Cardiology in the Young

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Postnatal short-term outcomes of pulmonary atresia with ventricular septal defect following prenatal diagnosis: a preliminary Vietnamese report

Linh Thuy Dinh[1] and Quang-Vinh Tran[2,3]

[1]Prenatal Diagnosis and Neonatal Screening Center, Hanoi Obstetrics and Gynecology Hospital, Hanoi, Vietnam; [2]Faculty of Surgery, Hanoi Medical University, Hanoi, Vietnam and [3]Cardiovascular Surgery, Heart Center, Vietnam National Children's Hospital, Hanoi, Vietnam

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Corresponding author: Quang-Vinh Tran; E-mail: vinhtran.taurus@gmail.com

Abstract

Introduction: Pulmonary atresia with ventricular septal defect (PA/VSD) is a complex cyanotic CHD that requires an early diagnosis for optimal management and outcomes. The purpose of this study is to evaluate the effectiveness of an inter-hospital management protocol utilising the prenatal CHD diagnosis for achieving favourable postnatal outcomes in PA/VSD patients in Vietnam. *Methods:* We described the protocol implemented between two tertiary medical settings in Vietnam for the prenatal diagnosis and postnatal management of PA/VSD infants. All PA/VSD patients with prenatal diagnosis between January 2016 and December 2022 were retrospectively reviewed. The primary outcome was postnatal survival, and the secondary outcome was the presence of major morbidities such as bleeding or the need for Extra Corporeal Membrane Oxygenation (ECMO) after total repair. *Results:* During the study period, 35 PA/VSD patients were identified including 29 infants who underwent surgical correction utilising a valved conduit and 6 infants who are still waiting for the next evaluation after the palliative surgery. No death prior to the surgery occurred. For 29 patients, one hospital death happened, two patients required ECMO initially in their postoperative course but both survived, one late mortality due to pneumonia, and three reoperations were due to conduit failure. In the mean follow-up time of 2.92 (0.51–7.92) years, all survivors had completed follow-up. *Conclusion:* Our protocol including a multidisciplinary management and a close follow-up has shown promising short-term results in achieving favourable postnatal outcomes for PA/VSD patients.

Introduction

Pulmonary atresia with ventricular septal defect (PA/VSD) is a rare complex cyanotic CHD defined as a discontinuity from the ventricle to the pulmonary arteries and a connection between ventricles. The term tetralogy of Fallot with pulmonary atresia has been used interchangeably with PA/VSD to better describe the intracardiac lesions.[1] Depending upon the source of the pulmonary blood flow, this CHD was classified into three types: A, B, and C according to the presence of the native pulmonary atresia, postnatal status of the ductus arteriosus, and the presence of major aorto-pulmonary collateral arteries (MAPCAs). PA/VSD remains a complex defect to manage with the many variations of pulmonary circulation contributing to differing management strategies among heart centres. Without surgery, less than 10% of PA/VSD patients will live through their first decade of life. Optimal surgical treatment aims to achieve a total biventricular repair including closure of the ventricular septal defect, establishment of continuity from the right ventricle to the pulmonary artery, and as needed, depending on the pulmonary anatomy, construction of a pulmonary bed able to manage the right ventricle's cardiac output.

A recent report shows that the Asian prevalence of CHD is 9.342 per 1000 live births, which is notably higher than CHD prevalence rates in Europe and the USA.[2] With the increased utilisation of prenatal echocardiographic (ECHO) to diagnose CHD, the postnatal outcomes of CHD have been shown to improve in low- and middle-income countries (LMICs). However, an increased detection rate of a prenatal CHD by ECHO may lead to a reduced birth incidence of severe complex CHD through a high rate of termination of pregnancy, which in turn may affect the actual prevalence of a specified defect.[3,4] PA/VSD can be detected prenatally by fetal ECHO with a high degree of accuracy, while the intracardiac septal anatomy and the primary source of the pulmonary blood supply may be visualised, whether by an arterial duct or by MAPCAs, if retrograde left-to-right flow is seen on colour flow Doppler.[5]

