±4.7 (p = 0.165), LVDs 26.9 ±4.6 vs 23.4 ±3.6 (p<0.001), LVPWd 6.3 ±1.2 vs 6.0 ±1.0 (p = 0.219), LVPWs 9.7 ±1.8 vs 9.7 ±1.9 (p<0.001), EF% 63.2 ±6.0 vs 67.7 ±8.3 (p<0.001), SF% 34.0 ±4.2 vs 37.9 ±6.2 (p<0.001). LVDs was found to be significantly higher resulting in significantly lower left ventricular function assessed by EF and SF. Interestingly LVPWd was thicker in DMD patients.

**Conclusions:** DMD is characterized by LV systolic dysfunction (higher systolic LV diameter) and mild thickening of the posterior

wall but no evidence of overt hypertrophic or dilatative cardiomyopathy. There is marked patient-to-patient variability in the observed indicates. Longitudinal observational studies accessing late clinical outcomes are necessary to understand prognostic value of this echocardiographic indices.

**Keywords:** Duchenne muscular dystrophy, cardiomyopathy, heart failure, echocardiography

#### P-154

##### Differences between myocardial deformation response to preload and exercise stress in the fontan circulation

Simone Goa Diab<sup>1</sup>, Assami Rösner<sup>2</sup>, Guro Grindheim<sup>3</sup>, Gaute Døhlen<sup>4</sup>, Henrik Brun<sup>4</sup>, Kanyalak Vithessonthi<sup>5</sup>, Thomas Möller<sup>4</sup>  
<sup>1</sup>Department of Paediatric Cardiology, Oslo University Hospital - Rikshospitalet, Oslo, Norway; <sup>2</sup>Institute of Clinical Medicine, Faculty of Medicine, University of Oslo, Oslo, Norway; <sup>3</sup>Department of Cardiology, University Hospital of North Norway, Tromsø, Norway; <sup>4</sup>Department of Clinical Medicine, UiT, The Arctic University of Norway, Tromsø, Norway; <sup>5</sup>Division of Emergencies and Critical Care, Oslo University Hospital - Rikshospitalet, Oslo, Norway; <sup>4</sup>Department of Paediatric Cardiology, Oslo University Hospital - Rikshospitalet, Oslo, Norway; <sup>5</sup>Department of Paediatric Cardiology, Oslo University Hospital - Rikshospitalet, Oslo, Norway; King Chulalongkorn Memorial Hospital, Bangkok, Thailand

**Background and Aim:** Limitation of cardiac output reserve in the Fontan circulation is well documented, but the mechanisms are not fully understood. We aimed to explore the limiting role of myocardial function in the single ventricle by investigating echocardiographic deformation measurements in response to both exercise and experimental preload increase in adolescent patients with Fontan circulation.

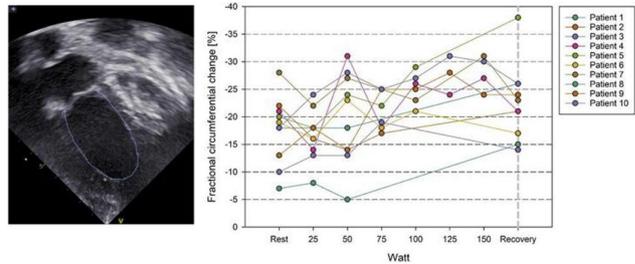
**Method:** We included 25 patients (median age 16.6 years, range 16.1–17.7 years, 8 females) during pre-transition diagnostic work-up. For 10/25 patients echocardiographic imaging was performed during exercise using a recumbent cycle ergometer with a 25W load increment every 3 minutes. We measured fractional circumferential change (FCC) in an apical view at rest, during exercise and at 1–3 minutes of recovery. For 20/25 patients echocardiographic imaging was also performed during heart catheterization, both at baseline, during a rapid 5 ml/kg body weight saline infusion and repeatedly until 6 minutes post infusion. We measured myocardial peak global longitudinal strain (GLS) in an apical view before saline infusion (baseline), at 1.00–2.00 minutes after saline infusion and at 6.00 minutes (steady state).

**Results:** During stress echocardiography, mean FCC was  $-17.6 (\pm 6.1)$  at baseline,  $-22.1 (\pm 7.9)$  at maximal load and  $-22.5 (\pm 6.9)$  at recovery. In half of the patients FCC declined (less negative) at the first workload of 25W but 8/10 patients had an increase (more negative) in FCC at maximal load (Figure 1). During heart catheterization, mean GLS was  $-17.4\% (\pm 4.5\%)$  at baseline,  $-18.0\% (\pm 3.8\%)$  after saline infusion, and  $-17.2\% (\pm 4.1\%)$  at steady state. In more than half of the patients, there was no change or even a decline (less negative) in GLS after saline infusion.

**Conclusions:** For the majority of patients, myocardial deformation imaging during exercise demonstrated an improvement in systolic function at maximum load. However, in half of the examined patients there was an initial decline in systolic function. Further, in half of the patients exposed to an invasive volume load, a persistent lack of improvement or even worsening of systolic function was seen. Whether these subsets of patients are overlapping and have a higher risk of a failing Fontan circulations is to be investigated.

**Keywords:** Fontan, echocardiography, strain, cardiac catheterization,

Fractional circumferential change during exercise stress



#### P-155

##### Congenital fistulas with particular origins from the right aortic CUSP and right atrial appendage to the pulmonary artery

Gabriela Doros<sup>1</sup>, Anca Popoiu<sup>1</sup>, Ana Maria Miinea<sup>2</sup>, Cristiana Stolojanu<sup>1</sup>, Andreea Vartop<sup>2</sup>, Andrada Mara Miclescu<sup>1</sup>

<sup>1</sup>Department of Pediatrics, “Victor Babes” University of Medicine and Pharmacy, Timisoara, Romania; <sup>2</sup>Department of Pediatric Cardiology, “Louis Turcanu” Emergency Hospital for Children, Timisoara, Romania; <sup>2</sup>Department of Pediatric Cardiology, “Louis Turcanu” Emergency Hospital for Children, Timisoara, Romania

**Background and Aim:** Pulmonary artery fistulas consist of abnormal vascular communications between the coronary arteries and pulmonary artery. It was considered to be a rare disorder, involving one or rarely, multiple coronary arteries. Our aim is to present two fistulas in the pulmonary artery trunk, with a particular origin. **Method:** A 13 years old girl was admitted into our clinic for cardiac evaluation due to a low-grade cardiac murmur in the upper left sternal border, irradiating to the upper right sternal border. She underwent clinical examination, ECG and echocardiography, followed by a 256-slice multidetector CT scanner (MDCT).

**Results:** The patient is clinically asymptomatic. The ECG was normal. Echocardiography revealed a subaortic membrane which did not produce outflow tract gradient and two fistulas in the pulmonary artery trunk. The fistulas were considered to be coronary artery fistulas. A 256 x 0.6-multislice MDCT scan coronarography, ECG modulated and synchronized, was performed, under betablockers in order to obtain a heart rate of 64 b/min, using a Cardiac Fast FLASH Protocol in diastole. The emergence of coronary arteries was normal. The coronary artery system was with right dominance. The coronary arteries were normal, with no stenotic lesions. However, we remarked two particular fistulas: one between the right atrial appendage and the pulmonary artery trunk, with a 0.39 cm diameter and visible over a 1.38 cm distance; a second fistula was seen between the right coronary artery cusp and the pulmonary artery trunk, with a 0.23 cm diameter. 3D reconstruction allows us to see the exact trajectory of the fistulas.

**Conclusions:** Noninvasive Computer Tomography Coronarography with contrast, using a 256 Multidetector CT scanner is the gold standard diagnostic tool for detecting the origin of pulmonary artery fistulas. Our patient presented two congenital fistulas with particular origins, one from right aortic cusp and the other from the right atrial appendage, both opening in the

pulmonary artery, instead of the classic coronary artery fistulas. Alongside the fistulas, our patient also presented a subaortic non-obstructive membrane. This performant exploration, together with the 3D reconstruction allow physicians to make a decision for further management.

**Keywords:** right aortic cusp, right atrial appendage, pulmonary artery fistulas

#### P-156

##### The prognostic value of the right atrial area in patients with repaired tetralogy of fallot

Alkistis Eleni Kalesi<sup>1</sup>, Maria Karakosta<sup>1</sup>, Despoina Ntiloudi<sup>1</sup>, Dimitrios Boukis<sup>1</sup>, Nikolaos Miaris<sup>1</sup>, Antonios Destounis<sup>1</sup>, Nearchos Kasinos<sup>1</sup>, Anastasios Theodosis Georgilas<sup>1</sup>, Sotiria Apostolopoulou<sup>2</sup>, George Vagenakis<sup>2</sup>, Stefanos Despotopoulos<sup>2</sup>, Niki Lama<sup>3</sup>, Dimitrios Barmpagiannis<sup>1</sup>, Evangelos Pismimis<sup>1</sup>, Panagiotis Zachos<sup>4</sup>  
<sup>1</sup>Echocardiography Training Center, Cardiology Department, "Tzanio" General Hospital, Piraeus, Greece; <sup>2</sup>Pediatric Cardiology and Adult Congenital Heart Disease Department, Onassis Cardiac Surgery Center, Athens, Greece; <sup>3</sup>Research Unit of Radiology and Medical Imaging, 2nd Radiology Department, School of Medicine, National and Kapodistrian University of Athens, Athens, Greece; <sup>4</sup>Pediatric Cardiology and Adult Congenital Heart Disease Department, Karditsa General Hospital, Karditsa, Greece; Department of Health Economics, School of Medicine, National and Kapodistrian University of Athens, Athens, Greece

**Background and Aim:** Cardiac Magnetic Resonance (CMR) is considered to be the gold standard imaging modality for the comprehensive assessment and risk stratification of patients with repaired Tetralogy of Fallot (rToF). Nevertheless, it is underutilized in the everyday assessment of patients with rToF, due to several limitations and a relatively high cost compared with transthoracic echocardiography (TTE). Thus, finding new prognostic echocardiographic indices is essential for better evaluating these patients. The present study aims to identify a possible prognostic value of the right atrial area (RA area) in the assessment of patients with rToF.

**Method:** The study population consisted of adults, with a history of rToF, who were followed up at the Echocardiography Department "D. Beldekos" of "Tzanio" Hospital of Piraeus between January 2019 and January 2022. All patients underwent TTE and cardiopulmonary exercise test (CPET). The RA area was measured by TTE, along with other parameters. Spearman's correlation was used for finding a possible correlation between the RA area and the variables measured during CPET since the variables didn't follow a normal distribution. The statistical significance level was set to 0.05 ( $p < 0.05$ ).

**Results:** A sample of 14 subjects with rToF (10 males, 4 females), of a mean age of 36.5 years (range 18 - 63 years), has been examined. A TTE was performed and the mean RA area was 18.8cm<sup>2</sup> (range 9cm<sup>2</sup>-32cm<sup>2</sup>). A CPET was also performed, revealing a mean value of maximum oxygen consumption (VO<sub>2</sub>max) of 24.1ml/kg/min (minimum = 12ml/kg/min, maximum = 36ml/kg/min), a mean value of minute ventilation/ carbon dioxide production slope (VE/VCO<sub>2</sub>) of 31 (minimum = 20, maximum = 42) and a mean value of anaerobic threshold (AT) of 22.7 (minimum = 12, maximum = 39). A negative correlation between RA area and VO<sub>2</sub>max values was found (Spearman's rho = 0.609,  $p = 0.035$ )

**Conclusions:** RA area measured by TTE seems to be a good predictor of a patient's exercise capacity. Higher values of RA area were associated with a lower exercise capacity.

**Keywords:** RA area, repaired Tetralogy of Fallot, CPET, rToF, Exercise Capacity

#### P-157

##### The prognostic value of right ventricular ejection fraction, measured by CMR, in the assessment of patients with repaired tetralogy of fallot

Alkistis Eleni Kalesi<sup>1</sup>, Maria Karakosta<sup>1</sup>, Nikolaos Miaris<sup>1</sup>, Maria Vassaki<sup>1</sup>, Dimitra Maritsa<sup>1</sup>, Dimitris Boukis<sup>1</sup>, Despoina Ntiloudi<sup>1</sup>, Nearchos Kasinos<sup>1</sup>, Antonios Destounis<sup>1</sup>, Anastasios Theodosis Georgilas<sup>1</sup>, George Vagenakis<sup>2</sup>, Niki Lama<sup>3</sup>, Sotiria Apostolopoulou<sup>2</sup>, Stefanos Despotopoulos<sup>2</sup>, Sotiris Mitsiadis<sup>1</sup>, Panagiotis Zachos<sup>4</sup>  
<sup>1</sup>Echocardiography Training Center, Cardiology Department, "Tzanio" General Hospital, Piraeus, Greece; <sup>2</sup>Pediatric Cardiology and Adult Congenital Heart Disease Department, Onassis Cardiac Surgery Center, Athens, Greece; <sup>3</sup>Research Unit of Radiology and Medical Imaging, 2nd Radiology Department, School of Medicine, National and Kapodistrian University of Athens, Athens, Greece; <sup>4</sup>Pediatric Cardiology and Adult Congenital Heart Disease Department, Karditsa General Hospital, Karditsa, Greece; Department of Health Economics, School of Medicine, National and Kapodistrian University of Athens, Athens, Greece

**Background and Aim:** Risk stratification in patients with repaired Tetralogy of Fallot (rToF) is often difficult due to the disease's complexity. Thus, finding new prognostic indices is essential for a more comprehensive approach to these patients. The present study aims to find if there is any correlation between the right ventricular ejection fraction (RVEF), measured by cardiac magnetic resonance (CMR), and the parameters measured during the cardiopulmonary exercise test (CPET) in patients with rToF.

**Method:** The study population consisted of adults, with a history of rToF, who were followed up at the outpatient clinic of the "Congenital Heart Diseases Department" of Tzanio Hospital of Piraeus between January 2019 and June 2021. All patients underwent CPET and CMR, and the RVEF was measured, along with other parameters. Spearman's correlation was used for finding a possible correlation between the variables since they didn't follow a normal distribution. The statistical significance level was set to 0.05 ( $p < 0.05$ ).

**Results:** A sample of 14 subjects with rToF (10 males, 4 females), of a mean age of 36.5 years (minimum = 18 and maximum = 63), has been examined. The mean right ventricular (RV) end-diastolic volume was 93.5ml/m<sup>2</sup> (minimum = 65ml/m<sup>2</sup>, maximum = 180 ml/m<sup>2</sup>), the mean RV end-systolic volume was 43.5ml/m<sup>2</sup> (minimum = 23ml/m<sup>2</sup>, maximum = 100ml/m<sup>2</sup>) and the mean RVEF was 48% (minimum = 45%, maximum = 68%). A CPET was also performed, revealing a mean value of maximum oxygen consumption (VO<sub>2</sub>max) of 24.1ml/kg/min (minimum = 12ml/kg/min, maximum = 36ml/kg/min), a mean value of minute ventilation/ carbon dioxide production slope (VE/VCO<sub>2</sub>) of 31 (minimum = 20, maximum = 42) and a mean value of anaerobic threshold (AT) of 22.7 (minimum = 12, maximum = 39). A positive correlation between RVEF and VO<sub>2</sub>max values was found (Spearman's rho 0.580,  $p = 0.048$ )

**Conclusions:** RVEF measured by CMR seems to be a good predictor of a patient's exercise capacity. Higher values of RVEF are associated with better exercise capacity in patients with rToF.

**Keywords:** Tetralogy of Fallot, ToF, RVEF, CPET, CMR, Exercise Capacity

## P-158

**Parameters of ventilation/perfusion matching at cardiopulmonary exercise test might detect asymmetric lung perfusion in fontan patients**

Alessia Callegari<sup>1</sup>, Jakob Usemann<sup>3</sup>, Alexander Moeller<sup>3</sup>, Christian Kellenberger<sup>2</sup>, Fraser Callaghan<sup>2</sup>, Julia Geiger<sup>2</sup>, Barbara Burkhardt<sup>1</sup>, Emanuela Valsangiacomo Büchel<sup>1</sup>

<sup>1</sup>University Children's Hospital Zurich, Pediatric Heart Centre – Division of Pediatric Cardiology, University of Zurich; Switzerland; <sup>2</sup>University Children's Hospital Zurich, Department of Diagnostic Imaging, University of Zurich; Switzerland; <sup>3</sup>University Children's Hospital Zurich, Division of Respiratory Medicine, University of Zurich; Switzerland

**Background and Aim:** Asymmetric distribution of the caval flow into the lung is a potential complication in Fontan patients and may lead to development of intrapulmonary shunts, aortopulmonary collaterals and elevated pulmonary vascular resistance. Echocardiographic assessment of pulmonary flow distribution is challenging and imprecise. Thus, additional non-invasive tests that would help clinicians to identify patients at risk are needed. We sought to explore the parameters of ventilation/perfusion matching at cardiopulmonary exercise (CPET) in patients with and without an asymmetric lung perfusion.

**Method:** Cardiovascular magnetic resonance (CMR), bodyplethysmography, spirometry, and CPET were prospectively performed in 36 single ventricle patients 11 years (8–15) after the Fontan operation. Differential lung perfusion was quantified using the 4D flow sequence. Lung perfusion was defined as asymmetric if the RPA/LPA flow difference was > 60%/40%. Ventilatory equivalents for O<sub>2</sub> (ventilatory efficiency; Eq VE/VO<sub>2</sub> peak) and of CO<sub>2</sub> (ventilatory drive; Eq VE/VCO<sub>2</sub> peak), and minimal partial end-tidal CO<sub>2</sub> tension (PEtCO<sub>2</sub> min) were used for assessment of gas exchange.

**Results:** The overall pulmonary net flow was RPA 25±11 (range 9–47) ml/beat and LPA 19±7 (range 5–31) ml/beat. Thirteen (27%) patients had an asymmetric lung perfusion. LPA stents were present in 17 (35%) patients. In patients with asymmetric lung perfusion we found higher respiratory equivalents, being Eq VE/VCO<sub>2</sub>peak 42.4±7.5 vs. 36.0±4.7 (p = 0.012) and Eq VE/VO<sub>2</sub>peak 44.6±9.4 vs. 37.4±6.0 (p = 0.015), but lower PEtCO<sub>2</sub> 33.7±4.2 vs. 38.6±4.7 mmHg (p = 0.012). The other cardiovascular and respiratory response parameters at CPET as well as lung volumes at spirometry and bodyplethysmography were similar in both groups.

**Conclusions:** In Fontan patients an asymmetric pulmonary blood flow is related to abnormal indicators of gas exchange and may express the presence of a ventilation/perfusion mismatch. The ventilation/perfusion parameters at CPET might be used as an additional non-invasive indicator for an asymmetric lung perfusion and indicate the need for further advanced imaging and subsequent intervention.

**Keywords:** Fontan, CPET, CMR

## P-159

**The prognostic value of myocardial work in the assessment of patients with repaired tetralogy of fallot**

Alkistis Eleni Kalesi<sup>1</sup>, Maria Karakosta<sup>1</sup>, Nearchos Kasinos<sup>1</sup>, Antonios Destounis<sup>1</sup>, Anastasios Theodosis Georgilas<sup>1</sup>, Despoina Ntiloudi<sup>1</sup>, Nikolaos Miaris<sup>1</sup>, Evangelia Grigoraki<sup>1</sup>, Sotiria Apostolopoulou<sup>2</sup>, Stefanos Despotopoulos<sup>2</sup>, Niki Lama<sup>3</sup>, George Vagenakis<sup>2</sup>, Sarantos Linardakis<sup>1</sup>, Dimitrios Bampagiannis<sup>1</sup>, Dimitra Maritsa<sup>1</sup>, Panagiotis Zachos<sup>4</sup>

<sup>1</sup>Echocardiography Training Center, Cardiology Department, "Tzanio" General Hospital, Piraeus, Greece; <sup>2</sup>Pediatric Cardiology and Adult Congenital Heart Disease Department, Onassis Cardiac Surgery Center, Athens, Greece; <sup>3</sup>Research Unit of Radiology and Medical Imaging, 2nd Radiology Department, School of Medicine, National and Kapodistrian University of Athens, Athens, Greece; <sup>4</sup>Pediatric Cardiology and Adult Congenital Heart Disease Department, Karditsa General Hospital, Karditsa, Greece; Second Department of Health Economics, School of Medicine, National and Kapodistrian University of Athens, Athens, Greece

**Background and Aim:** The evaluation of the left ventricle (LV) is often difficult in patients with repaired Tetralogy of Fallot (rToF), due to the disease's complexity and the ventricular interdependence. The prognostic value of left ventricular volumes and ejection fraction (LVEF), measured by Cardiac Magnetic Resonance (CMR), has been well established in previous studies. The present study aims to identify a possible prognostic value of myocardial work in the evaluation of patients with rToF.

**Method:** The study population consisted of adults with a history of rToF, followed up at the Echocardiography Department "D. Beldekos" of Tzanio Hospital of Piraeus between January 2019 and January 2021. Transthoracic echocardiogram (TTE), CMR and Cardiopulmonary Test (CPET) were performed in all patients. Measurements included the calculation of several myocardial work indices, such as Global Work Index (GWI), Global Constructive Work (GCW), Global Wasted Work (GWW) and Global Work Efficiency (GWE), by TTE and also LVEF by CMR. Spearman's correlation was used for finding a possible correlation between the variables since they didn't follow a normal distribution. The statistical significance level was set to 0.05 (p < 0.05).

**Results:** A sample of 14 subjects with rToF (10 males, 4 females), with a mean age of 36.5 years was examined. The mean values of myocardial work indices were GWI 1648mmHg%, GCW 1826.5mmHg%, GWW 99.0mmHg%, and GWE 93.5%. The mean left ventricular end-diastolic volume (LVEDV) by CMR was 75ml/m<sup>2</sup>, the mean end-systolic volume (LVESV) was 31ml/m<sup>2</sup> and the mean LVEF was 59%. The CPET revealed a mean value of maximum oxygen consumption (VO<sub>2</sub>max) of 24.1ml/kg/min, a mean value of minute ventilation/ carbon dioxide production slope (VE/VCO<sub>2</sub>) of 31, and a mean value of anaerobic threshold (AT) of 22.7. A positive correlation between GCW and VO<sub>2</sub>max values (Spearman's rho 0.721, p = 0.019) and a negative correlation between GWI and LVESV by CMR (Spearman's rho -0.724, p = 0.012) were found.

**Conclusions:** GCW measured by TTE seems to be a good predictor of a patient's exercise capacity. Higher values of GCW were associated with better exercise capacity, whereas higher values of GWI were associated with lower values of LV end-systolic volume.

**Keywords:** Myocardial Work, GWE, Tetralogy of Fallot, CPET, CMR, Exercise Capacity

## P-160

**Comparison between 3D echocardiography and CMR imaging in the assessment of right ventricular volumes in patients with repaired tetralogy of fallot**

Maria Karakosta<sup>1</sup>, Alkistis Eleni Kalesi<sup>1</sup>, Despoina Ntiloudi<sup>1</sup>, Antonios Destounis<sup>1</sup>, Nearchos Kasinos<sup>1</sup>, Anastasios Theodosis Georgilas<sup>1</sup>, Nikolaos Miaris<sup>1</sup>, Stefanos Despotopoulos<sup>2</sup>, Niki Lama<sup>3</sup>, George Vagenakis<sup>2</sup>, Sotiria Apostolopoulou<sup>2</sup>, Sotiris Mitsiadis<sup>1</sup>, Dimitrios Boukis<sup>1</sup>, Maria Vassaki<sup>1</sup>, Evangelos Pisis<sup>1</sup>, Panagiotis Zachos<sup>4</sup>

<sup>1</sup>Echocardiography Training Center, Cardiology Department, "Tzanio" General Hospital, Piraeus, Greece; <sup>2</sup>Pediatric Cardiology and Adult Congenital Heart Disease Department, Onassis Cardiac Surgery Center,

Athens, Greece; <sup>3</sup>Research Unit of Radiology and Medical Imaging, 2nd Radiology Department, School of Medicine, National and Kapodistrian University of Athens, Athens, Greece; <sup>4</sup>Pediatric Cardiology and Adult Congenital Heart Disease Department, Karditsa General Hospital, Karditsa, Greece; 2nd Department of Health Economics, School of Medicine, National and Kapodistrian University of Athens, Athens, Greece

**Background and Aim:** Measurement of right ventricular (RV) volumes and ejection fraction (RVEF) in patients with repaired Tetralogy of Fallot (rToF) is often challenging due to right ventricular remodeling. Cardiac magnetic resonance (CMR) is considered to be the gold standard imaging modality in the evaluation of those patients. Nevertheless, 3D echocardiography (3D echo) seems to be a promising imaging modality for the assessment of patients with rToF. The present study aims to compare 3D echo and CMR in the evaluation of RV volume and RVEF in those patients.

**Method:** The study population consisted of adults, with a history of rToF, who were followed up at the Echocardiography Department “D. Beldekos” of “Tzanio” Hospital of Piraeus between January 2019 and January 2022. In all patients, a complete transthoracic echocardiographic evaluation was performed, where RV volume and RVEF were measured by 3D echo, along with a CMR for the measurement of RV volume and RVEF. A Mann-Whitney U test was used to compare the values measured by the two different imaging modalities since the variables didn't follow a normal distribution. The statistical significance level was set to 0.05 ( $p < 0.05$ ).

**Results:** A sample of 14 subjects with rToF (10 males, 4 females), of a mean age of 36.5 years (range 18 – 63 years), was examined. The comparison of RVEF measured by 3D echo (mean value 47.6%) and CMR (mean value 49%) showed a strong correlation between the two values ( $p = 0.317$ ). On the contrary, 3D echo seems to underestimate RV volumes compared to CMR. Both end-diastolic RV volume (mean value 53.5ml/m<sup>2</sup> by 3D echo vs 94ml/m<sup>2</sup> by CMR,  $p < 0.001$ ) and end-systolic RV volume (mean value 25ml/m<sup>2</sup> by 3D echo vs 44ml/m<sup>2</sup> by CMR,  $p < 0.001$ ) were underestimated by 3D echo.

**Conclusions:** LVEF measured by 3D echo and LVEF measured by CMR strongly correlate. On the contrary, 3D echo seems to underestimate the RV volumes.

**Keywords:** RVEF, RV volume, CMR, 3D echo, Tetralogy of Fallot

## P-161

### Impact of ventricular geometrical characteristics on myocardial stiffness assessment using shear wave elastography in healthy children and young adult

Aimen Malik<sup>1</sup>, Jerome Baranger<sup>2</sup>, Minh Nguyen<sup>2</sup>, Cameron Slorach<sup>2</sup>, Wei Hui<sup>2</sup>, José Carlos Villalobos<sup>1</sup>, Mark Friedberg<sup>2</sup>, Luc Mertens<sup>2</sup>, Olivier Villemain<sup>1</sup>

<sup>1</sup>Department of Medical Biophysics, University of Toronto, Toronto, Canada; Division of Cardiology, Department of Pediatrics, The Hospital for Sick Children, Toronto, Canada.; <sup>2</sup>Division of Cardiology, Department of Pediatrics, The Hospital for Sick Children, Toronto, Canada

**Background and Aim:** Diastolic myocardial stiffness (MS) can serve as a key diagnostic parameter for congenital or acquired heart diseases. Using shear modulus and shear wave velocity (SWV), shear

wave elastography (SWE) is an emerging ultrasound-based technique that can allow non-invasive assessment of MS. However, MS extrinsic parameters such as left ventricular geometrical characteristics could impact on shear wave propagation.

To determine a range of normal values of MS using SWE in age groups of healthy children and young adults and to explore the impact of left ventricular geometrical characteristics on SWE.

**Method:** 60 healthy volunteers (HVs) were recruited in the study and divided into 2 groups: “neonates” (0–1-month-old,  $n = 15$ ), “>1 month old” (1 month – 45 years,  $n = 45$ ). SWE was performed using the Verasonics Vantage systems with a phased array ultrasound probe. The anteroseptal basal segment was assessed in two views. SWE was ECG-triggered during the end-diastolic phase. Conventional echocardiography was achieved to assess ventricular function and anatomy. Results are presented as mean velocity and SD. Simple and multivariate linear regression analysis were performed.

**Results:** For neonates, mean MS was  $1.87 \pm 0.79$  kPa [min = 0.59 kPa; max = 2.91 kPa] with high variability and no correlation with age ( $p = 0.239$ ). For this age group, no statistically significant correlation was found between MS and any demographic or echocardiographic parameters ( $p > 0.05$ ). For “>1 month old” group, mean MS value of  $1.67 \pm 0.53$  kPa was observed [min 0.6 kPa, max 3 kPa] for HV. When age-paired, no sex-related difference was observed ( $p = 0.55$ ). In univariate linear regression analysis, age ( $r = 0.83$ ,  $p < 0.01$ ), diastolic interventricular septum thickness (IVSd,  $r = 0.72$ ,  $p < 0.01$ ), and left ventricle end-diastolic diameter ( $r = 0.67$ ,  $p < 0.01$ ) were the parameters with the higher correlation coefficient with MS. In a multiple linear regression analysis incorporating these three parameters as cofounding factors, age was the only statistically significant parameters ( $r = 0.81$ ,  $p = 0.02$ ).

**Conclusions:** Diastolic MS has a linear increase in children and young adults. Diastolic MS correlates more robustly with age than with myocardial and left ventricular geometrical characteristics. However, the geometry impacts on the SWV implying the need to determine well established boundaries through future studies for the clinical application of the SWE.

**Keywords:** diastolic function, echocardiography, myocardial stiffness, myocardium

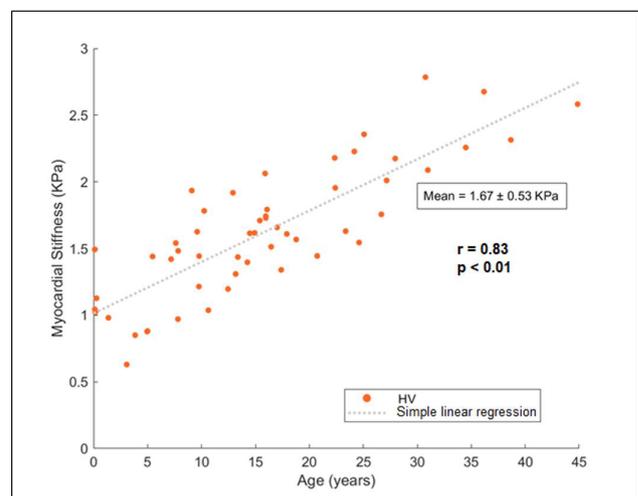


Figure 1. Myocardial Stiffness for “>1 month old” group

Figure 1. Myocardial Stiffness for “>1 month old” group

**P-163****Myocardial stiffness assessment by ultrasound in humans: comparison between shear wave elastography, natural mechanical waves, and myocardial stretch**

Jose Carlos Villalobos Lizardi<sup>1</sup>, Aimen Malik<sup>1</sup>, Jerome Baranger<sup>1</sup>, Minh Nguyen<sup>1</sup>, Maelys Venet<sup>1</sup>, Mathieu Pernot<sup>2</sup>, Luc Mertens<sup>1</sup>, Olivier Villemain<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology, The Hospital for Sick Children, Toronto, Canada; <sup>2</sup>Physic for Medicine, INSERM, Paris, France

**Background and Aim:** Myocardial stiffness (MS) is a key intrinsic property of the tissue that influences both diastolic and systolic cardiac function. Using wave velocity, different ultrasound methods exist to non-invasively assess MS during the cardiac cycle: shear wave elastography (SWE), natural mechanical waves (NMW), and intrinsic velocity propagation of myocardial stretch (iVP). The objective of this study is to compare these methods in human. **Method:** We performed a prospective study conducted in healthy children (n = 20; mean age = 5.8 years [1 week to 15 years]). SWE, NMW, and iVP were performed using the Verasonics Vantage systems with a phased array ultrasound probe. SWE was ECG-triggered with a 100ms incremental delay during 10 cardiac cycles. NMW velocities were assessed after mitral and aortic valve closure. iVP was assessed after the atrial kick.

**Results:** Using SWE, the mean velocity in systole was 1.85±0.58 m/s and 4.39±0.87 m/s in diastole and systole respectively (p<0.001) with a peak max velocity of 6.32 m/s in systole. Using NMW, the mean velocity after mitral valve closure was 3.3±0.75 m/s and 3.89±0.81 after aortic valve closure (p<0.001). These NSW were statistically different than mean SWE in diastole (p<0.005) and in systole (p<0.005). During the isovolumetric cardiac cycle periods, SWE and NMW were not different (p = 0.44 after mitral valve closure; p = 0.08 after aortic valve closure) and there was a significant linear correlation between the velocities obtained by SWE and NMW after mitral (r = 0.801, p<0.001) and aortic valve closure (r = 0.822, p<0.001). The iVP occurred 41.7±11.3 ms after the P wave onset (ECG) and were statistically different than the SWE at this moment (1.56±0.23 ms versus 1.2±0.37 ms respectively, p = 0.003).

**Conclusions:** In healthy volunteers, SWE has a significant physiological variation during cardiac cycle. NMW and SWE have similar velocities when temporally correlated. NMW occur during isovolumetric periods and are different than diastolic and systolic velocities observed by SWE.

**Keywords:** ultrafast ultrasound imaging, cardiology, shear wave imaging, myocardial stiffness

**P-164****Vascular and ventricular responses to exercise in pediatric marfan syndrome**

Nairy Khodabakhshian<sup>1</sup>, Cameron Slorach<sup>2</sup>, Wei Hui<sup>2</sup>, Cheryl Fackoury<sup>2</sup>, Barbara Cifra<sup>2</sup>, Olivier Villemain<sup>2</sup>, Vitor Guerra<sup>2</sup>, Luc Mertens<sup>1</sup>

<sup>1</sup>Institute of Medical Science, University of Toronto, Toronto, Canada; Division of Cardiology, The Hospital for Sick Children (SickKids), Toronto, Canada; <sup>2</sup>Division of Cardiology, The Hospital for Sick Children (SickKids), Toronto, Canada

**Background and Aim:** Cardiac clinical management of Pediatric Marfan Syndrome (MFS) focuses on slowing aortic dilation through pharmacological and surgical interventions coupled with

exercise restrictions. However, impact of the latter on aortic properties, ventricular function, and vascular function in pediatric MFS patients is poorly understood. We hypothesize that vascular and ventricular responses to exercise are abnormal in MFS patients when compared to healthy controls.

The objectives of this study are: 1) Study aortic, vascular, and ventricular properties in pediatric MFS at rest and during exercise; 2) Study aortic, vascular, and ventricular response to exercise and study the associations to the degree and rate of aortic dilatation in pediatric MFS.

**Method:** 35 pediatric MFS patients (ages 10-17) and 35 age-matched controls will undergo transthoracic echocardiography (TTE) during rest and exercise. Vascular function is assessed at rest using Flow-Mediated Dilatation (FMD), Carotid Intima-Media Thickness (CIMT), and endoPAT, and during exercise using carotid-femoral Pulse Wave Velocity (PWV). During exercise, participants pedal on a semi-supine bicycle at 60 revolutions per minute (rpm) against increasing resistance. Exercise concludes at 85% of age-adjusted maximum heart rate or upon participant fatigue.

**Results:** Compared to controls (n = 20) at rest, MFS patients (n = 23) have comparable aortic stiffness index (p = 0.98), FMD % change in diameter (p = 0.91), and CIMT (p = 0.64). MFS patients have significantly higher resting Reactive Hyperemia Index via endoPAT (p < 0.01) and comparable PWV at rest and peak exercise (p = 0.64 and p = 0.15, respectively). Resting systolic and diastolic functional parameters, such as mean longitudinal strain and mitral valve E/E' ratio, are significantly lower in MFS patients (all p < 0.01). All parameters in MFS patients, except mean longitudinal strain (p = 0.05), increase similarly to controls during submaximal exercise.

**Conclusions:** Our preliminary data suggests that when compared to healthy controls, aortic and vascular properties during rest and exercise are not significantly different in MFS. Furthermore, while MFS patients have lower resting parameters for systolic and diastolic function, these increase normally in response to submaximal exercise indicating a preserved exercise response in MFS.

**Keywords:** Cardiac imaging, echocardiography, vascular imaging, exercise physiology, Connective Tissue Disorders, pediatric cardiology

**P-165****Hemodynamics in children with partial anomalous pulmonary venous return and atrial septal defect using 4D-flow MRI**

Fraser Maurice Callaghan<sup>1</sup>, Barbara Burkhardt<sup>2</sup>, Emanuela Valsangiaco Buechel<sup>2</sup>, Christian Kellenberger<sup>3</sup>, Julia Geiger<sup>3</sup>  
<sup>1</sup>Center for MR Research, University Children's Hospital, Zurich, Zurich, Switzerland; Children's Research Center, University Children's Hospital Zurich, Zurich, Switzerland; <sup>2</sup>Division of Pediatric Cardiology, University Children's Hospital Zurich, Zurich, Switzerland; Children's Research Center, University Children's Hospital Zurich, Zurich, Switzerland; <sup>3</sup>Department of Diagnostic Imaging, University Children's Hospital Zurich, Zurich, Switzerland; Children's Research Center, University Children's Hospital Zurich, Zurich, Switzerland

**Background and Aim:** Partial anomalous pulmonary venous return (PAPVR) combined with atrial septal defect (ASD) is a rare congenital heart defect (CHD) causing a left to right shunt.

Left atrial (LA) hemodynamics are known to contribute to efficient left ventricle (LV) filling, and understanding the disruption of LA hemodynamics in these patients with PAPVR+ASD is important.

**Method:** We used 4D-flow magnetic resonance imaging (MRI) to assess LA hemodynamics in 10 patients (age  $3.8 \pm 1.9$  years), in comparison with 13 age-matched normal subjects.

Flow measurements at each pulmonary vein (PV), the ASD and mitral valve (MV) and flow particle tracking were used to classify flow transfer by source (PV) and destination (ASD or MV).

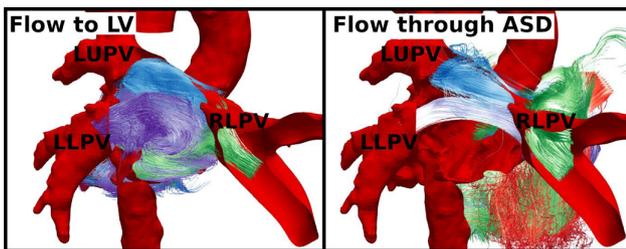
**Results:** All patients had a right upper anomalous pulmonary vein and a superior sinus venosus ASD. Patients had pulmonary to systemic flow ratios (Qp:Qs) of 2.2–3.5 (mean  $\pm$  standard deviation:  $2.7 \pm 0.4$ ). The fraction of total PV return flow per PV was calculated for each subject and no differences were found between normal and PAPVR+ASD subjects.

Right lower PV flow was more likely to pass through the ASD (75%) than the MV. Left lower PV flow was more likely to pass through the MV (75%) and left upper PV flow was almost evenly distributed to the ASD (44%) and MV (56%). The greatest amount of flow through the ASD occurred during systole. All patients' PVs showed a significantly greater proportion of total flow volume passing during systole compared with normal subjects (all  $p < 0.01$ ) who conversely showed a greater proportion of flow volume during diastole (all  $p < 0.01$ ). Pulmonary vein flow dynamics during the atrial contraction period of the cardiac cycle showed no difference. Left ventricular filling velocities were significantly reduced ( $p < 0.001$ ) in PAPVR+ASD patients compared to normal subjects, and Qp:Qs in patients ( $2.7 \pm 0.7$ ) correlated positively with ASD area ( $r = 0.8$ ).

**Conclusions:** Using 4D-flow MRI particle tracking allows classification and assessment of both PV and LA flow dynamics in patients with PAPVR and ASD. Flow through the ASD into the low pressure right atrium during systole disrupts normal hemodynamics and reduces LA conduit function leading to inefficient LV filling.

**Keywords:** 4D flow MRI, hemodynamics, Partial anomalous pulmonary venous return, atrial septal defect, left atrium

Left atrial flow dynamics assessment by 4D flow MRI particle tracking



An exemplar patient with partial anomalous pulmonary venous return and atrial septal defect showing flow paths classified by origin (pulmonary veins) and destination (left ventricle or atrial septal defect) allowing for complex analysis of flow dynamics.

## P-166

### Juxtaposition of the atrial appendages: recognition and clinical implications

Heba Abdllhamid<sup>1</sup>, Trevor Richens<sup>1</sup>, Nicola Viola<sup>2</sup>, Tara Bharucha<sup>1</sup>

<sup>1</sup>Pediatric Cardiology Department, University Hospital Southampton, Southampton, United Kingdom; <sup>2</sup>Congenital Cardiac Surgery Unit, University Hospital Southampton, Southampton, United Kingdom

**Background and Aim:** BACKGROUND: Juxtaposition of the atrial appendages (JAA) is an unusual anomaly in which the two atrial appendages lie on the same side, to the left or right of the great vessels instead of either side. JAA can be diagnosed by angiography, CT scan or echocardiography.

**AIM:** We highlight clinical implications of JAA and diagnostic approaches in a series of 3 cases.

**Method:** Case 1:

A 32 week gestation baby, birth weight 1.65 kg with antenatal diagnosis of transposition of great arteries (TGA) with intact ventricular septum, diagnosis confirmed postnatally, required balloon atrial septostomy for restrictive atrial septum.

Case 2:

Term baby with antenatal diagnosis of double outlet right ventricle with TGA and sub pulmonary stenosis. Postnatal echocardiography additionally identified JAA, and the balloon atrial septostomy technique was accordingly adapted.

Case 3:

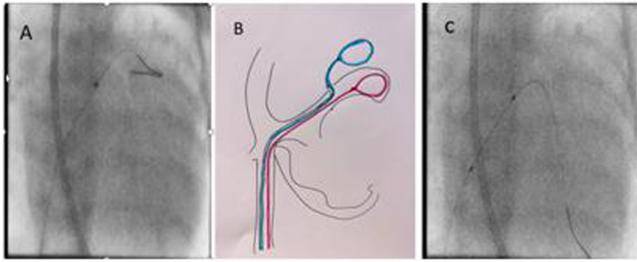
35+4 week gestation baby with antenatal diagnosis of TGA and bilateral superior vena cavae (SVC) was referred for arterial switch operation. Surgical inspection revealed severe hypoplasia of right SVC, and non-connected large left SVC. There was no identifiable atrial mass in the right fossa and right atrial appendage (RAA) and left atrial appendage (LAA) were juxtaposed to the left of the main pulmonary artery. The left coronary venous system drained directly to the RAA.

**Results:** Diagnostic approaches in a series of 3 cases.

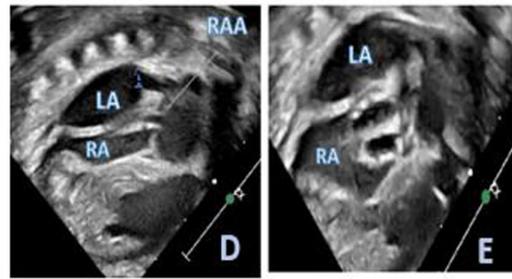
**Conclusions:** Although rare, JAA is often associated with severe congenital heart lesions, with significant implications for interventional or surgical treatment. Precise and complete description of this arrangement should be sought ahead of all interventions and surgeries.

**Keywords:** Juxtaposition of atrial appendages, Transposition of great arteries

Diagnostic approaches in a series of 3 cases



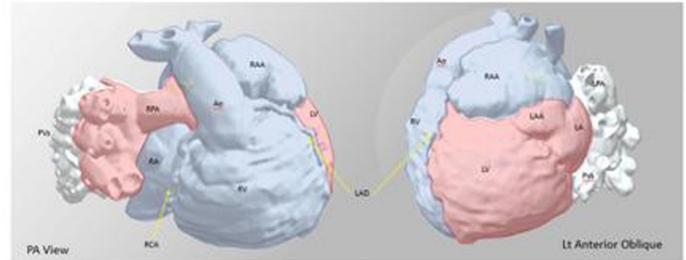
Case 1. Fluoroscopy guided images showing: A. The guidewire directed to the left and posteriorly, lying within the right atrial appendage (RAA). B. Diagram showing wire within RAA (pink) and across interatrial communication into left atrium (LA) (blue). Figure C. Guidewire very similar in position to A, but across atrial septum and entering pulmonary vein.



Case 1. Echo images - Figure D. Catheter in juxtaposed right atrial appendage. Figure E. Septostomy balloon inflated but lying in right atrium (RA).



Case 2. Figure F. Subcostal view showing right atrial appendage projecting beneath left atrium



Case 3. Postero-anterior and left anterior oblique views of 3D model printed from CT scan. The RAA and LAA are seen adjacent to each other to the left of the great arteries.

**P-167**

**Intracavitary blood flow dynamics in the systemic right ventricle: A 4D-flow MRI study**

Giulia Pasqualin<sup>1</sup>, Alessandra Riva<sup>2</sup>, Francesco Sturla<sup>3</sup>, Francesca Bevilacqua<sup>1</sup>, Mauro Lo Rito<sup>1</sup>, Daniel Giese<sup>4</sup>, Paolo Ferrero<sup>1</sup>, Alessandro Giamberti<sup>1</sup>, Antonio Saracino<sup>1</sup>, Alberto Redaelli<sup>2</sup>, Emiliano Votta<sup>2</sup>, Massimo Chessa<sup>1</sup>, Massimo Lombardi<sup>5</sup>

<sup>1</sup>Pediatric and Adult Cardiology Heart Centre, IRCCS Policlinico San Donato, Milan, Italy; <sup>2</sup>Department of Electronics, Information and Bioengineering, Politecnico di Milano, Milan, Italy; <sup>3</sup>3D and Computer Simulation Laboratory, IRCCS Policlinico San Donato, Milan, Italy; <sup>4</sup>Siemens Healthcare, Erlangen, Germany; <sup>5</sup>Multimodality Cardiac Imaging Section, IRCCS Policlinico San Donato, Milan, Italy

**Background and Aim:** Failure of the systemic right ventricle (SRV) is based on morphological differences between right and left ventricles (RVs and LVs). RV adaptation to systemic afterload includes increased circumferential over longitudinal global myocardial strain, with an unknown impact on intracavitary blood flow distribution. This study aimed to explore the SRV pattern of hemodynamic forces (HF).

**Method:** 4D-Flow cardiovascular magnetic resonance data were acquired using a prototype sequence on a 1.5-T MAGNETOM Aera (Siemens Healthcare, Erlangen, Germany). The ratio between transverse (inferior-anterior, HFIA and septal-lateral, HFSL) and longitudinal (basal-apical, HFBA) HFs (RRMS) was calculated as in Figure 1, for systole and diastole.

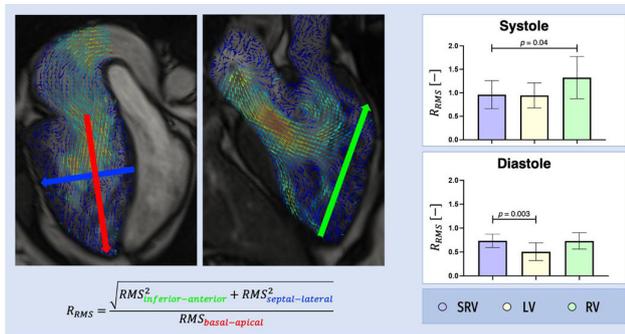
**Results:** We enrolled 12 adults with SRV (6 D-transposition of great arteries after atrial switch operation and 6 L-transpositions)

and 12 age-matched healthy subjects (41±12 vs 42±13, p=0.89). Although SRVs reported comparable end-diastolic volumes (83±18 ml/m<sup>2</sup>) and ejection fraction (60±8%) to control RVs (75±13 ml/m<sup>2</sup>, p=0.18; 64±5%, p=0.25), SRV mass (55±24 g/m<sup>2</sup>), free-wall circumferential strain (-18.9±7.8%) and longitudinal strain (-22.7±6.1%) were different than RVs (20±3 g/m<sup>2</sup>, p<0.001, -9.3[-11.0;-8.3], p<0.0001, -27.4[-30.2;-25], p=0.015) and comparable to control LVs (57±11 g/m<sup>2</sup>, p=0.7, -20.2±3.8%, p=0.3, 21.2±5.3, p=0.5 respectively).

The 4D-Flow analysis showed that SRV systolic RRMS (0.98 ±0.31) was similar to LVs (0.94±0.27, p=0.78) but lower than RVs (1.32±0.45, p=0.04). This reflected a significantly increased HFBA with respect to RVs (0.338±0.150 vs. 0.162 ±0.097, p=0.0025) and similar to LVs (0.462 ± 0.186, p=0.087). Concomitantly, a moderate correlation was demonstrated between SRV systolic HFBA magnitude and GCS (r<sup>2</sup>=0.47, p=0.013). During diastole, SRVs showed lower HFBA (0.173±0.086) than LVs (0.304±0.104, p=0.0028), revealing a diastolic RRMS (0.74 ± 0.14) comparable to RVs (0.73 ± 0.17, p=0.95) and significantly different from LVs (0.50 ± 0.19, p=0.003).

**Conclusions:** In SRVs, RRMS is similar to LVs during systole, revealing a prevalent longitudinal direction of the blood flow, possibly as a result of increased GCS. Inversely, the SRV filling appears to be closely related to ventricular morphology as suggested by a prevalent transversal blood flow direction during diastole and a diastolic RRMS comparable to RVs.

**Keywords:** Systemic Right Ventricle, 4D-Flow MRI



**Figure 1** On the left: ratio between the root mean square (RMS) of the transversal (i.e., septal-lateral and inferior-anterior) and longitudinal (basal-apical) components of the HDF (RRMS). On the right: RRMS systolic and diastolic values. Each box ranges between 25th and 75th percentile with a line pointing out the median values; whiskers indicate the 10th and 90th percentile, respectively.

### P-168

#### Left atrial strain in children with idiopathic dilated cardiomyopathy

Katarzyna Luczak Wozniak<sup>1</sup>, Cezary Niszczota<sup>2</sup>, Klaudia Obszajczyk<sup>2</sup>, Bożena Werner<sup>3</sup>

<sup>1</sup>Department of Pediatric Cardiology and General Pediatrics, Doctoral School, Medical University of Warsaw, Warsaw, Poland; <sup>2</sup>Department of Pediatric Cardiology and General Pediatrics, Jozef Polikarp Brudziński Public Pediatric Hospital, Warsaw, Poland; <sup>3</sup>Department of Pediatric Cardiology and General Pediatrics, Medical University of Warsaw, Warsaw, Poland

**Background and Aim:** In adults with dilated cardiomyopathies left atrial conduit strain on CMR has been shown to have a superior value for predicting unfavorable outcome than left ventricular ejection fraction or left ventricular global longitudinal strain. Strain analysis has also recently received attention in the pediatric population with cardiomyopathies, however the data is limited. The aim of the study was to assess differences between left atrial strain in pediatric patients with idiopathic dilated cardiomyopathy (DCM) and the control group, as well as its' association with unfavorable outcome.

**Method:** Patients with idiopathic dilated cardiomyopathy under the age of 18 and age-matched healthy control subjects were included into the study. 2D speckle tracking echocardiography (STE) was performed using Philips EPIQ ultrasound system 9.0.1., left atrial reservoir and conduit strain, left ventricular global longitudinal strain (LV GLS), left ventricular ejection fraction (LVEF) as well as left ventricular internal diastolic diameter (LVIDd) z-score were assessed. Unfavorable outcome was defined as qualification for or receiving heart transplant.

**Results:** There were 21 children with DCM included into the study; 14 boys and 7 girls, mean age 7 years (SD  $\pm 6.1$  years). Four patients (19%) were qualified for or received heart transplant. Children with dilated cardiomyopathy had a significantly lower left atrial reservoir strain ( $26 \pm 11.8$  vs.  $43 \pm 9.8$   $p < 0.01$ ) as well as a significantly impaired left atrial conduit strain ( $-22 \pm 12.8$  vs.  $-32 \pm 9.9$   $p = 0.01$ ). Furthermore, children with unfavorable outcome compared to those with DCM without adverse events had a significantly lower left atrial reservoir strain ( $13.1 \pm 7.4$  vs.  $29 \pm 10.7$   $p = 0.01$ ) as well as significantly different left atrial conduit strain ( $-10 \pm 3$  vs.  $-25.1 \pm 12.6$   $p = 0.03$ ). Patients with

unfavorable outcome had a significantly lower LV GLS  $-16\%$  vs.  $-7.6\%$  ( $p$ -value  $< 0.01$ ), LVEF  $18.9\%$  vs.  $42\%$  ( $p$ -value  $< 0.01$ ) as well as LVIDd z-score  $+5.0$  vs.  $+3.2$  ( $p$ -value  $< 0.01$ ). **Conclusions:** Left atrial strain seems to be an easily obtainable parameter that differentiates children with DCM from healthy control subjects. Furthermore, reduced LA reservoir function demonstrated by STE strain might be a useful marker for risk stratification in children with idiopathic DCM.

**Keywords:** pediatric idiopathic dilated cardiomyopathy, left atrial strain

### P-169

#### Genotype-phenotype interactions in left ventricular morphology, diastolic function, and systolic function in pediatric loeys-dietz syndrome

Nairy Khodabakhshian<sup>1</sup>, Alison Howell<sup>2</sup>, Pablo Perez Lopez<sup>3</sup>, Cameron Slorach<sup>2</sup>, Wei Hui<sup>2</sup>, Vitor Guerra<sup>2</sup>, Luc Mertens<sup>2</sup>

<sup>1</sup>Institute of Medical Science, University of Toronto, Toronto, Canada; <sup>2</sup>Division of Cardiology, The Hospital for Sick Children (SickKids), Toronto, Canada; <sup>3</sup>Division of Cardiology, The Hospital for Sick Children (SickKids), Toronto, Canada; <sup>3</sup>Faculty of Medicine, Universidad Autonoma de Madrid, Madrid, Spain; Hospital Universitario Puerta de Hierro Majadahonda, Majadahonda, Spain; <sup>2</sup>Division of Cardiology, The Hospital for Sick Children (SickKids), Toronto, Canada

**Background and Aim:** The mutations causing Loey-Dietz Syndrome (LDS) have pathological effects beyond aortopathy. These can be heterogenous in severity and phenotype between the mutations. Our group previously found LDS patients have significantly lower left atrial strain and left ventricular (LV) diastolic functional parameters compared to age-matched controls. The aim of this project is to study the impact of genotype-phenotype interactions on LV morphological and functional parameters in pediatric LDS.

**Method:** Patients were identified from an institutional database from 1/2001 - 12/2020. Inclusion criteria were: a pathogenic or likely pathogenic variant of TGF $\beta$ 2, TGF $\beta$ 3, TGF $\beta$ 1, TGF $\beta$ 2, SMAD2, or SMAD3; and  $> 2$  transthoracic echocardiogram (TTE) performed before age 18. LDS patients were divided into 2 groups: TGF $\beta$ 2 vs. Other (TGF $\beta$ 2, TGF $\beta$ 3, TGF $\beta$ 1, SMAD2, or SMAD3). Clinical parameters were collected via chart review of LDS patients and age-matched controls were extracted from our laboratory database. TTEs were reviewed offline and measurements were gathered on aortic Sinus of Valsalva (SoV), LV morphology, LV diastolic function, and LV systolic function.

**Results:** The median age at TTE was 12.3 (IQR: 6.85) for LDS-TGF $\beta$ 2 ( $n = 15$ ) and 12.8 (IQR = 10.7) for LDS-Other ( $n = 22$ ). LDS-TGF $\beta$ 2 patients had significantly larger mean SoV ( $2.18 \pm 0.32$  vs.  $1.89 \pm 0.42$ ,  $p = 0.026$ ) and SoV z-scores ( $3.04 \pm 0.45$  vs.  $2.70 \pm 0.68$ ,  $p = 0.053$ ). There were no significant differences in LV diastolic or systolic functional parameters between genotype groups.

**Conclusions:** Pediatric LDS patients with TGF $\beta$ 2 mutations have increased aortic dilatation compared to non-TGF $\beta$ 2 LDS patients, suggesting aortic genotype-phenotype differences. There are no significant differences in LV morphology and function between the mutation groups. Further studies are needed to better delineate genotype-phenotype relationships in LDS patients.

**Keywords:** Echocardiography, left ventricular diastolic function, left ventricular systolic function, left ventricular morphology, Connective Tissue Disorders, Pediatric Loey-Dietz Syndrome

General Characteristics	LDS: TGFβR2 (n=15)	LDS: Other (n=22)	p
Age at echo (years)*	12.3, 6.85	12.8, 10.7	0.745
Gender (#, % males)	8, 53	15, 72	0.573
Height (cm)*	152, 36	152, 62.5	0.898
Weight (kg)*	28.8, 25.9	39.9, 37.8	0.599
BSA (m <sup>2</sup> )	1.29 ± 0.47	1.32 ± 0.54	0.838
Aortic Sinus of Valsalva (cm)	2.18 ± 0.32	1.89 ± 0.42	<b>0.026</b>
Aortic Sinus of Valsalva Pediatric Heart Network (PHN) z-score	3.04 ± 0.45	2.70 ± 0.68	<b>0.053</b>
<b>Left Ventricular Structure</b>			
LV End-Diastolic Diameter, indexed*	3.92, 1.91	3.33, 2.2	0.795
LV End-Systolic Diameter, indexed*	2.56, 1.12	2.14, 1.40	0.927
LV Posterior Wall Thickness in End-Diastole, indexed*	0.52, 0.09	0.44, 0.11	0.089
LV Posterior Wall Thickness in End-Systole, indexed	0.95 ± 0.28	0.79 ± 0.27	0.093
<b>Left Ventricular Diastolic Function</b>			
Mitral Valve E-velocity (cm/s)	78.6 ± 24.9	78.5 ± 16.2	0.995
Mitral Valve A-velocity (cm/s)	45.1 ± 9.34	43.1 ± 8.01	0.502
Mitral Valve E/A ratio	1.81 ± 0.74	1.92 ± 0.58	0.644
Mitral Valve E'-velocity (cm/s)	16.1 ± 3.45	16.5 ± 3.53	0.722
Mitral Valve A'-velocity (cm/s)	6.10 ± 1.51	5.62 ± 1.69	0.376
Mitral Valve E/E'	4.86 ± 1.21	4.92 ± 1.34	0.905
Isovolumetric Relaxation Time (ms)*	60, 20	61, 20.5	0.526
Mitral Valve Deceleration Time (ms)*	172, 59.5	141, 58	0.710
Mitral Valve A-wave Time (ms)*	140, 84	119, 67	<b>0.058</b>
<b>Left Ventricular Systolic Function</b>			
Mitral Valve S'-velocity (cm/s)*	11.2, 2.05	10.8, 3.5	0.139
LV Ejection Fraction (%)	56.2 ± 7.01	53.6 ± 9.59	0.348
LV Shortening Fraction (%)	37.4 ± 3.68	35.5 ± 4.28	0.157
Global Longitudinal Strain (%)	18.8 ± 3.49	20.3 ± 3.62	0.317

**Table 1: General characteristics, LV morphology, LV diastolic function, and LV systolic function parameters in LDS-TGFβR2 patients vs. LDS-Other. The mean and standard deviations are listed for normally distributed variables, which were analyzed using Unpaired Student's t-test. Medians and interquartile ranges are listed for non-normally distributed parameters (marked by an asterisk), which were analyzed using Mann-Whitney U test.**

### P-170

#### Utilization of 3D-virtual cardiovascular models for operative planning of complex congenital heart defects: A single-center 1-year experience

Peter Olejnik<sup>1</sup>, Patricia Srnkova<sup>1</sup>, Marek Kardos<sup>2</sup>

<sup>1</sup>National Institute of Cardiovascular Diseases, Pediatric Cardiac Center, Bratislava, Slovakia; <sup>2</sup>Department of Pediatric Cardiology, Faculty of Medicine, Comenius University, Bratislava, Slovakia; <sup>2</sup>National Institute of Cardiovascular Diseases, Pediatric Cardiac Center, Bratislava, Slovakia

**Background and Aim:** The goal of this single-centre study was to evaluate the benefits of 3D virtual versicolour heart models in clinical management of patients with complex congenital heart defects.

**Method:** Between October 2021 and September 2022, 27 three-dimensional virtual heart models based on computed tomography data were reconstructed. The contribution of 3D virtual heart models for perioperative planning improvement was evaluated in all patients.

**Results:** 17 virtual models focused on imaging of complex intracardiac anatomical structures and 10 virtual models aimed on delineation of complex extracardiac vascular anatomy were created. Each model offered an improved anatomical orientation of cardiovascular structures. After models' analyses by pediatric cardiologists and cardiothoracic surgeons the most suitable cardiac surgery or catheter intervention was chosen in each individual case.

**Conclusions:** The use of 3D virtual models enabling high-quality spatial imaging of cardiovascular structures can facilitate

interventional procedures in complex congenital heart defects due to better preoperative planning.

**Keywords:** 3D-virtual models, congenital heart defects, operative planning

### P-171

#### Association between arterial stiffness of the common carotid artery and left ventricular function in children, adolescents and young adults

Franziska Sciuk<sup>1</sup>, Theresa Viltsmaier<sup>2</sup>, Marie Kramer<sup>1</sup>, Magdalena Langer<sup>1</sup>, Brenda Kolbinger<sup>2</sup>, Pengzhu Li<sup>1</sup>, André Jakob<sup>1</sup>, Nina Rogenhofer<sup>2</sup>, Robert Dalla-Pozza<sup>1</sup>, Christian Thaler<sup>2</sup>, Nikolaus Alexander Haas<sup>1</sup>, Felix Sebastian Oberhoffer<sup>1</sup>

<sup>1</sup>Division of Pediatric Cardiology and Intensive Care, University Hospital, LMU Munich, Munich, Germany; <sup>2</sup>Division of Gynecological Endocrinology and Reproductive Medicine, Department of Obstetrics and Gynecology, University Hospital, LMU Munich, Munich, Germany

**Background and Aim:** Arterial stiffness serves as an early marker for cardiovascular risk stratification. Two-dimensional speckle tracking (2DST) of the common carotid artery (CCA) has been recently introduced for the assessment of regional arterial stiffness. The literature suggests a link between increased arterial stiffness and cardiac dysfunction in adult patients. This study aimed to investigate the relationship between arterial stiffness of the CCA measured through 2DST and left ventricular (LV) function in a cohort of children, adolescents and young adults.

**Method:** Regional arterial stiffness of both CCA was evaluated through 2DST by determining peak circumferential strain (CS, %) and strain rate (SR, 1/s). LV function was assessed through M-Mode echocardiography, pulsed Doppler and tissue Doppler imaging. Moreover, a non-invasive LV pressure-volume loop was generated enabling the calculation of LV hemodynamics and efficiency parameters. For analysis, the Spearman and Pearson correlation coefficients were utilized.

**Results:** 149 subjects with a mean age of 13.19 years were included in this study. Average CS was 14.60±3.84 % and average SR 4.03 ±0.90 1/s. Myocardial early diastolic peak velocity (E', cm/s) measured over the interventricular septum correlated significantly with CS (r = 0.248, p = 0.002) and SR (r = 0.249, p = 0.002). Similar effects were demonstrated for E' measured over the LV wall and CS (r = 0.180, p = 0.028). Significant inverse correlations were displayed between CS and/or SR with interventricular septum thickness at end-diastole, LV posterior wall thickness at end-diastole, relative wall thickness and LV mass. For the remaining LV parameters including ejection fraction, global longitudinal strain, mitral inflow velocities, LV hemodynamics and efficiency parameters derived by generating a non-invasive pressure-volume loop, no significant correlations were found.

**Conclusions:** This study suggests a small but significant link between regional arterial stiffness of the CCA, LV diastolic function and LV morphology. Subjects with increased arterial stiffness might display a higher susceptibility for lower LV diastolic function.

Therefore, the implementation of 2DST of the CCA might provide additional information for early cardiovascular risk stratification.

**Keywords:** Arterial Stiffness, Two-Dimensional Speckle Tracking, Left Ventricular Function, Pediatric Cardiology

## P-172

**Regional elevation of liver T1 in fontan-associated liver disease**Edythe Tham<sup>1</sup>, Paul Greidanus<sup>1</sup>, Carolina Escudero<sup>1</sup>, Joseph Pagano<sup>1</sup>, Richard Thompson<sup>2</sup><sup>1</sup>Stollery Children's Hospital/University of Alberta; <sup>2</sup>Biomedical Engineering/University of Alberta

**Background and Aim:** Fontan associated liver disease (FALD) is a characterized by hepatic congestion and progressive hepatic fibrosis in patients with a single ventricle Fontan circuit. This condition is generally clinically silent until late in the disease necessitating techniques for early detection. Liver T1 mapping has been used to screen for FALD, though thus cannot reliably distinguish between congestion or fibrosis.

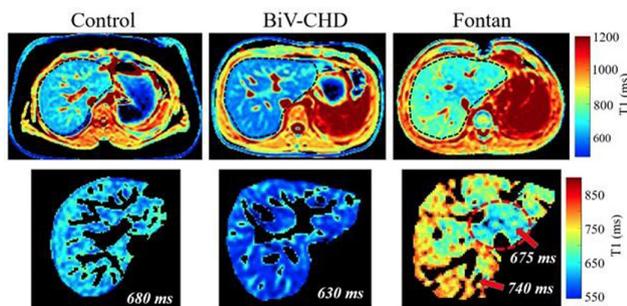
**Method:** This cohort study compares liver T1 measured with a liver-specific T1 mapping sequence (PROFIT1 - Proton Density Fat Fraction Imaging with Water-Specific T1) in Fontan patients to cohorts of patients with biventricular congenital heart disease (BiV-CHD) and controls with normal cardiac function and anatomy. Liver T1 was measured from several axial slices at the widest dimension of the liver. Ventricular volumes and ejection fraction (EF) were calculated from the systemic ventricle. Myocardial native T1 mapping (MOLLI) at a mid-ventricular short axis were quantified. Cohort means were compared with Kruskal Wallis test and Dunn's posthoc test and association was determined with Pearson correlation coefficient.

**Results:** Global Liver T1 was significantly elevated in the Fontan cohort (n = 20) compared to BiV-CHD patients (n = 12) and controls (n = 9) (785ms, 672ms, 671ms, p<0.001), with a consistent pattern of significantly elevated T1 values in the peripheral liver as compared to central liver regions in the Fontan group (change in T1 = 54 ms in Fontan, versus 6 ms in BiV-CHD and 13 ms in controls, p<0.001). PROFIT1 also yielded simultaneous T2\* maps, with similar values in all groups (Fontan 29.3 ms, BiV-CHD 24.7 ms and controls 24.4 ms) and PDFF (fat fraction) maps (Fontan 1.6%, BiV-CHD 1.9%, controls 1.8%). Fontan liver T1 values were also significantly elevated as compared to BiV-CHD and controls as measured with the cardiac (MOLLI) acquisitions (728ms, 583 ms, 583 ms, p<0.001) and values were correlated with PROFIT1 T1 values (r = 0.88).

**Conclusions:** Fontan patients have significantly increased liver T1 values as compared to BiV-CHD and controls, although the contributions from congestion and fibrosis remain unknown. Higher global T1 values are associated with a pattern of elevated values in the peripheral liver regions, which may be a clue to distinguishing liver fibrosis and congestion.

**Keywords:** T1 mapping, Fontan associated liver disease, MRI

T1 maps



PROFIT1 map showing regional variations in the liver

## P-173

**Role of speckle-tracking echocardiography and feature tracking-CMR in evaluation of myocardial deformation in repaired tetralogy of fallot patients**Annachiara Cavaliere<sup>1</sup>, Alice Pozza<sup>2</sup>, Elena Reffo<sup>2</sup>, Jolanda Sabatino<sup>2</sup>, Giovanni Di Salvo<sup>2</sup><sup>1</sup>Department of Neurosciences, Paediatric Radiology, Padua University, Italy; <sup>2</sup>Department of Women's and Children's Health, Paediatric Cardiology, Padua University, Italy

**Background and Aim:** Current surgical procedures guarantee a survival at 20 years of repaired Tetralogy of Fallot (r-TOF) patients post pulmonary valve replacement (PVR) around 90%. The growing adult r-TOF population is not free from adverse events and right ventricular dysfunction via ventricular-ventricular interactions might lead to left ventricular failure.

To identify possible correlation between cardiac magnetic resonance (CMR) and speckle-tracking echocardiography (STE) functional data in r-TOF patients pre and post-PVR; to early depict systolic dysfunction by global longitudinal strain (GLS) values and then optimize CMR and PVR timing; to identify correlation between GLS and feature tracking-CMR (FT-CMR) to detect differences in pre and post-PVR.

**Method:** In this retrospective and partly prospective single center study, r-TOF patients' inclusion criteria are: a) repaired TOF; b) available CMR and 2D-Echo data pre-PVR; c) underwent to PVR between July 2015 and September 2021; d) CMR and 2D-Echo data post-PVR. Multivariate analysis was performed.

**Results:** Analysis performed on data obtained from the 15 eligible patients (8 females, mean age 23.4 years, SD 12.4 years) showed a statistically significant reduction in telediastolic (p.004) and telesystolic (p.01) volumes after PVR, with no significant increase in ejection fraction (p.853). Right ventricular ejection fraction pre-PVR was found to be an early indicator of right ventricular GLS post-PVR (p.027). Right ventricular GLS calculated by FT-CMR showed statistically significant improvement pre- and post-PVR (p.005).

**Conclusions:** This study highlights that the sooner the valve replacement is performed, the better the GLS will be. Moreover, it depicts that right ventricular myocardial remodeling is well evaluated by STE and FT-CMR. Furthermore, it suggests an update of the current guidelines in order to include such parameters in the routine evaluation of r-TOF patients post-PVR.

**Keywords:** Tetralogy of Fallot, Pulmonary Valve Replacement, Cardiac Magnetic Resonance, Speckle-Tracking Echocardiography, Global Longitudinal Strain

## P-174

**Long term cardiovascular effects of COVID-19 infection in children**Dareilena Karaviti<sup>1</sup>, Marietta Charakida<sup>1</sup>, Dimitra Dimopoulou<sup>1</sup>, Irine Eleftheriou<sup>1</sup>, Marianthi Liaska<sup>1</sup>, Maria Papadaki<sup>1</sup>, Despoina Maritsi<sup>1</sup>, Irine Ikbale Sakou<sup>1</sup>, Antonios Marmarinos<sup>2</sup>, Margaritis Avgeris<sup>2</sup>, Dimitrios Gourgiotis<sup>2</sup>, Nikos Spyridis<sup>1</sup>, Maria Tsolia<sup>1</sup><sup>1</sup>2nd Department of Pediatrics, National and Kapodistrian University of Athens, Children's Hospital "P. & A. Kyriakou", Athens, Greece;<sup>2</sup>Laboratory of Clinical Biochemistry-Molecular Diagnostics, Second Department of Pediatrics, School of Medicine, National and Kapodistrian University of Athens, Children's Hospital "P. & A. Kyriakou", Athens, Greece

**Background and Aim:** COVID-19 infection has been associated with prolonged cardiovascular dysfunction in adults, however similar information in children is lacking. In this study, we performed

cardiovascular assessment in children who were infected by the SARS-CoV-2 up to one year from their infection and compared their results with healthy controls who have not been infected by SARS-CoV-2.

**Method:** A total of 234 children were evaluated. Group 1 consisted of 155 children with positive history of COVID-19 infection [105 recovered at home; mild disease, 40 required hospitalization; severe/moderate disease and 10 had multisystem inflammatory syndrome (MIS-C)]. The remaining were healthy controls with negative SARS-CoV-2 IgG antibodies (Group 2). In all children we performed anthropometric measurements and measured lipid profile and inflammatory markers. All children were examined with echocardiography to assess left and right ventricular function using conventional M-mode, colour Doppler and tissue Doppler and had aortic stiffness with pulse wave velocity (PWV) and carotid intima media thickness (IMT) measurements.

**Results:** Children in Group 1 were older (mean age 10.8 $\pm$ 3.2 years vs 8.5 $\pm$ 2.8 years,  $p < 0.001$ ) and had increased body mass index (20.3 $\pm$ 5.6 kg/m<sup>2</sup> vs 18.4 $\pm$ 3.5 kg/m<sup>2</sup>,  $p < 0.001$ ). Systolic blood pressure was also increased in Group 1 (118.9 $\pm$ 10.4 vs 114.1 $\pm$ 8.8 mmHg,  $p < 0.001$ ). Systolic and diastolic left ventricular functional indices using conventional and tissue Doppler modalities were comparable between groups. Left ventricular mass indexed for body surface area was also similar between groups (Group 1: 50.7 $\pm$ 10.8 vs 48.5 $\pm$ 10.2 in Group 2,  $p = 0.138$ ). PWV was increased in Group 1 (5.02 $\pm$ 0.7 m/sec vs 4.7 $\pm$ 0.6,  $p < 0.001$ ) whereas carotid IMT was similar between groups ( $p = 0.902$ ). The vascular changes between groups were abolished when analysis was accounted for differences in age, body mass index and blood pressure. Cholesterol levels and inflammatory markers were also comparable between groups.

**Conclusions:** This study demonstrates that there is no evidence of cardiovascular dysfunction in children up to one year following COVID-19 infection when comparison was made with children who have not been infected by SARS-CoV-2. These data contradict findings reported in adults and support the milder phenotype of COVID-19 infection in children.

**Keywords:** COVID-19 infection, children, cardiac involvement, echocardiography, SARS-CoV-2 IgG antibodies, PWV

#### P-175

##### **A rare case of abnormal origin of the left circumflex coronary artery from the right branch pulmonary artery**

Mohamed Metwally Abdelal<sup>1</sup>, Mustafa Hassan Alqabandi<sup>2</sup>

<sup>1</sup>Mohamed Abdelal Pediatric cardiology department chest Disease Hospital; <sup>2</sup>Mustafa AlQabandi Pediatric cardiology department Chest Disease Hospital

**Background and Aim:** We are reporting a case of 5 years old. Healthy and a symptomatic. He had a previous echo in which a coronary fistula from left anterior descending to the apex of left ventricle. The left ventricular function was normal, the left anterior descending coronary was dilated and there were no ECG changes.

**Method:** Repeated echo showed suspected left circumflex coronary from the right pulmonary artery branch. A cardiac catheterization was done (figure 1).

**Results:** Aortic root injection showed that the left circumflex coronary artery is filling retrogradely from the left anterior descending and draining at the right pulmonary artery branch. After discussion with the surgeon it was decided to re-implant the left circumflex to the aortic root.

**Conclusions:** Anomalous origin of the left circumflex from right pulmonary artery branch is a rare disease. Should be suspected if

the left circumflex was not seen clearly by echo. Confirmation by cardiac angiography is important before surgical repair.

**Keywords:** Anomalous coronary origin, Congenital heart disease, Case report



**Figure 1** Aortic root angiography showed Left circumflex artery filling retrogradely

#### P-176

##### **Early left atrial dysfunction by strain echocardiography in duchenne muscular dystrophy**

Jaroslav Meyer Szary<sup>1</sup>, Marcin Grytczuk<sup>2</sup>, Magdalena Bazgier<sup>1</sup>, Rafał Surmacz<sup>3</sup>, Marcin Pajkowski<sup>4</sup>, Joanna Kwiatkowska<sup>1</sup>, Waldemar Bobkowski<sup>3</sup>

<sup>1</sup>Department of Pediatric Cardiology and Congenital Heart Defects, Medical University of Gdańsk, Gdańsk, Poland; <sup>2</sup>Department of Neonatal Intensive Care and Congenital Malformations of the Neonates and Infants, Polish Mother's Memorial Hospital Research Institute, Łódź; <sup>3</sup>Department of Pediatric Cardiology, Nephrology and Hypertension, Poznań University of Medical Sciences, Poznań, Poland; <sup>4</sup>Department of Cardiac Diagnostics, Medical University of Gdańsk, Poland

**Background and Aim:** Duchenne muscular dystrophy (DMD) is a rare genetic disease. Dystrophin deficiency causes muscle dysfunction. With advances in respiratory support, cardiomyopathy related heart failure is becoming the leading cause of death. Currently, this patient group lacks suitable methods for early risk stratification aiding targeted cardiac prophylaxis and treatment in these patients. Our study aimed to analyze echocardiographic LA strain (LAS) indices in children with DMD.

**Method:** Design: multicenter retrospective case-control (1:1) observational study. DMD sample was collected from two pediatric clinical centers (Poznań, Gdańsk) based on genetically confirmed diagnosis. Age-matched healthy controls (CTR) were analyzed for comparison. Standard echocardiographic protocol and 2CV, 4CV LAS (zero strain reference at end-diastole) was used to acquire measurements. Data is given for DMD vs CTR respectively as mean  $\pm$ SD or median (min-max) appropriately for the

distribution. Statistical calculations facilitated appropriate parametric and non-parametric tests (Wizard 2.0.12).

**Results:** Forty-three cases and 44 controls were studied. There were no age ( $9.0 \pm 3.9$  vs  $9.0 \pm 3.9$  years,  $p = 0.966$ ), weight ( $30.0, 10.5\text{--}80.0$  vs  $32.7, 15.0\text{--}75.0$  cm,  $p = 0.403$ ), BMI ( $17.6, 12.8\text{--}31.2$  vs  $16.4, 13.2\text{--}26.0$  kg/m<sup>2</sup>,  $p = 0.169$ ) and BSA ( $1.0, 0.48\text{--}1.96$  vs  $1.1, 0.71\text{--}1.79$  m<sup>2</sup>,  $p = 0.522$ ) differences between the groups but they differed significantly in height ( $129 \pm 21.6$  vs  $141 \pm 25.2$  cm,  $p = 0.017$ ). The reservoir strain was significantly lower for DMD both in 4CV  $42.4\% \pm 17.3\%$  vs  $50.3\% \pm 15.6\%$ ,  $p = 0.010$  and 2CV  $48.2\% \pm 13.6\%$  vs  $55.1\% \pm 11.9\%$ ,  $p = 0.041$ . The conduit strain was significantly higher for DMD in 4CV  $-33.1\% \pm 14.7\%$  vs  $-38.5\% \pm 11.8\%$ ,  $p = 0.024$  but not in 2CV  $-36.9\% \pm 12.7\%$  vs  $-40.3\% \pm 10.4\%$ ,  $p = 0.127$ . Contractile strain was equal in 4CV  $-9.4\% \pm 5.0\%$  vs  $-11.8\% \pm 7.0\%$ ,  $p = 0.089$  but higher for DMD in 2CV  $-11.2\% \pm 6.6\%$  vs  $-14.8\% \pm 5.3\%$ ,  $p = 0.003$ .

Interestingly, we found that LA reservoir and conduit strain values are age-dependent in DMD patients (but not in controls). Consequently, in patients aged  $\geq 9$  years all LAS indices differed significantly between the groups.

**Conclusions:** This is first LA strain analysis in DMD patients. The LA strain differs in the DMD patient group and may be an early marker of cardiac dysfunction in these patients. Significant variability in the measurements was observed. Larger patient cohorts and longitudinal observational studies assessing the clinical value of LA strain analysis are necessary.

**Keywords:** left atrial strain, echocardiography, Duchenne Muscular dystrophy, cardiomyopathy, heart failure, risk stratification

## P-177

### Automated measurements based on artificial intelligence software in pediatric echocardiography: A feasibility study

Corina Maria Vasile<sup>1</sup>, Martina Avesani<sup>2</sup>, Camille Velly<sup>1</sup>, Camille Chan<sup>1</sup>, Xavier Paul Bouteiller<sup>3</sup>, Xavier Iriart<sup>1</sup>

<sup>1</sup>Department of Pediatric and Adult Congenital Cardiology, University Hospital of Bordeaux, France; <sup>2</sup>Department of Cardiac, Thoracic, Vascular, and Public Health Sciences, University of Padua, Padova, Italy; <sup>3</sup>IHU Liryc, Electrophysiology and Heart Modelling Institute, Bordeaux University Foundation, Pessac France

**Background and Aim:** Transthoracic echocardiography is the first-line non-invasive investigation for assessing pediatric patients' cardiac anatomy, physiology, and hemodynamics, based on its accessibility and portability, but complete anatomic and hemodynamic assessment is time-consuming. The study aimed to establish the feasibility, utility, and variability of automated software using artificial intelligence designed for adults in a cohort of pediatric patients.

**Method:** The study was carried out at the University Hospital of Bordeaux between August and September 2022 and included 45 patients with normal or near normal heart architecture who underwent a 2D TTE. We compared AI automatic measurements with manual measurements performed by a senior and a junior pediatric cardiologist. As the AI software has designed for adults, we decided to divide our patients into two subgroups: the first group (patients  $< 9$  years, 60%) and the second group (patients  $> 9$  years, 40%).

**Results:** The mean age of our patients at the time of evaluation was  $8.2 \text{ years} \pm 5.7$ , and the main reason for referral to our service was the presence of a heart murmur ( $n = 13$ ). Globally, we observed good Spearman correlations (Fig. 1) between two cardiologists ranging from 0.53 to 0.95, with only 2 values among 23 below

0.70 (FAC and EF). Whereas between cardiologists and software, correlations varied much more: from 0.00 to 0.96, with 19 among 46 values below 0.7. Bland Altman's analysis showed that globally there is a good agreement between both physicians with a mean error of 34.2% ( $< 9$  years old) and 33.5% ( $> 9$  years old), and the error value is lower than the 30% cut-off in respectively 13 and 14 traits among 23. When comparing software and senior physicians, the agreement is clearly better for the subgroup of patients over 9 years old (mean percentage of error of 43.3% and 51.0%). However, for 2 assessed parameters in the youngest group and 4 for the oldest, the error was below 30%.

**Conclusions:** In this preliminary study, we found that automated artificial intelligence software provided promising results for analyzing some of the main echocardiographic parameters, even for children.

**Keywords:** echocardiography, artificial intelligence, automated measurements, deep learning, pediatrics.

## P-178

### An echocardiographic score to predict central venous pressure in children with heart disease

Simone Bonetti<sup>1</sup>, Fiorentina Guida<sup>2</sup>, Rebecca Fogacci<sup>5</sup>, Maria Candelino<sup>4</sup>, Cristiano Conte<sup>3</sup>, Ambra Bulgarelli<sup>1</sup>, Tammam Hasan<sup>1</sup>, Rossana Zaroni<sup>1</sup>, Gaetano Domenico Gargiulo<sup>6</sup>, Andrea Donti<sup>1</sup>

<sup>1</sup>Pediatric Cardiology and Adult Congenital Heart Disease Program

Department of Cardio - Thoracic and Vascular Medicine IRCCS

Azienda Ospedaliero - Universitaria di Bologna, Italia (Italy);

<sup>2</sup>Specialization school in pediatrics, University of Bologna; <sup>3</sup>Specialization

school in pediatrics, University of Parma; <sup>4</sup>Specialization school in pedi-

iatrics, University of ChietiL; <sup>5</sup>Specialization school in anesthesiology and

resuscitation, University of Bologna; <sup>6</sup>Pediatric cardiac surgery Department

of Cardio - Thoracic and Vascular Medicine IRCCS Azienda

Ospedaliero - Universitaria di Bologna, Italia (Italy)

**Background and Aim:** Central Venous Pressure (CVP) invasive monitoring allows to directly evaluate haemodynamical changes in adult population with heart failure (HF), cardiogenic and obstructive shock. Nevertheless, evidence so far collected are not strong enough to support the use of the same parameters in children. We elaborated an echocardiographic model to indirectly estimate CVP in children using IVC diameters (IVCd), IVCd/Aortic diameters (Aod) ratio, and IVC collapsibility index (IVCCI) (i.e.: IVC maximal diameter - IVC minimum diameter)/IVC maximal diameter) based on pediatric z-scores recently published by Mannarino et al.

**Method:** we longitudinally evaluated children (0-18 years-old) with a diagnosis of chronic and acute HF, pulmonary hypertension, cardiogenic shock, and a biventricular cardiac physiology that accessed to our department from October 2021 to March 2022. CVP was invasively monitored due to cardiac surgery, heart transplantation and acute HF, without ventilatory assistance. IVCd, Aod, IVCd/Aod ratio, and IVCCI were simultaneously collected by 2-dimensional bed-side echocardiography.

**Results:** 80 children were enrolled. Table 1 shows the main echocardiographical results collected.

IVC diameter standardized per body surface area (BSA) ( $\pm 2$  SDS) showed a weak direct correlation to CVP ( $\rho = 0,36$ ;  $p = 0,001$ ). IVVCI standardized per BSA predicted CVP variation in patients with invasive CVP among 0-5 mmHg (sensitivity 96.5%) but it was inaccurate in patients with increased CVP. Univariate linear regression showed positive linear correlation between caval diameters, the cavo-aortic ratio, and CVP; while CVP and IVCCI showed a negative logarithmic relationship.

A predictive model was obtained through multiple linear regression ( $PVC = 1.8 \cdot IVC / AoSDS - 3, 9 \cdot \ln(IVCC) + 17, 8$ ) with a sensitivity of 85% and a simplified model was elaborated through the analysis of ROC curves:  $IVCC > 30\%$  and  $IVCmax/AoSDS < 1.55$  estimated a CVP between 0 and 5 mmHg;  $IVCCI < 30\%$  and  $IVCmax/AoSDS > 1.55$  estimated a CVP greater than 10 mmHg. Our simplified model showed high correlation with CVP measure (Spearman's rho 0.95, p value 0.000).

**Conclusions:** Our predictive model and simplified predictive model could be reliable tools to estimate CVP in children affected by HF, pulmonary hypertension, cardiogenic and obstructive shock. Further studies will be needed to confirm its validity.

**Keywords:** Predictive score, central venous pressure, cardiogenic shock, heart failure

**Table**

IVC minimal diameter (mean)	9,76 mm (+/- 4,61)		
IVC maximal diameter (mean)	12,33 mm (+/- 4,76)		
Aortic diameter (mean)	9,53 mm (+/- 2,36)		
IVCCI (mean)	23% (+/- 14)		
Thoracic aorta/IVC ratio (mean)	1,29 +/- 0,34		
CVP	0-5 mmHg	27	33,75%
	6-10 mmHg	34	42,5%
	11-15 mmHg	10	12,5%
	> 15 mmHg	9	11,25%
Correlation with CVP (linear regression)	Parameter	R2 corr	P value
	IVC min SDS	0,59	<0,001
	IVC max SDS	0,31	<0,001
	IVC max/ Ao SDS	0,79	<0,001
	IVCCI	0,68	<0,001

IVC, Inferior vena cava; IVCCI, (IVC maximal diameter - IVC minimum diameter)/ IVC maximal diameter; CVP, central venous pressure.

### P-179

#### Evaluation of natural mechanical wave velocities in children with pulmonary arterial hypertension

Matthew Henry<sup>1</sup>, Solveig Fadnes<sup>2</sup>, Kristian Sørensen<sup>4</sup>, Pablo Lopez<sup>5</sup>, Lasse Løvstakken<sup>2</sup>, Siri Ann Nyrnes<sup>3</sup>, Luc Mertens<sup>1</sup>

<sup>1</sup>Cardiology, The Hospital for Sick Children, Toronto, Canada;

<sup>2</sup>Department of Circulation and Medical Imaging, Norwegian University of Science and Technology (NTNU), Trondheim, Norway; <sup>3</sup>Children's Clinic, St. Olav's Hospital, Trondheim University Hospital, Norway;

<sup>4</sup>Division of Ålesund Hospital, Department of Pediatrics, Møre and Romsdal Hospital Trust; <sup>5</sup>Hospital Universitario Puerta de Hierro Majadahonda, Majadahonda, Spain

**Background and Aim:** Pulmonary arterial hypertension (PAH) is characterized by increased RV afterload leading to myocardial adaptation including interstitial fibrosis. There is growing evidence that fibrosis, and the resulting increase in myocardial stiffness play an important role in the development of right heart failure and may be used as an important prognostic factor. Myocardial stiffness has been non-invasively estimated using high frame rate imaging by quantifying the velocity of mechanical waves which relates to stiffness. These waves may be induced by valve closure, or during the atrial kick which is thought to reflect differences in end diastolic

stiffness. Our aim was to compare the mechanical wave velocity during atrial kick in children with pulmonary arterial hypertension with those in healthy children.

**Method:** Mechanical wave imaging was prospectively performed in children with PAH and controls with normal hearts using a commercially available clinical scanner with research software enabling high frame rate imaging. The interventricular septum was imaged in the 4 chamber view and mechanical wave velocity induced by the atrial kick (AK) was measured offline using custom made in-house software. Clinical data was obtained using conventional ultrasound as per institutional standard of care.

**Results:** Data from 28 children were analyzed (11 PAH, 17 controls) with no significant difference in age (9.5 years (7.4-16.7) vs 9.5 years (5.7 - 11.6) p = 0.61); height (133.5 cm (116.2-160.5) vs 146.3 cm (121.9-152.3) p = 0.86, weight (27.9 kg (20.8- 63.4) vs 32.4 kg (23.0- 49.2) p = 0.96) or interventricular septal thickness (0.62 cm (0.51-0.75) vs 0.63 cm (0.51- 0.67) p = 0.91). Patients with PAH showed a significant increase in atrial kick velocity when compared to controls (1.6 m/s (1.3-2.0) vs 1.1 m/s (1.0- 1.5) p = 0.003). When related to LV diastolic function assessed with LA strain, AK velocity significantly correlated to left atrial pump function (r = 0.81, p = 0.02) but not reservoir or conduit function.

**Conclusions:** Natural mechanical wave imaging is able to detect differences in wave velocities between children with PAH vs healthy controls and could be helpful in the surveillance and prognosis of pulmonary arterial hypertension.

**Keywords:** mechanical wave imaging, natural shear wave, myocardial stiffness, pulmonary arterial hypertension, high frame rate imaging,

### P-180

#### Beyond aortopathy: reduced left atrial strain and diastolic dysfunction in pediatric loeys-dietz and marfan patients

Alison Howell, Nairy Khodabakhshian, Wei Hui, Cameron Slorach  
Department of Cardiology, The Hospital For Sick Children, Toronto, Ontario, Canada

**Background and Aim:** Reports of cardiomyopathy and observed myocardial fibrosis in Loeys-Dietz (LDS) and Marfan's (MFS) suggest heart function beyond measured ejection fraction warrants closer observation. Left Atrial (LA) strain represents a novel approach to diagnosing diastolic dysfunction in Pediatrics.

**Method:** This is a single center retrospective cohort study of LDS & MFS patients from 2006 - 2020. Chart review identified clinical parameters. Coupled Cardiac MRI and Echocardiogram's were reviewed and comprehensive diastolic assessment performed and compared to matched controls (N = 32).

**Results:** We identified 37 LDS and 39 MFS patients; median overall age 15 years (IQR 6), 49.5% male. Mitral regurgitation  $\geq$  moderate was present in 2 LDS and 5 MFS. Mitral valve prolapse in 5 LDS and 23 MFS. Nine LDS and 5 MFS had previous aortic root replacements. No difference in left atrial or ventricular dimensions or heart rate (p = 0.6) between the LDS/MFS and controls.

Both LDS & MFS groups had significantly lower LA reservoir strain (LDS 34% (IQR13), MFS 32% (IQR 12) vs 42% (IQR15), p=0.001, <0.001), conduit strain (LDS 25% (IQR14), MFS 25% (IQR8.5) vs 35% (IQR1), p=0.002, p<0.001), peak strain rate (LDS 1.0 1/s (IQR0.4), MFS 1.0 (IQR0.5) vs 1.4 (IQR0.5), p= 0.004, p=0.001) and conduit strain rate (LDS 2.1 (IQR0.9), MFS 2.0 (IQR0.9) vs 3.01 1/s (IQR0.9), p = 0.002, p < 0.001).

Other diastolic parameters were significantly lower; mitral valve E (LDS 76±25, MFS 78±25.6 vs 66±13 cm/sec, p = 0.023, p 0.001),

lateral E' (LDS 16±6, MFS 15±5 vs 17.8±4 cm/sec,  $p = 0.335$ ,  $p = 0.007$ ), mitral valve E/E' (LDS 5±1.6, MFS 2±1 vs 14±10 cm/sec,  $p < 0.001$ ,  $p < 0.001$ ).

Reduced LA reservoir strain and strain rate were significantly correlated with reduced left ventricular strain ( $r = -0.602$ ,  $p < 0.001$  and  $r = -0.462$ ,  $p = 0.006$  respectively). There was no significant reduction in LA strain in 16/40 patients identified with fibrosis on T1 mapping. LDS patients receiving an angiotensin receptor blocker (ARB) showed a trend of improved LA reservoir function (LA reservoir strain: 40±17 Vs. 32±7%,  $p = 0.059$ . LA reservoir strain rate: 1.3±0.6 Vs. 1.1±0.2 1/s).

**Conclusions:** Pediatric LDS patients have reduced LA strain and lower diastolic function. Further prospective studies are warranted.

**Keywords:** Left Atrial Strain, Loeys-Dietz, Marfans, Diastolic function

### P-182

#### Investigating the prevalence of the patent ductus arteriosus in term neonates

Anton Friis Mariager<sup>1</sup>, Alberte Hammeken<sup>3</sup>, Mikkel Malham<sup>1</sup>, Anna Axelsson Raja<sup>3</sup>, Anna Selmer<sup>4</sup>, Raheel Altaf Raja<sup>5</sup>, Johan Navne<sup>5</sup>, Anne Sophie Sillesen<sup>2</sup>, Henning Bundgaard<sup>6</sup>, Kasper Iversen<sup>2</sup>, Ester Garne<sup>7</sup>, Dorthe Lisbeth Jeppesen<sup>1</sup>

<sup>1</sup>Department of Pediatrics and Adolescent Medicine, Copenhagen University Hospital - Amager and Hvidovre, Hvidovre, Denmark.;

<sup>2</sup>Department of Cardiology, Copenhagen University Hospital - Herlev Hospital, Herlev, Denmark.; <sup>3</sup>Department of Clinical Medicine, Faculty of Health and Medical Sciences, University of Copenhagen, Copenhagen, Denmark.; <sup>4</sup>Department of Pediatrics and Adolescent Medicine Aarhus University Hospital, Denmark; <sup>5</sup>Department of Pediatrics and Adolescents, Copenhagen University Hospital - Rigshospitalet, Copenhagen, Denmark; <sup>6</sup>The Capital Region's Unit of Inherited Cardiac Diseases, Copenhagen University Hospital - Rigshospitalet, Copenhagen, Denmark; <sup>7</sup>Department of Pediatrics and Adolescent Medicine, Lillebaelt Hospital, University Hospital of Southern Denmark, Kolding, Denmark

**Background and Aim:** The ductus arteriosus (DA) is a part of the fetal circulation. Normally the DA close in the neonatal cardiac transition period immediately after birth, but for some neonates the closure is delayed. Though a delayed closure is thought to be a common phenomenon, little is known about the prevalence of open DA and the timing of the spontaneous closure in term born children. We aim to investigate how often an open DA will develop into a persisting ductus arteriosus (PDA). Furthermore, we aim to assess associated factor with the delayed closure of DA based on echocardiographic variables and maternal factors.

**Method:** Echocardiograms were prospectively collected in the Copenhagen Baby Heart Study, a prospective cohort study with 24.522 echocardiographic examination in a database. All examinations made at baseline were evaluated to discover an open ductus arteriosus ( $n = 561$ ). Families to a child where an open DA was found at baseline examination was contacted by telephone and offered a follow-up examination. At initial contact, parents will be asked if the children have been treated for or still are being followed by a paediatric cardiologist for a PDA. Children with no known PDA, will be invited to a follow-up echocardiogram. A positive PDA will be defined by a retrograde jet in the parasternal SAX projection. The child with a PDA will be referred to a paediatric department to a clinical follow up.

In general, baseline data on maternal and obstetric factors will be extracted from the Copenhagen baby heart database. This will enable us to assess pre-, peri- and post-natal risk factors for developing a PDA.

**Results:** The project is ongoing. As for now we have contacted and included 52% ( $n = 294$ ). The inclusion-goal is 80%. Until now twenty-seven children out of 294 with a early open DA have been found with a PDA (8%). This is not adjusting for a small subpopulation of preterm born children.

**Conclusions:** Our preliminary result suggests a measurable prevalence of PDA in our population. A more reliably result will follow as we reach our inclusion criteria and statistically adjust for gestational age.

**Keywords:** Congenital Heart Defects (CHD), Ductus Arteriosus, Patent (PDA), Ductus Arteriosus, echocardiography, neonatology.

### P-183

#### Prevalence and time of closure of open ductus arteriosus in term born neonates

Anton Friis Mariager<sup>1</sup>, Alberte Hammeken<sup>3</sup>, Mikkel Malham<sup>1</sup>, Anna Axelsson Raja<sup>3</sup>, Anna Selmer<sup>4</sup>, Raheel Altaf Raja<sup>5</sup>, Johan Navne<sup>5</sup>, Anne Sophie Sillesen<sup>2</sup>, Henning Bundgaard<sup>6</sup>, Kasper Iversen<sup>2</sup>, Ester Garne<sup>7</sup>, Dorthe Lisbeth Jeppesen<sup>1</sup>

<sup>1</sup>Department of Pediatrics and Adolescent Medicine, Copenhagen University Hospital - Amager and Hvidovre, Hvidovre, Denmark.;

<sup>2</sup>Department of Cardiology, Copenhagen University Hospital - Herlev Hospital, Herlev, Denmark.; <sup>3</sup>Department of Clinical Medicine, Faculty of Health and Medical Sciences, University of Copenhagen, Copenhagen, Denmark.; <sup>4</sup>Department of Pediatrics and Adolescent Medicine Aarhus University Hospital, Denmark; <sup>5</sup>Department of Pediatrics and Adolescents, Copenhagen University Hospital - Rigshospitalet, Copenhagen, Denmark; <sup>6</sup>The Capital Region's Unit of Inherited Cardiac Diseases, Copenhagen University Hospital - Rigshospitalet, Copenhagen, Denmark; <sup>7</sup>Department of Pediatrics and Adolescent Medicine, Lillebaelt Hospital, University Hospital of Southern Denmark, Kolding, Denmark

**Background and Aim:** The ductus arteriosus (DA) is a part of the fetal circulation. Normally the DA close in the neonatal cardiac transition period immediately after birth, but for some neonates the closure is delayed. Though a delayed closure is thought to be a common phenomenon, little is known about the prevalence of open DA and the timing of the spontaneous closure in term born children. The aim of this study was to evaluate the prevalence of an open DA in term born neonates within the first 28 days after birth. **Method:** Echocardiograms were prospectively collected in the Copenhagen Baby Heart Study with just over 25.000 examination in the database. The present study included term born neonates with an echocardiogram performed in the neonatal period up to 28 days after birth. Neonates with findings of other congenital heart defects than atrial septal defects were excluded. All echocardiograms were analyzed to diagnose an open DA.

**Results:** A total of 21, 649 neonates were included in this study. The median age at examination was 11 days (IQR = 4–18). In 485 neonates, an open DA (2.3%) were identified. In those examined at day zero ( $n = 130$ ), day two ( $n = 1090$ ), and seven ( $n = 1080$ ), an open DA were found in 36%, 8% and 0.6%, respectively. After day seven the prevalence of an open DA was stable around 0.6%.

**Conclusions:** This large-scale echocardiography study in healthy neonates showed a high prevalence of open DA in the first day of life, with a rapid decline to a prevalence of less than 1% at day seven after birth. After day seven the prevalence of DA remained stable at around 0.6%, suggesting spontaneous closure after day seven is less likely.

**Keywords:** Congenital Heart Defects (CHD), Ductus Arteriosus, Patent (PDA), Ductus Arteriosus, echocardiography, neonatology

## P-184

**Impact of the maternal body mass index on the structure and function of the heart in newborns**

Mette Marie Olsen Noerregaard<sup>1</sup>, Saima Basit<sup>2</sup>, Anna Axelsson Raja<sup>3</sup>, Anne Sophie Sillesen<sup>1</sup>, Finn Stener Joergensen<sup>4</sup>, Kasper Iversen<sup>1</sup>, Heather Boyd<sup>2</sup>, Henning Bundgaard<sup>3</sup>, Otilia Voegg<sup>1</sup>

<sup>1</sup>Department of Cardiology, Herlev Hospital, Herlev, Denmark;

<sup>2</sup>Department of Epidemiological Research, Statens Serum Institut,

Copenhagen, Denmark; <sup>3</sup>Department of Cardiology, The Heart Centre,

Rigshospitalet, Copenhagen, Denmark; <sup>4</sup>Department of Obstetrics and Gynaecology, Hvidovre Hospital, Hvidovre, Denmark

**Background and Aim:** Maternal obesity has previously been associated with an increased risk of complex congenital heart defects in the offspring. Whether maternal obesity may also cause subtle abnormalities in infant cardiac structure and function is unknown. Our aim was to investigate whether different measurements of maternal body mass index (BMI) were associated with changes in left ventricle (LV) dimensions in the newborn child.

**Method:** Data was obtained as part of a population-based cohort study with prenatal inclusion and postnatal transthoracic echocardiography (TTE) of newborns within 60 days of birth. The TTE protocol included measurements of LV internal diameter at end-diastole (LVIDd), and LV internal diameter at end-systole (LVIDs) obtained from echocardiographic images from the parasternal long axis view. LV end-diastolic volume (EDV) and LV end-systolic volume (ESV) were calculated from LVIDd and LVIDs respectively, by use of Teicholz formula.

We included newborns with a gestational age at birth  $\geq 37$  weeks. The newborns were divided into subgroups depending on the pre-pregnancy BMI of their mothers in  $\text{kg}/\text{m}^2$ :  $\leq 18.4$ , 18.5–24.9 (reference group), 25.0–29.9, 30.0–34.9 and  $\geq 35.0$ . Association between maternal BMI and infant LV parameters were analysed by use of an adjusted linear mixed model.

**Results:** 24, 222 newborns were included in the study. The results reveal a tendency toward smaller measures of LVIDd in newborns to mothers with a BMI  $\geq 25.0$ , reaching statistical significance for maternal BMI 25.0–29.9 [ $-0.05 \pm 0.02$ ,  $p = 0.03$ ] and 30–34.9 [ $-0.10 \pm 0.04$ ,  $p = 0.02$ ]. LVIDs measures were significantly smaller for maternal BMI 25.0–29.9 [ $-0.05 \pm 0.02$ ,  $p = 0.02$ ], BMI 30.0–34.9 [ $-0.11 \pm 0.03$ ,  $p = 0.001$ ] and BMI  $\geq 35.0$  [ $-0.10 \pm 0.05$ ,  $p = 0.04$ ]. We saw a tendency toward smaller measures of EDV in newborns to mothers with a BMI  $\geq 25.0$ , reaching statistical significance for BMI 25.0–29.9 [ $-0.09 \pm 0.04$ ,  $p = 0.03$ ] and BMI 30.0–34.9 [ $-0.15 \pm 0.07$ ,  $p = 0.02$ ]. ESV was significantly smaller for maternal BMI 25.0–29.9 [ $-0.04 \pm 0.02$ ,  $p = 0.03$ ] and BMI 30.0–34.9 [ $-0.09 \pm 0.03$ ,  $p = 0.003$ ].

**Conclusions:** Systematic, population-based echocardiography of newborns showed that a maternal BMI  $\geq 25 \text{ kg}/\text{m}^2$  was associated with a pattern of smaller LV diameters and internal LV volumes in the offspring. The long-term effects of these novel findings are unknown.

**Keywords:** maternal risk factors, the newborn heart, epidemiology, echocardiography, cardiac structure and function

## P-185

**A rare truncus variant with pulmonary dominance and ascending aorta as a side branch visualized using open source software**

Bas Rebel<sup>1</sup>, Thomas Krasemann<sup>1</sup>, Ad Bogers<sup>2</sup>, Ingrid Van Beynum<sup>1</sup>

<sup>1</sup>Department of Paediatric Cardiology, Sophia Children's Hospital, Erasmus MC Rotterdam, Netherlands; <sup>2</sup>Department of Thoracic Surgery, Erasmus MC Rotterdam, Netherlands

**Background and Aim:** Pulmonary dominance is rare in truncus arteriosus communis. Segmentation of the imaging enables identification of different structures in a visual way.

**Method:** Case report. Antenatally diagnosed dextrocardia, situs solitus, normal systemic and pulmonary venous drainage, large perimembranous ventricular septal defect. The right ventricle gave rise to a common arterial trunc (CAT), and pulmonary arteries appeared to originate from this truncus. The ascending aorta was difficult to visualize and interruption of the arch was considered.

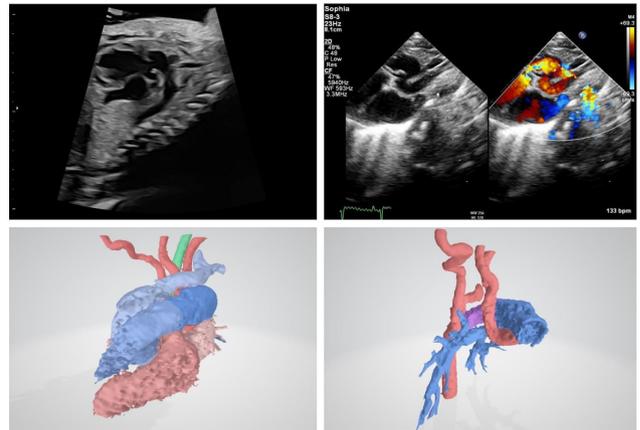
**Results:** Postnatal 2D and 3D echocardiography and a computed tomography scan with 3D reconstruction (open source software 3D Slicer 4.10.2 r28257) was performed. Echocardiography and CT scan showed a common arterial trunc completely arising from the relatively small right ventricle. The hypoplastic ascending aorta branching posteriorly from the truncus gave rise to both carotid arteries and the coronary arteries. The pulmonic branches originated from the posterior site of the trunc. A persistent arterial duct connected to the descending aorta which gave rise to both subclavian arteries (Lusoria).

Based on the clear anatomical findings combined with dysmaturity, treatment was abandoned.

**Conclusions:** Segmentation of complex congenital heart disease using open source software allows clear visualization of anatomical details. This helps to determine possible treatment strategies.

**Keywords:** Truncus, 3D segmentation, open source software

Segmented complex truncus



Echocardiographical images in comparison with 3D segmented CT

## P-186

**Univentricular heart in human heterotaxy syndrome: an anatomic study.**

Lucile Houyel<sup>1</sup>, Bettina Bessières<sup>2</sup>, Manon Hily<sup>1</sup>, Marie Gonzales<sup>2</sup>, Naima Talhi<sup>3</sup>, Mathilde Lambert<sup>4</sup>, Damien Bonnet<sup>1</sup>

<sup>1</sup>Medico-surgical Unit of Congenital and Pediatric Cardiology, Necker Hospital - M3C, Université Paris Cité, Paris, France; <sup>2</sup>Fetal pathology Unit, Necker Hospital, Paris, France; <sup>3</sup>Fetal Pathology Unit, Centre Hospitalier Intercommunal, Créteil, France; <sup>4</sup>Fetal Pathology Unit, Trousseau Hospital, Paris, France

**Background and Aim:** Heterotaxy corresponds to abnormal symmetry of bronchi and pectinate muscles (isomerism) and/or situs

discordance between visceral organs. Complex congenital heart defects are frequently associated, with a significant proportion of univentricular hearts (UVH). The aim of this study was to evaluate, in a cohort of fetal human heart specimens with heterotaxy, the incidence of UVH, and to describe morphologic particularities of UVH.

**Method:** Among the anatomic fetal hearts collection of the French Centre of Reference for complex CHD at Necker Hospital, we analyzed 85 heart specimens with heterotaxy, divided in right or left isomerism (RI, LI) according to the extent of the pectinate muscles within the atria or to the morphology of the bronchi. Among them, 31 had RI, 52 LI, 2 could not be classified as RI or LI. A ventricle was considered hypoplastic when it did not extend to the apex of the heart.

**Results:** UVH was found in 45/83 specimens (54.2%), 21/31 RI, 24/52 LI (68% vs 46%,  $p = ns$ ). It was morphologically right in 32 (hypoplastic left ventricle (LV)), left in 13 (71% vs 28.9%). Hypoplastic LV tended to be more frequent in RI than in LI (51.6% vs 30.8%,  $p = 0.06$ ), while the incidence of hypoplastic right ventricle was not different in RI and LI (16.1% vs 15.3%,  $p = ns$ ). Hypoplastic left heart syndrome (HLHS), defined as extreme LV hypoplasia with atrioventricular valve atresia or extreme hypoplasia, and atresia or severe stenosis of the arterial valve originating from the LV, was more frequent in RI than in LI (22.6% vs 5.8%,  $p < 0.04$ ). All UVH with RI but one had a common atrioventricular junction with complete atrioventricular canal (95%), vs 75% of UVH with LI.

**Conclusions:** UVH is present in more than half of fetuses with heterotaxy. Morphologically right UVH (hypoplastic LV) is twice more frequent than morphologically left UVH, mainly found in RI. The association of HLHS with heterotaxy is not rare (10/83) and is more frequent in RI. These results underline the severity of cardiac malformations associated with heterotaxy, and could explain the worse prognosis after Fontan-type surgery in these patients.

**Keywords:** Heterotaxy, Univentricular heart, hypoplastic left heart syndrome, isomerism

## P-187

### Multidisciplinary management in children with marfan syndrome and related disorders: why a dedicated marfan unit can make a difference?

Giovanni Parlapiano<sup>1</sup>, Monia Magliozzi<sup>2</sup>, Viola Alesi<sup>2</sup>, Michelina Armando<sup>3</sup>, Antonino Romanzo<sup>4</sup>, Gaetano Zinzanella<sup>4</sup>, Federica Cali<sup>1</sup>, Lorenzo De Luca<sup>1</sup>, Giulio Calcagni<sup>1</sup>, Benedetta Leonardi<sup>1</sup>, Gianluigi Perri<sup>1</sup>, Alessio Franceschini<sup>1</sup>, Lorenzo Galletti<sup>1</sup>, Fabrizio Drago<sup>1</sup>, Antonio Novelli<sup>2</sup>, Anwar Baban<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology and Cardiac Surgery, Heart and Lung Transplantation, Bambino Gesù Children Hospital and Research Institute, IRCCS, 00165 Rome, Italy, The European Reference Network for Rare, Low Prevalence and Complex Diseases of the Heart-ERN GUARD-Heart; <sup>2</sup>Laboratory of Medical Genetics, Bambino Gesù Children Hospital and Research Institute, IRCCS, 00165 Rome, Italy; <sup>3</sup>Department of Neuroscience and Neurorehabilitation, Bambino Gesù Children Hospital and Research Institute, IRCCS, 00165 Rome, Italy; <sup>4</sup>Ophthalmology Unit, Bambino Gesù Children Hospital and Research Institute, IRCCS, 00165 Rome, Italy

**Background and Aim:** Marfan Syndrome (MFS) is an autosomal dominant (AD) multisystemic condition characterized by variable multiorgan involvement. The main signs are generally cardiac, ocular, and skeletal, but any other organ or system with connective tissue background can be affected. This disorder has typically an age-related penetrance. The pediatric population of MFS

undoubtedly represents a real challenge for clinicians not only in terms of diagnosis but also in terms of management. In fact, different data in literature are available regarding MFS adult population. However, very few studies are reported regarding multisystemic involvement of MFS in children.

**Method:** Our tertiary academic pediatric center represents the referral point for the center and south of Italy for the multidisciplinary management of patients with MFS. In this study we include data from 43 patients with MFS who had multisystemic clinical assessment, multiorgan screening and genetic study through NGS sequencing, MLPA analysis and microarray analysis.

**Results:** We focused on MFS manifestations in children, reporting the main multisystemic signs and symptoms to be considered at diagnosis and during follow up. Cardiac manifestations is included in a comprehensive way both at structural, arrhythmic and myocardial level.

**Conclusions:** In the management of MFS pediatric population, it is important to consider not only the major signs classically associated with the disease such as aortic, ocular and skeletal abnormalities, but also the minor aspects that can have a significant progressive impact on the health in children. We emphasize the importance of the multidisciplinary and personalized approach to children with suspected or confirmed MFS. Early MFS diagnosis at multi-systemic level can prevent acute and chronic complications, offer tailored and improve the quality of life.

Cardiogenetic Center in Bambino Bambino Gesù Children Hospital and Research Institute is part of the referral center for Marfan Syndrome in Children (<https://www.salutelazio.it/malattie-rare-lazio>)

**Keywords:** Marfan syndrome, aortic root dilation, age-related penetrance, early diagnosis, multidisciplinary management, personalized approach

## P-188

### Treatment and outcome in patients with congenital idiopathic dilatation of the right or left atrium. a report of seven cases

Nicolas Marcel Forderer<sup>1</sup>, Joerg Michel<sup>2</sup>, Alexej Bobrowski<sup>4</sup>, Hakan Akintürk<sup>1</sup>, Christian Schlensak<sup>3</sup>, Migdat Mustafi<sup>3</sup>, Brigitte Stiller<sup>4</sup>, Michael Hofbeck<sup>2</sup>

<sup>1</sup>Department of pediatric Cardiology, Pediatric Heart Center, University of Giessen, Germany; <sup>2</sup>Department of Pediatric Cardiology, University Children's Hospital Tuebingen, Germany; <sup>3</sup>Department of Thoracic and Cardiovascular Surgery, University Hospital Tuebingen, Germany; <sup>4</sup>Department of Congenital Heart Disease and Pediatric Cardiology, University Heart Center Freiburg, University of Freiburg, Germany

**Background and Aim:** Idiopathic dilatation of the right (IDRA) or left atrium (IDLA) is a rare congenital malformation. There are no established guidelines for the management of this disease. It is defined by an isolated dilatation of the right or left atrium in absence of primary cause. This condition is associated with significant morbidity and mortality. We report on 7 cases of IAD.

**Method:** Data of patients with IDRA or IDLA were retrieved from the databases of 3 tertiary referral centers for pediatric cardiology. **Results:** The patients were admitted between July 2008 and July 2021. We identified 5 patients with IDRA and 2 with IDLA. 1 patient revealed additional left atrial dilatation after surgical reduction of IDRA. Patients presented at a median age of  $4 \pm 1487$  days. 3/7 were female. 5 patients were detected by prenatal ultrasound. 1 patient presented at the age of 18 years with recurrent atrial tachycardia and 1 patient with a heart murmur at the age of 10 years.

Rhythm disturbances were present in 3 patients. 5/7 patients received atrial reduction plasty (median age  $0.48 \pm 4.9$  years). 2 patients required second operations due to a demasked left atrial aneurysm (at the age of 1.4 years, 7 days after the first operation) and recurrent progressive dilatation (at the age 2.6 years, 24 months after the first operation). 1/5 patient showed SVTs 10 months after the operation and received an antiarrhythmic treatment with propranolol and propafenone.

During a mean follow-up period of  $10.38 \pm 6, 83$  years, the following complications were observed: SVT necessitating medication, ( $n = 1$ ), tricuspid insufficiency ( $n = 3$ ), mitral insufficiency ( $n = 1$ ), thrombo-embolism/stroke ( $n = 1$ ). 2 patients needed a second operation due to demasked second aneurysm and re-dilatation, respectively. Histology (2 patients) showed thin-walled fibrotic atrial tissue with scares myocytes.

**Conclusions:** In our cohort of 7 patients, 5 underwent atrial reduction surgery. Complications (rhythm disturbances, re-dilatation, re-operation, tricuspid insufficiency, mitral insufficiency, stroke) are prevalent and warrant long-term follow-up. Seldom, both atria can be affected. 2 patients had no clinical symptoms so far. Due to the low risk of surgical procedures prophylactic surgical resection should be recommended in asymptomatic patients to prevent future complications.

**Keywords:** idiopathic dilatation of the right atrium, idiopathic dilatation of the left atrium, atrial aneurysm, giant right atrium,

#### P-189

##### **Mitral insufficiency in adolescents and older children after rheumatic disease: two clinical observations from cameroon.**

Tony Nengom Jocelyn<sup>1</sup>, Essomba Aurore<sup>2</sup>, Sap Ngo Um Suzanne<sup>1</sup>, Epee Ngoue Jeannette<sup>2</sup>, Koki Ndombo Paul Olivier<sup>1</sup>, Chelo David<sup>1</sup>  
<sup>1</sup>Mother and Child Centre of the Chantal Biya Foundation, Yaounde Cameroon; Department of Pediatrics, Faculty of Medicine and Biomedical Sciences, University of Yaounde I, Cameroon; <sup>2</sup>Department of Pediatrics, Faculty of Medicine and Biomedical Sciences, University of Yaounde I, Cameroon

**Background and Aim:** Mitral insufficiency is the most frequently encountered post-rheumatic valve disease. We report here the cases of two severe post rheumatic mitral insufficiency in an 11 year old adolescent who was successfully operated on and a 6 year old child of recent discovery.

**Method:** clinical cases

**Results:** Case 1:

An 11 year old adolescent, brought in for a progressive exacerbation of respiratory difficulty evolving for several days without fever associated with abdominal distension, and oedema evolving for several months, without hourly predominance, without variation in hourly diuresis. The clinical examination on admission revealed a global cardiac insufficiency syndrome and a 3/6 systolic murmur at the mitral focus. Cardiac echography revealed severe mitral regurgitation. He was put on furosemide, spironolactone and captopril. The indication for surgery was retained: mitral plasty with placement of a Carpentier Edwards mitral ring. He was treated surgically three months later.

Case 2:

Large 6 year old female child with a history of 10 days of non-anginal precordialgia associated with rapidly progressing respiratory

distress 5 days later, prompting the consultation. With a history of febrile polyarthralgias and repeated angina in whom the clinical examination showed signs of global cardiac insufficiency, justifying diuretics. The echocardiography, C-Reactive Protein and Anti streptolysin O antigens were strongly positive, leading to the conclusion of a severe post-rheumatic mitral insufficiency. She is awaiting surgery and is cardiac stable.

**Conclusions:** Rheumatic fever is a major public health problem in developing countries. Primary prevention is the key to treatment. It is therefore important to raise awareness in the medical and paramedical community about the optimal management of Streptococcal infections.

**Keywords:** rhumatic heart disease, children, cameroon

#### P-190

##### **Local arterial stiffness measured by ultrafast ultrasound imaging in pediatric cancer survivors treated with anthracyclines**

Rahna Rasouli<sup>1</sup>, Jerome Baranger<sup>1</sup>, Cameron Slorach<sup>1</sup>, Maelys Venet<sup>1</sup>, Minh Nguyen<sup>1</sup>, Matthew Henry<sup>1</sup>, Josh Gopaul<sup>1</sup>, Paul Nathan<sup>2</sup>, Wei Hui<sup>1</sup>, Luc Mertens<sup>1</sup>, Olivier Villemain<sup>1</sup>

<sup>1</sup>Division of Cardiology, The Hospital for Sick Children, Toronto, Canada; <sup>2</sup>Division of Hematology Oncology, The Hospital for Sick Children, Toronto, Canada

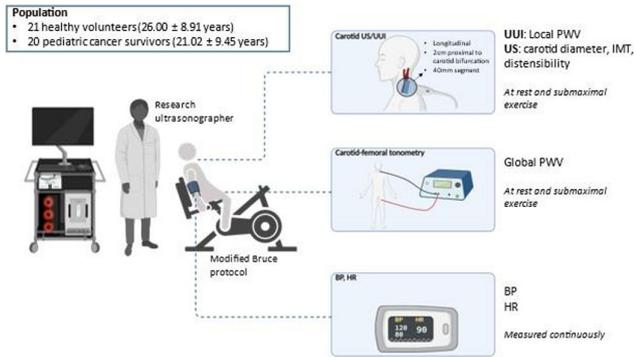
**Background and Aim:** There is conflicting literature regarding the long-term effect of anthracycline treatment on arterial stiffness. This study determined local arterial stiffness using ultrafast ultrasound imaging (UUI) in anthracycline treated pediatric cancer survivors for the first time, at rest and during exercise.

**Method:** 20 pediatric cancer survivors ( $21.02 \pm 9.45$  years) and 21 healthy controls ( $26.00 \pm 8.91$  years) were included. Participants completed a demographic survey, fasting bloodwork for cardiovascular biomarkers, and an exercise trial. Pulse wave velocity (PWV) was measured in the left common carotid artery by direct pulse wave imaging using UUI at rest and submaximal exercise. Both PWV at the systolic foot (PWV-SF) and dirotic notch (PWV-DN) were measured. Central (aortic) PWV was obtained by applanation tonometry. Other carotid measurements were taken by conventional ultrasound. Measures were compared using two-tailed Students t-test or Chi-squared test, as appropriate.

**Results:** There was no statistically significant difference ( $p > 0.05$ ) between pediatric cancer survivors and healthy controls in demographic parameters (age, sex, weight, height, BMI), blood biomarkers (total cholesterol, triglycerides, LDL-c, HDL-c, hs-CRP, fasting glucose, insulin, Hb A1c), cardiovascular parameters (IMT, SBP, DBP, HR, carotid diameters, distensibility) or PWV measured by UUI at rest or at exercise. There was also no difference in the cardiovascular adaptation between rest and exercise in the two groups ( $p > 0.05$ ). Multivariate analysis revealed age ( $p = 0.024$ ) and LDL-c ( $p = 0.019$ ) to be significant correlates of PWV-SF, in line with previously published data.

**Conclusions:** We did not identify any long-term impact of anthracycline treatment in pediatric cancers on local arterial stiffness in the left common carotid artery as measured by UUI. Additional larger cohort studies are needed to confirm our observations.

**Keywords:** Ultrafast ultrasound imaging, Arterial stiffness, Pulse wave velocity, Anthracyclines, Pediatric cancer survivor, Cardiotoxicity



## Methods

### P-191

#### A rare cause of paradoxical embolization in a child with operated sinus venosus atrial septal defect

Mario Giordano, Gianpiero Gaio, Raffaella Marzullo, Maurizio Cappelli Bigazzi, Fiorella Fratta, Raffaella Esposito, Maria Teresa Palladino, Maria Giovanna Russo

Department of Paediatric Cardiology, University of Campania "Luigi Vanvitelli", Monaldi Hospital, Naples, Italy

**Background and Aim:** Right-to-left shunt may be cause of paradoxical embolization (and cryptogenetic cerebral ischemic events). Patent foramen ovale and pulmonary arterio-venous malformations are the most common cause of paradoxical embolization; however, some rare cardiovascular lesions may cause a right-to-left shunt with consequent paradoxical embolization.

**Method:** We describe a case of young child with cryptogenetic cerebral ischemic attack due to a rare cause of paradoxical embolization.

**Results:** A 13th years-old, male child, with a caval superior sinus venosus atrial septal defect operated ten years ago. During a covid-19 infection, he developed a transient ischemic attack with speech and vision disorders. An interatrial residual shunt (or a patent foramen ovale) was suspected as the cause of a paradoxical embolization. A trans-oesophageal echocardiography with bubble-test from an upper-limb vein demonstrated a significant early presence of bubbles within the left side cardiac chambers. However, the bubble-test from a lower-limb vein negative. The cardiac catheterization demonstrated an occlusion of right superior vena cava with a dilated azygos vein and with the presence of multiple little collateral vessels between the superior vena cava and the pulmonary veins (figure). These collateral vessels were probably the cause of the paradoxical embolization phenomenon during covid-19 infection. The patient was discharged with antiplatelet therapy.

**Conclusions:** Patent foramen ovale and pulmonary arterio-venous malformation are the most common causes of paradoxical embolization; however, in the patients with significant stenosis of the superior vena cava, the development of collateral vessel linked with the pulmonary veins may be a rare cause of paradoxical embolization.

**Keywords:** paradoxical embolization, interatrial septal defect, ischemic cerebral event

### P-192

#### Subclinical vascular changes in children with well-regulated Type 1 diabetes

Ebba Bergdahl<sup>1</sup>, Linda Göransson<sup>3</sup>, Frida Sundberg<sup>4</sup>, Gun Forsander<sup>4</sup>, Frida Dangardt<sup>2</sup>

<sup>1</sup>Department of clinical and molecular medicine, Institute of medicine, at the Sahlgrenska academy University of Gothenburg, Gothenburg, Sweden; <sup>2</sup>Pediatric Heart Center, Queen Silvia Children's Hospital, Gothenburg, Sahlgrenska University Hospital, Region Västra Götaland, Sweden; <sup>3</sup>Department of Pediatrics, Queen Silvia Children's Hospital, Sahlgrenska University Hospital, Region Västra Götaland, Gothenburg, Sweden; <sup>4</sup>Department of Pediatrics, Institute of Clinical Sciences, Sahlgrenska Academy, University of Gothenburg, Sweden

**Background and Aim:** Patients with type 1 diabetes (T1D) have increased risk of cardiovascular disease (CVD). The impact of metabolic control and blood pressure on the cardiovascular system starts already in childhood. By using novel, highly sensitive methods for early CVD risk evaluation and treatment monitoring for paediatric patients with T1D, we aimed to establish the determinants for early vascular changes in children with T1D to facilitate cardiovascular prevention in the future.

**Method:** Fifty children (6-15, 99yr) with T1D duration of  $\geq 5$  years were randomly selected from the national pediatric diabetes registry SWEDIABKIDS. Forty-one age- and gender matched healthy controls were also included in the study. We used ultra-high frequency ultrasound, enabling separate visualization of the layers in the arterial wall for morphological assessment of the radial, dorsal pedal (DP) and carotid arteries. Office and ambulatory blood pressure measurements were performed and data from the T1D patients' continuous glucose monitoring (CGM) systems and HbA1c was collected.

**Results:** The children with T1D showed a 12% increase in radial intima thickness (IT) and a 22% increase in dorsal pedal (DP) intima-media thickness (IMT) ( $p = 0.002$  and  $p = 0.034$ , respectively) as compared to healthy controls. Increased Carotid IT was associated with HbA1c, time in range (TIR), time in target (TIT) and glucose variability (SD) (HbA1c:  $r_2 = 0.34$ ,  $p = 0.033$ , TIR:  $r_2 = -0.47$ ,  $p = 0.014$ , TIT:  $r_2 = -0.64$ ,  $p < 0.001$  and SD:  $r_2 = 0.40$ ,  $p = 0.004$ , respectively). Carotid intima/diameter ratio was associated to HbA1c, TIT and T1D duration (HbA1c:  $r_2 = 0.37$ ,  $p = 0.019$ , TIT:  $r_2 = -0.49$ ,  $p = 0.009$  and T1D duration:  $r_2 = 0.31$ ,  $p = 0.048$ , respectively). Positive correlations between blood pressure and SD ( $r_2 = 0.36-0.52$ ,  $p = 0.002-0.045$ ) and negative correlations between blood pressure and TIR and TIT, respectively, was also found ( $r_2 = -0.53- -0.42$ ,  $p = 0.002-0.017$ ).

**Conclusions:** Early morphological changes in both tunica intima and media were seen in the peripheral arteries in children with T1D. Strong associations between carotid IT as well as blood pressure with the clinically used markers for metabolic control including CGM data, supports the theory of vascular glucotoxicity in T1D and further stresses the importance of striving for normoglycemia.

**Keywords:** Cardiovascular prevention, vascular ultrasound, type 1 diabetes

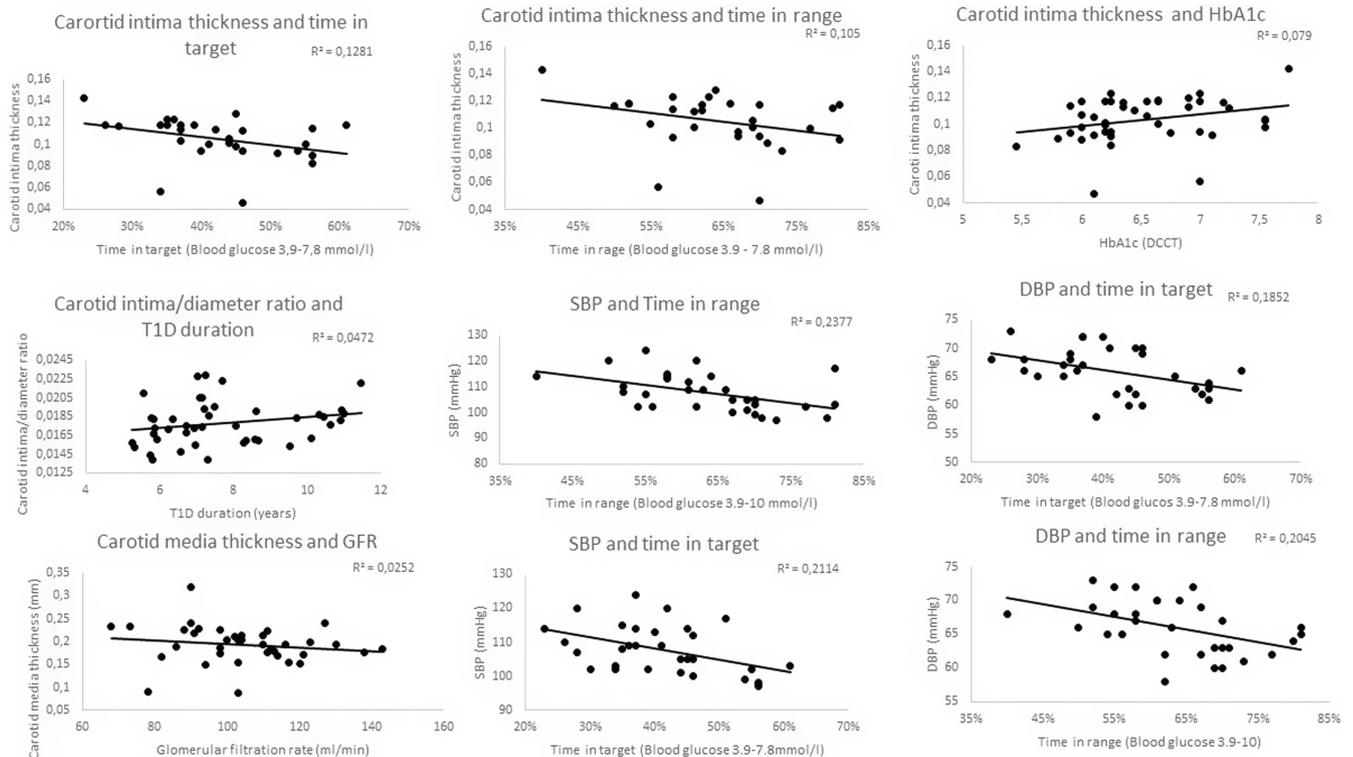


Figure showing correlations between vascular wall thickness, glycaemic control and blood pressure.

### P-193

#### Crisscross heart: an outflow tract defect? an anatomic study.

Segolene Bernheim<sup>1</sup>, Damien Bonnet<sup>2</sup>, Lucile Houyel<sup>2</sup>

<sup>1</sup>Congenital and pediatric cardiology, Necker-Enfants Malades-M3C, APHP, Paris, France; <sup>2</sup>Congenital and pediatric cardiology, Necker-Enfants Malades-M3C, APHP, Paris, France; Université Paris-Cité, Paris, France

**Background and Aim:** Crisscross heart (CCH) is a rare congenital malformation in which the atrioventricular inflow vectors are approximately orthogonal or perpendicular. CCH is associated with other defects including malposition of the great arteries, supero-inferior ventricles, and ventricular septal defects (VSD). A recent study in a mouse model demonstrates that CCH and associated malformations might be the result of a growth defect of the outflow tract. In order to confront this hypothesis, we studied 15 cases of crisscross heart with detailed anatomical description and clinical outcomes.

**Method:** All patients with crisscross heart in Necker –Enfants Malades from 1999 to 2022 were included in a retrospective study. Echocardiography, CT scans and anatomical MRIs were reviewed by a referent cardiologist to validate the diagnostic and the anatomical features.

**Results:** Segmental analysis according to Van Praagh was SDL in 11 patients (73%), SDD in 3 and SDA in 1. The ventricles were supero-inferior in 12 patients (80%). Ventriculo-arterial connections were always abnormal: double outlet right ventricle in 14/15 (93%) with a bilateral conus in 11 and a subaortic conus in 3, transposition of the great arteries in 1. The pulmonary valve was stenotic or atretic in 12 patients (80%). All patients had a VSD opening in the inlet of the right ventricle: inlet only in 8

patients (53%), confluent inlet/outlet in 5, inlet with muscular extension in 2. Other associated defects were: left superior vena cava (2), mitral cleft (1), straddling tricuspid valve (1), hypoplastic left ventricle (1), azygos venous return (1). Fourteen (93%) patients underwent surgery, univentricular repair in 12/14 (80%), biventricular in 2. There was no early nor late death with a median follow up duration of 10.7 years (0–23).

**Conclusions:** Crisscross heart is always associated with a malposition of the great vessels and a VSD, always of the inlet type. This might be in favor of a defective growth of the outflow tract, leading to an incomplete or absent rotation of the great arteries, as demonstrated in animal models. This also accounts for the difficulties encountered by the surgeons to repair this complex malformation, and for the high rate of univentricular repair.

**Keywords:** Crisscross heart, supero-inferior ventricles, outflow tract, morphogenesis

### P-195

#### The v-shaped double-layer patch technique for complete atrioventricular septal defect

Shanquan Sun<sup>1</sup>, Yangxue Sun<sup>2</sup>, Jingsi Huang<sup>1</sup>, Peng Zou<sup>1</sup>, Jiao Rao<sup>1</sup>, Weibin Xu<sup>1</sup>, Qin Liu<sup>1</sup>

<sup>1</sup>Cardiac Center of Guangdong Women and Children Hospital;

<sup>2</sup>Department of Pediatric Cardiac Surgery, National Center for Cardiovascular Disease and Fuwai Hospital, Chinese Academy of Medical Sciences, Peking Union Medical College

**Background and Aim:** Several surgical techniques for repair of a complete atrioventricular septal defect (CAVSD) have been developed. However, the postoperative complications with these methods may lead to reoperation during follow-up. The aim of this

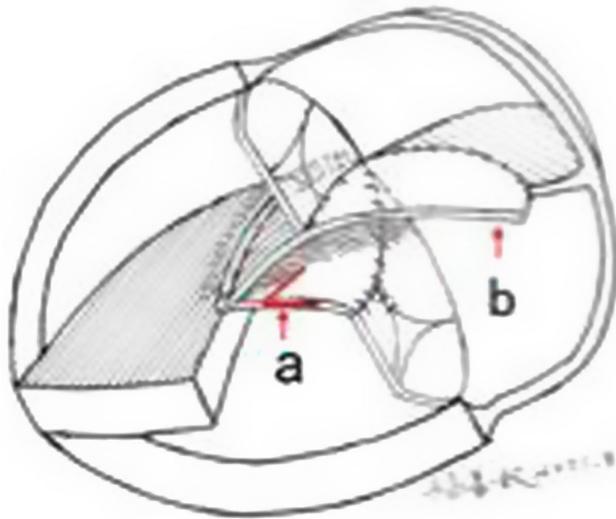
report is to share our experience with a novel surgical technique for CAVSD that has anatomic advantages postoperatively and could reduce the reoperation rate.

**Method:** 33 patients who underwent repair of CAVSDs between April 2011 and September 2021 were retrospectively investigated. All of these patients were repaired by the V-shaped double-layer patch technique.

**Results:** There were no deaths (0%) and only two reoperation (6.1%) in the series. The aortic cross-clamp and cardiopulmonary bypass times were  $65.9 \pm 18.1$  min and  $122.7 \pm 38.0$  min, respectively. To date, no significant residual ventricular septal defects have been detected and no left ventricular outflow tract obstruction has been seen on echocardiography in any patient. During follow-up, the left atrioventricular valve status was assessed as no incompetence in 9 patients (27.3%), trivial in 21 (63.6%), and mild in 3 (9.1%).

**Conclusions:** The V-shaped double-layer patch technique is a valuable surgical option for patients with CAVSDs. The mid-term results in our series document excellent performance of this technique, which augments the area of the anterior valve of the left atrioventricular valve to make it closer to a normal mitral valve and may also reduce the need for reoperation.

**Keywords:** V-shaped double-layer patch technique, complete atrioventricular septal defect, postoperative complications



**Figure 1** "V-shaped Double-layer Patch Technique" is formed by the patch(a) for repairing the root of the LAVV and the patch(b) for repairing the atrial and ventricular septal defect.

#### P-196

##### Novel mechanism of pulsus bisferiens encountered during pacemaker placement

Todd Glenski, Joseph Huffman, Christian Taylor  
Department of Anesthesiology, Children's Mercy Hospital, University of Missouri - Kansas City School of Medicine, Kansas City, United States

**Background and Aim:** Pulsus bisferiens (PB) is characterized by two arterial pressure waves separated by a mid-systolic dip. It is traditionally present in the setting of hypertrophic obstructive cardiomyopathy (HOCM) and left ventricular outflow tract (LVOT)

obstruction. Here we encounter a novel etiology of PB in a patient with congenital heart disease.

**Method:** A 26-year-old female with trisomy 21 and history of recent mechanical mitral and aortic valve replacement complicated by postoperative complete heart block presents for permanent pacemaker placement. Following an uneventful intravenous induction of general anesthesia, non-invasive blood pressure monitoring was inconsistent and a right radial arterial line was inserted. PB was then observed.

**Results:** TEE revealed a large echobright mass on the posterior/lateral leaflet of the mechanical mitral valve causing "at least moderate" mitral regurgitation, decreased valvular opening, and moderate mitral stenosis with an inflow gradient of 5mmHg. Although imaging was challenging through the mechanical valves, LV outflow was laminar and unobstructed. There was minimal gradient across the LVOT and mechanical aortic valve.

Encountered in this case is pulsus bisferiens in the absence of LVOT obstruction or aortic insufficiency. Rather, this new etiology of PB occurred in the setting of moderate mitral stenosis and regurgitation of a mechanical mitral valve. This finding may be the result of inconsistent, biphasic ventricular filling. The non-uniform preload of the left ventricle led to inconsistent, biphasic LV ejection during systole. The disorganized rhythm with ventricular pacing may have accentuated the compromised LV preload. Once the thrombus was removed, LV preload and stroke volume improved.

**Conclusions:** PB may not always be the result of LVOT obstruction, HOCM or AI and the true cause of PB is needed to guide any therapies and echocardiography is required for a definitive diagnosis. Traditional management of HOCM and LVOT obstruction with rate control and afterload support may be counterproductive. For our patient with mitral regurgitation, treatment would center around achieving AV synchrony and consistent LV filling with reduced afterload until thrombectomy is performed.

**Keywords:** Pulsus bisferiens, thrombus, mitral regurgitation



Pulses Bisferiens

#### P-197

##### Left ventricular outflow tract velocity time integral (LVOT-VTI) as marker of left ventricular function in post-op CHD patients

Samir Bhatia, Sitaraman Radhakrishnan, Sushil Azad  
Department of Pediatric Cardiology Fortis Escorts Heart Institute New Delhi India

**Background and Aim:** Left ventricular outflow tract velocity time integral (LVOT-VTI), a Doppler-derived measure of stroke distance has been used as an alternate marker of assessing left ventricular function in adults. LVOT-VTI is easily obtained and does not depend upon ventricular geometry and wall motion abnormalities.

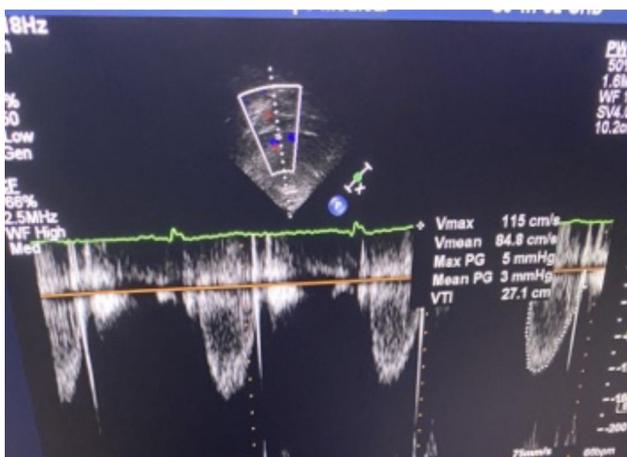
We investigated the relationship between LVOT-VTI and conventional measures of function on two dimensional echocardiography using biplane Simpson's method, and Global longitudinal strain (GLS) in children with post-tricuspid left to right shunt surgery in immediate post-operative period.

**Method:** A single institution observational study. Post-operated patients with post-tricuspid left to right shunt surgeries in the immediate post-operative period (till discharge) were enrolled in the study. Transthoracic 2D echocardiography along with Doppler evaluation in the immediate post-operative period was performed. Left ventricular function was evaluated using biplane Simpson's method. GLS measurement was made in three standard apical views and averaged. Left ventricular outflow tract velocity time integral (LVOT-VTI) was evaluated by Pulsed Wave Doppler Imaging (PWDI). LVOT-VTI was taken as average of 3 recordings and taken by 2 observers to overcome inter-observer variability.

**Results:** 95 patients fulfilling the inclusion and exclusion criteria were analyzed. Out of 95 patients 57.9% were males. Median age at surgery was 7 months. Most common post-tricuspid shunt lesion was peri-membranous VSD (62.1%). 70.5 % patients didn't have residual VSD lesion. Mean EF in immediate post-op period was 41.15(7.02) which improved to 52.05(5.15) at discharge. Corresponding FS at discharge was 25.83 (3.01), while mean GLS at discharge was -19.01(4.89). The mean LVOT-VTI in immediate post-op period was 13.5 (2.92) which improved to 16.47(1.94) at discharge. The mean duration of inotrope use was 4.16(1.86) days. LVEF and LVOT-VTI had significant correlation with each other both in immediate post op period and discharge. Immediate post-op LVOT-VTI and inotrope duration had negative correlation which was found significant. FS and LVOT-VTI had significant correlation with each other. LVOT-VTI and GLS had significant correlation with each other. Significant negative correlation is noted between GLS post op at discharge and LVEF post-op at discharge

**Conclusions:** LVOT-VTI has strong correlation with LVEF by Simpsons method, FS and GLS.

**Keywords:** LVOT-VTI, LV Function, Post tricuspid left to right shunts, CHD surgery, GLS



Left ventricular outflow tract velocity time integral (LVOT-VTI) evaluated by Pulsed Wave Doppler Imaging (PWDI)

## P-198

### Long-term follow-up of atrioventricular valve function in fontan patients

Eva Grootjans<sup>1</sup>, Romanie Naessens<sup>1</sup>, Thomas Martens<sup>1</sup>, Kristof Vandekerckhove<sup>2</sup>, Katya De Groot<sup>2</sup>, Joseph Panzer<sup>2</sup>, Julie De Backer<sup>2</sup>, Laurent Demulier<sup>2</sup>, Daniel De Wolf<sup>2</sup>, Katrien François<sup>1</sup>, Thierry Bove<sup>1</sup>  
<sup>1</sup>Department of Cardiac Surgery, University Hospital Ghent, Ghent, Belgium; <sup>2</sup>Department of Pediatric and Adult Congenital Cardiology, University Hospital Ghent, Ghent, Belgium

**Background and Aim:** The objective of this study is to evaluate the relationship between atrio-ventricular valve (AVV) and single ventricle function in Fontan survivors, including the effect of intercurrent AVV surgery.

**Method:** Outcome was analysed in 106 Fontan survivors operated between 1987 and 2021, focusing on transplant-free survival and need for AVV surgery along the Fontan pathway. Longitudinal echocardiographic examination was performed for long-term AVV and single ventricle function.

**Results:** Univentricular morphology was RV-dominant in 30%, LV-dominant in 66% and ambiguous in 4%. AVV morphology was tricuspid in 28%, mitral in 60%, while 12% had a common AVV. Within median follow-up of 13, 1y(IQR 7, 2-19, 4), survival was 100%. Transplant-free survival was 99±1% and 93±6% at 10 and 25 y respectively. Twenty AVV procedures were performed in 14 children, respectively pre-Fontan(n=10), per-Fontan(n=8), and post-Fontan(n=2), resulting in a freedom from AVV surgery of 94±2% and 88±3 % at 1 and 5 year.

Echocardiographic follow-up was available in 96%, yielding 7-25 examinations per patient. AVV function appeared to deteriorate over time (OR 1.10 (1.08-1.13), p<0.001), without difference for AVV morphology(p = 0.080 between mitral-cAVV, p = 0.928 between tricuspid-cAVV) or ventricular dominance(p = 0.088). However, AVV function degraded more in patients with prior AVV surgery(OR 10.5 (3.9-27.9), p = 0.001). Time-related worsening of single ventricle function was observed(OR 1.14 (1.10-1.18), p<0.001), and significantly more in RV-dominance(OR 3.12 (1.01-9.69), p = 0.049). There was a significant inverse correlation between degree of AVVR and ventricular function(Rho - 0.274, p<0.001). Of note, only 18% of the patients were taking ACE-inhibitors during FU.

**Conclusions:** In an homogenously treated Fontan population with increasing use of extracardiac conduits, the 20-year transplant-free survival is excellent. AVV surgery was necessary in 13%, resulting mostly in a durable AVV function. However, a slow time-related decline of AVV function together with degradation of ventricular function is worrisome, evoking a role for routine medical heart failure therapy in Fontan patients.

**Keywords:** atrioventricular valve function, Fontan, single ventricle physiology

## P-199

### Our 28 years experience with heart transplantation in paediatric patients and adults with congenital heart disease

María Luz Polo López<sup>1</sup>, Blanca Torres Maestro<sup>1</sup>, Bunty Kishore Ramchandani Ramchandani<sup>1</sup>, Raúl Sánchez Pérez<sup>1</sup>, Juvenal Rey Lois<sup>1</sup>, Álvaro González Rocafort<sup>1</sup>, Tomasa Centella Hernández<sup>1</sup>, María Jesús Lamas Hernández<sup>1</sup>, Carlos Labradero De Lera<sup>2</sup>, Juan José Menéndez Suso<sup>3</sup>, Ángel Aroca Peinado<sup>1</sup>

<sup>1</sup>*Pediatric Cardiac Surgery. La Paz-Ramón y Cajal University Hospital. Madrid. Spain;* <sup>2</sup>*Pediatric Cardiology. La Paz University Hospital. Madrid. Spain;* <sup>3</sup>*Pediatric Intensive Care Unit. La Paz University Hospital. Madrid. Spain*

**Background and Aim:** When conventional treatment is not possible to improve heart failure in a child or an adult with congenital heart disease, transplantation became the best option to achieve a better life. We present our whole experience with heart transplant in our congenital heart unit along 28 years.

**Method:** Retrospective study of 116 heart transplants in 116 patients operated from 1994 to 2022. Study approved by the ethics committee of our hospital. We analyse our short and long term follow-up results. Statistic analysis was performed with SPSS.20.  
**Results:** Median age: 7, 6 years [IQR: 1, 4-15, 8], 46% female. Diagnosis: 48% congenital heart disease and 52% cardiomyopathy (dilated-37%, restrictive-12% and hypertrophic-3%). Previous surgery: 65%. Functional class was poor (29% class-III, 70% class-IV). Transplants receptors were at home (48%), and others were hospitalized with intravenous inotropes (24%) or ventricular assistance (28%). Our initial surgical technique was biatrial-20%, and changed to bicaval-80% in 2008 until now. Along the years the complexity associated to our heart recipients has been increasing (p:0, 005) with more previous surgeries, end stage congenital heart disease and ventricular assistance in the last years respect to our beginnings. In-hospital mortality was 20, 7%, with risk factors related were biatrial technique (p:0, 015), long extracorporeal times (p:0, 05), and ECMO required in postoperative days (p<0, 001). Mean follow-up: 4, 75 ± 5, 4 years. Kaplan-Meier survival curve (excluded in-hospital mortality) showed that 92%, 82% and 71% of our patients were alive at 1, 5 and 10 years post-transplant respectively. Late mortality was 22, 8%, mainly owed to sudden cardiac death and oncologic reasons. Last follow-up of our living patients show that 90% are in functional class-I

**Conclusions:** Our own experience with paediatric and congenital heart disease transplant has improved along the years. Nowadays we offer good results to these complex patients to live longer with better quality of life.

**Keywords:** Pediatric, Heart transplantation, end stage congenital heart disease

## P-200

### Outcomes after ross procedure with pulmonary autograft reinforcement using reimplantation technique

*Lisa Guirgis, Sébastien Hascoet, Mohamed Ly, Bastien Provost, Jelena Radojevic, Isabelle Van Aerschot, Sarah Cohen, Emre Belli*  
*Department of Pediatric and Congenital Heart Disease, Marie Lannelongue Hospital, M3C, GHPSJ, University Paris-Saclay. Le Plessis Robindon France*

**Background and Aim:** Pulmonary autograft reinforcement to prevent dilatation and subsequent neo-aortic valve regurgitation was reported. However, the late functional outcome of the native pulmonary valve inside a rigid Dacron conduit requires to be documented. We aimed to assess the results of modified Ross procedure associated with autograft reinforcement reimplantation technique.

**Method:** Outcomes of 61 consecutive patients who underwent Ross procedure with reinforcement between 2009 and 2021 were analyzed. Majority of cases has presented with mono or bicuspid aortic valve (n = 52; 85%), predominant aortic valve regurgitation (n = 47; 77%) and dilatation (>30mm) of the ascending aorta (n = 33; 54%). Forty-seven patients (77%) had prior aortic valve procedure including 38 surgical repair (62%) and 9 balloon dilatation

(15%). Pulmonary autograft was reimplanted in a Dacron conduit of a median diameter = 25.6 mm (ranges: 20-30) using Tirone David's valve sparing aortic root replacement technique.

**Results:** No death occurred. Median age at procedure was 16.8 years (range: 6-38). Two patients (3%) required early neo-Aortic root revision, one other was reoperated later on resulting in neo-aortic valve replacement in 3 (4.9% IC 95% [0.34%; 12.7%]) because of respectively infective endocarditis, left ventricular false aneurysm and leaflet perforation. Six patients required right ventricular outflow conduit replacement including one percutaneous replacement. At mean 66 ± 50, 5 months postoperatively, the survival rate with freedom from reintervention was 83% [71, 9; 93, 5] and the deterioration of the initial neo-Aortic valve function (regurgitation or stenosis) was not observed.  
**Conclusions:** Autograft reinforcement by means of reimplantation technique allowed to extend the indications for Ross procedure to all patients requiring Aortic valve replacement and prevented neo-Aortic root dilatation. Failures were early and rare, and late controls confirmed the stability of the neo-aortic valve function in follow-up.

**Keywords:** Congenital Heart Disease, Aortic valve repair, Ross procedure

## P-201

### Echocardiographic predictors of atrioventricular valve regurgitation after surgical repair of complete atrioventricular septal defect

*Camilla Calvieri<sup>1</sup>, Mario Panebianco<sup>3</sup>, Corrado Di Mambro<sup>2</sup>, Fabrizio Drago<sup>2</sup>, Lorenzo Galletti<sup>3</sup>, Adriano Carotti<sup>3</sup>, Sonia Albanese<sup>3</sup>*  
*<sup>1</sup>Department of Clinical, Internal, Anesthesiological and Cardiovascular Sciences, "Policlinico Umberto I" Hospital, Sapienza University of Rome, Rome, Italy.; <sup>2</sup>Paediatric Cardiology and Cardiac Arrhythmia/Syncope Unit, Department of Paediatric Cardiology and Cardiac Surgery, Bambino Gesù Children's Hospital IRCCS, Rome, Italy.; <sup>3</sup>Department of Cardiac Surgery, Cardiology, Heart and Lung Transplantation Bambino Gesù' Children's Hospital, IRCCS, Rome, Italy.*

**Background and Aim:** Echocardiographic predictors of atrioventricular valve regurgitation (AVVR) after double sandwich technique surgical repair of complete Atrioventricular Septal Defect (cAVSD), age at surgery and the influence of Down Syndrome (DS) on post-surgical outcome have been widely debated. The aim of our study was to identify the impact of these factors on post-operative AVVR.

**Method:** Surgical and echocardiographic data of patients who underwent cAVSD biventricular repair at our Institution between 2015 and 2022 were retrospectively reviewed. Left AVV regurgitation (LAVVR) and Right AVV regurgitation (RAVVR) were assessed preoperatively, and postoperatively at 5, 30 and 90 days. All cAVSD patients were followed for a median of 2 years (IQR 1-4 years). Echocardiographic analysis was performed by 2 independent cardiologists.

**Results:** We enrolled 43 cAVSD patients, 24 (56%) female, 19 (44%) male, median weight of 4.3 kg (IQR 3.5-5 kg), median age of 4 months (IQR 2-6 months), 39 Rastelli type A (90%), 28 (65%) affected by DS. There were no early deaths. After repair, 14 (33%) showed early moderate to severe LAVVR and 6 (14%) early moderate RAVVR. Strong correlations between age and weight at repair ( $\rho:0.726$ ,  $p>0.001$ ) and between length of mural AVV leaflet and VSD size ( $\rho:0.654$ ,  $p<0.001$ ) were found. Corrected cAVSD patients with post-surgical moderate LAVVR had a lower pre-surgical mural leaflet length if compared with those without post-surgical moderate LAVVR (2, 4 ± 3, 5 vs 6, 6 ± 6 mm,  $p<0.001$ ). At univariate analysis length of AVV mural

leaflet was associated to early moderate LAVVR with an O.R. 0.832 (95% CI 0.705–0.983,  $p = 0.030$ ). Age at correction below 4 months was significantly associated to early moderate RAVVR, O.R. 0.159 (95% CI 0.027–0.931,  $p = 0.041$ ). Finally, cAVSD were analyzed on the basis of the presence of DS. A significant difference in left ventricular end-diastolic volume was observed according to DS ( $p < 0.05$ ). Among the 8 cases of early moderate severe RAVVR, a significant association was found with DS ( $p = 0.035$ ).

**Conclusions:** Length of mural leaflet seems to be a predictor of early development of LAVVR, meanwhile age at repair below 4 months of early moderate RAVVR occurrence. In DS children a strong association with early moderate RAVVR was found.

**Keywords:** complete atrioventricular septal defect, double sandwich surgical repair, atrioventricular valve regurgitation, echocardiographic parameters, down syndrome, age at repair

## P-202

### Double outlet right ventricle fallot type – is staged repair inferior to primary repair?

Jasmin Illig<sup>1</sup>, Julie Cleuziou<sup>1</sup>, Cordula Wolf<sup>2</sup>, Paul Philipp Heinisch<sup>1</sup>, Peter Ewert<sup>2</sup>, Juergen Hoerer<sup>1</sup>, Maria Von Stumm<sup>1</sup>

<sup>1</sup>Department for Congenital and Pediatric Heart Surgery, German Heart Center Munich, Munich, Germany; <sup>2</sup>Department for Congenital and Pediatric Cardiology, German Heart Center Munich, Munich, Germany

**Background and Aim:** In the current literature, outcome data of double outlet right ventricle Fallot type (DORV-TOF) is often mixed with other DORV subtypes despite differences in morphology and surgical treatment. Therefore, the optimal surgical management remains unknown. We compared the outcomes of DORV-TOF patients following staged and primary repair.

**Method:** The data is based on a single-centre retrospective cohort study. All patients with DORV-TOF who underwent repair at our institution between 2003 to 2020 were included in the study. The operative technique comprised transannular patch or conduit implantation for reconstruction of the right ventricular outflow tract (RVOT). The primary endpoint was all-cause mortality. The secondary endpoint was the composite of catheter-based re-intervention and re-operation.

**Results:** A total of 53 patients were included in the study, the median follow up time was 6 years [range:1month–18years]. The primary repair group (PRG) consisted of 25 (47%) patients and the staged repair group (SRG) of 28 (53%) patients. Palliative procedures comprised shunt operation ( $n = 20$ ), balloon pulmonary valvuloplasty ( $n = 5$ ), arterial duct stenting ( $n = 2$ ), and RVOT stenting ( $n = 1$ ).

At repair, right ventricle-to-pulmonary artery (RV-PA) continuity was achieved with a transannular patch (PRG = 23, 92%; SRG = 12, 43%;  $p < 0.01$ ) or with conduit implantation (PRG = 2, 8%; SRG = 16, 57%;  $p < 0.01$ ).

There were two early deaths (3.8%) in the PRG. No late deaths occurred, resulting in an overall survival rate of  $96 \pm 3\%$  at 10 years (SRG =  $100 \pm 0\%$ , PRG =  $92 \pm 5\%$ ,  $p = 0.13$ ).

Re-operations/re-interventions were necessary in 25 patients: 8 (32%) in PRG and 17 (61%) in SRG ( $p = 0.04$ ). Cumulative freedom of re-intervention/re-operation was  $68 \pm 11\%$  in the PRG and  $39 \pm 12\%$  in the SRG at 15 years. Re-operations included: takedown of repair, residual VSD closure, RV-PA conduit implantation, and conduit exchange. Re-interventions were: pulmonary artery angioplasty, embolization of aorto-pulmonary collaterals, and valve-implantation.

Re-operations/re-interventions were necessary early during the postoperative period in 8 (15%) patients (SRG = 3, 11%;

PRG = 5, 20%;  $p = 0.35$ ) and late in 17 (32%) patients (SRG = 14, 50%; PRG = 3, 12%;  $p < 0.01$ ).

**Conclusions:** Survival after repair of DORV-TOF is excellent. There were no differences regarding in-hospital events between staged vs. primary repair. Following discharge, staged repair was associated with an increased number of re-interventions and re-operations. Therefore, primary repair should be favoured if possible.

**Keywords:** double outlet right ventricle, tetralogy of fallot, right ventricular outflow tract obstruction, cono-truncal abnormalities, congenital heart surgery

## P-203

### Hand made TRI leaflet right ventricle to pulmonary artery conduits; a cost effective alternative

Faiz Rasool<sup>1</sup>, Mohammad Asim Khan<sup>2</sup>, Salman Ahmad Shah<sup>3</sup>

<sup>1</sup>Department of Pediatric Cardiac Surgery, Children Hospital Lahore / University of Child Health Sciences Lahore; <sup>2</sup>Department of Pediatric Cardiac Surgery, Children Hospital Lahore / University of Child Health Sciences Lahore; <sup>3</sup>Department of Pediatric Cardiac Surgery, Children Hospital Lahore / University of Child Health Sciences Lahore

**Background and Aim:** In congenital cardiac surgery, Right ventricle to Pulmonary artery conduits are inevitable in specific group of patients. Various strategies including homograft, valved conduit, and bovine jugular veins are used to restore continuity between right ventricle and pulmonary artery. In resource limited countries like Pakistan these are not easily available and affordable. We report the experience of our study of using a handmade trileaflet valve conduit to reconstruct the right ventricular outflow tract.

**Method:** From January 2012 to December 2021, a total of 150 patients with different congenital heart diseases underwent surgeries that required RV to PA conduit.

We used hand made Bovine pericardial tube with PTFE valve and used it as RV to PA conduit. More recently we are using PTFE tube with PTFE valve in bigger patients.

**Results:** Patients ranged from 1 month to 30 years. Diagnosis included transposition of great arteries with VSD and pulmonary stenosis, truncus arteriosus, aortic stenosis or regurgitation, tetralogy of fallots, and Double outlet right ventricle. Postoperative complications were observed in 23 patients including 7 in hospital deaths. No significant regurgitation was observed in this series. Overall postoperative gradients were stable with mean gradient 25.3 mmHg (8 mmHg – 40 mmHg). In follow up, most of the patients were doing fine with no significant complications. 6 patients developed calcification of the bovine pericardium.

**Conclusions:** The use of handmade valve conduits have excellent flow dynamics and are comparable results with any other conduit. These are cost effective alternatives in this part of the world, where well-established conduits have cost implications and uncertain availability.

**Keywords:** RV to PA conduits, hand made tri leaflet conduits

## P-204

### Tetralogy of fallot in toddler with left pulmonary artery agenesis – a challenging management. what means better?

Cristina Flavia Al Akel<sup>1</sup>, Horatiu Suci<sup>3</sup>, Ana Sorina Pasc<sup>4</sup>, Mihaela Badulescu<sup>4</sup>, Amalia Fagarasan<sup>2</sup>

<sup>1</sup>Department of Pediatric Cardiology, Emergency Institute of Cardiovascular Disease and Transplantation, Targu-Mures, Romania; <sup>2</sup>Department of Pathophysiology, University of Medicine, Pharmacy, Sciences and Technology of Targu Mures, Romania; <sup>3</sup>Department of

*Pediatric Cardiology, Emergency Institute of Cardiovascular Disease and Transplantation, Targu-Mures, Romania; Department of Pediatrics, University of Medicine, Pharmacy, Sciences and Technology of Targu Mures, Romania; <sup>3</sup>Department of Cardiovascular Surgery, Emergency Institute of Cardiovascular Disease and Transplantation, Targu-Mures, Romania; Department of Surgery, University of Medicine, Pharmacy, Sciences and Technology of Targu Mures, Romania; <sup>4</sup>Department of Pediatric Cardiology, Emergency Institute of Cardiovascular Disease and Transplantation, Targu-Mures, Romania*

**Background and Aim:** Tetralogy of Fallot can associate a great variety of lesions, also including more rarely encountered defects, such as left pulmonary artery agenesis, scenario that requires a particular approach. The aim of this paper is to present the case of a symptomatic toddler with tetralogy of Fallot and left pulmonary artery agenesis and bringing to light a particular therapeutic approach.

**Method:** We present the case of a two and a half years-old male toddler with tetralogy of Fallot, cyanotic (oxygen saturation of 74 percent), with clubbing, fatigue and reduced tolerance to physical activity. The patient presents in our service for therapeutic decision. Echocardiography exam and thoracic CT angiography documented the diagnosis: tetralogy of Fallot with left pulmonary artery agenesis and right-sided aortic arch with mirror image branching.

**Results:** The surgical approach was to perform a modified Blalock Taussig shunt, with postoperative evolution uneventful regarding hemodynamic status, with improvement in oxygen saturation up to 90 percent and also in Ross class staging, but complicated by an unique symptomatic seizure caused by otomastoiditis.

**Conclusions:** The particularity of this case consists in the difficulties encountered in making therapeutic decisions regarding a toddler with severe form of tetralogy of Fallot, left pulmonary artery agenesis and right-sided aortic arch.

**Keywords:** toddler, tetralogy of Fallot, pulmonary artery agenesis

## P-206

### Ductal stenting as a step towards cone reconstruction in ebstein anomaly

Julie Cleuziou<sup>1</sup>, Rüdiger Lange<sup>2</sup>, Nora Lang<sup>4</sup>, Sarah Hartmann<sup>3</sup>, Peter Ewert<sup>3</sup>, Jürgen Hörer<sup>1</sup>, Nicole Nagdyman<sup>3</sup>

<sup>1</sup>Congenital and Paediatric Heart Surgery, German Heart Centre Munich, Germany; <sup>2</sup>Cardiovascular Surgery, German Heart Centre Munich, Germany; <sup>3</sup>Paediatric Cardiology and Congenital Heart Disease, German Heart Centre Munich, Germany; <sup>4</sup>Department of Paediatric Cardiology, University Hospital Hamburg, Germany

**Background and Aim:** Neonatal Ebstein anomaly is characterized by functional pulmonary atresia associated with a varying degree of tricuspid regurgitation. These patients have a duct dependent pulmonary perfusion and require an early intervention to achieve adequate saturations. Ductal stenting has shown excellent results in the palliation of neonates with duct dependent pulmonary circulation but is not routinely utilized in neonates with Ebstein anomaly.

**Method:** We report on 2 patients who were treated with ductal stenting as neonates and consecutively received a Cone reconstruction of the tricuspid valve as definitive treatment.

**Results:** Both patients were born at term and weighed 3.4 and 3.1 kg, respectively. Saturation was 83 and 88%, respectively. On echocardiography, the tricuspid valve was severely displaced with moderate tricuspid regurgitation in both neonates. The atrialized portion of the right ventricle was very large and the functional size reduced. Both had an atrial septal defect with right-to-left shunt

and required intravenous prostaglandine infusion to preserve adequate pulmonary perfusion. Initial treatment was ductal stenting with two Onyx stents in both patients at the age of 7 and 10 days. Both interventions were uneventful although the next step, i.e. the possibility of a biventricular repair was not clear. The saturations increased to 95% in both patients and after 5 and 6 months respectively, antegrade pulmonary flow was visible on echocardiography. The saturations started to decrease at one year of age with an increase of body weight. At the same time, the tricuspid regurgitation increased in both patients to severe.

A Cone reconstruction of the tricuspid valve with additional bidirectional cavo-pulmonary anastomosis was performed at 14 and 28 months, respectively. The operation and postoperative course was uneventful with minimal and trivial tricuspid regurgitation at discharge. At last follow-up (7 and 23 months post Cone reconstruction), both patients are clinically well with minimal tricuspid regurgitation but residual large right atrium and right ventricle.

**Conclusions:** Ductal stenting is a good initial treatment option in patients with Ebstein anomaly. Caregivers can gain time to decide on the best definitive treatment strategy. This might increase the chance of achieving a biventricular repair in selected patients.

**Keywords:** Ebstein, Cone procedure, Neonate, Ductal Stenting

## P-207

### Incidental diagnosis of large aortopulmonary window post patent ductus arteriosus ligation: A case report

Megha Bharatkumar Unadkat<sup>1</sup>, Aliasgher Mukhtar Saajan<sup>1</sup>, Julieth Nachone Kabirigi<sup>1</sup>, Gloria Reginald Mbwile<sup>1</sup>, Zawadi Edward Kalezi<sup>2</sup>, Stella Mongella Sharau<sup>2</sup>, Godwin Godfrey Sharau<sup>2</sup>, Sulende Daudi Kubhoja<sup>2</sup>, Deogratias Arnold Nkya<sup>1</sup>, Naizihijwa Gadi Majani<sup>2</sup>

<sup>1</sup>Department of Paediatrics & Child Health, Muhimbili University of Health & Allied Sciences, Dar es Salaam, Tanzania; <sup>2</sup>Department of Paediatric Cardiology, Jakaya Kikwete Cardiac Institute, Dar es Salaam, Tanzania

**Background and Aim:** Aortopulmonary window (APW) is a rare congenital heart disease resulting from the deficiency of a septum between the ascending aorta and the main pulmonary artery resulting in an abnormal communication between the two major arteries. Patients with APW usually present with early signs and symptoms of congestive heart failure and pulmonary hypertension (PH).

