Cardiology in the Young

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The treatment process of a case with dilated cardiomyopathy, severe mitral insufficiency, and coaptation defect in the context of decompensated clinical condition with left ventricular assist device placement

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Brief Report

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Abstract

Heart failure in children is a clinical and pathophysiological syndrome arising from ventricular dysfunction and pressure or volume overload of the circulatory system. Features of paediatric heart failure include feeding problems, poor weight gain, exercise intolerance, or dyspnoea. The aetiology of heart failure in children is complex, with the primary causes being CHD and cardiomyopathies. Cardiomyopathies occur at an incidence of 1.13–1.24 cases per 100,000 children. The prevalence of cardiomyopathy is estimated to be 7.8–8.3 cases per 100,000 infants, particularly common in patients under one year of age presenting with severe heart failure symptoms. Mitral valve insufficiency is a significant source of morbidity in children with dilated cardiomyopathy. Severe mitral insufficiency can lead to a decrease in cardiac output, independent of the left ventricular ejection fraction, exacerbating the clinical course of heart failure in patients with dilated cardiomyopathy. As ventricular systolic function deteriorates, the options for treating mitral insufficiency decrease, leading to a loss of surgical intervention opportunities and making heart transplantation the only viable option. Close monitoring of mitral valve insufficiency in children with dilated cardiomyopathy is essential, as it may lead to decompensated heart failure. In patients who have lost the chance for valve surgery due to decompensation, the application of left ventricular assist device can help improve the decompensatory state and contribute to the reduction of left ventricular diastolic and systolic dimensions, consequently leading to improvements in the dilation of the mitral annulus and severe mitral insufficiency findings. Further studies are needed to determine the optimal timing for surgery in patients who have not missed the chance for valve surgery due to a decrease in ejection fraction.

Introduction

Heart failure in children is a clinical and pathophysiological syndrome associated with ventricular dysfunction and pressure or volume overload in the circulatory system. The characteristics of paediatric heart failure include feeding difficulties, poor weight gain, exercise intolerance, or shortness of breath. The aetiology of heart failure in children is complex. The primary causes are CHD and cardiomyopathies, while less common causes include cardiac arrhythmias, myocarditis, acquired heart diseases such as Kawasaki disease, or heart failure secondary to oncological treatments. In the United States, approximately 12,000 to 35,000 children suffer from heart failure due to CHD or cardiomyopathy, indicating an heart failure incidence of 16.4–48 cases per 100,000 children in the paediatric population.[1] Each year, 14,000 of these patients require hospitalisation, with 65% of cases arising from CHD. According to a study conducted in Belgium, about 50% of hospital admissions due to heart failure exacerbations are attributable to CHD. Fourteen percent of patients with single ventricle physiology require multiple hospital stays due to heart failure exacerbations. Most of these patients will need mechanical circulatory support or heart transplantation in the future.[2] Cardiomyopathies occur at an incidence of 1.13–1.24 cases per 100,000 children. The prevalence of cardiomyopathy in infants under one year of age is estimated to be 7.8–8.3 cases per 100,000, with severe heart failure more commonly observed in this age group.[3,4] Not all children with cardiomyopathy will develop heart failure . The prevalence of heart failure attacks in cardiomyopathic children is estimated to be 0.87 cases per 100,000 children under 16 years old. Seventy-one percent of heart failure attacks occur in patients with dilated cardiomyopathy.[5] Heart transplantation is considered the preferred method for treating end-stage heart failure in paediatric patients.

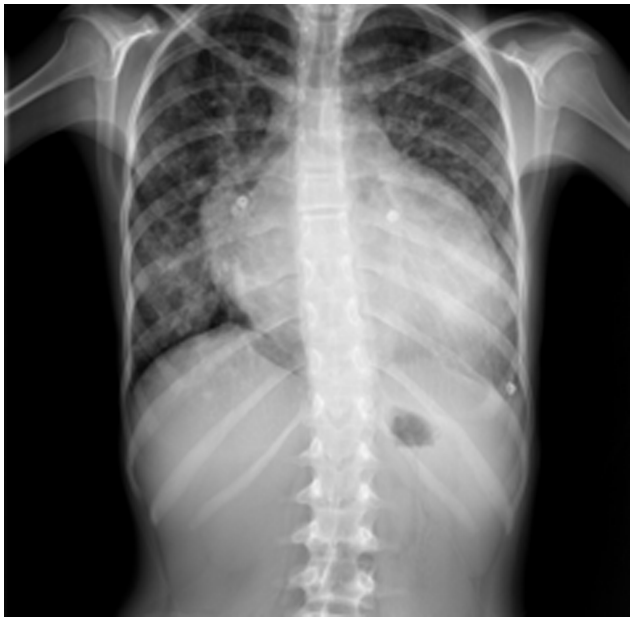

Figure 1. Telecardiogram taken at presentation.