There are a few reports on the outcomes of PA/VSD in LMICs.[6] One possible explanation for this is that the use of fetal ECHO in pregnancy is not well established and could be more challenging to perform in LMICs due to equipment needs and training. In this report, we

described our protocol, which we implemented between two tertiary care centres in Vietnam since 2016, in order to better standardise management of CHD patients, including PA/VSD patients from prenatal diagnosis to postnatal care. Additionally, we focus our evaluation on the specific cardiac defect—PA/VSD—which might potentially benefit from the implementation of this protocol. The primary outcome of our study was overall postnatal survival, while the secondary outcome was the presence of major surgical morbidities and/or the need for reoperation or reintervention after a total biventricular repair.

Materials and methods

Subjects

This retrospective study was approved by the institutional review board of Vietnam National Children's Hospital (VNCH) (IRB – VN/01037/00011976). Written informed consent for any procedure (amniocentesis, surgery, genetic testing, CT scan with anaesthesia) was obtained from the legal guardians of all patients. Due to the retrospective nature of the study, informed consent for the study was exempt, but the study ensured that the rights and well-being of patients were protected throughout the study. Inclusion criteria for participation in the study included (1) having a confirmed prenatal ECHO diagnosis of PA/VSD at the Hanoi Obstetrics and Gynecology Hospital (HOGH) and (2) having a surgical intervention (biventricular repair or palliative aorto-pulmonary shunt) done between January 2016 and December 2022. Exclusion criteria included surgical treatment at a non-study institution. Eventually, 35 PA/VSD patients were enrolled in the study, including 29 infants who underwent total biventricular repair including ventricular septal defect closure and placement of an right ventricle to pulmonary artery valved conduit and 6 infants who are still waiting further diagnostic evaluation after palliative aorto-pulmonary artery shunt surgery.

PA/VSD management protocol

The prenatal ECHO diagnosis of possible CHD was initially made by an obstetrician at HOGH during routine second-trimester morphology scanning ultrasound (between 18 and 22 weeks of gestation). If CHD was suspected, the patient was then referred for further fetal echocardiography evaluation by a paediatric cardiologist from VNCH. Postnatal diagnosis and principal management of these patients were conducted at the Heart Center. In order to discuss management options and to provide appropriate care and support, the families received counselling from our multidisciplinary team, including a geneticist, obstetrician, paediatric cardiologist, cardiac surgeon, and neonatologist. Further, if the diagnosis was made before 27 weeks of gestational age (GA), the mother was asked to provide consent and then underwent amniocentesis and genetic testing. However, if patients were referred from other medical facilities with a GA greater than 27 weeks, the pregnancy followed the expectant standard of care obstetric management without amniocentesis. Those who chose to continue their pregnancies after diagnosis had regular check-ups that were conducted every 2 weeks at HOGH until delivery. Newborns with PA/VSD were initially examined by a neonatologist at HOGH, then referred to VNCH's neonatal ICU (NICU) for further assessment, and were cared for in the NICU until undergoing surgical interventions as indicated.

Surgical procedures

The decision to do a complete biventricular repair or palliative surgery was chosen based on the evaluation of pulmonary artery anatomy type. In type A, pulmonary blood flow is provided solely by a Patent Ductus Arteriosus (PDA). In type B, pulmonary blood flow is provided by both native pulmonary arteries and by MAPCA(s). In type C, there are only MAPCA(s).[1]

Palliative surgery consisted of either a modified Blalock–Taussig shunt or a central aorto-pulmonary shunt, specifically in cases where severe hypoplasia of both pulmonary artery branches was seen or in conjunction with concurrent unifocalisation of MAPCAs if these were the sole blood supply for the lungs. At VNCH, PDA stenting is not considered an alternative to surgical shunting due to the lack of interventional expertise in this procedure. However, this strategy may represent a valuable option in centres equipped with interventional cardiology expertise. Total biventricular repair of the PA/VSD defect was performed either primarily as a single-stage procedure or in a staged manner after first receiving a palliative aorto-pulmonary shunt, followed later by a detailed assessment that confirmed favourable pulmonary artery anatomy. The total biventricular correction technique, utilising a valved conduit, was described in our previous article.[7]