**Method:** We present a case of a three-year-old boy of African descent who presented to our outpatient clinic with features of heart failure and was diagnosed to have a large patent ductus arteriosus (PDA) by transthoracic echocardiography (ECHO). He subsequently underwent PDA surgical ligation but one month later, he still had persistent symptoms of heart failure. A repeat ECHO revealed a large undetected APW, which was confirmed by a cardiac computed tomography (CT) scan. A diagnostic right heart catheterization ruled out irreversible PH and the patient subsequently underwent successful surgical patch repair.

**Results:** In our case, the diagnosis of APW was initially missed by ECHO. It is difficult to differentiate APW from other left-to-right shunts like a PDA therefore it can be easy to miss APW in the presence of other conditions. APW can also lead to irreversible PH early in infancy therefore, un-repaired or late repair of APW can lead to poor outcomes and mortality. Our case was unique in the sense that PH was still reversible even at the age of three years. Hence, even delayed diagnosis of APW might still be operable beyond infancy therefore thorough assessment of operability must be done in all patients, regardless of their age at presentation.

**Conclusions:** Clinical and transthoracic echocardiographic diagnosis of an APW can be challenging especially if it is large or associated with other large left-to-right shunt lesions like PDA. Therefore, a high index of suspicion is required for patients who present with early signs of congestive heart failure and pulmonary hypertension. A thorough and systematic transthoracic echocardiogram is recommended to avoid missing this rare lesion. Other noninvasive imaging modalities such as cardiac CT scan or magnetic resonance imaging with angiography may be required to confirm the diagnosis.

**Keywords:** Aortopulmonary window, Case report, Patent ductus arteriosus, Pulmonary hypertension

#### P-209

##### **Pre-glenn lymphatic abnormalities on MRI in single-ventricle congenital heart defects**

Rasmus Kristensen<sup>1</sup>, Benjamin Kelly<sup>2</sup>, Vibeke E. Hjortdal<sup>1</sup>, Yoav Dori<sup>3</sup>  
<sup>1</sup>Department of Cardiothoracic Surgery, Copenhagen University Hospital – Rigshospitalet, Copenhagen, Denmark.; <sup>2</sup>Department of Cardiothoracic Surgery, Aarhus University Hospital, Skejby, Denmark.; <sup>3</sup>Division of Cardiology, Children’s Hospital of Philadelphia, Philadelphia, Pennsylvania, USA

**Background and Aim:** Lymphatic abnormalities such as plastic bronchitis and protein-losing enteropathy are seen after staged surgery for single ventricle defects. Lymphatic abnormalities in the neck and thorax have been shown on T2-weighted MRI after the Glenn- and prior to the Fontan operation. The type of lymphatic abnormality correlates with the surgical outcomes following the Fontan operation. It is unknown if the lymphatic abnormalities are already present prior to the Glenn operation.

**AIM:** Determining if lymphatic abnormalities in the neck and thorax are present in children with single-ventricle physiology prior to their Glenn operation.

**Method:** Patients with a single-ventricle defect and a T2-weighted MRI-scan prior to their Glenn operation (Superior Cavopulmonary Connection) at The Children’s Hospital of Philadelphia from 2012 to 2022 were reviewed. Lymphatic abnormalities were categorized from type 1 (little T2 signal in supraclavicular region) to type 4 (supraclavicular, mediastinal and lung parenchymal T2-signal). Outcomes were the distribution of lymphatic abnormalities on T2-weighted MRI prior to Glenn operation, and secondary; Glenn take down, Fontan takedown, heart transplant, plastic bronchitis, chylothorax, prolonged pericardial effusion, chylous pericardial effusion, mortality. Comparison was done using analysis of variance, the Kruskal-Wallis test and Fisher’s exact test.

**Results:** The study population included 71 children: 30 (42%) with hypoplastic left heart syndrome and 41 (58%) with non-hypoplastic left heart syndrome. A total of 42 (59%) were classified as type 1-2 lymphatic abnormality, 15 (21%) as type 3 and 14 (20%) as type 4. Genetic abnormalities were identified in 39 (55%) patients. Pre-Glenn Chylothorax was recorded in 12 (17%, type 3 or 4) patients, and chylous pericardial effusion in 2 (3%) patients. No children had plastic bronchitis. Ten (14%) patients were deceased; two with type 1-2 lymphatic abnormality, four with type 3 and four with type 4,  $p = 0.01$ .

Mortality was significantly different between types 1-2 of lymphatic abnormalities vs type 3 ( $p = 0.04$ ) and vs. type 4 ( $p = 0.03$ ).  
**Conclusions:** Lymphatic abnormalities in the neck and thorax can be found on T2-weighted MRI in children with single-ventricle physiology prior to their Glenn-operation and mortality is more prevalent with advanced grades of abnormalities.

**Keywords:** congenital heart defect, lymphatic abnormalities, staged palliation, hypoplastic left heart syndrome, magnetic resonance imaging, MRI.

#### P-210

##### **Re-intervention after repair of aortic coarctation or aortic ARCH reconstruction**

Yangxue Sun, Yuanrui Gu, Shuo Dong, Haitao Xu, Chuhao Du, Jie Dong, HuaWei Pei, Yang Liu, Hongwei Guo, Jun Yan  
 Fuwai Hospital, National Center for Cardiovascular Diseases, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing, China

**Background and Aim:** Aortic coarctation is still faced with reoperation due to restenosis, hypertension and other complications even after the first intervention treatment, which affects the long-term survival rate of patients. We assessed the outcomes of late re-intervention after repair of aortic coarctation or aortic arch reconstruction.

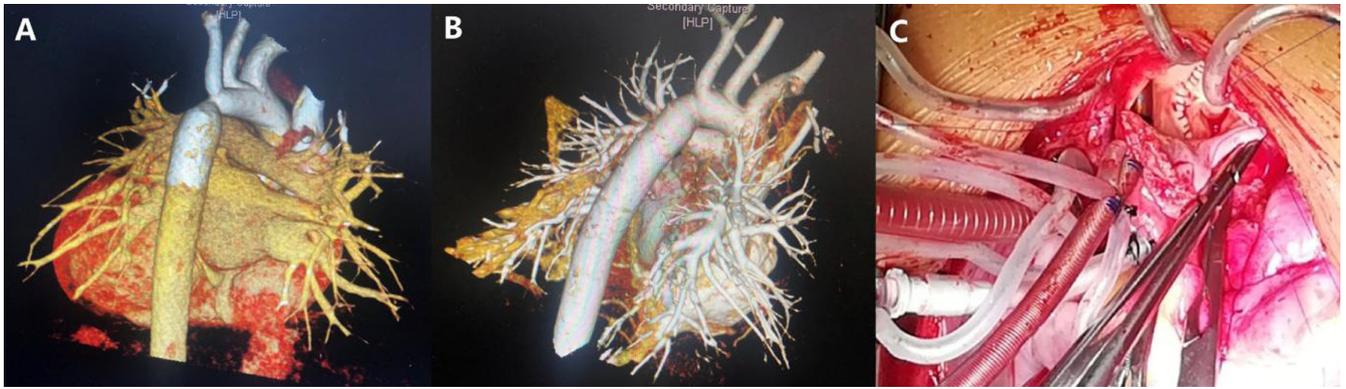
**Method:** From October 2007 to July 2022, 67 patients had operations or endovascular procedures after previous coarctation repair or aortic arch reconstruction. We excluded patients who underwent Norwood operation for hypoplastic left heart syndrome, double aortic arch and who required early reoperation for bleeding and incision infection.

**Results:** Middle age at first intervention was re-intervention was 1 year old (4 days to 52 years old). The average interval between coarctation repair and first re-intervention was  $56.7 \pm 67.46$  months. Reasons for re-intervention were restenosis ( $n = 31$ ), aortic valve re-intervention ( $n = 15$ ), stenosis of the left ventricular outflow tract ( $n = 13$ ), mitral valve re-intervention ( $n = 11$ ), aneurysm ( $n = 3$ ), vascular compression of bronchi ( $n = 2$ ), stenosis of the left ventricular outflow tract ( $n = 2$ ), coronary stent implanting ( $n = 2$ ), cardiac arrhythmia ( $n = 2$ ), pseudoaneurysm ( $n = 1$ ). There was 2 early mortality. There are 10 patients had twice re-intervention. In restenosis group, 14 patients (45.2%) underwent operative procedures including an extra-anatomic conduit ( $n = 6$ ), patch aortoplasty ( $n = 5$ ), interposition graft ( $n = 3$ ). 17 patients underwent balloon dilatation treatment. In the vascular compression of bronchi group, all patients underwent descending aortic translocation.

**Conclusions:** Although the early mortality of aortic coarctation is low, multidisciplinary cooperation is still needed to conduct lifelong health management for patients to further improve the long-term survival rate and avoid re-intervention. The selection of re-intervention treatment should consider the severity of the disease, the patient’s physical condition and the incidence of related complications.

**Keywords:** Aortic coarctation, aortic arch reconstruction, re-intervention, re-coarctation

Autologous pericardium patch aortoplasty



A. Computer tomography angiography before re-intervention of re-coarctation of aorta. B. Computer tomography angiography after re-intervention of re-coarctation of aorta. C. Picture of using untreated autologous pericardium underwent patch aortoplasty

**P-211**

**The mechanistic relationship between shape and function in repaired coarctation of the aorta**

Liam Swanson<sup>1</sup>, Jan Lukas Bruse<sup>2</sup>, Emilie Sauvage<sup>1</sup>, Malebogo Ngoepe<sup>3</sup>, Tain Yen Hsia<sup>4</sup>, Silvia Schievano<sup>1</sup>

<sup>1</sup>Institute of Cardiovascular Science, University College London and Great Ormond Street Hospital for Children, London, United Kingdom; <sup>2</sup>Vicomtech Foundation, Basque Research and Technology Alliance (BRTA), Donostia-San Sebastian, Spain; <sup>3</sup>Mechanical Engineering, University of Cape Town, Cape Town, South Africa; <sup>4</sup>Heart Center at Arnold Palmer Hospital for Children, Orlando, FL, USA

**Background and Aim:** Specific aortic arch shapes are known to be associated with worse left ventricular function and chronic hypertension late after aortic coarctation (CoA) repair, even in the absence of residual obstruction or stenosis. A previously validated, 3D statistical shape model of successfully repaired CoA arches (53 asymptomatic patients, 12-38 years post-repair) showed that elongated ascending aortas with a high arch height-to-width ratio, a relatively short proximal transverse arch, and a relatively dilated descending aortas are associated to poorer late outcomes. In this study, we aimed to simulate blood flow through these shapes using computational fluid dynamics (CFD), to investigate possible mechanistic explanations for these phenomenological findings.

**Method:** CFD was carried out on the shapes associated with the following high (+2 standard deviations, SD) and low (-2SD) cardiovascular function parameters: left ventricular ejection fraction (LVEF), indexed mass (iLVM), indexed end diastolic volume (iLVEDV) and systolic blood pressure (BP). A typical cardiac cycle was simulated in all 8 shapes to calculate and compare the maximum pressure gradient ( $\Delta P$ ) between the ascending and descending aorta and the rate of viscous energy loss (ELv). All arches had the same input blood flow conditions so that the differences in shape would be the driving factor of any resulting haemodynamic variations.

**Results:** Overall pressure gradients were clinically insignificant (<8 mmHg) in all 8 arch shapes. However, in all 4 shapes related to worse cardiovascular function (low LVEF, high iLVEDV, high iLVM and high BP),  $\Delta P$  and ELv were uniformly higher than in the 4 shapes related to better cardiovascular performance (Figure). The largest  $\Delta P$  difference was recorded between the BP shape modes (3.4 mmHg) and the maximum peak and average ELv percentage differences were between the BP shape modes (27%), and EF shape modes (32%), respectively.

**Conclusions:** This study sheds light into the links between cardiovascular shape and function late after successful CoA repair. Higher viscous energy losses persist in those aortic arch shapes associated with poorer cardiovascular performance. Thus, medical and surgical strategies may be needed to mitigate this insidious, but unremitting liability in CoA patients.

**Keywords:** Coarctation of the Aorta, Computational Fluid Dynamics, Statistical Shape Modelling, Simulation, Congenital Heart Disease

SSM		Max Pressure Gradient [mmHg]	Peak Viscous Energy Loss [mW]	Ave Viscous Energy Loss [mW]
EF	Low	30.3	86.8	14.9
	High	28.8	72.8	11.3
		5 %	19 %	32 %
iLVM	Low	26.2	68.0	10.9
	High	30.0	91.0	15.4
		13 %	25 %	29 %
BP	Low	27.8	67.0	11.6
	High	31.0	91.6	13.6
		11 %	27 %	15 %
iLVEDV	Low	28.3	74.9	11.5
	High	30.1	82.5	13.9
		6 %	9 %	17 %

**Results Figure** CFD results for the 8 CoA anatomies associated to high and low cardiovascular performance parameters. Red indicates worse and green, better, cardiovascular conditions. Percentage differences are calculated with respect the high state of each cardiovascular function parameters.

## P-212

**Impact of surgical strategy and post-repair transverse aortic ARCH size on late hypertension after coarctation repair during infancy**

Sanam Saji<sup>1</sup>, David Hoganson<sup>2</sup>, Sitaram Emani<sup>2</sup>, Elia Eleni<sup>3</sup>, Lynn Sleeper<sup>5</sup>, Tor Biering Sørensen<sup>4</sup>, Ashwin Prakash<sup>5</sup>

<sup>1</sup>Department of Cardiology, Boston Children's Hospital, Boston, MA, USA; <sup>2</sup>Department of Cardiology, Copenhagen University Hospital - Herlev and Gentofte, Hellerup, Denmark; <sup>3</sup>Department of Cardiac Surgery, Boston Children's Hospital, Boston, MA, USA; <sup>4</sup>Department of Surgery, Harvard Medical School, Boston, MA, USA; <sup>5</sup>Department of Cardiology, Boston Children's Hospital, Boston, MA, USA; <sup>6</sup>Department of Cardiology, Copenhagen University Hospital - Herlev and Gentofte, Hellerup, Denmark; Department of Biomedical Sciences, Faculty of Health and Medical Sciences, University of Copenhagen, Copenhagen, Denmark; The Copenhagen City Heart Study, Copenhagen University Hospital - Bispebjerg and Frederiksberg, Copenhagen, Denmark; <sup>7</sup>Department of Cardiology, Boston Children's Hospital, Boston, MA, USA; <sup>8</sup>Department of Pediatrics, Harvard Medical School, Boston, MA, USA; <sup>9</sup>Department of Cardiology, Boston Children's Hospital, Boston, MA.

**Background and Aim:** Transverse aortic arch (TAA) hypoplasia is common in patients with coarctation of the aorta (CoA) and is usually not addressed during the initial repair unless severe. Despite adequate treatment, 30% - 60% of adults with repaired CoA develop hypertension (HTN) which contributes to higher rates of morbidity and mortality. Previously, an association between late TAA hypoplasia and HTN has been found, but its relationship with surgical strategy is unclear. To study the impact of the surgical strategy on late HTN, we assessed the relationship between TAA size immediately after surgery and late HTN.

**Method:** We retrospectively reviewed patients who underwent surgical repair of CoA during infancy at a single institution between 1980-2010 with ≥10 years follow-up after repair, excluding those with atypical coarctation, major associated heart defects, and residual isthmus narrowing. TAA diameter and its z-score was measured on echocardiography immediately post-repair. Systemic HTN at latest follow-up was assessed using standard task-force criteria.

**Results:** A total of 130 patients (mean current age 19 years, range 10-40 years; 58% male) underwent surgical repair of CoA (76% via thoracotomy, 24% via sternotomy; type of repair – resection and end-to-end anastomosis 62%, extended end-to-end anastomosis 30%, subclavian flap 5%, extensive arch repair with patch 4%), at a median age of 14 (IQR 7-62) days. Median post-repair TAA diameter z-score was -2.04 (IQR -2.69, 1.24). After a median follow-up duration of 18 years (10-30 years), 43/130 (33.1%) patients developed HTN. After controlling for age at repair, gender, and presence of a genetic syndrome, HTN was not associated with immediate post-repair TAA diameter z-score, ( $p = 0.42$ ), type of surgical incision ( $p = 0.99$ ), and type of surgical repair ( $p = 0.56$ ).

**Conclusions:** In patients undergoing surgical repair of CoA during infancy, late HTN was not associated with immediate post-repair TAA size or type of surgical repair. Since late HTN has previously been associated with late TAA hypoplasia, these results suggest that factors other than surgical strategy, such as differential growth of the TAA during childhood, may have a larger impact on development of late HTN.

**Keywords:** Coarctation, Congenital heart disease, Transverse Aortic Arch, Hypoplasia, Hypertension

## P-213

**Clinical correlates of impaired individualised perioperative brain growth in infants with congenital heart disease**

Alexandra Bonthron<sup>1</sup>, Daniel Cromb<sup>1</sup>, Alessandra Maggioni<sup>1</sup>, Paul Crawley<sup>2</sup>, Ralica Dimitrova<sup>3</sup>, Christopher Kelly<sup>1</sup>, Lucilio Cordero Grande<sup>4</sup>, Olivia Carney<sup>1</sup>, Alexia Egloff<sup>1</sup>, Emer Hughes<sup>1</sup>, Joseph Hajnal<sup>1</sup>, John Simpson<sup>5</sup>, Kuberan Pushparajah<sup>5</sup>, Mary Rutherford<sup>1</sup>, A David Edwards<sup>2</sup>, Jonathan O'muircheartaigh<sup>6</sup>, Serena Counsell<sup>1</sup>

<sup>1</sup>Centre for the Developing Brain, School of Biomedical Engineering and Imaging Sciences, King's College London, London, UK; <sup>2</sup>Centre for the Developing Brain, School of Biomedical Engineering and Imaging Sciences, King's College London, London, UK and MRC Centre for Neurodevelopmental Disorders, King's College London, London, UK; <sup>3</sup>Centre for the Developing Brain, School of Biomedical Engineering and Imaging Sciences, King's College London, London, UK and Department for Forensic and Neurodevelopmental Sciences, Institute of Psychiatry, Psychology and Neuroscience, King's College London, London, UK; <sup>4</sup>Centre for the Developing Brain, School of Biomedical Engineering and Imaging Sciences, King's College London, London, UK and Biomedical Image Technologies, ETSI Telecomunicación, Universidad Politécnica de Madrid and CIBER-BBN, ISCIII, Madrid, Spain; <sup>5</sup>Paediatric Cardiology Department, Evelina London Children's Healthcare, London, United Kingdom; <sup>6</sup>Centre for the Developing Brain, School of Biomedical Engineering and Imaging Sciences, King's College London, London, UK, MRC Centre for Neurodevelopmental Disorders, King's College London, London, UK and Department for Forensic and Neurodevelopmental Sciences, Institute of Psychiatry, Psychology and Neuroscience, King's College London, London, UK

**Background and Aim:** Infants with Congenital Heart Disease (CHD) are at risk of impaired neurodevelopment. We aimed to characterize perioperative brain growth in individual infants with CHD and assess the relationship with clinical risk factors.

**Method:** 36 infants with critical or severe CHD [19 male, median (IQR) gestational age (GA) at birth 38.5 (38.1-39.0) weeks] underwent pre- and postoperative 3T brain MRI on a Philips Achieva system situated on the neonatal unit at St Thomas' Hospital London [Preoperative scan GA 39.4 (38.7-39.9); Postoperative scan GA 41.9 (41.0-42.7)]. T2-weighted images were segmented using a neonatal-specific algorithm. Normative curves of typical volumetric brain development were generated using a data-driven technique applied to 219 healthy infants from the developing Human Connectome Project. Z-scores, representing the degree of positive or negative deviation from the normative mean for postmenstrual age at scan, days of life and sex, were calculated for each brain volume from each infant with CHD before and after surgery. The degree of change in Z-scores was correlated with clinical risk factors using partial Spearman's rank correlations.

**Results:** Perioperative growth was impaired in cortical grey matter, white matter, cerebellum, brainstem, right thalamus, right lentiform, total tissue volume and extracerebral CSF spaces. The degree of impaired brain growth was associated with a longer postoperative intensive care stay (Table 1). Higher preoperative serum creatinine levels were associated with impaired brainstem, caudate nuclei and right thalamus growth. Older postnatal age at surgery was associated with impaired brainstem and right lentiform growth (Table 1). Longer cardiopulmonary bypass duration was associated with impaired brainstem and right caudate growth (Table 1).

**Conclusions:** Brain growth was impaired in the acute postoperative period. Brainstem growth was particularly vulnerable to clinical course, with preoperative creatinine levels, later age at surgery, longer duration of cardiopulmonary bypass, and longer time on intensive care postoperatively associated with impaired growth. Impaired growth in the right lentiform and right caudate nuclei was associated with later age at surgery and longer time on

cardiopulmonary bypass respectively. This may reflect a particular vulnerability to acute and chronic hypoxic injury. Overall, these results provide novel opportunities to optimize perioperative brain growth in individual infants with CHD.

**Keywords:** Magnetic Resonance Imaging, Brain Growth, risk factors, intensive care, perioperative care

Region	Relationship between Z-score change and duration of intensive care stay   age at surgery		Relationship between Z-score change and preoperative creatinine   age at sample		Relationship between Z-score change and postnatal age at surgery		Relationship between Z-score change and time on cardiac bypass   intensive care stay	
	p	PFDR	p	PFDR	p	PFDR	p	PFDR
Extracerebral CSF	0.022	0.977	-0.094	0.712	-0.174	0.599	-0.162	0.478
Cortical grey matter	<b>-0.507</b>	<b>0.015</b>	-0.308	0.175	-0.381	0.237	-0.080	0.703
White matter	-0.349	0.086	-0.230	0.286	-0.399	0.126	-0.162	0.478
Ventricles	0.002	0.990	-0.066	0.774	-0.069	0.861	-0.185	0.478
Cerebellum	<b>-0.495</b>	<b>0.015</b>	-0.345	0.128	-0.333	0.173	-0.164	0.478
Brainstem	<b>-0.419</b>	<b>0.039</b>	<b>-0.503</b>	<b>0.033</b>	<b>-0.481</b>	<b>0.042</b>	<b>-0.518</b>	<b>0.026</b>
Right Caudate	-0.214	0.336	<b>-0.434</b>	<b>0.039</b>	-0.232	0.864	<b>-0.587</b>	<b>0.009</b>
Left Caudate	-0.134	0.593	<b>-0.476</b>	<b>0.033</b>	-0.029	0.903	-0.247	0.350
Right Lentiform	-0.338	0.089	-0.032	0.859	<b>-0.477</b>	<b>0.042</b>	-0.011	0.949
Left Lentiform	-0.097	0.698	0.173	0.435	-0.268	0.331	0.091	0.695
Right Thalamus	-0.391	0.053	<b>-0.436</b>	<b>0.038</b>	-0.348	0.162	-0.390	0.335
Left Thalamus	<b>-0.456</b>	<b>0.027</b>	-0.279	0.189	-0.219	0.467	-0.261	0.109
Total Tissue Volume	<b>-0.590</b>	<b>0.004</b>	-0.279	0.189	-0.403	0.126	-0.211	0.412

Results in bold are significant

## P-214

### Sternal opening width is associated with increased risk for capillary leak syndrome and death in neonates and infants after open-chest cardiac surgery

Christoffer Lindbom<sup>1</sup>, Lars Lindberg<sup>2</sup>, Phan Kiet Tran<sup>3</sup>, Katarina Hanséus<sup>1</sup>, Petru Liuba<sup>1</sup>

<sup>1</sup>Cardiology, Pediatric Heart Centre, Skåne University Hospital, Lund, Sweden; <sup>2</sup>Anaesthesiology and Intensive Care, Pediatric Heart Centre, Skåne University Hospital, Lund, Sweden; <sup>3</sup>Cardiac Surgery, Pediatric Heart Centre, Skåne University Hospital, Lund, Sweden

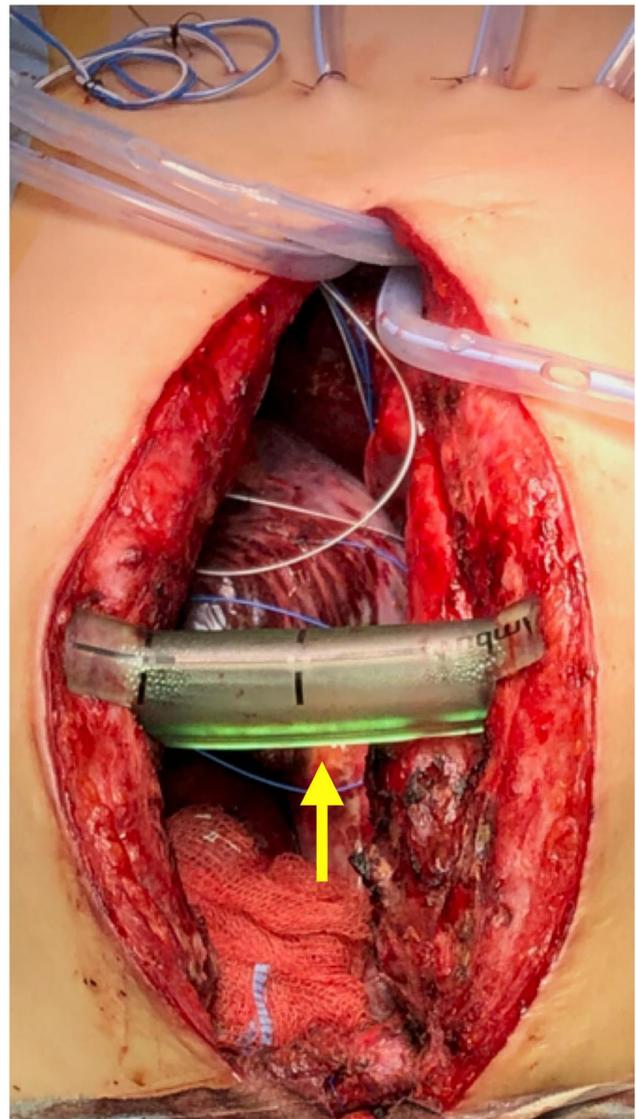
**Background and Aim:** Capillary leak syndrome (CLS) after open-heart surgery is a major cause of postoperative morbidity and mortality in neonates and infants with congenital heart disease (CHD). Since previous studies targeting inflammatory mediators and complement system abnormalities have not fully explained the prevalence of postoperative CLS, there is a need to investigate other factors that may contribute to it. Delayed sternal closure (DSC), which involves stenting the sternal halves apart with a modified plastic tube (Figure 1), has previously been speculated to be associated with increased risk for CLS. This retrospective study aimed to determine the relationship between postoperative CLS and stent length in neonates and infants undergoing open-chest cardiac surgery with DSC.

**Method:** We reviewed 164 neonates and infants (< 1 year of age at surgery; median age 7 days, range 1-249 days; median body weight 3.4 kg, range 1.6-6.2 kg) who underwent open-heart surgery for CHD with DSC between 2016 and 2021. Eligibility criteria for CLS were clinical diagnosis by a pediatric intensive care unit (PICU) physician, DSC > 3 days, and postoperative PICU length of stay > 10 days.

**Results:** Of the 164 patients included, 12 (7.3%) fulfilled the eligibility criteria for CLS. Nine postoperative deaths were observed in the cohort, including three in the CLS group. The mean difference in stent length per kg (SL/kg) between patients with and without postoperative CLS was 1.8 mm ( $p = 0.07$ ). Univariate analysis of the entire cohort revealed a correlation between SL/kg and postoperative mortality ( $r = 0.16$ ;  $p < 0.05$ ). In the multivariate analysis, higher age at surgery ( $r = 0.019$ ;  $p < 0.05$ ), increased cardiopulmonary bypass (CPB) time ( $r = 0.013$ ;  $p < 0.05$ ), and greater SL/kg ( $r = 0.20$ ;  $p < 0.05$ ) were significantly associated with increased risk of mortality.

**Conclusions:** Neonates and infants who developed postoperative CLS tended to have a greater sternal opening width. The latter, along with age at surgery and CPB time were identified as risk factors for postoperative mortality.

**Keywords:** congenital heart disease, capillary leak, cardiac surgery, young age, delayed sternal closure, mechanical factors



**Figure 1:** Sternal wound after stenting with a modified plastic tube (the stent is marked with an arrow).

## P-215

**Ross operation in children**

Petr Bukovský, Václav Chaloupecký, Jan Janoušek, Roman Gebauer  
Children's Heart Centre, 2nd Faculty of Medicine, Charles University  
and Motol University Hospital, Prague, Czech Republic

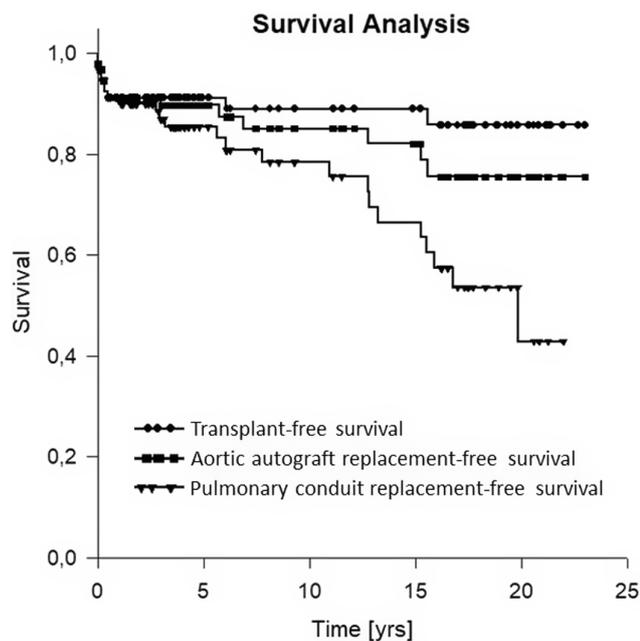
**Background and Aim:** Data on long-term outcomes of Ross operation in children are limited. We decided to evaluate long-term results in a single-centre cohort of patients after Ross operation performed in childhood and adolescence.

**Method:** Retrospective analysis of all patients who underwent Ross/Ross-Kono operation at our institution between 1990-2020. Data was crossmatched with the National Death Registry and registries of cardiovascular interventions. Death from any cause/heart transplantation and need for surgical or catheter reintervention on the aortic autograft/conduit in pulmonary position were considered as study endpoints. Kaplan-Meier curves were used to assess long-term outcomes.

**Results:** Ninety-five patients, median age 4.2 (IQR 0.2-7.9 years, neonates 11.5%), who underwent Ross operation or its Konno modification and were followed-up for a median of 4.1 (IQR 1.5-16.5) years were included in the study. Previous procedures (N = 130) were performed in 81 (85.3%) patients with balloon aortic valvuloplasty being most frequent (69.2%). There were 4 early and 3 late deaths. Four patients underwent heart transplantation. Long-term transplant-free survival at 10 and 20 years after surgery was 88.9% and 85.8%, resp. Surgical (N = 37) and transcatheter (N = 7) reinterventions were performed in a total of 23 patients (24.2%). Ten and 20-years survival free from reintervention on the aortic autograft/conduit in pulmonary position was 85.0/78.3 and 75.4/42.9%, resp

**Conclusions:** Ross operation represents the only option for aortic valve replacement in neonates, infants and younger children with favourable transplant-free survival. Freedom from reinterventions is negatively affected mainly by reinterventions on the conduit in pulmonary position. Aortic autograft reinterventions are relatively rare.

**Keywords:** Ross, Konno, children, neonates, autograft



Survival Analysis

## P-216

**Use of a postoperative care management pathway reduces the incidence of chylothorax post fontan surgical palliation**

Alanna Ash<sup>1</sup>, Lindsay Ryerson<sup>2</sup>, Vijay Anand<sup>2</sup>, Jennifer Conway<sup>1</sup>,  
George Slim<sup>1</sup>, Iman Naimi<sup>1</sup>, Luke Eekersley<sup>1</sup>

<sup>1</sup>Division of Cardiology, Stollery Children's Hospital, Edmonton, Alberta, Canada; <sup>2</sup>Division of Critical Care, Stollery Children's Hospital, Edmonton, Alberta, Canada

**Background and Aim:** Pleural fluid drainage is a normal consequence of Fontan palliation. Use of a multifaceted postoperative management strategy reduces pleural drainage days and length of stay (LOS) in Fontan patients; however, the efficacy of individual elements has not been identified.

The aims of this study were to: 1) evaluate the effect of a post Fontan care pathway on incidence of chylothorax and median time to chest tube removal (TTCTR); and 2) the effect on these endpoints of individual factors within this bundled approach.

**Method:** Between 01/17 and 04/22 our institutional approach to post Fontan care fell into three categories: Group 1 (n = 36) followed no standardized care approach, group 2 (n = 30) received a prophylactic chylothorax diet (fat content <5%) postoperatively, and group 3 (n = 57) followed a Fontan care pathway which included a chylothorax diet, fluid restriction, supplemental O<sub>2</sub> and aggressive diuresis. The incidence of chylothorax and TTCTR was compared between groups. Predictors of TTCTR were analyzed using a linear regression model, adjusting for covariates

**Results:** Groups were similar by Fontan type, fenestration rates and ventricular morphology, cardiopulmonary bypass time and conduit size. Laboratory confirmed chylothorax decreased significantly with use of a bundled approach (Group 3, 9% vs Groups 1 and 2, 28% and 33% respectively, p = 0.011), with no change in TTCCR (median 7, IQR 6-12 days). Univariate factors associated with median TTCTR included chylothorax ( $\beta = 13.7$ days, p<0.001), additional procedures at time of Fontan ( $\beta = 2.4$  per procedure p = 0.017), need for Fontan revision or takedown ( $\beta = 11.7$ days, p = 0.018) and minor ( $\beta = 5.1$ , p=0.01) or major ( $\beta = 8.6$ , p = 0.001) complications. On multivariable analysis, chylothorax ( $\beta = 10.6$ days, p<0.001) and major complications ( $\beta = 8.6$ days, p = 0.017) were associated with increased TTCTR. When chylothorax was excluded from multivariable analysis, Group 3 membership showed a trend toward a decrease in TTCTR (p = 0.075).

**Conclusions:** A bundled therapy approach was associated with reduced laboratory confirmed chylothorax, whereas diet change alone was not. Additional studies with larger sample sizes are needed to determine if a decrease in the incidence of chylothorax using this management approach will translate into better short-term outcomes post Fontan.

**Keywords:** chylothorax, Fontan, postoperative management

## P-217

**Outcomes of isolated right aortic ARCH surgery in the young. A 6 year experience in a United Kingdom Tertiary paediatric cardiac centre**

Peter John Lillitos, Carin Van Doorn, Elspeth Brown

Department of Congenital Cardiology, Leeds Teaching Hospitals NHS Trust, United Kingdom

**Background and Aim:** Prevalence of right aortic arch (RAA) is traditionally 0.1% but rising with improved antenatal screening. Associated vascular ring may cause tracheal-oesophageal compression and surgical relief is indicated with feeding and/or breathing

symptoms. Operative mortality is low, but little is known about other operative complications or resolution of symptoms.

**Method:** Retrospective review of isolated RAA vascular-ring operations performed in children in our Centre between 2016–2021. **Results:** There were 27 patients with isolated RAA (out of 145 antenatally diagnosed) and 1 postnatally diagnosed that were symptomatic. Eight were excluded from the study as postnatal diagnosis were double-aortic-arch, and another had additional diagnosis of ventricular-septal-defect requiring concomitant repair. Of the 19 patients (11 males) undergoing isolated RAA repair, symptoms were respiratory (10 patients), feeding difficulties (2) and combined (7). Additional diagnoses were chromosome-8-inversion (1) and Smith-Magenis syndrome (1). Mean gestational age at birth was 38.9 weeks (SE 0.26). All underwent cross-sectional imaging. Surgery was via left thoracotomy, performed at mean 46.6 weeks (SE 7.7) after birth. There was no mortality. Postoperative complications occurred in 5 (26%): chylothorax (3), vocal-cord (VC) palsy (1), wound infection (1). Mean length of stay for uncomplicated recovery was 3.7 days vs 8.6 days in those with complications ( $p0.04$ ). All are alive at mean follow up 59 weeks (SE 13). The patient with VC palsy continues to have respiratory symptoms and has been diagnosed with laryngeal cleft. All others are free from pre-operative symptoms, mean time to symptom resolution of 6.8 weeks (SE 1.9). However, 4 have different respiratory symptoms despite relief of external vascular compression demonstrated on bronchoscopy. These are bronchomalacia (1, with resolution of feeding problems 1 week postoperatively), asthmatic wheeze (1, resolution of stridor at 8 weeks) and lower respiratory tract symptoms with distal airway trapping (2, both with resolution of swallowing and stridor symptoms respectively at 3 weeks).

**Conclusions:** Repair of isolated RAA can be offered at low operative risk and offers good resolution of presenting symptoms. However, despite adequate relief of external vascular compression on the airway some have ongoing respiratory problems and the nature of this in relation to RAA pathology requires further investigation.

**Keywords:** right aortic arch, symptoms, surgery, outcomes, complications, children

## P-220

### Decision support for patients with complex univentricular physiology via digital twins

Jan Brüning, Pavlo Yeutushenko, Marie Schafstedde

Institute of Computer-assisted Cardiovascular Medicine; Charité - Universitätsmedizin Berlin

**Background and Aim:** Treatment of congenital heart defects usually aims at facilitating biventricular repair by either surgical or interventional procedures. However, depending on the type and complexity of the heart defect as well as the patient-specific anatomy, univentricular palliation might be the only viable treatment option. In those patients, the superior and inferior vena cava are connected to the pulmonary artery via staged surgeries, facilitating the so-called total cavopulmonary connection (TCPC). Both anatomy and hemodynamics of the TCPC are subject to severe patient-specific differences. Furthermore, univentricular patients often require additional surgical and/or interventional treatments adding to this heterogeneity. We aim to assess whether digital twins of the TCPC allow to provide additional anatomical and functional information for both diagnosis and treatment planning tailored to a specific patient.

**Method:** The anatomical digital twin of the TCPC is reconstructed from different imaging sources, as for example MRI, CT and

angiography. To also provide functional information, the patient-specific blood flow within the TCPC is calculated via computational fluid dynamics. This approach allows to provide detailed insight on relevant hemodynamic information such as pressures and velocities. Furthermore, the digital twins can be virtually altered to mimic different treatment strategies, such as interventional implantation of a covered stent or surgical implantation of a vascular graft.

**Results:** The project is currently ongoing and only preliminary results are available. The patient-specific anatomy of 8 univentricular patients was reconstructed. Treatment was required to either facilitate an even distribution of hepatic blood towards the left and right pulmonary artery or to reduce the energy loss caused by Fontan stenosis. For both applications, the digital twins were able to provide the necessary functional information and allowed to compare different treatment strategies.

**Conclusions:** Digital twins are a promising approach to overcome the obstacles imposed by the heterogeneity found in patients with univentricular palliation. The outcome prediction of different treatment strategies holds the promise to provide additional objective information, allowing to better stratify treatment risks and potential outcomes on a patient-specific level.

**Keywords:** Fontan, congenital heart disease, computational fluid dynamics, magnetic resonance imaging, computed tomography

## P-222

### Early mortality in infants born with neonatally-operated congenital heart defects and low or very-low birthweight: systematic review and meta-analysis

Neil Deridj<sup>1</sup>, Ali Ghanchi<sup>2</sup>, Damien Bonnet<sup>3</sup>, Pauline Adnot<sup>4</sup>, Laurent J. Salomon<sup>5</sup>, Jérémie F. Cohen<sup>6</sup>, Babak Khoshnood<sup>2</sup>

<sup>1</sup>Université de Paris, CRESS, INSERM, INRA, F-75004 Paris, France; <sup>2</sup>M3C-Paediatric Cardiology, Necker Enfants malades, AP-HP, Université de Paris, Paris, France; <sup>3</sup>Université de Paris, CRESS, INSERM, INRA, F-75004 Paris, France; <sup>4</sup>M3C-Paediatric Cardiology, Necker Enfants malades, AP-HP, Université de Paris, Paris, France; <sup>5</sup>Department of General Pediatrics and Pediatric Infectious Diseases, APHP, Université Paris Cité, Hôpital Necker-Enfants malades, Paris, France; <sup>6</sup>Department of Obstetrics and Fetal Medicine, APHP, Université Paris Cité, Hôpital Necker-Enfants malades, Paris, France; <sup>7</sup>Université de Paris, CRESS, INSERM, INRA, F-75004 Paris, France; Department of General Pediatrics and Pediatric Infectious Diseases, APHP, Université Paris Cité, Hôpital Necker-Enfants malades, Paris, France.

**Background and Aim:** Mortality outcomes of children with isolated (i.e., not associated with chromosomal anomalies, malformations from other systems or syndromes) neonatally-operated congenital heart defects (CHD) born low, moderately-low or very-low birthweight (i.e., LBW < 2500g, MLBW 1500–2499g and VLBW <1500 g, respectively) remain ambiguous. To evaluate early mortality outcomes of infants born with neonatally-operated isolated CHD and LBW, MLBW or VLBW

**Method:** We searched Medline and Embase (inception until October 2021) and included studies that evaluated early mortality. Risk of bias was assessed using the Critical Appraisal Skills Programme cohort checklist. Meta-analysis involved random-effects models. We explored variability in mortality across birthweight subgroups, CHD types, and study designs.

**Results:** From 2, 035 reports, we included 23 studies in qualitative synthesis, and the meta-analysis included 11 studies (1, 658 CHD cases), divided into 30 subcohorts. Risk of bias was low in 4/11 studies. Summary mortality before discharge or within one month

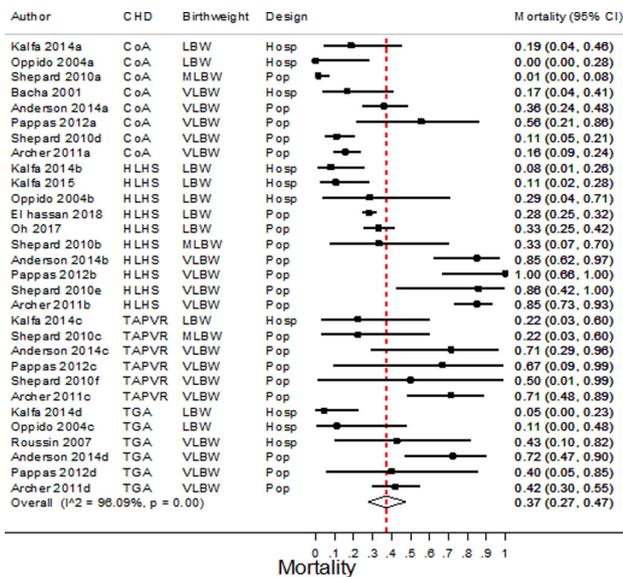
after surgery was 37% (95%CI 27-47). Early mortality varied by birthweight (VLBW 56%, MLBW 15%, LBW 16%;  $p = 0.003$ ) and CHD types (hypoplastic left heart syndrome [HLHS] 50%, total anomalous pulmonary venous return [TAPVR] 47%, transposition of the great arteries [TGA] 34%, coarctation of the aorta [CoA] 16%;  $p = 0.13$ ). Mortality was higher in population-based studies (49% vs. 10%;  $p = 0.006$ ).

**Conclusions:** One-third of infants born with neonatally-operated isolated CHD and LBW, MLBW, or VLBW die within 30 days after surgery. Mortality varies across infant and study characteristics. These results may help clinicians when assessing neonatal prognosis.

Registration: PROSPERO, CRD42020170289.

**Keywords:** Early mortality, low or very-low birthweight, meta-analysis, metaregression

Early mortality in infants born with severe neonatally-operated congenital heart defects and low or very-low birthweight: Meta-analysis



Legend: CHD, Congenital Heart Defect; LBW, Low birthweight (i.e., <2500g); MLBW, moderately low birthweight (i.e., 1500-2499g), VLBW, Very low birthweight (i.e., <1500g); HLHS, hypoplastic left heart syndrome; TAPVR, total anomalous pulmonary venous return; CoA, coarctation of the aorta; TGA, transposition of the great arteries

**P-223**

**3D surgical planning of aortic arch and coronary anomalies: how personalised decision tools using 3D technologies can help in achd complex cases**

István Hartyánszky<sup>1</sup>, László István Hartyánszky<sup>2</sup>, Sándor Varga<sup>1</sup>, Márta Katona<sup>3</sup>, Krisztina Kádár<sup>1</sup>, Andrea Kőszegi<sup>2</sup>, Gábor Kapus<sup>2</sup>, Bálint Csikós<sup>2</sup>, Andrea Székely<sup>2</sup>, Anikó Nagy<sup>2</sup>, Luca Kuthi<sup>2</sup>, Attila Tóth<sup>2</sup>, Hajni Vágó<sup>2</sup>, Levente Molnár<sup>2</sup>, István László Hartyánszky<sup>2</sup>

<sup>1</sup>University of Szeged, Department of Internal Medicine, Szeged, Hungary; <sup>2</sup>Heart Center, Semmelweis University, Budapest, Hungary; <sup>3</sup>University of Szeged, Department of Paediatrics, Szeged, Hungary

**Background and Aim:** The number of adult congenital heart disease patients in the world is growing, thanks to constant evolution and increasing number of modern surgical and catheter techniques. This patient group may consist of anatomically special, complex cases, with individual pathology. Surgical decision making in this challenging patient group is difficult and needs multidisciplinary approach. The use of modern imaging, 3D engineering softwares and 3D printing may help in indication, intraoperative decision making, and preoperative surgical planning. In 3 cases we would like to show our method of personalised decision making.

**Method:** Case 1 was a 22-year-old female patient with a history of non-operated Coarctation, case 2 was a 51-year-old female patient with a malignant type of anomalous origin of the right coronary, and case 3 was a 59-year-old female patient with Bland-White-Garland syndrome. In all cases we made 3D reconstructions from CT angiogram dicom files using 3D Slicer program and printed the models in real size using Flashforge Creator3 printer. In case 1 the model showed a severe hypoplasia of the descending aorta, and we decided not to operate. In case 2 we were able to plan the new orifice of the right coronary and avoided CABG surgery. In case 3 we were able to make a personalised surgical plan using off-pump left internal mammary artery bypass and ligation of the left anterior descending artery.

**Results:** In all cases decision making was enhanced with 3D modelling, and printing. In case 2 and 3 operations were uneventful, and control investigations at 3 and 12 months were negative.

**Conclusions:** 3D technologies can enhance visualisation, thus helping not only the understanding the anatomy or pathology, but giving the opportunity to make personalised, individual surgical solutions.

**Keywords:** ACHD, heart surgery, 3D reconstruction, 3D printing

**P-225**

**Ruptured sinus of valsalva aneurysms after surgical closure of ventricular septal defect: A lucky surgeon case report**

Gama Satria<sup>1</sup>, Bermansyah Boestham<sup>1</sup>, Ahmat Umar<sup>1</sup>, Aswin Nugraha<sup>1</sup>, Arie Hasiolan<sup>1</sup>, Fredi Heru Inwanto<sup>3</sup>, Ria Nova<sup>2</sup>, Desti Handayani<sup>4</sup>

<sup>1</sup>Department of Surgery, Division of Thoracic and Cardiovascular, Dr.Moh. Hoesin Hospital, Universitas Sriwijaya, Palembang, Indonesia; <sup>2</sup>Department of Child Health, Division of Pediatric Cardiology, Dr.Moh. Hoesin Hospital, Universitas Sriwijaya, Palembang, Indonesia; <sup>3</sup>Department of Anesthesiology and Intensive Care, Dr.Moh. Hoesin Hospital, Universitas Sriwijaya, Palembang, Indonesia; <sup>4</sup>Department of Child Health, Division of Pediatric Intensive Care, Dr.Moh. Hoesin Hospital, Universitas Sriwijaya, Palembang, Indonesia

**Background and Aim:** Sinus of Valsalva aneurysm (SVA) is a rare congenital heart disease that can cause severe clinical presentations when the aneurysm ruptures. SVA is frequently associated with other congenital defects, particularly ventricular septal defect (VSD), and is consequently seldom diagnosed. This study aims to report a previously unknown case of a ruptured sinus Valsalva aneurysm in a patient who had undergone VSD closure surgery.

**Method:** Case Report

**Results:** In this case report, we describe a 13-year-old girl with a previously undetected ruptured non-coronary sinus of the Valsalva aneurysm. This patient underwent surgical closure of the VSD, with minimal residual but no significant hemodynamic disturbance. However, one month after the operation, the patient complained of severe heart failure. She presented shortness of breath, chest pain, fatigue, and vibration in her chest.

Transthoracic echocardiogram (TTE) only showed what appeared to be an eccentric severe aortic regurgitant jet and residual VSD. Initially, it was thought of as a large residual VSD only, so it was decided to do re-operative VSD closure. However, upon reoperation of the residual VSD, a rupture of the sinus of Valsalva was also found. It was decided to close the residual VSD with a running suture technique, and aortic valve replacement was performed using a bioprosthetic valve. Finally, after aortic valve replacement was successfully performed, the patient was discharged with no residual symptoms.

**Conclusions:** Rupture of SVA, though rare, must be considered in a patient with severe aortic regurgitation and sudden onset of severe heart failure. Aortic valve replacement resulted in an excellent outcome for the patient.

**Keywords:** Sinus of Valsalva, rupture of aneurysm, VSD, surgical closure

### P-227

#### Extracorporeal membrane oxygenation in postcardiotomy pediatric patients: 18 years of experience

Bunty Ranchandani<sup>1</sup>, Inigo Velasco<sup>2</sup>, Blanca Torres<sup>1</sup>, Raul Sanchez<sup>1</sup>, Juvenal Rey<sup>1</sup>, Alvaro Gonzalez<sup>1</sup>, Luz Polo<sup>1</sup>, Paula Burgos<sup>2</sup>, Jorge Utanda<sup>2</sup>, Pascual Sanabria<sup>3</sup>, Laura Sanchez<sup>4</sup>, Luis Guereta<sup>5</sup>, Juan Jose Menendez<sup>6</sup>, Angel Aroca<sup>1</sup>

<sup>1</sup>Congenital Cardiovascular Surgery, Hospital Universitario La Paz and Ramon & Cajal, Madrid, Spain.; <sup>2</sup>Perfusion, Congenital Cardiovascular Surgery, Hospital Universitario La Paz, Madrid, Spain.;

<sup>3</sup>Anesthesiology and Reanimation, Hospital Universitario La Paz, Madrid, Spain.; <sup>4</sup>Neonatology, Hospital Universitario La Paz, Madrid, Spain.; <sup>5</sup>Paediatric Cardiology, Hospital Universitario La Paz, Madrid, Spain.; <sup>6</sup>Paediatric Intensive Care Unit, Hospital Universitario La Paz, Madrid, Spain.

**Background and Aim:** Postcardiotomy extracorporeal life support (PC-ECLS) is a life saving resource for patients with cardiopulmonary failure after cardiac surgery. We aim to report our experience of the last 18 years in paediatric patients.

**Method:** Single centre, retrospective study of all paediatric patients on PC-ECLS for postcardiotomy shock from January 2004 to June 2022. Primary outcomes were survival to ECMO and survival to discharge. Secondary outcomes were postoperative complications which were analysed following the ELSO data registry definitions. **Results:** 98 PC-ECLS runs in 88 paediatric patients; 27 neonates and 61 infants being d-TGA (N = 12) and tetralogy of Fallot (n = 12) the most frequent diagnosis respectively. One fifth of the patients had single ventricular physiology. Average ECMO duration was of 6.2 days. 76.3% of the ECLS runs were due to failure to wean from cardiopulmonary bypass. In 92.8% of the cases a veno-arterial ECMO was preferred. 90.8% of the cannulations were central. In 82.6% the therapy was initiated in the operation theatre. Survival to ECMO was of 70.4%; 75.8% in neonates and 68.1% in infants. Survival to discharge was of 53.4%; 51.8% in neonates and 54.1% in infants. Secondary outcomes: 45.9% patients had mediastinal cannulation site bleeding, 21.4% patients required reoperation, 29, 6% cohort had a positive blood culture.

**Conclusions:** Despite the high complication rate and dismal prognosis, PC-ECLS support provide encouraging survival benefit for patients who would otherwise die.

**Keywords:** Postcardiotomy, ECMO, Congenital heart disease, Congenital heart surgery

### P-228

#### Fulminant plastic bronchitis immediately post fenestrated fontan surgery treated with thoracic duct embolization

Sharon Katz<sup>1</sup>, Asaf Mandel<sup>2</sup>, Avital Rabinovitz<sup>3</sup>

<sup>1</sup>Department of Pediatric Cardiology, Hadassah Hebrew University Medical Center, Jerusalem, Israel.; <sup>2</sup>Pediatric Cardiac Critical Care Unit, Hadassah Hebrew University Medical Center, Jerusalem, Israel.;

<sup>3</sup>Congenital Cardiovascular Surgery Department, Hadassah Hebrew University Medical Center, Jerusalem, Israel.

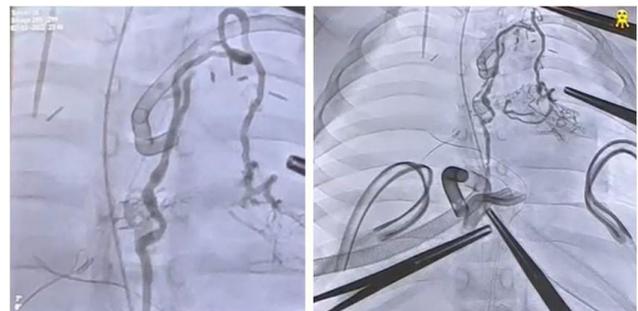
**Background and Aim:** Plastic bronchitis type II is a serious complication after surgeries for congenital heart defects, occurring most frequently after Fontan and less common after Glenn, arterial switch, and Tetralogy of Fallot's repair operations. Significant mortality rates of up to 30-60% have been reported. According to the current literature, plastic bronchitis usually presents months to years after surgery.

**Method:** We here describe a case of 4 years old girl, with single right ventricle physiology, who underwent fenestrated Fontan. Extensive, mainly fibrinoid casts started accumulating in her bronchi early in post-operative day 1, obstructing the airways.

**Results:** Due to insufficient ventilation and oxygenation and hemodynamic instability she required veno-arterial ECMO support. Treatment with inhaled anti-fibrinolytic or anti-coagulants was contra-indicated due to airway bleeding. Albeit recurrent bronchoscopies with casts removal, there was no significant improvement and large casts were continuously and rapidly formed. Angiography demonstrated no stenosis in the Glenn-Fontan-pulmonary arteries circulation. Therefore, on post-operative day 8 she underwent lymphatic catheterization demonstrating an obstruction of the thoracic duct, and a large collateral draining into the left lung. A percutaneous embolization of the thoracic duct was performed, after which there was no formation of new bronchial casts, and we were able to wean her from ECMO support. Nonetheless, other complications, including severe acute kidney injury occurred.

**Conclusions:** Though pathophysiology of fulminant casts formation is not yet understood, lymphatic drainage anomalies should be recognized before Fontan completion is performed.

**Keywords:** Fontan, Plastic bronchitis, lymphatic anomaly, thoracic duct embolization



**Image 1** Lymphangiography demonstrating thoracic duct obstruction and large collateral draining to the left lung, before (left panel) and during (right panel) embolization.

## P-230

**Successful surgical resection of a complicated left ventricular diverticulum in a neonate presented with unexplained anemia: A case report**

Mohammed Abutaqa<sup>1</sup>, Odai Shrath<sup>2</sup>, Mutaz Abusamra<sup>2</sup>, Majdi Abudaoud<sup>2</sup>, Moayad Srouf<sup>2</sup>, Samer Abdelraziq<sup>1</sup>, Nidal Haymouni<sup>1</sup>, Iyad Sbeitan<sup>1</sup>, Nizar Hijeh<sup>1</sup>

<sup>1</sup>Department of Pediatric cardiology, Al-Makassed Islamic Charitable Hospital, Jerusalem, Palestine; <sup>2</sup>Al-Quds University-School of Medicine, Abu-Dis, Jerusalem, Palestine.

**Background and Aim:** INTRODUCTION: Congenital left ventricular diverticulum (LVD) is a rare congenital cardiac anomaly and may be complicated by fatal adverse events such as diverticulum rupture. Most LVD cases are asymptomatic and often discovered incidentally. Herein, we describe an unusual and peculiar clinical presentation with felicitous surgical management of ruptured LVD. **Method:** Case presentations: A 10-day-old male infant presented with severe, intractable, and unexplained anemia associated with respiratory distress. Upon admission, the patient was clinically shocked with a hemoglobin level of 6.0 g/dL. As chest imaging showed cardiomegaly, echocardiography was performed and revealed a 9 x 10 mm diverticulum arising from the posterolateral wall of the left ventricle along with blood and clot collection in the pericardium. The patient underwent an urgent surgical resection of the diverticulum. He was followed up for 2 years without any readmissions or cardiac complaints.

**Results:** Clinical DISCUSSION: Systemic thromboembolism, heart failure, infarction, and tachyarrhythmias have all been reported as complications of LVD. The most serious complication is diverticulum rupture, which can result in death. As a result, this congenital defect should be discovered early in order to determine the potential risks and plan appropriate treatment.

**Conclusions:** CONCLUSION: Congenital heart defects such as LVD should be suspected in neonates presenting with unexplained and intractable anemia. To avoid the diagnosis confusion and risk of serious complications in LVD patients, such as spontaneous rupture of the diverticulum, we advocate immediate surgical management of LVD in children.

**Keywords:** Left ventricular diverticulum, congenital heart defects, unexplained anemia, diverticulum rupture, surgical resection, case report

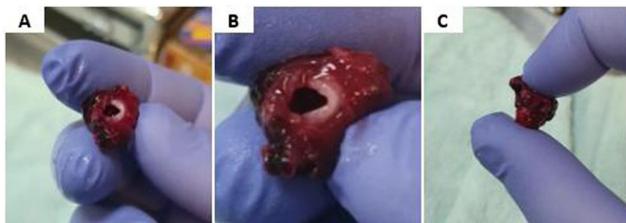


Figure 3 shows the resected left ventricular diverticulum

## P-231

**An unusual case of “Aortic Coarctation” treated with intrapericardial ascending to descending aortic graft.**

Sara Moscatelli<sup>1</sup>, Darryl Shore<sup>2</sup>, Ulrich Rosendahl<sup>2</sup>, Claudia Montanaro<sup>3</sup>  
<sup>1</sup>Paediatric Cardiology Department, Royal Brompton Hospital, London, UK; <sup>2</sup>Cardiothoracic Surgery, Royal Brompton Hospital, London, UK; <sup>3</sup>Adult Congenital Department, Royal Brompton Hospital, London, UK

**Background and Aim:** Coarctation of the aorta (CoA) is defined as a narrowing of the aorta at the level of the isthmus. Most commonly,

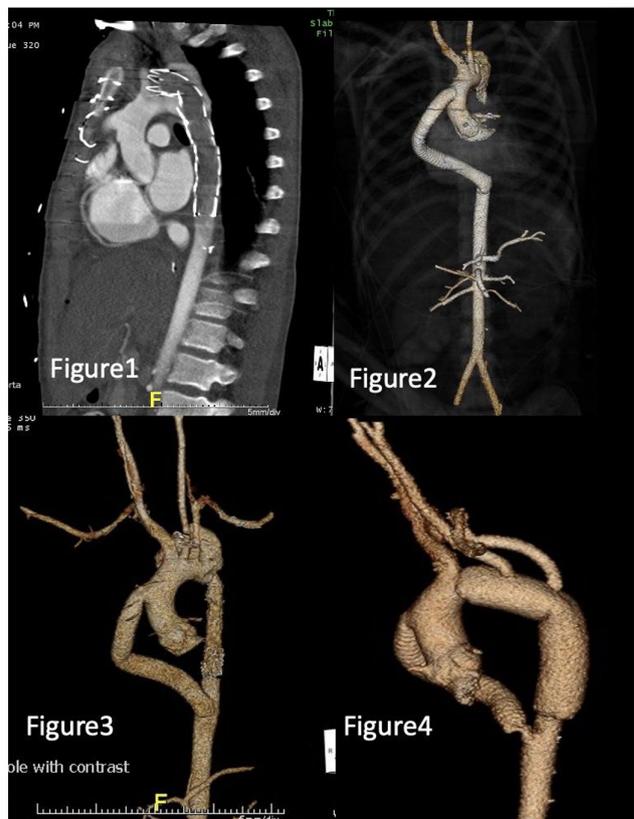
CoA is due to a congenital abnormality, but sometimes it can be due to an iatrogenic cause. We are presenting the rare case of a CoA in the context of a polytrauma.

**Method:** A 16-year-old boy was involved in a car accident on the 21.12.2020. He was immediately transferred to the nearest Trauma Centre for assessment and stabilisation. A total body CT showed multiple fractures, the most relevant at the pelvis, together with compound crural ones. In addition, CT showed a transection of the aorta at the isthmus level. For this reason, he was brought to the cath lab for stenting of the aorta. The postoperative course was complicated by sepsis, feeding intolerance, 2 cardiac arrests and renal failure with filter dependency. The echocardiography showed a gradient of 110mmHg proximal and distal to the stent, highly suspicious for severe CoA. Urgent CT showed stent migration outside and above the descending aorta, causing a complete obstruction (Figure1). The patient was transferred to the RBH on the 8.01.2021 for further specialised management. Due to the current fractures, it could not be heparinised, and consequently, cardiopulmonary bypass (CBP) could not be used during an eventual surgery. In addition, thoracotomy could not be performed due to bilateral crural external osteosynthesis.

**Results:** After a multidisciplinary discussion, despite the high risks, he was accepted for surgery performed on the 09.01.2022. An intrapericardial ascending to descending aorta grafting was implanted through a midline sternotomy off CBP using octopus' stabiliser (Figure2). The operation was successful and the patient restarted to produce urine after 36 hours. He was discharged on 05.2021 and a follow-up CT demonstrated rehabilitation of the stent (Figure3). Patient was elected to further operation for anatomical repair of the aorta few months later (Figure4). After one year, he is completely asymptomatic with well-controlled blood pressure and he has a normal life.

**Conclusions:** This case shows that in the scenario of an infrequent and life-threatening cause of CoA, surgically treated with intrapericardial ascending to descending aortic grafting without CBP.

**Keywords:** Coarctation, Congenital Heart Surgery



## P-232

**The technical performance score and its association with surgical outcomes across a range of severity of cardiac lesions in pediatric patients**

Francesca De La Cruz, Eftychia Kapravelou, Pearl Waraich, Astrid De Souza, James Potts, Derek Human, Kathryn Armstrong  
Division of Cardiology, Department of Pediatrics, British Columbia Children's Hospital, Vancouver, Canada

**Background and Aim:** The Technical Performance Score (TPS) assesses the adequacy of congenital heart surgery operative repairs and has been used to predict post-operative complications. This study aims to: (1) evaluate TPS in a cohort of pediatric surgical patients; and (2) determine if the TPS predicts surgical outcomes in these patients.

**Method:** This is a single centre, retrospective cohort study. All surgeries on patients with complete clinical and echocardiographic data were included (January 2019 to September 2019). The TPS was applied to intraoperative transesophageal echocardiograms (TEE) and postoperative transthoracic echocardiograms (TTE) and was correlated to morbidity, mortality and length of stay.  $\chi^2$  tests and logistic regression were used to determine the association between TPS (optimal, adequate and inadequate) and surgical outcome, with p values  $<0.05$  considered statistically significant.

**Results:** 106 patients were included (median age = 2.9 years; weight = 12.5 kg). Median ICU stay was 1 day and total hospital stay was 4 days. On TEE, 83 (78.3%) had an optimal TPS, 12 (11.3%) adequate and 11 (10.4%) inadequate scores. On TTE 82 (77.4%) had an optimal TPS, 13 (12.3%) adequate and 11 (10.4%) inadequate scores. There was one mortality with an optimal TPS. An inadequate TPS on TEE was associated with the need for a second bypass run ( $\chi^2 = 33.209$ ;  $P < 0.0001$ ) and postoperative ECMO ( $\chi^2 = 7.185$ ;  $P = 0.0275$ ), but not with delayed chest closure, incidence of arrhythmia or the need for early or late re-intervention. An inadequate TPS on pre-discharge TTE was associated with the need for a second run of bypass ( $\chi^2 = 7.557$ ;  $P = 0.0229$ ) and delayed chest closure ( $\chi^2 = 7.113$ ;  $P = 0.0285$ ), but not with the need for postoperative ECMO, incidence of arrhythmia or the need for early or late re-intervention. Almost 43% of surgeries were RACHS-1 category 2, 39.8% were category 3. TPS on both TEE and TTE demonstrated a decreasing incidence of optimal score to increasing risk-adjustment of the procedure (optimal scores for RACHS category 1 was 100%, 2: 81%, 3: 74.4% and 4: 0%).

**Conclusions:** TPS on TEE is a good predictor of early postoperative complications such as the need for a second run of bypass or postoperative ECMO.

**Keywords:** Technical Performance Score, TPS, RACHS-1

## P-233

**Shrill screaming as a sign of a malignant coronary artery anomaly? a case of an uneventful PAVSD-correction with an unexpected postoperative course**

Ida Hüners<sup>1</sup>, Martin Munz<sup>1</sup>, Henning Carstens<sup>1</sup>, Sebastian Harms<sup>2</sup>, Dennis Klinkenberg<sup>2</sup>, Urda Gottschalk<sup>2</sup>, Rainer Kozlik Feldmann<sup>2</sup>, Jörg Siegmair Sachweh<sup>1</sup>, Michael Hübler<sup>1</sup>, Daniel Biernann<sup>1</sup>

<sup>1</sup>Children's Heart Clinic, Congenital and Pediatric Heart Surgery, University Heart & Vascular Center Hamburg, University Medical Center Hamburg-Eppendorf, Hamburg, Germany.; <sup>2</sup>Children's Heart Clinic, Pediatric Cardiology, University Heart & Vascular Center Hamburg, University Medical Center Hamburg-Eppendorf, Hamburg, Germany.

**Background and Aim:** Coronary artery anomalies (CAA) in children are extremely rare. They can occur alone or in conjunction with other congenital heart defects.

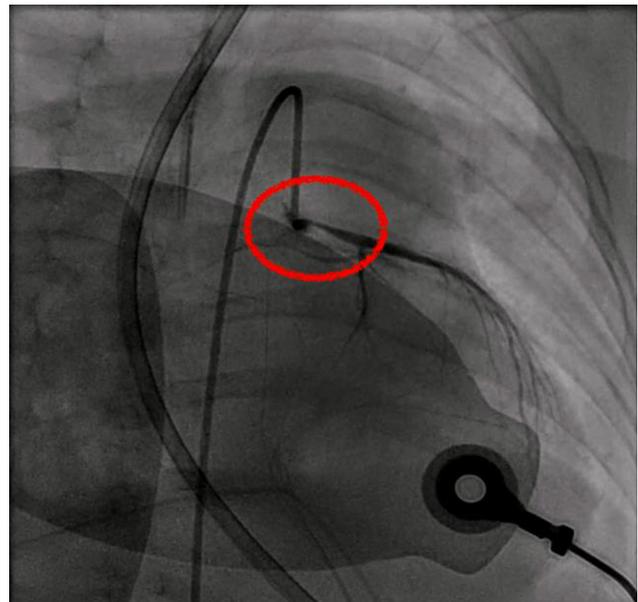
**Method:** We present the case of a 7-month-old boy (70 cm, 8 kg) who underwent a correction of a partial atrioventricular septal defect by ASD-closure with a pericardial patch and cleft closure.

**Results:** Echocardiography demonstrated good biventricular function (EF: 56%). Despite the positive outcomes, the patient required multiple doses of dipidolone, which did not provide pain relief. Analgesia was escalated to a continuous morphine perfusor. The parents indicated that the child had a history of shrill screaming and crying attacks for no apparent reason already before the operation. On the second postoperative day, the patient's pain episodes and restlessness worsened. Morphine was doubled, and ketamine perfusor was added. Episodes of decompensation followed with hypotension (MAP 35–45 mmHg), bradycardia (90–100 bpm), and lactate elevation (up to 5.2 mmol/l). ST-segment elevations in derivations II and III and depressions in V3, as well as a rise in troponin could be detected (from 3268 pg/ml to 12808 pg/ml). A coronary angiography was conducted, revealing left coronary artery (LCA) with suspected coronary spasm at the outflow. Symptoms persisted after administration of continuous infusion of nitroglycerin. Subsequently, a cardiac CT was conducted, showing a short-segment exit stenosis right after the relatively high exit of the LCA ostium. Reoperation on the same day revealed an intramural course of the LCA. The LCA was mobilized and unroofed, followed by a pericardial patch augmentation of the neo-ostium.

The postoperative course after the second surgery was uneventful. After 14 days the patient was discharged with no residual symptoms of coronary malperfusion and is in good health. The screaming and crying attacks have adjourned.

**Conclusions:** This case depicts that there is no straightforward correction of a congenital heart defect. An intramural course of coronary arteries is rare and the detection of CAA is crucial for administering the appropriate treatment to prevent subsequent myocardial ischemia. The clinical presentation of children with CAA may be heterogenous, ranging from absence of symptoms to severe, potentially life-threatening symptoms.

**Keywords:** congenital heart disease, congenital heart surgery, malignant coronary artery anomaly, angina pectoris



Coronary angiography revealing left coronary artery (LCA) with suspected coronary spasm at the outflow

## P-234

**Streamlined interdisciplinary management of a pediatric patient with severe heart failure and sickle cell hemoglobinopathy**

Ida Hüners<sup>1</sup>, Martin Munz<sup>1</sup>, Henning Carstens<sup>1</sup>, Frithjof Lemme<sup>1</sup>, Maria Grafmann<sup>2</sup>, Corinna Güsmer<sup>3</sup>, Thomas Mir<sup>2</sup>, Jörg Siegmar Sachweh<sup>1</sup>, Matthias Bleeke<sup>4</sup>, Daniel Biermann<sup>1</sup>, Michael Hübler<sup>1</sup>

<sup>1</sup>Children's Heart Clinic, Congenital and Pediatric Heart Surgery, University Heart & Vascular Center Hamburg, University Medical Center Hamburg-Eppendorf, Hamburg, Germany.; <sup>2</sup>Children's Heart Clinic, Pediatric Cardiology, University Heart & Vascular Center Hamburg, University Medical Center Hamburg-Eppendorf, Hamburg, Germany.; <sup>3</sup>Institute of Transfusion Medicine, University Medical Center Hamburg-Eppendorf, Hamburg, Germany.; <sup>4</sup>Department for Pediatric Hematology and Oncology, University Medical Center Hamburg-Eppendorf, Hamburg, Germany.

**Background and Aim:** Sickle cell disease (SCD) is a severe anemic disease characterized by abnormal hemoglobin (Hb) S in erythrocytes. Multiple medical complications may arise, necessitating an interdisciplinary team approach for optimal treatment. In Europe, people with SCD and those who additionally require open-heart surgery (OHS) are extremely rare, but their prevalence is increasing due to the global migration of refugees for various reasons.

**Method:** We present the case of a three-year-old west-African girl (87 cm, 11 kg) with restrictive cardiomyopathy and severe atrioventricular valve (AV) regurgitation, as well as homozygous SCD.

**Results:** The child's initial symptoms included tachypnea, palpitations, and abnormal fatigue. Our team decided for OHS with CPB to treat the regurgitation of tricuspid and mitral valve. Preoperatively, Hb was 8.5 g/dl, HbS was 68.7% and HbA was 16.8%. Exchange transfusion (ET) was performed to achieve a HbS fraction of <30% by an apheresis system connected to CPB supervised by both our perfusionist and our doctor of transfusion medicine. The patient's initial venous return was sequestered after priming the circuit with packed red blood cells (RBC) with plasma to hematocrit level of 50%. Through this setup, an automated ET with three RBCs was performed. Echocardiography revealed good AV-function following reconstruction. Postoperatively, Hb was 11.6 g/dl and HbS fraction decreased to 12%. Despite persistent tachypnea, the patient's general condition significantly improved since admission; she was discharged and is now seeing her cardiologist on a regular basis.

**Conclusions:** Cardiopulmonary bypass (CPB) is associated with numerous sickle cell crisis risk factors, including hypothermia, hypoxia, hypoperfusion, and acidosis resulting in hemolysis, microvascular obstruction, decreased erythrocyte survival, and decreased oxygen-carrying capacity. SCD patients needing OHS with CPB need specialized care and monitoring to prevent vaso-occlusive events. It is indispensable to lower HbS fraction through ET in SCD patients undergoing OHS with CPB to prevent postoperative mortality. Current recommendations propose reducing on-pump HbS fraction to less than 30%; some studies even advocate less than 10%.

Our case depicts an individualized concept combining OHS with CPB and simultaneous ET by connecting an apheresis system to CPB executed by an interdisciplinary team approach leading to a favorable patient outcome.

**Keywords:** congenital heart surgery, sickle cell disease, cardiopulmonary bypass, apheresis

## P-235

**Successful LVAD bridge-to-recovery 472 days after the correction of ALCAPA syndrome**

Martin Munz<sup>1</sup>, Ida Hüners<sup>1</sup>, Henning Carstens<sup>1</sup>, Jörg Siegmar Sachweh<sup>1</sup>, Sebastian Hams<sup>2</sup>, Dennis Klinckenberg<sup>2</sup>, Urda Gottschalk<sup>2</sup>, Thomas Mir<sup>2</sup>, Rainer Kozlik Feldmann<sup>2</sup>, Daniel Biermann<sup>1</sup>, Michael Hübler<sup>1</sup>

<sup>1</sup>Children's Heart Clinic, Congenital and Pediatric Heart Surgery, University Heart & Vascular Center Hamburg, University Medical Center Hamburg-Eppendorf, Hamburg, Germany.; <sup>2</sup>Children's Heart Clinic, Pediatric Cardiology, University Heart & Vascular Center Hamburg, University Medical Center Hamburg-Eppendorf, Hamburg, Germany.

**Background and Aim:** Late diagnosis of malignant coronary anomalies, such as an anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), can lead to severe heart failure. Ventricular dilatation and dysfunction, as well as mitral valve insufficiency, are typical clinical hallmarks and should lead clinicians to suspect coronary anomalies. ALCAPA is treated surgically immediately after diagnosis to preserve as much myocardium as possible. Mechanical circulatory support can be beneficial in both the short- and long-term for the sub-cohort of patients who do not recover myocardial function after corrective surgery.

**Method:** We want to highlight the case of a 15-month-old boy who required a left ventricular assist device (LVAD) intended as a bridge-to-transplantation approach due to a lack of recovery after primary repair.

**Results:** The patient's ejection fraction (EF) preoperatively was 15%. After the initial surgery on day 403 of life, the EF was 18% and the fractional shortening (FS) 8%. In the days following primary surgery, cardiac function recovered only marginally, and the boy showed signs of developing end-organ damage. Accordingly, we decided to implant a pneumatically powered extracorporeal pump with a 15-mL ventricle size as a LVAD. On day 53 of treatment, cannulas were placed in the left apex and ascending aorta. The child gradually throve (9 kg on admission; 14 kg at discharge) within 472 days while under LVAD therapy and overcame significant side effects such as bleeding and infection at the cannula exit sites. Fortunately, we could switch to a bridge-to-recovery strategy as the heart's function markedly recovered (EF, FS, LVEDD). After a fast recovery from the uncomplicated explantation of the LVAD, systolic and diastolic biventricular cardiac function was excellent, with a FS of approximately 28–30% and an EF between 55–60% after 20 days of follow-up. The patient was discharged 26 days after explantation of the system and will be reevaluated regularly by the referring cardiologist.

**Conclusions:** The cardiac performance of patients under LVAD therapy within a bridge-to-transplant approach must be reevaluated, preferably based on a standard protocol, to not overlook the potential for successful weaning.

**Keywords:** congenital heart surgery, lvad, alcapa, htx

## P-236

**3D imaging improves surgical planning for unifocalization procedures in children with pulmonary atresia, VSD and MAPCAS**

Martin Munz<sup>1</sup>, Ida Hüners<sup>1</sup>, Philipp Schneider<sup>2</sup>, Thomas Mir<sup>2</sup>, Henning Carstens<sup>1</sup>, Jörg Siegmar Sachweh<sup>1</sup>, Daniel Biermann<sup>1</sup>, Michael Hübler<sup>1</sup>

<sup>1</sup>Children's Heart Clinic, Congenital and Pediatric Heart Surgery, University Heart & Vascular Center Hamburg, University Medical Center Hamburg-Eppendorf, Hamburg, Germany.; <sup>2</sup>Children's Heart Clinic, Pediatric Cardiology, University Heart & Vascular Center Hamburg, University Medical Center Hamburg-Eppendorf, Hamburg, Germany

**Background and Aim:** Planning unifocalization procedures in patients with pulmonary atresia with a ventricular septal defect

and major aortopulmonary collateral arteries (PA/VSD-MAPCAs) is challenging. Due to their excellent contrast and spatial resolution, computed tomography (CT) and magnetic resonance imaging (MRI) are the most suitable imaging modalities. Whereas preoperative planning based on 2D images only is rather old-fashioned and sometimes inconclusive, 3D imaging allows the congenital heart team to receive an in-depth understanding of the morphology of each individual patient with PA/VSD-MAPCAs. *Method:* We identified three patients from our cohort with CT scans prior to surgery. 3D models were created utilizing DICOM (Digital Imaging and COmmunications in Medicine) datasets derived from thin-slice CT examinations. Via medical image processing software, the 3D models were created.

*Results:* In total, three patients with PA/VSD-MAPCAs were operated after bicaval cannulation, cardioplegic arrest (Bretschneider 30 ml/kg bodyweight), and induction of moderate hypothermia (28 °C).

Patient 1 (male, age 16 months, 13.6 kg, 91 cm) formerly palliated with one central ap-shunt mainly feeding the right lung received a step towards unifocalization via the union of further MAPCAs to the left lung and was discharged at postoperative day 10.

Patient 2 (female, age 17 months, 9.3 kg, 77 cm) was unifocalized and discharged home after 14 days.

Patient 3 (female, age 2 months, 3.9 kg, 55 cm) died after unifocalization on the 21st postoperative day after a complication during cardiac catheterization. Prior to that, an early reoperation and a cardiac catheterization were conducted to improve the blood flow to the left lung.

*Conclusions:* The surgical team found the relevant vessels in these three patients effortlessly. Primary correction or the switch from palliative to corrective surgery in congenital heart disease can be simplified through the use of 3D models, both in the form of 3D printed models and visualization through virtual reality glasses.

**Keywords:** congenital heart surgery, mapca, pulmonary atresia with ventricular septal defect, major aortopulmonary collateral arteries

### P-237

#### Left coronary birth defects from the aorta: diagnostic and therapeutic challenge of a sudden death risk in a limited-resource country. A case report

Tony Nengom Jocelyn<sup>1</sup>, Sap Ngo Um Suzanne<sup>2</sup>, Epee Ngoue Jeannette<sup>1</sup>, Chelo David<sup>2</sup>

<sup>1</sup>Department of Pediatrics, Faculty of Medicine and Biomedical Sciences, University of Yaounde I, Cameroon; <sup>2</sup>Mother and Child Centre of the Chantal Biya Foundation, Yaounde Cameroon; Department of Pediatrics, Faculty of Medicine and Biomedical Sciences, University of Yaounde I, Cameroon

*Background and Aim:* Coronary artery birth and pathway anomalies, particularly those where the artery originates from the contralateral coronary sinus with a pathway between the aorta and the pulmonary artery, represent a high risk of sudden death. Early diagnosis and management are essential for a good prognosis.

*Method:* Clinical CASE:

*Results:* 9 year old boy brought to a paediatric cardiology consultation for loss of consciousness during physical effort with cardiac arrest, having required in situ cardiopulmonary resuscitation two days ago. It was the second such episode within a month. No particular personal or family history.

Physical examination revealed good general condition with 100% room air saturation, stable haemodynamic parameters and normal anthropometric parameters for age. Heart sounds are regular, no

murmur, symmetrical femoral pulses regular and of good volume. The rest is unremarkable.

In view of this normal physical examination and the notion of malaise during the effort, the suspicion of anomaly of the coronary artery pathway is evoked and a cardiac echography is carried out: left coronary artery coming from the right aortic sinus with a trunk pathway between the aorta and the pulmonary artery. Normal flow in both coronary arteries. Good overall cardiac function with an ejection fraction of 64%. A cardiac CT scan confirmed the origin of the left coronary from the right coronary sinus at 2 o'clock, which travels along the anterior aspect of the aorta in the inter-aorto-pulmonary, then divides into the anterior inter-ventricular and circumflex arteries at the level of the left atrioventricular fold. As the technical platform did not allow for surgical management in Cameroon, a medical evacuation was organised for unroofing of the left coronary.

*Conclusions:* We presented a case revealed by a sudden cardiac arrest, in a country where diagnosis and management remain a real challenge due to limited technical facilities.

**Keywords:** coronary anomaly, child, sudden death, cameroon

### P-238

#### Coronary artery anomalies in transposition of great arteries: their impact on postoperative outcomes after arterial switch operation.

Blanca Torres Maestro, Bunty Ramchandani Ramchandani, Luz Polo López, Álvaro González Rocafort, Juvenal Rey Lois, Raúl Sánchez Pérez, María Jesús Lamas Hernández, Tomasa Centella Hernández, Ángel Aroca Peinado  
*Congenital Cardiovascular Surgery. La Paz-Ramón y Cajal University Hospital. Madrid. Spain*

*Background and Aim:* Coronary anomalies are relatively frequent in patients presenting transposition of great arteries. Such anomalies definitely increase the difficulty of the arterial switch operation. We sought to analyse clinical results and the influence of coronary artery anatomy on mortality after the arterial switch operation.

*Method:* Between January 2007 and September 2022, 130 patients underwent an arterial switch operation for transposition of great arteries. We analysed preoperative and intraoperative characteristics. Patient coronary patterns were recorded from operative reports (using the Leiden Convention) and analysed to determine their association with adverse postoperative outcomes. The statistical analysis was made with STATA IC 15.

*Results:* The mean age at operation was 12±9 days (range 1-84). A total of 91 patients (70%) had an associated ventricular septal defect. Nine patients (6.9%) had an aortic arch obstruction. Coronary anatomy anomalies were present in 46 patients (35.4%). The most common type of anomalous coronary anatomy was the circumflex coronary artery arising from sinus 2 (23 patients [17%]), followed by a single sinus pattern (11 patients [8.5%]). Four patients (3.1%) had an intramural coronary artery. Extracorporeal membrane oxygenation (ECMO) after surgery was needed by 12 patients (9.2%), eight of them had coronary artery anomalies, which was statistically significant ( $p = 0.03$ ). Early mortality was 4.8% (4 out of 84) for patients with normal coronary pattern and 13% (6 out of 46) in patients with any coronary artery anomaly, although it was not statistically significant ( $p = 0.17$ ). Likewise, there was not a statistically significant difference in mortality between normal and anomalous coronary pattern after separating the sample in two eras (2007-2014,  $p = 0.13$ ; 2015-2022,  $p = 1.00$ ).

*Conclusions:* Patients with transposition of great arteries that present coronary pattern anomalies had higher rates of early mortality after the arterial switch operation, although not statistically significant

neither globally nor within the two study groups independently (2007–2014, 2015–2022). On the other hand, unusual coronary pattern was found to be significantly associated with the need of extracorporeal membrane oxygenation on the postoperative period.

**Keywords:** arterial switch operation, anomalous coronary pattern, transposition of great arteries

#### P-239

##### **Destruction and remodeling – analysis of equine pericardium used for AVR**

Armin Darius Peivandi<sup>1</sup>, Sven Martens<sup>1</sup>, Klaus Michael Müller<sup>2</sup>, Sabrina Martens<sup>1</sup>

<sup>1</sup>University Hospital Muenster, Department of Cardiothoracic Surgery, Muenster, Germany; <sup>2</sup>University Hospital Muenster, Gerhard-Domagk Institute of Pathology, Muenster, Germany

**Background and Aim:** Autologous pericardium is currently gold standard for reconstructive surgery of the aortic valve in children with congenital heart disease. However, the use of industrially processed equine pericardium has recently become more popular amongst cardiac surgeons. In this regard, we analyzed explanted equine pericardium histologically to provide insights in the formal pathomechanism of patch degeneration.

**Method:** Equine pericardial tissue was gathered, handled according to usual histologic procedures and stained with Elastica van Gieson staining (EvG) to visualise collagen as well as elastic fibres. Alizarin red S allowed detection of calcification. Light microscopy pictures were taken in different magnifications to analyse tissue structures.

**Results:** Due to aortic valve dysfunction, re-operation was necessary in 5 patients after a median duration of implantation of 5.2 years (IQR 10.1–3). Degenerative developments included tissue edema, destructive fiber structure, dehiscence, nodose-like fiber arrangements and loco regional myxoid swelling. Two explants were severely calcified. Florid inflammation could not be detected. **Conclusions:** Explanted equine patches show complex degenerative developments. These include different destruction and remodeling patterns as well as chronic foreign body reactions and calcification. To assess long-term durability, large-scale registry studies are advisable.

**Keywords:** aortic valve reconstruction, equine pericardium, histopathology, remodeling

#### P-240

##### **Double-arterial cannulation perfusion provides better brain protection during surgical correction of aortic coarctation with ventricular septal defect**

Jiachen Bi, Shixin Su, Jianbin Li, Minghui Zou, Xinxin Chen, Jia Li, Li Ma

Department of Cardiovascular Surgery, Guangzhou Women and Children's Medical Center, Guangzhou Medical University, Guangdong, China & Guangdong Provincial Key Laboratory of Research in Structural Birth Defect Disease, Department of Pediatric Surgery, Guangzhou Women and Children's Medical Center, Guangzhou Medical University, Guangdong, China

**Background and Aim:** DHCA is the gold standard perfusion strategy for CoA-VSD correction, but associated with risk of brain injury. Double arterial cannulation (DAC), firstly introduced in aortic arch reconstruction due to aortic dissection or aneurysm, is now an alternative perfusion strategy. The purpose of this study was to evaluate the effectiveness of brain protection of DAC compared with DHCA. **Method:** A single-center pilot study of RCT was carried out from 2020 to 2021. Patients with CoA-VSD ready for surgical

correction were enrolled in and randomly assigned into 2 groups, namely DHCA without cerebral perfusion (n = 5) and DAC (n = 6). Brain MRI and TCD were applied before and after operation; EEG and cerebral oxygen saturation were continuously monitored before, during, and after surgery until 48 hours post-operation.

**Results:** The operation time, CPB time and cross-clamp time were all numerically shorter in DAC group. The duration of intraoperative isoelectric state was shorter in DAC group (22±31min vs. 64±30min, p<0.05). Post-operative cerebral oxygen saturation was higher in DAC group (66±10% vs. 49±2%, p = 0.04 18h post-operation; 65±5% vs. 56±7%, p = 0.04, 27h post-operation, and 72±5% vs. 65±2%, p = 0.04, 36h post-operation; separately). The cerebral vascular resistance was lower in DAC group, with both pulsatility index and resistivity index were lower 15h post-operation (1.15±0.17 vs. 1.51±0.09, p<0.01; 0.65±0.04 vs. 0.72±0.02, p<0.01; separately). Post-operative intracranial hemorrhage was numerically lower in DAC group. Though without statistically difference due to limited number, the non-hemorrhage cases were only seen in DAC group. 1 seizure case was seen in each group, with comparable seizure time. Mixed linear regression indicated better systematic hemodynamic characteristics in DAC group: lower intraoperative (P = 0.03) and post-operative (p = 0.01) systematic vascular resistance index, and higher post-operative cardiac index (p = 0.01) were found; lower post-operative pulsatility index and resistivity index still existed (p<0.01); and lower pulse pressure variation were seen (p<0.01).

**Conclusions:** DAC is a better perfusion strategy during surgical CoA-VSD correction, which could improve cerebral perfusion and reduce brain injury.

**Keywords:** Double-arterial cannulation, brain protection, aortic coarctation, ventricular septal defect

#### P-242

##### **Technique of deferring decannulation in pediatric ECMO – 10 years of experience**

Inigo Velasco<sup>1</sup>, Paula Burgos<sup>1</sup>, Jorge Utanda<sup>1</sup>, Luz Polo<sup>2</sup>, Álvaro González<sup>2</sup>, Juvenal Rey<sup>2</sup>, Raúl Sánchez<sup>2</sup>, Buntty Ramchandani<sup>2</sup>, Blanca Torres<sup>2</sup>, José Encinas<sup>3</sup>, Ane Miren Andrés<sup>3</sup>, Carlos De La Torre<sup>3</sup>, Carlos Labrandero<sup>4</sup>, Ángela Uceda<sup>4</sup>, Luis Castro<sup>5</sup>, Raquel Pérez<sup>5</sup>, Laura Sánchez<sup>6</sup>, Paloma López<sup>6</sup>, Juan José Menéndez<sup>7</sup>, Irene Amores<sup>7</sup>, Eva Barahona<sup>8</sup>, Carolina Muñoz<sup>8</sup>, Ángel Aroca<sup>2</sup>

<sup>1</sup>Pediatric Perfusion. Cardiovascular Surgery Department. Hospital Universitario La Paz, Madrid; <sup>2</sup>Pediatric Cardiovascular Surgery. Hospital Universitario La Paz, Madrid; <sup>3</sup>Pediatric Surgery. Hospital Universitario La Paz, Madrid; <sup>4</sup>Pediatric Cardiology, Hospital Universitario La Paz, Madrid; <sup>5</sup>Pediatric Anesthesia, Hospital Universitario La Paz, Madrid; <sup>6</sup>Neonatal Intensive Care Unit, Hospital Universitario La Paz, Madrid; <sup>7</sup>Pediatric Intensive Care Unit, Hospital Universitario La Paz, Madrid; <sup>8</sup>Pediatric Surgery Nurse Department, Hospital Universitario La Paz

**Background and Aim:** The removal of the ECMO cannulas and the necessary systemic anticoagulation once the therapy has been withdrawn, constitutes a difficult and risky decision.

In our center, for more than 10 years now, we generally defer the removal of the cannulae for a minimum of 24 hours after conclusion of the ECMO therapy.

This fact does not cause great harm to the patient, and presents some benefits in case of needing a rapid restitution of therapy in patients with risk of cardiogenic shock or respiratory failure after discontinuing the ECMO run.

**Method:** Between November 2012 and november 2022, 145 ECMO therapies were applied in 128 pediatric patients for

different reasons (82 cases of inability of weaning from cardiopulmonary bypass, 20 cases of low cardiac output syndrome, 36 cases of acute respiratory failure, 7 for other causes (4 for septic shock, 2 for IO procedures, 1 to isolate the pulmonary circulation due to hemorrhagic reasons)

**Results:** From the 128 patients, 43 did not survive to ECMO therapy, and 22 of the remaining, did not survive to hospital discharge. Main duration of ECMO run was 8, 11 days, and the main BSA 0, 41m<sup>2</sup>. From the 85 patients who survived to therapy and decannulation, 102 ECMO runs were established; bridge to recovery, to cardiac surgery, to bridge, to transplant. From the ECMO runs bridge to recovery and cardiac surgery, (n = 84), 62 were decannulated with this technique (39 central cannulation and 20 cervical cannulation). The duration from ECMO support conclusion to decannulation was 20, 2 hours, and in 10 patients, reinstitution of therapy was conducted.

**Conclusions:** Our technique is safe and advantageous because the heparin dose to maintain the cannulae permeability is low and is rapidly accessible to reinstitute ECMO therapy in case of low cardiac output syndrome or respiratory failure.

**Keywords:** Pediatric, ECMO- Extracorporeal Membrane Oxygenation, Postcardiotomy, Respiratory Failure, Low cardiac output syndrome, Decannulation

#### P-243

##### Outcome after arterial switch operation in patients with transposition of the great arteries with left ventricular outflow tract obstruction

Abe Den Ouden<sup>1</sup>, Ingmar Knobbe<sup>2</sup>, Monique Rm Jongbloed<sup>3</sup>, Berto J Bouma<sup>4</sup>, Mark Mg Hazekamp<sup>5</sup>, Roel Lf Van Der Palen<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology, Leiden University Medical Center, Leiden, Netherlands; <sup>2</sup>Department of Pediatric Cardiology, Amsterdam UMC, University of Amsterdam, Amsterdam, Netherlands;

<sup>3</sup>Department of Cardiology and Department of Anatomy & Embryology, Leiden University Medical Center, Leiden, Netherlands; <sup>4</sup>Department of Cardiology, Amsterdam UMC, University of Amsterdam, Amsterdam, Netherlands; <sup>5</sup>Department of Cardiothoracic Surgery, Leiden University Medical Center, Leiden, Netherlands

**Background and Aim:** Selected patients with transposition of the great arteries (TGA) and left ventricular outflow tract obstruction (LVOTO), usually associated with ventricular septal defect (VSD), may still undergo arterial switch operation (ASO). We aimed to review the clinical outcomes of TGA-VSD-LVOTO patients after ASO and compare the spectrum of LVOTO with a historical series of cardiac specimens with TGA-VSD-LVOTO.

**Method:** Of 516 patients who underwent ASO at our institution between 1977–2022, 11 had TGA-VSD-LVOTO. Patient records and operative reports were reviewed. Moreover, a series of cardiac specimens with TGA-VSD-LVOTO from the University collection of congenital anomalies who did not undergo cardiac surgery was examined and used as illustration.

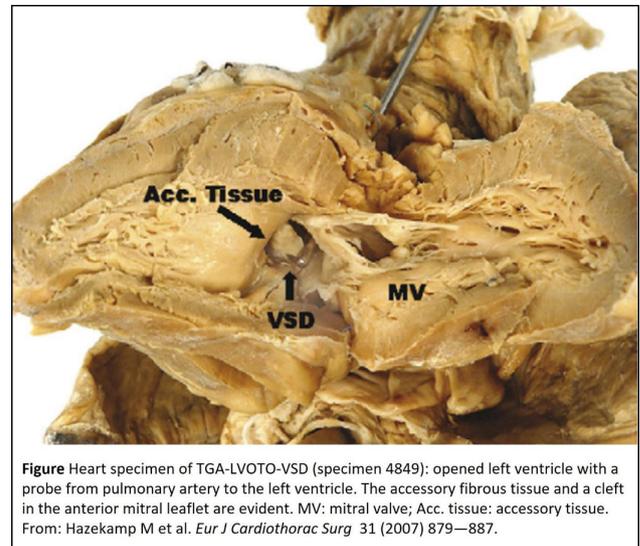
**Results:** Eight patients had TGA-VSD, 3 DORV type Taussig-Bing anomaly (TBA). LVOTO mechanisms were multifactorial, ranging from posteriorly deviated infundibular septum to fibrous tissue tags/masses and fibromuscular membrane. Median age at ASO was 0.4 (IQR 0.07–1.8) years. One-stage ASO was performed in 3, two-stage ASO in 8 (modified Blalock-Thomas-Taussig shunt(6), pulmonary banding(1), coarctectomy(1)). Primary LVOTO resection was performed in 10/11 patients during ASO; in one TBA patient LVOT was considered as being potentially narrow but acceptable. No in-hospital mortality occurred. Two patients died: 3.1 months (sudden death) and 6.0 years post-ASO (unknown cause).

Median follow-up of survivors: 17.1 (IQR 10.3–25.7) years, all in NYHA class I. No reoperation for LVOTO was necessary after primary LVOTO relief during ASO. One patient in whom the LVOTO was not primarily addressed required surgery for progressive LVOTO 3 months post-ASO. At latest follow-up, one patient had moderate residual LVOTO (peak-gradient 45mmHg). No patients developed significant neo-aortic valve regurgitation.

From the cardiac specimen series, 10 out of 23 TGA-VSD-LVOTO specimen were retrospectively judged eligible for ASO (8 TGA-VSD, 2 TBA). A similar spectrum of LVOTO causes as in the clinical patients was identified: fibrous ridge (2), malalignment outlet septum (3), obstructive muscular tissue (2), aneurysmatic tissue of membranous septum (1) and accessory fibrous tissue attached to the mitral valve (2)(Figure).

**Conclusions:** Arterial switch operation is in selected cases possible in patients with TGA-VSD and LVOTO of different etiology. Survivors showed good functional ability, without reoperations for LVOTO and preserved neo-aortic valve function.

**Keywords:** Transposition of the great arteries, Left ventricular outflow tract obstruction, Arterial switch operation, Outcome



**Figure** Heart specimen of TGA-LVOTO-VSD (specimen 4849): opened left ventricle with a probe from pulmonary artery to the left ventricle. The accessory fibrous tissue and a cleft in the anterior mitral leaflet are evident. MV: mitral valve; Acc. tissue: accessory tissue. From: Hazekamp M et al. *Eur J Cardiothorac Surg* 31 (2007) 879–887.

#### P-246

##### The experience of subaortic obstruction repair concomitantly with total cavopulmonary connection

Yuliya Pozniak<sup>1</sup>, Elshad Bairamov<sup>1</sup>, Oleksandr Romaniuk<sup>2</sup>, Ihor Ihor Lebid<sup>1</sup>, Nadiia Rudenko<sup>2</sup>, Illya Yemets<sup>2</sup>

<sup>1</sup>Ukrainian Children's Cardiac Center, Kyiv, Ukraine; <sup>2</sup>Ukrainian Children's Cardiac Center, Kyiv, Ukraine, Shupyk National Medical Academy of Postgraduate Education, Kyiv, Ukraine

**Background and Aim:** Systemic ventricle outflow obstruction (SVOTO) could result in unfavorable hemodynamic conditions that might worsen the prognosis of patients with total cavopulmonary connection (TCPC)

**Method:** From January 2005 to December 2016 137 consecutive children with variable single ventricle pathologies underwent TCPC procedure, 10, 9% (n = 15) of which was presented with SVOTO. We used two main options for surgical relief of SVOTO: the first consisted in ventricular septal defect enlargement, the second one – in modified Damus-Kaye-Stansel procedure (DKS). There was 2 (1, 4%) early death in the control group and no mortality in study groups through the study period. Two patients in study groups underwent a permanent pacemaker

implantation. SVOT gradient decreased from  $21 \pm 5$ , 82 mmHg preoperatively to  $8 \pm 4$ , 33 mmHg after the procedure

**Results:** Our results indicate that the SVOTO relief at the moment of TCPC does not cause significant increase in morbidity and mortality in the early postoperative period

**Conclusions:** The results suggest that the DKS is the method of choice with regard to avoidance of heart block or adequate removal of SVOTO

**Keywords:** total cavopulmonary connection, systemic ventricle outflow obstruction, subaortic obstruction, single ventricle, the Damus-Kaye-Stansel procedure

#### P-247

##### The outcomes of the extracardiac total cavopulmonary connection in 137 consecutive patients with single-ventricle physiology

Yuliya Pozniak<sup>1</sup>, Elshad Bairamov<sup>1</sup>, Oleksandr Romaniuk<sup>2</sup>, Ihor Lebid<sup>1</sup>, Nadiia Rudenko<sup>2</sup>, Illya Yemets<sup>2</sup>

<sup>1</sup>Ukrainian Children's Cardiac Center, Kyiv, Ukraine; <sup>2</sup>Ukrainian Children's Cardiac Center, Kyiv, Ukraine, Shupyk National Medical Academy of Postgraduate Education, Kyiv, Ukraine

**Background and Aim:** Fontan procedure has gone through several modifications and today is the primary palliation to treat a broad spectrum of complex congenital heart defects. The aim of this study is to present the results of treatment of children and adults with a single ventricle.

**Method:** Since 2005 to 2017 years, in 137 consecutive patients with a single ventricle morphology aged 9,  $97 \pm 4$ , 37 years the extracardiac total cavopulmonary connection (TCPC) was performed. The most common heart pathology was tricuspid atresia (27.1%). In 125 (91, 2%) patients, the operation was carried out with the moderately hypothermic cardiopulmonary bypass (CPB), without aortic cross-clamp. The average CPB was 94,  $3 \pm 56$ , 7 min. In the remaining twelve patients extracardiac TCPC was carried out without CPB.

**Results:** There were three (2, 1%) early deaths. The median duration of the mechanical ventilation was 9,  $6 \pm 16$ , 2 h (1–71 h). The average length of stay was 17,  $4 \pm 18$ , 3 days (8, 0–81, 0 days).

**Conclusions:** Improved understanding of single-ventricle physiology, patient preparation, surgical techniques, and postoperative care has allowed us to reach a steady state in the early outcomes after TCPC.

**Keywords:** single ventricle, Fontan procedure, total cavopulmonary connection, extracardiac conduit, cardiopulmonary bypass

#### P-249

##### A rare case of type iv aorta-left ventricular tunnel with intra-aneurysmal thrombus

Vivek Bagaria, Channabasavaraj Hiremath

Department of Cardiothoracic and Vascular Surgery, Sri Sathya Sai Institute of Higher Medical Sciences, Bangalore

**Background and Aim:** Aorto-left ventricular tunnel (ALVT) in adult patients is an exceedingly rare congenital anomaly. We report a case of Type IV ALVT arising from the left coronary sinus, opening into the lateral wall of the left ventricle with a thrombus present in the aneurysmal tunnel.

**Method:** An 18-year-old female patient with easy fatigability, on echocardiography, was diagnosed to have aortic insufficiency with an additional paravalvular regurgitant jet. Contrast computed tomography demonstrated an aorto-left ventricular tunnel with aneurysmal dilatation of both the extracardiac and intracardiac

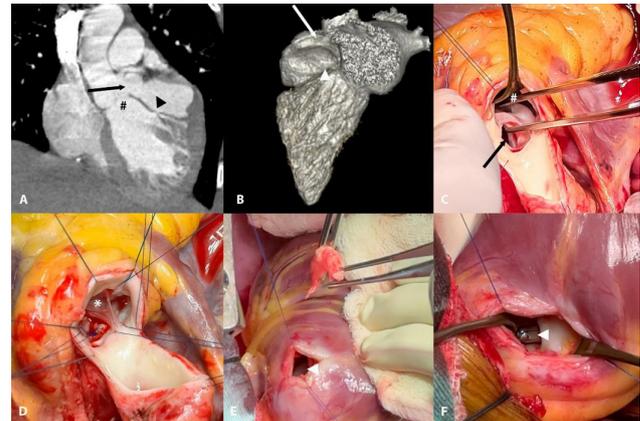
segments (Type IV ALVT). Magnetic resonance imaging confirmed the presence of a thrombus within the tunnel. The intra-operative assessment demonstrated the aortic end of the tunnel arising at the level of the sinotubular junction of the left coronary sinus below the displaced left coronary ostia, in addition, the lateral wall of the left ventricle overlying the distal aneurysmal end of the tunnel was thinned out. Also, a perforation was identified in the non-supported left aortic cusp. The patient underwent bicameral closure of the ALVT, removal of the thrombus within the aneurysmal tunnel and autologous pericardial neocusp creation at the left aortic leaflet position.

**Results:** Aorto-left ventricular tunnel, a paravalvular communication between the aorta and left ventricle, has an incidence of less than 0.1% in infants born with congenital heart disease. Associated anomalies of the coronary artery and aortic valve anatomy have been described. Aortic valve regurgitation requiring valve repair or replacement remains a major long-term concern, with a reported incidence as high as 50% on 5-year follow-up. Our patients underwent aortic valve repair in view of the perforation of the LCC and had mild aortic insufficiency at the 1-month follow-up. Longer surveillance will be essential to determine the progression of aortic regurgitation and determine the timing of intervention if needed.

**Conclusions:** Aorto-left ventricular tunnel is a very rare congenital cardiac anomaly. Accurate diagnosis is essential in guiding appropriate management strategies. Surgical treatment is relatively simple and entails the closure of one or both ends of the tunnel. Aortic insufficiency on long-term follow-up remains a concern, with up to 50% of patients requiring intervention for the same.

**Keywords:** Aorto-left ventricular tunnel, congenital heart disease

#### Computed tomography and intra-operative images



A: Oblique view CT - Demonstrating aortic end of aorto-left ventricular tunnel, ventricular end of the tunnel, left ventricular outflow tract, aneurysmal dilation of both the extracardiac and intracardiac components on the tunnel and superiorly displaced left coronary artery ostia. B: VRT CT - Showing aneurysmal aorto-left ventricular tunnel with its aortic and ventricular ends. C: Intraoperative image after aortotomy showing the aortic end of the tunnel, the unsupported left coronary cusp and the left ventricular outflow tract. D: Intraoperative image after pericardial patch closure of the aortic end of the tunnel and perforation of the left aortic cusp. E: Intraoperative imaging showing removal of clot from the distal end of the tunnel after opening it externally. F: Intraoperative image showing a right angle passed from the LVOT to demonstrate the ventricular end of the tunnel. Arrow - Aortic end of the tunnel, Arrow head - Ventricular end of the tunnel, Hash - left ventricular outflow tract and Asterisk - Perforation of left aortic leaflet

## P-250

**Management of percutaneous pulmonary valve endocarditis: A multidisciplinary approach**

Micol Rebonato, Mara Pilati, Gianfranco Butera, Adriano Carotti, Lorenzo Galletti, Sonia Albanese  
Ospedale Pediatrico Bambino Gesù, Rome, Italy

**Background and Aim:** Infective endocarditis (IE) after transcatheter pulmonary valve (PPV) replacement in CHD is a major concern and its therapeutic management still remains challenging. The incidence of IE after PPVI is estimated at 3% per year for the Melody valve and slightly lower for the Sapien valve. Herein we describe our institutional approach for the management of IE developed on percutaneous bioprosthetic pulmonic valves.

**Method:** Between January 2017 and October 2022, a total of 86 patients with complex CHD received PPV at our center, Melody valve in 30 cases and Sapien valve in 56. Of them, 4 patients required surgical treatment for IE on PPV. Clinical and therapeutic decisions were managed by a multidisciplinary team.

**Results:** Three patients had an RV-PA conduit, in two cases for PA/VSD repair and in one case of Nikaidoh for TGA/VSD/PS, while one had received transannular repair of Fallot's tetralogy. The devices used for PPV replacement were the Melody valve in 3 cases and the Sapien valve in one, defining an IE rate of 0, 1% and 0, 01% for the respective device type. The median time from transcatheter valve implantation to the onset of IE was 4.5 years (IQR 2-9). The diagnosis was confirmed in each patient based on both the Duke's criteria and a positive PET/CT scan. Blood cultures showed growth of *Granulicatella Adiacens* in one case, *Streptococcus hemolyticus* in one case and *Corynebacterium propinquum* in one case, while one patient presented with a rare form of aseptic endocarditis. Patients received at least 6 weeks of targeted or broad-spectrum antibiotic therapy. All patients underwent subsequent surgical replacement of the infected devices with pulmonary homografts. At a median follow-up of 5 months (IQR 3-7) no signs of recurrence of IE were detected.

**Conclusions:** Our data confirm the EI rate after PPV reported in the literature and the variety of infectious agents involved. The use of PET/CT and the collaboration of a multidisciplinary team is essential for diagnostic and therapeutic purposes. Furthermore, early surgical replacement of infected devices with pulmonary homografts after an adequate period of antimicrobial therapy has been shown to be safe and effective for the management of this complex condition.

**Keywords:** infective endocarditis, PPVI, homograft

## P-253

**The ross-konno procedure in patients with severe LVOTO/VSD and interrupted or hypoplastic aortic ARCH**

Eva Sames Dolzer<sup>1</sup>, Anna Maria Mittermair<sup>1</sup>, Michaela Kreuzer<sup>1</sup>, Roland Mair<sup>1</sup>, Gregor Gierlinger<sup>1</sup>, Fabian Seiber<sup>1</sup>, Andreas Tulzer<sup>2</sup>, Rudolf Mair<sup>1</sup>

<sup>1</sup>Division of Pediatric and Congenital Heart Surgery, Kepler University Hospital Linz, Linz, Austria; <sup>2</sup>Department of Pediatric Cardiology, Kepler University Hospital Linz, Linz, Austria

**Background and Aim:** Patients with ventricular septal defect and interrupted or hypoplastic aortic arch can show a severe left ventricular outflow tract obstruction, aortic valve dysplasia and ductus dependent systemic circulation. Function and size of the left ventricle is usually not impaired as the left ventricle can eject via the VSD. The neonatal Ross-Konno procedure offers a promising radical biventricular treatment option for the LVOT stenosis.

**Method:** Between 2003 and 2021 14 babies with the above described congenital lesion have received an early Ross-Konno procedure, VSD closure and concomitant aortic arch repair (13 pts; one had arch repair and VSDC previously) at our center. 9 patients presented with an interrupted arch Typ B mostly associated with an aberrant right subclavian artery, 5 patients had an hypoplastic arch. Two patients had a previous palliation with a Norwood procedure plus isthmus stent and a ductus stent and bilateral PAB from other centers.

**Results:** Median age at surgery was 14 days, median weight was 3, 3 kg. The aortic arch reconstruction was done without foreign material in 6 cases, the other patients received a homograft or pericardial patch enlargement, in two cases an additional reversed subclavian flap technique was used. Median bypass time was 341 min, median XCT was 165 min. 12 pts. had delayed sternal closure, 4 needed ECMO therapy for 5-7 days. Median ICU stay was 24 days, median hospital stay was 34 days. During the follow up period of maximum 18, 8 years (median FU 6, 4 years) there was no mortality and at last FU left ventricular function was considered good in 12 pts. and slightly reduced in 2 children. There was no case of LVOTO or autograft stenosis or insufficiency during FU. One patient received a David's procedure due to autograft dilatation 18 years after the initial operation.

**Conclusions:** The early Ross-Konno procedure with concomitant aortic arch repair and VSD closure is a challenging complex operation requiring a long aortic cross clamp time and a prolonged intensive care therapy. Nevertheless, the overall outcome of this small subset of patients was very good without mortality and with excellent long term performance of the autograft.

**Keywords:** neonatal Ross-Konno procedure, left ventricular outflow tract obstruction, interrupted aortic arch

## P-254

**Comparative analysis of fenestrated and non-fenestrated extracardiac conduit fontan in kazakhstan: A single center experience**

Tatyana Ivanova Razumova<sup>1</sup>, Aizhan Manabay<sup>1</sup>, Almira Baigalkanova<sup>1</sup>, Dmitriy Gorbunov<sup>2</sup>, Aikerim Zhanatkyzy<sup>1</sup>

<sup>1</sup>Department of pediatric cardiology, National research cardiac surgery center, Astana, Kazakhstan; <sup>2</sup>Department of pediatric cardiac surgery, National research cardiac surgery center, Astana, Kazakhstan

**Background and Aim:** The main method of surgical treatment of univentricular congenital heart diseases is hemodynamic correction based on a complete "right heart bypass": total cavapulmonary connection. There are several modifications of the Fontan anastomosis. Since 1990 extracardiac conduit has been mainly used, which is divided into two subtypes: non-fenestration and fenestration. Currently, the latter used widely in many countries, referring to the low index of postoperative pleural effusion and the number of days spent in the hospital, whereas in our country it prefers non-fenestrated conduit. There are a limited number of cardiac surgery centers in the world that perform Fontan surgery in the modification of non-fenestrated extracardiac conduit. In this regard, there are few works in the world literature devoted to the comprehensive study of long-term results with this non-fenestrated Fontan.

**Method:** Cross-sectional study with retrospective analysis. Data were analyzed with a predominance of mean and standard deviations.

The present study included 60 patients who underwent hemodynamic correction -Fontan surgery in the period from 2011 to 2022. The children were divided into two groups: in the first group, patients with the formation of fenestration of extracardiac conduit-14 (23%) and in the second group in the modification of

non-fenestrated extracardiac conduit - 46 (77%) patients, respectively ( $p < 0.01$ ).

**Results:** The number of days of pleural effusions and hospital stay among patients of the first group is 18 and 32 days, then as in the second group of 13 and 27 days. Early postoperative SaO<sub>2</sub> 87% in the first group and in the second group SaO<sub>2</sub> 90%, time of cardiopulmonary bypass and artificial lung ventilation 176 and 2946 minutes in children of the first category and in patients of the second category 152 and 2942 minutes respectively ( $p < 0.01$ ). Other parameters did not show significant differences between the two groups.

**Conclusions:** Fontan operation in the modification of non-fenestrated extracardiac conduit does not lead to a longer pleural effusion and hospital stay, the time spent on cardiopulmonary bypass is significantly less.

**Keywords:** fontan, congenital heart disease, non-fenestration, single ventricle

#### P-256

##### Extra corporal membrane oxygenation therapy in a neonatal patient with giant rhabdomyoma arising of the left ventricular wall – a case report

Roland Mair<sup>1</sup>, Eva Sames Dolzer<sup>1</sup>, Michaela Kreuzer<sup>1</sup>, Gregor Gierlinger<sup>1</sup>, Fabian Seeber<sup>1</sup>, Bruno Schachner<sup>2</sup>, Paiman Nawrozi<sup>2</sup>, Rudolf Mair<sup>1</sup>

<sup>1</sup>Department of Pediatric and Congenital Heart Surgery, Kepler University Hospital, Linz, Austria; <sup>2</sup>Department of Cardiac, Vascular and Thoracic Surgery, Kepler University Hospital, Linz, Austria

**Background and Aim:** Cardiac rhabdomyoma account for about 20% of all benign tumors but are the most common cardiac neoplasia in children. The disease is caused by a mutation of the TSC1- or TSC2-gene, therefore an association with tuberous sclerosis complex is common. In most cases cardiac rhabdomyomas do not cause any symptoms and need no further treatment, as they tend to regress with age.

In some patients, depending on the size and location of the rhabdomyoma, they can cause arrhythmia, cardiac failure or ventricular inflow- or outflow tract obstructions.

Surgical treatment is limited. Conservative treatment with mTOR inhibitors has been reported.

**Method:** We report a case of a patient with giant cardiac rhabdomyoma arising from the left ventricular free wall, causing total compression of the left lung and severe arrhythmia, with wide QRS complex tachycardia, resulting in low cardiac output.

The fetus was monitored over the last trimester of gestation. Ultrasound showed one big tumor arising from the left ventricular wall with 49 x 42 x 33 mm.

The male infant was delivered by cesarean section at 35 + 0 GA because of fetal tachycardia.

The patient had to be intubated right away, amiodarone infusion was started and inotropic support with epinephrine infusion had to be induced.

Due to the worsening respiratory situation the patient had to be taken on ECMO on day one. Intraoperative view of the tumor showed, that it was supplied by branches of the left anterior descending coronary artery and was not amenable to resection or surgical reduction, because too much left ventricular myocardium and coronary vessels would have been resected.

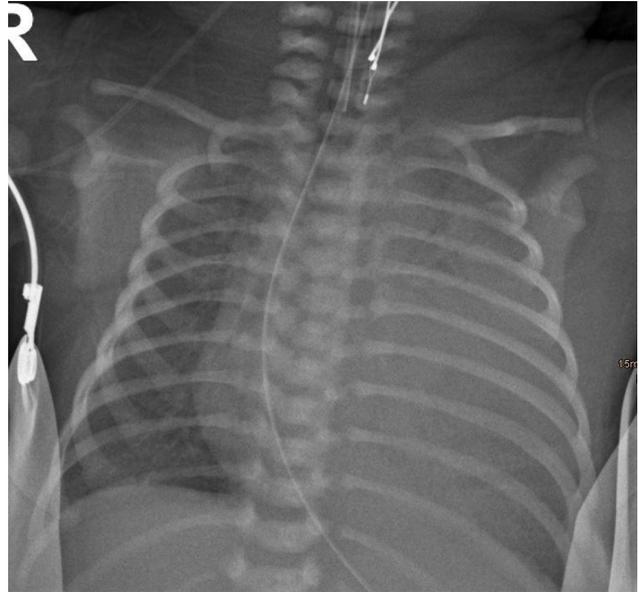
Oral medication with mTOR inhibitor everolimus was initiated. **Results:** Tumor showed remarkable regression during first two weeks (CT-scan). Chest x-ray almost normalized.

After 28 days sinus rhythm was observed for the first time. Episodes of sinus rhythm extended over the following days.

At day 35 the patient was successfully weaned from ECMO.

**Conclusions:** In patients with giant rhabdomyoma, causing low cardiac output, respiratory failure and not suitable for resection, awaiting tumor regression with everolimus therapy on ECMO support is a therapeutic option.

**Keywords:** rhabdomyoma, everolimus



First chest x-ray

#### P-257

##### Aortic valve neocuspidization – bailout procedure or upcoming standard in pediatric patients ? – first experiences and midterm follow up

Ina Michel Behnke<sup>1</sup>, Dominik Wiedemann<sup>2</sup>, Erhan Urganci<sup>2</sup>, Maja Daurach<sup>1</sup>, Julian Heno<sup>1</sup>, Daniel Zimpfer<sup>3</sup>

<sup>1</sup>Department of Pediatric Cardiology, Pediatric Heart Center Vienna, Medical University Vienna, Austria; <sup>2</sup>Department of Heart Surgery, Pediatric Heart Center Vienna, Medical University Vienna, Austria; <sup>3</sup>Department of Heart Surgery, Medical University Graz, Austria; <sup>4</sup>Department of Heart Surgery, Pediatric Heart Center Vienna, Medical University Vienna, Austria

**Background and Aim:** Surgical techniques for aortic valve disease (AVD) in pediatric patients include repair techniques as first option, followed by Ross operation (golden standard in many units). Finally aortic valve replacement by biological or mechanical substitutes has to be performed. All techniques have advantages and drawbacks in terms of longevity reintervention and anticoagulation. Neocuspidization as described by Ozaki adds to the armamentarium for a wide spectrum of congenital or infectious AVD as promising alternative also in children. Our experience in several entities of AVD is described.

**Method:** Five patients with a median age of 5 y (4 w - 12 y) and a body weight of 3.6 - 45.2kg (med 24 kg) underwent the Ozaki-type operation. Three had bicuspid aortic valve, two had AR III, two combined AS/AR and one had Truncus arteriosus type 1

(TAC). Two patients were postoperative (TAC) and post balloon valvuloplasty respectively. Autologous pericardium was used in 4 patients for restoration of aortic cusps, in one a Matrix patch was used (TAC).

**Results:** All patients survived; ICU-stay was 2–7 days (med 2 d), hospital-stay 9–40 d (med 13 d). Anticoagulation: Warfarin (n = 2), LMW-Heparin (n = 1) for three months, Aspirin thereafter. The two smallest patients received Clopidogrel. LV size/-function normalized in all. During FUP (max 3 y), three patients had no further surgery with a residual gradient of 10–15 mmHg. AR in these patients was mild with a slight increase starting in the second year after surgery. The TAC-patient developed significant AS/AR within 9 mo and underwent implantation of a decellularized homograft in aortic position 14 mo after surgery. One patient developed severe stenosis 2 y postop within a few weeks - intraoperatively diverse cusp morphology/function was observed: one cusp completely normal, one calcified immobile and one degenerated/shrunked. This patient underwent Ross operation.

**Conclusions:** Neocuspidization is feasible also in young patients and can be offered as Bailout procedure, accepting limited longevity. Heterologous tissue should be avoided. Frequency of reoperation compares to Ross operation in this age group. Temporary anticoagulation with Warfarin must be advised. For older children it might be a reasonable alternative when Ross operation can not be offered.

**Keywords:** Aortic stenosis, aortic regurgitation, Ozaki, neocuspidization, Ross operation

#### P-259

##### **A handsewn pericardial valved pulmonary conduit: pulsatile flow loop in vitro and acute porcine in vivo evaluation**

Lisa Carlson Hanse<sup>1</sup>, Marcell Juan Tjornild<sup>1</sup>, Zarmiga Karunanithi<sup>3</sup>, Johannes Høgfjeldt Jędrzejczyk<sup>1</sup>, Lejla Islamagic<sup>2</sup>, Nynne Emilie Hummelshøj<sup>2</sup>, Malene Enevoldsen<sup>2</sup>, Peter Johansen<sup>4</sup>, Mette Høj Lauridsen<sup>5</sup>, Vibeke Elisabeth Hjortdal<sup>6</sup>

<sup>1</sup>Department of Cardiothoracic and Vascular Surgery, Aarhus University Hospital, Aarhus, Denmark; <sup>2</sup>Department of Clinical Medicine, Aarhus University Hospital, Aarhus, Denmark.; <sup>3</sup>Department of Anaesthesiology, Viborg Regional Hospital, Viborg, Denmark;

<sup>4</sup>Department of Electrical and Computer Engineering, Section for Biomedical Engineering, Aarhus University, Aarhus, Denmark.;

<sup>5</sup>Department of Paediatrics and Adolescent Medicine, Aarhus University Hospital, Aarhus, Denmark.; <sup>6</sup>Department of Cardiothoracic and Vascular Surgery, Rigshospitalet, Copenhagen, Denmark.

**Background and Aim:** Right ventricle to pulmonary artery anatomic discontinuity is common in complex congenital heart malformations. Handsewn conduits are a practiced method of repair. We performed pulmonary valve replacement with a handsewn pericardial valved pulmonary conduit in vitro and in vivo.

**Method:** A pulsatile flow-loop model (in vitro) and an acute 60-kilogram porcine model (in vivo) were used. With echocardiography and pressure catheters, baseline geometry and hydro-/hemodynamics were measured. The pulmonary valve was

replaced with a handsewn glutaraldehyde treated pericardial valved pulmonary conduit corresponding to a 21-mm prosthetic valve. Geometry and hydro-/hemodynamics were then reassessed.

**Results:** In vitro, 15 pulmonary trunks at 4 L/min and 13 trunks at 7 L/min, and in vivo, 11 animals were investigated. The valved pulmonary conduit was easy to produce at the table and to stitch in. All valves were sufficient in vitro and in vivo. The mean transvalvular pressure gradient in the native valve and the conduit was 8 mmHg (SD: 2) and 7 mmHg (SD: 2) 4 L/min in vitro, 19 mmHg (SD:3) and 17 mmHg (SD: 4) at 7 L/min in vitro, and 3 mmHg (SD: 2) and 6 mmHg (SD: 3) in vivo.

**Conclusions:** Acute in vitro and in vivo investigations demonstrated a pericardial valved pulmonary conduit with no clinically significant regurgitation or stenosis. The handsewn conduit can be produced fast and easy at the operating table.

**Keywords:** Valved conduit, congenital heart disease, experimental

#### P-260

##### **Probability of truncal valve replacement within 30 years after the correction**

Václav Chaloupecký Jr., Roman Gebauer, Rudolf Poruban, Ondřej Materna, Jan Kovanda, Jan Janoušek, Václav Chaloupecký  
Children's Heart Centre, Motol University Hospital, Prague, Czech Republic

**Background and Aim:** Truncus arteriosus is severe, but manageable congenital heart disease with high morbidity associated with multiple reinterventions. The aim of this study was to evaluate long-term survival and to identify the risk factors for truncal valve replacement.

**Method:** This single-centre nation-wide retrospective study included consecutive children who underwent truncus arteriosus repair in the Czech Republic between 1991 and 2020 (N = 109). Long-term outcomes were obtained by cross-mapping individual data with the National Death Registry and the National Registry of Cardiovascular Interventions for adults.

**Results:** The median patient age at complete truncus arteriosus repair was 38 days (IQR 22.5–60 days) and median weight 3.3 kg (2.8–3.7 kg). Eleven patients (10.1%) who died ≤ 30 days after the correction were excluded from the study. Median follow-up of 98 early survivors was 10.2 years (IQR 0.1–29.3 years). Late mortality was 10.1% (10 of 98 pts) with the survival probability at 5, 15, and 25 years of 91%, 88%, and 86%, respectively. Twenty patients (20.4%) underwent 29 truncal valve replacements. Probability of survival without truncal valve replacement at 5, 15, and 25 years was 82%, 70% and 61%, respectively. Independent risk factors for truncal valve replacement were moderate or greater truncal valve regurgitation at repair (HR, 4.34; 95% CI, 1.88 – 10.06, p < 0.001) and the presence of quadricuspid truncal valve (HR, 4.13; 95% CI, 1.92 – 8.86, p < 0.000).

**Conclusions:** Truncal valve replacement is frequently needed after truncus arteriosus repair during long-term follow-up. The presence of quadricuspid truncal valve and/or at least moderate truncal valve regurgitation at primary repair are significant risk factors.

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**Keywords:** congenital heart disease, truncus arteriosus, truncal valve function

### P-262

#### Treatment of pediatric patients with isolated right ventricular hypoplasia

Olena Stohova, Tatyana Hura, Danylo Shypov, Oleksandra Motrechko, Andrii Kurkevych  
Ukrainian Children's Cardiac Center, Kyiv,

**Background and Aim:** Isolated right ventricular hypoplasia (IRVH) is a rare anomaly with the underdeveloped trabeculated part of the ventricle and without important valvular abnormalities. Patients with IRVH usually have variable manifestations. The aim of this study was to analyze of the anatomical features of right heart that affect the choice of treatment management.

**Method:** From 2017 to 2022 seven patients with IRVH from neonatal period to 6 years old were observed. The apex of the heart was formed by the LV due to hypoplasia of the trabecular part of the RV in all children. Patients were divided into three groups according to the RV/LV ratio by echocardiography.

**Results:** In the first and second groups, a mild or moderate hypoplasia of the RV was diagnosed, Z-score TV and PV were within the normal range. In the first group (n = 3), RV/LV ratio was 0.8–0.85, saturation of 97%–98%, left-to-right shunt across the PFO and had no signs of impaired pulmonary blood flow. Two of them were premature and required PDA closure. The second group (n = 2), had moderate hypoplasia of the RV. The RV/LV ratio was 0.68 and 0.75, saturation was 82% and 93%, respectively. Glenn shunt and atrioseptectomy was performed in the first of them at the age of 14 months. The second patient received medical treatment with b-blockers. In the third group (n = 2) there was severe hypoplasia of the RV: RV/LV ratio was 0.58 and 0.4, Z-score TV -2.62 and -2.97 and saturation - 60% and 62%, respectively. The first patient underwent PDA stenting at the age of 4 days to ensure adequate pulmonary blood flow, the second patient underwent Blalock-Taussig shunt on the first day of the life and an atrioseptostomy on the 19th day. The last patient died at the age of 38 days.

**Conclusions:** Isolated right ventricular hypoplasia is a rare anomaly with very different manifestations. Patients with severe hypoplasia of the RV have significant clinical signs in the neonatal period and need urgent interventions. While another patients may be asymptomatic during many years and require only observation.

**Keywords:** Isolated right ventricular hypoplasia, congenital heart defects

### P-263

#### Association between reported activity level, objective exercise capacity and cardiac magnetic resonance imaging parameters in tetralogy of fallot

Benedetta Leonardi<sup>1</sup>, Federica Gentili<sup>2</sup>, Marco Alfonso Perrone<sup>1</sup>, Fabrizio Sollazzo<sup>3</sup>, Stefani Silva Kikina<sup>1</sup>, Rachel Wald<sup>4</sup>, Vincenzo

Palmeri<sup>3</sup>, Aurelio Secinaro<sup>5</sup>, Maria Giulia Gagliardi<sup>1</sup>, Attilio Turchetta<sup>2</sup>, Lorenzo Galletti<sup>1</sup>, Massimiliano Bianco<sup>3</sup>, Fabrizio Drago<sup>1</sup>  
<sup>1</sup>Department of Cardiology and Cardiac Surgery, Bambino Gesù Children's Hospital IRCCS, 00165 Rome, Italy; <sup>2</sup>Department of Sport Medicine, Bambino Gesù Children's Hospital IRCCS, 00165 Rome, Italy; <sup>3</sup>Unità Operativa Complessa di Medicina dello Sport e Rieducazione Funzionale, Fondazione Policlinico Universitario Agostino Gemelli IRCCS, Università Cattolica del Sacro Cuore, 00168 Rome, Italy; <sup>4</sup>Peter Munk Cardiac Centre and Joint Department of Medical Imaging, University Health Network, University of Toronto, Toronto, ON M5S 1A1, Canada; <sup>5</sup>Advanced Cardiothoracic Imaging Unit, Department of Imaging, Bambino Gesù Children's Hospital IRCCS 00165 Rome, Italy

**Background and Aim:** Patients with repaired Tetralogy of Fallot (rToF) typically report having preserved subjective exercise tolerance. Chronic pulmonary regurgitation (PR) with varying degrees of right ventricular (RV) dilation as assessed by cardiac magnetic resonance imaging (MRI) is prevalent in rToF and may contribute to clinical compromise. Cardiopulmonary exercise testing (CPET) provides an objective assessment of functional capacity, and the International Physical Activity Questionnaire (IPAQ) denotes physical activity level. Our aim was to assess the association between CPET values, IPAQ measures, and MRI parameters.

**Method:** All rToF patients who had both an MRI and CPET performed between March 2019 and June 2021 were selected. Clinical data were extracted from electronic records (including demographic, surgical history, New York Heart Association (NYHA) functional class, QRS duration, arrhythmia, MRI parameters, and CPET data). Physical activity level, based on the IPAQ, was assessed at the time of CPET.

**Results:** Eighty-four patients (22.8 ± 8.4 years) showed a reduction in exercise capacity (median peak VO<sub>2</sub> 30 mL/kg/min (range 25–33); median percent predicted peak VO<sub>2</sub> 68% (range 61–78)). Peak VO<sub>2</sub>, correlated with biventricular stroke volumes (SV) (RVS: β = 6.11 (95% CI. 2.38–9.85), p = 0.002; LVS: β = 15.69 (95% CI. 10.16–21.21), p < 0.0001) and LVEDVi (β = 8.74 (95% CI. 0.66–16.83), p = 0.04) on multivariate analysis adjusted for age, gender, and physical activity level. Other parameters which correlated with SV included oxygen uptake efficiency slope (OUES) (RVS: β = 6.88 (95% CI. 1.93–11.84), p = 0.008; LVS: β = 17.86 (95% CI. 10.31–25.42), p < 0.0001) and peak O<sub>2</sub> pulse (RVS: β = 0.03 (95% CI. 0.01–0.05), p = 0.007; LVS: β = 0.08 (95% CI. 0.05–0.11), p < 0.0001). On multivariate analysis adjusted for age and gender, physical activity level correlated significantly with peak VO<sub>2</sub>/kg (β = 0.02, 95% CI. 0.003–0.04; p = 0.019).

**Conclusions:** A reduction in objective exercise tolerance in rToF patients was observed. Biventricular SV and LVEDVi were associated with peak VO<sub>2</sub> irrespective of RV size. OUES and peak O<sub>2</sub> pulse were also associated with biventricular SV. While physical activity level was associated with peak VO<sub>2</sub>, the incremental value of this parameter should be the focus of future studies.

**Keywords:** Tetralogy of Fallot, cardiopulmonary exercise testing, physical activity level

Characteristics of 84 rToF patients.

Overall (N=84)		
<b>Demographic features</b>	Male sex, n (%)	41 (48.8%)
	Age at repair (months) (median, IQR)	13.4 (0.0 – 87.0)
	Age at CPET (yrs) (median, IQR)	21.1 (15.0-30.0)
	Time between surgery and CPET (yrs) (median, IQR)	20.2 (10.2 – 28.9)
	BSA (m <sup>2</sup> ) (median, IQR)	1.7 (1.1 – 2.2)
	BMI (median, IQR)	22.6 (15.6 -33.3)
<b>MRI parameters</b>	RVEDV (ml) (median, IQR)	205.0 (106.0 – 327.9)
	RVEDVi (ml/m <sup>2</sup> ) (median, IQR)	122.1 (63.3 – 174.3)
	RVESV (ml) (median, IQR)	92.2 (43.0 – 152.0)
	RVESVi (ml/m <sup>2</sup> ) (median, IQR)	54.9 (25.7 – 84.6)
	RVEF (%) (median, IQR)	55.0 (45 – 69.0)
	RVSV (ml/beat) (median, IQR)	109.7 (6.0 – 182.5)
	LVEDV (ml) (median, IQR)	125.0 (69.0– 206.5)
	LVEDVi (ml/m <sup>2</sup> ) (median, IQR)	75.4 (48.8 – 113.7)
	LVESV (ml) (median, IQR)	54.1 (27.0 – 86.0)
	LVEDVi (ml/m <sup>2</sup> ) (median, IQR)	33.5 (19.9 – 52.9)
	LVEF, (%) (median, IQR)	57.0 (45.5 – 66.0)
	LVSV (ml/beat) (median, IQR)	69.8 (42.0 – 108.3)
	PR (%) (median, IQR)	37.4 (27.0 – 60.0)
	RVOTO and/or PAs branches stenosis, n (%)	8 (9.5)
	<b>CPET values</b>	Total test duration (s) (median, IQR)
Peak HR (bpm) (median, IQR)		176 (167 – 182)
Peak HR (%) (median, IQR)		88 (85 – 92)
HR at AT (bpm) (median, IQR)		128 (119 – 138)
Peak VO <sub>2</sub> (ml/min) (median, IQR)		1885 (1530 – 2166)
Peak VO <sub>2</sub> /Kg (ml/min/Kg) (median, IQR)		31 (25 – 33)
Peak VO <sub>2</sub> /Kg (% of predicted) (median, IQR)		68 (61 – 78)
VO <sub>2</sub> at AT (ml/min/Kg) (median, IQR)		19.7 (18.3 – 23.7)
Peak RER (median, IQR)		1.09 (1.02 – 1.15)
Peak O <sub>2</sub> pulse [ml/bpm] (median, IQR)		10.4 (8.81; 12.28)
Peak O <sub>2</sub> pulse (% of predicted) (median, IQR)		81 (71; 93)
O <sub>2</sub> pulse trend n (%)		
– Increasing		7 (10.3%)
– Flattening		58 (85.3%)
– Decline		3 (4.4%)
OUES (ml/min/l/min) (median, IQR)		1.908 (1.665 – 2.538)
OUES (% of predicted) (median, IQR)		79 (68 – 85)
VE/VCO <sub>2</sub> at AT (median, IQR)		27.5 (25.0 – 29.9)
VE/VCO <sub>2</sub> slope (AT) (median, IQR)		25.4 (23.6 – 28.9)
Peak VE/VCO <sub>2</sub> (median, IQR)		31.1 (27.3 – 34.3)
VE/VCO <sub>2</sub> slope (VCP) (median, IQR)		28.3 (25.5 – 31.5)
VE/VCO <sub>2</sub> slope (stop) (median, IQR)		30.7 (27.5 – 34.5)
FVC (l) (median, IQR)		3.6 (2.97 – 4.26)
FEV1 (l) (median, IQR)		3.1 (2.77 – 3.79)
Peak VE (l/min) (median, IQR)		61 (54 – 76)
Peak VE/VO <sub>2</sub> (median, IQR)		33.0 (28.9 – 38.2)
BR (%) (median, IQR)	50 (40 – 57)	

**Table 1:** Data are expressed as median values and interquartile ranges for continuous variables and as counts and percentages for categorical variables. Legend: AT: anaerobic threshold; BR: breathing reserve. BSA: Body surface area; HR: heart rate; LVEDV: left ventricular end-diastolic volume; LVEDVi: left ventricular end-diastolic volume indexed to body surface area; LVEF: left ventricular ejection fraction; LVESV: left ventricular end-systolic volume; LVESVi: left ventricular end-systolic volume indexed to body surface area; LVSV: left ventricular stroke volume; OUES: oxygen uptake efficiency slope; PR: pulmonary regurgitation; RER: respiratory exchange ratio; RVEDV: right ventricular end-diastolic volume; RVEDVi: right ventricular end-diastolic volume indexed to body surface area; RVEF: right ventricular ejection fraction; RVESV: right ventricular end-systolic volume; RVESVi: right ventricular end-systolic volume indexed to body surface area; RVSV: right ventricular stroke volume; VE/VCO<sub>2</sub>: ventilatory equivalent for CO<sub>2</sub>; VE: ventilation; VO<sub>2</sub>: oxygen consumption.

## P-264

**Is fallot corrective surgery forever? late reoperations during childhood and adulthood**

María Luz Polo López<sup>1</sup>, Bunty Kishore Ramchandani Ramchandani<sup>1</sup>, Blanca Torres Maestro<sup>1</sup>, Juvenal Rey Lois<sup>1</sup>, Raúl Sánchez Pérez<sup>1</sup>, Álvaro González Rocafort<sup>1</sup>, Tomasa Centella Hernández<sup>1</sup>, María Jesús Lamas Hernández<sup>1</sup>, Montserrat Bret Zurita<sup>4</sup>, Ángela Uceda Galiano<sup>2</sup>, José Ruiz Cantador<sup>3</sup>, Ángel Aroca Peinado<sup>1</sup>

<sup>1</sup>Congenital cardiac surgery. La Paz-Ramón y Cajal University Hospital. Madrid. Spain; <sup>2</sup>Pediatric Cardiology. La Paz University Hospital. Madrid. Spain; <sup>3</sup>Congenital Adult Cardiology. La Paz University Hospital. Madrid. Spain; <sup>4</sup>Pediatric Radiology. La Paz University Hospital. Madrid. Spain

**Background and Aim:** Fallot corrective surgery has an excellent prognosis and pulmonary insufficiency-(PI) is usually well tolerated until adulthood. We analyse late reoperations of these patients during their childhood or adulthood along the period 2003-2022 in our Congenital Heart Unit, and present our results.

**Method:** Retrospective study of 225 late reoperations in 209 patients (70 children, 155 adults). Reoperations were done with extracorporeal circulation and moderate hypothermia. Right heart lesions were operated on beating heart, and the aorta was clamped in cases with left heart pathology or residual septal defects. We analyse our short and long term follow-up results. Statistic analysis was performed with SPSS.20. Study approved by the ethics committee of our hospital.

**Results:** Median age: adults 40 years [IQR: 30-45], children 8, 9 years [IQR: 5-13], Preoperative median NYHA functional class: adults 2 [IQR: 2-3], children 1 [IQR: 1-2]. Main indications for surgery were PI-70% in adults, and children had PI-41% and pulmonary stenosis-41%. Surgical techniques: 91% adults and 44% children received a porcine pulmonary bioprosthesis, and 46% children had a bovine jugular vein connection between right ventricle and pulmonary artery. Associated surgery in 62% adults and 41% children: mainly closure of septal defects and tricuspid valvuloplasty. In-hospital mortality was 2, 6% in adults and 1, 4% in children. Median follow-up was 5 years (maximum 11 years). Late mortality was 1, 3% in adults and 2, 9% in children. Children had more percutaneous and surgical reinterventions and more endocarditis than adults, and also had less need of arrhythmia treatment with pacemaker or internal automatic defibrillator (see Table).

**Conclusions:** Late reoperations after Fallot corrective surgery can be done safely in an experienced Congenital Heart Unit while in childhood and adulthood. We recommend interposing a pulmonary bioprosthesis in adult age whenever possible, because the durability and late results are better in comparison to be operated in children age. Rhythm stimulation in the follow-up after reoperation is needed with more frequency in adults than in children.

**Keywords:** Fallot reoperation, pulmonary bioprosthesis

	Adults	Children
Late reoperation	2 %	17,4 %
Pulmonary stenosis and/or insufficiency	11 %	45 %
Interventional catheterization	4 %	29 %
Endocarditis	3,3 %	15 %
Pulmonary prosthesis (percutaneous or surgical)	2,2 %	36 %
Pacemaker o defibrillator	26 %	4,4 %

**Table 1** Long term follow up in adults and children with Fallot after reoperations

## P-267

**Double aortic arch: A rare cause of respiratory distress in infants a case report and review of the literature**

Marwa Jarraya<sup>1</sup>, Rania Gargouri<sup>1</sup>, Hela Fendri<sup>2</sup>, Mohamed Ali Zghal<sup>3</sup>, Mariem Jabeur<sup>1</sup>, Selma Charfeddine<sup>1</sup>, Leila Abid<sup>1</sup>

<sup>1</sup>cardiology departement, University of medecine of Sfax, Sfax, Tunisia; <sup>2</sup>radiology departement, University of medecine of Sfax, Sfax, Tunisia; <sup>3</sup>pediatrics departement, University of medecine of Sfax, Gabes, Tunisia

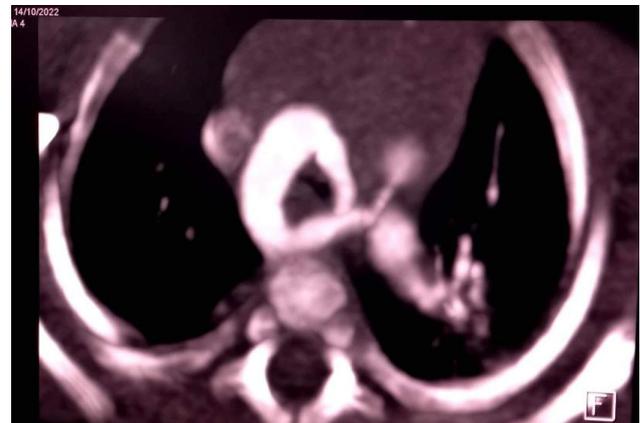
**Background and Aim:** Double aortic arch (DAA) is the most common vascular ring anomaly. Symptoms are related to tracheal and oesophageal compression and may cause respiratory distress in some children. However, the symptomatology may be limited to a simple stridor. It is therefore prudent to have a high index of suspicion when evaluating cases of paediatric stridor

**Method:** We report a case of a 2-month-old infant that was born at full term and had no medical or surgical history who presented with progressive stridor and dyspnea.

On examination, we noticed a stridor, with suprasternal and intercostal recessions. Oxygen saturation on room air was 93% and chest auscultation revealed bilateral transmitted stridor. A chest X-ray was normal. Echocardiography was performed and revealed a left-right shunting atrial septal defect, a restrictive persistent ductus arteriosus with an aortic arch.

**Results:** Thoracic computed tomography (CT) was performed and revealed duplication of the aortic arch encircling the thoracic trachea and oesophagus. The CT confirm the double aortic arch, its location and the severity of airway and oesophageal compression. Three-dimensional reconstruction shows that the right side arch was bigger and dominant and then was helpful to plan surgical intervention. The patient was then referred to the cardiothoracic surgery and underwent surgical repair with resection of the left arch by left posterolateral thoracotomy.

**Conclusions:** Vascular ring is reported to account for <1% of congenital heart disease. DAA is the most common form of it. It can be diagnosed in early childhood due to symptoms related to oesophageal and/or tracheal compression. DAA is usually isolated but can be rarely associated with cardiac malformations including ventricular septal defect and tetralogy of Fallot. Classically, DAA has three types: right dominant aortic arch (75%), left dominant aortic



Axial section in mediastinal window focused on the aortic ring: Evidence of two aortic arches, one predominant right and one rudimentary left. Each ring gives rise to a carotid artery and a subcalvarial artery. In the back, there is a persistent ductus arteriosus. The whole makes a real aortic ring which encircles and exerts a mass effect on the trachea.

arch (20%), and balanced type (5%) Left untreated, it may lead to significant morbidity and mortality due to airway obstruction. Surgical repair remains the mainstay of treatment and is indicated for symptomatic patients or as a supplementary procedure in other cardiac surgery. The principle of surgery is to relieve the compression on the trachea and/or esophagus by the division of the lesser arch.

**Keywords:** computed tomography, Compression, Surgery, stridor

## P-268

### Cardiopulmonary parameters during exercise in patients with tetralogy of fallot

Benedetta Leonardi<sup>1</sup>, Elettra Pomiato<sup>1</sup>, Federica Gentili<sup>2</sup>, Arthur Gavotto<sup>3</sup>, Stefani Silva Kikina<sup>1</sup>, Attilio Turchetta<sup>2</sup>, Maria Giulia Gagliardi<sup>1</sup>, Giulio Calcagni<sup>1</sup>, Rosalinda Palmieri<sup>1</sup>, Fabrizio Drago<sup>1</sup>  
<sup>1</sup>Department of Cardiology and Cardiac Surgery, Bambino Gesù Children's Hospital IRCCS, 00165 Rome, Italy; <sup>2</sup>Department of Sport Medicine, Bambino Gesù Children's Hospital IRCCS, 00165 Rome, Italy; <sup>3</sup>Paediatric and Congenital Cardiology Department, M3C Regional Reference CHD Centre, University Hospital, Montpellier, France

**Background and Aim:** The use of CPET as in many cardiovascular diseases, whether congenital or acquired, has become increasingly significant. However, while the values of CPET parameters considered normal for the healthy population are well known, there are still uncertainties regarding the "normal" values for individuals with a specific pathology. In Tetralogy of Fallot (ToF), where this test is becoming increasingly important in their follow-up, it is not yet clear at which values these parameters should be considered abnormal. Therefore, our aim was to evaluate these parameters in a large cohort of ToF patients.

**Method:** All ToF patients who were able to perform CPET on a cycle ergometer between 2020–2022 were enrolled. Age at first corrective surgery and the type of surgery were recorded.

**Results:** Two hundred seventy-nine patients were enrolled (58% male, mean age 19.6±8.0 years old at CPET). Oxygen consumption was statistically lower in patients who underwent corrective surgery after the first year of age (31.3+8.3 vs 25.6+6.5; p = 0.001), but surprisingly not different in relation to the type of surgery. Furthermore, in our data, oxygen consumption decreased with patient age from the second/third decade of life (Table 1), in contrast to oxygen pulse, VE/VCO<sub>2</sub> slope, VE/VCO<sub>2</sub> ratio and finally the OUES.

	VO <sub>2</sub> /kg peak (mL/min/Kg)		O <sub>2</sub> pulse peak (mL/beat)		VE/VCO <sub>2</sub> slope		VE/VCO <sub>2</sub> peak		OUES	
	Mean	SD	Mean	SD	Mean	SD	Mean	SD	Mean	SD
≤ 10 yrs (29)	41.4	7.0	na	na	38.0	7.6	na	na	1034	166
10.1-15 yrs (51)	30.5	6.6	9.6	1.6	32.3	4.4	33.2	3.9	1788	412
15.1-20 yrs (88)	28.1	7.2	10.2	2.7	28.8	5.3	32.6	4.6	1985	605
20.1-30 yrs (77)	26.4	5.4	5.4	3.4	27.4	4.1	32.1	5.7	1872	471
30.1-40 yrs (28)	23.2	5.1	10.9	4.1	28.1	4.3	31.2	4.7	1967	733
40.1-50 yrs (6)	21.1	3.5	9.5	2.6	31.5	6.7	33.2	4.7	1809	502

**Table 1:** CPET parameters according to age

Legend: OUES: oxygen uptake efficiency slope; VE/VCO<sub>2</sub>: ventilatory equivalent for CO<sub>2</sub>; VE: ventilation; VO<sub>2</sub>: oxygen consumption.

**Conclusions:** The oxygen peak, expression of the functional capacity of the patient with Tetralogy of Fallot, tends to be influenced by the age at the surgical correction, but not by the type of the repair. It seems to start to get worse in the second/third decade of life. Further studies on a larger case series are mandatory to establish benchmarks in this population.

**Keywords:** cardiopulmonary exercise stress, Tetralogy of Fallot

## P-269

### Ross procedure in paediatric patients: long term follow up in an italian centre.

Luca Zanella<sup>1</sup>, Alice Pozza<sup>2</sup>, Elena Reffo<sup>2</sup>, Alvise Guariento<sup>1</sup>, Biagio Castaldi<sup>2</sup>, Domenico Sirico<sup>2</sup>, Giovanni Di Salvo<sup>2</sup>, Vladimiro Vida<sup>1</sup>, Massimo Padalino<sup>1</sup>

<sup>1</sup>Paediatric and Congenital Cardiac Surgery Unit, Department of Cardiac, Thoracic, Vascular Sciences and Public Health, University of Padova, Padova, Italy.; <sup>2</sup>Paediatric Cardiology Unit, Department of Health of Woman and Child, University of Padova, Padova, Italy.

**Background and Aim:** Ross procedure consists of re-placement of diseased aortic valve with the pulmonary autograft. The overall survival after it is comparable with general population, giving better quality of life and long-term outcomes compared with other aortic valve replacement techniques in young patients. The aim of this study is to analyse outcomes of Ross procedure on patients aged less than 18 years.

**Method:** This is a single-centre observational study. Patients and data were collected from 1997 to 2022 by using a hospital-based informatic system and archival research.

**Results:** Since 1997, 34 patients aged less than 18 years old treated with Ross procedure were collected. The median age at intervention was 10, 75 years. Indications for surgery were aortic stenosis in 18 cases (52, 9%), aortic regurgitation in 5 (14, 7%), aortic steno-regurgitation in 11 (32, 35%). Seven patients had associated congenital heart diseases (20, 5%) and 11 (32, 35%) had previous cardiac surgery. The Ross techniques were 28 total free standing root replacement (81, 5%) and 6 Ross-Konno procedures (18, 5%). At a follow up time of 65, 5 months (Interquartile 12, 5–202, 5) the mortality consists of 2 patients (5, 9%) characterized by the presence of a complex congenital heart disease. Only 1 child had an atrio-ventricular block requiring a permanent pace-maker implantation. Reinterventions on left side heart occurred in 4 patients (11, 8%) at median of 76 months (Interquartile 7, 25–159), due to aneurism or severe aortic regurgitation. At a median of 204 months (Interquartile 30, 75–218), reinterventions on right heart appeared in 8 cases (29, 6%). One patient with a complex combined congenital heart disease undergone to a heart transplant.

On patients free from reinterventions, at a follow up time of 65, 5 months (Interquartile 12, 5–202, 5), the autograft and pulmonary functions were good in all of them.

**Conclusions:** Ross procedure in patients aged less than 18 years old could be a good option for aortic valve replacement in selected patients, because in this study deaths occurred in patients with associated complex congenital heart diseases. Moreover, the few patients experienced reinterventions had good results and reinterventions occurred mainly after Ross performed in very young patients. Children who remain free from reintervention had good function of autograft and homograft.

**Keywords:** Ross procedure, aortic disease, paediatric, autograft, homograft

**Education/Training****P-271****Creating and implementing a training programme for neonatologist-performed echocardiography: How, Who, What, Why?**

Maaïke ten Westenend<sup>1</sup>, Michiel Schokking<sup>2</sup>, Willem Pieter De Boode<sup>3</sup>  
<sup>1</sup>Department of Paediatrics, Radboudumc, Nijmegen, The Netherlands;  
<sup>2</sup>Department of Paediatric Cardiology, Radboudumc Amalia Children's Hospital, Nijmegen, The Netherlands; <sup>3</sup>Department of Neonatology, Radboudumc Amalia Children's Hospital, Nijmegen, The Netherlands

**Background and Aim:** Neonatologist Performed Echocardiography (NPE) is a relatively new and essential part of hemodynamic assessment of newborns. However, teaching and learning echocardiography is a complex process. Currently, no formal and accredited NPE training programme has been implemented in Europe. This led to the initiation of the NPE project within the ESPR. The aim is to create a safe and competency-based NPE training programme, by doing comprehensive preparation of investigating design requirements and current practices. Recently, infant manikins became available for simulation training of echocardiography. The research question is how experienced NPE-instructors use infant manikins for learning to perform basic and safe echocardiography.

**Method:** In 2021, the ESPR launched the NPE projectgroup, consisting of a researcher, a paediatric cardiologist and NPE-neonatologist. The concept of Design Based Research (DBR) is used to build a NPE training programme, that ensures the highest quality of NPE. To investigate current training practices, semi-structured interviews were performed with experienced NPE instructors. Thematic analyses was performed on the transcripts of the interviews by two researchers. By discussion a definitive list of codes was produced.

**Results:** From the eleven interviews a range of themes were generated: preparation, use of models, instruction style, study load per session, personalized education, time investment, competency over hours, feedback approaches, automation of movements, independent discovery or mimic movements, unawareness of teaching style, and use of clinical context. Simulation training was considered useful as a pre-clinical training tool, and providing relevant clinical context will enhance the trainee's motivation.

**Conclusions:** An optimal training programme should be designed based upon comprehensive evaluation of current practices and experiences with NPE education, and relevant evidence from the literature. Multiple education styles were identified, but a shared vision could be distilled regarding 1) preparation, 2) personalised training, 3) competency based, 4) limited information per session, and 5) practising on manikin before scanning live infants. These themes are translated into design recommendations for the European NPE training programme.

**Keywords:** Neonatologist Performed Echocardiography, training, design based research

**P-273****Illustrating heart defects with 3D-printed models – a new principle of student teaching**

Sabrina Martens<sup>1</sup>, Jonas Weber<sup>1</sup>, Daniela Kiski<sup>2</sup>, Armin Darius Peivandi<sup>1</sup>

<sup>1</sup>University Hospital Muenster, Department of Cardiothoracic Surgery, Muenster, Germany; <sup>2</sup>University Hospital Muenster, Department of Pediatric Cardiology, Muenster, Germany

**Background and Aim:** In curricular student teaching, transfer of knowledge about congenital malformations is highly challenging. This is mainly due to heterogeneity and complexity. Industrially made models are only available for common defects. The aim of the described teaching concept is to evaluate potential additional benefits of 3-D printed models.

**Method:** After pre testing knowledge (10 min) on 3 common heart defects (ventricular septum defect, fallot tetralogy and coarctation of the aorta), oral presentations on these defects are delivered by experts of the field. Then, participants are randomly assigned in 2 groups. One group studies heart defects with standard 2D teaching material. The other group (intervention group) is provided with additional 3D-printed models. Finally, transfer of knowledge is assessed through a second test. Study design was developed in cooperation with the Institute of Biostatistics and Clinical Research of the University Hospital Muenster.

**Results:** Through a time span of 3 half-terms, a total number of 288 students from 2 separate medical schools will be included in the study. The goal is not only to evaluate a gain of knowledge, but also to assess the students' subjective evaluation of this alternative teaching method.

**Conclusions:** We hypothesize, that a haptic experience enhances knowledge on complex congenital heart defects and encourages students to deepen their interests in the field.

**Keywords:** 3D-Printing, student teaching, congenital malformations

**P-274****Leveraging technology for the assessment and monitoring of fragile infants with complex congenital heart disease in the community**

Kerry Louise Gaskin<sup>1</sup>, Chris Bowers<sup>1</sup>, Jo Wray<sup>2</sup>, Suzie Hutchinson<sup>3</sup>  
<sup>1</sup>University of Worcester, Worcester, United Kingdom; <sup>2</sup>Great Ormond Street Hospital for Children NHS Foundation Trust, London, United Kingdom; <sup>3</sup>Little Hearts Matter, United Kingdom

**Background and Aim:** Babies with complex heart conditions have their first heart surgery in the first few days of life and require several further stages of surgery. They remain fragile after surgery and in the early weeks after discharge home. The paper based Congenital Heart Assessment Tool (CHAT2) was developed to help parents to monitor their baby's condition at home using a traffic light system (red, amber, green) to indicate the need for involvement of healthcare professionals, based on the daily monitoring of their baby's condition and behaviour. Evidence gained from parents' forums and studies about CHAT2 have highlighted parental and clinicians' fears about taking sick infants' home from hospital and venturing out into normal life. Additionally, barriers to the success of CHAT2 include the anxieties of parents in managing frequent monitoring and reliable transmission of information.

The project will develop a prototype mobile application for smartphones or tablets, that helps these parents to make life saving decisions about their baby wherever they are.

**Method:** This is a six-month quality improvement project, funded by NIHR Invention 4 Innovation FAST

Phase 1 – Focus groups (Sept–Oct 2022) to ascertain parents and healthcare professionals' using CHAT2 as a guide, to identify essential functional (e.g. CHAT21 parameters) and non-functional (e.g. plain English) requirements to ensure the application meets the everyone's needs.

Phase 2 – Prototype Design and Development (Oct–Jan 2023). The mobile application is being specified, designed, and developed based on the gather requirements, ensuring relevant guidelines on

accessibility and usability were met, taking advantage of the commercial human-computer interaction research, mobile application development, and distributed system experience of the team.

Phase 3 – User study (Feb 2023) to test the prototype mobile application with parents and healthcare professionals to ensure that it meets their needs

**Results:** The focus groups have identified key requirements of parents and healthcare professionals, and these are currently being used to develop the mobile application

**Conclusions:** Results from the user testing being conducted in Feb 2023 will be presented along with key conclusions emerging about the prototype application

**Keywords:** parents, education, discharge, infants, congenital heart disease, monitoring

### P-275

#### Evaluation of a national virtual heart camp using novel educational strategies for children with a fontan circulation

Carolina Andrea Escudero<sup>1</sup>, Alanna Livia Ash<sup>2</sup>, Lucy Harris<sup>3</sup>, Lily Lin<sup>1</sup>, Danielle Harake<sup>4</sup>, Edythe Tham<sup>1</sup>

<sup>1</sup>Department of Pediatrics, Division of Cardiology, University of Alberta and Stollery Children's Hospital, Edmonton, Canada; <sup>2</sup>Stollery Children's Hospital, Edmonton, Canada; <sup>3</sup>University of Alberta, Edmonton, Canada; <sup>4</sup>The Permanente Medical Group, California, USA

**Background and Aim:** Children with a Fontan circulation have unique physiology with lifelong implications on their medical and physical well-being. Many children with a Fontan circulation may not know peers with a similar heart disease or understand their heart disease.

**Method:** We organized a national Canadian virtual camp for children with a Fontan circulation ("Fontan camp"), which expanded from previous Fontan camps hosted for smaller regions within Canada. Recreational activities were organized for the children and they were able to meet and talk to peers living with a Fontan circulation. Educational activities were incorporated into the camp including painting of a 3D model of a heart with a Fontan procedure, online quizzes which included questions pertaining to the heart, discussions with adults living with a Fontan circulation, and participants' questions being answered by cardiologists. The 3D heart models were painted in blue and red acrylic paint to indicate the path of deoxygenated (blue) vs oxygenated (red) blood within the Fontan circulation (Figure 1). Feedback was elicited using an anonymized online survey for children and their caregivers to assess the satisfaction and self-perceived learning experience from the camp. A 5-point Likert scale was used to assess satisfaction.

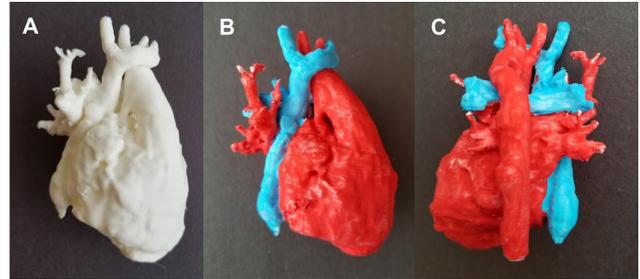
**Results:** Forty-five children (aged 8-15 years) attended the camp from 8/10 Canadian provinces with 57% attending their first Fontan camp. Twenty-one (47%) of children and families completed the survey. The children reported enjoying the 3D heart painting, online quizzes, discussion with adult Fontan survivors, and cardiologist answers to questions in 100%, 67%, 81%, and 57% respectively ( $p = 0.002$ ). Seventy-six percent of children reported learning something new about their heart during the camp and 80% learned something new from the 3D heart painting. Ninety-five percent of children reported feeling more connected to other similar children and 100% reported that they would attend a future Fontan camp.

**Conclusions:** Implementation of a national virtual Fontan camp is feasible with high rates of satisfaction from the attendees. The majority of children with a Fontan circulation reported learning something new about their heart and enjoying novel educational

strategies including speaking with adult Fontan survivors and painting of a 3D model of a heart similar to theirs.

**Keywords:** Fontan, virtual, education, 3D heart models

#### 3D Heart Model Painting



Depiction of the 3D heart models of a heart with a Fontan circulation (A) that were painted by participants to indicate the flow of deoxygenated (blue) and oxygenated (red) blood in the Fontan circulation (B - anterior view, C- posterior view)

### P-276

#### Aorto-pulmonary window in a preterm neonate: A case for pulse oximetry screening on the neonatal unit?

Bryony Dunning Davies, Wei Ling Amelia Lee, Ramona Onita  
Paediatric and Neonatal Department, Colchester General Hospital, Colchester, United Kingdom

**Background and Aim:** The most recent consultation from the UK National Screening Committee in 2019 concluded that further research was needed to provide sufficient evidence to recommend the addition of pulse oximetry (PO) to the national newborn screening programme. Despite this, a national survey conducted in 2020 reported that 51% of neonatal units in the UK were already using routine pulse oximetry to screen for critical congenital heart defects (cCHD).

We present a case of twin babies delivered by elective Caesarean section at a gestational age of 34+1 due to concerns regarding growth restrictions in twin 2. This baby had an initial normal systemic examination during newborn screening and there was no suspicion of congenital heart disease. At 10 hrs of age, a routine pre- and post-ductal saturation check revealed preductal saturation of 99% and post-ductal saturation in high 80s. Bedside echocardiography done by a Paediatrician with expertise in Cardiology (PEC) revealed significant aorto-pulmonary window, large PDA, hypoplastic aortic arch and interrupted aortic arch type A. Baby was commenced on low dose Prostaglandin infusion and baby had an uneventful transfer to a tertiary cardiac unit without requirements for significant clinical interventions prior.

**Method:** We carried out a literature search for PO monitoring as a routine screening tool in newborns. Our particular focus was on looking at the usage of both pre- and post-ductal saturation screening in babies admitted to neonatal/ neonatal intensive care units (NNU/NICU).

**Results:** There is an established amount of literature, most of which is supportive of the use of PO as a routine screening tool to detect cCHD. Most studies however have excluded babies admitted to NNU/NICU and focused on term/near term babies on the post-natal wards. This is due to additional considerations that will need

factoring in, including oxygen supplementation, extreme prematurity and potentially higher false positive rates due to the aforementioned.

**Conclusions:** Our case study highlights the importance of developing PO screening in NNU/NICU babies to detect cCHD which is a relatively new area of research.

**Keywords:** neonatal, congenital heart disease, oxygen saturation monitoring

#### P-278

##### **Fetal vessel anomalies- a rare case of a persistent fifth ARCH, total anomalous pulmonary venous return, persistent left superior vena cava and coarct**

*Elisabeth Seidl Mlczech, Sabine Greil, Sulaima Albinni, Erwin Kitzmueller, Ina Michel Behnke*

*Pediatric Heart Venter Vienna, Department of Pediatrics and Adolescent Medicine, Division of Pediatric Cardiology, Medical University of Vienna, Austria*

**Background and Aim:** A persistent fifth aortic arch is a rare congenital anomaly that can be diagnosed in the fetus or postnatally. It originates from the distal ascending aorta and is distally connected with the descending aorta. It can present as an incidental finding or in association with complex congenital heart anomalies involving other vessels. Symptoms may be obscure and diagnosis eventually delayed.

**Method:** We report a rare case of a complex vessel anomaly diagnosed in the fetus.

**Results:** A 34 year old pregnant woman was referred for suspicion of coarctation at 23+0. A detailed fetal echocardiogram revealed ventricular asymmetry, coarctation and a membrane in the left atrium. A cesarean section was performed at GA 38+0 with a birth weight of 2520gramm. Postnatal echocardiography showed intracardiac total anomalous pulmonary venous return, persistent left superior vena cava to coronary sinus, a persistent fifth arch and coarctation. To determine the diagnosis a cardiac catheter was performed on the first day of life followed by a surgical corrective procedure on day 2.

**Conclusions:** Persistent fifth arch is a rare congenital anomaly. This the first case in the literature presenting with intracardiac total anomalous pulmonary venous return, as well as coarctation and persistent left superior vena cava to coronary sinus which was successfully repaired.

**Keywords:** persistent fifth arch, TAPVD, CoA

#### P-279

##### **Fetal persistent left superior vena cava- postnatal clinical implication?**

*Elisabeth Seidl Mlczech<sup>1</sup>, Sulaima Albinni<sup>1</sup>, Doris Luckner<sup>1</sup>, Christiane Pees<sup>1</sup>, Barbara Ulm<sup>2</sup>, Ina Michel Behnke<sup>1</sup>*

*<sup>1</sup>Pediatric Heart Center Vienna, Division of Pediatric Cardiology, Department of Pediatrics and Adolescent Medicine, Medical University of Vienna, Vienna, Austria; <sup>2</sup>Division of Obstetrics and Fetomaternal Medicine, Department of Obstetrics and Gynaecology, Medical University of Vienna, Vienna, Austria*

**Background and Aim:** Persistent left superior vena cava (PLSCV) has become a more frequent prenatal diagnosis due to introduction of the 3-vessel view (3VV) and 3-vessel and trachea view (3VT). It is the most common variant of the thoracic venous system. It usually drains via the coronary sinus to the right atrium. In this study, we aimed to identify prenatally diagnosed cases of PLSVC in our

clinic, to evaluate the associated cardiac, extracardiac, and chromosomal anomalies, and to review their outcomes

**Method:** This is a retrospective case series study of fetuses diagnosed with situs solitus and persistent left superior vena cava (PLSVC) at the Pediatric Heart Center Vienna, from 2015-2021. Prenatal and postnatal outcome data were collected. Prenatal diagnosis was based on abnormal presentation of the 3-vessel view (3VV) and/or 3-vessel and trachea view (3VT) showing an additional vessel to the left of the pulmonary artery or absence of the usual vessel to the right of the ascending aorta, the right superior vena cava (RSVC) and presence of a LSVC draining to the coronary sinus. **Results:** During the study period 47 fetuses were identified. All of them were liveborn. Isolated PLSVC was present in n = 37, single left superior vena cava n = 1, associated with coarctation n = 2, associated with other complex cardiac disease in n = 7. Genetic anomalies were found in 2 fetuses (T21, Di George Syndrom). **Conclusions:** Prenatal detection of a persistent left superior vena cava should be followed by a detailed fetal echocardiography to rule out associated anomalies. Isolated findings of a persistent left superior vena cava or single left superior vena cava are benign and have an excellent outcome.