Mitral insufficiency in individuals with left ventricular dysfunction is a significant source of morbidity. The prognosis for adults with severe mitral valve insufficiency and associated left ventricular dysfunction or dilated cardiomyopathy is poor.[6–8] The presence of valvular insufficiency leads to progressive deterioration in left ventricular function and clinical symptoms, ultimately resulting in death if no intervention is performed.[6] Until the mid-1990s, the most common recommendation for adults with severe MI following medical treatment failure was heart transplantation.[9–11] This approach has also been supported in paediatric literature, advocating for listing for organ transplantation after medical treatment has failed.[8,11] However, recent studies in adults have supported mitral valve repair or replacement as an alternative treatment for MI in the presence of left ventricular dysfunction.[12] There are limited data on the use of mitral valve surgery in children with severe MI and left ventricular dysfunction.[13] The standard treatment process for children with dilated cardiomyopathy and severe mitral insufficiency includes medical treatment and heart transplantation when medical treatment fails.[14] Mitral valve repair in children with dilated cardiomyopathy and mitral insufficiency has rarely been reported.[15,16]

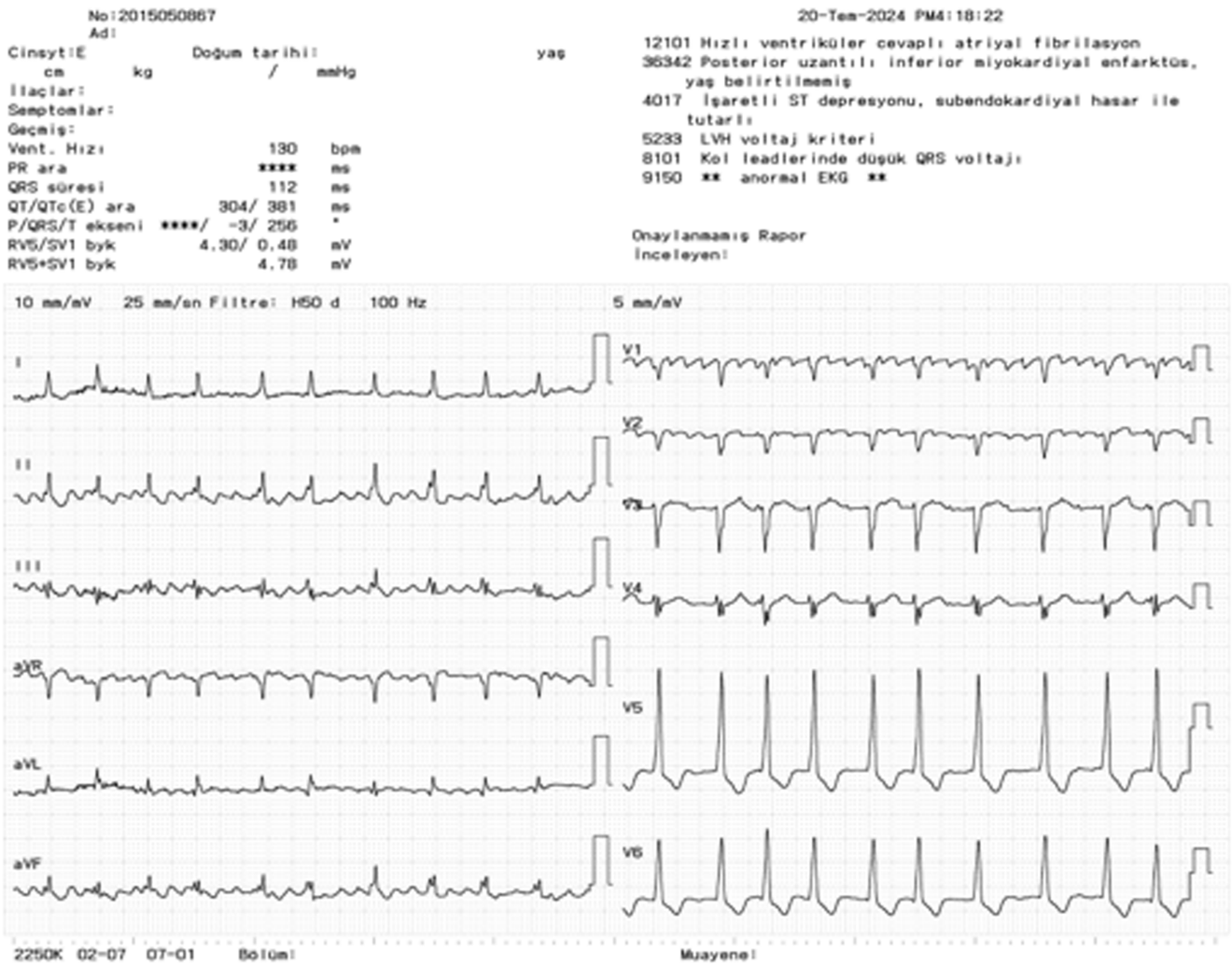

Figure 2. EKG at presentation indicating atrial fibrillation.

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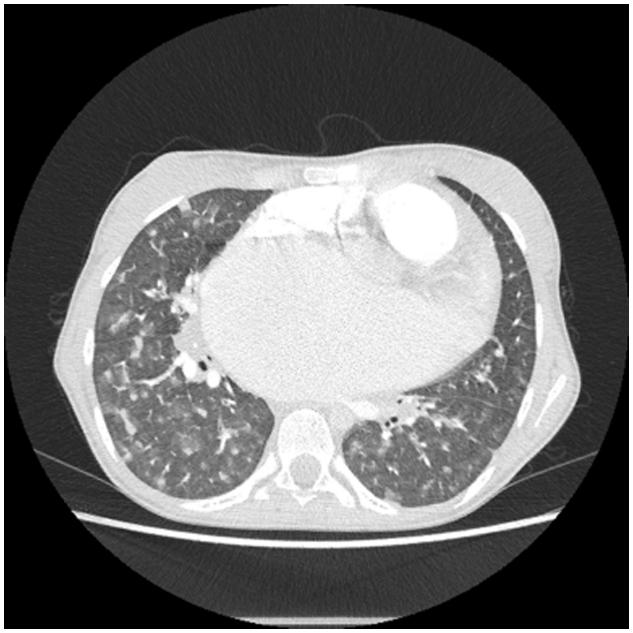

Figure 3. Chest angiography showing bilateral interstitial edema and bilateral lung findings.

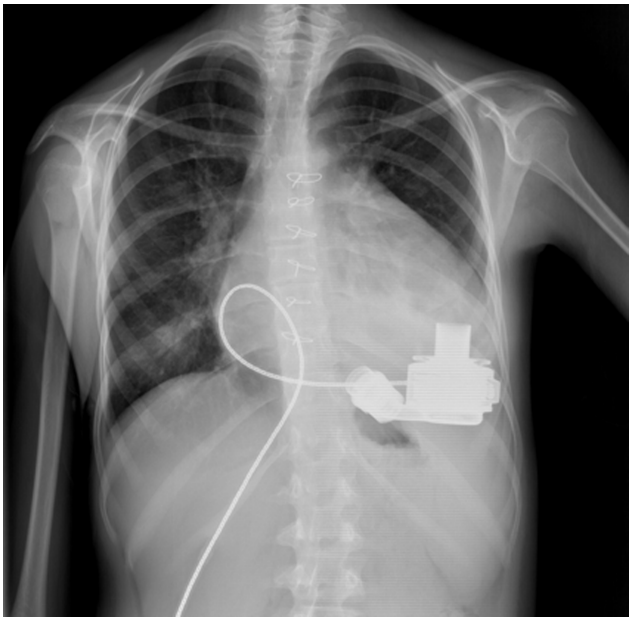

Figure 5. Telecardiogram following LVAD placement showing marked regression in cardiomegaly compared to preoperative status.