Variables and statistical analysis

Demographic and clinical data at the time of total repair with a valved conduit were collected from the patient's medical records. R studio was utilised to perform statistical analysis, and descriptive statistics was utilised to summarise the demographic and clinical characteristics of the study population. Non-normally distributed quantitative variables are presented as the median and interquartile ranges. The Kaplan–Meier curve was utilised for survival analysis. The primary outcome was postnatal survival, while the secondary outcome was the presence of major complications and reoperation or reintervention.

Results

Flow of the study

In total, between January 2016 and December 2022, 1774 pregnancies were prenatally suspected of having a fetus with a CHD. GA at the time of CHD diagnosis ranged from 18 to 36 weeks of pregnancy. A total of 219 fetuses were identified with conotruncal abnormalities including tetralogy of Fallot, common truncus arteriosus, PA/VSD, interrupted aortic arch, and transposition of great arteries. Among this group, termination of pregnancy with or without genetic results was decided in 53 cases. Of those who chose to continue their pregnancy, there were no in utero fetal deaths during the study period. Eventually, 35 PA/VSD babies were delivered and managed at the two tertiary care study centres. This accounted for 15.98% of the conotruncal anomalies found in utero, with PA/VSD accounting for 1.97% among all the CHD cases in this group. No mortality was seen in the PA/VSD study patients prior to surgery. Eighteen patients underwent a single-stage primary total biventricular repair, while 17 patients underwent aorto-pulmonary shunt as their first surgery. Of those in the staged group, 11 patients underwent total biventricular repair as their second surgery, and the remaining 6 patients are still waiting for further diagnostic evaluation after palliative aorto-pulmonary artery shunt surgery. All survivors had complete follow up, with a mean follow-up time of 2.92 (0.51–7.92) years. Patient flow is displayed in Figure 1.

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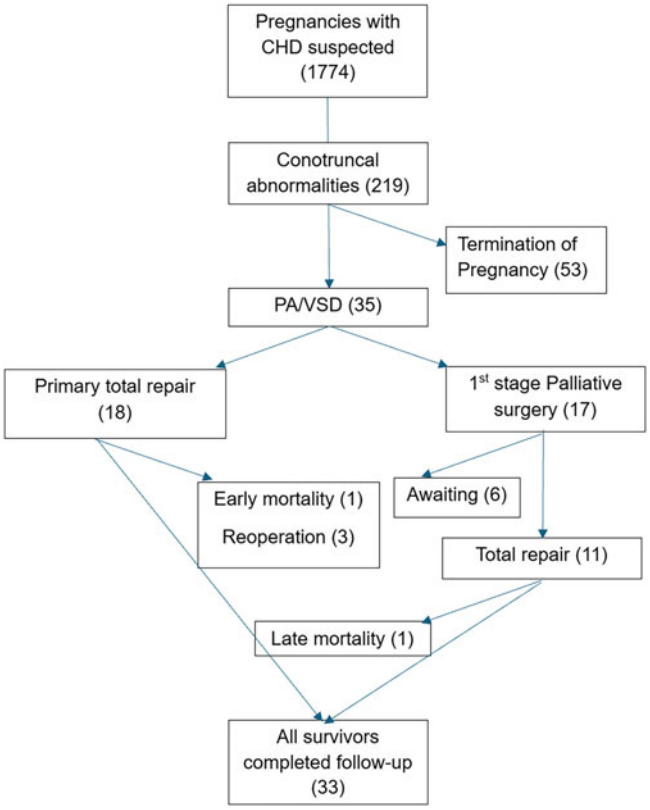

Figure 1. Flow of the prenatal diagnosis and postnatal management of PA/VSD infants. PA/VSD = pulmonary atresia with ventricular septal defect.