**Keywords:** LSVc, CoA, Fetal, Outcome

#### P-280

##### **First trimester fetal arrhythmias**

*Agata Michalczyk<sup>1</sup>, Bartosz Czuba<sup>2</sup>, Stanisław Włoch<sup>3</sup>, Wojciech Cnota<sup>2</sup>, Agata Włoch<sup>2</sup>*

*<sup>1</sup>Department of Congenital Heart Defects and Pediatric Cardiology, Silesian Centre for Heart Diseases in Zabrze, Medical University of Silesia; <sup>2</sup>Department of Obstetrics and Gynecology in Ruda Śląska, Medical University of Silesia; <sup>3</sup>Department of Obstetrics and Gynecology in Katowice, Medical University of Silesia*

**Background and Aim:** Fetal arrhythmias diagnosed during the first trimester scan are not well described in the literature. They occur much more rarely than in second or third trimester. Many reports are available on FHR as a first trimester marker of chromosomal abnormalities while none has focused on the arrhythmias as an issue.

**Method:** Fifty fetuses with bradycardia or tachycardia diagnosed at the first trimester scan examined in the reference center were analyzed retrospectively. The chromosomally abnormal fetuses were excluded from the study. The follow-up is complete in examined group.

Fetal bradycardia was recognized when the fetal heart rhythm (FHR) was diagnosed below the 10th and tachycardia above the 90th percentile for gestational age. Atrial and ventricular rhythm as well as AV and VA times were measured and analyzed separately.

**Results:** Bradycardia was found in twenty fetuses (20/50-40 %). Fourteen (14/20-70%) presented the regular atrial and ventricular rhythm, with AV conduction 1:1 and normal AV time. Four fetuses presented the congenital heart block with coexisted structural heart defect (4/20-20%), one presented the atrial rhythm with shortened AV time, in the next case the blocked atrial premature contractions were recognized. In two cases LQTs was recognized after birth.

Among the fetuses with tachycardia (30/50-60 %), one was diagnosed as SVT with FHR 300 bpm with spontaneous return to the sinus rhythm. Twenty nine fetuses (29/30-96 %) presented tachycardia with regular atrial and ventricular rhythm and 18/29 of them had normal FHR later in pregnancy with normal AV time whereas in 11/29 fetuses (37%) tachycardia was observed as well as during the 2nd or 2nd and 3rd trimester. Two of them presented

SVT later in time (one in the 3rd trimester one in the neonatal Period).

**Conclusions:** Fetal cardiac rhythm abnormality detected in the first trimester, requires identification of its cause if possible, establishing its mechanism and hemodynamic consequences. Most of arrhythmias are transient and don't affect fetuses later in pregnancy. In studied group there is a higher risk of cardiac disorders, therefore the fetal echocardiography examinations and fetal monitoring is strongly recommended

**Keywords:** Fetal, arrhythmia, examination, pregnancy, abnormality, cardiac

## P-281

### Epicardial fat thickness in infants of diabetic mothers

Ma Martha Vela Huerta<sup>1</sup>, Norma Amador Licona<sup>2</sup>, Rigoberto Dominguez Damian<sup>1</sup>, Antonio Heredia Ruiz<sup>1</sup>, Helena Victoria Orozco Villagomez<sup>1</sup>, Juan Manuel Guizar Mendoza<sup>2</sup>, Salvador Alarcon Santos<sup>1</sup>  
<sup>1</sup>Department of Neonatology, Hospital General, Leon, Secretaria de Salud de Guanajuato, Mexico; <sup>2</sup>Direccion de Investigacion, Universidad De La Salle Bajio, Leon, Mexico

**Background and Aim:** In adults and children epicardial fat thickness (EFT) has been related to metabolic syndrome and other cardiovascular risks factors. This study compares the epicardial fat thickness (EFT) between infants of diabetic mother (IDM) and infants of non-diabetic mother (INDM) and its association with other echocardiographic parameters.

**Method:** A cross sectional study was performed in 64 IDM and 29 INDM (93 in total). In both groups an echocardiogram was performed within the first 24h of extrauterine life, for the purpose of measuring the EFT dimensions

**Results:** The EFT (3.6 vs. 2.5 mm,  $p < 0.0001$ ), the interventricular septum thickness (IVST) (6.2 vs. 5.2 mm,  $p < 0.0001$ ) and the IVST / left ventricle posterior wall (LVPW) (1.3 vs. 1.1,  $p = 0.001$ ) were higher in the IDM; while the left ventricular ejection fraction [LVFE] (71.1 vs. 77.8;  $p < 0.0001$ ) was lower than in the INDM, respectively. There was a positive correlation between EFT with (IVST) ( $r = 0.577$ ;  $p = 0.0001$ ), left ventricle posterior wall (LVPW) ( $r = 0.262$ ;  $p = 0.011$ ), IVST/LVPW index ( $r = 0.353$ ;  $p = 0.001$ ), and a negative correlation with LVFE was observed ( $r = -0.376$ ;  $p = 0.0001$ ).

**Conclusions:** IDM presented higher EFT than INDM. This was positively related with echocardiographic parameters of left ventricular thickness and negatively related with left ventricular ejection function.

**Keywords:** Epicardial fat, infants of diabetic mothers, left ventricle

Table 4. Correlation between epicardial fat thickness with other echocardiographic variables in the entire group.

	IVST	LVPW	IVST/LVPW	LVFE	Mitral Integral Velocity	Early
EFT	0.577	0.262	0.353	-0.376	0.313	
	$p=0.0001$	$p=0.011$	$p=0.001$	$p=0.0001$	$p=0.002$	

There was a positive correlation between EFT with (IVST), left ventricle posterior wall (LVPW), IVST/LVPW index, and a negative correlation with LVFE.

## P-282

### Prenatal detection rate and postnatal incidence of major congenital heart defects in an unselected national population from 1991 to 2018

Viktor Tomek<sup>1</sup>, Hana Jičínská<sup>2</sup>, Jan Pavlíček<sup>3</sup>, Jan Kovanda<sup>1</sup>, Petr Jehlička<sup>4</sup>, Eva Klásková<sup>5</sup>, Jan Janoušek<sup>1</sup>, Jan Marek<sup>6</sup>

<sup>1</sup>Children's Heart Centre, 2nd Faculty of Medicine, Charles University in Prague and Motol University Hospital, Czech Republic; <sup>2</sup>Department of Pediatric Cardiology, University Hospital, Brno and Faculty of Medicine, Masaryk University, Czech Republic; <sup>3</sup>Department of Pediatrics and Prenatal Cardiology, University Hospital Ostrava, Ostrava, Czech Republic; <sup>4</sup>Department of Pediatrics, University Hospital in Pilsen, Charles University Prague; <sup>5</sup>Department of Pediatrics, University Hospital Olomouc and Faculty of Medicine and Dentistry, Palacky University Olomouc, Czech Republic; <sup>6</sup>Great Ormond Street Hospital for Children and Institute of Cardiovascular Sciences UCL, London, UK

**Background and Aim:** We aimed to determine the pre- and postnatal incidence of major congenital heart disease (CHD) in a setting of centralized health care with a single tertiary institution and the impact of cardiac screening on postnatal outcome.

**Method:** 3215 fetuses diagnosed with major CHD in the Czech Republic (population 10.7 million) between 1991 and 2018 were prospectively evaluated with known outcomes and associated comorbidities. Pre- and postnatal incidence of CHD in non-selected population was assessed by comparison with a retrospective analysis of all children born alive with major CHD in the same period (N = 5025) using national data registry.

**Results:** Major CHD was identified in 3215 fetuses, 1323 (41.2%) were born, 1798 (55.9%) terminated (TOP) and 94 (2.9%) died in utero. Detection rate increased from 6.2% to 79.9% ( $p < 0.0001$ ). TOP decreased from 70% to 43% ( $P < 0.001$ ). Out of 468 fetuses diagnosed in the first trimester (introduced in 2007), 342 were terminated (73.1%). From 1842 fetuses diagnosed in the second trimester since 2007, 799 (43.4%) fetuses were terminated yielding an odds ratio of decision to TOP of 3.5 (95% CI 2.8–4.5,  $p < 0.001$ ) in favor of first trimester screening. Mean postnatal incidence of major CHD was 1.69 per 1000 live born declining from 2.1% to 1.4% ( $p < 0.0001$ ), total incidence of was 0.23% without change during the study period.

Number of TOP with univentricular heart morphology remained unchanged (0.10 cases per 1year,  $p = 0.63$ ) and number of TOP with non-cardiac anomalies increased during the study period (0.51 cases per 1year,  $p = 0.006$ ). TOP of isolated heart defects allowing for biventricular repair dropped from 61% in 1992 to 1–3% ( $p < 0.001$ ) in current era.

**Conclusions:** Since 1991, overall incidence of major CHD did not change significantly. A nationwide centralized prenatal detection of CHD is leading to increasing detection rates from 6% to 80% of all major CHD. TOP of major CHD has decreased from 70% to 43% but remains high in fetuses with univentricular heart and those with associated comorbidities. The introduction of the first trimester screening, despite its high termination rate, did not increase the total number of terminated pregnancies.

**Keywords:** congenital heart defect, echocardiography, prenatal diagnosis, termination of pregnancies, epidemiology

## P-283

**Trisomy 18 and congenital heart defects: dealing with an ethical dilemma**

Marta Gamba Arzo<sup>1</sup>, Lucía Deiros Bronte<sup>1</sup>, Cristina Arribas Sánchez<sup>2</sup>, Ángela Uceda Galiano<sup>1</sup>, Viviana Arreo Del Val<sup>1</sup>, Roberto Rodríguez<sup>3</sup>, Luis García Guereta Silva<sup>1</sup>, Felipe Garrido Martínez De Salazar<sup>2</sup>, Carlos Labradero De Lera<sup>1</sup>, Laura Muñoz Saa<sup>4</sup>, Federico Gutiérrez Larraya Aguado<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology, Hospital Universitario La Paz, Madrid, Spain; <sup>2</sup>Department of Pediatrics, Clínica Universidad de Navarra, Madrid, Spain; <sup>3</sup>Department of Obstetrics, Hospital Universitario La Paz, Madrid, Spain; <sup>4</sup>Department of Obstetrics, Clínica Universidad de Navarra, Madrid, Spain

**Background and Aim:** Trisomy 18 is characterized by variable but severe clinical manifestations, with involvement of multiple organs (congenital heart defects in 80%), mental retardation and low survival rates. Some small sample studies support that heart surgery can improve life expectancy, allow the hospital discharge and improve the quality of life of patients and their families. However, there are no specific guidelines for the management of these patients. The aim is to review clinical-management and survival rates in our patients diagnosed of trisomy 18 during antenatal follow up.

**Method:** We conducted a retrospective observational study over a 6-year period, from January 2016 to May 2022; involving fetuses diagnosed of trisomy 18 in two hospitals (Hospital Universitario La Paz-Madrid, and Clínica Universidad de Navarra-Madrid).

**Results:** Ten pregnancies with presumed trisomy 18 were enrolled. Nine showed a congenital heart defect, and seven other organs malformations. Ventricular septal defect was observed in seven patients, atrioventricular septal defect in one, and hypoplastic left heart syndrome in another. In one of the pregnancies parents decided the termination. In the remaining nine, a planned delivery was settled, considering the prognosis of the baby, the internal organ malformations in each fetus, and parental expectations. Three fetuses did not survive to the end of the gestation. Those who were born alive were supported following a planned delivery shared with parents (compassionate care in five, and noninvasive ventilation support in one). Three patients died in the first 48 hours of life; two patients were discharged and supported by regional palliative care services. Survival rates of 17 and 21 days were observed. Surgical closure of ventricular septal defect was performed in one patient without no other internal organ malformation and is still alive (45 months).

**Conclusions:** Fetuses and babies suffering from trisomy 18 have a poor prognosis with low reported survival rates. There are no specific guidelines for the management of these patients. Small sample case reports, state that heart surgery can improve life expectancy. Management of these pregnancies continues to be a clinical challenge, and we think a multidisciplinary approach, with active involvement of parents in decisions, is a good practice approach.

**Keywords:** Trisomy 18, congenital heart defects, survival rates

## P-284

**Right aortic ARCH: prenatal diagnosis, associated anomalies and postnatal management.**

Alejandro Domarco Manrique<sup>1</sup>, José Ignacio Camuña Correa<sup>1</sup>, Lucía Sanjuán Benita<sup>1</sup>, Patricia Alonso Rivero<sup>1</sup>, Constanco Medrano López<sup>1</sup>, Teresa Álvarez Martín<sup>1</sup>, Ana Pita Fernández<sup>2</sup>, Juan De León Luis<sup>3</sup>, María Antonia García Casillas<sup>4</sup>

<sup>1</sup>Department of Pediatric Cardiology, Hospital General Universitario Gregorio Marañón, Madrid, Spain; <sup>2</sup>Department of Cardiovascular Surgery, Hospital General Universitario Gregorio Marañón, Madrid,

Spain; <sup>3</sup>Department of Gynecology and Obstetrics, Hospital General Universitario Gregorio Marañón, Madrid, Spain; <sup>4</sup>Department Pediatric Surgery, Hospital General Universitario Gregorio Marañón, Madrid, Spain

**Background and Aim:** The right aortic arch is characterized by an abnormal laterality of the aorta and supra-aortic trunks, located to the right of the trachea. It may form a vascular ring, compressing the airway and the digestive tract. The improvement of prenatal diagnosis in recent years has made it possible to establish an early follow-up and management plan.

Our aim is to describe our experience in patients with right aortic arch with left ductus, and the associated anomalies, the evolution and the diagnostic-therapeutic procedures.

**Method:** Retrospective case series including 70 patients diagnosed in utero of right aortic arch with left ductus throughout ten years by pediatric cardiologists specialized in Fetal Cardiology. The data was obtained from the clinical history.

**Results:** The mean gestational age at diagnosis was 21 weeks, identifying right arch type I in 35/70 and type II in 25/70 (with aberrant left subclavian artery). The most frequently associated prenatal echocardiographic abnormalities were: ventricular septal defect, double vena cava system, hyperechoic foci in the left ventricle, and ductal stenosis; and postnatal: patent foramen ovale, ductus arteriosus and ostium secundum atrial septal defect. 1/71 had major extracardiac malformations prenatally (megacisterna magna), postnatally 9/71 (anorectal and craniofacial anomalies); and minor extra cardiac malformations were diagnosed prenatally in 5/71, and postnatally in 17/71 (mainly related to the genitourinary tract and skeletal anomalies). Prenatal genetic study was performed in 48/70, detecting trisomy 21 in 3/70 and 22q11 microdeletion in 1/70.

Symptoms of vascular ring were reported in 10/70. Computerized tomography was performed in 28/70 patients, showing airway and/or digestive tract compression in 24/28 (imprint or mild stenosis in 21/24, moderated stenosis in 3/24), bronchoscopy was performed in 7/70 patients and esophagogram in 4/70, with none to mild stenosis in all of them.

Surgical correction was performed in three patients, with a mean age at the time of surgery of 43 months. All of them had moderate stenosis on imaging techniques.

**Conclusions:** Right aortic arch with left ductus is frequent, it is usually isolated and asymptomatic, although follow-up and evaluation of complementary tests and surgery must be carried out in patients with airway compression.

**Keywords:** fetal, aortic, arch, right, sling, vascular

## P-285

**Impact of fetal growth restriction on perinatal cardiac adaptation and cardiovascular health outcomes in offspring**

Conrado Milani Coutinho<sup>1</sup>, Veronica Giorgione<sup>2</sup>, Basky Thilaganathan<sup>2</sup>, Olga Patey<sup>3</sup>

<sup>1</sup>Fetal Medicine Unit, St George's University Hospitals NHS Foundation Trust, University of London, London, UK. <sup>2</sup>Department of Gynaecology and Obstetrics, Hospital das Clínicas, Ribeirão Preto Medical School, University of São Paulo, Ribeirão Preto, São Paulo, Brazil; <sup>3</sup>Fetal Medicine Unit, St George's University Hospitals NHS Foundation Trust, University of London, London, UK. <sup>4</sup>Vascular Biology Research Center, Molecular and Clinical Sciences Research Institute, St George's University of London, London, UK; <sup>5</sup>Fetal Medicine Unit, St George's University Hospitals NHS Foundation Trust, University of London, London, UK. Royal Brompton and Harefield Hospitals, Guy's & St Thomas' NHS Foundation Trust, London, UK

**Background and Aim:** Fetal growth restriction (FGR) associated with chronic hypoxaemia from uteroplacental insufficiency results in fetal cardiac remodeling and myocardial dysfunction. These adverse developmental conditions could persist leading to susceptibility to cardiovascular disease in offspring. Our study aimed to investigate fetal perinatal cardiac adaptation and cardiovascular outcomes in offspring of FGR pregnancies.

**Method:** Prospective follow up study of 37 normal control and 35 FGR pregnancies assessed as fetuses (mean age 37±1 weeks) and again in infancy (mean age 8±2 months). Conventional echocardiography techniques, tissue Doppler imaging and speckle tracking echocardiography were applied for assessment of left (LV) and right ventricular (RV) geometry and function. Echocardiographic parameters were normalised by ventricular size adjusting for differences in body weight between groups.

**Results:** Compared to controls, FGR term fetuses showed significant alterations in cardiac geometry with more globular LV chamber (LV sphericity index, 0.56 vs 0.52), increase in biventricular global longitudinal systolic contractility (MAPSE, 0.29 vs 0.25mm; SAPSE, 0.27 vs 0.22mm; TAPSE, 0.42 vs 0.37mm) and elevated biventricular cardiac output (combined CO: 592 vs 497ml/min/kg,  $p<0.01$  for all). Whereas LV diastolic function in FGR fetuses was significantly impaired: decreased myocardial diastolic velocities (LV A', 0.30 vs 0.26cm/s; IVS E', 0.19 vs 0.16cm/s; IVS A', 0.25 vs 0.22cm/s) and reduced LV torsion (1.2 vs 3.5deg/cm,  $p<0.01$  for all). Relative to controls, FGR offspring revealed persistently increased SAPSE (0.27 vs 0.24mm), LV longitudinal strain (-19.0 vs -16.0%), reduced LV torsion (1.6 vs 2.1deg/cm) and elevated CO (791 vs 574ml/min/kg,  $p<0.01$  for all).

**Conclusions:** Perinatal cardiac remodeling and myocardial dysfunction in term FGR fetuses is most likely due to chronic placental hypoxaemia. Persistent changes in cardiac geometry and function in FGR offspring may reflect cardiovascular maladaptation that could predispose to long-term cardiovascular complications.

**Keywords:** echocardiography, fetal cardiac adaptation, fetal growth restriction, hypoxaemia, perinatal cardiac remodeling

#### P-286

##### **Impact of maternal gestational diabetes on fetal perinatal cardiac adaptation and cardiovascular health outcomes in offspring**

Conrado Milani Coutinho<sup>1</sup>, Veronica Giorgione<sup>2</sup>, Basky Thilaganathan<sup>2</sup>, Olga Patey<sup>3</sup>

<sup>1</sup>Fetal Medicine Unit, St George's University Hospitals NHS Foundation Trust, University of London, London, UK. Department of Gynaecology and Obstetrics, Hospital das Clínicas, Ribeirão Preto Medical School, University of São Paulo, Ribeirão Preto, São Paulo, Brazil; <sup>2</sup>Fetal Medicine Unit, St George's University Hospitals NHS Foundation Trust, University of London, London, UK. Vascular Biology Research Center, Molecular and Clinical Sciences Research Institute, St George's University of London, London, UK; <sup>3</sup>Fetal Medicine Unit, St George's University Hospitals NHS Foundation Trust, University of London, London, UK. Royal Brompton and Harefield Hospitals, Guy's & St Thomas' NHS Foundation Trust, London, UK

**Background and Aim:** Maternal gestational diabetes mellitus (GDM) is associated with fetal hypersulinemia, increased metabolic rate, erythropoietin level and hypoxaemia. GDM increases fetal cardiac remodeling and myocardial dysfunction that could potentially predispose to cardiovascular disease in offspring. Our study aimed to investigate fetal perinatal cardiac adaptation and cardiovascular outcomes in offspring of GDM pregnancies.

**Method:** Prospective follow up study of 37 normal control and 24 GDM pregnancies assessed as fetuses (mean age 37±1 weeks) and again in infancy (mean age 8±2 months). Conventional echocardiography techniques, tissue Doppler imaging and speckle tracking echocardiography were applied for assessment of left (LV) and right ventricular (RV) geometry and function. Echocardiographic indices were normalised by ventricular size adjusting for differences in body weight between groups.

**Results:** Compared to controls, term fetuses from GDM pregnancies demonstrated significant alterations in cardiac geometry with more globular RV chamber (RV sphericity index, 0.66 vs 0.60), thickened LV (4.0 vs 3.07mm) and RV (4.1 vs 3.1mm) walls and IVS (3.9 vs 3.1mm). GDM fetuses also had increased biventricular global longitudinal systolic contractility (MAPSE, 0.29 vs 0.26mm; TAPSE, 0.46 vs 0.37mm), impaired diastolic function (IVS E', 0.20 vs 0.16cm/s; LV torsion, 5.89 vs 3.45deg/cm) and elevated biventricular cardiac output (LV CO, 247 vs 219ml/min/kg; RV CO, 330 vs 264ml/min/kg;  $p<0.01$  for all). Although geometrical changes normalised in maternal GDM offspring, there were persistently increased MAPSE (0.30 vs 0.27mm), SAPSE (0.29 vs 0.24mm), IVS late diastolic velocity A' (0.26 vs 0.21cm/s) and altered myocardial deformation (basal radial strain, 29 vs 19%; apical radial strain, 31 vs 39%).

**Conclusions:** Perinatal cardiac remodeling and myocardial dysfunction in term fetuses of GDM pregnancies most likely reflect a response to the adverse hyperglycemic intrauterine environment. Evidence of persistent alterations in ventricular geometry and function could indicate a predisposition to cardiovascular morbidity in the offspring of diabetic mothers in future life.

**Keywords:** echocardiography, fetal cardiac adaptation, diabetes mellitus, hyperglycaemia, hypoxaemia, perinatal cardiac remodeling

#### P-287

##### **Maternal hypertensive disorders of pregnancy and electrocardiographic findings in newborns – a copenhagen baby heart study**

Emil Hoeygaard Noerskov<sup>1</sup>, Maria Paerregaard<sup>1</sup>, Anna Axelsson Raja<sup>2</sup>, Anne Sophie Sillesen<sup>1</sup>, Alex H Christensen<sup>1</sup>, Heather Boyd<sup>3</sup>, Henning Bundgaard<sup>2</sup>, Kasper Iversen<sup>1</sup>, Ottilia Voegg<sup>1</sup>

<sup>1</sup>Department of Cardiology, Copenhagen University Hospital Herlev, Copenhagen, Denmark; <sup>2</sup>Department of Cardiology, Copenhagen University Hospital Rigshospitalet, Copenhagen, Denmark; <sup>3</sup>Department of Epidemiology Research, Statens Serum Institut, Copenhagen, Denmark

**Background and Aim:** Hypertensive disorders of pregnancy (HDP) - preeclampsia in particular - are important causes of pregnancy complications and death for both mother and child. Preeclampsia is also associated with both congenital heart defects and more subtle cardiac structural and functional changes in the offspring. However, whether HDP also affects the newborn's cardiac conduction system is un-known. We will assess whether the electrocardiograms (ECGs) of infants born after pregnancies complicated by HDPs differ from those of infants born after uncomplicated pregnancies.

**Method:** The Copenhagen Baby Heart Study (CBHS) is a population-based cohort study of newborn and childhood cardiac function that performed echocardiography and ECG on more than 25,000 newborns in the period April 2016–October 2018. The present study population included 17,480 newborn who had a validated ECG performed within 30 days of birth, and who did not have congenital heart disease at the CBHS examination. Using a linear regression analysis, we will compare mean ECG

parameter values for offspring exposed to maternal HDPs (761 mothers with preeclampsia, and 344 with gestational hypertension) and unexposed offspring. The following ECG variables will be investigated: heart rate, PR interval, QRS axis, QRS duration, uncorrected/corrected QT intervals, Q peak amplitudes, and maximum R- and S-wave amplitudes in V1 and V6, respectively. Analyses will be adjusted for the child's sex, gestational age, length and weight at birth, and age at ECG examination.

**Results:** Preliminary results will be presented at AEPC 2023.

**Conclusions:** The described project will fill gaps in our knowledge regarding whether maternal HDPs are associated with alterations in the newborn ECG.

**Keywords:** hypertensive disorders of pregnancy, preeclampsia, electrocardiography, newborn, risk factors

### P-290

#### Impact of early detection and steroid treatment on fetal ventricular heart rate and pacemaker implantation in congenital heart block

Hakan Eliasson<sup>1</sup>, Sven Erik Sonesson<sup>3</sup>, Aurelie Ambrosi<sup>4</sup>, Marie Wahren Herlenius<sup>2</sup>

<sup>1</sup>Department of Women's and Children's Health, Karolinska Institutet and Department of Pediatric Cardiology, Karolinska University Hospital, Stockholm, Sweden; <sup>2</sup>Department of Medicine, Karolinska Institutet, Stockholm, Sweden, Department of Clinical Science, University of Bergen, Bergen, Norway; <sup>3</sup>Department of Women's and Children's Health, Karolinska Institutet, Stockholm, Sweden; <sup>4</sup>Department of Medicine, Karolinska Institutet, Stockholm, Sweden

**Background and Aim:** Immune-mediated congenital heart block (CHB) is a rare life-threatening disease that affects fetuses exposed to maternal anti-Ro/SSA antibodies, with an inflammatory response leading to cardiac damage in its most severe cardiac manifestation. The effect of fetal treatment with fluorinated steroids on outcome is unclear.

Our aim was to study the effects of timing of detection and steroid treatment on ventricular heart rate (HR) and age at pacemaker (PM) implantation in fetal third-degree atrioventricular block (AVB III).

**Method:** Twenty-five of 31 fetuses diagnosed with autoimmune AVB II-III at our tertiary fetal cardiology centre from 2000 to 2020, with AVB III as a final outcome were reviewed. Fluorinated steroids were used to improve atrioventricular conduction and/or limit cardiac inflammatory damage

**Results:** Our surveillance program detected 7 cases approximately 5 weeks earlier than 18 cases referred for bradycardia. Fetuses detected before 24 weeks had a higher HR ( $63.3 \pm 6.9$  vs.  $57.2 \pm 6.9$  bpm,  $p = 0.042$ ) and a larger proportion with HR  $>60$  bpm (80 vs. 33%,  $p = 0.041$ ) than those detected at later gestation. Steroid treatment was commenced in 17/25; within 1 week after AVB III development in 7 and with an unknown delay in 10. Treated cases were diagnosed earlier, with a higher HR at diagnosis ( $61.7 \pm 7.1$  vs.  $54.7 \pm 6.3$  bpm,  $p = 0.026$ ), 1-2 weeks after diagnosis/treatment and before birth ( $65.4 \pm 12.4$  vs.  $54.9 \pm 5.7$  bpm,  $p = 0.030$ ), than the untreated. Overall, only 2/24 (8%) surviving babies were born preterm and neonatal PM treatment was used in 4 (17%). Age at PM implantation correlated significantly with HR before birth, and fetuses with HR  $>60$  bpm had a lower rate of PM implantations before 3 (10 vs. 57%,  $p = 0.033$ ) and 12 months of age (20 vs. 86%,  $p = 0.003$ ). A trend for a higher percentage of infants alive without PM at 3 months of age (71 vs. 38%) in those prenatally exposed to fluorinated steroids was observed

**Conclusions:** Early detection of AVB III and initiation of fluorinated steroid treatment may offer some benefits, such as maintaining a

higher ventricular HR to the end of gestation, as well as delayed PM implantation.

**Keywords:** Anti-Ro/SSA, congenital heart block, treatment, heart rate, pacemaker

### P-291

#### Outcome of fetuses with hemodynamically significant heart defects delivering in a non collocated maternity unit

Leila Rittey<sup>1</sup>, Andrew Salmon<sup>2</sup>, Michelle Carr<sup>1</sup>, Kumal Babla<sup>2</sup>, Pranav Pandya<sup>3</sup>, Robert Yates<sup>1</sup>, Victoria Jowett<sup>1</sup>

<sup>1</sup>Department of Paediatric and Fetal Cardiology, Great Ormond Street Hospital for Children, London, England; <sup>2</sup>Department of Neonatology, University College London Hospitals, London, England; <sup>3</sup>Department of Fetal Medicine, University College London Hospitals, London, England

**Background and Aim:** Although the optimal model for delivery of high-risk CHD fetuses is collocation of delivery and cardiac care, this is not universally available. We have a well-established clinical pathway for high-risk fetuses, in collaboration with our local tertiary neonatal unit. The aim of this study was to review early postnatal morbidity and mortality of neonates with hemodynamically-significant CHD to determine if delivery at a non-collocated maternity unit is associated with adverse outcome.

**Method:** Electronic records of fetal assessment and perinatal outcome of high risk fetuses were reviewed from 1/1/18-31/12/21. **Results:** 83 fetuses with complete data were included; 59 (71%) had TGA IVS, 10 (12%) CCHB and 3 (3.6%) HLHS with restrictive atrial septum. The delivery was as planned in 64 (77%). In 19 (23%), changes included emergency caesarian section in 10 (12%); 4 (4.8%)  $<37$  weeks gestation. Spontaneous delivery prior to planned occurred in 9 (10.8%).

Transfer to the cardiac centre occurred after 24 hours in 35 (42.2%); 15 (18.1%) 24-72 hours, 16 (19.3%) 3-6 days and 4 (4.8%)  $>7$  days. In all cases this was due to bed capacity at the cardiac centre. 3/83 (3.6%) required transfer to the cardiac centre for urgent assessment or management.

30/59 (50.8%) TGA patients required balloon atrial septostomy (BAS). The was undertaken at the non-collocated neonatal unit in 26/30 (86%) and the cardiac centre in 4/30 (13.3%). 20 patients (33.9%) had saturations  $<70\%$ ; 10 (16.9%) saturations  $<50\%$ . Delivery out of hours following induction of labour occurred in 2 of these (2.4%). Final fetal cardiology review was 30+3 to 36 +0 weeks gestation. Complication rate following BAS was 20% (6/30). All neonates had successful arterial switch procedure (71.2% within 8 days).

In the 10 CCHB patients, 4 (40%) had a heart rate  $<60$ bpm at final assessment (34-36 weeks). 1 had a neonatal death.

**Conclusions:** Most high risk fetuses with CHD can be safely delivered in a non-collocated tertiary delivery unit with established prenatal assessment pathways and coordinated perinatal care allowing prompt and adequate intervention where required. Further examination of patient pathways and comparison with collocated units are needed to optimise care for this high risk group.

**Keywords:** fetal cardiology, non collocated maternity, haemodynamically significant

### P-292

#### Prenatal constriction of arterial duct causing right heart dysfunction – a case report

Krzysztof Kocot<sup>1</sup>, Justyna Doniec<sup>2</sup>, Luiza Zalewska<sup>2</sup>, Lesław Szydłowski<sup>1</sup>, Iwona Marumiak Chudek<sup>3</sup>, Agata Włoch<sup>4</sup>

<sup>1</sup>Department of Pediatric Cardiology, Medical University of Silesia in Katowice, Katowice, Poland; <sup>2</sup>Department of Pediatric Cardiology, John

Paul II Upper Silesian Child Health Centre in Katowice, Katowice, Poland; <sup>3</sup>Department of Neonatology and Neonatal Intensive Care, Medical University of Silesia in Katowice, Katowice, Poland; <sup>4</sup>Department of Obstetrics and Gynecology, Medical University of Silesia in Katowice, Ruda Śląska, Poland

**Background and Aim:** Fetal constriction of the arterial duct (DA) in the absence of a cardiac defect is a rare cause of severe right heart dysfunction. It results in pulmonary hypertension (PH), right ventricle (RV) dilatation and tricuspid regurgitation (TR). The aim was to present a case of patient with DA constriction diagnosed prenatally.

**Method:** The patient was treated in a paediatric reference centre with neonatal intensive care unit. The data was collected retrospectively and prospectively with one year follow up.

**Results:** A woman in first pregnancy with no past medical history was referred to the fetal cardiologist in 32nd week of pregnancy. Fetal echocardiography showed dilated RV with signs of pressure overload, TR and signs of DA constriction. Low-polyphenol diet and referral to a reference centre for the birth was recommended. The baby was born by caesarean section after 37th week of gestation with birth weight of 3415g. Postnatal echocardiography showed enlarged right atrium, dilated hypokinetic RV, hyperechogenic tricuspid valve with moderate TR, constricted DA and signs of PH. Oxygen and prostaglandin E1 were successfully introduced. Prostaglandin was discontinued on day 6 and PH and RV dysfunction were treated with milrinone, sildenafil, spironolactone and furosemide. Echocardiographic follow up showed systematic improvement of RV function (fractional area change (FAC) 19% on day 5 to 35% on day 30) and PH signs withdrawal. Laboratory tests excluded metabolic errors as potential causes of RV dysfunction. Cardiac magnetic resonance (MRI) on day 22 showed impaired RV contractility (ejection fraction (EF) = 38%) with no signs of cardiomyopathy. The child was discharged home after one month of hospitalisation in a good health with the recommendation of oral diuretics use. At the age of 7 weeks Holter ECG monitoring showed numerous premature ventricular contractions with episodes of bigeminy and trigeminy. The arrhythmia was treated successfully with propranolol. After one year of follow up the child is in good clinical status with satisfying RV systolic function and no signs of PH on echocardiography.

**Conclusions:** Prenatal DA constriction may result in right heart dysfunction that may require treatment aiming at reduction of RV overload and PH.

**Keywords:** Arterial duct constriction, fetal echocardiography, pulmonary hypertension, right ventricular dysfunction

## P-293

### Maternal anti-RO & anti-LA antibodies and cardiac manifestations: more than complete congenital heart block?

Olga Panagiotopoulou<sup>1</sup>, Laurie Anderson<sup>2</sup>, Lindsey Hunter<sup>1</sup>

<sup>1</sup>Department of Paediatric Cardiology, Royal Hospital for Children, Glasgow, UK; <sup>2</sup>Department of Fetal Medicine, Queen Elizabeth University Hospital, Glasgow, UK

**Background and Aim:** Transplacental passage of maternal anti-Ro and anti-La antibodies can lead to fetal complete atrioventricular block (CAVB). In some cases, CAVB is associated with endocardial fibroelastosis (EFE). Isolated EFE, in the absence of CAVB secondary to maternal anti-Ro and anti-La antibodies, is a rare association and the clinical significance remains unclear.

**Method:** We report two fetal cases of isolated EFE diagnosed over the period of 2016–2022 in a national fetal cardiology centre.

Maternal antibody status was identified during pregnancy following EFE diagnosis. Retrospective data from maternal and offspring's history as well fetal and postnatal echocardiograms were collected.

**Results:** Case 1: A 36-year-old gravida 3 para 2+1 was referred for fetal echocardiography at 20+5 weeks gestation due to an abnormal fetal anomaly scan (FAS). Fetal echocardiogram showed increased endocardial echo-brightness around aortic and pulmonary annuli suggesting EFE. No valvular regurgitation, impaired ventricular function or pericardial effusion were detected. Fetal heart rate was normal with 1:1 atrioventricular conduction. Findings were stable at 36+6 weeks. The mother was screened for anti-Ro and anti-La antibodies and was found to be positive for Ro52 and Ro60. A healthy infant weighing 3.56 kg was born by elective caesarean-section at 39 weeks. Cardiac follow-up until 3 years of age showed no progression of EFE or development of CAVB. Systolic and diastolic ventricular performance was normal. Case 2: A 32-year-old gravida 3 para 2+0 was referred at 27+2 weeks gestation due to an abnormal FAS. Echogenic myocardium of tricuspid papillary muscles and aortic valve annulus was diagnosed without related valvar insufficiency, no ventricular dysfunction/dilatation, and no fetal heart rhythm abnormality. Fetal echocardiogram remained unchanged at 31+2 weeks. High maternal antibody titres of Ro52 (>240.0 U/ml), Ro 60 (>282 U/ml) and La (15 U/ml) were detected. Early and long-term postnatal cardiac assessment is anticipated.

**Conclusions:** Isolated EFE is a rare but a recognised feature in fetuses of mothers with autoimmune antibodies. Limited literature evidence suggests a poor pre and postnatal outcome, however in our experience, fetal and post-natal outcomes have been favourable. Increased awareness is essential as progression to CAVB and ventricular dysfunction have been described.

**Keywords:** endocardial fibroelastosis, maternal anti-Ro antibodies, maternal anti-La antibodies

## P-294

### Role of fetal echocardiography in double outlet right ventricle in accuracy of prenatal diagnosis and prediction of surgical approach

Anna Balducci<sup>1</sup>, Caterina Cecilia Lerosè<sup>2</sup>, Bartolacelli Ylenia<sup>1</sup>, Ferrè Cristina<sup>2</sup>, Gesuete Valentina<sup>1</sup>, Ciuca Cristina<sup>1</sup>, Palleri Daniela<sup>1</sup>, Montedoro Chiara<sup>3</sup>, Perolo Antonella<sup>3</sup>, Pilu Gianluigi<sup>3</sup>, Hasan Tammam<sup>1</sup>, Ragni Luca<sup>1</sup>, Gargiulo Gaetano<sup>4</sup>, Donti Andrea<sup>1</sup>

<sup>1</sup>Department of Paediatric Cardiology and Adult Congenital Heart Disease Program, IRCCS Azienda Ospedaliero-Universitaria S.Orsola-Malpighi, Bologna, Italy.; <sup>2</sup>Department of Medical Sciences, Neonatal and Intensive care Unit and Pediatric Section, University of Ferrara, Italy.; <sup>3</sup>Department of Medical and Surgical Sciences, Obstetric Unit, University of Bologna and IRCCS Azienda Ospedaliero-Universitaria S.Orsola-Malpighi, Bologna, Italy; <sup>4</sup>Department of Pediatric and Adult Congenital Heart Cardiac Surgery, University of Bologna and IRCCS Azienda Ospedaliero-Universitaria S.Orsola-Malpighi, Bologna, Italy

**Background and Aim:** Double outlet right ventricle (DORV) is a complex conotruncal cardiac malformation which affect less than 1% of all congenital heart disease. For complex cardiac malformations, paediatric cardiologists' counselling is essential. Recently literature has proved that an accurate fetal diagnosis and a proper parental counselling play a key role in defining clear prognosis and postnatal surgical approach. The aim of this study was to determine whether improvements in fetal echocardiography can result in increasing accuracy of diagnosis of DORV and also in predicting the type of surgical intervention, in terms of uni- or bi-ventricular repair.

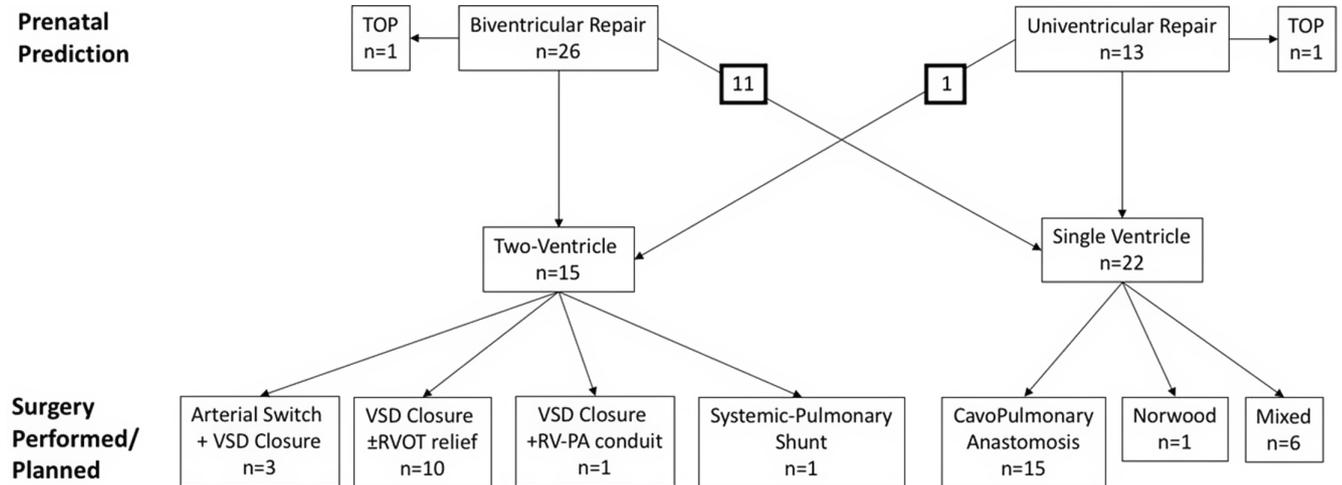
**Method:** Medical records of patients with fetal echocardiography diagnosis of DORV at our institution from 2009 to 2022 were collected. Pre and post-natal diagnosis were matched. We assessed the agreement between fetal echocardiography and postnatal evaluation in determining on one hand DORV type depending on VSD location, great artery orientation, and ventricular size, and on the other hand surgical prognosis distinguishing between two-ventricle repair and single-ventricle palliation.

**Results:** Our population consisted of 39 fetuses with none in utero deaths. There were 2 terminations of pregnancy. Median gestational age at first fetal study at our institution was 21 weeks and 3 days of gestation (range: 16–38 weeks). Prenatal diagnosis observed 16 patients (41%) with DORV Transposition of Great Arteries type; 9 patients (23%) with DORV non-committed VSD type, 7 patients (17.9%) with DORV Tetralogy of Fallot type, 5 patients (12.8%) with DORV-VSD type and 2 patients (5%) with DORV Atrio-Ventricular Septal Defect/Heterotaxy type. Diagnosis was confirmed in 94% (35 out of 37 living born) with prenatal accuracy and sensibility of certain diagnosis of 79.5% and 81%. Agreement in prenatal accuracy of type of DORV was 71.8%. Among patients who underwent surgery, 15 (40.5%) had two-ventricle repair and 22 (59.5%) had single-ventricle palliation. (Fig. 1) Agreement in prenatal prediction of surgical approach (single versus biventricular surgery) was 67.6%.

**Conclusions:** Double Outlet Right Ventricle is a rare and often complex cardiac anomaly that can be diagnosed prenatally with high precision nowadays. By implementing diagnostic accuracy of fetal echocardiography, important anatomical details can be determined enabling accurate prenatal counselling.

**Keywords:** fetal echocardiography, double outlet right ventricle, prenatal diagnosis, congenital heart disease

Prenatal surgical prediction compared with surgery performed/planned.



TOP: Termination of Pregnancy; VSD: Ventricular Septal Defect; RVOT: Right Ventricle Outflow Tract; RV-PA: Right Ventricle - Pulmonary Artery

Numbers in think-line square represent number of patients who changed categories between prenatal prediction and neonatal plan

**P-295**

**Hypoplastic left heart syndrome associated with total anomalous pulmonary venous return: case series.**

Ana Cécilia Mandirola<sup>1</sup>, Paula Randanne<sup>1</sup>, Fuensanta Escudero<sup>2</sup>, Olga Gómez Del Rincón<sup>3</sup>, Flavio Zuccarino<sup>4</sup>, Bosco Moscoso<sup>5</sup>, Fredy Prada<sup>1</sup>, Joan Sánchez De Toledo<sup>1</sup>, María C. Escobar Díaz<sup>1</sup>

<sup>1</sup>Department of Cardiology, Hospital Sant Joan de Deu, Barcelona, Spai; <sup>2</sup>Department of Cardiology, Hospital Clínico Universitario Virgen de la Arrixaca, Murcia, Spai; <sup>3</sup>Department of Maternal-Fetal Medicine, Hospital Sant Joan de Deu, Barcelona, Spain; <sup>4</sup>Department of Radiology, Hospital Sant Joan de Deu, Barcelona, Spain; <sup>5</sup>Department of Cardiovascular Surgery, Hospital Sant Joan de Deu, Barcelona, Spain

**Background and Aim:** The association of Hypoplastic left heart syndrome (HLHS) with Total Anomalous Pulmonary Venous Return (TAPVR) is extremely rare. Its timely recognition allows planning postnatal management.

**OBJECTIVE:** To describe the evolution of 4 cases of HLHS (mitro-aortic atresia) diagnosed prenatally with TAPVR.

**Method:** case series study

**Results:** Case 1: Male late preterm baby with low birth weight born due to fetal distress requires advanced resuscitation in the delivery room. Prenatally, a prominent levoatrial cardinal vein with a restrictive foramen ovale (FO) was observed, which was confirmed postnatally. The hemodynamic study prior to atrioseptostomy evidenced supracardiac TAPVR with hypoplasia and severe stenosis. The patient died 6 hours later after deciding comfort care.

Case 2: Female term newborn with a postnatal echocardiographic diagnosis of TAPVR with non-obstructive retrocardiac collector. She was maintained with prostaglandins, milrinone, non-invasive ventilation with hypoxic mixture. On the 5th day, Norwood type surgery was performed with anastomosis of the collector to the left atrium. At 4 months, she presented severe coarctation and percutaneous aortic angioplasty was performed, requiring

post-procedure ECMO. Due to neurological complications, comfort care was offered, and the patient died 15 days later

Case 3: Female term newborn. Postnatally, Non-obstructive TAPVR was diagnosed. She requires prostaglandins, milrinone, and non-invasive ventilation with hypoxic mixture. On the 7th day, Norwood-type surgery was performed with TAPVR repair. Glenn procedure was done at 4 months of age. She is now 21-months-old and in good general conditions.

Case 4: Male late preterm newborn. Restrictive FO was confirmed postnatally and required percutaneous atrioseptostomy that was not effective. The patient presented clinical worsening. Echocardiography/ CT angiography evidenced obstructive TAPVR and surgery was indicated. Hybrid procedure was decided due to the unfavorable anatomy. However, comfort care was started due to the impossibility of weaning extracorporeal circulation and the documented early-onset sepsis. The patient died few hours later

**Conclusions:** The association of HLHS with TAPVR is infrequent, but it is important to maintain a high level of suspicion for timely prenatal diagnosis. Postnatal management depends on the type of TAPVR. If it is obstructive, it manifests similarly to HLHS with restrictive FO showing high morbidity and mortality.

**Keywords:** hypoplastic left heart syndrome, total anomalous pulmonary veins drainag

#### P-296

##### Maternal hyperoxygenation test to predict postnatal urgent interventions

Maria C Escobar Díaz<sup>1</sup>, Olga Gómez Del Rincón<sup>2</sup>, Laura Nogué<sup>2</sup>, Leticia Benítez<sup>2</sup>, Jose María Martínez<sup>2</sup>, Joan Sánchez De Toledo<sup>1</sup>  
<sup>1</sup>Department of Cardiology, Hospital Sant Joan de Deu, Barcelona, Spain; <sup>2</sup>BCNatal, Fetal Medicine Research Center (Hospital Clinic and Sant Joan de Deu), Barcelona, Spain

**Background and Aim:** Maternal hyperoxygenation test (MHT) is a safe and effective tool to evaluate pulmonary vascular reactivity (PVR). It promotes pulmonary vasodilation, mimicking postnatal physiology. Some congenital heart diseases (CHD) require urgent interventions in the first days of life. Echocardiographic parameters during MHT may predict the need for these interventions.

MHT was performed to better characterize the PVR, and help to predict the need for urgent interventions, and the presence of pulmonary hypertension.

**Method:** Ten fetuses had MHT: 2 Ebstein's anomaly (EA), 3 transpositions of great arteries (TGA), 3 hypoplastic left heart syndrome (HLHS), and 2 aneurismatic foramen ovale (FO). PVR and physiologic and anatomic changes with MH and outcomes were recorded.

**Results:** Mean gestational age at MHT was 36, 1 weeks (range 35, 1-37, 2). One patient with EA persisted without antegrade pulmonary flow after the test and needed pulmonary valvuloplasty postnatally. The other patient who did not have antegrade flow pre-test developed antegrade flow and postnatally had antegrade flow and no need for any intervention.

In two fetuses with TGA, the MHT showed increased velocity at the FO and postnatally they needed urgent balloon atrioseptostomy (BAS). In the other patient, there were no changes in the flow through the FO, and did not need BAS after birth. All of them showed PVR and did not have pulmonary hypertension postnatally.

Pre-test, both patients with aneurismatic FO had retrograde flow at the aortic arch. After MHT, the aortic flow was antegrade and

the mitral inflow increased because of a reposition of the atrial septum. After birth, none of them had coarctation.

All 3 patients with HLHS showed PVR after MHT and postnatally did not have signs of pulmonary hypertension. One of them had increased velocity at the FO with MHT and required BAS after birth.

**Conclusions:** In our cohort, MHT was useful to predict the presence of pulmonary hypertension and the need for urgent intervention in some CHD. In some others, may help to elucidate the probability of developing aortic coarctation. Rutinary use of this test in these specific CHD may help to better counsel families and neonatal care planning.

**Keywords:** Maternal hyperoxygenation, congenital heart disease, pulmonary hypertension

#### P-297

##### Prenatal diagnosis and postnatal outcome of criss-cross heart – a single-center case series

Agnieszka Grzyb<sup>1</sup>, Joanna Szymkiewicz Dangel<sup>2</sup>  
<sup>1</sup>Department of Perinatal Cardiology and Congenital Anomalies, Center of Postgraduate Medical Education, Warsaw, Poland; Department of Cardiology, The Children's Memorial Health Institute, Warsaw, Poland.; <sup>2</sup>Department of Perinatal Cardiology and Congenital Anomalies, Center of Postgraduate Medical Education, Warsaw, Poland

**Background and Aim:** Criss-cross heart (CCH) is a very rare, complex congenital cardiac anomaly, defined by crossing of the ventricular inlets, however highly variable in terms of detailed segmental anatomy. Echocardiographic imaging of CCH is challenging, especially in the fetus.

We aimed to describe the prenatal findings and postnatal outcome of patients diagnosed with CCH in a single tertiary fetal cardiology center.

**Method:** Fetal echocardiographic database was retrospectively reviewed for cases of CCH diagnosed between 2011-2022. Segmental heart anatomy was described based on stored images and echocardiographic reports. Data on referral indications, coexisting anomalies, counseling and postnatal outcome were retrieved from medical records.

**Results:** 10 patients with CCH (5 males and 5 females) were identified within the study period, the diagnosis was confirmed postnatally in all cases. The median time of diagnosis was 22 weeks (range 17-24), usually after the mid-trimester obstetrics scan, where either single ventricle or conotruncal defects were suspected. Genetic testing was performed in 6 cases, with no abnormalities found. All patients presented with atrial situs solitus, in 3 cases – with dextrocardia. Atrioventricular connections were concordant in 6 cases, ventriculoarterial connections were discordant or double-outlet in all but one case. All fetuses had ventricular septal defects; other associated cardiac anomalies resulted in postnatal ductal-dependency in 9 patients. All neonates were born at term, in good clinical condition, with mean body weight 3440 gram. 9 patients are alive at follow-up, at different stages of single ventricle pathway, one died after surgery.

**Conclusions:** Despite complex anatomy, criss-cross heart can be precisely diagnosed in the prenatal period. Postnatal management is challenging; most patients require single ventricle palliation due to either ventricle hypoplasia or difficulty with intracardiac baffling.

**Keywords:** fetal cardiology, prenatal diagnosis, congenital heart defect, criss-cross heart

Patient	Diagnosis [weeks]	Referral indication	Sex	Genetics	Heart position	Atrial situs	AV connections	VA connections	VSD	Associated anomalies	Prostaglandin	Treatment	Outcome
1	22	TOF	M	46,XY	R	solitus	concordant	DORV	+	critical pulmonary stenosis	yes	BTS → BDG → Fontan	alive (9 yo)
2	17	SV	M	not tested	L	solitus	discordant	discordant	+	hypoplastic aortic arch	yes	Norwood → BDG → Fontan	alive (9 yo)
3	22	TOF	F	not tested	L	solitus	concordant	discordant	+	hypoplastic aortic arch	yes	aortic arch reconstruction, PAB	died at 2 months
4	24	CAT	F	aCGH (1-22,X)x2	L	solitus	concordant	DORV	+	hypoplastic RV	no	PAB → BDG	alive (5 yo)
5	23	heterotaxy	F	46,XX	R	solitus	discordant	discordant	+	pulmonary stenosis	yes	pulmonary balloon valvuloplasty, BTS → BDG	alive (4 yo)
6	20	SV	F	not tested	L	solitus	concordant	discordant	+	subpulmonary stenosis	yes	Rashkind, BTS → BDG	alive (3 yo)
7	23	SV	F	aCGH (1-22,X)x2	L	solitus	discordant	DORV	++	pulmonary stenosis	yes	BTS → BDG	alive (2 yo)
8	24	SV	M	aCGH (1-22)x2 (X,Y)x1	L	solitus	concordant	concordant	+	pulmonary atresia, straddling tricuspid valve	yes	BTS → BDG	alive (2 yo)
9	21	heterotaxy	M	aCGH (1-22)x2 (X,Y)x1	R	solitus	discordant	discordant	+	pulmonary atresia	yes	BTS → BDG	alive (2 yo)
10	23	complex CHD	M	not tested	L	solitus	concordant	discordant	+	straddling tricuspid valve, coarctation of the aorta, mild pulmonary stenosis	yes	Rashkind → Norwood → BDG	alive (2 yo)

**Table 1** Prenatal findings and postnatal outcome of criss-cross heart patients. Abbreviations: aCGH – array comparative genomic hybridization, BDG – bidirectional Glenn, BTS – Blalock-Taussig shunt, CAT – common arterial trunk, CHD – congenital heart disease, DORV – double outlet right ventricle, PAB – pulmonary artery banding, SV – single ventricle, TOF – tetralogy of Fallot, yo – years old.

## P-298

### Antenatal echocardiographic predictive parameters of postnatal outcome in neonates with ebstein anomaly

Sylvie Di Filippo<sup>1</sup>, Jérôme Massardier<sup>1</sup>, Fanny Deville<sup>1</sup>, Diane Ouvrier Buffet<sup>1</sup>, Olivier Metton<sup>1</sup>, Sylvie Di Filippo<sup>2</sup>

<sup>1</sup>Department of Congenital Heart Disease, Claude Bernard University, Lyon, France; <sup>2</sup>Congenital Cardiology, Cardiothoracic Center of Monaco

**Background and Aim:** Ebstein tricuspid valve anomaly is a rare CHD with uncertain postnatal prognosis. Criteria to predict outcome are still a matter of debate. The aim of this study was to determine antenatal echocardiographic predictive parameters.

**Method:** Retrospective multicentric analysis of fetus with diagnosis of Ebstein anomaly. Echocardiographic measurements of ventricles, atria, great vessels and tricuspid regurgitation were collected. Comparisons were made between group I (poor outcome = death occurred in utero or within the first 3 months of life) and group II (favourable outcome: postnatal survival > 3 months).

**Results:** 16 fetuses were included in the study: 10 in group I (62.5%: 2 terminations of pregnancy, 2 fetal deaths, 6 postnatal deaths) and 6 in group II (37.5%). Mean gestational age at diagnosis was 29 weeks (22 to 38). The mean number of echocardiographic records per patient was 2 (1 to 6). LV to RV ratio, tricuspid valve regurgitation grade and retrograde or anterograde ductal flow did not differ between the 2 groups. Significant differences were found between groups I and II regarding the presence of pulmonary flow (none or mild RV to PA flow: 8 of 9 cases died = 89%), AO to PA ratio (75% death if > 97°p vs 25% if 3-97°p), RA diameter (77.3% death if > 97°p vs 0%), PA diameter (100% death if < 3°p) and pericardial effusion (80% death vs 0%). Only 1 case had arrhythmia and died.

**Conclusions:** this small sample size study showed that the absence of RV to PA flow and/ or pulmonary valve opening, increased AO to

PA ratio, increased RA and decreased PA diameter and the presence of pericardial effusion might represent prognosis factors in fetus with Ebstein anomaly. These results should be confirmed by large scale prospective study.

**Keywords:** antenatal diagnosis, Ebstein anomaly, prognosis, risk factors

## P-299

### Anusual fetal case of suspected PAPVD with LPA sling – added value of fetal cardiac mri

Mari Nieves Velasco Forte<sup>1</sup>, Patricia Caldas<sup>1</sup>, Trisha Vigneswaran<sup>2</sup>, Laura Konta<sup>1</sup>, Kelly Ann Eastwood<sup>3</sup>, Vico Schot<sup>4</sup>, Giovanni Biglino<sup>4</sup>, Massimo Caputo<sup>4</sup>, Srinivas Narayan<sup>4</sup>, Milou Van Poppel<sup>5</sup>, Kuberan Pushparajah<sup>5</sup>, David Lloyd<sup>5</sup>

<sup>1</sup>Fetal & Paediatric Cardiology Department, Bristol Royal Hospital for Children, Bristol; <sup>2</sup>Fetal & Paediatric Cardiology Department, Evelina Children's Hospital, London; <sup>3</sup>Fetal Medicine Department, St Michael's Hospital, Bristol; <sup>4</sup>Bristol Medical School, University of Bristol; <sup>5</sup>School of Biomedical Engineering & Imaging Sciences, King's College London

**Background and Aim:** Left pulmonary artery (LPA) sling is a rare form of congenital heart disease, in which the LPA arises from the right pulmonary artery, passing between the trachea and the oesophagus to reach the left lung. We present a case in which PAPVD plus LPA sling was initially suspected, but further imaging involving fetal Cardiac Magnetic Resonance (CMR) revealed different findings.

**Method:** 35-year-old woman referred to Fetal Cardiology in view of her 20-week anomaly scan showing abnormal 3 vessel view (3VV).

**Results:** After fetal echocardiogram, LPA sling, persistent left superior vena cava (LSVC) and possible right-sided PAPVD were suspected, the latter based on the finding of venous drainage from the

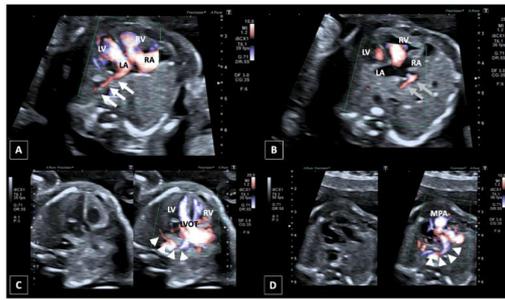


Figure 1

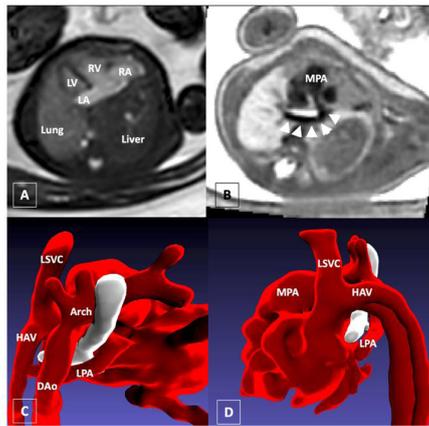


Figure 2

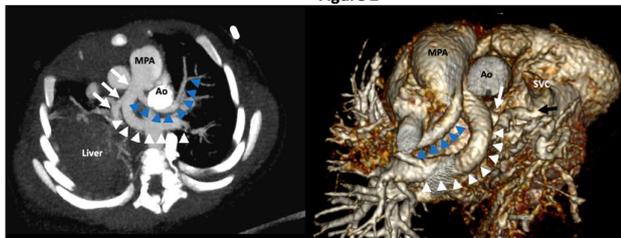


Figure 3.

**Figure 1.** Fetal echocardiogram. A) 4-chamber view showing left pulmonary veins (long white arrows) into the left atrium. B) 4-chamber view at a higher level, showing hepatic venous return (grey arrows) into the RA (initially thought to be right pulmonary veins). C) LVOT view including LPA sling (white short arrows). D) Modified MPA/3-vessel-view demonstrating LPA sling (white short arrows). LA= left atrium, LV= left ventricle, LVOT= left ventricular outflow tract, RA= right atrium, RV= right ventricle

**Figure 2.** Fetal CMR. A) Uncorrected bSSFP image showing axial view at level of four chamber. B) Motion-corrected HASTE image showing left pulmonary artery sling in axial plane (white arrows). C) Segmentation of motion-corrected HASTE data (right superior oblique view). D) Segmentation of motion-corrected HASTE data (left lateral view) B= bronchi, DAo= descending aorta, HAV = hemi-azygos vein, LA= left atrium, LPA= left pulmonary artery, LSVC= left superior vena cava, LV= left ventricle, T= trachea

**Figure 3.** Postnatal MDCT scan demonstrating unusual anatomy of the pulmonary circulation: left upper pulmonary branch arising normally from MPA (blue arrows), left middle and left lower lobe vascularized through artery arising as an LPA sling (short white arrows); RPA arising from MPA (long white arrows) and partial anomalous pulmonary drainage with right upper pulmonary vein draining into SVC (black arrow). Right image: 2D axial multiplanar reformat view. Left image: 3D reconstruction from MDCT. Ao= aorta, MPA= main pulmonary artery, SVC= superior vena cava

right thorax to the right atrium with ventricular asymmetry (Figure 1). Due to the unusual combination of findings, the patient was referred for a fetal CMR to further delineate the anatomy, which was performed at 31 weeks. Motion-corrected 3D fetal CMR confirmed LPA sling but also revealed a large right-sided congenital diaphragmatic hernia (CDH) on T2-weighted imaging, with intra-thoracic hepatic veins from the herniated liver explaining the unusual ultrasound appearance. The right lung was visible in the upper thorax but severely hypoplastic and the right pulmonary veins were not visualised (Figure 2). Multi-disciplinary input was sought from fetal medicine, paediatric cardiology and paediatric surgery and the parents were counselled accordingly. Postnatally, the CDH was repaired, and a CT scan was performed, which confirmed an LPA sling with an additional left upper branch arising more proximally from the MPA, as well as PAPVD with the RUPV draining to the SVC, with mild to moderate compression of the distal trachea and left bronchus (Figure 3). Surgical repair of the sling/PAPVD has not yet been performed and symptoms are being monitored.

**Conclusions:** Right-sided CDH can be difficult to diagnose in fetal life due to the similar echodensity of lung and liver prenatally, whereas these tissues can have markedly different contrast on prenatal MRI. Whilst not a replacement for postnatal imaging, fetal CMR can help to delineate complex anatomy when the diagnosis is uncertain in fetal life, allowing for more reliable parental counselling and more comprehensive postnatal planning.

**Keywords:** Fetal CMR, Fetal echocardiography, LPA sling, PAPVD, prenatal diagnosis

### P-301

#### Perinatal and delivery management in prenatally diagnosed congenital heart disease; an analysis of practice in northern Ireland

Nicola Mcay<sup>1</sup>, Edith Crawford<sup>2</sup>, Frank Casey<sup>2</sup>

<sup>1</sup>Children's Health Ireland, Crumlin, Dublin, Ireland; <sup>2</sup>Paediatric Cardiology, Royal Belfast Hospital for Sick Children, Belfast, Northern Ireland

**Background and Aim:** Subjective assessment in the Paediatric Cardiology department in the Royal Belfast Hospital for Sick Children between 2020–2021 was that an increasing number of deliveries of patients with a prenatal diagnosis of congenital heart disease were delayed and subsequently occurring 'out of hours' or at weekends. This resulted in sub-optimal timing for clinical management and increased difficulties arranging transfer to Ireland's surgical centre in Dublin.

Our study aimed to analyse the delivery pathway for patients with a prenatal diagnosis of congenital heart disease in Northern Ireland. Through definition and analysis of current practice we felt this would better enable us to understand and implement change to improve future practice. Our aim was to create a fetal cardiac delivery pathway to optimise delivery of patients with a prenatal diagnosis of congenital heart disease.

**Method:** Retrospective chart and electronic record review of all patients diagnosed antenatally with congenital heart disease in Northern Ireland between 2019–2021. Telephone reviews were conducted with families impacted by delivery delays to gain their perspective of events.

**Results:** • 87-fetal cardiac patients were reviewed between 2019–2021 with 86-live born infants.

• Planned delivery method for patients with prenatal diagnosis of congenital heart disease in 69%—was induction of labour and 25%—elective caesarean section.

- 30.6%–of deliveries that were planned for induction delivered spontaneously prior to their due date.
- 27%–of deliveries occurred between Friday–Sunday.
- 54%–of all deliveries and 76% of induction deliveries occurred ‘out of hours’ between 5pm–9am.
- 24.7%–of all planned deliveries were delayed and 55% of induction deliveries were delayed.
- 95%–of deliveries were delayed due to lack of neonatal bed availability as a consequence of skilled nursing shortages.
- Delivery delays lead to difficulties in 66%–of patients including stress, subsequent hospital transfer and weekend delivery. Longer delivery delays were associated with more significant congenital heart disease.

**Conclusions:** Identification and analysis of this sub-optimal delivery of patients with an antenatal diagnosis of congenital heart disease enabled us to engage the neonatal and obstetric teams to implement a multi-speciality fetal cardiac delivery pathway. Better delivery planning should reduce delay in planned births and facilitate improvements in surgical planning leading to better outcomes for babies and their families.

**Keywords:** fetal, cardiac, delivery

### P-302

#### **Fetal cardiology in northern ireland; analysis of antenatal diagnosis, postnatal management and transfer of patients requiring early neonatal surgery**

Nicola McCay<sup>1</sup>, Edith Crawford<sup>2</sup>, Frank Casey<sup>2</sup>

<sup>1</sup>Children’s Health Ireland, Crumlin, Dublin, Ireland; <sup>2</sup>Paediatric Cardiology, Royal Belfast Hospital for Sick Children, Belfast, Northern Ireland

**Background and Aim:** Paediatric Cardiology in Northern Ireland is unique as a previous centre for Paediatric Cardiology Surgery it has had to adjust following discontinuation of surgical services for patients born in NI with a diagnosis of congenital heart disease that require immediate surgical management post-delivery.

The Paediatric Cardiology Consultant team have continued to offer fetal cardiac diagnosis and delivery of these patients within Northern Ireland with an aim to transfer post-delivery when stable to the allied surgical centre in Children’s health Ireland, Dublin. There has been a transition of surgical services from England to Dublin in recent years.

The aim of our study was to review fetal cardiology diagnoses, accuracy of antenatal detection and immediate postnatal management including transfer to a surgical centre when indicated.

**Method:** Retrospective chart and electronic record review of all patients diagnosed antenatally with congenital heart disease in Northern Ireland between 2019–2021.

**Results:** • 87 fetal cardiac patients were reviewed between 2019–2021 with 86 live born infants; 58.2% male, 41.8% female.

- Mean gestation at cardiac diagnosis 23+4 weeks.
- 100% accuracy of antenatal diagnosis of patients correctly diagnosed with TGA, isomerism, truncus arteriosus, right heart obstruction, TV anomalies and double inlet ventricle.
- Accuracy of 19/20 patients with left heart obstruction, 11/15 tetralogy/DORV diagnoses and 20/22 septal defects (100% accuracy with AVSD).
- Of those that were felt antenatally to be suspicious of coarctation only 42.8% were confirmed postnatally.

- All patients with an antenatal diagnosis of TGA were delivered in Dublin close to the surgical centre.
- 34.9% of patients required early neonatal surgical transfer within 2 weeks of life.
- Patients with duct-dependent circulation were maintained on Prostaglandin infusion. Single ventricle patients were transferred to a surgical centre within a mean time of 2.32days, coarctation of aorta transfer time 4.67days,
- Mean single ventricle transfer time to England 2.36days versus average transfer to Dublin 2.09days.

**Conclusions:** There is a high accuracy rate of antenatal diagnosis of severe congenital heart disease in Northern Ireland which enables delivery in a tertiary neonatal centre allowing families to deliver closer to home. Babies are then stabilised in Belfast prior to transfer to Dublin within an average transfer time of 2.09days.

**Keywords:** fetal, cardiac, diagnosis, transfer

### P-304

#### **A novel ratio to differentiate fetal right aortic ARCH from double aortic ARCH pilot study**

Pia Cassanello<sup>1</sup>, Laura Nogué<sup>2</sup>, Olga Gómez<sup>2</sup>, Flavio Zuccharino<sup>3</sup>, Mar Bennasar<sup>2</sup>, Marta Camprubi-Camprubi<sup>4</sup>, Jose María Martínez<sup>2</sup>, Joan Sánchez-de-Toledo<sup>5</sup>, Maria Clara Escobar-Díaz<sup>6</sup>

<sup>1</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain; <sup>2</sup>BCNatal, Fetal Medicine Research center (Hospital Clínic and Sant Joan de Déu), Barcelona, Spain; <sup>3</sup>Department of Pediatric Radiology, Hospital Sant Joan de Déu, Barcelona, Spain; <sup>4</sup>Department of Radiology, Hospital del Mar, Barcelona, Spain; <sup>5</sup>BCNatal, Fetal Medicine Research center (Hospital Clínic and Sant Joan de Déu), Barcelona, Spain; <sup>6</sup>Institut de Recerca Sant Joan de Déu, Barcelona, Spain; <sup>7</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain; <sup>8</sup>Institut de Recerca Sant Joan de Déu, Barcelona, Spain; <sup>9</sup>Department of Critical Care Medicine, University of Pittsburgh, Pittsburgh, USA; <sup>10</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain; <sup>11</sup>Institut de Recerca Sant Joan de Déu, Barcelona, Spain

**Background and Aim:** Currently, prenatal diagnosis of right aortic arch (RAA) is feasible. However, differentiating RAA from double aortic arch (DAA) might be challenging. In DAA the carotid and subclavian artery arise separately from each aortic arch. Therefore, we hypothesized that patients with DAA have a smaller transverse right arch (TRA) than RAA patients due to blood flow differential distribution. This study aims to explore the utility of the TRA/ascending aorta (AA) ratio to prenatally differentiate between DAA and RAA regardless of gestational age.

**Method:** Single-center, retrospective cohort pilot study of patients with isolated DAA or RAA prenatally diagnosed between 2015 and 2022. Confirmatory measurements and ratio values were obtained from the post-natal computed tomographic angiography (CTA) in all patients. AA diameter was measured in a sagittal view above the aortic valve, and TRA at its larger diameter between the first and second supra-aortic branches. Fetal measurements were made blinded to the diagnosis by two expert echocardiographers. Prenatally, AA diameter was measured above the aortic valve in the 5-chamber view, and TRA in the 3- vessel and trachea view (3VTV) at its larger diameter. We established cut-off points for TRA/AA ratio, and AUC were calculated. Data is presented as median and IQR.

**Results:** We included 17 patients: 12 RAA, 5 DAA. Median gestational age at diagnosis was 25 weeks (20–30). Postnatal CTA TRA/AA ratio in the RAA group was 0.97 (0.8–1) vs 0.74 (0.67–0.82) in the DAA group (p = 0.03). Prenatal ultrasound TRA/AA ratio in the RAA group was 0.87 (0.78–0.89) vs 0.71

(0.67–0.79) in the DAA group ( $p = 0.08$ ). Prenatal TRA/AA AUC was 0.78 (0.50–0.93) with an estimated cut-off value of 0.77 with a sensitivity of 83% and specificity of 60% to differentiate DAA from RAA.

**Conclusions:** In our cohort, patients with DAA have smaller TRA compared to AA than those with RAA, both pre- and postnatally. A prenatal cut-off ratio of 0.77 between these diameters was found to be helpful to differentiate DAA from RAA. Further studies encompassing larger series should confirm these preliminary results that suggest that prenatal TRA/AA ratio might be used as a diagnostic tool to differentiate between DAA and RAA.

**Keywords:** Double aortic arch, right aortic arch, fetal diagnosis

### P-305

#### Management of fetal atrial flutter: A 5 cases study

Manel Charfi, Bouraoui Amira, Gargouri Rania, Rguaieg Chiraz, Kolsi Nedja, Ben Hmed Amel, Abdennadher amina, Kammoun Dhrouha, Hmida Nedja, Abid Leila, Ben Thabet Afef, Gargouri Abdeltif  
departement of cardiology an neonatology, HEDI CHAKER university hospital, SFAX, TUNISIA

**Background and Aim:** Fetal arrhythmias can be observed in 1 to 3% of pregnancies. Their diagnosis is still a challenge for physicians. Fetal atrial flutter is a rare condition that represents about 30% of fetal tachyarrhythmias. It's a serious condition that can lead to cardiac dysfunction an even fetal demise. Intrauterine therapy is possible. Through this study we aim to remind, clinical features, management and outcome of fetal atrial flutter.

**Method:** It's a retrospective study of all cases of documented fetal atrial flutter hospitalized in the neonatal intensive care unit of Sfax between January 2004 and October 2022.

**Results:** We registered 5 fetuses diagnosed prenatally with atrial flutter. A male predominance was noted with a sex ratio of 1.5. Prenatal diagnosis was performed in all cases by fetal echocardiography which was indicated because of hydrops fetalis in 2 cases, a dilatation of the right heart cavities in one case and a fetal tachycardia in 2 cases. Intrauterine treatment was administered in the 3 cases. It was based on amiodarone associated to digoxin in one case, digoxin in one case and digoxin associated with sotalol then relayed by flecainide in one case. For the other fetuses we opted for clinical surveillance as they were full term and they have no signs of heart failure. After birth, 3 newborns presented signs of heart failure. For the others, the clinical manifestation was tachycardia. Diagnosis was confirmed in all cases by postnatal electrocardiogram. Echocardiography was performed for all patients. It showed tight pulmonary narrowing in one case. Spontaneous conversion to sinus rhythm was observed in one newborn. In the other cases, the first line treatment was amiodarone. It was successful in only one case. An external electric conversion was then necessary in 3 cases. Mean age of conversion to sinus rhythm was 5 days [0 to 11 days]. Outcome was favorable for all newborns. No recurrence of the flutter was noted with an average follow-up of 2 years.

**Conclusions:** The outcome of fetal atrial flutter depends on the prompt of diagnosis which can be challenging. Management must be rapid and multidisciplinary. Hence we insist on the importance of prenatal diagnosis.

**Keywords:** fetal, flutter, diagnosis, management

### P-306

#### Right ventricular outflow tract anomalies in twin-to-twin transfusion syndrome – a single-centre experience.

Katarzyna Szaflik<sup>1</sup>, Katarzyna Janiak<sup>2</sup>, Krzysztof Szaflik<sup>2</sup>, Paweł Dryżek<sup>1</sup>, Tomasz Moszura<sup>1</sup>

<sup>1</sup>Department of Cardiology, Polish Mother's Memorial Hospital – Research Institute, Łódź, Poland; <sup>2</sup>Department of Gynecology, Fertility and Fetal Therapy, Polish Mother's Memorial Hospital – Research Institute, Łódź, Poland

**Background and Aim:** To evaluate the prenatal course and perinatal and postnatal outcome of fetuses with twin-to-twin transfusion syndrome (TTTS) and right ventricular outflow tract (RVOT) abnormalities and find possible risk factors of severe pulmonary stenosis after birth.

**Method:** This was a retrospective study of 19 fetuses (16 recipients, 3 donors) diagnosed with TTTS and RVOT anomalies treated with selective fetoscopic laser photocoagulation (SFLP) at our institution from 2015 to 2021. Prenatal RVOT anomalies were defined as abnormal flow velocity waveform across the pulmonary valve and two subgroups were identified: functional/anatomical pulmonary atresia and pulmonary stenosis. We evaluated fetal echocardiography data, data regarding perinatal outcome, neonatal echocardiography and cardiac management including need for trans-catheter intervention.

**Results:** Pulmonary stenosis was most common (89, 5%), followed by pulmonary atresia (10, 5%). The survival rate was around 78, 9%: one fetus died in utero at 28 weeks of gestation, three newborns died within 1 week owing to prematurity combined with cardiac failure, before cardiac intervention could be performed. Fetal therapy resulted in improvements in 14/15 recipient twins following SFLP, but four newborns (21% of the entire cohort) had persistent pulmonary valve abnormalities at birth, requiring balloon dilatation of the pulmonary valve. In this group of patients the time-interval between diagnosis of TTTS and antenatal RVOT anomalies was significantly shorter than in newborns who did not require percutaneous treatment ( $p < 0, 05$ ). One patient with clinical manifestation of heart failure and cyanosis presented hypoplasia of tricuspid valve, pulmonary valve and right ventricle with signs of its fibroelastosis. Bidirectional cavopulmonary shunt in this case is considered.

**Conclusions:** Postnatal severe RVOT anomalies are associated with early gestational age at onset of TTTS and RVOT anomalies. Abnormal tricuspid valve and right ventricle development may probably result from disturbance in intracardiac flow during the period of embryogenesis in TTTS. Extended postnatal follow-up of these high-risk cases is mandatory.

**Keywords:** twin-to-twin transfusion syndrome, selective fetoscopic laser photocoagulation, right ventricular outflow tract obstruction, pulmonary atresia, pulmonary stenosis, balloon valvuloplasty

### P-307

#### Prenatal diagnosis of aortic ARCH anomalies, vascular rings in the last seven years in a pediatric cardiac tertiary center: presentation and outcome.

Lucía Deiros Bronte<sup>1</sup>, Ángela Uceda<sup>1</sup>, Roberto Rodríguez<sup>2</sup>, Beatriz Herrero<sup>2</sup>, Carlos Labrandero<sup>1</sup>, Diana Salas<sup>1</sup>, Alvaro Lafuente<sup>1</sup>, Mercedes Aristoy<sup>1</sup>, Marta Gamba<sup>1</sup>, Raul Sanchez<sup>4</sup>, Jesus Diez Sebastian<sup>3</sup>, Luz Polo<sup>4</sup>, Eugenia Antolin<sup>2</sup>, Luis García Guereta<sup>1</sup>, Federico Gutierrez Laraya<sup>1</sup>

<sup>1</sup>Department of pediatric cardiology, La Paz University Hospital, Madrid, Spain; <sup>2</sup>Department of obstetrics and gynaecology, La Paz University Hospital, Madrid, Spain; <sup>3</sup>Department of clinical epidemiology, La Paz University Hospital, Madrid, Spain; <sup>4</sup>Department of pediatric cardiac surgery, La Paz University Hospital, Madrid, Spain

**Background and Aim:** After adopting a three-vessel and three-vessel-trachea view (3VT), the diagnosis of fetal aortic arch anomalies

(AAA) improved. These alterations may be related to vascular rings (VR), intracardiac, and chromosomal abnormalities.

**AIM:** to describe the frequency of fetal AAA and VR subtypes and analyse the associated malformations and diagnostic accuracy in our centre.

**Method:** Retrospective study of AAA (except cases of aberrant right subclavian artery) and VR, and the association with intracardiac / extracardiac / genetic alterations, diagnosed during 2015–2022. Postnatal evolution was collected.

**Results:** 53 cases of fetal AAA (median gestational age: 25 weeks) were diagnosed during the study period: 25 (47, 2%) with right aortic arch (RAA) + aberrant left subclavian artery (ALSCA) / left-sided ductus arteriosus (DA), 12 (22, 6%) patients with double aortic arch (DAA), 11 (20.8%) RAA with mirror image branching (MIB) without VR, 4 (7, 5%) RA+ MIB with left DA (VR) and 1 (1, 9%) with pulmonary artery sling.

RAA + MIB without VR were significantly associated with intracardiac defects ( $p < 0.000$ ), while most of the other AAA subtypes had a normal heart. Extracardiac anomalies were found in 20% of RAA+ALSCA cases. Genetic anomalies were checked in 33/53 (63, 4%). In four (12%) a genetic anomaly was detected, of these only one had isolated AAA.

Pregnancy was interrupted in two fetuses (both of them had a major cardiac anomaly). Two died in utero, three are currently unborn, 46 babies were delivered and 100% were postnatally confirmed to have an AAA. In VR diagnosis, a postnatal CT was done in 30/36 cases. In 87%, the fetal diagnosis of VR subtype was concordant with that of the CT.

Currently, the follow-up average is 3.2 years and 55% of patients born with VR, either due to symptoms or tracheal/esophageal compression, required surgery (double aortic arch up to 88% of cases).

**Conclusions:** AAA and VR subtype prenatal diagnoses are remarkably accurate and allow parents to be correctly informed of possible associated fetal abnormalities. RAA + MIB without VR is significantly associated with intracardiac defects. Cases of isolated vascular ring should be followed up in paediatric cardiac centres with adequate information to parents on symptoms.

**Keywords:** Aortic arch anomalies, vascular rings, prenatal diagnosis, right aortic arch.

### P-308

#### Regression of cardiac rhabdomyomas producing severe aortic stenosis. case report

Vlasta M. E. Fessler<sup>1</sup>, Luciane Piazza<sup>2</sup>, Antonio Saracino<sup>2</sup>, Andreea Andronache<sup>2</sup>, Alessandro Varrica<sup>3</sup>, Alessandro Giamberti<sup>3</sup>, Alessandro Frigiola<sup>3</sup>

<sup>1</sup>Center Fetal Cardiology, IRCCS Policlinico San Donato, Milan, Italy;

<sup>2</sup>Department Pediatric Cardiology, IRCCS Policlinico San Donato, Milan, Italy; <sup>3</sup>Department Pediatric Cardiosurgery, IRCCS Policlinico San Donato, Milan, Italy

**Background and Aim:** Fetal cardiac rhabdomyomas (R) are known to regress after birth. We report an unusual case.

**Method:** A 30 years old healthy women at her 2nd pregnancy, was sent at 32 weeks gestation (wg) for echocardiography for a finding at obstetrical scan of a mass in the heart. No familiar or obstetrical scan problems were reported.

**Results:** A voluminous echodense mass was evident in the inlet left ventricle (LV) close to the mitral valve and a smaller mass protruding into the aortic outflow with a small free space around the valve and accelerated aortic flow, with pulsed Doppler velocity at upper limits. Mitral valve showed slightly reduced flow. No other nodes were visualized. The LV contractility was normal. Diagnosis of

multiple R was done, without abnormal findings in other fetal organs. CNS MRI was postponed. At 36 w.g. a cesarean section was done fearing a progression of the masses. A girl baby was born in good general conditions, birth weight 3.150 kg, Apgar 8–9. The appearance of the mass in the LV inflow was unchanged, while a smaller mass in the aortic area obstructed aortic flow with maximum gradient 50 mmHg that remained unchanged thereafter. There were no rhythm problems. Neurological evaluation was normal, CNS MRI did not show specific lesions and tuberous sclerosis was excluded by negative genetics. No immediate surgery was considered necessary and the baby was followed up closely, the aortic gradient remaining stable. The large cardiac mass progressively regressed throughout the years, as well as the aortic mass, but with apparently damaged aortic valve. At 9 years the LV mass almost disappeared but the aortic valve looked more severely damaged, with increased aortic gradient reaching 88–100 mmHg. Therefore it was decided to plan a correction. It was possible to perform a plasty of the valve resecting a fibrous circumferential rim. After then only mild aortic regurgitation and mild systolic gradient of 20 mmHg with good LV function remained. The girl is now 12 years old and well.

**Conclusions:** Our case is unusual showing a progressive spontaneous regression of R masses but with a subsequent development of severe aortic valve stenosis.

**Keywords:** fetus, fetal echocardiography, rhabdomyoma, aortic stenosis

### P-310

#### Prenatal diagnosis of absent pulmonary valve syndrome: anatomical features and perinatal outcomes

Andrii Kurkevych, Anna Didkowska

Research and Diagnosis Department, Ukrainian Children's Cardiac Center, Kyiv, Ukraine

**Background and Aim:** Absent pulmonary valve syndrome (APVS) is a rare congenital heart disease that occurs in about 1 in 100 000 fetuses. There are 2 different types of APVS: with ventricular septal defect (APVS-VSD) and with intact ventricular septum (APVS-IVS). The aim of this study was to analyze the anatomical features of these two types with an assessment of their perinatal outcomes.

**Method:** According to the prenatal database of our center from 2010 to 2021, 36 fetuses with APVS were identified. We retrospectively reviewed echocardiographic data of all fetuses with APVS. Perinatal outcomes analyzed according to clinical data.

**Results:** All 36 fetuses at the initial diagnosis (mean age 24, 1±5, 7 weeks) were divided into two groups: I group – APVS-VSD (n = 31; 86, 1%) and II group – APVS-IVS (n = 5; 13, 9%). In the I group the ductus arteriosus (DA) was absent in 29 (93.5%) cases and right aortic arch (RAA) was present in 14 (45.2%) ones. Thymus hypoplasia/agenesis was diagnosed in 8 (25.8%) fetuses. Conversely, in the II group DA was present in all cases (100%), but there were no RAA and thymus abnormalities. Analyzing fetuses with absence (n = 29) or presence (n = 7) of DA, we found a significant increase in Z scores of PA branches in the first group without DA: RPA (5, 6±1, 9 and 3, 2±1, 7;  $p < 0, 01$ ) and LPA (5, 4±1, 7 and 4, 0±1, 4;  $p < 0, 05$ ) relatively. Whereas there was no significant difference in diameters of the pulmonary fibrous ring and the pulmonary trunk. Of all fetuses with APVS (n = 36), in 11 (30, 6%) cases the parents decided to terminate the pregnancy. In 3 (8.3%) cases there was intrauterine death, and in another 3 (8.3%) the follow-ups were lost. Nineteen (52, 8%) children were born, but 4 (11, 1%) of them died in the pre-operative period, and 12 (33, 3%) patients were operated on with a mortality rate 8, 3%

(1 death). In total, 14 patients (73, 7% of all born children) survived after 1 year of life.

**Conclusions:** Two types of APVS have different anatomical features and the presence of DA reliably reduces the diameters of the pulmonary artery branches. Perinatal outcomes remain controversial, with high level of pregnancy termination at early diagnosis and perinatal death.

**Keywords:** absent pulmonary valve syndrome, fetal diagnosis, anatomical features, perinatal outcome

### P-312

#### Early fetal cardiovascular adaptation to an artificial placenta in a sheep model initial approach to extrauterine support for extreme prematurity.

Paula C Randanne<sup>1</sup>, Sergio Sanchez Martinez<sup>2</sup>, Ameth Hawkins Villareal<sup>4</sup>, Kambiz Rezaei<sup>4</sup>, Miguel Moran<sup>4</sup>, Raquel Ficho<sup>4</sup>, Sara Bobillo Perez<sup>4</sup>, Victor Gomez<sup>3</sup>, Mireia Gispert<sup>3</sup>, Maite Mata<sup>3</sup>, Elisenda Bonet\_carne<sup>2</sup>, Miriam Illa<sup>4</sup>, Elisenda Eixarch<sup>4</sup>, Bart Bijnen<sup>2</sup>, Joan Sanchez De Toledo<sup>5</sup>, Fatima Crispi<sup>2</sup>, Eduard Gratacos<sup>4</sup>

<sup>1</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain; BCNatal Fetal Medicine Research Center (Hospital Clinic and Hospital Sant Joan de Déu). Universitat de Barcelona, Barcelona, Spain; <sup>2</sup>BCNatal Fetal Medicine Research Center (Hospital Clinic and Hospital Sant Joan de Déu). Universitat de Barcelona, Barcelona, Spain; Institut d'Investigacions Biomèdiques August Pi i Sunyer (IDIBAPS), Barcelona, Spain; <sup>3</sup>Hospital Clinic, Barcelona, Spain; <sup>4</sup>BCNatal Fetal Medicine Research Center (Hospital Clinic and Hospital Sant Joan de Déu). Universitat de Barcelona, Barcelona, Spain; <sup>5</sup>Department of Pediatric Cardiology Hospital Sant Joan de Déu, Barcelona, Spain

**Background and Aim:** To describe the early fetal cardiovascular adaptation after connection to an artificial placenta (AP) in fetal sheep, measured by ultrasound and invasive hemodynamics.

**Method:** An experimental study of 12 fetal lambs (109-117 days), which were transferred to an AP system (pumpless circuit with umbilical cord connection). The study was designed to collect in utero and post-cannulation measures. The six first consecutive fetuses were instrumented with intravascular catheters and perivascular probes to obtain key physiological invasive data (arterial and venous intravascular pressures and blood flows) These experiments aimed at 1-3 hours survival. The second set of 6 fetuses were not instrumented and belonged to experiments aiming at 3-24 hours survival. Echocardiography-derived anatomical and functional measurements were collected in most animals, as well as the blood flow and pre-membrane and post-membrane pressures measured in the AP system. These data were acquired at different stages of our experimental setting, namely, in utero, 5', 30' (instrumented animals) and in utero, 30' and 180' (non-instrumented animals) after transfer into the AP system.

**Results:** There was a reduced pulsatility index in the umbilical artery (UA-PI in utero median 1.36 (IQR1.06-1.5) vs. 30' 0.38 (0.31-0.5) vs. 180' 0.36 (0.29-0.41),  $p < 0.001$ ) and ductus venosus, together with increased umbilical venous peak velocity and flow (UV peak velocity in utero 20.3 cm/s (18.2-22.4) vs. 5' 39 cm/s (30.7-43.2) vs 180' 43 cm/s (34-54),  $p < 0.001$ ) becoming pulsatile after connection.

Intravascular measures showed transiently increased arterial and venous pressures (mean arterial pressure in utero 43 mmHg (35-54) vs. 5' 72 mmHg (61-77) vs. 30' 58 mmHg (50-64),  $p = 0.02$ ) and fetal heart rate (in utero 145 bpm (142-156) vs. 30' 188 bpm (171-209) vs. 180' 175 bpm (165-190),  $p = 0.001$ ). The fetal heart structure and function was mainly preserved (right fractional area

change in utero 36% (34-40.9) vs. 30' 38% (30-40) vs. 180' 37% (33.3-40),  $p = 0.807$ ).

**Conclusions:** Connection to an AP resulted in a transient fetal hemodynamic response that tended to normalize over hours. In this short-term evaluation, cardiac structure and function was preserved. However, the system results in non-physiologically elevated venous pressure and pulsatile flow, which should be corrected to avoid later impairment in cardiac function.

**Keywords:** artificial placenta, fetal sheep model, fetal sheep echocardiography, fetal hemodynamics,

#### INVASIVE MONITORING AP

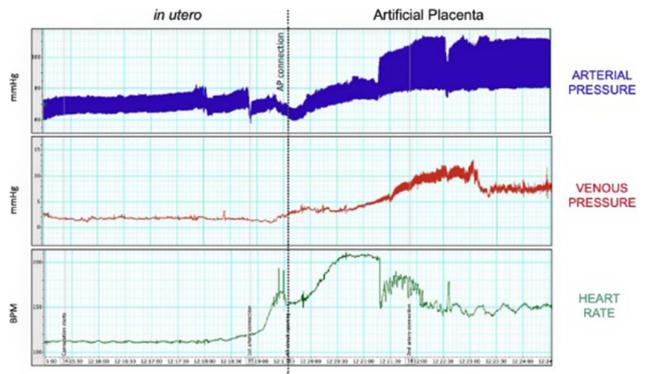


Figure 1. shows the changes observed in the continuous monitoring signal of arterial (top, in red) and venous (middle, in blue) pressure and heart rate (bottom, in green) during the in utero period, circuit connection to an artificial placenta and subsequent early adaptation, in one illustrative example.

### P-314

#### Entrustable professional activities and their relevance to pediatric cardiology training

Michael Kim<sup>1</sup>, Justin Tretter<sup>2</sup>, Ivan Wilmot<sup>1</sup>, Eunice Hahn<sup>1</sup>, Andrew Redington<sup>1</sup>, Colin McMahon<sup>3</sup>

<sup>1</sup>Department of Pediatric Cardiology, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA; <sup>2</sup>Department of Pediatric Cardiology, Cleveland Clinic Children's, Cleveland, OH, USA; <sup>3</sup>Department of Paediatric Cardiology, Children's Health Ireland at Crumlin, Dublin, Ireland

**Background and Aim:** Entrustable Professional Activities (EPAs) have become a popular assessment framework for medical trainees and a supplemental component for milestone and competency assessment. EPAs were developed to facilitate assessment of competencies and furthermore translate into clinical practice. Its efficacy in accomplishing these tasks is not well known and is implemented in various forms throughout North American programs (Canada and United States). In this review we explore the rationale for the introduction of EPAs, examine whether they fulfill the promise expected of them, and contemplate further developments in their application with specific reference to training in pediatric cardiology.

**Method:** We conducted a metanarrative review of the literature pertaining to EPAs, pediatric cardiology medical education, and EPAs within pediatric cardiology training through an online search

engines (PubMed, Google Scholar). In total, 46 articles were cited and summarized.

**Results:** Following the outlined themes we described an overarching trend and challenges unique to pediatric cardiology education, how the Covid-19 pandemic affected pediatric cardiology education, and how EPAs could function within these type of programs. We also found the majority of studies published at this juncture focus on the development and implementation of EPAs with no formal studies examining the efficacy of EPAs in transitioning trainees into practice successfully.

**Conclusions:** While EPAs are a very promising assessment framework with its practical approach in more task-oriented evaluations, its long-term utility is not well known. There is reasonable concern that adding another assessment framework could result in more work and resources for programs and trainees without proving EPAs are successful at accomplishing their original purpose. Future studies should be directed at understanding how programs are currently using EPAs and assessing its efficacy amongst trainees.

**Keywords:** medical education, entrustable professional activities, pediatric cardiology, competency assessment

substance abuse, trauma, complications of CHD surgery, among other rare conditions.

**Method:** We present the case of a previously healthy 7-year-old boy, with no relevant cardiovascular familiar history, evaluated in the emergency department because of chest pain. The day before admission, while playing, he suffered a minor chest compression and since then presented retrosternal pain and discomfort. No other symptoms were reported. He was hemodynamically stable and the electrocardiogram (ECG) evidenced major repolarization abnormalities in inferior and left precordial leads. The echocardiogram showed mild global left ventricle dysfunction and a dyskinetic apical interventricular septum, cardiac morphology and coronary arteries origin and initial course were normal. Analytic study showed an isolated, but significant, elevation of cardiac markers. Considering these findings, a cardiac catheterization was promptly performed showing an occlusion of the distal segment of the left descending artery. Cardiac magnetic resonance performed during hospitalization indicated a recent transmural infarction of the apical septal segment. He was discharged after ten days, after a favorable clinical evolution and normalization of the cardiac biomarkers, under anti-aggregation therapy.

Authors	Country	Purpose	Type of source	Major themes
16. Ross, RD, et al. (2015)	USA	SPCTPD/ACC/AAP/AHA training guidelines for pediatric cardiology programs	Original research	Introductory statement by the council created for providing curriculum guidelines for pediatric cardiology fellowships including the introduction of EPAs
4. Frank, L (2017)	USA	Primer on domains, competencies, and EPAs within pediatric cardiology	Original research	A summative study defining and examining the differences between competency assessments, EPAs, and domains within the context of pediatric cardiology versus other subspecialties
15. Srivastava, S, et al. (2017)	USA	Review of curriculum components of EPAs within pediatric cardiology	Original research	General primer on the full-length description of EPAs specifically pertaining to pediatric cardiology as developed by the ABP and CoPS
14. Baffa, JM, et al. (2018)	USA	Assessing graduation readiness using EPAs within pediatric cardiology	Conference abstract / quantitative study	Fellowship program directors in USA surveyed about PC trainees – consensus is that imaging skills and non-ICU level care should yield higher EPA scores than other areas; overall the expectation is that trainees upon graduation are not expected to practice independently based on EPA progress; emphasis on need for support structure/guidance for new graduates
41. Beeri, R, et al. (2020)	Europe	7 <sup>th</sup> ESC Education Conference discussion surrounding EPA use and trusting cardiology trainees	Conference paper	Majority of attendants agreed trainees should be certified only once they can perform clinical duties unsupervised; highlighted importance of providing supervision and mentorship to trainees and identified barriers to providing continuity of assessments/education; 100% thought EPAs were valuable, 69% thought EPAs were applicable to clinical practice
38. Werbo, DK et al. (2022) <sup>33</sup>	USA	Creating EPAs for the pediatric cardiac intensive care unit	Mixed methods study	Creation of 9 EPAs specific to CICU training; next steps include integration into curricula and assessing EPA quality

Table of published papers dedicated to EPAs within pediatric cardiology

### P-315

#### Acute myocardial infarction in a previously healthy child: A case report

Catarina Maria Almeida<sup>1</sup>, Marisa Pereira<sup>1</sup>, Sofia Granja<sup>1</sup>, João Carlos Silva<sup>2</sup>, Jorge Moreira<sup>1</sup>

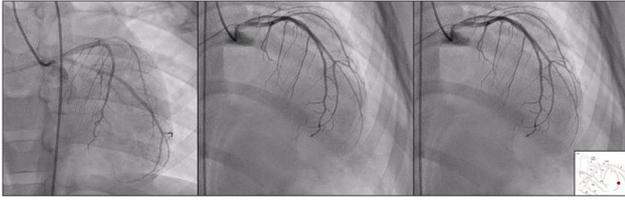
<sup>1</sup>Pediatric Cardiology Department, Centro Hospitalar Universitário de São João, Porto, Portugal; <sup>2</sup>Cardiology Department, Centro Hospitalar Universitário de São João, Porto, Portugal

**Background and Aim:** Acute myocardial infarction (AMI) is a rare life-threatening event in children. Although more frequent in those with congenital heart disease (CHD) and/or abnormal coronary anatomy, it can occur in healthy children. Pediatric AMI etiologies include Kawasaki disease, myocarditis, cardiomyopathy,

**Results:** After a month he kept asymptomatic. On his last visit, the ECG presented changes in repolarization compatible with a subacute myocardial infarction. The echocardiogram showed a hyperrefringency of the distal part of the septum and right ventricle apex and a slight dilatation of the left ventricle apex.

**Conclusions:** Myocardial ischemia and infarction are uncommon in children with normal coronary anatomy. Nevertheless, while facing an acute typical chest pain presentation in a child, this etiology cannot be forgotten. AMI is not a common entity in children. The diagnosis requires a high index of suspicion and should prompt a meticulous cardiac workup.

**Keywords:** pediatric cardiology, acute myocardial infarction, distal left anterior descending artery occlusion



Cardiac catheterization exam showed an occlusion of the distal segment of the left descending artery.

### P-316

#### Speckle tracking echocardiography in healthy newborns and in neonates with coarctation of aorta

Liliana Gozar<sup>1</sup>, Andreea Cerghit Paler<sup>1</sup>, Dorottya Gabor Miklosi<sup>1</sup>, Daniela Toma<sup>1</sup>, Claudiu Marginean<sup>2</sup>, Mihaela Iancu<sup>3</sup>, Amalia Fagarasan<sup>1</sup>

<sup>1</sup>Emergency Institute of Cardiovascular Diseases and Transplantation, Department of Pediatric Cardiology, Târgu-Mureş, Romania, Department of Pediatrics, "George Emil Palade" University of Medicine, Pharmacy, Science, and Technology of Târgu-Mureş, Romania;

<sup>2</sup>Department of Obstetrics and Gynecology, "George Emil Palade" University of Medicine, Pharmacy, Science, and Technology of Târgu-Mureş, Romania; <sup>3</sup>Department of Medical Informatics and Biostatistics, Faculty of Medicine, "Iuliu Hațieganu" University of Medicine and Pharmacy Cluj-Napoca, Romania

**Background and Aim:** Neonatal aortic coarctation (CoA), defined as narrowing of the isthmic aorta, due to its significant contribution to neonatal mortality and morbidity, remains a widely debated topic. Although prenatal diagnosis has helped reduce neonatal mortality, postnatal diagnostics remain difficult, as the diagnostic algorithm lacks parameters for the evaluation of the ventricular function. Data regarding the evaluation of myocardial function by speckle-tracking are limited, but must find its place in the hemodynamic evaluation of fetuses and newborns affected by this pathology. Our aims were to compare speckle-tracking strain values in neonates with CoAo and control group, respectively to establish reference intervals for strain measurement in assessing myocardial function in healthy newborns.

**Method:** Our study consisted of two stages. Firstly, a retrospective single-center study was conducted for a period of 3 years, in which echocardiographic evaluations of all newborns with intrauterine suspicion of CoA were reviewed (41 newborns). These patients were postnatally subdivided: patients with confirmed diagnosis (13 patients) and infirmed (control group, 28 patients). A comparison of peak longitudinal strain (pGLS) was performed between the two groups. In the second stage, conducted for a period of 14 months, with the aim of establishing reference intervals, a total of 103 healthy, full-term newborns were included. In both stages, two-dimensional echocardiographic acquisitions from apical four-chamber view were used and analyzed with the autostrain function offline.

**Results:** We noticed that the overall longitudinal deformation of the left ventricle (LV) pGLS had a lower value in CoA group ( $-13.72 \pm 3.88$ ) than in those with infirmed diagnosis ( $-17.29 \pm 3.83$ ). We found that an optimal cut-off value of  $-16.6\%$  for overall longitudinal deformation of the LVpGLS demonstrated a good ability in discriminating between neonates with CoA and those without CoA, having a sensitivity, specificity, and a positive

likelihood ratio value of 92.31%, 71.43%, and 3.23 with 0.79 area under the curve.

The reference values obtained in the second stage from the healthy group for LVpGLS were between  $[-24.65$  to  $-14.62]$ .

**Conclusions:** The speckle-tracking LVpGLS parameter can aid the differential diagnosis of neonates with CoA from newborns with a false positive intrauterine diagnosis, respectively represents a good alternative for cardiac evaluation of healthy newborns.

**Keywords:** coarctation of aorta, speckle-tracking, myocardial strain, neonate

### P-317

#### Association of gastrointestinal symptoms and presentation and outcomes for kawasaki disease versus multisystem inflammatory syndrome in children

Marianna Fabi<sup>1</sup>, Ashraf S Harahsheh<sup>2</sup>, Geetha Raghuvver<sup>3</sup>, Sundaram Balasubramanian<sup>4</sup>, Simon Lee<sup>5</sup>, Supriya S Jain<sup>6</sup>, Pei Ni Jone<sup>7</sup>, Angela T Yetman<sup>8</sup>, Ana C Barri<sup>9</sup>, Elizabeth Braunlin<sup>10</sup>, Manaswita Khare<sup>11</sup>, Kimberly E Mchugh<sup>12</sup>, Suryakant Shah<sup>13</sup>, Pedrom Farid<sup>14</sup>, Brian W McCrindle<sup>14</sup>

<sup>1</sup>Pediatric Emergency Unit, IRCCS Azienda Ospedaliero-Universitaria di Bologna, Bologna, Italy; <sup>2</sup>Children's National Hospital, The George Washington University School of Medicine and Health Science, Washington, USA; <sup>3</sup>Children's Mercy Hospital, Kansas City, USA; <sup>4</sup>Kanchi Kamakoti CHILDS Trust Hospital, Chennai, India; <sup>5</sup>The Heart Center, Nationwide Children's Hospital, Columbus, USA; <sup>6</sup>New York Medical College, Maria Fareri Children's Hospital, Valhalla, USA; <sup>7</sup>Children's Hospital Colorado, University of Colorado School of Medicine, Aurora, USA; <sup>8</sup>University of Nebraska Medical Center, Children's Hospital and Medical Center, Omaha, USA; <sup>9</sup>Pediatric Cardiology Unit, Department of Pediatrics, Hospital Universitario 12 de Octubre, Madrid, Spain; <sup>10</sup>University of Minnesota Children's Hospital, Minneapolis, USA; <sup>11</sup>Department of Pediatrics, Rady Children's Hospital/US San Diego, San Diego, USA; <sup>12</sup>Medical University of South Carolina, Charleston, USA; <sup>13</sup>Memorial University of Newfoundland, St. John's, Canada; <sup>14</sup>Labatt Family Heart Centre, The Hospital for Sick Children, University of Toronto, Toronto, Canada

**Background and Aim:** Gastrointestinal (GI) symptoms are a common presenting feature of Multisystem Inflammatory Syndrome in Children (MIS-C) associated with COVID-19 and Kawasaki disease (KD), two clinically similar conditions. We sought to compare the association of GI symptoms with demographics, clinical presentation, and outcomes of MIS-C versus KD patients.

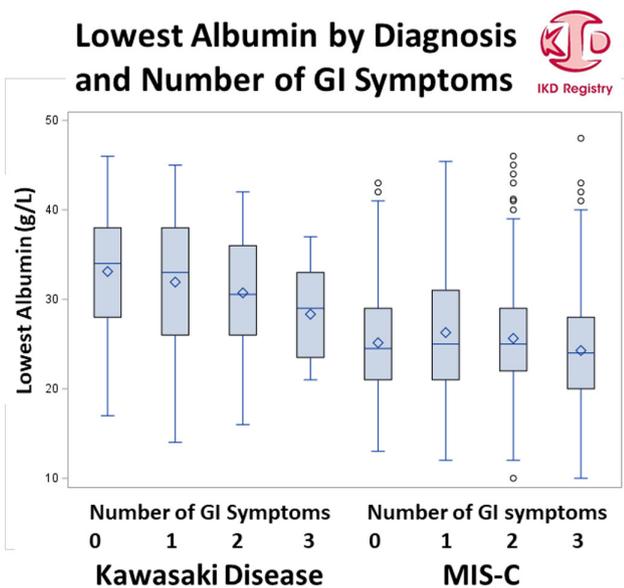
**Method:** The International KD Registry (IKDR) enrolled contemporaneous patients with acute MIS-C or KD from January 2020 to July 2022. Of 1708 presumed MIS-C patients, 1318 were included who met strict US Centers for Disease Control case definition criteria and had documentation of prior COVID-19 or exposure. Of 904 presumed KD patients, 387 were included who met American Heart Association diagnostic criteria and were confirmed not to have prior COVID-19. The association of number of GI symptoms of abdominal pain, diarrhea and vomiting at presentation (0-3) with diagnosis, clinical features and outcomes was determined using general linear regression models.

**Results:** When compared to KD, MIS-C patients had both significantly greater individual GI symptoms (abdominal pain, 67% vs. 21% respectively,  $p < 0.001$ ; diarrhea, 50% vs. 30%,  $p < 0.001$ ; and vomiting, 65% vs. 39%,  $p < 0.001$ ) and a greater number of symptoms ( $p < 0.001$ ). Greater number of GI symptoms was not associated with sex, but was significantly associated with older age and greater adiposity for both MIS-C and KD. Shock and ICU admission were more likely for MIS-C, and more likely with greater number of GI symptoms in both groups. Time from

admission to first IVIG treatment was longer with more GI symptoms for KD patients only. Likewise, lowest albumin was lower for MIS-C but only associated with greater GI symptoms for KD patients (FIGURE). Higher peak NTproBNP and Troponin I levels and lower worst LV ejection fraction were noted for MIS-C and also related to greater number of GI symptoms. Peak coronary artery Z score was greater for KD with no association with number of GI symptoms.

**Conclusions:** Greater number of GI symptoms is associated with delayed treatment for KD but not MIS-C patients. GI symptoms are likely a marker of more severe disease, particularly for MIS-C, and are associated with worse ventricular function but not with coronary artery involvement.

**Keywords:** Kawasaki disease, Multisystem inflammatory syndrome in children, ventricular function, coronary artery



**P-321**

**Electrocardiographic characteristics of neonates with atrial septal defects**

Anna Maria Dehm<sup>1</sup>, Maria Munk Pærregaard<sup>2</sup>, Anna Sellmer<sup>1</sup>, Sofie Dannesbo<sup>3</sup>, Elisabeth Blixenkroner Møller<sup>2</sup>, Anne Sophie Sillesen<sup>2</sup>, Anna Axelsson Raja<sup>3</sup>, Kasper Karmark Iversen<sup>2</sup>, Henning Bundgaard<sup>3</sup>, Alex Hørby Christensen<sup>3</sup>, Vibeke Hjortdal<sup>1</sup>

<sup>1</sup>Department of Cardiothoracic Surgery, The Heart Center, Rigshospitalet, Copenhagen University Hospital, Copenhagen, Denmark; <sup>2</sup>Department of Cardiology, Herlev-Gentofte Hospital, Copenhagen University Hospital, Copenhagen, Denmark; <sup>3</sup>Department of Cardiology, The Heart Center, Rigshospitalet, Copenhagen University Hospital, Copenhagen, Denmark

**Background and Aim:** Arrhythmias and electrocardiographic (ECG) abnormalities are common among patients with atrial septal defects (ASDs). We studied a large cohort of neonates with ASDs to investigate whether ECG abnormalities are present at this early stage or develop later, secondary to hemodynamic changes.

**Method:** We analyzed echocardiograms and ECGs from the Copenhagen Baby Heart Study; a multicenter, population-based

cohort study. We included 438 neonates with secundum ASDs. ECG characteristics in neonates with ASD were compared to 1,314 matched controls. In subgroup analyses we investigated if electrocardiographic characteristics were associated with age at examination.

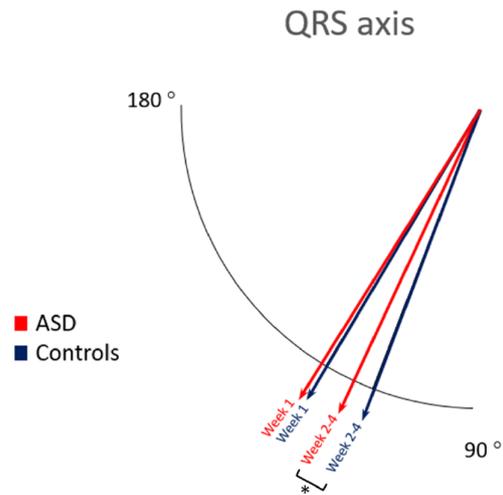
**Results:** Neonates with an ASD (median age 11 days, 51% boys) had longer P-wave durations (58 vs. 56 ms,  $p = 0.000057$ ), PR intervals (100 vs. 96 ms,  $p = 0.000027$ ) and a more rightward shifted QRS axis (116 vs. 114 degrees,  $p = 0.032$ ) than controls (median age 10 days, 51% boys). There were no differences between cases and controls in the P-wave area, amplitude, or axis. Subgroup analyses showed that the differences in P-wave duration and PR interval were present in neonates examined in the first week after birth. The difference in QRS axis was not found in neonates examined this early but was found in neonates examined at age two to four weeks.

**Conclusions:** ASDs are associated with ECG changes from the neonatal phase. Prolongation of P-wave duration and PR interval are found as early as the first week after birth, indicating that these changes are not purely secondary, but that neonates with an ASD have altered cardiac electrical activity.

**Keywords:** Atrial septal defect, electrocardiography, echocardiography, neonates

**The QRS axis for neonates with atrial septal defects and controls, divided in subgroups by age at examination.**

The figure illustrates the gradual leftward shift of the QRS axis from week 1 to week 2-4 after birth. Arrows depict median values for the QRS axis in neonates with an ASD (red arrows) and controls (blue arrows). In week 1 after birth, both newborns with ASD and controls have a QRS axis that is physiologically shifted rightwards. During week 2-4 after birth, the QRS axis of neonates with ASD stays significantly more rightwards shifted than the QRS axis of controls. \* =  $p < 0.05$



**P-322**

**Myokines predicted heart failure with preserved ejection fraction in type 2 diabetes mellitus patients**

Alexander A Berezin<sup>1</sup>, Ivan M Fushchey<sup>1</sup>, Alexander E Berezin<sup>2</sup>  
<sup>1</sup>Zaporozhye Medical Academy of Postgraduating Education, Internal Medicine Department; <sup>2</sup>Zaporozhye State Medical University, Internal Medicine Department

**Background and Aim:** The aim of the study was to investigate whether serum levels of both irisin and apelin predict HF with preserved ejection fraction (HFpEF) in patients with T2DM.

**Method:** One hundred and eight HF patients with T2DM having HFpEF (EF>50%; n = 58), HF with mildly reduced ejection fraction (HFmrEF, EF<40%; n = 22), HF with reduced ejection fraction (HFrEF, EF<40%; n = 28) aged from 41 to 62 years and 20 non-HF patients with T2DM. Healthy control group was consisted of 25 individuals matched with age and sex.

**Results:** The levels of irisin were significantly higher in HFpEF patients than in HFrEF individuals, whereas healthy volunteers and T2DM non-HF patients demonstrated lower concentrations of these peptides. Apelin levels were significantly increased in HFrEF. There were not significant differences between these biomarkers in HFrEF and HFmrEF. We revealed that cut-off points for irisin and apelin that distinguished HFpEF from HFrEF/HFmrEF were (6.50 ng/mL; AUC = 0.78; 95% confidence interval [CI] = 6.85 - 10.66 ng/mL and 4.12 ng/mL, AUC = 0.72; 95% CI = 3.90-5.75 ng/mL, respectively). Then we divided all patients with HF having elevation of NT-proBNP > 750 pmol/mL into three subgroups depending on the biomarkers' levels. Patients from subgroup A had both irisin and apelin levels higher cut-off points, individuals from group B had higher concentration of one of two biomarkers, and patients from subgroup C demonstrated levels of both peptides lower cut-off points. Multivariate logistic regression analysis revealed that discriminative value of irisin and apelin to predict HFpEF in subgroup B (Odds Ratio [OR] = 2.18; 95% CI = 1.26-3.14; P = 0.001) were substantially higher compared with subgroups A and C (OR = 1.03; 95% CI = 1.00-1.05; P = 0.64 and OR = 0.92; 95% CI = 0.89-1.01; P = 0.62, respectively).

**Conclusions:** We found that multidirectional changes in the levels of irisin and apelin along with increased levels of NT-proBNP in T2DM patients had better predictive value for HFpEF than any changes in the circulating levels of each peptide alone.

**Keywords:** Heart failure; diabetes mellitus; irisin, apelin, natriuretic peptides

### P-323

#### Clinical profile of congenital heart disease and outcome after surgical treatment: A single-center experience in a developing country

Ria Nova<sup>1</sup>, Bermansyah Boestham<sup>2</sup>, Denny Salverra Yosy<sup>1</sup>, Gama Satria<sup>2</sup>, Ahmat Umar<sup>2</sup>, Aswin Nugraha<sup>2</sup>, Arie Hasiolan<sup>2</sup>

<sup>1</sup>Department of Child Health, Division of Pediatric Cardiology, Dr.Moh. Hoesin Hospital, Universitas Sriwijaya, Palembang, Indonesia;

<sup>2</sup>Department of Surgery, Division of Thoracic and Cardiovascular, Dr.Moh. Hoesin Hospital, Universitas Sriwijaya, Palembang, Indonesia

**Background and Aim:** Mortality rates after cardiac surgery in children with congenital heart disease (CHD) differ markedly depending on the severity of cases, comorbidities, and postoperative care. This study aimed to evaluate CHD's clinical profile and surgical therapy outcome in our hospital.

**Method:** This retrospective data were collected from medical records of children who underwent CHD surgery in the Moh Husin Hospital Palembang, Indonesia, between 2019 and 2021. The primary variable analyzed was in-hospital mortality and the distribution of mortality by age, nutritional state, heart failure, pulmonary hypertension, duration of CPB, aortic clamp time, and high inotropic support.

**Results:** There were 64 cases of CHD undergoing surgical treatment. In total, nine patients had ASD, 36 had VSD, ten had PDA, and nine had TOF. Outcomes of surgery were 82.8% alive, and 17.2% died during a hospital stay. Of the patients who died, there were 90% over one year, 63.6% were underweight, 81.8% were heart failure, 81.8% were pulmonary hypertension, aortic

clamps for more than 30 minutes, CPB time of more than 60 minutes, and using three inotropic drugs were 75%.

**Conclusions:** The mortality rate is still high related to being underweight, having heart failure, pulmonary hypertension, longer duration of aortic clamps and CPB time, and high inotropic support.

**Keywords:** Clinical profile, congenital heart disease, surgical treatment, outcome

### P-324

#### Long-term prophylactic effect of angiotensin receptor blockers in children with genetic aortopathies: early bird catches the worm

Johanna Josephine Kanitz, Jakob Olffe, Veronika Stark, Fridrike Stute, Daniel Biermann, Rainer Gerhard Kozlik-Feldmann, Michael Hübler, Thomas Mir

Department of Pediatric Cardiology, University Heart & Vascular Center Hamburg, University Medical Center Hamburg-Eppendorf, Hamburg, Germany

**Background and Aim:** Genetic aortopathies (GA) manifest with various symptoms. Particular attention is paid to aortic root dilatation which has impact on morbidity and mortality. Some trials have been conducted in children and showed similar effectiveness of angiotensin receptor blockers (ARB) and beta blockers (BB). This study focuses on long term effects of therapy with ARB or BB on aortic root growth and the question which of the drugs to choose as first line therapy at different ages.

**Method:** Since 1998 we diagnosed 203 patients with GA (165 FBN1, 19 LDS, 19 others). 82 patients between 3 mth and 16 yrs receiving either ARB or BB therapy were included. We retrospectively analyzed the progression of the dilatation of Sinus Valsalva (SV) using calculated z-scores before and after therapy initiation and compared BB and ARB treatment.

**Results:** This study proved that both ARB and BB (p<0.05) therapy showed significant benefit by reducing the growth of the aortic root, while the effect is significantly more pronounced in ARB (p<0.01) independent of age and genetic causes. A detailed comparison of the two drug groups showed a more sustained effect in limiting the progression of the dilatation of the aortic root in patients treated with ARB. Long term progression of dilatation of the SV was significantly lower in children treated with ARBs compared to BB (delta z-score, p<0.05). In addition, ARB were better tolerated and had a significant lower discontinuation rate (3%) compared to betablockers (53%) (p<0.001). Independently of age at initiation of prophylaxis all children and adolescents were able to reach the target dose under ARB.

**Conclusions:** In our cohort we demonstrate a significant benefit with both treatment options, but the effect was more pronounced and longer lasting in ARB. In contrast to betablockers a very good tolerance of the medication was observed in ARB making it our therapy of choice in children. As a next step it would be important to define what dose to start with and which target dose to reach. At this point, it is no longer a question of which medication we use but in which dosage and at what time.

**Keywords:** Marfan Syndrom, Sartan, Beta-Blocker

### P-325

#### Head circumference in neonates with atrial septal defect

Line Høffner<sup>1</sup>, Anna Maria Dehn<sup>1</sup>, Sofie Dannesbo<sup>2</sup>, Elisabeth Blixenkroner Møller<sup>2</sup>, Anna Axelsson Raja<sup>3</sup>, Anne Sophie Sillesen<sup>2</sup>, Kasper Iversen<sup>2</sup>, Henning Bundgaard<sup>3</sup>, Vibeke Hjortdal<sup>1</sup>

<sup>1</sup>Department Of Cardiothoracic Surgery, Copenhagen University Hospital Rigshospitalet; <sup>2</sup>Department of Cardiology, Herlev-Gentofte Hospital, Copenhagen University Hospital; <sup>3</sup>Department of Cardiology, The Heart Center, Rigshospitalet, Copenhagen University Hospital

**Background and Aim:** Neurodevelopmental disorders occur in up to 50% of children with congenital heart disease (CHD) including patients with simple cardiac defects. Small head circumference (HC) at birth is a risk factor for impaired neurodevelopment in children with CHD and brain size is smaller in infants with more complex CHD. The objective of this study is to investigate if the head circumference in neonates with a secundum atrial septal defect (ASD) is different from the HC in neonates without an ASD.

**Method:** This study is part of the Copenhagen Baby Heart Study (CBHS); a prospective, population-based cohort study including neonates (n>25, 000) born at the three largest maternity wards in Copenhagen, Denmark in the period April 1st, 2016 to October 31st, 2018. Included neonates were examined with a transthoracic echocardiogram within the first 30 days after birth. Echocardiograms were analyzed for interatrial communications using a published algorithm. We excluded neonates with other cardiac malformations and chromosomal anomalies as well as neonates born preterm. The HC of neonates with a secundum ASD was compared to the HC of neonates without an ASD from the same birth cohort.

**Results:** We investigated the HC at birth for 673 neonates with secundum ASD (47% male, median gestational age (GA) 281 days; mean weight 3531 g ± 470, 2; mean length 51.6 cm ± 2.1) and compared it to 11, 107 neonates without ASD (53% male; median GA 282 days; mean weight 3538 g ± 482, 7; mean length 51.7 cm ± 2.2). In neonates with an ASD, the mean HC was 34.78 cm ± 1.5, and neonates without an ASD had a mean HC of 34.72 cm ± 1.6 (difference = 0.06 cm, 95%CI = -0.06-0.19, p-value: 0.34). We also investigated the relation between birth weight and HC (BW/HC ratio). In neonates with an ASD, mean BW/HC ratio was 101.31 g/cm ± 11.7, whereas neonates without an ASD had a mean BW/HC ratio of 101.74 g/cm ± 11.7 (difference = 0.43 cm, 95%CI = -1.34-0.48, p-value: 0.36).

**Conclusions:** The head circumference at birth in term neonates with ASD does not differ from the head circumference in neonates without ASD.

**Keywords:** Head circumference, Congenital heart disease., Atrial septal defect, Neurodevelopment.

### P-326

#### Successful treatment of cardiac rhabdomyomas using sirolimus; with the challenges of fluctuating levels due to theorised metabolic pathway development

Teresa Jean Brooks<sup>1</sup>, Antigoni Deri<sup>2</sup>, Sarah Joanne Rebecca Whiteley<sup>1</sup>  
<sup>1</sup>Department of Pharmacy, Leeds Children's Hospital; <sup>2</sup>Department of Congenital Cardiology, Leeds Children's Hospital

**Background and Aim:** Sirolimus (mTOR inhibitor) has been described to play a significant role in promoting the natural regression of cardiac rhabdomyomas. Sirolimus is unlicensed in this indication/cohort and dosing experience is limited. We describe the use and monitoring of sirolimus in a term baby with multiple, extensive rhabdomyomas causing outflow tract obstruction and significant arrhythmias. Due to their extent and distribution, these were inoperable and sirolimus was used as an alternative.

**Method:** Dosing of 1mg/m<sup>2</sup> daily with a target blood level of 5-15ng/mL was used based on the prospective cohort study (1). Sirolimus levels and full blood count (FBC) were checked every

few days until the levels were in target range (based on rejection data) and every 2 weeks thereafter. Echocardiogram and ambulatory Electrocardiogram (ECG) were performed at monthly intervals to monitor the cardiac tumours and arrhythmias.

**Results:** See from the graph the changes to sirolimus levels and dosing as the child develops from a neonate to an infant:

Mild neutropenia (1.33 10<sup>9</sup>/L) was observed with high levels of sirolimus, but resolved when sirolimus level returned to the target range.

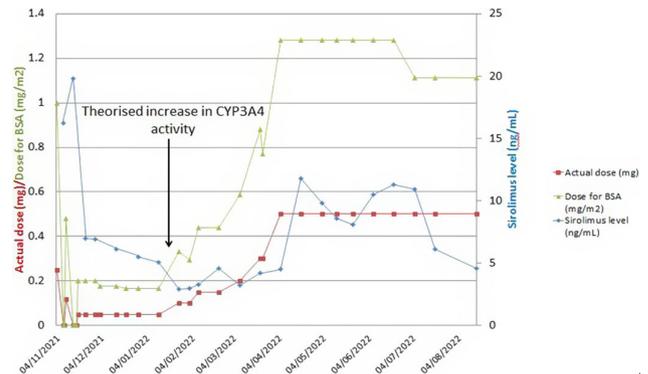
Sirolimus assisted in >50% tumour debulking by 2 months and complete resolution of tumours and arrhythmias by 8 months of age, even with out of target range levels.

**Conclusions:** The large variation of sirolimus dosing and levels is potentially due to theorised metabolic pathway maturing (CYP3A4) during the neonatal to infant period, causing an increased clearance rate [2]. There is a large increase in activity of CYP3A4 during the first 3 months of life, with nearly no activity during the first few days [3] which may have contributed to the initial high sirolimus levels observed and then the long period of suboptimal levels, resulting in the significant increase of dose. Interestingly, even at suboptimal target levels sirolimus assisted in tumour and arrhythmia resolution.

Sirolimus treatment can be very successful in resolution of cardiac rhabdomyomas. Close blood level monitoring is required for neonates and infants due to the development of metabolic pathways. Future work would be required to ascertain the ideal target range of sirolimus blood levels in this indication.

**Keywords:** sirolimus, cardiac rhabdomyoma

Graph to show changes of dosage and levels overtime



See from the graph the changes to sirolimus levels and dosing as the child develops from a neonate to an infant.

### P-328

#### Utilising the modified ross score in children with single ventricle physiology-is it clinically relevant?

Claire McGinn<sup>1</sup>, Louise Morrison<sup>2</sup>, Chris Watson<sup>1</sup>, Frank Casey<sup>1</sup>  
<sup>1</sup>Wellcome-Wolfson Institute for Experimental Medicine, Queen's University Belfast.; <sup>2</sup>Royal Belfast Hospital for Sick Children, Belfast.

**Background and Aim:** Paediatric single ventricle patients are at risk of ventricular dysfunction. The Modified Ross Score has been designed to describe severity of paediatric heart failure but is not widely used in routine practice. It considers the influence of age, feeding, growth, heart rate, respiratory rate, hepatomegaly,

NT-proBNP and echo parameters of ejection fraction and AV insufficiency.

We hypothesised that calculating the Modified Ross score for children with single ventricle physiology may be useful in clinical care for assessment of heart failure.

**Method:** Children aged 2 months to 15 years with a diagnosis of single ventricle physiology were recruited. All participants had a detailed echocardiogram focusing on measures of systolic and diastolic function including fractional area change, myocardial performance index, global longitudinal strain and tissue doppler indices. They also completed a Modified Ross Score. Not all patients had NT-proBNP routinely measured.

**Results:** Fifty-eight children participated; all were outpatients. Two patients were on the cardiac transplant waiting list. 81% (N = 47) of patients had undergone their TCPC. 84% (N = 49) of patients were on ACE inhibitors and 55% (N = 32) were receiving at least once daily diuretics. On echocardiogram 30% (N = 18) of patients had mild AV valve regurgitation, 12% (N = 7) had moderate AV valve regurgitation and 1 patient had severe AV valve regurgitation. 23% (N = 17) of patients had at least mildly reduced systolic ventricular function. Using the Modified Ross Score 15% (N = 9) of patients had evidence of mild heart failure but none had moderate or severe heart failure.

**Conclusions:** This study suggests the Modified Ross Score may not fully correlate with clinical evidence of paediatric heart failure. Despite 23% of this cohort having at least mild ventricular dysfunction on echocardiogram, only 15% met the criteria for mild heart failure using the score. The two patients on the cardiac transplant list were both classed as having only mild heart failure using the score. The study highlights that the score doesn't include medications or activity levels and is influenced by parent and clinician subjectivity. A limitation of this study was lack of NT-proBNP assays for all patients so further correlation between Modified Ross Score, NT-proBNP and ventricular function may be useful.

**Keywords:** Heart Failure, Fontan, Modified Ross Score

### P-329

#### Treatment individualization in low weight birth patients with transposition of great arteries and intact ventricular septum: two-stage approach

Mihaela Bădulescu<sup>1</sup>, Sorina Ana Pasc<sup>1</sup>, Lacramioara Eliza Chiperi<sup>1</sup>, Cristina Flavia Al Akel<sup>2</sup>, Daniela Toma<sup>3</sup>

<sup>1</sup>Department of Pediatric Cardiology, Emergency Institute for Cardiovascular Diseases and Transplantation, Targu Mures, Romania;

<sup>2</sup>Department of Pediatric Cardiology, Emergency Institute of Cardiovascular Disease and Transplantation, Targu-Mures, Romania; Department of Pathophysiology, University of Medicine, Pharmacy, Sciences and Technology of Targu Mures, Romania; <sup>3</sup>Department of Pediatric Cardiology, Emergency Institute of Cardiovascular Disease and Transplantation, Targu-Mures, Romania; Department of Pediatrics, University of Medicine, Pharmacy, Sciences and Technology of Targu Mures, Romania

**Background and Aim:** Management of patients with transposition of great arteries with intact ventricular septum (TGA-IVS) is challenging in low birth weight (LBW) patients in which the timing for surgical treatment is debatable due to technical difficulties and a high risk for irreversible decompensation of the left ventricular (LV) function. This case presentation debates the optimal timing for arterial switch operation (ASO) in a newborn with LBW and the need for staged approach.

**Method:** We present a case of a newborn with LBW diagnosed postnatally with TGA-IVS and an unrestricted atrial septal defect (ASD) initially, in which patent ductus arteriosus (PDA) was

maintained with prostaglandin. In the fifth day of life, due to an ASD which became restrictive, we performed atrioseptectomy and prostaglandin was weaned. Considering the LBW of 2.480gr, efficient intracardiac mixing, preserved LV systolic function, hemodynamic stability, ASO was planned for around 10 days of life. However, the daily echocardiographic evaluation highlighted, starting with the ninth day, dilatation of right cavities, deconditioning of the systolic-diastolic function of the LV with banana appearance, a free wall thickness of 3mm and decreased LV indexed mass of 24g/m<sup>2</sup>.

**Results:** In this context, a two-stage approach was adopted. In the twelfth day of life BAP was performed combined with prostaglandin for maintaining the PDA. The patient presented contractile dysfunction for which inotropic support was used and supraventricular dysrhythmia for which antiarrhythmics were initiated. In evolution, gradual normalization of the LV systolic function was observed with an increase in LV indexed mass to 70.4gr/m<sup>2</sup> and free wall thickness of 5mm. In the eighth day after BAP, debanding and ASO were performed, necessitating mechanical ventilation and open sternum for three days. A severe LV systolic dysfunction was described after surgery for which he received inotropic support with gradual resumption of biventricular systolic function which allowed the discharge of the patient at 33 days of life.

**Conclusions:** In LWB patients, is necessary to individualize surgical correction, keeping in mind the possibility of two-stage surgical approach, with BAP and then ASO, in order to allow the patient to gain weight and also to improve systolic function of systemic ventricle when needed.

**Keywords:** transposition of the great arteries, pulmonary artery banding, low birth weight, two-stage approach, arterial switch operation

### P-330

#### Cardiac involvement across the diagnostic spectrum from kawasaki disease to multisystem inflammatory syndrome in children associated with COVID-19

Nilanjana Misra<sup>1</sup>, Luis Martin Garrido-Garcia<sup>2</sup>, Mona Mostafa El-Ganzoury<sup>3</sup>, Aishwarya Venkataraman<sup>4</sup>, Deepa Prasad<sup>5</sup>, Elif Seda Selamet Tierney<sup>6</sup>, Deepika Thacker<sup>7</sup>, Mark D Hicar<sup>8</sup>, Tyler H Harris<sup>9</sup>, Matthew D Elias<sup>10</sup>, Daniel Mauriello<sup>11</sup>, William B Orr<sup>12</sup>, Cedric Manlhiot<sup>13</sup>, Pedrom Farid<sup>14</sup>, Brian W McCrindle<sup>14</sup>

<sup>1</sup>Cohen Children's Medical Center of New York, Northwell Health, New York, USA; <sup>2</sup>Hospital Angeles Lomas, Huixquilucan, Mexico; <sup>3</sup>Ain Shams University, Cairo, Egypt; <sup>4</sup>Kanchi Kamakoti CHILS Trust Hospital, Chennai, India; <sup>5</sup>Banner Children's at Desert Medical Center, Mesa, USA; <sup>6</sup>Stanford University, Stanford, USA; <sup>7</sup>Nemours Children's Health, Wilmington, USA; <sup>8</sup>Department of Pediatrics, Jacobs School of Medicine and Biomedical Sciences, University at Buffalo, Buffalo, USA; <sup>9</sup>UPMC Children's Hospital of Pittsburgh, University of Pittsburgh School of Medicine, Pittsburgh, USA; <sup>10</sup>Division of Cardiology, Children's Hospital of Philadelphia, Philadelphia, USA; <sup>11</sup>Johns Hopkins All Children's Hospital, Division of Pediatric Cardiology, St. Petersburg, USA; <sup>12</sup>Washington University School of Medicine, St. Louis, USA; <sup>13</sup>Department of Pediatrics, Johns Hopkins School of Medicine, Baltimore, USA; <sup>14</sup>Labatt Family Heart Centre, The Hospital for Sick Children, University of Toronto, Toronto, Canada

**Background and Aim:** Case definitions for Kawasaki disease (KD) and Multisystem Inflammatory Syndrome in Children associated with COVID-19 (MIS-C) show considerable overlap, creating the potential for misclassification. We sought to define the spectrum of classification of KD and MIS-C patients, and its association with cardiac involvement.

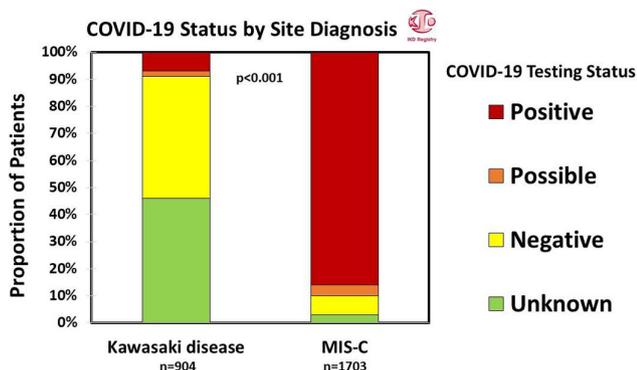
**Method:** The International KD Registry (IKDR) enrolled contemporaneous patients with clinical features of either acute MIS-C or KD during the course of the pandemic. Patients were classified by site diagnosis, American Heart Association (AHA) case definition for KD, US Centers for Disease Control (CDC) case definition for MIS-C, and COVID-19 status (Unknown, Negative, Possible, Positive, based on testing and exposure status). Associations of classification type with lowest LV ejection fraction a maximum coronary artery Z score and were explored using general linear regression modeling.

**Results:** From January 2020 to July 2022, 904 patients with a site diagnosis of KD were enrolled, of which 49% met AHA KD criteria only and 49% met both AHA KD and CDC MIS-C criteria. Of 1708 patients enrolled with a site diagnosis of MIS-C, 7% met criteria for AHA KD only and 40% for CDC MIS-C only, 6% did not meet criteria for either, and 47% met criteria for both. COVID-19 status by site diagnosis is shown in FIGURE. From regression modeling, lower LV ejection fraction was independently associated with COVID-19 status (Unknown Parameter Estimate [PE] 0, Negative 0.84, Possible 0.93, Positive -1.15;  $p = 0.02$ ), site diagnosis of MIS-C (PE -5.23;  $p < 0.001$ ) and CDC MIS-C diagnosis (PE -1.64;  $p = 0.003$ ). After controlling for these 3 classification types, AHA KD diagnosis was not significantly associated ( $p = 0.53$ ). Higher maximum coronary artery Z score was independently associated with site diagnosis of KD (PE 0.64;  $p < 0.001$ ), AHA diagnosis of KD (PE 0.28;  $p = 0.02$ ) and CDC diagnosis of MIS-C (PE 0.39;  $p < 0.001$ ). After controlling for these 3 classification types, COVID-19 status was not significantly associated (0.23).

**Conclusions:** Classification of patients using multiple methods is predictive of cardiac involvement with some variation. MIS-C and positive COVID-19 status but not AHA KD diagnosis are associated with ventricular dysfunction. Both MIS-C and KD but not COVID-19 status are associated with coronary artery involvement.

**Keywords:** Kawasaki disease, Multisystem Inflammatory Syndrome in Children, ventricular function, coronary artery

#### Site Diagnosis by COVID Testing Status



#### P-331

##### Association of sex and age with presentation and outcomes of kawasaki disease versus multisystem inflammatory syndrome in children

Nagib Dahdah<sup>1</sup>, Michael A Portman<sup>2</sup>, Michael Khoury<sup>3</sup>, Audrey Dionne<sup>4</sup>, Arash A Sabati<sup>5</sup>, Nadine F Choueiter<sup>6</sup>, Benjamin T Barnes<sup>7</sup>,

Tapas Mondal<sup>8</sup>, Robert Lowndes<sup>9</sup>, Geetha Raghuveer<sup>10</sup>, P Sulochana Putli Bai<sup>11</sup>, Jacqueline R Szmuszkovicz<sup>12</sup>, Dongngan T Truong<sup>13</sup>, Frederic Dallaire<sup>14</sup>, Kambiz Norozi<sup>15</sup>, Shae Merves<sup>16</sup>, Pedrom Farid<sup>17</sup>, Brian W McCrindle<sup>17</sup>

<sup>1</sup>Division of Pediatric Cardiology, CHU Ste-Justin, Universite de Montreal, Montreal, Canada; <sup>2</sup>Seattle Children's Hospital, Seattle, USA; <sup>3</sup>Stollery Children's Hospital, University of Alberta, Edmonton, Canada; <sup>4</sup>Boston Children's Hospital, Harvard University, Boston, USA; <sup>5</sup>Phoenix Children's Hospital, Phoenix, USA; <sup>6</sup>Children's Hospital at Montefiore, Albert Einstein College of Medicine, Bronx, USA; <sup>7</sup>Department of Pediatrics, Johns Hopkins University, Baltimore, USA; <sup>8</sup>McMaster Children's Hospital, Hamilton, Canada; <sup>9</sup>Children's National Hospital, George Washington University School of Medicine and Health Sciences, Washington, USA; <sup>10</sup>Mercy Children's Hospital, Kansas City, USA; <sup>11</sup>Kanchi Kamakoti CHILDS Trust Hospital, Chennai, India; <sup>12</sup>Children's Hospital of Los Angeles, University of Southern California, Los Angeles, USA; <sup>13</sup>Primary Children's Hospital, University of Utah, Salt Lake City, USA; <sup>14</sup>Centre de recherche du Centre hospitalier universitaire de Sherbrooke, Sherbrooke, Canada; <sup>15</sup>Children's Hospital of Western Ontario, Western University, London, Canada; <sup>16</sup>Arkansas Children's Hospital, university of Arkansas for Medical Sciences, Little Rock, USA; <sup>17</sup>Labatt Family Heart Centre, The Hospital for Sick Children, University of Toronto, Toronto, Canada

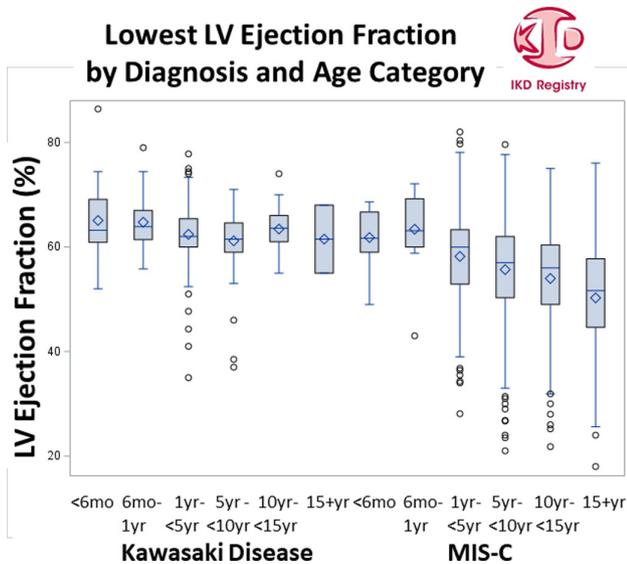
**Background and Aim:** Kawasaki disease (KD) and Multisystem Inflammatory Syndrome in Children associated with COVID-19 (MIS-C) both have a male preponderance but different age profile. We sought to determine associations of sex and age with clinical features and outcomes for KD versus MIS-C.

**Method:** The International KD Registry (IKDR) enrolled contemporaneous patients with clinical features of either acute MIS-C or KD from January 2020 to July 2022. For this analysis, of 1708 presumed MIS-C patients, 1318 were included who met strict US Centers for Disease Control case definition criteria and had documentation of prior COVID-19 or exposure. Of 904 presumed KD patients, 387 were included who met American Heart Association diagnostic criteria and were confirmed not to have prior COVID-19. Associations of sex and age with clinical features and outcomes for KD vs. MIS-C were explored using general linear regression modeling.

**Results:** MIS-C and KD groups had a similar male preponderance (62% vs 61%, respectively;  $p = 0.69$ ), but MIS-C patients were significantly older (median 8.6 vs 2.6 years;  $p < 0.001$ ). Presentation with shock was rare for KD (3% vs. 35%;  $p < 0.001$ ), but for MIS-C shock was associated with older age ( $p < 0.001$ ) but not sex. ICU admission, more common for MIS-C (59% vs 6%;  $p < 0.001$ ), was not associated with sex, but had a U-shaped association with age ( $p < 0.001$ ; higher for both younger and older patients) for KD and MIS-C. C-reactive protein and ferritin levels were higher with MIS-C, not related to sex, and were higher for both younger and older patients. Peak creatinine was higher with MIS-C, not related to sex, and higher for older patients. Both peak NTproBNP and troponin I were higher for MIS-C, with higher NTproBNP in females, and higher troponin I with older age. LV ejection fraction was lower with MIS-C, not related to sex but lower with older age only for MIS-C (FIGURE). Maximum coronary artery Z score was higher for KD, not related to sex, and decreased with older age for KD only.

**Conclusions:** There were few associations for clinical features and outcomes with sex, and associations with age, while often present, existed in linear and non-linear patterns and were specific to diagnosis.

**Keywords:** Kawasaki disease, multisystem inflammatory syndrome in children, ventricular function, coronary artery



LVEF by Diagnosis and Age Category

**P-332****Left and right ventricular dysfunction in propionic acidemia – preliminary results of a single-center cross-sectional study**

Alexander Kovacevic<sup>1</sup>, Sven F Garbade<sup>2</sup>, Friederike Hörster<sup>2</sup>, Georg F Hoffmann<sup>2</sup>, Matthias Gorenflo<sup>1</sup>, Derliz Mereles<sup>3</sup>, Stefan Kölker<sup>2</sup>, Christian Stauffer<sup>2</sup>

<sup>1</sup>Department of Pediatric and Congenital Cardiology, Heidelberg University Hospital, Heidelberg, Germany; <sup>2</sup>Department of General Pediatrics, Division of Neuropediatrics and Metabolic Medicine, Heidelberg University Hospital, Heidelberg, Germany; <sup>3</sup>Department of Cardiology, Angiology and Pulmology, Heidelberg University Hospital, Heidelberg, Germany

**Background and Aim:** Propionic acidemia (PA) is an organic aciduria caused by deficiency of propionyl-CoA carboxylase. Besides e.g., progressive neuromuscular disease, left ventricular (LV) dysfunction is common, leading to cardiomyopathy or acute heart failure, which strongly contributes to mortality in PA. We sought to assess echocardiographic parameters of left and right ventricular (RV) systolic and diastolic function that indicate early signs of cardiac disease manifestation in PA.

**Method:** This is a cross-sectional single-center study conducted at a Tertiary Medical Care Center. Systolic and diastolic functional parameters of the LV and RV were assessed by echocardiography: LV fractional shortening (FS), LV ejection fraction (EF), mitral annular plane systolic excursion (MAPSE), LV-global longitudinal strain (GLS), mitral valve (MV) E/A ratio, MV E/e', LV-myocardial performance index (LV-MPI), MV deceleration time (DT-E), tricuspid annular plane systolic excursion (TAPSE), RV-global longitudinal strain (GLS)/-free wall strain (FWS), RV-fractional area of change (FAC), tricuspid valve (TV) S', RV-myocardial performance index (RV-MPI), TV E/A, TV e'/a', and TV E/e'. Besides descriptive analyses we assessed frequency, onset, and combinations of echocardiographic parameters.

**Results:** N = 18 patients with PA were enrolled (mean age at assessment 13.1 years). Abnormal parameters were LV-GLS (72.2%) LV-EF (61.1%), MAPSE (50%), MV E/e' (44.4%), LV-MPI (33.3%), LV-FS (33.3%), MV E/A (27.8%), TV S' (16.7%), TAPSE (11.1%), RV-GLS/FWS (11.1%), TV E/A (16.7%), and

TV E/e' (33.3%). The most prevalent combinations of pathological parameters were MAPSE+LV-GLS, LV-EF+LV-GLS, TV E/A+TV E/e', and TAPSE+RV S'. With age, the probability of developing abnormal LV/RV function increases.

**Conclusions:** We demonstrate a high rate of cardiac disease manifestation in PA, higher than in all previous studies regarding LV dysfunction in PA, where only LV-FS was measured (FS abnormal in 9–40%). LV-GLS seems to be a robust parameter to indicate early cardiac disease manifestation, which is shown here for the first time in literature for this disease. In particular systolic LV dysfunction is more often found than RV dysfunction. It remains to be evaluated, whether earlier detection of cardiac disease in PA leads to improved outcome, e.g., by earlier implementation of cardiac therapies or liver transplantation, which can reverse cardiomyopathy in PA.

**Keywords:** propionic acidemia, metabolic cardiomyopathy, left ventricular function, right ventricular function

**P-333****Modified right ventricular stroke work index for predicting MID- and long-term prognosis after right ventricular outflow tract reconstruction**

Takashi Honda<sup>1</sup>, Yoichiro Hirata<sup>1</sup>, Rumi Watanabe<sup>1</sup>, Manabu Takanashi<sup>1</sup>, Atsushi Kitagawa<sup>1</sup>, Fumiaki Shikata<sup>2</sup>, Kagami Miyaji<sup>2</sup>, Kenji Ishikura<sup>1</sup>

<sup>1</sup>Department of Pediatrics, Kitasato University School of Medicine, Sagami-hara, Japan; <sup>2</sup>Department of Cardiovascular Surgery, Kitasato University School of Medicine, Sagami-hara, Japan

**Background and Aim:** Prediction of the timing for reoperation after right ventricular outflow tract reconstruction (RVOTR) is essential for patients. However, it is often difficult to determine the optimal time because stenosis and regurgitation lesions are intricately involved. In the present study, we evaluated the utility of the right ventricular stroke work index (RVSWI), which is an integrated index reflecting volume and pressure load, for predicting the timing of reoperation.

**Method:** This was a retrospective cohort study of 67 patients who underwent RVOTR, including 35 patients with tetralogy of Fallot and 32 with double outlet right ventricle. Based on the results of catheterization 1 year after RVOTR, we calculated the modified RVSWI (mRVSWI) based on the following equation: mRVSWI = right ventricular stroke volume/body surface area × (right ventricular systolic pressure – right atrial pressure) × 0.0136. We evaluated the influence of the RVSWI on the timing of reoperation using the Kaplan–Meier method.

**Results:** Catheterization was performed at 1.9 ± 0.9 years (1.2 ± 0.6 years after surgery). RVSWI was 18.8 ± 8.9. We divided the 67 patients into the high RVSWI group (Group H, RVSWI > 19, N = 29), moderate RVSWI group (Group M, 14–19, N = 30), and low RVSWI group (Group L, <14, N = 28). There was a significant difference in intervals from the catheterization to the reoperation between the 3 groups (p < 0.0001). Compared with Group M, Group H had a significantly lower avoidance rate for reoperation, and Group L had a higher avoidance rate (p = 0.0261 and p = 0.0489, respectively). The 10-year and 15-year reoperation avoidance rates in Groups H, M, and L were 56, 90, 100% and 32, 54, 100%, respectively. **Conclusions:** As mRVSWI is theoretically proportional to the area enclosed by the pressure–volume loop, this parameter is considered to comprehensively evaluate the pressure and volume load due to right ventricular outflow tract stenosis and regurgitation. Our results revealed that we could predict the timing for reoperation in the early phase after RVOTR based on mRVSWI. We

therefore propose RVSWI as a novel parameter for predicting prognosis after RVOTR.

**Keywords:** Right ventricular stroke work index, Right ventricular outflow tract reconstruction, Reoperation, Pressure load, Volume load

### P-334

#### Complete atrioventricular septal defect: preoperative risk factors

Zornitsa Vassileva, Anna Kaneva

Department of Pediatric Cardiology, University National Heart Hospital, Sofia, Bulgaria

**Background and Aim:** Complete atrioventricular septal defect (CAVSD) is a common congenital heart defect (CHD) whose preoperative diagnosis relies entirely on echocardiography and rarely requires other imaging modalities.

The aim of our study is to identify the preoperative risk factors for unfavorable outcome by CAVSD.

**Method:** We have reviewed the medical records of 75 patients with balanced CAVSD who underwent two-ventricle repair at the Pediatric Cardiology Department, Sofia, Bulgaria, during the period January 2014–December 2021.

**Results:** 44 (60%) of the operated children were male, and 29 (40%) – female; 40 patients (56%) had Down syndrome. The following surgical techniques were used: “Australian” technique – in 45 cases (60%), two-patch technique – in 27 cases (36%), single-patch – in 3 children (4%). All patients had some form of AVV plasty – of both the right and the left AVV (n = 63, 84%), only of the left AVV (n = 12, 16%), the cleft of the left AVV was closed in most patients (n = 63, 84%). 10 patients required reoperations, the reasons for which were as follows: residual VSD closure (n = 3), additional left AVV plasty (n = 5), VSD closure and left AVV plasty (n = 1), permanent pacemaker implantation (n = 1). 4 of the 75 children (5.3%) died in the early postoperative period.

No significant difference in the degree of reoperations between children with Down syndrome and those with normal karyotype was demonstrated (p = 0.676). There was no statistically significant correlation between the degree of preoperative and postoperative AVV regurgitation (p = 0.283); the suture of the left AVV cleft was not linked to the degree of postoperative AVV insufficiency (p = 0.151). The larger the VSD, the greater the probability for lethal outcome (p < 0.05). The “Australian” technique is utilized most often by smaller indexed VSD and the difference reaches statistical significance (p = 0.000).

**Conclusions:** The size of the VSD is the only significant preoperative risk factor for unfavorable outcome by CAVSD which could be identified. Nevertheless, accurate preoperative echocardiographic assessment with detailed description of the AVV anatomy and degree of regurgitation remains important for surgical planning and undoubtedly contributes to favorable patient outcome.

**Keywords:** atrioventricular septal defect, echocardiography, risk factors

### P-335

#### Fontan-associated liver and renal disease: A prospective single center study to early detect long-term complications

Alessia Basso<sup>1</sup>, Roberta Biffanti<sup>1</sup>, Jolanda Sabatino<sup>1</sup>, Liliana Chemello<sup>2</sup>, Davide Meneghesso<sup>3</sup>, Andrea Gasperetti<sup>4</sup>, Giovanni Di Salvo<sup>1</sup>, Domenico Sirico<sup>1</sup>

<sup>1</sup>Paediatric Congenital Cardiology Unit, Women and Child Health Department, Padova University Hospital, Padova, Italy; <sup>2</sup>Internal

Medicine 5, Padova University Hospital, Padova, Italy; <sup>3</sup>Paediatric Nephrology Unit, Women and Child Health Department, Padova University Hospital, Padova, Italy; <sup>4</sup>Sports Medicine, Padova University Hospital, Padova, Italy

**Background and Aim:** Fontan operation represents the surgical palliative option for congenital heart disease with single ventricle physiology. With the improvement of surgical techniques, we are facing a growing population with multiorgan complications: in particular, hepatic and renal dysfunction remain mostly unknown.

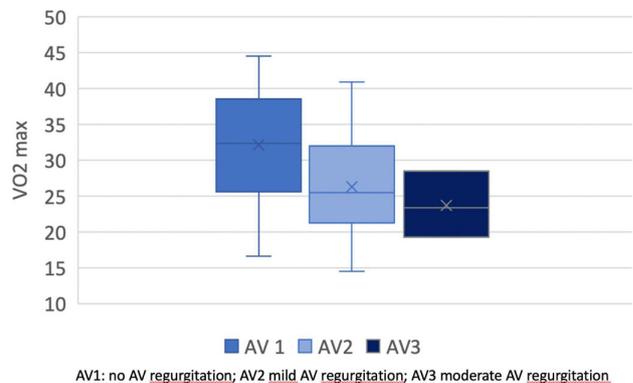
**Method:** We enrolled patients that underwent Fontan palliation in our Centre between 1985 and 2016 were included in this prospective study. We excluded patients with major congenital renal anomalies, those that underwent cardiac transplantation and redo-Fontan patients. All the subjects underwent clinical evaluation, laboratory exams with complete renal and hepatic function, transient hepatic elastography and complete cardiac evaluation. We aimed at 1) evaluate the cardiovascular, renal and hepatic dysfunction and 2) show the relationship between renal and hepatic results and cardiovascular status and the effect of time on multiorgan involv

**Results:** We enrolled 45 patients, 60% male. Medium time from Fontan completion was 13 years 6 months (167 months), SD ± 6.8 years (82 months). Data from echocardiographic evaluation shows that patients with functionally single right ventricle have worse diastolic function (p < 0.05); AV valve insufficiency is associated with low VO<sub>2</sub> max at cardiopulmonary testing (p 0.035). 23% patients have a mildly reduced glomerular filtration rate and 3% moderately reduced. Renal evaluation shows glomerular dysfunction (associated with systolic dysfunction p < 0.05) and chronic tubular alterations (associated with diastolic dysfunction p < 0.05). Renal dysfunction doesn't appear to be worse in patients with a past history of dialysis. Hepatic tests show that GGT alterations are more frequent than AST elevation (66% vs 9%). Hepatic stiffness higher than reference value is present in 32% of the population, with a lower VO<sub>2</sub> max at cardiopulmonary testing (p < 0.05).

**Conclusions:** Fontan patients show hepatic and renal dysfunctions, that worsen with time and appear to be related with functional ad cardiological tests. This suggests that Fontan patients need a multidisciplinary follow-up.

**Keywords:** Fontan, Follow-up, Echocardiography

#### Role of systemic AV regurgitation



AV valve insufficiency is associated with low VO<sub>2</sub> max (p 0.035)

P-336

### Outcomes of dilated and non- compaction cardiomyopathy presenting during childhood in sweden 1991 – 2020 a preliminary report

Shalan Fadl<sup>1</sup>, Stefan Sämrblad<sup>1</sup>, Håkan Wåhlander<sup>2</sup>, Jan Sunnegårdh<sup>2</sup>  
<sup>1</sup>Fadl S. School of Medical Sciences, Faculty of medicine and Health, Örebro University, Örebro, Sweden; <sup>2</sup>Sämrblad S. School of Medical Sciences, Faculty of Medicine and Health, Örebro University, Örebro, Sweden; <sup>3</sup>Wåhlander H. The Queen Silvia Children's Hospital, Sahlgrenska University Hospital, Gothenburg, Sweden; <sup>4</sup>Sunnegårdh J. Department of Paediatric Cardiology, Institution of Clinical Science, Sahlgrenska Academy, Gothenburg University, Sweden

**Background and Aim:** National data on the incidence and outcome in dilated cardiomyopathy (DCM) during childhood are rare. Incidence and outcome of DCM and non-compaction of the left ventricle (LVNC) in children in Sweden 1991 – 2020 is presented. **Method:** Hospital records of children and adolescents, 0 – 18 years, diagnosed with dilated cardiomyopathy (DCM) and/or left ventricle non-compaction (LV-NC) over a 29-year period were reviewed. Inclusion criteria for DCM were LV shortening fraction (LVFS) < 27% and symptoms of congestive heart failure. In LVNC inclusion criteria were according to international definition of the disease. Clinical data including echocardiography findings were registered. Incidence was cross-checked against the Swedish Population Registry in October 2022. Standard statistical measures determined survival, normalization of cardiac function and morbidity in those with and without transplant or death

**Results:** Fem-hundred forty patients were identified; 340 (63%) males and 200 (37%) females. The combined incidence of DCM and LVNC in children and young adults during this time period was 0.73 per 100 000 per year. Mean age at diagnosis was 3, 3 years (1 day – 17.9 years). Children with DCM associated with neuromuscular disease presented mostly after 10 years of age. Transplant-free survival was 87.5%, 83, 1% and 81, 2% at one, two and five years after diagnosis, respectively. Combined mortality and transplant rate was 27.2% over the studied period. This is a preliminary results.

**Conclusions:** The incidence of DCM and LVNC (0.73/100 000/year) in Sweden was similar to reports from other countries. The majority of children with so called idiopathic DCM and presented during the first year of life. Mortality was highest during the first year after diagnosis. The majority of children with DCM associated with other diseases were diagnosed beyond 10 years of age.

**Keywords:** Dilated Cardiomyopathy, non-compaction, incidence, survival

P-337

### Normal values of echocardiographic subcostal aortic and pulmonary artery velocity time integrals in neonates and infants

Nathalie Oeffl<sup>1</sup>, Martin Koestenberger<sup>1</sup>, Ante Burmas<sup>1</sup>, Gernot Grangl<sup>1</sup>, Sabrina Schweintzger<sup>1</sup>, Daniela Baumgartner<sup>1</sup>, Andrea Fandl<sup>1</sup>, Hannes Sallmon<sup>2</sup>, Stefan Kurath Koller<sup>1</sup>

<sup>1</sup>Division of Pediatric Cardiology, Department of Pediatrics, Medical University Graz, Austria; <sup>2</sup>Department of Pediatric Cardiology, Deutsches Herzzentrum Berlin and Charité Berlin

**Background and Aim:** Subcostal views are an easily accessible option, especially in neonates and infants where cardiac structures are in proximity to the ultrasound probe as well as in emergency assessment of critically ill patients. They are further useful under special conditions such as mechanical ventilation or CPAP, where imaging may be limited to subcostal views only. Normative values of

subcostal view measurements remain scarce. To quickly identify hemodynamic impairment and to support clinical decision making and therapy, velocity time integrals (VTI) are well established transthoracic echocardiographic parameters to estimate left and right heart function.

We aim to establish normative values of subcostal aortic and pulmonary VTI in neonates and infants.

**Method:** The target population were neonates and infants composed of a healthy study group and a validation study group of children suffering from cardiac shunt lesions, pulmonary hypertension or low cardiac output. Echocardiography was performed following a study protocol for subcostal views and collected data were compared to existing normative parameters of standard parasternal views.

**Results:** In this ongoing study we have included 300 patients to date. Our preliminary data show that subcostal aortic VTI and pulmonary VTI values are in good correlation with already existing parasternal normative values.

**Conclusions:** With this study we confirm that subcostal views in echocardiography provide a good imaging quality and are easily accessible. By establishing normative values in this vulnerable neonatal population the practical approach to investigate cardiac function from subcostal view will gain increasing clinical value.

**Keywords:** echocardiography, subcostal approach, neonatal intensive care, critical care

P-338

### A case of kawasaki-like syndrome in a 4.5 months old infant after COVID-19 infection

Sofia Loukopoulou<sup>1</sup>, Evangelia Grigoraki<sup>1</sup>, Aikaterini Giannakopoulou<sup>1</sup>, Nikolaos Eleftherakis<sup>1</sup>, Nikolaos Andreou<sup>1</sup>, Ioanna Maligianni<sup>2</sup>, Theodora Xanthaki<sup>2</sup>, Zoi Stavroula Siska<sup>2</sup>, Elena Tsitsami<sup>2</sup>, Eleni Pergantou<sup>3</sup>, Evangelos Karanasios<sup>1</sup>, Irene Orfanou<sup>2</sup>  
<sup>1</sup>Department of Cardiology, Aghia Sophia Childrens' Hospital, Athens, Greece; <sup>2</sup>Second Paediatric Department of Capodistrian University of Athens, Aghia Sophia Childrens' Hospital, Athens, Greece; <sup>3</sup>Department of bleeding disorders, Aghia Sophia Childrens' Hospital, Athens, Greece

**Background and Aim:** Kawasaki disease (KD) is a vasculitis of unknown etiology, usually affecting children between the age of 6 months to 5 years old. Multisystem inflammatory syndrome (MIS-C) is a pediatric hyperinflammation disorder associated with COVID-19 infection. Clinical manifestations of MIS-C can sometimes seem like KD. Kawasaki-like syndrome (KLS) is a terminology that has been used to correlate these two conditions. Complications of KD include coronary artery aneurysms, rupture, thrombosis and myocardial infarction. Early treatment is necessary for complete recovery.

**Method:** A 4.5 months old male infant proceeded to our Emergency Department because of fever, poor feeding, rash and bilateral conjunctival redness. There was no significant past medical history apart from a recent family infection from SARS-CoV2.

**Results:** The patient was admitted to a pediatric clinic and was initially treated with intravenous antibiotics. Because of high clinical suspicion of MIS-C, he received intravenous immunoglobulin (2g/kg), methylprednisolone (2mg/kg) and per os aspirin (3mg/kg). The patient responded well with recession of fever and rash. Every second day, his ECG and cardiac echocardiography were normal. On the 12th day of hospitalization a new clinical worsening took place with erythema, cracking of lips, maculopapular rash and desquamation of the tips of the fingers and toes, while laboratory tests showed high rise of WBC, PLT, CRP and hs-troponin.

A new cardiac echocardiography revealed a giant aneurysm of the right coronary artery (RCA) and a medium saccular aneurysm of the left coronary artery. Enoxaparin and prednisolone were started, immunoglobulin was repeated and aspirin was increased to 5mg/kg. 10 days later, echocardiogram pointed out a thrombus formation in the giant aneurysm of the RCA. The patient was admitted in the intensive care unit and was treated with intravenous thrombolysis (alteplase). Partial thrombus resolution followed, while subcutaneous anakinra was added to the patient's medication. A gradual clinical and echocardiographic improvement was observed.

**Conclusions:** Both KD and MIS-C are related to a significant cytokine storm that results in systemic inflammation. Clinical manifestations of MIS-C and KD overlap. More research is needed to clarify how new immunosuppressive drugs like anakinra affect coronary aneurysms.

**Keywords:** Kawasaki disease, Kawasaki-like syndrome, Multisystem inflammatory syndrome in children, giant aneurysms, Covid-19

### P-339

#### A case report of lysosomal acid lipase deficiency (LAL-D) and cardiovascular manifestations in a 13 year old child

Sofia Loukopoulou<sup>1</sup>, Kleopatra Skourti<sup>2</sup>, Elisabeth Georgiadou<sup>2</sup>, Evangelia Grigoraki<sup>1</sup>, Nikolaos Eleftherakis<sup>1</sup>, Evangelia Lykopolou<sup>2</sup>, Evridiki Drogari<sup>2</sup>

<sup>1</sup>Department of Cardiology, Aghia Sophia Childrens' Hospital, Athens, Greece; <sup>2</sup>First Paediatric Department, Capodistrian University of Athens, Aghia Sophia Childrens' Hospital, Athens, Greece

**Background and Aim:** Lysosomal acid lipase deficiency is an ultra-rare, progressive, autosomal recessive disorder. The disease spectrum ranges from the historically described infantile Wolman Disease to the later-onset cholesteryl ester storage disease (CESD). It is associated with mutations of the LIPA gene that encodes LAL enzyme, which catalyzes hydrolysis of cholesteryl esters and triglycerides and is characterized by hyperlipidemia, hepatomegaly and hepatic fibrosis. We report a case of a 13-year-old girl, who suffers from the disease and, as being untreated with enzyme replacement therapy, has already abnormal cardiovascular manifestations.

**Method:** The patient, when 5 years old, presented with hypertransaminasemia and hyperlipidemia. She underwent an Ultrasound Scan, that revealed moderate liver steatosis and liver elasticity measurement of 5, 54 kPa. In addition, during the following liver biopsy, we found micro vesicular steatosis without the presence of fibrosis. The diagnosis of the disease was certified by the measurement of LAL activity (0, 6 nmoles/mg/min) and a karyotype examination, that revealed homogeneity for the c.894G>a mutation.

**Results:** The patient commenced on anti-hyperlipidemic treatment with ezetimibe and simvastatin. The newly accepted enzyme replacement therapy with Sepelibase alfa was not available at the time. Upon presentation at the 12 years of age, the newly conducted liver biopsy revealed progression of the disease with presence of liver fibrosis. A following ultrasound scan showed elevation of liver elasticity to 7 kPa with co-existing hepatomegaly. The astonishing finding was, although, the presence of progressive atheromatic changes during the imaging examination of carotid arteries.

**Conclusions:** Cardiovascular abnormalities are thought to be a later presented manifestation of that rare disease. Normal anti-lipidemic treatment is probably not able to counter the progressive

cardiovascular disorders. Treatment with the newly certified enzyme replacement therapy may be the key to stop and even alter the prognosis of the condition.

**Keywords:** Lysosomal acid lipase deficiency, hyperlipidemia, atheromatosis, enzyme replacement therapy

### P-340

#### Severe adverse events related to vitamin K antagonists in children with congenital or acquired heart disease

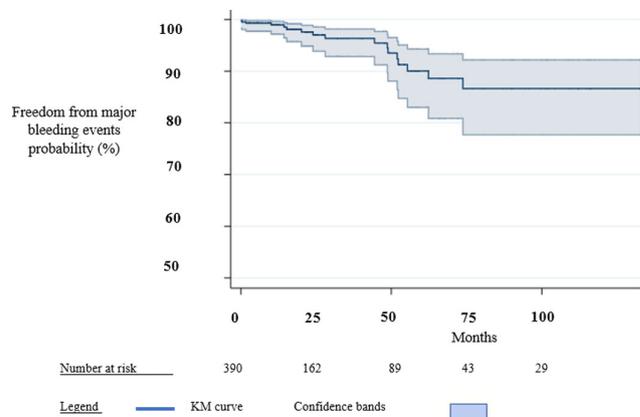
Fanny Bajolle<sup>1</sup>, Neil Derridj<sup>1</sup>, Joan Bitan<sup>1</sup>, Dominique Lasne<sup>2</sup>, Damien Bonnet<sup>1</sup>

<sup>1</sup>M3C-Necker, Hôpital Universitaire Necker-Enfants malades, AP-HP, Université de Paris Cité, Paris, France; <sup>2</sup>Hematology Laboratory, Hôpital Necker Enfants-malades, AP-HP, F-75015 Paris, France  
Pharmacogenetic HEGP

**Background and Aim:** Although increasingly used, Vitamin K antagonists (VKA) remain difficult to manage for children with congenital or acquired heart disease (CAHD). To assess the occurrence of thrombosis and major bleeding events in children with CAHD treated with VKA and to determine associated risk factors.

**Method:** All children enrolled in the VKA dedicated educational program between 2008 and 2020 were prospectively included. Clinically suspected thrombotic events were confirmed by imaging, while major bleedings were defined according to the ISTH. Time in therapeutic range (TTR) was calculated to evaluate the stability of anticoagulant therapy. Statistical analysis included Cox proportional hazard models.