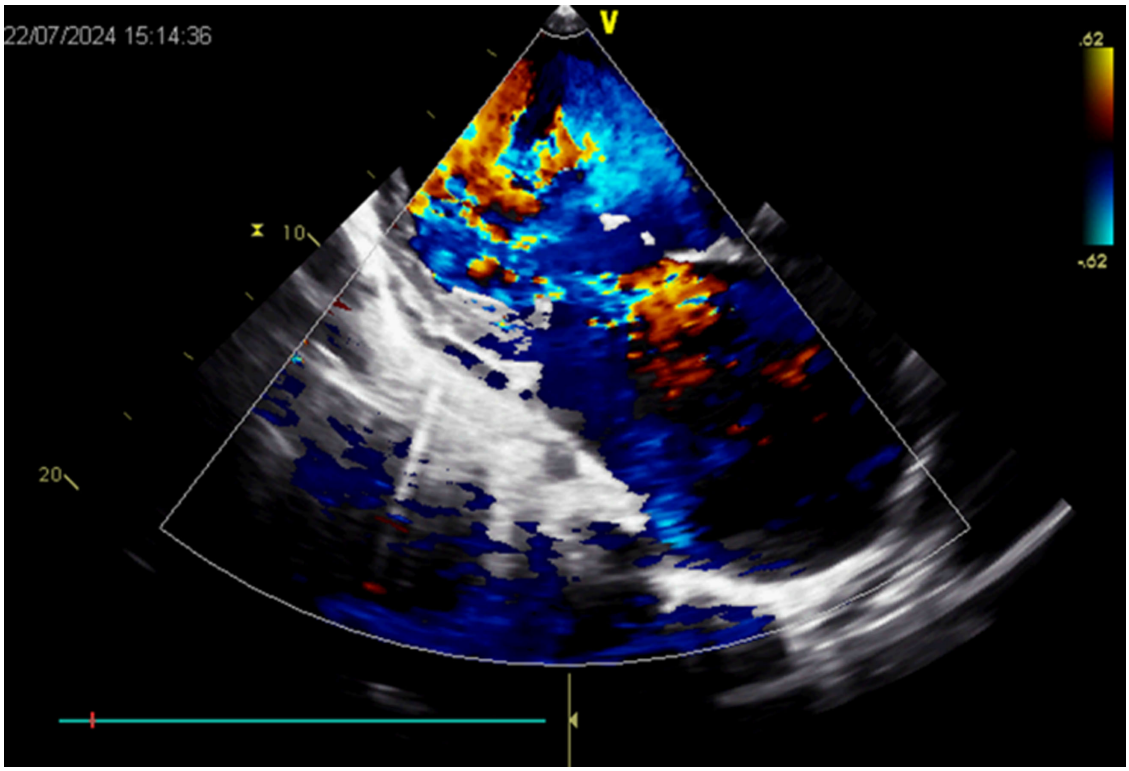

Figure 4. Severe MY before LVAD operation.

Case presentation

A 16-year-old female patient presented at 2.5 months of age with feeding difficulties, sweating, and restlessness and was referred to paediatric cardiology due to the presence of a heart murmur. An echocardiogram conducted at an external centre revealed an ejection fraction of 30%, and the patient was diagnosed with dilated cardiomyopathy.

Regarding the evaluation of dilated cardiomyopathy aetiology, comprehensive metabolic screening and genetic testing were performed at the initial referring centre. The results revealed no

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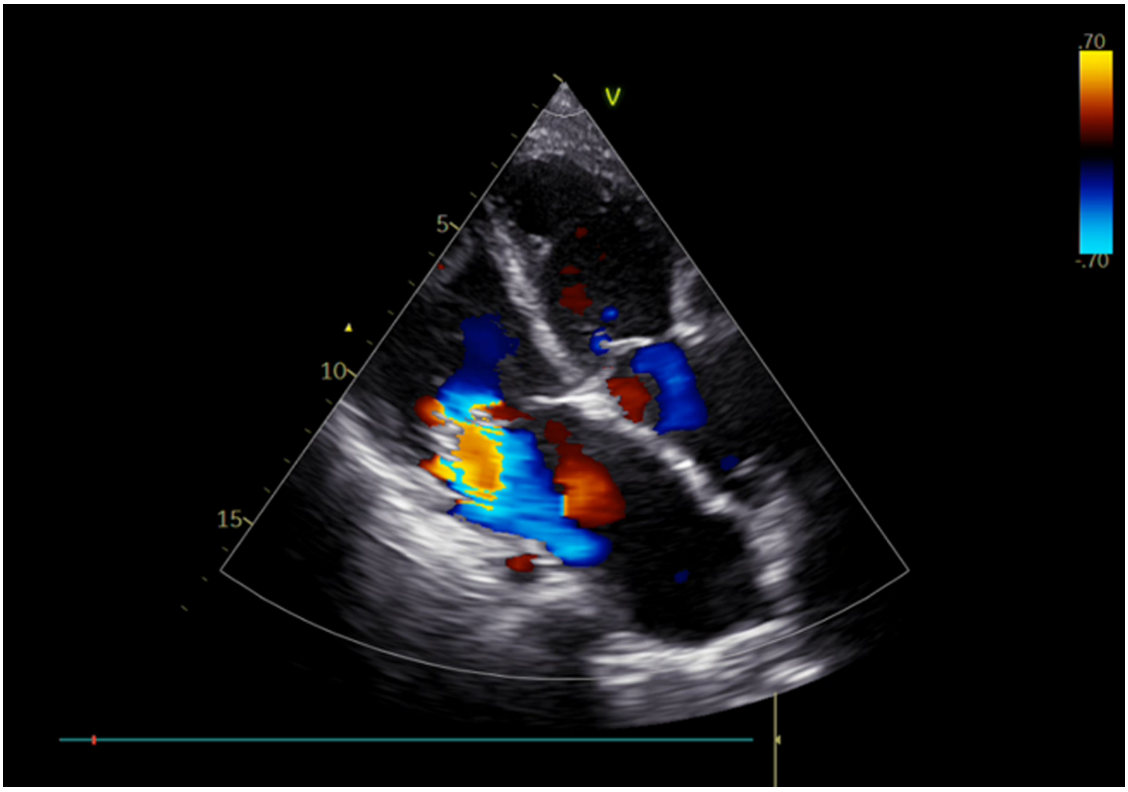