Table 1. Characteristics of PA/VSD patients with prenatal diagnosis

Characteristics	*n* = 35 (%)
Gender	
Male	23 (65.7)
Female	12 (34.3)
Diagnosis	
Type A Type B Type C	24 (68.6) 9 (25.7) 2 (5.7)
Premature	4 (11.4)
Preoperative ventilation	4 (11.4)
22q11	3 (8.6)
Extracardiac abnormalities	8 (22.8)
Right Aortic Arch	13 (37.1)
Left Superior Vena Cava	6 (17.1)

PA/VSD = pulmonary atresia with ventricular septal defect.

Table 2. Surgical variables

Variables	Primary total repair (*n* = 18)	Staged repair (*n* = 17)
Age at the first surgery (months)	1.6 (0.8–2.6)	1.7 (1–3)
Waiting time until repair (months)		12.8 (7.9–20.2)
	Total repair (*n* = 29)	
Age at the total repair (months)	3.5 (1.3–12)	
Weight at the total repair (kg)	4.3 (3.5–7.7)	
Conduit size (mm)	12 (10–16)	

Characteristic variables

Demographics and variables of the 35 study patients are described in Tables 1 and 2. Twenty-three of the study patients were male (65.7%), and type A pulmonary morphology accounted for 68.6% of the study group. Median age at the first palliative aorto-pulmonary shunting procedure was 1.7 months, and the median age for primary one-stage biventricular repair was 1.6 months. There were four (11.4%) premature babies managed at our centres prior to any surgeries, but none of them had extremely low birth weight. Of this premature group, one patient died after the single-stage total biventricular repair, while the remaining three survived, resulting in a survival rate of 75%. Genetic testing was performed in 16 patients (45.7%) who had not undergone maternal amniocentesis during pregnancy. DiGeorge syndrome was identified in 3 patients (8.6%), and an additional 8 patients (22.8%) had extra-cardiac abnormalities such as hypospadias, urinary tract defects, ano-rectal malformations, limb defects, and cleft lip and/or palate.

Survival

In the study group, there was no fetal death or postnatal mortality prior to any surgical intervention. In total, there were 29 patients (82.9%) who underwent total biventricular repair with the use of a valved conduit for restoration of pulmonary blood flow between the right ventricle and the pulmonary artery. There was one in-hospital death after total biventricular repair. In this patient, PA/VSD and abnormal right arm bones were detected at 22 weeks of gestation, but the family refused amniocentesis. The patient was born preterm at 35 weeks via vaginal delivery, with a birth weight of 2.7 kg. Despite an abnormal phenotypic appearance on clinical examination raising the suspicion of a genetic disorder, karyotyping and FISH tests showed a male (46,XY) karyotype and no 22q11 deletion. When the neonate weighed approximately 2.8 kg at 39 days of age, the one-stage biventricular surgical correction was performed. After surgery, the patient experienced dysrhythmias requiring AAI mode pacing, fluid therapy resuscitation, and significant inotropes support due to persistent hypotension and metabolic acidosis. Despite pacing, the patient later experienced refractory bradycardia, which did not respond to CPR and expired on Post-operative day (POD) 3.

There was one late death in our study. A 20-month-old male died approximately 6 months after a total biventricular repair due to sepsis and pneumonia. The patient was a full-term infant with a birth weight of 3.0 kg. The prenatal diagnosis of PA/VSD was made at 35 weeks of gestation, so amniocentesis was not indicated. The patient presented with an abnormal facial feature and an aberrant right subclavian artery arising from a left-sided aortic arch, which raised a strong concern for a genetic disorder. However, karyotyping showed a normal male karyotype (46,XY), but no further genetic testing was performed. He underwent a 4 mm left Blalock–Taussig shunt via a left thoracotomy at 1 month of age, followed at 14 months of age by a total biventricular repair. His ICU course was unstable with a pericardial effusion requiring