**Results:** We included 390 patients, median age at initiation was 6.6 (IQR:12.8-3.2) years. Median follow-up was 17.8 (47.6 - 9.1) months. The median TTR was 85.2% (IQR:95.3%-74.4%). Thirteen thrombosis in 9 (2.3%) children and 24 major bleedings in 17 (4.4%) children were reported. The incidence of major bleeding and thrombosis events were 2, 26% (95%CI 0.74-3.51) and 1, 22% (95% CI 0.12-2.15) per patient year, respectively. At 6 years, 86.6% (CI95%[75.6%-92.4%]) and 95% (CI95%[89%-97.8%]) of patients were free of major bleeding and thrombotic events, respectively. Although mitral valve replacement (MVR) was associated to major bleeding events (HR 3.32 [1.16-9.44], p = 0.025) in univariate analysis only recurrent minor bleeding events ( $\geq 3$ ) (HR 4.9 [1.7-13.8], p = 0.002) and global TTR under 70% (HR: 3.6, [1.1-10.9], p = 0.04) were risk factors associated to major bleeding events in multivariable analysis. Kawasaki disease (HR: 11.84 [2.7-52.6], p = 0.001) and global TTR under 80% (HR: 5.05,



[1.2–20.7],  $p = 0.025$ ) were main risk factors for thrombotic events in multivariable analysis

**Conclusions:** Overall, VKA use appears to be safe and effective in children with CAHD. Patients with Kawasaki disease and those with a TTR under 80% are at highest risk of thrombosis. A TTR under 70% and minor recurrent bleeding events are the main risk factors for major bleeding events

**Keywords:** vitamin K antagonists, congenital heart disease, major bleeding events, mitral valve replacement

### P-341

#### Cardiac manifestations of paediatric multisystem inflammatory syndrome temporally associated with SARS-COV-2: romanian tertiary referral hospital data

Iolanda Muntean<sup>1</sup>, Carmen Corina Suteu<sup>2</sup>, Amalia Fagarasan<sup>1</sup>  
<sup>1</sup>University of Medicine, Pharmacy, Science and Technology „George Emil Palade“ of Târgu Mureş, Romania & Emergency Institute for Cardiovascular Diseases and Transplantation of Targu Mures, Romania; <sup>2</sup>Emergency Institute for Cardiovascular Diseases and Transplantation of Targu Mures, Romania

**Background and Aim:** One of the rare complications of SARS-CoV-2 infection in children is an inflammatory illness called paediatric inflammatory multisystemic syndrome (PIMS). The aim of this study is to describe a single center experience in relation to cardiac manifestations of PIMS temporally associated with SARS-CoV-2 infection, in Romanian children without coronavirus disease 2019 vaccination.

**Method:** This was a study performed at, a referral pediatric cardiology hospital in Romania. The study included all cases of PIMS that also associated cardiac manifestations between September 2020 to June 2022.

**Results:** We identified a total of 14 patients with a median age of 7.2 years old, 8 (57%) boys. All patients had positive PCR, serology test, or COVID-19 exposure. A previously healthy status was recorded in 13 (93%) children, 1 of the patients was previously diagnosed with autoimmune pathology. The most common presenting symptoms were fever (100%), digestive symptoms (abdominal pain, nausea, vomiting) (100%), fatigue (100%), rash (85.7%), conjunctivitis (71.4%), lymphadenopathy (57.1%), edema (57.1%), stomatitis (28.6%), arthralgia (28.5%), and respiratory symptoms (14.3%). The inflammatory markers: C-reactive protein (125mg/dL), ferritin (440.37ng/mL) and also Troponin I (141.12ng/L) and NT-proBNP (10732.65pg/mL) were elevated in all children. Electrocardiogram revealed sinus tachycardia, diffuse T wave inversions, and ST segment elevation. All children underwent echocardiography. The observed echocardiographic characteristics were basal septal dyskinesia (100%), mitral regurgitation (100%), left ventricular systolic dysfunction (87.5%) with an average ejection fraction of 45%, and decreased left ventricular longitudinal strain, left ventricular diastolic dysfunction (20%), pericardial effusion (87.5%), and coronary involvement (25%). Two patients needed inotropes and intensive care unit admission. During their hospitalization, all patients received methylprednisolone and intravenous immunoglobulin. No deaths were reported. The average total hospitalization was 12 days. The outpatient follow-up revealed left ventricular systolic dysfunction in 2 patients  
**Conclusions:** PIMS temporally associated with SARS-CoV-2 can present a wide spectrum of cardiac manifestations. Thorough cardiac evaluation and prompt anti-inflammatory therapy could

probably contribute to a favorable outcome; we highlighted the null mortality.

**Acknowledgements:** “This work was supported by the University of Medicine, Pharmacy, Science and Technology „George Emil Palade“ of Târgu Mureş, Research Grant number 511/5/17.01.2022”. Iolanda Muntean, Carmen Suteu contributed equally to the present study

**Keywords:** paediatric inflammatory multisystemic syndrome, Sars-Cov-2, cardiac manifestations

### P-342

#### Augmenting patient care: implementing a text messaging (SMS) service to improve communication and engagement in a pediatric cardiology clinic

Kathryn Armstrong<sup>1</sup>, Claire Galvin<sup>1</sup>, Astrid De Souza<sup>1</sup>, James Potts<sup>1</sup>, Richard Lester<sup>2</sup>  
<sup>1</sup>Children’s Heart Centre, Department of Pediatric Cardiology, British Columbia Children’s Hospital, Vancouver, Canada; University of British Columbia, Vancouver, Canada; <sup>2</sup>Department of Medicine, Vancouver General Hospital and Health Sciences Centre, Vancouver, Canada; University of British Columbia, Vancouver, Canada

**Background and Aim:** Increasing communication between patients and their health care providers (HCP) may improve care for many pediatric cardiology patients and their families. For adolescent patients, poor knowledge of their heart condition, their medications or how to access answers to their health care questions compromises their overall care. Fostering communication between these adolescent patients and their HCP may help to improve their engagement and resolve this issue. For parents, an SMS service provides convenient access to their child’s HCP. **AIM:** We implemented a bidirectional text messaging service (SMS) to improve communication between patients (families) and their HCP and extended the utility of the platform during the Covid 19 pandemic. This is a review of our experience.

**Method:** Inclusion Criteria: Patients aged 10–18 years and parents of children <10 years with access to a cell phone and text messaging plan were recruited. Demographic data was recorded. Weekly Messages: Every Monday at 12:00pm, participants received a text message asking “How are you?”. Participants could respond with “Ok”, “Not Ok”, or with any question, comment, or concern they may have related to their clinical care. Participants responding with “Not Ok” or with a concern were contacted by a HCP within 48 hours via text message. The number of care conversations, defined as more than two messages between patient and the HCP, were recorded

**Results:** A total of 721 patients (51% female) have been enrolled on to the SMS platform. Five hundred and forty-five virtual consultations have been conducted through the platform and 176 patients have signed up to receive weekly text messages. This has generated 47, 439 text messages to support clinical follow-up. The average number of care conversations are 2.6 per patient.

**Conclusions:** The SMS platform has allowed for increased communication and engagement to provide supplemental patient care. The functionality of the platform allowed for an immediate virtual care solution at the onset of the COVID-19 pandemic and promises to continue to enhance care for patients seen in our pediatric cardiology clinic.

**Keywords:** Text messaging, SMS, pediatric cardiology, engagement, communication, adolescent

## P-343

**The environmental impact of a pediatric cardiology outreach program on travel-related greenhouse gas emissions.**

James Potts<sup>1</sup>, Derek Human<sup>1</sup>, Astrid De Souza<sup>1</sup>, Jan Frew<sup>2</sup>, Connie Ens<sup>2</sup>, Pearl Waraich<sup>1</sup>, Christopher Buckland<sup>1</sup>, Sarah Wissmann<sup>1</sup>, Cindy Sha<sup>1</sup>, Kathryn Armstrong<sup>1</sup>

<sup>1</sup>Children's Heart Centre, Department of Pediatric Cardiology, British Columbia Children's Hospital, Vancouver, Canada; University of British Columbia, Vancouver, Canada; <sup>2</sup>Children's Heart Centre, Department of Pediatric Cardiology, British Columbia Children's Hospital, Vancouver, Canada

**Background and Aim:** British Columbia Children's Hospital (BCCH) serves the children and families of British Columbia and the Yukon, a region of 5 million people with a land mass of over 1.5 million km<sup>2</sup>. The geographical diversity of the region provides logistical difficulties for families to travel to Vancouver to receive care for their children with heart disease. Since 1994, the Children's Heart Centre has provided outreach services to children with congenital and acquired heart disease in their own communities. **AIM:** We report on the environmental impact of providing community outreach services on travel-related greenhouse gas emissions (GHG).

**Method:** All patient visits to Cardiology outreach program clinics between April 1, 2018 and March 31, 2020 were reviewed. Driving distance, driving time and GHG emissions were estimated using specialized geocoding software. Carbon dioxide equivalents (CO<sub>2</sub>e) were used as a surrogate for GHG emissions and estimated for five different fuel efficiencies. Flight distance, time, and CO<sub>2</sub>e were also calculated for air travel.

**Results:** There were 206 clinics held in 11 communities involving 2, 653 patients and 3, 163 patient visits. Patient Motor Vehicle Travel: If all patients drove from home to BCCH, travel distances would approximate 2.41 million km with travel times of 27, 344hrs. The resulting GHG emissions would be 981.161 metric tonnes (t) of CO<sub>2</sub>e. Patient Airline Travel: If patients were to fly from home to BCCH, travel distances would approximate 1.38 million km with travel times of 3, 127hrs. The resulting GHG emissions would be 975.877t of CO<sub>2</sub>e. Medical Team Travel: All medical teams flew to community outreach clinics, except to one clinic located 34km from BCCH. They flew an estimated 47, 406km with flying times of 106hrs or drove 10, 540km with travel times of 186hrs. The resulting GHG emissions were an estimated 146.873t of CO<sub>2</sub>e. Reduction in GHG Emissions: The community outreach service resulted in an estimated 85% reduction in GHG emissions for both motor vehicle and airline travel when patients are seen in their local communities.

**Conclusions:** The Cardiology outreach service had a profound effect on reducing GHG emissions by an estimated 85% as well as reducing the distance and time families spend travelling.

**Keywords:** green house gas emissions, environment, outreach clinic, pediatric cardiology

## P-344

**Hypertrophy of the heart in an infant.**

Anna Kadłubiska, Aleksandra Szatyłowicz, Kornel Semeran, Artur Tadeusz Bossowski

Department of Paediatrics, Endocrinology, Diabetology with Cardiology Unit, Medical University of Białystok, Poland

**Background and Aim:** Myocardial hypertrophy is a heterogeneous group of diseases. In the group of newborns and infants,

cardiomyopathy, congenital heart defects, gestational diabetes, arterial hypertension and metabolic diseases should be taken into account.

**Method:** This case report presents a 9-month-old infant with a severe hypertrophy of the left ventricles.

**Results:** A male infant, born from pregnancy III complicated by arterial hypertension, delivery I, by caesarean section at 38 hbd with a body weight of 3500 g with 10 points on the Apgar scale. At 5 weeks old the patient got urinary tract and COVID-19 infections. During hospitalization at the 3 months old, because of urosepsis, a chest X-ray showed an enlarged cardiac silhouette. The diagnostics was extended with echocardiography and generalized hypertrophy of the myocardium was found, especially of the left ventricle, without narrowing the outflow tract of the left ventricle. ECG showed generalized hypertrophy of both ventricles. Proteinuria was found in the urinalysis. Because of that propranolol was used in the treatment. Initially, blood pressure (RR) was 90-100/40-65mmHg. Differential diagnosis was performed. During observation, elevated RR values of 137-144/88-112mmHg were measured, and further antihypertensive drugs were added to the treatment. Complex ventricular arrhythmia was recorded on the 24-hours ECG. Despite the normal Doppler USG result in the neonatal period, the examination was repeated and the tardus-parvus spectrum was recorded of the right kidney. In angioCT of the kidneys, bilateral critical stenosis of the kidneys arteries was found, as well as anomalies of the blood supply to the lower poles of the kidneys. The boy was transferred to the Department of Nephrology, Children's Memorial Health Institute, where kidney scintigraphy was performed, antihypertensive treatment was modified, resulting in normalization of blood pressure values. Due to the size of the vessels and renal vascular anomaly, the patient was disqualified from surgical treatment

**Conclusions:** Renal vascular hypertension can cause significant myocardial hypertrophy and, as a result, further complications may occur. There is a chance that hypertrophy of the heart will decrease when the normal range of blood pressure will be maintained. The impact of viral and bacterial infections on renal vascular endothelial damage is not fully understood.

**Keywords:** hypertrophy, arterial hypertension

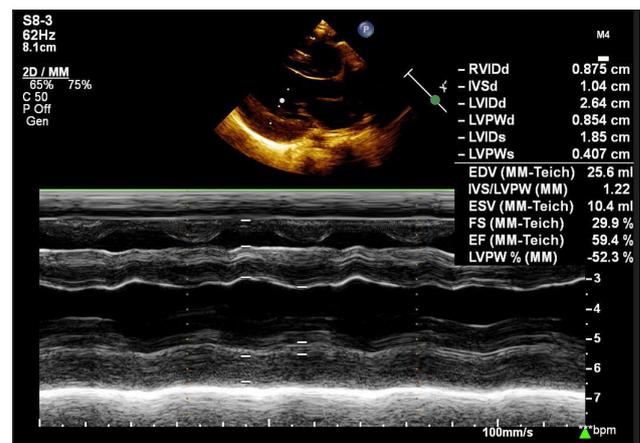


Figure 1. Echocardiography

P-348

**Down syndrome: prevalence and distribution of congenital heart disease on a tertiary centre**Ana Raquel Marques<sup>1</sup>, Madalena Von Hafe<sup>2</sup>, Joana Pimenta<sup>1</sup><sup>1</sup>Department of Pediatric Cardiology, Centro Hospitalar Universitário de São João, Porto, Portugal; <sup>2</sup>Department of Pediatrics, Centro Hospitalar Universitário de São João, Porto, Portugal

**Background and Aim:** Down syndrome (DS) is the most common genetic disorder, affecting one in every 700 live births. Because of the presence of extra genetic material from chromosome 21, children with DS have multiple malformations and medical conditions. Among the clinical findings, one constant concern is the high prevalence of congenital heart disease (CHD), which is reported to be approximately 50%. The aim of the study was to determine the prevalence and patterns of CHD in pediatric patients with DS.

**Method:** We conducted a descriptive and retrospective study of patients with DS followed in a tertiary center with at least one outpatient visit between 2008 and 2021. Clinical data were retrospectively reviewed and evaluated. Statistical analysis was performed with IBM SPSS Statistics for Windows, Version 27.0.

**Results:** Ninety-three patients with DS were included in the study (50.5% female). Maternal and paternal age in years was  $33.8 \pm 8.12$  and  $36.0 \pm 7.10$ , respectively, with no differences between the patients with or without congenital heart disease. Among the 93 patients, 51 (54.8%) were diagnosed with CHD; 27 (52.9%) were female and 24 (47.1%) were male. There was no gender difference in the prevalence of CHD. It was found that 61.7% of patients didn't had prenatal diagnosis of CHD, but patients with prenatal diagnosis of heart disease had also significantly higher rates of prenatal diagnosis of DS. Atrioventricular septal defect was the most frequent lesion, identified in 31.4% of patients, followed by ventricular septal defect and atrial septal defect, detected in 23.5% and 17.6% of patients, respectively; 9.8% had patent ductus arteriosus beyond the neonatal period. Regarding treatment approaches, 36 patients (70.6%) underwent cardiac surgery and 3 (5.9%) were submitted to a percutaneous procedure, within an average of  $21, 5 \pm 7$ , 3 months after birth.

**Conclusions:** The high prevalence of congenital heart disease among patients with DS was similar to findings from other cohorts. It justifies investigation during the prenatal period and/or early referral, in order to optimize the standard of care desirable to these patients.

**Keywords:** Down syndrome, Congenital heart disease, Prenatal diagnosis

P-349

**Complexity of medication regimens for children with heart failure – pilot study**Milica Bajcetic<sup>1</sup>, Milica Bajcetic<sup>2</sup><sup>1</sup>Department of pharmacology, clinical pharmacology and toxicology, Faculty of Medicine, University of Belgrade, Serbia; <sup>2</sup>University Children's Hospital Belgrade

**Background and Aim:** The unavailability of commercial paediatric drug formulations labelled for the treatment of HF in children significantly influences the quality and efficacy as well as the complexity of medication regimens. The aim of this investigation was to characterize the medication regimen complexity index (MRCI) in different ages of children with heart failure.

**Method:** This cross-sectional analysis was conducted in the cardiological department of the two university children's hospitals. Children age from birth up to 18 years with HF was included.

The age groups were defined according to the European Agency for the Evaluation of Medical Products. Drugs were classified according to the Anatomic and Therapeutic Chemical Classification. The degree of complexity of the medication regimen was identified using MRCI scores (low, medium, and high score) in the different age groups of children. The total MRCI score is composed of 3 subscores: dosage form, dose frequency, and specialized instructions.

**Results:** The data of 70 children, mostly male (51%), were analyzed. Drugs were prescribed to children with HF due to congenital heart diseases (91.4% of patients) and dilated cardiomyopathy (8.6% of patients). The average number of prescriptions on a per-child basis was 3.7 (range: 1–15). Furosemide, captopril, digoxin, and spironolactone represent 75% of drug utilisation (DU75%). More complex regimens were observed in the group of neonates, toddlers, and children. The highest MRCI score (58) was calculated in neonates, followed by the medium scores in toddlers (46) and children (38) and low score tertiles in adolescents (29). The total median (IQR) counts for the subscores were 4 (2–6) dosage forms per patient, 6 (4–8) dose frequencies per patient, and 4 (3–7) instructions per patient.

**Conclusions:** Higher MRCI scores were observed in neonates, toddlers, and children compared to adolescents with HF. High MRCI was associated with multiple-dose frequencies, complicated by different dosage forms and instructions. Our findings suggest that the simplification of dosing schedules is essential for managing complex medication regimens. The introduction of drug formulations designed for children with more dose flexibility, easier administration, and uncomplicated instructions is essential for simplification of the medical regimen in children with HF.

**Keywords:** Heart failure pharmacotherapy, medication regimen complexity index, polypharmacy

P-350

**Chd infant mortality disparities in the usa: positive changes in the worst performing state after creation of a joint heart programme**Eimear McGovern<sup>1</sup>, David P Nelson<sup>1</sup>, David Winlaw<sup>2</sup><sup>1</sup>Department of Pediatrics, University of Kentucky, Lexington, USA;<sup>2</sup>Department of Cardiothoracic Surgery, Cincinnati Children's Medical Center, Cincinnati, USA

**Background and Aim:** As per “Geographical Variation in Infant Mortality due to Congenital Heart Disease in the USA: a Population-Based Cohort Study” by Udine et al, from 2006 to 2015, Kentucky was the worst performing state for infant mortality from congenital heart disease (CHD).

Proximity to a US top 50 pediatric congenital cardiac center and poverty level were two variables associated with mortality.

A pediatric cardiac center in the state of Kentucky joined with a top 20 US pediatric cardiac center in nearby Ohio in 2017, to create a “Joint Heart Programme.” All procedural cases are discussed at biweekly conferences between the two centers, where timing and location of such procedures are decided. Imaging, intensive care, procedural and surgical skills and expertise are shared between both centers.

With the permission of the original authors, I will replicate this study focusing on the state of Kentucky from 2017 to 2021, with the aim of assessing how CHD infant mortality has changed with time and since the Joint Heart Programme has been in place.

**Method:** A population-based retrospective cohort study using the US National Center for Health Statistics de-identified livebirth and infant death data. A cohort of live births from Jan 1st 2017 to Dec 31st 2021 for Kentucky will be assessed. Observed rates

of infant mortality due to CHD are calculated for Kentucky and across various covariates: sex; gestational age; maternal ethnicity; maternal education; poverty; and proximity to a top 50 ranked pediatric cardiac centre. Outcome is death attributable to CHD as the underlying cause on the death certificate. Mortality trends and associations will be compared to 2006–2015 data.

**Results:** Livebirth and infant death data is still awaited from the CDC. To be received in the coming 1–2 weeks.

**Conclusions:** Analysing variation in CHD infant mortality across geographical boundaries and over time, can help identify health-care disparities. This can then inform the relevant medical and social bodies where funds and efforts need to be channeled. These findings may also support regionalization of care, a debate also pertinent to the EU.

**Keywords:** Healthcare disparity, Infant Mortality, Regionalization of Care

### P-351

#### **Myocarditis in pediatric population, a challenge for diagnosis and management**

*Erika Rezola, Ainhoa Izquierdo, Maria Angeles Izquierdo, Marta Iglesias, Miren Eizemendi, Ane Etxeberria*  
Pediatric Cardiology Department. Donostia University Hospital. San Sebastian. Spain.

**Background and Aim:** Myocarditis is defined as a cardiac muscle inflammatory disease. Despite etiology can be very variable, infectious disease is the most common one. Clinical presentation varies from subclinical disease to cardiogenic shock and sudden cardiac death.

The aim of this study is to review clinical presentation, course, etiology, treatment and follow up of pediatric myocarditis admitted in our hospital

**Method:** We performed a retrospective analysis of all children admitted to our hospital with diagnosis of myocarditis between 2016 and 2022. Patient characteristics are summarized using frequencies and percentages for categorical variables and medians with percentiles for continuous variables

**Results:** 10 patients were analyzed (5 male, 5 female) with a median age of 40, 13 months. The most common symptom was respiratory distress (60%), followed by stomach pain (20%), chest pain (10%) and shock (10%). Hepatomegaly was found in 60% of patients, third heart sound was found in 60% and auscultation revealed murmurs in 20%. Cardiomegaly was identified on chest X-ray in 70% patients, every patient showed elevated troponin levels, 90% had an abnormal ECG and 90% had alterations on the echocardiogram. The most common cause of myocarditis was a viral infection. The echocardiogram showed impaired left ventricular function in 66, 6%, biventricular dysfunction in 22, 2% and 55, 5% of patients associated mitral regurgitation. All patients received medical therapy and some of them needed intensive therapy such as inotropic and respiratory support (70%). In the follow up, ventricular function returned to normality in most patients (66, 6%). However, 3 patients maintained an impaired ventricular function, 1 patient required heart transplantation and 2 patients died

**Conclusions:** Myocarditis in children remains a challenging condition requiring high index of suspicion. Prognosis in acute myocarditis is generally good and patients usually respond well to standard therapy. However, it is an important cause of morbimortality in pediatric population. Many patients need intensive circulatory support or heart transplantation and some progress to chronic cardiomyopathy or death

**Keywords:** Myocarditis, pediatrics

### P-352

#### **Evaluation of late cardiac effects after multisystem inflammatory syndrome in children.**

*Rik De Wolf<sup>1</sup>, Nicolas Arribard<sup>2</sup>, Wendy Dewals<sup>3</sup>, Laura Muiño Mosquera<sup>4</sup>, Kaoru Tanaka<sup>5</sup>, Kristof Vandekerckhove<sup>4</sup>, Mahmoud Zaqout<sup>6</sup>, Daniel De Wolf<sup>7</sup>*

<sup>1</sup>Department of Pediatric Cardiology, Brussels University Hospital, Brussels, Belgium; <sup>2</sup>Department of Pediatric Cardiology, Queen Fabiola Children's University Hospital, Brussels, Belgium; <sup>3</sup>Department of Pediatric Cardiology, Antwerp University Hospital, Antwerp, Belgium; <sup>4</sup>Department of Pediatric Cardiology, Ghent University Hospital, Ghent, Belgium; <sup>5</sup>Department of Radiology, Brussels University Hospital, Brussels, Belgium; <sup>6</sup>Department of Pediatric Cardiology, Antwerp University Hospital, Antwerp, Belgium; <sup>7</sup>Department of Pediatric Cardiology, Queen Paola Children's Hospital, Antwerp, Belgium; <sup>7</sup>Department of Pediatric Cardiology, Brussels University Hospital, Brussels, Belgium; Department of pediatric Cardiology, Ghent University Hospital, Ghent, Belgium

**Background and Aim:** Multi Inflammatory Syndrome in Children (MIS-C) is associated with important cardiovascular morbidity. On short-term follow-up most patients recover. However, a small portion of patients has persistent cardiac sequelae at mid-term, but data are scarce. The goal of our study was to assess cardiac outcomes of MIS-C at mid-term by echocardiography, cardiac MRI (CMR), NT-proBNP and 24-hour Holter monitoring.

**Method:** A prospective observational multicenter study was performed in children admitted with MIS-C and cardiac involvement between April 2020 and March 2022. Follow-up by echocardiography, 24-Hour Holter monitoring and NT-proBNP measurement and CMR was performed at least 6 months after MIS-C diagnosis.

**Results:** We included 36 children with a median age of 10.0 (8.0–11.0) years who could undergo CMR without sedation. At diagnosis, all patients had an elevated NT-proBNP and 40% had a decreased left ventricular ejection fraction (LVEF; < 55%). Follow-up visit was done at a mean time of 12.1 (± 1.0) months after diagnosis. One patient kept a severely reduced LVEF and myocardial fibrosis on CMR requiring heart transplantation. He was our first pediatric patient with MIS-C of the SARS-CoV-2 pandemic and he did not receive immunoglobulins nor corticosteroids. All other patients had normal NT-proBNP and normal echocardiographic LVEF at follow-up. LV global longitudinal strain, as marker of subclinical myocardial dysfunction, was decreased ( $z < -2$ ) in 35%. CMR identified one patient with borderline LVEF, another patient had moderate mitral valve insufficiency and no patients showed signs of myocardial fibrosis. 24-Hour Holter monitoring was normal in all except one patient with a supraventricular tachycardia.

**Conclusions:** The majority of MIS-C patients have no clinically significant cardiac sequelae at mid-term follow-up, however a subgroup has persistent subclinical myocardial dysfunction and a small minority shows clinically relevant residual lesions.

**Keywords:** MIS-C, Cardiac MRI, SARS-CoV-2

### P-354

#### **Idiopathic dilatation of the right atrium: A case report**

*Catarina Maria Almeida<sup>1</sup>, João Sarmento<sup>1</sup>, Joana Miranda<sup>1</sup>, Joana Miranda<sup>2</sup>*

<sup>1</sup>Pediatric Cardiology Department, Centro Hospitalar Universitário de São João, Porto, Portugal; <sup>2</sup>UnIC@RISE, Department of Surgery and Physiology, Faculty of Medicine of the University of Porto, Portugal

**Background and Aim:** Idiopathic dilatation of the right atrium (IDRA) is a rare congenital anomaly that presents as an isolated enlargement of the right atrium (RA). A wide clinical spectrum has been reported, from asymptomatic patients to sudden death. Thrombus formation and atrial arrhythmias are the major complications. Management with antiplatelet therapy is recommended. Reduction plasty is reserved for patients that present a rapid growth of the RA, refractory arrhythmias or compression of neighboring structures.

**Method:** We report a case of IDRA diagnosed prenatally with a 9 year follow-up. A previously healthy pregnant woman was referred to a pediatric cardiology center due to cardiomegaly on obstetric ultrasound. The fetal echocardiogram confirmed cardiomegaly as a result of a significant RA enlargement. No other alterations were found. A male infant was delivered at term with an Apgar Score of 8/10. The physical examination after birth was normal. Postnatal transthoracic echocardiogram confirmed an aneurismatic dilatation of the RA with no other abnormalities. Thrombosis prophylaxis with acetylsalicylic acid was started.

**Results:** During follow-up the patient was regularly evaluated with serial echocardiograms that presented an evolution of the RA enlargement. Serial 24 hour Holter reports showed sinus rhythm sometimes alternating with junctional rhythm and premature atrial contractions (less than 2% of heartbeats) without pathological events. At 5 years of age, cardiac magnetic resonance imaging confirmed the significant RA enlargement (calculated area of 52 cm<sup>2</sup>, Z-Score 11.60, range: 4.65-9.35). However, it was decided to continue medical management due to clinical stability and absence of complications. Currently, at 9 years old, echocardiogram

revealed a stable RA area and the patient is asymptomatic. Throughout the years he maintained therapy with salicylic acid.

**Conclusions:** Considering the possible complications of IDRA, periodic follow-up should be focused on detecting progressive and rapid atrial enlargement, new symptoms and uncontrolled arrhythmias. Despite being asymptomatic, our patient underwent serial evaluations and maintained antiplatelet prophylaxis. We also want to highlight the importance of the prenatal diagnosis that in this case allowed an anticipated strategy to prevent complications and to improve this congenital heart disease outcome.

**Keywords:** pediatrics, cardiology, right atrium, dilatation

### P-355

#### Use of global longitudinal strain in detecting early cardiac dysfunction in children with duchenne muscular dystrophy: A meta-analysis

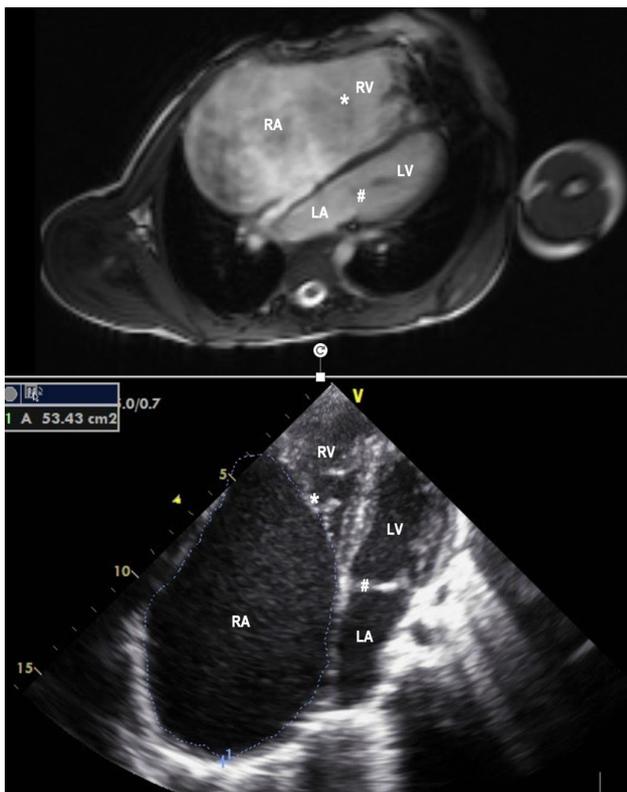
Zhia Ning Lim<sup>1</sup>, Amy Yi Lin Ng<sup>3</sup>, Sheng Chia Chung<sup>1</sup>, Juan Pablo Kaski<sup>2</sup>

<sup>1</sup>University College London, London, UK; <sup>2</sup>Great Ormond Street Hospital for Children NHS Foundation Trust, Great Ormond Street, London, UK; <sup>3</sup>Barts and the London School of Medicine and Dentistry, Queen Mary University of London, London, UK

**Background and Aim:** Duchenne Muscular Dystrophy (DMD)-associated Cardiomyopathy is characterised by progressive heart muscle loss, insidious onset of heart failure, and is a leading cause of death for patients with DMD. Functional assessment by cardiac Magnetic Resonance Imaging (cMRI) is not well tolerated in this patient cohort. This systematic review aims to compare the use of standard echocardiographic measurements of ejection fraction with strain to detect early left ventricular systolic dysfunction in children with DMD.

**Method:** This systematic review was registered with PROSPERO (CRD42022308511) prior to initiation. A restricted literature search was conducted on 5 databases with a focus on 'Duchenne Muscular Dystrophy', 'speckle tracking echocardiography', and 'strain'. A total of 10,352 publications from 1974 to 2022 were screened. Using prespecified inclusion and exclusion criteria, 15 original studies had sufficient data to be included for data extraction. Standardised mean difference (SMD with 95% confidence intervals (CI) were estimated for left ventricular ejection fraction (LVEF), fractional shortening (FS), and global longitudinal strain (GLS). Newcastle Ottawa Scale (NOS) was used for risk of bias assessment. RevMan and R were used for conducting the meta-analysis.

**Results:** 483 children with Duchenne Muscular Dystrophy and 352 controls were included from all 15 studies. The sample size of the studies ranged from 13 subjects to 63 subjects. Mean age ranges from 5-15.6 years old and 5-12.6 years old for DMD and controls respectively. Diagnosis of DMD were obtained from clinical phenotype and genetic confirmation. Parameters used to define cardiac dysfunction were LVEF<50-55%, FS<28%, and GLS <18-20.5%. There was a significant heterogeneity reported in LVEF and FS measurements (SMD = -0.57, 95% CI -1.02, -0.12, p = 0.01, I<sup>2</sup> = 84%; SMD = -0.69; -1.35, -0.03, p = 0.04, I<sup>2</sup> = 89%), whereas global longitudinal strain measurements were more consistent (SMD = 3.93, 95%CI 2.96, 4.91, p = 0.001, I<sup>2</sup> = 71%). A subgroup analysis categorising studies that investigated LVEF and GLS parameters in patient cohort <10 years old showed that there was GLS was abnormal in context of a normal LVEF (SMD = 3.27, 95%CI 2.48, 4.07, P<0.00001 I<sup>2</sup> = 1%; SMD = -0.48, 95%CI -1.14, 0.18, p = 0.15, I<sup>2</sup> = 82% respectively).



Cardiac Magnetic Resonance (superior image) and Transthoracic Echocardiogram (inferior image). RA - right atrium; LA - left atrium; RV - right ventricle; LV - left ventricle; \* - tricuspid valve; # - mitral valve.

**Conclusions:** GLS is useful screening tool for early detection of left ventricular dysfunction in children with DMD.

**Keywords:** Duchenne, Muscular Dystrophy, Speckle tracking, strain

### P-356

#### **San filippo syndrome: A case report with bicuspid aortic valve**

*Evmorfia Ladopoulou<sup>1</sup>, Aikaterini Boli<sup>1</sup>, Labrini Poulidou<sup>1</sup>, Alkistis Kalesi<sup>2</sup>, Maria Karakosta<sup>2</sup>, Antonios Destounis<sup>2</sup>, Nearchos Kasinos<sup>2</sup>, Anastasios Theodosios Georgilas<sup>2</sup>, Niki Lama<sup>3</sup>, Efthimios Brachos<sup>1</sup>, Nikolaos Tziolas<sup>1</sup>, Konstantinos Tsatiris<sup>1</sup>, Sotiria Apostolopoulou<sup>4</sup>, Panagiotis Zachos<sup>5</sup>*

<sup>1</sup>Department of Cardiology, General Hospital of Karditsa; <sup>2</sup>Department of Cardiology, Tzaneio General Hospital of Piraeus; <sup>3</sup>Research Unit of Radiology and Medical Imaging, 2nd Department of Radiology, Medical School of Athens, Greece; <sup>4</sup>Pediatric Cardiology and Adult Congenital Heart Disease Department, Onassis Cardiac Surgery Center; <sup>5</sup>Pediatric Cardiology and Adult Congenital Heart Disease Department, General Hospital of Karditsa

**Background and Aim:** Mucopolysaccharidosis type III (MPS III or Sanfilippo disease) is a rare autosomal recessive lysosomal storage disorder, with incidence 0.28–4.1 cases per 100,000 births, caused by an enzyme deficiency leading to accumulation of a glycosaminoglycan (GAG). It is characterized by progressive neurocognitive decline, caused by GAG's accumulation in the central nervous system. Typically, somatic disease is limited whereas the cardiac involvement (CD) is less common and milder compared with other types of MPS (I, II, IV).

**Method:** We report a case of MPS III and cardiac anomalies who visit our Pediatric Cardiology and Adult Congenital Heart Disease Department for regular cardiac screening.

**Results:** The case concerns a boy 10 years old with confirmed MPS III with quantitative analysis of urinary GAGs followed by genetic testing for the identification of the type, during one of his hospitalizations for multiple respiratory infections. The boy presents with adenoid face, bilateral sensorineural deafness, multiple dysostosis, speech and behavior disorders and severe mental retardation. The echocardiogram unveils bicuspid aortic valve with a small degree of regurgitation with eccentric jet and normal blood flow velocity in aortic isthmus with no other pathological findings.

**Conclusions:** CD is prevalent in MPS III, affecting the myocardium, the valves, the conduction system and rarely the coronary arteries. Subclinical systolic or diastolic dysfunction and valvular heart disease, mostly mild, were observed. Most studies report that mitral valve is more often affected in comparison with aortic valve and regurgitation is more common than stenosis. Our case seems to be the first case to our knowledge that describes bicuspid aortic valve in MPS III.

**Keywords:** San Filippo syndrome, bicuspid aortic valve, Mucopolysaccharidosis

### P-357

#### **Cardiovascular abnormalities in a paediatric population of cleft palate carriers in yaounde-cameroon: clinical and echocardiographic aspects.**

*Tony Nengom Jocelyn<sup>1</sup>, Atanga Leonel<sup>2</sup>, Meyebe Thibaut<sup>1</sup>, Sap Ngo Um Suzanne<sup>1</sup>*

<sup>1</sup>Department of Pediatrics, Faculty of Medicine and Biomedical Sciences, University of Yaounde I, Cameroon; <sup>2</sup>Yaounde Gynaeco-Obstetric and Paediatric Hospital, Cameroon

**Background and Aim:** Cleft palate are congenital malformations whose incidence is estimated at 1 per 1200 births by the World Health Organisation in Africa. They are frequently associated with other congenital malformations, particularly cardiac ones, impacting the overall management of patients. There are no data in our setting on the association of cleft palate with congenital heart defects.

The objective was to describe the clinical and echocardiographic aspects found in children with cleft palate in the city of Yaounde. **Method:** We carried out a cross-sectional analytical study with retrospective and prospective data collection between January 1, 2017 and May 1, 2022, at the Yaounde Gynaeco-Obstetric and Paediatric Hospital. All patients under 18 years of age who consulted for cleft palate were included in our study.

**Results:** We enrolled 76 patients. The median age was 3 months with an interquartile range of [2–6] months. Primary cleft palate was found in 42 patients (55.26%), secondary cleft palate in 4 patients (5.26%) and primary and secondary cleft palate in 30 patients (39.47%). A congenital heart defect was found in 20 of the 76 patients (26.32%). Among these cardiopathies, all were simple and non-cyanogenic. The most common was atrial septal defect (17.10%) followed by patent ductus arteriosus (5.26%). Clinical Holt-Oram syndrome was found in 4 patients (5.26%). There was no significant association between the type of cleft and the type of cardiac anomaly.

**Conclusions:** The prevalence of congenital heart defects in children with cleft palate in Cameroon is high. Although all were simple anomalies not contraindicating facial reconstructive surgery, cardiac evaluation of these patients is necessary to identify potentially high risk patients.

**Keywords:** cleft palate, congenital heart defects, children, cameroon

### P-358

#### **Abnormal coagulation of single ventricle patients prior to stage I relates to complicated clinical course and invasive haemodynamic at stage II**

*Alessia Callegari<sup>1</sup>, Manuela Albisetti<sup>2</sup>, Stefano Di Bernardo<sup>3</sup>, Regina Wespi<sup>1</sup>, Oliver Kretschmar<sup>1</sup>, Daniel Quandt<sup>1</sup>*

<sup>1</sup>University Children's Hospital Zurich, Pediatric Heart Centre – Division of Pediatric Cardiology, University of Zurich; Switzerland;

<sup>2</sup>University Children's Hospital Zurich, Division of Hematology, University of Zurich; Switzerland; <sup>3</sup>Lausanne University Hospital (CHUV), Woman-Mother-Child Department, Paediatric Cardiology Unit, Lausanne University Hospital (CHUV), Lausanne; Switzerland.

**Background and Aim:** Clotting factor abnormalities leading to a prothrombotic state are frequent in single ventricle patients (SVs). Whether the coagulation imbalance precedes or is consequence of univentricular palliation, its evolution during staged palliation, and relation to haemodynamics factors remain unknown.

**Method:** This is an ongoing multicenter, prospective, longitudinal, controlled study with 36 SVs and 34 healthy controls (HCs). Coagulation profile was assessed prior to any surgical/interventional procedure at stage-I and at cardiac catheterization prior to stage-II. Mean±SD age at first and second sample was 11.4±12.9 days and 5.0±2.6 months in the SVs; 15.9±19.8 days and 5.2±7.5 months in the HCs (p=0.09; p=0.9). Severe reduction of coagulatory factors in SVs was defined with values below the 5. percentile of the HCs.

**Results:** Prior to stage-I SVs had significantly reduced levels of prothrombin (PT) (p<0.001), free-protein-S (PS) (p<0.001), antithrombin (AT) (p<0.001), coagulation factor (F) II (p<0.001), V

( $p < 0.001$ ), VII ( $p < 0.001$ ), X ( $p = 0.001$ ), XI ( $p = 0.05$ ), and plasminogen ( $p < 0.001$ ) compared to HCs.

SVs with a severe reduction of PC had higher Rp ( $p = 0.009$ ); of PS higher mPAP ( $p = 0.01$ ) and tPG ( $p = 0.05$ ); of AT higher mPAP (0.05), tPG ( $p = 0.004$ ), and Rp ( $p < 0.001$ ) at the cardiac catheterization prior to stage-II.

Prior to stage-II SVs had significantly reduced levels of PC ( $p < 0.001$ ), AT ( $p < 0.001$ ), FV ( $p < 0.001$ ), FVII ( $p = 0.03$ ), and plasminogen ( $p = 0.2$ ) compared to HCs.

A severe reduction of PC prior to stage-I persisted at stage-II in 9/12 (75%); of PS in 2/12 (16%); of AT in 8/10 (80%); of FV in 3/11 (27%) SVs. Persistence of very reduced PC ( $p = 0.01$ ), PS ( $p = 0.05$ ), and AT ( $p = 0.03$ ) was more frequent in patients with a complicating clinical course after stage-I (defined as ECMO, surgical/interventional reintervention, no ICU-discharge until stage-II).

**Conclusions:** Abnormalities in coagulation profile are present prior to any intervention in SVs, seem to persist at stage-II in patients with a complicating clinical course, and are associated with worse invasive hemodynamics prior to stage-II. These abnormalities do not seem to be caused by inflammatory processes, but a decreased liver synthetic capacity and/or a reduced vitamin K level could be of relevance.

**Keywords:** Coagulation, thromboembolic events, Single Ventricle (SV), invasive hemodynamic

### P-359

#### Central role of cardiovascular manifestations in multisystem inflammatory syndrome associated with COVID -19 in children- a single centre experience

Diana Jecan Toader<sup>1</sup>, Alina Grama<sup>1</sup>, Alexandra Maris<sup>2</sup>, Laura Bodea<sup>2</sup>, Mihai Militaru<sup>1</sup>, Marta Ginga<sup>2</sup>, Beatrice Tanasa<sup>2</sup>, Alexandra Mititelu<sup>1</sup>, Vlad Florin Chelaru<sup>3</sup>, Tudor L Pop<sup>1</sup>, Simona S Cainap<sup>1</sup>

<sup>1</sup>Emergency Clinical Hospital for Children Cluj-Napoca Romania & University of Medicine and Pharmacy "Iuliu Hatieganu" Cluj-Napoca Romania; <sup>2</sup>Emergency Clinical Hospital for Children Cluj-Napoca Romania; <sup>3</sup>University of Medicine and Pharmacy "Iuliu Hatieganu" Cluj-Napoca Romania

**Background and Aim:** Multisystem inflammatory syndrome in children (MIS-C) is a rare but serious condition often requiring intensive care. Cardiac involvement, ranging from myocarditis to coronary abnormalities, is a key feature of the syndrome.

The aim of our study was to describe the cardiac involvement in MIS-C in reference to our centre's experience.

**Method:** We conducted an observational study that included 37 patients aged less than 18 years old, diagnosed with MIS-C according to CDC and WHO criteria, treated in our centre between January 2020-September 2022.

**Results:** The most frequent cardiac clinical manifestations were: tachycardia (62, 16%), 29, 73% presented hypotension, 18.91% developed shock and 30.81% had signs of cardiac insufficiency. 10 patients required ICU admission and inotropic support. From a biochemical point of view: 89.19% presented high levels of NT-proBNP and 41.1% raised troponins. ECG abnormalities were found in 55.55% of patients: sinus tachycardia (30.55%), ST segment abnormalities (22.22%), prolonged QTc (13.88%), sinus bradycardia (13.88%), premature contractions (supraventricular - 2.77% and ventricular - 2.77%), grade I atrioventricular block (2.77%) and left branch block (2.77%). Serial echocardiographic assessments revealed decrease ejection fraction (EF) in 6 patients (16.6%), left ventricular hypertrophy (16.66%), dilated left ventricle (11.11%) and right ventricle (8.33%), mitral insufficiency (72.22%) and pericardial effusion (41.66%). 11 patients (29.72%)

presented dilation of the coronary arteries with a Z-score > 2.5, with higher incidence in the right artery. Of the patients who did not present dilation of the coronary arteries, 40.62% presented a greater than 1 decrease in the coronary artery Z-score after treatment. By discharge, 5 patients (15.63%) still presented dilated coronary arteries and 2 patients still presented decreased EF, of which one requiring heart transplant.

Treatment consisted of IVIG in 43.24% patients, corticotherapy alone in 16.22% and a combination of IVIG and corticotherapy in 40.54%.

CMRI performed after 6 months in selected patients (6) revealed no myocardial edema, but variable degrees of fibrosis. However, stress tests revealed normal exercise capacity.

**Conclusions:** Heart involvement is a common feature of MIS-C. Despite rapid resolution of systolic dysfunction and coronary anomalies in the majority of patients, a long term follow up program should be established considering the numerous unknowns of the disease.

**Keywords:** MIS-C, cardiac involvement, systolic dysfunction, coronary artery dilation, myocardial fibrosis

### P-360

#### Travel-related costs associated with a pediatric cardiology outreach program: shifting the burden away from families

James Potts, Derek Human, Astrid De Souza, Jan Frew, Connie Ens, Pear Wairach Wairach, Christopher Buckland, Sarah Wissmann, Cindy Sha, Kathryn Armstrong

Children's Heart Centre, Pediatric Cardiology, British Columbia Children's Hospital, Vancouver, Canada; The University of British Columbia, Vancouver, Canada

**Background and Aim:** Since 1994 the Children's Heart Centre has provided outreach services to children with congenital and acquired heart disease in their own communities. Given the vastness and geographical diversity of our region, travel can be expensive, especially for families making multiple clinic visits each year. **AIM:** We report on travel costs associated with patient and medical staff (cardiologist/echocardiographer/nurse) travel to attend pediatric Cardiology outreach clinics and compare those to the costs of having families attend clinics at British Columbia Children's Hospital (BCCH) in Vancouver.

**Method:** All patient visits to outreach clinics between April 1, 2018 and March 31, 2020 were reviewed. Costs associated with motor vehicle travel, car rental, flights, ferries, accommodation, meals and parking were calculated for each clinic location.

**Results:** There were 206 clinics held in 11 communities involving 2, 653 patients and 3, 163 patient visits. **Patient Travel by Motor Vehicle:** If all patients drove from home to BCCH, travel distances would approximate 2.41 million km with travel times of 27, 344hrs. The estimated travel costs would be \$3, 219, 043 CAD or €2, 341, 629. **Patient Travel by Air/Motor Vehicle:** If patients were to fly from home to BCCH, travel distances would approximate 1.38 million km with travel times of 3, 127hrs. The resulting travel cost would be \$2, 878, 188 or €2, 101, 078. **Local Motor Vehicle Travel for Patients:** Patients seen at outreach clinics in their local communities travelled 160, 369km with total driving times of 2, 417hrs at a cost of \$294, 393 or €214, 907. **Medical Staff Travel:** All medical staff flew to community outreach clinics, except to one clinic located 34km from BCCH. They flew an estimated 47, 406km with flying times of 106hrs or drove 10, 540km with travel times of 186hrs. The estimated travel costs were \$231, 626 or €169, 087. **Reduction in Patient Travel Costs:** The community outreach service resulted in a net cost reduction of 91% and 90% for motor vehicle and mixed air/motor vehicle travel, respectively.

**Conclusions:** The Cardiology outreach service had a profound effect by reducing travel-related costs and shifting the financial burden of travel away from families.

**Keywords:** cost, financial burden, outreach clinic, pediatric cardiology, cost saving

### P-361

#### Assessment of nutritional status evolution of children with congenital heart disease: A single tertiary center study

Ana Raquel Marques<sup>1</sup>, Madalena Von Hafe<sup>2</sup>, Joana Pimenta<sup>1</sup>, Susana Corujeira<sup>3</sup>, Paula Guerra<sup>3</sup>

<sup>1</sup>Department of Pediatric Cardiology, Centro Hospitalar Universitário de São João, Porto, Portugal; <sup>2</sup>Department of Pediatrics, Centro Hospitalar Universitário de São João, Porto, Portugal; <sup>3</sup>Pediatric Nutrition Unit, Department of Pediatrics, Centro Hospitalar Universitário de São João, Porto, Portugal

**Background and Aim:** Patients with congenital heart disease (CHD) are susceptible to malnutrition, which is multifactorial and associated to mechanisms like cyanosis or pulmonary overcirculation. The risk of growth failure may persist in the postoperative period and catch-up growth may be impaired. This study aims to assess the prevalence of malnutrition in patients with CHD.

**Method:** Retrospective study performed by reviewing medical records of children with CHD born between 2014 and 2022 and followed in a tertiary center, that underwent a formal evaluation from the Pediatric Nutrition Unit. Patients' diagnosis, birth weight and surgery related data were extracted, and anthropometric measurements were recorded in the first evaluation and again on the 3, 6, 9 and 12-month outpatient visit. Z-scores for weight for age (WAZ), length for age (LAZ) and weight for length (WLZ) were calculated using Anthro software.

**Results:** Fifty-one patients were included (47, 1% female). Mean birth weight was 2705±754 g. Forty-two patients (82.4%) underwent cardiac surgery, at a mean age of 8.7±12.75 months. The average WAZ at surgery was -2.75, with an overall prevalence of malnutrition of 83.3%. The first evaluation occurred at a mean age of 7.9±12.30 months, in the preoperative phase in 57.1% of patients. The average WAZ, LAZ and WLZ was -3.02, -2.83 and -1.50, respectively. There were no differences in the z-scores of the first assessment regarding the timing of evaluation (pre vs. postoperative period) and the type of CHD (cyanotic vs. acyanotic). Patients were submitted to several interventions, including enteral nutrition (45.1%), modular fortification (19.6%) and dietary adjustments (27.5%). Seventeen patients (33.3%) required tube feeding. WAZ lowered from the first evaluation throughout all follow up visits (Table 1), and also from 3 to 6-month (-2.57 ±1.86 vs -2.09±1.34, p<0.001) and 6 to 9-month (-2.09±1.34 vs -1.89±1.38, p = 0.001) visit. WLZ lowered only from the first to the 12-month visit (-1.49±1.53 vs -0.52±1.12, p = 0.001).

**Conclusions:** While malnutrition is a very common problem in children with CHD, nutritional management could result in remarkable improvement, including better surgical outcomes. This study highlights the central role of nutritional support for these patients and reinforce the need of an early and time-sustained intervention.

**Keywords:** Congenital heart disease, cardiac surgery, nutritional status, weight for age z-score

**Table 1 – Weight for age z-score evolution**

WAZ at first evaluation (n=51)		p value
-3.08 ± -1.64 vs.	WAZ at 3-month visit (n=45) -2.57 ± 1.86	0.019
	WAZ at 6-month visit (n=41) -2.09 ± 1.34	< 0.001
	WAZ at 9-month visit (n=25) -1.89 ± 1.38	< 0.001
	WAZ at 12-month visit (n=27) -1.39 ± 1.66	< 0.001

### P-362

#### An audit for the cardiac management of paediatric patients with duchenne's muscular dystrophy in a UK tertiary centre

Stavros Christoforides, Maria Iliina

Royal Hospital for Children - Glasgow - UK

**Background and Aim:** The cardiac management of children with dystrophinopathy in UK has been variable as there has not been a clear national guidance. The latter has changed with the circulation of a UK-wide cardiac guidance on the management of children with dystrophinopathy in 2022. With this opportunity, we performed an audit of the paediatric population with Duchenne's muscular dystrophy known to our hospital.

**Method:** This study involved retrospective data collection for the 20 younger patients with Duchenne's muscular dystrophy known to Royal Hospital for Children (Glasgow) for the last 15 years. The data collection was from December 2021 to April 2022.

**Results:** The mean age of this group of patients was 10.6 years (range 5 – 15 years) and the mean age at diagnosis was 2.9 years (range 6 days – 5 years). 12/20 patients (60%) were commenced and maintained on ACEi. The mean age of ACEi introduction was 6.8 years (range 4 – 13 years). 9/12 patients were started on ACEi prophylactically at a mean age 6.5 years, 2/12 patients due to impaired LV systolic function and 1/12 patient for systemic hypertension. There was a significant variability between individual Cardiologists in terms of the age introducing ACEi for their patients (5.6 – 13 years). Regarding other classes of medications, 1/20 patient was treated with b-blockers for WPW and documented SVT and no patients received mineralocorticoid antagonist or angiotensin receptor blocker. All 20 patients were started on steroids at a mean age of 4.9 years (range 4 – 9 years). A cardiac MRI was electively performed in one patient at the age of 13 years and a 24-hour-ECG tape was done in 6/20 patients.

**Conclusions:** Steroids was the most consistently used class of medications in our group of patients, with ACEi being the second most common. The two patients in this cohort with documented LV systolic impairment had not been escalated onto additional cardiac medications. Finally, we found variable practice between different Cardiologists in terms of medicines introduction and surveillance cardiac investigations. All above indicate that there is significant space for improvement in the cardiac management of DMD patients, with more timely and consistent interventions.

**Keywords:** Duchenne's muscular dystrophy, dystrophinopathy

## P-363

**Characteristics and long-term outcomes of Ebstein anomaly in children and adults**Dimitri Herrera<sup>1</sup>, Bozio Andre<sup>1</sup>, Olivier Metton<sup>1</sup>, Julia Mitchell<sup>1</sup>, Loic Bousset<sup>1</sup>, Francis Bessiere<sup>1</sup>, Sylvie Di Filippo<sup>2</sup><sup>1</sup>Department of Congenital Heart Disease, Claude Bernard University, Lyon, France; <sup>2</sup>Congenital Cardiology, Cardiothoracic Center of Monaco

**Background and Aim:** The objective of the study was to assess characteristics and outcomes over time of a population of patients with Ebstein anomaly

**Method:** All patients diagnosed with Ebstein anomaly were included in the study. Demographics, clinical data, transthoracic echocardiographic data, outcomes and events were assessed.

**Results:** Seventy-six patients were included, 34 males (45%). Mean age at diagnosis was 21.9 years, median 13.9, max = 67.8 (neonates 22%, adult 46%, antenatal 5%). Symptoms at diagnosis included: dyspnea (39%), cyanosis (30%), palpitations (17%), syncope (4%). NYHA class at diagnosis was I, II, III and IV in respectively 55%, 35%, 7% and 3%. SpO<sub>2</sub> was lower in NYHA class III-IV and in neonates. Age at diagnosis of severe Ebstein anomaly was lower (mean 13.8 years) than minor/moderate Ebstein (21 to 25 years). ASD was present in 71%, WPW in 16%, obstructive RV outflow tract lesion in 12%, VSD in 10%. RV/LV ratio was lower in minor Ebstein (1.1) than in severe/moderate (1.7). Mitro-tricuspid distance was 15.5mm in minor Ebstein, 28.3mm in moderate and 48mm in severe Ebstein ( $p = 0.0034$ ). Atrial defect was absent in 31.5%, PFO in 42% or ostium secundum ASD in 26.5%, with bidirectional shunt in 15.3%, right to left in 36%. Follow-up was 2.8 to 66 years, mean 28, median 25 years. Supraventricular tachycardia occurred in 43.5%, more frequent if WPW present (41% vs 14%,  $p = 0.06$ ). Age at reentrant tachycardia was 25 years, at IART was 45 years ( $p = 0.0003$ ). Right bundle branch was present in 63% of cases, less frequent if WPW (41% vs 67%,  $p = 0.09$ ). Pacemaker implantation was performed in 12%. Three deaths occurred (4%), due to uncontrolled HF, at age 24 days, 37 and 52 years. HF occurred in 26% of cases, thromboembolic events in 9%, stroke in 10.5%. Mean age at event was 39 to 41 years. Procedures were performed over follow-up in 58% of cases (ASD closure, electrophysiology procedures). Freedom from procedure was 85%, 80%, 70%, 50%, 20% at respectively 10, 20, 50 and 70 years of age.

**Conclusions:** Long-term survival rates of Ebstein anomaly patients were favorable. Severe Ebstein was associated with early diagnosis and events and more than half of patients underwent one or more procedure over follow-up

**Keywords:** Ebstein anomaly, outcomes, children, adult

## P-364

**Discrepancies in quantifying coronary dilatation among standard z score nomograms in kawasaki disease and multisystem inflammatory syndrome**Akshay Aggrawal<sup>1</sup>, Sachit Ganapathy<sup>2</sup>, N.sreekumaran Nair<sup>2</sup>, Jaikummar Gr<sup>3</sup>, Narayanan Parmeswaran<sup>3</sup>, Raja Selvaraj<sup>1</sup>, Santhosh Satheesh<sup>1</sup>, Avinash Anantharaj<sup>1</sup>

<sup>1</sup>Department of Cardiology, Jawaharlal Institute of Post graduate Medical education and Research (JIPMER) Pondicherry, India; <sup>2</sup>Department of Biostatistics, Jawaharlal Institute of Post graduate Medical education and Research (JIPMER) Pondicherry, India; <sup>3</sup>Department of Paediatrics, Jawaharlal Institute of Post graduate Medical education and Research (JIPMER) Pondicherry, India

**Background and Aim:** Coronary artery (CA) dilatation is seen in Kawasaki disease (KD) and Multisystem inflammatory syndrome in

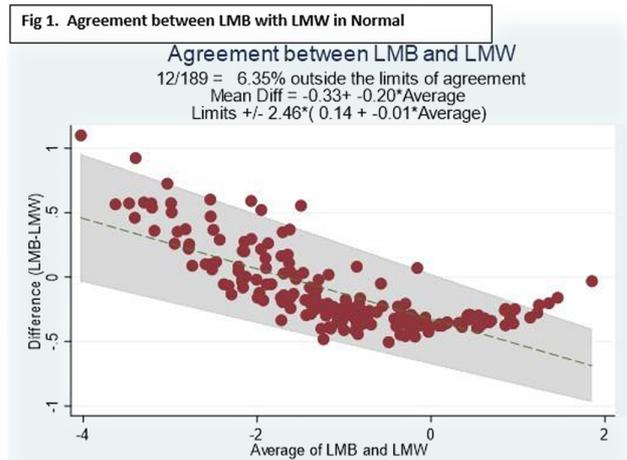
children (MISC). Various standard z score nomograms are available but there is no consensus as to which standard is appropriate for Indian children. The aim is to determine agreement between z scores derived from the existing nomograms for CA dimensions including that derived from coronary aorta index (CAI) in children belonging to the age group of 1 month- 10 years who are/were diagnosed with KD or MISC and also in normal population.

**Method:** CA dimensions were measured in normal, MISC and KD children. Z scores were calculated for each by the five standard nomograms: Boston, Washington, Montreal, Kobayashi and Pediatric Heart Network (PHN). CAI (coronary artery to aortic annulus ratio) derived z scores were also compared with z scores derived from body surface area (BSA) regression equations. The agreement between CA z scores was assessed using Bland-Altman plots.

**Results:** Amongst normal children, a high agreement was found between LM z scores by Boston and Washington methods using intraclass correlation (ICC) (ICC r value = 0.983;  $P < 0.001$ ). The best agreement was between Boston and PHN methods for LM and LAD and between Boston and Montreal methods for RCA in the normal group. In both KD and MISC groups the best agreement was between Boston and PHN methods for LAD and between Boston and Montreal for RCA. For LM in KD group agreement was better between Boston and Montreal methods and between Boston and Washington methods in the MISC group. For LCx, agreement was seen between the Z scores derived from Montreal regression equations with the square root of BSA and those derived from equations using aortic annulus diameter. This assessment suggested a comparatively lower agreement in the normal group (ICC r = 0.882) compared to that in the KD and MISC group (ICC r = 0.987, 0.909). For RCA, LAD and LM also the highest agreement was seen in the KD group for the 2 methods.

**Conclusions:** Various CA z score formulas have high agreement with Boston z scores in all three groups of normal, KD and MISC children. On comparing z scores derived from aortic annulus diameter equations to that derived from Montreal BSA regression equations highest agreement was seen in the KD group.

**Keywords:** Boston, Washington, Montreal, Kobayashi and Pediatric Heart Network (PHN), Coronary aorta index (CAI)



Amongst normal children, the agreement between LMB and LMW were assessed using intraclass correlation (ICC). It was found that the ICC r value was 0.983 (95% confidence interval: 0.978, 0.987) which was found to be statistically significant ( $P$  value  $< 0.001$ ). This suggests a very high agreement between the methods. A Bland Altman (BA) plot was plotted to visually represent the agreement between the methods. It was found that 12 (6.35%) of children were outside the limit of agreement between these two methods.

## P-366

**An *in vitro* analysis of a fenestration connecting the total cavopulmonary connection in a hypoplastic left heart syndrome patient specific phantom**Liam Morris<sup>1</sup>, Shane V Mchugo<sup>1</sup>, Lars Nolke<sup>3</sup>, Colin Mahon<sup>2</sup><sup>1</sup>Medical and Engineering Technology Gateway, Department of Mechanical and Industrial Engineering, Atlantic Technological University, ATU Galway City, Galway, Ireland.; <sup>2</sup>Department of Pediatric Cardiology Children's Health Ireland, Crumlin, Dublin 12, Ireland; <sup>3</sup>Department of Cardiothoracic Surgery, Children's Health Ireland at Crumlin, Dublin 12, Ireland

**Background and Aim:** The introduction of a fenestration into the total cavopulmonary connection for treating hypoplastic left heart syndrome has been shown to benefit patients with improved clinical outcomes and shorter postoperative care. There is limited knowledge on the impact of fenestration size and geometry on the circulation. Fenestration geometries can vary by size (3 – 6 mm diameter shunts) and shape (patch – box shaped fenestration). The main aim of this study was to examine three fenestration type geometries within a simulated *in vitro* environment.

**Method:** A set of DICOM format datasets of one anonymised post Fontan model of the total cavopulmonary connection (TCPC) circulation for a 3.7 years old male was acquired by a Toshiba Asteion CT scanner, with a pixel size of 0.78mm and a slice thickness of 0.5mm. The open-source image reconstruction software 3D slicer generated a virtual three-dimensional model. A series of thin-walled flexible *in vitro* phantom models were fabricated by 3D printing and the lost wax methodologies comprising of the superior/inferior vena cava, left/right pulmonary arteries and the univentricular heart. These phantom models were positioned within a biostimulator capable of replicating pulsatile flows, pressures and heart motion. Both 3mm and 6mm non-compliant circular fenestrations were incorporated between the TCPC and right atrium (RA). The fenestration patch was created by cutting a 4 mm window in both the TCPC and RA of the heart model and suturing a 5-0 Prolene using a running stitch. Various flow scenarios left/right PA flow splits were simulated.

**Results:** There was a reduction in pressure within the patch, 3mm and 6mm fenestration shunts when compared with the no fenestration scenario by 6.7%, 5.4% and 9.2% respectively with a corresponding reduction in energy losses of 26.5%, 9.8%, 39.2% respectively.

**Conclusions:** The inclusion of the fenestration reduces overall TCPC pressures and energy losses when compared to a non-fenestrated case. This shows that the fenestration is effective in reducing

high risk TCPC pressures while also making the improving circuit efficiency.

**Keywords:** Total cavopulmonary connection, fenestration, hemodynamics, shunt, energy losses, hypoplastic left heart syndrome.

## P-369

**Audit investigating the use of clinical investigations for children with chest pain following COVID-19 vaccination**

Danyal Memon, Ibrahim Dafalla, Ibraheem Krebit

Department of Paediatric Emergency Medicine, Children's Health Ireland at Tallaght, Dublin, D24 TN3C, Ireland

**Background and Aim:** Chest pain following mRNA vaccination for COVID-19 has been a documented symptom in the adult population, with reports of pericarditis and myocarditis in some adult cases. As national vaccination programmes became available for younger age groups, it was noted there were presentations of paediatric patients to the Emergency Department due to chest pain following COVID-19 mRNA vaccination. There have been reports of pericarditis and myocarditis diagnoses in some of these adolescent patients. This audit investigates these cases, and in particular explores the recommended investigations and guidelines for these cases. The primary aim of this audit project was to investigate the department's compliance with performing the recommended investigations in this sub-group of patients. The secondary aim was to assess if any of these investigations were consistently abnormal. We also assessed if any further investigations or clinical review was needed for these patients.

**Method:** This was a retrospective audit, assessing the data over a 11 month period between August 2021 (when vaccinations for paediatric patients aged between 12-15 years first began in Ireland) to June 2022. In total, 30 patients met the criteria for involvement in this study.

**Results:** The majority of the patients were found to be male, and symptoms were more common after a second or further vaccine dose. Regarding investigations performed, all patients had an ECG performed and reviewed in the ED. Twenty eight of the thirty patients had a troponin level performed in the ED. Only twenty patients had a chest radiograph, and only twelve patients had a creatine kinase level measured in the ED.

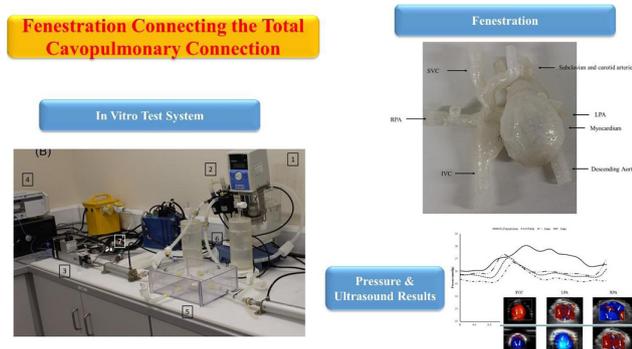
**Conclusions:** In conclusion, initial investigations of ECG and troponin are key in informing whether specialist referral and further investigations such as echocardiogram or cardiac MRI are necessary. Chest radiograph should be considered in individual cases when seeking other differentials or if assessing for pericardial effusion. Creatine kinase measurement has not been a routinely recommended investigation of choice by the international guidelines.

**Keywords:** COVID-19, chest pain, vaccination, myocarditis, pericarditis

## P-371

**All Island congenital heart network brings diagnosis closer to home**Daragh Finn<sup>1</sup>, Samy Allawendy<sup>2</sup>, Eugene Dempsey<sup>1</sup>, Colin J Mc Mahon<sup>3</sup><sup>1</sup>Department of Paediatrics and Child Health, University College Cork, Ireland; <sup>2</sup>Department of Paediatrics and Child Health, Cork University Maternity Hospital, Ireland; <sup>3</sup>Department of Paediatric Cardiology, Children's Health Ireland, at Crumlin, Ireland

**Background and Aim:** The All-Island congenital heart network appointed paediatricians with expertise in cardiology in regional centres. Prior to these appointments children with suspected



Showing the *in vitro* (experimental) setup and flow and pressure results for Fenestration Connecting the Total Cavopulmonary Connection within a Hypoplastic Left Heart Syndrome Patient Specific Phantom

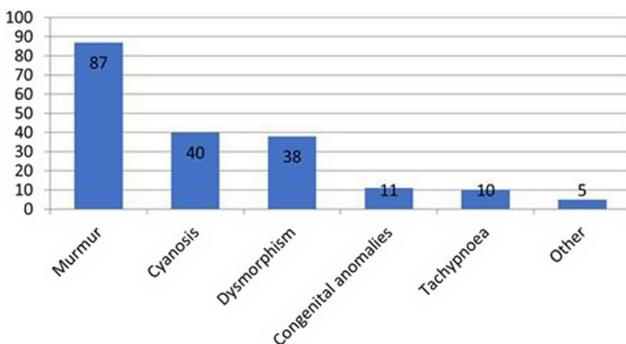
congenital heart disease were referred to the national children's heart centre for investigation. The aim of this study is to quantify paediatric cardiology activity in a regional Irish centre over the first year of service provision.

**Method:** Data was collected retrospectively on all inpatient neonatal referrals over a 12-month period (January 2019 to January 2020).

**Results:** There were 268 neonatal referrals. Premature infants (< 37 weeks gestation) accounted for 26% (n = 69) of total neonatal referrals. Congenital cardiac disease was identified in 58.5% (n = 113) of referrals. Cardiac intervention in the first year of life was required in 24 infants, 12.2% of referrals (5.6% catheter and 6.6% surgery).

**Conclusions:** Our report displays how clinical networks of care can reduce hospital transfers from regional neonatal centres for non-invasive cardiology investigations.

**Keywords:** Congenital cardiac disease, Neonatal, Clinical Network



Indication for paediatric cardiology referral

### P-372

#### Staphylococcus lugdunensis endocarditis and pseudoaneurysm formation in a patient with bicuspid aortic valve.

Georgios Meridis, Edward Peng  
Scottish Paediatric Cardiac Service, Royal Hospital for Children,  
Glasgow, Scotland, UK

**Background and Aim:** Acute left sided endocarditis with aortomitral curtain involvement is not uncommon, but formation of a sizeable pseudoaneurysm cavity is rare.

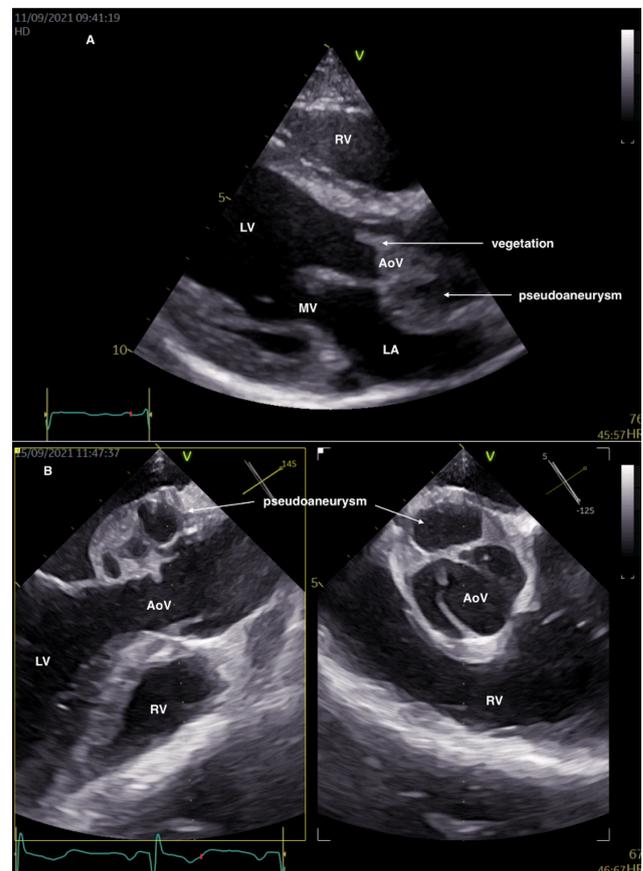
**Method:** This abstract reports the case of a patient with Turner syndrome (TS) and bicuspid aortic valve (BAV), who developed aortic root endocarditis, abscess and pseudoaneurysm, requiring surgery during the acute phase.

**Results:** A 14-year-old, previously healthy patient with TS and BAV, presented to a District General Hospital (DGH) with prolonged pyrexia. She was initially treated for Paediatric Inflammatory Multisystem Syndrome Temporally associated with SARS-COV-2 (PIMS-TS) before getting transferred to the Royal Hospital for Children (RHC) in Glasgow. On admission, she was found to have new onset moderately severe aortic regurgitation (AR). Transthoracic echocardiography (TTE) demonstrated aortic valve vegetation, root abscess with evidence of posterior left ventricular outflow tract obstruction (LVOT) pseudoaneurysm. Transoesophageal echocardiography (TOE) further delineated a complex root abscess with Doppler flow into this pseudoaneurysm cavity in the aorto-mitral curtain. Most aortic regurgitation flow

was seen to be through the cystic connection, rendering it essentially a left ventricle to aorta tunnel, resulting in progressively more severe aortic regurgitation (AR). Staphylococcus Lugdunensis was isolated from blood culture, which is a coagulase negative staphylococcus, well known to be associated with endocarditis. Despite appropriate antibiotic therapy serial CT Angiogram imaging showed progressive enlargement of the aortic root abscess-pseudoaneurysm complex, in addition to splenic infarct, multiple cerebral septic emboli and new hyperdense lesion raising concern of focal intracranial haemorrhage. Further CT brain suggested likelihood of mycotic aneurysm rather than bleed. Patient underwent urgent repair of LVOT pseudoaneurysm using aortic wall autograft to reconstruct aorto-mitral curtain, debridement of aortic root endocarditis, and bicuspid aortic valve repair. Patient had uneventful recovery and was extubated on postoperative night.

**Conclusions:** Surgery in acute phase and without local infection control is associated with friable tissues, higher risk of dehiscence and recurrent infection. Timing of surgery is further compounded by intracerebral lesions and risks of intracranial bleed during cardiopulmonary bypass. Progressive enlargement of abscess, false aneurysm cavity, risk of rupture, and worsening of AR dictated an emergent surgical intervention, balanced against significant risk of peri-operative surgical and neurological complications.

**Keywords:** Endocarditis, Staphylococcus Lugdunensis, pseudoaneurysm



**Figure 1.** Echocardiography. A. Transthoracic mode, parasternal long axis view of the vegetation and pseudoaneurysm. B. Transoesophageal mode, long and short axis views. AoV, aortic valve, LA, left atrium, LV, left ventricle, MV, mitral valve, RV, right ventricle.

## P-374

**Analysis of heart rate variability in patients with williams-beuren syndrome**

Heike Elisabeth Schneider, Assunta Schiebel, Thomas Paul, Matthias Johann Mueller, Verena Dorothea Gravenhorst  
Department of Pediatric Cardiology, Intensive Care Medicine and Neonatology, University Medicine Goettingen, Goettingen, Germany

**Background and Aim:** Arterial hypertension and tachycardia are common features in patients with Williams-Beuren Syndrome (WBS). In addition, heart rate variability is thought to be decreased. Data concerning this observation are sparse. Underlying mechanisms such as increased sympathetic tone that may lead to this phenomenon have not yet been studied in detail. We evaluated data collected from 24-hour-Holter monitoring (HM) from WBS patients at our institution.

**Method:** Screening our medical records from 2011–2022, we identified 24 patients with WBS in whom HM had been performed for different reasons. Patients with significant supraventricular and ventricular extrasystoles were excluded. We evaluated 4 typically used time domain measures: pNN50, RMSSD, SDNN, SDANN and mean heart rate (HR) and compared it to values available for the general population.

**Results:** In the first 3 decades of life, mean HR and all 4 time domain measures are significantly decreased in WBS patients with no overlap to values from healthy subjects (Table 1). This difference becomes less with age. With the 4th decade mean heart rate, pNN50% and RMSSD are similar in WBS and healthy controls, while SDNN and SDANN remain different but to a lesser degree. Interestingly, while in the healthy populations all these parameters decrease with increasing age, in WBS patients pNN50 and RMSSD remain in the same range, SDANN and SDNN rather increase slightly.

**Conclusions:** It has been hypothesized that the loss of medial elastin results in baroreflex insufficiency contributing to vagal inhibition and in some degree to sympathetic activation. Potential clinical significance of this phenomenon remain subject of necessary research in this rare disease. If decreased heart rate variability may be associated with accelerated aging remains to be elucidated.

**Keywords:** Williams-Beuren syndrome, heart rate variability, elastin arteriopathy

Age		Heart rate per min	pNN50% %	RMSSD ms	SDANN ms	SDNN ms
Age <= 19 years	WBS subjects n=7	92 ± 14	4 ± 3	25 ± 12	42 ± 22	87 ± 37
	Healthy controls *	80 ± 10	25 ± 13	53 ± 17	159 ± 35	176 ± 38
20-29 years	WBS subjects n=7	91 ± 10	4 ± 3	26 ± 8	40 ± 12	97 ± 34
	Healthy controls *	79 ± 10	18 ± 13	43 ± 19	137 ± 43	153 ± 44
30-39 years	WBS subjects n=7	83 ± 10	4 ± 2	34 ± 13	48 ± 12	91 ± 20
	Healthy controls *	78 ± 7	13 ± 9	35 ± 11	130 ± 33	143 ± 32
40-59 years	WBS subjects n=3	85 ± 13	4 ± 2	30 ± 8	51 ± 18	102 ± 36
	Healthy controls *	78 ± 7	10 ± 9	31 ± 11	116 ± 31	132 ± 30
50-59 years		76 ± 9	6 ± 6	25 ± 9	106 ± 27	121 ± 27

\*Reference: Umetani K, et al., JACC 1998

Data are presented as mean and standard deviation

## P-375

**Neonatal giant fibromas. an overview of surgical strategies**

Pia Cassanello, Carin Walter, Cecilia Mandirola, Juan Carretero  
Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain

**Background and Aim:** Primary cardiac tumors in childhood are rare and usually benign. Cardiac fibromas (CF), the second most common tumors following rhabdomyomas, are rarely

encountered in neonates. Although benign, CF prognosis depends on its characteristics, size, and location. The most frequent location is the left ventricle (LV), and less commonly the right ventricle or atrium. Patients might remain asymptomatic or present with inflow/outflow obstruction, arrhythmias, congestive cardiac failure, and even sudden death. Surgical resection is indicated, especially in symptomatic patients with hemodynamic impairment.

**Method:** We report two neonates with giant CF prenatally diagnosed and discuss their surgical management.

**Results:** Patient 1:

A 41 year-old primigravida was referred at 32 weeks of gestation with diagnosis of a large ventricular mass. Fetal echocardiogram revealed a giant heterogeneous mass (30x30mm) outgrowing the interventricular septum to the LV. Genetic test ruled out tuberous sclerosis (TS). Postnatal echo confirmed a highly echogenic solid mass with poor vascularization. The patient remained asymptomatic until 5 months of age, when episodes of sustained ventricular tachycardia were observed. Cardiac MR revealed a giant cardiac interventricular septum mass (65x63x57mm) occupying practically the entire left endocavitary lumen. Surgical resection was performed at another institution (parents' decision), achieving total tumor resection but the patient died 15 days later.

Patient 2:

A 29 year-old multipara was referred at 36 weeks with a diagnosis of cardiac mass. Genetic testing was negative for TS. Postnatal echocardiogram and Cardiac MR showed a giant cardiac mass located at the lateral wall of the LV measuring 23x38x42mm with left coronary involvement. Cardiac function was normal without obstructive signs. MR tissue characterization was consistent with CF diagnosis. The patient underwent successful partial tumor resection at three months of life due to severe mitral regurgitation. During the immediate post-operative period, he presented ventricular arrhythmias requiring amiodarone. The patient is currently nine months, remains asymptomatic, free of arrhythmias and no tumor growth evidenced in follow-up echocardiograms.

**Conclusions:** CF surgical strategies recommend total or subtotal tumor resection as extensive Mass debulking might be sufficient to reverse its arrhythmogenic substrate. Long-term follow-up is necessary to monitor arrhythmias, RV inflow/outflow obstruction and valvar function. 3D-technology might assist preoperative planning and decision making.

**Keywords:** Pediatric cardiac tumors, cardiac fibroma

## P-376

**Current management practices in giant coronary artery aneurysm and kawasaki disease: A global perspective**

Mia Chen<sup>1</sup>, Audrey Dionne<sup>2</sup>, Nadine F Choueiri<sup>3</sup>, Ashraf S

Harahsheh<sup>4</sup>, Elif Seda Selamet Tierney<sup>5</sup>, Nagib Dahdah<sup>1</sup>

<sup>1</sup>CHU Sainte-Justine, University of Montreal; <sup>2</sup>Boston Children's Hospital, Harvard Medical School; <sup>3</sup>Children's Hospital at Montefiore, Albert Einstein College of Medicine; <sup>4</sup>Children's National Hospital, The George Washington University School of Medicine & Health Sciences; <sup>5</sup>Stanford University

**Background and Aim:** Management of giant coronary artery aneurysms (CAA) a rare complication of Kawasaki disease (KD) varies due to limited evidence-based data. We describe the variation in surveillance, pharmacological and interventional management, in KD patients with giant CAA.

**Method:** An international survey of clinical vignettes was sent to physicians caring for KD patients through various distribution lists and contacts. Chi-squared test was used, p<0.05 was significant.

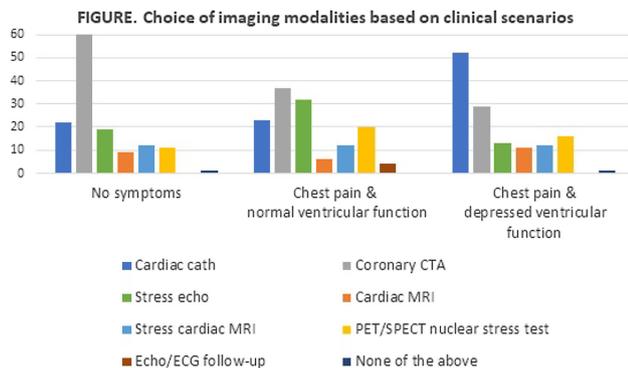
**Results:** 134 physicians responded from North-America (35%), South-America (37%), Asia (15%), Europe (8%) Middle-East

and North-Africa (5%); 89% pediatric cardiologists (29% of them interventionalists), and 11% other specialties. 53% followed patients every 6 months, 35% every 3 months and 11% every 9–12 months; 6 months follow-up was more common in North-America (80% vs 39%,  $p < 0.001$ ). Coronary CT-scan (CTA) was the preferred imaging, with incremental preference for invasive imaging (Cardiac cath) with worsening scenarios (FIGURE). Region ( $p = 0.327$ – $0.948$ ) or interventionalist vs. non-interventionalist ( $p = 0.138$ – $0.489$ ) did not skew preferences. 80% used combination of aspirin and anticoagulants, regardless of patient's age or presence of CA stenosis. Beta-blockers were used preferentially in those with CA stenosis (52% vs 18%,  $p < 0.001$ ) regardless of age. Statin use varied with age (43% in 12 years-old vs 12% in 2 years-old,  $p < 0.001$ ) and CA stenosis status (35% if stenosis present vs 19% if no stenosis,  $p < 0.001$ ). 31% relied on symptoms as an indication for coronary interventions, 18% on presence of inducible ischemia, 27% on abnormal perfusion, and 21% on CA imaging. Percutaneous coronary intervention (PCI) was preferred in CA stenosis (69% PCI vs 31% CABG) and thrombosis (40% PCI, 31% intra-coronary thrombolysis, 19% systemic thrombolysis, 11% CABG), regardless of specialty ( $p = 0.394$ – $0.437$ ) or region ( $p = 0.664$ – $0.536$ ). Interventions done by a team of pediatric and adult interventionalists were preferred, regardless of age (80% for 6-year-old vs 82% for 16-year-old). A pediatric center was preferred in the young for both PCI (92% pediatric center vs 8% adult center) and CABG (89% pediatric center vs 11% adult center).

**Conclusions:** Physician practices varied in the management of giant CAA in KD regardless of specialty or location. Robust data is needed to measure impact and guide management of these patients.

**Keywords:** Kawasaki disease, coronary aneurysm intervention, work-up

Imaging modalities based on advanced clinical scenarios



Responders selected their preferred imaging modalities for cases with asymptomatic giant CA aneurysms, or with chest pain, or chest pain and depressed ventricular function. (CTA = Coronary CT-scan Angiography; Cardiac Cath = invasive selective coronary angiography).

### P-377

#### Parental understanding of their child's cardiac medical therapy

Michaela Pentony<sup>1</sup>, Esme Dunne<sup>1</sup>, Kathryn Shaw<sup>2</sup>, Orla Franklin<sup>1</sup>, Adam James<sup>1</sup>, Terence Prendiville<sup>1</sup>, Colin J McMahon<sup>3</sup>

<sup>1</sup>Department of Paediatric Cardiology, Children's Health Ireland, Crumlin, Dublin 12, Ireland; <sup>2</sup>Pharmacy Department, Children's Health Ireland, Crumlin, Dublin 12, Ireland; <sup>3</sup>Department of Paediatric Cardiology, Children's Health Ireland, Crumlin, Dublin 12, Ireland;

School of Medicine, University College Dublin, Belfield, Dublin 4, Ireland

**Background and Aim:** In the cardiology outpatient department, we have frequently encountered parents and caregivers lacking certainty of what cardiac medications and/or what dosing regimen their child has been prescribed. We have also encountered issues where accidental overdoses have been given where the parent does not know what to do or who to contact for help. We aimed to assess parental knowledge and understanding of their child's cardiac medications in a national tertiary paediatric cardiology centre. **Method:** A 24-part questionnaire was created to assess knowledge regarding indication for medication, dosing in milligrams (mgs) and millilitres (mls) and potential side effects. Parental perception of confusion with their child's medication and satisfaction with hospital discharge information was assessed. Other information obtained included details of pharmacy education provided. The questionnaire was conducted in a face-to-face interview style to enable investigators to qualify answers given, and also allow parents the opportunity to provide commentary on specific questions in order to gain valuable qualitative information.

**Results:** Fifty interviews were conducted with parents and caregivers when attending their cardiology outpatients appointment. The majority of parents (36/50, 72%) stated they received education from a hospital pharmacist, and most (42/50, 84%) clearly understood the information they received. Despite most parents, (37/50, 74%) stating that they were not confused regarding their child's medications, only 13/37 (35%) knew the correct name, frequency, and dose in milligrams and/or millilitres. Two-thirds of all parents interviewed (33/50, 66%) answered one or more of these questions incorrectly. The majority 35/50 (70%) of parents were aware of resources to inform themselves about their child's medications, including asking their pharmacist, or online resources. Less than half of parents knew the correct procedure if they inadvertently administered an incorrect dose of medication, 21/50 (42%).

**Conclusions:** Despite having received prior medicine education, a significant percentage of parents fail to understand the specific dosing regimen for their child. This study has shown that further strategies are required to improve parental knowledge. Development of medication mobile application (app) would represent one particular strategy. Additional qualitative data will aid in development of such an educational tool for informing parents.

**Keywords:** Medication, education, parents, caregivers

### P-378

#### Infants with hypoplastic left heart syndrome and their outcomes before and after stage 1 surgery: the belfast experience

Claire McGinn<sup>1</sup>, Ian Scott Kendall<sup>2</sup>, Rachel Bates<sup>2</sup>, Frank Casey<sup>1</sup>  
<sup>1</sup>Royal Belfast Hospital for Sick Children, Belfast, UK; <sup>2</sup>Queen's University, Belfast, UK.; <sup>2</sup>Royal Belfast Hospital for Sick Children, Belfast

**Background and Aim:** Hypoplastic Left Heart Syndrome (HLHS) accounts for 5–10% of critical congenital heart disease and is associated with significant mortality in the first year of life. The Royal Hospital, Belfast, is a Level 2 centre which facilitates fetal echocardiography, delivery of infants with HLHS and long-term postoperative care. It transfers patients to surgical centres in the UK and Ireland for staged palliation. This study covered a significant time period where termination of pregnancy was illegal for cardiac disease in Northern Ireland. Aim was assessment of the centre's initial

diagnosis, stabilisation, and early surgical outcomes in patients with HLHS.

**Method:** Retrospective analysis of all live births in Northern Ireland diagnosed with HLHS from 01/01/2002 to 01/09/2022. Data collated included antenatal diagnosis, sex, birthweight, gestation, echocardiogram findings, decision to proceed to surgery, surgical centre, surgical procedure, mortality in first 30 days postoperatively and mortality prior to 2nd stage.

**Results:** Seventy patients were included. 49/70(70%) had an antenatal diagnosis and 54/70(77%) were male. The mean birthweight was 3.11kg and mean gestation was 39 weeks. 6/70(9%) had a low birth weight <2.5kg.

All patients underwent echocardiography. A restrictive atrial septum was identified in one patient, significant tricuspid regurgitation in five (7%) and reduced right ventricular function in seven (10%).

11/70(16%) of patients died before progressing to Stage 1 surgery. Of those who proceeded to surgical palliation, 49/59(83%) underwent a Norwood-Sano, 6/59(10%) a Norwood modified BT-shunt and other procedures in 4/59(7%). There was one patient death in the immediate perioperative period resulting in 98% 30-day-postoperative survival. Following initial stabilisation post-surgery, all surviving patients returned to our centre. The mortality rate between stage 1 and stage 2 Norwood was 8/70(12%).

**Conclusions:** This retrospective study demonstrates comparable early outcomes to cardiac centres worldwide. One limitation of this study is that our centre does not electively deliver infants requiring earlier surgical intervention e.g. presence of restrictive atrial septum. The majority of our cohort had favourable anatomy, birthweight >2.5kg, progressed to Stage 1 and had excellent 30-day postoperative survival. Importantly, our outcomes highlight that carefully selected patients delivered in a non-surgical centre can be optimised prior to transfer for Stage 1 surgery.

**Keywords:** Norwood, Hypoplastic, fetal, palliation

### P-379

#### **Multisystem inflammatory syndrome in children (MIS-C) related to a SARS-COV-2: tunisian experience**

Rania Gargouri, Ines Maaloul, Kolsi Roeya, Elleuch Amal, Hajer Aloulou, Thouraya Kammoun, Leila Abid

department of cardiology and department of pediatrics, HEDI CHAKER university hospital, Sfax, TUNISA

**Background and Aim:** multisystem inflammatory syndrome in children (MIS-C) related to a novel coronavirus (SARS COV2) is a new emerging disease in childhood and may be serious engaging the prognosis because of cardiac complications. The aim of this study is to analyze cardiac complications in MIS-C.

**Method:** a retrospective study was conducted in the pediatric department of Hedi Chaker Hospital in Sfax(TUNISIA.) All children under 15 years and met the criteria of MIS-C related to SARS COV2 were included from January 2020 to March 2022.

**Results:** twenty-seven patients were included. They were 15 boys and 12 girls. The median age was 5, 58 ans +/-3, 37and the sex-ratio was 1, 25. Echocardiography was performed in all children and revealed abnormalities in 24 patients. Cardiac involvement included in the majority of cases coronary artery lesions. Myocarditis with low left ventricular ejection fraction was noted in seven children and pericarditis in six patients.

**Conclusions:** the prognosis of MIS-C in the short term depends on cardiac involvement. The mean cardiac complications are represented by myocarditis and coronary artery lesions

**Keywords:** MIS-C; COVID19, cardiac involvement; echocardiography; management

### P-380

#### **Characteristics of heart rate variability in children and adolescents diagnosed with psychiatric disorders**

Loriana Kilaitė<sup>1</sup>, Kamilė Plėšnytė Čapkauskienė<sup>2</sup>

<sup>1</sup>Vilnius university hospital Santaros Klinikos, Paediatrics Centre, Vilnius, Lithuania; <sup>2</sup>Vilnius university hospital Santaros Klinikos, Child development center, Vilnius, Lithuania

**Background and Aim:** Psychiatric disorders are well known to affect heart rhythm via sympathetic stimulation and impulse propagation in the atrioventricular node via  $\beta$ -receptor stimulation. This may lead to the prescribing of cardiovascular medication. But good monitoring and treatment of psychiatric disorders could allow us to avoid unnecessary cardiologic drugs. Our aim is to evaluate the heart rhythm control of psychiatric disorders, with or without medication.

**Method:** This is an ongoing pilot study. We are gathering information on children aged 7 to 17 who have psychiatric illnesses, focusing in particular on heart rhythm, ECG, 24 h - holter monitoring, and treatment. The study is being carried out at the Vilnius University Hospital's Child Development Center from 2022-10 to 2022-11.

**Results:** We gathered 30 patients data, yet there is no clinical significans between age groups or disorders. But individual cases already show the importance of this study. A 16-yo female was being treated at the Child Development Center for a moderate depressive episode. She was also referred to a paediatric cardiologist for tachycardia. On examination the ECG showed a sinus rhythm of 133 beats per minute, on 24h holter monitoring, the heart rhythm varies between 65 and 152 beats per minute. After 6 month of psychotherapy and treatment with sertraline 150 mg/d and quetiapine prolonged release 200 mg/d, her heart rhythm was at age average (norm)

**Conclusions:** Understanding the harm caused by polypharmacy is important given the prevalence of mental and behavioral issues in children and adolescents and the sensitivity of this population. As a result, any medical treatment option should be thoroughly assessed and supported. As a result, if heart rate variability could be better distinguished, it would be easier to assess the choice and strategies of both drug and non-drug treatment, as well as predict long-term effects.

**Keywords:** irritable heart, tachycardia, paediatric, psychosomatic symptoms

### P-381

#### **Prevalence and significance of captopril cough in paediatric population - a retrospective telephone study**

Michael Coffey<sup>1</sup>, Mike Shields<sup>2</sup>, Brian Mccossan<sup>1</sup>, Rachel Moore<sup>1</sup>

<sup>1</sup>Royal Belfast Hospital for Sick Children, Belfast, United Kingdom;

<sup>2</sup>Royal Belfast Hospital for Sick Children, Belfast, United Kingdom, Queen's University Belfast, Belfast, United Kingdom

**Background and Aim:** Captopril cough (cc) is a well-known side effect of ace-inhibitors (ACE-i) in an adult population with an incidence up to 35% and is the most frequent indication for discontinuation. Anecdotally, there is a feeling that it is not as prevalent in the paediatric population but there is little evidence to confirm this. The aim of this study was to survey a paediatric population prescribed regular ACE-i to assess the prevalence of cc and whether it impacted upon patient management.

**Method:** This was a retrospective telephone study performed in 2020 in a tertiary centre in the United Kingdom. Patients aged less than 16 years old who were prescribed regular ACE-I therapy were eligible for inclusion. The main outcomes measures were (1) presence of chronic cough & (2) whether ACE-I was discontinued due to presence of cough.

**Results:** 100 consecutive patients were enrolled and reported their experiences with ACE-i in this study. 59 patients received captopril, 21 received lisinopril, 12 received enalapril, and 6 received perindopril. 54 patients were aged between 0-4, 14 aged between 5-9, 12 aged between 10-14 and 20 aged between 15-20. Fifteen patients (15/100) experienced symptoms typical of cc. 11/59 (18.6) of patients taking captopril, 2/21 (9.5%) of patients taking lisinopril, 2/12 (16.7%) of patients taking enalapril, and 0 patients taking perindopril reported cc. Only one patient (1%) experienced a chronic cough resulting in discontinuation of ACE-i (captopril).

**Conclusions:** Children also frequently experience chronic cough in association with ACE-i therapy, however, in this cohort rarely did it necessitate a change in management.

**Keywords:** Ace-inhibitor, captopril, cough

### P-382

#### Medication management and ECG screening in children with ADHD

Michaela Pentony<sup>1</sup>, Zainab Ali<sup>2</sup>, Kieran Moore<sup>2</sup>, Fiona McNicholas<sup>3</sup>, Adam James<sup>1</sup>

<sup>1</sup>Department of Paediatric Cardiology, Children's Health Ireland, Crumlin, Dublin 12, Ireland; <sup>2</sup>Department of Psychiatry, Children's Health Ireland, Crumlin, Dublin 12, Ireland; <sup>3</sup>Department of Psychiatry, Children's Health Ireland, Crumlin, Dublin 12, Ireland; CAMHS Lucena Clinic; Department of Academic Child & Adolescent Psychiatry, UCD

**Background and Aim:** Pharmacological treatment for children with ADHD (Attention deficit hyperactivity disorder) has a robust evidence base, with approved medications including stimulant (eg. Methylphenidate) and non-stimulant options (eg. Atomoxetine). International guidelines (NICE) exist for cardiovascular (CVS) screening prior to medication use in youth with ADHD and specialist cardiology opinion and/or ECG screening is not recommended without a clinical indication (1). We aimed to quantify the number of referrals sent to the Crumlin Cardiology Department prior to commencement or modifying ADHD medication, assess the number of which detected a clinically significant abnormality, and ascertain whether the referrals were indicated as per best practice guidelines.

**Method:** A prospective audit was performed over a 6-month period, from November 2021 – April 2022 inclusive. Referrals sent via OPD triage letters, ECG department email (ecg.review@olchc.ie) and walk-in ECG service were screened for those pertaining to commencing or modifying medication for children with ADHD. Each referral was coded against NICE guideline to determine the degree of clinical details given. Reported abnormalities, recommended management and correspondence were recorded.

**Results:** Ninety-one referrals were received in total during the 6-month audit period. There was no clinical indication for referral in 53/91 (58.2%). Fewer than one third, twenty six (26/91, 28.5%) met NICE criteria for referral for cardiology opinion including concern regarding clinical symptoms, personal or family history of cardiac disease. Of the 76 clinically reviewed referrals, seventy-one (93.4%) were reported as normal with no cardiology follow up required. Five referrals (6.6%) required further cardiology review, 2 were considered incidental and not a contraindication

for ADHD medication use. Three referrals required further cardiology opinion prior to medication use, all of which met NICE criteria for referral.

**Conclusions:** Routine screening prior to ADHD medication prescription in the absence of clinical indications (as per NICE) contributed to delays in medication initiation and has resource implications for clinical teams involved. Creation of a CVS referral checklist will improve adherence to NICE guidelines and should provide benefits for patients and clinicians.

**References:**

1. National Institute for Health and Care Excellence: Guidelines. Attention deficit hyperactivity disorder: diagnosis and management. London: National Institute for Health and Care Excellence (NICE) 2018.

**Keywords:** ECG, screening, ADHD, medication

### P-383

#### Additional challenges of feeding and growing infants with trisomy 21 and congenital heart disease

Michaela Pentony<sup>1</sup>, Sarah Hayes<sup>2</sup>, Marie Lavelle<sup>1</sup>, Noeleen Allen<sup>2</sup>, Allan Jenkinson<sup>1</sup>, Orla Franklin<sup>1</sup>

<sup>1</sup>Department of Paediatric Cardiology, Children's Health Ireland, Crumlin, Dublin 12, Ireland; <sup>2</sup>Department of Nutrition and Dietetics, Children's Health Ireland, Crumlin, Dublin 12, Ireland

**Background and Aim:** Ireland has the highest incidence of Trisomy 21 (T21) in Europe, 1 in 546 live births. Congenital heart disease (CHD) occurs in 50% of patients with T21. The optimal time for surgical repair of complete AVSD (cAVSD) or VSD is between 3 and 6 months of life with a target weight of 5kg (1). We describe the growth trajectory and dietetic intervention strategies in preoperative infants with T21 to optimise them and compared them with infants with matched CHD with a normal chromosome complement.

**Method:** The Children's Health Ireland at Crumlin NICOR database identified patients who had surgical repair of cAVSD or VSD between 1st January 2019 – 31st December 2020. Retrospective chart review was undertaken to obtain demographic, medical and nutritional data.

**Results:** Sixty patients with T21 and 50 patients with a normal chromosome complement (non-T21) met inclusion criteria. The majority of infants required calorie supplementation pre-operatively – 51/60, (85%) in the T21 group and 40/50 (80%) in the non-T21 group. Nasogastric tube (NGT) placement was required for 37/60 (62%) patients with T21 and 26/50 (52%) of non-T21 group. Seventeen patients 17/60, (28%) were receiving some breastmilk at surgery in the T21 group compared with 10/50 (20%) in the non-T21 group. Three patients in the T21 group (3/60, 5%) and two patients in the non-T21 (2/50, 4%) exclusively received breastmilk at time of surgery. On average, ten dietetic contacts were required to wean NGT post-operatively in the T21 group compared with 3.5 contacts in the non-T21 group.

**Conclusions:** Infants with T21 who require cardiac surgery have significant additional nutritional needs and require multidisciplinary professional input to achieve target weights for surgical intervention. This data will enable us to accurately counsel parents regarding the expected challenges surrounding feeding their child prior to cardiac surgery.

(1) Xie O, Brizard CP, d'Udekem Y, Galati JC, Kelly A, Yong MS, et al. Outcomes of repair of complete atrioventricular septal defect in the current era. *Eur J Cardiothorac Surg.* 2014;45(4):610-7.

**Keywords:** Trisomy 21, nutrition, feeding, surgery

## P-384

**Reversible string of pearls-like arterial spasms during cardiac catheterization in patients with williams-beuren syndrome - a reason for concern?**

Verena Dorothea Gravenhorst, Matthias Johann Mueller, Thomas Paul, Heike Elisabeth Schneider

Department of Pediatric Cardiology, Intensive Care Medicine and Neonatology, University Medicine Goettingen, Germany

**Background and Aim:** Williams-Beuren Syndrome (WBS) is a contiguous gene syndrome as a result of a microdeletion in 7q11.23 leading to an elastin arteriopathy which can cause stenosis or other alteration in all arteries. Patients with WBS have been reported to be at high risk for anesthesia-related adverse events in medical interventions. However, to date, the reasons for this effect are not finally known.

**Method:** Analyzing our medical records on heart catheterization, we identified 2 patients in our WBS-cohort who developed arterial spasms during the examination.

**Results:** Heart catheterization for further clarification and therapy planning was performed in a 25-year-old woman with WBS suffering from abdominal angina due to severe stenosis of the superior mesenteric artery visualized by computed tomography. Apart from severe long segment stenosis of superior mesenteric artery and minimal supravalvular aortic stenosis, this examination showed mild mid-aortic syndrome and moderate stenosis of the truncus celiacus and left renal artery. Injection of contrast agent into the right femoral artery led to a string of pearls like spasm of right iliac artery, which was nearly reversible following the administration of intervals of 2 ml Xylocain 1% in the artery. Similar findings were observed in another 26-year-old female with WBS.

**Conclusions:** After receiving contrast agent, these patients with WBS displayed a string of pearls-like spasm in an otherwise morphologically normal right iliac artery. It may be hypothesized, that a local reaction to pharmacologic or mechanic stimuli may also cause transient functional stenosis of the coronary arteries in this susceptible group potentially leading to fatal outcome.

**Keywords:** Williams-Beuren syndrome, arterial spasm, elastin arteriopathy

## P-385

**Eating disorder with swallowing difficulty – should we investigate for vascular ring?**

Laura Konta<sup>1</sup>, Andrew Tometzki<sup>1</sup>, Alison Hayes<sup>1</sup>, Vico Schol<sup>2</sup>, Giovanni Biglino<sup>2</sup>, Massimo Caputo<sup>2</sup>, Mari Nieves Velasco Forte<sup>1</sup>

<sup>1</sup>Bristol Royal Hospital for Children; <sup>2</sup>Bristol Medical School, University of Bristol

**Background and Aim:** Aberrant right subclavian artery (ARSA) is quite common, occurring in 0.5% to 1.8% of the population. Most cases are asymptomatic but extrinsic compression of the oesophagus may lead to dysphagia in adults, with few cases described in childhood. We report two adolescents who were followed up in the eating disorder clinic. They suffered from swallowing difficulty for years before they were investigated for an organic cause.

**Method:** Clinical history and imaging for these patients were retrospectively reviewed.

**Results:** A 14-year-old girl with history of swallowing difficulty for four years and weight loss was followed up in the eating disorder clinic. She complained of dysphagia, which was more prominent with large pieces of food. Several years after the initial complaints, she underwent an oesophageal video fluoroscopy, which showed an extrinsic compression of the thoracic oesophagus at the level of

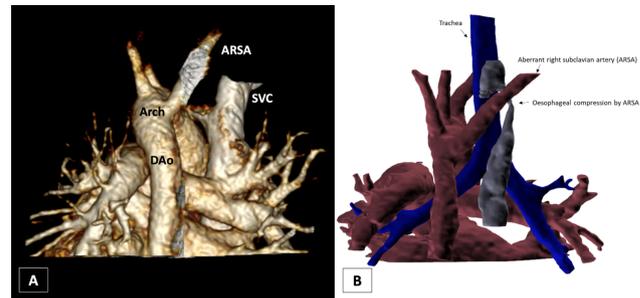
the aortic arch with holdup of liquids and solids. A CT angiogram confirmed left aortic arch and ARSA causing oesophageal compression. She underwent ARSA re-implantation directly into the right carotid artery from right thoracotomy. She developed right sided Horner syndrome following the procedure which has improved up to date. She reported that the swallowing difficulty immediately improved and she is now managing solids without symptoms.

A 15-year-old girl had difficulty and discomfort swallowing since the age of 7, subsequently developing rejection of food and vomiting. She was referred and followed up in the eating disorder clinic. Several years after the initial complaint, she had a barium swallow at the age of 14, which showed the external compression of the oesophagus, and a CT angiogram confirmed left aortic arch with ARSA. She underwent re-implantation of the ARSA directly into the right carotid artery from right posterolateral thoracotomy. As in the previous case, her swallowing improved immediately after the procedure.

**Conclusions:** Although eating disorders are common, it is important to consider an organic cause when there are symptoms of swallowing difficulty at presentation. Dysphagia lusoria should be investigated accordingly, as reimplantation of the ARSA can successfully relieve symptoms and immensely improve the quality of life of these patients.

**Keywords:** aberrant right subclavian artery, dysphagia lusoria, vascular ring, eating disorder

Oesophageal compression by aberrant right subclavian artery



A) 3D reconstruction from MDCT showing left aortic arch with aberrant right subclavian artery. B) 3D segmentation including arch anatomy and its relationship with the trachea and oesophagus.

## P-386

**Anthracycline chemotherapy and cardiac toxicity measurement in pediatric patients- a single center experience**

Derya Karpuz<sup>1</sup>, Begümhan Demir<sup>2</sup>, Derya Duman<sup>1</sup>, Fatih Sağcan<sup>2</sup>, Sevan Tuğ Bozdoğan<sup>3</sup>

<sup>1</sup>Department of Paediatric Cardiology, Mersin University Hospital, Mersin, Türkiye; <sup>2</sup>Department of Paediatric Hematology and Oncology Mersin University Hospital, Mersin, Türkiye; <sup>3</sup>Department of Genetics, Çukurova University Hospital, Adana, Türkiye

**Background and Aim:** To investigate the cardiotoxicity indexes in children with malignant tumors after the administration of anthracycline (ANT) chemotherapy.

**Method:** Data from 63 children with malignant tumors who were treated using ANT chemotherapy at our hospital from January

2010 to September 2019 were collected to analyze the echocardiography parameters, blood pressure and serologic tests (troponin and BNP levels) cumulative dose of doxorubicin. As a result of the examinations, 17 healthy children with normal Echo, ECG and cardiovascular conditions or no other chronic disease were included in the study as the control group. All the patients were grouped based on their cumulative doses of ANT: namely groups 1 <200 mg/m<sup>2</sup> (low dose) in 27 patients and ≥200 mg/m<sup>2</sup> (high dose) in 19 patients.

**Results:** Diastolic blood pressure and troponin levels were statistically different between the groups. While the diastolic blood pressure of the control group was higher (77, 8±15, 5) than the other two groups (p <0, 001), the troponin level of the low-dose group was found to be lower than the troponin level of the other two groups (p 0, 008). When the patient group and the control group were compared, the diastolic blood pressure of the patient group were found to be lower than the control group p <0, 001, a higher rate of cardiotoxicity was observed in the patient group compared to the control group p <0, 001.

There was a statistical difference between the groups in terms of the presence of cardiotoxicity (no difference was found between low dose and high dose in terms of cardiotoxicity rate).

EF and FS values are higher in the control group than in the other two groups (p 0, 001). EF and FS values of patients with cardiotoxicity were found to be lower (p <0, 001). EF, FS and IVS Em values of patients with cardiotoxicity were found to be lower and MPI values were higher (p <0, 001).

**Conclusions:** EF, FS and IVS Em values showed significant changes when the ANT dose was >200 mg/m<sup>2</sup>. Diastolic blood pressure and troponin levels were statistically different between the groups. While the diastolic blood pressure of the control group was higher than the other two groups (p <0, 001).

**Keywords:** Anthracycline, cardiotoxicity, echocardiography, tumor.

### P-387

#### Miocardial function in newborns with transposition of great arteries, conventional ecocardiography versus speckle-tracking

Daniela Toma<sup>1</sup>, Liliana Gozar<sup>1</sup>, Andreea Cerghit Paler<sup>1</sup>, Dorottya Gabor Miklosi<sup>1</sup>, Carmen Corina Suteu<sup>1</sup>, Marius Cosma<sup>1</sup>, Claudiu Marginean<sup>2</sup>, Mihaela Iancu<sup>3</sup>, Amalia Fagarasan<sup>1</sup>

<sup>1</sup>Department of Pediatrics, "George Emil Palade" University of Medicine, Pharmacy, Science, and Technology of Târgu-Mureş, 540139, Târgu-Mureş, Romania, and Department of Pediatric Cardiology, Emergency Institute of Cardiovascular Diseases and Transplantation, 540139, Târgu-Mureş, Romania; <sup>2</sup>Department of Obstetrics and Gynecology, "George Emil Palade" University of Medicine, Pharmacy, Science, and Technology of Târgu-Mureş, 540139, Târgu-Mureş, Romania;

<sup>3</sup>Department of Medical Informatics and Biostatistics, Faculty of Medicine, "Iuliu Hațieganu" University of Medicine and Pharmacy Cluj-Napoca, Romania

**Background and Aim:** Transposition of great arteries (TGA) is one of the most severe and the fourth most common type of major heart defect, with a prevalence of 3.54/10, 000 live births. Its most common form, D-TGA is defined by ventriculoarterial discordance with atrioventricular concordance, resulting two parallel circulations. To ensure an efficient mixing between the two circulations, the immediate postnatal therapy aims to maintain the permeability of the arterial duct and to ensure an unrestricted atrial septal communication. Until the surgical correction is performed, echocardiographic monitoring with a careful evaluation of the performance of the left ventricle (LV) is required.

Reduction of the indexed LV mass <35 g/m<sup>2</sup> is one of the non-invasive criteria for arterial switch intervention in two stages. In conventional echocardiography, the determination of the ejection fraction (EF) is based on changes concerning the ventricular cavity and is unable to capture myocardial dysfunction.

Our aim was to evaluate the myocardial function using 2D-strain analysis to identify a more sensitive parameter for LV function.

**Method:** In this retrospective, single-center study, 16 newborns with confirmed simple D-TGA were included and subjected to a thorough echocardiographic evaluation. Two-dimensional, apical 4-chamber view images of both ventricles were acquired and analyzed offline with the autostrain function.

**Results:** Descriptive statistical measures showed a median age of 2 days for the included newborns, with a gender ratio of 3:1 with a male preponderance, a gestational age of 39 weeks, 9 newborns being born through C-section. APGAR score at 1 minute, respectively at 5 minutes showed median values of 8 and 9. EF obtained through classic echocardiography had a mean of 69.06, with 3.68 standard deviation (SD). Regarding the classical echocardiographic parameters, the included newborns with simple D-TGA showed good indexed LV mass values with a median value of 70.33g/m<sup>2</sup>, (53.74 and 75.96 ranges). With reference to speckle-tracking parameters LV peak longitudinal strain (LVpGLS) showed a mean value of -18.08 with 4.26 SD.

**Conclusions:** LVpGLS parameter was found to have a strong potential in the evaluation of the myocardial dysfunction of newborns with D-TGA, and may represent an area of interest for future studies.

**Keywords:** Transposition of great arteries, echocardiography, speckle-tracking, myocardial function, congenital heart disease

### P-389

#### Miocardial function in preterm infants using speckle-tracking echocardiography

Andreea Cerghit Paler, Daniela Toma, Liliana Gozar, Dorottya Gabor Miklosi, Marius Cosma, Amalia Fagarasan

Department of Pediatrics, "George Emil Palade" University of Medicine, Pharmacy, Science, and Technology of Târgu-Mureş, 540139, Târgu-Mureş, Romania, and Department of Pediatric Cardiology, Emergency Institute of Cardiovascular Diseases and Transplantation, 540139, Târgu-Mureş, Romania

**Background and Aim:** The cardiopulmonary transition from fetal to neonatal circulation represent the most important adaptation during lifetime. This process is responsible for dramatic changes in cardiovascular physiology and left ventricle (LV) loading conditions, generating progressive modification in the LV geometry and function. Premature newborns are a special category of newborns which undergo more hemodynamic challenges in the post-natal period. Besides the difficult physiological adaptation, the myocardial function can be affected by many factors (medications, immaturity of system, hypoxia, sepsis, e.g.). Nowadays, left ventricular ejection fraction (LVEF) is most commonly used in standard echocardiography for measuring the systolic function. In adults and children it is demonstrated that speckle-tracking assessment of miocardial function overcomes many of the limitations of LVEF, such as reproducibility and early detection of LV dysfunction.

Our aim was to study myocardial function in preterm infants using speckle-tracking echocardiography.

**Method:** This is a prospective single-centre study. Twenty-two preterm infants ≤36 weeks' gestation underwent cardiac ultrasound evaluation. For the assessment of the LV systolic function were used LVEF as classic parameter and LV peak global

longitudinal strain (LVpGLS) as speckle-tracking parameter. The speckle-tracking analysis was made from selected 2D-images of the apical four-chamber clips and then were analyzed offline using a PhilipsQlab 13 software with LV autostrain. The cardiac cycle was validated from the operator. The epicardial border was automatically traced by the software and corrected manually from the operator.

**Results:** The gestational weeks (GW) median was 32.5 [28, 36] with a mean birth weight of 1.79 kg.  $\pm 0.4$ kg. 36.3% neonates were very preterm (28.1–32.0 GW) and 14 low preterm (32.1–34 GW). The gender distribution was equally, 1:1. For LV systolic function, the mean LVEF was 66% with 7.6 standard deviation (SD), and the mean LVpGLS obtain was  $-20.05$ , 3.3 SD. There was no significant relationship of EF and LVpGLS with gestational age, gender and birth weight (Pearson correlation coefficients  $<0.4$ ). Deformation parameter was smaller in premature infants with EF above 65%.

**Conclusions:** Echocardiographic assessment of myocardial deformation parameters is feasible in preterm infants. LVpGLS overcomes many of the limitations of LVEF in this age category, such as reproducibility.

**Keywords:** speckle tracking, myocardial function, premature, pediatric cardiology

#### P-390

##### **Neonatal management and outcomes of congenital tricuspid valve anomalies with anatomic pulmonary atresia: A multi-center experience**

Yalin Lin<sup>1</sup>, Maria Clara Escobar Diaz<sup>2</sup>, Edgar Jaeggi<sup>1</sup>, Wayne Tworetzky<sup>3</sup>, Lindsay Freud<sup>1</sup>

<sup>1</sup>Division of Pediatric Cardiology, Department of Pediatrics, Hospital for Sick Children, Toronto, Canada; <sup>2</sup>Department of Pediatric Cardiology, Sant Joan de Deu, Barcelona, Spain; <sup>3</sup>Department of Pediatric Cardiology, Boston Children's Hospital, Boston, USA

**Background and Aim:** Neonates with congenital tricuspid valve anomalies, including Ebstein anomaly (EA), tricuspid valve dysplasia (TVD), and unguarded tricuspid valve (UGTV), with anatomic pulmonary atresia (PA) are challenging to manage with poor outcomes described from single center experiences. We sought to describe the neonatal management and outcomes from a multi-center cohort of prenatally diagnosed patients.

**Method:** For this retrospective study, we utilized a database of 243 fetuses who were diagnosed with EA/TVD/UGTV from 23 North American centers. We described the fetal and neonatal course and echocardiographic findings of those with anatomic PA. Data are presented as count (%) or median (interquartile range).

**Results:** Of 243 fetuses, 28 were diagnosed with anatomic PA: 12 EA (43%), 9 TVD (32%), 7 UGTV (25%). 4 (14.2%) had genetic diagnoses: Trisomy 21 (n = 2), mosaic Trisomy 21 (n = 1), and unbalanced chromosome 2:3 translocation (n = 1). Only 1 (3.7%) developed hydrops and 3 (12%) had arrhythmias. There were 5 terminations, 4 fetal demises, and 1 lost to follow-up; 2 died within 24 hours of life and 1 (with T21 and congenital chylothorax) had comfort care.

Of the 15 remaining neonates, the median birth weight and gestational age were 2.7 kg (IQR 2.5–2.8) and 38 (IQR 37–38.6) weeks. Median tricuspid regurgitant jet velocity was 2.45 m/s (IQR: 2.15–2.6), 40% had mild RV dysfunction, and 6.7% had mild LV dysfunction. The median main pulmonary artery z-score was  $-2.6$  (IQR:  $-3.2$  to  $-1.5$ ). 14 (93.3%) had surgical intervention: 5 had attempted biventricular repair and 3 (60.0%) died prior to hospital discharge, 5 had aortopulmonary shunt placement and

1 (20.0%) died, and 4 underwent right ventricular exclusion with no deaths. 1 patient had pulmonary valve dilation in the catheterization lab and survived to discharge. Overall, 4 of 15 patients died (26.7%) at a median age of 21 days.

**Conclusions:** EA/TVD/UGTV with anatomic PA is a rare condition across multiple centers with high perinatal and post-interventional mortality, similar to those without anatomic PA. Patients who underwent palliative approaches, particularly right ventricular exclusion, had lower mortality than those who underwent biventricular repair.

**Keywords:** tricuspid valve anomalies, pulmonary atresia

#### P-391

##### **Coarctation of the aorta and trisomy 21; a single centre review.**

Ross Foley, Frank Casey

Department of Cardiology, RBHSC, Belfast, Northern Ireland, United Kingdom

**Background and Aim:** In our tertiary centre, all neonates with a diagnosis of Trisomy 21 (T21) undergo an echocardiogram (echo) within the first two weeks of life, given the estimated 45% risk of congenital heart disease. Not infrequently the echo raises concern about the calibre of the aortic arch and the risk of evolving coarctation of the aorta (coarctation). This often results in admission and serial echos until a definitive diagnosis can be made. Data regarding association between T21 and coarctation is variable. We therefore sought to analyse the prevalence of coarctation of the aorta within our T21 population.

**Method:** We analysed all our patients who received a diagnosis of T21 over a 20 year period from 2002 to 2022. We then examined how many of these received a diagnosis of coarctation, if they required an intervention on the aortic arch and whether this was a catheter or surgical intervention. We also analysed whether these patients had other cardiac diagnoses requiring catheter or surgical intervention.

**Results:** From 2002–2022, 11 or 814 (1.4%) patients with T21 also received a diagnosis of coarctation. 9 of these patients required intervention on the aortic arch, all of which were surgical repair. No patients received catheter intervention. 9 patients also had other cardiac diagnoses. 8 patients had an aortic arch repair in combination with another cardiac surgical procedure. Only one of the 11 patients underwent isolated surgical coarctation repair.

**Conclusions:** Our data confirms previous findings that the association between T21 and coarctation of the aorta requiring intervention is rare and is usually found in the presence of other cardiac anomalies. It is not uncommon for echocardiography to indicate a mild degree of hypoplasia of the aortic arch in T21, but this large series confirms that clinically significant coarctation of aorta is rare in this patient group.

This should give some reassurance to physicians when evaluating the aortic arch in patients with T21.

**Keywords:** Trisomy21, Coarctation, Aorta

#### P-392

##### **Novel insights into aspirin non-responsiveness in a national congenital heart disease population with effective use of laboratory assessment**

Irene E Regan<sup>1</sup>, Sean T Kelleher<sup>2</sup>, Jennie Ryan<sup>3</sup>, Terence Prendiville<sup>3</sup>, Orla Franklin<sup>3</sup>, Adam James<sup>3</sup>, Damien Kenny<sup>3</sup>, Dermot Cox<sup>4</sup>, Colin J McMahon<sup>2</sup>

<sup>1</sup>Coagulation/Haematology Department, CHI at Crumlin, Dublin, Ireland; <sup>2</sup>School of Medicine, UCD, Dublin, Ireland; <sup>3</sup>Cardiology

Department, CHI at Crumlin, Dublin, Ireland; School of Medicine, UCD, Dublin, Ireland; <sup>3</sup>Cardiology Department, CHI at Crumlin, Dublin, Ireland; <sup>4</sup>School of Pharmacology, RCSI, Dublin, Ireland

**Background and Aim:** The incidence of aspirin non-responsiveness in children with CHD has been reported to range from 2.3 up to 80% through different laboratory assessments, cut-off values and patient cohorts (long-term aspirin versus recent exposure and surgery). The aim of this prospective observational study was to define aspirin non-responsiveness in 2 paediatric cardiology cohorts; cohort 1 (C1) outpatients on long-term aspirin (minimum of 3 months) and cohort 2 (C2) in-patients who are post-surgery having recently commenced aspirin (minimum 5 days). Patients were on a standard therapeutic dose (1–5mg/kg/day) not to exceed 75 mg/day.

**Method:** This study was performed at the National Cardiac Centre, Children's Health Ireland at Crumlin, Dublin, Ireland. Inclusion criteria; cardiology patients on aspirin therapy. Exclusion criteria; known coagulation disorder or syndrome interfering with platelet function/number, haemoglobin <100g/l, platelet count <150 x10<sup>9</sup>/l. Laboratory criteria for aspirin non-response was defined as platelet inhibition below 50% by thromboelastography (TEGPM) and Light Transmission platelet aggregation (LTA-AA) (in patients ≥ 2 years of age) above 20% using arachidonic acid (AA). TEGPM data was confirmed using surrogate AA. Cut-off ≥40mm for TEGPM MA AA was also applied. Patients had a FBC, coagulation screen, CRP and recent echocardiogram. Variables evaluated included platelet inhibition, age, dose/kg, and type of surgical procedure.

**Results:** In total, 137 patients were recruited (101 in C1, 36 in C2). C1 median age was 9.3 years (0.1–18.2 years) with 60 boys, 41 girls. There was good agreement between assays. Two patients had discrepant results and are due repeat follow-up. Mean aspirin dose 2.41 ± 1.27 mg/kg/day. Five percent (5/101) of C1 patients demonstrated poor aspirin response. Three of the five patients had a bio-prosthetic valve.

C2 median age was 0.5 years (0.04–13.2 years) with 13 boys and 23 girls. The mean aspirin dose was 3.14 ± 1.46 mg/kg/day. Twenty-five percent of (9/36) patients demonstrated poor aspirin response and raised CRP (18 ± 12mg/l p<0.05).

**Conclusions:** Twenty-five percent of the in-patient cohort demonstrated poor aspirin response compared to 5% of the outpatient population. Given this high rate in this at-risk group, robust testing such as this should be considered especially following events such as surgery and sepsis.

**Keywords:** Aspirin non-responsiveness, CHD, cardiac surgery, general cardiology, platelet inhibition, platelet aggregation

### P-393

#### The nest study: nicom vs echo in the screening of transient hypertrophic obstructive cardiomyopathy (HOCUM)

Bryan Padraig Finn<sup>1</sup>, Neidin Bussmann<sup>1</sup>, Daragh Finn<sup>2</sup>, Eugene Dempsey<sup>1</sup>

<sup>1</sup>Department of Neonatology, Cork University Maternity Hospital, Wilton, Cork, Ireland; <sup>2</sup>Department of General Paediatrics, Cork University Hospital, Cork, Ireland

**Background and Aim:** Gestational Diabetes Mellitus (GDM) is the most common metabolic disorder of pregnancy. An increasing number of infants are exposed to hyperglycaemia antenatally, with the national prevalence estimated at 10.1–12.4%. Transient HOCUM and interatrial/interventricular communications remain the most common cardiac sequelae for such infants. In

particular, transient HOCUM has an estimated global incidence of 13–44%.

We aimed to determine if NICOM (non-invasive cardiac output monitoring) technology could be used as an alternative screening tool to echocardiography in detecting transient hypertrophic cardiomyopathy (HOCUM) in infants born to mothers with GDM.

**Method:** A single-centre prospective observational cohort study was conducted. All infants were recruited in the postnatal period prior to the routine newborn baby check assessment. Fifty infants underwent echocardiography, while 29/50 were paired with a NICOM assessment. A sample size of 29 patients was required to detect a significant correlation (Cohen's r = 0.5) between both variables, with a power of 80%.

**Results:** 4/46 patients (8.7%) were noted to have a z-score of >2 for both the interventricular septal wall thickness and left posterior ventricular wall in diastole, consistent with a diagnosis of transient HOCUM. 26 out of 46 infants born to mothers with diabetes had significant thickening (Z-score >2) of either the interventricular septum or the left ventricular posterior wall. The incidence of transient HOCUM was not associated with the degree of maternal glycaemic control, maternal BMI or the method of diabetes management (all P > 0.05). NICOM was not significantly correlated with ECHO LVO values (r(25) = 0.2, p = -0.31) or Bland Altman analysis.

**Conclusions:** Our cohort's incidence of transient HOCUM was lower than in previous studies, 8.7% versus 13–44%. NICOM is not an effective screening tool for transient HOCUM based on a poor Pearson correlation and the broad limits of agreement found with the Bland Altman plot. Infants born to mothers with GDM would not benefit from routine echocardiography screening beyond the current standard of care. This is because all cases of clinically relevant congenital heart disease would have been detected with the current standard of care – newborn bedside examination followed by targeted ECHO as clinically indicated.

**Keywords:** Neonatal, Cardiomyopathy, Echocardiogram, NICOM, GestationalDiabetes

### P-395

#### Current clinical profile of acute rheumatic fever and rheumatic recurrence in pakistan

Noor Masood Sadiq, Gul Afshan, Ahmad Usaid Qureshi, Uzma Kazmi, Najam Hyder, Masood Sadiq

Department of Paediatric Cardiology, The Children's Hospital and University of Child Health Sciences, Lahore, Pakistan

**Background and Aim:** BACKGROUND: Rheumatic Fever (RF) and Rheumatic heart disease (RHD) is still an important cause of morbidity and mortality in low and middle-income countries (LMICs) like Pakistan. The clinical profile of acute rheumatic fever however, seems to have changed. Inclusion of echocardiography and arthralgia as major criteria is likely to enhance the diagnostic ability of the physicians.

**AIM:** To review the clinical profile of patients presenting with acute rheumatic fever and rheumatic recurrence referred to a single tertiary care children's hospital.

**Method:** One hundred and thirty consecutive patients with rheumatic activity (acute rheumatic fever or rheumatic recurrence) were included in the study from August 2019 to March 2022. The diagnosis was based on modified Jones criteria 2015. The socio-demographic variables, family and clinical details were recorded. All patients underwent 12 lead ECG, chest x-ray, and a detailed echocardiography. Laboratory tests included complete

blood count, ESR, CRP, and ASOT. Treatment during admission, duration of stay and outcome were also recorded.

**Results:** Mean age was 13.5 + 0.3yrs (range 6–26yrs). Male to female ratio was 1.6:1. Majority were of low socioeconomic status, from a rural background and more than 5 family members in a house. Out of total 130 patients, 27 (20.8%) were diagnosed with acute rheumatic fever while 103 (79.2%) were cases with rheumatic heart disease and now presenting with recurrence. Carditis was the most common presenting feature (n = 122, 93.8%), followed by arthralgia (n = 53, 40.8%), arthritis (n = 42, 32.3%), subcutaneous nodules (n = 10, 7.7) and chorea (n = 5, 3.8%). Carditis was more common in recurrence (n = 22, 81.5%) than first episode of ARF (n = 100, 97.1%), p = 0.01. Congestive cardiac failure was the most common complication (n = 24, 18.5%) followed by arrhythmias (n = 4, 3.1%). Twenty four patients (18.5%) presented in heart failure out of which 23 patients (22.3%) had RHD with recurrence and only 1 patient (3.7%) with acute rheumatic fever (p = 0.001).

**Conclusions:** Rheumatic recurrence with predominantly carditis is now a more common presentation of acute rheumatic fever. This often is the cause of deterioration of symptoms in otherwise stable or undiagnosed patients with RHD.

**Keywords:** Rheumatic fever, Rheumatic heart disease, clinical profile, Low middle income countries

#### P-396

##### **A 36-week new-born baby with calcified thrombus in ductus aneurism extending to LPA**

Nadezhda Georgieva Dzhelepova<sup>1</sup>, Krasimir Atanasov<sup>1</sup>, Amit Maniyar<sup>1</sup>, Liz Shakley<sup>2</sup>

<sup>1</sup>Manchester Children's Hospital; <sup>2</sup>Stepping Hill Hospital

**Background and Aim:** We present a case of a 36-week-old baby who was diagnosed with LPA thrombus and Ductal thrombus on 5th day of life. The baby was born from first uneventful IVF pregnancy. There was no maternal diabetes, no infection, no clotting disorder.

**Method:** Noted restricted growth and delivery was induced. Delivered in good condition at 36 weeks via emergency CS for foetal bradycardia with birth weight of 2.4 kg. At 12 h of age had low blood sugar and was screened for infection. Chest XR raised concerns about boot shaped heart and local echo found suspicion of LPA thrombus. The baby was started on enoxaparin and transferred to our centre. Further imaging -echocardiogram, CT and MRI demonstrated calcified thrombus in ductal aneurism extending to LPA. The left pulmonary artery was compressed but flow void could be seen.

**Results:** After starting enoxaparin, the flow in LPA seemed to improve. The case was discussed at JCC, and the decision was to continue conservative treatment with enoxaparin as the baby was haemodynamically stable. The patient was discharged home on enoxaparin treatment for 6 months.

**Investigations:** Coagulation screen:

Anti-thrombin activity 38.3 (normal range 39–93)

Free protein S and protein C - normal

Lupus anti-coagulant screen - insufficient

Factor 5 Leiden and prothrombin - pending

Lipoprotein A - pending

Factor 10A - 0.71

ECG, CT MRI – confirming thrombus in Ductus arteriosus aneurism extending to LPA. Arthropathy and connective tissue genes panel pending.

**Conclusions:** Neonatal ductus arteriosus aneurism is a rare entity. It can be incidental finding, but it can also lead to complications like thrombosis, infection, aneurism rupture, infection, or compression of nearby structures (1). The possible risk factors for neonatal ductus arteriosus aneurism and clotting are genetic syndromes, elastin tissue disorders, clotting disorders (1). Our patient has a large ductus arteriosus aneurism with a calcified thrombus in it and the decision was for conservative anticoagulation treatment.

**Keywords:** DUCTUS ARTERIOSUS ANEURISM, THROMBUS

#### Ductus arteriosus thrombus



A calcified thrombus in PDA

#### P-397

##### **Scimitar syndrome in children: an uncommon cause of recurrent bronchopneumonia**

Houda Ajmi<sup>1</sup>, Rihab Jabnoun<sup>1</sup>, Salsabil Nouir<sup>1</sup>, Fadoua Majdoub<sup>1</sup>, Sameh Mabrouk<sup>1</sup>, Ayoub Fatnassi<sup>1</sup>, Safa Hannachi<sup>1</sup>, Ibtissem Hasni<sup>2</sup>, Saoussen Abroug<sup>1</sup>, Jalel Chemli<sup>1</sup>

<sup>1</sup>Department of Pediatrics, Sahloul Teaching Hospital, 4054, Sousse, Tunisia; <sup>2</sup>Department of Radiology, Sahloul Teaching Hospital, 4054, Sousse, Tunisia

**Background and Aim:** The Scimitar syndrome is a rare congenital Heart defect. It represents a combination of pulmonary hypoplasia and partial anomalous pulmonary venous return which can be noted on X-ray and documented angiographically. This malformation occurs in 1 to 3 per 100.000 live births. The aim of this study was to describe the clinical presentation, echocardiographic, and angiographic findings of Scimitar syndrome in infancy.

**Method:** Here we report two cases of infantile Scimitar syndrome who were hospitalized in the pediatric department of Sahloul hospital.

**Results:** The first case is about a 2-year-old boy who was admitted several times for dyspnea. The chest X-ray revealed cardiomegaly and right post-basal pulmonary opacity. Echocardiography showed dilation of the right cavities with no pulmonary arterial hypertension. Chest computed tomography showed right basal lung sequestration and partial anomalous pulmonary venous return in the inferior vena cava. The heart correction surgery of this malformation has been delayed until the pulmonary condition of the child became better. The second case is about a 2-year-old female infant who was referred to our department for recurrent respiratory infections. The physical examination revealed a well youth in no apparent distress. Right pulmonary veins were not visible

on echocardiography and pulmonary arterial pressure was estimated as in normal level. Cardiac catheterization confirmed the diagnosis of Scimitar syndrome with pulmonary sequestration. The patient underwent surgery and was discharged in good condition on medical treatment

**Conclusions:** Scimitar syndrome is a little-known disease with insidious and non-specific clinical expression. No treatment is required in asymptomatic patients, however surgical treatment should be proposed in the case of severe right left shunt, sequestration or recurrent lung infections.

**Keywords:** Scimitar syndrome, bronchopneumonia

#### P-398

##### **Congenital heart disease spectrum in williams and beuren syndrome: report of three cases**

Houda Ajmi<sup>1</sup>, Hana Daoud<sup>1</sup>, Sameh Mabrouk<sup>1</sup>, Abir Daya<sup>1</sup>, Fadoua Majdoub<sup>1</sup>, Salsabil Nouir<sup>1</sup>, Sarah Dimassi<sup>2</sup>, Saoussen Abroug<sup>1</sup>, Jalel Chemli<sup>1</sup>

<sup>1</sup>Department of Pediatrics, Sahloul Teaching Hospital, Sousse, Tunisia;

<sup>2</sup>Department of Cytogenetics and Reproductive Biology, Farhat Hached University Hospital, Sousse, Tunisia

**Background and Aim:** Williams and Beuren syndrome (WBS) is a congenital, multisystem disorder involving the cardiovascular, connective tissue, and central nervous systems. It is a relatively rare microdeletion. Because of elastin deficiency, cardiovascular abnormalities are the leading cause of morbidity and mortality in patients with WBS.

**Purpose:** The present study aimed to review the clinical and echocardiographic particularities of congenital heart disease in children with WBS.

**Method:** Review of charts of all patients with WBS who were hospitalized in the pediatric department of Sahloul hospital during the period between 2000 and 2022 revealed three cases with WBS and affected by congenital heart disease.

**Results:** Three cases of patients with WBS were included in our study: two boys and one girl. The average age in the diagnosis was 3, 4 months-old [5 days -7 months]. All the patients presented dysmorphic features of WBS: elfin face, hypoplastic nails, short stature, and microcephaly. Clinical findings-signs showed severe hypotrophy in 2 cases, dyspnea in 2 cases and heart murmur in the 3 cases. Congenital heart defect was diagnosed in all cases: the first one had severe supra-aortic valve stenosis and severe sub-valvular pulmonary stenosis, the second had severe multiple severe supra-valvular and valvular pulmonary stenosis associated with severe supra-aortic valve stenosis and atrial septal defect, the third had mild supra-aortic valve stenosis. Their genetic analysis revealed microdeletion on chromosome 7q11.23.1. Only one case had been operated on for his heart defect (case 1): sub pulmonary myomectomy and enlargement aortoplasty with one pericardial patch repair. The outcome of this child was good over 10 years of follow up. He remains asymptomatic, with good bi-ventricular performance and no aortic valve regurgitation on echocardiography. The second case presented a severe dyspnea and died. The third one was still asymptomatic and has been followed regularly by echocardiography.

**Conclusions:** WBS is a complex, multisystem progressive disorder with significant cardiovascular manifestations. Arterial stenoses make up the large majority of cardiovascular issues in patients with WBS. Advances in surgical techniques and medical therapeutic options hold promise for significant improvements in the cardiovascular outcomes of these patients.

**Keywords:** Williams and Beuren syndrome, aortic stenosis, pulmonary stenosis

#### P-399

##### **Between the hammer and the anvil: gigantic coronary aneurisms in the context of EBV and SARS-COV-2 co-infection, in a patient with coronary anomalies**

Simona Sorana Căinap<sup>1</sup>, Diana Jecan Toader<sup>1</sup>, Diana Borcau<sup>2</sup>, Simona Oprea<sup>3</sup>, Angela Eni<sup>3</sup>, Bianca Simionescu<sup>1</sup>

<sup>1</sup>Emergency Clinical Hospital for Children Cluj-Napoca Romania & University of Medicine and Pharmacy "Iuliu Hatieganu" Cluj-Napoca Romania; <sup>2</sup>Emergency Clinical Hospital for Children Cluj-Napoca Romania <sup>3</sup>Nicolae Stăncioiu Heart Institute Cluj-Napoca Romania

**Background and Aim:** Coronary artery dilatation has been observed with coronavirus disease 2019 (COVID-19)-associated multisystem inflammatory syndrome in children (MIS-C), which is more common in those with Kawasaki-like disease.

**Method:** A seven-year-old boy was referred to our centre for the investigation of prolonged fever.

**Results:** The current symptomatology first developed 1 month before referral to our service, when he presented high fever, unilateral latero-cervical adenopathy, and hepatosplenomegaly.

Inflammatory markers were 6 times the normal values, but the EBV serology was negative at that moment. The patient improved under symptomatic treatment, but after a month the high fever relapsed, associated with asthenia and myalgia. In our service, the physical exam was notable for bilateral non-purulent conjunctivitis, bilateral submandibular adenopathy, red strawberry tongue, grade 1 systolic murmur (AV = 117 bpm, BP = 100/65 mmHg), but no organomegalies. The laboratory investigations revealed high inflammatory parameters (ESR, CRP, but negative procalcitonin), IgG SARS-COV2 antibodies (750 BAU/ml, NV <33.8 BAU/ml) and positive IgM EBV antibodies, but normal values for cardiac markers, ferritin, and D-Dimers. The echocardiography revealed the origin of the right coronary artery from the left coronary sinus. At distance from the origin, a fusiform aneurism of 6/10 mm (Z-Score = 9.99) was noted, with no suggestive images for thrombosis. The left coronary artery had 3 mm at the origin (Z score = 0.72), 4 mm at bifurcation (Z score = 3.04), without patent aneurysmal images. Cardiac AngioCT confirms the abnormal origin of the right coronary artery, that emerges from the left coronary sinus with a path at a sharp angle post-origin, partially intraparietal. It is noted the presence of aneurysmal dilations at the level of the right coronary artery (segment I and II) and the ectasia of the left coronary trunk. The treatment consisted of IVIG (2g/kg), corticotherapy, Aspirin and Enoxaparin, under which the fever remitted and the inflammatory parameters normalised. The serial echocardiographic evaluations revealed the persistence of coronary aneurysms at the same dimensions even at the 6 months follow-up.

**Conclusions:** Concerning the long term implications, our patient presents an increased risk of acute cardiac events, due to the two associated pathologies: the persistence of giant aneurysms and the presence of coronary origin anomaly.

**Keywords:** Coronary aneurysm, coronary anomaly, SARS-COV2, EBV

#### P-400

##### **Experimental model of cardiopulmonary bypass and cardiac arrest in neonatal piglets**

Madeleine Ball<sup>1</sup>, Sergio Benito<sup>2</sup>, Marta Camprubí Camprubí<sup>3</sup>, Cristina Ruiz Herguido<sup>4</sup>, Joan Sanchez De Toledo<sup>5</sup>

<sup>1</sup>Institut de Recerca Sant Joan de Déu, Barcelona, Spain; <sup>2</sup>Vanderbilt University School of Medicine, Nashville TN, U.S.A.; <sup>3</sup>Institut de Recerca Sant Joan de Déu, Barcelona, Spain; <sup>4</sup>Department of Pediatric Critical Care Medicine, Hospital Sant Joan de Déu, Barcelona, Spain;

<sup>3</sup>Institut de Recerca Sant Joan de Déu, Barcelona, Spain; BCNatal—Barcelona Center for Maternal Fetal and Neonatal Medicine, Hospital Sant Joan de Déu, Barcelona, Spain; <sup>4</sup>Institut de Recerca Sant Joan de Déu, Barcelona, Spain; <sup>5</sup>Institut de Recerca Sant Joan de Déu, Barcelona, Spain; Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain; Department of Critical Care Medicine, University of Pittsburgh, Pittsburgh PA, U.S.A.

**Background and Aim:** Comprehensive investigation of congenital heart disease requires appropriate and validated animal models. Piglets are commonly used for translational research due to similar physiological traits. The aim of this study was to describe and validate a neonatal piglet model of cardiopulmonary bypass (CPB) with circulatory and cardiac arrest (CA) as a tool for studying severe brain damage and other complications of pediatric cardiothoracic surgery (CTS).

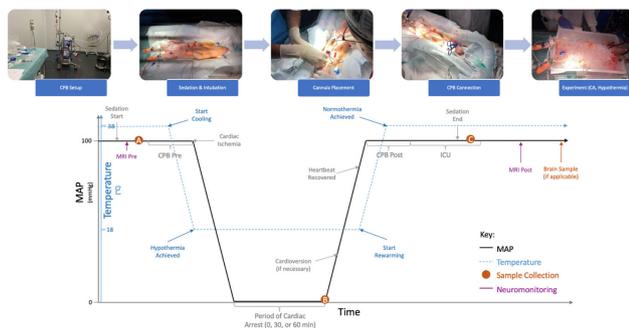
**Method: CPB Circuit Preparation:** The CPB circuit was primed with saline solution and fresh donor adult pig blood, followed by heparin, sodium bicarbonate, and calcium gluconate.

**Animal Preparation:** Species *Sus scrofa domestica* was used. Orotracheal intubation was performed and mechanical ventilation was started. Sedation and paralysis were maintained with sevoflurane.

Bilateral neck vessels were exposed. A 6 Fr arterial and an 8 Fr venous cannula were placed in the left carotid artery and the right external jugular, respectively. The cannulas were connected to the CPB circuit. Initial flow rate was set at 80–85 ml/kg/min and increased to 150 ml/kg/min. CPB was maintained for as long as the experiment required. To initiate CA, potassium chloride was administered and arrest was confirmed with echocardiography. Following CA, CPB flow was reduced to zero to initiate circulatory arrest. After experimental condition was met (30–60 min, hypothermia, etc.), resuscitation was started using calcium gluconate and bicarbonate, with dose increases and cardioversion as necessary.

**Results:** Eight consecutive experiments were performed and analyzed. Piglets were 4.6 days old [4–6] and weighed 2746g [2400–2900]. 25% were female. Time to from sedation to cannulation was 3.3 hours [2.75–3.56]. Vessel cannulation and CPB was successfully achieved in 88% of experiments. The single failure occurred during cannulation, likely secondary to small piglet size. MRI demonstrated early brain damage. The main steps of the protocol are outlined in Figure 1.

**Conclusions:** We describe a step-wise strategy to institute CPB with CA in neonatal piglets to further study neonatal CTS, including intra-operative brain damage. This method uses materials readily



**Figure 1** Schema demonstrating key steps in the model of cardiopulmonary bypass with cardiac arrest and experimental conditions. CPB = cardiopulmonary bypass, MAP = mean arterial pressure, ICU = intensive care unit

**Figure 1** Schema demonstrating key steps in the model of cardiopulmonary bypass with cardiac arrest and experimental conditions. CPB = cardiopulmonary bypass, MAP = mean arterial pressure, ICU = intensive care unit.

available in most hospital settings, is reliable and reproducible, and may be widely employed by those using animal models to enhance translational research in children undergoing heart surgery.

**Keywords:** translational cardiology research, cardiopulmonary bypass, animal model, pediatric cardiothoracic surgery

#### P-401

##### Pericardial effusion in children after pediatric cardiac surgery: A retrospective analysis of the current burden of disease.

Noelia Gisele Pastene<sup>1</sup>, Maria Lucia Rossi<sup>1</sup>, Madeleine Ball<sup>2</sup>, Patricia García Canadilla<sup>2</sup>, Roger Domingo Espinós<sup>3</sup>, Joan Sánchez De Toledo<sup>4</sup>, Carin Cristina Walter<sup>5</sup>

<sup>1</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Esplugues de Llobregat, Spain.; <sup>2</sup>Cardiovascular Diseases and Child Development, Institut de Recerca Sant Joan de Déu, Esplugues de Llobregat, Spain. BCNatal—Barcelona Center for Maternal-Fetal and Neonatal Medicine, Hospital Sant Joan de Déu and Hospital Clínic, University of Barcelona, Barcelona, Spain.; <sup>3</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Esplugues de Llobregat, Spain. Additum Blockchain, SL. Madrid, Spain.; <sup>4</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Esplugues de Llobregat, Spain. Cardiovascular Diseases and Child Development, Institut de Recerca Sant Joan de Déu, Esplugues de Llobregat, Spain. Department of Critical Care Medicine, University of Pittsburgh, Pittsburgh, United States.;

<sup>5</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Esplugues de Llobregat, Spain. Cardiovascular Diseases and Child Development, Institut de Recerca Sant Joan de Déu, Esplugues de Llobregat, Spain

**Background and Aim:** Post-operative pericardial effusion(PPE) are a frequently reported complication after pediatric heart surgery. This study aims to describe the incidence of PPE in our institution and to assess the overall burden of disease.

**Method:** This retrospective study included 308 children who required cardiac surgery from January/2021 to September/2022. Surgical complexity was stratified according to the STAT score and divided into Low(STAT 1, 2) and High(STAT 3, 4, 5)complexity. PPE was classified as mild(<10mm), moderate(10–20 mm) and severe(>20mm). Hospital(H) and ICU-Length of stay(LOS), need for ICU and/or Hospital readmission and the need for acute interventions were analyzed. Results are presented as mean(Interquartile range), frequency or percentages.

**Results:** During the study period, 38/308(12%) children (age 2 years [0–7]) developed PPE. Twenty(52%) were mild, 12(31%) moderate and, 6(15%) severe. The incidence based on surgical complexity was: Low 30/225(26%), High 7/83(8%). Seven(19%) were diagnosed in the ICU and 31(81%) after ICU-discharge. Among ICU PPEs, 3/7(40%) developed in patients with mediastinal tube in place and resolved with standard maneuvers; 2/7(28%) required pericardiocentesis and 2/7(28%) were medically managed. Among non-ICU PPEs, 2/38(5%) required ICU-readmission, and only one(3%) required pericardiocentesis.

Children who developed PPE had longer extracorporeal circulation times[75(53–108) vs 127(95–140) minutes,  $p = 0.006$ ]. Overall, those who developed PPE in the ICU had longer ICU-LOS[9(6–21) vs 3(2–7) days,  $p = 0.008$ ] and hospital-LOS[20(16–28) vs 7(6–12) days,  $p < 0.001$ ] than children without PPE.

ICU-LOS was longer(days) in the severe [27(21–55)] than in moderate [2(2–4)] or mild PPE [4(2–6)],  $p = 0.02$ ; Non-ICU-LOS was

also longer in the severe [40(28-71)] than in moderate [8(5-9)] or mild [8(6-11)], ( $p = 0.03$ ).

Children with STAT 2 had more number of follow-up visits in those with PPE [4(3-5) vs 6(4-7),  $p = 0.05$ ]. In other STAT groups there were no differences.

**Conclusions:** In our cohort, 12% of surgical cases developed PPE and interestingly enough, 30/38 (79%) were patients undergoing Low Complexity score surgeries. In our cohort, PPE is associated with prolonged LOS and higher number of follow-ups post-discharge. Patient diagnosed with PPE during ICU admission, and those with larger effusions, had longer ICU and HLOS. Future studies should focus in identification of early predictors of PPE to elaborate personalized clinical pathways to avoid the burden of the PPE in children undergoing heart surgery.

**Keywords:** pericardial effusion, critical care, congenital heart defects

#### P-402

##### Cardiovascular risk stratification in children undergoing chemotherapy

Esther Aurensanz Clemente<sup>1</sup>, Patricia Garcia Canadilla<sup>2</sup>, Natalia Gimenez De León<sup>3</sup>, Joan Sanchez De Toledo<sup>4</sup>

<sup>1</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona. Spain. Cardiovascular Diseases and Child Development, Institut de Recerca Sant Joan de Déu, Esplugues de Llobregat, Spain;

<sup>2</sup>Cardiovascular Diseases and Child Development, Institut de Recerca Sant Joan de Déu, Esplugues de Llobregat, Spain 3BCNatal—Barcelona Center for Maternal-Fetal and Neonatal Medicine, Hospital Sant Joan de Déu and Hospital Clínic, University of Barcelona, Barcelona, Spain;

<sup>3</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona. Spain.; <sup>4</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona. Spain. Cardiovascular Diseases and Child Development, Institut de Recerca Sant Joan de Déu, Esplugues de Llobregat, Spain Department of Critical Care Medicine, University of Pittsburgh

**Background and Aim:** Advances in oncological therapies have significantly improved patients' outcomes; however associated toxicities remain a matter of concern. The aim of this study is to analyze the incidence of cardiotoxicity in a pediatric cancer population.

**Method:** Prospective study including pediatric patients undergoing chemotherapy between September 2020 and March 2022. Patients were divided into four groups according to the treatment phase they were allocated at the time of evaluation: Baseline; Early (3 months after treatment); Late ((6 months after treatment) 4) Final (end of treatment) and, 5) Relapse (before starting a new treatment). The comprehensive cardiovascular evaluation included anthropometric measurements, electrocardiogram (EKG) and functional echocardiography. Patients were stratified into four different cardiotoxicity groups according to clinical practice guidelines.

**Results:** 265 patients were included. Mean age was  $7.9 \pm 5.2$  years, 58.9% were males, 62.3% had solid tumor, 37.8% hematologic. The incidence of overweight was 4.5% and arterial hypertension 6.41%. While the global incidence of ventricular dysfunction was only 2.2%, a decreased of  $> 10\%$  of the baseline left ventricular ejection fraction (LVEF) was observed in 24.9% of the patients during follow-up. Late and relapse evaluations showed a higher decrease of LVEF (29.58% and 28.57% respectively). A significant elevation of Troponin was observed at both Early and Late phases compared to baseline ( $0.012 \pm 0.014$  and  $0.006 \pm 0.008$ ,  $p < 0.001$ ). A significant change in the global longitudinal strain was documented in all study groups. EKG abnormalities were documented in 16.3% of the patients. Prolongation of QT interval and changes

in repolarization were predominantly seen at the final and relapse evaluation (18.4% and 22.6%,  $p = 0.007$ ) (13.2% and 18.9%,  $p = 0.007$ ). About the stratification according to the guidelines: the percentage of patients with subclinical damage is significantly higher in groups Early and Late (21.1% and 20.8%, respectively) compared to the other groups; while the amount of patients classified as healthy high-risk is significantly higher in groups Late and Final (13.2% and 22.2%,  $p = 0.009$ ).

**Conclusions:** This study show that pediatric patients undergoing chemotherapy have changes in the EKG during their treatment. These results highlight the importance of the EKG evaluation during cardiovascular follow-up of pediatric cancer patients.

**Keywords:** Cardiotoxicity, electrocardiogram, functional echocardiogram, cancer.

#### P-403

##### Coronary artery dilation in febrile infants: incomplete kawasaki disease, multisystem inflammatory syndrome or febrile exanthematous illness?

Salva Addilami, Laura Prisacariu

Paediatric department, National Institute for Mother and Child Health "Alessandrescu Rusescu", Bucharest, Romania

**Background and Aim:** Kawasaki disease is the most common cause of vasculitis in pediatric population. It primarily affects children between the age of 6 month and 5 years. Atypical or incomplete Kawasaki disease is more commonly diagnosed in infants under 6 months of age. Since the COVID 19 pandemic there were reported outbreaks of Kawasaki-like multisystem inflammatory syndrome in pediatric population.

The most important complications and sequelae of Kawasaki disease are coronary abnormalities that are also the leading cause of acquired heart disease in children. The American Heart Association and American Academy of Pediatrics guidelines include an algorithm for evaluation and treatment of suspected patients with incomplete or atypical Kawasaki disease. Although 40 % of children with Kawasaki disease have a concurrent infection, the incomplete Kawasaki disease treatment algorithm includes children with fever for at least 5 days without an alternative explanation making the diagnosis and the decision of treatment dependent on clinical judgment.

Our aim is to emphasize the importance of clinical judgement when evaluating a febrile infant.

**Method:** We present a series of three case reports of young infants who presented in our clinic with febrile disease in a period of 6 months during the covid pandemic.

**Results:** The course of diagnosis, treatment and evolution was different but all three of them had medium and large coronary artery aneurysms. The first patient received intravenous immunoglobulin and corticosteroids as additional therapy being a high risk patient but developed severe coronary involvement and needed a second line treatment, in this case an interleukin 1 inhibitor was administered. The second patient received a second dose of intravenous immunoglobulin as salvage therapy. The third patient was referred to the pediatric cardiologist in the 14th day of the disease and at more than 72 hours of normal body temperature so immunosuppressive or anti-inflammatory treatment was no longer recommended.

**Conclusions:** This case report series underlines the correlation of severe cardiac involvement with delayed diagnosis and incomplete forms of disease but also the importance of clinical judgement in the diagnosis of Kawasaki disease.

**Keywords:** kawasaki disease, infant, coronary aneurysms

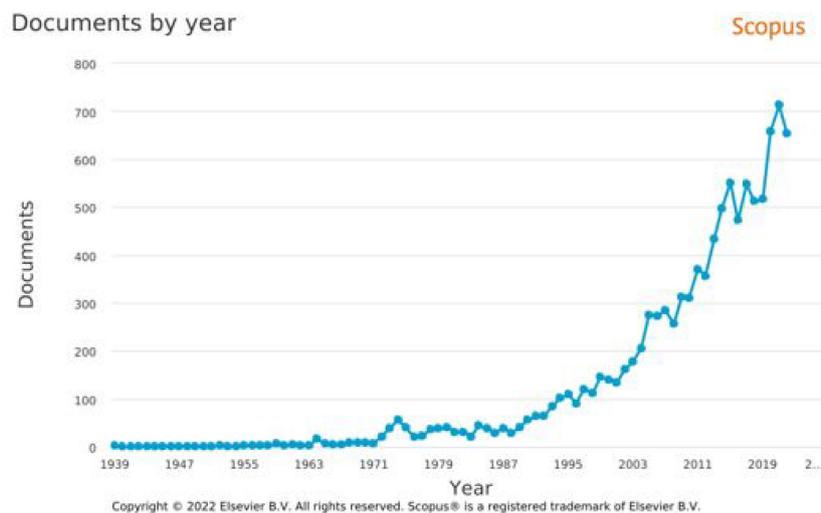
**P-404**  
**How did the pediatric cardiology research community react to COVID-19?**

Jernej Završnik<sup>1</sup>, Ivan Malčič<sup>2</sup>, Helena Blažun Vošner<sup>1</sup>, Peter Kokol<sup>3</sup>  
<sup>1</sup>Community Healthcare Center dr. Adolf Drolc Maribor; <sup>2</sup>Medical Faculty, University of Zagreb; <sup>3</sup>Faculty of Electrical Engineering and Computer Science, University of Maribor

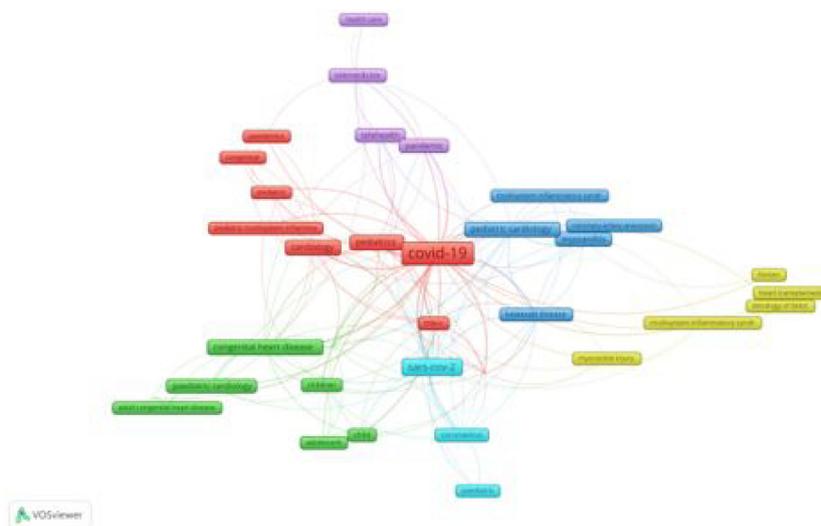
**Background and Aim:** The coronavirus disease (COVID-19) has significant cardiovascular implications. Given the increased risk for in adults, there is a raised concern that patients with pediatric and congenital heart disease (CHD) may likewise be at increased risk for severe infection, however, limited data exist regarding pediatric patients.

**Method:** To fill this gap and analyse how the pediatric cardiology research community reacted to the above threat we performed a bibliometric-based synthetic knowledge synthesis study based on published research indexed in the Scopus bibliographic database.

**Results:** The search in Scopus resulted in 10068 documents. First indexed paper on the topic of pediatric cardiology as such was published in 1939 and till 1971 the research literature production was sparse, on average less than five papers per year. In the period 1971-1987, production increased but the growth was still linear. In 1988 the exponential growth began reaching its peak in 2015 with 542 publications. Then the product stabilized at around 500 publications, however, strongly increased in 2020 to almost 700 hundred publications (Figure 1). This sudden rise in productivity coincides with the emergence of the Covid-19 pandemic. Our analysis showed that among 1959 papers published in the period 2020-2022, 157 were devoted to Covid-19. Interestingly, no “Corona virus paper” related to pediatric cardiology was published before, despite some previous corona virus epidemics (SARS, MERS). The synthetic knowledge synthesis (Figure 2) showed that the following themes were researched: telemedicine, pediatric multisystem inflammations, myocarditis, rare diseases in children and congenital heart diseases. By far the most productive country



**Figure 1.** Indexed papers on the topic of pediatric cardiology through time



**Figure 2.** The scientific landscape of pediatric cardiology research in relation to Covid-19

was United State (n = 71), followed by Italy (n = 22) and United Kingdom (n = 18). The most productive institutions were Children's hospital Boston (n = 14), Harvard Medical School (n = 13) and University of Cincinnati College of Medicine (n = 12). The core journals were Cardiology in The Young (n = 14), Pediatric Cardiology (n = 12) and Frontiers in Pediatrics (n = 5). Most publications were funded by National Institutes of Health USA (n = 8) and National Heart, Lung and Blood Institute USA (n = 4).

**Conclusions:** Our study showed that pediatric cardiology research community reacted to Covid-19 pandemic with increased and focused research and an increased number of research publications.

**Keywords:** Public health, pediatric cardiology, Covid 19

#### P-406

##### **Efficacy of cibenzoline for hypertrophic obstructive cardiomyopathy in pediatric patients with RAS/MAPK pathway syndromes**

Kota Watanabe<sup>1</sup>, Yuji Maruo<sup>1</sup>, Atsuhito Takeda<sup>2</sup>

<sup>1</sup>Kitami Red Cross Hospital; <sup>2</sup>Hokkaido University Graduate School of Medicine

**Background and Aim:** RASopathies—caused by mutations in the RAS/MAPK signaling pathway—are frequently associated with cardiac diseases, such as hypertrophic obstructive cardiomyopathy (HOCM). Recently, cibenzoline has been reported to be effective in adults with HOCM; however, little is known about its effects in children. Here, we present the efficacy of cibenzoline in HOCM related to RASopathies in children with case reports.

**Method:** We present two cases of HOCM related to RASopathies in children who improved with cibenzoline.

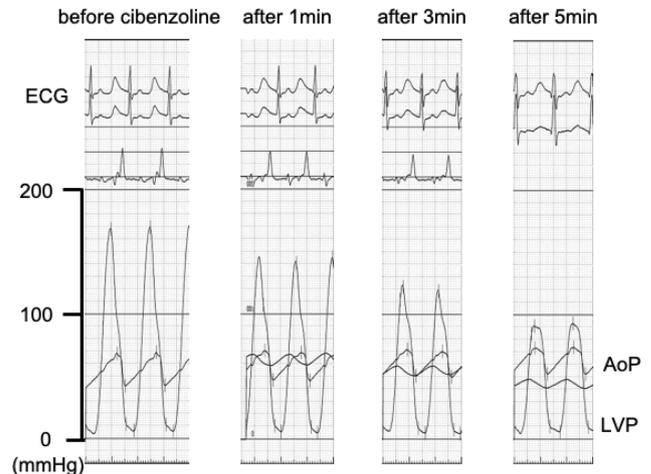
**Results:** Case 1: A 2-year-old boy with HOCM associated with Noonan syndrome with multiple lentiginos with a mutation in the PTPN11 gene (p.Y279C). Administration of propranolol was started for progressive left ventricular outflow tract stenosis (LVOTS) at the age of 1 year, but there was insufficient improvement. At the age of 2 years, a cibenzoline stress test was performed under cardiac catheterization, and the left ventricular pressure gradient improved from 86 to 23 mmHg, 5 minutes after cibenzoline injection (Fig). He has been successfully treated with oral cibenzoline.

Case 2: A 6-year-old boy with HOCM associated with Noonan syndrome with a mutation in the RAF1 gene (p.S257L). Administration of propranolol was started at 5 months of age for LVOTS with an estimated pressure gradient of 34 mmHg on echocardiography. At 19 months of age, oral cibenzoline was added to prevent further LVOTS progression. To date, no LVOTS progression has been observed.

**Conclusions:** In both cases of HOCM with RASopathies, cibenzoline was administered effectively and safely. Cibenzoline has the advantage that a loading test can be performed by cardiac catheterization and the dose can be adjusted while monitoring blood levels during oral administration. Moreover, it can be applied prophylactically to prevent the progression of HOCM, which is exacerbated during growth hormone treatment for patients with RASopathies. Cibenzoline is thus safe and effective for HOCM in children.

**Keywords:** hypertrophic obstructive cardiomyopathy, RASopathies, cibenzoline

The results of cardiac catheterization in case 1



Effect of cibenzoline administration on the LVP gradient. The maximum LVP gradient attenuated from 86 mmHg to 23 mmHg following intravenous injection of cibenzoline. No QT prolongation was observed in the ECG. ECG, electrocardiogram; AoP, aortic pressure; LVP, left ventricular pressure

#### P-408

##### **Significance of cardiac troponin in hypertrophic cardiomyopathy in the young**

Hirokuni Yamazawa, Atsuhito Takeda, Ayako Nagai Chida, Daisuke Sasaki, Takao Tsujioka, Yuji Maruo

Department of Pediatrics, Hokkaido University Hospital, Sapporo, Japan

**Background and Aim:** Cardiac troponin has been useful for diagnostic and prognostic evaluation in coronary artery disease, but its significance has rarely been reported in hypertrophic cardiomyopathy (HCM) in young patients. The possible causes of elevated myocardial troponin include microvascular ischemia, increased myocyte turnover, and fibrotic replacement, which are presumably related to factors such as wall thickness and late-gadolinium enhancement (LGE) in magnetic resonance imaging (MRI).

**Method:** Thirty-five patients with HCM were included in this study. Cardiac troponin I (cTnI) and T (cTnT) were log-transformed (LncTnI, LncTnT) and their relationship with major cardiovascular events (MACE), left ventricular ejection fraction (LVEF), maximum wall thickness (mWT), relative wall thickness (RWT), left atrial diameter (LAD), left ventricular outflow tract pressure gradient (LVPG) in echocardiography, left ventricular myocardial mass index (LVMI), extent of LGE in MRI, and the score of HCM Risk-Kids model was investigated. To determine the cutoff value of cardiac troponin for MACE, a receiver-operating characteristic (ROC) curve was constructed. The log-rank test was used for comparisons in the time-to-event analysis. The values have been described as median [interquartile range], and  $p < 0.05$  was statistically significant.

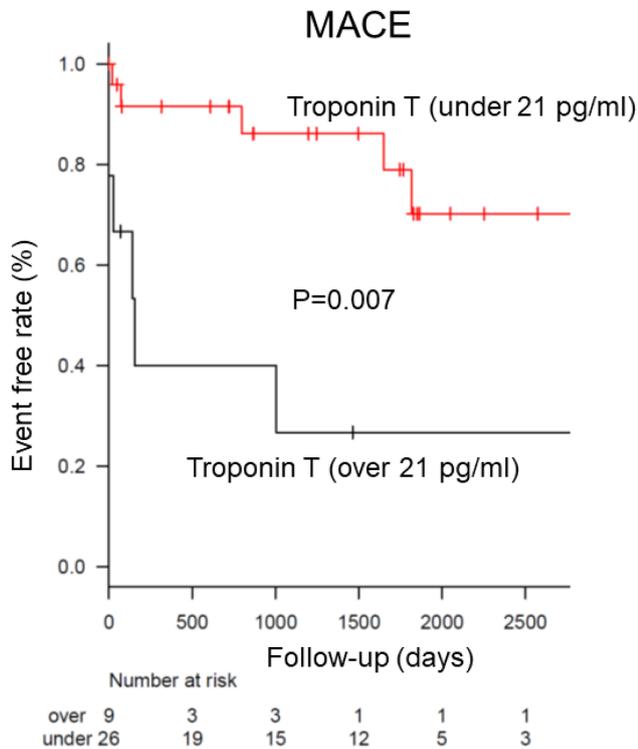
**Results:** The age of the subjects was 16 [12–18] years. The values of LncTnI and LncTnT with and without MACE were 3.7 [2.9–6.5] and 2.9 [2.3–4.1];  $p = 0.09$ , and 2.9 [2.4–3.9] and 2.4 [1.9–2.7];  $p = 0.04$ , respectively. These values showed a significantly positive correlation with mWT, RWT, LVPG, LVMI, and the score of the HCM Risk-Kids model. LncTnI also correlated significantly positively with the extent of LGE, and LncTnT with LAD, but LncTnT had a significantly negative correlation with the LVEF.

The ROC curve of cTnT levels to predict MACE showed that the cutoff value was 21 pg/ml, and the area under the curve was 0.72. The group with a cTnT level of 21 pg/ml or higher had more MACE than the group with a cTnT level of less than 21 pg/ml (log-rank  $p = 0.007$ ).

**Conclusions:** In the young, troponin levels are associated with the development of MACE and with risk factors for sudden cardiac death in HCM, such as mWT. This information may be useful for managing patients with HCM.

**Keywords:** cardiac troponin, hypertrophic cardiomyopathy, major cardiovascular events

Time-to-event analysis for MACE



**P-410**

**Is valve morphology associated with the risk for aortic dilation in patients with turner syndrome?**

Steve Bibeovski<sup>1</sup>, Mark Ruzmetov<sup>1</sup>, John Dentel<sup>1</sup>, Roman Yusupov<sup>2</sup>, Frank G. Scholl<sup>1</sup>, Kak Chen Chan<sup>3</sup>

<sup>1</sup>Division of Cardiothoracic Surgery, Joe DiMaggio Heart Institute, Hollywood, FL, USA; <sup>2</sup>Division of Genetics, Joe Dimaggio Children's Hospital, Hollywood, FL, USA; <sup>3</sup>Division of Pediatric Cardiology, Joe DiMaggio Children's Hospital, Hollywood, FL, USA

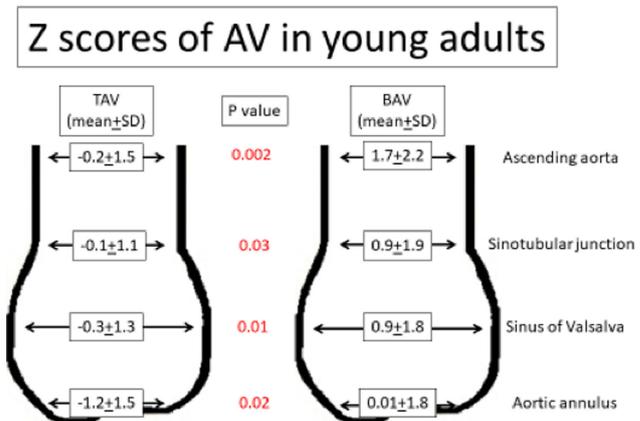
**Background and Aim:** Aortic dilation and dissection occur more frequently in patients with Turner syndrome (TS) than in the general population. Dilation of the aorta is also a frequent complication in patients with bicuspid aortic valve (BAV). The aim of this study was to determine the relationship between the aortic leaflet morphology, BAV versus tricuspid aortic valve (TAV) and the patterns of aortic dilation and valve dysfunction in patients with TS.

**Method:** We performed a retrospective review of all patients who presented with TS between 2012 and 2021 ( $n = 48$ ). All patients underwent cross-sectional M-mode and Doppler echocardiography. Aortic dimensions were compared with published norms and expressed as Z-scores relative to body surface area (BSA).

**Results:** Median age was 16 years. TAV was the most prevalent leaflet morphology (TAV,  $n = 25$ ; 52% vs. BAV,  $n = 23$ ; 48%), and right and left leaflet fusion was most prevalent subtype of BAV (22/23, 96%). The two groups were similar with respect to age, weight, height, BSA, diameter of aortic dimensions, presence of coarctation of the aorta, and previous cardiac procedures ( $p = NS$ ). The peak gradient on aortic valve (BAV,  $10.0 \pm 8.1$  vs. TAV,  $5.6 \pm 1.7$ ;  $p = 0.02$ ) and grade of aortic insufficiency (BAV,  $1.4 \pm 0.6$  vs. TAV,  $1.0 \pm 0$ ;  $p = 0.008$ ) were seen significantly more often in patients with BAV. Similarly, Z-score of all four aortic dimensions was significantly greater in patients with BAV (Figure).

**Conclusions:** In young patients with TS, aortic valve morphology may be associated with the propensity for aortic dilation and valve dysfunction. Patients with BAV were more likely to have aortic dilatation with the greatest risk being in the ascending aorta. Recognition of the increased risk in this subgroup may eventually be helpful for patient counseling and the planning of follow-up.

**Keywords:** bicuspid aortic valve, congenital heart surgery, Turner syndrome



**P-411**

**Dysregulation of hypoxia-inducible factor 1α in the sympathetic nervous system accelerates diabetic cardiomyopathy**

Petra Hrabalova<sup>1</sup>, Romana Bohuslavova<sup>2</sup>, Katerina Matejkova<sup>2</sup>, Frantisek Papousek<sup>3</sup>, David Sedmera<sup>4</sup>, Pavel Abaffy<sup>5</sup>, Frantisek Kolar<sup>3</sup>, Gabriela Pavlinkova<sup>2</sup>

<sup>1</sup>Laboratory of Molecular Pathogenetics, Institute of Biotechnology CAS, BIOCEV, Vestec, Czechia & Faculty of Science, Charles University, Prague, Czechia; <sup>2</sup>Laboratory of Molecular Pathogenetics, Institute of Biotechnology CAS, BIOCEV, Vestec, Czechia; <sup>3</sup>Institute of Physiology CAS, Prague, Czechia; <sup>4</sup>Institute of Anatomy, First Faculty of Medicine, Charles University, Prague, Czechia; <sup>5</sup>Laboratory of Gene Expression, Institute of Biotechnology CAS, BIOCEV, Vestec, Czechia

**Background and Aim:** The heart is innervated by the sympathetic, parasympathetic and sensory nerves. The cardiac sympathetic

nervous system plays an important role in the regulation of cardiac functions, such as increasing heart rate, myocardial contraction and conduction velocity. A change in sympathetic nerve fiber density leads to an imbalance that results in many cardiac pathologies, ranging from sudden infant death syndrome to common diseases of adulthood such as hypertension, myocardial ischemia and infarction, cardiac dysfunction and arrhythmias, sudden cardiac death and heart failure.

**Method:** This project uses a transgenic mouse model with a conditional deletion of *Hif1a* (*Hif1aCKO*) in sympathetic neurons. We evaluated how the diabetic environment in combination with HIF-1 $\alpha$  dysregulation affects the cardiac sympathetic system and heart function.

**Results:** Our results show that the two-month-exposure to the diabetic environment leads to the reduction of sympathetic innervation in the hearts of *Hif1aCKO* mice; the branching and thickness of innervation, together with the density of sympathetic neurons are affected. Additionally, we identified aberrant structural remodeling of the myocardium, vasculature changes, macrophage infiltration, and increased expression of cardiac injury markers in our diabetic *Hif1aCKO* mice compared with non-diabetic mice. Compared to the control, RNA sequencing of neurons isolated from *Hif1aCKO* sympathetic ganglia show differentially expressed genes associated with sympathetic neurons, heart function, and hypoxia.

**Conclusions:** We demonstrated that the elimination of *Hif1a* in the sympathetic nervous system in combination with diabetes results in changed transcriptome, compromised cardiac innervation, worsened heart function, and adverse myocardial remodeling.

**Keywords:** cardiac function, inflammation, diabetic cardiomyopathy, nervous system, sympathetic innervation

#### P-413

##### **Clinical characteristics and disease progression of sarcomeric infant-onset hypertrophic cardiomyopathy: A 10 year follow-up study**

Asvathy Ajith, Ella Field, Gabrielle Norrish, Juan Pablo Kaski  
Centre for Paediatric Inherited and Rare Cardiovascular Disease, UCL Institute of Cardiovascular Science, London, UK; Centre for Inherited Cardiovascular Diseases, Great Ormond Street Hospital, London, UK

**Background and Aim:** Up to one quarter of childhood hypertrophic cardiomyopathy (HCM) presents in infancy and early onset is associated with worse prognosis. Disease progression in this patient group is poorly understood, but may have important clinical implications. This study describes the clinical characteristics and disease progression in a cohort of patients with infant-onset sarcomeric HCM.

**Method:** A multicentre, retrospective European cohort of children presenting with non-syndromic HCM under the age of 1 year was formed. Clinical phenotype data (symptoms, medications, and echocardiography) were collected from baseline and over 10 years follow up.

**Results:** 64 patients (male n = 43, 67.2%) with sarcomeric HCM diagnosed <1 year of age were included. At baseline, 16 (34.8%) were symptomatic (heart failure symptoms, hypoxia,

neonatal respiratory distress, HIE with seizures) but the proportion of patients who remained symptomatic decreased over follow up (baseline vs 1 year n = 5 (11.6%) vs 5 years n = 6 (11.1%) vs 10 years n = 8 (19.5%), p = 0.020). At baseline, mean maximal left ventricular wall thickness (MLVWT) Z score was +11.7 (SD 5.2) with 47 (78.3%) asymmetric vs 4 (6.7%) concentric hypertrophy. Over follow up, 5 (21.7%) experienced an increase, 15 (65.2%) a decrease, and 3 (13.0%) no change in hypertrophy. The greatest rate of change occurred in the first year after diagnosis (mean rate of -1.9 [SD 5.8] per year).

**Conclusions:** The disease progression of infantile sarcomeric HCM in this cohort was highly variable over time. The highest symptom burden was in infancy but improved over time.

**Keywords:** child, sudden death, heart failure

#### P-414

##### **Echocardiographic functional examination in pediatric patients with becker muscular dystrophy**

Jihye You<sup>1</sup>, Mi Jin Kim<sup>2</sup>, Seulgi Cha<sup>2</sup>, Jae Suk Baek<sup>2</sup>, Mi Sun Yum<sup>2</sup>, Beom Hee Lee<sup>2</sup>, Jeong Jin Yu<sup>2</sup>

<sup>1</sup>Department of Pediatrics, Jeonbuk National University Children's Hospital, Jeonju, Korea; <sup>2</sup>Department of Pediatrics, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Republic of Korea

**Background and Aim:** Becker muscular dystrophy (BMD) is rare genetic disease, caused by mutations in the dystrophin gene. Cardiac involvement is a predominant feature in BMD. Little is known about the time of onset of subclinical cardiac function abnormalities. We conducted this study to compare the results of echocardiographic function in BMD patients with age, gender-matched control and to analyze the starting point of subclinical functional abnormality in BMD group.

**Method:** Seventeen pediatric patients with BMD who visited Seoul Asan Medical Center from January 1, 2002 to December 31, 2018 with analytic echocardiographic images were enrolled. 17 normal control subjects whose age and sex matched with BMD group were selected.

**Results:** Fractional shortening (FS) (32.9 $\pm$ 3.1 vs. 38.8 $\pm$ 3.7, %, P<0.001), the absolute value of global longitudinal strain (GLS) (-19.5 $\pm$ 2.5 vs. -22.1 $\pm$ 3.7, %, P = 0.043) were lower in BMD patients than control groups. The absolute values of four-chamber LS (P = 0.027), GLS (P = 0.043), global longitudinal strain rate (GLSR) (P = 0.022), RV free wall LS (P = 0.009), RV fractional area change (FAC) (P = 0.015) tend to decrease with aging. Even in BMD subgroup of under 10 years of age, FS (P = 0.010) and the absolute value of basal inferior segmental LS (P = 0.028) were lower than control subgroup.

**Conclusions:** Echocardiographic parameters including deformation values are decreased in BMD patients compared to control group. This change depends on patient's age. Early intervention with prophylactic angiotensin-converting enzyme inhibitor (ACEi) might be suggested to protect ventricular dysfunction even in patients under 10 years of age.

**Keywords:** Echocardiography, Becker muscular dystrophy, Left Ventricular Mass, Pediatric, Strain

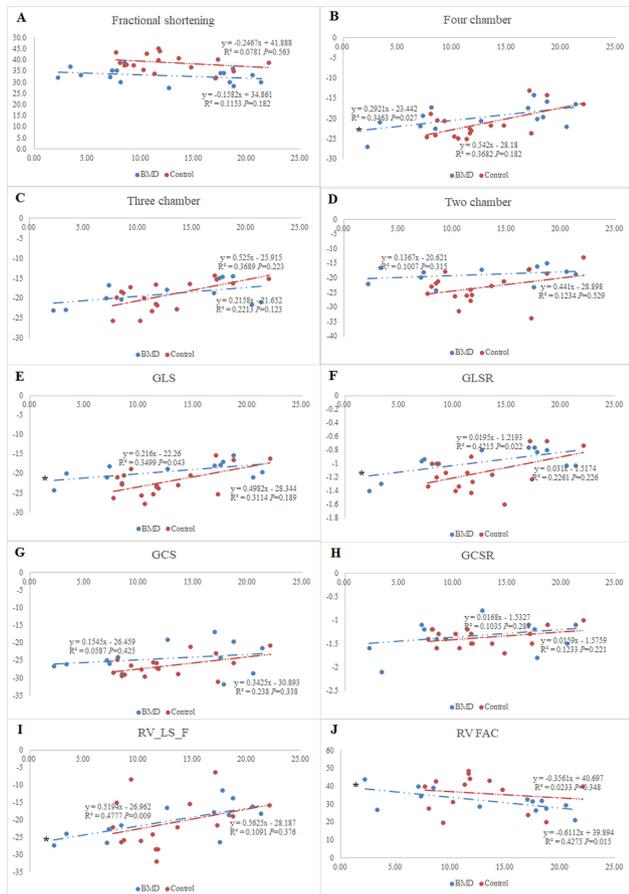


Image 1 Echocardiographic parameters depending on age

P-415

**A novel MYH7 variant in a five-generation-family with a severe presentation of hypertrophic cardiomyopathy**

Magda Franke<sup>1</sup>, Tomasz Marcin Ksiaczczyk<sup>2</sup>, Dorota Czupczak<sup>3</sup>, Urszula Demkow<sup>3</sup>, Bożena Werner<sup>2</sup>

<sup>1</sup>Department of Pediatric Cardiology and General Pediatrics, Doctoral School, Medical University of Warsaw, Warsaw, Poland; <sup>2</sup>Department of Pediatric Cardiology and General Pediatrics, Medical University of Warsaw, Warsaw, Poland; <sup>3</sup>Department of Laboratory Diagnostics and Clinical Immunology of Developmental Age, Medical University of Warsaw, Warsaw, Poland

**Background and Aim:** Hypertrophic cardiomyopathy (HCM) composes the most frequent inherited cardiac disease with a prevalence of 1:500. Variants in genes encoding sarcomeric proteins are most commonly present. Clinical presentation of the disease strongly varies between affected individuals. The aim of the study was to report a novel MYH7 variant responsible for HCM with left ventricular outflow tract obstruction in a 5-generation-family.

**Method:** Based on two-dimensional Doppler echocardiogram and cardiovascular magnetic resonance diagnosis of HCM in members of the presented family was established following the ESC HCM criteria. Genetic testing of all individuals, using Next Generation Sequencing or Sanger method, were then performed to ascertain the diagnosis.

**Results:** Medical history of the family begun with a prenatal diagnosis of HCM in a first child (female) of a family with so far healthy

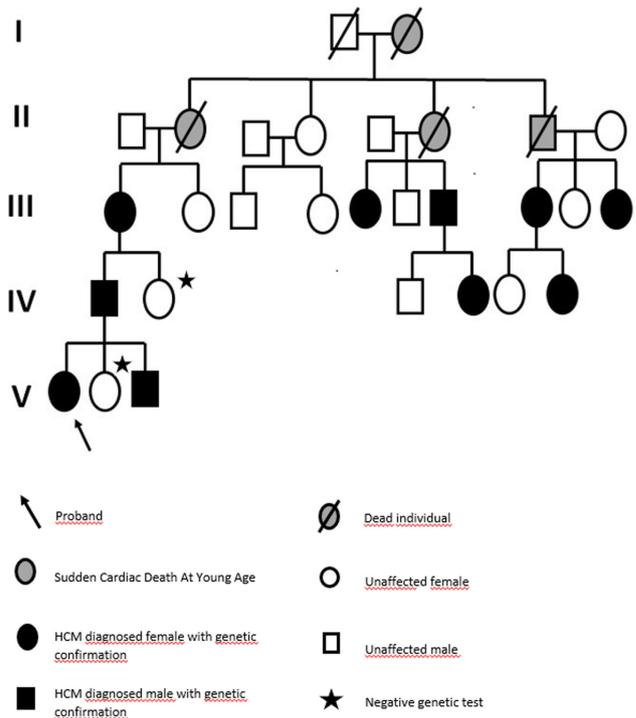
parents. A thorough interview revealed a long family history of SCD and cardiological problems.

Presence of a novel heterozygous variant, NM\_000257.2: c.[2342T>A(p.Leu781Gln)] was detected in the MYH7 gene in the proband. It is a single base exchange resulting in a single amino acid substitution in the 781 codon altering Leucine for Glycine. The variant was genetically confirmed in 10 members of the family, all of whom have a positive phenotype. Unaffected members of the family were proven not to carry the variant. The clinical course was severe: 3 members of the family died of a sudden cardiac death under the age of 40, 3 members had ICDs implanted, 1 needed a heart transplant and 3 had a septal myectomy performed. Two other members were qualified for an ICD implantation, including our proband.

**Conclusions:** In this study, we suggest that the novel missense mutation is responsible for the phenotype observed in the affected members of the described family. The presented family shows full spectrum of possible complications of the HCM. Genetic basis verification is crucial in order to anticipate the course and severity of the disease and to determine strategy for the family members of the affected individual.

**Keywords:** HCM, genetic variant, MYH7

Pedigree of a 5-generation-family with HCM



P-416

**Metoprolol has beneficial effect on heart rate variability in hypertrophic cardiomyopathy: A randomised study**

Ingegerd Östman Smith<sup>1</sup>, Daniel Petersson<sup>2</sup>

<sup>1</sup>Institute of Clinical Sciences, Sahlgrenska Academy and Childrens Heart Center, Queen Silvia Childrens Hospital, Gothenburg, Sweden; <sup>2</sup>Department of Clinical Physiology, Sahlgrenska University Hospital, Gothenburg, Sweden

**Background and Aim:** Hypertrophic cardiomyopathy (HCM) is a disease with poly-genetic etiology, where it has been hypothesized that excessive sympathetic nervous activity may have a role in inducing the pathological hypertrophy. Disordered heart rate variability (HRV) with a dominance of sympathetic activity has been correlated with an increased risk of sudden cardiac death (SCD) in HCM and other cardiac pathologies. HCM is the most common cause of unexpected SCD in older children and teenagers. Beta-blocker therapy is the treatment of choice for symptomatic HCM-patients, but its use in asymptomatic HCM-patients is controversial. We aimed to study how the HRV changed over time in asymptomatic HCM-patients, and if beta-blocker treatment would affect HRV.

**Method:** Asymptomatic patients with familial HCM newly diagnosed by family screening, median age 17y, with no risk factors for SCD, were randomised to life-style advice (CONTROLS; n = 16) or life-style advice plus treatment with metoprolol (BBL-group; n = 15). Metoprolol is a relatively selective beta-1 receptor blocker which is also lipophilic. The beta-blockade was increased stepwise over 9–12 months with a standardized protocol. Final median dose was 325 mg [IQR 300–400] per day, corresponding to 3.7 [3.4–4.3]mg/kg BW/day, and median follow-up was 6y. All patients underwent 5 min HRV analysis with Welch–Allyn CardioPerfect software after minimum 20 min supine rest yearly. **Results:** At base-line the low-frequency to high-frequency power-ratio (LF/HF ratio) showed a wide range, with no significant difference between the groups (Controls 0.71 [IQR 0.42–1.00], BBL-group 0.89 [0.56–1.98; p = 0.3]. After two years of follow-up the LF/HF ratio had increased significantly in CONTROLS (p = 0.017), whereas it had reduced significantly in BBL-group to 0.44 [0.26–0.67], p = 0.001, and the difference between the two-year values of the groups was significant (p = 0.035), with even clearer differences after five-year follow up (p = 0.003), when the BBL-group had an LF/HF ratio of 0.36 [0.19–0.54], p = 0.005 versus base-line. Improvement in HRV in BBL-group was confirmed by a significant increase in proportion of successive heart-beat interval-difference exceeding 50 msec (PNN50) at two-year follow-up to 0.46 [0.19–0.62], (p = 0.013).

**Conclusions:** Treatment with high-dose metoprolol leads to a significant improvement in autonomic balance in HCM patients, in a direction potentially reducing risk for SCD.

**Keywords:** Hypertrophic cardiomyopathy, heart rate variability, LF/HF ratio, PNN50

#### P-417

##### Heart failure with preserved ejection fraction (HFpEF) in children

Sophie Quennelle<sup>1</sup>, Lucile Houyel<sup>2</sup>, Zahra Belhadjer<sup>2</sup>, Julien Grynblat<sup>2</sup>, Antoine Neuraz<sup>3</sup>, Nicolas Garcelon<sup>4</sup>, Damien Bonnet<sup>5</sup>

<sup>1</sup>M3C-Necker Enfants malades, AP-HP, Paris, France; HeKA Team, Inria, Inserm UMRS1138, Paris, France; Sorbonne université, Paris, France; Université de Paris Cité, Paris, France; <sup>2</sup>M3C-Necker Enfants malades, AP-HP, Paris, France; <sup>3</sup>M3C-Necker Enfants malades, AP-HP, Paris, France; HeKA Team, Inria, Inserm UMRS1138, Paris, France; Université de Paris Cité, Paris, France; <sup>4</sup>HeKA Team, Inria, Inserm UMRS1138, Paris, France; Data Science Platform, Imagine Institute, Université de Paris Cité, Paris, France; <sup>5</sup>M3C-Necker Enfants malades, AP-HP, Paris, France; Université de Paris Cité, Paris, France

**Background and Aim:** Heart failure (HF) with preserved ejection fraction (HFpEF) is a complex cardiac condition that results from a structural or functional impairment of filling of the heart. It has a high prevalence in elderly but has rarely been analyzed in children. Our objective was to identify HFpEF in children with

biventricular physiology without shunt or outflow tract obstruction, through a dedicated algorithm applied to a large tertiary hospital database, and to describe the causes, hemodynamic profiles, treatment, and hard outcomes of this population.

**Method:** We applied a combination of clinical, echocardiographic, and hemodynamic measurements to confirm the diagnosis of HFpEF to identify children with a confirmed HFpEF phenotype extracted from a pediatric hospital data warehouse.

**Results:** 137 HFpEF cases were included. Mean age was 7±5.7 years. The cardiovascular causes of HFpEF were hypertrophic cardiomyopathy (44%), restrictive cardiomyopathy (RCM) (32%), cardiomyopathy due to repeated cardiac surgeries (10%), dilated cardiomyopathy (7%), and constrictive pericarditis (7%). Children with RCM had the worst prognosis (27% ten years transplant-free survival, p < 0.005). Mean patient follow-up was 5.3 years. 43% of the patients were hospitalized for acute HF, 12% were transplanted and 31% died. N-terminal pro b-type natriuretic peptide (NT-proBNP) increase was found in all groups and was associated with mortality and transplantation, HR 1.91 (95% CI 1.31, 2.76, p-value < 0.001). Based on echocardiographic evaluation, 56% of the patients were diagnosed with pulmonary hypertension (PH), associated with mortality and transplantation, HR 2.84 (95% CI 1.58, 5.10, p < 0.001). In the 48 patients who underwent right heart catheterization, post-capillary or combined PH were the most frequent but the 15% with isolated pre-capillary PH had poorer outcomes, HR 4.60 (95% CI, 1.46, 14.5, p = 0.009).

**Conclusions:** HFpEF is a rare and concerning condition in children. Younger age, RCM, acute heart failure episodes, and increased NT-proBNP are associated with hard outcomes. A precapillary component in patients with pulmonary hypertension is associated with poorer outcomes.

**Keywords:** children, cardiology, cardiomyopathies, diastolic heart failure, pulmonary hypertension

#### P-418

##### Influence of pulmonary valve regurgitation on right-ventricular function and morphology in a juvenile porcine model

Philippe Grieshaber<sup>1</sup>, Thomas Rösch<sup>2</sup>, Moritz Hippchen<sup>2</sup>, Nils Herion<sup>2</sup>, Peter Rose<sup>1</sup>, Florian André<sup>3</sup>, Gregor Warnecke<sup>1</sup>, Matthias Karck<sup>1</sup>, Tsvetomir Loukanov<sup>1</sup>, Matthias Gorenflo<sup>2</sup>

<sup>1</sup>Department of Cardiac Surgery, Heidelberg University Hospital;

<sup>2</sup>Department of Congenital Heart Disease and Pediatric Cardiology;

<sup>3</sup>Department of Cardiology

**Background and Aim:** Right-ventricular (RV) failure secondary to RV volume or pressure overload is frequently encountered in patients with congenital heart disease. The functional and structural features of RV failure are currently insufficiently understood. This study aimed to characterize the influence of experimentally induced pulmonary valve (PV) regurgitation (PR) on RV morphology and function in a juvenile porcine model.

**Method:** One-month-old piglets underwent surgical PR induction. The animals were randomly assigned to the PR or the control group. After left-lateral thoracostomy, the PV cusps of the lateral hemi-circumference were attached to the pulmonary artery wall by external placement of plication sutures, thus creating PR. In control group animals, a sham procedure without placement of the plication sutures was conducted. After 10 weeks, the animals underwent cardiac magnetic resonance imaging (MRI) in order to objectify the degree of PR, static and dynamic measurements of the RV and the LV dimensions and functions, respectively.

**Results:** 12 piglets (weight 15±2.5kg) were included in the experiment. 4 animals died, thereof 2 because of immediate perioperative RV failure, 1 because of ventricular arrhythmia after central line placement and 1 due to pulmonary infection 9 days postoperatively. The remaining 8 animals (PV group n = 5, control group n = 3) underwent MRI after a median of 70±2 days with a median weight of 47±7.9kg. The PR group showed a significantly increased RV stroke volume (95±20 vs. 64±13ml; p = 0.041) and pulmonary valve regurgitation fraction (23±17 vs. 0.3 ±0.02%; p = 0.031) compared with the control group. The RV enddiastolic volume (PR:130±21ml, control:87±16ml; p = 0.012), endsystolic volume (PR:35±10ml, control:23 ±2.4ml; p = 0.043) and myocardial mass (PR:34±4g, control:23 ±4g; p = 0.05) were increased in the PR group, while the RV ejection fractions were comparable in both groups (PR:73±8% control:73±3%; p = 0.046). Circumferential Of note, the differences in in RV volumes were not reflected by the RV end-diastolic (PR:33±7mm, control:28±6mm; p = 0.19) and endsystolic (PR:13±3mm, control:10±1.6mm; p = 0.14) diameters. The left-ventricular and left- and right-atrial parameters did not differ between the groups.

**Conclusions:** Despite high vulnerability in this young animal model, the model results in reproducible induction of PR and RV volume overload. The effects can be objectified using cardiac MRI.

**Keywords:** Pulmonary regurgitation, right heart failure, exerimental, animal model

**P-419**  
**Cardiomyopathies in children with systemic disorders can cardiogenetic approach influence outcome in clinical practice?**

Anwar Baban<sup>1</sup>, Giovanni Parlapiano<sup>1</sup>, Viola Alesi<sup>2</sup>, Monia Magliozzi<sup>2</sup>, Maria Gnazzo<sup>2</sup>, Francesca Romana Lepri<sup>2</sup>, Federica Cali<sup>1</sup>, Lorenzo De Luca<sup>1</sup>, Marianna Cicienia<sup>1</sup>, Pasqualina Bruno<sup>1</sup>, Luca Di Chiara<sup>1</sup>, Enrico Bertini<sup>3</sup>, Carlo Dionisi Vici<sup>4</sup>, Antonio Novelli<sup>2</sup>, Fabrizio Drago<sup>1</sup>  
<sup>1</sup>The European Reference Network for Rare, Low Prevalence and Complex Diseases of the Heart-ERN GUARD-Heart, Department of Pediatric Cardiology and Cardiac Surgery, Heart and Lung Transplantation, Bambino Gesù Children Hospital and Research Institute, IRCCS, 00165 Rome, Italy; <sup>2</sup>Laboratory of Medical Genetics, Bambino Gesù Children Hospital and Research Institute, IRCCS, 00165 Rome, Italy; <sup>3</sup>The European Reference Network for Neuromuscular Disorders (ERN NMD), Unit of Neuromuscular and Neurodegenerative Disorders, Bambino Gesù Children Hospital and Research Institute, IRCCS, 00146 Rome, Italy; <sup>4</sup>Division of Metabolism, Bambino Gesù Children Hospital and Research Institute, IRCCS, 00165 Rome, Italy

**Background and Aim:** Etiology of CMPs in children is extremely heterogenous. It can be secondary to myocarditis, metabolic, toxic, malformative, or genetic background. On one hand, isolated CMP in children is a frequent finding. On the other hand, complex multisystemic forms of children CMP are even more heterogenous. Few studies take into consideration this topic as the main core since it represents a rarity (systemic CMP) within a rarity (pediatric population CMP).

**Method:** Our tertiary academic pediatric center represent the referral point for the center and south of Italy for the multidisciplinary management of patients with CMPs. In this study we include data from 450 patients who received multisystemic clinical assessment, multiorgan screening, metabolic investigations and genetic study through and NGS sequencing and/or microarray.

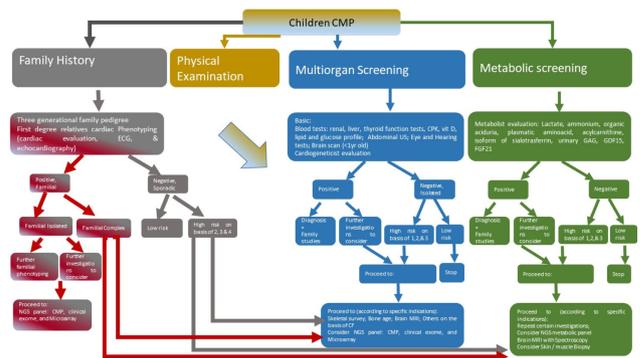
The main focus of this abstract is to discuss rare multisystemic manifestations that were diagnosed, clinical approach, and therapeutic options on the basis on underlying background.

**Results:** multisystemic involvement in children CMPs was variable according to the type of underlying CMPs. In other words, it was more observed in hypertrophic CMP compared to arrhythmogenic CMP. Restrictive CMP pediatric onset showed a wide range from subtle musculoskeletal manifestations up to diffuse arthrogryposis.

**Conclusions:** Identifying etiology in this cohort requires a dedicated team work. It might be essential for understanding prognosis, risk stratification, eligibility to heart transplantation and/or mechanical-assisted procedures, preventing multiorgan complications, and relatives' recurrence risk calculation. The previous points represent a cornerstone in patients' empowerment and personalized medical care approach. The aim of this work is to propose a new approach for an algorithm in the setting of the diagnostic framework of systemic pediatric CMP.

Part of this topic is reviewed in: PMID: 35200700 PMID: PMC8877723 DOI: 10.3390/jedd9020047

**Keywords:** CMP and systemic disorders, CMP & short stature syndromes; CMP and dysmorphic features and ID, CMP and immunological involvement, CMP and ectodermal abnormalities, CMP and high inbreeding.



**Figure 1** shows the proposed algorithm for systemic screening of children with CMP.

**P-420**  
**Paediatric dilated cardiomyopathy in khartoum state, sudan: A prospective study**

Osama Hafiz Elshazali<sup>1</sup>, Marian Zaki<sup>1</sup>, Ekhlas Abdalla<sup>2</sup>  
<sup>1</sup>Department of Paediatrics, University of Khartoum, Sudan; <sup>2</sup>Ahmed Gasim Paediatric Hospital, Khartoum North, Sudan

**Background and Aim:** Dilated Cardiomyopathy (DCM) is a disease of the heart muscle that affects ventricular function. It is a debilitating disease that can lead to heart failure and death in children. There is a lack of good epidemiological data about paediatric dilated cardiomyopathy in Sudan or in Sub-Saharan Africa.

Practically all the data about Paediatric DCM came from high income countries. This research attempts to provide insight about the incidence, prevalence, causes and the outcome of paediatric DCM in Khartoum state, Sudan.

**Method:** A Prospective cross-sectional study that was conducted in three paediatric hospitals in Khartoum state, Sudan over six months

**Results:** During the study period 55 children were seen with the diagnosis of DCM. The female: male ratio was 1.9:1. The incidence of DCM in Khartoum state was found to be 1.4 cases per 100, 000 child per year, 10 of the children (18.2%) were diagnosed in the first year of life. 27 of the children (49.1%) showed evidence of failure to thrive being below the 5th percentile for weight. The most common cause of DCM was found to be post viral in 27 children (49%) followed by Idiopathic in 25 children (45.5%). The most common presentation was shortness of breath in 53 children (97%), on echocardiography 26 children (47.%) showed evidence of severe left ventricular impairment and 23 children(42%) showed moderate ventricular impairment.

Regarding the outcome eight children (14.5%) recovered, the condition of 11 children (20%) remained static, and there were 36 deaths (65.5%).

**Conclusions:** The incidence of DCM in Sudanese children was found to higher when compared to the developed world. The children in Sudan tend to present older, malnourished, with severe symptoms and with significantly impaired ventricular function all of these factors contribute to the high mortality. Improvements in the entire health system are needed to tackle this condition

**Keywords:** Dilated Cardiomyopathy, Children, Sudan

#### P-421

##### Genetic analysis in childhood-onset dilated aortic root in a consanguineous population

Abeer Essa Almostafa<sup>1</sup>, Mohammed Saleh Alhabdan<sup>2</sup>, Nadiyah Salem Alruvaili<sup>2</sup>, Maarab Mohammed Alkorashy<sup>1</sup>, Abdullah Mohammed Alquae<sup>3</sup>, Dimpna C Albert Brotons<sup>2</sup>, Naji Ali Alkholajif<sup>2</sup>, Abdulrahman Abdulaziz Almesned<sup>3</sup>, Zuhair Nasser Alhassnan<sup>1</sup>  
<sup>1</sup>Center of Genomic Medicine, King Faisal Specialist Hospital & Research Centre, Riyadh; <sup>2</sup>Heart center Department, King Faisal Specialist Hospital & Research Centre, Riyadh; <sup>3</sup>Prince Sultan Cardiac Center, Qassim, Saudi Arabia

**Background and Aim:** Childhood-onset thoracic aortic dilatation (TAD)

is a heterogenous group of genetic condition the inheritance of which is largely in a dominant pattern. To our knowledge, the influence of consanguinity on the genetics of childhood-onset TAD has not been addressed with a systematic approach. We aim to study a cohort of children with TAD in our highly consanguineous population.

**Method:** Saudi children with TAD were consecutively recruited and underwent categorized genetic analysis. Based on the likelihood of a founder mutation and on the clinical diagnosis, genetic test was categorized to either targeted mutation test (TMT), single gene test, multi-gene panel, or whole exome sequencing.

**Results:** Thirty children with TAD, from 23 families, were recruited in the study. Genetic analysis detected 21 variants, 6 of which were novel. Homozygous variants were detected in 14 cases (47%) while 16 cases (53%) had heterozygous variants. A single founder variant in EFEMP2 accounted for most of the cases (n = 13 in 5 families). However, de novo variants in FBN1 were detected in more probands (n = 6, 26%).

**Conclusions:** Our work illustrates the impact of consanguinity on the genetics of childhood-onset TAD and the value of adopting a categorized genetic approach in a consanguineous population. Our data suggest that if a founder mutation is not suspected, adopting next generation sequencing as a first-line test should be considered.

**Keywords:** Dilated Aortic Root, Target mutation

#### P-422

##### Impact of pulmonary artery banding on the prognosis of genetic dilated cardiomyopathy in toddlers

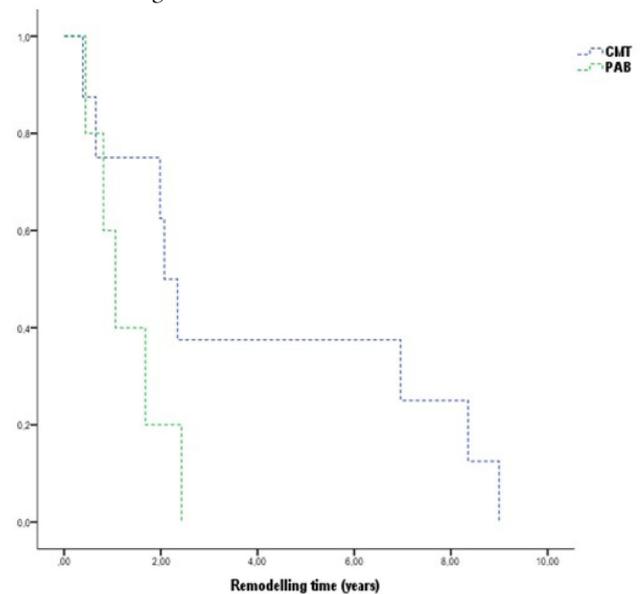
Elena Panaoli, Margaux Pontailier, Antoine Legendre, Regis Gaudin, Olivier Raisky, Damien Bonnet, Diala Khraiche  
 Unité médico-chirurgicale de cardiologie congénitale et pédiatrique, centre de référence des maladies cardiaques congénitales complexes - M3C, Hôpital universitaire Necker-Enfants Malades, Université de Paris, France

**Background and Aim:** Dilated cardiomyopathy (DCM) in children is a rare disease with poor prognosis when diagnosed within the first year of life. The limited availability of heart transplantation in infants has promoted alternative therapeutic strategies such as pulmonary artery banding (PAB). However, the impact of such strategy on survival is still unknown. The first aim of our study is to compare the prognosis of toddlers with genetic DCM or congenital left ventricular aneurysm (CLVA) with severe cardiac dysfunction, treated with conventional medical treatment (CMT) or with PAB. The second aim is the evaluation of left ventricular remodelling.

**Method:** We analysed 39 children with a diagnosis of DCM (genetic or CLVA) in our institution between 2010 and 2022. Among them, 12 patients received PAB. The median time of follow up in the overall population is 4 (1.6-7, 4 years). We collected demographic, biological and echocardiographic data at the diagnosis, at 6 months and 1 years of follow up. We compared the survival and left ventricular (LV) remodelling defined as the rate of decrease of the LV end diastolic diameter (LVEDD) between the group of patients treated by PAB and the group treated by CMT.

**Results:** The median age at diagnosis was 0.32 years (0-1.36 years). 46% of patients were in WHOFC class 4 needing an ICU's admission. 50% of patients needed inotropic and ventilation support in the PAB group vs 44% in the CMT group (p = 0.825). LVEDD was higher in the PAB group than in the CMT group (Z score 9, 6 vs 6, 12; p = 0, 05) with no difference in LV function (26, 2 vs 27, 5; p = 0, 75). There was a trend towards a lower mortality in the

LV remodelling time



Time to left ventricle remodelling in terms of reduction of left ventricle dilatation and/or improvement of left ventricle ejection fraction

PAB group (20% vs 48%  $p = 0.065$ ). There was a significant difference in terms of rate of LV remodelling at 6 months between the PAB group (Delta Zscore of LVEDD at 6 months  $6, 9+/-4$ ) and the CMT group ( $3, 4 +/-5$ ;  $p 0.02$ ).

**Conclusions:** We found a trend towards a lower mortality in toddlers with genetic DCM or CVLA treated by PAB and a better rate of LV remodelling. These promising results need to be confirmed in a prospective study.

**Keywords:** dilated cardiomyopathy, pulmonary artery banding, heart failure

#### P-423

##### **Potential genes involved in nonsyndromic ventricular septal defects of congenital heart disease**

Chaitra Shanthithadda

Department of Studies in Genetics and Genomics, University of Mysore

**Background and Aim:** Congenital heart disease (CHD) is mainly characterized by defects in septation, proper partitioning of the great vessels, and valve formation during cardiac development. The ventricular septal defect (VSD) is the common type of CHD and accounts for 20–40%. The differential expression of the genes at different stages of development is responsible for normal heart development. It is regulated by interactions of transcription factors, ligands, receptors, contractile proteins and signalling pathways. Any disruption in these events will result in congenital malformations of the heart. Although several studies have been made, the pathogenic key factors involved in manifesting nonsyndromic VSD of CHD need to be understood. Therefore, we aim to understand the high-confidence genes and their pathogenicity for VSD of CHD.

**Method:** A systematic study was conducted to identify the high-confidence genes through text mining by employing different databases, in silico analysis using SIFT, PolyPhen, Mutation Taster, Mutation assessor, and CADD prediction tools to predict the pathogenicity of gene variations. To check the protein–protein interactions, the pathway network analysis was employed.

**Results:** We identified 42 genes through text mining using different databases. Of the 42 genes, 24 genes were found coding for transcription factors and 18 genes for enzymes, ligands, receptors and other proteins. Among these, 29 genes were found in 42 Chinese population studies. About 19 variations were observed in GATA4; five variations showed a high level of pathogenicity. The most frequent variant, c.1325C>T, in GATA4 was found in the Chinese and Iranian populations with a high level of pathogenicity. Pathway analysis also confirmed the involvement of these genes in the manifestation of CHD.

**Conclusions:** This study revealed that more transcription factors are involved in heart development, indicating their major role in the manifestation of VSD. This will help us to characterize these genes further to create a gene panel for a better diagnosis.

**Keywords:** Congenital heart disease, Ventricular septal defect, Genetics

#### P-424

##### **Woolly hair palmoplantar keratoderma cardiac abnormalities: naxos and naxos-like syndrome**

Maha Mubarak Binfadel<sup>1</sup>, Dimpna Calila Albert Brotons<sup>1</sup>, Zuhair Nasser Alhassnan<sup>2</sup>, Olga Vriz<sup>3</sup>, Nadiyah Salem Alnuwaili<sup>4</sup>

<sup>1</sup>Pediatric cardiology department, King Faisal Specialist Hospital, Riyadh, Saudi Arabia; <sup>2</sup>Genetic department, King Faisal Specialist Hospital, Riyadh, Saudi Arabia; <sup>3</sup>Adult cardiology department, King Faisal

Specialist Hospital, Riyadh, Saudi Arabia; <sup>4</sup>Research department, King Faisal Specialist Hospital, Riyadh, Saudi Arabia

**Background and Aim:** Naxos disease is a rare autosomal recessive genodermatosis that was first reported in the families descending from the Hellenic Island of Naxos in Greece. Mutations in the genes encoding the desmosomal proteins have been identified as the cause. A syndrome with the same cutaneous phenotype and predominantly LV involvement has been described in families from India and Ecuador (Carvajal syndrome). Naxos disease associates arrhythmogenic RV cardiomyopathy/dysplasia with woolly hair and palmoplantar keratoderma. It is usually with an autosomal recessive inheritance; however, dominant forms have been reported. Woolly hair is usually present from birth. Skin histology reveals nonspecific features of keratoderma. Usually there are no signs of heart disease up to the age of 12 years. Although heterozygous carriers do not develop the disease. Few heterozygotes present with woolly hair with or without mild palmoplantar keratoderma and minor cardiac abnormalities not fulfilling the diagnostic criteria for ARVC/D. The hair/skin phenotype in a child should alert for signs of cardiomyopathy. Annual follow-up with ECG, 24-hour Holter and echocardiography to detection of early signs consistent with ARVC/D should be applied.

**Method:** a cross-sectional retrospective cohort study of children treated in our Pediatric Heart Failure Clinic with depressed function and clinical signs of Naxos Syndrome and variants (woolly hair + mucocutaneous abnormalities)

**Results:** We are reporting 10 cases from 6 Saudi families with very rare genetic cardio-cutaneous Syndrome, all patients have woolly hair +/- palmoplantar keratoderma with severely dilated and depressed LV function +/- depressed RV function. Genetic study for all came homozygous for DSP gene. Some of those patients has family history of sudden deaths. 4 patients needed ICD implantation because uncontrolled ventricular tachycardia despite maximum medical therapy. 4 patients needed heart transplantation. 3 of them died suddenly while waiting for heart transplantation and 3 waiting for heart transplantation.

**Conclusions:** Nexos disease is a very rare genetic cardiocutaneous syndrome manifesting with very rapid progressing cardiomyopathy and arrhythmia. It manifests with woolly hair and keratosis of the extremities with high risk for arrhythmia and sudden cardiac deaths. Those patients need extensive evaluation and close follow up and as early as possible heart transplant work up.

**Keywords:** Naxos disease, Carvajal syndrome, arrhythmogenic right ventricular cardiomyopathy/dysplasia, desmosomal proteins

#### P-425

##### **Successful rescue therapy with everolimus in an infant with noonan syndrome with multiple lentigines**

Martin Cremer<sup>1</sup>, Martin Glöckler<sup>2</sup>, Alexander Kadner<sup>3</sup>, Cordula Wolf<sup>4</sup>, Birgit Donner<sup>1</sup>

<sup>1</sup>Department for Pediatric Cardiology, University Children's Hospital Basle (UKBB), Basel, Switzerland; <sup>2</sup>Center for Congenital Heart Disease, Department of Cardiology, Pediatric Cardiology, Inselspital, Berne University Hospital, University of Berne, Berne, Switzerland; <sup>3</sup>Centre for Congenital Heart Disease, Department for Cardiovascular Surgery, Inselspital, Berne University Hospital, University of Berne, Berne, Switzerland; <sup>4</sup>Department of Congenital Heart Defects and Pediatric Cardiology, German Heart Center Munich, Technical University of Munich, School of Medicine, Munich, Germany

**Background and Aim:** Noonan syndrome with multiple lentigines (NSML) formerly known as LEOPARD-Syndrome is a rare genetic condition frequently associated with hypertrophic

obstructive cardiomyopathy (HOCM). With increasing knowledge about underlying mutations novel therapeutic options arise. *Method:* We report about a boy with a heterozygous de novo-mutation of the PTPN11 gene, C.836A>C, P. (Tyr279Ser). The congenital HCM progressed to significantly biventricular outflow tract obstructions and a modest mitral valve insufficiency. Despite a muscle resection in both outflow tracts at the age of 18 weeks a progressive, critical left ventricular outflow tract obstruction (LVOTO) developed. Considering basic research studies and a case report (Hahn A et al., 2015, Am J Med Genet Part A 167A:744–751) everolimus was started as an individual therapeutic approach at the age of 26 weeks for 11 months. *Results:* In the following months a decrease of the LVOTO gradient from 80mmHg to less than 20mmHg and a drop from NT-proBNP levels from >36'000 ng/l preoperatively to 14'000 ng/l postoperatively to lastly 1200 ng/l occurred. Minimal side effects were mild aphthosis and a clinically insignificant platelet dysfunction.

*Conclusions:* To our knowledge this is the second published case with everolimus as a treatment in NSML with HOCM and the first case with a notable decrease of the obstruction. This case adds to the growing experience with intracellular inhibition of signaling molecules in the treatment of HOCM in RASopathies based on the knowledge of exact mutations. Similar examples were described using MEK-Inhibitors (e.g., trametinib) in Noonan Syndrom with RIT mutations and NSML with PTPN11 gain of function mutations. These cases emphasize the growing importance of genetic analysis and the potential of translational medicine.

**Keywords:** Noonan Syndrom with Multiple Lentiginos, Hypertrophic Obstructive Cardiomyopathy, Everolimus, Genetics, Rasopathies

#### P-426

##### **Risk stratification for ascending aortic dilatation in young Turner syndrome patients - is it possible to predict?**

Julian Heno, Christiane Pees, Ina Michel Behnke

Department of Pediatric Cardiology, Medical University of Vienna, Austria

*Background and Aim:* Cardiac implications of Turner Syndrome (TS) include coarctation of the aorta (CoA) and bicuspid aortic valves (BAV). Ascending aortic dilatation is detected as early as in childhood even in patients without congenital heart defects, predisposing these patients to aortic dissection and indicative of an underlying vessel pathology. We report on the longitudinal findings of aortic root dimensions and elasticity in our pediatric Turner Syndrome outpatient clinic over a period of up to 7.6 years.

*Method:* 49 Turner Syndrome patients (median age at baseline 9.7 ±5.9 years, range 0–19.8) were followed-up for on average 2.9±1.1 examinations and a median time of 3.4±1.6 years. Aortic root (AoR) diameters and corresponding Z-scores were determined echocardiographically, and elasticity parameters as well as annual progression rates were calculated.

*Results:* At baseline, 16.3% of patients showed z-scores >2SD at one or more levels of the AoR (35.7% of patients with BAV, corresponding to an Odds Ratio of 4.2) and there was net progression to be noted at all measuring levels, leading to 28.6% of patients (50% of patients with BAV) exhibiting aortic dilatation at the end of follow-up. Progression correlated with the presence of BAV, non-mosaic monosomy and age. A levelling-off of progression was seen with the onset of adolescence.

*Conclusions:* Marked progression of aortic diameters leading to the development of dilatation can be observed in children and stresses

the importance of close surveillance during childhood. Main risk factors are BAV and complete monosomy 45X0. A beneficial influence of estrogen substitution can be suspected but needs to be investigated further.

**Keywords:** aortopathy, genetic vasculopathy, Turner Syndrome, aortic dilatation, bicuspid aortic valve

#### P-427

##### **Genetic studies on congenital heart disease for two decades in India: an overview**

Ramachandra Basappa Nallur

Department of Studies in Genetics and genomics, University of Mysore, Manasagangotri

*Background and Aim:* With the advent of technology, unravelling the genetic factors causing congenital heart disease (CHD) is fascinating. This was supported by the prevalence of different types of CHD cases globally. In view of this, in my laboratory, we conducted systematic genetic studies on CHD over two decades.

*Method:* For this study, we employed pedigree analysis, prevalence analysis, chromosomal anomaly studies, consanguinity studies, candidate gene studies, and whole exome sequence analysis.

*Results:* Some of the novel findings are as follows: (a) Majority of the studies revealed the autosomal recessive patterns of inheritance. (b) A ventricular septal defect is the most common CHD in India. (c) The uncle-niece marriage and first-cousin marriages are the prominent causes of CHD. (d) Chromosomes 18 and 9 are involved in CHD cases. (e) The candidate genes, GATA4 and NKX2.5, showed nonsynonymous variations in VSD cases. (f) The exome sequence analysis revealed the high-risk genes, namely, NOTCH1, NCOR1, HEY1, HEY2 and others.

*Conclusions:* These studies indicate that consanguineous marriages increase the prevalence of CHD in India and help us to educate families and prevent the birth of children with abnormal cardiac development. These studies will also guide us to create a gene panel for better diagnosis for possible treatments.

**Keywords:** Congenital heart disease, Genetics

#### P-428

##### **Maternal medication during pregnancy and cardiac abnormalities in the offspring**

Giaan Ninh<sup>1</sup>, R. Ottilia B. Vøgg<sup>1</sup>, Sofie Dannesbo<sup>1</sup>, Magdalena Hansson<sup>1</sup>, Line Cordua Stormly<sup>1</sup>, Anna Axelsson Raja<sup>2</sup>, Anne Sophie Sillesen<sup>1</sup>, Heather Boyd<sup>3</sup>, Kasper Iversen<sup>1</sup>, Henning Bundgaard<sup>2</sup>  
<sup>1</sup>Department of Cardiology, Copenhagen University Hospital Herlev, Herlev, Denmark; <sup>2</sup>Department of Cardiology, The Heart Centre, Copenhagen University Hospital Rigshospitalet, Copenhagen, Denmark; <sup>3</sup>Department of Epidemiology Research, Statens Serum Institut, Copenhagen, Denmark

*Background and Aim:* More than half of pregnant women use prescription drugs. Some of these drugs might expose the fetus to teratogenic effects including risk of developing congenital heart diseases (CHDs). In this study we will assess characteristics and prevalence of maternal medication use during pregnancy in mothers of newborns included in the Copenhagen Baby Heart Study (CBHS). We will then investigate whether newborns born to mothers with use of prescription drug during pregnancy differs in electro- and echocardiographic parameters at birth compared to non-exposed newborns. Furthermore, we will assess whether it is possible to detect drug metabolites using umbilical cord blood samples at birth.

**Method:** CBHS is a prospective, multi-center population study of newborns, born at the three main maternity wards in Copenhagen, between April 2016–October 2018. During this period more than 25, 000 new-borns had a cardiac examination consisting of echocardiography and electrocardiography performed within 60 days of life. At birth, an umbilical cord blood sample was drawn for biobank storage. All newborns in CBHS born to mothers with prescription drug use during pregnancy and the periconceptional period will be included. Exposure will be investigated as both groups of prescription drugs as well as individual prescription drugs. Echocardiographic parameters of interests include structural defects, measured structural and functional parameters of the ventricles, great arteries, and valves. Electrocardiographic parameters of interests include rhythm, rate, conduction, intervals, amplitudes, and waveform descriptions.

**Results:** Preliminary results are expected to be ready by spring 2023.

**Conclusions:** A greater insight into the etiology, including identification of modifiable factors, is essential to the implementation of preventative strategies and earlier detection of CHDs. Using the comprehensive data collected in the CBHS we will be able to further address any potential association between maternal medication use and cardiac structure and function in the offspring, both neonatally and later in life through longitudinal follow-up. This substudy is expected to generate prospective, new knowledge that may impact both clinical guidelines in maternal drug use during pregnancy.

**Keywords:** Maternal medication, congenital heart disease, echocardiography, electrocardiography, etiology, epidemiology

#### P-429

##### **Pharmacological reduction of oxidative stress counteracts cardiac dysfunction in a murine model of dystrophic cardiomyopathy**

Irene Cattapan<sup>1</sup>, Elena Cuppini<sup>2</sup>, Jolanda Sabatino<sup>2</sup>, Angela Di Candia<sup>2</sup>, Ruth Jephchirchir Arusei<sup>3</sup>, Marco Brugnaro<sup>4</sup>, Fabio Di Lisa<sup>4</sup>, Nina Kaludercic<sup>5</sup>, Giovanni Di Salvo<sup>2</sup>

<sup>1</sup>Pediatric Cardiology, Department of Woman's and Child's Health, University of Padua, Italy; <sup>2</sup>PhD School on Developmental Medicine and Health Planning Sciences, University of Padua, Padua, Italy; <sup>3</sup>Institute for Pediatric Research Città della Speranza, Padua, Italy; <sup>4</sup>Pediatric Cardiology, Department of Woman's and Child's Health, University of Padua, Italy; <sup>5</sup>Institute for Pediatric Research Città della Speranza, Padua, Italy; <sup>6</sup>Institute for Pediatric Research Città della Speranza, Padua, Italy; <sup>7</sup>Department of Woman's and Child's Health, University of Padua, Italy; <sup>8</sup>Department of Biomedical Sciences, University of Padua, Italy; <sup>9</sup>Department of Biomedical Sciences, University of Padua, Italy; <sup>10</sup>Institute for Pediatric Research Città della Speranza, Padua, Italy; <sup>11</sup>Department of Biomedical Sciences, University of Padua, Italy; <sup>12</sup>Neuroscience Institute, National Research Council of Italy (CNR), Padua, Italy

**Background and Aim:** Dystrophic cardiomyopathy culminates in heart failure and arrhythmias and is a major burden for Duchenne muscular dystrophy (DMD) patients. Mechanism-driven therapies designed to contrast the development of muscle and cardiac dysfunction are still missing. Antioxidant treatments counteract myocyte injury in mdx mice, a genetic model of DMD, supporting the possible role of enhanced reactive oxygen species (ROS) in the pathophysiology of muscular dystrophies. Previous evidence shows that monoamine oxidases (MAO) represent an important source of ROS, thus contributing to cardiomyocyte damage and dysfunction in different models of heart disease. Therefore, the aim of the study was to test whether

MAO-induced ROS formation contributes to the progression of pathology in dystrophic hearts.

**Method:** Wild type (WT) and mdx mice (a well known model of DMD) were studied at 3 and 12 months of life. Cardiac function was determined measuring fractional shortening, ejection fraction and left ventricular strain. Cardiac structure and fibrosis amount were assessed through histology (H&E, Masson's Trichrome). To test whether reduction in ROS burden could ameliorate cardiac structure and function in mdx mice, Safinamide, an inhibitor of MAO-B, was administered to WT and mdx mice respectively at 3 and 12 months of age.

**Results:** Fractional shortening (FS) and ejection fraction (EF) were significantly reduced in mdx mice at both 3 and 12 months of age, if compared to the wild type counterpart. In addition, left ventricular strain was already impaired in mdx mice at 3 months of age. This functional impairment was accompanied by a 5-fold increase in fibrosis in mdx hearts, evident already at 3 months. Safinamide administration for 30 days led to a significant reduction of levels of fibrosis among 3-month-old mdx mice and partial recovery of function among 12 months old mdx mice with significant improvement of circumferential strain and a trend toward increasing ejection fraction and fractional shortening.

**Conclusions:** These results suggest that pharmacological MAO-B inhibition improves cardiac function in a genetic model of DMD and may represent a clinically relevant target for the treatment of dystrophic cardiomyopathies.

**Keywords:** Duchenne Dystrophy, dystrophic cardiomyopathy, myocardial non-compaction, safinamide

#### P-430

##### **Personalized stenting for the treatment of aortic coarctation in paediatric patients: design, computational evaluation and 3D printing**

Robert Daniel Johnston<sup>1</sup>, Ciara Mckenna<sup>2</sup>, Samuel Geraghty<sup>1</sup>, Niall Linnane<sup>4</sup>, Damien Kenny<sup>3</sup>, Caitriona Lally<sup>1</sup>

<sup>1</sup>Trinity Centre for Biomedical Engineering, Trinity Biomedical Sciences Institute and Department of Mechanical, Manufacturing & Biomedical Engineering, School of Engineering and Advanced Materials and Bioengineering Research Centre (AMBER), Royal College of Surgeons in Ireland and Trinity College Dublin, Dublin, Ireland; <sup>2</sup>Trinity Centre for Biomedical Engineering, Trinity Biomedical Sciences Institute, and Department of Mechanical, Manufacturing & Biomedical Engineering, School of Engineering, Trinity College Dublin, Dublin, Ireland; <sup>3</sup>Our Lady's Children's Hospital, Department of Cardiology, Children's Health Ireland at Crumlin, Dublin 12 and Royal College of Surgeons, St Stephen's Green, Dublin, Ireland; <sup>4</sup>Trinity Centre for Biomedical Engineering, Trinity Biomedical Sciences Institute and Department of Mechanical, Manufacturing & Biomedical Engineering, School of Engineering, Trinity College Dublin, Dublin, Ireland and Royal College of Surgeons, St Stephen's Green, Dublin, Ireland

**Background and Aim:** Coarctation of the aorta (CoA) is a congenital disease that is characterized by a narrowing in the aorta [1]. Endovascular stenting to restore lumen gain at the location of the coarctation is a common treatment method, however, due to limited paediatric stents, adult stents intended for other applications are commonly used off-label. The use of stents not designed specifically for paediatric aortic tissue may lead to suboptimal treatment for patients and contribute to long-term hypertension [2]. This study demonstrates the feasibility of using finite element analysis (FEA) to perform a patient specific computational simulation of a stenting procedure to investigate and optimize stent design, material and location to improve clinical outcomes. Furthermore, we show that optimised patient specific stent designs

can be achieved using 3D printing alongside a robust post processing protocol to reduce strut size sufficiently to be in line to those of commercially available stents.

**Method:** Patient specific finite element models were reconstructed from clinically acquired CT images pre-stenting. The stenting procedure was simulated in four steps, as shown by Figure 1A, to mimic the clinical procedure.

**Results:** Stent design, material and location influence lumen gain, stresses on the vessel wall and stresses in the stent, see Figure 1B. Finally, novel stent designs produced using 3D printing and a robust post-processing protocol [3] demonstrate the feasibility of producing bespoke stent designs for pediatric patients to treat aortic coarctation, see Figure 1C.

**Conclusions:** The work presented in this study demonstrates how simulating the aortic coarctation stenting procedure in a finite element framework can (1) assist clinical decisions and treatment planning and (2) inform the development of patient specific stent designs to improve clinical outcomes.

#### References

- [1] Rao, P. S. (2005). Coarctation of the Aorta. *Current Cardiology Reports*, 7, 425–434.
- [2] Morgan, G. J., et al. (2013). Systemic blood pressure after stent management for arch coarctation implications for clinical care. *JACC. Cardiovascular Interventions*, 6(2), 192–201. <https://doi.org/10.1016/j.jcin.2012.10.009>
- [3] McGee, O. M., et al. (2022). An investigation into patient-specific 3D printed titanium stents and the use of etching to overcome Selective Laser Melting design constraints. *Journal of the Mechanical Behavior of Biomedical Materials*, 134. <https://doi.org/10.1016/j.jmbbm.2022.105388>

**Keywords:** Aortic coarctation, Patient specific, Computational modelling, 3D printing

#### P-431

##### **Sudden cardiac death in rasopathy-associated hypertrophic cardiomyopathy: validation of HCM risk-kids model and predictors of event**

Olga Dimitra Boleti<sup>1</sup>, Sotirios Roussos<sup>2</sup>, Gabrielle Norrish<sup>1</sup>, Ella Field<sup>1</sup>, Gauri Nepali<sup>3</sup>, Vinay Bhole<sup>3</sup>, Orhan Uzun<sup>4</sup>, Piers Edward Francis Daubeney<sup>5</sup>, Graham A Stuart<sup>6</sup>, Precyia Fernandes<sup>7</sup>, Karen McLeod<sup>7</sup>, Maria Iliina<sup>7</sup>, Muhammad Najih Ali Liaqath<sup>8</sup>, Tara Bharucha<sup>8</sup>, Grazia Delle Donne<sup>9</sup>, Katie Linter<sup>10</sup>, Bernadette Khodaghalian<sup>11</sup>, Jonathan Searle<sup>12</sup>, Sujeev Mathur<sup>13</sup>, Nicola Boyd<sup>14</sup>, Terence Prendiville<sup>15</sup>, Satish Adwani<sup>12</sup>, Martin Zenker<sup>17</sup>, Cordula Wolf<sup>16</sup>, Juan Pablo Kaski<sup>1</sup>

<sup>1</sup>Institute of Cardiovascular Sciences, University College London, London, United Kingdom; <sup>2</sup>Department of Hygiene, Epidemiology and Medical Statistics, Medical School, National and Kapodistrian University of Athens, Athens, Greece; <sup>3</sup>The Heart Unit, Birmingham Children's Hospital, Birmingham, United Kingdom; <sup>4</sup>Children's Heart Unit, University Hospital of Wales, Cardiff, United Kingdom; <sup>5</sup>Department of Paediatric Cardiology, Royal Brompton and Harefield NHS Trust, London, United Kingdom; <sup>6</sup>Department of Paediatric Cardiology, Bristol Royal Hospital for Children, Bristol, United Kingdom; <sup>7</sup>Department of Paediatric Cardiology, Royal Hospital for Children, Glasgow, United Kingdom; <sup>8</sup>Department of Paediatric Cardiology, Southampton General Hospital, Southampton, United Kingdom; <sup>9</sup>Department of Paediatric Cardiology, Leeds General Infirmary, Leeds, United Kingdom; <sup>10</sup>Department of Paediatric Cardiology, Glenfield Hospital, Leicester, United Kingdom; <sup>11</sup>Department of Paediatric Cardiology, Alder Hey Children's Hospital, Liverpool, United Kingdom; <sup>12</sup>Department of Paediatric Cardiology, John Radcliffe Hospital, Oxford, United Kingdom; <sup>13</sup>Children's Heart Service, Evelina Children's Hospital,

London, United Kingdom; <sup>14</sup>Department of Paediatric Cardiology, The Freeman Hospital, Newcastle, United Kingdom; <sup>15</sup>The Children's Heart Centre, Our Lady's Children's Hospital, Dublin, Republic of Ireland; <sup>16</sup>Department of Pediatric Cardiology and Congenital Heart Disease, German Heart Center Munich, Technical University Munich, Munich, Germany; <sup>17</sup>Institute of Human Genetics, University Hospital, Magdeburg, Germany

**Background and Aim:** RASopathies represent an important proportion of hypertrophic cardiomyopathy (HCM) presenting in childhood, with a significant risk of suffering sudden cardiac death (SCD). Despite this, there are no published risk factors for SCD in this cohort of patients. Our primary aim was to validate HCM Risk-Kids, a SCD prediction model published and validated in 2019, for children with RASopathy-associated HCM, with a secondary aim to investigate for independent predictors of SCD in this group.

**Method:** One-hundred-and-sixty-nine (185) patients from 15 international paediatric cardiology centres (UK, Ireland and Germany)  $\leq 18$  years with a RASopathy syndrome and HCM were identified. Patients with more than 50% missing data in predictor variables used in the initial model were excluded, leaving a total of 169 patients for analysis. Multiple imputation by chained equations was used to impute missing values of baseline variables and clinical parameters.

**Results:** Eleven (6.5%) patients suffered a SCD equivalent event (3 (27.3%) suffered SCD, 5 (45.5%) an aborted cardiac arrest, 1 (9.1%) had an appropriate ICD shock and 2 (18.2%) had sustained VT) at a median age of 12.5 months (IQR 7.7–28.64). The calculated SCD equivalent event incidence per 100 patient years was 0.78 (95% CI 0.43 – 1.41). Six patients (54.55%) who had a SCD equivalent event were in the low-risk category according to the HCM Risk-Kids model. Harrell's C index was 0.5959 (95% CI 0.26–1.22) with a sensitivity of 9.09%, specificity of 63.92%, positive predictive value of 1.72%, and negative predictive value of 90.99% as well as poor distinction between the different risk groups. Unexplained syncope (HR 102.63 (95% CI 14.94 – 704.99),  $p < 0.001$ ) and NSVT (HR 5.67 (95% CI 1.30 – 24.70),  $p = 0.021$ ) were shown to be independent predictors of SCD.

**Conclusions:** HCM Risk-Kids model should not be used for patients with RASopathy-associated HCM but further, larger centre collaborative studies should be organised to investigate predictors of SCD in this group of patients.

**Keywords:** sudden cardiac death, hypertrophic cardiomyopathy, RASopathies, paediatrics, risk

#### P-432

##### **Critical insights from the mechanical properties and histology of neonatal aortic coarctation tissue to inform bespoke patient therapies**

Niall Linnane<sup>1</sup>, Robert Johnston<sup>2</sup>, Damien P Kenny<sup>3</sup>, Cairiona Lally<sup>2</sup>  
<sup>1</sup>Trinity Centre for Biomedical Engineering & School of Engineering, Trinity College Dublin. Royal College of Surgeons, St Stephen's Green, Dublin 2. Children's Health Ireland at Crumlin, Dublin 12; <sup>2</sup>Trinity Centre for Biomedical Engineering & School of Engineering, Trinity College Dublin. Advanced Materials and Bioengineering Research Centre (AMBER), Royal College of Surgeons in Ireland and Trinity College Dublin, Dublin, Ireland.; <sup>3</sup>Royal College of Surgeons, St Stephen's Green, Dublin 2. Children's Health Ireland at Crumlin, Dublin 12

**Background and Aim:** Coarctation of the aorta has an incidence of 0.3 to 0.6 in 1000 live births, accounting for 5–12% of all congenital heart disease [1] Ho et al demonstrated that ductal tissue is a

cause of coarctation and is histologically different to surrounding aorta [2]. The aim of this study is to ascertain the mechanical properties of harvested neonatal aortic coarctation specimens and to investigate the relationship with its histological structure as a precursor to understanding the potential tissue impact of neonatal stenting.

**Method:** Five specimens of neonatal aortic coarctation tissue were harvested during surgery from Children's Health Ireland. The vessels were cut into ring samples and uniaxial tensile testing was performed to predetermined stretch levels before being fixed for histological analysis.

**Results:** Five sections were harvested of which three (samples 3–5) were further subdivided into two test specimens. Samples were labelled "A, B and C" with "B" being at the ductal-insertion point and "A" and "C" being proximal and distal, respectively.

Figure 1 shows the force–diameter change behaviour of the samples and highlights the inter- and intra-specimen variability. Specimens appear to have quite different mechanical properties depending on their relative location with respect to the ductus arteriosus insertion point (i.e. proximal or distal). Preliminary histological assessment of the ductus arteriosus tissue demonstrates the different histological structure of the ductal tissue when compared to the aortic wall tissue (Figure 2). The ductal tissue has very little elastin or collagen thus inferring that its load bearing role is likely limited when undergoing stretch.

**Conclusions:** Neonatal coarcted aortic tissue has variable mechanical properties and the amount of ductal tissue which invades into the healthy aortic tissue likely plays a significant role in this variability and needs to be accounted for in any bespoke device-based patient treatments.

#### References:

- Mai, C.T., et al., National population-based estimates for major birth defects, 2010–2014. *Birth Defects Res*, 2019.
- Ho, S.Y. and R.H. Anderson, Coarctation, tubular hypoplasia, and the ductus arteriosus. *Histological study of 35 specimens. Heart*, 1979. 41(3): p. 268–274.

**Keywords:** Coarctation of the aorta, Histology, Bioengineering, Interventional Cardiology

Mechanical testing and histology images

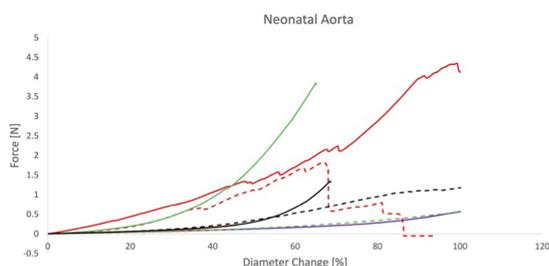


Figure 1: Force-Diameter change (%) curves from uniaxial testing of neonatal coarcted tissue



Figure 2: Sample 3B. (a) Gross, (b) H&E (cell content), (c) Picrosirius red (collagen), (d) Verhoeff's (elastin).

#### P-433

##### Doxorubicin-induced muscle myopathy involves inflammation mediated cellular mechanisms in diabetes

Dinender K Singla

Burnett School of Biomedical Sciences, University of Central Florida, Orlando Florida

**Background and Aim:** Patients diagnosed with cancer have pre-existing diabetes which results in higher rate of muscle weakness and cardiac dysfunction. A significant increase in hospitalization in diabetic patients following cancer treatment is due to lack in understanding on the pathophysiological mechanisms. Post cancer treatments develop major side effects such as skeletal muscle myopathy and cardiomyopathy in post Doxorubicin treatment. is the major Aim of this study to investigate. Therefore, we designed a study to examine pathophysiological mechanisms of doxorubicin-induced muscle and cardiomyopathy in diabetic animals.

**Method:** In the present study, we developed a DOX-induced diabetic animal model. We divided C57BL/6J mice into four groups; control, diabetic (streptozotocin; STZ), DOX, and DOX+STZ. Animals were sacrificed and heart function were evaluated. Markers for apoptosis (Caspse-3, Bax, and Bcl2) and various inflammatory cytokines (IL-6, TNF- $\alpha$ ).

**Results:** We demonstrated significant increase in Caspse-3, Bax, and Bcl2 and apoptosis with TUNEL staining. Increased apoptosis increased infiltration of monocytes that initiated inflammation and further worsening the tissue damage. Next, our RNA sequencing data shows infiltrated inflammatory cells (Dendritic cells, neutrophils, monocytes, and macrophages) which further confirmed with immunohistochemistry. We also examined cellular interactions and their role in DOX-induced cardiomyopathy. Our data shows significant increase in cell death (non-inflammatory-apoptosis and inflammatory-necroptosis). This set of data was further confirmed with various molecular biology, histology, and RNA sequencing gene analysis. Further, we determined presence of inflammatory cells have interactions with cardiac and muscle cells which causes inflammation mediated fibrosis, adverse cardiac remodeling and decreased cardiac function.

**Conclusions:** Overall, we present data which shows complexity in cellular mechanisms as well as role of inflammation in developed cardiac and muscle myopathy following Dox treatment in diabetic animals.

**Keywords:** Doxorubicin, cardiomyopathy, inflammation, diabetes, heart

#### P-434

##### Exploring the proteomic characteristics of aortic isthmus stenosis by mass spectrometry

Martin Misailovski<sup>1</sup>, Christof Lenz<sup>2</sup>, Theodor Tirilomis<sup>3</sup>, Christian Dullin<sup>4</sup>, Matthias Sigler<sup>1</sup>, Thomas Paul<sup>1</sup>, Katja Eildermann<sup>1</sup>

<sup>1</sup>Pediatric Cardiology and Intensive Care Medicine, Georg-August-University Göttingen, Germany; <sup>2</sup>Bioanalytical Mass Spectrometry, Max Planck Institute for Multidisciplinary Sciences, Göttingen, Germany; <sup>3</sup>Pediatric Heart Surgery, Dept. of Cardiac, Thoracic, and Vascular Surgery, University Medical Center Göttingen, Germany; <sup>4</sup>Institute for Diagnostic and Interventional Radiology, University Medical Center Göttingen, Germany

**Background and Aim:** Coarctation of the aorta (CoA) is a common congenital heart defect in newborns. Given the spatial proximity of the aortic narrowing to the ductus arteriosus (DA), a relation between the two structures is plausible. However, the exact pathogenesis is still under debate. Aim of the study was to obtain the

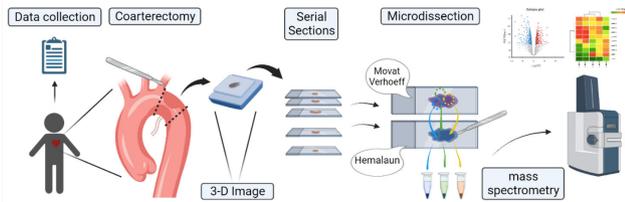
proteomic footprint of I) unchanged aorta, II) the stenotic part of the aorta, and III) the closing ductus arteriosus to gain insight into disease mechanism.

**Method:** Material from nine patients who underwent CoA-resection with end-to-end anastomosis was fixed in formalin and embedded in paraffin. High resolution 3-dimensional imaging was achieved in the Elettra Synchrotron in Trieste (Italy) to obtain a general overview of the tissue and to guide the selection of the regions of interest (ROI). Specimens were serially sectioned and Movat Verhoeff (MV) staining was applied every 200  $\mu\text{m}$ . Material from ROIs was manually dissected from deparaffinized, Hemalaun stained sections using the closest consecutive MV-staining as guide. Proteins were isolated using pressure-cycling technology and analyzed by data-independent acquisition mass spectrometry.

**Results:** 3926 proteins were consistently detected and quantified across all samples with a False Discovery Rate (FDR) of <1%, and quantitatively compared between groups. The analysis revealed an enrichment of contractile proteins in the DA and enriched extracellular matrix components and proteins related to secretory activity in changed aorta.

**Conclusions:** While histologically, ductal tissue and changed aorta present very similar, their proteomic footprint shows distinct differences. However, the impact of these differences remains to be further elucidated.

**Keywords:** Mass spectrometry, CoA, Isthmus stenosis, congenital heart defect, proteome



Workflow. Created with Biorender

#### P-435

##### Optimization of support materials for the cultivation of engineered heart tissue (EHT) using 3D printing technology

Daniel Biermann<sup>1</sup>, Luca Josefina Jochheim<sup>2</sup>, Maria Köhne<sup>2</sup>, Thomas Eschenhagen<sup>2</sup>, Jörg Siegmund Sachweh<sup>1</sup>, Florian Weinberger<sup>2</sup>, Michael Hübler<sup>1</sup>

<sup>1</sup>Congenital & Pediatric Heart Surgery, Children's Heart Clinic, University Heart & Vascular Center Hamburg, University Medical Center Hamburg-Eppendorf, Germany; <sup>2</sup>Department of Experimental Pharmacology and Toxicology, University Medical Center Hamburg-Eppendorf, German

**Background and Aim:** The ability to replace malformed or revitalize damaged cardiac structures with vital, contractile engineered heart tissue (EHT) could revolutionize treatment options. One major limitation is that most support materials for culturing larger EHTs (> 1x10<sup>6</sup> cardiomyocytes (CMs)) have to be manufactured with extensive manual labor, which makes their production very time-consuming and leaves little flexibility for optimization. We intend to address these limitations by using 3D printing technology to fabricate components used for the EHT cultivation system in a standardized and simplified manner.

**Method:** In a previous project of our group a cultivation system for tubular EHTs (inner diameter (ID): 6 mm, length (L): 18 mm, wall thickness (WT): ~ 1 mm; 18x10<sup>6</sup> CMs / ml) was created. To standardize and optimize this system, we initially developed several computational designs for some of its parts.

**Results:** As a prototype, the cultivation chamber for tubular EHTs was printed from polylactic acid (PLA), a biocompatible and biodegradable plastic. Our current approach is to print constructs from water-soluble polyvinyl alcohol (PVA). These formats should then serve as casting molds for Polydimethylsiloxane (PDMS), which is widely used for EHT culture. With this technique, even complex and fine structures can be produced. As an example, the production of the silicone perfusion tube (WT: 200  $\mu\text{m}$ ) that served as support for the EHT during cultivation was standardized and optimized using this technique. We developed a perfusion tube model that is radially equipped with flexible silicone columns (L: 2 mm, D: 1 mm). After casting, this results in a tubular net shape. Further, the flexible columns might produce a higher mechanical load on the tissue during culture. These facts could promote enhanced uniform cell distribution and maturation.

**Conclusions:** The 3D printing technology offers a variety of opportunities to simplify and optimize the production of culture materials for EHTs. The new setting needs to be evaluated during and after EHT cultivation in the next steps. This will include the cultivation of tubular EHTs in the optimized setting and subsequent histological and functional analysis.

**Keywords:** Tissue Engineering, Engineered Heart Tissue, 3D Printing, Univentricular Heart

#### P-436

##### Acute myocarditis in children in the SARS-COV2 pandemic area

Cloe Huet<sup>1</sup>, Fanny Deville<sup>1</sup>, Diane Ouwrier Buffet<sup>1</sup>, Olivier Metton<sup>1</sup>, Julia Mitchell<sup>1</sup>, Sylvie Di Filippo<sup>2</sup>

<sup>1</sup>Department of Congenital Heart Disease, Claude Bernard University, Lyon, France; <sup>2</sup>Congenital Cardiology, Cardiothoracic Center of Monaco

**Background and Aim:** The objective of the study was to assess the features of acute myocarditis in the area of SARS-Cov2 pandemic. **Method:** Patients < 18 years of age with acute myocarditis (proved by virology and/or MRI and/ or complete recovery of myocardial function) were included. Clinical data, echocardiographic parameters and outcomes were collected.

**Results:** From 1983 to 2021, 140 patients were included: 68 males and 72 females. Mean age at diagnosis was 6.8 years. Heart failure (HF) was present at onset in 65% of cases. Mean left ventricular shortening fraction (LVSF) at diagnosis was 24%. Mitral regurgitation was present in 64% of cases, pericarditis in 16%. Thromboembolic events occurred in 7% and arrhythmias in 10%. Virus was retrieved in 66% of the cases. Inotropic support was needed in 47%, mechanical circulatory support in 8%. Death occurred in 11 patients (7.8%). Complete recovery occurred in 74% of all cases. Sixty-four cases were due to SARS-Cov2 (45.7%). Male gender was more frequent (57% vs 40%), patients were older (9.9 vs 4.2 years), severe HF less frequent, chest pain more frequent, mean left ventricular shortening fraction higher (31.6% vs 18.4%) in SARS-Cov2 cases. Mean time to recovery was 2 weeks to 2 years. Mean LVSF improved from 18% at onset, to 25% at 1st month, 26.5% at 3rd month, 31% at 6th month and 38% at last follow-up in the non-Covid cases.

**Conclusions:** Myocardial dysfunction and heart failure were less frequent in SARS-Cov2 cases, while myocardial improvement progressed slowly over time in non-COVID cases.

**Keywords:** acute myocarditis, children, SARS-Cov2, outcomes

#### P-437

##### **Rare genetic background in familial hypertrophic cardiomyopathy – KLHL24 as a disease gene for HCM**

Franziska Seidel<sup>1</sup>, Jirko Kühnisch<sup>2</sup>, Friederike Danne<sup>1</sup>, Annemarie Krauß<sup>1</sup>, Nele Rolfs<sup>1</sup>, Bernd Oppen Rhein<sup>3</sup>, Josephine Dartsch<sup>2</sup>, Thomas Pickardt<sup>5</sup>, Oliver Miera<sup>1</sup>, Karin Klingel<sup>4</sup>, Felix Berger<sup>1</sup>, Sabine Klaassen<sup>2</sup>

<sup>1</sup>German Heart Center Berlin, Department of Congenital Heart Disease and Pediatric Cardiology, Berlin, Germany; <sup>2</sup>Experimental and Clinical Research Center, a cooperation between the Max-Delbrück-Center for Molecular Medicine in the Helmholtz Association and the Charité-Universitätsmedizin Berlin, Germany; <sup>3</sup>Charité-Universitätsmedizin Berlin, Department of Pediatric Cardiology, Berlin, Germany; <sup>4</sup>Cardiopathology, Institute for Pathology and Neuropathology, University Hospital Tübingen, Tübingen, Germany; <sup>5</sup>Competence Network for Congenital Heart Defects, Berlin, Germany

**Background and Aim:** Hypertrophic cardiomyopathy (HCM) may cause heart failure and sudden cardiac death. Assessing its genetic cause might be important for diagnosis and further therapy. Generally, in 60% pathogenic variants are found when testing children genetically, the remaining cases remain unclear. We present a case report of a consanguine family with rare genetic background. **Method:** We performed in depth assessment of an 11-years-old girl with HCM, her consanguine parents, and an older sister. The family was analyzed with whole-exome sequencing (WES) and filtered for rare (minor allele frequency <10<sup>-4</sup>) likely pathogenic or pathogenic L(P) genetic variants. Endomyocardial biopsy (EMB) samples of the index patient were analyzed histologically and with immunostaining.

**Results:** The girl presented with HCM, left ventricular (LV) outflow obstruction, ventricular tachycardia, and severe heart failure with cardiac decompensation. The LV ejection fraction was 25%. NT-proBNP level and Troponin Ihs levels were elevated with 25940 ng/l and 240 pg/nl, respectively. Accordingly, she needed biventricular assist device implantation and heart transplantation. Her father was also affected and heart transplanted. WES of the family identified in the index patient three homozygous variants: 1) a L(P) missense variant in dihydropyrimidinase (DPYS, p.S379R) causing pyrimidine deficiency; 2) a L(P) non-sense variant in patatin like phospholipase domain containing 2 (PNPLA2, p.L205Pfs\*102) resulting in defective triglyceride metabolism, lipid storage disease, and myopathy; 3) a truncating variant in Kelch like family member 24 (KLHL24, p.T496Kfs\*5). Mutation of KLHL24 was recently linked to HCM with glycogen storage disease. The affected father also carried the homozygous variant in KLHL24 suggesting this genetic alteration as primary disease cause. EMB of the index patient showed diffuse, focal, and perivascular fibrosis. Myocardial cardiomyocytes were hypertrophic, irregularly structured, and showed enlarged nuclei. Very noticeable were masses of small vacuoles in the cytoplasm of the cardiomyocytes. In PAS staining there was increased intercalated glycogen.

**Conclusions:** This is a rare autosomal recessive familial HCM leading to characteristic histopathological changes in EMB with increased glycogen storage highlighting the importance of storage disease as potential cause of HCM. The genetic diagnosis in this previously unresolved case of cardiomyopathy supports the role of KLHL24 as a disease gene for HCM.

**Keywords:** hypertrophic cardiomyopathy, pathogenic variants, KLHL24

#### P-438

##### **One family, with two types of cardiomyopathies in first degree relatives and one sudden cardiac death – a familial genetic overlap?**

Gabriela Doros<sup>1</sup>, Anca Popoiu<sup>1</sup>, Ana Maria Miinea<sup>2</sup>, Cristiana Stolojanu<sup>1</sup>, Andreea Vartop<sup>2</sup>, Andrada Mara Micescu<sup>1</sup>

<sup>1</sup>Department of Pediatrics, “Victor Babes” University of Medicine and Pharmacy, Timisoara, Romania; Department of Pediatric Cardiology, “Louis Turcanu” Emergency Hospital for Children, Timisoara, Romania; <sup>2</sup>Department of Pediatric Cardiology, “Louis Turcanu” Emergency Hospital for Children, Timisoara, Romania

**Background and Aim:** To present a family with two different types of cardiomyopathies, one hypertrophic with sudden cardiac death and one noncompaction of the left ventricle (LVNC), in two first degree relatives, as a possible genetic overlap syndrome. LVNC is a primary genetic cardiomyopathy, associated with high risk of sudden cardiac death, heart failure, thromboembolisms and severe arrhythmias. It can coexist with dilated, hypertrophic, restrictive and arrhythmogenic cardiomyopathy.

**Method:** A 14 years old girl was referred by her mother, after her father’s sudden death. Postmortem, her father was diagnosed with hypertrophic cardiomyopathy, genetically tested and confirmed to be with heterozygous DSG2 gene mutation of uncertain significance which may be responsible for sudden cardiac death. We evaluated his daughter and his sister was referred to the adult cardiology department to be assessed. Both of them were genetically screened for the same mutation with father.

**Results:** The DSG2 gene mutation confirmed in the father was tested in his daughter and his sister, however both came back negative. Thorough cardiological examination was performed for both daughter and sister: clinical, ECG, Echocardiography and angio-MRI. Surprisingly, both were diagnosed with noncompaction cardiomyopathy. Father’s sister has both ventricles affected by noncompaction cardiomyopathy. The daughter is symptomatic, with effort intolerance. Echocardiography confirmed left ventricle noncompaction cardiomyopathy. Speckle tracking had regional hypokinesia in the noncompaction areas. Cardiac MRI confirmed the diagnosis. Holter ECG did not reveal ventricular arrhythmias. Because the ejection fraction of the daughter was borderline, at the lower limit, we started treatment with Lisinopril and low dose Aspirin. We retested the daughter for a larger panel of cardiomyopathy inducing genes. Both of them are included in a follow up program.

**Conclusions:** Coexistence of hypertrophic cardiomyopathy and Noncompaction of the left ventricle cardiomyopathy is a rare combination in the same family, but it has been cited in literature. Noncompaction cardiomyopathy of both ventricles is an extremely rare occurrence

**Keywords:** cardiomyopathy, genetic cardiomyopathy, non-compaction cardiomyopathy

#### P-439

##### **Endomyocardial biopsy in myocarditis identifies factors that predict outcome in children:**

Rachele Adorisio<sup>1</sup>, Gessica Ingrassiotta<sup>1</sup>, Aurora Ionata<sup>2</sup>, Nicoletta Cantarutti<sup>1</sup>, Elisa Bellettini<sup>1</sup>, Erica Mencarelli<sup>1</sup>, Maria Grandinetti<sup>1</sup>, Mara Pilati<sup>1</sup>, Richard Kirk<sup>1</sup>, Antonio Amodeo<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology and Cardiac Surgery, Bambino Gesù Children’s Hospital, IRCCS, Rome, Italy; <sup>2</sup>University of Rome “Tor Vergata”, Rome, Italy

**Background and Aim:** Pediatric myocarditis is a rare and challenging disease characterised by a broad spectrum of clinical presentations.

The aim of this study is to identify prognostic factors for death and/or cardiac transplantation or myocardial recovery in children with biopsy proven myocarditis and referred to Bambino Gesù Children's Hospital from 1988 to 2022.

**Method:** This is a single-center retrospective study. Inclusion criteria included age <18 years and a diagnosis of biopsy proven myocarditis.

**Results:** 113 patients (mean age  $5 \pm 5.2$  years; M 49) underwent endomyocardial biopsy (EMB), 38% showed a viral myocarditis. The most frequently detected was Parvovirus B19 (24%). Medication used included heart failure therapy (32%), inotropes (78%), immunosuppressive therapy (69%), antiviral therapy (6%) and, finally, mechanical circulatory support (9%) for pts with cardiogenic shock. 13% died or required cardiac transplantation. Multivariate analysis showed negative prognostic factors were: age <1 years ( $p = 0.024$ ) and LVEF <40% at onset ( $p = 0.011$ ) and the presence of virus at EMB ( $p = 0.037$ ). In addition, the presence of fibrosis already at first EMB ( $p = 0.023$ ) as well as the viral co-infection (defined as the presence of  $\geq 2$  virus) were significantly related to clinical outcome ( $p = 0.013$ ). In contrast, the absence of virus at EMB was significantly related with ventricular recovery ( $p = 0.019$ ).

**Conclusions:** This study identified endomyocardial biopsy parameters which help to predict outcome. The presence and number of viruses, and fibrosis at EMB in the acute phase of the disease are negative prognostic factors in children.

**Keywords:** Myocarditis, Endomyocardial Biopsy, Fibrosis

#### P-440

##### High dosage of propranolol prolongs survival in newborns with hypertrophic cardiomyopathy associated to Noonan syndrome and mitochondrial diseases

Rachele Adorisio<sup>1</sup>, Nicoletta Cantarutti<sup>1</sup>, Gessica Ingrassiotta<sup>1</sup>, Erica Mencarelli<sup>1</sup>, Elisa Bellettini<sup>1</sup>, Maria Grandinetti<sup>1</sup>, Fabrizio Drago<sup>1</sup>, Alessandra Toscano<sup>2</sup>, Cosimo Marco Campanale<sup>2</sup>, Marco Masci<sup>2</sup>, Diego Martinelli<sup>3</sup>, Andrea Dotta<sup>2</sup>, Antonio Amodeo<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology and Cardiac Surgery, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy; <sup>2</sup>Department of Medical and Surgical Neonatology, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy; <sup>3</sup>Division of Metabolism, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy

**Background and Aim:** Hypertrophic Cardiomyopathy (HCM) presenting during the neonatal period has different etiologies, risk factors, and clinical course comparing to HCM with onset during childhood, adolescence or adult age. Noonan and mitochondrial disease have the worst prognosis among all neonatal HCM.

**Method:** To describe the impact of early use of beta-blockers on survival (defined as freedom from cardiac death or heart transplantation) and clinical course of Noonan and mitochondrial HCM with neonatal onset during long term follow up (FU).

**Results:** In this single-center retrospective study, we reviewed 43 HCM with neonatal onset. From this group, we selected 17 neonates with Noonan syndrome (40%) and 9 with mitochondrial diseases (21%). At the time of diagnosis, early treatment with beta-blockers was started and up-titrated to highest tolerated dosage (propranolol mean dosage  $24.5 \pm 10$  mg/kg). Kaplan Meier analysis showed a survival at 6 months of 95% in patients with Noonan syndrome and of 88% in patients with mitochondrial disorders. After the first 6 months of FU, surviving patients were stabilized and no other cases of death or heart transplantation were recorded during the next FU (median FU  $4.7 \pm 1.6$  yrs). During long-term FU, 4 Noonan patients (23%) underwent myectomy before 4 years of age and ICD was implanted in 2 patients (12%) during

adolescence. No cases of myectomy or ICD implantation were recorded in the mitochondrial group.

**Conclusions:** Noonan and mitochondrial disorders represent the great majority of neonatal HCM. Early treatment and high dosage of beta-blockers allow a good prognosis with a high rate of survival in these patients compared to data previously reported in literature.

**Keywords:** HCM, Neonatal Cardiomyopathy, Mitochondrial Disease, Beta-Blockers

#### P-441

##### Success rate of percutaneous balloon dilatation as first treatment option in children with pulmonary stenosis associated with Noonan syndrome

Lukas Jozef E. Rubbens<sup>1</sup>, Laura Muiño Mosquera<sup>3</sup>, Joseph Panzer<sup>2</sup>, Daniel De Wolf<sup>2</sup>, Hans De Wilde<sup>2</sup>, Katya De Groot<sup>2</sup>, Ilse Meerschaut<sup>2</sup>, Wendy Dewals<sup>1</sup>, Luc Bruyndonckx<sup>1</sup>, Kristof Vandekerckhove<sup>2</sup>, Mahmoud Zaqout<sup>1</sup>

<sup>1</sup>Department of Pediatrics, division of pediatric cardiology, Antwerp University Hospital, Antwerp, Belgium; <sup>2</sup>Department of Pediatrics, division of pediatric cardiology, Ghent University Hospital, Ghent, Belgium; <sup>3</sup>Department of Pediatrics, division of pediatric cardiology, Ghent University Hospital, Ghent, Belgium & Center of Medical Genetics, Ghent University Hospital, Ghent, Belgium

**Background and Aim:** Noonan syndrome (NS) can be associated with a number of congenital heart defects (CHD) of which a (supra-)pulmonary valve stenosis ((SV)PS) is the most frequent. Possible treatment options are percutaneous balloon pulmonary valvuloplasty (BVP) or surgical intervention. However, anatomical location of the PS may help predict BVP failure.

We aimed to evaluate the outcome of treatment with BVP of PS in children with Noonan syndrome.

**Method:** All medical records of children with a clinical diagnosis of Noonan syndrome and in follow-up at the Antwerp- and the Ghent University Hospitals were retrospectively reviewed. The study was approved by the medical ethical committee of both institutions (EC2022/0141).

**Results:** 50 children (median age 6 months, IQR 1.7-54, 54% female) were included in the study (28 PTPN11, 8 SOS1, 5 RIT1, 1 LZTR1, 1 RAF1, 1 KRAS, 1 BRAF, 1 SHOC2, 4 unknown). Of these children 39 (78%) had a congenital heart disease (CHD) of which 32 (64%) a (SV)PS, either isolated or in combination with other CHD. 69% of the children with PS had a SVPS. The prevalence of PS and SVPS was similar for all genes. A surgical or percutaneous intervention was necessary in 17/32 patients with PS (53%). Except for 2 children with pulmonary valve stenosis, all had SVPS. Only 2 of these 17 children had a surgical repair as first option. The remaining 15 (13 SVPS) underwent a percutaneous balloon dilatation. 10 of these 15 patients (66%) needed a second balloon dilatation, all of them ultimately converted to surgical repair due to persistent stenosis. Except for one patient, all had SVPS. Median time to reintervention was 1 month. The global success rate of percutaneous intervention in children with Noonan and SVPS was (30, 7%).

**Conclusions:** (SV)PS is the most frequent CHD in children with NS. In our cohort, the prevalence of (SV)PS was similar for all genes. The success rate of BVP in patients with NS is low. The most determinant factor of treatment failure seems to be the presence of SVPS. However, BVP might still be useful in selected cases and might be considered to clarify the anatomical location of PS.

**Keywords:** Noonan syndrome, (supra) pulmonary valve stenosis, balloon pulmonary valvuloplasty

**P-442****Seckel syndrome: A case report with multiple heart defect**

*Evmorfia Ladopoulou<sup>1</sup>, Aikaterini Boli<sup>1</sup>, Nikias Milaras<sup>1</sup>, Labrini Pouliou<sup>1</sup>, Alkistis Kalesis<sup>2</sup>, Maria Karakosta<sup>2</sup>, Antonios Destounis<sup>2</sup>, Nearchos Kasinos<sup>2</sup>, Anastasios Theodosis Georgilas<sup>2</sup>, Niki Lama<sup>3</sup>, Efthimios Brachos<sup>1</sup>, Nikolaos Tziolas<sup>1</sup>, Konstantinos Tsatiris<sup>1</sup>, Sotiria Apostolopoulou<sup>4</sup>, Panagiotis Zachos<sup>5</sup>*

<sup>1</sup>Department of Cardiology, General Hospital of Karditsa; <sup>2</sup>Department of Cardiology, Tzaneio General Hospital of Piraeus; <sup>3</sup>Research Unit of Radiology and Medical Imaging, 2nd Department of Radiology, Medical School of Athens, Greece; <sup>4</sup>Pediatric Cardiology and Adult Congenital Heart Disease Department, Onassis Cardiac Surgery Center; <sup>5</sup>Pediatric Cardiology and Adult Congenital Heart Disease Department, General Hospital of Karditsa

**Background and Aim:** Seckel syndrome is a rare genetic condition with autosomal recessive inheritance affecting 1/10000 that equally affects men and women. Patients present with dwarfism, microcephaly with special facial appearance and mental retardation. The presence of heart defects is unusual.

**Method:** We report a case of Seckel Syndrome and cardiac anomalies who visit our Pediatric Cardiology and Adult Congenital Heart Disease Department for regular cardiac screening.

**Results:** The case concerns a girl 4 years old with confirmed Seckel Syndrome with genetic testing. It was born preterm with a diagnosis of intrauterine growth restriction (IUGR) and hospitalized to neonatal intensive care unit because of hemodynamic instability. The girl presents with dwarfism, microcephaly, micrognathia, small triangular beak-like nose, large eyes with strabismus, dental dysplasia, cleft palate and mental retardation. The echocardiogram unveils small atrial septal defect (ASD) with left-to-right shunt and a perimembranous ventricular septal defect (VSD) with a slightly overriding aorta. Dimensions and function of both ventricles are normal.

**Conclusions:** There are only a few cases with confirmed Seckel Syndrome and cardiac involvement. May present with complex congenital heart defects such as Tetralogy of Fallot and atresia of tricuspid valve. Some present with ASD, VSD or patent ductus arteriosus and few cases have present with complete heart block and severe sinus bradycardia. Our case might be the first case to our knowledge that describes secondary ASD and perimembranous VSD in association with Seckel Syndrome.

**Keywords:** Seckel syndrome, multiple heart defect, ASD, VSD

**P-443****Cardiac troponin I as cardiomyopathy biomarker in duchenne muscular dystrophy**

*Iulia Rodoman<sup>1</sup>, Lucia Pîrîtu<sup>2</sup>, Alexandr Dorif<sup>1</sup>, Victoria Sacară<sup>1</sup>, Ina Paliu<sup>1</sup>*

<sup>1</sup>PMHI Institute of Mother and Child, Chisinau, Republic of Moldova; <sup>2</sup>PI Nicolae Testemitanu State University of Medicine and Pharmacy, Chisinau, Republic of Moldova

**Background and Aim:** Duchenne muscular dystrophy (DMD) is the most common type of childhood muscular dystrophy, caused by an X-linked mutation in the DMD gene, resulting in loss of muscle membrane stability and progressive muscle necrosis and fibrosis inclusive in the heart. In the current era, cardiomyopathy is the leading cause of death in DMD. Elevated serum troponin levels (cTnI) are observed in DMD and may change with disease progression. In this study, we aimed to reveal the cTnI levels and their relation to echocardiography parameters.

**Method:** serum cTnI values obtained from 50 DMD patients were analyzed retrospectively. The relationship between cTnI and echocardiography data was assessed.

**Results:** this study registered 50 male subjects with DMD varying from 4 to 17 years old. Only 23 DMD patients were in the ambulatory phase at enrollment. Of all the patients, 28 were set on glucocorticosteroid medication with additional therapies consisting of an angiotensin-converting enzyme inhibitor (57.1%), angiotensin receptor blocker (10.7%), and beta-blockers (39.2%) at the time of data collection. Totally, there were 37 DMD children with normal ejection fraction (EF) (age 10.4 ± 2.7 yo; EF% 65 ± 3.2) and troponin I level of 0.04 ± 0.09 ng/ml, of which just two had an abnormal troponin level (over 0.04 ng/ml); 9 DMD patients with preserved EF (age 13.2 ± 3.2 yo, EF% 51.3 ± 8.2) and troponin I level of 0.32 ± 0.22 ng/ml, of which three had an abnormal troponin level; and 4 DMD subjects with low EF (age 14.2 ± 3.5 yo; EF% 48 ± 2.9) and troponin I level of 0.1 ± 0.13 ng/ml, of which three had an abnormal troponin level. In addition, troponin I levels in DMD subjects with low EF were increased compared to subjects with normal EF (p = 0.04).

**Conclusions:** further understanding of cTnI levels in DMD is necessary to best monitor and treat the cardiomyopathy associated with DMD.

**Keywords:** duchenne, cardiomyopathy, troponin

**P-445****Undetectable serum troponin in dilated cardiomyopathy, an unusual presentation of TNNI3 mutation**

*Alvaro Lafuente-Romero<sup>1</sup>, Guillermo Santos Simarro<sup>2</sup>, Luis Fernández García Moya<sup>3</sup>, Fernando Santos Simarro<sup>3</sup>, Juan J Menéndez Suso<sup>2</sup>, Juvenal Rey Lois<sup>4</sup>, Luis García Guereta<sup>1</sup>, Federico Gutierrez Larraya<sup>1</sup>*  
<sup>1</sup>Pediatric Cardiology Department, Hospital Universitario La Paz, Madrid, Spain.; <sup>2</sup>Pediatric Intensive Care Department, Hospital Universitario La Paz, Madrid, Spain.; <sup>3</sup>Institute of Medical & Molecular Genetics (INGEMM), Idipaz, CIBERER, Hospital Universitario La Paz, Madrid, Spain.; <sup>4</sup>Congenital Cardiac Surgery Department, Hospital Universitario La Paz, Madrid, Spain.

**Background and Aim:** Cardiac troponin (cTp) is the main serum biomarker for myocardial injury diagnosis, due to its high sensibility and specificity. Undetectable levels are frequently used to rule out diagnosis. Cardiomyopathies, which are the leading cause of cardiac transplantation in childhood, usually present with troponin rises during decompensations. On the other hand, troponin function is implicated in the pathophysiology of many familial cardiomyopathies. The understanding of cTp serum analysis is essential to interpreting laboratory results and making diagnosis.

**Method:** We present a case of dilated cardiomyopathy due to cardiac troponin I mutation (*TNNI3*) that showed unusual but pathophysiologically expected laboratory behavior.

**Results:** A 10 month old infant born to consanguineous parents was admitted to the pediatric intensive care unit due to cardiogenic shock with no relevant past medical history. Bedside echocardiography confirmed the diagnosis of dilated cardiomyopathy of unknown etiology with severe systolic dysfunction. Myocarditis and other causes were discarded due to undetectable cTnI levels. After weeks of supportive treatment and lack of improvement with functional severe mitral regurgitation, a surgical mitral prosthesis implantation was performed. Nevertheless, eventually the patient required mechanical assistance as a bridge to heart transplantation. Despite these procedures and events that implied direct myocardial damage (cardiotomy . . .), cTnI remained below detectable limits during the whole admission. Meanwhile, other nonspecific myocardial markers such as CK or LDH did peak during these episodes.

During etiologic work-up, genetic/familial origin was suspected. A protein-truncating mutation in cTnI gene was hypothesized and subsequently confirmed by a massive sequencing genetic panel: an homozygous change in *TNNI3* gene ((NM\_000363.4) exon 5: c.204del; p.(Arg69Alafs\*8)) was detected and classified as pathogenic, inherited from each of his parents. This mutation explained the impossibility to detect cTnI by usual laboratory arrays.

Finally, as a confirmation of our hypothesis, the child was transplanted and cTnI rose from then on as expected.

**Conclusions:** Mutations in sarcomeric genes are a frequent cause of cardiomyopathies. Laboratory findings can help elucidate the etiology, though their deep understanding is key. Undetectable troponin I is an uncommon entity explained by *TNNI3* gene mutation that had not been previously reported, it was a defining clue for the case.

**Keywords:** dilated cardiomyopathy, troponin I, myocardial injury, cardiac transplantation, mutation

#### P-446

##### Myocarditis and acute-onset dilated cardiomyopathy in the youngest: A diagnostic challenge.

Roger Esmel Vilomara<sup>1</sup>, Lucía Rianza<sup>3</sup>, Paola Dolader<sup>2</sup>, Pedro Betrian<sup>2</sup>, Marta Garrido<sup>4</sup>, Sandra Navarro<sup>4</sup>, Anna Sabaté Rotés<sup>2</sup>, Ferran Rosés Noguera<sup>2</sup>, Ferran Gran<sup>2</sup>

<sup>1</sup>Pediatric Cardiology, Vall d'Hebron Hospital Campus, Barcelona, Spain; <sup>2</sup>Pediatric Cardiology, Hospital de la Santa Creu i Sant Pau, Barcelona, Spain; <sup>3</sup>Pediatric Cardiology, Vall d'Hebron Hospital Campus, Barcelona, Spain; <sup>4</sup>Pathological Anatomy, Vall d'Hebron Hospital Campus, Barcelona, Spain

**Background and Aim:** The etiological diagnosis of infants and toddlers with acute-onset heart failure may be challenge at presentation. The aim of this study is to analyze the capacity of magnetic resonance imaging (MRI) in differentiating between myocarditis and dilated cardiomyopathy (DCM) and whether it correlates with a greater inflammation detected by endomyocardial biopsy (EMB).

**Method:** Prospective study including infants and toddlers (median age 0.93years, IQR 0.5-3.07) with acute-onset heart failure that underwent MRI with parametric mappings and EMB; lately diagnosed as DCM (n = 6) or myocarditis (n = 4) depending on evolution and genetic tests. A control group (n = 17) of patients without heart failure who had undergone EMB and MRI (cardiac transplant recipients free of rejection) was also analyzed.

**Results:** MRI accurately detects myocardial inflammation, showing a significant difference between cases and controls using the Lake Louise criteria or parametric mappings. When it comes to differentiating myocarditis from DCM, both present severe dysfunction and no statistically significant differences are either observed in clinical, echocardiographic characteristics nor cardiac biomarkers. Lake Louise criteria may differentiate between groups, but the sample is too small to reveal significant RESULTS: myocarditis present T2-weighted hyperintensity (3/4), early (2/4) and late gadolinium enhancement (4/4), DCM present T2-weighted hyperintensity (5/6), early (1/6) and late gadolinium enhancement (3/6). Parametric MRI mappings were elevated in myocarditis: native T1 (median 1184.5, IQR 1104.5-1236), extracellular volume fraction (ECV) (40.48, IQR 33.21-53.15) and T2 (61.5ms, IQR 57.5-65.5); also in DCM: native T1 (1177ms, IQR 1100.25-1239.25), ECV (35.29, IQR 32.14-38.52) and T2 (64.5ms, IQR 58-70.5). Only T2 mappings revealed a significant difference between groups (p = 0.047). A patchy affection, detected as greater intramyocardial variability in ECV, could also

be discriminatory in favour of myocarditis (p = 0.027). EMB differentiates myocarditis from DCM (p = 0.038), but greater inflammation on EMB doesn't significantly imply greater inflammation on MRI and is not associated with any mapping in this study.

**Conclusions:** EMB is the gold standard test to guide the diagnosis in acute-onset heart failure. MRI can also detect myocardial inflammation but it may be misleading to differentiate DCM from myocarditis. T2 mappings or variability in ECV may have diagnostic value. Larger studies are needed to confirm these results.

**Keywords:** Myocarditis, dilated cardiomyopathy, magnetic resonance imaging, mapping, endomyocardial biopsy

#### P-448

##### Arterial tortuosity syndrome: phenotypic features and cardiovascular manifestations in 4 newly identified patients.

Roger Esmel Vilomara<sup>1</sup>, Irene Valenzuela<sup>4</sup>, Lucía Rianza<sup>5</sup>, Benjamín Rodríguez Santiago<sup>6</sup>, Ferran Rosés Noguera<sup>2</sup>, Susana Boronar<sup>2</sup>, Anna Sabaté Rotés<sup>2</sup>

<sup>1</sup>Department of Pediatric Cardiology, Vall d'Hebron Hospital Campus, Barcelona; <sup>2</sup>Pediatric Cardiology, Department of Pediatrics, Hospital de la Santa Creu i Sant Pau, Barcelona; <sup>3</sup>Department of Pediatric Cardiology, Vall d'Hebron Hospital Campus, Barcelona, Spain; <sup>4</sup>Department of Pediatrics, Hospital de la Santa Creu i Sant Pau, Barcelona, Spain; <sup>5</sup>Department of Pediatric Radiology, Vall d'Hebron Hospital Campus, Barcelona, Spain.; <sup>6</sup>Department of Genetics, Hospital de la Santa Creu i Sant Pau, Barcelona, Spain.

**Background and Aim:** Arterial tortuosity syndrome (ATS) is an autosomal recessive connective tissue disease caused by biallelic variants in the *SLC2A10* gene. It is considered an extremely rare disease, only 106 individuals with genetically confirmed ATS have been identified to date. It is characterized by tortuosity and elongation of the aorta and medium-sized arteries, focal stenosis and propensity for aneurysm formation. The aim of this case series is to contribute to the phenotypic and cardiovascular description of this disease, as well as its genetic characterization.

**Method:** Four cases of ATS from two families are described. At the time of diagnosis, all patients were evaluated by clinical geneticists and pediatric cardiologists. Clinical and molecular information and MRI (magnetic resonance imaging) data were retrospectively obtained.

**Results:** We report 3 siblings born from healthy consanguineous parents with an homozygous variant in *SLC2A10*: c.510G>A (p.Trp170Ter) and a single child with two heterozygous variants: c.417T>A (p.Tyr139Ter), classified as pathogenic, and c.899T>G (p.Leu300Trp), not previously described as pathogenic but with an allegedly deleterious effect in compound heterozygosis.

The presented clinical spectrum is wide, two cases with diaphragmatic hernia and one with a complex uropathy are highlighted. Regarding the vascular involvement, a predominant supra-aortic affection stands out with less involvement in abdominal aorta, visceral branches and lower extremities. No aneurysms were observed and only 1 case of focal stenosis (renal artery). All presented severe intracranial tortuosity. No case of intracardiac involvement is described. To reduce hemodynamic stress on the arterial wall, beta-adrenergic blocking treatment was prescribed.

**Conclusions:** Four novel pediatric cases are described, contributing to the clinical delineation of the entity. A case with complex uropathy, reported in the literature only 11 times, is included. Moreover, a not previously described mutation is presented; with an allegedly deleterious effect in association with another pathogenic variable.

**Keywords:** Arterial tortuosity syndrome, SLC2A10, GLUT10, Genotype, Pyelectasis

#### P-449

##### **Prevalence of 22q11.2 deletion and immunological profile in children with cono-truncal anomalies**

Souvik Das, Arun Kumar Baranwal, Amit Rawat, Ankur Jindal, Anupriya Kaur, Sanjeev Naganur, Anand Kumar Mishra, Anit Kaur Postgraduate Institute of Medical Education and Research (PGIMER), Chandigarh, India

**Background and Aim:** Cono-truncal anomalies constitute 12% of congenital heart disease (CHD) (1) and are often associated with 22q11.2 deletion. Immunological abnormalities are common feature of 22q11.2 deletion (2). As formation of conotruncus and differentiation of branchial arches to thymus are governed by the same set of neural crest cells, (3) conotruncal anomalies are likely to be associated with T cell defects

**Method:** Preoperative children with conotruncal anomalies were planned to be evaluated for clinical dysmorphism, lymphocyte subsets by flowcytometry, immunoglobulin levels by nephelometry and 22q11.2 deletion by multiplex ligand-dependent probe amplification (MLPA) for 22q11.2DS during Jan'21-Mar'22 in a tertiary care teaching hospital.

**Results:** 101 patients were enrolled (age, 1 day-11 years); cardiac defects being d-TGA (n = 42), TOF (n = 38), DORV (n = 14), TA (n = 5), IAA type B (n = 1) and DOLV (n = 1). 17.5% (17/97) children had clinical dysmorphism. Flowcytometry (n = 82) revealed low percentages of lymphocytes (n = 9, 9%), T cells (n = 17, 20.7%), CD4+ cells (n = 20, 24.4%) and CD8+ cells (n = 19, 23.2%) in significant number of cases, as were the absolute counts of these subsets: lymphocyte count (n = 32, 33%), T cells (n = 42, 51.2%), CD4+ cells (n = 41, 50%) and CD8+ cells (n = 42, 51.2%). However, only 14.1% (10/71) patients had low IgG levels. MLPA (n = 101) revealed 22q11.2 deletion in eight patients, of whom 7 had subtle dysmorphism. TBX1-2 and TBX1-7 genes were deleted in all patients, while other 19 genes were deleted in various combination in different patients. 66 patients could be contacted at the end of study period, of whom 32 survived. There was no association between survival outcome and low T cell, CD4, CD8 cell counts. There was no association between 22q11.2 deletion and low cell counts or survival outcome.

**Conclusions:** 22q11.2 deletion was present only in 8% patients. However, low T cell subsets were widely prevalent. Defining its clinical significance needs larger and long-term studies.

**Keywords:** Di George Syndrome, Conotruncal anomalies, 22q11.2 deletion, immunological profile, lymphocyte counts.

#### P-450

##### **The assembly is also essential: complex v deficiency due to TMEM70 defect, a literature review and peculiarities in a case with cardiac manifestations**

Alina Botezatu<sup>1</sup>, Alexandru Necula<sup>1</sup>, Gabriella Horvath<sup>2</sup>, Cecilia Lazea<sup>3</sup>, Melinda Baizat<sup>4</sup>, Adina Chiş<sup>5</sup>, Romana Cleopatra Vulturar<sup>5</sup>  
<sup>1</sup>Faculty of Medicine, "Iuliu Hațieganu" University of Medicine and Pharmacy Cluj-Napoca, Romania; <sup>2</sup>Division of Biochemical Genetics, Department of Pediatrics, University of British Columbia, BC Children's Hospital, Vancouver, BC, Canada; <sup>3</sup>Pediatric Clinic 1, Emergency Pediatric Hospital, Cluj-Napoca, Romania & Department Mother and Child, "Iuliu Hațieganu" University of Medicine and Pharmacy, Cluj-Napoca, Romania; <sup>4</sup>Zalau Emergency County Hospital, Department of Neonatology, Zalau, Romania; <sup>5</sup>Department of Cell and Molecular

Biology, "Iuliu Hațieganu" University of Medicine and Pharmacy Cluj-Napoca, Romania,

**Background and Aim:** Deficiencies in the oxidative phosphorylation system (OXPHOS) are an important cause of a large and diverse group of disorders affecting the brain, muscles and heart. TMEM70 mutation leads to the nuclear-encoded ATP synthase deficiency, resulting in mitochondrial complex V assembly defect syndrome characterized by a neonatal onset disorder with severe lactic acidosis, muscular hypotonia, cardio-myopathy, variable central nervous system involvement, accompanied by 3-methylglutaconic aciduria (3-MGA-uria) and hyperammonemia. Through this comprehensive analysis that we are doing, as well as the case of our patient with TMEM70 deficiency that we are presenting, we want to provide important information regarding the diagnosis, prognosis and optimal management of metabolic crises, encountered in these patients.

**Method:** Retrospective clinical data, metabolic profiles and therapy methods of about 62 patients, from European countries, also, Japan, Turkey, Israel and Russia with confirmed TMEM70 mutations, were collected and included in a targeted literature review; several laboratory peculiarities in our newborn case with homozygous TMEM mutations are included. This is a pan-ethnic disease, but frequently in the Roma population, with a variable outcome.

**Results:** An increasing number of affected individuals, many from consanguineous parents of Roma ethnic background, have been reported, due to a founder allele effect in this population, since its first description by Cízková et al., 2008. The disease outcome is severe, and more than half of affected individuals die in early childhood. The three most common clinical features associated with these mutations are lactic acidosis, hypotonia, and hypertrophic cardiomyopathy. Anaplerotic therapy is recommended for patients with the TMEM70 defect, especially during metabolic crises, although clinical trials for this are still lacking. In our case, the clinical and laboratory data have improved considerably after introducing the special milk formula (Galacto start).

**Conclusions:** There are many learning lessons from studying this defect causing complex V deficiency with morphological stigmata, hypertrophic cardiomyopathy, and metabolic crises. Raising awareness of this disease among pediatric cardiologists will help to diagnose and introduce supportive therapy sooner in future cases. TMEM70 defect is considered amenable to treatment; there are cases with long-term survival based on anaplerotic therapy that includes Sodium citrate, L-carnitine, Coenzyme Q10, vitamin C and others.

**Acknowledgement:** Project 'Research, diagnosis and education in inborn errors of metabolism in Romania'.

**Keywords:** TMEM70, hypertrophic cardiomyopathy, mitochondria, ATP synthase assembly defect.

#### P-453

##### **Cardiovascular involvement in friedreich's ataxia: outcomes from children to young adults**

Sergi Cesar Diaz<sup>1</sup>, Jose Cruzalegui Gomez<sup>1</sup>, Mar O'callaghan<sup>2</sup>, Alejandra Darling<sup>2</sup>, Andrea Greco<sup>1</sup>, Victoria Fiol<sup>1</sup>, Patricia Cerralbo<sup>3</sup>, Oscar Campuzano Larrea<sup>4</sup>, Josep Brugada Tarradellas<sup>1</sup>, Fredy Chipa Ccasani<sup>1</sup>, Estefania Martinez Barrios<sup>3</sup>, Georgia Sarquella Brugada<sup>1</sup>  
<sup>1</sup>Arrhythmia, Inherited Cardiac Diseases and Sudden Death Unit, Hospital Sant Joan de Déu, University of Barcelona, Barcelona, Spain; <sup>2</sup>European Reference Network for Rare, Low Prevalence and Complex Diseases of the Heart (ERN GUARD-Heart), 1105 AZ Amsterdam, The Netherlands.; <sup>3</sup>Paediatric Neurology Department, Hospital Sant Joan de Déu, University of Barcelona, Barcelona, Spain; <sup>4</sup>Arritmies Pediàtriques, Cardiologia Genètica i Mort Sobtada, Malalties

Cardiovasculars en el Desenvolupament, Institut de Recerca Sant Joan de Déu, Esplugues de Llobregat, 08950 Barcelona, Spain.; <sup>4</sup>Medical Science Department, School of Medicine, Universitat de Girona, 17003 Girona, Spain; Cardiovascular Genetics Center, University of Girona-IDIBGI, 17190 Girona, Spain

**Background and Aim:** Comprehensive cardiovascular follow-up with clinical, electrophysiologic and echocardiographic data is essential in this group of patients since heart failure and life-threatening arrhythmias are the main cause of mortality in Friedreich's Ataxia (FRDA). We present a retrospective description of cardiovascular involvement and outcomes during long follow-up from children to adulthood.

**Method:** We analysed retrospectively a cohort of 32 patients diagnosed with Friedreich's Ataxia, from 1990. Demographic, genetics, clinical, electrophysiologic and echocardiographic data were collected.

**Results:** Thirty-two patients (5-35y, 68% females) were analysed. FRDA cardiomyopathy was diagnosed in 85%. Echocardiography: LVPW showed a significant reduction over-time: 13mm at onset (10.5, 14; z+2.58) and 11.5mm at last control (10, 13; z+2.63) (p = 0.005). RWT value was  $\geq 0.42$  in the majority of patients suggesting concentric LV remodelling. Longitudinal strain showed decreased values (median -14%) despite of preserved EF. One patient showed transient severe diastolic dysfunction related to adverse drug event. LVOT was detected in 1 patient. Atrial fibrillation was detected in two patients (20 and 22y), 5 patients showed asymptomatic atrial tachycardia (3 children), 1 both atrial tachycardia and AVNRT requiring an ablation, and 1 showed pre-excitation syndrome requiring an ablation. Four patients died (19, 25, 28&29y): acute heart failure (2), respiratory insufficiency and severe weakness (1) and unknown reason (1). All patients were under different combinations of treatment (iron chelators, vitamins, Q10coenzyme and idebenone).

**Conclusions:** Cardiomyopathy is highly present in pediatric FRDA patients, characterized by concentric remodelling and preserved EF. Echocardiography showed normal LVMI and increased RWT values, so RWT could be more sensitive than LVMI for detecting FRDA cardiomyopathy. Myocardial strain is more accurate. Cardiomyopathy showed slow progression and, in some cases, showing wall thinning over time. Atrial tachycardia is present early during pediatric age. Atrial fibrillation is present in young adult population. No ventricular arrhythmia was detected. Acute heart failure is the main cause of mortality of our cohort but it was present in young adults. Questions remain about the progression, nature and frequency of the patterns of cardiac involvement in FRDA.

**Keywords:** Friedreich's Ataxia, cardiomyopathy, pediatric

#### P-454

##### Characterization of early-onset pediatric cardiomyopathies

Sergi Cesar Diaz<sup>1</sup>, Jose Cruzalegui Gomez<sup>1</sup>, Estefania Martinez Barrios<sup>2</sup>, Fredy Chipa Ccasani<sup>1</sup>, Andrea Greco<sup>1</sup>, Victoria Fiol<sup>1</sup>, Patricia Cerralbo<sup>2</sup>, Oscar Campuzano Larrea<sup>3</sup>, Josep Brugada Tarradellas<sup>1</sup>, Georgia Sarquella Brugada<sup>1</sup>

<sup>1</sup>Arrhythmia, Inherited Cardiac Diseases and Sudden Death Unit, Hospital Sant Joan de Déu, University of Barcelona, Barcelona, Spain; European Reference Network for Rare, Low Prevalence and Complex Diseases of the Heart (ERN GUARD-Heart), 1105 AZ Amsterdam, The Netherlands.; <sup>2</sup>Arritmies Pediàtriques, Cardiologia Genètica i Mort Sobtada, Malalties Cardiovasculars en el Desenvolupament, Institut de Recerca Sant Joan de Déu, Esplugues de Llobregat, 08950 Barcelona, Spain.; <sup>3</sup>Medical Science Department, School of Medicine, Universitat de

Girona, 17003 Girona, Spain; Cardiovascular Genetics Center, University of Girona-IDIBGI, 17190 Girona, Spain

**Background and Aim:** Pediatric cardiomyopathy is a rare heart condition sometimes present from birth and related to very heterogeneous etiologies. The aim of this review is to present a cohort of early onset of cardiomyopathy in small children.

**Method:** Retrospective analysis of children with cardiomyopathies were reviewed (2014-2022). Neuromuscular and movement disorders patients were excluded from this analysis.

**Results:** Fifty-eight pediatric patients (80% male, mean age at diagnosis was 8 (0;14)

**Genotype-phenotype:**

HCM (29/58 patients, 1m-17y), age at diagnostic 1m-17y. MYH7 was responsible for 8 (2 with double mutation), MYBPC3 (4), LAMP2 (3), FLNC (1), GAA (1), HADHA (1), TPM1 (1), TNNT3 (1), and LZTR1 (1).

DCM (12/58 patients, 1m-17y). Genetics: MYH7 (3), LMNA (1), TNNT2 (1), TTN (1), FLNC (1), and a deletion in 1p36.

NCMC (6/58, 1m-15y), MYH7(1), HCN4 (1), TAZ (1). Three patients did not have genetic diagnosis.

RCM (3/58, 1-4y). Genetics showed double mutation in MYBPC3 in two patients (sisters) and the other case did not have genetic diagnosis.

ACM (8/58 patients, 5-18y), 5p with desmosomal genes (DSP, PKP2), FLNC(1) and unknown(2).

Onset <6m (12p, 20.6%), 6m-5y (13p, 22.4%), 5-10t (11p, 19%), 10-18y (38%).

Early onset (<6m): syndromic and metabolic HCM (Rasopathy, PMM2, Pompe, LCHAD), sarcomeric DCM (MYH7 -3p-, TNNT2 -1p-), and NCMC (Barth Syndrome -1p-).

ICD (24p, 41%, 3-17y).

Heart transplantation in childhood (5p, 8.6%): Danon (2p), MYH7 (2p DCM, 1p HCM)

Mortality (7p, 12%): metabolic disease (5p), ACM double mutation DSP-PKP2 (1p), RCM MYBPC3 (1p).

**Conclusions:** In our cohort, early onset cardiomyopathies had poorest prognosis, especially those manifesting before 1 year of age and those related with metabolic disorders.

Genetics is key to evaluate the prognosis in these rare and severe pediatric conditions that often does not follow any rule and could be present with double mutations that can imply severe phenotypes. DCM with severe heart failure in <2 years old and younger adults with HCM in relation with MYH7 and Danon disease were the two groups of age with more risk for heart transplantation. Mortality is high in our cohort (12%), especially increased by metabolic diseases, restrictive cardiomyopathy and arrhythmogenic cardiomyopathy.

**Keywords:** pediatric, cardiomyopathies

#### P-455

##### Rasopathies – can't judge a book by its cover? experience of a tertiary center

Catarina F Silva<sup>1</sup>, Andreia Constante<sup>1</sup>, Sílvia Gomes<sup>1</sup>, Sofia Nunes<sup>2</sup>, Inês Carvalho<sup>2</sup>, Petra Loureiro<sup>1</sup>, Conceição Trigo<sup>1</sup>, Margarida Venâncio<sup>2</sup>, Fátima Pinto<sup>1</sup>, Diana Antunes<sup>2</sup>

<sup>1</sup>Pediatric Cardiology Department, Hospital de Santa Marta, CHULC, Lisbon, Portugal; <sup>2</sup>Medical Genetics Department, Hospital Dona Estefânia, CHULC, Lisbon, Portugal

**Background and Aim:** RASopathies refers to a clinical spectrum of disorders typically associated with dysmorphic features, short

stature, cognitive impairment and cardiac disease (CD). This group of diseases, namely Noonan (NS), Leopard and Cardiofacialcutaneous Syndrome, is caused by germline mutations in genes associated with RAS/MAPK pathway and have a worldwide prevalence of 1:1000, being NS the most common one.

Our aim is to characterize phenotypically and genetically the patients followed in our center.

**Method:** All patients followed in the Genetic Clinic (1998–2022) were reviewed and the 45 cases with molecularly confirmed RASopathy were included in this study.

**Results:** Based on these results, the prevalence of RASopathies in our center is 1:40000. In this sample, 82% (N = 37) of the patients have dysmorphisms, 29% (N = 13) have delayed psychomotor development and 40% (N = 18) have short stature. Regarding cardiac phenotype, 73% (N = 29) have CD: 66% (N = 19) were diagnosed with pulmonary valve stenosis and 34% (N = 10) with hypertrophic cardiomyopathy. Relevant family history was present in 44% (N = 20) – out of the 12 non-index cases, only 66% (N = 8) had the appropriate screening and, of those, 50% (N = 4) were diagnosed with CD. The average age of the genetic diagnosis index cases is 6 years old (0–46 years old). Despite 22% (N = 10) having prenatal ultrasound abnormalities, only one case had prenatal molecular diagnosis. Two of those families were offered Preimplantation Genetic Diagnosis in subsequent pregnancies. The most prevalent mutations in our sample were located in the PTPN11 gene – 58% (N = 26) – and in the RAF1 gene – 13% (N = 6). From the 10 different reported mutations in our sample, only one was new (LZTR1 gene).

**Conclusions:** Our results showed a lower number of confirmed cases than we expected. These results reinforce the underdiagnosis pattern of RASopathies, possibly related to subtle forms of disease. The lower percentage of cardiac phenotype in non-index cases is due to the fact that CD is one of the red flags that prompts genetic diagnosis, with facial dysmorphism being often overlooked. A higher awareness of these diseases and better communication between clinical specialties can improve diagnostic rates and offer better clinical care to these patients. Follow-up research studies are needed to better understand the genetic basis and its influence in the cardiac phenotype of these patients.

**Keywords:** RASopathies, Pulmonary Valve Stenosis, Hypertrophic Cardiomyopathy, Cardiac Disease, Genetics,

#### P-456

##### Red flags in children FLNC-pathies

Anwar Baban<sup>1</sup>, Giovanni Parlapano<sup>1</sup>, Viola Alesi<sup>2</sup>, Monia Magliozzi<sup>2</sup>, Silvia Genovese<sup>2</sup>, Marianna Cicienia<sup>1</sup>, Sara Loddo<sup>2</sup>, Luca Di Chiara<sup>3</sup>, Fabiana Fattori<sup>4</sup>, Adele D'amico<sup>4</sup>, Paola Francalanci<sup>5</sup>, Antonio Amodio<sup>6</sup>, Antonio Novelli<sup>2</sup>, Fabrizio Drago<sup>1</sup>

<sup>1</sup>The European Reference Network for Rare, Low Prevalence and Complex Diseases of the Heart—ERN GUARD-Heart. Department of Pediatric Cardiology and Cardiac Surgery, Heart and Lung Transplantation, Bambino Gesù Children Hospital and Research Institute, IRCCS, 00165 Rome, Italy; <sup>2</sup>Laboratory of Medical Genetics, Translational Cytogenomics Research Unit, Bambino Gesù Children's Hospital, IRCCS, 00165 Rome, Italy; <sup>3</sup>Pediatric Cardiac Intensive Care Unit, Department of Cardiology and Cardiac Surgery, Bambino Gesù Children's Hospital, IRCCS, 00165 Rome, Italy; <sup>4</sup>Unit of Muscular and Neurodegenerative Disorders, Bambino Gesù Children's Hospital, IRCCS, 00165 Rome, Italy; <sup>5</sup>Department of Pathology, Bambino Gesù Children's Hospital and Research Institute, IRCCS, 00165 Rome, Italy; <sup>6</sup>Heart Failure and Transplant, Mechanical Circulatory Support Complex Unit, Bambino Gesù Children's Hospital, IRCCS, 00165 Rome, Italy

**Background and Aim:** Filamin C is a protein specifically expressed in myocytes and cardiomyocytes and is involved in several biological functions, including sarcomere contractile activity, signaling, cellular adhesion, and repair. FLNC variants are associated with different disorders ranging from striated muscle (myofibrillar distal or proximal) myopathy to cardiomyopathies (CMPs) (restrictive, hypertrophic, and dilated), or both. The outcome depends on functional consequences of the detected variants, which result either in FLNC haploinsufficiency or in an aberrant protein, the latter affecting sarcomere structure leading to protein aggregates. Cardiac manifestations of filaminopathies are most often described as adult onset CMPs and limited reports are available in children or on other cardiac spectrums (congenital heart defects—CHDs, or arrhythmias).

**Method:** Our tertiary academic pediatric center represents the referral point for the center and south of Italy for the multidisciplinary management of patients with CMPs. In this study we include data from 14 children with filaminopathy C out of 500 pediatric patients with early-onset different cardiac features ranging from CMP to arrhythmias and CHDs. Patients had multisystemic clinical assessment, multiorgan screening and genetic study through NGS sequencing and microarray analysis.

**Results:** In one patient, we identified a deletion encompassing FLNC detected by microarray, which was overlooked by NGS. We established a potential genotype–phenotype correlation of the p.Ala1186Val variant in severe and early-onset restrictive cardiomyopathy (RCM) associated with a limb-girdle defect (two new patients in addition to the five reported in the literature). Moreover, in three patients (21%), we identified a relatively frequent finding of long QT syndrome (LQTS) associated with RCM (n = 2) and a hypertrabeculated left ventricle (n = 1).

**Conclusions:** RCM and LQTS and musculoskeletal involvement in children might represent specific red flags for FLNC variants. Further studies are warranted in pediatric cohorts to delineate potential expanding phenotypes related to FLNC.

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**Keywords:** FLNC, restrictive cardiomyopathy, musculoskeletal changes, congenital heart defects, long QT syndrome

#### P-457

##### Myocardial blood fractional flow reserve (FFR) testing in giant coronary aneurysm of Kawasaki disease

Yoshiaki Hashimoto, Makoto Watanabe, Ryosuke Matsui, Kanae Shimada, Koji Hashimoto, Masanori Abe, Mitsuhiro Kamisago, Ryuji Fukazawa

Department of Pediatrics, Nippon Medical School, Tokyo, Japan

**Background and Aim:** FFR is a well-established indicator of interventional and surgical intervention in adult coronary artery disease. FFR data for GAN of Kawasaki Disease are still scarce.

**Method:** A retrospective study was conducted on patients with GAN who underwent cardiac catheterization at our institution from 1990 to 2022. The significance of FFR in GAN was investigated. Coronary artery lesions were limited to the left coronary artery, and the presence of ischemia was determined by treadmill, scintigraphy and PET scan. The significance of FFR in GAN was examined.

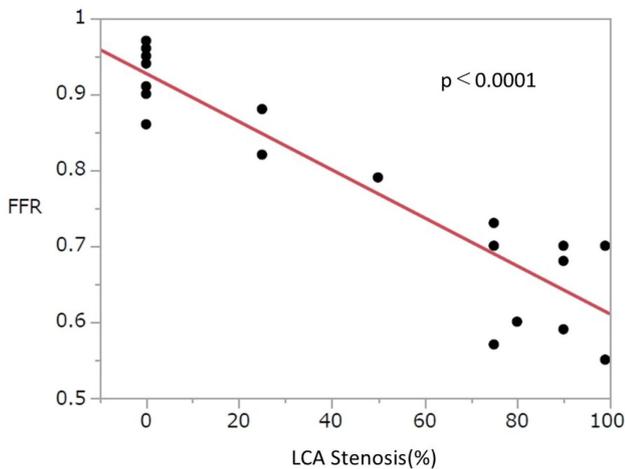
**Results:** Forty-nine applicable patients were examined, and 22 patients were examined excluding those who did not undergo FFR (n = 27), such as LAD occlusion. The mean (median) age was 3 years and 10 months (3 years and 6 months) at the onset of Kawasaki disease and 12 years (11 years and 8 months) at the time of examination. All patients in the GAN alone group were within the normal range, and all patients showed a decrease in

FFR  $\leq 0.8$  from  $>50\%$  stenosis, showing a strong correlation between degree of stenosis (%) and FFR value ( $p < 0.0001$ ). There were 0 ischemia-positive cases in the GAN alone group and 8 ischemia-positive cases in the GAN + stenosis group ( $p = 0.0055$ ), while there were 3 ischemia-negative cases with low FFR values.

**Conclusions:** FFR testing is useful in GAN management and understanding coronary hemodynamics.

**Keywords:** Kawasaki Disease, GAN, FFR, ischemia

Bivariate relationship between left coronary artery stenosis and FFR



#### P-458

##### The shifting anatomy of the patent ducts arteriosus towards a new paradigm for unmet needs

Nagib Dahdah<sup>1</sup>, Iñaki Navarro Castellanos<sup>2</sup>, Jawad Al Kassmy<sup>3</sup>, Joaquim Miro<sup>4</sup>

<sup>1</sup>Nagib Dahdah; <sup>2</sup>Iñaki Navarro-Castellanos; <sup>3</sup>Jawad Al-Kassmy; <sup>4</sup>Joaquim Miro

**Background and Aim:** Following birth, the ducts arteriosus undergoes physiological dynamic closure via a prostaglandin receptor mechanism. There is an under-development of the prostaglandin receptor cells in the ductal tissue of the prematurely born baby. A sharp increase in survival rate of the extreme premies led to increased proportion of atypical patent ducts arteriosus (PDA). PDA occlusion devices initially developed for conal shaped duct are unfortunately neither adapted nor designed for the elongated tubular PDA of the premie. We sought to assess inherent risks of device instability, embolization, and protrusion of current PDA devices, including those adapted to tubular shapes carry.

**Method:** We reviewed PDA anatomy, device selection and consequential inadvertent events in a single institution between 10/2010 and 12/2020. For simplicity of data expression, ADO-1 is used for amplatzer ADO-1 and for Occlutech PDA device.

**Results:** There were 206 cases (75% females), 23% ex-premies (28  $\pm$  3 Weeks). Interventions performed at 2.7  $\pm$  3.3 years for the whole population, and at 16  $\pm$  16 months for premies. Weight 13.8  $\pm$  13.6 Kg (1.3-96). Non-type-A PDA represented 24% of cases compared to historical 15% (1989-Krichenko's classification) most with type E, C or D (mostly in premies n = 44; or

extreme-premies  $< 31$  Weeks; n = 37). ADO-1 devices represented 39%, ADO-2AS 44%, and ADO-2, Plug-2 or Plug-4 devices 17%. In situ complications included left pulmonary stenosis by device protrusion requiring subsequent stenting (n = 3), aortic protrusion exceeding 10% or 20% diameter in 32% and 21% of the cohort respectively. This was independent from prematurity status ( $p = 0.5$ ). Aortic protrusion of  $> 20\%$  was recorded in 41 (20%) cases; [20/81 (24%) in ADO-1, 12/90 (13%) ADO-2AS, and 9/21 (43%) Plug-2; ( $p = 0.008$ )], and none in the rarest used ADO-2 (n = 9) or Plug-4 (n = 5). PDA anatomy implicated in protrusions were 22/94 (23%) type-A, 0/4 type-B, 9/39 (23%) type-C, 3/9 (33%) type-D, and 11/68 (16%) type-E ( $p = 0.5$ ).

**Conclusions:** Current PDA devices are efficient in resolving the shunt. Protrusion of the device remains problematic overall at different proportion according to device used, including FDA approved ones. Our data are challenging in the absence of better current options but should be considered seriously in order to improve patient care and to adapt to new realities.

**Keywords:** Patent Ductus Arteriosus, Device Closure, Protrusion

#### P-459

##### Virtual reality for assessment of suitability for transcatheter correction of sinus venosus atrial septal defects – a retrospective study

Natasha Stephenson<sup>1</sup>, Eric Rosenthal<sup>2</sup>, Matthew Jones<sup>2</sup>, Shujie Deng<sup>1</sup>, Gavin Wheeler<sup>1</sup>, Julia A. Schnabel<sup>3</sup>, Kuberan Pushparajah<sup>4</sup>, John M. Simpson<sup>2</sup>

<sup>1</sup>School of Biomedical Engineering and Imaging Sciences, King's College London, United Kingdom; <sup>2</sup>Department of Congenital Heart Disease, Evelina Children's Hospital, London, United Kingdom; <sup>3</sup>School of Biomedical Engineering and Imaging Sciences, King's College London, United Kingdom; Faculty of Informatics, Technical University of Munich, Munich, Germany; Institute of Machine Learning in Biomedical Engineering, Helmholtz Centre, Munich, Germany; <sup>4</sup>School of Biomedical Engineering and Imaging Sciences, King's College London, United Kingdom; Department of Congenital Heart Disease, Evelina Children's Hospital, London, United Kingdom

**Background and Aim:** Sinus venosus atrial septal defect (SVASD) can now be treated by transcatheter correction (TCC), using a covered stent deployed in the SVC to direct systemic venous blood to the right atrium and anomalous pulmonary venous (PV) blood to the left atrium. Assessing suitability for TCC requires in-depth anatomical understanding, usually via production of segmented virtual or 3D-printed models. "3D Heart" is a virtual reality (VR) system which rapidly displays multimodality imaging data without complex pre-segmentation and enables placement of virtual stents. We assessed whether this system could assist interventionists to evaluate suitability for TCC.

**Method:** Two experienced interventional cardiologists reviewed 10 anonymised CT datasets in VR from patients with SVASD who had previously undergone surgery or TCC. Virtual stents were placed within the VR image to assist planning. Interventionists predicted patients' suitability for TCC and whether procedural PV protection (PVP) was required. Review time for each case was recorded.

**Results:** 6/10 patients underwent successful TCC. 3 required pulmonary vein protection. 4 patients underwent surgery; 2 were deemed unsuitable for TCC on standard imaging and 1 following diagnostic catheterisation. The 4th patient chose surgery although deemed suitable for TCC based on imaging.

Using VR, both interventionists were concordant in 8 cases correctly predicting suitability in 6, unsuitability in 1 and uncertainty

in 1. Interventionist 1 predicted suitability correctly in eight cases, incorrectly in one and uncertainty in 1. Interventionist 2 was correct in eight cases and uncertain in 2 cases. In case 8, both interventionists were uncertain – this patient underwent surgery because standard imaging had suggested unsuitability. In case 4, interventionist 2 was uncertain of success – this patient underwent successful TCC. The positive predictive value of VR was 93% and negative predictive value was 100%. Both interventionists were concordant for the need for PVP in 5 of 6 cases undergoing TCC and found placement of virtual stents useful for visualising the PV pathway. Median (IQR) VR review time was 10 (10) minutes.

**Conclusions:** The “3D Heart” VR prototype shows great potential to assist selection of patients with SVASD for TCC. Use of virtual stents in VR was rated highly by interventionists.

**Keywords:** Virtual reality, procedure planning, catheter intervention

Table 1: Case characteristics and results of VR review

Case	TCC offered	TCC attempted	TCC successful	PV protection	Surgery	VR review			
						Interventionist 1		Interventionist 2	
						Suitable	PV protection	Suitable	PV protection
1	Y	Y	Y	Y	N	Y, correct	Y, correct	Y, correct	Y, correct
2	Y	Y	Y	Y	N	Y, correct	Y, correct	Y, correct	Y, correct
3	Y	Y	Y	N	N	Y, correct	Y, incorrect	Y, correct	Y, incorrect
4	Y	N	-	-	Y	Y, correct	-	U	-
5	Y	Y	Y	Y	N	Y, correct	Y, correct	Y, correct	Y, correct
6	Y	Y	Y	N	N	Y, correct	N, correct	Y, correct	N, correct
7	N	N	-	-	Y	N, correct	-	N, correct	-
8	N	N	-	-	Y	U	-	U	-
9	Y	Y	Y	N	N	Y, correct	N, correct	Y, correct	N, correct
10	Y	Y	N	-	Y	Y, incorrect	-	N, correct	-

Abbreviations: Y – yes, N – no, U – uncertain

interventionists but data on their clinical application and performance remain limited. We aim to evaluate the performance of balloon-expandable, cobalt-chrome, hybrid-cell designed Optimus stents in CHD interventions.

**Method:** Retrospective multicenter review of CHD patients receiving Optimus stents. Stent mechanical behavior, clinical indications, and outcomes were comprehensively assessed.

**Results:** 183 stents (49.2% XXL/15-ZIG, 33.3% XL/12-ZIG, 17.5% L/9-ZIG) were implanted (98.9% success rate, 2.3% serious procedural complications rate) in 170 patients (57.6% males, 64.1% adults), median age 23.6 years (IQR, 15.2–39.2), and weight 63.5Kg (IQR, 47–75.7). Indications were right ventricular outflow tract standalone stenting or before reevaluation (62.4%), aortic coarctation treatment (15.3%), Fontan-circuit fenestration closure (12.4%), and miscellaneous (10%). 86/170 (50.6%) patients had PTFE-covered stenting (50% prophylactic). In 86/170 (50.6%) patients with stenotic lesions, median percentage of achieved stent expansion was 93.4% (IQR, 85.5–97.7), median gradient decreased from 28mmHg (IQR, 19–41) to 5mmHg (IQR, 1–9) ( $p < 0.001$ ), median vessel diameters increased from 13mm (IQR, 7.9–17) to 18.9mm (IQR, 15.2–22) ( $p < 0.001$ ), and percentage of vessel expansion was 45.2% (IQR, 19.8–91.3%). In 30/36 (83.3%) patients with graft, median dilation of 2mm (IQR, 2–5) above nominal diameter was achieved. Median stent shortening was 10.9% (IQR, 6.1–15.1) and was associated only with expansion diameter (OR: 0.66, 95%CI: 0.38–0.93). No clinically relevant fracture, stent embolization, or dysfunction occurred on a median follow-up of 9 (IQR, 4–14) months.

**Conclusions:** Optimus stents are effective tools for transcatheter treatment of simple and complex CHD. Optimus stents' reliable mechanical behavior and particular covering design can promote widespread use. A formal protocol-driven monitored study is required to evaluate outcomes of Optimus stents in the long run.

**Keywords:** congenital heart disease, new device, Optimus, stent, transcatheter interventions.

## P-462

### Multicenter experience with optimus balloon-expandable cobalt-chromium stents in congenital heart disease interventions

Raymond N. Haddad<sup>1</sup>, Sébastien Hascœt<sup>2</sup>, Clément Karsenty<sup>3</sup>, Ali Houeijeh<sup>4</sup>, Alban Elouen Baruteau<sup>5</sup>, Caroline Ovaert<sup>6</sup>, Estibaliz Valdeolmillos<sup>2</sup>, Zakaria Jalal<sup>7</sup>, Damien Bonnet<sup>1</sup>, Sophie Malekzadeh Milani<sup>1</sup>

<sup>1</sup>Centre de Référence Malformations Cardiaques Congénitales Complexes - M3C, Hôpital Universitaire Necker-Enfants malades, AP-HP, Paris, France.; <sup>2</sup>Department of Pediatric Cardiology and Congenital Heart Diseases, Marie Lannelongue Hospital, Groupe Hospitalier Paris Saint Joseph, Centre de Référence Malformations Cardiaques Congénitales Complexes - M3C, Le Plessis-Robinson, France.; <sup>3</sup>Department of Pediatric and Adult Congenital Cardiology, University Hospital of Toulouse, and Clinique Pasteur, Toulouse, France. Institut Des Maladies Métaboliques Et Cardiovasculaires, Université de Toulouse, Toulouse, France.; <sup>4</sup>Pediatric Cardiology and Congenital Heart Disease, Institut Coeur Poumon, Lille University, Lille, France.; <sup>5</sup>Department of Pediatric Cardiology and Pediatric Cardiac Surgery, CHU Nantes, Nantes, France.; <sup>6</sup>Pediatric Cardiology Department, Marseille University Hospital, Marseille, France.; <sup>7</sup>Department of Pediatric Cardiology, CHU Haut Leveque, centre de reference cardiopathies congénitales complexes M3C, IHU Lyric, Bordeaux, France.

**Background and Aim:** Bare-metal Optimus™ and PTFE-covered Optimus-CVS® (AndraTec, Germany) are new promising additions to the armamentarium of congenital heart disease (CHD)

## P-463

### Transvenous retrograde thoracic duct embolization for effective treatment of recurrent plastic bronchitis after fontan palliation

Raymond N Haddad<sup>1</sup>, Raphael Daruty<sup>2</sup>, Damien Bonnet<sup>1</sup>, Sophie Malekzadeh Milani<sup>1</sup>

<sup>1</sup>M3C-Necker, Hôpital Universitaire Necker-Enfants malades, Assistance Publique-Hôpitaux de Paris (AP-HP), Paris, France.;

<sup>2</sup>Department of Radiology, Hôpital Cochin, Assistance Publique-Hôpitaux de Paris (AP-HP), Paris, France.

**Background and Aim:** Plastic bronchitis (PB) is a rare but potentially detrimental complication of the Fontan circulation. Understanding of this disorder has improved over the past years with the advances in lymphatic system imaging and encouraging outcomes of the lymphatic transcatheter interventions. Thoracic duct embolization (TDE) has been proposed as a treatment strategy for lymphatic dysfunction. We report an interesting pediatric case of PB relapse 2 months after retrograde transvenous embolization of the caudal end of the thoracic duct (TD) and requiring end-to-end TDE for complete and sustainable resolution of symptoms.

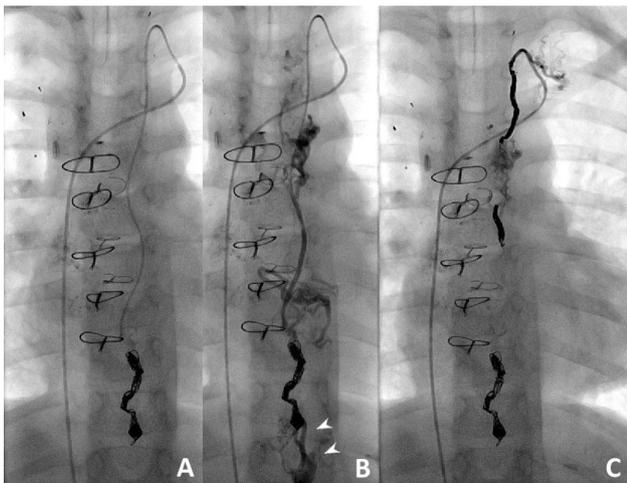
**Method:** A 5.5-year-old male patient (16 Kg/105 cm) was referred to our institution for severe PB refractory to conservative treatment 3 months after completion of Fontan palliation. Bi-inguinal transnodal fluoroscopy-guided lymphangiogram confirmed the chylous leak originating from the thoracic duct into the chest and did not opacify any target central lymphatic vessel for direct

transabdominal puncture. Retrograde approach from the femoral vein was adopted to catheterize the thoracic duct and selectively embolize its caudal portion using microcoils and liquid embolic adhesive.

**Results:** Recurrence of symptoms after 2 months indicated a redo catheterization to occlude the thoracic duct over its entire length using the same technique. Procedure was technically successful and the patient was uneventfully discharged after 2 days (Fig. 1). Sustained clinical improvement without subsequent bronchial cast recurrence was confirmed after 24 months of follow-up. The patient did not report any TDE-related complications such as chronic leg swelling, abdominal swelling, or chronic diarrhea.

**Conclusions:** We demonstrate that the etiology of recurrent PB in a child with Fontan physiology was retrograde flow from the TD into peribronchial lymphatic collateral networks. Fluoroscopy-guided lymphangiography was essential to understand the anatomy and flow pattern of the lymphatic system and to plan the lymphatic intervention. End-to-end TDE is an effective minimally-invasive treatment in Fontan patients with PB and can be necessary in case of relapse after selective embolization of the TD caudal section.

**Keywords:** Bronchitis, Fontan circulation, heart defects, congenital, lymphography, thoracic duct.



**Fig. 1** Transvenous retrograde TD cannulation (A) and ductography (B) showing residual leak and persistent dilated paratracheal lymphatic ducts above the previously embolized caudal section (white pointed arrows). End-to-end embolization of the TD with intercalation of glue and coils (C).

#### P-464

##### Early spontaneous closure of large arterial ducts in two term neonates with ebstein anomaly after failed attempts of transcatheter closure

Raymond N Haddad, Damien Bonnet, Sophie Malekzadeh Milani M3C-Necker, Hôpital Universitaire Necker-Enfants malades, Assistance Publique-Hôpitaux de Paris (AP-HP), Paris, France.

**Background and Aim:** The unique features of neonatal Ebstein's anomaly (EA) are the presence of pulmonary atresia, arterial duct (AD) patency, and the variability of pulmonary vascular resistance.

Promoting early ductal closure after ruling out or relieving anatomic pulmonary obstruction might improve neonatal survival. There is no opportunity to determine the timing and the occurrence of spontaneous closure or diminution in the size of the AD. Transcatheter ductal closure is an interesting option with several advantages but can be technically challenging in tiny neonates with large tortuous AD due to the limited available sizes of low-profile approved devices. We report, for the first time, on 2 consecutive term newborns with EA and large AD in whom mechanical stimulus of the AD during failed attempts of transcatheter closure led after 2 days to definitive closure with good outcomes.

**Method:** Two consecutive term neonates with EA were approached from the femoral vein for an attempted ductal closure. An 8mm Amplatzer Vascular Plug II (AVPII) was implanted in the AD (pulmonary end: 6.3mm, length 8.9mm) of the first 9-day-old patient (3Kg) and was removed before release for important leak and device instability. The delivery of 10mm AVPII was laborious and the procedure was aborted after 70ml of bleeding. A microvascular plug-9Q was implanted in the AD (pulmonary end: 7.6mm, length 16.1mm) of the second 14-day-old patient (2.8Kg) and was removed before release after immediate migration to the pulmonary artery trunk. The procedure was aborted in the absence of a larger low-profile occlusion device in the armamentarium.

**Results:** Patients were sent back to the intensive care unit for surveillance and a re-discussion of the management strategy. Follow-up ultrasound showed a spontaneous and progressive diminution in the AD size until complete closure two days postoperative with good follow-up outcomes.

**Conclusions:** Transcatheter closure should be attempted in similar cases requiring early ductal closure. Surgical ligation can be postponed a couple of days after failed transcatheter interventions when clinically possible. Further well-conducted studies seem interesting to evaluate whether mechanical stimulation of the ductal wall can be a useful alternative to initiate spontaneous ductal closure in preterm and term neonates.

**Keywords:** Arterial duct, Neonatal Ebstein Anomaly, spontaneous closure, term newborn.

#### P-465

##### Impact of adenosine and rapid ventricular pacing, on occurrence of aortic regurgitation after balloon aortic valvuloplasty

Filip Tyc, Michał Galeczka, Mateusz Knop, Sebastian Smerdziński, Roland Fiszer

Department of Congenital Heart Defects and Pediatric Cardiology, Faculty of Medical Sciences in Zabrze, Medical University of Silesia, Silesian Center for Heart Diseases, Zabrze, Poland

**Background and Aim:** Adenosine and rapid ventricular pacing (RVP) are techniques to stabilise the position of a balloon during interventional procedures. Both are used in balloon aortic valvuloplasty (BAV) to reduce possible aortic regurgitation (AR), however, available data on their impact is inconsistent and in case of adenosine very scarce. The aim of the study is to present the impact of RVP and adenosine on postprocedural AR.

**Method:** Between 1998-2021 over 150 patients underwent primary BAV in our centre, of which 111 pts (4.5 +/- 5.9 years) were included. Inclusion criteria were: primary BAV and good left ventricle (LV) function. Exclusion criteria were low-flow low-gradient aortic stenosis (AS). RVP was performed from a venous approach, pacing rate was chosen to decrease the LV pressure by 50%. Adenosine was administered to cause hemodynamically significant bradycardia or cardiac standstill in infants and neonates.

Due to the heterogeneity, the study group was divided into 2 sub-groups, group I: patients <10 kg (n = 57), of whom in 18 patients (31.6%) adenosine was used as a stabilising method, group II: patients >10 kg (n = 54) of whom in 17 patients (31.5%) RVP was used. Due to the lack of compelling data, use of a stabilising factor was operator dependent.

**Results:** Significant AR occurred in 15 patients (13.5%): group I - 8 patients (14%), group II - 7 patients (13%). In group I AR occurred in 1 patient (5.6%) who received adenosine vs. 7 patients (18%) who did not ( $p>0.05$ ). In group II AR occurred in 3 patients (17.7%) with RVP vs 4 patients (10.8%) without RVP ( $p>0.05$ ). Overall, AR occurred in 4 out of 35 patients (11.4%) in whom balloon stabilising technique was used vs. 11 patients (14.5%) without any balloon stabilising technique ( $p>0.05$ ). We did not observe a significant impact of neither stabilising technique on gradient reduction.

**Conclusions:** The study did not prove a positive effect of balloon stabilising techniques on overall successfulness of BAV. Presented data did not show a favourable effect of RVP on post-procedural aortic regurgitation, use of adenosine is promising, however it requires further study on bigger cohort.

**Keywords:** balloon aortic valvuloplasty, aortic stenosis, ventricular pacing, interventional cardiology

#### P-466

##### **Percutaneous revaluation of biological - stented and stentless valves in pulmonary position**

Roland Fiszer, Michał Gateczka

Department of Heart Defects and Pediatric Cardiology Silesian Center For Heart Diseases, Medical University of Silesia, Katowice, Poland

**Background and Aim:** Pulmonary stenosis and Pulmonary regurgitation (PR) as the result of previous surgery in right ventricular outflow tract obstruction may be treated by surgical valve implantation usually biological stented or stent less valves. Their viability is limited and in the patient lifetime will require either surgical replacement or interventional revaluation. The aim of the study was the evaluation of percutaneous revaluation of biological stented and stent less valves in pulmonary position

**Method:** Between 2010 - 2022 years 9 patients after surgical valve implantation in pulmonary position required valve exchange. There were two groups of patients after surgical procedure - group I (stent less valves) - 4 patients and group II (stented valves) - 5 patients. In both groups the main dysfunction was RV-PA gradient - from 37 to 50 median 46 mmHg in group I and from 8 to 53 median 51 mmHg in group II. Group I (Free-Style and Biopulmonic valves) was treated in usual manner with presenting and valve implantation in all patients. Group II (Magna 23 and 21 mm, Melody) was treated with valve cracking first in 4 patients and subsequent valve implantation

**Results:** All procedures in both groups were successful. Post procedure residual gradient in group I was from 7 to 20 median 17, 5 mmHg and group II was from 7 to 25 median 20 mmHg. There was no significant post procedure PR. No major complications were observed. In group I in all 4 patients prestenting procedure was performed and in all cases Melody valve was implanted. In group II 4 patients had first biological valve cracking with Atlas an Atlas Gold Balloons in 4 cases and subsequent valve presenting in 4 patients. Two of them had Edwards-Sapien valve and 2 had Melody valve implanted. One patient in the group had Melody as the first valve and had next Melody valve implanted in the procedure. All implanted Melody valves were on 22 mm balloon ensemble.

**Conclusions:** Percutaneous revaluation after surgical implantation of biological valve is feasible in typical manor in stent less and with previous frame stent cracking in stented valves

**Keywords:** pulmonary valve implantation, biological pulmonary valve

#### P-468

##### **Headaches in children after transcatheter device closure of atrial septal defects: A single centre experience**

Joshua D Griesman, Audrey C Marshall

Department of Paediatrics, University of Toronto and The Hospital for Sick Children, Toronto, Canada

**Background and Aim:** Transcatheter device closure (TDC) is the most common treatment for isolated atrial septal defects (ASD). The incidence of new-onset migraine headache in adults after TDC is well-recognized, and is estimated at 15%. This has led to practice change favoring post-procedural dual-antiplatelet therapy at some centres. New-onset headache is not well documented or described in children. We reviewed our single center experience to estimate the rate of headache complaints among pediatric patients undergoing TDC.

**Method:** We performed a single center retrospective review of all children undergoing TDC between January 1, 2018 and December 31, 2021. Patients under four years of age and patients with developmental delay, were excluded, based on the anticipated unreliability of headache reporting. We also excluded patients undergoing additional interventions at the index catheterization. Among included patients, we comprehensively reviewed the electronic medical record, including documentation of routine post-procedural follow-up calls and ambulatory appointments, to identify patients reporting headache.

**Results:** 165 consecutive patients underwent TDC during the study period. Of these, 134 met inclusion criteria. 20 (14.9%) patients had documented headache. Of those, 7 (5%) had persistent headaches (lasting greater than four weeks) or required further investigations. One child with persistent headache underwent brain MRI due to weeks of headache. One patient with a prior history of migraine required admission for migraine exacerbation. Another required emergency room management for status migrainosus the week following his procedure. Management approaches varied, with 3 patients having their anti-platelet changed to clopidogrel. Note, all patients with persistent headache were female.

**Conclusions:** Our study revealed new onset or worsening of persistent headaches in 5% of children who underwent TDC, in some cases requiring additional medical encounters or testing. It is important for practitioners to discuss this association prior to TDC. Determination of the true incidence will require prospective data collection.

**Keywords:** ASD, TDC, interventional, headache, migraine

#### P-469

##### **Pre-stenting with self-expandable stents in pulmonary valve replacement: A single-center experience**

Lisa Bianco, Gerard Martí Aguasca, Yassin Belahnech Pujol, Pedro Betrián Blasco

Pediatric Interventional Cardiology Unit. Vall d'Hebron University Hospital. Barcelona. Spain.

**Background and Aim:** Percutaneous pulmonary valve implantation is a known valid alternative to surgery. It is usual to place a stent prior to valve implantation (pre-stenting) in patients with more complex anatomy, especially in those with highly dilated and

distorted native outflow tract without significant stenosis/optimal anchorage area for the valve.

Most of the stents used in this scenario are balloon-expandable and require large-caliber introducers. The Sinus XL is a self-expanding stent with a high radial strength within the field of self-expanding stents and good flexibility, requiring only a 10Fr sheath.

The objective of this study is to present our experience in pulmonary pre-stenting with the Sinus XL stent.

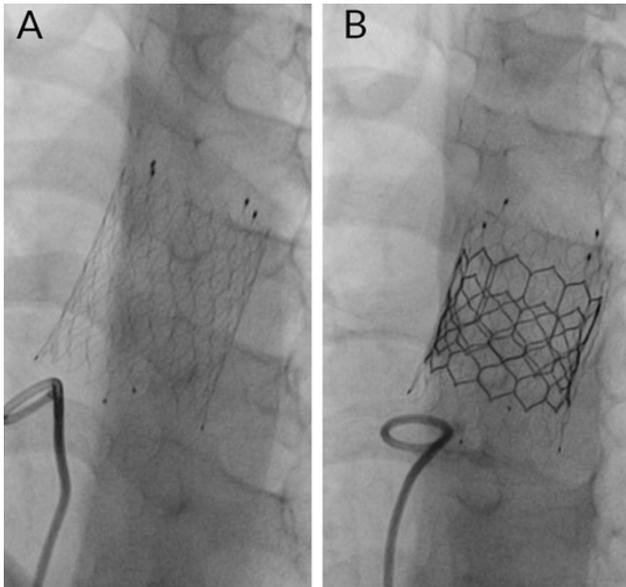
**Method:** This is a retrospective, descriptive study that includes all patients in which a pre-stenting with a Sinus XL was realized at our hospital over 5 years.

**Results:** A total of 10 Sinus XL were implanted with a wide range of sizes (30x40mm in 4 patients, 34x40mm and 28x30mm in 2 cases respectively, and 30x30mm or 28x40mm in other 1 patient each). The indication for valve replacement was severe insufficiency in the native tract in 9 patients, and severe stenosis and regurgitation in the remaining one. No complications were recorded during the implantation. The median hospital stay after the procedure was 24 (24–38) hours. No data compatible with fracture, significant displacement, or deformation of the stent were observed during follow-up. Following the pre-stenting, a 29-mm Edwards Sapien was correctly implanted in 7 patients (in 2 patients the implant took place during the first procedure, and one is still awaiting valvulation). With a median follow-up of 40 (22–47) months, patients have shown a good evolution. The valve was normally functioning in all cases, none required reintervention or hospitalization. We register only an exitus not related to the procedure in a patient suffering from an oncological illness.

**Conclusions:** In our case series, pre-stenting with Sinus XL was safe and effective, with low procedure complexity and the advantage of utilizing relatively small caliber introducers. The results support the use of the technique as a possible alternative to balloon expandable stents.

**Keywords:** pre-stenting, PPVI, Sinus XL

Pre-stenting with Sinus XL



(A) fluoroscopic image of a Sinus XL stent implanted at the level of the right outflow tract (B) image of the stent after implantation of an Edwards S3 valve

#### P-470

##### **Percutaneous vs surgical approach to the right ventricular outflow tract dysfunction. retrospective 5 years clinical and cost-analysis cohort study**

Pia Cassanello<sup>1</sup>, Juan Carretero<sup>1</sup>, Fredy Prada<sup>1</sup>, Stefano Congiu<sup>2</sup>, Joan Sanchez De Toledo<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain; <sup>2</sup>Department of Pediatric Cardiovascular Surgery, Hospital Sant Joan de Déu, Barcelona, Spain

**Background and Aim:** Over the last two decades, management of right ventricular outflow tract (RVOT) dysfunction with transcatheter pulmonary valve replacement (TPVR) has rapidly evolved, providing similar survival rates and freedom of reintervention than surgical pulmonary valve implantation (SPVI). This study aims to compare clinical short and mid-term outcomes as well as hospital-derived costs of the two procedures.

**Method:** Single-center, retrospective cohort study of children undergoing either TPVR or SPVI since implementation of the TPVR procedure in our institution in 2018. Patients' demographic and clinical data were collected including pre- and post-procedural echocardiography and cardiac MR measurements. Total hospital costs of TPVR were compared to costs of SPVI. Age, length of stay (LOS), hospital charges and lost parental wages (societal costs) were compared between groups using t test or Mann-Whitney U test, as appropriate.

**Results:** Seventy-three patients required PVR: 13(16%) TPVR and 68(84%) SPVI. TPVR patients were older than SPVI [15.9 years (14.3–17.3) vs 12.2(9.35–15.7),  $p = 0.006$ ]. At the time of the procedure, TPVR was most frequently performed in patients who had a previously valved conduit surgically implanted [7/13 (53.8%) vs 2/68 (2.9%),  $p < 0.001$ ]. Main indication for TPVR was pulmonary valve stenosis [9/13 (69.2%)] and pulmonary regurgitation (PR) for SPVI [46/68 (67.6%),  $p = 0.012$ ]. Pre-procedural cardiac MR right ventricular end-diastolic volume index was larger in the SPVI group compared to TPVR [154ml/m<sup>2</sup> (135–165) vs 133ml/m<sup>2</sup> (64–147) respectively,  $p = 0.018$ ]. TPVR implantation was successful in all patients. There was no in-hospital mortality or unplanned reinterventions in the TPVR group vs 1/68 (1.4%) and 2/68 (2.9%) respectively in the SPVI group. Median LOS for SPVI was longer than TPVR [6 days (6–8) vs 2 (2–4),  $p < 0.001$ ]. SPVI direct hospital costs were higher than TPVR [38, 079€ vs 32, 478€ respectively,  $p < 0.001$ ]. During follow-up no IE was reported. Peak RVOT gradient was significantly reduced in both groups, while moderate PR was noted in both SPVI and TPVR [5/68 (7.3%) vs 1/13 (7, 6%) respectively,  $p = 0.550$ ].

**Conclusions:** TPVR is a safe and less invasive alternative that provides similar excellent short-midterm clinical outcomes and entails a significant cost advantage over the surgical approach.

**Keywords:** transcatheter valve implantation, congenital heart disease

#### P-471

##### **Use of dobutamine to assess haemodynamic significance in borderline coarctation of aorta.**

Charalampos Kotidis, Rizwan Rehman, Atul Kalantre, Marinos Kantzis

Department of Paediatric Cardiology, University Hospitals of Leicester NHS Trust, Leicester, UK

**Background and Aim:** Current guidelines indicate that a peak to peak pressure gradient (PtP-PG) >20mmHg during invasive assessment of patients with coarctation of the aorta (CoA) is an indication for stenting. However, there are patients with systemic

hypertension or exercise hypertension with PtP-PG <20mmHg. Moreover, general anesthesia can mask borderline haemodynamically significant CoA lesions. Dobutamine administration may help assessing these borderline patients and unmask the haemodynamic impact of the CoA during stress. We report our experience using dobutamine for assessing the hemodynamic significance of CoA. *Method:* This was a retrospective observational study. All patients received general anesthesia and standard procedures were used for obtaining PtP-PG across the CoA. In patients with CoA PtP-PG <20mmHg and systemic arterial hypertension, intravenous dobutamine was administered with an aim to increase the baseline heart rate by 75% or increase systemic blood pressure by 30%. Decision for stenting was made if the PtP-PG was higher than 25mmHg. SPSS was used for the statistical analysis and median and interquartile range for descriptive statistics.

*Results:* Ten patients (six male) with [median (IQR)] age 16 years (15) and weight 56kg (24) were assessed. Dobutamine administration significantly increased the PtP-PG from 10 (14) to 20mmHg (22) when all patients analysed [ $P = 0.013$  (Wilcoxon Signed Ranks Test)]. Only two patients with baseline PtP-PG of 10 and 20mmHg increased PtP-PG above 25mmHg to 50 and 30mmHg respectively. Patients with low baseline PtP-PG had a PtP-PG increase from 8.5 (5) to 15mmHg (12) and did not receive a stent.

*Conclusions:* In a population of patients with CoA and systemic hypertension, dobutamine administration unmasked haemodynamically significant CoA in 20% of the patients. Larger prospective study with end point clinical outcomes assessment is needed to further validate our hypothesis.