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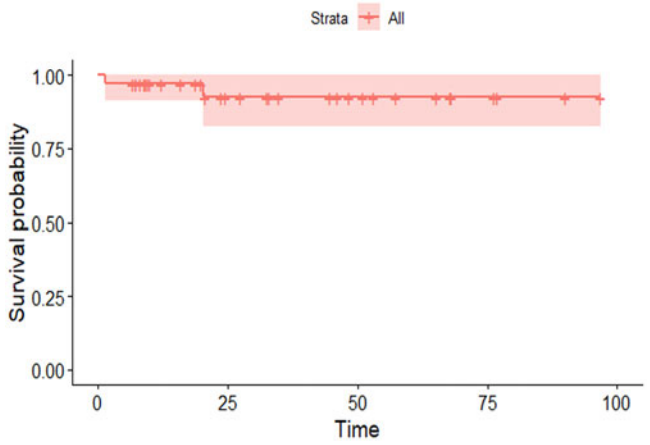

Figure 2. Kaplan–Meier survival curve (time in months). The estimated survival rate at 3 years was 94.3%.

Table 3. Major morbidities

Complications	Total repair surgery (n = 29) (%)	Palliative surgery (n = 17) (%)
Bleeding	5 (17.2)	3 (17.6)
Arrrythmias	8 (27.6)	0
Diaphragmatic plication	1 (3.4)	1 (5.9)
ECMO	2 (6.8)	0
Sepsis	3 (10.2)	2 (11.8)
Surgical site infection	9 (31.0)	3 (17.7)
Early mortality	1 (3.4)	0
Reintervention	4 (13.8)	0
Reoperation	3 (10.3)	0

emergent drainage on POD 7 and positive blood cultures for *Enterococcus faecalis* requiring multiple antibiotic therapies. He recovered and was able to be discharged on POD 20. Six months later, he was re-admitted to the paediatric ICU with severe respiratory distress secondary to pneumonia and sepsis due to *Klebsiella* and *Enterococcus faecalis*. He was placed on ECMO due to hypotension and severe respiratory distress but did not improve and expired on paediatric ICU day 3.

At the final study follow-up in June 2023, with a mean follow-up of 2.92 (0.51–7.9) years, all surviving patients were found to be in good cardiovascular health. The overall survival rate at 3 years utilising our multidisciplinary PA/VSD management protocol was 94.3% (Figure 2).

Morbidities

Nosocomial infection accounted for a large proportion of the morbidities—a third of the cohort.(Table 3). In the total biventricular repair group, two patients required ECMO in their postoperative courses due to low cardiac output but were successfully weaned off ECMO and were discharged home in stable condition. At their last check-up, they were both found to be in good cardiovascular health.

Four patients required balloon dilatation via catheterised reintervention due to peripheral pulmonary branch stenosis. Three patients required conduit replacement due to conduit failure. In these three patients, the second conduit was surgically implanted at a mean of 12 months after total biventricular repair.

Discussions

In the setting of LMICs, suboptimal management of neonates with complex cardiac defects during and after transport to paediatric cardiac centres can result in adverse clinical outcomes.[8] In our two-centre study setting, there was no obstetrics unit at VNCH and no cardiac ICU at HOGH. While HOGH and VNCH are two separate hospitals, they are physically located very close to each other, with a geographical distance of about 500 m (0.31 miles). Both the maternal patient and the neonate could access high-quality medical care from both centres (the obstetric centre and the cardiac centre), almost as if they were on the same campus. Our prenatally identified complex CHD infants' policy was for the obstetrician to utilise the normal standard of obstetric care in monitoring pregnant women at HOGH, in order to keep the baby until 39 weeks GA (if possible) and then preferably a vaginal delivery. C-section was only done when clinically indicated for obstetric reasons. In our study, 35 neonates including 4 preterm babies were safely transferred from HOGH to VNCH. No mortalities occurred prior to either complete biventricular repair or palliative aorto-pulmonary shunt surgery. This result shows that our interhospital programme and multidisciplinary team care approach presents as a good example of referred centres in which the "intra-uterine transfer" could be implemented in case of the prenatal diagnosis of a complex CHD, both in Vietnam and other LMICs where obstetric and paediatric cardiac hospitals are in relatively close proximity.