**Keywords:** Coarctation of aorta, dobutamine

#### P-472

##### **Automatic processing of electronic health records to calibrate a predictive score of major adverse outcomes of patients with congenital heart disease**

Sophie Quennelle<sup>1</sup>, Sophie Malekzadeh Milani<sup>2</sup>, Hassan Faour<sup>3</sup>, Nicolas Garcelon<sup>4</sup>, Anita Burgun<sup>5</sup>, Damien Bonnet<sup>6</sup>, Antoine Neuraz<sup>5</sup>  
<sup>1</sup>Inserm, UMR\_S1138, Centre de Recherche des Cordeliers, Sorbonne Université, Paris, France; Inria, HeKA Team, PariSantéCampus, Paris, France; M3C-Necker, Hôpital Universitaire Necker-Enfants malades, Assistance Publique-Hôpitaux de Paris (AP-HP), Paris, France; <sup>2</sup>M3C-Necker, Hôpital Universitaire Necker-Enfants malades, Assistance Publique-Hôpitaux de Paris (AP-HP), Paris, France; <sup>3</sup>Data Science Platform, Imagine Institute, Université de Paris Cité, Paris, France.; <sup>4</sup>Inserm, UMR\_S1138, Centre de Recherche des Cordeliers, Sorbonne Université, Paris, France; Inria, HeKA Team, PariSantéCampus, Paris, France; Data Science Platform, Imagine Institute, Université de Paris Cité, Paris, France; <sup>5</sup>Inserm, UMR\_S1138, Centre de Recherche des Cordeliers, Sorbonne Université, Paris, France; Inria, HeKA Team, PariSantéCampus, Paris, France; Université de Paris Cité, Paris, France; <sup>6</sup>M3C-Necker, Hôpital Universitaire Necker-Enfants malades, Assistance Publique-Hôpitaux de Paris (AP-HP), Paris, France; Université de Paris Cité, Paris, France

*Background and Aim:* We present a method based on natural language processing and machine learning to automate the computation of a risk score of major adverse event in the context of cardiac catheterization. The method first automates the data collection from the electronic health records and, secondly, computes and calibrates the score on the patients of our hospital.

*Method:* We analyzed the electronic health records of patients who had been hospitalized in Necker-Enfants malades Hospital for cardiac catheterization. We used the automated extracting method provided by the Necker data warehouse, Dr Warehouse, to extract

the information required to calculate an existing score: the IMPACT-score. We developed two natural language processing algorithms to detect the data of interest in medical notes. Active learning was used to obtain a set of annotated sequences of text to train these algorithms. To illustrate the quality of our data, we computed and calibrated the IMPACT-score on the Necker population.

*Results:* We extracted clinical data from 2,980 patients who had a cardiac catheterization between 2010 and 2016. This model identified adverse events in clinical texts with an accuracy of 94.8%. In total, 211 (7.1%) patients suffered at least one procedural complication. The IMPACT model applied on our study cohort showed an area under the receiver-operating characteristic curve of 0.65 whereas it achieved an area under the receiver-operating characteristic curve of 0.76 on the IMPACT Registry cohort (i.e., in the original publication). We calibrated a logistic regression on our study cohort and the area under the receiver-operating characteristic curve reach 0.72.

*Conclusions:* The risks associated with cardiac catheterization can be automatically assessed using medical records, including clinical notes. In house calibrated scores may achieve better predictions than significantly bigger external cohorts. Further work is required to improve the performance of our model and to propose an automatized individual risk calculator for use in healthcare services.

**Keywords:** Cardiac catheterization, Congenital heart disease, Risk adjustment, Natural language processing, Electronic health records, Active learning

#### P-473

##### **Rare or frequent complication? – the need for multicentre studies and registries**

Alexandra Kiess<sup>1</sup>, Thomas Krasemann<sup>2</sup>, Franziska Markel<sup>1</sup>, Florentine Graefe<sup>1</sup>, Frank Thomas Riede<sup>1</sup>, Ingo Dähnert<sup>1</sup>  
<sup>1</sup>Department of Pediatric Cardiology, Heart Center Leipzig, Leipzig, Germany; <sup>2</sup>Division of Pediatric Cardiology, Sophia Kinderziekenhuis, ErasmusMC Rotterdam, Dept. of Pediatrics, Rotterdam, The Netherlands

*Background and Aim:* CASE: A male neonate presented on day 1 of life with a murmur which led to the diagnosis of critical aortic stenosis. Echocardiography showed a tricuspid, dysplastic aortic valve (diameter 6-7 mm, -1.8 - -0.6z) with a peak gradient of 74 mmHg (mean gradient 39 mmHg) and impaired LV-function (FS 22%). Balloon aortic valvotomy (BAV) was successfully performed with a 5 x 20 mm balloon (invasive gradient decreased from 64 to 17 mmHg). No immediate complications occurred and the child recovered. On day 14 routine echo showed an aortic wall injury with an intimal tear from the aortic valve into the descending aorta. The child was initially managed conservatively and later underwent uneventful Ross-Konno procedure for progressive aortic stenosis, as well as resection of the intima flaps. *Discussion:* To date there are only a few studies analyzing aortic wall injuries following BAV in neonates. The incidence of aortic wall injuries varied from <1% to 15% in one study where only a retrospective re-evaluation revealed the complication in more than half of the affected patients. Risk factors for complications included age, severe LV-dysfunction, novice interventionalist, and number of attempts, as well as retrograde approach.

*Method:*

*Results:*

*Conclusions:* To further analyse the main pitfalls and complications in BAV and improve the treatment strategies for critical aortic stenosis a multicentre study is needed. Therefore, we are in process to organize a Europe-wide study to investigate aortic wall injuries

and general complications and outcomes in these children following BAV.

**Keywords:** critical aortic stenosis, balloon aortic valotomy, multicentre study

#### P-474

##### Transcatheter closure of coronary artery fistula in pediatrics

Doaa Mohamed Abdel Aziz, Hala Salah Hamza, Walid Musbah Elhabeshi, Mohamed Ali Hegazi

Department of Pediatric Cardiology, Cairo University, Cairo, Egypt

**Background and Aim:** Coronary artery fistulas (CAFs) are rare congenital malformations that can be defined as direct vascular connections from a coronary artery to a cardiac chamber or major central blood vessel without an intervening capillary bed. Our aim was to review the limited available information and discuss our current management strategies for patients with this rare malformation.

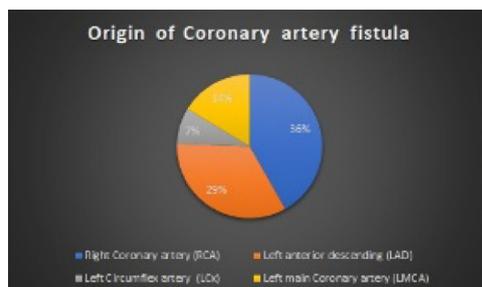
**Method:** A cross sectional observational study was conducted that included (21) patients referred to Cardiac catheterization unit - Pediatric Cardiology section - Cairo University Pediatric Hospital with suspected coronary fistula for further investigations.

**Results:** The age of the studied patients ranged from 4 Mo to 14 Yrs. Clinical examination revealed tachypnea (n = 9), murmur (n = 7), tachycardia (n = 6), congestive heart failure (n = 6). About (10) patients had abnormality in the ECG in the form of sinus tachycardia (n = 5), left axis deviation with left ventricular hypertrophy (n = 3) and ST segment abnormality (n = 2). Most of the studied patients (n = 14, (66 %)) were confirmed by cardiac catheterization to have Coronary artery fistula, while (5) patients diagnosed to have Anomalous left coronary from pulmonary (ALCAPA) and (2) patients had normal angiography. The coronary artery fistula originated in most cases from the right coronary artery (RCA) (n = 5), and left anterior descending branch of the left coronary artery (LAD) (n = 4). The fistula was found to open most commonly in the right ventricle (RV) in 7 cases (50%), the right atrium (RA) in 5 cases (36%) and the pulmonary artery (PA) in 2 cases (14%). Device closure was attempted in (7) patients (50%) and was successful in (5) patients using the Amplatzer device in (3) patients and Vascular plugs (AVPs) in (2) patients, while in (2) patients coil embolization of the fistula was not successful and the patients were referred for surgery.

**Conclusions:** Coronary artery fistula in the neonatal and pediatric age group is a rare disease and difficult to diagnose because of diversity of symptoms that can mimic other congenital or acquired heart diseases. Cardiac catheterization plays a key role in diagnosis and management of Coronary artery fistula and there is a good chance for total cure if the patients were diagnosed early.

**Keywords:** Coronary artery fistula, ST segment, Right Coronary artery, cardiac catheterization, Amplatzer device

Origin of CAFs among the studied patients



#### P-475

##### Safety and feasibility of same-day discharge after cardiac catheterization intervention in children

Stefano Di Bernardo<sup>1</sup>, Damien Schaffner<sup>1</sup>, Sylvain Mauron<sup>2</sup>, Yann Boegli<sup>2</sup>, Nicole Sekarski<sup>1</sup>

<sup>1</sup>Women-Mother-Child Department, University Hospital and University of Lausanne, Lausanne, Switzerland; <sup>2</sup>Anesthesiology and Surgery Department, University Hospital and University of Lausanne, Lausanne, Switzerland

**Background and Aim:** Nowadays, percutaneous cardiac intervention in children with congenital heart disease offers many possibilities: from the closure of shunts to stent implantation. Most of these interventions are performed in children under general anesthesia. In our institution, percutaneous intervention is performed on a same-day discharge mode, allowing shortening the hospital stay for patients and family. The scope of this study was to analyze if ambulatory catheterization is feasible without particular risks in children.

**Method:** Retrospective, monocentric study. Children with a planned elective interventional cardiac catheterization under general anesthesia were included. Outcomes were any hospitalization or consultation during the seven following days of the planned intervention.

**Results:** One hundred ninety-three patients included with a median age of 4.5 [2.2-9.2] years (range three months-16 years old), a median weight of 17 [11-27] kg. Balloon vascular angioplasty (37%) and shunt closure (36%) were the main type of interventions. Ninety (48%) patients had only venous access, 36 (19%) arterial access, and 64 (33%) both. The median intervention duration was 73 [52-94] min. Overall, 23 (12%) patients have to stay at the hospital the night after the intervention: eleven (5.6%) because the time allowed for post-intervention observation was insufficient for the type of vascular access, five (2.5%) for a complication related to the intervention itself, and seven (3.5%) for conditions related to the anesthesia. Only one patient (0.5%) had a consultation at the emergency department of another hospital not related to the intervention.

**Conclusions:** Ambulatory care for interventional cardiac catheterization is feasible in children. After the intervention, the monitoring protocol allows to identify patients with complications related to the intervention or the anesthesia, and a majority of them can go back home without particular risks.

**Keywords:** Cardiac intervention, congenital heart disease, ambulatory treatment

#### P-476

##### Balloon atrial septostomy in transposition of the great arteries with intact ventricular septum – CUI bono?

Anouska Lerner, Zdenek Slavik, Anke Furck, Melanie Holtrup

Department of Paediatric Cardiology, Royal Brompton Hospital, London, UK

**Background and Aim:** Balloon atrial septostomy (BAS) has been used in the neonates with transposition of the great arteries and intact ventricular septum (TGA/IVS) for 55 years. BAS rates vary widely between centres (1, 2). We reviewed recent indications and outcomes of BAS in TGA/IVS in a tertiary pediatric cardiac centre. **Method:** Digital patients' records were reviewed to compare data between BAS and non-BAS group. Within the BAS group further data interrogation was performed between the 'elective' and 'emergency' BAS group.

**Results:** Between 2009 and 2019, from 130 neonates with TGA/IVS admitted 76 (58%) had BAS performed. Complete data set for

review was available in 64 patients with BAS (84%). BAS was performed as an “emergency” treatment due low oxygen saturation (57 +/-12%; range 35–80%) in 41 patients (64%). In the remaining patients BAS was performed “electively” while oxygen saturation was 81 +/- 7% (range 75–90%;  $p = 0.01$ ).

No mortality was attributable to BAS. Dysrhythmia and vascular complication were present in 3 patients (13%) undergoing “elective” BAS. Prostaglandin infusion was continued for more than 24hrs after BAS in 19 patients (30%) and it had to be restarted in 30 patients (47%) due to low oxygen saturations.

**Conclusions:** Indication for BAS should be considered carefully in well neonates with TGA/IVS whose oxygen saturations are stable and can be managed by multi-disciplinary team using less invasive means or by surgery performed early given complication risks related to BAS and the fact that BAS does not guarantee subsequent improved oxygen saturations.

**Keywords:** Balloon atrial septostomy, transposition of the great arteries

#### P-477

##### **Drug eluting duct stenting in neonates is safer than mbt and central aortopulmonary shunts and of lower interventional complexity in right aortic ARCH**

Jenna Boettcher<sup>1</sup>, Daniel Biermann<sup>2</sup>, Nora Lang<sup>1</sup>, Phillip Schneider<sup>1</sup>, Michael Huebler<sup>2</sup>, Rainer Kozlik Feldmann<sup>1</sup>, Goetz Christoph Mueller<sup>1</sup>  
<sup>1</sup>Department of pediatric cardiology, University Heart Center, Hamburg, Germany; <sup>2</sup>Department of congenital heart surgery, University Heart Center, Hamburg, Germany

**Background and Aim:** Infants with duct depending pulmonary circulation are often palliated by more than 70 years known shunts to safe pulmonary perfusion after birth. Modified Blalock-Taussig (mBT) and central aortopulmonary shunts (AP) are well known surgical procedures but feared for postprocedural instability and early mortality. Duct stenting by drug eluting stents (DS) is emerging as a modern interventional alternative but of different complexity dependend on aortic arch anatomy. The purpose of this study was to compare surgical and interventional palliations and to evaluate the impact of different aortic arch anatomies.  
**Method:** In the UHZ-Hamburg 121 neonates with duct-dependent pulmonary bloodflow undergoing palliation by cardiac surgery (since 2008) and congenital interventions (since 2018) were retrospectively analyzed concerning procedures, aortic arch anatomy, hospital stay, morbidity and mortality  
**Results:** Modified Blalock-Taussig shunts were performed in 63 infants (26 female), central aortopulmonary shunts were performed in 36 infants (18 female). Duct stenting was performed in 22 patients (8 female). Survival advantage appeared in the DS group compared to surgical palliation (mortality DS 0% vs AP 2, 78% and mBT 12, 67%) combined with lower ventilation time (DS 1, 11d vs mBT 13, 1d and AP 11, 4d;  $p < 0.05$ ) and hospital stay (DS 19, 4d vs mBT 40, 9d and AP 33, 7d;  $p < 0.05$ ). Reintervention and re-surgery rate were comparable between the groups. Analysis of aortic arch anatomy in neonates receiving surgical palliation or drug-eluting duct stenting revealed no differences in mortality, hospital stay, peri- or postprocedural extracorporeal support or reintervention rate, whereas subanalysis of interventional procedure complexity showed shorter procedure time (right aortic arch 60, 9min vs left aortic arch 127, 6min;  $p < 0.05$ ) compared with less radiation exposure (fluoroscopy time 8, 6 vs 33, 9min;  $p < 0.05$ ) for neonates with duct stenting in mirror image right aortic arch anatomies compared to left aortic arch anatomies.

**Conclusions:** Ductus stenting is emerging as a serious alternative to a surgical shunt for neonatal palliation with evidence for shorter hospital stay, less morbidity and mortality. Mirror image right aortic

arch anatomy seems favourable in case of shorter procedure time and less radiation exposure compared to left aortic arch

**Keywords:** ductus stenting, drug-eluting, palliation, shunt, intervention

#### P-478

##### **Transcatheter pulmonary valve replacement after rvot reconstruction with a composite porcine valved conduit of bovine pericardium**

Jan Hinrik Hansen<sup>1</sup>, Jill Jussli Melchers<sup>2</sup>, Gunther Fischer<sup>3</sup>, Arne Martin Eide<sup>2</sup>, Tim Attmann<sup>2</sup>, Jens Scheewe<sup>2</sup>, Inga Voges<sup>1</sup>, Anselm Uebing<sup>1</sup>

<sup>1</sup>Department of Congenital Heart Disease and Pediatric Cardiology, University Hospital Schleswig-Holstein, Kiel, Germany; DZHK (German Center for Cardiovascular Research), Partner Site Hamburg/Kiel/Lübeck, Germany; <sup>2</sup>Department of Cardiovascular Surgery, University Hospital Schleswig-Holstein, Kiel, Germany; <sup>3</sup>Department of Congenital Heart Disease and Pediatric Cardiology, University Hospital Schleswig-Holstein, Kiel, Germany

**Background and Aim:** We report our experience with transcatheter pulmonary valve replacement into a composite conduit consisting of a porcine valve assembled inside a bovine pericardium tube. The tube is made from a patch and contains a longitudinal suture line. It is crimped to achieve a stable tubular configuration and is glutaraldehyde treated. Conduit degeneration has been shown to go along with heavy calcifications. The suture line might be a weak point resulting in a higher risk for rupture. Conduit rupture along the suture line has been demonstrated in ex-vivo experiments.

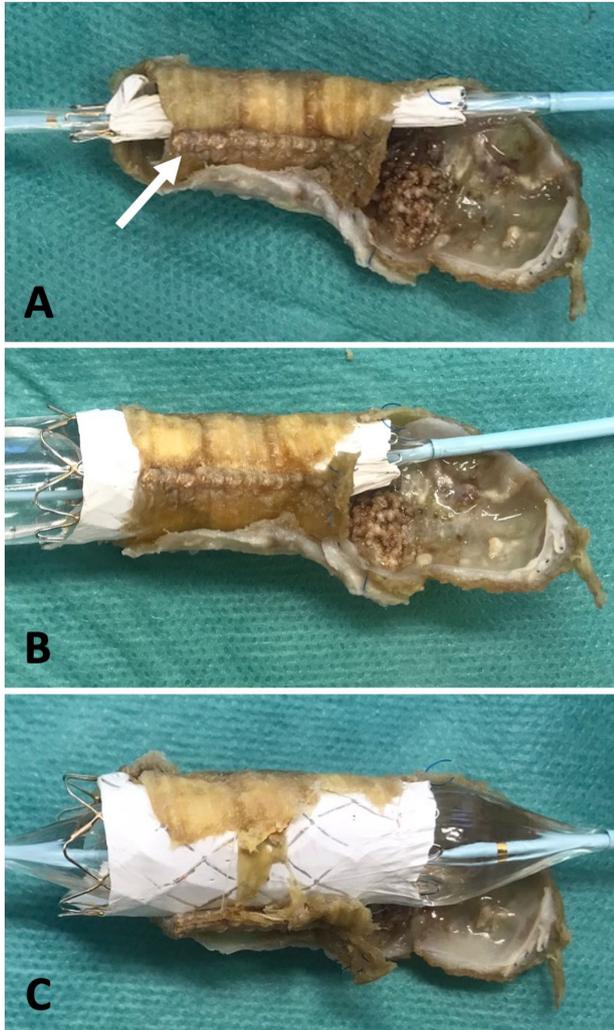
**Method:** Patient records, pre-procedural imaging and catheter reports were reviewed.

**Results:** Eight patients with a dysfunctional conduit underwent percutaneous pulmonary valve replacement at a median age of 12.2 (9.3–35.1) years. All patients had at least 2 (2–5) previous surgeries. Conduits were implanted for a median duration of 5.9 (5.0–9.4) years. Stenosis was noted in all cases with a pressure gradient of 57 (35–90) mmHg. The minimal conduit diameter was 9.5 (9.0–12.0) mm compared to the nominal diameter of 21 (15–21) mm at implantation. The first patient experienced conduit rupture during interrogation with a semi-compliant balloon evaluating for coronary compression. The rupture could be sealed with a covered stent followed by uncomplicated valve implantation. In all remaining cases, the spatial relation between coronary arteries and conduit was delineated by pre-procedural CT imaging. In amenable cases, the entire conduit including the proximal and distal anastomosis was lined-out with covered stents, which were incrementally expanded to a diameter of 18 to 22 mm. Transcatheter valve implantation with a stented bovine jugular vein graft was performed using a 20 mm or 22 mm delivery system without any periprocedural complications. The residual gradient was 10 mmHg or less in all cases.

**Conclusions:** We noted heavy calcifications and shrinkage in degenerated composite conduits consisting of a porcine valve assembled inside a bovine pericardium tube. Balloon interrogation should therefore be performed with caution. Pre-procedural cross-sectional imaging is of special importance to delineate the relationship between conduit and coronary arteries. In amenable cases, percutaneous valve implantation can be safely performed with a good result after pre-stenting of the entire conduit with covered stents.

**Keywords:** Transcatheter pulmonary valve replacement, Conduit dysfunction, Tetralogy of Fallot

## Ex-vivo Conduit rupture



Implantation of a covered stent into an surgically explanted conduit results in rupture along the suture line of the pericardial tube.

## P-479

### Outcome of valve sparing stenting of the right ventricular outflow tract (RVOT) in tetralogy of fallots and variants

Ahmed Reda Sanad Arafa Afifi, Rizwan Rehman, Chetan Mehta, Vinay Bhole, Milind Chaudhari, Oliver Stumper  
Department of paediatric cardiology, Birmingham Children's Hospital, Birmingham, UK

**Background and Aim:** Stenting of the right ventricular outflow tract (RVOT) has been established in the staged management of Tetralogy of Fallot and its variants. Our institutional practice is to avoid placing the stent across the pulmonary valve, to prevent free pulmonary regurgitation and to allow for pulmonary valve growth, potentially enabling complete valve-sparing repair.

We compare outcome data of patients with Fallot type lesions who underwent RVOT stenting depending on whether the stent placed crossed the valve (Group I) or spared the valve (Group II). **Method:** Retrospective single centre review of patients with Tetralogy of Fallot type lesions, who underwent non-randomised

RVOT stenting followed by complete repair between 2005–2021. Pulmonary valve growth was assessed by serial echocardiography. **Results:** Ninety-six patients were studied, 50 patients (52.1%) in Group I and 46 patients (47.9%) in Group II. There was no significant difference in the age and weight at stent or surgical repair in the 2 groups. Patients in Group II had more associated lesions (AVSD or DORV) [14/46 vs 5/50,  $P = 0.01$ ]. Stents used in group II were larger in diameter, median 6 mm (4–10 mm) vs 5 mm (4–8 mm),  $P < 0.01$ . There were less interstage planned or un-planned catheter reinterventions in group II, however, that was statistically not significant [18/46 (39.1%) vs 28/50 (56%)]. Pulmonary valve annulus improved from  $z$ -4.1 to  $z$ -2.87 [ $p < 0.01$ ] in the spared group, whereas there was no change in the crossed group ( $z$ -4.3 to  $z$ -4.1).

Complete valve sparing surgical repair was achieved in 5 cases (10.9%) of group II compared to none in group I [ $p = 0.017$ ]. There was no difference in the rate of trans-annular patch repair between the 2 groups. Fewer group II cases needed a surgical conduit but that was statistically not significant (21.7% vs 36%) [ $p = 0.1$ ]

**Conclusions:** Stenting the RVOT in Fallot type lesions sparing the pulmonary valve is feasible. It not only promotes growth of the pulmonary arteries but also the pulmonary valve, allowing for later valve sparing surgical correction in selected cases. Thus, this approach should be favoured.

**Keywords:** Fallots, RVOT, stenting, infants



Valve sparing RVOT stenting (A: angiography, B & C: Echocardiography)

Valve spared RVOT stenting, Angiography and Echocardiography pictures

## P-481

### Atrial unloading percutaneous shunts: new devices in congenital heart disease, a palliative technique reinvented

María De Las Mercedes Aristoy, Alvaro Lafuente Romero, Enrique Balbacid, Cesar Abelleira, Luis García Guereta, Federico Gutiérrez Laraya  
Pediatric Cardiology Department, Hospital Universitario La Paz, Madrid, Spain.

**Background and Aim:** Advanced heart failure and refractory pulmonary hypertension (PHT) are the main cause of morbidity and mortality in congenital heart disease (CHD). Therapeutic options are limited with scarce new techniques and alternatives to transplant. The creation of interatrial shunts percutaneously has been recently reconsidered as a palliative option for left and right heart failure. There are a couple of methods to keep an atrioseptostomy open: off-label use of stents or the new specific device Atrial Flow Regulator (AFR). Trials in adults endorse its safety and efficacy. But reported experience in CHD and pediatrics is almost nonexistent.

Thus the objective of our study is to evaluate its use in these populations.

**Method:** We present our experience in a single center performing long-term palliative atrioseptostomy (with device placement) in CHD and PHT. We collected clinical findings, hemodynamic data and outcomes from patients catheterized between 2013 and 2022. **Results:** A total of 10 patients were recruited (4 stents and 6 AFR). 60% of them were under 18 years old (range: 2 months - 66 years). The unloading indication was equally distributed in two groups: 50% required left chamber unloading for pulmonary congestion reduction while 50% needed right heart unloading for amelioration of systemic congestion and guarantee adequate left cardiac output (at the expense of mild cyanosis). No periprocedural complications nor mortality were reported, with adequate mid and long term results and no sequelae related to the intervention. One of the most illustrative cases was a 12 month old infant (10 kg) that required an AFR size 4 due to unmanageable right heart failure and systemic congestion. There were no technical issues and the infant showed immediate improvement. AFR showed a higher long-term permeability (100%) compared to stents.

**Conclusions:** Percutaneous atrioseptostomy is a safe and useful procedure in CHD. AFR provides a predictable and durable fenestration adapted to patient size and to specific clinical situations. Its use is feasible even in small infants.

**Keywords:** Pulmonary hypertension, Congenital heart disease, Percutaneous atrioseptostomy, Devices, Unloading

Table patients

RIGHT-LEFT SHUNT				
Diagnosis	Age	Weight	Device	Outcomes
Pulmonary atresia	23 years	65 kg	AFR 8 mm	Improvement in functional class (NYHA class II) and echocardiographic data.
Tricuspid atresia with normally related arteries - Fontan	30 years	70 kg	Formula stent 8x24 mm	Progressive improvement in functional class (II to III) and quality of life. Desaturation to 93%
Heart transplant (HT): Restrictive CMP with moderate-severe PHT.	5 years	14 kg	AFR 4 mm	Improvement of pericardial edema. Progressive healing of pulmonary vasculature. Late chronic humoral rejection. Good functional capacity.
Heart transplant (HTM). Graft failure (long ischemia, diastolic failure and tricuspid regurgitation).	12 months	10 kg	AFR 4 mm	Better edema management with possibility to decrease diuretic burden. Late death due to hepatic complications
Idiopathic pulmonary arterial hypertension.	12 years	45 kg	AFR 6 mm	Decrease in hospital admissions and syncope episodes. Bridge to lung transplant.

LEFT-RIGHT SHUNT				
Diagnosis	Age	Weight	Device	Outcomes
Shone Complex with severe PHT. Kabuki syndrome	2 months	3.8 kg	Stent 6 x 12 mm	Death by 3 years due to PHT crisis. (Good outcome)
Total supraaortic pulmonary venous anomalous connection (date surgery by 33 years with no ligation of vertical veins). Moderate PHT.	66 years	75 kg	AFR 8 mm	Progressive functional class worsening. Combined PHT, right ventricular dysfunction and severe tricuspid regurgitation. NYHA III.
Shone complex. Combined PHT and restrictive atrial septal defect.	5 months	5 kg	VASCO stent 8 x 26 mm	Death by 2 years due to PHT crisis/cardiac shock.
Neonatal coarctation, aortic stenosis, mitral anomaly with combined mild lesion and restrictive left ventricle. Combined PHT.	40 years	95 kg	AFR 8 mm	Functional class improvement (initially NYHA class III to class II) and echocardiographic and hemodynamic data.
Mitral valve combined disease. Genetic developmental disease (XO, hepatic transplant...)	23 months	9 kg	Stent 5 x 20 mm	Improvement in fluid and respiratory management. Decrease of hospital admissions.

Details of the 10 patients in whom shunt devices were implanted

**P-482**  
**Transcatheter closure of right to left interatrial shunt in children with right ventricle dysfunction and cyanosis – long-term observation**

Michał Goleczka, Sebastian Smerdzinski, Filip Tyc, Roland Fiszer  
 Department of Congenital Heart Defects and Paediatric Cardiology,  
 Faculty of Medical Sciences in Zabrze, Medical University of Silesia,  
 Silesian Center for Heart Diseases, Zabrze, Poland

**Background and Aim:** Right to left (R-L) shunt through atrial septal defect (ASD) or persistent foramen ovale (PFO) in patients with

right ventricle (RV) dysfunction due congenital RV abnormalities is a rare finding and without clear guidelines. Exclusion of such shunt terminates systemic hypoxemia, but has the potential to aggravate RV failure and decrease the systemic output. The objective is to evaluate impact on patient oxygenation and long-term results of transcatheter closure of R-L interatrial shunt in paediatric population.

**Method:** Among nearly 3500 procedures of ASD or PFO transcatheter closure performed in a single tertiary, all consecutive paediatric patients with R-L interatrial shunting were included in the retrospective analysis. Medical records, haemodynamic data, echocardiographic findings and 11.3 years observation (range: 1-14) in the out-patient clinic were obtained from our registry. Oxygen saturation of <85% at rest was considered to be contraindication to such procedure.

**Results:** Overall 7 children were included with median age of 3 years (2-13) and median weight of 16.7 kg (8.5-38). The cause of R-L shunting were: pulmonary atresia with intact ventricular septum (n = 4), severe pulmonary stenosis (n = 1), tetralogy of Fallot (n = 1), hypoplastic RV (n = 1). All but 1 patient had previous corrective surgery or transcatheter intervention performed. Cyanosis was evident in 6 patients at rest and in all during exercise. ASD in 6 and PFO in 1 patient were successfully closed with nitinol wire mesh occluders. Right atrium pressure increased insignificantly by 0.5±1.1 mmHg after closure, in none of patients systemic output decreased. Oxygen saturation increased in all patients from median 88% (85-96) to 97% (96-99) (p<0.001). Decreased exercise tolerance was noted in 4 patients before procedure and improved in 3 out of them, RV failure did not appear or aggravate in any patient. RV size did not normalize during follow-up among patients with hypoplastic RV at the time of procedure (n = 5).

**Conclusions:** Transcatheter closure of R-L interatrial shunt in appropriately selected children with RV dysfunction is a safe and effective method of restoration of normal oxygen saturation without haemodynamic compromise in long-term observation.

**Keywords:** right ventricle dysfunction, atrial septal defect, cyanosis, transcatheter closure, long-term observation

**P-483**  
**Simulating atrioventricular valves using fluid-structure interaction, a validation study against phantom experiments**

Lea Christerson<sup>1</sup>, Petter Frieberg<sup>4</sup>, Tania Lala<sup>4</sup>, Johannes Töger<sup>4</sup>, Petru Liuba<sup>2</sup>, Johan Revstedt<sup>5</sup>, Hanna Isaksson<sup>3</sup>, Nina Hakacova<sup>2</sup>  
<sup>1</sup>Children's Heart Centre of Skåne University Hospital, Lund, Sweden;  
<sup>2</sup>Department of Biomedical Engineering, Lund University, Lund, Sweden;  
<sup>3</sup>Children's Heart Centre, Skåne University Hospital, Lund, Sweden;  
<sup>4</sup>Department of Biomedical Engineering, Lund University, Lund, Sweden;  
<sup>5</sup>Department of Energy Science, Lund University, Lund, Sweden

**Background and Aim:** Children born with an atrioventricular valve defect often suffer from an imbalanced valve opening. The degree of imbalance varies and is a key factor in deciding if the child will undergo a univentricular or biventricular repair, thus it has a great impact on the short and long-term consequences for the child. Our long-term aim is to develop a computer simulation model for predicting the surgical outcome in these cases, using patient-specific flow data and valve geometries for easier treatment decision-making. Several attempts at simulating heart valves have previously been made, however, the flow is often approximated as an evenly

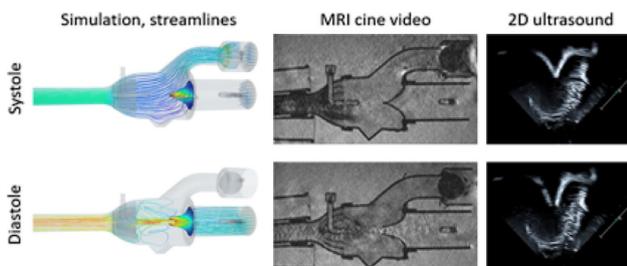
distributed force across the leaflets and only a few validation attempts have been made. Therefore, this study aims to thoroughly validate the computer model framework in a simplified scenario mimicking the left heart, against phantom data.

**Method:** The computer model was based on fluid-structure interaction (FSI) which utilizes a combination of finite element modeling and computational fluid dynamics. The experimental phantom was 3D-printed, allowing the computational model to use the same geometry. A programmable pump was used to create a pulsatile flow condition corresponding to 60 beats per minute and 4.4 l/min. Using magnetic resonance imaging (MRI), ultrasound, and pressure measurements, clinically important parameters such as velocity, pressure, and opening of the valve were measured and compared to the numeric results (Fig. 1).

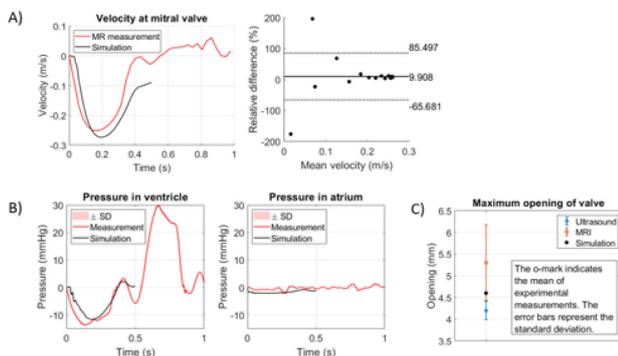
**Results:** Preliminary results of the diastolic phase (Fig. 2) show that the FSI model can capture the general behavior of the valve. The simulated velocity (Panel A) is on average overestimated by 9.9% compared to MRI measurements. Comparing simulation with experimental results obtained from pressure probes, peak pressure (Panel B) during mid-diastole differs by 15%. The simulated valve opening (Panel C) differs by 11% and 13% compared to ultrasound and MRI measurements respectively.

**Conclusions:** To date, we show that FSI models can capture the valve opening, during the diastolic phase of the heart cycle, and that peak flow and pressure differ by an acceptable error margin. This study is the first step toward predicting the surgical outcome in patient-specific valves.

**Keywords:** Fluid-structure interaction, atrioventricular valves, validation, phantom experiment, hemodynamics



**Figure 1:** Results from the computer simulation model compared to images from MRI and 2D ultrasound during peak systole and diastole.



**Figure 2:** Comparison of A) the simulated velocity against MRI measurements. Bland-Altman: bias indicated by the solid line, standard deviation indicated by the dashed lines. B) The simulated pressure against pressure measurements, and C) the simulated opening degree against simulation results and ultrasound and MRI measurements during diastole.

Figure: Comparison of simulation results against phantom data.

#### P-484

##### Covered coronary stent for implantation into the right ventricular outflow tract in infants with TOF/PA+VSD – a new idea in palliative treatment

Rafat Surmacz<sup>1</sup>, Elisabeth Beran<sup>3</sup>, Daniel Tanase<sup>2</sup>, Stanimir Georgiev<sup>2</sup>, Katarzyna Gendera<sup>2</sup>, Peter Ewert<sup>2</sup>, Jürgen Hörer<sup>3</sup>, Andreas Eicken<sup>2</sup>  
<sup>1</sup>Department of Pediatric Cardiology, Poznan University of Medical Science, Poznan, Poland; <sup>2</sup>Department of Paediatric Cardiology and Congenital Heart Defects, German Heart Centre, Munich, Germany; <sup>3</sup>Department of Cardiovascular Surgery, German Heart Centre, Munich, Germany

**Background and Aim:** Right ventricular outflow tract (RVOT) stenting is one of the palliative treatment strategies in symptomatic infants with tetralogy of Fallot (ToF) or pulmonary atresia and ventricular septal defect (PA+VSD). We present our experience in patients treated with covered coronary stent (CCS) – BeGraft (Bentley InnoMed, Hechingen, Germany) in the RVOT.

**Method:** Between 11/2017 and 07/2021, the CCS was used to widen the RVOT in 20 patients. We evaluate patients' characteristics, indications for the procedure, efficacy and safety of the catheter intervention, and additional procedures performed between stent implantation and surgical correction.

**Results:** The study group consists of 20 patients (PA+VSD – 5, ToF – 15), 6 patients with duct-dependent pulmonary circulation. The median age of the patients was 32 (4–306) days, the median body weight was 3, 0 (1, 89–6, 5) kg, and the median height was 50, 5 (42, 0–74, 0) cm. Nine patients were newborns. The diameters of implanted CCS were in a range from 3, 5x8mm to 5, 0x16, 0mm. Five patients had an additional stent(s) implanted during the initial procedure. All stents were implanted successfully. The saturation increased from a median of 81, 5 to 89, 5 ( $p < 0, 05$ ). There were three complications: RVOT perforation, stent embolization to the aorta, and pulmonary artery aneurysm. The median time of palliation was 156, 5 (50–835) days. 10 patients had an additional stent implanted in the median time of 40 (3–834) days after initial palliation. 10 patients had a later redilatation of the implanted CCS to adapt its size to the increased size of the pulmonary arteries in a median time of 226 (43–835) days. All CCS were widely open during the follow-up time. There was a significant increase in the diameters of the pulmonary artery and its branches after the procedure ( $p < 0, 02$ ). 17 patients had corrective surgery performed. Cardiac surgeons evaluated the removal of the implanted CCS as complete and easy in all cases.

**Conclusions:** Stenting the RVOT with CCS in infants with diminutive pulmonary arteries not being amenable to primary surgical correction was safe and effective, with a possibility of further redilatation. Utilization of the CCS may facilitate surgical removal of the implanted stent during the surgical correction.

**Keywords:** covered coronary stent, RVOT stenting, ToF palliation, PA+VSD palliation

#### P-485

##### Zero-fluoroscopic transeptal puncture using transesophageal echocardiography for catheter ablation of left-sided accessory pathways in children

Mustafa Gülgün<sup>1</sup>, Stefan Ostermayer<sup>1</sup>, Sevinc Özcan<sup>1</sup>, Jens Hanten<sup>1</sup>, Majed Kanaan<sup>1</sup>, Gunter Kerst<sup>2</sup>  
<sup>1</sup>Department of Pediatric Cardiology, RWTH University Hospital, Aachen, Germany; <sup>2</sup>Department of Pediatric Cardiology, Klinikum Stuttgart, Stuttgart, Germany

**Background and Aim:** Transeptal puncture (TSP) is a routine procedure to access the left atrium for catheter ablation of left-sided

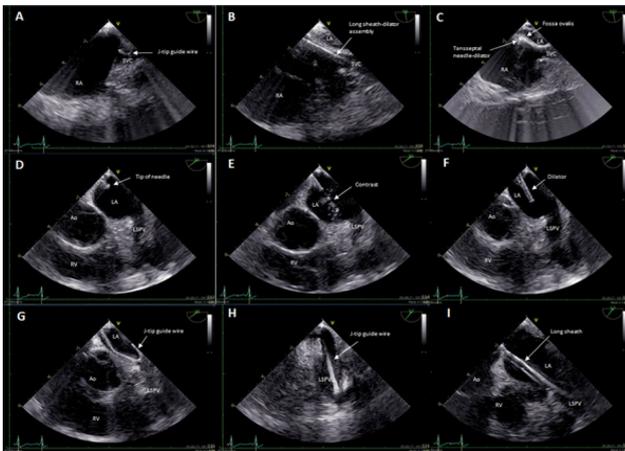
arrhythmia. However, it may still be challenging in children due to small body size, more septal flexibility and sub-optimal equipment. Long-term carcinogenic effects of X-rays request an abolition of X-rays whenever safe and feasible. The aim of this study was to investigate the safety and efficacy of transeptal punctures guided by transesophageal echocardiography (TEE) for a complete avoidance of fluoroscopy during catheter ablation of left-sided accessory pathways (APs) in children.

**Method:** A total of 63 consecutive children with left-sided AP catheter ablation between July 2016 and December 2021 were analyzed retrospectively.

**Results:** A total of 50/63 (79.4%) patients underwent TSP for left-sided catheter ablation using TEE and no or limited fluoroscopy. Mean age, weight and BMI were  $11.1 \pm 4.1$  years (range 3–19),  $47.7 \pm 20.3$  kg (range 15–88) and  $20.3 \pm 4.5$  (range 11.8–27.5), respectively. 32/50 (64%) patients were male. 28/50 (56%) patients had manifest left-sided APs. Mean total procedure time of catheter ablation was  $182.4 \pm 57.0$  min (range 75–323). Mean follow-up was  $34.6 \pm 36.0$  months (range 2–68). 28/50 (56%) patients underwent TSP with zero fluoroscopy and limited fluoroscopy was used in 22/50 (44%) patients. Transeptal access to the left atrium was performed successfully in all 50 patients. No complication occurred. Comparison of 28 patients with no fluoroscopy with the 22 patients using limited fluoroscopy showed no statistically difference in terms of acute success, recurrence and complication rate ( $p > 0.05$ ). The mean procedure time ( $161.0 \pm 52.3$  vs.  $209.7 \pm 51.70$ ) and follow-up ( $28.5 \pm 21.8$  vs.  $42.3 \pm 44.0$  months) were significantly lower in the group of patients with no fluoroscopy ( $p = 0.006$  and  $p = 0.02$ , respectively).

**Conclusions:** Transeptal puncture for ablation of left-sided APs in children using TEE and thereby completely abandoning the use of fluoroscopy appears safe and effective.

**Keywords:** transeptal puncture, children, catheter ablation, echocardiography



**Figure 1.** Sequential steps of TEE-guided transeptal puncture: A. J-tipped guidewire insertion into the SVC in bicaval view, B. Advancement of the long sheath-dilator assembly into the SVC, C. Tenting of the interatrial septum with transeptal needle-dilator assembly, D. Tenting of the needle's tip in short axis view, E. Contrast injection via the needle to confirm left atrial access, F. Dilator tip in the left atrium, G-H. J-tip guidewire placement in the left atrium and in the LSPV, I. The long sheath in the LSPV. Ao = Aorta, LA = Left atrium, LSPV = Left superior pulmonary vein, RA = Right atrium, RV = Right ventricle, SVC = Superior vena cava, TEE = Transoesophageal echocardiography.

#### P-486

##### Multicentre report on device closure of post-myocardial infarction ventricular septal rupture – in search of risk factors of early mortality

Michał Galeczka<sup>1</sup>, Filip Tyc<sup>1</sup>, Grzegorz Smolka<sup>2</sup>, Adam Sukiennik<sup>3</sup>, Maciej Chojnicki<sup>3</sup>, Jacek Kubica<sup>3</sup>, Sebastian Smerdzinski<sup>1</sup>, Małgorzata Szkutnik<sup>1</sup>, Jacek Bialkowski<sup>1</sup>, Roland Fiszer<sup>1</sup>

<sup>1</sup>Department of Congenital Heart Defects and Paediatric Cardiology, Faculty of Medical Sciences in Zabrze, Medical University of Silesia, Silesian Center for Heart Diseases, Zabrze, Poland; <sup>2</sup>Department of Cardiology and Structural Heart Diseases, 3rd Division of Cardiology, Faculty of Medical Sciences in Katowice, Medical University of Silesia, Katowice, Poland; <sup>3</sup>Department of Cardiology and Internal Diseases, University Hospital No. 1, Collegium Medicum in Bydgoszcz, Nicolaus Copernicus University in Toruń, Toruń, Poland

**Background and Aim:** Post-myocardial infarction ventricular septal rupture (VSR) is a rare and one of the most severe complications of myocardial infarction (MI). The objective is to find risk factors of early mortality (<30 days) after device closure of VSR as well as to evaluate its long-term outcome.

**Method:** Multicentre retrospective analysis of all 46 consecutive patients with device closure of VSR (percutaneous or hybrid) in years 2000–2020. Medical records, hemodynamic data, procedure results, short- and long-term ( $4.8 \pm 3.7$  years, range: 0.1–15; available in 61.7% patients) observations were analysed. All patients qualified for device VSR closure were disqualified from surgical closure or had significant residual shunt after previous VSR surgery (17.4%). Period of 21 days after VSR occurrence was considered an acute phase.

**Results:** The analysed cohort consisted of 46 patients with a median age of 67 years (40–85). Median interval between VSR occurrence to closure was 31 days (7 days – 4 years; 35% patients in acute phase). VSR was closed by percutaneous approach in 43 and by hybrid approach in 3 patients. Technical success rate was 80.4%. Major peri-procedural complications were noted in 23.9% patients (2 patients died due to further mechanical damage of left ventricle). More than moderate residual shunt was observed in 18.9% patients. Early mortality was 26.1% (18.9% in patients with successful vs. 55.6% in patients with unsuccessful VSR closure;  $p = 0.025$ ). Early surgical reintervention was necessary due to severe residual shunt in 3, device embolisation/dislocation in 2, and significant haemolysis in 1 patient. Older age, need for IABP before procedure, failure to deploy the device and severe complication were found to be the risk factors of early mortality. Three patients needed second device deployment due to residual shunt after 1–5 months. NYHA class improved in 88.2% patients at the latest follow-up.

**Conclusions:** Procedure of VSR device closure presents an acceptable technical success rate, however, number of severe complications and early mortality is high. Older patients and in poor hemodynamic condition as well as those with unsuccessful occluder deployment are particularly at a higher risk of fatal outcome. Although the clinical state of patients improves in long-term observation, reinterventions are not infrequent.

**Keywords:** acute myocardial infarction, ventricular septal defect, transcatheter closure, hybrid procedure, early mortality

#### P-487

##### Feasibility, safety and efficacy of atrial flow regulator in children: A single center experience

Elena Cuppini, Domenico Sirico, Irene Cattapan, Anna Gozzi, Francesco Prati, Biagio Castaldi

*1*Department for Women's and Children's Health, University Hospital of Padova, Pediatric and Congenital Cardiology Unit, Via Nicolò Giustiniani 2, 35128 Padova, Italy

**Background and Aim:** The Atrial Flow Regulator (AFR, Occlutech International, Helsingborg, Sweden) is a self-expandable nitinol device implanted into the interatrial septum in order to establish an interatrial communication with a predetermined diameter. The creation of such shunt is advisable in several cardiovascular diseases. In pediatric population, the experience is limited to a few case reports describing patients with failing Fontan, pulmonary hypertension or severe heart failure. We aim to report the initial single center experience of the AFR implantation in children with congenital and acquired heart disease.

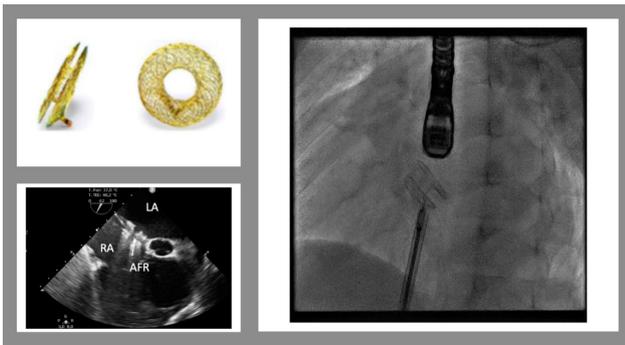
**Method:** This is a retrospective, single-center analysis of pediatric patients admitted to our institution between December 2021 to October 2022. AFR implantation was proposed for compassionate use to all patients with symptomatic heart failure, not responding to maximal medical therapy and a history of complex congenital or acquired cardiac diseases.

**Results:** We enrolled 5 patients (6 months to 12 years), indications were LV systolic dysfunction in 3 patients, restrictive cardiomyopathy with pulmonary hypertension in 1 and post-operative RV dysfunction after surgical repair of a native TOF in a 12-year-old child. AFR implantation was successfully reached in all 5 consecutive patients. In one case, the smallest child, we performed a hybrid procedure during pulmonary artery surgical banding, with peratrial approach through the right atrial free wall. Atrial Septostomy was needed in 3 cases. Balloon pre-dilation was performed in 4 cases. AFR-8mm device was implanted in all 5 cases. Mean time of procedure was 50 minutes, median fluoroscopy time was 13 minutes, median radiation exposure dose was 2.86 Gy/cm<sup>2</sup>. No complications were reported during the procedure. No shunt occlusion, stroke or new-onset right HF was observed. One patient died of sepsis not related to the procedure (16 days after the procedure); one patient presented a cardiac arrest due to a cerebral ischemia 24 hours after the hybrid procedure. Heparin or aspirin were administered to all patients for at least 6 months. Clinical improvement was observed in all patients with a reduction of LA dilation, postcapillary pulmonary hypertension, and HF symptoms.

**Conclusions:** AFR implantation is safe and feasible in children with specific congenital and acquired heart diseases, allowing right/left cavities unloading and improvement of hemodynamics and symptoms.

**Keywords:** AFR, interatrial shunt device, cardiomyopathy, chronic heart failure

AFR Elena CUPPini



## P-489

### Case series on transcatheter closure of large perimembranous ventricular septal defects during charity medical missions

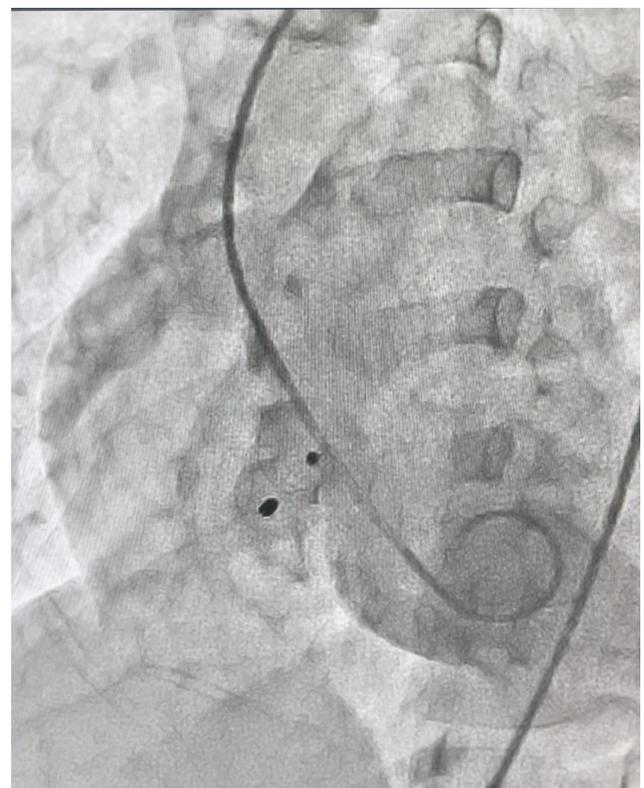
Ahmed Adel Hassan<sup>1</sup>, Mahmoud Abdul Latif Alsoufi<sup>1</sup>, Mohamed Kasem<sup>1</sup>, Obaid Aljassim<sup>2</sup>

<sup>1</sup>Pediatric Cardiology Department Heart Center of Excellence, Al Jalila Childrens Specialty Hospital, Dubai Academic Health Corporation, Dubai, United Arab Emirates; <sup>2</sup>Dubai Hospital, Dubai Academic Health Corporation, Dubai, United Arab Emirates

**Background and Aim:** Transcatheter Perimembranous VSD (PmVSD) Device closure is relatively a safe solution to prevent pulmonary overflow and its sequelae (pulmonary hypertension, heart failure, failure to thrive). It provides a shorter hospital stay and procedure time with lower cost compared to surgical closure. During charity missions the aim is to treat more unwell patients utilizing limited resources and time.

**Method:** We are reporting two patients with large PmVSD that underwent successful transcatheter closure during the charity mission held by NABADAT initiative under the umbrella of Mohammed Bin Rashid Al Maktoum Humanitarian and Charity Establishment. Patients were given a trial of catheter closure as there was no available surgical slots. Procedure done under deep sedation. We used single plane Fluoroscopy and transthoracic echocardiogram. LV angiogram done in left anterior oblique projection to delineate the PmVSD. Device type and size selection was based on both echocardiographic and angiographic measurements. In both patients we used MemoPart™ Patent Ductus Arteriosus (PDA) Occluder.

**Results:** Large PmVSD was closed successfully in 2 patients with no procedural or short term complications. First patient was 16 years



Post deployment LV angiogram

old boy, 40 Kg, his PmVSD was closed using (PDA) Occluder 18/16. Second patient was 4 years old girl, 15 Kg, her PmVSD was closed using (PDA) Occluder 16/14. Patients showed clinical improvement post procedure. Echocardiogram next day showed stable device with no leak in the first patient, and minimal residual leak though the device in the second patient. ECG done for both of them next day and showed normal sinus rhythm. Both patients were discharged on Aspirin 5mg /kg daily for 6 months

**Conclusions:** Transcatheter PmVSD device closure for large defects using large PDA occluder device provides a safe and effective solution in selected cases in immediate and short term follow up. It can be a good alternative to surgery during charity missions to allow more surgical slots for other complex cardiac anomalies.

**Keywords:** PmVSD, Charity Missions, Transcatheter PmVSD device closure

#### P-490

##### **Total repair of pulmonary atresia with major aortopulmonary collateral arteries, diminutive pulmonary artery branches after endovascular interventions**

Oleksandra Motrechko<sup>1</sup>, Anzhelika Mykhaylovska<sup>2</sup>, Anna Pavlova<sup>2</sup>, Andriy Maksimenko<sup>1</sup>, Ilya Yemets<sup>3</sup>

<sup>1</sup>Department of interventional Cardiology, Ukrainian Children's Cardiac Center, Kyiv, Ukraine; <sup>2</sup>Department of Pediatric Cardiology, Ukrainian Children's Cardiac Center, Kyiv, Ukraine; <sup>3</sup>Department of Cardiac Surgery, Ukrainian Children's Cardiac Center, Kyiv, Ukraine

**Background and Aim:** This prospective study examined the effect of an staged endovascular interventions on growth of diminutive pulmonary artery branches (DPAs) sufficient for total repair in patients with pulmonary artery atresia type I with ventricular septal defect, major aortopulmonary collateral arteries and diminutive pulmonary artery branches (PA I/VSD/MAPCAs/DPAs).

**Method:** From 2018 to 2022 staged endovascular optimization of pulmonary blood flow was performed in our center in four patients with PA I/VSD/MAPCAs/DPAs. Average indicators before the first intervention: Nakata index –  $57.4 \pm 25.8$  mm<sup>2</sup>/m<sup>2</sup>, McGoon index –  $0.96 \pm 0.4$ . The native pulmonary artery supplied blood to  $12 \pm 2.6$  segments on average. As the first stage, all patients (underwent perforations and balloon pulmonary valvuloplasty (BPV). The median of age at the time of the BPV was 4, 4 months (from the 1st to 9th months). Further, repeated BPV. In cases where the RVOT was less than 2, 5 mm, or the Z-score of the pulmonary artery valve was less than -5, the RVOT stenting was performed. MAPCA embolization were performed too. Embolization of MAPCAs was necessary to eliminate competing blood flow, ensure antegrade filling of the communicating MAPCAs from the native pulmonary artery and formation of developed native pulmonary blood flow.

**Results:** All 4 patients achieved total repair. The median of age at the time of the total repair was 27, 5 months (from the 12th to 34th months). The average Nakata index increased from  $57.4 \pm 25.8$  mm<sup>2</sup>/m<sup>2</sup> to  $160 \pm 42.5$  mm<sup>2</sup>/m<sup>2</sup>, the McGoon index increased from  $0.96 \pm 0.4$  to  $1.8 \pm 0.5$ . At the time of total repair, the native pulmonary artery supplied blood to  $16 \pm 2.5$  segments on average. The average pressure in the right ventricle 6-12 months after the total repair was 40-50% of the systemic arterial pressure, and the pressure in the pulmonary artery was 25-30 mm Hg.

**Conclusions:** Through a series of consecutive endovascular interventions in patients with pulmonary artery atresia type I with ventricular septal defect, major aortopulmonary collateral arteries and diminutive pulmonary artery branches it is possible to ensure the growth of the pulmonary arteries allowing complete repair, without any open palliative cardiac surgery.

**Keywords:** Diminutive pulmonary artery branches, endovascular optimization of pulmonary blood flow

#### P-491

##### **Initial single-centre experience with the optimus stent family in paediatric cardiology**

Katarzyna Gendera, Andreas Eicken, Peter Ewert, Dunja Renner, Kristina Borgmann, Judith Schopen, Stanimir Georgiev, Rafal Surmacz  
<sup>1</sup>. Department of Paediatric Cardiology and Congenital Heart Defects, German Heart Centre Munich, Technische Universität München, Munich, Germany

**Background and Aim:** A perfect stent for small, but also for adolescent patients with congenital heart diseases should be implantable through a small sheath, should have enough radial force to deal with even long-segment and tortuous vessels narrowing, and should be dilatable to an adult size diameter in the future. The balloon-expandable AndraTec Optimus stents meet all above-mentioned requirements. This study presents the initial single-centre experience with this stent in field of congenital heart diseases.

**Method:** There were 32 patients in this study group (male:n = 18; The median age at the time of implantation was 12, 7(min.0, 3; max.66, 9) years. The median weight was 49, 5(min.5, 1; max.150) kg and the median height was 156(min.58; max.189) cm. Indications for stent implantation: CoA (n = 9); LPA stenosis (n = 5), PA stenosis (n = 2); Fontan tunnel stenose (n = 2), SVC stenose (n = 1) pre-stenting before PPVI (n = 11), fenestration closure (n = 1), interventional PCPC (n = 1). In this study group 24 bare metal stents (Optimus L, n = 5; XL, n = 9; XXL, n = 10) and 8 covered stents (Optimus covered L, n = 1; XL, n = 5; XXL, n = 2) were implanted. The length of the stents varied between min. 13mm and max. 57mm. In 5 patients stents were crimped and implanted on Powerflex balloons, in 19 on balloon-in balloons, in one on an Advance Cook balloon, in two on a Gemini balloon, in two on an Atlas balloon and in 3 patients on a Mustang balloon. In nine patients the arterial femoral access was used, in 21 Patients stents were implanted through the femoral vein and in 2 through the jugular vein. In the smallest patients it was possible to implant the Optimus stents through a 6F sheath.

**Results:** All stents were implanted successfully. One stent dislocated but it was possible to reposition it in the proper position again and in one patient the balloon ruptured at the time of implantation. There were no other complications.

**Conclusions:** Our study shows that Optimus stents (L, XL and XXL) enlarge interventional treatment possibilities in the field of paediatric cardiology. The stents are suitable for many various indications. It is also a safe and feasible management option for the smallest patients due to its low profile and wide expansion range.

**Keywords:** Optimus stent, paediatric cardiology, interventions, stent implantation

#### P-492

##### **Right ventricle outflow tract stenting in tetralogy of fallot patients as bridging to full repair during charity medical missions**

Ahmed Adel Hassan<sup>1</sup>, Mahmoud Abdul Latif Alsoufi<sup>1</sup>, Obaid Aljassim<sup>2</sup>, Mohamed Kasem<sup>1</sup>

<sup>1</sup>Pediatric Cardiology Department Heart Center of Excellence, Al Jalila Childrens Specialty Hospital, Dubai Academic Health Corporation, Dubai, United Arab Emirates; <sup>2</sup>Dubai Hospital, Dubai Academic Health Corporation, Dubai, United Arab Emirates

**Background and Aim:** Tetralogy of Fallot (TOF) is a cyanotic heart disease with a variable spectrum from mild malalignment up to pulmonary atresia. It is corrected surgically during the first year of life and if left untreated can be debilitating and fatal. During frequent charity missions it was found that some patients with TOF pass away in between visits when unoperated due to lack of surgical slots. RVOT stenting provides an effective bridging procedure allowing somatic and pulmonary artery growth. We are reporting a case series of 10 patients who underwent RVOT stenting during the charity mission held by NABADAT initiative under the umbrella of Mohammed BinRashid Al Maktoum Humanitarian and Charity Establishment.

**Method:** TOF patients that were symptomatic with frequent cyanotic spells and have no surgical repair opportunity or have small pulmonary arteries were offered RVOT stenting. Age range was 3 months to 6 years with mainly patients above 2 years of age. We used single plane fluoroscopy and all done through RJJV access. Introducer sheath sizes were 5 and 6 Fr. We used BeSmooth Peripheral Stent System (6x28) (Bentley InnoMed GmbH, Germany) and Formula® 418 Vascular Balloon-Expandable Stent (6x20) (P.O. Box 489, IN 47402-0489, USA). Procedure done under general anesthesia and all patient admitted to ICU post catheterization for slow extubation over 4 hours and IV heparin infusion with bridging of Aspirin and Clopidogrel

**Results:** All patients started with saturation in the 60s and post procedure saturation went up to 90s. All succeeded to extubation same day. All stayed in ICU for 24 hours and total of 48 hours in the hospital. Follow up after 6 and 12 months showed improvement of NYHA score, absence of spells and growth of the pulmonary arteries. 6 patients underwent surgical repair with short ICU stay and no RV failure.

**Conclusions:** RVOT stenting provides a safe bridging option and life saving procedure for older TOF patients in absence of surgical option or poor anatomy. RVOT stents improve the pulmonary

artery size and reduces ICU stay post later surgical repair which is important element during charity missions.

**Keywords:** RVOT stent, Severe TOF, Cyanotic attacks, Charity missions

#### P-493

#### Echocardiographic predictors of biventricular repair post decompression of the right ventricle in pulmonary atresia with intact ventricular septum

Abinas Sriskandarajah<sup>1</sup>, Ahmed Afifi<sup>2</sup>, Vinay Bhole<sup>2</sup>, Rami Dhillon<sup>2</sup>, Milind Chaudhari<sup>2</sup>, Chetan Mehta<sup>2</sup>

<sup>1</sup>Institute of Clinical Sciences University of Birmingham, Birmingham, UK; <sup>2</sup>Paediatric cardiology department, Birmingham Children's Hospital, Birmingham, UK

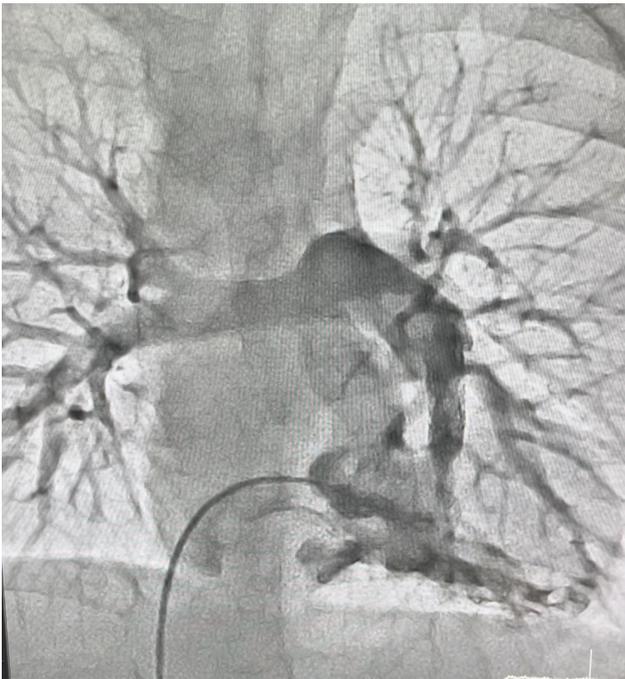
**Background and Aim:** Majority of infants diagnosed with 'Pulmonary atresia with Intact Ventricular Septum' (PA/IVS) with an imperforate pulmonary valve, undergo decompression of the right ventricle (RV) with the aim of achieving biventricular repair (BVR), despite the great morphological diversity displayed by this congenital heart defect. We aim to identify baseline echocardiographic variables which could help predict whether a patient would achieve a biventricular or univentricular circulation following RV decompression.

**Method:** A retrospective cohort study was conducted on 19 patients who underwent RV decompression via transcatheter radiofrequency perforation between 2000-2020, comparing baseline echocardiographic parameters between patients who achieved BVR following decompression against those who established a non-biventricular circulation. Univariate analysis and a receiver-operator-characteristic curve were utilised to identify baseline echocardiographic parameters predictive of BVR. Patients with an associated Ebstein's anomaly were excluded.

**Results:** The median baseline tricuspid valve/ mitral valve (TV/MV) ratio was significantly greater in patients who achieved BVR following RV decompression in comparison to those who established non-biventricular repair (1.00 (IQR 0.85-1.13) vs 0.79 (IQR 0.66-0.87),  $p=0.013$ ). A baseline TV/MV ratio  $>0.82$  was identified as a good predictor of BVR. A greater baseline severity of tricuspid regurgitation (TR) was also significantly associated with achieving BVR ( $p=0.006$ ). Lastly, a baseline tripartite RV was significantly associated with achieving BVR following RV decompression ( $p=0.038$ ).

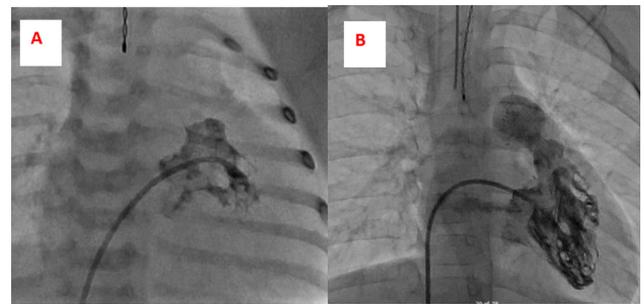
**Conclusions:** Though patients with PA/IVS with imperforate pulmonary valve undergo RV decompression, a baseline TV/MV ratio  $>0.82$ , presence of moderate-to-severe TR and a tripartite

#### RVOT stenting



RV angiogram showing the stented RVOT with good forward flow

#### PA-IVS



Patient with Pulmonary atresia + intact ventricular septum who had radiofrequency (RF) perforation of the pulmonary valve in week 2 of life (picture a, bipartite RV) and balloon redilatation at age of 2.5 years (picture b, tripartite RV).

RV are likely to achieve biventricular circulation. Further prospective research pooling a larger study population is necessary to validate the role of these echocardiographic parameters as predictors of BVR.

**Keywords:** pulmonary atresia, intact interventricular septum, right ventricle decompression

#### P-494

##### **Off-label Edwards sapien 3 ULTRA for tricuspid valve in valve implantation: A single unit experience.**

*Daniel Sew, Atul Kalantre, Marinos Kantzis*

*East Midlands Congenital Heart Centre, University Hospitals of Leicester NHS Trust, UK*

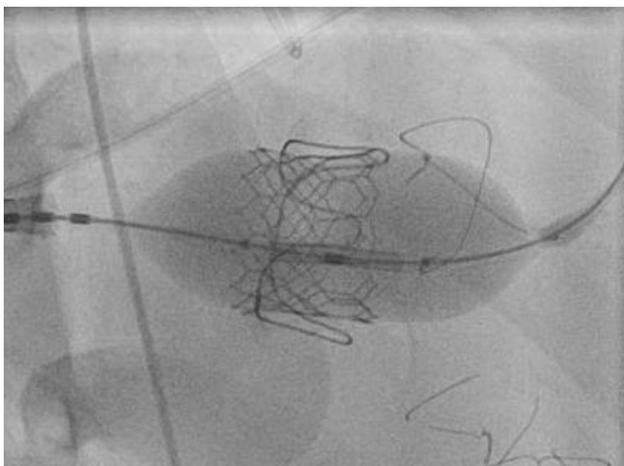
**Background and Aim:** The management of severe tricuspid valve (TV) disease remains challenging in the most experienced of centers. Since the introduction of transcatheter tricuspid valve-in-valve (ViV) implantation by Dr. Van Garsse et al in 2011, cardiac departments worldwide are reporting successful outcomes with promising short and medium-term results for high risk patients unsuitable for surgical re-intervention.

We report our experiences with TV ViV implantation (Edwards SAPIEN 3 ULTRA valve) in two young patients who have presented with significant progressive TV disease.

**Method:** The techniques utilised were similar to that of percutaneous pulmonary valve implantation (PPVI) through a transfemoral approach. For valve implantation, we used the Edwards® eSheath introducer set in one patient, whilst the other through a DrySeal® sheath. Key considerations for optimal valve alignment was a side-view-projection. To avoid paravalvular leak, we intentionally positioned the covered skirt of the new bioprosthesis partially within the right atrium.

**Results:** Both Invasive gradient post-deployment and mean gradient at short-term follow-up showed compelling results. No major complications were reported both post-procedurally and at initial follow-up. No paravalvular leak was noted. Total length of stay in the hospital was 2 days. Both patients reported notable improvement in exercise tolerance post-procedure.

Tricuspid valve in valve deployment



Final position of the new TV bioprosthesis implanted with the covered skirt extending into the right atrium

**Conclusions:** We conclude that the use of either eSheath or DrySeal sheath through the transfemoral approach are safe and sustainable for TV ViV implantation of the Edwards SAPIEN 3 ULTRA valve. The additional 40% of texture offered an easier implantation landing zone. Short term outcomes are promising with low morbidity and mortality. Further follow-up will be required for validating medium and long term outcomes.

**Keywords:** Transcatheter Tricuspid valve implantation, valve in valve implantation, Edwards SAPIEN 3 ULTRA,

#### P-495

##### **Successful rheolytic thrombectomy of near occlusive pulmonary embolism in a neonate**

*Ingrid M. Van Beynum<sup>1</sup>, Ewout P. Boesaard<sup>2</sup>, Jeroen M. Wilschut<sup>3</sup>*

*<sup>1</sup>Department of Pediatric Cardiology, Erasmus University Medical Center, Rotterdam, The Netherlands.; <sup>2</sup>Department of Pediatric Cardiology, Radboud University Medical Center, Nijmegen, the Netherlands; <sup>3</sup>Department of Cardiology, Erasmus University Medical Center, Rotterdam, The Netherlands.*

**Background and Aim:** Acute primary pulmonary embolism is very uncommon in neonates. We present a 3 weeks old male neonate, 3 kg, part of a gemelli, born at 37 weeks, with a life threatening near occlusive saddle back pulmonary embolism successfully treated with rheolytic thrombectomy with the AngioJet catheter.

**Method:** The boy presented with cardiorespiratory insufficiency at the emergency department due to a saddle back pulmonary embolism with right ventricular failure. The patient was put on (venous-arterial extracorporeal membrane oxygenation (VA-ECMO) through jugular access by the general pediatric surgeon. Systemic thrombolysis was started with recombinant tissue plasminogen activator (rTPA). After 24 hours the subtotal occlusion of pulmonary arteries remained, with dilated right ventricle with poor function and right to left atrial shunting. On VA-ECMO echo guided venous access of right femoral vein (6 Fr Merit Prelude) was obtained. Angiography with multipurpose catheter (MPA2) 4Fr in right ventricle outflow tract demonstrated complete occlusion of the right pulmonary artery (RPA) and subtotal occlusion of the left pulmonary artery (LPA). A 6Fr Launcher guiding catheter was advanced over a MPA1 4Fr and a Grandslam 0.014 coronary wire and positioned deep in the RPA and LPA, respectively. The 4 Fr AngioJet was advanced through the Launcher catheter over the coronary wire to the RPA and LPA. In total, 15 passes of with the AngioJet were performed. The patient experienced bradycardia during the sessions, which resolved spontaneously between the passes and was hemodynamically tolerated well on VA-ECMO.

**Results:** Post-intervention angiography showed patency of the major pulmonary artery branches. The patient was continued on systemic low molecular weight heparin. In two days' time the patient was weaned from VA-ECMO with recovery of the right ventricle function. Further analysis did not reveal any underlying cause for pulmonary embolism until now.

**Conclusions:** In neonate, only two case reported treatment of post-operative thrombosis of one pulmonary artery with the AngioJM. et catheter with success. This is the first neonate with a massive pulmonary embolism, of unknown cause, treated with Rheolytic thrombectomy successfully without complications.

**Keywords:** pulmonary embolism, neonate, AngioJet catheter, thrombectomy

P-496

**Purely echocardiography guided device closure of a persistent arterial duct in a 1.1kg premature**

Thomas Krasemann, Michiel Dalinghaus, Ingrid Van Beynum, Bas Rebel, Gert Van Den Berg

Department of Paediatric Cardiology, Sophia Children's Hospital, Erasmus MC Rotterdam, Netherlands

**Background and Aim:** Device closure of persistent arterial ducts in premature babies has gained acceptance as an alternative to surgical and pharmacological closure. The Amplatzer Piccolo device is used most frequently and is mainly placed under fluoroscopy, or under fluoroscopy combined with echocardiography.

**Method:** We report our experience with closure of PDAs in premature babies under 2kg. One of the babies weighing 1.1 kg showed a large PDA of 4mm diameter which could be visualized very clearly by transthoracic echocardiography.

**Results:** Device closure was performed in the cathlab (to have fluoroscopy as backup) under echocardiographic guidance alone. The catheters and wires used showed different echogenicity: Common Cordis 4F Judkins right 2.5 could be seen easily as the 2.5 Cantata microcatheter (which was used to cross valves atraumatically). Guidewires (Asai SionBlue and GrandSlam) were far less echogenic. Device deployment through a 4F Torqvew delivery catheter was well visualized.

The techniques employed were also used for subsequent premature babies in whom the combination of fluoroscopy and echocardiography were used for PDA device closure.

**Conclusions:** Knowledge of the echogenic properties of materials used in the cathlab guided the choice of catheters and wires. Atraumatic passage of the vulnerable valves can be achieved with microcatheters which can be visualized very well. Telescope techniques ease the intervention and need to be planned well in advance to be able to carry out PDA closure in premature neonates in short time.

**Keywords:** persistent ductus arteriosus, premature, echocardiographical guidance, device closure

P-497

**CMR- guided cardiac catheterization: A useful tool for the management of congenital heart disease**

Micol Rebonato, Mara Pilati, Aurelio Secinaro, Francesca Iodice, Carmelita Varano, Mattia Privato, Silvia Ceccarelli, Paolo Toma', Gianfranco Butera

Ospedale Pediatrico Bambino Gesù', Rome, Italy

**Background and Aim:** Cardiovascular magnetic resonance is an accurate imaging technique for the investigation and management of patients with congenital heart diseases. It provides accurate tissue characterization and quantification of ventricular volumes and vascular flow. However, invasive catheter derived measurement of pressures and calculation of vascular resistances are still necessary for optimum management. One way of combining the benefits of CMR with catheter-derived pressure measurement is CMR guided catheterization. Aim of this study is to describe our early experience of MR-guidance cardiac catheterization in a population of children and young adults.

**Method:** Between May 2022 and November 2022 six patients affected by complex congenital heart disease and idiopathic or secondary cardiomyopathy underwent cardiac catheterization guided by MRI. All procedures were performed in a suite equipped with a 1.5 T CMR scanner. A CMR-compatible monitor was used to mirror the scanner display for in-room catheter visualization. A single lumen balloon wedge-pressure end-hole catheter, a pigtail

catheter and the Emery Glide Straight tip MR Wire were used for all cardiac catheterizations

**Results:** A complete CMR- guided cardiac catheterization was performed successfully in six patients. Three of them were univentricular heart palliated with Glenn and Fontan, three had cardiomyopathies with various degree of pulmonary hypertension. Mean age was 15 years (6-30), mean body weight was 41 kg (17-70). Jugular or femoral venous access and arterial femoral access were obtained in all patients. Procedural time ranged between 120 and 280 minutes and became significantly shorter with increasing procedural experience. Measurements of cardiac output using phase contrast CMR and derived PVR were compared with those calculated with the Fick principle and our results showed no significant differences between measurements calculated by Fick's method and flow quantification from MRI (IC MRI median 2, 3 L/min/m<sup>2</sup>, 95%CI 1, 05-3.94; Fick IC median 3L/min/m<sup>2</sup> 95% CI 0, 29-3, 6).

**Conclusions:** This study shows that CMR guided cardiac catheterization using standard technology can be feasible in a selected group of young patients with congenital heart disease. It makes the procedure radiation-free and it is a validated method to better assess cardiac output and PVR in a variety of complex heart diseases

**Keywords:** CMR, Cardiac cath, flow quantification, derived PVR

P-498

**Transcutaneous closure of persistent ductus arteriosus: complication rates and long term follow up, a single centre retrospective study**Ian Scott Kendall<sup>1</sup>, Gail Davison<sup>1</sup>, Emma Mccann<sup>2</sup>, Oisín Fearon<sup>2</sup>, Neil Kennedy<sup>3</sup>, Brian Mccrossan<sup>1</sup><sup>1</sup>Royal Belfast Hospital for Sick Children; <sup>2</sup>Queen's University Belfast; <sup>3</sup>Royal Belfast Hospital for Sick Children & Queen's University Belfast

**Background and Aim:** Transcutaneous closure of PDA in childhood is a common cardiovascular procedure. Longer term follow-up by paediatric cardiologists is variable. Identification and classification of post-operative complications may enable targeted patient follow-up and timelier discharges. This study aimed to characterise complication rates and assess when patients can be safely discharged

**Method:** Single centre retrospective study of paediatric patients (aged 0-15 years) who underwent a transcutaneous closure of a PDA between January 2006 and December 2015. Data was gathered from Heartsuite©. Patients who had significant additional congenital heart disease or those aged 16 years or more were excluded. Complications were classified according to severity and timing of identification (1) low (mild), medium (possible clinical significance but no re-intervention) or high severity (re-intervention required) and (2) immediate (within 24 hours), short term (≤2 years) or late (2years).

**Results:** The mean age & weight at time of procedure was 44 (s.d.+/-33) months and 15.9 (s.d.+/-8.5, range = 3.9-80) kg. Ducts were occluded with Cook Coils (CC) = 87/156 (56%) and Amplatzer Ductal Occluders: ADO1 = 63/156 (40%) and ADO2 = 4/156 (2.5%) or a combination of ADO and CC = 2/156 (1.2%). There were no procedural deaths or significant vascular trauma.

Complications were seen in 18/156(12%). High-grade complications occurred in 8/156(5.1%): Device embolisation 3/156(1.9%), failure to close PDA 2/156(1.3%), and residual shunt requiring repeat procedure 3/156(1.9%). Moderate-grade complication 1/156(0.6%): residual aortic arch obstruction 3.1m/s. Low-grade complications 9/156(6.4%): mild left pulmonary artery obstruction 3/156(2.6%) and mild aortic turbulence 6/156(3.8%).

Immediate complications were evident in 14/156(9%), short term 1/156 (0.6%), late 3/156(1.9%) -Mild LPA turbulence 2.3m/s & 3 patients with mild aortic obstruction ( $V_{max} = 2.5-2.9\text{m/s}$ ). Late obstruction was seen in the ADO group only. Average follow-up time for all patients was 81(+/-47) months.

**Conclusions:** PDA occlusion is associated with a not insignificant major complication rate (5.1%) which is evident within 24 hours. However, a further 2.5% (all ADO1 occluders) developed between mild and moderate LPA/ aortic obstruction at least 1 year post procedure, which to date has not required intervention. It may be prudent to continue longer term surveillance of patients who have undergone PDA occlusion with the ADO 1 device

**Keywords:** PDA, Transcatheter Closure, Complications, Follow-up

#### P-499

##### **Initial experience with venous p valve implantation: the difficult way to find the right patient.**

Mara Pilati, Veronica Bordonaro, Micol Rebonato, Roberto Formigari, Aurelio Secinaro, Enrico Piccinelli, Gianfranco Butera  
Ospedale Pediatrico Bambino Gesù

**Background and Aim:** Venous P Valve is a self-expandable valve designed for native outflow tract valvulization. Characteristics and dimensions of the valve will permit to expand the numbers of patients candidable to percutaneous pulmonary valvulization. Worldwide clinical Experience with this valve is just beginning with encouraging results but criteria of eligibility are still restrictive. We are presenting our initial experience with the use of Venous P valve in a group of patients with native outflow tract dysfunction.

**Method:** From June to November 2022, 7 patients (median age 19 years, range 16-24 years; 4/3 male/female) were selected for Venous P valve implantation on the basis of a significant pulmonary valve insufficiency and enlarged outflow tract (RVOT > 29 mm). Patients with pulmonary artery stenosis were excluded. All the patients underwent MRI, CT and/or catheterization and all the images were sent to an Imaging review board of the manufacturing company and only the patients that met all the inclusion criteria were implanted.

**Results:** Among 7 patients, 3 (43%) were considered not eligible for Venous P Valve implantation. The exclusion criteria were: RVOT conical shape with a diameter more than 34 mm, pulmonary arteries under-over and proximity of the left coronary artery to the RVOT. One patient was accepted for Venous P valve implantation but was excluded at the time of the procedure because of the under-over appearance of pulmonary arteries not detected at angio CT. Two patients underwent Venous P valve implantation. Both patients were affected by Tetralogy of Fallot and were corrected with transannular patch. At sizing balloon, maximum RVOT diameters were 31 mm and 29 mm, respectively. In both patients, a Venous P Valve 34/25 mm was implanted with good result and without complication. Two patients are on a waiting list.

**Conclusions:** Percutaneous pulmonary valvulization with Venous P valve is a very good option for large native RVOT and in the future it will enrich the already available armamentarium for the transcatheter treatment of dysfunctioning RVOT. Hopefully the increasing worldwide experience will lead to less selective inclusion criteria.

**Keywords:** Right ventricular outflow tract, Venous P Valve

#### P-500

##### **An unusual complication post neonatal aortic balloon valvuloplasty**

Stavros Christoforides, Mark Danton, Patrick Noonan  
Royal Hospital for Children - Glasgow - UK

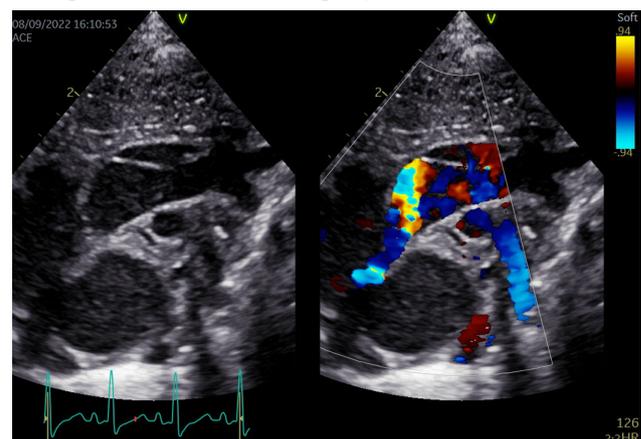
**Background and Aim:** Aortic dissection following balloon aortic valvuloplasty is a reported but rare procedural complication. We describe a case of a 3-week-old infant who sustained a massive aortic dissection after balloon dilation of the aortic valve for severe aortic stenosis associated with bicuspid aortic valve.

**Method:** A term male infant with bicuspid aortic valve and severe stenosis was planned for aortic balloon valvuloplasty. Patient weighed 3.6 kg and aortic annulus measured 7mm on echocardiography (8mm on angiography). At catheterisation the valve was crossed with a hydrophilic wire and dilated with 7mm angioplasty balloon. The balloon was inflated by hand to allow rapid deflation. Invasive gradient was reduced from 50mmHg to 18mmHg but there was moderate-severe aortic regurgitation on the post-balloon angiogram. A sizeable dissection flap was seen originating 19mm above the aortic valve and extending to origin of brachiocephalic artery. Echo and CT findings were consistent with angiogram and the flap prolapsed through the aortic valve during diastole. There was a moderate degree of obstruction seen in the aortic arch due to the flap. The patient remained clinically well at all times and treatment options were considered. As the valve was not repairable, the patient went for Ross procedure. The aorta was dilated with a segment of denuded intima that required replacement with a short ascending interposition graft of non-valved contegra(12mm). Patient recovered well following surgery and was discharged home on post-operative day 12.

**Results:** Aortic dissection has been reported as a complication of balloon aortic valvuloplasty. The reported incidence appears higher than has been noted with local experience, however dissection of this magnitude is extremely rare. Genetic associations with bicuspid aortic valve are uncommon and no abnormality has been identified on aortopathy panel. The aforementioned surgical solution has achieved an excellent short-term result, but this will need careful surveillance in the future.

**Conclusions:** Aortic injury post balloon aortic valvuloplasty is a rare but possibly under-recognised complication. Dissection of this

Echo post balloon aortic valvuloplasty



A large intimal flap is seen at the distal ascending aorta

magnitude has not been described to date and raises questions about associated pathology however the cardiac surgical repair performed in our patient has been successful in the short term but will require careful life-long surveillance.

**Keywords:** balloon aortic valvuloplasty, congenital aortic stenosis, aortic injury, aortic dissection

#### P-501

##### Single ventricle patients with and without interventional mapca closure: comparison of pulmonary artery dimension, oxygen saturation and hemodynamics

Alessia Callegari<sup>1</sup>, Walter Knirsch<sup>1</sup>, Robert Cesnjevar<sup>2</sup>, Hitendu Dave<sup>2</sup>, Oliver Kretschmar<sup>1</sup>, Daniel Quandt<sup>1</sup>

<sup>1</sup>University Children's Hospital Zurich, Pediatric Heart Centre – Division of Pediatric Cardiology, University of Zurich; Switzerland; <sup>2</sup>University Children's Hospital Zurich, Pediatric Heart Centre – Division of Congenital Cardiothoracic Surgery, University of Zurich; Switzerland

**Background and Aim:** Comparison of single ventricle (SV) patients with (group 1) and without (group 2) interventional closure of MAPCAs after stage I or/and II of SV palliation.

**Method:** Single center, retrospective (2006–2021) longitudinal study on 136 SV patients (exclusion criteria: congenital abnormalities of the PAs, pulsatile partial BDCPC). Patient characteristics, oxygen saturation (SpO<sub>2</sub>) pre stage II, pulmonary artery dimensions and invasive hemodynamics (both assessed during cardiac catheterization pre stage II and pre stage III), and their surgical procedures were compared between the two groups. Median follow-up was 7.17 (3.25–9.74) years after stage III.

**Results:** A total of 52/136 (38%) patients underwent MAPCA closure after stage II or/and III, with 14 patients (10%) having had several interventional procedures with occlusion of MAPCAs. 45/136 (33%) underwent MAPCA closure after stage II and 17/136 (12%) after stage III. Male patients ( $p = 0.04$ ), patients with single right ventricle ( $p = 0.04$ ) or a Giessen approach at stage I ( $p = 0.02$ ) had higher incidence of MAPCA closure after stage II. Patients that had whether surgical nor interventional procedure at stage I had less frequent MAPCA closures after stage II ( $p = 0.03$ ). Patients with MAPCA closure after stage II ( $p < 0.001$ ) and stage III ( $p = 0.01$ ) had lower SpO<sub>2</sub> prior to stage II. Patients with smaller central pulmonary arteries pre stage II (McGoon  $p = 0.04$ ; Nakata  $p = 0.05$ ) or a larger ascending aorta (mm/BSA) ( $p = 0.03$ ) had more frequent interventional MAPCA closures after stage II. SV-EDP, mPAP, tPG, and Qp:Qs assessed prior to stage II were similar in group 1 and 2. Patients with smaller central and peripheral pulmonary arteries pre stage III (McGoon  $p = 0.001$ ; Nakata  $p = 0.003$ ; Lower-lobe-Index  $p = 0.02$ ) had more frequent interventional MAPCA closures after stage III and/or II. SV-EDP ( $p = 0.01$ ), mPAP ( $p = 0.05$ ), and tPG ( $p = 0.02$ ) prior to stage III were higher in group 1. Creation of a fenestration at stage III was more frequent ( $p < 0.001$ ) in group 1.

**Conclusions:** MAPCA closure is frequent in this cohort of SV patients, especially after stage II. The presence of small pulmonary arteries and hypoxemia might lead to MAPCAs formation after stage II and stage III of single ventricle palliation.

**Keywords:** Single ventricle, MAPCAs, hemodynamic, Fontan palliation

#### P-502

##### Comparison of hemodynamic parameters and pulmonary artery dimension in single ventricle patients with and without closure of veno-venous collaterals

Alessia Callegari<sup>1</sup>, Walter Knirsch<sup>1</sup>, Robert Cesnjevar<sup>2</sup>, Hitendu Dave<sup>2</sup>, Oliver Kretschmar<sup>1</sup>, Daniel Quandt<sup>1</sup>

<sup>1</sup>University Children's Hospital Zurich, Pediatric Heart Centre – Division of Pediatric Cardiology, University of Zurich; Switzerland; <sup>2</sup>University Children's Hospital Zurich, Pediatric Heart Centre – Division of Congenital Cardiothoracic Surgery, University of Zurich; Switzerland

**Background and Aim:** Comparison of single ventricle patients with (group 1) and without (group 2) interventional closure of veno-venous collaterals (VVCs) after stage I or/and II of single ventricle (SV) palliation.

**Method:** Single center, retrospective (2006–2021) longitudinal study on 136 SV patients (exclusion criteria: congenital abnormalities of the PAs, pulsatile partial BDCPC). Patient characteristics, invasive hemodynamics and pulmonary artery dimensions (both assessed during cardiac catheterization pre stage II and pre stage III), and their surgical procedures were compared between the two groups. Median follow-up was 7.17 (3.25–9.74) years after stage III.

**Results:** A total of 30/136 (22%) patients underwent VVC closure after stage II or III, with 10 patients (7%) having had several interventional procedures with occlusion of VVCs. 25/136 (18%) patients underwent VVC closure after stage II and 10/136 (7%) after stage III. Patient characteristics were similar between group 1 and 2. Patients with smaller central and/or peripheral pulmonary arteries in the cardiac catheter assessments pre stage II (McGoon  $p = 0.03$ ; Nakata  $p = 0.01$ ; LPA/m2BSA  $p = 0.002$ ; RPA/m2BSA  $p = 0.03$ ; Lower-lobe-Index  $p = 0.004$ ) or a wider ascending aorta (mm/BSA) ( $p = 0.01$ ) had more frequent interventional VVC closures after stage II and/or III. Similarly, patients with smaller central and/or peripheral pulmonary arteries in the cardiac catheter assessments pre stage III (McGoon  $p = 0.005$ ; Nakata  $p = 0.009$ ; LPA/m2BSA  $p = 0.002$ ; RPA/m2BSA  $p = 0.007$ ; Lower-lobe-Index  $p < 0.001$ ) had more frequent VVCs closures after stage III and/or II. Prior to stage II SV-EDP, mPAP, and tPG were similar between the groups. Prior to stage III tPG was lower ( $p = 0.04$ ) in group 1, while SV-EDP and mPAP were similar between the groups. Creation of a fenestration at stage III was more frequent ( $p < 0.001$ ) in group 1. Patients that had VVC closure after stage II had smaller diameter of extracardiac Fontan conduits ( $p = 0.03$ ) but similar age at stage III ( $p = 0.1$ ).

**Conclusions:** Smaller pulmonary arteries prior to stage II and III seem to relate to VVC formation. In patients after VVC closure special attention should be drawn on further pulmonary artery development and Fontan hemodynamics.

**Keywords:** Single ventricle, VVCs, hemodynamic, Fontan palliation

#### P-503

##### Pediatric transcatheter closure of multi fenestrated secundum atrial septal defects

Francesco Prati, Domenico Sirico, Elena Cuppini, Jennifer Fumanelli, Angela Di Candia, Biagio Castaldi, Giovanni Di Salvo  
Department for Women's and Children's Health, University Hospital of Padova, Pediatric and Congenital Cardiology Unit

**Background and Aim:** Atrial septal defects (ASDs) are among the most common congenital heart diseases. Transcatheter closure is

nowadays considered as the first line treatment for ostium secundum ASDs, however the implanting technique remains challenging in complex anatomies. In particular there is still debate regarding the use of single vs. multiple devices implantation for closure of multifenestrated ASDs.

The aim to evaluate the efficacy and the safety of multifenestrated ASDs closure with single or double devices implantation.

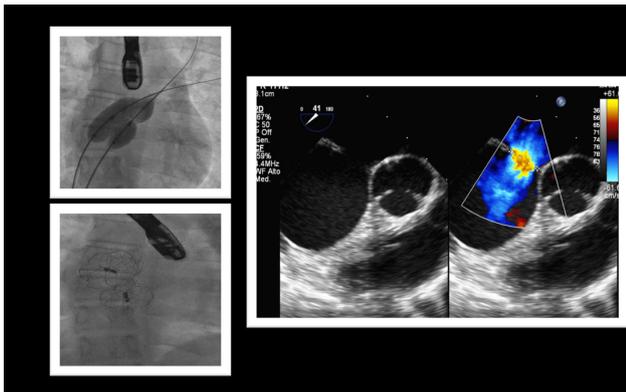
**Method:** Between January 2020 and May 2022, 110 patients with haemodynamically significant ASD underwent transcatheter closure at the pediatric cardiology unit in Padua. Pre-operative evaluation was based on 2D and 3D transthoracic (TTE) and transesophageal (TEE) echocardiography. Procedure was guided by intra-operative TEE and static balloon sizing, while residual shunt was assessed post-operatively with 2D-TTE, 24 hours and 6 months after device placement. The devices were selected on the basis of ASD anatomic features among GORE® Cardioform ASD Occluder (GCA), GORE® Cardioform Septal Occluder (GSO), Amplatzer® septal occluder (ASO) and Occlutech® Figulla Flex II (FFII).

**Results:** The procedure was successful in all 110 patients. ASO device was used in 17 cases, FFII in 24 cases, GSO in 33 cases and GCA in 36 cases. Sixteen patients (14, 5%) presented a multifenestrated ASD. In these cases two devices were used in 4 cases (GSA+GSO in all cases). In the remaining 12 cases closure was obtained with a single device (5 GCA 42%; 7 GSO 58%) by slightly oversizing the device in the main defect. No significant complication was reported in our cohort. The prevalence of residual shunt at 24 hours after device implantation was 18, 7% in multifenestrated vs. 3% in single ASDs. Six months after the procedure no residual shunt was detected in both groups.

**Conclusions:** In pediatric patients, closure of multifenestrated ASDs was achieved with a single device in approximately 75% of cases thanks to reduced septum dimensions. When this approach is not sufficient, the implantation of a second smaller device represents a safe and effective option to correct these defects.

**Keywords:** Atrial septal defect, ostium secundum, double device, transcatheter closure, multiple atrial septal defects

#### Double device closure of atrial septal defect



#### P-504

##### Predictors of left ventricular hypertrophy at long-term follow-up after effective stent implantation for aortic coarctation

Isabel Sampaio Graca, Miguel Mata, Maria Ana Esteves, Joao Rato, Susana Cordeiro, Mafalda Sequeira, Rui Anjos

Department of Pediatric Cardiology, Santa Cruz Hospital, Lisbon, Portugal

**Background and Aim:** Left ventricular hypertrophy (LVH), defined echocardiographically as an increased LV mass index (LVMI), is a well-established risk factor for cardiovascular mortality and morbidity, as well as a marker for arterial hypertension-mediated organ damage. We evaluated LV mass at long-term follow up of effectively stented aortic coarctation patients, assessing possible targets to decrease this risk factor.

**Method:** Study population included 86 patients with aortic coarctation and no significant aortic valve disease, who had undergone stent implantation, with a follow up of 2 to 24 years (mean 11.5 years). Evaluation included clinical data, transthoracic echocardiogram, office blood pressure (BP) measurement, 24-hour BP monitoring. LV mass was measured echocardiographically as per published guidelines and indexed to body surface area. Simple linear regression was used to assess correlations for LV mass. Significant variables were used to build a multivariable model.

**Results:** At the time of stenting, 59 patients (69%) had native coarctation; mean age was 29 years (SD 15.5); 42% had bicuspid aortic valve. Invasive aortic gradient decreased from mean 42.3mmHg (SD 21.2) to 4.7mmHg (SD 6.5) immediately after stenting. There were no major complications.

At last follow-up: mean age was 40.5 (SD 15.5) and 12% were over 60 years old; all patients had an echocardiographic isthmus gradient of less than 20mmHg, mean 12.2mmHg (SD 4.8). Fifty-two (60%) patients were on anti-hypertensive medication. Mean systolic office BP was 132mmHg (SD 11); and at 24-hour monitoring was 125mmHg (SD 17.9). Mean LVMI decreased from 129.7g/m<sup>2</sup> (SD 53.6) at time of stenting, to 105.6g/m<sup>2</sup> (SD 31.9) at last follow-up; 36% had LVH as per published guidelines.

The multivariable regression model predicted LVMI using age at first stenting procedure ( $p < 0.001$ ), mean systolic BP at 24-hour Ambulatory BP Monitoring ( $p = 0.005$ ) and sex ( $p = 0.007$ ).

**Conclusions:** Despite an effective stenting procedure, patients with aortic coarctation may still have elevated LVMI at long-term follow up. Early identification and treatment of the disease is key to reduce this burden, while the 24-hour BP monitoring is the single best exam to predict LVMI, and should be used to guide treatment.

**Keywords:** aortic coarctation, stenting, left ventricular hypertrophy, arterial hypertension

#### P-506

##### Coarctation stent compression after high-velocity trauma

Kristoffer Jonathan Steiner<sup>1</sup>, Gunnar Sjöberg<sup>2</sup>  
<sup>1</sup>Department of Paediatric Cardiology, Astrid Lindgren Children's Hospital, Stockholm, Sweden; <sup>2</sup>Department of Women's and Children's Health, Karolinska Institute, Stockholm, Sweden

**Background and Aim:** We present a case of a 17 year old female patient with Noonan's syndrome. The patient underwent surgery with end-to-end anastomosis of a Coarctation in infancy.

At 16 years of age, the patient underwent stent implantation of a 12x48 mm Bentley BeGraft Aortic covered stent with complete elimination of the gradient across the re coarctation from 40mmHg to 3 mmHg. Angiography demonstrated excellent flow across the stent with no extravasation and good flow to the left subclavian artery.

**Method:** Echo follow up 3 months after implantation demonstrated good stent position with normal flow in the descending aorta. A CT scan was performed as a regular follow up 6 months demonstrated a significant compression and re coarctation in the lower half of the stent. At follow up in our outpatient clinic following

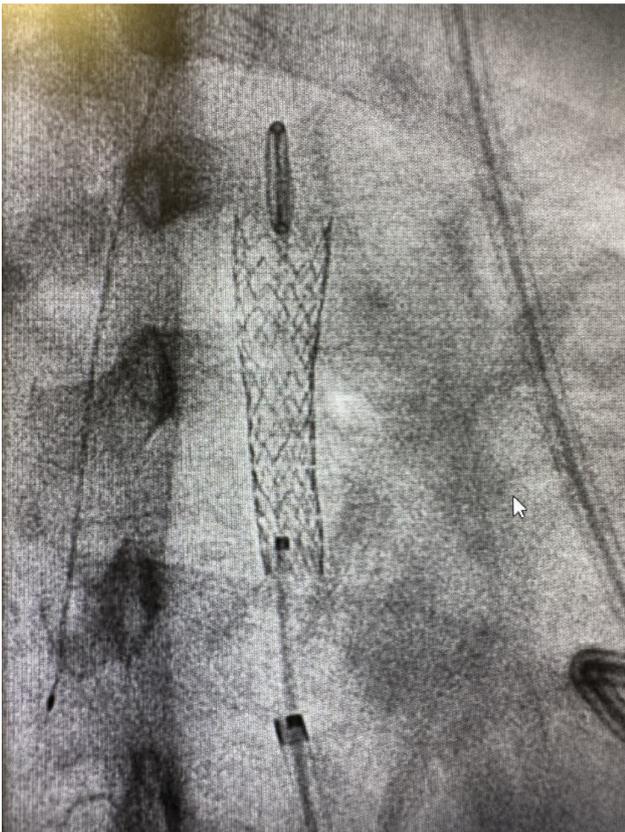
the CT scan, the patient reported two accidents with an electric scooter, the second one with high velocity when the patient crashed with the chest onto a table in an outdoor café.

**Results:** Catheterization was performed and demonstrated a significant compression anteroposterior of the stent in the lower part, the minimal diameter was 5.5x 8.2 mm. Redilation was performed with a high pressure AtlasGold 16x40mm. Angiography post dilatation demonstrated good flow with improved diameter of 12.8 x 13.4 mm. Peak-to-peak gradient was reduced from 40mmHg to 6 mmHg. Mild stent ingrowth was seen with no extravasation or stent fracture.

**Conclusions:** Stent implantation is an effective treatment for aortic coarctation. The Bentley BeGraft Aortic stent with Cobalt Chromium and ePTFE covering has a high radial force and is effective for treatment of aortic coarctation. However, reaction of the metal stent to trauma is different than in native tissue that can restore its natural form after external compression. This should be kept in consideration when informing patients of suitable activities and sports as well as the importance of investigating stents after trauma to the chest.

**Keywords:** coarctation, covered stent, compression, trauma

Stent compression trauma



#### P-507

##### Determination of factors affecting the development of aortopulmonary collaterals in patients after the glenn procedure

Jacek Kusa<sup>1</sup>, Pawel Czesniewicz<sup>2</sup>, Lukasz Chodorowski<sup>2</sup>, Elzbieta Skiba<sup>2</sup>, Dominika Gardynska<sup>2</sup>, Magdalena Slupska<sup>2</sup>

<sup>1</sup>Pediatric Cardiology Department, Medical University of Silesia, Katowice, Poland; <sup>2</sup>Pediatric Cardiology Department, Regional Specialist Hospital - Research and Development Centre, Wroclaw, Poland

**Background and Aim:** Aortopulmonary collaterals are a frequent consequence of single-chamber tract procedures: Glenn and hemiFontan. In the immediate postoperative period, they can be an important adaptation factor, but they can also cause problems during the completion of the Fontan procedure. The aim of this work is to determine the hemodynamic and echocardiographic factors affecting their formation.

**Method:** 98 patients aged 3, 6 ± 3, 4 years (range: 0, 8-17, 8) with single ventricle anatomy after Glenn shunt were analyzed. They underwent cardiac catheterization from 2013 to 2019 at our institutions. We identified 82 significant aorto-pulmonary collaterals in 60 patients. All collaterals were embolized. Imaging and hemodynamic data were compared in two groups: patients with aortopulmonary collaterals versus patients without aorto-pulmonary collaterals.

**Results:** Of the many factors analyzed, only 3 were statistically significant. Echocardiographic measurements did not reveal any group differences. Pulmonary arteries were equally developed in both groups. Patients with aorto-pulmonary collaterals had lower mean pulmonary artery pressure (13, 1 mmHg vs. 15, 3 mmHg, p<0, 05) and end diastolic pressure (10, 65 mmHg vs. 12, 18 mmHg, p<0, 05). They had also lower pressure gradient between ascending and descending aorta (3, 4 mmHg vs. 6, 7 mmHg, p<0.05). Patients without aorto-pulmonary collaterals and with higher pressure gradients in aortic isthmus had more often aorto-aortic collaterals. The remaining haemodynamic parameters did not show statistical significance.

**Conclusions:** Hypoperfusion of the pulmonary segments has the most significant effect on the development of the aortopulmonary collaterals.

We also hypothesize that these vessels more often develop with laminar flow through the neoisthmus of the aorta. Collateral circulation in the case of stenosis at the level of the neoisthmus is most often formed from the same vessels and this is an element of competition.

**Keywords:** Glenn, complication, collaterals

#### P-508

##### Results of periventricular device closure of ventricular septal defects in children without cardiopulmonary bypass.

Shravan Kanaparthy<sup>1</sup>, Shreesha Maiya<sup>1</sup>, Sudesh Prabhu<sup>2</sup>, Suresh Pv<sup>1</sup>, Prakash Ramachandra<sup>1</sup>, Sathesh Siddaiah<sup>1</sup>

<sup>1</sup>Department of Paediatric Cardiology, Narayana Institute of Cardiac Sciences, Narayana Health, Bengaluru, India; <sup>2</sup>Department of Paediatric Cardiac Surgery, Narayana Institute of Cardiac Sciences, Narayana Health, Bengaluru, India

**Background and Aim:** Open surgical and percutaneous transcatheter closure of Ventricular septal defects (VSD) are two well-known methods. Off-late, a new hybrid approach, periventricular closure of VSDs is gaining importance given the benefits it has over both these procedures. We analyzed the interventional results after periventricular device closure (PVDC) of VSD without cardiopulmonary bypass (CPB) in children and compared with standard surgical closure of VSD under CPB.

**Method:** It was a prospective and retrospective observational study (Sep 2017-August 2022). Children with isolated VSD, growth failure, recurrent LRTI'S, signs of heart failure, LV Dilatation Z scores >+2SD and VSD gradient <4m/sec and satisfying the following criteria were included: VSDs deemed suitable for the

procedure (after trans-thoracic & trans-esophageal echocardiography), after failed percutaneous VSD device closures (that are considered suitable for PVDC). Children who had VSDs with inlet-extension/ significant prolapsed aortic valve/ associated lesions requiring open-heart surgery (OHS) and guardians refusing consent were excluded. Age, sex and weight-matched, consecutive children with isolated VSD undergoing traditional OHS with CPB from August-2018 to August-2022 were included in the surgical group.

**Results:** A total of 38 children (M: F = 16:22) in the periventricular and 84 (M: F = 42:42) in the surgical group were included. 31(81.5%) children had successful PVDC. Significant hemolysis, embolisation to the Left pulmonary artery, severe tricuspid regurgitation with additional VSD, new aortic regurgitation, inability to deploy the device due to malaligned IVS, significant new-onset TR and multiple additional VSDs in one case each were the reasons for conversion to surgery on CPB. The median age and weight of the two groups were comparable. The duration of the procedure was significantly lower in the periventricular compared to the surgical group ( $p < 0.01$ ). There was no significant difference in arrhythmias, valvular regurgitations, post-op ventilation time and hospital stay.

**Conclusions:** In selective cases, per ventricular device closure of VSD is an alternative to traditional VSD closures with a complication rate similar to traditional surgical repair.

**Keywords:** heart septal defects, ventricular; heart defects, congenital; heart block; child; aortic valve insufficiency; tricuspid valve insufficiency; echocardiography, trans esophageal

#### P-509

##### Transesophageal echocardiography guided per-atrial atrial septal stenting and pulmonary artery banding

Prerna Bhat

Pediatric Cardiac Services, Narayana Institute of Cardiac Sciences, Bengaluru, India

**Background and Aim:** Pulmonary artery banding (PAB) is a surgical procedure performed for the palliation of severe PAH in congenital cardiac defects with increased pulmonary blood flow (PBF). Adequate atrial level mixing of pulmonary and systemic venous return is essential to maintain a reasonable systemic oxygenation in DORV and TGA spectrum, even in the presence of increased PBF, necessitating the creation of an unrestrictive inter atrial communication (either surgical septectomy or interventional septostomy) before attempting PAB in such lesions.

**Method:** A 6 week old male with DORV and a remote VSD (non-routable) with intact interatrial septum (IAS) with juxtaposed atrial appendages and severe PAH, presented with dyspnea and cyanosis, where a hybrid procedure was planned under transesophageal echocardiography (TEE) guidance.

Via a sternotomy and 3 mg/kg heparinization, a 6F sheath was introduced into right atrium (RA) through a purse-string. Under TEE guidance a stiff wire was passed through the sheath, pierced the IAS and advanced into left atrium (LA). The sheath was advanced over the guide wire into the LA, the position of which was confirmed by 'tram track appearance' and agitated saline injection. A balloon mounted stent was passed through the sheath, and placed across the IAS (position confirmed by echogenicity) and deployed across the IAS, creating an unrestrictive interatrial communication. The sheath was then removed and purse-string tied. Pulmonary artery (PA) was banded using a 3 mm wide polytetrafluoroethylene strip, to get a mean PA pressure of 20 mmHg. Heparin was not reversed. The child was extubated

after 12 hours with minimal inotropes and discharged with aspirin and furosemide on POD-10.

**Results:** Successful PAB was performed with per-atrial atrial septal stenting under TEE guidance, thus avoiding CPB, cardioplegic arrest and radiation exposure.

**Conclusions:** A hybrid procedure in the form of PAB with TEE guided per-atrial atrial septal stenting, is an alternative both to surgical atrial septectomy (ameliorates the need for CPB) and interventional atrial septal stenting (avoids radiation and vascular access related complications). Furthermore, hybrid procedure decreases the risk of stent embolization as it can be suture fixed.

**Keywords:** admixture lesions, pulmonary plethora

#### P-510

##### Percutaneous treatment of svc stenosis after surgical repair of sinus venosus defect

Atul Prabhu, Prashant Thakur, Kanupriya Chaturvedi, Vishwas Kodapu

Department of Paediatric Cardiology, Sri Sathya Sai Sanjeevani Hospital, Nava Raipur, India

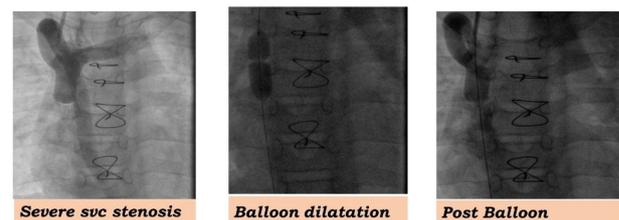
**Background and Aim:** Stenosis or obstruction of the superior vena cava (SVC) is a rare complication of surgery to correct sinus venosus (SV) atrial septal defect (ASD) with partial anomalous pulmonary venous drainage (PAPVD). We report case series that demonstrate the utility of percutaneous treatment of SVC stenosis

**Method:** Superior vena cava catheterization interventions between August 2020 and April 2022 were reviewed.

**Results:** Out of 7 patients with median age of 4 to 13 years (range) and weight of 15 to 60kg were treated. Only 20% had symptoms suggestive of SVC obstruction. The mean gradient and SVC diameter improved from 20mmHg ( $\pm 5.8$  mmHg) to 13mmHg ( $\pm 2.2$ mmHg) ( $p < 0.001$ ) and 5mm ( $\pm 2.7$  mm) to 8mm ( $\pm 3.5$ mm), respectively ( $p < 0.001$ ). The obstruction was adequately relieved in all patients who had balloon dilation alone. All had Freedom from re-intervention so far till 6 months -18 months follow up.

**Conclusions:** Post operative Superior vena cava-related obstruction occur in only 10% of patients with hemodynamically significant SVC obstruction. Endovascular therapy is successful in relieving the stenosis and associated symptoms with good long-term results.

**Keywords:** Sinus Venosus defect, svc stenosis, balloon dilation



Angio images

#### P-511

##### Successful percutaneous re-permeabilization of fontan circuit with stent implantation after conduit thrombosis

Miguel Fogaça Da Mata, Maria Ana Esteves, Isabel Sampaio Graça, Mariana Lemos, Duarte Martins, Rui Anjos

Pediatric Cardiology Department, Hospital de Santa Cruz, Centro Hospitalar Lisboa Ocidental, Lisbon, Portugal

**Background and Aim:** The Fontan circuit predisposition for thromboembolic complications is multifactorial and accounts for significant morbidity and mortality. Poor survival after thromboembolic complications has been reported, with mortality rates as high as 25% in pediatric series and 38% in adult series.

**Method:** We report two cases of Fontan conduit thrombosis treated by percutaneous stenting.

**Results:** Patient A, male, 43 years-old, with history of right isomerism with complete atrioventricular septal defect and hypoplastic left ventricle, underwent Fontan procedure with extracardiac conduit at age 15. He presented to ED with epigastric pain, nausea, syncope and hypoxemia. Echocardiography and angio-CT confirmed conduit thrombosis. Shortly thereafter he developed portosystemic encephalopathy and cardiogenic shock with need for mechanical ventilation and ICU admission. Conduit replacement surgery was deemed of too high-risk. Conduit re-permeabilization, with tandem implantation of 4 stents in the IVC-PA conduit, covering from distal to proximal ends, was performed. A 45mm CP covered stent, two 39 mm CP covered stents and a 45 mm CP bare stent were implanted and dilated to 22mm, with final absence of gradient in the Fontan system. The patient recovered and was discharged home after 17 days. At 9 months follow-up he is asymptomatic with patent conduit.

Patient B, male, 15 years-old, with history of double inlet left ventricle and transposition of the great arteries submitted to Fontan procedure, with extracardiac conduit implanted at 6 years of age. At age 15, on routine follow-up echocardiogram, thrombosis of the conduit with a 50% stenosis was diagnosed. He was asymptomatic and under anticoagulation with warfarin. Extensive thrombosis of the conduit was confirmed by MRI. Surgical replacement of the conduit was considered, but due to subacute and organized nature of the thrombus percutaneous intervention was attempted. Conduit dilation with a Mullins 18/40mm balloon was followed by implantation of two covered 45 mm CP stents. The procedure was successful, with no residual gradient in the conduit and the patient was discharged after 2 days. At 6 months follow up, he maintains a patent conduit with no residual thrombus.

**Conclusions:** In conclusion, conduit thrombosis is amenable to percutaneous stenting, which is an effective and safe option for re-permeabilization.

**Keywords:** Fontan, Interventional cardiology, Stenting, Conduit thrombosis

#### P-513

##### “Percutaneous closure of coronary fistulas”

Romina Mabel Aroca, Juan Manuel Carretero Bellón, Fredy Prada, Ana Cecilia Mandirola, Roman Anibal Gestoso  
Departament of Pediatric Cardiology of Hospital Sant Joan de Deu, Barcelona, Spain

**Background and Aim:** Coronary artery fistula (CAF) is a rare congenital anomaly that is defined as an abnormal connection between coronary arteries and a cardiac chamber or great vessel. The most common type arises from the right coronary artery (RCA) and drain into the right heart chambers or pulmonary artery. Earlier treatment of CAF in childhood is recommended to avoid complications

In the past, surgical ligation was the only option, today percutaneous closure has emerged as a less invasive alternative strategy. This study aimed to review our experience in transcatheter closure of CAF and midterm clinical follow-up.

**Method:** We describe our experience of 5 patients in a period of ten years. Two patients CAF were closed by surgery, requiring later on

transcatheter closure due to residual leakage, and in 3 patients percutaneous closure was the first option. The mean age 4.62 (5 months to 13 years), mean weight 23, 3 (6, 5–53 kg)

The most common presenting symptom was heart failure and a typical murmur in two patients. 3 patients were asymptomatic.

Echocardiography and electrocardiogram were done before and after the procedure. Selective coronary angiography was performed to delineate the anatomy of the fistula. In 3 patients CAF originated from the RCA. In 4 patients CAF drained into the right ventricle and in another into the left ventricle.

Closure was performed with Amplatzer® Duct Occluder II in 3 patients and Amplatzer® Vascular Plug II in two.

**Results:** Anterograde percutaneous closure of CAF was performed in 4 patients using an arteriovenous circuit. Complete occlusion of the CAF was achieved in all patients in a single procedure, but one with a giant coronary fistula who required two procedures where three devices were implanted, despite which he persisted with a small residual flow. Patients were treated with acetylsalicylic acid for 6 months. The mean follow-up was 5.8 years. No fistula recanalization has been observed.

**Conclusions:** Percutaneous closure of CAF has proven to be a safe and effective procedure in selected patients, even in children. In our experience, although limited, there were no cases of mortality or significant complications.

**Keywords:** coronary fistulas, percutaneous closure

#### P-514

##### Extracorporeal membrane oxygenation standby utilization in the paediatric cardiac catheterization LAB

Sarah Pradhan<sup>1</sup>, Myunghyun Lee<sup>2</sup>, Minako Sano<sup>3</sup>, Audrey Marshall<sup>1</sup>  
<sup>1</sup>Division of Cardiology, Labatt Family Heart Centre, The Hospital for Sick Children, Toronto, Canada; <sup>2</sup>Department of Paediatrics, University of Toronto, Toronto, Canada; <sup>3</sup>Division of Cardiovascular Surgery, Labatt Family Heart Centre, The Hospital for Sick Children, Toronto, Canada; <sup>4</sup>Department of Surgery, University of Toronto, Toronto, Canada; <sup>5</sup>Division of Cardiac Anesthesia, Labatt Family Heart Centre, The Hospital for Sick Children, Toronto, Canada; <sup>6</sup>Department of Anesthesiology and Pain Medicine, University of Toronto, Toronto, Canada

**Background and Aim:** Extracorporeal membrane oxygenation (ECMO) provides lifesaving support in cases of circulatory failure that can occur during cardiac catheterization. Institutions aim to identify patients at risk of decompensation when determining if ECMO standby (ES) is needed. ES is activated by the attending interventionalist prior to the case, with some consensus-based practice but no explicit guidelines. This results in a diversion of resources, whilst emergent ECMO cannulation without standby requires gathering of available personnel. We describe ES utilization in a quaternary care center paediatric cardiac catheterization lab.

**Method:** Single-center, retrospective chart review of all patients who underwent ES in the catheterization lab over a 5-year period (January 2016 to July 2021). Patient characteristics included age, congenital heart disease diagnosis and presence of ventricular dysfunction ( $\geq$ mild) at time of catheterization. Clinical parameters included ECMO type, catheterization procedure and mortality.

**Results:** A total of 99 patients underwent cardiac catheterization with ES, of which 1 was cannulated, and 14 patients were emergently cannulated without standby. Commonest procedures performed with standby were: hypoplastic left heart syndrome interstage interventions (19%), elevated right ventricular systolic pressure (15%), right ventricular outflow tract (RVOT) obstruction (11%), majority (64%) were RVOT stents, securing ductal

dependent pulmonary blood flow (10%), mostly (60%) ductal stents, and coronary assessments (9%). Median age at intervention was 88 days [IQR 21; 455], with 39% of single ventricle physiology, and ventricular dysfunction was present in nearly a third.

**Conclusions:** We demonstrate that with ECMO standby for catheterizations deemed at higher risk of circulatory decompensation, cannulation rates are very low. This results in excessive healthcare resource costs with personnel remaining on standby in the lab, which may not translate to improved patient outcomes after cannulation. This serves as a foundation for a quality improvement initiative such as creation of an intermediary status of “ECMO aware” including an available designated surgeon and a primed circuit in the room.

**Keywords:** ECMO, interventional catheterization, quality improvement

### P-515

#### Renal perfusion changes after aortic coarctation stenting

Sigitas Cesna<sup>1</sup>, Kamile Cerlinskaite Bajore<sup>1</sup>, Augustinas Bielinis<sup>2</sup>, Virgilijus Tarutis<sup>3</sup>

<sup>1</sup>Clinic of Cardiac and Vascular Diseases, Institute of Clinical Medicine, Faculty of Medicine, Vilnius University, Vilnius, Lithuania; <sup>2</sup>Center of Radiology and Nuclear Medicine, Department of Nuclear Medicine, Vilnius University Hospital Santaros Clinics, Vilnius, Lithuania; <sup>3</sup>Center of Cardiothoracic Surgery, Clinic of Cardiac and Vascular Diseases, Institute of Clinical Medicine, Faculty of Medicine, Vilnius University, Vilnius, Lithuania

**Background and Aim:** Success of percutaneous aortic coarctation (CoA) treatment mainly is evaluated by angiographic and invasive hemodynamic results. However, some of the patients remain hypertensive. In fact, long standing renal hypoperfusion impact for residual hypertension is not appreciated in CoA. The aim of this review is to evaluate percutaneous CoA treatment influence to renal perfusion changes.

**Method:** Five patients with significant native aortic coarctation or re-coarctation were treated with percutaneous stent implantation between 2020–2022. Effectiveness of the procedure was evaluated by hemodynamic and angiographic parameters. The <sup>99m</sup>Tc-MAG3 captopril scintigraphy was performed before stent implantation and 3–6 month after procedure. Time to peak, time to half-peak, peak to half-peak, 30 min/peak and 20 min/3 min count ratios were determined for whole-kidney ROIs.

**Results:** Median age of patients was 26 years old (18–70). All patients after CoA stenting had residual gradient less than 5mmHg and angiographic images showed well expanded stents. However, renal scintigraphy in two patients showed that time to peak after captopril test was elevated in both kidneys after stenting compared to basic measurements (patient 1 and patient 4) (Figure 1, A). What is more, time to ½ peak did not shorten only in patient 1, which suggests renal parenchymal hypoperfusion (Figure 1, B). Conversely, ACE inhibitor scintigram show decrease in 30 min/peak uptake ratio in all patients after stenting of coarctation (Figure 1, C).

**Conclusions:** Overall, renal perfusion scan shows promising data regarding effectiveness of percutaneous CoA treatment with improved renal perfusion and function. However, there are signs of renal parenchyma injury due to longstanding renal hypoperfusion, which may not recover after gradient reduction at isthmus level.

**Keywords:** aortic coarctation, percutaneous stent implantation, renal scintigraphy, renal hypoperfusion, residual hypertension, ACHD.

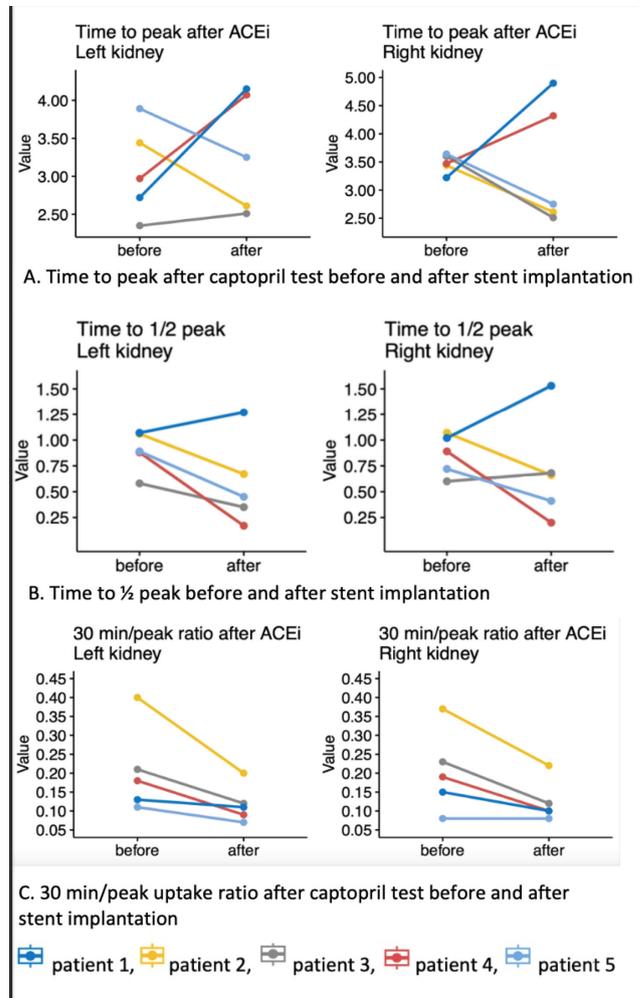


Figure 1. Renal scintigraphy data.

### P-516

#### Anatomical variabilities of sinus venosus defect: contribution of multiplanar analysis and 3D reconstruction.

Efthymia Gkogkou, Clement Batteux, Gregoire Albenque, Florence Leceuf, Vlad Ciobotaru, Regine Roussin, Emre Belli, Sebastien Hascoet, Department of Congenital Heart Diseases, Hôpital Marie Lannelongue, Plessis-Robinson, Paris, France

**Background and Aim:** Surgical repair of sinus venosus defects (SVD) is the standard of care but a transcatheter approach using a long tailored covered stent has emerged as an alternative.

As the anatomy of each patient is unique, we aim to propose a new anatomical description's approach to optimize the suitability assessment for transcatheter correction.

**Method:** In 130 consecutive SVD patients, who were referred in our tertiary center since 2002, we analyzed cardiac CT and searched for reliable criteria using 2D multiplanar analysis and 3D reconstruction (figure 1).

**Results:** Median age at CT was 12.1 years old (1 to 75). Persistent left superior vena cava (L-SVC) was found in 11% of the patients (figure 2). Diameter of right SVC (R-SVC) was narrower in patients with persistent L-SVC ( $p < 0.01$ ) and increased caudally toward the right atrium (RA). Mean R-SVC diameter at the

cavo-atrial junction was 20mm in patients <15 years versus 27 mm in  $\geq 15$  years,  $p < 0.01$ ).

A biatrial connection of the SVC was found in 83 (64%) patients (68 patients had < 50% of overriding of the SVC to the atrial septum and 15 patients had > 50% of overriding). Forty-seven patients (36%) had no overriding of the SVC (figure 3).

For 42 (32%) patients, the distance between the innominate vein and the lower edge of the defect was more than 60mm. Most patients (51%) had two anomalously connected right upper pulmonary veins (RPVs) in SVC, 10 (8%) patients had one, 16 (35%) patients had three and 8 (6%) patients had four RPVs (figure 5). Nineteen (15%) patients had a high connection of the right upper PV (>20mm higher than the cavo-atrial junction). Two different shapes of defect were observed: a cone shape in 70 (53.8%) patients and a hole shape in 60 (46.2%) patients (figure 4, 6).

In 7 patients (5%) SVD was associated with an atrial septal defect. **Conclusions:** We observed a wide spectrum of anatomical configuration of SVD with heterogeneity in the number and location of PVs, defect's morphology, and cavo-atrial connection.

Anatomical variabilities of sinus venous defect (SVD).

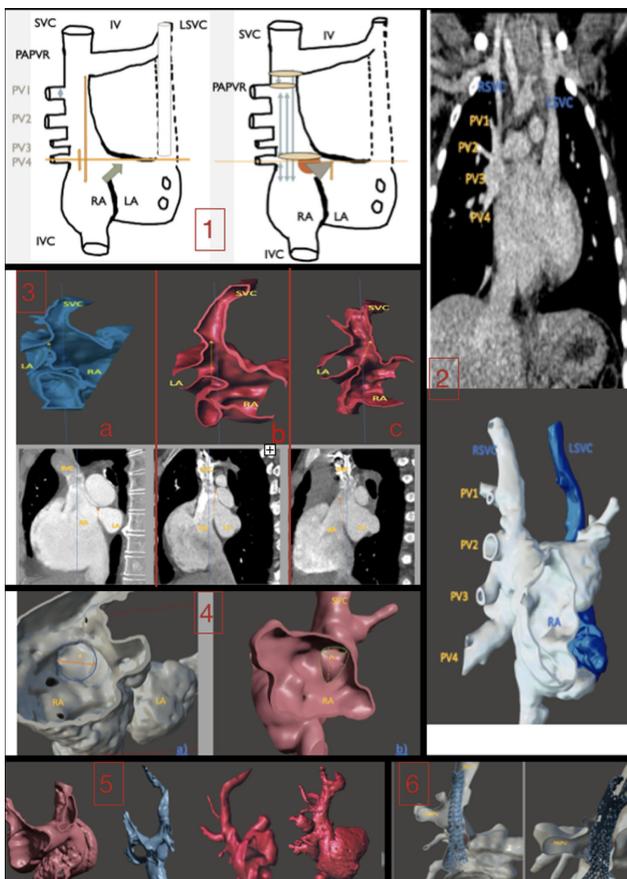


Figure 1: Methods, anatomic analysis and measurements. Figure 2: SVD associated with 4 anomalous connected right pulmonary veins and a persistent left superior vena cava. Figure 3: Different degree of the override of the superior vena cava (SVC) to the atrial septum. a) SVC draining exclusively to the right atrium (no override), b) <50% override of the SVC c) > 50% override of the SVC towards the left atrium. Figure 4: Analysis of the shape of SVD. Figure 5: Variability of the number of anomalous connected right pulmonary veins. Figure 6: Virtual simulation of percutaneous closure. SVD with a shape like a cone and risk of residual shunt.

A precise understanding and classification of these anatomical elements is mandatory for screening and decision to perform transcatheter correction.

**Keywords:** Sinus venous defect, partial anomalous pulmonary venous return, transcatheter closure

## P-517

### Results of right ventricular outflow tract stenting as a primary palliation in symptomatic children having fallot type of lesion

Pratibha Rai<sup>1</sup>, Sathesh S<sup>1</sup>, Shreesha Shankar Maiya<sup>1</sup>, P V Suresh<sup>1</sup>, Prakash R<sup>1</sup>, Sudesh Prabhu<sup>2</sup>

<sup>1</sup>Department of Paediatric cardiology, Narayana Institute of cardiac sciences, Bengaluru, India; ; <sup>2</sup>Department of CTVS, Narayana Institute of cardiac sciences, Bengaluru, India

**Background and Aim:** The initial management of symptomatic children with tetralogy of Fallot, similar lesions with restriction of antegrade pulmonary blood flow and adverse risk factors for corrective surgery is still controversial. Lately (RVOT) stenting has emerged as an alternative that bypasses the side effects of open heart surgery at an early age.

**AIMS:** To analyze the interventional results after RVOT stenting in symptomatic patient of Fallot's physiology with normally related great vessels ( in children less than 5 year of age).

**Method:** This is a single centre retro-prospective observational study conducted at the Department of Paediatric Cardiology, Narayana Institute of Cardiac Sciences, Bengaluru over a period of 14 months from August 2020 to November 2021. Fifty five patients severely symptomatic children (age less than 5 years of age) with TOF physiology and having normally related great vessels with restriction of antegrade pulmonary blood flow and risk factors for correctives surgery were included. Procedure was performed from femoral or from the right internal jugular vein. A pre-mounted balloon expandable stent with a nominal diameter 2 mm greater than the narrowest diameter across RVOT was implanted in the RVOT.

**Results:** 55 patients were included in the study, with a successful stenting in 46 patients (83%).

→ Median age was 11 months. Mean weight at stent implantation was 7kg Mean procedure time was 151.

→ Median fluoroscopy 24 minutes and fluoro dose was 120 cGy m2. Most common access was RIJV (78%).

→ Median ICU stay was 4 days and hospital stay was 7 days. Saturations increased from 67 % to 87 % [ $p < 0.001$ ]. Median RPA z scores increases from -1.03 to 0.4 with [ $p < 0.001$ ]. Median LPA z score increases -1.08 to 0.13 with a [ $p < 0.001$ ].

→ Two cases died (1.6%), two had persistent desaturation (3.6%).

**Conclusions:** → The success of the right ventricular outflow tract stenting is dependent on the selection of an appropriate patient and stent.

→ It can be considered as a considered as first line palliative treatment in patients who are very bad substrate for surgical repair or palliation.

→ Studies with larger sample size and longer follow-up duration would further help in formulating guidelines for the appropriate use of this procedure.

**Keywords:** Cyanosis, RVOT stenting

**P-518****Interventional extracardiac fontan – technique, feasibility, immediate results and short term follow up – a case series.**Prakash Ramachandra<sup>1</sup>, Shreesha Shankar Maiya<sup>1</sup>, Sudesh Prabhu<sup>2</sup>, Suresh Venkatesh Pujar<sup>1</sup><sup>1</sup>Department of Pediatric Cardiology, Narayana Institute of Cardiac Sciences, Bangalore, India.; <sup>2</sup>Department of Pediatric Cardiac Surgery, Narayana Institute of Cardiac Sciences, Bangalore, India.

**Background and Aim:** Interventional Fontan is a technique wherein catheter based intervention is done to complete the Fontan as stage-2. During preparatory stage-1, along with a bidirectional cavopulmonary connection, a polytetrafluoroethylene (PTFE) tube graft is fashioned to connect the inferior caval vein to pulmonary artery (PA) in an end-to-side fashion, with the PA end closed with a pericardial membrane and the side of the tube graft connected to right atrium through an atriotomy. To describe the techniques and feasibility of Interventional Extracardiac Fontan in patients presenting late in childhood and in adults. To describe the immediate procedural success and short term follow up.

**Method:** We describe a series of 7 patients who presented late in the natural history of single ventricle physiology, who successfully underwent the Interventional Fontan after preparatory stage-1 of Interventional Fontan. During the procedure, the pericardial membrane was perforated with a Brokenbrough needle by applying bipolar cautery to the needle externally. The membrane was dilated with a balloon and an appropriate sized covered stent was inflated across the membrane in the tube graft in such a manner that the graft-atriotomy connection was covered and the inferior caval vein was routed to PA, thus completing the total cavopulmonary connection.

**Results:** The mean age at stage-1 was 13.5 years, range 7 years to 23 years. The average time interval between stage-1 and stage-2 was 1 year 6 months, with 9 months being the earliest and 2 years 6 months being latest. The average mean pulmonary artery pressure (Glenn) was 13mmHg which increased by 2-3 mmHg after stage-2. The average baseline saturation was 75% after stage-1, which improved to >95% after stage-2. The procedure was done under general anaesthesia in all but one patient (under sedation). There was 100% procedural success with one patient requiring additional covered stents due to malfunctional PTFE sleeve of the first covered stent. All patients are in functional class 2 with saturations >90% at a mean follow up of 1.5 years.

**Conclusions:** Interventional Fontan is a technically feasible option with low complications, even in older patients if the hemodynamics are appropriate.

**Keywords:** Interventional Fontan, Covered stent

**P-519****Interventional closure of the ductus arteriosus in preterm infants: our experience in the last three years.**

Ana Cecilia Mandirola, Romina Mabel Aroca, Fredy Prada, Juan Manuel Carretero Bellon, María Clara Escobar Díaz, Joan Sanchez De Toledo

Hospital Sant Joan de Deu, Barcelona, Spain

**Background and Aim:** The patent ductus arteriosus (PDA) in preterm infants can have clinical consequences and increase the risk of morbidity and mortality. There is a subgroup of infants who will likely benefit from intervention and the transcatheter device closure of the PDA has recently become more widely available. This study aimed to review our experience in percutaneous closure of PDA in preterm infants.

**Method:** Descriptive study. Case series of preterm infants who had intervention of PDA in the period between August 2019 and November 2022 in our catheterization laboratory.

**Results:** Transcatheter closure was done in 17 patients, 64% were male. The median gestational age at birth was 28 weeks (IQR 26-31) and the median birth-weight was 800 g. (IQR 670-1000).

The median age at the procedure was 42 days (IQR 31-60), and median procedural weight was 1330 g. (IQR 1000-1700).

Median fluoroscopy time was 7:51 minutes (IQR 6:04-14:30) and median total procedural time was 1:10 hours (IQR 0:55-1:37)

As complications: one patient had a device migration to the right pulmonary artery 48 hours after the procedure that was recovered in a second procedure before implantation of a new device, one patient had a residual shunt that resolved 3 weeks later and 2 patients had post-ligation syndrome.

After the procedure, 9 patients were extubated at a median of 5 days (IQR 4-8), 6 patients required invasive ventilation upon transfer to their original center. There are 2 patients who are currently hospitalized, one of them still intubated.

A total of fourteen patients were referred from another hospital for the procedure, of which 10 patients returned to their center after a median time of 3 days (IQR 1-17).

There were no procedural deaths. Survival to discharge was 93% (14/15) with a single death unrelated to the procedure.

**Conclusions:** Percutaneous closure in premature infants is a safe and feasible therapeutic alternative to surgical ligation. In our growing experience, the complication rate was low, and one of them could be resolved at the catheterization laboratory. The acquisition of the skill to perform this type of procedure is necessary for centers of excellence to achieve better results.

**Keywords:** PDA, Intervention closure, Preterm infants

**P-520****A novel technique for physiologic determination of right ventricle-dependent coronary circulation in pulmonary atresia intact ventricular septum**

Peter W Guyon, Kak Chen Chan, Larry Latson

Division of Pediatric Interventional Cardiology, Joe DiMaggio Children's Hospital- Memorial Health System, Hollywood FL, United States of America

**Background and Aim:** Pulmonary atresia with intact ventricular septum (PAIVS) is a heterogenous disease with a wide spectrum of presentation. A key determination is a patient's candidacy for RV decompression. Patients with right ventricular dependent coronary circulation (RVDCC) are at risk of sudden death and should not undergo decompression. Current determination of RVDCC is made primarily by angiography and electrocardiogram (EKG), but these modalities are imprecise in this scenario. A physiologic determination would be preferred.

**Method:** Our center uses a Zero Tip Nitinol Stone Retrieval Basket (Boston Scientific, Marlborough, MA) deployed open across the tricuspid valve annulus during cardiac catheterization, effectively simulating RV decompression. By measuring RV pressure and monitoring for EKG changes or other types of clinical instability, we have an invasive physiologic test for RVDCC. We share a case report of this technique.

**Results:** Patient was a term infant born to a G1P1 female weighing 3.31kg. Echocardiogram confirmed the diagnosis of tricuspid stenosis, PAIVS, unrestricted ASD, and large, tortuous patent ductus arteriosus (PDA). Patient initially underwent PDA stenting for primary palliation then returned to the catheterization laboratory at 5 months of age for a diagnostic catheterization, angiography, and testing for RVDCC. The procedure was performed with

percutaneous access of the right femoral and right internal jugular veins (4 Fr), and arterial monitoring via a 20 gauge access in the right femoral artery. Angiograms of the right ventricle were performed by hand injection (Fig. 1).

Patient underwent a temporary RV decompression with the nitinol basket deployed across the tricuspid valve and the glide catheter (transducing pressure) in the RV body (Fig. 2). Pressure waveforms with original RV pressure and RV pressure with basket in position confirm temporary RV decompression with no changes in the EKG and no hemodynamic instability (Fig. 3).

Subsequently, the patient underwent a hemi-Fontan, atrial septectomy, and avulsion of the tricuspid valve to render it incompetent at just over 6 months of age. At the time of most recent follow-up, he was doing well.

**Conclusions:** Temporary RV decompression can be performed as a physiologic test for RVDCC and may help clarify which patients can undergo permanent RV decompression. Further study is warranted.

**Keywords:** Pulmonary Atresia, Interventional Cardiology

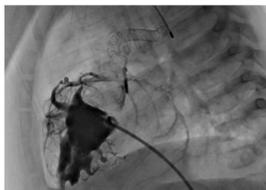


Figure 1. Angiogram performed with a 4 Fr Angle Glide catheter (Terumo, Tokyo, Japan) positioned in the RV. There is opacification of the RV with several large anastomoses with systemic connections to the coronary circulation with brisk filling of the left anterior descending coronary and circumflex branches with delayed filling of the right coronary branch (with competitive filling from the aorta). These findings could be consistent with physiologic RVDCC according to some criteria.



Figure 2. Lateral fluoroscopy with the nitinol basket across the tricuspid valve (white arrow) and the glide catheter within the RV apex.

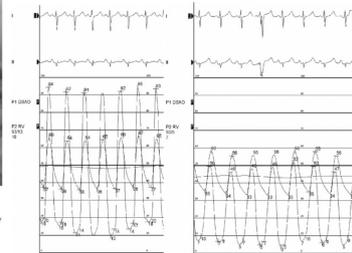


Figure 3. Pressure tracings of the Right Ventricle and Descending Aorta baseline (left) and the decompressed right ventricle and descending aorta during physiologic testing for RVDCC.

## Descriptive Figures

### P-522

#### Results of intraoperative hybrid cardiac interventions in patients with various congenital heart defects.

Judyta Szeliga<sup>1</sup>, Andrzej Rudziński<sup>1</sup>, Tomasz Mroczek<sup>2</sup>, Jacek Koltcz<sup>2</sup>, Anna Grondalski<sup>4</sup>, Michał Parzniewski<sup>3</sup>, Sebastian Górczynski<sup>1</sup>

<sup>1</sup>Department of Paediatric Cardiology, University Children's Hospital, Faculty of Medicine, Jagiellonian University Medical Collage, Cracow, Poland; <sup>2</sup>Department of Paediatric Cardiac Surgery, University Children's Hospital, Faculty of Medicine, Jagiellonian University Medical Collage, Cracow, Poland; <sup>3</sup>Students Scientific Circle, Department of Paediatric Cardiology, University Children's Hospital, Faculty of Medicine, Jagiellonian University Medical Collage, Cracow, Poland; <sup>4</sup>Students Scientific Circle, Department of Paediatric Cardiology, University Children's Hospital, Faculty of Medicine, Jagiellonian University Medical Collage, Cracow, Poland & Medical University of Szczecin, Poland

**Background and Aim:** The intraoperative hybrid procedure brings significant benefits in the treatment of patients with congenital heart defects. We present a single-centre experience in intraoperative hybrid cardiovascular procedures for the treatment of various congenital heart defects.

**Method:** A retrospective analysis of intraoperative hybrid procedures performed between October 2020 and October 2022. All procedures were performed together by interventional cardiologists and cardiac surgeons in a dedicated hybrid operating room. The vascular access was achieved with the surgical assist in the favourable location; directly through the ventricular wall, big vessels or through a cannula of the cardiopulmonary bypass circuit. **Results:** Within 2 years, 44 hybrid procedures were performed in 40 patients (3 patients underwent hybrid treatment more than once). Nine patients were under 1 month old, 19 patients between 1 and 12 months and 16 patients were above 1 year old. The youngest patient was 2 days old and the eldest was 7 years old. Thirty nine procedures included transcatheter interventions and the remaining five were diagnostic. In the former group, 54 interventions were performed. Interventions on pulmonary arteries concerned 16 stents implantations, 12 balloon dilations of previously implanted stents and 3 isolated balloon angioplasties. Ten interventions were performed on the aorta including 5 stent dilations, 3 stent implantations and 2 isolated balloon angioplasties. During 9 of the procedures, arterial duct stenting was conducted either for duct dependent systemic (6) or pulmonary (3) circulation. A hybrid closure of muscular ventricular septal defect was performed in 2 patients. One patient after Fontan operation underwent stenting of the fenestration. The major aortopulmonary collateral artery was closed in one case. All but one cardiac interventions performed during the hybrid procedure were successful without major complications.

**Conclusions:** The intraoperative hybrid procedure can be successfully and safely applied in the treatment of various congenital heart defects. A joint approach enables optimal benefits during single anaesthesia.

**Keywords:** congenital heart disease, cardiovascular intervention, hybrid intervention

### P-524

#### Profiles of EEG background and discharge abnormalities in children undergoing cardiac surgery

Rouyi Lin, Na Du, Li Ma, Shuyou Ning, Minghui Zou, Xinxin Chen, Jia Li

Guangzhou Women and Children's Medical Center, Guangzhou Medical University, Guangdong Province, China

**Background and Aim:** We investigated the characteristics of electroencephalogram (EEG) abnormal patterns before, during and for 48 hours after cardiac surgery in patients with congenital heart disease in relation to demographic and perioperative variables and early outcomes.

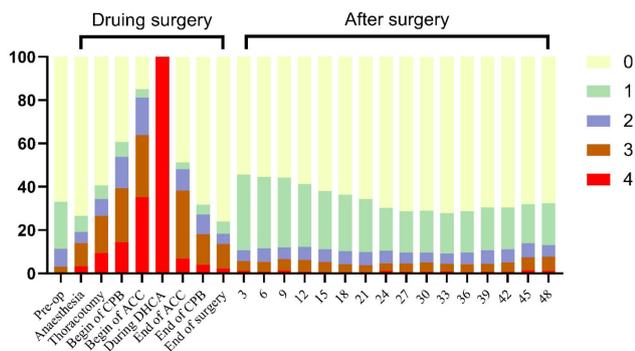
**Method:** This cohort included 443 patients (aged 1-895 days, median 94) undergoing cardiac surgery. Perioperative EEG recording was evaluated for background, sleep-wake cycling, and abnormal discharges (seizures, spikes/sharp waves, delta brushes). Arterial venous pressure, arterial blood gases, doses of inotropic, vasoactive drugs and serum lactate were recorded 3-hourly. Postoperative magnetic resonance imaging of brain was performed. Demographic, STS-EACTS Morbidity and Mortality Categories and early outcome measures (durations of mechanic ventilation, CICU and hospital stay, death) were also collected.

**Results:** Preoperatively, 95(66.4%) patients had EEG abnormalities. Intraoperatively, 107 (49.1%) patients progressed into an isoelectric state and lasted 5282 minutes (median 47). Postoperatively, background abnormalities occurred in 224 (50.5%) patients and gradually lessened ( $P_{time} < 0.0001$ ). 125(55.8%) patients did not have complete recovery by 48th hour after surgery. Spikes/sharp

waves occurred in 365 (82.4%) patients and the number significantly reduced over 48 hours ( $P_{\text{time}} < 0.0001$ ). Seizures occurred in 37 (8.3%) patients. The median time to first seizure was 24 hours (range, 0-45 hours) after surgery and lasted 1.6–1980.1 minutes. Patients with severe preoperative background abnormalities and longer duration of intraoperative isoelectric state had higher degree of postoperative background abnormalities ( $P < 0.0001$ ). After adjusted by time, EEG abnormalities were significantly correlated with all demographic and perioperative variables ( $P \leq 0.09$ ). Patients with EEG abnormalities had longer CICU and hospital stay and death.

**Conclusions:** Perioperative EEG abnormalities were common, and gradually reduced in the first 48 hours after cardiac surgery. Preoperative EEG background abnormalities and longer duration of intraoperative isoelectric state were significantly correlated with postoperative background abnormalities. Numerous perioperative variables adversely correlated with postoperative EEG abnormalities which in turn correlated with early outcomes. Careful clinical management in the CICU may reduce EEG abnormalities and improve outcomes.

**Keywords:** Congenital heart disease, Cardiac surgery, EEG background, EEG abnormal discharges, Isoelectric state, Outcome



The changes of the degree of background abnormalities in the perioperative period

#### P-525

### Psychosocial functioning in adolescents with a congenital heart disease: “the role of attachment and emotion regulation”.

Saskia Mels<sup>1</sup>, Lien Goossens<sup>2</sup>, Katya De Groot<sup>3</sup>, Kristof Vandekerckhove<sup>3</sup>

<sup>1</sup>Ghent University Hospital, Department of Pediatrics, Pediatric Psychology, Ghent, Belgium; <sup>2</sup>Ghent University, Faculty of Psychology and Educational Sciences, Department of Developmental, Personality and Social Psychology, Ghent, Belgium; <sup>3</sup>Ghent University Hospital, Department of Pediatrics, Pediatric Cardiology, Ghent, Belgium

**Background and Aim:** Because of their medical vulnerability, youngsters with a congenital heart disease (CHD) are not only at an increased risk for developing psychological problems but already at young ages they may also face more challenging parent-child interactions. It is the aim of the present study to examine whether insecure attachment relationships with parents and the use of maladaptive emotion regulation strategies explain the psychosocial functioning in adolescent patients with a CHD.

**Method:** A sample of 11 to 18-year-old adolescents who were born with a CHD (percutaneous and/or invasive surgically corrected) was recruited (“N” = 106; 48% boys and 52% girls). Participants completed online questionnaires about emotion regulation (FEEL-KJ), attachment towards mother and father (ECR-RC) and psychosocial wellbeing (SDQ).

**Results:** Results show that higher levels of attachment anxiety towards mother and attachment avoidance towards mother were significantly associated with worse psychosocial functioning. In addition, higher levels of attachment anxiety towards father and attachment avoidance towards father were also significantly associated with worse psychosocial functioning. In line with the hypotheses, the adolescent’s use of maladaptive emotion regulation strategies mediated the associations between insecure attachment and psychosocial functioning.

**Conclusions:** Future research should further unravel the role of emotional processing in explaining psychosocial functioning in youth with CHD. Clinical practice may benefit from strengthening parent-child relationships and emotion regulation training in order to strengthen this vulnerable sample’s wellbeing.

**Keywords:** adolescents, congenital heart disease, psychosocial functioning, attachment, emotion regulation, tripartite model

#### P-526

### Correlations between psychomotor developmental and neuromarkers in children with unrepaired congenital heart defects

Lacramioara Eliza Chiperi

Doctoral School of George Emil Palade University of Medicine, Pharmacy, Sciences and Technology of Targu Mures, Romania

**Background and Aim:** Congenital heart defects (CHD) are the most frequent birth defects and they associate with psychomotor developmental impairment in a percentage of 10–50%, both in those with cyanotic CHD and in those with adequate cerebral tissue oxygenation (non-cyanotic CHD).

This study aim was to evaluate neuromarkers such as neuron specific enolase (NSE), s100 protein (pS100), glial fibrillary acidic protein, tau protein, myelin basic protein as indicators of impairment in the dynamics of psychomotor development.

**Method:** The study included 50 children with CHD excluding those with suspected genetic syndromes based on clinical appearance, genetically confirmed defects, association of other birth defects or born premature. Patients’ psychomotor development was assessed using Denver Developmental Screening Test II, a scale with 125 items that evaluate 4 domains: personal-social behavior, fine motor- adaptive function, gross motor function and language.

Neuromarkers were determined and correlated with psychomotor developmental scores.

**Results:** Four levels of psychomotor development were assessed: all passed through level, baseline level of competence, highest item passed before consistent failure and the upper limit of children’s development. Based on these, domain-specific and overall developmental functioning estimates were calculated and a developmental quotient score was derived. Delays in psychomotor development were observed in 80% of patients.

NSE was over the upper limit in 96% of cases and pS100 in 77% of cases. Correlation between neuromarkers and psychomotor developmental delay (included all four domains and levels of neurodevelopment) were found as follows: in non-cyanotic group with NSE and pS100 and in the cyanotic group only with pS100.

**Conclusions:** Psychomotor developmental delay was found in a great proportion of children clinically considered as normal and it correlated with neuromarkers value, in both groups, cyanotic and non-cyanotic CHD.

**Keywords:** psychomotor development, congenital heart defect, neuromarkers, neuron specific enolase, protein S100

#### P-527

##### **A family-tailored early motor intervention in infants (EMI-heart) with complex congenital heart disease: preliminary results of a feasibility RCT**

Elena Mitteregger<sup>1</sup>, Tineke Dirks<sup>2</sup>, Manuela Theiler<sup>3</sup>, Oliver Kretschmar<sup>4</sup>, Beatrice Latal<sup>1</sup>

<sup>1</sup>Child Development Center, University Children's Hospital Zurich, Switzerland; <sup>2</sup>Lecturer emerita, paediatric physiotherapy, Groningen, Netherlands; <sup>3</sup>Swiss parents' association for the child with heart disease (Elternvereinigung für das herzkrankte Kind); <sup>4</sup>Department of Pediatric Cardiology, University Children's Hospital Zurich, Switzerland

**Background and Aim:** Children with congenital heart disease (CHD) undergoing open-heart surgery are at risk for developmental impairments with motor delay manifesting first and contributing to parental concerns. Only a few interventional studies aim to improve motor development in CHD infants with inconclusive results. We thus developed a family-tailored early motor intervention (EMI-Heart), which aims to promote postural control enhancing motor development and partnering with parents to encourage family well-being.

The primary aim is to evaluate the feasibility of EMI-Heart. The secondary aim is to evaluate differences between the intervention (EMI-Heart) and control group (standard of care) in motor outcomes and family well-being at baseline, post-treatment, and follow-up.

**Method:** This single-centre feasibility RCT compares two groups of CHD infants after open-heart surgery. Infants meeting inclusions criteria are randomly allocated to EMI-Heart or the control group. Infants assigned to EMI-Heart receive early motor intervention for 3 months. Feasibility outcomes are a) clinical recruitment rates and percentage of families completing, b) average duration and number of sessions, and c) acceptability using parental questionnaires post-treatment. Secondary outcomes are a) motor performance (e.g., General Movement Assessment, Alberta Infant Motor Scale) and b) family well-being (e.g., PedsQL, SF36) at baseline (3–5 months), post-treatment (6–8 months) and follow-up (12 months). We evaluate the feasibility using descriptive statistics and assess differences between the two groups using non-parametric statistical analysis at baseline, post-treatment, and follow-up.

**Results:** Regarding feasibility we found a) a recruitment rate of 59% (10/17), all families completed the study (100%, 10/10), b) average intervention duration was 3–4 months, the number of EMI-Heart sessions was 9 sessions, c) the average Likert scale of parental acceptability was 3, 7 (range 1 not agree to 4 totally agree). Regarding secondary outcomes we found that motor performance scores of the intervention and control group were not different at baseline.

**Conclusions:** EMI-Heart is a feasible early motor intervention with a good recruitment rate and high parental adherence and acceptability. This feasibility RCT provides information about a newly developed early motor intervention in infants with complex CHD and provides a foundation for a future large-scale RCT.

**Keywords:** congenital heart disease, open-heart surgery, early motor intervention, physiotherapy, family-tailored, family well-being

#### P-528

##### **Executive functions in preschool children with congenital heart disease and controls**

Andrew T M Chew<sup>1</sup>, Alexandra F Bonthron<sup>1</sup>, Christopher Kelly<sup>1</sup>, Kuberan Pushparajah<sup>2</sup>, John Simpson<sup>2</sup>, Mary Rutherford<sup>1</sup>, A. David Edwards<sup>1</sup>, Chiara Nosarti<sup>3</sup>, Serena J Counsell<sup>1</sup>

<sup>1</sup>Centre for the Developing Brain, School of Biomedical Engineering and Imaging Sciences, King's College London, London, UK; <sup>2</sup>Paediatric Cardiology Department, Evelina London Children's Healthcare, London, UK; <sup>3</sup>Department of Child and Adolescent Psychiatry, Institute of Psychiatry, Psychology and Neuroscience, King's College London, London, UK

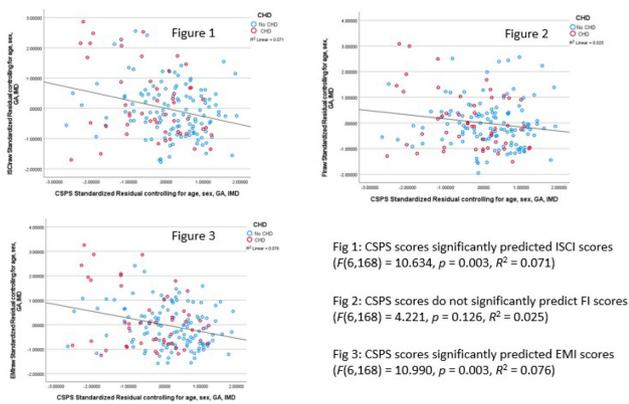
**Background and Aim:** Executive functions (EF) in preschool children are related to successful learning during school years and beyond. Children born with congenital heart disease (CHD) are at risk of impaired EF but, to date, few studies have assessed EF in preschoolers with CHD. We aimed to investigate the association between EF and demographic, environmental and clinical factors in children with CHD and controls aged between 4 to 6 years.

**Method:** Parents completed the Behavior Rating Inventory of Executive Function-Preschool Version (BRIEF-P) questionnaire and Cognitively Stimulating Parenting Scale (CSPS) questionnaire. Index of multiple deprivation (IMD) was calculated from participants' postal address as a measure of socioeconomic status. Children with critical or serious CHD who had surgery in their first year were included. Controls were recruited from participants in the developing Human Connectome Project. Children born before 31 weeks gestational age (GA) were excluded. In the newborn period infants underwent presurgical brain MRI on a 3-T scanner, and any brain injury rated based on previously published criteria.

**Results:** Fifty children with CHD (Transposition of Great Arteries  $n = 20$ , truncus arteriosus  $n = 1$ , coarctation of the aorta  $n = 11$ , Tetralogy of Fallot  $n = 11$ , pulmonary stenosis  $n = 3$ , pulmonary atresia  $n = 3$ , Tricuspid atresia  $n = 1$ ) and 118 controls were included. There were no significant differences in all three composite EF scores (Inhibitory Self-Control Index (ISCI), Flexibility Index (FI), Emergent Metacognition Index (EMI)) between CHD and control children. Higher CSPS scores were significantly associated with lower ISCI ( $p = 0.003$ ) and EMI scores ( $p = 0.003$ ) after controlling for age of assessment, sex, GA at birth and IMD. There was no relationship between EF scores and CHD type, surgical factors or brain MRI injury rating. **Conclusions:** There were no significant differences in EF scores between children with CHD and controls. All preschool children have better EF scores (ISCI and EMI) when within a more stimulating home environment. There were no significant relationships between EF scores and surgical factors, CHD type or brain MRI injury scores in the CHD group. Encouraging parents to provide a stimulating home environment may support executive function development in both children with CHD and typically developing children.

**Keywords:** congenital heart disease, preschool, executive function, stimulating environment, MRI, outcome

Relationship of CSPS and EF scores



CSPS scores significantly predicted ICSI and EMI scores but not FI scores

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Visual attention profiles in toddlers with congenital heart disease: an eye-tracking study

Alexandra Bonthron<sup>1</sup>, Vanessa Kyriakopoulou<sup>1</sup>, Luke Mason<sup>2</sup>, Andrew Chew<sup>1</sup>, Shona Falconer<sup>1</sup>, Christopher Kelly<sup>1</sup>, John Simpson<sup>3</sup>, Kuberan Pushparajah<sup>3</sup>, A. David Edwards<sup>1</sup>, Chiara Nosarti<sup>4</sup>, Emily Jones<sup>2</sup>, Serena Counsell<sup>1</sup>

<sup>1</sup>Centre for the Developing Brain, School of Biomedical Engineering and Imaging Sciences, King's College London, London, UK; <sup>2</sup>Centre for Brain and Cognitive Development, Department of Psychological Sciences, Birkbeck, University of London, London, UK; <sup>3</sup>Paediatric Cardiology Department, Evelina London Children's Healthcare, London, UK; <sup>4</sup>Centre for the Developing Brain, School of Biomedical Engineering and Imaging Sciences, King's College London, London, UK and Department of Child and Adolescent Psychiatry, Institute of Psychiatry, Psychology and Neuroscience, King's College London, London, UK

**Background and Aim:** Infants born with Congenital Heart Disease (CHD) are at increased risk of neurodevelopmental impairments. Executive functions are a set of abilities encompassing information processing, cognitive flexibility, attentional control and goal setting which support daily life and academic achievement and are particularly impaired in school-aged children with CHD. However, it is not yet known if antecedents of impaired executive functions are detectable in younger children with CHD. The aim of this study was to characterise subdomains of visual attention: cognitive control, social attention, and attentional orienting and searching abilities, in toddlers with severe CHD compared to typically developing children.

**Method:** 30 toddlers with CHD (transposition of the great arteries  $n = 13$ , coarctation of the aorta  $n = 8$ , tetralogy of Fallot  $n = 3$ ; pulmonary stenosis  $n = 3$ ; pulmonary atresia  $n = 1$ ; aortic stenosis with coarctation of the aorta  $n = 1$ ; tricuspid atresia  $n = 1$ ) were recruited from St Thomas' Hospital, London. 66 typically developing toddlers recruited as part of the Developing Human Connectome Project (dHCP) were included as a control group. At 22 months, all children completed an eye-tracking battery, in which the gaze was tracked using a Tobii TX-300 during a series of passive and gaze-contingent viewing tasks designed to assess cognitive control, visual searching, orienting attention, and social attention. Accuracy and reaction times were extracted and

compared between groups using repeated measures analysis of covariance.

**Results:** Toddlers with CHD were born at a younger gestational age [Median (IQR) 38.71 (38.32–39.00) weeks] than typically developing children [40 (39–40.71) weeks] but did not differ on corrected age at assessment, sex, socioeconomic status or eye-tracking data quality. Toddlers with CHD were slower at orienting and searching attentional tasks and less accurate on searching and cognitive control tasks compared to typically developing children (Table 1). In contrast, social attention was not impaired.

**Conclusions:** Here we provide the first evidence using objective measures derived from eye-tracking that toddlers with CHD have impaired attentional orienting, searching and cognitive control. These abilities likely scaffold the development of executive functions in childhood. Further research is needed to determine if eye-tracking metrics represent an early marker for impaired executive functions at school-age in children with CHD.

**Keywords:** Executive functions, toddlers, eye-tracking, attention

Table 1. Eye-tracking tasks and differences between children with CHD and controls

Task Description	Visual attention ability targeted	Metrics of interest	Overall effect of CHD	Interaction between CHD and task condition
A cartoon appears in the center of the screen, then another cartoon appears at the left or right edge of the screen during the following conditions: 1. while the center cartoon remains 2. as the center cartoon disappears 3. 200ms after the center cartoon disappears	Attentional orienting	Time to look at cartoon at the side of the screen during each condition	$F(1, 78) = 7.1$ $pFDR = 0.034$	$F(2, 162) = 0.002$ $pFDR = 0.921$
Child is rewarded for identifying a red apple amongst: 1. 8 blue apples or red slices of apple 2. 8 or 12 blue apples and red slices of apple	Visual searching	Time to finding red apple in each condition Proportion of correctly found apples in each condition	$F(1, 82) = 19.0$ $pFDR < 0.001$ $F(1, 84) = 2.9$ $pFDR = 0.180$	$F(2, 171) = 4.9$ $pFDR = 0.034$ $F(2, 174) = 4.1$ $pFDR = 0.047$
The screen is divided into two: 1. nine 2s cartoons are played on one side 2. eight 2s cartoons are played on the other	Cognitive control and reversal learning	Reaction time to look at video during each condition Proportion of times the correct side of the screen is anticipated in each condition	$F(1, 50) = 0.01$ $pFDR = 0.863$	$F(1, 55) = 0.30$ $pFDR = 0.863$ $F(1, 55) = 7.0$ $pFDR = 0.034$
Five images are presented on screen: 1. face with direct gaze 2. scrambled face image 3. car 4. mobile phone 5. bird	Social attention	Longest look at each image Proportion of time spent looking at each image	$F(1, 61) = 2.8$ $pFDR = 0.180$ $F(1, 61) = 0.005$ $pFDR = 0.946$	$F(2, 138) = 0.48$ $pFDR = 0.863$ $F(2, 138) = 0.06$ $pFDR = 0.946$

Covariates included when significantly associated with dependent variable: Gestational age at birth (all models); eye-tracking degrees of precision and accuracy (visual searching, attentional orienting, social attention); multiple deprivation index (cognitive control, social attention); Results in bold are significant

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Neurodevelopmental outreach project trial at east midlands congenital heart network (EMCHN)

Vicky Elliott<sup>1</sup>, Felicity Larson<sup>2</sup>, Emily Hancox<sup>3</sup>  
<sup>1</sup>East Midlands Congenital Heart Network, UK; <sup>2</sup>University Hospitals of Leicester; <sup>3</sup>Leicestershire Partnership NHS Trust

**Background and Aim:** There is a recognition that children and young people growing up with congenital heart disease (CHD) are at greater risk of neurodevelopmental challenges, such as diagnoses of attention deficit/hyperactivity disorder (ADHD), autism spectrum disorder (ASD), as well as specific cognitive impairments. It is recommended that children with congenital heart disease (CHD) are offered regular neurodevelopmental follow-up during childhood to screen for emerging psychosocial issues and offer early support.

**Method:** Children most at risk of neurodevelopment complications were identified from a database of patients of the East Midlands Congenital Heart Centre and invited to complete the PedsQL-C (Cardiac Module) and Strengths and Difficulties Questionnaire (SDQ) to screen for difficulties. Where difficulties were noted, or families asked to speak with a psychologist, appointments were offered.

**Results:** A total of 138 young people were invited across three age ranges (Young Child, Middle Childhood, and Adolescent). Response rates were similar across groups and all were lower than expected (ranging from 6–18%). Respondents reported a mix of many different levels of functioning. Assessments were undertaken with 12 patients and their families, resulting in primarily onward referrals for neurodevelopmental assessment (33%) and/or mental health support (25%). Three cases were taken on for continued work of different types.

**Conclusions:** While the offer was clearly helpful to some families, the majority of patients invited did not respond. This leads us to believe that a “cold-calling” opt-in approach may not be the most appropriate, and is certainly not an effective use of limited psychology resource. Suggestions are made regarding how clinicians in EMCHN can support with identification of children at risk of neurodevelopmental difference and in need of support.

**Keywords:** Congenital Heart Disease, Neurodevelopment, Screening, Early Intervention

#### P-532

##### **Structured psychosocial assessment during hospitalisation for adults with congenital heart disease – a pilot study**

Edward Callus<sup>1</sup>, Valentina Fiolo<sup>2</sup>, Enrico Giuseppe Bertoldo<sup>2</sup>, Silvana Pagliuca<sup>2</sup>

<sup>1</sup>Department of Biomedical Sciences for Health, University of Milan, Milan, Italy; Clinical Psychology Service, IRCCS Policlinico San Donato Research and University Hospital, San Donato Milanese, Milan, Italy; <sup>2</sup>Clinical Psychology Service, IRCCS Policlinico San Donato Research and University Hospital, San Donato Milanese, Milan, Italy

**Background and Aim:** Adults with congenital heart disease (ACHD) often face many psychosocial challenges including possible frequent hospitalisations, in which their condition is evaluated, and they could require surgical, cardiac catheterizations and/or implantation of cardiac devices. Nevertheless, there are often shortcomings in the healthcare system to identify, address and treat psychological distress in this population. In an effort to address this important issue, a protocol of psychosocial screening is proposed according to the pertinent literature.

**Method:** The main patient reported outcomes selected are perceived health and quality of life measured with a numerical rating scale ranging from 0–100%, 100 being the best perceived health and highest level of quality of life, anxiety (General Anxiety Disorder -7), depression (Patient Health Questionnaire -9) and post-traumatic stress disorder (Impacts of Events Scale- revised). The questionnaires were administered during the pre-operative/pre-interventional phase to a total of 24 patients (10 females, average age 41, 96, and the following distribution regarding cardiac diagnosis; 8 mild, 10 moderate and 6 severe).

**Results:** The average perceived health was 66, 39/100 and quality of life 75, 38/100 (2 patients scored lower than 33% for perceived health and 1 patient for quality of life). 41, 66% of patients manifested significant psychological distress (moderate or severe anxiety 37%, moderate or severe depression 12% with 8, 3% presenting

both moderate and/or severe anxiety and depression). 45, 83% resulted as having a probable presence of PTSD.

**Conclusions:** These preliminary results confirm that this population presents specific psychological fragilities which require a special attention and monitoring during this period. In this manner, personalized and targeted interventions to improve the patient's overall health and limit the stressful or potentially traumatic effects of the hospitalisation itself can be structured and implemented.

**Keywords:** Congenital heart disease in adults, hospitalised patients, clinical psychology, psychosocial, patient reported outcomes

#### P-533

##### **Neurodevelopment of children with congenital heart defect born preterm or growth restricted at birth: A systematic review and meta-analysis**

Neil Derridj<sup>1</sup>, Johanna Calderon<sup>3</sup>, Damien Bonnet<sup>2</sup>, Ali Ghanchi<sup>5</sup>, Babak Khoshnood<sup>5</sup>, Romain Guedj<sup>4</sup>

<sup>1</sup>Université de Paris, CRESS, INSERM, INRA, F-75004 Paris, France; <sup>2</sup>M3C-Necker, Hôpital Universitaire Necker-Enfants malades, AP-HP, Université de Paris Cité, Paris, France; <sup>3</sup>M3C-Necker, Hôpital Universitaire Necker-Enfants malades, AP-HP, Université de Paris Cité, Paris, France; <sup>4</sup>UMR 1046 INSERM CNRS PhyMedExp, Université de Montpellier, Montpellier, France; Harvard Medical School, Department of Psychiatry, Harvard University, Boston, USA; <sup>5</sup>Pediatric Emergency Department, AP-HP, Armand Trousseau Hospital, Sorbonne Université, Paris, France; Université de Paris, CRESS, INSERM, INRA, F-75004 Paris, France; <sup>5</sup>Université de Paris, CRESS, INSERM, INRA, F-75004 Paris, France

**Background and Aim:** Children born preterm (PT), small for gestational age (SGA), low birth weight (LBW) or with a congenital heart defect (CHD) are independently at higher risk to develop neurodevelopmental (ND) impairments. The combined effect between CHD and one of these neonatal vulnerability factors (PT, SGA, and/or LBW) is suggested but not clearly established. To systematically review and meta-analyze ND outcomes in children born with the combination of CHD and these neonatal vulnerability factors.

**Method:** Medline and Embase were searched for eligible studies that reported ND outcomes in children born with CHD and neonatal vulnerability factors. Data extraction was performed by two blinded reviewers. Risk of bias was assessed using the Critical Appraisal Skills Programme cohort checklist. Meta-analysis involved use of random-effects models.

**Results:** From 2926 reports, we included 12 studies in qualitative synthesis, and the meta-analysis included 6 studies. Risk of bias was low in 4/12 studies. Regarding children born PT with CHD, 26% (95%CI: 20%–32%,  $I^2 = 0\%$ ) of them had overall cognitive impairment and 19% (95%CI: 7%–35%,  $I^2 = 82\%$ ) had intellectual disability. Regarding children born SGA with CHD, a single study reported lower IQ score for CHD requiring surgery. Regarding children born LBW with CHD, a single study reported a lower score in communication, daily living and motor skills for children born with hypoplastic left heart syndrome

**Conclusions:** There is a lack of robust evidence that prematurity, SGA or LBW significantly increase the neurodevelopmental risk that is already associated with CHD. Further studies with sufficient

power are needed to further explore the impact of these associations.

Registration: PROSPERO, CRD42020201414

**Keywords:** Neurodevelopment, congenital heart defect, preterm, growth restricted at birth, meta-analysis

#### P-534

##### **Similarities and differences in the neurodevelopmental outcome of children with congenital heart disease and children born very preterm**

Flavia Wehrle<sup>1</sup>, Timm Bartal<sup>1</sup>, Mark Adams<sup>2</sup>, Dirk Bassler<sup>2</sup>, Cornelia Hagmann<sup>3</sup>, Oliver Kretschmar<sup>4</sup>, Giancarlo Natalucci<sup>5</sup>, Bea Latal<sup>1</sup>

<sup>1</sup>Child Development Center, University Children's Hospital Zurich, Zurich, Switzerland; <sup>2</sup>Newborn Research, Department of Neonatology, University Hospital Zurich, Zurich, Switzerland; <sup>3</sup>Department of Neonatology and Intensive Care, University Children's Hospital Zurich, Zurich, Switzerland; <sup>4</sup>Department of Cardiology, University Children's Hospital, Zurich, Switzerland; <sup>5</sup>Family Larsson-Rosenquist Center for Neurodevelopment, Growth and Nutrition of the Newborn, Department of Neonatology, University Hospital Zurich, Zurich, Switzerland

**Background and Aim:** Infants with congenital heart disease (CHD) undergoing cardiopulmonary bypass surgery and infants born very preterm (VPT) are critically ill when they are born, and these high-risk neonates may experience neurodevelopmental impairments as they grow up. Here, similarities and differences in their neurodevelopmental outcome and therapy utilization at preschool age are described.

**Method:** 155 children with complex CHD were assessed as part of a prospective, single-center longitudinal study, and compared to 254 children born VPT who were assessed at the same center as part of a national follow-up register. Neurodevelopmental outcome included IQ, motor abilities, and behaviour. Further, parents reported on therapy utilization (i.e., motor therapies, early interventions and other types of therapies). Group differences were tested using independent t-tests and K<sup>2</sup>-tests. Equivalence testing was used to investigate similarities between the groups.

**Results:** Learning disabilities (i.e., 70 ≤ IQ < 85) and intellectual impairments (i.e., IQ < 70) occurred in 17.4% and 4.5% of children with CHD compared to 23.1% and 5.5% in children born VPT. Motor functions were impaired in 57.0% of children with CHD compared to 37.8% of children born VPT, and behavioral problems were apparent in 15.3% and 11.5% of the children, respectively. Children with CHD had poorer global motor abilities ( $d = -0.26$ ) and poorer dynamic balance ( $d = -0.62$ ) than children born VPT, and children born VPT had poorer fine motor abilities than children with CHD ( $d = 0.34$ , all  $p < .023$ ). Static balance and peer problems were statistically similar between the groups (both  $p < .049$ ). Children with CHD received significantly fewer therapies compared to children born VPT (23.4% versus 41.0%,  $p < .001$ ).

**Conclusions:** Children with CHD undergoing cardiopulmonary bypass surgery and children born VPT share an overall risk for neurodevelopmental impairments while the developmental problems may manifest as different impairment profiles, including unique relative strengths and weaknesses in each group. Despite this, children with CHD receive fewer therapies, indicating a lack of awareness of the neurodevelopmental burden these children face. Long-term evaluation programs should be established for all high-risk children surviving neonatal critical illness to ensure adequate follow-up care and initiation of therapies.

**Keywords:** neurodevelopmental outcome; IQ, motor abilities, behavioural problems, preterm children, therapy utilization

#### P-535

##### **Placental histology, perioperative brain development, and neurodevelopmental outcome at one year of age in infants undergoing neonatal cardiac surgery**

Céline Steger<sup>1</sup>, Alexander Bögeholz<sup>2</sup>, Beatrice Latal<sup>3</sup>, Maria Feldmann<sup>3</sup>, Cornelia Hagmann<sup>4</sup>, Tanja Restin<sup>5</sup>, Ruth Tuura<sup>6</sup>, Michael Reinehr<sup>2</sup>, Walter Knirsch<sup>7</sup>

<sup>1</sup>Pediatric Cardiology, Pediatric Heart Center; Center for MR Research; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland; <sup>2</sup>Institute of Pathology and Molecular Pathology, University Hospital Zurich, Switzerland; <sup>3</sup>Child Development Center; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland; <sup>4</sup>Neonatology and Pediatric Intensive Care; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland; <sup>5</sup>Neonatology, University Hospital Zurich, Switzerland; <sup>6</sup>Center for MR Research; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland; <sup>7</sup>Pediatric Cardiology, Pediatric Heart Center; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland

**Background and Aim:** Neurodevelopmental (ND) outcome of neonates operated for complex type of congenital heart disease (CHD) may be altered. Multiple factors contribute to an impaired ND outcome either before birth including placental function and altered brain development or after birth due to need of cardiopulmonary bypass surgery and long-stay perioperative intensive care. The specific impact of pathological placental findings in CHD infants on ND outcome at one year of age remains unclear.

**Method:** We studied placenta pathology reports of infants born with complex type of CHD (> 36 week of gestation) undergoing neonatal cardiopulmonary bypass surgery taken from two prospective long-term follow-up studies cohorts (2009 to 2021), including perioperative cerebral MRI and ND outcome data (Bayley III) at one year of age.

**Results:** 50 infants with complete data set from the former Heart and Brain (n = 44) resp. BrainCHD study (n = 6) were analysed. Cardiac diagnosis included infants with D-TGA (n = 23), single ventricle (n = 14), LVOTO (n = 5), RVOTO (n = 6) or others (n = 2). Placental findings were scored as normal (n = 25), or pathologic (n = 25). The mean (±SD) birthweight was slightly reduced in children with pathologic placental findings (3250±380g vs. 3586±624g,  $p = 0.04$ ), while gestational age at birth did not differ (39.3 vs. 39.2 weeks of gestation,  $p = 0.71$ ). Perioperative cMRI were scored as normal (57%) or pathologic (43%) consisting of white matter injury, stroke, sinus venous thrombosis, or other. Pathologic placental findings was not associated with pathologic cMRI, neither before nor after surgery. One year ND outcomes were similar for motor composite score with vs. without pathological placenta findings (90 vs. 88,  $p = 0.74$ ), language composite score (98 vs. 91,  $p = 0.15$ ), and cognitive composite score (105 vs. 104,  $p = 0.83$ ).

**Conclusions:** Although the placental function plays an important role in energy and oxygen supply during fetal life, placenta pathology in infants with complex type of CHD was associated with lower birth weight, but not with structural brain lesions or ND outcome at one year of age. Future research in larger CHD cohorts is needed to better understand its role in altered brain development during fetal life.

**Keywords:** neurodevelopmental outcome, neonatal cardiac surgery, perioperative brain imaging, placental findings

### P-536

#### Cerebral perfusion in neonates with severe congenital heart disease

Alexandra De Silvestro<sup>1</sup>, Giancarlo Natalucci<sup>2</sup>, Maria Feldmann<sup>3</sup>, Cornelia Hagmann<sup>4</sup>, Thi Dao Nguyen<sup>2</sup>, Seline Coraj<sup>2</sup>, Beatrice Latal<sup>3</sup>, Ruth Tuura<sup>5</sup>, Walter Knirsch<sup>6</sup>

<sup>1</sup>Pediatric Cardiology, Pediatric Heart Center; Center for MR Research; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland; <sup>2</sup>Newborn Research, Department of Neonatology, University Hospital Zurich, Switzerland; <sup>3</sup>Child Development Center; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland; <sup>4</sup>Neonatology and Pediatric Intensive Care; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland; <sup>5</sup>Center for MR Research; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland; <sup>6</sup>Pediatric Cardiology, Pediatric Heart Center; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland

**Background and Aim:** Infants with severe congenital heart disease (CHD) are at risk for perioperative brain injuries and neurodevelopmental impairment. An abnormal cerebral blood supply may limit optimal brain development. The aim of this study was to compare regional and whole brain perfusion in CHD and healthy neonates.

**Method:** Analysis of subjects recruited in two prospective cohort studies between 2013 and 2020. Patients with severe CHD undergoing cardiac surgery within the first six weeks of life and healthy controls were included. Cerebral MRI was conducted pre- and/or postoperatively, and at one neonatal time point in controls.

Cerebral perfusion images were acquired with a background-suppressed pulsed continuous arterial spin labeling (pCASL) sequence. Ten grey matter regions (basal ganglia, thalamus, paracentral region, frontal, temporal, parietal and occipital lobe, cingulate gyrus, hippocampus, insula), and whole brain (average of regional values) perfusion were analyzed, controlling for postmenstrual age at scan.

**Results:** The study group consisted of 78 neonates (55 CHD, 23 controls). 47 patients had biventricular, 8 univentricular CHD. Ninety MRI scans (30 pre-, 37 postoperative, 23 controls) were available. Systemic-to-pulmonary shunt was present in 33 (49%) CHD scans. Postmenstrual age at scan was comparable (CHD:  $41.6 \pm 1.8$  weeks, controls:  $41.9 \pm 2.1$  weeks,  $p = 0.55$ ).

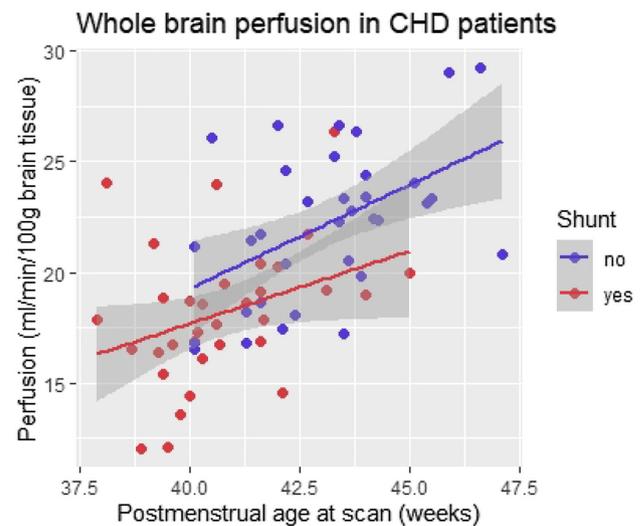
Age-dependent increase of whole brain perfusion was similar between CHD and controls, but regional perfusion was increased in CHD in basal ganglia ( $p = 0.034$ ), hippocampus ( $p = 0.006$ ), insula ( $p = 0.026$ ) and temporal lobe ( $p = 0.028$ ). Furthermore, regional redistribution of cerebral perfusion was found in CHD patients, manifested as higher relative hippocampal perfusion and lower relative frontal perfusion as compared to controls ( $p = 0.014$  and  $p = 0.020$ ). Patients with systemic-to-pulmonary shunt had lower whole brain ( $p = 0.044$ ) and frontal perfusion ( $p = 0.035$ ) as compared to patients without.

**Conclusions:** The age-dependent increase of cerebral perfusion is already well-described in healthy neonates and discussed to represent high metabolic brain activity. Whereas whole brain perfusion was similar between the groups, localized changes and redistribution of regional perfusion in CHD confirm the influence of cardiac disease on cerebral blood supply. The presence of a systemic-to-pulmonary shunt was detected as one causal factor for cerebral perfusion alterations in CHD patients. However, effects

on brain maturation and neurodevelopment need to be further studied.

**Keywords:** Neurodevelopmental outcome, brain imaging, neonatal cardiac surgery

Rplot\_perfusionshunt



### P-537

#### Treatment of congenital heart disease plus prematurity and/or low birth weight - european association of brain in congenital heart disease (EUR-ABC)

Alexandra De Silvestro<sup>1</sup>, Walter Knirsch<sup>1</sup>, Sarah Bless<sup>1</sup>, Sieker Julia<sup>2</sup>, Willemijn Hollander<sup>3</sup>, Sergei Chin On<sup>3</sup>, Cornelia Hagmann<sup>4</sup>, Christian Meierhofer<sup>2</sup>, Bettina Reich<sup>2</sup>, Nijman Joppe<sup>3</sup>, European Association Of Brain In Congenital Heart Disease EUR-ABC<sup>5</sup>

<sup>1</sup>Pediatric Cardiology, Pediatric Heart Center; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland; <sup>2</sup>German Heart Center, Pediatric Cardiology and Congenital Heart Disease, Munich, Germany; <sup>3</sup>University Medical Center Utrecht, Pediatric Intensive Care, Netherlands; <sup>4</sup>Neonatology and Pediatric Intensive Care; Children's Research Center, University Children's Hospital, University of Zurich, Switzerland; <sup>5</sup>on behalf of the European Association of Brain in Congenital Heart Disease

**Background and Aim:** Treatment of preterms and low birthweight (LBW) neonates with congenital heart disease (CHD) may be limited due to early need of cardiac procedures. We aimed on analysing clinical outcome in this combined high-risk patient group.

**Method:** The analysis based on data of the European Association of Brain in Congenital Heart Disease (EUR-ABC), a European-wide collaboration. We analysed clinical characteristics, morbidity, mortality and neurodevelopmental (ND) outcome of preterms (<37 weeks GA) and LBW (<2500g) born between 2016 and 2020, requiring cardiac procedure (catheter or surgery) during first year of life, treated in three European centers (Zurich, CH; Utrecht, NL; Munich, GE). Solitary closure of patent arterial duct was excluded.

**Results:** 308 Neonates (51% male) of which 237 (77%) preterms and 71 (23%) LBW term-newborns were born at median

(IQR) GA of 35.4 (33.3–36.9) weeks, mean (SD) birth weight of 2016 (580) g. CHD were mild (17%), moderate (60%), or severe (24%) including cyanotic (39%), and single ventricle CHD (10%). Rate of associated extra-cardiac anomalies was high (42%). Surgical (90%) as well as catheter-based procedures (10%) were performed at chronological age of 68 (12–155) days, resp. post-menstrual age of 43.1 (38.0–56.8) weeks at 3275 (2383–5143) g. One year mortality was high (19%), both before (7%) or after first intervention (13%) and was associated with perinatal factors such as lower 5min-Apgar score ( $p < 0.001$ ), smaller head circumference at birth and at first intervention (both,  $p < 0.05$ ), more complicative course including bronchopulmonary dysplasia ( $p < 0.05$ ), more severe type of CHD, cyanotic diagnosis, resp. duct dependent CHD (all,  $p < 0.01$ ), genetic anomalies ( $p < 0.05$ ), lower body weight, and younger age at time of intervention (both,  $p < 0.001$ ). Preoperative neuroimaging was performed by cerebral ultrasound (76%), combined with MRI (12%). Data on ND outcome after one year were scarce (19%).

**Conclusions:** Treatment of congenital heart disease plus prematurity and low birth weight remains challenging due to perinatal, cardiac, genetic factors as well as age and body size at first intervention. Low number of ND outcome data in this combined high-risk population shows the need of prospective structured follow-up programs including neuroimaging, neuromonitoring, and functional ND outcome in Europe.

**Keywords:** Neurodevelopmental outcome, neonatal cardiac surgery, prematurity, risk population

#### P-539

##### **Plasma biomarkers of brain injury and the relationship with altered neuroimaging in neonates with congenital heart disease undergoing cardiac surgery**

Ana Remón, Débora Cañizo, Mónica Rebollo, Alba Rivas, Stephanie Handley, Lucía Rossi, Joan Sánchez De Toledo, Marta Camprubí BCNatal, Barcelona, Center for Maternal Fetal and Neonatal Medicine, Hospital Sant Joan de Déu and Hospital Clinic, University of Barcelona, Barcelona, Spain Cardiovascular Disease and Child Development, Institut de Recerca Sant Joan de Déu, Esplugues de Llobregat, Spain.

**Background and Aim:** In the last decades, the survival of patients with congenital heart disease (CHD) has improved and as a result, it has shown up the high incidence of adverse neurologic outcome in these patients. Brain injury biomarkers (BIB) are useful to predict high-risk patients. The aim of this study is to determine the relationship between the main magnetic resonance imaging (MRI) alterations in the postoperative period and the levels of the neuronal biomarkers.

**Method:** Prospective observational study. Forty-four newborns that underwent cardiac surgery during the first month of life were included. MRI was performed per clinical protocol postoperatively. Images were reviewed for multifocal (watershed, white matter injury (WMI)) and focal injury (stroke, single white matter lesion). Two BIB, S100B protein and neuron-specific enolase (NSE), were collected during the perioperative period.

**Results:** In our study, the prevalence of brain injury in MRI was 41.86%, being WMI the more frequent presentation (25%). No significant differences were detected considering the different congenital heart defects or kind of surgery. Levels of both BIB increase immediately after surgery, especially S100B protein ( $p < 0.001$ ). Neonates with stroke presented increased levels of both BIB. WMI was associated with increased levels of NSE with a cut-off value of 45 mg/dl for prediction.

**Conclusions:** Patients with CHD present a high prevalence of brain injury in the postoperative MRI. WMI is the most prevalent alteration and post-operative values of NSE could be useful to identify high-risk patients. Post-operative BIB were also increased in patients with ischemic stroke.

**Keywords:** biomarkers, magnetic resonance imaging, brain injury

#### P-540

##### **A challenging interdisciplinary meeting along with parents before complex heart transplantation with severe pulmonary hypertension (PHT)**

Luis García Guereta<sup>1</sup>, Alvaro González Rocafort<sup>2</sup>, Eduardo Fernandez Jimenez<sup>3</sup>, Juan Jose Menendez<sup>4</sup>, Carlos Labrandero De Lera<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology, Hospital Universitario La Paz, Madrid, Spain; <sup>2</sup>Department of Pediatric Cardiac Surgery, Hospital Universitario La Paz, Madrid, Spain; <sup>3</sup>Department of Psychiatry, Clinical Psychology and Mental Health. Hospital Universitario La Paz, Madrid, Spain; <sup>4</sup>Department of Pediatric Intensive Care, Hospital Universitario La Paz, Madrid, Spain

**Background and Aim:** Informed consent regarding life-threatening procedures for children is a very demanding task for cardiologists and cardiac surgeons in transplantation teams. This work aims to underscore this difficult scenario.

**Method:** A. is a female diagnosed at birth with a non-compacted cardiomyopathy. She presented an asymptomatic course for many years. At age 12 a new mitral regurgitation was detected with restrictive physiology. She denied any symptoms and physical exam only showed a murmur without other signs of heart failure. A cardiac cath showed moderate pulmonary hypertension (PHT) (PVR 3 WU). The medical team decided her inclusion in the heart transplantation waiting list (HTWL) but she refused to consent. Then, an aggressive relationship with her mother was detected and she was referred to mental health department as part of a protocolized assistance for all candidates of pediatric organ and hematopoietic stem cell transplantation. Parents finally signed informed consent and she was included in HTWL. Two months later her clinical situation worsened rapidly and she was admitted to the Intensive Care Unit (ICU). A new cath disclosed severe PHT that contraindicated HT. She lost a significant amount of weight, was bedridden and had a very fragile situation.

**Results:** A new meeting that included a cardiologist, a cardiac surgeon and a clinical psychologist discussed 3 options with her parents: Heart-Lung transplantation (HLT), mechanical circulatory support (MCS) with a paracorporeal device to decrease PVR, or compassionate care. A very difficult scenario had to be transmitted to them: the possibility of failure after device placement that would lead to contraindication of further therapy and lead also to a difficult management of compassionate care with a device that should need to be switched off. The meeting lasted 90 minutes and we all felt the parents would choose compassionate care but two days after they decided MCS as a bridge to HT. 70 days later she is still in ICU, has gained weight, improved her mood, and has been included again in the HTWL. She still has to face transplantation but her mind is set for it.

**Conclusions:** A biopsychosocial approach is key during the transplantation process.

**Keywords:** Pediatric Heart Transplantation, Informed consent refusal, Compassionate care, Mechanical circulatory support

P-541

### Executive functions and primary neurodevelopmental processes in adolescents with congenital heart disease after cardiopulmonary bypass surgery

Alenka Sarah Schmid<sup>1</sup>, Melanie Ehrler<sup>1</sup>, Nadja Näf<sup>1</sup>, Oliver Kretschmar<sup>2</sup>, Ruth O'gorman Tuura<sup>3</sup>, Flavia Wehrle<sup>4</sup>, Beatrice Latal<sup>5</sup>  
<sup>1</sup>Child Development Center, University Children's Hospital Zurich, Zurich, Switzerland; <sup>2</sup>Department of Cardiology, University Children's Hospital Zurich, Zurich, Switzerland; <sup>3</sup>Children's Research Center, University Children's Hospital Zurich, Zurich, Switzerland; <sup>4</sup>University of Zurich, Zurich, Switzerland; <sup>5</sup>Center for MR Research, University Children Hospital Zurich, Zurich, Switzerland; <sup>6</sup>Child Development Center, University Children's Hospital Zurich, Zurich, Switzerland; <sup>7</sup>Children's Research Center, University Children's Hospital Zurich, Zurich, Switzerland; <sup>8</sup>Department of Neonatology and Intensive Care, University Children's Hospital Zurich, Zurich, Switzerland; <sup>9</sup>Child Development Center, University Children's Hospital Zurich, Zurich, Switzerland; <sup>10</sup>Children's Research Center, University Children's Hospital Zurich, Zurich, Switzerland; <sup>11</sup>University of Zurich, Zurich, Switzerland

**Background and Aim:** Children with congenital heart disease (CHD) are at risk for neurodevelopmental impairments, in particular in executive functions (EF). Inhibition, working memory and cognitive flexibility constitute core EF. In addition primary neurodevelopmental processes (PNP), namely, attention, processing speed, and fine motor abilities can also be impaired. This study aimed to investigate the association between PNP and EF in CHD patients. **Method:** EF were assessed in 95 adolescents with CHD and 103 typically developing peers using the Corsi Block Tapping-Test, subtests of the Wechsler Intelligence Scale-IV (WISC-IV), and the Delis-Kaplan Executive Function System. PNP were assessed using subtests of the Test of Attentional Performance (attention divided into phasic and intrinsic alertness), subtests of the WISC-IV and the Zurich Neuromotor Assessment-2. Differences between groups and correlations between functions were assessed using multiple regression analyses (adjusted for SES, age and sex and corrected for multiple testing).

**Results:** Compared to healthy peers, adolescents with CHD showed deficits in all three EF ( $\beta_{\text{inhibition}} = .25$ ,  $\beta_{\text{flexibility}} = .33$ ;  $\beta_{\text{working\_memory}} = .33$ ; all  $p < .01$  and  $R^2$  between 0.10 to 0.24). They performed poorer in intrinsic alertness and processing speed ( $\beta_{\text{intrinsic\_alertness}} = .25$ ,  $p < .001$ ,  $R^2 = 12.01$ ;  $\beta_{\text{processing\_speed}} = .29$ ,  $p < .001$ ,  $R^2 = 0.17$ ), but not in phasic alertness and the fine motor task ( $\beta_{\text{phasic\_alertness}} = -0.06$ ;  $\beta_{\text{fine\_motor}} = 0.09$ ;  $p > .05$ ). CHD severity was not significantly associated with EF or PNP. The three EF were correlated with intrinsic alertness, processing speed and fine motor skills ( $\beta$  between 0.04 to 0.57, all  $p < .05$  and  $R^2$  between 0.10 to 0.51). Further, inhibition was correlated with phasic alertness ( $\beta = 0.13$ ,  $p < 0.001$  and  $R^2 = 0.12$ ).

**Conclusions:** Adolescents with CHD show poorer performance in core EF, in processing speed, and intrinsic alertness with a significant interrelations between these functions. A better understanding of the evolution of impairments will help to improve early detection and tailored interventions.

**Keywords:** executive functions, congenital heart disease, adolescents, processing speed, attention, fine motor

P-542

### Regional cerebral oxygen saturation and serum biomarkers to predict neurological outcome in children undergoing cardiac surgery

Déborá Cañizo<sup>1</sup>, Marta Pérez<sup>2</sup>, Alba Rivas<sup>1</sup>, Sergio Benito<sup>3</sup>, Madeleine Ball<sup>4</sup>, Lucía Rossi<sup>4</sup>, Joan Sánchez De Toledo<sup>4</sup>, Marta Camprubí<sup>1</sup>

<sup>1</sup>Department of Neonatology, Hospital Sant Joan de Déu, Barcelona, Spain.; <sup>2</sup>Department of Psychology, Hospital Sant Joan de Déu, Barcelona, Spain.; <sup>3</sup>Department of Pediatric Intensive Care Unit, Hospital Sant Joan de Déu, Barcelona, Spain.; <sup>4</sup>Department of Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain.

**Background and Aim:** One of the most prevalent morbidities in patients with congenital heart disease (CHD) is neurocognitive impairment. Noninvasive cerebral regional oxygen saturation (CrSO<sub>2</sub>) in the perioperative period is extensively used. Serum biomarkers (SB) such s100B protein and lactate have also been described to predict brain damage.

The aim of this study was to evaluate the capacity of CrSO<sub>2</sub> and SB after cardiac surgery (CS) to predict neurodevelopmental outcome (NO).

**Method:** Patients 6 months or younger undergoing CS were included in this prospective, observational study.

CrSO<sub>2</sub> was recorded in the perioperative period and the amount of time below 40% and 50% was calculated. SB were collected after surgery at 0, 24 and 72 hours. NO at 24 month of age was assessed with Bayley Scales of Infant Development or Vineland test.

**Results:** Forty-six patients were included. CrSO<sub>2</sub> values during surgery do not seem to have an impact on NO.

Twenty-four hours after CS, time of CrSO<sub>2</sub> below 40% was a good indicator of NO (AUC of 0.677,  $p = 0.017$ ), with a cut-off of 49 minutes. If we include levels of s100B at 72 hours and lactate after surgery the predictive AUC increased to 0.897 ( $p = 0.033$ ).

Time of CrSO<sub>2</sub> below 50% was also a strong indicator of NO (AUC 0.749,  $p = 0.032$ ), similarly, levels of s100B at 72 hours and lactate after surgery increased the AUC to 0.8409 ( $p = 0.052$ ).

**Conclusions:** Time of CrSO<sub>2</sub> below 40% and 50% 24 hours after CS combined with levels of lactate and s100B 72 hours after surgery are a great tool to predict NO in children with CHD.

**Keywords:** Cerebral regional oxygen saturation, cardiac surgery, serum biomarkers, neurodevelopmental outcome, congenital heart disease

P-543

### Assessment of the corpus callosum by ultrasound in patients with congenital heart disease.

Alba Rivas<sup>1</sup>, Déborá Cañizo<sup>1</sup>, Patricia García<sup>2</sup>, Ana Remón<sup>1</sup>, Miriam Pérez<sup>3</sup>, Lucía Rossi<sup>4</sup>, Joan Sánchez De Toledo<sup>4</sup>, Marta Camprubí<sup>1</sup>  
<sup>1</sup>Department of Neonatology, Hospital Sant Joan de Déu, Barcelona, Spain.; <sup>2</sup>Department of Technology and Innovation, Hospital Sant Joan de Déu, Barcelona, Spain.; <sup>3</sup>Department of Gynecology and Obstetrics, Hospital Sant Joan de Déu, Barcelona, Spain.; <sup>4</sup>Department of Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain.

**Background and Aim:** Congenital heart disease (CHD) are associated with adverse neurodevelopment. Many CNS alterations has been described in these patients, being white matter (WM) damage the most consistent pattern of brain injury. Corpus callosum (CC), a WM commissure, has been reported to be one of the main biomarkers to assess WM in fetuses with CHD.

Our aim is to describe CC biometry in CHD newborns, and try to establish how the type of CHD and its surgery could influence in its development.

**Method:** Fifty-five newborns that underwent cardiac surgery during their first month of life were included. Brain ultrasound was performed previous to surgery and after surgery. CC was identified in the midsagittal plane as a slightly curved hypoechoic structure. Offline evaluation measurements were done using a specific Matlab 2009b®. The analysis was performed grouping patients

according to clinical variables such as CHD and the kind of surgery.

**Results:** No significant differences in the biometry were detected in pre-surgical scans. After surgery, those CHD with aortic obstruction presented significantly smaller posterior part of the CC (posterior midbody, isthmus and splenium) ( $p < 0.0308$ ). Interestingly, an increase in global area and by segments was found in those patients that underwent extracorporeal circulation during surgery ( $p = 0.0124$ ).

**Conclusions:** Patients with CHD could present modifications in CC area, especially after surgery. Those with aortic obstruction had smaller posterior part of the CC, suggesting that hypoperfusion and cyanosis could modify CC development. Other changes in CC area could appear depending on the surgery and the CHD.

**Keywords:** Corpus Callosum, Congenital heart disease, Brain ultrasound, cardiac surgery

#### P-544

##### **Health related quality of life outcomes in children and adults with fontan circulation - association with physical capacity and clinical variables**

*Kristina Marlene Svensson, Friederike Danne, Anastasia Schleiger, Hannes Sallmon, Felix Berger, Stanislaw Ovroutski, Peter Kramer*  
Department of Congenital Heart Disease / Pediatric Cardiology, German Heart Center Berlin, Berlin, Germany

**Background and Aim:** Children with univentricular heart disease have been reported to have reduced health related quality of life (HRQOL) and therefore, assessment of HRQOL in Fontan patients has been included in our routine follow-up program in 2018. We now sought to evaluate HRQOL in our cohort of Fontan patients and characterize factors that are associated with reduced HRQOL.

**Method:** We retrospectively analyzed all patients that had completed Pediatric Quality of Life Inventory (PedsQL) core scales and cardiac module questionnaires from 2020-2022 during outpatient visits or hospital admission in our institution. Anthropometric data and additional cardiopulmonary exercise, laboratory and clinical data were recorded.

**Results:** A total of 115 patients were identified (median age 17.6 [IQR 11.7-26.3] years, median follow-up after Fontan 13.2 [IQR 6.7-23.0] years) with available self-reported ( $n = 101$ , 88%) or proxy-reported ( $n = 14$ , 12%) HRQOL questionnaires. Overall PedsQL summary, physical and psychosocial health domains and cardiac module scores were significantly lower than published values for healthy children and adults (69.9 [IQR 52.8-80.4], 68.8 [IQR 50.0-84.4], 71.7 [IQR 53.3-82.1] and 71.3 [IQR 59.3-82.4], all  $p < 0.001$ ). PedsQL summary, physical health and cardiac module scores did not correlate with age ( $p = 0.09-0.77$ ), however, psychosocial health score showed a weak correlation ( $r = 0.19$ ,  $p = 0.038$ ), with adults showing higher scores (75.0 [IQR 56.7-82.4]). Physical health scores were correlated with several parameters of cardiopulmonary exercise testing including peak oxygen uptake, heart rate reserve, VE/VCO<sub>2</sub> slope, resting and minimum oxygen saturation, vital capacity ( $p = 0.001-0.046$ ) but also laboratory parameters such as pro-BNP ( $p = 0.048$ ). Parameters that correlated best with all PedsQL domains and summary scores were number of cardiovascular medications, VE/VCO<sub>2</sub> slope and minimum oxygen saturation during exercise (all  $p < 0.001-0.010$ ). No correlations were found for type of Fontan modification, presence of pacemaker and number of re-operations or catheterizations. Estimated GFR correlated significantly with PedsQL summary, physical health and cardiac module

scores ( $p = 0.003-0.006$ ) while hepatic enzymes and bilirubin did not.

**Conclusions:** Fontan patients of all age groups report lower HRQOL scores in comparison to the general population, reflecting reduced functional, physical and psychosocial health. Beyond improvement of Fontan hemodynamics, future studies should focus on targeted preventive/supporting interventions to ameliorate the psychosocial burden of disease in these vulnerable patients.

**Keywords:** Fontan patients, Health related quality of life, cardiopulmonary exercise testing

#### P-545

##### **Quality of life and VO<sub>2</sub> in children and young adults with marfan and related conditions**

*Aitor Guitarte Vidaurre<sup>1</sup>, Thomas Edouard<sup>1</sup>, Fernanda Bajanca<sup>1</sup>, Bertrand Chesneau<sup>1</sup>, Eric Garrigue<sup>1</sup>, Pascal Amedro<sup>2</sup>, Yves Dulac<sup>1</sup>*  
<sup>1</sup>Centre de Référence du syndrome de Marfan et des syndromes apparentés, Hôpital des Enfants, CHU de Toulouse, Toulouse, France; <sup>2</sup>Cardiologie pédiatrique et congénitale, CHU de Bordeaux, Bordeaux, France.

**Background and Aim:** Marfan syndrome (MFS) is characterized by the association of multisystem damage related to connective tissue fragility. The risk of aortic dissection has diverted attention from significant musculoskeletal damage despite its perceived impact on the patient's quality of life.

We have analysed the impact of the disease on the quality of life (QoL) of patients from a young age until early adulthood, and investigated the correlation with the performance at a cardiopulmonary exercise test.

**Method:** This study included 63 Marfan or associated syndrome patients between 5 and 21 years (average 12.4 years). Their responses to the generic health-related quality of life questionnaires PedsQLTM 4.0 and KidScreen were compared with those of a healthy age-equivalent cohort. Socio-demographic parameters were also considered. VO<sub>2</sub>max, as measured by cardiopulmonary exercise test, was analysed in 28 patients of the Marfan cohort between 7 and 20 years (average 12.6 years).

**Results:** Preliminary results confirm the disease's significant impact on the life quality of Marfan patients from a young age. The VO<sub>2</sub>max values were severely impaired in this population, on average reaching 63.8% of the expected values. A correlation between the QoL and cardiopulmonary exercise test performance parameters was demonstrated.

**Conclusions:** The impact of the Marfan syndrome's multisystemic damage and deconditioning on the patient's QoL is evident from a young age and should not be neglected. Namely, the care provided to patients should include adapted physical activity programs from a young age.

**Keywords:** QoL, Marfan syndrome, functional capacities,

#### P-546

##### **Non-cardiac co-morbidities in children with congenital heart defects who underwent cardiac surgery in their first year of life**

*Aoife Golden<sup>1</sup>, Colin J Mc Mahon<sup>2</sup>, Daragh Finn<sup>1</sup>*  
<sup>1</sup>Department of Paediatrics and Child Health, University College Cork, Ireland; <sup>2</sup>Department of Paediatric Cardiology, Children's Health Ireland, at Crumlin, Ireland

**Background and Aim:** Almost 1 in 100 children are born with a congenital heart defect. While advancements in cardiac surgery since the 1980s has led to an improvement in survival rates, there

remains a lack of research surrounding co-morbidities and the needs these patients have throughout childhood. The aim of this study is to investigate the non-cardiac comorbidities in children who underwent cardiac surgery in their first year of life.

**Method:** A cross sectional study was conducted in Cork University Hospital. Thirty participants were recruited through a paediatric cardiology patient list. Inclusion criteria necessitates children between the ages of 4–17 who underwent cardiac surgery in their first year of life. Parent completed questionnaires—including a standardised Strengths and Difficulties questionnaire which serves as an

emotional and behavioural screening tool—were analysed with SPSS software using descriptive statistical methods.

**Results:** Thirty per cent of participants had a genetic diagnosis of which 67% had Down Syndrome, 22% a 22q11 deletion, and 11% a Q14 deletion. 46.7% reported medical problems in areas including: ear, nose, and throat (50%), renal (14.2%), respiratory (14.2%), gastrointestinal (14.2%), musculoskeletal (7.1%), neurological (7.1%), cardiovascular (7.1%), ophthalmological (7.1%) and, rheumatological (7.1%). 63.3% attended any one or multiple of early intervention, occupational therapy, speech and language therapy, psychology. 63.3% require special needs assistance. The scores of the Strengths and Difficulties questionnaires indicated that, 43.3% of the children had 'total difficulties' scores within the 'raised' categories suggesting increased emotional and behavioural difficulties. Having additional medical problems ( $p > 0.24$ ) or a genetic diagnosis ( $p > 0.94$ ) were not associated with increased 'total difficulties' scores.

**Conclusions:** The results depict the prevalence and variety of non-cardiac comorbidities highlighting the importance of interdisciplinary and multidisciplinary input to provide optimal care for these children. Interestingly, genetic diagnosis was not associated with increased difficulties scores,

**Keywords:** Congenital cardiac, medical co morbidity

#### P-547

##### **Impact of a child with congenital heart defect on mother's quality of life – is there any improvement after surgical correction of the heart defect?**

*Lacramioara Eliza Chiperi*

*Doctoral School of George Emil Palade University of Medicine, Pharmacy, Sciences and Technology of Targu Mures, Romania. Department of Pediatric Cardiology, Emergency Institute for Cardiovascular Diseases and Heart Transplant, Targu Mures, Romania*

**Background and Aim:** Mothers have a crucial role in their child physical and psychomotor development: a depressed mother due to affected life quality, will be less stimulating and willing to care for the child.

The aim of this study was to assess if the impact of raising a child with congenital heart defect on maternal quality of life is improved after surgical correction of the child's heart defect.

**Method:** Our study was a prospective, longitudinal one which included 52 mothers of children with unrepaired CHD.

In order to assess life quality of the mother, several domains were evaluated through ICCAP questionnaire (Impact of a Child with Congenital Anomaly on Parents) developed by dr. Petra Honig-Mazer and her team from Erasmus University in Rotterdam and used with permission.

Statistical level of significance was considered  $p < = 0.05$ . Data normality was evaluated with Shapiro-Wilk test and Pearson rank test was used for assessing the correlation between variables in Stata version 13.

**Results:** Six domains were evaluated in each mother: contact with caregivers, social network, partner relationship, state of mind, child acceptance, fears and anxiety. ICCAP questionnaire was completed before the surgery of the child and also after the correction of child's heart defect at an interval of time ranging between 6 and 12 months.

Mean age of evaluated mothers was 27.7 years (limits of 16–39 years) when they have completed the questionnaire. Majority of mothers had only middle school education and were living in the country area. In only 34% of cases the child's heart defect was suspected prenatally. The mean age of their children was 11 months (limits of 1–47 months).

State of mind significantly correlated with number of children per family ( $-0.44$ ), maternal education level ( $0.38$ ) and maternal age at child birth ( $-0.42$ ), all parameters improving at the evaluation done after surgical correction of child's heart defect. Fears and anxiety category turned out to be negatively correlated with number of children per family ( $-0.38$ ) and child age ( $-0.37$ ) with a significant improvement at the second evaluation.

**Conclusions:** Our study demonstrated improvement in mother quality of life after the surgical correction of their child congenital heart defect.

**Keywords:** maternal quality of life, congenital heart defect, Impact of a Child with Congenital Anomaly on Parents questionnaire

#### P-549

##### **Development and implementation of a comprehensive pediatric cardiology transition program**

*Navreet Gill, Sandra Aiello, Rafael Alonso Gonzalez, Conall Thomas Morgan*

*Division of Cardiology, The Labatt Family Heart Centre, The Hospital for Sick Children, Toronto, Canada*

**Background and Aim:** More than 90% of children born with congenital heart disease are now surviving into adulthood, necessitating life-long cardiac follow up. Unfortunately, there exists a lack of structured programs to support adolescents with the transition from pediatric to adult cardiac care, resulting in delayed care, improper timing and transfer of care, emotional distress, and patients who are lost to follow up. This lapse in cardiac management predisposes patients to delayed recognition of cardiac complications, resulting in emergency room visits, hospital admissions and emergent procedures. Thus the aim of this quality improvement (QI) project, was to improve the transition process of adolescent patients aged 14 to 17 years old with congenital heart disease (CHD), through the development and implementation of a cardiac transition program within the Labatt Family Heart Centre at The Hospital for Sick Children.

**Method:** The pediatric cardiac transition program was established in conjunction with the Toronto Adult Congenital Heart Disease Program at the University Health Network Peter Munk Cardiac Centre, using both a QI process and Got Transition's Six Core Elements of Health Care Transition™ approach.

**Results:** Program development included creation of an organizational cardiac transition policy, transition registry, EMR-integrated transition flowsheet, and transfer checklist. Program implementation consisted of delivering in-person or virtual, one-on-one, nurse-led transition sessions. Tailored education sessions focus on CHD disease-specific education (e.g., cardiac anatomy), self-management behaviours (e.g., medication management), and self-advocacy (e.g., knowledge of patient rights). Patients are assessed on their cardiac knowledge, transition readiness, quality of life and are screened for both anxiety and

depression. Sessions are scheduled to coincide with patients' cardiac follow up appointments beginning at 14 years old, with final sessions occurring at 17 years old. With each transition session, adolescents are encouraged to increase their responsibility of their healthcare management over time.

**Conclusions:** Development and implementation of the cardiac transition program are the first steps towards improving the transition experience of adolescents with CHD. Future endeavours include development of parent-focused transition sessions, program expansion to include patients aged 12–25 years old, and the evaluation of the program's impact on patient transition readiness, satisfaction, and CHD knowledge.

**Keywords:** transition, adolescence, transfer

#### P-550

##### **Chdsi questionnaire makes measuring disease-specific quality of life in children/adolescents with congenital heart disease reliable for the first time**

Paul Christian Helm<sup>1</sup>, Anna Lena Ehmann<sup>1</sup>, Constanze Pfitzer<sup>2</sup>, Jia Li<sup>3</sup>, Ulrike Bauer<sup>1</sup>, Peter Ewert<sup>3</sup>, Julia Remmele<sup>3</sup>

<sup>1</sup>National Register for Congenital Heart Defects, Berlin, Germany;

<sup>2</sup>Department of Congenital Heart Disease-Paediatric Cardiology, Deutsches Herzzentrum Berlin, Berlin, Germany;

<sup>3</sup>German Heart Center Munich, Technical University of Munich, Department of Congenital Heart Disease and Pediatric Cardiology, Munich, Germany

**Background and Aim:** Health-related quality of life (HrQoL) and disease-specific quality of life (DsQoL) are to be understood as part of quality of life (QoL). Compared to the HrQoL, the DsQoL does not primarily focus on general health-related aspects of the QoL but additionally takes into account disease-specific issues of a particular patient group, which is confronted with special challenges, problems and concerns due to their disease. Currently, there is no measurement tool to capture the DsQoL of children and adolescents with congenital heart disease (CHD). To change this, the Congenital Heart Disease Specific Inventory (CHDSI) was developed.

**Method:** Validation and standardization of the CHDSI was performed online by the German National Register for Congenital Heart Defects. Study participants were invited to participate by e-mail/post. A total of 1,201 patients were included in the statistical analyses (52% female). The participating patients received – depending on age – the child or adolescent version of the CHDSI in the form of an online questionnaire.

**Results:** The 6 postulated DsQoL subscales consistently show good to very good measures (e. g. Cronbach's alpha:.683 -.914; MIC:.401 -.587; discriminatory power:.478 -.772), and the proposed model of overall DsQoL could be confirmed for both the child and adolescent versions of the CHDSI using confirmatory factor analysis (CFA). CFA showed good to very good fit indices (CFI/TLI: >.95; RMSEA: <.06; relative/normalized  $\chi^2$ : < 3 and factor loadings: >.50). Both sex and CHD severity can be taken into account when interpreting total DsQoL scores and when assessing subscale sum scores.

**Conclusions:** With the validated and standardized CHDSI for the assessment of the DsQoL of children and adolescents with CHD, the individual subjective DsQoL can be reliably assessed. For the first time, the CHDSI provides a measurement instrument that can be used to develop a comprehensive treatment concept taking the DsQoL into account. Only by recording and addressing the individual subjective DsQoL of children and adolescents with CHD in the medical care process is it possible to achieve the best possible general, health-related and disease-specific QoL for the respective patient.

**Keywords:** congenital heart defect, quality of life, CHDSI, questionnaire, validation

#### P-551

##### **Investigation of potential factors influencing disease-specific quality of life in children with congenital heart disease using CHDSI questionnaire**

Paul Christian Helm<sup>1</sup>, Anna Lena Ehmann<sup>1</sup>, Constanze Pfitzer<sup>2</sup>, Jia Li<sup>3</sup>, Ulrike Bauer<sup>1</sup>, Peter Ewert<sup>3</sup>, Julia Remmele<sup>3</sup>

<sup>1</sup>National Register for Congenital Heart Defects, Berlin, Germany;

<sup>2</sup>Department of Congenital Heart Disease-Paediatric Cardiology, Deutsches Herzzentrum Berlin, Berlin, Germany;

<sup>3</sup>German Heart Center Munich, Technical University of Munich, Department of Congenital Heart Disease and Pediatric Cardiology, Munich, Germany

**Background and Aim:** Disease-specific quality of life (DsQoL) does not primarily focus on general health-related aspects of the quality of life (QoL) but additionally takes into account disease-specific issues of a particular patient group, which is confronted with special challenges, problems and concerns due to their disease. Currently, there is no measurement tool to capture the DsQoL of children and adolescents with CHD. To change this, the Congenital Heart Disease Specific Inventory (CHDSI) was developed. The aim of the present statistical analyses was to identify relevant factors influencing the individual DsQoL of a patient.

**Method:** Validation and standardization of the CHDSI was performed online by the German National Register for Congenital Heart Defects. Study participants were invited to participate by e-mail/post. A total of 530 children (6–13 years) were included in the statistical analyses (46.8% female). The participating patients received the child version of the CHDSI in the form of an online questionnaire.

**Results:** Slight negative associations were present between the number of siblings and the DsQoL total score ( $p < .01$ ;  $r = -.117$ ) and for 2 out of 6 subscales ( $p < .01/.05$ ;  $r = -.131/-.103$ ). There was a slight significant association between parental occupational/educational level and DsQoL total score ( $p < .001$ ;  $r = .256$ ) and for 5 out of six subscales ( $p < .001$  and  $p < .01$ ;  $r = .265/.263/.162/.163/.176$ ). Further, mild to moderate significant associations were found between subjective CHD knowledge and DsQoL total score ( $p < .001$ ;  $r = .275$ ) and for all six subscales ( $p < .001$ ;  $r = .196/.162/.276/.231/.326/.225$ ).

**Conclusions:** In particular, the presence of siblings, parental education/occupation level, and subjective heart defect knowledge seem to have a relevant influence on the DsQoL of young and adolescent CHD patients, although the correlations are rather low except in one case with moderate correlation. The results found should be considered when treatment plans are developed. Only by recording and taking into account the individual life situation of children with CHD and possible influences on DsQoL in the medical care process is it possible to achieve the best possible general, health-related and disease-specific QoL for the respective patient.

**Keywords:** congenital heart defect, quality of life, children, CHDSI, questionnaire, validation

#### P-552

##### **Potential factors influencing disease-specific quality of life in adolescents with congenital heart disease using the chdsi questionnaire**

Paul Christian Helm<sup>1</sup>, Anna Lena Ehmann<sup>1</sup>, Constanze Pfitzer<sup>2</sup>, Jia Li<sup>3</sup>, Ulrike Bauer<sup>1</sup>, Peter Ewert<sup>3</sup>, Julia Remmele<sup>3</sup>

<sup>1</sup>National Register for Congenital Heart Defects, Berlin, Germany; <sup>2</sup>Department of Congenital Heart Disease-Paediatric Cardiology, Deutsches Herzzentrum Berlin, Berlin, Germany; <sup>3</sup>German Heart Center Munich, Technical University of Munich, Department of Congenital Heart Disease and Pediatric Cardiology, Munich, Germany

**Background and Aim:** Disease-specific quality of life (DsQoL) does not primarily focus on general health-related aspects of the quality of life (QoL) but additionally takes into account disease-specific issues of a particular patient group, which is confronted with special challenges, problems and concerns due to their disease. Currently, there is no measurement tool to capture the DsQoL of children and adolescents with CHD. To change this, the Congenital Heart Disease Specific Inventory (CHDSI) was developed. The aim of the present statistical analyses was to identify relevant factors influencing the individual DsQoL of a patient.

**Method:** Validation and standardization of the CHDSI was performed online by the German National Register for Congenital Heart Defects. Study participants were invited to participate by e-mail/post. A total of 625 adolescents (14 – 17 years) were included in the statistical analyses (52.3% female). The participating patients received the youth version of the CHDSI in the form of an online questionnaire.

**Results:** There was no significant association between the number of siblings and the DsQoL total score or any of the subscales. Parental occupational/educational level showed a slight significant association ( $p < .001$ ;  $r = .170$ ) only with one of the six subscales. Further, slight significant associations were present between subjective CHD knowledge and DsQoL total score ( $p < .001$ ;  $r = .289$ ) as well as between the subjective CHD knowledge and all six subscales ( $p < .001$ ;  $r = .234/.184/.240/.251/.267/.245$ ).

**Conclusions:** The parental education/occupation level, and subjective heart defect knowledge seem to have a relevant influence on the DsQoL of adolescent CHD patients, although the correlations are rather low. The results found should be considered when treatment plans are developed. Only by recording and taking into account the individual life situation of children with CHD and possible influences on DsQoL in the medical care process is it possible to achieve the best possible general, health-related and disease-specific QoL for the respective patient.

**Keywords:** congenital heart defect, quality of life, adolescents, CHDSI, questionnaire, validation

## P-553

### Evolution of cerebral perfusion in infants with congenital heart disease

Albert Batista<sup>1</sup>, Alba Rivas<sup>2</sup>, Débora Cañizo<sup>2</sup>, María Clara Escobar<sup>3</sup>, Joan Sánchez De Toledo<sup>3</sup>, Marta Camprubí<sup>2</sup>

<sup>1</sup>Department of Neonatology, Hospital General de l'Hospitalet, Hospitalet de Llobregat, Spain; <sup>2</sup>Department of Neonatology, Hospital Sant Joan de Déu, Esplugues de Llobregat, Spain; <sup>3</sup>Department of Cardiology, Hospital Sant Joan de Déu, Esplugues de Llobregat, Spain

**Background and Aim:** Congenital heart disease (CHD) are associated with adverse neurodevelopment Cerebral blood flow disturbance in these patients could affect their normal brain develop. Moreover, this situation may be exacerbated during hemodynamic instability, chronic hypoxia and requirement of a cardiac surgery (CS) during first weeks of life. Doppler ultrasound study of the anterior cerebral artery (ACA), as a part of a brain ultrasonography, is useful to evaluate cerebral perfusion. The aim of this study is to describe, in CHD newborns, the Pulsatility index (PI) and Resistance index (RI) of the ACA in four different moments,

and try to establish how the type of CHD and its surgery could influence these parameters.

**Method:** This is a prospective observational study. Newborn infants undergoing CS, with or without cardiopulmonary bypass (CPB), were eligible for inclusion. Brain ultrasound and doppler study was performed at birth, pre-surgery, post-surgery and previous to discharge. PI and ACA were measured in a sagittal view.

**Results:** Ninety-seven infants were included. Patients were classified in different types of CHD according to oxygen perfusion to the brain (normal, mixed, and low) and aortic flow (normal or obstructed) characteristics.

Table 1 and Table 2 shows PI and RI at four moments of study by groups:

RI of newborns with Transposition of Great Arteries (Group 3) decreases after surgery ( $p < 0.05$ ). We don't observe other differences statistically significantes in RI and PI in our population.

**Conclusions:** In our population of infants with CHD, parameters of cerebral flow (PI and RI) are increased when compare to those described in healthy newborns. Cerebral flow tends to normalized after the surgery. More studies are needed to investigate the relation between alterations in cerebral flow of the newborns with CHD and their neurologic outcome.

**Keywords:** Congenital Heart Disease, Doppler Study, Pulsatility Index, Resistance Index

Table 1

CHD PI (median and IQR)	N	Birth	PreSurgery	PostSurgery	Discharge
Group 1	20	1.92 1.47-2.21	1.69 1.46-2.07	1.76 1.55-2.15	1.8 1.54-2.14
Group 2	19	1.87 1.37-1.96	1.8 1.45-2.36	1.63 1.16-3.01	2.17 1.16-2.41
Group 3	25	1.77 1.65-2.03	1.72 1.59-2.13	1.78 1.45-2.15	1.78 1.32-1.95
Group 4	24	1.42 1.39-1.59	1.52 1.28-1.58	1.67 1.33-1.9	1.48 1.32-1.84
Group 5	7	1.76 1.37-2.26	1.60 1.18-2.26	1.48 1.46-1.82	1.89 1.75-2.04
Group 6	2	1.73 -	1.97 1.73-2.2	1.81 1.62-1.99	2.08 1.39-2.76
Total	97	1.73 1.42-2.08	1.61 1.45-2.07	1.67 1.46-2.12	1.89 1.33-2.08

Table 2

CHD RI (median and IQR)	N	Birth	PreSurgery	PostSurgery	Discharge
Group 1	20	.85 .75-.88	.81 .75-.85	.78 .76-.82	.81 .76-.85
Group 2	19	.74 .69-.81	.77 .69-.82	.77 .67-.96	.82 .66-.92
Group 3	25	.88 .81-.92	.85 .78-.91	.78 .72-.84	.76 .7-.83
Group 4	24	.76 .72-.78	.76 .71-.77	.77 .73-.81	.75 .72-.8
Group 5	7	.8 .79-.85	.81 .68-.91	.78 .72-.79	.83 .79-.83
Group 6	2	.82 -	.89 .82-.95	.82 .77-.86	.81 .72-.89
Total	97	.81 .73-.86	.79 .72-.87	.77 .73-.82	.77 .72-.83

## P-555

### The psychological impact of the diagnostic pathway for inherited cardiac conditions in children and adolescents: A systematic review of the literature

Amanda Potterton<sup>1</sup>, Sanjay Sharma<sup>1</sup>, Marcus Wotton<sup>2</sup>

<sup>1</sup>St George's University Hospitals NHS Trust, London, UK; <sup>2</sup>London Southbank University, London, UK

**Background and Aim:** Inherited cardiac conditions are a group of life-threatening cardiovascular disorders genetically passed down through families. Several avenues can be taken to investigate these

conditions, leading to a diagnosis or requirement for long-term surveillance. Diagnosed individuals are at increased risk of sudden cardiac death. Therefore, lifestyle restrictions and/or modifications and possible medical interventions are implemented to reduce risk. Young individuals with a new diagnosis need to come to terms with its impact on their lives, relationships, families, recreational activities and employment.

**Method:** A systematic review was utilised to evaluate the psychological impact of screening for, and diagnosis of an inherited cardiac condition in children and adolescents, to provide increased awareness and guidance for healthcare professionals. A systematic search identified 1139 abstracts, of which 14 met the inclusion criteria; five qualitative, seven quantitative (observational) and one mixed-method. Critical appraisal identified 10 out of the 14 studies as high-strength of evidence, using Joanna Briggs Institute checklists. A convergent-segregated narrative synthesis approach was used to evaluate the research and address the identified studies' heterogeneity.

**Results:** Seven factors were identified to impact illness outcomes, including age, parental influence, family history, familial nature, diagnosed condition, management strategies, and support and understanding. These factors can positively impact, leading to adaptation and improved outcomes; or negatively impact, leading to uncertainty and poor outcomes. The most significant of the negatively impacting factors were family history of sudden cardiac death or adverse event, physical activity restrictions and lack of support and understanding.

**Conclusions:** Psychological challenges were identified throughout the diagnostic and management pathway, impacting children and adolescents on an individualised basis. Further research is necessary to determine more specific factors that impact psychological outcomes for each area of the diagnostic pathway and age and condition-specific challenges. Specialised support through a multidisciplinary team is crucial in promoting understanding, which helps remove uncertainty and improve health outcomes.

**Keywords:** inherited cardiac conditions, sudden cardiac death, psychological impact

#### P-556

##### **What adolescents with congenital heart disease want to know about their heart disease and health including sexual health and contraception**

Åsa Burström<sup>1</sup>, Annika Bay<sup>2</sup>, Malin Berghammer<sup>3</sup>

<sup>1</sup>Department of Neurobiology, Care Science and Society, Karolinska Institutet, Stockholm, Sweden; <sup>2</sup>Department of Nursing, Umeå University, Sweden; <sup>3</sup>Department of Health Science, University West, Trollhättan, Sweden

**Background and Aim:** In adolescence, adolescents with congenital heart disease (CHD) are preparing for the transition to adulthood and adult care. Adolescents need to learn about their CHD and health and develop self-management skills to be active partners in their care. International guidelines have recognized the necessity for health care providers (HCP) to educate and encourage adolescents with CHD to self-management and self-advocacy. However, studies show that adolescents lack knowledge both about their CHD and their health including sexual as well as reproductive health. Before creating health intervention programs, more knowledge of the information needed and how to reach adolescents is a prerequisite.

The aim of this project was to explore what adolescents with CHD describe as important information regarding (i) health, (ii) heart disease, and (iii) sexual health and contraception.

**Method:** Data was collected at/in a workshop at a Nordic youth camp for adolescents with CHD. The participants (n = 35) were 13–19 years old and from all five Nordic countries. They were divided into groups and were asked to discuss and prioritize the three most essential topics they considered important information.

**Results:** The adolescents discussed and prioritized their opinions about important information about health, heart disease, and sexual health, and contraception. Information about risks and restrictions regarding smoking, vaping, alcohol, food, supplementary diets, physical activity, and the possible effect on CHD was considered essential. Further, mental health was mentioned as an important topic to address during the consultations. It was important to start early on with information about CHD, medication, and treatment and it had to be repeated. They asked for information about safe contraception, whether a pregnancy is possible, hereditary, risks with abortion, intercourse, and sexually transmitted diseases (STD). Generally, they wanted specific information related to their heart disease, given both together with peers and individually. However, at the same time, the given information from the HCP must be balanced, where too detailed information could be frightening.

**Conclusions:** Balanced information about health, heart disease, sexual health and contraception is uninterruptedly warranted for adolescents with CHD. To encourage adolescents' self-management and shared decision-making HCP must address these needs in their practice.

**Keywords:** Adolescents, CHD, Knowledge, health, sexual health and contraception

#### P-557

##### **Trametinib for chylothorax management in Noonan syndrome: discussion about efficacy, safety and tolerability**

Teresa Jean Brooks<sup>1</sup>, Alix Xanthe Dunlop Jones<sup>2</sup>, Sarah Joanne Rebecca Whiteley<sup>1</sup>

<sup>1</sup>Department of Pharmacy, Leeds Children's Hospital; <sup>2</sup>Department of Paediatric Dietetics, Leeds Children's Hospital

**Background and Aim:** 3 year old with recurrent chylothoraces. Background of Noonan syndrome (RIT1 mutation), hypertrophic obstructive cardiomyopathy, multiple congenital cardiac abnormalities, and spontaneous bowel perforation with ileostomy. Previous failed treatments for the chylothoraces included medium-chain triglyceride diet, parenteral nutrition (PN) and octreotide.

Noonan syndrome is linked to dysregulation of the mitogen-activated protein kinase (MAPK) pathway (1). Mitogen-activated protein kinase enzyme (MEK) inhibitors disrupt the MAPK signalling pathway. One case demonstrated lymphatic vasculature remodelling and resolution of recurrent chylothoraces with the provision of MEK inhibitor, trametinib, in a patient with Noonan's syndrome (2).

**Method:** Trametinib was accessed through Novartis' compassionate scheme with dosing based on oncology (0.032mg/Kg/day), for 12 weeks of treatment (3). A side-effect management protocol was designed targeting the most common side effects: skin rashes, increased stoma losses, pneumonitis, hypertension and cardiac impairment.

Baseline assessment included: blood testing, tibial growth plate and left wrist X-ray, ophthalmology assessment, physical examination. Throughout treatment monitoring included; weekly bloods and daily physical examination, stoma output measurement and weight. A skin emollient regime was proactively initiated. All baseline tests were normal.

**Results:** Skin irritation worsened within 4 weeks of treatment. It was managed with steroid creams and dressings under dermatology guidance.

Stoma output escalated within 24hrs of starting treatment, to a maximum 45mL/kg, normalising 14days post-treatment. Stoma losses remained high (>20mL/kg) despite loperamide and withholding most enteral nutrition, moving to PN to meet nutritional needs. Enteral intake was limited to small bites of fat-free food, and rehydration solution for stoma loss replacement. Diuretics and weight were reviewed daily, ensuring a neutral fluid balance, appreciating large insensible losses.

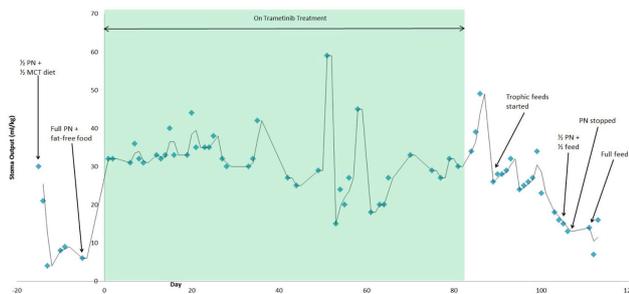
Nausea and vomiting were seen throughout, leading to the provision of anti-emetics and proton pump inhibitors.

ALT and serum triglycerides increased, improving with a reduction in PN lipid provision and infusion time. Gamma GT increased within 1 week of treatment and were not back to baseline at discharge.

There has been no chylothorax recurrence 31 weeks post-treatment.

**Conclusions:** Our experience has demonstrated that it is possible to manage the side effects of trametinib in a patient with multiple comorbidities using a patient centred and MDT approach.

**Keywords:** Trametinib, chylothorax, Noonan syndrome



**Graph:** stoma output throughout admission

## P-558

### Clinical nurse specialist led telephone clinics and the use of the innovative digital platform: ISLA for surveillance of children with a vascular ring

Grace Moriarty<sup>1</sup>, Alice Hurn<sup>1</sup>, Trisha Vigneswaran<sup>1</sup>, Maya Asir<sup>3</sup>, Miriam Giacobbe<sup>1</sup>, Jane Heraghty<sup>2</sup>, Simona Turcu<sup>2</sup>, Kirsty Brennan<sup>4</sup>  
<sup>1</sup>Department of Paediatric Cardiology, Evelina London Children's Hospital, London, England; <sup>2</sup>Department of Paediatric Respiratory, Evelina London Children's Hospital, London, England; <sup>3</sup>Department of Paediatric Speech and Language, Evelina London Children's Hospital, London, England; <sup>4</sup>Department of Paediatric General Surgery, Evelina London Children's Hospital, London, England

**Background and Aim:** To report the use of a symptom questionnaire delivered in a nurse-led telephone triage clinic (NTC) and the ISLA care visual health record ([www.islacare.co.uk](http://www.islacare.co.uk)) for follow-up of infants with a prenatal diagnosis of a vascular ring (right aortic arch with left sided arterial duct (RAA-LD) or double aortic arch (DAA)).

**Method:** From June 2022 the new pathway commenced after first postnatal review, with NTC at 2 and 4 months for DAA and 6 and 12 months for RAA. A questionnaire designed to elicit symptoms of tracheal or oesophageal compression is completed at every

NTC. If symptomatic, patients were referred to an in-person multidisciplinary consultant clinic (MDCC). DAA patients are seen in the MDCC at 4 months. From July 2022, before each NTC, a text link is sent to parents to upload a video of their child breathing and feeding. Submissions are reviewed and actioned by dedicated CNS' for the presence of symptoms. We examined the number of patients followed up in the NTC, versus the number referred to the MDCC, and how NTC/ISLA enabled accurate review without need for an in-person outpatient appointment. Parental feedback was sought.

**Results:** From June to October 2022, 37 patients were referred to the NTC (29 RAA-LD 8 DAA). 17/37 (46%) utilised ISLA, with abnormal respiratory signs present in 6/17 (35%). In 4/37 (11%), symptoms were elicited solely from the questionnaire. 11/37 (30%) were referred to the MDCC, whereas 26/37 (70%) were asymptomatic and managed solely in the NTC. 3/37 (8%) had contacted the CNS team to report symptoms prior to the NTC. Feedback was collected via a multiple choice questionnaire: 100% could easily contact the CNS team if needed. 100% said their overall experience of the service was 'Very Good'. Of the parents that submitted videos to ISLA, 100% stated it was 'Easy' or 'Very Easy' to use.

**Conclusions:** The implementation of a NTC, dedicated CNS team and ISLA has enabled infants with symptoms to be identified and assessment expedited, in addition to avoiding hospital visits and when required, facilitating escalation of care.

**Keywords:** Vascular ring, Nurse-Led, Telephone triage,

## P-559

### Patient and public involvement (PPI) with families affected by sudden cardiac death - considerations and challenges

Jennifer Tollit<sup>1</sup>, Ella Field<sup>2</sup>, Jo Wray<sup>2</sup>, Juan Pablo Kaski<sup>1</sup>  
<sup>1</sup>Institute of Cardiovascular Science, University College London; <sup>2</sup>Great Ormond Street Hospital Foundation NHS Trust, London, UK

**Background and Aim:** Patient and Public Involvement (PPI) is widely acknowledged as fundamental to designing and carrying out high quality research which places patients and their families at its centre. As part of a new research fellowship at the Great Ormond Street Hospital Centre for Inherited Cardiovascular Diseases, a decision was made to form a Patient Advisory Group (PAG) for families of paediatric patients who are undergoing cardiac evaluation due to a family history of Sudden Arrhythmic Death Syndrome (SADS), Sudden Infant Death Syndrome (SIDS) and Sudden Cardiac Arrest (SCA). It is understood that sudden death puts relatives at increased risk of problematic bereavement and poor subsequent adjustment and therefore, it is vital that further research is performed in this area. The aim of the PAG is for patients and their families to help guide research priorities and offer advice on aspects such as approach and methodology to ensure the research is conducted in the most effective and sensitive manner possible.

**Method:** This poster will explore the key themes that emerged from multi-disciplinary exploration of the ethical, psychological, and practical considerations when carrying out PPI with this patient group (parents and carers of paediatric first-degree relatives who have suffered a sudden death or cardiac arrest). Although vulnerable to further psychological distress, existing literature suggests that research participation for bereaved individuals can be a positive experience, providing therapeutic benefits or even attributing a sense of meaning.

**Results:** The main themes for further discussion are:

- Recruitment
- Ethical Considerations and Managing Risk

-Grief, Bereavement and Psychological Implications

-Lessons learned from previous PPI within inherited cardiology

**Conclusions:** PPI with vulnerable groups such as those who have suffered a sudden and unexpected death or cardiac event of a family member requires additional considerations to avoid secondary trauma or adverse psychological impacts in participants. Factors such as participant selection, support offered by the research team and preparing participants for research are key in managing risks to this patient population.

**Keywords:** Sudden Death, Screening, Ethics, PPI

#### P-560

##### **An audit and re-audit of post-transplant blood tests in paediatric cardiology**

*Catriona Drake, Terry Hewitt, Zdenka Reinhardt*

*Department of Cardiology, Newcastle upon Tyne Hospitals NHS Foundation Trust, Newcastle upon Tyne, United Kingdom*

**Background and Aim:** During the Covid-19 Pandemic face-to-face appointments were reduced, resulting in post-transplant patients being reviewed less frequently than routine. This highlighted that some routine blood tests were either delayed or missed and there was, at times, a delay in obtaining results. The aim of the original audit, and subsequent re-audit, was to identify whether delayed or missed blood tests were a problem, to propose a change and to reevaluate.

**Method:** A retrospective eight-week period from 18.01.2021–14.03.2021 was used in the original audit. Electronic notes and blood results for all patients were reviewed. If no blood results were recorded the patient notes were reviewed to determine whether this had been followed up. Changes were made to practice including further education for patients and families, as well as implementing a clinical report system to chase missed tests. A re-audit of the same patients was conducted at one year (17.01.2022–13.03.2022).

**Results:** 124 patients were included. In the initial audit the mean age was 11.1 years and mean time post-transplant 6.7 years. 107/124 patients (86%) had blood tests. 12 patients had a gap >10 weeks between blood tests, 2/12 (17%) were followed up. Patients who missed blood tests were older than 11 years and more than four years post-transplant. In the reaudit 116/124 patients (94%) had blood tests. Seven patients had a gap >10 weeks between blood tests, all (7/7) were followed up. All missed blood tests had reasons documented.

**Conclusions:** With further patient and family education and introduction of our clinical reporting system compliance to the standard has greatly improved with 100% of post-transplant blood tests being followed up.

**Keywords:** Heart Transplantation, Paediatric, Children, Adolescents, Cardiac Transplant

#### P-561

##### **”The heart keeps my child alive”- experience of parents to children with complex rvot anomalies during assessment for cardiac reoperation**

*Birgitta Svensson<sup>1</sup>, Petru Liuba<sup>1</sup>, Anne Wennick<sup>2</sup>, Malin Berghammer<sup>3</sup>*

*<sup>1</sup>Department of Cardiology Pediatric Heart Center, Skåne University hospital, Lund, Sweden; <sup>2</sup>Department of Clinical Sciences Pediatrics, Lund University, Lund, Sweden; <sup>3</sup>Department of Care Science, Faculty of Health and Society, Malmö University, Malmö, Sweden; <sup>3</sup>The Queen Silvia Children’s Hospital, Gothenburg, Sweden; The Department of Health Sciences, University West, Trollhättan, Sweden*

**Background and Aim:** Parents to children with complex right ventricle outflow tract (RVOT) anomalies are confronted with their child’s need for heart surgery early in life and repeated reoperations later on. Detailed preoperative assessment is important in selecting the time for reoperations. This recurrent assessment and uncertainty about the timing of reoperation could cause anxiety and become a burden for the parents. The aim was to illuminate experiences of parents to children diagnosed with RVOT anomalies and how they experience their children’s heart disease and everyday life during the assessment and after the decision on whether to perform reoperation.

**Method:** Individual interviews (n = 27) were conducted with nine parents at three occasions between 2014–2016 and the interviews were analyzed using reflexive thematic analysis.

**Results:** The analysis resulted in five main coexisting themes. “The heart keeps my child alive” illuminate parents’ experiences during and after the examination and emphasize that the heart is central for their child’s survival. There were key moments like the way the doctor’s decision after assessment was handed over, the waiting time from decision to heart surgery, preparing their older child for heart surgery and facilitating the communication during the whole process. “The everyday struggles” illuminate the different struggles’ parents had to face for their child to be found in the best possible condition given. It implied a need to manage a worry far beyond the usual parent-worry and sometimes even the need of a superpower. “The unconditional love”, “The trust to life” and “The way is togetherness” illuminates how and in what way the parents through love, trust and togetherness gain strength and power in their everyday life.

**Conclusions:** Although the parents were grateful for the assessment and have learned to navigate among aroused fears, they experienced several difficult situations during the assessment process that must be addressed. By understanding the parents’ experiences and their sources of strength, the healthcare providers can better aid the parents to find these. Furthermore, by inviting both the parents and their child to participate in the child’s care an individualized support that takes into account both the parent’s and the child’s needs can be created.

**Keywords:** Parents, Children, Right ventricle outflow tract anomalies, Everyday life, Cardiac reoperation, Reflexive Thematic Analysis

#### P-562

##### **Support for families of a child in a palliative situation in the cardiac pediatric intensive care unit**

*Natalie Wieden<sup>1</sup>, Andrea Barbara Eissler<sup>2</sup>, Yvonne Kröger<sup>1</sup>*

*<sup>1</sup>University Children’s Hospital Zurich; <sup>2</sup>University Hospital Bern*

**Background and Aim:** The birth prevalence of children with congenital heart disease is about one percent a year. This might mean that a palliative diagnosis maybe directly exists from birth due to the complexity of the congenital heart disease. The required intensive care stay that may follow after birth presents a challenge for parents. To care for the family, the concept of family-centered care is presented and the involvement of the pediatric palliative care team is considered. The aim was to identify nursing measures which support families of a child in a palliative situation in the cardiac pediatric intensive care unit.

**Method:** This literature search was conducted between January 1, 2022 and May 31, 2022 in Medline via PubMed, CINAHL and Cochrane research databases and was based on defined inclusion and exclusion criteria. Studies from the PICU and NICU as well as studies focusing on end-of-life care were considered, as it can be

assumed that the results may be transferable. Studies with an exclusive oncological focus or specific rare diseases were excluded.

**Results:** Seven main categories could be identified to support the parents. The communication, the parental participation in the decision-making process, continuity of care and relationship building. Also the grieving process and memory making takes place. At least challenges in the intensive care unit and satisfaction with care and unmet needs are highlighted.

**Conclusions:** The included studies suggest important features of communication and can be partially transferred to the implementation of nursing measures with the help of family-centered care. The necessity of the need to involve parents in the care of the child can be demonstrated in the majority of the studies. Therefore, appropriate communication and parental participation in the whole process should be considered as the focus of care. For holistic care, the involvement of the palliative pediatric care team should be evaluated early. Due to the limitations of the studies and the low level of evidence, the results must be viewed with caution. Further research is needed to comprehensively map the specific area of the cardiac pediatric intensive care.

**Keywords:** pediatric intensive care units, family-centered care, pediatric palliative care

#### P-563

##### **An evaluation of a cardiology department's post-operative pain management pathway for children undergoing cardiac implantable electronic devices.**

Helen Walsh<sup>1</sup>, Jasveer Mangat<sup>1</sup>, Ella Field<sup>2</sup>, Juan Kaski<sup>2</sup>

<sup>1</sup>Great Ormond Street Hospital, London UK; <sup>2</sup>University College London Institute of Cardiovascular Science, London UK

**Background and Aim:** Implantation of cardiac electronic devices has been shown to be a painful procedure in recent studies. There has been little research into this area and guidelines to manage this type of procedure do not currently exist. The cardiology department of a large tertiary centre has its own pathway to manage these patients post-operatively. This pathway has never been evaluated; a service evaluation was therefore undertaken to assess the efficacy of the pathway.

**Method:** A retrospective chart review was undertaken, data was collected from 81 patients that had undergone a permanent pacemaker/ implantable cardioverter defibrillator (PPM/ICD) implant in the last 3 years. Patient demographics, characteristics, pain scores, analgesia use and admission lengths were collected.

**Results:** Average pain scores (2.25) indicate pain is tolerable for most patients (70%), however, 69% patients experience moderate-to-severe pain during the first 24-hours. Descriptive statistics showed patients following the pathway had reduced pain scores and fewer opioid doses, although not statistically significant. Unmanageable pain accounted for 54% of delayed discharges, 83.3% of these had not followed the pathway. Pathway adherence was more likely for subcutaneous ICD implantations ( $p = 0.013$ ). Patients were more likely to receive more doses of ibuprofen if the medication was prescribed regularly ( $p = <0.001$ )

**Conclusions:** This evaluation showed the pathway helps to reduce patients' pain and improve the patient journey. Due to the large proportion of patients experiencing moderate-to-severe pain, work is required to improve accessibility to the pathway, improve prescribing practices to ensure around the clock multimodal pain relief is administered and reduce gaps in pain management pathway.

**Keywords:** CIED, ICD, PPM, Paediatrics, Pain

#### P-564

##### **The development of a surgical site infection (SSI) surveillance programme for an Irish paediatric cardiothoracic service**

Malena McLoone, Aisling Feeney, Niamh O Sullivan, Jonathan McGuinness

CHI Crumlin, Dublin 12, Ireland

**Background and Aim:** ECDC PPS 2017 reported surgical site infection rates in Irish Hospitals of over 18%. Any SSI has an adverse impact on both patients and service providers impacting on patient flow, waiting times and outcomes. An absence of routine cardiothoracic SSI surveillance was noted in CHI. In order to meet mandatory requirements and facilitate quality improvement projects, a cardiothoracic SSI surveillance programme was implemented. The introduction of this SSI surveillance programme is reviewed here along with findings on surgical site infection rates.

**Method:** A collaborative approach was used involving the surveillance nurse, surveillance scientist and lead clinicians in both microbiology and cardiothoracic surgery. A paediatric cardiothoracic focused surveillance programme was developed. A robust database was created with customised surveillance form and collection of specific data from both internal and external sources, e.g. NICOR, ICCA, TMS.

**Results:** Data collection and feedback of results promoted buy-in from key stakeholders. Having an identified governance structure and open inter-departmental communication was vital to the success of this SSI surveillance programme. An overall reduction in SSI rates of approximately 80% has been observed to date. Evaluation of the data collected identified a high risk cohort. Microbiological analysis demonstrated our most common isolates and the appropriate antimicrobial surgical prophylaxis required to impact on SSI rates.

A targeted approach for high risk groups and subsequent adaptation to the surgical prophylaxis was agreed and implemented.

Not having an integrated ICT system across the different departments involved has resulted in a reliance on the paper surveillance form.

**Conclusions:** Europe wide paediatric cardiothoracic surgical site infection data is not readily available. Any reduction in infection rates will yield financial and human resource savings. Ongoing multidisciplinary collaboration, team discussions and presentation of meaningful statistics has resulted in a greater awareness and promotion of SSI prevention. Significant improvement in infection rates and a change in antibiotic prophylaxis has occurred in this first year of surveillance. Many more opportunities for pre, peri and post-operative quality improvements will be undertaken using the customised and verified dataset created during the surveillance programme.

**Keywords:** Surveillance, Cardiothoracic, Surgery, Quality assurance, Clinical governance, Healthcare-acquired infection

#### P-566

##### **The heart transplantation - and suddenly, the world is upside down**

Gabriela Oeler, Gaby Stoffel

Children's University Hospital, Zurich, Switzerland

**Background and Aim:** For many children and adolescents with congenital or acquired heart disease, heart transplantation is a treatment option for terminal heart failure. However, heart transplantation goes far beyond surgical intervention and involves a lifelong commitment accompanied by a complex medical

regimen. During their journey, patients and their families face a wide variety of challenges. Before, during and after transplantation, they go through different stages of stress. After leaving the hospital, parents resume the role of primary caregivers for their child and must now care for a child once again with new medically but also emotionally complex needs. The whole family routine is turned upside down and requires adaptation to the new situation. Patients and their families have to learn what it means to cope with everyday life with a new heart and to care for a transplanted child.

**Method:** A nurse-led consultation was established several months ago in a Children's University Hospital. These consultations are led by an Advanced Practice Nurse (APN) with many years of expertise in the care of heart transplanted children, adolescents, and their families. She knows these families previously. The nursing consultations are an integral part of the treatment concept and are coordinated with the medical consultations.

**Results:** The first results confirmed that the nursing consultations contribute to a more holistic support and continuity of care. The families appreciate the opportunity to ask questions about symptom and/or medication management in a familiar atmosphere and to discuss their fears. Individual solutions are discussed with the patients and their families. If necessary, learning sequences are also carried out. It has been shown that the scheduled consultations lead to fewer telephone calls and to higher patient and family satisfaction, thus relieving the treatment team.

**Conclusions:** For a successful outcome, continuity of care and a professional support of these patients and their families, the nursing consultations are part of a holistic care concept. The consultations are appreciated by the families, serve to clarify questions, and can be used for learning sequences if necessary. The family is supported in returning to a normal everyday family life.

**Keywords:** heart transplantation, children, family nursing, advances practice nursing

#### P-567

##### **Developing a psychologically informed paediatric cardiac surgery guide for all north and south of the country of Ireland.**

*Catherine Emma Matthews*

author 1 *catherine matthews, Children's Health Ireland at Crumlin, Dublin, Ireland*

**Background and Aim:** While parents naturally focus on survival and complication rates in surgery they can struggle to find the words for the event they fear the most. A practical psychologically informed guide to paediatric cardiac surgery was developed to support the whole family to work as a team and to emotionally survive cardiac surgery. The All Ireland Cardiac Network performs all of the children of Ireland's cardiac surgery in Dublin. This hospital and the cardiac team may be unknown to families from the North of the country. This guide was developed to familiarise all families North and South to the environs of Children's Health Ireland's cardiac surgery wards and teams.

**Method:** Clinical practise and paediatric psychological research informs the importance of procedure specific and psychologically informed preparation for all medical and surgical procedures. Medically induced trauma is noted as a real risk when embarking on a cardiac surgical journey with unknown timelines in icu and cardiac wards.

A psychologically informed guide for families on what to prepare for and how to prepare was developed.

Parents, children and staff North and South were asked to review and provide feedback to ensure information was accessible and effective.

**Results:** A professional and family review and feedback model ensured the highest degree of relevant and relatable content and mode of communication.

**Conclusions:** A how to guide to prevent trauma and promote resilience when undergoing open heart paediatric surgery was developed in order to best support all of Ireland's cardiac families survive and thrive.

**Keywords:** paediatric, cardiac surgery, psychological trauma, resilience

#### P-568

##### **Thematic classification of paediatric cardiology care conversations by the text analysis software conVIscope**

*Christopher Channing Buckland<sup>1</sup>, Claire Galvin<sup>1</sup>, Astrid De Souza<sup>2</sup>, Richard Lester<sup>3</sup>, Matthew Manson<sup>3</sup>, Fanan Fattah<sup>3</sup>, Raymond Chau<sup>4</sup>, James Potts<sup>2</sup>, Kathryn Armstrong<sup>2</sup>*

<sup>1</sup>*Children's Heart Centre, British Columbia Children's Hospital, Vancouver, Canada;* <sup>2</sup>*Children's Heart Centre, British Columbia Children's Hospital, Vancouver, Canada;* <sup>3</sup>*University of British Columbia, Department of Pediatrics (Cardiology), Vancouver, Canada;* <sup>4</sup>*University of British Columbia, Division of Infectious Diseases, Department of Medicine, Vancouver, Canada;* <sup>4</sup>*University of British Columbia, Institute for Computing, Information and Cognitive Systems, Vancouver, Canada*

**Background and Aim:** The implementation of a SMS (text messaging) platform has been used to enhance the care of adolescents seen in our pediatric cardiology clinic. Characterizing the scope of conversations between adolescents and their healthcare providers (HCP) helps to better understand patients' needs and may influence clinical resource allocation. Artificial intelligence (AI) technology may help to characterize these conversations. WelTel Inc, a global leader in clinical digital messaging, has developed conVIscope; a visual text analytic system trained from a dataset of healthcare conversations to characterize them into pre-defined themes. The aim of our study is to compare the characterization of text message conversations using the pre-defined themes in conVIscope versus manual categorization.

**Method:** This was a retrospective review of text message conversations between a cohort of pediatric cardiology patients and their HCP between Dec 2018 and June 2020. Text messages were uploaded into conVIscope and the software defined care conversations as >2 text messages per interaction. Care conversations were sorted into pre-defined themes. All patient text messages were also analyzed manually and sorted into conVIscope pre-defined themes. Care conversations could have multiple themes.

**Results:** The study cohort included 26 (88% female) patients with a median age of 16.8 years (15.7-17.4). ConVIscope identified more care conversations than manual characterization (384 vs 206). Conversations were classified into the following pre-defined themes: 160 (42%) Symptoms, 23 (6%) Diagnostic, 39 (10%) Treatment, 17 (4%) Lifestyle, 30 (8%) Social, 46 (12%) Logistical, 2 (1%) Health Education, 24 (6%) Service Quality and 0 (0%) Special Topics. ConVIscope misidentified 72% of conversations in the Treatment theme, 62% in the Logistical theme and 41% in the Service Quality theme. It under-reports conversations by 72% in Social, 72% in Lifestyle and 100% in Special Topics. The time commitment for analyzing conversations via conVIscope was considerably lower than manual classification (1.5 hours vs 15 hours).

**Conclusions:** ConVIscope can identify major healthcare themes discussed in care conversations within a pediatric cardiology patient population. Further training of the tool may improve its ability to identify pediatric specific themes.

**Keywords:** digital health, cardiology, pediatric, conversations, artificial intelligence, thematic classification

### P-569

#### 'Expanding the scope of nurse referring for radiological procedures; a collaborative children's health Ireland initiative'

Fionnuala O'Neill<sup>1</sup>, Frieda Clinton<sup>1</sup>, Dr Siobhan Hoare<sup>2</sup>

<sup>1</sup>Children's Health Ireland at Crumlin, Dublin, Ireland; <sup>2</sup>Children's Health Ireland at Temple Street, Dublin, Ireland

**Background and Aim:** The legislation supporting Nurse Referring for Radiological Procedures was enshrined in Irish law in 2008, and NMBI guidelines were developed to support this practice. CHI implemented ANP referring for x-ray examinations in 2015. The utility and effectiveness of nurse referring had thus been well demonstrated in our paediatric setting.

Updated NMBI Guidelines in October 2021 built upon recent legislative changes to support a wider scope of referral including CT, MRI, US and fluoroscopy. Institutional adoption of these new responsibilities in CHI demanded a multidisciplinary approach. Our main drivers were a strong focus on the patient journey and quality safe nursing care.

**Aim/OBJECTIVES:**

- To support the nurse referrer in developing a patient-centred scope
- To develop One CHI policies and pathways for referrers
- To determine audit schedules and feedback mechanisms
- To promote interdisciplinary relationship building to safeguard patient care via improved communication.

**Method:** A multi-disciplinary implementation group was convened, with representation from Nursing, Radiography, Medical Physics and Radiology. A Plan Do Check Act (PDCA) approach was implemented, utilising the Deming cycle method.

- A dedicated session was convened to finalise documentation
- A scope of practice document was developed by each referrer in consultation with their mentor
- It was agreed that nurse referrers meet with modality radiographers and/or medical physics experts to build relationships and knowledge.

**Results:** In July 2022, the first two Registered Advanced Nurse Practitioners were approved to refer for radiological procedures to include CT, MRI and Fluoroscopy. The specific referral scopes were devised by the referrers in collaboration with their mentors initially, and shared in advance. Suggestions from nursing, radiology and medical physics could be incorporated prior to approval as appropriate.

**Conclusions:** • To share this collaborative initiative with our colleagues nationally

- To continue to improve service delivery via safe effective nurse referring

**Keywords:** Nurse, Referring, Radiological Referring

### P-571

#### Developing new pathway for patients with congenital heart disease undergoing non-cardiac surgery and how to plan this

Anna Wieland<sup>1</sup>, Marie Haack<sup>2</sup>

<sup>1</sup>Anna Wieland, Nurse Specialist, department of Adult Congenital Heart Disease; <sup>2</sup>Marie Brix Haack, Nurse Specialist, department of Adult Congenital Heart Disease

**Background and Aim:** We know that adult with moderate and severe congenital heart disease (ACHD) have an increased risk when undergoing non-cardiac surgery and general anesthesia. More than 90 % of children born with congenital heart disease reach adulthood. With that in mind the incidence of non-cardiac surgery will also rise.

The aim is how a multidisciplinary intervention can ensure to prevent cancellations and reduce the patient's risk of complications while undergoing non-cardiac surgery – both pre- peri- and postoperative.

In 2021 a model was introduced at Rigshospitalet (Copenhagen), to identify ACHD patients with risk prior to non-cardiac surgery. This model is published by cardiologists and cardiothoracic anesthesiologists in a Danish medical journal also in 2021.

The purpose is to secure the patient pathway with a minimal risk using this model and how to plan and coordinate.

**Method:** Multidisciplinary meetings were held to discuss and generate ideas for intervention in practice and how to use the model. The multidisciplinary team was consisting of a consultant cardiologist, consultant cardiothoracic anesthesiologists and specialist nurses. All specialized in congenital heart disease.

**Results:** • The patient pathway was considered important. This provides the patient with a sense of being safe.

- Easy access to the department of cardiothoracic anesthesia
- Easy access at specialized knowledge and assessment. Using the model.
- Supervision of collages between medical specialties to increase safety for all health care professionals involved in the patient pathway
- Ensure identification of patient with increased risk of perioperative complications
- Before undergoing anesthesia, the specialist nurses ensure that the right cardiologic assessment has been done.

**Conclusions:** Contributed to an increased focus on ACHD undergoing any type of non-cardiac surgery and general anesthesia. The collaboration between the consultant cardiologist, Consultant cardiothoracic anesthesiologists and specialist nurses has been reinforced.

**Keywords:** Patient pathway, non-cardiac surgery in ACHD,

### P-572

#### Exploring the implementation of a nurse-LED warfarin service for children with congenital heart disease in Ireland – a process evaluation

Jennie Ryan

Department of Cardiology, CHI @ Crumlin, University College Dublin

**Background and Aim:** International advances in healthcare options for children with congenital heart disease (CHD), have resulted in increased survival rates for this patient population. A resulting challenge is the increasing need to use oral anti-coagulation therapy (OAT) for primary thromboprophylaxis amongst this group. The most common indication for OAT in paediatrics is among the CHD population who have complex shunt or valve dependent surgical anatomies. There is an identified need for dedicated OAT clinics to maximise patient safety outcomes.

Guided by implementation science (IS) methods, this project aimed to address an identified quality gap within a national referral centre for children with CHD, requiring treatment with OAT. Its objective was to develop a model of service delivery in line with evidence-based research to empower professionals within the organisation to deliver robust patient-centred care to this patient cohort.

**Method:** Using a descriptive exploratory design and participatory co-design approach, theoretically underpinned and grounded in methods guided by implementation science (IS), a formative process evaluation using the Consolidated Framework for Implementation Science (C-FIR) was conducted. This identified key barriers, facilitators and implementation strategies most valuable to achieving the outcome of 'acceptability' in the pre-implementation stage of this project.

**Results:** The findings demonstrate that implementing a multi-component intervention into healthcare practice is complex. Significant time spent on 'diagnostic analysis' is required to identify pre-implementation organisational barriers and facilitators. Once identified, focused implementation strategies are useful to overcome barriers and achieve acceptability.

This project presents some salient theoretical and professional debates surrounding the implementation of complex interventions, specifically focusing on the crucial role of 'diagnostic analysis' and importance of establishing stakeholder perception in the pre-implementation stage of healthcare projects.

**Conclusions:** In the absence of a best-practice guideline to guide improvements in practice, this project has identified the strategies of 'audit and feedback' and 'facilitation' within the C-FIR framework, as catalysts to achieving the pre-implementation outcome of 'acceptability'. Nursing leadership in the form of skilled facilitation to create opportunity for organisational collaboration was core to this outcome.

**Keywords:** Implementation science, Leadership, Organisational collaboration

#### P-573

##### **Implementation of nurse referral for ionising and non-ionising procedures: A cross site multidisciplinary collaborative project**

*Niamh Maire Maguire, Jennifer Emily Ryan, Fionnuala Gardiner  
Department of Cardiology, Children's Health Ireland at Crumlin,  
Dublin, Ireland*

**Background and Aim:** The All-Island Congenital Heart Disease Network was established in 2015 to provide a single paediatric cardiology service across Ireland delivered through a surgical centre in Dublin, a Children's Heart Centre in Belfast and supported by enhanced paediatric cardiology services at local hospitals, making it the first collaborative cross jurisdictional service of its kind on the island of Ireland.

As part of this network, the Children's Heart Centre @ CHI Crumlin is the tertiary referral centre for paediatric patients who require treatment for congenital and acquired heart disease on the island of Ireland. Practicing within this cross jurisdictional service are 3 registered Advanced Nurse Practitioners (RANPs) who provide expert knowledge and advanced clinical decision-making skills for paediatric cardiac patients. A need was identified for RANPs working within the service to refer for CT, MRI, Ultrasound and fluoroscopy, to deliver a safe and consistent referral pathway for cardiac patients.

Legislation supporting Nurse Referral for ionising radiation procedures was introduced into Irish law in 2009. The aim of this project was to successfully implement nurse referral for CT, MRI, Ultrasound and Fluoroscopy, to delineate the medical and surgical trajectories of patients within this All-Island service.

**Method:** A collaborative quality improvement approach was utilised engaging key stakeholders across CHI sites and disciplines. A multi-disciplinary cross-site committee was established chaired by the Chief Director of Nursing. A PDSA cycle from Deming was employed to progress the implementation process. Stakeholders

engaged in extensive collaborative meetings to standardise documentation across all CHI sites. The Policy on Nurse Referring for Radiological Procedures was completed to support practice using the National Guideline and legislation as a resource, it is supported with governance from both Nursing and Radiology to ensure a safe robust system.

**Results:** All RANPs working within this service can now refer patients for CT, MRI, Ultrasound and Fluoroscopy within their scope of practice.

**Conclusions:** Building collaborative relationships between disciplines was key to the success of this project. It has resulted in the implementation of nurse referral, with the network achieving the first nurse referrers in paediatrics to refer CT, MRI, Ultrasound and Fluoroscopy.

**Keywords:** leadership, communication, imaging modalities

#### P-574

##### **Nutrition intakes and bowel management of paediatric patients on extracorporeal membrane oxygenation (ECMO) to CHI at crumlin**

*Anne Marie Shine<sup>1</sup>, Roisin Mcdonagh<sup>2</sup>, Sunimol Joseph<sup>3</sup>, Mong Hoi Tan<sup>3</sup>, Michaela Pentony<sup>5</sup>, Suzanne Crowe<sup>4</sup>*

<sup>1</sup>Department of Clinical Nutrition and Dietetics, CHI at Crumlin, Dublin

<sup>2</sup>University College Dublin

<sup>3</sup>Department of Critical Care Nursing, CHI at Crumlin, Dublin

<sup>4</sup>Department of Paediatric Critical Care Medicine, CHI at Crumlin, Dublin

<sup>5</sup>Department of Paediatric Cardiology, Children's Health Ireland at CHI at Crumlin, Dublin

**Background and Aim:** The importance of adequate nutrition support for ECMO patients is well documented. Enteral nutrition (EN) is the preferred mode of nutrient delivery to critically ill children. There are many challenges to the provision of EN to this cohort. Poor feed tolerance, feed interruptions and concerns regarding gut perfusion can lead to delayed commencement and sub optimal delivery of nutrition. The two main objectives of this research were to quantify nutrition intakes and to investigate the incidence of constipation.

**Method:** Ethical approval was obtained for this retrospective, observational study. Data from children who received va ECMO for >48hrs during a 12 month period (2020) were reviewed. Patients were identified using the Extracorporeal Life Support Organisation database. Demographic, clinical and anthropometrical data were recorded. Energy and protein intakes via parenteral and EN routes were assessed during ECMO and up to 14 days post cannulation. The frequency of bowel movements while on ECMO were reviewed. Laxative information such as type, frequency of prescription and delivery were recorded.

**Results:** Seventeen patients met the inclusion criteria. The median (IQR) age at the commencement of ECMO was 107 days (4-3564). The primary indications for ECMO were myocarditis (12%), complex CHD (76%) and pulmonary hypertension (11%). The median weight for age Z score at start of ECMO was -1.14 with 17% of subjects having WAZ < -2 on admission. Ninety-one percent of paediatric and 67% of neonatal patients received EN. PN was administered to 82% of patients. The median (IQR) number of days to reach; estimated average energy requirement, basal metabolic rate and recommended protein intakes for the cohort was 16 (14.5-18.5), 6 (6-7) and 7 (5-15) respectively. Of the 17 patients, 94% (n = 16) and 88% (n = 15) had constipation for greater than 3 and 5 days respectively. Forty-seven percent were prescribed laxatives with a cumulative number of 53 prescribed doses and 17 (32%) delivered doses.

**Conclusions:** This study highlights the extended number of days to reach nutrition requirements and the prevalence of undernutrition. Constipation featured frequently and prescribed laxatives were often not administered. These findings support the development of feeding and bowel management protocols for this high nutrition risk cohort.

**Keywords:** Nutrition, ECMO, Constipation

#### P-575

##### **Patient feedback for a new, local paediatric cardiology service; what we can learn from this**

Abby Lauren Nelson, Clare Mckeown, Sinead Callaghan  
Paediatric Cardiology Service, Craigavon Area Hospital, Southern Health & Social Care Trust, Northern Ireland, United Kingdom

**Background and Aim:** A new Cardiology service has been set up in Craigavon Area Hospital in the Southern Trust, this service was local and having longer appointments was a 'one stop shop' for patients. The aim was to record patient feedback to ensure the new service was a benefit to patients and to identify areas of improvement by giving service users a voice for suggestions ensuring it was a positive experience for both patients and their carers.  
**Method:** A patient questionnaire was identified as the best method to record patient feedback. Several drafts of the questionnaires were distributed amongst the team for comments, to develop a questionnaire that was easy to read and would collect useful data. With agreement on an approved version, an online questionnaire was created using Citizen Space, Northern Ireland Government online survey tool. 9 key questions were used for this qualitative study design. The online questionnaire was piloted for 2 weeks to identify improvements. Patients were provided with a leaflet explaining the questionnaire when they arrived at clinic. To eliminate selection bias everyone attending clinic was offered the opportunity to complete the questionnaire. Depending on the age of the patient, either the patient or carer completed the questionnaire.

**Results:**

- 91.3% of patients were satisfied with ease of making appointment and waiting times
- 100% of patients rated the benefit of a local appointment as good or excellent
- 95.65% of patients felt that the 'one stop shop' appointments were helpful
- 100% of patients were satisfied with their consultation, 95.65% rating it excellent and 4.35% rating it good
- Patient comments identified the need for play equipment
- Most common recommendation for improvement was parking facilities at Craigavon Area Hospital

**Conclusions:** Overall, patient satisfaction has been high, with positive feedback on the new service, highlighting the importance of consideration of the impact clinic appointments and access to medical care can have on patient's daily lives. Plans have been put in place for facilities to be equipped with sensory and play equipment to help with waiting times. Further improvement involves information leaflets to new patients explaining procedures and what to expect at clinic in advance of attending appointments.

**Keywords:** Patient Feedback, Service, Improvement

#### P-576

##### **A multidisciplinary collaborative quality improvement approach for children requiring sedation to achieve ECHO imaging in a tertiary referral centre.**

Kathryn Shaw<sup>1</sup>, Jennie Ryan<sup>2</sup>, Adam James<sup>2</sup>, Jacinta Mcginley<sup>3</sup>

<sup>1</sup>Pharmacy Department, Children's Health Ireland, Crumlin, Dublin 12, Ireland; <sup>2</sup>Department of Paediatric Cardiology, Children's Health Ireland, Crumlin, Dublin 12, Ireland; <sup>3</sup>Department of Anaesthesia & Critical Care Medicine

**Background and Aim:** Paediatric patients frequently have difficulty remaining still for transthoracic echo imaging. This can lead to challenges obtaining detailed images required to make accurate clinical decisions and diagnoses.

This project is situated in the national children's heart centre at CHI, Crumlin where approximately 70 patients per annum require sedation for echo imaging. A review of practice revealed significant failure rates in achieving optimal sedation levels using oral chloral hydrate and midazolam. This was associated with a high level of parental dissatisfaction and failure to achieve quality echo imaging.

The aim of this project was to review existing practice and improve quality outcomes relating to this practice.

**Method:** A multidisciplinary collaborative meeting to review practice in other departments occurred and a trial sedation protocol was agreed for all suitable patients > 1 year and over 10kilograms.

A retrospective chart review of patient records before and after the change in sedation protocol was conducted.

**Results:** Of the 10 patients who were given sedation based on existing practice (EP), sedation was effective in 30% of patients (3/10) versus 100% (9/9) using new protocol (NP).

Of EP, 90% (9/10) received chloral hydrate. 6 of the 9 patients received midazolam. 1/10 received midazolam only.

NP – 100% received clonidine, 6 of the 9 patients required buccal midazolam.

20% (2/10) receiving the EP required further admission for a TOE under general anaesthesia, one of which required an overnight admission. None of the patients receiving the NP required overnight admission or escalation of care to obtain imaging.

Good images were obtained in 30% of the patients on the EP versus 100% on NP.

Guardian satisfaction in patients receiving EP was 50% versus 100% on NP.

**Conclusions:** Our results suggest that clonidine and buccal midazolam are more effective than chloral hydrate and oral midazolam for procedural sedation in children with diagnosed or suspected heart disease.

**Keywords:** Treatment, Diagnosis, Medication, Teamwork

#### P-581

##### **Protocolised feeding of infants with congenital heart disease in PICU: A closed loop audit**

Abhishek Oswal<sup>1</sup>, Devika Arambepola<sup>2</sup>, Elizabeth Silverwood<sup>1</sup>, Jessica Brayley<sup>2</sup>, Alice Grove Smith<sup>2</sup>, Kumal Gadhvi<sup>2</sup>

<sup>1</sup>Department of Paediatric Cardiology, Bristol Royal Hospital for Children; <sup>2</sup>Paediatric Intensive Care, Bristol Royal Hospital for Children

**Background and Aim:** Infants with congenital heart disease (CHD) are at risk of malnutrition, due to their increased metabolic requirement, fluid restriction, feed intolerance, impaired absorption and concerns around necrotising enterocolitis (NEC) precluding routine enteral feeding. Studies have shown the benefits of protocolised introduction of feeds, such as reduced rate of infections and quicker establishment of feeds. We audited our practice before and after implementation of a new feeding protocol, to review whether children were able to be fed earlier, and whether it affected rates of serious adverse events.

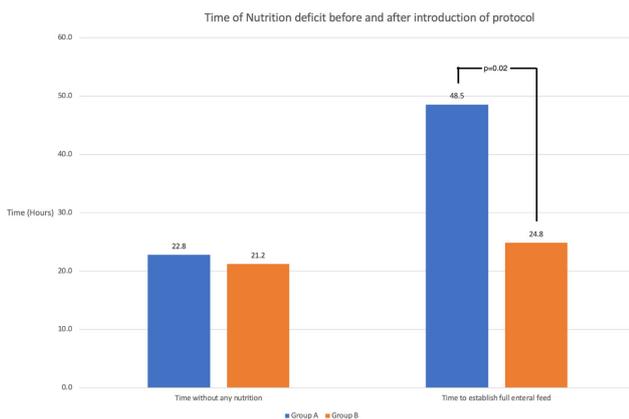
**Method:** We performed a retrospective audit of Paediatric Intensive Care Unit (PICU) patients with CHD over 6 weeks, with defined

audit standards based on departmental consensus. We published a guideline with multi-disciplinary input for risk assessment of infants with CHD and protocols for establishing feeds in both pre- and post-operative infants. We re-audited the same standards 2 months following introduction of the guideline, and performed appropriate statistical analysis to compare the groups, excluding outliers.

**Results:** There were 29 care episodes pre-implementation (Group A) and 22 post-implementation (Group B), each over a period of 5–6 weeks. Group A were younger (0.36 vs 1.65 months) and smaller (3.5kg vs 4.7kg). Group A appeared less malnourished, comparing mean Z score for weight (−0.61 vs −1.23), though not meeting statistical significance ( $p = 0.10$ ). The rates of death, cardiac arrest and NEC were similar in group A and B. The time without nutrition in both groups was similar (22.8 vs 21.2 hours), however the post-implementation group reached full feeds significantly more quickly (48.5 vs 24.8 hours,  $p = 0.02$ ). Documentation of feeding plan, and review from the nutrition support team was less in group B than group A.

**Conclusions:** Infants with CHD admitted to PICU are often malnourished with low weight Z scores. Allowing for our small sample size, our audit has shown that our protocol may improve nutrition by allowing more rapid increases in enteral feeding without an increase in adverse events. We plan to repeat this audit following a staff education programme to improve awareness and compliance with the protocol and clear documentation between the MDT.

**Keywords:** Nutrition, PICU, Feeding, NEC



Key results

#### P-582

##### **Ventricular-ventricular interactions in a desaturated single ventricle patient: A case report**

Joshua D Griesman<sup>1</sup>, David J Barron<sup>2</sup>, Lee N Benson<sup>1</sup>, Steven M Schwartz<sup>3</sup>

<sup>1</sup>Department of Paediatrics, Division of Cardiology, University of Toronto and The Hospital for Sick Children, Toronto, Canada; <sup>2</sup>Department of Cardiovascular Surgery, University of Toronto and The Hospital for Sick Children, Toronto, Canada; <sup>3</sup>Department of Paediatrics and Critical Care Medicine, University of Toronto and The Hospital for Sick Children, Toronto, Canada

**Background and Aim:** Cyanosis can be an important clinical problem in the single ventricle patient. Here, we describe a case of a

newborn with single ventricle physiology and an unusual mechanism to explain cyanosis and low cardiac output. This case highlights the importance of ventricular morphology and ventricular-ventricular interactions.

**Method:** This is the case of a term infant with an antenatal diagnosis of pulmonary atresia with intact ventricular septum. The baby was started on prostaglandin E1 at birth. A post-natal echocardiogram confirmed the diagnosis and documented a large hypertensive right ventricle with a bowing interventricular septum. Attempted wire perforation of the pulmonary valve in the catheter laboratory was unsuccessful, and therefore the arterial duct was stented to secure pulmonary blood flow (Qp).

Within hours, the baby had profound and persistent desaturations to approximately 30% despite multiple maneuvers by the intensive care team. An x-ray showed oligemic lung fields and an emergent echocardiogram showed that the ductal stent was patent, and the hypertensive right ventricle continued to compress the left ventricle. Intravenous fluid boluses consistently had the transient effect of increasing the oxygen saturation for 15–30 minutes at a time. Because this was not sustained, the baby was urgently brought to the operating room for transannular patch repair to allow for right ventricular decompression.

**Results:** The echocardiogram was instrumental in understanding the physiology. The hypertensive right ventricle caused a septal shift which resulted in left ventricular outflow tract obstruction and maintained high left-sided filling pressures. Third space fluid loss from high right-sided filling pressure and decreased left-to-right flow across the stented ductus reduced preload, cardiac output and Qp. Fluid boluses would transiently increase preload, and cardiac output and Qp, but these effects were short-lived as high central venous pressures limited venous return.

This case highlighted the importance of using echocardiography images to construct potential explanations for bedside phenomena. Morphology and ventricular-ventricular interactions may be overlooked when troubleshooting complex physiology, but recognition of these interactions can be the key to finding successful therapeutic options.

**Conclusions:** “I hereby confirm that the consent of the relevant patient(s) has been obtained to submit this Case Reports / Case Series abstract.”

**Keywords:** echocardiography, critical care, single ventricle, physiology

#### P-584

##### **Clinical profile and pre-operative critical care needs of children with transposition of great arteries: A retrospective study**

Navpreet Kaur, Arun Kumar Baranwal, Rakesh Kumar Pilonia, Priyanka Goyal

Department of Paediatrics, Advanced Paediatrics Center, Post Graduate Institute of Medical Education and Research, Chandigarh, India-160012

**Background and Aim:** BACKGROUND: Transposition of great arteries (TGA) is the commonest cyanotic congenital heart disease (CHD) in neonatal period. There is scarcity of data on their pre-operative clinical profile and critical care needs from lower-middle income countries.

AIM: Current study is conducted to identify clinical profile, pre-operative critical care needs and outcome among patients with TGA in a newly commissioned pediatric cardiac intensive care unit.

**Method:** Medical records of all admitted children with diagnosis of TGA during its first 4 years of commissioning (November 2018–September 2022) were reviewed. Data regarding demographic and

clinical details, their critical care needs and final outcome were collected and analyzed.

**Results:** Out of 920 patients admitted, 93 had TGA (male:female; 1.9:1). 87(93.5%) had d-TGA, while 6(6.5%) had congenitally corrected TGA. They were diagnosed to have TGA at postnatal age (IQR) of 19(55) days, however median age of presentation to us was 30(58.5) days. Ten (10.8%) children were detected antenatally. Respiratory distress (75.3%) and cyanosis (69.9%) were common presenting symptoms. Ventricular septal defect (VSD) was present in 55(59.1%), while 32(34.4%) had intact ventricular septum. Critical care requirements included mechanical ventilation (invasive; n = 55, 59.1%; non-invasive; n = 41, 44.1%), inotropic support (n = 46, 49.5%), peritoneal dialysis (n = 1, 1.1%), and red blood cell transfusion (n = 17, 18.3%). 44(47.3%) patients needed prostaglandin E1 for duct-dependency. Balloon atrial septostomy was done in 11 patients at 3(2) days of presentation. 32/93(34.4%) underwent arterial switch operation, while 5 underwent palliation; 18/37(48.6%) of them died. Admission-surgery interval was 19(21) days. Among those who could not be operated upon (n = 56), 23/56(41.1%) died, 21/56(37.5%) left against medical advice, while 12/56(21.4%) patients were discharged for surgery on follow up or due to un-operability.

**Conclusions:** Identification, presentation to the tertiary care center, and subsequent surgery is significantly delayed in our setting. A significant number of these patients needed intensive care pre-operatively.

**Keywords:** Transposition of great arteries, Critical care needs, Clinical profile, Outcome, Lower-middle income country.

#### P-585

##### **Cor triatriatum sinistrum in an adolescent presenting as fulminant cardiogenic shock and resuscitated with extracorporeal membrane oxygenation (ECMO)**

Swathy Subhash<sup>1</sup>, Ajay Desai<sup>1</sup>, Susanna Price<sup>2</sup>, Andres Rico Armada<sup>3</sup>, Richard Trimlett<sup>4</sup>, Barnabe Rocha<sup>5</sup>, Thomas Semple<sup>6</sup>, Jonathan Weale<sup>7</sup>, Anke Furck<sup>8</sup>, Amy Chan Dominy<sup>9</sup>

<sup>1</sup>Department of Paediatric Intensive Care, Royal Brompton Hospital, Guy's & St Thomas NHS Foundation Trust, London, UK;

<sup>2</sup>Department of Adult Cardiac Intensive Care, Royal Brompton Hospital, London, UK; <sup>3</sup>Department of Paediatric Cardiology, Royal Brompton Hospital, London, UK; <sup>4</sup>Department of Adult Cardiac Surgery & Cardiac Intensive Care, Royal Brompton Hospital; <sup>5</sup>Department of Paediatric and Congenital Heart Surgery, Royal Brompton Hospital, London, UK; <sup>6</sup>Department of Radiology, Royal Brompton Hospital, London, UK; <sup>7</sup>Department of Anaesthesia, Royal Brompton Hospital, London, UK; <sup>8</sup>Department of Paediatric Intensive Care and Department of Paediatric Cardiology, Royal Brompton Hospital, London, UK; <sup>9</sup>Department of Paediatric Intensive Care and Department of Anaesthesia, Royal Brompton Hospital, London, UK

**Background and Aim:** Cor triatriatum sinistrum (CTS) is a rare congenital heart defect usually diagnosed in childhood. Cases may remain asymptomatic into adulthood, depending on degree of flow obstruction from the pulmonary venous system into the left ventricle by an intra-atrial fibromuscular membrane, and presence of other anomalies. We report an adolescent who received extracorporeal-membrane-oxygenation(ECMO) resuscitation for fulminant cardiogenic shock and newly diagnosed CTS with major intra-cardiac thrombi.

**Method:** Case report

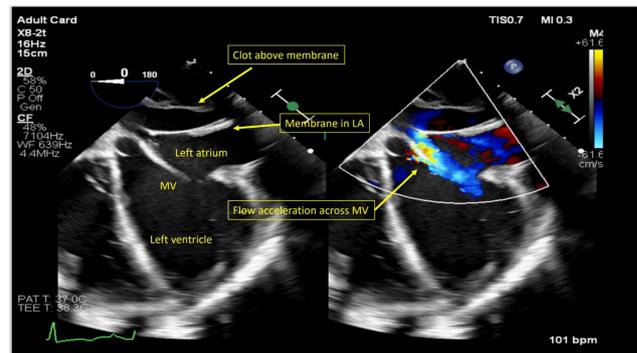
**Results:** A previously well 13-year-old schoolgirl (38kg) presented after several days of cough then breathlessness to local hospital periarrest with profound hypoglycaemia and hyperlactataemia (>22mmol/L). She was referred to our tertiary paediatric cardiac

centre with mobile adult ECMO facility in extremis, vasoactive-inotropic score 150 with profound metabolic acidosis (pH 6.8, base excess -25). Within 2 hours of activation of the adult shock team with paediatric critical care, she was established on peripheral veno-arterial(VA) ECMO via bifemoral cannulation for the 21-mile interhospital transfer. Echocardiogram showed restrictive cor triatriatum with large obstructive left atrial thrombus extending into the 3mm communication between the accessory and true left atrial chambers. CT scan confirmed large filling defect within much of the left atrium part obstructing right pulmonary venous return, and extensive arterial irregularity suggesting a widespread vasculitis, but no intracranial pathology. To optimise cerebral perfusion and mitigate worsening limb ischaemia, she underwent urgent sternotomy for hybrid femoral-aorta VA-ECMO with up-sized central arterial cannula and an additional left ventricular venting cannula, and removal of femoral artery cannula. Extracorporeal life support including renal replacement therapy offered some metabolic stability during evolving multi-organ ischaemic injury. Oseltamivir for intercurrent influenza A and pulsed methylprednisolone were given. Interval imaging and EEG by 64 hours showed no intracranial pathology or seizure activity, and she could localise external stimuli during reduced sedation. Given the high risk of clot embolization with varying flow-filling dynamics, she underwent surgical repair resection of intra-atrial membrane, clot evacuation, and creation of an inter-atrial communication. She remains on VA-ECMO for post-cardiotomy support.

**Conclusions:** This case highlights the potentially life-threatening course of CTS after an asymptomatic childhood. In refractory cardiogenic shock, expedient ECMO offers a mechanical circulatory bridge to diagnosis and treatment including surgery.

**Keywords:** Cor triatriatum, adolescent, cardiogenic shock, ECMO

Echocardiogram findings in cor triatriatum



Echocardiogram showing the opening in the intra-atrial membrane with flow acceleration across and left atrial thrombus

#### P-586

##### **Vitamin C levels in pediatric patients undergoing cardiovascular surgery: A prospective cohort study**

Irene Cattapan<sup>1</sup>, Luca Marchetto<sup>2</sup>, Giovanna Cendon<sup>3</sup>, Marco Manfredini<sup>4</sup>, Fabio Scattolin<sup>5</sup>, Massimo Padalino<sup>5</sup>, Vladimiro Vida<sup>5</sup>, Alvisio Tosoni<sup>6</sup>

<sup>1</sup>Pediatric Intensive Care Unit, Department of Women's and Children's Health, University Hospital of Padua, Padua, Italy; PhD School on

Developmental Medicine and Health Planning Sciences, University of Padua, Padua, Italy; Pediatric Residency Program, Department of Women's and Children's Health, University Hospital of Padua, Padua, Italy; <sup>2</sup>Pediatric Intensive Care Unit, Department of Women's and Children's Health, University Hospital of Padua, Padua, Italy; PhD School on Developmental Medicine and Health Planning Sciences, University of Padua, Padua, Italy; Paediatric Critical Care Fellowship, Department of Paediatric Critical Care, Hospital for Sick Children, Toronto, Canada; <sup>3</sup>Pediatric Intensive Care Unit, Department of Women's and Children's Health, University Hospital of Padua, Padua, Italy; Pediatric Residency Program, Department of Women's and Children's Health, University Hospital of Padua, Padua, Italy; <sup>4</sup>Department of Medical Sciences, University of Ferrara, Ferrara, Italy; <sup>5</sup>Pediatric Cardiac Surgery Unit, Department of Cardio-Thoracic-Vascular Sciences and Public Health, University of Padua, Padua, Italy; <sup>6</sup>Pediatric Intensive Care Unit, Department of Women's and Children's Health, University Hospital of Padua, Padua, Italy

**Background and Aim:** Patients undergoing cardiovascular surgery experience a complex systemic inflammatory response syndrome with an important oxidative stress, generally associated with the application of cardiopulmonary bypass (CPB). Vitamin C (vit C) is a water-soluble micronutrient, not synthesized by humans. It is known for its antioxidant properties, protecting biomolecules from reactive oxygen species (ROS) and preserving endothelial function. In some adult studies, cardiac surgery decreased vit C levels, consistent with increased oxidative stress. No data are available about vit C in pediatric cardiac surgery. The primary objective of this study is to determine pre-operative and post-operative plasma values of vit C in pediatric patients undergoing cardiac surgery.

**Method:** A single-center prospective cohort study is being conducted since May 2021 on children undergoing cardiovascular surgery in our Pediatric Cardiac Surgery Unit in Padua. A blood sample was taken pre-operatively and a second one in the immediate post-operative phase. Plasma concentration of vit C were measured pre-operatively and in the immediate post-operative phase using high performance liquid chromatography and mass spectrophotometry. We considered vit C deficiency as concentration  $<11 \mu\text{mol/L}$  and hypovitaminosis  $<23 \mu\text{mol/L}$ .

**Results:** We prospectively recruited 56 pediatric patients undergoing open heart surgery. Median age was 43 months. Median pre-operative and post-operative vit C were, respectively, 61 (IQR 43.3 – 80.8) and 23  $\mu\text{mol/L}$  (IQR 11 – 35.3). Vit C decreased significantly by 58.8% (IQR 35.6 – 77.1,  $p < 0.01$ ) corresponding to a median perioperative loss ( $\Delta$ ) of 29  $\mu\text{mol/L}$  (IQR 20 – 52). Twenty-seven (46.5%) patients had post-operative hypovitaminosis C, 14 (25.9%) had vit C deficiency. Comparing patients according to their perioperative loss ( $\Delta$ ) (Table 1), duration of the surgical procedure, CPB and Aortic cross-clamp (AxC) were significantly higher in those patients that experienced  $\Delta > 50\%$ .

**Conclusions:** Our preliminary data suggest that cardiovascular surgery induces a significant reduction in perioperative vit C in the pediatric population, similar to what reported in adults. Longer surgical, CPB and AxC time were significantly associated with perioperative loss  $> 50\%$ . This is a preliminary report of an ongoing study whose objective is to determine if post-operative hypovitaminosis is associated with relevant outcomes in the post-operative period.

**Keywords:** Vitamin C, Pediatric Cardiac surgery

Vitamin C levels pre and post-surgery

Table 1. Demographic and intraoperative characteristics of patients according to their pre-operative vitamin C level and perioperative loss of vitamin C ( $\Delta$ )

Baseline characteristic	N	Preoperative vitamin C level			Perioperative loss of vitamin C ( $\Delta$ )		
		$> 50 \mu\text{mol/L}$ N = 19	$< 50 \mu\text{mol/L}$ N = 37	P-value	$\Delta < 50\%$ N = 19	$\Delta > 50\%$ N = 37	P-value
Age (months)	56	61 (2 – 89.5)	13 (5 – 82)	0.768	43 (4.5 – 86)	17 (5 – 82)	0.904
Sex (female)	56	14 (73.7%)	19 (51.4%)	0.186	14 (73.7%)	19 (51.4%)	0.186
BSA ( $\text{m}^2$ )	56	0.72 (0.30 – 0.89)	0.46 (0.33 – 0.84)	0.665	0.63 (0.41 – 0.89)	0.46 (0.31 – 0.84)	0.604
Duration of surgical procedure (minutes)	56	240 (195 – 285)	240 (200 – 285)	0.775	195 (172.5 – 245)	255 (225 – 290)	0.011
Duration of CPB (minutes)	56	122 (72 – 155)	114 (82 – 142)	0.672	74 (58 – 112)	130 (106 – 157)	0.003
Duration of AxC (minutes)	48	83.5 (47.5 – 119.5)	84 (50.75 – 101)	0.844	50 (40 – 90)	92 (62.5 – 105)	0.018
MUF (yes)	56	12 (63.2%)	27 (73.0%)	0.653	11 (57.9%)	28 (75.7%)	0.288

Abbreviations: N, number of patients; CPB, Cardiopulmonary Bypass; AxC, Aortic cross-clamping; MUF, Modified ultrafiltration. Values presented as median (interquartile range) or amount (%).

Demographic and intraoperative characteristics of patients according to their pre-operative vitamin C level and perioperative loss of vitamin C ( $\Delta$ )

## P-587

### Management of patients with left ventricular diastolic dysfunction after radical surgical correction of congenital heart diseases

Yurii Shaviak<sup>1</sup>, Vadym Tkachuk<sup>1</sup>, Ilya Yemets<sup>2</sup>

<sup>1</sup>Department of Anesthesiology and intensive care, Ukrainian Children's Cardiac Center, Kyiv, Ukraine; <sup>2</sup>Department of Cardiac Surgery, Ukrainian Children's Cardiac Center, Kyiv, Ukraine

**Background and Aim:** To change the approaches to the treatment of heart failure with preserved ejection fraction after radical surgical correction of congenital heart diseases in which signs of diastolic dysfunction of the left ventricle appear.

**Method:** In 2020–2022, in our center, 10 patients in the neonatal period after radical surgical correction of a congenital heart diseases (coarctation of the aorta, aortic stenosis) developed clinical signs characteristic of heart failure (tachycardia, shortness of breath, oligoanuria, edema, gastrointestinal disorders, arrhythmia, limitation of physical activity) but according to the echocardiographic examination, the ejection fraction was within the normal range. At the same time, adrenomimetic support was not effective and only infusion of levosimendan gave a slight improvement in the patient's condition. After thorough additional examination of patients by echocardiographic METHOD: Index LA  $\geq 34\text{ml/m}^2$ , PRV  $\uparrow$ , E/e'  $> 15$ ; Diagnostic catheterization of the heart cavities with further determination of the pressures in the heart chambers: end-diastolic pressure of the LV  $\uparrow 14 \text{mmHg}$ . diastolic dysfunction of the left ventricle was diagnosed and adrenomimetic support was radically changed to beta-blockers, namely infusion of metoprolol 1mcg/kg/min.

**Results:** 32 $\pm$ 3 hours after setting up the infusion of beta-blockers, the patient's condition improved, heart rate normalized, diuresis was restored to 6.9 ml/kg/h, the need for oxygen support disappeared, and enteral nutrition began to be absorbed.

**Conclusions:** If there are signs of heart failure but the ejection fraction of the left ventricle is preserved, it is worth investigating the diastolic function of the LV. With diagnosed diastolic dysfunction of the left ventricle, it is better to prescribe not beta-agonists, but beta-blockers.

**Keywords:** dysfunction, beta-blockers

## P-589

**Retrograde flow to the aortic root affects coronary perfusion and impedes cardiopulmonary performance in young fontan patients**Henri Pyykkönen<sup>1</sup>, Otto Rahkonen<sup>2</sup>, Heikki Tikkanen<sup>1</sup>, Päivi Pürilä<sup>3</sup>, Olli Pitkänen Argillander<sup>2</sup><sup>1</sup>Faculty of Health Sciences, School of Medicine, University of Eastern Finland; <sup>2</sup>New Children's Hospital, Helsinki University Central Hospital and University of Helsinki; <sup>3</sup>Unit of Clinical Physiology of the HUS Medical Diagnostic Center, Helsinki University Central Hospital

**Background and Aim:** Single ventricle function in Fontan circulation (FC) depends on systemic venous pressure, pulmonary vascular resistance and ventricular function. Since myocardial function is dependent on coronary perfusion we studied whether in FC retrograde flow from the Damus-Kaye-Stensel anastomosis to aortic root (AoR) affects cardiopulmonary performance (CPP).

**Method:** 26 stable Fontan patients (14.4 + 2.4 years) with right (RV, n = 17) and left systemic ventricle morphology (LV, n = 9) were studied. All RV patients had HLHS and were subdivided according to postnatal flow to the hypoplastic AoR being antegrade (RV-aAoR) or retrograde due to valve atresia (RV-rAoR). Anaerobic threshold (AT), maximal oxygen uptake maxVO<sub>2</sub>, forced vital capacity (FVC), forced expiratory volume in 1 second (FEV1) were measured. These data were correlated with the postnatal size of aorta and current branch pulmonary artery size index (McGoon index). Mann-Whitney U-test and one-way ANOVA tested statistical difference

**Results:** In RV group VO<sub>2</sub>max negatively correlated with postnatal AoR size (R<sup>2</sup> = 0, 6148, p<0.0382). McGoon index did not correlate with VO<sub>2</sub>max. All patients had restrictive non-obstructive lungs. Lung restriction was emphasized in RV-rAoR. **Conclusions:** Young Fontan patients with LV had better CPP over patients with HLHS. In HLHS small postnatal AoR size anticipated lowest maxVO<sub>2</sub> and most restrictive lungs.

**Keywords:** Fontan, Retrograde flow, Maximal oxygen uptake, Lung function

Abstract results table 1

	LV-SVM (n9)	RV-aAoR (n8)	RV-rAoR (n9)	One-way ANOVA
SpO <sub>2</sub> %	94 + 3	94 + 4	93 + 6	p=0.7412
AT (ml/kg/min)	21.5 + 4.3	21.1 + 4.3	18.6 + 5.2	p=0.2795
VO <sub>2</sub> max (ml/kg/min)	32.5 + 5.8	30.0 + 4.2	25.4 + 5.9	p=0.0447
McGoon	2.3 + 0.4	2.0 + 0.3	1.66 + 0.4	p=0.0110
FVC (Z)	-1.20 + 1.3	-2.7 + 1.9	-2.9 + 0.9	p=0.0440
FEV1 (Z)	-1.0 + 1.2	-3.19 + 2.7	-3.3 + 1.1	p=0.0193
FEV1/FVC, %	92	83	85	p=0.8432
Ao-asc (mm)	normal aorta & flow	5.9 ± 1.8	2.8 ± 1.1	p=0.0221
RPA (Z)	0.8 + 1.0	0.6 + 1.2	-0.09 + 1.4	p=0.6769
LPA (Z)	2.0 + 6.5	0.4 + 0.7	-0.5 + 1.7	p=0.4761

Table to clarify the results

## P-590

**Family-centered care and the relationship with parent and infant well-being in two german pediatric cardiac intensive care units**Katharina Rose Luise Schmitt<sup>1</sup>, Hannah Ferentzi<sup>1</sup>, Magdalena Blendemann<sup>1</sup>, Ralph C Rippe<sup>2</sup>, Jos M Latour<sup>3</sup>, Maika Böhm<sup>1</sup>, Michaela Jönebratt Stocker<sup>1</sup>, Alona Girch<sup>1</sup>, Felix Berger<sup>5</sup>, Stephan Schubert<sup>4</sup>, Katharina Hofmann<sup>1</sup><sup>1</sup>Department of Congenital Heart Disease / Pediatric Cardiology, Deutsches Herzzentrum Berlin, Berlin, Germany; <sup>2</sup>Research Methods and Statistics, Institute of Education and Child Studies, LeidenUniversity, Leiden, Netherlands; <sup>3</sup>School of Nursing and Midwifery, Faculty of Health, University of Plymouth, Plymouth, United Kingdom; <sup>4</sup>Department of Pediatric Cardiology, Heart and Diabetes Center NRW, Bad Oeynhausen, Germany; <sup>5</sup>Charité – Universitätsmedizin Berlin, Augustenburger Platz 1, Berlin, Germany

**Background and Aim:** Infants with Congenital Heart Disease (CHD) are at risk for neurodevelopmental delays, emotional, social and behavioral difficulties. Family-centered care (FCC) is a healthcare approach in inpatient care for children. FCC has shown to be effective in improving health outcomes and reducing parental stress, anxiety and depression in neonatology. The aim of this study is to investigate the relationship between parent satisfaction with FCC, parental mental well-being, as well as child physical and mental well-being.

**Method:** In this bicentric cross-sectional study, we included 280 families with infants aged ≤12 months, who were admitted to a pediatric cardiac intensive care unit. We explored socio-demographic factors, complexity of the congenital heart defect (Aristotle Comprehensive Complexity Score), parental satisfaction with the (EMPATHIC-30), quality of life of children (PEDSQL), parental anxiety and depression (DASS-21) and stress (PSS: NICU). Using multivariate statistics and multilevel modeling, we investigated the interplay between socio-demographic factors, severity of the congenital heart defect, parental and child well-being, and parent satisfaction with FCC.

**Results:** Analyses show that in over 80% of the cases, the mothers filled out the questionnaire and 10% of the participants were not a native speaker of German. More than 50% of the children had siblings and the families lived 70 km apart from the hospital, on average. In the analyses, we found a complex relationship between parental satisfaction, parent mental well-being and child physical and mental well-being.

**Conclusions:** This is the first bi-centric study investigating FCC practices in German pediatric cardiac ICUs. Our results show the importance of family-centered care in pediatric cardiology for parent and child well-being.

**Keywords:** Pediatric cardiac intensive care unit, family centered care, congenital heart disease, quality of life, children, cardiac surgery

## P-591

**Summary of ten-year experience with using tissue plasminogen activator (TPA) in children at the gottsegen national cardiovascular center in hungary**Gábor Simon<sup>1</sup>, Brigitta Pongrácz<sup>2</sup>, László Ablonczy<sup>1</sup><sup>1</sup>Paediatric Cardiac Intensive Care Unit, Gottsegen National Cardiovascular Center, Budapest, Hungary; <sup>2</sup>Department of Paediatrics, András Jóna Teaching Hospital, Nyíregyháza, Hungary

**Background and Aim:** Temporary or permanent use of thrombosis prophylaxis is becoming common in paediatric cardiac care due to the increasing number of catheter interventions and prosthetic valve implantations. Nevertheless, serious thromboembolic complications can occur in some cases. If the thrombus formation is intra-cardiac, major damage of the nervous system may develop. In the case of intravascular occlusions, there may be a risk of limb loss. In these cases, thrombolysis with tPA is required to eliminate the circulatory failure. Wildly accepted guidelines, however, are missing for the paediatric age group.

**Method:** In this retrospective review we collect the data of our Paediatric Cardiac Intensive Care Unit patients over the past 10 years from the 1th of May 2012 to the 31th of May 2022.

**Results:** There were 35 cases which required tPA-lysis. Twenty-nine of them were younger than one year old, while eight were between one and 20 years old. Two patients had prosthetic valve thromboses, while 33 children needed thrombolytic therapy following intravascular cannulation. Twenty-seven of all interventions were partially or fully successful. Partial success was defined as improvement of the circulation without total elimination of the blockage according to vascular ultrasound examination. Haemorrhagic complications developed in four patients.

**Conclusions:** More data and development of an international guideline would be necessary for the thrombolysis in children with cardiac disease. In this presentation we would like to give an overview of the thrombolytic therapy strategy used in our institute and to outline our treatment experiences.

**Keywords:** tissue plasminogen activator, thrombolysis

### P-595

#### Mobility and freedom of movement: A novel out-of-hospital treatment for children with terminal cardiac insufficiency and a ventricular assist device

Kathrin Rottermann<sup>1</sup>, Sven Dittrich<sup>1</sup>, Oliver Dewald<sup>2</sup>, Andreas Teske<sup>2</sup>, Nicola Kwapil<sup>2</sup>, Steffen Bleck<sup>1</sup>, Ariawan Purbojo<sup>2</sup>, Frank Muench<sup>2</sup>

<sup>1</sup>Department of Pediatric Cardiology; University Hospital Erlangen, Friedrich-Alexander-University Erlangen-Nuremberg; <sup>2</sup>Department of Pediatric Cardiac Surgery; University Hospital Erlangen, Friedrich-Alexander-University Erlangen-Nuremberg

**Background and Aim:** Due to rapid medical and technological progress, more and more pediatric patients with terminal heart failure are being implanted with a ventricular assist device as a bridge to transplant without legal approval for hospital discharge. EXCOR® Active is a recently developed mobile driving unit for the EXCOR® ventricular assist device (EXCOR® VAD) with a long-lasting battery life that can manage small blood pumps, offering improved mobility for pediatric patients. This study strives to elaborate the requirements necessary for a safe home healthcare environment (HHE) for pediatric patients on EXCOR® VAD powered by the EXCOR® Active driving unit.

**Method:** Patient- and device-related preconditions (medical, ethical, psychological, technical, structural, organizational) were analyzed with regard to feasibility and safety in three individual patient cases. Included were pediatric patients with terminal cardiac insufficiency in a stable medical condition receiving in-hospital treatment with a univentricular or biventricular EXCOR® VAD powered by EXCOR® Active. This single-center analysis included patients between 05/2020-02/2022.

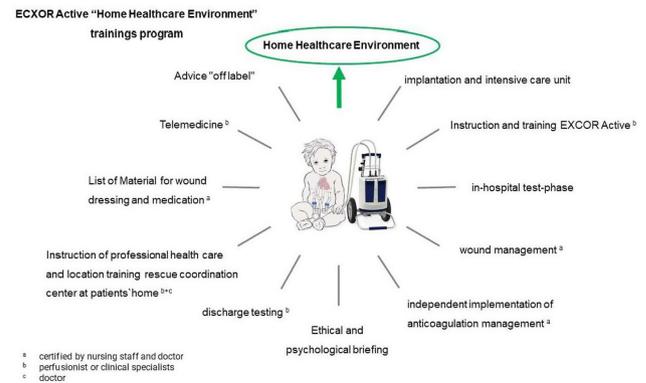
**Results:** A total of three patients on EXCOR® VAD were identified for HHE treatment with the EXCOR® Active driving unit. Switch was performed safely and increased mobility led to improved psychomotor development and improved quality of life. Ethical approval for off-label use was obtained and patients and parents were given the required technical training and psychological support. Caregivers and medical professionals involved in the patients' care at home were briefed intensively. No complications directly related to HHE-treatment occurred. One patient recently underwent an orthotopic heart transplant, one patient remains in HHE, and one patient died due to a complication not related to the HHE. Remote consultations were implemented and interdisciplinary in-hospital checks reduced to a 4-week-scheme.

**Conclusions:** While it is challenging to discharge pediatric patients being treated with a paracorporeal ventricular assist device (EXCOR® VAD) from hospital, it is feasible and can be managed safely with the novel driving unit EXCOR® Active. A home healthcare environment may help to improve patients'

psychomotor development, offer normalized social contacts and strengthen both patients' and parents' physical and mental resources. Legal approval and another study with a larger sample size are warranted.

**Keywords:** Home care environment, ventricular assist device (VAD), EXCOR, pediatric, Heart Failure, tele-medicine

“Clock of action”



“Clock of action” until official hospital discharge with EXCOR® Active

### P-597

#### Liver transplantation for pediatric portopulmonary hypertension

Julie Wacker<sup>1</sup>, Raphael Joye<sup>1</sup>, Valerie Mclin<sup>2</sup>, Barbara Wildhaber<sup>3</sup>, Christian Toso<sup>4</sup>, Leon Genecand<sup>5</sup>, Frederic Lador<sup>5</sup>, Maurice Beghetti<sup>1</sup>

<sup>1</sup>Pediatric Cardiology Unit, University Hospitals of Geneva, Geneva, Switzerland; <sup>2</sup>Pediatric Gastroenterology, Hepatology and Nutrition Unit, Swiss Pediatric Liver Center, University Hospitals of Geneva, Geneva, Switzerland; <sup>3</sup>Division of Pediatric Surgery, University Center of Pediatric Surgery of Western Switzerland, University Hospitals of Geneva, Geneva, Switzerland; <sup>4</sup>Division of Abdominal Surgery, University Hospitals of Geneva, Geneva, Switzerland; <sup>5</sup>Division of Pulmonary Medicine, Department of Medicine, University Hospitals of Geneva, Geneva, Switzerland

**Background and Aim:** Portopulmonary hypertension (PoPH) is a serious complication occurring in 1–5% of children with portal hypertension and in 11% of children with congenital portosystemic shunt. Outcome of pediatric PoPH remains poor. Liver transplant (LT) is indicated in adults with PoPH fulfilling hemodynamic criteria, but data in children are lacking.

**Method:** We retrospectively reviewed all children that underwent LT for PoPH at our center, and described their characteristics and outcome.

**Results:** Among the 217 pediatric LT performed at our center, 4 adolescents (aged 14–19 years) underwent LT for PoPH (due to NASH, biliary atresia with liver cirrhosis, and congenital or surgical porto-systemic shunt). Baseline hemodynamics ranged: mPAP 30–62mmHg, PVR 5–19 WU. Following POPH diagnosis, all patients were treated with pulmonary vasodilators (ERA (1), ERA+PDE-5i (1), ERA+PDE-5i+epoprostenol (2)), and reassessed by catheterization. LT was considered according to adult suggested haemodynamic criteria (mPAP < 35 mmHg) and multidisciplinary discussions when above that cut-off value (low right atrial pressure and normal RV function were then of paramount importance). There was no mortality at a mean follow-up of 5

years. Two patients improved their PAH following LT: one patient completely normalized his haemodynamics; one had a complicated post-LT course with sepsis and severe PH requiring circulatory assistance, but her hemodynamics have then improved, allowing epoprostenol wean. Two patients worsened their POPH: one has significant developmental delay and poor medical adherence, rendering the course of her post LT hemodynamics difficult to interpret; the other had initial post-LT improvement allowing for epoprostenol wean, but then progressively worsened requiring lung transplant 7 years after LT.

**Conclusions:** Liver transplantation may be considered in selected cases of pediatric PoPH, after optimization of hemodynamics with aggressive treatment using PAH-specific therapies.

Hemodynamic criteria for LT need to be better define, but pre-LT right ventricular function seems to be a critical factor. Post-LT outcome based on our small series is encouraging, and deserves further research. Although some patients may worsen their hemodynamics a few years after LT, postponing the need for lung transplantation may result in better outcome than upfront combined lung and LT considering the survival prospect following lung transplantation for PoPH.

**Keywords:** Pediatric, pulmonary hypertension, portopulmonary hypertension, liver transplantation

#### P-598

##### Mid-term results of a nationwide paediatric heart transplantation programme

Karel Koubský, Roman Gebauer, Tomáš Matějka, Tomáš Tláškal, Pavel Vojtovič, Peter Kubuš, Denisa Jičínská, Jan Janoušek  
Children's Heart Centre, 2nd Faculty of Medicine, Charles University in Prague and Motol University Hospital

**Background and Aim:** Heart transplantation (HTx) is an effective treatment for terminal heart failure in children. The aim was to evaluate the outcomes of patients who underwent HTx at a tertiary centre with a nationwide paediatric HTx programme.

**Method:** Retrospective observational study including all patients who underwent HTx from the beginning of the HTx programme in June 2014 till October 2022. Data from the institutional database were used for descriptive statistics and survival analysis.

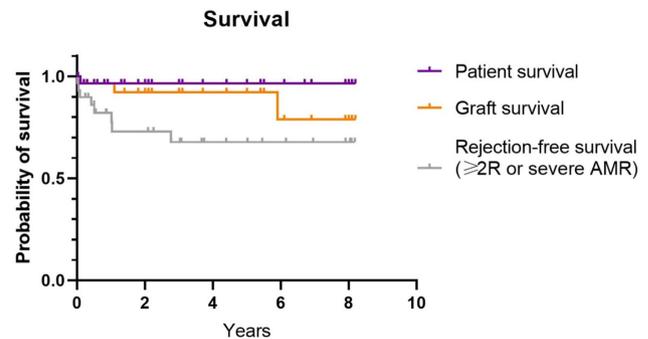
**Results:** A total of 30 HTx were performed in 29 patients with congenital heart disease (CHD, N = 15, single ventricular physiology in 10 patients, median 4 prior cardiac surgeries per patient) and cardiomyopathy (CMP, N = 14). Median age at HTx was 10.2 years (IQR 2.5–14.4) and median time spent on the waiting list was 84 days (IQR 42–228). Ten patients had implanted durable left ventricular assist devices (LVAD) before HTx (Berlin Heart Excor N = 3, Heartware N = 4, Heartmate 3 N = 3) for a median duration of 98 days (IQR 27–150 days). Compared to the CMP group, the CHD group had significantly longer surgery time (median 480 min vs 300 min,  $p < 0.0001$ ) and cardiopulmonary bypass time (median 259 min vs 145 min,  $p < 0.0001$ ), but identical graft ischemic time (median 136 vs 125 min,  $p = 0.27$ ). One patient died early after HTx. There was no late mortality during median follow-up of 3.1 (IQR 0.9–5.7) years. Survival probability at 5 years after HTx was 97%. Two patients underwent re-transplantation (one of them in an adult centre) for late graft failure. Significant rejection-free survival (with significant rejection defined as cellular  $\geq 2R$  or severe antibody-mediated rejection) at 1, 3 and 5 years after HTx was 73%, 68% and 68%, resp.

**Conclusions:** Paediatric HTx programme reflects the complexity of the treated population with about half of patients having complex congenital heart disease and multiple prior cardiac surgeries. One third of the cohort underwent LVAD implantation before HTx.

Mid-term results show excellent survival and modest rejection-free survival. (Supported by Ministry of the Czech Republic – RVO, Motol University Hospital 00064203)

**Keywords:** heart transplantation, cardiomyopathy, univentricular circulation

#### Survival after heart transplantation



#### P-599

##### Speckle tracking echocardiography (STE) imaging for prediction of survival in paediatric patients with pah: A prospective observational study

Shatabdi Giri, Prashant Bobhate  
Children Heart Centre, Kokilaben Dhirubhai Ambani Hospital and Medical Research Institute, Mumbai, India

**Background and Aim:** Pulmonary arterial hypertension (PAH) is a progressive disease with varying ages of presentation and is usually irreversible. Non-invasive assessment of RV longitudinal systolic strain predicts future right-sided heart failure, clinical deterioration, and mortality in patients with PAH. However, its prognostic value for paediatric PH population remains poorly defined. We aimed to use Speckle tracking echocardiography (STE) imaging to assess outcomes in pediatric patients with pulmonary arterial hypertension.

**Method:** Prospective observational study done in single centre tertiary care hospital. Patients  $< 18$  years with PAH were included in the study and those with significant intra/extra cardiac shunt or transient PH were excluded. Study duration: June 2009–June 2022. STE was used to assess RV function at first visit. Primary end point included all cause mortality, patients who underwent Potts shunt or were referred for heart and lung transplant. Statistical analyses were performed using SPSS 20 software and  $p$  value of  $< 0.05$  was considered significant.

**Results:** Out of a total 198 patients in pediatric PAH registry, 155 belonged to Group 1 PAH. 65 patients had significant intra-cardiac and extra-cardiac shunt lesion. Out of the remaining 90 patients 12 patients were either lost to follow-up or had incomplete data, so 78 patients were included in the study cohort. The median age at diagnosis was  $6.97 \pm 5.1$  years with male:female ratio of 1.1:1. Majority presented with symptoms of easy fatigue and right heart failure. RVGLS  $< -12.4\%$ , right atrial strain  $> 19.5\%$ , RVFAC  $> 23.4\%$  and RVEF  $> 24.89\%$ , were associated with better survival. Mean event free survival of study cohort was  $4.2 \pm 0.3$  years with 1, 3 and 5 year survival of 68%, 64% and 64% respectively. RVGLS was found to be most sensitive and RVFAC was found to be more specific marker for predicting survival.

**Conclusions:** Predicting all-cause mortality and classifying PAH patients aides clinical decision-making. The ability for repeated assessment of RV function enables evaluation of disease progress which is feasible with Echo parameters. Noninvasive assessment of RV and RA strain predicts future right-sided heart failure, clinical deterioration, and mortality in paediatric patients with PAH. STE can be a useful marker to predict survival benefits in pediatric patients with PAH

**Keywords:** Speckle Tracking Echocardiography(STE), Pulmonary arterial hypertension(PAH)

Table showing survival in pediatric PAH patients and sensitivity and specificity of STE markers

		P VALUE	SURVIVAL			MEDIAN SURVIVAL (IN YEARS)	SENSITIVITY	SPECIFICITY
			1 YEAR	3 YEAR	5 YEAR			
RVGLS	GROUP 1	0.013	80	80	80	5	78	54
	GROUP 2		47	39	39	1.2		
RA STRAIN	GROUP 1	0.011	73	70	70	6	77	57
	GROUP 2		44	35	35	0.9		
RVFAC	GROUP 1	0.001	79	76	76	6	82	64
	GROUP 2		40	27	27	0.8		
RVEF	GROUP 1	0.004	84	79	79	6	63	74
	GROUP 2		45	34	34	0.9		
MEAN			60	55	55			

RVGLS < -12.4%, right atrial strain > 19.5%, RVFAC >23.4% and RVEF >24.89%, were associated with better survival. Mean event free survival of study cohort was 4.2±0.3 years with 1, 3 and 5 year survival of 68%, 64% and 64% respectively. RVGLS was found to be most sensitive and RVFAC was found to be more specific marker for predicting survival.

#### P-600

##### **Pediatric pulmonary hypertension registry: an Indian scenario**

Shatabdi Giri, Prashant Bobhate

Children Heart Centre, Kokilaben Dhirubhai Ambani Hospital and Medical Research Institute, Mumbai, India

**Background and Aim:** Pulmonary hypertension (PH) is rare and barring few causes, usually irreversible disease eventually leading to progressive rise in pulmonary vascular resistance and right heart failure. There are multiple PH registries available for patients with pulmonary hypertension but majority of them are based on adult data and very few are the Pediatric pulmonary hypertension registries with scarcity of data from developing countries like India.

**Method:** We reviewed data of PH patients presenting to PH clinic. We enrolled all patients with PH between 2months to 18yrs of age. PH was defined by ECHO and cardiac catheterization. We excluded patients with transient PH like, large left to right shunt associated hyperkinetic PH, persistent pulmonary hypertension of newborn (PPHN), single ventricle physiology etc

**Results:** Our registry included 198 patients from January 2012 to June 2021 with mean age 6.6±5.3 years and nearly equal sex ratio (M:F = 0.9). Easy fatigability followed by features of right heart failure were most common presenting features. WHO functional class was III in 26% and IV in 17% at presentation. 78% of the patients had group 1 PH and 69% from this group had associated congenital heart disease. 14% belonged to group 2 and in nearly 1/3rd (31%) patients, etiology was multifactorial PH. Almost 1/3rd (31%) children were not on any medications at the time of presentation and 42% children were on dual vasodilators. Median follow up duration was 17months (range 3 to 83 months) and median survival was 17months (range 1-83months). There were 44 deaths and there was no significant difference between the different

groups of NICE classification of PAH (p value = 0.07). On multivariate analysis, lower age and weight at diagnosis, higher functional class at presentation and higher mean right atrial pressure were associated with increased mortality risk.

**Conclusions:** Pediatric PAH may have different type of presentation and prognosis compared to adults depending upon the underlying etiology. Further long term data and multicenter studies are needed to provide more insight into the disease.

**Keywords:** Pulmonary Hypertension, Pediatric

#### P-601

##### **Selective eating habits lead to pulmonary hypertension and neurological impairment in a child: rare case of thiamin deficiency in the developed world**

Mieke Roggen, Sophie Groignet

Department of Pediatric Cardiology, Cliniques Universitaires Saint Luc, Bruxelles, Belgium

**Background and Aim:** A 3 year old boy had been hospitalized for 1, 5 months for limping and later refusal of walking, when he was referred to our hospital. Marked sinus tachycardia was noted upon arrival and lead to cardiac investigations. Echography showed severe pulmonary arterial hypertension and dilatation of right heart. The child was admitted on the pediatric intensive care unit and immediate treatment (Sildenafil and Bosentan) was started with improvement of the right heart pressure but persistence of the neurological symptoms. Further anamnesis revealed very selective eating habits.

**Method:** They combination of those three elements lead to the suspicion of a Vitamine B1 (thiamine) deficiency.

Diagnosis was confirmed when the child started moving again within 24 hours after Thiamine administration and regained the ability to walk later.

The right heart pressures normalised and the child could be discharged with nutritional supplementations.

After 2 months, Sildenafil and Bosentan were stopped and cardiac catheterization returned normal without pulmonary hypertension.

**Results:** Cardiac beriberi due to thiamine deficiency is considered rare in the developed world. Thiamine is an important cofactor for production of energy and present in meat and vegetables. Severe thiamine deficiency can cause cognitive impairment (Wernicke's encephalopathy), peripheral neuropathy ("dry beriberi") or heart failure (cardiac or wet beriberi).

**Conclusions:** This case confirms that, although a very rare and almost forgotten disease, it's very important to rule out all known causes of pulmonary hypertension as some of them are, although rare, easily treatable.

**Keywords:** pulmonary hypertension, thiamine deficiency, neurological deficiency

#### P-602

##### **Serial echocardiographic indices in relation to transplant-free survival in hypoplastic left heart syndrome**

Aine Lynch<sup>1</sup>, Aamir Jeeva<sup>1</sup>, Chun Po Steve Fan<sup>2</sup>, Wei Hui<sup>1</sup>, Emilie Jean St Michel<sup>1</sup>, Mark K Friedberg<sup>1</sup>

<sup>1</sup>Division of Pediatric Cardiology, The Hospital for Sick Children, University of Toronto, Toronto, ON, Canada; <sup>2</sup>Ted Rogers Computational Program, Peter Munk Cardiac Centre, University Health Network, Toronto, ON, Canada

**Background and Aim:** Systemic right ventricular (RV) dysfunction after stage 1 (S1) palliation is a risk factor for interstage death or transplant in patients with hypoplastic left heart syndrome

(HLHS). However, the trajectory of echocardiographic parameters over time and impact on transplant-free survival (TFS) is poorly understood. We sought to describe echocardiographic trends in a large cohort of HLHS patients and their long-term survival with and without early RV dysfunction.

**Method:** We retrospectively analyzed 668 echocardiograms on 148 HLHS patients who underwent S1 at The Hospital for Sick Children. Echocardiograms were analyzed at 4 pre-specified time points; (1) initial anatomic study prior to S1 palliation, (2) pre-discharge echocardiogram after S1, (3) pre-operative echocardiogram prior to stage 2 or Glenn palliation (S2), (4) pre-operative echocardiogram prior to Fontan(S3). 20 patients were excluded from echo analysis for lack of complete follow up echocardiogram. The primary outcome was TFS and secondary outcomes included recovery of RV function as measured by fractional area change (FAC). Descriptive statistics and Kaplan Meier survival analysis were used.

**Results:** The overall TFS of our total cohort of 128 HLHS patients was 59.3% at 1 year and 47.4% at 5 years. Of our total cohort, 65 (51%) had FAC <35% on Norwood pre-discharge echocardiogram. TFS at 5 years was 41% for those with FAC<35% at S1 discharge, and 57% for those with FAC>35%. Of those with FAC <35% at S1 discharge; 18 (28%) had improvement in FAC to >35% prior to S2 and 9(14%) had normal FAC prior to S3.

Echocardiographic parameters on initial pre-S1 study associated with later dysfunction (FAC<35%) were RV end-systolic area indexed to BSA (21.7cm<sup>2</sup>/m<sup>2</sup> vs 19.9 cm<sup>2</sup>/m<sup>2</sup>, p=0.011), RV basal diameter (2.46cm vs 2.21cm, p=0.001) and mid-RV diameter (2.42cm vs 2.13cm, p=0.005).

**Conclusions:** Our data demonstrates a subgroup of HLHS patients with early RV dysfunction demonstrate improvement in FAC prior to ventricular unloading at time of Glenn procedure. Patients with increased RV dilation on initial echocardiogram were more likely to have longstanding dysfunction. We will further assess the impact of heart failure medication use on RV functional parameters over time in this cohort.

**Keywords:** Hypoplastic left heart syndrome, heart failure, outcomes

### P-603

#### Doppler tissue imaging in assessment of pulmonary hypertension secondary to congenital heart disease

Doaa Mohamed Abdel Aziz<sup>1</sup>, Hala Mounir Agha<sup>1</sup>, Mohamed Ibrahim El Kalioby<sup>2</sup>, Badr Elden Mohamed Mesbah<sup>2</sup>, Waleed Attia Diab<sup>2</sup>, Mohamed Samir Eid<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology, Cairo University, Cairo, Egypt;

<sup>2</sup>Department of Pediatric Cardiology, Suez Canal University, Ismailia, Egypt

**Background and Aim:** Pulmonary hypertension is defined as a mean pulmonary artery pressure equal to or more than 25 mm Hg at rest or 30 mm Hg on exercise. Doppler tissue imaging (DTI) is a relatively new echocardiographic technique that uses Doppler principles to measure the velocity of myocardial motion. We aimed in this study to emphasize the role of Doppler tissue echocardiography as a non-invasive technique in assessment of ventricular function and ventricular-ventricular interaction in patients with pulmonary hypertension secondary to congenital heart disease (PAH).

**Method:** A cross sectional analytic study was conducted among (34) children aging between 2 and 16 Yrs, who had congenital heart disease with left to right shunt and did diagnostic cardiac catheterization at the Cardiac catheterization laboratory, Cairo University

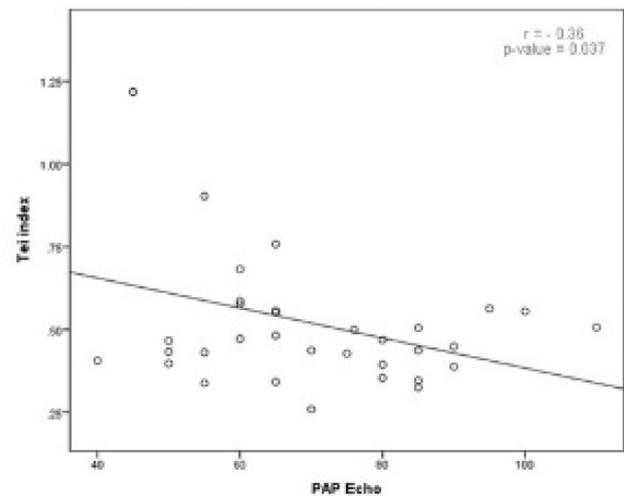
Specialized Pediatric Hospital. Echocardiographic assessment of the pulmonary artery pressure was done and DTI variables were calculated to estimate it's the effect on the right ventricular functions.

**Results:** The most common congenital heart disease was Ventricular septal defect (VSD) in 65% of patients followed by Complete A-V canal in 24% of patients. The Pulmonary arterial pressure had a mean of 69.59±17.17 mmHg as measured by Echocardiography and a mean of 67.47±19.25 mmHg as measured by cardiac cath with a positive direct significant correlation ( r= 0.56, p value = 0.001\*). Analysis of the data on the Pulmonary artery pressure (PAP) and Doppler tissue outcome variables on the right ventricle (RV) revealed a positive significant correlation between isovolumetric contraction time (IVCT) and pulmonary artery pressure (PAP) (r= 0.38) (P= 0.025). Tei index showed a negative significant correlation with PAP (r= -0.36, P= 0.037). Comparison of the different grades of Pulmonary hypertension (PAH) and DTI variables revealed a statistically significant difference in the peak late diastolic velocity (a') outcome measures among mild (11.75±4.59), moderate (14.93±3.12), and severe (17.33±4.56) grades of PAH (P= 0.018).

**Conclusions:** Doppler tissue imaging can be used as a reliable method for follow up of patients with PAH associated with congenital heart disease and to predict it's adverse outcomes. However, cardiac catheterization remains the gold standard in the diagnosis and evaluation of PAH

**Keywords:** Doppler tissue imaging, Pulmonary hypertension, isovolumetric contraction time (IVCT), Tei Index, peak late diastolic velocity (a')

Correlation analysis between Tei Index of the RV and PAP



### P-604

#### The right ventricle longitudinal strain relation to determinants of risk in children with pulmonary arterial hypertension - preliminary experience.

Monika Kowalczyk Domagala, Malgorzata Zuk, Dorota Jagiellowicz, Alicja Mirecka Rola

Pediatric Cardiology Department The Children's Memorial Health Institute Warsaw, Poland

**Background and Aim:** The right ventricle (RV) function is an important part of evaluation patients with pulmonary arterial

hypertension (PAH). The aim of this study was to evaluate global RV and regional free wall RV two-dimensional strain and relation to determinants of risk in children with PAH according to 2019 Updated Consensus Statement on the Diagnosis and Treatment of Pediatric Pulmonary Hypertension.

**Method:** Twenty six children with PAH were studied. We divided patients into 2 groups: group 1 lower risk (11pts) and group 2 intermediate and higher risk (15pts). Data were presented as median (interquartile range). RV global and regional free wall (basal, middle, apical) longitudinal strain (LS) was assessed in children with PAH. Strain analysis was performed during last echo.

**Results:** Median follow-up duration was 20.3 months (19.4-21.6) for all children; 20.4 mo (19.5 – 21.5) for group 1 and 20.2 mo (19.4 – 21.5) for group 2, NS. Median age was 12.1 years (9.8 – 14.1) in whole group; 12.8 (10.4 – 14.2) in group 1 and 12.1 (9.8 – 13.9) in group 2, NS. Median RV global strain was -16% (-17.7 – -11.7%) for all patients; -17.7% (-23.1 – -17.1%) for group 1 and -12.3% (-15.5 – -10.7%) for group 2. There were significant difference between group 1 vs group 2 ( $p < 0.001$ ). Median RV free wall strain was -20.2% (-25.1 – -17.6%) for all children; -25.5% (-27.3 – -20.7%) for group 1 and -18.6% (-21.6 – -13.5%) for group 2. There were significant difference between group 1 vs group 2 ( $p = 0.002$ ). Significant differences between groups were also observed in basal, middle and apical strain (group 1 vs group 2,  $p$ ):

- Basal: -27.2% (-30.2– -23.0) vs -18.8% (-21.7– -14.5), 0.002
- Middle: -24.3% (-26.5– -20.4) vs -17.1% (-22.2– -12.8), 0.01
- Apical: -20.6% (-25.8– -18.2) vs -18.5% (-20.8– -15.6), 0.03

**Conclusions:** RV global and regional longitudinal strain were significantly reduced in children with PAH with intermediate and higher risk. RV strain may be useful as an additional parameter in monitoring a patient with PAH.

**Keywords:** Right ventricle 2-D strain, echocardiography, pulmonary arterial hypertension, children

**P-605**

**Understanding right ventricular functional response to exercise in children with pulmonary arterial hypertension by comparison to elite athletes**

Dan M. Dorobantu<sup>1</sup>, Jessica E. Caterini<sup>2</sup>, Curtis A. Wadey<sup>3</sup>, Diane Ryding<sup>4</sup>, Steve McNally<sup>4</sup>, Dave Perry<sup>4</sup>, Barbara Cifra<sup>5</sup>, Luc Mertens<sup>5</sup>, A. Graham Stuart<sup>6</sup>, Craig A. Williams<sup>2</sup>, Guido E. Pielers<sup>7</sup>, Mark K. Friedberg<sup>5</sup>

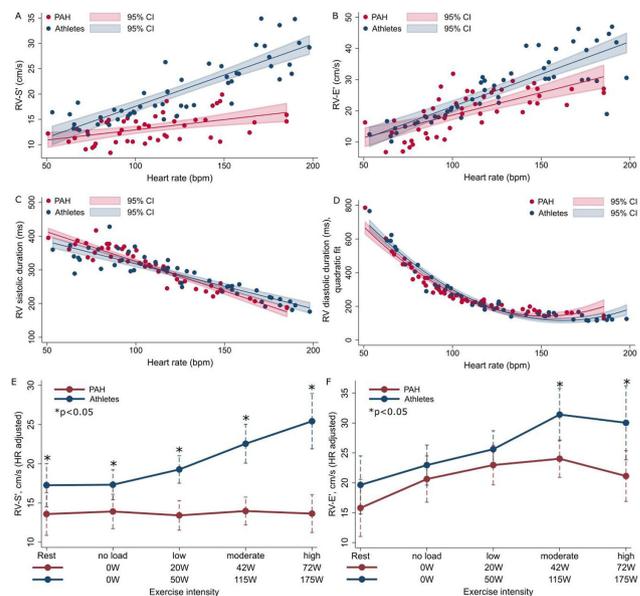
<sup>1</sup>Children’s Health and Exercise Research Center (CHERC), University of Exeter, Exeter, UK; <sup>2</sup>Congenital Heart Unit, Bristol Royal Hospital for Children and Heart Institute, Bristol, UK; <sup>3</sup>Queen’s Medical School, Kingston, Canada; <sup>4</sup>Children’s Health and Exercise Research Center (CHERC), University of Exeter, Exeter, UK; <sup>5</sup>Manchester United Football Club, Football Medicine & Science Department, Manchester, UK; <sup>6</sup>Labatt Family Heart Centre, Hospital for Sick Children, University of Toronto, ON, Canada; <sup>7</sup>Congenital Heart Unit, Bristol Royal Hospital for Children and Heart Institute, Bristol, UK; National Institute for Health Research Cardiovascular Biomedical Research Centre, University of Bristol, UK; <sup>7</sup>Sports Medicine Department and the Athlete Health and Performance Research Centre, ASPETAR Qatar Orthopaedic and Sports Medicine Hospital, Doha, Qatar; Institute of Sport Exercise and Health, University College London, London, UK; National Institute for Health Research Cardiovascular Biomedical Research Centre, University of Bristol, UK

**Background and Aim:** Children with pulmonary arterial hypertension (PAH) have right ventricular (RV) dysfunction and prolonged systolic duration at rest. Exercise induced tachycardia, which may worsen relative systole-to-diastole prolongation, could limit

exercise capacity. Athletes are a physiologically relevant population to understand cardiac responses to exercise, and by contrast PAH maladaptive mechanisms. Consequently, we aimed to compare RV functional responses to exercise in paediatric PAH patients versus athletes, hypothesising that limited exercise capacity in PAH is associated with disproportionate increases in systolic duration.

**Method:** Elite football players and idiopathic PAH patients <16 years-old (10-pairs matched by age, resting heart rate (HR) and exercise duration) underwent cardiopulmonary exercise testing with concomitant tissue Doppler echocardiography at 50W/20W increments. Exercise intensity domains were defined based on the individual gas exchange threshold (GET). Peak tricuspid annular systolic and diastolic velocities (RV-S’, RVE’) and RV systolic and diastolic durations were measured at each exercise intensity. Differences between groups were evaluated using repeated measures mixed models, in relation to HR and exercise intensity. **Results:** Mean age of participants (n = 20) was 13.9 ± 2.2 years (males PAH/athletes = 4/10). Peak oxygen consumption (20.1 ± 5.8 vs 42.7 ± 6.4 mL•min<sup>-1</sup>•kg<sup>-1</sup>,  $p < 0.001$ ), work-rate (72 ± 16 vs 219 ± 40 W,  $p < 0.001$ ) and HR (154 ± 25 vs 191 ± 5 bpm,  $p < 0.001$ ) were reduced in PAH vs athletes. The HR–RV-S’ relationship throughout exercise was blunted in PAH patients vs. athletes ( $p < 0.001$ , Figure A) and to a lesser degree the HR–RVE’ relation ( $p = 0.01$ , Figure B). Mean HR was lower in PAH compared to athletes at and above low intensity exercise ( $p < 0.05$ ). Compared to athletes, systolic functional response was impaired in PAH patients even at low intensity exercise, while diastolic functional response was impaired at ≥ moderate intensity (Figure E, F,  $p = 0.2$  and  $p = 0.8$ , respectively). The systolic and diastolic duration–HR relations were similar between groups throughout exercise ( $p = 0.9$  and  $0.3$ , Figure C and D).

**Conclusions:** RV impairment during exercise in PAH versus athletes appears related to reduced systolic and diastolic functional reserve rather than disproportionate prolongation of systolic duration. This suggests that despite chronotropic insufficiency, exercise associated tachycardia is tolerated in children with PAH, and may allow them to exercise beyond the GET into high-intensity domain, even with reduced RV functional reserve.



Relation between RV-S' (A), RV-E' (B), systolic duration (C) diastolic duration (D) and heart rate (HR) during exercise in pulmonary hypertension (PAH) and athletes. Mean and 95% confidence interval RV-S' (E) and RV-E' (F) by exercise intensity (with mean work-rate) in PAH and athletes, adjusted by HR at each step.

**Keywords:** pulmonary arterial hypertension, right ventricular function, exercise physiology, adolescents, athletes, tissue Doppler imaging

#### P-606

##### **Pediatric heart transplantation in different age groups**

Csaba Vilmányi, Edgár Székely, Zsolt Prodan, László Ablonczy  
Gottsegen National Cardiovascular Center

**Background and Aim:** Cardiac transplantation (Tx) can be considered as the ultimate and sole therapy of end-stage heart failure in children. Children of varying ages, due to their non-uniform body size, face different health and psychosocial issues not just while on the waiting list but also following transplantation. Smaller body-size supposedly associated with longer waiting times, would more often need bridging to transplantation by MCS, while at the same time their immature and permissive immune system predisposes them to less rejection. Based on these premises our study focused on the data-analysis of our enlisted patients, while waiting for a suitable donor organ, their eventual requirement for MCS and different outcome variables.

**Method:** Since the establishment of the pediatric cardiac Tx program in our institution in 2007, we performed 61 procedures. From the 66 enlisted patients 1 was delisted due to improvement of his cardiac status and 4 deceased prior to Tx (three on MCS and one without MCS). Median age at the time of enlisting was 8.9ys (36 kg, 131 cm). Patients were divided in two groups (less than 6 and older than 6 years of age). We studied the days spent on the waiting-list, the need and time spent on MCS, as well as the complications occurring on MCS. Further, the occurrence of hemodynamically relevant rejection episodes were also analysed.