In developed countries, PA/VSD patients undergo neonatal assessment in order to evaluate the clinical status and the pulmonary circulation's anatomy, leading to excellent outcomes in these patients after the surgery.[9] On the contrary, difficulties such as hospital overcrowding, late presentations due to social or economic conditions, and shortages of well-trained healthcare staff make optimal surgical care of these patients a major challenge in LMICs.[10] By implementing our multidisciplinary team approach programme to CHD at our institutions, we are able to detect CHD earlier in pregnancy and complex CHD such as PA/VSD patients in particular. This approach then results in a prompt and complete assessment of the CHD fetus and initiates planning for which surgical intervention postnatally will be of the greatest benefit for the infant. In our study, 65.7% of our cohort had PA/VSD type A pulmonary morphology which had ductal dependent pulmonary circulation required PGE1 infusion soon after birth. By initiating this inter-hospital multidisciplinary team collaboration, we allowed for the sharing of expertise, resources, and best practices among physicians at the two participating hospitals. This is not the usual educational experience in most LMICs but implementing a programme like ours could lead to a more coordinated and effective approach to the care of PA/VSD and other complex CHD patients in other LMIC settings.

In developed countries, early repair of a PA/VSD is preferred with excellent outcomes reported.[11,12,13] Surgical management options for PA/VSD patients vary depending upon the pulmonary artery anatomy encountered, presence or absence of MAPCAs, and different centres' surgical strategies.[14] While we face the same challenging PA/VSD anatomy as developed countries, we have the additional burden of a challenging socio-economic situation, which is encountered in most other LMICs as well. Still, we

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continuously tried to improve our surgical skills and tailored our approach for PA/VSD in order to achieve an early total repair with the use of a valved conduit. Similar to our previous report,[7] the findings of this study showed good early outcomes with low mortality after total repair of PA/VSD. By learning to overcoming elevated rates of peri-surgical morbidities such as surgical site infections, sepsis, the need for ECMO support, and the potential need of reintervention or reoperation on the right ventricle outflow tract due to the use of artificial conduits and to the complex nature of PA/VSD lesions, we hope our results will give us further insights to guide parent counselling about PA/VSD surgical management in the future. In Vietnam, currently, when a prenatal diagnosis of complex CHD such as PA/VSD is made, the decision of many couples often leads to termination of the pregnancy. It is our belief, now backed up by our PA/VSD outcomes and management experience, that every neonate or child with CHD such as PA/VSD deserves a chance to obtain advanced heart care in our healthcare system. By informing families about the benefits of early diagnosis and treatment during and after the baby's birth, expectant parents can have a more comfortable experience and reliance on the process.

Complex CHD defects such as PA/VSD require a lifetime of close follow-up, as a growing child will eventually outgrow the initial valved conduit, making conduit replacement, whether by surgery or in the catheterisation laboratory, unavoidable. In our study cohort, no patients were lost to follow-up. We do believe that by providing sufficient information, the parents will be more compliant with the management pathway and their child's need for follow-up. Again, the results of our study highlight the importance of a multidisciplinary approach, which includes an accurate CHD prenatal diagnosis, and intensive surgical planning for repairing the morphology of the infant's PA/VSD pulmonary bed, which will then result in optimal postnatal outcomes of PA/VSD patients. Our findings support the need for healthcare authorities and policymakers to prioritise multidisciplinary collaboration between hospitals and paediatric subspecialities when caring for newborns with complex CHD like PA/VSD.

Limitations of the research

Due to the nature of retrospective research and the low number of patients included, as well as the proximal location of the two study hospitals, there is potential bias towards the group of patients in the capital city and nearby areas, where access to medical care is easier. It also raises concerns about the generalisability of our findings to other hospitals in LMICs that take care of CHD infants. Our study also faced limitations in confirming a PA/VSD diagnosis in cases of termination of pregnancy or maternal patients lost to follow-up during the pregnancy. The absence of a comparative group further limits our ability to draw definitive conclusions regarding the effectiveness of the multidisciplinary programme we implemented. Future research into multidisciplinary approaches to complex CHD in LMICs will be beneficial in potentially demonstrating that this approach has a positive impact on overall CHD patient outcomes.

Conclusion

In conclusion, the results of our study demonstrate that inter-hospital collaboration, multidisciplinary team approach coupled with a prenatal diagnosis, and optimal surgical management of PA/VSD in a LMIC like Vietnam can result in optimal patient outcomes. Our findings also provide important insights for healthcare providers, policymakers, and future parents, which can ultimately lead to better care and outcomes for newborns with PA/VSD.

References

1. Tchervenkov CI, Roy N. Congenital heart surgery nomenclature and database project: pulmonary atresia—ventricular septal defect. Ann Thorac Surg 2000; 69: 97–105.
2. Liu Y, Chen S, Zühlke L, et al. Global birth prevalence of congenital heart defects 1970–2017: updated systematic review and meta-analysis of 260 studies. Int J Epidemiol 2019; 48: 455–463.
3. Bonnet D. Impacts of prenatal diagnosis of congenital heart diseases on outcomes. Transl Pediatr 2021; 10: 2241–2249.
4. Liu H, Zhou J, Feng Q-L, et al. Fetal echocardiography for congenital heart disease diagnosis: a meta-analysis, power analysis and missing data analysis. Eur J Prev Cardiol 2015; 22: 1531–1547.
5. Vesel S, Rollings S, Jones A, et al. Prenatally diagnosed pulmonary atresia with ventricular septal defect: echocardiography, genetics, associated anomalies and outcome. Heart 2006; 92: 1501–1505.
6. Lertsakulpiriya K, Vijarnsorn C, Chanthong P, et al. Current era outcomes of pulmonary atresia with ventricular septal defect: a single center cohort in Thailand. Sci Rep 2020; 10: 5165.
7. Vinh TQ, Truong NLT, Hung DQ. 26. Definitive repair of pulmonary atresia with ventricular septal defect using valved conduit for low body weight infant. TCNCYH 2022; 161: 227–237.
8. Karmegaraj B, Kappanayil M, Sudhakar A, et al. Impact of transport on arrival status and outcomes in newborns with heart disease: a low-middle-income country perspective. Cardiol Young 2020; 30: 1001–1008.
9. Holly Bauser-Heaton, Alejandro Borquez, Brian Han, et al. Programmatic approach to management of tetralogy of Fallot with major aortopulmonary collateral arteries. Circ Cardiovasc Interv 2017; 10: e004952.
10. Rao SG. Pediatric cardiac surgery in developing countries. Pediatr Cardiol 2007; 28: 144–148.
11. Soquet J, Barron DJ, d'Udekem Y. A review of the management of pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries. Ann Thorac Surg 2019; 108: 601–612.
12. Derby CD, Pizarro C. Routine primary repair of tetralogy of Fallot in the neonate. Expert Rev Cardiovasc Ther 2005; 3: 857–863.
13. Reddy VM, Petrossian E, McElhinney DB, et al. One-stage complete unifocalization in infants: when should the ventricular septal defect be closed? J Thorac Cardiovasc Surg 1997; 113: 858–866. discussion 866–868.
14. Garg A, Sharma R. Ventricular septal defect with pulmonary atresia: approaches, results, prognosticators and current status. Indian J Thorac Cardiovasc Surg 2022; 38: 28–37.

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