**Results:** The gender ratio (11/10 vs 22/18 M/F) was almost identical in the two groups. The time spent on the waiting list was significantly longer for patients in group 1 (206 vs. 108 days  $p = 0, 045$ ). Neither the requirement (6/21 cases 28% vs. 11/40 27%) nor the duration (150 vs. 113 days), and major complications encountered during MCS (3/6 cases vs. 5/11 cases) differed significantly. The incidence of at least grade 2 or hemodynamically relevant rejections occurred in 4/21(19%) and 7/40 (17.5%) patients in the compared groups respectively.

**Conclusions:** In view of the Hungarian data, the time spent on waiting list is significantly longer for younger patients, but neither the incidence, nor the major complications of the MCS showed inter-group differences. The post tx rejections were rare overall, without differences between the groups.

**Keywords:** waiting list, heart transplantation, age group

#### P-608

##### **Sacubitril/valsartan in treatment of heart failure with left ventricle systolic dysfunction in infant with congenital LV aneurysm: case report**

Julia Kalinicheva, Zhareydat Gagieva, Anastasia Kharisova, Kamil Kadyrov, Denis Petrushenko  
Department of Cardiac Surgery, Children's Medical Center, Kazan, Tatarstan, Russia

**Background and Aim:** Heart failure (HF) in pediatric patients has a significant impact on morbidity and mortality. Treatment strategies of pediatric HF are based on extrapolation from adults. One of those is the application of ARNI (neprilysin inhibitor + Rc AT II inhibitor), which showed good results in children during the PANORAMA - HF study.

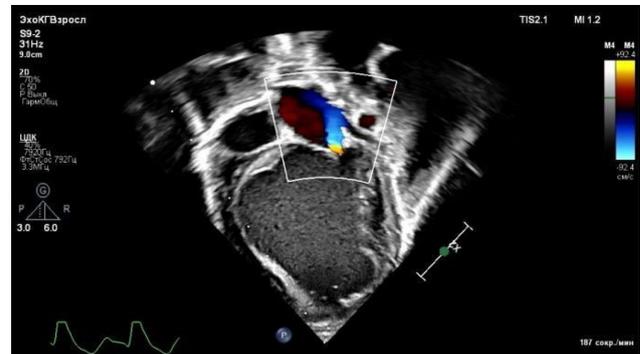
**Method:** 3.2 kg boy was born at term with prenatal history of COVID-19 at 30 week g.a. Large LV aneurysm was revealed at week 32. The child was stable at birth. US, CT coronary angiography, cardiac MRI showed a pronounced dilatation of the LV with the large zone of dyskinesia on the inferior and lateral LV walls 27\*28 mm with transmural edema of the myocardium (myocardial infarction (EF 25 %). Coronary arteries were enlarged, but without signs of stenoses or aneurisms. High level of Ig G to COVID-19 were revealed, as well as increased NT-proBNP up to 35, 000 pg /ml. Signs of HF had increased progressively in spite of escalation of conventional treatment: ACE inhibitors, diuretics, b-blockers, digoxin and aldosterone inhibitors. Patient became ROSS III functional class. The size of the LV increased, mitral insufficiency complicated with venous pulmonary hypertension appeared. Surgical resection of aneurysm was impossible, because of its large size Pulmonary artery banding was denied because of the irregular shape of LV. Due to the ineffectiveness of conventional therapy, it was decided to switch it to sacubitril/valsartan off-label at an initial dose of 0.8 mg/kg/day.

**Results:** Improvement of symptoms was noted in 2 to 4 days: functional class ROSS II, weight gain, reduction of the LV size and mitral valve insufficiency, decrease of NT-pro BNP level to 5000 pg/ml. Blood pressure was slightly decreased after initiation of the first dosage. Nowadays the child is 5 months old, body weight 6.1 kg, the dosage of sacubitril/valsartan is 1.8 mg/kg/day.

**Conclusions:** Clinical case demonstrates the effectiveness and safety of a new class of drug for treating pediatric population with HF due to severe systolic LV dysfunction in case of ineffectiveness of the conventional therapy and as a bridge to heart transplant or VAD implantation.

**Keywords:** heart failure, LV systolic dysfunction, congenital LV aneurysm, sacubitril/valsartan

#### Congenital LV aneurysm



#### P-609

##### **Ukrainian pediatric center for pulmonary arterial hypertension: organization of work in times of war**

Valentyna Khanenova<sup>1</sup>, Arkadii Dovgaliuk<sup>1</sup>, Anna Razinkina<sup>1</sup>, Nadiia Rudenko<sup>2</sup>, Ilya Yemet<sup>1</sup>

<sup>1</sup>Ukrainian Children's Cardiac Center; <sup>2</sup>Shupyk National Healthcare University of Ukraine

**Background and Aim:** The Ukrainian Center for Pulmonary Arterial Hypertension (UCPAH) in Children was created at the Ukrainian Children's Cardiac Center in 2015. The main objectives of the UCPAH were: creation of a general register of patients in the pediatric group with PAH, conducting highly specialized PAH

diagnostics, optimizing drug therapy, monitoring its effectiveness and assessing the dynamics of the clinical condition of patients, conducting cardiac surgery, providing emergency care to patients with PAH complications in the ICU, increasing the level of knowledge on the diagnosis and treatment of children with PAH. The purpose of the work: to analyze the main problems, methods to improve the efficiency of the UCPAH and changes in the demographic composition of the UCPAH's patients, taking into account the military situation in the country.

**Method:** The military invasion of the troops of the Russian Federation into the territory of sovereign Ukraine began on February 24, 2022. Under martial law, the organization of medical care in Ukraine required special measures to ensure the work of medical institutions.

**Results:** During 7 years 207 patients with PAH were registered in UCPAH. There were 112 females and 95 males. All patients were with diagnosis of idiopathic PAH (iPAH) – 44 (21, 3%) patients and PAH – associated with congenital heart disease (PAH-CHD) – 163 (78.7%) patients. In group of PAH-CHD 41 (25 %) was after total repair of CHD, 72 (44.1%) – after palliative operations, 50 (30, 9%) – non operative.

According to the data of the check of the demographic composition of the patients of the UCPAH for the period from February 24 to November 1, 2022, 98 (47.3%) patients changed their place of residence due to hostilities, of which 63 (64.2%) became federal emigrants, and 37 (35.8%) – internally displaced persons. 10 (4.8%) patients died as a result of disease progression or infection.

**Conclusions:** During the period of martial law in the country, the main problems in the work of the UCPAH were: saving the staff of the clinic, ensuring patients' access to medical care, and finding resources to resume the supply of specific therapy drugs, because of problems with the state budget program

**Keywords:** Pulmonary Arterial Hypertension, martial law, demographic composition, medical care.

## P-610

### Pulmonary hypertension and right ventricular remodeling in children with severe bronchopulmonary dysplasia in the first 5 years of life

Devi Gnanam<sup>1</sup>, Eline Smorenburg<sup>1</sup>, Tarik Karamass<sup>2</sup>, Irwin Reiss<sup>3</sup>, Liesbeth Duijts<sup>4</sup>, Beatrijs Bartelds<sup>1</sup>, Laurens Koopman<sup>1</sup>

<sup>1</sup>Division of Pediatric cardiology, Department of Pediatrics, Erasmus MC - Sophia Children's Hospital, University Medical Center Rotterdam, Rotterdam, The Netherlands.; <sup>2</sup>Department of Pediatrics, Division of Neonatology, Department of Pediatrics; Erasmus MC - Sophia Children's Hospital, University Medical Center Rotterdam, Rotterdam, The Netherlands.; <sup>3</sup>Division of Neonatology, Department of Pediatrics; Erasmus MC - Sophia Children's Hospital, University Medical Center Rotterdam, Rotterdam, The Netherlands.; <sup>4</sup>Division of Respiratory Medicine and Allergology, Department of Pediatrics, Erasmus MC - Sophia Children's Hospital, University Medical Center Rotterdam, Rotterdam, The Netherlands.

**Background and Aim:** BACKGROUND: Preterm-born infants with BPD are at risk to develop pulmonary hypertension (PH), which is associated with cardiac remodeling.

AIM: 1) Assess the prevalence of PH in preterm-born infants with BPD after discharge 2) Assess right ventricular (RV) structure and function in these patients.

**Method:** We prospectively studied 161 preterm-born infants with severe BPD (supplemental oxygen required  $\geq 28$  days, and need for  $\geq 30\%$  O<sub>2</sub>, > 1litre min/flow and/or continuous airway support at 36 weeks gestational age at 6 months corrected age). Follow-up visits at 6 months and 5 year using standard

echocardiography and measurement of strain. PH defined as tricuspid regurgitation jet velocity  $\geq 2.8\text{m/s}$  and/or flattening of the interventricular septum in systole.

**Results:** The prevalence of PH at the age of 6 months was 6% (10/161). There was no differences in gestational age, birthweight and weight at follow-up between infants with PH and without PH. There was no differences in LV circumferential and longitudinal strain between infants with PH and without PH. Patients with PH at 6 months corrected age had lower RV lateral wall longitudinal strain as compared to those without PH. So far 48 out of 161 children reached the age of 5 years, 6 previously diagnosed with PH. Of those followed-up until age 5 years none had signs of PH. Yet, RV end-diastolic dimension (RVEDD) Z-score of those with early PH remained higher compared to patients without early PH ( $1.6 \pm 0.4$  vs  $0.9 \pm 0.5$   $p = 0.003$ ).

**Table:** Baseline characteristics and echocardiographic measurements at 6 months corrected age.

**Conclusions:** The prevalence of PH in preterm infants severe BPD in our prospective cohort study was low (6%) in infancy and absent in childhood. Infants with PH had increased RV dimensions and subtle signs of reduced RV function. The preliminary medium-term follow-up at age 5 implicate persistence of RV remodeling.

**Keywords:** Bronchopulmonary dysplasia, pulmonary hypertension, strain echocardiography, right ventricle function, right ventricle dimensions.

TABLE: Baseline characteristics and echocardiographic measurements at 6 months corrected age.

	Infants with PH (at 6 months) n=10	Infants without PH (at 6 months) n=151	p value
Gestational age (weeks)	26(24-26)	26(25-27)	0.084
Birth weight (grams)	755(518-927)	770(645-910)	0.589
Weight at 6 months (grams)	7.2 $\pm$ 1.4	7.0 $\pm$ 1.0	0.457
RVEDD (cm)	1.6 $\pm$ 0.3	1.4 $\pm$ 0.3	<b>0.013</b>
RVEDD Z-score	1.2 $\pm$ 0.7	0.7 $\pm$ 0.8	<b>0.032</b>
RVESA (cm <sup>2</sup> )	3.7(3.1-4.2)	3.2(2.6-3.7)	<b>0.048</b>
Mean RV lateral wall LS	-20.6( $\pm$ 5)	-24.5( $\pm$ 5.5)	<b>0.03</b>

Values: mean (standard deviation) or median (25-75 interquartile range), p value: t-test or Mann Whitney test as appropriate reflecting differences baseline and echocardiographic measurements between infants with and without pulmonary hypertension (PH). RVEDD, right ventricle end-diastolic dimension; RVESA, Right ventricle end systolic area; RV, right ventricle; LS, longitudinal strain.

## P-613

### An accessory donation: uncovering a wpw pattern in a heart transplant child recipient

Alejandro Martín Asenal Schaffi, Luis García Guereta, Antonio Cartón Sanchez, Federico Gutierrez Laraya, Alvaro Lafuente Romero  
Pediatric Cardiology Department, Hospital Universitario la Paz, Madrid, Spain.

**Background and Aim:** Arrhythmias in the post-transplant period are not uncommon, although less frequent since the introduction of bi-caval suture technique. Moreover, preexcitation syndrome is fairly common in the general population and a potential substrate for arrhythmias, hence the importance of knowing if a heart suitable for donation is affected by this condition. Our aim is to raise awareness of the importance of EKG evaluation prior to transplant.

**Method:** We present the case of a 6-year-old girl with restrictive cardiomyopathy that, after heart transplantation from a donor with no reported history of heart disease, experienced self-limited episodes of supraventricular tachycardia (SVT).

**Results:** Initial postoperative EKGs showed no clear signs of WPW pattern or other arrhythmogenic findings except for intermittent small slurring at the beginning of the QRS in lead II. However, first attempts to wean from the atrial temporary pacemaker would trigger wide QRS tachycardias (suspected to be SVTs for its clinical tolerability and EKG characteristics).

Due to the unknown origin of these arrhythmias, further inquiries on the donor revealed EKG findings consistent with an overt left lateral accessory pathway that wasn't reported during the organ-donor briefing. No history of palpitations nor arrhythmias was reported in the donors chart.

Two weeks after transplant, pacemaker could be removed with no tachyarrhythmia recurrences and invasive testing showed no high-risk characteristics. Months later a new SVT episode was detected and the family reported frequent palpitations episodes, so intervention was decided.

An electrophysiology study (EPS) confirmed the presence of the accessory pathway and inducible AV reentrant tachyarrhythmias. Pathway ablation was successfully performed with no further signs of preexcitation in follow-up EKGs.

**Conclusions:** There is few available data regarding transmission and behaviour of preexcitation syndrome in pediatric transplant. We consider that revision of donor's EKG during comprehensive briefing is crucial to ensure the best outcomes, since any overlooking may cause significant morbimortality. Wolff-Parkinson-White syndrome has been considered in some studies as contraindication for donation. However it should not preclude transplantation as accessory pathways may be ablated if needed.

**Keywords:** Transplant, Preexcitation, WPW, Donor, Arrhythmia

#### P-615

##### Neonatal dilated cardiomyopathies: diagnosis and outcomes at a tertiary center.

Cosimo Marco Campanale<sup>1</sup>, Rachele Adoriso<sup>2</sup>, Gessica Ingrassiotta<sup>2</sup>, Valeria Donato<sup>1</sup>, Patrizio Moras<sup>3</sup>, Marco Masci<sup>1</sup>, Maria Carolina Colucci<sup>4</sup>, Yeva Guk<sup>1</sup>, Andrea Dotta<sup>5</sup>, Luca Di Chiara<sup>6</sup>, Antonio Amodio<sup>2</sup>, Luciano Pasquini<sup>1</sup>, Alessandra Toscano<sup>1</sup>

<sup>1</sup>Perinatal Cardiology Unit, Department of Medical and Surgical for Fetus-Neonate, Bambino Gesù Children's Hospital IRCCS, Rome, Italy; <sup>2</sup>Unit of Heart Failure and Transplant, Department of Pediatric Cardiology and Cardiac Surgery, Bambino Gesù Children's Hospital IRCCS, Rome, Italy; <sup>3</sup>Pediatric Department, University of Tor Vergata, Rome, Italy; <sup>4</sup>Unit of Pediatrics, NEMOS Department, Sapienza University, c/o Sant'Andrea University Hospital, Rome, Italy;

<sup>5</sup>Neonatal Intensive Care Unit, Department of Medical and Surgical for Fetus-Neonate, Bambino Gesù Children's Hospital IRCCS, Rome, Italy; <sup>6</sup>Cardiac Intensive Care Unit, Department of Pediatric Cardiology and Cardiac Surgery, Bambino Gesù Children's Hospital IRCCS, Rome, Italy.

**Background and Aim:** Dilated Cardiomyopathies (DCMs) are a heterogeneous group of myocardial diseases, resulting in ventricular dilation, cardiac dysfunction, heart failure and even death. Scarce evidence is available about DCMs with neonatal onset (Neonatal DCMs, NDCMs). We described our experience with NDCMs in terms of time at diagnosis, clinical and outcomes in the first year of life.

**Method:** All patients from January 2019 to August 2022 presenting with NDCMs diagnosed in fetal life or within the first 2 months were collected; fetal and post-natal echocardiographic ultrasounds and clinical data were reviewed.

**Results:** Among 12 NDCMs, 5 (42%) were diagnosed pre- and 7 (58%) post-natally. One interruption of pregnancy was performed

and 1 was not born yet. At birth mean weight was  $2.785 \pm 0.6$  kg and GA  $36 \pm 2$  weeks. Length of hospital stay at first admission varied from 12 to 428 days (median 60 days). Etiology was genetic in 5, secondary to coronary anomaly in 1 and the rest still of uncertain origin. Cardiac phenotypes included: global LV dilation and dysfunction in 7 (58%), Ventricular apex aneurysm in 3 (25%), LV dilations with multiple ventricular septal defects in 1 (8.5%) and LV dilation within significant mitral regurgitation in 1 (8.5%). Mean Fetal GLS and LVEF were reduced ( $-8.4 \pm 2.4\%$  and  $24.4 \pm 6.4\%$  respectively). Mean LVEF was  $34 \pm 15\%$  at admission and  $41 \pm 15\%$  at 1 year. At birth none was intubate and only 2 started inotropes: these were patients with fetal diagnosis who were born inside our hospital for predicted clinical instability at birth. Intubation was needed for 1 pt at one week and 2 pts at one month, inotropes in 1 at one week and 4 at one months. ECMO/VAD were present in 1 at one month, in two pts at 6 months and in one at 12 months. One pt was transplanted and one was dead at one year.

**Conclusions:** NDCMs have heterogenous cardiac phenotype and etiology. After a stable presentation at birth a rapid deterioration at 1 month can be observed (inotropes dependency). The endpoint of death/transplant is about 20% at 1 year.

**Keywords:** heart failure, dilated cardiomyopathy, neonates

#### P-616

##### COR triatriatum sinister and aortic coarctation, a rare association

Maria Daniela Romero Espinoza<sup>1</sup>, Flavio Zuccarino<sup>2</sup>, Fredy Prada<sup>1</sup>, Stefano Congiu<sup>3</sup>, Joan Sánchez De Toledo<sup>1</sup>, Maria Clara Escobar Díaz<sup>1</sup>  
<sup>1</sup>Department of Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain; <sup>2</sup>Department of Radiology, Hospital Sant Joan de Déu, Barcelona, Spain; <sup>3</sup>Department of Cardiovascular Surgery, Hospital Sant Joan de Déu, Barcelona, Spain

**Background and Aim:** Cor triatriatum sinister (CTS) is a rare congenital heart disease (CHD) in which the atrium is divided into two distinct chambers by a fibro-muscular septum. The proximal chamber drains the pulmonary venous blood while the distal chamber contains the atrial appendage. The physiologic consequences of CTS are related to the size of the orifice between the chambers, with pulmonary hypertension when the orifice is obstructed.

CTS may be associated with other cardiac anomalies in up to 80% of cases, but is rare its association with aortic coarctation.

**Method:** Case Report

**Results:** We present a fetus who was diagnosed at 20 weeks of gestation with high suspicion of coarctation and persistent left superior vena cava (PLSVC) with dextrocardia. Genetic study was normal. A female newborn was born at 39 weeks with 3100 grams. Postnatal echocardiography confirmed the coarctation and showed also a membrane dividing the left atrium above the left atrial appendage diagnosing a CTS. Signs of pulmonary hypertension were present since birth. A CT-scan confirmed the diagnosis and showed that the PLSVC was draining directly to the left atrium. Surgical correction was performed at 9 days of life with intra-atrial membrane resection and extended coarctation repair. During the postoperative period, the patient persisted with pulmonary hypertension and signs of systemic venous congestion with chylous ascites, pleural and pericardial effusion, requiring drainage and parenteral nutrition. A hemodynamic study confirmed a high left ventricle end-diastolic and post-capillary pulmonary pressure. Treatment with diuretics and milrinone was continued until pressure normalized.

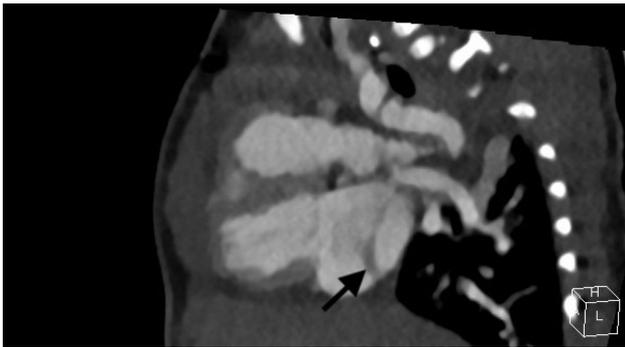
After 2 months of treatment, the patient was discharged. Currently (3 months) she is asymptomatic.

In this case, the location of the heart (dextrocardia) with a posterior left atrium, made difficult the diagnosis. The presence of persistent pulmonary hypertension and a high suspicion of obstructed pulmonary venous drainage helped us to diagnose CTS. The CT-scan, made a more accurate diagnosis.

**Conclusions:** Cor triatriatum sinister is rare and its association with aortic coarctation even more. CTS should be suspected in patients with persistent pulmonary hypertension and obstruction to pulmonary venous return. CT-scan may help to better define the anatomy of the left atria.

**Keywords:** Cor triatriatum, aortic coarctation, pulmonary hypertension

#### Atrial Membrane



Thin oblique membrane that divides the left atrium above the appendage

#### P-617

##### **The development of idiopathic pulmonary hypertension during treatment with methylphenidate in pediatric patient with attention-deficit hyperactivity D**

Soumia Fillal<sup>1</sup>, Katarzyna Polak<sup>2</sup>, Jose Felix Coserria<sup>1</sup>, Inmaculada Guillén<sup>1</sup>

<sup>1</sup>Paediatric Cardiology department, Hospital Universitario Virgen del Rocío, Sevilla, Spain; <sup>2</sup>Pediatric department, Hospital Universitario Nuestra Señora la Candelaria, Tenerife, Spain

**Background and Aim:** Methylphenidate (MPH) is federal drug administration approved central nervous system stimulant commonly used for attention deficit hyper-activity disorders (ADHD) in children. The pulmonary hypertension (PHA) is a known adverse effect of MPH overdose due to intravenous abuse yet extremely rare in patients treated with therapeutic doses.

**Method:** We report the case of our patient, who presented PHA during therapeutic treatment with MPH. Consequently, using the PRISMA protocols, we performed a review of literature published within the last 20 years.

**Results:** We present a male patient who underwent annual follow-up visits in pediatric cardiology consult since birth due to the family history of a brother who died at 15 months of age because of severe PHA. The annual echocardiographic (ECHO) controls were performed and resulted normal until 9 years of age. The patient presented to consult at 10 years of age referring episodes of hypotonia, dizziness, hearing loss and blurred vision and ECHO revealed signs of PHA with increased tricuspid regurgitation pressure gradient (59 mmHg) and flattened interventricular septum in systole (I/II).

The guided interview revealed that 18 months prior the patient was diagnosed with ADHD and started treatment with MPH.

Diagnostic catheterization revealed elevated mean pulmonary pressure (22 mmHg) and pulmonary artery resistance (3.34 U/m<sup>2</sup>) with a negative vasoreactivity test. The genetic tests were conducted and resulted negative.

Treatment included bosentan and sildenafil and suspension of MPH, with complete remission of symptoms. Currently at 17 years of age the patient remains asymptomatic and doesn't present complications.

The literature review identified 2 articles reporting PHA and its asociacion to therapeutic use of MPH, prescribed for ADHD and narcolepsy with concomitant features of obstructive sleep apnea in males of 15 and 17 years of age, respectively.

**Conclusions:** Although the exact cause of PHA could not be determined in this case, the literature describes association of PHA and therapeutic use of MPH, our patient presented normal ECHO controls until 18th month of treatment with MPH and presented good outcomes after its suspension, we posit a possible correlation in between therapeutic MPH use and PHA development, especially in predisposed patients with family history of PHA.

**Keywords:** Pulmonar hypertension, ADHD, metilphenidate

#### P-618

##### **The development of idiopathic pulmonary hypertension during treatment with methylphenidate in patient with attention-deficit hyperactivity disorder**

Soumia Fillal<sup>1</sup>, Katarzyna Polak<sup>2</sup>, Jose Felix Coserria<sup>1</sup>, Inmaculada Guillén<sup>1</sup>

<sup>1</sup>Paediatric Cardiology department, Hospital Universitario Virgen del Rocío, Sevilla, Spain; <sup>2</sup>Pediatric department, Hospital Universitario Nuestra Señora la Candelaria, Tenerife, Spain

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**Keywords:** pulmonary hypertension, methylphenidate, ADHD

**P-619**

**Beyond 100 days: single center experience of outcomes and complications of long-term vad as a bridge to cardiac transplantation in children**

Nicola Boyd<sup>1</sup>, David Blundell<sup>1</sup>, Emma Simpson<sup>2</sup>

<sup>1</sup>Paediatric Cardiology, Freeman Hospital, Newcastle-upon-Tyne, UK;

<sup>2</sup>Paediatric cardiac intensive care, Freeman Hospital, Newcastle-upon-Tyne, UK

**Background and Aim:** Ventricular Assist Devices (VADs) sustain life in children with cardiac failure as a bridge to transplantation. Over recent years, patients awaiting transplantation have experienced longer VAD runs, due to scarcity of organs. As one of two centers in the UK offering VAD therapy, we analysed the complications experienced by children with paracorporeal (Berlin Heart EXCOR™, BH) VADs whose run lasted beyond 100 days.

**Method:** We performed a retrospective analysis of consecutive patients who had BH implanted between December 2014 and May 2021 (following conversion from a short run of Levitronix VAD or ECMO). We compared mortality and outcome in long run patients (>100 days) with that of short run patients. Morbidity was defined as renal if dialysis > 1 month, gastroenterological if TPN >one month, NEC or laparotomy, sepsis if positive blood cultures and neurological if seizures or stroke.

**Results:** 14 patients had a long BH run (mean 249 days, range 106–732). Mean age and weight of this cohort was 20 months (range 4–

67) and 9.6kg (4.3–16.5) respectively. Of the 14 patients: 7 died (1 following transplantation), 7 were discharged following successful heart transplant. The mortality rate of long BH runs was higher than with short runs (50% vs 35%). The burden of stroke (9/14) and pump exchange (10/14 patients (mean changes 2.3; range 1–8)) was also high. Blood stream infections were ubiquitous within this group. A summary of the complications is included in Table 1.

**Conclusions:** Low organ donation rates and an increase in new transplant list registrations have led to longer VAD runs in children awaiting transplantation. This data demonstrates the complications of VAD runs over 100 days and the consequences regarding mortality and morbidity. Whilst advances in anticoagulation strategies may reduce CVA and VAD change burden, infection remains a significant issue both with resultant time off the transplant list and as a contributory factor to mortality.

**Keywords:** Transplant, Mechanical support, Berlin Heart

**P-621**

**Risk factors of heart failure and prognosis in ebstein anomaly**

Dimitri Herrera Nativi<sup>1</sup>, Fanny Deville<sup>1</sup>, Diane Ouwrier Buffet<sup>1</sup>, Julia Mitchell<sup>1</sup>, Olivier Metton<sup>1</sup>, Sylvie Di Filippo<sup>2</sup>

<sup>1</sup>Department of Congenital Heart Disease, Claude Bernard University, Lyon, France; <sup>2</sup>Congenital Cardiology, Cardiothoracic Center of Monaco

**Background and Aim:** Ebstein anomaly outcomes often complicate with heart failure (HF). The objectives of the study were to analyze the occurrence and risk factors of heart failure in children and adults with Ebstein anomaly.

**Method:** Single-center study including patients diagnosed with Ebstein anomaly over a 20-year period of time.

**Results:** 76 patients were included, 34 males (45%), aged 21.9 ± 21.4, median 13.9 (21 neonates (27.5%), 20 children (26.5%) and 35 adults (46%)). Symptoms at diagnosis in 47 (62%) included cyanosis (23), dyspnea and heart failure (35), palpitations (13), while 29 (38%) had no symptom. NYHA class at diagnosis was I, II, III and IV in respectively 55%, 35.5%, 6.5% and 3% of cases. Age at diagnosis was higher in class III (p < 0.05). Median pulse oxymetry at diagnosis was 98% (60% to 100%), the lowest in NYHA class III (88%) and class IV (75%) patients. NYHA functional class worsened over time. Ebstein anomaly was major type in 19% (14 cases), minor in 35%, intermediate in 46%. ASD was present in 54 (71%), VSD in 8 (10%), pulmonary hypoplasia in 5 (6.6%), accessory pathway in 12 (16%). Supraventricular tachycardia occurred in 33 (43.4%). Mean age at SVRT was 25.3 years, 45.3 years at atrial fibrillation (p = 0.0003). Pacemaker was implanted in 9 (12%). HF occurred in 20 (26.3%). Mean age at first HF episode was 39.9 years. At least one procedure was performed in 44 (58%): 11 tricuspid valve replacement (14.5%), 15 ASD closure (19.7%), 12 ablations (15.8%). Freedom from HF was 90%, 80%, 50%, and 30% at respectively 40, 50, 60 and 75 years of age. Risk factors for HF include: major type (43.7% vs 20%, p = 0.05), neonates (41%) and adult age at diagnosis (28.5% vs childhood (10%), p = 0.09, arrhythmias (43.7% vs 13.6%, p = 0.03), TR grade III–IV vs I–II (38.5% vs 16.3%, p = 0.03), number of interventions (12.5% if none, 22 to 27% if 1–2, 40% if 3 and 83% if 4 interventions, p = 0.006). Three patients died (4%), all from heart failure.

**Conclusions:** HF complicates outcomes of Ebstein anomaly. Anatomical type, youngest or oldest age at diagnosis, arrhythmias, tricuspid regurgitation and number of interventions are risk factors for HF.

Patient	Age at VAD implantation (months)	Weight at VAD implantation (kg)	Original cardiac diagnosis	RVAD/BIVAD/LVAD	Duration of BH (days)	Tracheostomy (Y/N)	VAD pump exchanges	Renal impairment (Y/N)	GI complication Operation (O)/ NEC (N)	Infection (Y/N)	CVA (Y/N)	Outcome: Death (D) Transplanted (T)
A	5	5.1	DCM	LVAD	190	N	1	N	N	Y	Y	D
B	53	16.5	DCM	LVAD	106	N	2	N	-	Y	N	T
C	39	15	DCM	BIVAD	331	Y	3	Y	N	Y	Y	D
D	20	11.8	DCM	BIVAD	106	Y	1	Y	-	Y**	N	T
E	5	6.18	DCM	LVAD	732	N	8	N	-	Y	Y	T
F	37	14	DCM	BIVAD	180	N	0	N	N	Y	Y	D
G	7	7.6	RCM	BIVAD	343	N	1	N	-	Y	Y	D
H	12	8.3	DCM	BIVAD	266	N	1	Y	-	Y	Y	T+D
I	8	7.3	DCM	LVAD	181	N	0	N	O (PR bleed)	Y	Y	T
J	4	5	DCM	LVAD	306	N	1	N	-	Y	Y	D
K	8	6.28	DCM	LVAD	167	N	1	N	-	Y	Y	D
L	67	16.1	DCM	RVAD*	116	N	0	Y	-	Y	N	T
M	4	4.3	DCM	LVAD	286	Y	0	N	-	Y	N	T
N	18	10.2	Shone complex	BIVAD	173	N	4	N	N	Y	N	T

DCM = dilated cardiomyopathy; RCM = restrictive cardiomyopathy; LVAD = left ventricular assist device; RVAD = right ventricular assist device; BIVAD = biventricular assist device; VAD = ventricular assist device; CVA = cerebrovascular accident

\* RVAD BH whilst on Heartware LVAD

\*\* Endocarditis requiring aortic valve replacement

**Table 1** Morbidity & Mortality Outcomes for Long BH Runs

**Keywords:** Ebstein anomaly, heart failure, risk factor, prognosis

**P-622**

**Long-term survival and functional status of adult patient with Eisenmenger syndrome**

Sylvie Di Filippo<sup>1</sup>, Fanny Deville<sup>1</sup>, Diane Ouvrier Buffet<sup>1</sup>, Julia Mitchell<sup>1</sup>, Annandre Bozio<sup>1</sup>, Sylvie Di Filippo<sup>2</sup>

<sup>1</sup>Department of Congenital Heart Disease, Claude Bernard University, Lyon, France; <sup>2</sup>Congenital Cardiology, Cardiothoracic Center of Monaco

**Background and Aim:** This study aimed to assess the functional status and long-term outcomes of patients with Eisenmenger Syndrome reaching adulthood.

**Method:** This is a single-centre retrospective review of all patients diagnosed with Eisenmenger Syndrome. Demographics, clinical data, underlying cardiac disease, functional status, therapeutics and outcomes were collected.

**Results:** 159 patient were included (94 females: 59%), aged 27.7 ± 14.8years at end-follow up, and 60 with Down syndrome (38%). Underlying cardiac disease was: AVSD in 30%, VSD in 35%, ASD in 9%, PDA in 5%, associated shunts in 5%, complex CHD in 10%, left heart obstruction in 2.5%, pulmonary veins anomaly in 2.5% and TGA in 1%. CHD was native in 122 cases (77%), 7 had palliation (4%) and 30 complete repair (19%). Pulse oxygen saturation was 84 ± 12% (range 44 to 98%), lower in non-operated or palliated cases (81%) than in repaired cases (92%, p = 0.002). Patients were in NYHA class I (18%), class II (42%), class III (37%) or IV (3%), not different with previous repair or not. Target therapy agents were given in 35% of the cases (1 agent in 20%, 2 associated in 13%, intravenous epoprostenol in 1.5%). Death occurred in 26 patients (16%) at the age of 29.3 ± 17.8years. Complications occurred in 64% including: hemorrhages events, syncope, thrombo-embolia, cerebral abscess, infective endocarditis, heart failure or arrhythmias. NYHA class did not differ between patients with or without target therapy. Survival rates were: 98% at 10-years, 93% at 20-years, 87% at 30-years, 83% at 40-years, 73% at 50-years and 53% at 60-years of follow-up. Survival was lower in Down patients (p = 0.0023), in males (p = 0.04) and tends to be higher up to 50-years of age in patients under target therapy (p = 0.05).

**Conclusions:** Survival rates of adult patients with Eisenmenger Syndrome seem to improve up to 50-year of age with target therapy agents.

**Keywords:** Congenital heart disease, Eisenmenger syndrome, pulmonary antihypertensive therapy

**P-624**

**The concept of right ventricular-pulmonary arterial coupling explored in chronic pulmonary regurgitation**

Rahi Alipour Symakani<sup>1</sup>, Thierry Bové<sup>2</sup>, Willem A Helbing<sup>3</sup>, Annemarie Prins Verzijl<sup>4</sup>, Yannick J H J Taverne<sup>5</sup>, Daphne Merkus<sup>4</sup>, Beatrijs Bartelds<sup>2</sup>

<sup>1</sup>Department of Pediatrics, Division of Pediatric Cardiology, Erasmus Medical Center, Sophia Children's Hospital, Rotterdam, the Netherlands; <sup>2</sup>Department of Cardiothoracic Surgery, Erasmus Medical Center, Rotterdam, the Netherlands; <sup>3</sup>Department of Cardiology, Division of Experimental Cardiology, Erasmus Medical Center, Rotterdam, the Netherlands; <sup>4</sup>Department of Cardiac Surgery, Ghent University Hospital, Ghent, Belgium; <sup>5</sup>Department of Pediatrics, Division of Pediatric Cardiology, Erasmus Medical Center, Sophia Children's Hospital, Rotterdam, the Netherlands; <sup>6</sup>Department of Cardiology, Division of Experimental Cardiology, Erasmus Medical Center,

Rotterdam, the Netherlands; <sup>5</sup>Department of Cardiothoracic Surgery, Erasmus Medical Center, Rotterdam, the Netherlands

**Background and Aim:** Right ventricular (RV) failure is an important determinant of outcome in congenital heart disease. Chronic pulmonary regurgitation (PR) increases the risk of developing RV failure, e.g. in tetralogy of Fallot. So far, clinical markers fail to predict which patients are at risk and require interventions like pulmonary valve replacement. Right ventricular-pulmonary arterial (RV-PA) coupling has been identified as a marker of early RV dysfunction in pulmonary hypertension. In this study, longitudinal assessment of RV-PA coupling was performed in a model of RV hypertrophy and chronic PR.

**Method:** A porcine model of sequential RV overload was developed. Neonatal pulmonary artery banding induced RV hypertrophy during 1 month. Subsequently, banding was removed and PR was created by a transannular patch (TAP group, n = 4). Control animals (n = 2) underwent sham surgeries. After 2, 3 and 4 months of follow-up, invasive pressure-volume loop measurements were performed to assess RV volumes, load-independent contractility, afterload and end-diastolic stiffness. RV-PA coupling is the ratio of contractility to afterload. Hypertrophy was assessed as the right to left ventricular mass ratio.

**Results:** Contractility, afterload and RV-PA coupling did not change over time in the TAP group and were not significantly different from sham (figure 1). Stroke volume (indexed to body

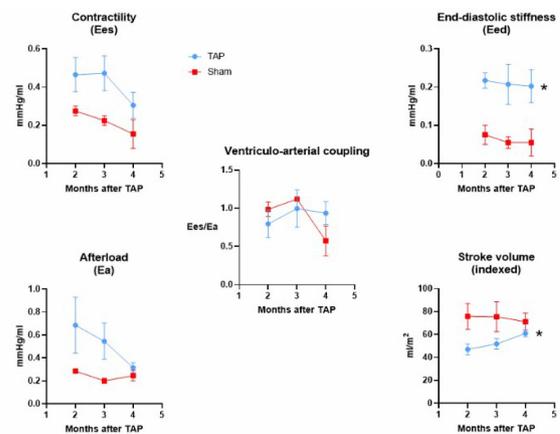


Figure 1: progression of hemodynamic parameters after transannular patch surgery

Data expressed as mean ± standard error of the mean. Ea: effective arterial elastance; Eed: end-diastolic elastance; Ees: end-systolic elastance; TAP: transannular patch. The asterisk marks a significant difference between the transannular patch and sham groups.

Table 1: additional data at each time point after transannular patch surgery

		2 months		3 months		4 months	
		Mean	SEM	Mean	SEM	Mean	SEM
Body mass (kg)	TAP	48	2	69	4	100	2
	Sham	54	3	76	8	113	3
Heart rate (beats/min)	TAP	87	2	85	3	80	3
	Sham	85	10	89	15	77	6
RV ESV <sub>i</sub> (ml/m <sup>2</sup> )	TAP	62.8	13.4	70.3	14.3	78.6	17.2
	Sham	99.7	12.9	97.6	25.0	85.2	1.1
RV EDV <sub>i</sub> (ml/m <sup>2</sup> )	TAP	109.8	17.8	122.2	18.6	139.5	17.1
	Sham	175.5	24.2	173.0	12.0	156.3	6.4
RV/LV mass ratio	TAP					58.5	1.9
	Sham					46.0	3.0

EDV<sub>i</sub>: end-diastolic volume indexed to body surface area; ESV<sub>i</sub>: end-systolic volume indexed to body surface area; LV: left ventricle; RV: right ventricle; SEM: standard error of the mean; TAP: transannular patch group

surface area) was initially lower in the TAP group and tended to normalize at 4 months. End-diastolic stiffness was increased in the TAP group at each time point, consistent with persistent RV hypertrophy (table 1).

**Conclusions:** Preliminary data in chronic PR suggest preservation of RV-PA coupling over time. Sufficient contractility is retained after remodeling to prior pressure overload. Although not significant, both contractility and afterload appear to decrease over time after TAP, leaving the coupling ratio unchanged. Stroke volume was initially lower due to pressure-induced hypertrophy but increases progressively during follow-up. This characterizes the decrease in afterload in chronic PR and might (pseudo)normalize RV-PA coupling. Additionally, pulmonary valve incompetence means that the RV is also directly coupled to the pulmonary artery in diastole. Predictive markers of early RV dysfunction should therefore assess the RV filling phase and diastolic dysfunction, which is supported by persistently increased end-diastolic stiffness.

**Keywords:** congenital heart disease, pulmonary regurgitation, right ventricular dysfunction, myocardial adaptation, contractility, afterload

#### P-625

##### **A thirty-year review of shared care paediatric cardiac transplantation outcomes in republic of ireland**

Helene Murchan<sup>1</sup>, Esmé Dunne<sup>1</sup>, Ross Foley<sup>2</sup>, Adam James<sup>1</sup>, Craig Laurence<sup>3</sup>, Jacob Simmonds<sup>3</sup>, Michael Burch<sup>3</sup>, Zdenka Reinhardt<sup>4</sup>, Matthew Fenton<sup>3</sup>, Colin McMahon<sup>5</sup>

<sup>1</sup>Paediatric Cardiology Department, Children's Health Ireland at Crumlin, Dublin 12, Ireland; <sup>2</sup>Paediatric Cardiology Department, Royal Children's Hospital Belfast, Northern Ireland; <sup>3</sup>Great Ormond Street Hospital for Children NHS Foundation Trust and Cardiothoracic Transplant Unit, London, WC1N 3JH, England, United Kingdom; <sup>4</sup>Department of Cardiac Transplantation, Freeman Hospital, Newcastle upon Tyne, England, United Kingdom; <sup>5</sup>School of Medicine, University College Dublin, Belfield Dublin 4, Ireland

**Background and Aim:** This study reviewed demographics and shared care outcomes of paediatric heart transplantation patients in Ireland over thirty years.

**Method:** We conducted a retrospective review of paediatric patients of Children's Health Ireland at Crumlin who underwent cardiac transplantation between January 1992 and March 2022. Data was obtained from patient charts and correspondence received from the shared care transplant centres (Great Ormond Street Hospital London, Freeman Hospital Newcastle).

**Results:** Forty-six patients were identified who met criteria for inclusion. Thirty (65%) patients were female. Ten patients (22%) had underlying single ventricle physiology. Fourteen (30%) patients required mechanical bridging, inclusive of ECMO or VAD implantation and nineteen (41%) required milrinone prior to transplantation. The mean length of stay on the transplant list was 218 days (range 5 -1063 days). The mean age at time of transplantation was 75 months (range 3-199 months). At the time of transplantation offer thirteen (28%) were at home, sixteen (35%) were inpatients in the local centre and sixteen (35%) were inpatients at the transplant centre.

Forty-seven heart transplants were performed in 46 patients. Two transplants were from DCD donors and the remainder were from DBD donors. Nine (20%) were ABO-incompatible, nine (20%) were size-mismatched and one was HLA-mismatched. Nine (20%) were EBV mismatched and nineteen (41%) were CMV mismatched. Fourteen (29%) patients required treatment for CMV infection.

In terms of post-transplant complications, five (11%) patients developed antibody mediated rejection at a range of 7 days to

13 years post-transplant. Twenty-four (52%) developed cellular rejection at a median time point of ten days (range 3 - 4872 days). Post-transplant lymphoproliferative disorder was identified in three (7%) patients. Sixteen (35%) patients had developed coronary vasculopathy at the time of the study period. Two patients were re-listed and one patient received a second organ. At the time of this review there had been seven deaths (15%) amongst the patient group at a range of 42 - 41, 636 days.

**Conclusions:** Although Ireland has adopted a shared care transplant model with a significant patient population, outcomes in terms of survival, freedom from graft rejection and graft vasculopathy are excellent over a thirty year period.

**Keywords:** paediatric heart transplantation, post-transplant lymphoproliferative disorder, mechanical bridging; outcomes; shared care transplantation

#### P-626

##### **Paradigm shift in management of shared care paediatric heart transplantation patients during the COVID-19 pandemic**

Esmé Dunne<sup>1</sup>, Helene Murchan<sup>1</sup>, Adam James<sup>1</sup>, Zdenka Reinhardt<sup>2</sup>, Craig Laurence<sup>3</sup>, Jacob Simmonds<sup>3</sup>, Matthew Fenton<sup>3</sup>, Michael Burch<sup>3</sup>, Damien Kenny<sup>1</sup>, Colin McMahon<sup>4</sup>

<sup>1</sup>Paediatric Cardiology Department, Children's Health Ireland at Crumlin, Dublin, Ireland; <sup>2</sup>Department of Paediatric Cardiac Transplantation, Freeman Hospital, Newcastle upon Tyne Foundation, NHS England; <sup>3</sup>Great Ormond Street Hospital for Children NHS Foundation Trust and Cardiothoracic Transplant Unit, London, WC1N 3JH, England, United Kingdom; <sup>4</sup>School of Medicine, University College Dublin

**Background and Aim:** The impact of the COVID-19 pandemic on shared care transplant management between Dublin and two transplant centres in the United Kingdom has been unexplored.

**Method:** A retrospective review of changes in management of paediatric heart transplant patients before and during the COVID-19 pandemic to date, defined from 1st March 2020 until 1st November 2022.

**Results:** Thirty paediatric heart transplant patients were actively managed during study period. Six patients underwent transplantation. There was a high level of acuity during this period with 2 new cases of antibody-mediated rejection, 6 episodes of cellular mediated rejection and 4 case of post-transplant lymphoproliferative disorder. Each of these patients was managed in the local centre with transplant centre MDT involvement.

Prior to the COVID-19 pandemic, patients were managed with a shared care model between CHI at Crumlin and the transplant centre. Following the onset of the pandemic, care was largely based in CHI at Crumlin, with regular discussion and review with the managing transplant centre. Furthermore, 3 patients were transitioned to local adult services.

Procedures involved in the annual review and continued transplant monitoring including coronary angiography, intravascular ultrasound and endomyocardial biopsy were undertaken by an interventional cardiologist in Ireland instead of the patient travelling to the transplant centre. In the study period 27 patients underwent IVUS, coronary angiography, diagnostic cardiac catheterisation and endomyocardial biopsy. No patient required travel to the transplant centre for one of these procedures to be performed.

Of note 27 (90%) patients had a documented COVID-19 infection and 17 (57%) had undergone a primary course of COVID-19 vaccination. There were no ICU admissions amongst the group

with COVID-19 infection. Finally COVID-19 resulted in establishment of

a virtual transplant clinic with the MDT involvement from local and transplant teams.

**Conclusions:** The global COVID-19 pandemic caused a paradigm shift in the service provision of shared care with majority of diagnostic, testing and management shifting to the local centre in addition to virtual clinic care.

**Keywords:** COVID-19, shared care, paediatric heart transplantation

## P-627

### Evaluation of left ventricular diastolic flow dynamics in children with pulmonary arterial hypertension using blood speckle tracking

Matthew Henry<sup>1</sup>, Solveig Fadnes<sup>2</sup>, Kristian Sørensen<sup>4</sup>, Pablo Lopez<sup>5</sup>, Lasse Lovstakken<sup>2</sup>, Luc Mertens<sup>1</sup>, Siri Ann Nyrnes<sup>3</sup>

<sup>1</sup>Cardiology, The Hospital for Sick Children, Toronto, Canada;

<sup>2</sup>Department of Circulation and Medical Imaging, Norwegian University of Science and Technology (NTNU), Trondheim, Norway; <sup>3</sup>Children's Clinic, St. Olav's Hospital, Trondheim University Hospital, Norway;

<sup>4</sup>Division of Ålesund Hospital, Department of Pediatrics, Møre and Romsdal Hospital Trust; <sup>5</sup>Hospital Universitario Puerta de Hierro Majadahonda, Majadahonda, Spain

**Background and Aim:** Pulmonary arterial hypertension (PAH) can lead to progressive RV dilation and dysfunction, as well as LV diastolic dysfunction due to an interventricular interaction. Assessment of diastolic function in children is particularly challenging which has led to the search for novel methods. Blood speckle tracking (BST) uses high frame rate imaging and speckle correlation which allows for the assessment and quantification of complex blood flow. Our aim was to compare left ventricular diastolic flow dynamics using BST in children with pulmonary hypertension to those in healthy children.

**Method:** Clinical and routine echocardiographic parameters were prospectively collected for children with PAH and controls with normal hearts. An apical 4 chamber view was acquired using a commercially available clinical scanner with research software enabling the acquisition of high-frame rate imaging and blood speckle tracking. Left ventricular rate of energy loss, vorticity, kinetic energy, and vector complexity were derived using in-house custom made software.

**Results:** Data from 34 children were analyzed (15 PAH, 19 controls) with no significant difference in age, height, weight or heart rate between groups (table 1). There were no qualitative differences in the location of maximum rate of energy loss, vorticity or kinetic energy.

Patients with PAH had significantly reduced average vorticity (6.2 Hz (8.3) vs 13.2 Hz (8.4)  $p = 0.01$ ), kinetic energy (0.05 J/m (0.05) vs 0.08 J/m (0.04);  $p = 0.01$ ) and vector complexity (0.16 (0.2) vs 0.25 (.2)  $p = 0.03$ ) (Figure 1). Average kinetic energy significantly correlated with LV septal  $e'$  ( $r = 0.76$ ,  $p < 0.01$ ) and lateral  $e'$  ( $r = 0.636$ ,  $p < 0.05$ ), as well as septal  $E/e'$  ( $r = -0.673$ ,  $p < 0.05$ )

**Conclusions:** Differences in LV diastolic flow are detectable in patients with pulmonary hypertension using blood speckle tracking which shows promise as a tool in the assessment of diastolic function in children.

**Keywords:** Pulmonary hypertension, blood speckle imaging, diastolic function, high frame rate imaging

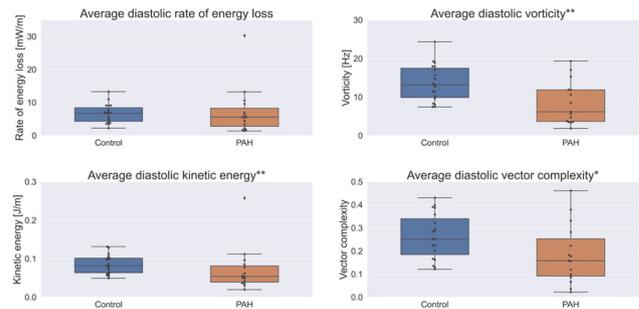
Results

Table 1

	Pulmonary Hypertension	Controls	P value
Age (years)	8.0 (13)	6.75 (7)	0.445
Height (cm)	120 (72)	126 (45)	0.649
Weight (kg)	24.2 (40)	22.8 (26)	0.703
Heart rate (bpm)	83 (42)	82 (47)	0.425
Gender (m:f)	3:12	9:10	0.152

Data shown in as median and interquartile range ( )

Figure 1



LV diastolic parameters between groups. \*  $p < 0.05$ ; \*\*  $p < 0.01$

## P-628

### Mid and long-term efficacy of macitentan in children with advanced pulmonary hypertensive vascular disease - a single centre experience

Sulaima Albinni, Julian Heno, Imre Pavo, Erwin Kitzmueller, Ina Michel Behnke

Medical University of Vienna, Department of Paediatrics and Adolescent Medicine, Paediatric heart centre

**Background and Aim:** Macitentan is a safe and effective treatment option in adults with pulmonary arterial hypertension. Data on its use in paediatric patients is limited. In this prospective single centre study, we investigated mid- and long-term effects of macitentan in children with advanced pulmonary hypertensive vascular disease (PHVD).

**Method:** 24 patients with either IPAH (6), CHD-PAH (10), CTEPH (1) or PAH associated with other conditions (6) were enrolled for treatment. Efficacy was determined by echo (RVSP, RVED, TAPSE, VTI, PAAT), BNP levels and 6MWT at 3 months and 1-year. For detailed analysis, the entire cohort was subgrouped in CHD-PAH and non-CHD-PAH patients respectively.

**Results:** Mean age was 10.7 +/- 7.6 years, median observation period was 36 months. 18/24 pts had systemic or suprasystemic PA pressures. 20/24 pts were on additional sildenafil and /or prostacyclines. 2 pts discontinued because of peripheral edema.

Overall, there was mid and long-term decrease of BNP levels ( $p < 0, 05$ ). On echo, there was improvement of all parameters after 3 months (RVSP, RVED, TAPSE, VTI, PAAT;  $p < 0, 05$ ), after 1 year only changes of VTI (+14%) and PAAT (+11%) remained significant ( $p < 0, 05$ ).

Non-CHD-PAH patients showed significant improvement of TAPSE (+15%), VTI (+13%), PAAT (+36%) and BNP (-57%) at 3 months ( $p < 0, 005$ ) and after 1 year ( $p < 0, 05$ ). Decrease of RVSP (-23%) and RVED (-11%) was significant at 3 months ( $p < 0, 01$ ) but did not reach statistical significance after 1 year. In CHD - PAH patients mid and long-term changes were not significant. 6-MWT slightly increased but was not statistically evaluated. **Conclusions:** Data presented herein comprise the largest cohort of severely affected paediatric patients receiving Macitentan. Overall,

Macitentan was safe and associated with beneficial mid-term effects, whereas after 1 year these improvements persisted only in non-CHD - PAH patients. Our data suggest that efficacy was mainly driven by improvements in patients with PAH not related to CHD, whereas in CHD -PAH effects were limited. Larger studies are needed to verify these preliminary results and to prove efficacy of this drug in different paediatric PHVD entities.

**Keywords:** pulmonary arterial hypertension, infants, congenital heart disease, endothelin receptor antagonist, pediatric

## P-629

### Additional interventional and surgical treatment in pediatric patients with pulmonary arterial hypertension – single center experience

Joanna Młodawska, Paweł Dryżek, Katarzyna Ostrowska, Justyna Topolska Kusiak, Patrycja Gładysz Piestrzyńska, Tomasz Moszura  
Department of Pediatric Cardiology, Polish Mother's Memorial Hospital, Lodz, Poland

**Background and Aim:** Treatment of pediatric patients with pulmonary hypertension is extremely challenging. Drug options are limited, and Polish national healthcare medical regimens are age based which can limit therapeutic options. We present our single center experience in treating patients with different forms of moderate to severe pulmonary arterial hypertension.

**Method:** The aim of this study was to present single center experience and outcome of treatment in patients requiring pulmonary vasodilators with emphasis on additional interventional and surgical treatment complementing pharmacotherapy in cases with moderate to severe PAH and clinical manifestation.

**Results:** Between 2018-2022 16 pts with moderate/severe pulmonary arterial hypertension were treated with targeted pharmacotherapy. Most of these patients underwent additional interventional or surgical treatment. Interventions in the Cath-Lab: 1 pt underwent PDA stenting and in time balloon redilatation of implanted coronary stents; 1pt ASD II device closure; 1pt MAPCA occlusion due to hemoptysis; 1pt PDA device closure. Surgical shunt closure: VSD+ASD+PDA:1pt; ASDII: 2pts. Included in the study were patients with severe PH (RV systolic pressure > 2/3 systolic) in the NICU that required surgical PDA ligation: 6pts. Included is 1 patient with Eisenmenger syndrome (sort-pulmonary window+ severe aortic stenosis) that underwent aortic valvuloplasty due to recurring syncope spells and chest pain which improved quality of life, this patient awaits heart and lung transplant. The study group included 3 patients without additional intervention: 1 patient with small ASD II after congenital diaphragmatic hernia repair presently prequalified for lung transplant, 1 patient with Eisenmenger Syndrome (VSD)- referred from Ukraine. Pharmacological treatment regimens included: Sildenafil monotherapy: (10pts); Sildenafil + Bosentan (3pts); Sildenafil+Bosentan+Treprostynil (3pts) with no severe side-effects. 12/13 patients additionally treated interventional or surgically required escalation of PH-therapy during anesthesia or in postprocedural period with epoprostenol or iNo.

**Conclusions:** PAH treatment with pulmonary vasodilators is well tolerated and each additional procedure whether surgical or interventional led to clinical improvement and better quality of life.

**Keywords:** pulmonary arterial hypertension, interventional therapy, surgical treatment, PDA, ASD

Tabel 1 Patient population, study group

Study group characteristics

No pt	Drug	Congenital defect	Interventional treatment	Additional Information
1	II	(-)	Implantation of 2 coronary stents into residual PDA	Microdeletion 17q22a23.2
1	III	Aortopulmonary window severe aortic stenosis	BAV (TYSHAK angioplasty balloon)	Eisenmenger syndrome
1	III	VSD+ASD+PDA after congenital correction	MAPCA occlusion using coil after hemoptysis	Growth hormone treatment short stature
1	I	ASD II	Occlutech ASD Occluder	
1	II	ASD II	Surgical closure	(-)
1	I	ASD II	Surgical Closure	Noonan Syndrome
1	I	PDA	ADO II	(-)
6	I	PDA	Surgical ligation	iNO postprocedure in the NICU
1	III	A residual ASD II	(-)	Neonatal congenital diaphragmatic hernia
1	III	VSD	(-)	(-)
1	I	(-)	(-)	Sick Sinus Syndrome

Pharmacotherapy: I: Sildenafil; II: Sildenafil+Bosentan; III: Sildenafil+Bosentan+Treprostynil

## P-630

### The unintended consequence: obesity in children following a diagnosis of arrhythmogenic right ventricular cardiomyopathy

Alison Howell, Shufan Wang, Barbara Cifra, Dawn Nicolson, Sunghoon Minn, Robert Hamilton, Emilie Jean St Michel, Amir Jeewa, Luc Merterns, Vitor Guerra  
Department of Cardiology, The Hospital For Sick Children, Toronto, Ontario, Canada

**Background and Aim:** Current guidelines restrict patients with a diagnosis of arrhythmogenic right ventricular cardiomyopathy (ARVC) from competitive sport and high impact athletic activity. However, exercise plays an important part in preventing obesity, which is associated with an increased risk of cardiovascular (CV) events. We sought to evaluate the prevalence of obesity in children diagnosed with ARVC.

**Method:** We retrospectively reviewed patients 0-18 years of age with a definite or possible borderline ARVC diagnosis at our institution between 2002-2019. Patients who were diagnosed as obese (defined as body mass index > 95%) were identified and their outcomes compared to ARVC patients without obesity.

**Results:** We identified 184 patients with ARVC (51 definite, 133 possible/borderline). 56% (n = 102) were males. Median age was 12.9 years (IQR 9.0-15.4 years). The median time from diagnosis to last follow-up was 2.7 years (IQR 1.1-6.2 years). Prior to diagnosis, the prevalence of obesity was 2% (3/184). At last follow-up this had increased to 15% (27/184 p < 0.0001). There was no significant difference in obesity between diagnosis types (11.1% (6/54) definite vs 15.8% (21/133) possible/borderline p = 0.49). Obesity was associated with a diagnosis at an earlier age (9.9 years (IQR 6.3-14.4) vs 13.2 years (IQR 9.7 - 15.6, p = 0.006). There was a trend to higher resting heart rates (HR) in the obese group (91 (IQR 78-99) vs 82 (IQR 73-93) p = 0.09) but no difference in systolic or diastolic blood pressure (p = 0.34 and p = 0.84 respectively). There was a trend towards fewer obese patients having tachyarrhythmia (11% vs 28.6% p = 0.06) but no difference in ICD implantation (22% vs 23% p = 0.95). There was no statistical difference between the groups in regard to listing for heart transplant and no deaths.

**Conclusions:** The prevalence of obesity in childhood significantly increases following ARVC diagnosis and is associated with diagnosis at younger age. Obesity is a modifiable risk factor for future CV related events. Future prospective studies should evaluate how to properly promote physical activity at the time of diagnosis and the impact of personalized exercise program for these patients.

**Keywords:** arrhythmogenic right ventricular cardiomyopathy, obesity, cardiomyopathy

## P-631

**Assessment of orthostatic intolerance in adolescents with myalgic encephalomyelitis/ chronic fatigue syndrome (ME/CFS) and healthy controls**Ariane Leone<sup>1</sup>, Renate Oberhoffer<sup>2</sup>, Uta Behrends<sup>1</sup>, Andrea Maier<sup>3</sup><sup>1</sup>Department of Pediatrics, Technical University Munich, Munich, Germany; <sup>2</sup>Department of Preventive Pediatrics, Technical University Munich, Munich, Germany; <sup>3</sup>Department of Neurology, University Hospital RWTH Aachen, Aachen, Germany

**Background and Aim:** In childhood and adolescence, Epstein-Barr-Virus is held responsible for 50% of postinfectious myalgic encephalomyelitis/ chronic fatigue syndrome (ME/CFS) cases. Currently, ME/CFS after SARS-CoV-2-infection is increasingly reported. Symptoms include orthostatic intolerance (OI) such as postural tachycardia syndrome (PoTS). PoTS in ME/CFS is decisive for everyday functioning and treatment, yet data in adolescents are rare and inconsistent. This study aimed to better characterize OI in adolescent ME/CFS and standardize diagnostics.

**Method:** We performed a pilot-case-control-study on adolescents with ME/CFS according to CCC 2003, IOM 2015, Rowe 2017 or Jason 2006 compared to healthy controls (HC). We developed a standardized interview to assess the medical history of OI (HOI). NASA Lean test (leaning 10-min-standing-test) with registration of heart rate (HR) and blood pressure every minute was performed in the morning on empty stomach without morning medication. Data were evaluated according to current PoTS-criteria by 2019 National Institutes of Health Expert Consensus Meeting and ICD-11 (8D89.2) and analyzed by Fisher's exact test and independent t-test.

**Results:** 15 ME/CFS-patients (15.2±0.8 years) and 17 HC (15.2±1.1 years, p>0.05) were included. HR supine and standing differed significantly between ME/CFS (supine 79.5±4.6, standing 112.2±7.6 beats/min) and HC (supine 66.6±4.3, standing 101.9±0.5 beats/min, p<0.05). HOI was reported in 17 subjects (13 ME/CFS, 4 HC). Of those, definite PoTS was diagnosed in five (5 ME/CFS, 0 HC). In seven participants (5 ME/CFS, 2 HC) HOI was positive, but without tachycardia during standing. Of those, two ME/CFS had symptoms during testing. Absolute HR greater than 120 beats/min during standing discriminated better between ME/CFS and HC than a HR-increase of 40 beats/min from supine to standing.

**Conclusions:** These preliminary results suggest that adolescent ME/CFS-patients had higher HR supine and upright, and higher prevalence of OI than HC. Significantly more patients than HC had a diagnosis of PoTS. Data suggest that a HR greater than 120 beats/min as criterion for PoTS can reduce false positive results compared to the 40 beats/min-limit. In clinical practice we suggest performing a standardized interview in combination with the NASA Lean test to diagnose PoTS in ME/CFS patients. Further analysis with more participants will show if these results can be generalized.

**Keywords:** ME/CFS, chronic fatigue syndrome, POTS, postural tachycardia syndrome, orthostatic intolerance, post-COVID

## P-633

**Prevalence, determinants and clinical characteristics of "super fontan"**Hideo Ohuchi<sup>1</sup>, Aki Mori, Aki Mori, <sup>1</sup>, Kenichi Kurosaki<sup>1</sup>, Michikazu Nakai<sup>2</sup><sup>1</sup>Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Suita, Japan; <sup>2</sup>Department of Medical and Health Information Management, National Cerebral and Cardiovascular Center, Suita, Japan

**Background and Aim:** "Super Fontan" (SF) is an excellent phenotype of patients with Fontan circulation (FC) who have normal exercise capacity [ $\geq 80\%$  predicted peak oxygen uptake (PVO<sub>2</sub>)]. This study was to clarify the prevalence, predictors, and clinical characteristics of SF.

**Method:** We reviewed 404 FC patients (21 ± 10 years old) who had undergone cardiopulmonary exercise testing (CPX) between 2005 and 2021 and the latest results were compared with clinical profiles.

**Results:** Of these, 77 (19%) FC patients were SF. Prevalence of SF at 5, 10, 15, 20, and  $\geq 25$  years after the operation were 16 (35%), 30 (39%), 18 (19%), 13 (14%), and 0 (0%), respectively. Compared to non-SF, SF patients were younger (14 ± 5 vs. 23 ± 10, p < 0.001) and 69% were male (p < 0.05). Although there were no differences in ventricular function or hemodynamics, except for high arterial blood pressure and oxygen saturation (SaO<sub>2</sub>), SF was characterized by favorable body composition (less fatty mass and greater muscle mass), superior pulmonary function, preserved hepatorenal and hemostatic functions as well as better glucose tolerance (p < 0.05 – 0.001) at the time of CPX. Interestingly, better ventricular function, low pulmonary artery resistance, and high SaO<sub>2</sub> before Fontan operation were associated with current SF (p < 0.05 – 0.01). Furthermore, past positive trajectory of PVO<sub>2</sub> during childhood (n = 134) was associated with their current adult SF (n = 22, p < 0.05). During mean follow-up of 3.7 years after the latest CPX, 25 patients died and 74 patients were unexpectedly hospitalized. There were no death in the SF and the rate of hospitalization was 67% lower in the SF than that in the non-SF (p < 0.01 – 0.001).

**Conclusions:** Prevalence of SF was 19% and decreased over time, especially  $\geq 15$  years after Fontan operation. SF characterized by better multi-organ function and excellent prognosis, rather than hemodynamics, may have been shaped by better pre-operative hemodynamics and long-term post-operative favorable stress to multi-organs. Life-long management strategy is required to prevent the late rapid decline of SF prevalence.

**Keywords:** Fontan, Super-Fontan, exercise capacity, multi-organ, mortality

## P-634

**Impaired aerobic capacity in adolescents and young adults after treatment for cancer or non-malignant haematological disease**Pascal Amedro<sup>1</sup>, Vincent Dubard<sup>3</sup>, Sophie Guillaumont<sup>2</sup>, Gregoire De La Villeon<sup>2</sup>, Nicolas Sirvent<sup>5</sup>, Anne Sirvent<sup>5</sup>, Anne Requirand<sup>1</sup>, Helena Huguet<sup>4</sup>, Marie Christine Picot<sup>4</sup>, Stefan Matecki<sup>3</sup>, Arthur Gavotto<sup>3</sup><sup>1</sup>Pediatric and Congenital Cardiology Department, M3C National Reference CHD Centre, Bordeaux University Hospital, IHU Liryc, INSERM 1045, University of Bordeaux, Bordeaux, France; <sup>2</sup>Pediatric Cardiology and Rehabilitation Unit, St-Pierre Institute, Palavas-Les-Flots, France.; <sup>3</sup>Pediatric and Congenital Cardiology Department, M3C Regional Reference CHD Centre, Montpellier University Hospital, Montpellier, France; <sup>4</sup>Epidemiology and Clinical Research Department, University Hospital, Montpellier, France.; <sup>5</sup>Pediatric Oncology Department, Montpellier University Hospital, Montpellier, France

**Background and Aim:** Childhood cancer survivors are at increased risk for cardiovascular disease. Maximal oxygen uptake (VO<sub>2</sub>max) is a major determinant of cardiovascular morbidity. The aim of this study was to compare aerobic capacity, measured by cardiopulmonary exercise test (CPET), of adolescents and young adults in remission with that of healthy controls and to identify the predictors of aerobic capacity in this population.

**Method:** In this cross-sectional multicentric study, a total of 477 subjects aged from 6 to 25 years, were included and underwent a complete cycle-ergometer CPET (Figure). Two groups were identified: adolescents and young adults in complete remission after treatment for cancer or transplanted for non-malignant haematological disease during young age (i.e., the “remission group”), and the controls subjects. Subjects from both groups were enrolled during the same period. A multiple linear regression was used to identify the explanatory factors for maximum oxygen uptake (VO<sub>2</sub>max) and ventilatory anaerobic threshold (VAT) in the remission group. The study was approved by our institutional review board and registered on ClinicalTrials.gov (NCT04815447).

**Results:** In the remission group, the mean VO<sub>2</sub>max was significantly lower than in controls (37.3±7.6 vs. 43.3±13.1 mL/Kg/min, P<0.01, respectively), without any clinical or echocardiographic evidence of heart failure. The VAT was significantly lower in the remission group (26.9±6.0 mL/Kg/min vs. 31.0±9.9 mL/Kg/min, P<0.01, respectively).

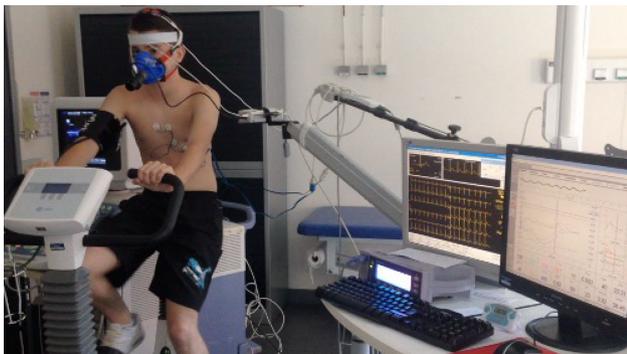
In both univariate and multivariate analyses, a lower VO<sub>2</sub>max in the remission group was associated with female sex, higher BMI, and previous treatment with radiotherapy or hematopoietic stem cells transplantation. The final multivariate model explained 47% of the VO<sub>2</sub>max variability in the remission group.

In both univariate and the multivariate analyses, a lower VAT in the remission group was associated with female sex, older age, higher BMI, and previous treatment with hematopoietic stem cells transplantation. The final multivariate model explained 43% of the VAT variability in the remission group.

**Conclusions:** Impaired aerobic capacity had higher prevalence in adolescents and young adults in cancer remission. This impairment was primarily related to physical deconditioning and not to heart failure.

**Keywords:** childhood cancer survivor, cardiopulmonary fitness, onco-cardiology, physical deconditioning, paediatric heart failure, anthracycline

#### Paediatric CPET



Cardiopulmonary exercise test (CPET)

#### P-635

##### Reference values of aerobic fitness in the contemporary paediatric population

Arthur Gavotto<sup>1</sup>, Jonathan Rhodes<sup>2</sup>, Suellen Moli Yin<sup>2</sup>, Sophie Guillaumont<sup>3</sup>, Thibault Mura<sup>4</sup>, Alfred Hager<sup>5</sup>, Julia Hock<sup>5</sup>, Stephan Matecki<sup>1</sup>, Pascal Amedro<sup>6</sup>

<sup>1</sup>Pediatric and Congenital Cardiology Department, M3C Regional Reference CHD Centre, Montpellier University Hospital, Montpellier, France; <sup>2</sup>Department of Cardiology, Boston Children's Hospital, Boston, USA; <sup>3</sup>Paediatric Cardiology and Rehabilitation Unit, St-Pierre Institute, Palavas-Les-Flots, France; <sup>4</sup>Epidemiology and Clinical Research Department, Nimes University Hospital, Nimes, France; <sup>5</sup>Clinic for Paediatric Cardiology and Congenital Heart Diseases, German Heart Centre, Munich, Germany; <sup>6</sup>Pediatric and Congenital Cardiology Department, M3C National Reference CHD Centre, Bordeaux University Hospital, IHU Liryc, INSERM 1045, University of Bordeaux, Bordeaux, France

**Background and Aim:** Aerobic physical fitness (VO<sub>2</sub>max assessed by cardiopulmonary exercise testing (CPET) has shown a growing interest in the follow-up of paediatric chronic diseases. The dissemination of CPET in paediatrics requires valid paediatric VO<sub>2</sub>max reference values to define the upper and lower limits of the norm. This study aimed to establish VO<sub>2</sub>max reference Z-scores values, from a large cohort of children representative of the contemporary paediatric population, including extreme weights.

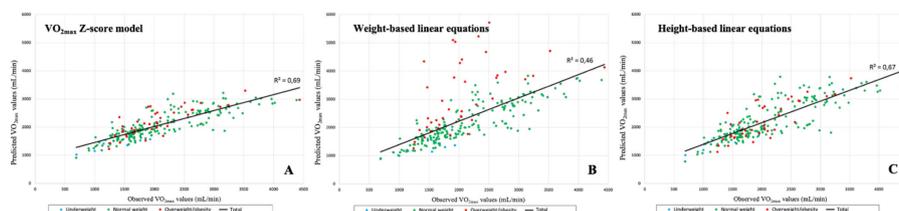
**Method:** In this cross-sectional study, 909 children aged 5 to 18 years old from the general French population (internal cohort), and 232 children from the general German and US populations (external cohort) underwent a CPET, following the guidelines on high-quality CPET assessment. Linear, quadratic, and polynomial mathematical regression equations were applied to identify the best VO<sub>2</sub>max Z-score model. Predicted and observed VO<sub>2</sub>max values using the VO<sub>2</sub>max Z-score model, and the existing linear equations were compared, in both internal and external cohorts.

**Results:** For both genders, the mathematical model using natural logarithms of VO<sub>2</sub>max, height, and BMI was the best fit for the data (Figure). This Z-score model could be applied to normal and extreme weights and was more reliable than the existing linear equations, in both internal and external validity analyses. The following open-source application can be used to calculate the VO<sub>2</sub>max Z-score: <https://play.google.com/store/apps/details?id=com.d2l.zscore>.

**Conclusions:** This study established reference Z-score values for paediatric cycleergometer VO<sub>2</sub>max using a logarithmic function of VO<sub>2</sub>max, height and BMI, applicable to normal and extreme weights. Providing Z-scores to assess aerobic fitness in the paediatric population should be useful in the follow-up of children with chronic diseases.

**Keywords:** cardiopulmonary exercise test, VO<sub>2</sub>, paediatrics, Z-score, aerobic fitness

Correlation between observed and predicted VO<sub>2</sub>max values using the Z-score model and Cooper's weight-based and height-based linear equations (external cohort).



Legend: the “underweight” group was represented by blue points, the “normal weight” group by green points, and “overweight/obesity” group by red points. The figure panels A, B and C represented the correlation between predicted and measured VO<sub>2</sub>max values (in mL/min) using the Z-score model and Cooper's weight-based and height-based linear equations, respectively.

**P-636**

**Comparison of the results of CPET in the fontan population versus the population without congenital heart disease**

Roman Gestoso<sup>1</sup>, Esther Aurensanz<sup>2</sup>, Emma Grau Sió<sup>2</sup>, Joan Sanchez De Toledo<sup>3</sup>

<sup>1</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain; <sup>2</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain; Institut de Recerca Sant Joan de Déu, Barcelona, Spain; <sup>3</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain; Institut de Recerca Sant Joan de Déu, Barcelona, Spain; Department of Critical Care Medicine, University of Pittsburgh, Pittsburgh, Pennsylvania, United States

**Background and Aim:** Patients with congenital heart disease have some impairment of functional capacity, compared to the population without congenital heart disease. Patients with Fontan surgery are a unique group, which requires further study to understand the functionality of univentricular physiology during exercise by CPET.

**Method:** Prospective study in 7-18 years old patients. Comparison CPET between patients with Fontan surgery (n = 24) and without congenital heart disease (n = 20).

**Results:** In both groups, the RER max was >1.05, considered the maximum test, with no statistically significant differences. None of the patients had a diagnosis of respiratory problems.

In Fontan patients, the VO2max% reached was 54.25%; the relationship between VO2max% and VO2 AT% reached 99.07% of VO2max% in the anaerobic threshold (VO2 AT%); the respiratory reserve was maintained at normal values; HR max% was 77.5% (±7.46) and VE/VCO2 Slope was 36.03 (±6.99).

Significance difference between both groups was found at VO2max% (p<0.001), VE/VO2 and VE/VCO2 (p<0.001) and HR max% (p = 0.010). 21 of the 24 Fontan patients (87.50%) presented VE/VCO2 Slope 30 (ventilatory insufficiency), against 9 of the 20 patients (40%) in the control group.

**Conclusions:** In the CPET, we observed that Fontan patients reached VO2max% close to the VO2 AT%. Considering that all had a normal respiratory reserve, the result could be interpreted as a sign of ventilatory insufficiency. PulseO2 max% as equivalent to stroke volume, showed that PulseO2 decreased when the stroke volume had declined in Fontan patients who were incapable of passing AT. VE/VCO2 Slope analysis also demonstrated signs of ventilatory insufficiency. Low HRmax% values suggest that some Fontan patients have chronotropic insufficiency.

**Keywords:** CPET, Fontan, VE/VCO2-Slope, VO2peak

**P-637**

**Impact of the COVID-19 pandemic on the mental health and physical activity of children and adolescents in germany**

Christina Sitzberger<sup>1</sup>, Christina Holzzapfel<sup>2</sup>, David Ebert<sup>3</sup>, Renate Oberhoffer Fritz<sup>4</sup>

<sup>1</sup>Institute of Preventive Pediatrics, Faculty of Sport and Health Sciences, Technical University of Munich; <sup>2</sup>Clinical Nutritional Medicine, TUM School of Life Sciences, Technical University Munich; <sup>3</sup>Professorship of Psychology & Digital Mental Health Care, Faculty of Sports and Health Sciences, Technical University Munich; <sup>4</sup>Institute of Preventive Pediatrics, Faculty of Sport and Health Sciences, Technical University of Munich; German Heart Centre, Department of Paediatric Cardiology and Congenital Heart Defects, Munich, Germany

**Background and Aim:** The Covid-19-Pandemic in 2020 led to many changes in children's and adolescents' life. Rising infection as well as mortality rates forced governments to declare massive restrictions to counteract the pandemic state. Children and adolescents in particular are affected by measures such as school closures and regulations on social distancing. The aim of this study is to investigate the associated and so far insufficiently clarified effects on the mental health and physical activity of children and adolescents.

**Method:** Data collection took place at the end of 2021 from October to December through a prospective multi-centric survey. A questionnaire was designed for this purpose, which was completed in written form by the participating children and adolescents independently. The study population consisted of n = 188 students of different school types in Germany from the 5th grade onwards.

**Results:** For more than one third (36.5 %) of the subjects, mental health deteriorated as a result of the pandemic and the restrictions imposed in connection with it. Worries about the illness of one's own social contacts (59.0 %), bans on activities (33.0 %) and worries about school or work (33.0 %) were perceived as very stressful. The percentage of children and adolescents meeting WHO activity recommendations decreases from 29.8% before the pandemic to 24.5% during the pandemic. Analogously, the percentage of non-compliance increases from 68.6% before to 73.4% during the pandemic. In each case, 23.9% reported being more or less physically active during the week than before the pandemic, while 41.5% recorded no change.

**Conclusions:** The results of this work show that the effects of the COVID-19 pandemic can have a negative impact on the mental well-being and physical activity of children and adolescents. Even though this negative development only affects a minority, it is advisable to implement health promotion strategies as well as preventive approaches to maintain and improve mental and physical health during and after the COVID-19 pandemic. This is the only

**RESULTS**

CPET RESULTS Fontan vs Control without heart disease

CPET	FONTAN (n= 24)	Without heart disease (n= 20)	p
	Media SD	Media SD	
VO2 max	25,79 ±4,70	31,84 ±8,92	0,006
VO2 max%	54,25 ±11,02	72,25 ±17,41	<0,001
VO2 AT	13,63 ±1,86	16,77 ±5,25	0,009
VO2 AT%	53,75 ±8,43	52,77 ±7,55	0,625
RER max	1,08 ±0,130	1,05 ±0,08	0,468
RER max%	88,83 ±10,40	86,95 ±6,71	0,493
HR AT	112,33 ±24,97	123,40 ±9,24	0,068
HR max%	77,50 ±7,46	84,70 ±7,98	0,010
Pulse O2max	7,02 ±2,62	9,44 ±4,66	0,036
Pulse O2%	71,33 ±15,78	91,10 ±29,82	0,007
BR	45,63 ±12,39	38,30 ±22,15	0,170
VE/VO2 max	38,30 ±5,43	30,05 ±4,83	<0,001
VE/VCO2 max	35,27 ±5,28	28,52 ±3,91	<0,001
Slope VE/VCO2	36,03 ±6,99	28,37 ±5,38	<0,001

Comparative analysis VE/VCO2 Slope

Ventilatory class	VE/VCO2 Slope	FONTAN (n= 24)	Without heart disease (n= 20)
1	<30	3	11
2	30-35,9	11	9
3	36-44,9	5	
4	>45	5	

way to protect the young generation from the negative effects and to support them in their mental recovery and increase their physical activity.

**Keywords:** covid 19 pandemic, mental health, physical activity, children and adolescents

### P-638

#### Digital health nudging in pediatric patients with congenital heart disease: A randomized controlled trial

Laura Willinger<sup>1</sup>, Renate Oberhoffer-Fritz<sup>2</sup>, Peter Ewert<sup>1</sup>, Jan Müller<sup>2</sup>

<sup>1</sup>Department of Congenital Heart Disease and Pediatric Cardiology, Deutsches Herzzentrum München, Technische Universität München;

<sup>2</sup>Institute of Preventive Pediatrics, Technische Universität München

**Background and Aim:** A previous study from our institution showed that particularly adolescents with more severe congenital heart disease (CHD) are rather inactive. Digital health nudging is a modern e-health intervention and was shown to increase physical activity (PA) in healthy adolescents. This randomized-controlled trial examines if digital health nudging increases PA, activity-related self-efficacy (ArSE) and health-related quality of life (HrQoL) in adolescents with CHD.

**Method:** From May 2021 to April 2022, 97 patients (15.1 ± 2.0 years, 50% girls) with moderate or severe CHD were randomly allocated 1:1 to an intervention group (IG) or control group (CG). Daily PA was objectively assessed in minutes of moderate-to-vigorous PA (MVPA) by the wrist worn wearable "Garmin Vivofit jr. ®" over the entire study period. ArSE and HrQoL were assessed with questionnaires at baseline and after the intervention. The IG received daily smartphone messages based on Bandura's social cognitive theory on over a period of three months. Intention-to-treat analysis was performed with multiple imputed data. The first two weeks were compared to the last two weeks of the intervention with Mann-Whitney-U test. Results are expressed as median [25 quartile; 75 quartile].

**Results:** MVPA decreased from 78.1 [62.5; 86.5] to 71.4 [56.6; 75.2] min/day in the IG and from 80.2 [64.7; 102.9] to 76.0 [68.0; 94.0] min/day in the CG over the study period. There was no significant difference in MVPA when comparing the IG with the CG group (IG: Δ -6.4 [-18.6; 3.0] min/day vs. CG: -2.5 [-12.8; 8.5] min/day, p = 0.199). Physical well-being significantly increased over the study period (p = 0.043), but not the total HrQoL score (p = 0.518). ArSE (IG: Δ-0.1 [-0.38; 0.13] vs. CG: Δ-0.05[-0.36; 0.25], p = 0.544) did not change during the digital health nudging intervention compared to the CG.

**Conclusions:** Three months of digital health nudging did not increase PA, but improved feelings of physical well-being in adolescents with CHD. Our results might indicate that although wearables and digital health nudging initially have a positive influence on PA, activity declines over time in both groups and digital health nudging apparently does not have the potential to stop or slow down this phenomenon.

**Keywords:** digital health, physical activity, pediatric, congenital heart disease

### P-639

#### Exercise echocardiography combined with cardiopulmonary exercise testing in small children

Isabelle Schöffl, Weigelt Annika, Rottemann Kathrin, Dittrich Sven, Wällisch Wolfgang

Department of Pediatric Cardiology, Friedrich-Alexander-Universität Erlangen-Nürnberg

**Background and Aim:** Exercise echocardiography in combination with cardiopulmonary exercise testing (CPET) allows for evaluation of cardiac function evaluated using echocardiography during physical exertion while at the same time measuring cardiopulmonary function. As heart rate can only be increased to maximal levels during true physical exercise this is the only method to evaluate cardiac function during maximal exercise while at the same time allowing for objectifying the extent of physical exertion. So far, exercise echocardiography is mainly limited to adults tall enough to fit on a tilt-recline ergometer.

**Method:** We evaluated 6 children (3 girls and 3 boys) after arterial switch operation or Kawasaki Disease, below the required height for tilt-recline ergometers, for exercise echocardiography in combination with CPET on a treadmill. We used an adapted Bruce treadmill protocol. All Echocardiography assessments were undertaken with the child standing upright, bent slightly forward. The evaluations were undertaken before CPET, after the 4th, and the 8th step, and directly as well as after 2 and 3 minutes after ending the exercise. The treadmill was stopped for echocardiography for 30 seconds.

**Results:** All children were able to perform the exercise tests up to maximal exertion. The CPET results are presented in table 1. Exercise echocardiography allowed for good image quality even for evaluating global longitudinal strain. Five children presented with normal cardiac function even at peak exercise. One child showed reduced cardiac function which worsened over the course of the CPET. A catheter investigation revealed stenosis of the right coronary artery.

**Conclusions:** Exercise echocardiography in combination with cardiopulmonary exercise testing in children too small for being able to perform on a tilt-recline ergometer is feasible and safe. Image quality during upright echocardiography is very high and allows for the estimation of global longitudinal strain. This is a first study showing the possible inclusion of exercise echocardiography for evaluating coronary insufficiency in children during exertion.

**Keywords:** Exercise physiology, treadmill protocol, functional echocardiography

Parameter	Mean ± standard deviation
$\dot{V}O_2$ peak (ml/kg/min)	48.0 ± 4.9
Peak RER	1.3 ± 0.1
Peak heart rate (beats/minute)	195 ± 8.6
Exercise time (min)	15.3 ± 1.8
Peak O <sub>2</sub> pulse (ml/min)	6.7 ± 1.0

Mean and standard deviation of exercise variables recorded during exercise echocardiography on the treadmill

### P-640

#### Exercise capacity, endothelial function, muscle mass and strength in pediatric fontan patients

Tania Davila<sup>1</sup>, Daniel Bernstein<sup>1</sup>, Mary Leonard<sup>3</sup>, Jin Long<sup>3</sup>, John P Cooke<sup>2</sup>, Inger N Olson<sup>1</sup>, Tulsı Damase<sup>2</sup>, Mavis Lui<sup>1</sup>, Jonathan Myers<sup>4</sup>, Latha Palaniappan<sup>5</sup>, Seda Tierney<sup>1</sup>

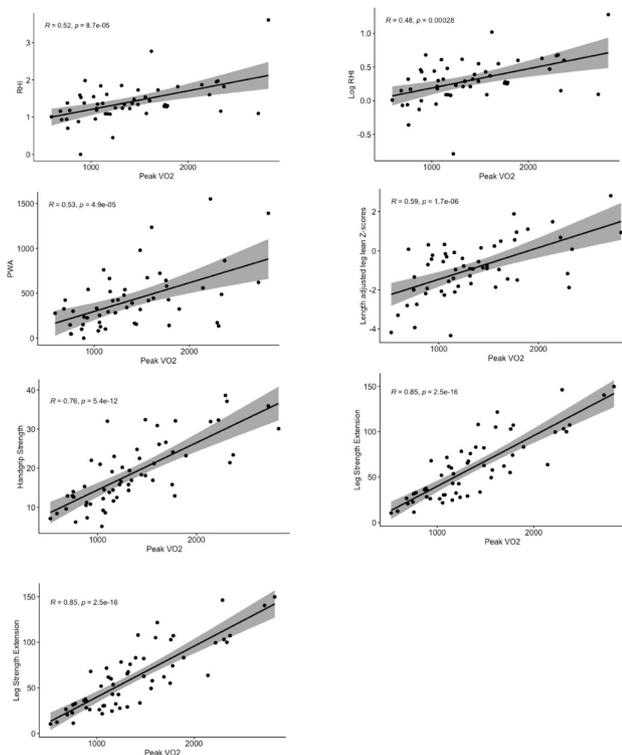
<sup>1</sup>Department of Pediatrics, Division of Cardiology, Lucile Packard Children's Hospital, Stanford University, School of Medicine, Palo Alto, USA; <sup>2</sup>Department of Cardiovascular Sciences, Research Institute, Methodist Hospital, Texas, USA; <sup>3</sup>Department of Pediatrics, Division of Nephrology, Lucile Packard Children's Hospital, Stanford University, School of Medicine, Palo Alto, USA; <sup>4</sup>Palo Alto VA Health Care System, Research Institute, Palo Alto, USA; <sup>5</sup>Department of Medicine, Stanford Hospital, Stanford University, School of Medicine, Palo Alto, USA

**Background and Aim:** By age 40, 50% of Fontan patients will have died or undergone heart transplantation. Poor exercise capacity, low muscle mass and strength, and endothelial dysfunction impact disease progression. Here, we present the baseline data of pediatric Fontan patients prior to entering an exercise intervention (RE-ENERGIZE FONTAN) and assess the relationship between exercise capacity, endothelial function, muscle mass, and strength.

**Method:** This randomized controlled trial in Fontan patients utilizes live-video conferencing to deliver a supervised exercise intervention. Inclusion criteria include: (1) age 9–19 years; (2) exercise clearance by primary cardiologist; (3) English-speaking patient. Exclusion criteria include: (1) NYHA Class IV, (2) Acute illness within the past three months; (3) Active protein losing enteropathy (albumin <2.5 mg/dL); (4) Pacemaker; and/or (5) Cognitive delay impeding participation in the intervention. At baseline, patients underwent a cardiopulmonary exercise test, endothelial pulse amplitude testing, blood work, a DXA scan, and isokinetic dynamometry to measure VO<sub>2</sub> peak, reactive hyperemia index (RHI), baseline pulse wave amplitude (PWA), nitrogen oxide (NO<sub>x</sub>), leg lean mass, and handgrip and leg strength. Leg lean mass was expressed as sex and age-specific z-score adjusted for leg length.

**Results:** 57 Fontan patients with a median age of 13.17 years [IQR, 10.78, 15.58] completed the baseline visit. The median time from Fontan operation was 9.2 years [IQR, 7.2, 12.0]. 34 (60%) had a single right ventricle. VO<sub>2</sub> peak was 1367±520 ml/min. RHI and lnRHI were 1.40±0.53 and 0.30±0.32, respectively. PWA was 423±330 (au). NO<sub>x</sub> level was 15.9±5.9 umol/L. Leg lean mass z-score was -0.89±1.45. Lean handgrip strength was 18.8±8.6 kg, while leg strength extension and flexion were 60.6±35.8 and 27.91±16.3 ft bls, respectively. VO<sub>2</sub> peak was significantly and positively associated with RHI, lnRHI, PWA, lean leg muscle mass z-score, and muscle strength (Figure 1).

**Figure 1.** VO<sub>2</sub> peak and its relation to endothelial function, arterial tone, and muscle mass and strength



**Figure 1**

**Conclusions:** In pediatric Fontan patients, exercise capacity correlates positively with endothelial function, arterial tone, muscle mass, and muscle strength. Our exercise intervention is designed to demonstrate the impact of aerobic activity and strength training on these parameters with the ultimate goal of instituting an “exercise prescription” in Fontan patients to decrease long-term morbidity and mortality.

**Keywords:** Fontan, children, exercise, endothelial function, muscle mass, muscle strength

## P-641

### Myocarditis and sports in the young: data from a nationwide registry on myocarditis – mykke-sport

Isabelle Schöffel<sup>1</sup>, Sven Dittrich<sup>1</sup>, Thomas Pickhardt<sup>2</sup>, Franziska Seidel<sup>3</sup>, Bernd Oppen Rhein<sup>4</sup>, Martin Boehme<sup>5</sup>, Bardo Wannemacher<sup>6</sup>, Katja Reineker<sup>7</sup>, Gesa Wiegand<sup>8</sup>, Tobias Hecht<sup>9</sup>, Axel Rentzsch<sup>10</sup>, Michael Kaestner<sup>11</sup>, Annika Weigelt<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology, Friedrich-Alexander-Universität Erlangen-Nürnberg, Germany; <sup>2</sup>Kompetenznetz Angeborene Herzfehler, Berlin, Germany; <sup>3</sup>German Heart Center Berlin, Department of Congenital Heart Disease and Pediatric Cardiology, Berlin, Germany; <sup>4</sup>Institute for Imaging Science and Computational Modelling in Cardiovascular Medicine; Charité-Universitätsmedizin Berlin, Germany; <sup>5</sup>Department of Paediatric Cardiology and Intensive Care Medicine, Hannover Medical School, Hannover, Germany; <sup>6</sup>Clinic for Paediatric Cardiology, Heart Centre, University of Leipzig, Leipzig, Germany; <sup>7</sup>Department for Paediatric Cardiology, University Heart Center Freiburg, Freiburg, Germany; <sup>8</sup>Department for Paediatric Cardiology, University Hospital Tübingen, Tübingen, Germany; <sup>9</sup>Center for Congenital Heart Disease/Pediatric Cardiology, Heart- and Diabetescenter NRW, University Clinic of Ruhr University Bochum, Bad Oeynhausen, Germany; <sup>10</sup>Department for Paediatric Cardiology, Saarland University Medical Center, Homburg, Germany; <sup>11</sup>Pediatric Cardiology, University Hospital Ulm, Ulm, Germany

**Background and Aim:** Myocarditis affects primarily young adults and children. It is the most common cause of Sudden Cardiac Death (SCD) in this cohort. In animal models, myocardial involvement during viral infections has been shown to be higher with intensive exertion. The current recommendations for returning to sports after myocarditis are based on cohort and case studies. Often, the restrictions chosen by the medical community are arbitrary and not based on these recommendations and thus lead to poor compliance with serious consequences if not adhered to.

**Method:** This study is a subproject of the MYKKE registry, a German multicenter registry for children with suspected myocarditis. The observation period for this analysis was 84 months (June 2014 to June 2021). Every patient fulfilling the criteria of myocarditis (biopsy proven, positive cardiac biomarker or ECG abnormality) was sent a questionnaire regarding physical activity before, during and after the onset of myocarditis. Furthermore, the current recommendations for return to sport were included. History, cardiac MRI, echocardiography, biopsy, and laboratory records from every patient were retrieved from the MYKKE registry database.

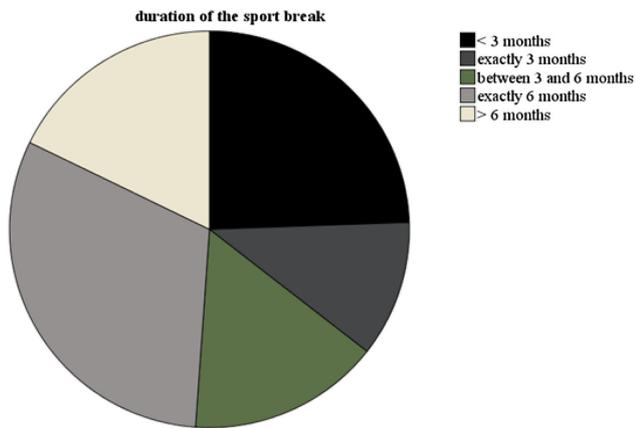
**Results:** 58 patients (average age 14.6 years) from 9 centers participated (65.5% male). Most subjects (84.5%) participated in curricular physical activity, 36% in competitive sports prior to the onset of myocarditis. There was no difference of heart function between the physically active and inactive subjects. Most participants (60.3%) observed symptoms of an infectious disease in the two weeks leading to the myocarditis. Continuing with physical activity despite these symptoms did not lead to a worse outcome. The recommendations regarding the return to sports varied widely and followed current guidelines in merely 45%. Most patients (84.5%)

did not receive the recommended exercise test before returning to sports.

**Conclusions:** Physical activity before and after the onset of myocarditis was not associated with a more severe outcome. There is still a discrepancy between current guidelines and the actual recommendations provided by health care providers. Especially the fact that only a minority of patients received the recommended exercise test before returning to sport is noteworthy, given the fact that malignant arrhythmias must be ruled out.

**Keywords:** myokarditis, acquired heart disease, infectious disease, sudden cardiac death

duration of sport break



#### P-642

##### A new treadmill protocol for cardiopulmonary exercise testing in schoolchildren using field-derived data

Annika Weigelt, Kathrin Rottermann, Sven Dittrich, Isabelle Schöffl  
Department of Pediatric Cardiology, Friedrich-Alexander-Universität, Erlangen-Nürnberg, Germany

**Background and Aim:** Cardiopulmonary exercise testing in young children represents a challenge. So far, mainly the Bruce protocol, has been used for testing children on the treadmill. However, long exercise times in combination with very low starting speeds represent a challenge for most children. Furthermore, because of the weak knee extensors, achieving cardiopulmonary exertion is challenging. Based on data from mobile cardiopulmonary exercise testing in a park, the mean speeds achieved by the children were used to develop a protocol on the treadmill, for reaching maximal exertion based on natural running speeds in a reasonable time-frame.

**Method:** A cardiopulmonary exercise step test on a treadmill was performed by a group of 7 – 10-year-old children. Each step was two minutes long and represented the mean speeds recorded during outdoor testing. The starting speed was set at 3 km/h, representing a slow walk. The second step was increased to 6 km/h according to a slow jog, then to 8 km/h like a strenuous running speed and to 9 km/h, which represented the instruction to run “all out”. Further steps increased the speed by 1 km/h every 2 minutes. The inclination was set at 1%.

**Results:** We were able to test 52 children. All children reached a reasonable exertion level above an RER of 1.0 which was significantly higher for girls who also reached significantly higher peak heart rates. There were no significant differences with respect to

the other cardiopulmonary exercise variables (s. table 1). The duration of the tests was within the recommended 6 – 10 minutes.

**Conclusions:** Transferring natural running speeds determined during outdoor exercise testing onto an indoor treadmill test proved to be feasible for healthy children. The recommended exercise times for reaching maximal exertion were well below exercise times using the Bruce protocol in that age-group but within the recommended time. Interestingly, there were no significant differences between peakVO<sub>2</sub> for girls and boys, contrary to previous studies. This could be attributable to the fact that girls seemed to push themselves harder, reaching significantly higher peak RER and peak heart rates. However, the self-reported extracurricular activity levels were significantly lower for girls.

**Keywords:** cardiopulmonary exercise testing, pediatric cardiology

#### P-644

##### Open field exercise stress testing in very young children with congenital heart disease – reference values for 4–8 year old children

Christian Paech, Philipp Kalden, Roman Gebauer, Ingo Dähnert, Susan Brosig, Franziska Markel, Anna Michaelis  
University of Leipzig, Heart Center, Pediatric Cardiology, Leipzig, Germany

**Background and Aim:** Cardiopulmonary exercise testing represents the diagnostic tool for determining cardiopulmonary function. Especially in small children, exercise testing is challenging. To address this problem, field testing has been implemented using small mobile devices and reference values for healthy children have been established. This study aims at using this protocol for developing normal values for cardiopulmonary exercise testing in very young children with congenital heart disease.

**Method:** Children aged 4–8 years with congenital heart disease were recruited. Three groups were assigned (Tetralogy of Fallot, Fontan and d- Transposition of the great arteries). All children were tested according to an outdoor protocol, in which they were instructed to walk, then run slowly, then a little harder and at last run at full speed. Each step lasted for 2 minutes, except the last step, in which the children were instructed to maintain as long as possible.

**Results:** A total of 78 children (33 female/45 male, mean age 6.2 years) performed outdoor cardiopulmonary exercise testing using a mobile device. Almost all tests were completed successfully (98%). Anthropometric values did not differ between boys and girls. VO<sub>2</sub>peak/kg, respiratory exchange ratio, VT1, heart rate at VT1, and time of exercise were also comparable between sexes. Generally, a tendency of higher maximal oxygen uptake could be observed in older children. And overall lower values than in healthy controls were documented as expected.

**Conclusions:** Open field mobile cardiopulmonary exercise testing represents a novel approach in very young children. In this study, we were able to determine normal values of maximal oxygen uptake and OUES/kg for 4–8-year-old children with congenital heart disease in completion of normal values in healthy children.

**Keywords:** exercise testing, children, congenital heart disease, spirometry

#### P-645

##### First real life data on the diving reflex in healthy children

Christian Paech<sup>1</sup>, Mareike Rixen<sup>1</sup>, Johannes Weidemann<sup>1</sup>, Roman Gebauer<sup>1</sup>, Michael Weidenbach<sup>1</sup>, Franziska Markel<sup>1</sup>, Anna Michaelis<sup>1</sup>, Isabelle Schöffl<sup>3</sup>, Ingo Dähnert<sup>1</sup>, Peter Rüdrieh<sup>2</sup>, Bernd Wolfahrt<sup>2</sup>, Jan Wüstenfeld<sup>2</sup>

<sup>1</sup>University of Leipzig, Heart Center, Pediatric Cardiology, Leipzig, Germany; <sup>2</sup>University of Leipzig, Institute of Applied Exercise Sciences, Leipzig, Germany; <sup>3</sup>University of Erlangen, Pediatric Cardiology, Erlangen, Germany

**Background and Aim:** Swimming and diving are popular recreational activities, representing an effective option in maintaining and improving cardiovascular fitness in healthy people. Additionally, both skills are essential in the prevention of drowning incidents. Yet, immersion into the water confronts the human body and cardiovascular system with several challenges. To date there are almost no data on diving physiology in children. Two recent studies by our group could demonstrate first physiologic data in children with congenital heart disease. Yet, these lack normal values in healthy children to correctly assess the data. Therefore, the current study aims to evaluate the effect of submersion on the heart rate, i.e. diving reflex in healthy children.

**Method:** Patient selection: 50 healthy children, 25 trained and 25 untrained children aged 8–12 years (prepubertal) will be included. Anthropometric data, baseline cardiologic and pulmonary parameters will be obtained.

Measurements on diving reflex and oximetry:

1. Patient at rest on a chair / continuous pulseoximetry / 2 minutes rest / than static apnea in normal air.
2. Patient at rest on a chair / continuous pulseoximetry / 2 minutes rest / than static apnea with the face immersed in cold (same temperature as pool = 28° celsius) water.
3. Patient standing in the pool immersed up to the neck / continuous pulseoximetry / 2 minutes rest / static apnea at a depth of 1m. Transcutaneous oxygen saturation, heart rate, and perfusion index, as an index of peripheral vascular tone, were recorded.

**Results:** The study protocol has been approved by the ethics committee of the University of Leipzig. Currently patient inclusion is not fully closed until 01/2023. Data shall be presented as preliminary data at AEPC Dublin 2023.

**Conclusions:** The current study will present the first structured data on the effect of a submersion on the heart rate and vascular tone, i.e. diving reflex.

**Keywords:** diving reflex, swimming, children, diving

## P-646

### Blood pressure profiles and cardiovascular risk profiles in 8–12-year-old children following pre-eclampsia

Michelle Anna-Katarina Renlund<sup>1</sup>, Tiina Johanna Jääskeläinen<sup>2</sup>, Anni Sofia Emilia Kivelä<sup>3</sup>, Seppo Tapani Heinonen<sup>4</sup>, Hannele Maaret Laivuori<sup>5</sup>, Taisto Askonpoika Sarkola<sup>1</sup>

<sup>1</sup>Children's Hospital, University of Helsinki and Helsinki University Hospital, Helsinki, Finland; <sup>2</sup>Minerva Foundation Institute for Medical Research, Helsinki, Finland; <sup>3</sup>Medical and Clinical Genetics, University of Helsinki and Helsinki University Hospital, Helsinki, Finland; <sup>4</sup>Department of Food and Nutrition, University of Helsinki, Helsinki, Finland; <sup>5</sup>Medical and Clinical Genetics, University of Helsinki and Helsinki University Hospital, Helsinki, Finland; <sup>6</sup>Department of Obstetrics and Gynecology, Helsinki University Hospital, Helsinki, Finland; <sup>7</sup>Medical and Clinical Genetics, University of Helsinki and Helsinki University Hospital, Helsinki, Finland; <sup>8</sup>Department of Obstetrics and Gynecology, Tampere University Hospital and Tampere University, Faculty of Medicine and Health Technology, Tampere Center for Child, Adolescent, and Maternal Health Research, Tampere, Finland

**Background and Aim:** Studies show that pre-eclampsia (PE), and early-onset PE (diagnosis before 34+0 weeks of gestation) in particular, is associated with hypertension and cardiovascular disease later in life in offspring. The aim of this study was to investigate

whether children following PE develop alterations in blood pressure (BP) already early in life and if this is reflected in their cardiovascular phenotype.

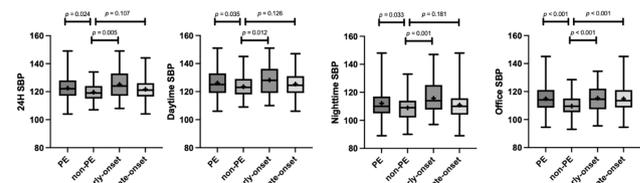
**Method:** Cardiovascular disease risk profiles in 185 PE (46 early-onset and 136 late-onset PE) and 85 non-PE children from the Finnish Genetics of Pre-eclampsia Consortium (FINNPEC) were in a cross-sectional study setting (FINNCARE) assessed 8–12 years from delivery. Office and 24-hour ambulatory BP, body composition, anthropometrics, lipids, glucose, inflammatory markers and pulse wave velocity (PWV) were assessed.

**Results:** Office and 24-hour systolic BP and pulse pressure were significantly higher in PE compared with non-PE. Early-onset PE children had the highest systolic BP values and loads. PE children displayed a greater portion of nondipping during nighttime and significantly higher carotid-femoral PWV, negative augmentation index and central systolic BP and pulse pressure. Early-onset PE was associated with prematurity and small for age birth weight at birth, and smaller height, higher weight and BMI z-scores, as well as higher waist-hip ratio and body fat percentage, and lower high-density lipoprotein concentration at follow-up.

**Conclusions:** Children born following PE develop an adverse BP profile and signs of arterial stiffness already early in life. The alteration in the cardiovascular profile is pronounced in the early-onset form of PE.

**Keywords:** Pre-eclampsia, Blood pressure, Cardiovascular disease, Adiposity, Arterial stiffness

Office and 24-hour systolic blood pressure profiles



Office and 24-hour systolic blood pressure profiles. PE = pre-eclamptic group, non-PE = control group, early-onset = pre-eclampsia diagnosis before 34+0 gestational weeks, late-onset = pre-eclampsia diagnosis at 34+0 gestational weeks or after. Boxplots with whiskers: mean = the + sign, median = the horizontal line of the box, interquartile range = the box limits, minimum and maximum values = the extent of the whiskers.

## P-647

### The impact of the COVID-19 pandemic on recruitment and follow-up in a prospective exercise intervention in young people with congenital heart disease

Lynsey C Forsythe<sup>1</sup>, Nurul H Amir<sup>2</sup>, Dan M Dorobantu<sup>3</sup>, Curtis A Wade<sup>3</sup>, Guido E Piele<sup>4</sup>, Craig A Williams<sup>3</sup>, A. Graham Stuart<sup>5</sup>

<sup>1</sup>Congenital Heart Unit, Bristol Royal Hospital for Children, University Hospitals Bristol and Weston NHS Foundation Trust, Bristol, UK; <sup>2</sup>Department of Translational Health Sciences, Bristol Medical School, University of Bristol; Faculty of Sports Science and Recreation, Universiti Teknologi MARA Cawangan Perlis, Kampus Arau, Perlis, Malaysia; <sup>3</sup>Children's Health and Exercise Research Centre (CHERC), University of Exeter Medical School, University of Exeter, Exeter, UK; <sup>4</sup>Athlete Health and Performance Research Centre and the Sports Medicine Department, Aspetar Qatar Orthopaedic and Sports Medicine Hospital, Doha, Qatar; <sup>5</sup>Congenital Heart Unit, Bristol Royal Hospital for Children, University Hospitals Bristol and Weston NHS Foundation Trust, Bristol, UK; National Institute for Health Research (NIHR), Cardiovascular Biomedical Research Centre, Bristol Heart Institute, Bristol, UK

**Background and Aim:** The pandemic has reduced both the volume of exercise undertaken by young people, especially during lockdowns, and recruitment to non-Covid-19 related trials. The

aim of the study was to describe the effect of the pandemic on recruitment and follow-up in a 6-month, prospective, exercise prescription trial in adolescents with congenital heart disease (CHD).

**Method:** Adolescents, 12-18 years with CHD were invited to participate. Recruitment commenced January 2021 (coinciding with 3rd UK national lockdown) and ended June 2022. Recruitment rate was calculated from the total number of pre-screened, eligible patients. Reasons for non-recruitment were identified and scrutinised by age and sex.

**Results:** Onset of recruitment was delayed by the pandemic leading to a reduced recruitment period. 92 patients were screened, of which 74 (80%) were eligible, 28 patients (38%) were recruited (male  $n = 16$ , female  $n = 12$ ; 12-15 years  $n = 21$ , 16-18 years  $n = 7$ ). 89% were recruited via clinic, 11% via postal invitation. 46 patients (62%) declined participation (male  $n = 28$ , female  $n = 18$ , 12-15 years  $n = 35$ , 16-18 years  $n = 11$ ). 43% of non-recruited patients did not reply or declined invitation (non-specific reasons), were predominantly male and those 12-15 years (Figure 1). Other common reasons for non-participation included; time constraints (predominantly females and those 16-18 years), travel (predominantly female and those 12-15 years), anxiety/mental health (predominantly male and those 16-18 years). Those not wanting extra hospital appointments were all 12-15 years and predominantly male. Only one female, 12-15 years, cited a Covid-19 specific reason.

Concerning Covid-19, 6 participants (21%) tested positive during follow-up. 5 face-to-face appointments were cancelled and rebooked (1 = positive test, 4 = symptoms/close contact). Trial extensions were afforded to 4 participants ( $n = 3$  positive and  $n = 1$  had two periods of symptomatic isolation). All face-to-face researchers ( $n = 4$ ) had a positive test at some point during the follow-up period without impact on booked appointments. Although recruitment rate was low, participant retention was excellent (96%).

**Conclusions:** A less than desirable recruitment rate may have been accentuated by the pandemic but primarily this exaggerated the difficulty of interventional studies in adolescents. Future studies should minimise the identified barriers to participation. Despite the impact of the pandemic on follow-up, initial participant reports are encouraging.

**Keywords:** Exercise, Physical Activity, Adolescents, Congenital Heart Disease, Covid-19

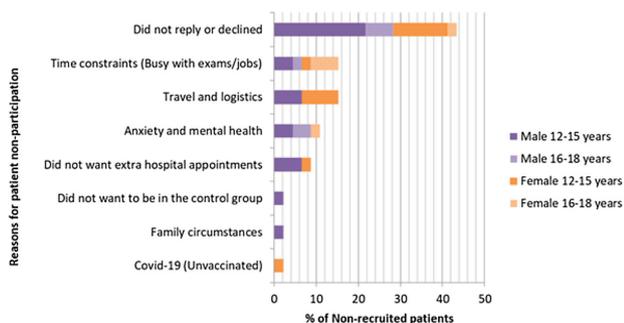


Figure 1: Age and sex responses by non-recruited patients following an invitation to take part in a 6-month, prospective exercise prescription trial in adolescents with congenital heart disease (CHD).

## P-648

### Elevated lipoprotein(a) in the youth – effects on the vascular system

Magdalena Langer<sup>1</sup>, Theresa Vilismaier<sup>2</sup>, Marie Kramer<sup>1</sup>, Franziska Sciuk<sup>1</sup>, Brenda Kolbinger<sup>2</sup>, André Jakob<sup>1</sup>, Pengzhu Li<sup>1</sup>, Nina Rogenhofer<sup>2</sup>, Nikolaus Alexander Haas<sup>1</sup>, Robert Dalla Pozza<sup>1</sup>, Christian Thaler<sup>2</sup>, Felix Sebastian Oberhoffer<sup>1</sup>

<sup>1</sup>Department of Pediatric Cardiology and Intensive Care, University Hospital, LMU Munich, 81377 Munich, Bavaria, Germany; <sup>2</sup>Division of Gynecological Endocrinology and Reproductive Medicine, Department of Obstetrics and Gynecology, University Hospital, LMU Munich, 81377 Munich, Bavaria, Germany

**Background and Aim:** Approximately 1.4 billion people worldwide are affected by elevated lipoprotein(a) (Lp(a)) >50mg/dL. Increased Lp(a) levels are strongly associated with the early onset of adult cardiovascular disease. In recent years, the clinical management of subjects with elevated Lp(a) has gained remarkable interest. Limited data on Lp(a) and its impact on vascular function in young subjects is available. Hence, this study aimed to investigate if elevated Lp(a) levels already impact vascular integrity in young subjects.

**Method:** Young subjects with a Lp(a) >50mg/dL and controls with a Lp(a) ≤50mg/dL were selected from the Munich heARTerY study cohort. This cohort consists of subjects conceived through assisted reproductive technologies and spontaneously conceived peers. Lp(a) was assessed in all study participants at the day of vascular screening. Vascular function was assessed by an oscillometric blood pressure device measuring brachial systolic blood pressure (SBP), brachial diastolic blood pressure (DBP) and pulse wave velocity (PWV). Utilizing the EndoPAT2000 device, the reactive hyperemia index (RHI) was recorded. Carotid intima-media thickness (cIMT) was evaluated sonographically. Continuous and normally distributed variables were compared using the independent-samples t-test. The Mann-Whitney-U-test was utilized for non-normally distributed variables. A  $p$ -value <0.05 was considered as statistically significant.

**Results:** In total, 21 study participants with a Lp(a) >50mg/dL (female: 42.9%) and 42 controls (female: 42.9%) were included in this study. There were no statistically significant differences in age between both groups (11.93 (5.05/24.34) years vs. 11.94 (4.99/24.28) years,  $p = 0.759$ ). No significant differences were detected in SBP (113.86±8.97 mmHg vs. 111.96±9.48 mmHg,  $p = 0.448$ ), DBP (65.02±8.65 mmHg vs. 64.95±6.93 mmHg,  $p = 0.976$ ) and cIMT (0.44±0.03 vs. 0.44±0.02,  $p = 0.876$ ). In addition, both groups did not differ significantly in PWV and RHI. Absolute Lp(a) levels did not correlate significantly with parameters of vascular function.

**Conclusions:** Young subjects with Lp(a) levels >50mg/dL did not show vascular alterations compared to controls. As the current study was limited by its sample size and the relatively young age of study participants, larger follow-up studies are required to detect potential vascular alterations at advanced age. Nevertheless, young subjects with a Lp(a) >50mg/dL should be encouraged to implement healthy lifestyle habits to minimize their cardiovascular risk.

**Keywords:** Lipoprotein(a), Vascular Function, Youth, Prevention

## P-649

**Comparison of total physical activity levels between device-based and self-reported measurement among young people with congenital heart disease**

Nurul Hidayah Amir<sup>1</sup>, Lynsey Forsythe<sup>2</sup>, Curtis A Wadey<sup>3</sup>, Dan Mihai Dorobantu<sup>3</sup>, Massimo Caputo<sup>4</sup>, Guido E Piele<sup>5</sup>, A. Graham Stuart<sup>6</sup>, Craig A Williams<sup>3</sup>

<sup>1</sup>Department of Translational Health Sciences, Bristol Medical School, University of Bristol, Bristol, United Kingdom; <sup>2</sup>Faculty of Sports Science and Recreation, Universiti Teknologi MARA Cawangan Perlis, Kampus Arau, Perlis, Malaysia; <sup>3</sup>Congenital Heart Unit, Bristol Royal Hospital for Children, University Hospitals Bristol and Weston NHS Foundation Trust, Bristol, United Kingdom; <sup>4</sup>Children's Health and Exercise Research Centre (CHERC), University of Exeter Medical School, University of Exeter, Exeter, United Kingdom; <sup>5</sup>Department of Translational Health Sciences, Bristol Medical School, University of Bristol, Bristol, United Kingdom; <sup>6</sup>National Institute for Health Research Cardiovascular Biomedical Research Centre, Bristol Heart Institute, Bristol, United Kingdom; <sup>7</sup>Athlete Health and Performance Research Centre and the Sports Medicine Department, Aspetar Qatar Orthopaedic and Sports Medicine Hospital, Doha, Qatar; <sup>8</sup>Institute of Sport, Exercise and Health, University College London, London, United Kingdom; <sup>9</sup>Congenital Heart Unit, Bristol Royal Hospital for Children, University Hospitals Bristol and Weston NHS Foundation Trust, Bristol, United Kingdom; <sup>10</sup>National Institute for Health Research Cardiovascular Biomedical Research Centre, Bristol Heart Institute, Bristol, United Kingdom

**Background and Aim:** Young people with congenital heart disease (CHD) are less active than their healthy peers negatively affecting quality of life and long-term outcomes. Despite no specific guidelines, a commonly used measure of physical activity (PA) is moderate-to-vigorous PA (MVPA), which has the disadvantage of neglecting light-intensity activity and thus does not reflect total PA. Therefore, the aim of this study was to assess total PA including light, moderate, and vigorous PA (LMVPA), average MVPA, and sedentary time; and compare device-based and self-reported PA levels among young people with CHD.

**Method:** Twenty-eight adolescents with CHD (12–17 years) were recruited in a prospective, randomised controlled trial. Participants wore a GENEActiv accelerometer for 7-days, data being included if wear time was  $\geq 16$  hours/day and  $\geq 4$  days. Total LMVPA, average MVPA and sedentary time were calculated using validated age-specific cut-off points by Phillips (2013). Participants also completed self-reported PA diary. Independent and paired sample t-tests analysed and compared PA intensities and sedentary time. Pearson's correlation coefficients analysed the relationships between both PA instruments.

**Results:** Total 7-days LMVPA (Table 1) did not differ by NYHA class ( $p = 0.243$ ). Participants in NYHA I recorded higher average MVPA ( $p = 0.017$ ) and lower sedentary time ( $p = 0.007$ ) than other classes. Average weekday MVPA was higher ( $p = 0.001$ ) than on weekends. Self-reported total LMVPA in NYHA I participants was higher (34%) than NYHA class II-III ( $p < 0.005$ ). A positive-moderate correlation was found between device-based and self-reported total LMVPA ( $r = 0.486$ ,  $p = 0.014$ ). Bland-Altman analysis demonstrated a proportional bias ( $p < 0.001$ ) indicating that self-reported PA tended to underestimate light PA and overestimated vigorous PA (Figure 1).

**Conclusions:** Despite variations in sedentary time and MVPA patterns, participants are more active during the weekdays compared to the weekends and total LMVPA was similar across the groups. The discrepancies between device-based and self-reported total LMVPA were likely caused by young people's propensity to exaggerate vigorous and underestimate light PA in self-reported diary.

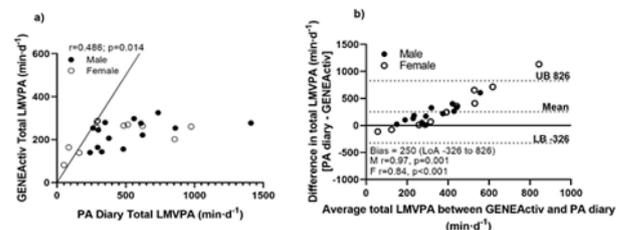
Based on current WHO PA recommendations for healthy youth, it is unknown whether PA levels for young people with CHD is optimal. Therefore, including light, moderate and vigorous PA is needed to reflect the overall total PA of young people with CHD.

**Keywords:** physical activity, total LMVPA, MVPA, accelerometer, self-reported PA, young people with CHD

**Table 1:** Participants' demographics and comparison of activity between accelerometer and self-reported PA diary.

	Total (N=28)	NYHA Class I (n=10)	NYHA Class II & III (n=18)	p-value
<b>Participants' characteristics</b>				
Gender, male (%)	16 (59.3%)	5 (55.6%)	11 (64.7%)	0.59
Age (years)	14.21 $\pm$ 1.60	14.40 $\pm$ 1.58	14.11 $\pm$ 1.64	0.66
Height (cm)	165.7 $\pm$ 9.8	162.9 $\pm$ 7.3	167.3 $\pm$ 10.8	0.27
Body Mass (kg)	60.0 (50.3-62.1)	61.5 $\pm$ 16.4	60.0 (50.4-61.6)	0.32
Fat Percentage (%) <sup>a</sup>	25.6 $\pm$ 10.7 <sup>a</sup>	27.1 $\pm$ 12.1 <sup>a</sup>	24.8 $\pm$ 10.0 <sup>a</sup>	0.60
Fat Mass (kg) <sup>a</sup>	12.1 (8.3-19.7) <sup>a</sup>	12.70 (9.93-22.95) <sup>a</sup>	14.43 $\pm$ 7.68 <sup>a</sup>	0.39
Free fat mass (kg) <sup>a</sup>	42.5 $\pm$ 6.0 <sup>a</sup>	43.5 $\pm$ 7.1 <sup>a</sup>	40.7 (37.0-45.4) <sup>a</sup>	0.49
<b>Device-based accelerometry (N=28)</b>				
Total 7-days LMVPA (min-d <sup>-1</sup> )	223.31 $\pm$ 64.97	242.85 $\pm$ 70.40	212.46 $\pm$ 61.08	0.243
Average MVPA (min-d <sup>-1</sup> )	57.03 $\pm$ 28.16	73.58 $\pm$ 26.91	47.84 $\pm$ 25.01	0.017*
Average LPA (min-d <sup>-1</sup> )	167.27 $\pm$ 44.24	171.79 $\pm$ 50.47	164.76 $\pm$ 41.74	0.70
Sedentary time (min-d <sup>-1</sup> )	763.01 $\pm$ 105.42	694.13 $\pm$ 72.69	801.27 $\pm$ 102.53	0.007*
Weekday average MVPA (min-d <sup>-1</sup> )	61.59 $\pm$ 30.83**	81.29 $\pm$ 29.53*	50.64 $\pm$ 26.31	0.009*
Weekend average MVPA (min-d <sup>-1</sup> )	44.78 $\pm$ 28.01	52.20 $\pm$ 25.14	40.66 $\pm$ 29.35	0.31
<b>Self-reported diary (N=25)<sup>†</sup></b>				
Total 7-days LMVPA (min-d <sup>-1</sup> )	482.00 $\pm$ 309.36	692.00 $\pm$ 376.92	363.88 $\pm$ 189.76	<0.005*

Values are presented as mean ( $\pm$  standard deviation) or median (interquartile range) as appropriate. Total LMVPA = total light, moderate, vigorous physical activity; MVPA = moderate-to-vigorous physical activity; NYHA Class = New York Heart Association Classification. <sup>a</sup>1 missing value for these variables due to ICD in-situ. \*Significant difference  $p < 0.05$ . <sup>†</sup>3 missing values due to participants not returning the self-reported diary. <sup>‡</sup>Meeting 2020 WHO PA guidelines: at least an average of 60 minutes per day of moderate-to-vigorous intensity, mostly aerobic, physical activity, across the week; should incorporate vigorous-intensity aerobic activities, as well as those that strengthen muscle and bone, at least 3 days a week; should limit the amount of time spent being sedentary, particularly the amount of recreational screen



**Figure 1:** a) Pearson's correlation coefficient and b) Bland-Altman plot displaying agreement between accelerometry and PA diary.

## P-650

**Return to competitive sport after oncohematological disease: one will not fit all**

Giulia Cafiero<sup>1</sup>, Giordano Ugo<sup>1</sup>, Tranchita Eliana<sup>1</sup>, Girardi Katia<sup>2</sup>, Gentili Federica<sup>1</sup>, Turchetta Attilio<sup>1</sup>

<sup>1</sup>Department of Cardiac Surgery, Cardiology and Heart Lung Transplant, Sport Medicine Unit, Bambino Gesù Children's Hospital, Rome, Italy.;

<sup>2</sup>Department of Pediatric Hemato-Oncology and Cell and Gene Therapy, Scientific Institute for Research and Healthcare (IRCCS) Bambino Gesù Children's Hospital, Rome, Italy.

**Background and Aim:** Although since years there has been talk of the fundamental role of sport in oncohematological patients, the reality in the pediatric field is quite different. There are still few projects that envisage the practice of physical activity as an integral part of the therapeutic path. And the situation does not improve at the end

of the treatment: scant scientific data support the safety of a early return to physical activity even at a competitive level, especially among the youngest. The aim of this study is the evaluation of cardiovascular complications following the early return to competitive physical activity in a girl undergoing therapy for Hodgkin's lymphoma.

**Method:** A 15-year-old girl with Hodgkin's lymphoma, who underwent chemotherapy according to the EURONET PHL C2 protocol, was followed from a cardiological point of view in the two years following the end of the therapy. The protocol provided for: monthly electrocardiogram (EKG) for the first three months. Subsequently EKG, echocardiocolordoppler, maximal ergometric test, spirometry at the 3rd, 6th, 10th, 12th, 18th and 24th months from the end of therapy. The girl resumed controlled training already in the final stages of the therapeutic protocol and the first competitive certificate for athletics was issued 3 months after the therapy was stopped.

**Results:** No cardiovascular complications have been detected during follow-up to date.

**Conclusions:** Although the data refer to a single patient, the evidence should stimulate increasingly in-depth studies in relation to the return to physical activity even at a competitive level. Although the role of sport in the treatment of pediatric oncohematological patients is evident, it is necessary to support these evidences more in order to favor a peaceful recovery of these children not only physically but also psychologically.

**Keywords:** children, physical activity, oncohematological disease

#### P-651

##### Medical care of patients with congenital heart disease (CHD) during the first 18 months of the covid-19 pandemic in germany

Paul Christian Helm, Cornelia Tremblay, Ulirike Bauer  
National Register for Congenital Heart Defects, Berlin, Germany

**Background and Aim:** With the onset of the COVID-19 pandemic, it was unclear how the pandemic would affect the medical care of CHD patients, what a COVID-19 infection would mean for CHD patients, and what behavioral recommendations should be made.

**Method:** We performed an online survey of a representative cohort of CHD patients (04-07/2021) to collect information about the medical care they received. 3, 655 patients participated in the survey [3, 179 (87%) completed the questionnaire]: 458 (14.4%) simple CHD, 1, 244 (39.1%) moderate CHD, 1, 200 (37.7%) complex CHD, 277 (8.7%) other/unclassified CHD.

**Results:** Since the COVID 19 pandemic (January 2020 to June 2021), 1, 264 patients (39.8%) had at least one visit to a cardiologist/pediatric cardiologist office, and 1, 482 patients (46.6%) had at least one visit to a hospital/cardiac center cardiologist/pediatric cardiologist. 2, 092 patients (65.8%) saw another physician during that time period. Only 218 patients (6.9%) did not see a physician during the COVID-19 pandemic. In 304 cases (9.6%), patients/parents canceled a medical appointment or follow-up visit with a pediatric cardiologist/cardiac clinic/hospital due to Corona. Cardiac catheterization was canceled in 26 (0.8%) and surgery in 30 cases (0.9%) by patients/parents because of Corona. In 315 cases (9.9%), the physician canceled a medical appointment or check-up with a pediatric cardiologist/cardiac clinic/hospital. Cardiac catheterization was canceled in 33 cases (1%) and surgery in 52 cases (1.6%) by the hospital. 1, 722 patients/parents have

spoken with their physician about Corona and/or received information about Corona. 955 individuals (30%) reported that they called the physician because they wanted to know about Corona and CHD. Overall, in 1, 472 cases (85.5%), the physician answered CHD related questions about Corona satisfactorily.

**Conclusions:** It was shown that during the first one and a half years of the COVID-19 pandemic, that medical care, even cardiac surgeries, were cancelled/postponed due to patient/parent's concerns, but also due to the high burden on the German healthcare system. In addition, the treating physicians often could not answer questions about COVID-19 and CHD.

**Keywords:** Corona, CHD, medical care, online survey

#### P-652

##### Cardiopulmonary screening after SARS-COV-2 infection in children and adolescents returning to sports: A retrospective analysis

Maximilian Dettenhofer<sup>1</sup>, Miriam Reuter<sup>1</sup>, Christine Stratmann<sup>1</sup>, Tobias Engl<sup>1</sup>, Katharina Köble<sup>1</sup>, Mikiko Harada<sup>1</sup>, Renate Oberhoffer Fritz<sup>2</sup>, Frauke Mühlbauer<sup>2</sup>

<sup>1</sup>Institute of Preventive Pediatrics, Department of Sport and Health Sciences, Technical University of Munich, Germany; <sup>2</sup>Institute of Preventive Pediatrics, Department of Sport and Health Sciences, Technical University of Munich, Germany & Department of Congenital Heart Defects and Pediatric Cardiology, German Heart Center Munich, Germany

**Background and Aim:** The COVID-19 pandemic has raised concern about cardiopulmonary risks for athletes returning to sport (RTS) after an infection with the virus. While initial protocols recommended evaluation of all young athletes, current guidelines diversify screening procedures based on disease severity. However, there are few systematic studies addressing RTS-screening in the paediatric age group.

**Method:** We conducted a retrospective analysis of athletes seen at our institute within 3 months after infection with SARS-CoV-2 (09/2020-10/2022). They received resting electrocardiogram (R-ECG), echocardiography, and – in a subset – cardiopulmonary exercise testing (CPET).

**Results:** 136 recreational and competitive athletes were included (22.8% female; aged 13.2 ± 2.7 years, range 6-17). 53 subjects (39.0%) presented within one month after infection, 63 (46.3%) within the second, and 20 (14.7%) within the third month. The majority (n = 127, 93.4%) reported asymptomatic or mild infection, 9 (6.6%) reported moderate disease. Residual symptoms were noted in 39 athletes (28.7%), 29 of whom (21, 3%) reported exercise intolerance, which in 12 cases (8.8%) persisted after 28 days. 15 athletes (11.0%) reported other symptoms, including unspecific chest pain, dyspnea, palpitations, or vertigo during exercise.

R-ECG and echocardiography were performed in all and CPET in 82 subjects (60.3%). Abnormal findings included premature atrial contractions (PAC) in R-ECG or CPET in 3 cases (2.2%), 1 of which reported moderate COVID-19. Further, premature ventricular contractions (PVC) during CPET were noted in 2 cases (1.5%), 1 of which reported residual symptoms. Otherwise, ECG and CPET were classified as normal. Echocardiography identified no pathologies in any of the cases.

Using Fisher's exact test, no statistically significant association was found between PAC in R-ECG and disease severity (p = 0.13) or residual symptoms (p = 1.00), nor between PAC / PVC during exercise and disease severity (p = 0.14) or residual symptoms (p = 1.00).

**Conclusions:** None of our screened athletes presented with myo- or pericarditis after SARS-CoV-2 infection; however, residual

symptoms were reported. For some, mild cardiac involvement remained plausible. Our results support current RTS-guidelines based on an individual approach depending on disease severity and presence of cardiac symptoms.

**Keywords:** COVID-19, return-to-sport, cardiac risks, athletes, paediatric

#### P-653

##### **Resilience as a protective factor- study in children and adolescents with and without chronic diseases visiting a summercamp**

Nicola Stöcker<sup>1</sup>, Dominik Gaser<sup>2</sup>, Renate Oberhoffer Fritz<sup>2</sup>, Christina Sitzberger<sup>2</sup>

<sup>1</sup>Applied Sport Sciences, TU Munich, Germany; <sup>2</sup>Preventive Pediatrics, TU Munich, Germany

**Background and Aim:** Children with congenital heart diseases and other chronic diseases have lower life quality and a greater risk of depression, especially after two years of covid lockdowns. In turn, resilience and a high life quality can act as protective factors against depression. Therefore, this study evaluates the association between the participation in a recreation camp for children with chronic diseases and their resilience before and after the camp. **Method:** In this pre-post-study 33 children and adolescents (10, 47 ± 3, 01 years), thereof 16 male and 15 with chronic diseases, participated in the kidsTUMove-summercamp (08.08.-13.08.22) in Germany. The intervention had social, physical, and psychical elements. The life quality was measured with kidscreen questionnaire and the resilience with a validated questionnaire (Schumacher et al.).

**Results:** There is a tendency for higher scores in this study population concerning resilience after attending the camp (pre: 62.91 ± 10.42, post: 63.61 ± 10.89%;  $p > 0, 05$ ). No significant differences showed between sex (male pre: 60.53 ± 9.21, post: 61.07 ± 9.8 and female pre: 62.88 ± 7.51, post: 63.76 ± 8.3;  $p > 0, 05$ ), age (<12 years, pre: 61.9 ± 9.42, post: 61.75 ± 9.88 and ≥12 years, pre: 61.58 ± 6.37, post: 63.75 ± 7.51;  $p > 0, 05$ ) and with/without chronic diseases (pre: 59 ± 7.99, post: 60.47 ± 9.43/ pre: 64 ± 8.19, post: 64.56 ± 8.65;  $p > 0, 05$ ). Life quality results are outstanding.

**Conclusions:** Special elements in a camp can help to reach a good health boost. It would be important to generate even more targeted content in order to offer children and young people more protective resources for processing in traumatic experiences such as in the Covid pandemic.

**Keywords:** chronic diseases, resilience, protective factor

#### P-654

##### **Parental recommendations and exercise attitudes in congenital hearts (preach)**

Curtis A Wadey<sup>1</sup>, Fiona Leggat<sup>2</sup>, Nurul Amir<sup>3</sup>, Julia Potter<sup>4</sup>, Guido E Pieleś<sup>5</sup>, Lynsey Forsythe<sup>6</sup>, Graham Stuart<sup>6</sup>, Alan R Barker<sup>1</sup>, Craig A Williams<sup>1</sup>

<sup>1</sup>Children's Health & Exercise Research Centre (CHERC), Public Health and Sport Science, Faculty of Health and Life Sciences, University of Exeter, Exeter, UK.; <sup>2</sup>Population Health Research Institute, St George's University of London, London, UK.; <sup>3</sup>Department of Translational Health Sciences, Bristol Medical School, University of Bristol, Bristol, UK & Faculty of Sports Science and Recreation, Universiti Teknologi MARA Cawangan Perlis, Kampus Arau, Perlis, Malaysia; <sup>4</sup>Department of Physical Education, University of Chichester, Chichester, UK.; <sup>5</sup>Athlete Health and Performance Research Centre and the Sports Medicine Department, Aspetar Qatar Orthopaedic and Sports Medicine Hospital, Doha, Qatar; <sup>6</sup>Bristol Congenital Heart Centre, The

Bristol Heart Institute, University Hospitals Bristol NHS Foundation Trust, Bristol, UK.

**Background and Aim:** Young-people with congenital heart disease (ConHD) benefit from regular physical activity (PA) and their parents have been reported as potential facilitators and barriers to their PA behaviour. PA promotion is often neglected by clinicians, which may also influence the PA behaviours in young-people with ConHD. The aim of this study was to explore parental factors (demographics, PA, and attitudes), their child's factors (demographics, clinical severity), and clinical experience on PA behaviours in young-people with ConHD.

**Method:** A online questionnaire was co-developed with parents (n = 5) who have children with ConHD. The questionnaire had closed (87%) and open-ended (13%) questions and was distributed nationally by UK ConHD networks and social media. A mixed-methods approach was utilised for data-analysis. Specifically, frequency analysis, Chi<sup>2</sup>, Fishers Exact, and reflexive thematic analysis were used to analyse the data.

**Results:** There were 83 respondents of which 94% were mothers. Young-people with ConHD were 53% female, aged between 0-21 years, and performed moderate (84%) and/or vigorous PA (65%). Parental demographics were not associated to young-people's PA, however, parental participation in PA ( $\chi^2(1) = 6.9$ ,  $P < 0.05$ ) and parents viewing PA as important for their child was associated with young-people's PA (Fisher's Exact,  $P < 0.05$ ). Some parents (~15%) were unsure if exercise was safe, and most (~70%) were unsure where they could receive additional information. Parents reported that their children had never received an exercise test (n = 61, 85%) or PA advice (n = 52, 72%). Of those who received PA advice (n = 20), 50% said it was not consistent between staff or visit. Qualitative analysis produced the theme of "Knowledge is power and comfort", many parents described not knowing what PA was appropriate, nor the impact of PA both in the short and long-term. Where information was provided in ConHD clinics, this could contribute to parental confusion.

**Conclusions:** Whilst a large proportion of young-people performed PA, parental PA and attitudes likely influence their child's PA. Uncertainty remains in what activity young-people should undertake, possibly due to the lack and inconsistency of advice offered by ConHD clinics. Young-people with ConHD may benefit from PA prescription with their families in ConHD clinics.

**Keywords:** physical activity promotion, parental influences, exercise prescription, congenital heart disease.

#### P-655

##### **Detection of risk factors for atherosclerosis in families with premature acute coronary syndrome**

Kristina Rücklová<sup>1</sup>, Veronika Urbanová<sup>2</sup>, Markéta Pavlíková<sup>3</sup>, Aleš Hořínek<sup>4</sup>, Petr Toušek<sup>5</sup>

<sup>1</sup>Klinikum Stuttgart, Stuttgart, Germany and <sup>3rd</sup> Faculty of Medicine, Charles University, Prague, Czech Republic; <sup>2</sup>3rd Faculty of Medicine, Charles University, Prague, Czech Republic; <sup>3</sup>Faculty of probability and medical statistics, Prague, Czech Republic; <sup>4</sup>Institute of biology and medical genetics, Prague, Czech Republic; <sup>5</sup>Dept. of cardiology, University Hospital Královské Vinohrady, Prague, Czech Republic

**Background and Aim:** Atherosclerosis has been demonstrated already in childhood and many risk factors (RF) have a cumulative negative effect. Some of the RF usually affect several family members including children and may be modifiable, such as unhealthy lifestyle, obesity, hypertension, diabetes mellitus, familial hypercholesterolaemia (FH) and psychosocial stress. Other, less

known RF include hyperhomocysteinaemia, antiphospholipid syndrome, chronic inflammatory conditions, malignancies and environmental pollution and noise.

**Method:** Individuals (total 32, men 23, women 9) after acute coronary syndrome (ACS) aged <50 years (median 43) and their children (total 39, men 23, women 16) were included in the study. The RF were detected by means of a detailed questionnaire, a physical and a laboratory examination. The latter was focused on lipid analysis, fasting glycaemia, HbA1c, homocysteine, anti-phospholipid autoantibodies, molecular genetic analysis of genes associated with FH, etc. The incidence of RF in patients with premature ACS was compared with healthy age and sex matched controls. Children with detected RF were offered a multidisciplinary intervention.

**Results:** Among patients with premature ACS, smoking, psychosocial stress, sedentary lifestyle and obesity represented the most prevalent RF and were detected in 24/32 (75%), 23/32 (72%), 21/32 (66%) and 15/32 (42%), respectively. No significant difference was found between men and women in the prevalence of the respective RF. Almost three quarters (74%) of the offspring had at least one preventable RF including sedentary lifestyle (49%), unhealthy diet (50%), smoking (27%) and hypercholesterolaemia (26%).

**Conclusions:** Based on our results a systematic detection of RF in children from families with premature ACS has a high yield and this approach may potentially decrease the risk of cardiovascular deaths in these children in the future. Our results also confirmed the hypothesis that male gender as such represents a cardiovascular RF.

Supported by institutional grant Cooperatio "Pediatrics", from Charles University.

**Keywords:** premature atherosclerosis, risk factors

#### P-656

##### Evaluation of fitness capacities of children and young adults with marfan and related conditions

Aitor Guitarte Vidaurre, Thomas Edouard, Bertrand Chesneau, Maud Langeois, Eric Garrigue, Yves Dulac

Centre de Référence du Syndrome de Marfan et des Syndromes Apparentés, Hôpital des Enfants - Toulouse (France)

**Background and Aim:** Early diagnosis of Marfan syndrome (MFS) leads us to a comprehensive approach to the multisystem damage related to connective tissue fragility. Impairment in exercise capacity has a great impact on quality of life (QoL) and rehabilitation programs need to address his ultimate causes. We have analysed fitness capacities of patients from a young age until early adulthood, and analysed the causes of limitation.

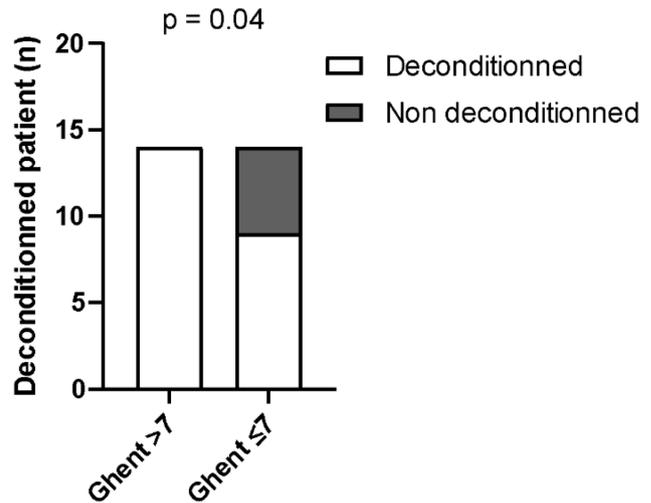
**Method:** Early diagnosis of Marfan syndrome (MFS) leads us to a comprehensive approach to the multisystem damage related to connective tissue fragility. Impairment in exercise capacity has a great impact on quality of life (QoL) and rehabilitation programs need to address his ultimate causes. We have analysed fitness capacities of patients from a young age until early adulthood, and analysed the causes of limitation.

**Results:** Early diagnosis of Marfan syndrome (MFS) leads us to a comprehensive approach to the multisystem damage related to connective tissue fragility. Impairment in exercise capacity has a great impact on quality of life (QoL) and rehabilitation programs need to address his ultimate causes. We have analysed fitness capacities of patients from a young age until early adulthood, and analysed the causes of limitation.

**Conclusions:** Early diagnosis of Marfan syndrome (MFS) leads us to a comprehensive approach to the multisystem damage related to

connective tissue fragility. Impairment in exercise capacity has a great impact on quality of life (QoL) and rehabilitation programs need to address his ultimate causes. We have analysed fitness capacities of patients from a young age until early adulthood, and analysed the causes of limitation.

**Keywords:** Marfan syndrome, CPET, sports, QoL,



**Figure 1** Fisher test showing that all patients in our study with a positive systemic score by Ghent criteria had a muscular deconditioning

#### P-657

##### Does surgical complexity influence aerobic capacity or sports participation in children with congenital heart disease?

Astrid M De Souza, David C Clarke, Pearl Waraich, James E Potts, Cindy Sha, Kathryn R Armstrong  
Children's Heart Centre, Pediatric Cardiology, British Columbia Children's Hospital, Vancouver, Canada; The University of British Columbia, Vancouver, Canada

**Background and Aim:** Previous work by our group has shown that children with congenital heart disease (CHD) who regularly participate in extracurricular sport ( $\geq 2-3$  days/week) have a higher peak aerobic capacity (VO<sub>2</sub>peak) than those who do not participate or only partake in their physical education class at school. There were no differences in VO<sub>2</sub>peak based on underlying lesion. AIM: We sought to determine whether surgical course affected VO<sub>2</sub>peak in a cohort of CHD patients.

**Method:** We performed a 2-year, single-centre, retrospective review (May 2016–November 2018) on CHD patients with aortic valve disease (AoV), surgically-corrected coarctation (CoA), Ross procedures (Ross), tetralogy of Fallot (ToF), and transposition of great arteries (TGA). The Risk Adjustment for Congenital Heart Surgery-1 (RACHS-1) was calculated for all patients. Sports participation was assessed by an ACSM Clinical Exercise Physiologist at the time of testing and categorized into 3 groups: 0-1 day/week; 2-3 days/week;  $\geq 4$  days/week. A validated institutional treadmill protocol (BCCH) was used. The criteria for a maximal test included: VO<sub>2</sub> plateau, respiratory exchange ratio >1.0, and/or a peak heart rate >200 bpm. VO<sub>2</sub>peak z-scores and gas exchange threshold (GET) were calculated. Data are reported in frequency

tables and as medians and interquartile ranges. Kruskal-Wallis and Mann Whitney U tests were used to test for group differences. A  $P < 0.05$  was considered statistically significant.

**Results:** Fifty-three CHD patients (68% male) were included based on diagnosis: 5 AoV; 12 CoA; 6 Ross; 20 ToF, and 10 TGA. RACHS-1 scores in this cohort are as follows: 1 = 6%; 2 = 36%; 3 = 41%; 4 = 17%. VO<sub>2</sub>peak for the respective RACHS-1 scores was as follows: 1 = 36.7; 2 = 36.6; 3 = 39.8; 4 = 44.7 mL/min/kg. There were no significant differences in VO<sub>2</sub>peak, VO<sub>2</sub>peak z-score or VO<sub>2</sub>GET based on RACHS-1 score. There were also no significant differences in those who participated in sport based on their RACHS-1 score: (1: n = 3; 2: n = 14; 3: n = 14; 4: n = 7).

**Conclusions:** There is no relationship between RACHS-1, VO<sub>2</sub>peak, and sport participation in this cohort of CHD patients.

**Keywords:** aerobic capacity, pediatrics, sports participation, exercise, RACH-1 score, congenital heart disease

#### P-660

##### How well are CHD patients and their twin siblings vaccinated in times of the covid-19 pandemic?

Jia Li<sup>1</sup>, Paul Christian Helm<sup>2</sup>, Renate Oberhoffer Fritz<sup>3</sup>, Ulrike M M Bauer<sup>4</sup>, Peter Ewert<sup>5</sup>, Julia Remmele<sup>6</sup>

<sup>1</sup>German Heart Center Munich, Technical University of Munich, Department of Congenital Heart Disease and Pediatric Cardiology, Munich, Germany; <sup>2</sup>Ludwig Maximilian University of Munich, Munich, Germany; <sup>3</sup>National Register for Congenital Heart Defects, Berlin, Germany; <sup>4</sup>Technical University of Munich, Chair of Preventive Pediatrics, Munich, Germany; <sup>5</sup>National Register for Congenital Heart Defects, Berlin, Germany; <sup>6</sup>Competence Network for Congenital Heart Defects, Berlin, Germany; <sup>7</sup>German Heart Center Munich, Technical University of Munich, Department of Congenital Heart Disease and Pediatric Cardiology, Munich, Germany; DZHK (German Centre for Cardiovascular Research), Partner Site Munich Heart Alliance, Munich; Competence Network for Congenital Heart Defects, Berlin, Germany; <sup>8</sup>German Heart Center Munich, Technical University of Munich, Department of Congenital Heart Disease and Pediatric Cardiology, Munich, Germany; <sup>9</sup>Technical University of Munich, Chair of Preventive Pediatrics, Munich, Germany

**Background and Aim:** Due to hospitalizations, surgeries, interventions and contact with family and friends, patients with CHD are more susceptible to infectious diseases from an early age and therefore are often at risk of a severe course of infection. Especially in times of the Corona pandemic, protection through vaccination is more important than ever before. The aim is to investigate if vaccination recommendations are met by CHD patients and their twin siblings and the differences between them. **Method:** In total, 73 twin pairs (85 girls (58.2%), 12.37 ± 8.38 years) were recruited nationwide since August 2019 within the framework of the “Same Same, but different?” twin study. Six out of the 73 pairs both twins have CHD. Data on immunization status and medical history were collected through medical documents and compared with the German recommendations of the Standing Committee on Vaccination at the Robert Koch Institute. **Results:** In 35 pairs (58.3%) both met the recommendations for basic vaccination, while in 4 pairs only the CHD twin was fully vaccinated. All pairs were completely vaccinated against Measles, Mumps and Rubella. Patients with CHD reached

complete basic immunization at the mean age of 4.3 ± 3.8 years, while their siblings without CHD reached basic immunization at 4.0 ± 3.9 years ( $p = 0.092$ ). In 32 pairs (43.8%) both received at least one flu vaccination. Out of 29 pairs recruited after the approval of the first COVID-19-vaccination on December 21st, 2020, 15 pairs (51.7%) from the age of 5 years were vaccinated against COVID-19 after a mean time of 1.03 ± 0.42 years. While 75% of the twins with complex CHD were vaccinated against COVID-19, only 33.3% of the simple and moderate CHD twin pairs received a vaccination.

**Conclusions:** The COVID-19-vaccination coverage is higher among the complex CHD group than the simple and moderate CHD group. Vaccination recommendations were met by most of the twin pairs, especially against Measles, which is obligatory for attending German kindergarten and school. COVID-19 and influenza vaccination rates in the immediate circle of patients with CHD must be increased through better education of parents, patients and medical staff for successful prevention.

**Keywords:** chd, vaccination, immunization, twin study

#### P-661

##### Energy drinks and adverse cardiovascular events in minors: A review of case reports

Pengzhu Li, Robert Dalla Pozza, André Jakob, Nikolaus Alexander Haas, Guido Mandilaras, Felix Sebastian Oberhoffer  
Division of Pediatric Cardiology and Intensive Care, University Hospital, LMU Munich, Munich, Germany

**Background and Aim:** Energy Drinks (ED) are sweetened beverages containing stimulant substances such as caffeine or taurine. Approximately 30 to 50 % of teenagers and young adults consume EDs. In correlation with their popularity, reports on adverse cardiovascular events in minors associated with ED consumption are rising. This review of case reports aimed to summarize adverse cardiovascular events in minors associated with the consumption of EDs.

**Method:** The Pubmed database was searched for articles published before August 23, 2022 using the following text words individually or in combination: “Energy drink\*” and “Child\*” or “Adolescent\*” or “Teen\*” or “Youth\*” and “Heart” or “Cardiovascular” and “Case\*” or “Case Reports” or “Letter” or “Comment”. Inclusion criteria were as follows: (1) reported patients were <18 years of age, (2) the consumption of EDs was confirmed, (3) case reports were written in English.

**Results:** In total, seven cases were included (age: 13 to 17 years; sex: 5 male). The following adverse cardiovascular events were reported: spontaneous coronary artery dissection with ST-segment elevation myocardial infarction; acute coronary artery vasospasm; long QT syndrome unmasked through ED consumption; narrow-complex tachycardia with atrial fibrillation and occasional atrial flutter; atrial tachycardia and atrial fibrillation with a rapid ventricular response; sinus tachycardia and arterial hypertension; postural tachycardia syndrome. In two cases, additional trigger factors (e.g., physical activity, alcohol consumption) were described. Three subjects displayed preexisting health conditions including recurrent fainting episodes, questionable history of myopericarditis, and ADHD.

**Conclusions:** This review of case reports suggests that the consumption of EDs in combination with additional trigger factors or in the

presence of preexisting health conditions seems critical. To prevent adverse health events in the future, children and adolescents should be informed about ED-linked risk factors and responsible ED consumption behaviors.

**Keywords:** Energy Drinks, cardiovascular events, children

**P-662**

**QTc at brisk standing in children with psychiatric drug treatment**

Estelle Naumburg, Linn Bromée, Björn Reynisson  
Department of clinical sciences, pediatrics, Umeå University, Sweden

**Background and Aim:** The prescription rate of psychotropic drugs to children and adolescents in Sweden has increased over the last decades, as well as the incidence of syncope and other cardiac symptoms. Some psychotropic drugs are known to be pro-arrhythmic, which may be caused by a prolonged QT-interval. Prolongation of the QT-interval may be concealed on standard ECG in a supine position, while it can be prolonged when standing. The aim of this study was to assess the QTc interval in supine and brisk standing position in children and adolescents with psychotropic drug treatment in relation to age and number of drugs.

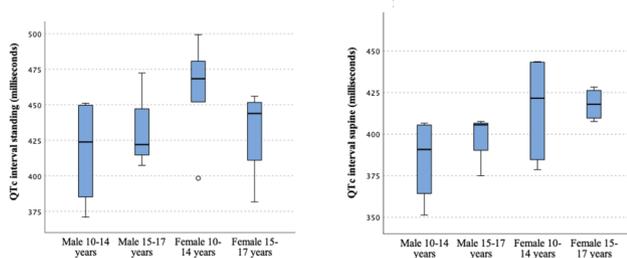
**Method:** Patients aged 10-17 years with at least one psychotropic drug treatment were included. An ECG in an active standing position was registered immediately after a resting supine ECG. QTc was calculated, using Bazette's formula, for each participant in both body positions. The QTc difference between supine and standing position was calculated for number of psychiatric drugs, gender, and age. Information on cardiac related symptoms was retrieved.

**Results:** Overall 16 patients (7 male and 9 female) were included in the study, age 10-17 years. Mean QTc interval in supine position was 404.5 ms, while standing was 437.2. None of the patients included had a QTc interval over 450 ms in supine position. In seven patients the QTc was more than 450 ms while standing. For girls age 10-14 years the QTc interval in varied the most between the patient and increased more compared to other groups in brisk standing. There was no difference in QTc with regard to number of drugs. Seven patients had experienced palpitations and five at least one event of unexplained syncope.

**Conclusions:** Prolongation of QTc interval in young patients with psychotropic drug treatment was not seen this small study. However, there was a great variation between the patients and especially in younger girls. An ECG in supine as well as standing position can be recommended for children and adolescents with psychiatric drug treatment in presence of clinical symptoms, but the clinical impact has to be studied further.

**Keywords:** QTc prolongation, Psychotropic drug treatment, Cardiac symptoms

Figure 1. QTc in standing and supine position with regard to age and sex.



**Figure 1** QTc in standing and supine position with regard to sex and age

**P-663**

**Exercise capacity in children with d-loop transposition of the great arteries: can we identify factors related to worse exercise performance?**

Erika Rezola Arcelus<sup>1</sup>, Marta Iglesias Barroso<sup>1</sup>, Begoña Manso García<sup>2</sup>, Esteban Peiro Molina<sup>3</sup>

<sup>1</sup>Paediatric Cardiology Department. Donostia University Hospital. San Sebastian, Spain.; <sup>2</sup>Paediatric Cardiology Department. Virgen del Rocío University Hospital. Seville, Spain.; <sup>3</sup>Paediatric Cardiology Department. La Fe University Hospital. Valencia, Spain.

**Background and Aim:** Arterial switch operation (ASO) has replaced atrial switch operation as the preferred approach for infants with D-loop of the great arteries (D-TGA). Survivors of this surgery have been found to have better functional capacity and exercise performance than previous atrial switch operation survivors, assessed by cardiopulmonary exercise testing (CPET). However, risk factors in ASO related to inferior exercise performance need to be evaluated.

**Method:** Multicentre retrospective review of patients with D-TGA status post ASO who underwent CPET between march 2017 and June 2022. We compared CPET results in patients with simple D-TGA (group 1) and D-TGA and previous ventricular septal defect (VSD) repair (group 2). All underwent standardised CPET, using Godfrey ramp protocol as recommended by the European Society of Cardiology (ESC). Measured variables were: minute ventilation (VE), breath reserve (BR), peak heart rate (pHR), peak systolic arterial pressure (pSAP), rate pressure product (RPP), peak oxygen consumption (pVO2) and peak oxygen pulse (pOxygenPulse). Exclusion criteria included palliative surgery prior to ASO, severe ventricular or valve dysfunction (stenosis or regurgitation) and severe coronary anomalies.

**Results:** We identified 43 patients who met entry criteria, 25 in group 1 and 18 in group 2. Both groups were homogeneous in terms of age, body mass index (BMI) and gender distribution. Table 1 resumes demographic characteristics and CPET results in both groups.

**Conclusions:** In our series, group 1 patients had a significantly better ventilatory, chronotropic and tensional response than group 2, and a better functional capacity (higher pVO2 and pOxygenPulse), although results are not statistically significant. Thus, simultaneous VSD is related to a worse exercise performance.

**Keywords:** Cardiopulmonary exercise test, D-loop Transposition of the Great Arteries, Exercise performance

	Group 1 (n=25)	Group 2 (n=18)	p
Age (y) (mean±SD)	13,3±3,3	12,4±2,7	0,361
BMI (kg/m2) (mean±SD)	19,1±2,9	20,8±3,9	0,092
Gender: men (%)	20 (80%)	9 (50%)	0,149
women (%)	5 (20%)	9 (50%)	
VE (l/min) (mean±SD)	66,3±16,7	53,8±12,8	0,011
BR (%) (mean±SD)	27,8±1,8	38,8±1,1	0,033
pHR (bpm) (mean±SD)	170,9±14,6	160,2±23,1	0,042
pSAP (mmHg) (mean±SD)	152,3±29,8	129,5±20,65	0,008
RPP (bpm*mmHg) (mean±SD)	26049±60003,2	20793,4±4819	0,004
pVO2 (ml/kg) (mean±SD)	34,7±8,4	30,4±6,6	0,085
pOxygenPulse (ml) (mean±SD)	12,1±4,3	10,1±2	0,445

Table 1. Demographic characteristics and CPET results in group 1 and 2.

**Table 1.** Demographic characteristics and CPET results in group 1 and 2.

## P-664

**Perception of functional capacity and physical activity and correlation with response to physical exercise in pediatric patients with fontan surgery**

Roman Gestoso<sup>1</sup>, Esther Aurensanz<sup>2</sup>, Emma Grau Sió<sup>2</sup>, Joan Sanchez De Toledo<sup>3</sup>

<sup>1</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain; <sup>2</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain; Institut de Recerca Sant Joan de Déu, Barcelona, Spain; <sup>3</sup>Department of Pediatric Cardiology, Hospital Sant Joan de Déu, Barcelona, Spain; Institut de Recerca Sant Joan de Déu, Barcelona, Spain; Department of Critical Care Medicine, University of Pittsburgh, Pittsburgh, Pennsylvania, United States

**Background and Aim:** Fontan Surgery has drastically changed the long-term prognosis of children born with univentricular congenital heart disease (CHD). Nevertheless, decrease functional capacity (FC) and significant loss of quality of life (QOL) continues to affect children surviving total cavopulmonary (TCP) surgery. Data on self-reported outcomes and physical activity habits is scant.

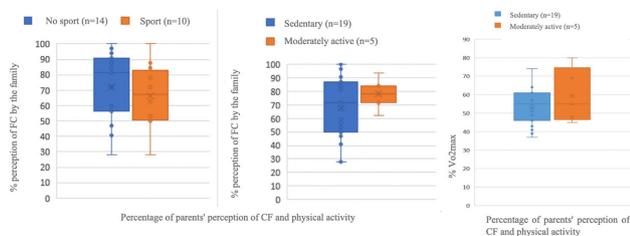
**Method:** Prospective observational study of patients aged 7-18 years (n = 24) who underwent TCP surgery at a tertiary referral heart center. Comparison between self-reported QOL and FC measured by PedsQL of patients and their relatives was compared with FC measured by CPET and physical activity assessed via the "Assesment of Physical Activity Levels" (APALQ) scale.

**Results:** Patients were evaluated at a median age of fifteen years old. Fourteen (58, 3%) did not practice any sport. They had similar parental FC perception by PedsqQL (72% (SD 6.04) compare to 10 patients who practiced 66.5%(SD 6.87) p = 0.550). By APALQ, parents of the 19 sedentary patients perceived FC 67.59%(SD 5.4), while 5 non-sedentary patients 78.12% p = 0.350. FC assessment by CPET demonstrated similar VO2max% in patients who practice sport compared to those who did not (51.79% (SD 2.33) vs. 57.70%(SD 4.21) p = 0.200). No VO2Max% differences were observed comparing sedentary vs. non-sedentary patients assessed by APALQ [52.89%(SD 2.26) and 59.40%(SD 6.60) p = 0.249]. no positive relationship was found between the minutes of activity/week and the VO2max% reached (R<sup>2</sup> 0.003) p = 0.870. A positive correlation between the self-perceived FC and parents perception using the PedsQL in the PHYSICAL aspect (R<sup>2</sup> 0.420) p = 0.030

**Conclusions:** Our study shows that no correlation exists between FC perceived by parents and the physical activity patients perform. Surprisingly, parents who consider better FC corresponded to the children with less physical activity. There is a correlation between the perception of the parents FC and self-perceived by the patients, but this does not occur when we compare their perceptions with the real FC measured by CPET. Patients who practiced sports had a better VO2max%, such as the non-sedentary ones, although it was not significant. Our sample has shown that many of our patients are sedentary, but we have not been able to clarify the reasons that lead to this sedentary habit.

**Keywords:** CPET, Fontan, PedsQL, APALQ, QOL

## RESULTS



## P-666

**Nutrition in children and adolescents with congenital heart defects during the covid-19 pandemic**

Paul Christian Helm<sup>1</sup>, Dominik Tobias<sup>2</sup>, Claudia Niessner<sup>3</sup>, Anna Lena Ehmman<sup>1</sup>, Sigrid Hahn<sup>4</sup>, Christian Apitz<sup>2</sup>, Ulrike Bauer<sup>1</sup>, Jannos Siaplaouras<sup>2</sup>

<sup>1</sup>National Register for Congenital Heart Defects, Berlin, Germany; <sup>2</sup>Division of Pediatric Cardiology, Children's Hospital, University of Ulm, Ulm, Germany; <sup>3</sup>Institute of Sports and Sports Science, Institute of Technology, Karlsruhe, Germany; <sup>4</sup>Fulda University of Applied Sciences, Fulda, Germany

**Background and Aim:** Appropriate nutrition is important for healthy physical and mental development in children and adolescents with congenital heart defects (CHD). The still ongoing COVID-19 pandemic has also affected dietary patterns. Lockdown measures, the closing of food service establishments, and the partial elimination of school lunches, for example, due to school closures has a direct impact on dietary behavior.

**Method:** The aim of the study was to learn more about the nutrition and physical activity of young and adolescent CHD-patients. CHD patients registered in the German National Register for Congenital Heart Defects were invited by email/post to join the study. In total, 894 patients have finished the online questionnaire (average age: 12.5±3 years; 47.2 % female; 23.8 % simple CHD, 37.8 % moderate CHD, 38.4 % complex CHD).

**Results:** Nutrition counseling has been provided in 146 cases (simple CHD: 10.3%; moderate CHD: 13.3%; complex CHD: 23%; p < .001). 165 patients (18.5%) reported eating more sweets, chips, or the like at the time of the interview than before the COVID-19 pandemic, 86 patients (9.6%) ate more fruits and vegetables than before the pandemic, 79 patients (8.8%) ate more meals per day than before the pandemic (simple CHD: 12.7%; moderate CHD: 8%; complex CHD: 7.3%; p < .01) and 64 patients (7.2%) drank more soda/juices than before the pandemic. Overall, 125 patients (14%) reported having become fatter during the COVID-19 pandemic, 43 patients (4.8%) had become slimmer, and 726 (81.2%) reported having become neither fatter nor thinner as a result of the pandemic (simple CHD: fatter (16.4%), thinner (8.5%), neither nor (75.1%); moderate CHD: fatter (13%), thinner (4.7%), neither nor (82.2%); complex CHD: fatter (13.4%), thinner (2.6%), neither nor (84%); p < .05).

**Conclusions:** Overall, young and adolescent CHD patients' assessment of their own dietary behavior seems to depend to a small extent on the severity of CHD. The pandemic has a major impact on the dietary behavior of young CHD patients. Unfortunately, during the pandemic period, it seems only partially successful to support and encourage young patients especially in a good, balanced and healthy diet to avoid secondary diseases.

**Keywords:** congenital heart defects, nutrition, COVID-19, pandemic, online survey

## General Cardiology

## P-318

**Acute COVID 19 infection in patients with congenital heart disease (CHD)**

Cornelia Tremblay, Paul Christian Helm, Ulrike Maria Margarethe Bauer

National Register for Congenital Heart Defect, Berlin, Germany

**Background and Aim:** Many patients with CHD are considered to have an increased risk for a severe course of COVID. Which patients are affected?

*Method:* We performed telephone interviews with COVID positive patients with CHD. Baseline information was extracted from our nationwide data bank, with further details from hospital discharge letters.

*Results:* 152 (4.2 %) out of 3,655 “online questionnaire” patients with CHD have been infected with COVID. Ninety-nine were interviewed (male 50.5%): 28 children, 32 young adults (up to 29 years), and 39 “older adults”. Mild CHD was present in 16, moderate in 36, and complex in 41 patients (cyanotic 10.1 %). Twelve had native CHD and ten underwent univentricular palliation. 37 patients had additional non-cardiac risk factors (immunocompromise, chronic lung disease, etc). The course of COVID was mild in 50, moderate in 38, and severe in 3 patients, who had to be admitted to the hospital (one to the ICU).

Around half of the patients had fever and cold-associated symptoms (runny nose, sore throat, cough), 55.6 % flu-like symptoms (malaise, muscle, joint pains). 46 patients reported a loss of smell and/or taste, 55 had dizziness, 57 headache, and 7 confusion. More severe respiratory symptoms like shortness of breath were reported by 19 patients, 6 had decreased oxygen saturations, and 5 pneumonia. Gastrointestinal Symptoms (decreased appetite, weight loss, nausea, vomiting, diarrhea) were present in 52 patients. Two reported chest tightness, 8 patients had arrhythmias, and 2 impaired heart function. One patient had myocarditis. Two patients reported impaired pulmonary function, one patient reported a temporary compromise of renal function and liver function. Other severe complications (ARDS, PIMS, ) were not reported. Two patients received steroids, 4 antibiotic treatment, 2 oxygen, and one needed mechanical ventilation. The average time to recover was 12 days.

*Conclusions:* Despite an underlying CHD, the course of COVID in our cohort seems to be comparable to the general population. Even patients with complex CHD, who underwent univentricular palliation or those with pulmonary hypertension, had mostly a mild to moderate course.

**Keywords:** Congenital Heart Disease, COVID, children, adults, univentricular palliation

### P-319

#### Long-COVID in patient with congenital heart disease (CHD)

Cornelia Tremblay, Paul Christian Helm, Ulrike Maria Margarethe Bauer

*National Register for Congenital Heart Defect, Berlin, Germany*

*Background and Aim:* According to general published data between 7.5% and 41% of Covid patients have problems with long-COVID-symptoms. Are patients with CHD as likely to be affected with Long-COVID?

*Method:* We interviewed COVID-positive patients with CHD by phone. Baseline information was taken from our data bank, further details were extracted from rehabilitation letters.

*Results:* Ninety-nine COVID-positive patients were included (male 50.5%): 28 children, 32 young adults, and 39 older than 29 years. 16 patients had mild, 36 moderate, and 41 complex heart lesions. A univentricular form was present in 11 patients, overall, 10.1 % were cyanotic. Thirty-six patients reported temporary symptoms in the weeks following the acute phase. Thirty-three percent complained of fatigue, 18 mentioned temporary concentration and memory problems, and 16 patients had trouble finding the right words. Long-COVID (ongoing symptoms after 12 weeks) was reported by 31 patients (2 children, 9 young adults, and 20 (51, 3 %) older adults. Twenty-three reported impaired activity level, 22 decreased exercise tolerance, 14 shortness of breath, 18 post-exertional fatigue, and/or 4 postural orthostatic tachycardia syndrome. Sixteen patients described cognitive dysfunction (“brain fog”), 13 ongoing loss of smell/taste, 3 sleeping difficulties, and 2 anxiety and depression. Three patients underwent rehabilitation for Long-Covid. One patient had a significant reduction of the lung capacity. The second one suffered from decreased exercise tolerance, shortness of breath, Fatigue-Syndrome, and ongoing muscle pain. The last one had shortness of breath, Fatigue-syndrome, brain fog, and ongoing sleeping difficulties. The only described symptoms in children with long-covid were impaired activity level, decreased exercise tolerance, and shortness of breath. For the adult population the occurrence of Long-Covid symptoms was more frequent in older (51 %) than in younger adults (20 %).

*Conclusions:* Surprisingly the number of patients with CHD and Long-Covid-symptoms are in the higher range compared to the general population. Children in our cohort had primarily physical symptoms; cognitive issues were rarely reported. Our conclusions are limited by the small sample size and that most of the data are self-reported.

**Keywords:** Long-COVID, Congenital Heart Disease, children, adults