Figure 6. Modarate MY after LVAD operation.

identifiable metabolic disorder or pathogenic genetic variant, suggesting an idiopathic form of the disease.

Treatments with Aldactone, digoxin, enalapril, and carnitine were initiated. She attended regular follow-up visits for six months. At eight months of age, another echocardiogram indicated an ejection fraction of 18%. The family was informed of the necessity for a transplant and the challenges involved in the process. Believing their child would die, the family ceased all treatments and follow-ups except for digoxin. The father began administering herbal remedies (e.g., ***Alchemilla vulgaris***, which was used until the age of 6, and Alpharice, last used in 2020). The patient was seen for follow-ups at the age of 6 and 9, where she had an ejection fraction of 35%. She had not received any follow-up care from a paediatric cardiology clinic for the past seven years, continuing to take digoxin (*215 gtt), Omega 3 (1*1000 mg), and Coenzyme Q (1 × 100 mg). The father reported that in the last year, she had conflicts with the family and had regularly neglected her treatments. The family concealed the child's illness from her. Two months before her admission to the emergency department, the patient had a history of influenza, during which her cough persisted, and her dyspnoea progressively worsened, followed by the development of orthopnea. In the last month, her respiratory rate increased to an intermittent tachypnoeic pattern (30–35 breaths/min) Figure 1. Over the past week, tachycardia (130–140 beats/min) was observed. Upon presenting to the emergency department due to a severe worsening of her symptoms, she was admitted with decompensated heart failure and atrial fibrillation rhythm on Elekcrocardiography (EKG) Figure 2 . While under treatment, she was given Aldactone, Lisinopril, Bisoprolol, Coumadin, Ferrosanol, Digoxin, and Lasix. After three days of hospitalisation at the external centre, she discharged against medical advice and presented to our emergency department of her own accord. Upon examination, her vital signs showed a respiratory rate of 32 breaths/min with tachypnoea, orthopnoea, and dyspnoea Figure 3. She was afebrile with a pulse of 128 beats/min and hypotension (68/45 mmHg, MAP 52). Cardiac monitoring and EKG displayed an atrial flutter/atrial fibrillation rhythm. Laboratory tests showed normal renal function (BUN: 69 mg/dL, Creatinine: 0,72 mg/dL), elevated troponin T (18.3 ng/L), and NT-Pro BNP (11,612 ng/L). Initial echocardiography revealed significant left ventricular dilation ; left ventricular end diastol diameter (**8.5 cm + 6.6 SDS**) consistent with dilated cardiomyopathy, poor mitral valve coaptation, and the presence of severe mitral insufficiency Figure 4. Left atrial dilation was notable and bulging towards the right side. The ejection fraction was 40%. There was mild aortic valve insufficiency and mild tricuspid valve insufficiency. Tricuspid annular plane systolic excursion (TAPSE) measured 23 mm, and the interatrial septum and interventricular septum were intact. No coarctation of the aorta or patent ductus arteriosus was observed, and the inferior vena cava showed spontaneous contrast without collapse.

On the 20th day of hospitalisation in the paediatric intensive care unit (PICU), the patient experienced sudden haemoptysis and desaturation, resulting in severe decompensation. Despite single inotropic therapy, her hypotension necessitated a switch to dual inotropic support (dobutamine + adrenaline), and it was decided to proceed with left ventricular assist device placement. The heart transplant team performed left ventricular assist device surgery. After a two-day observation in the cardiac intensive care unit, the patient was transferred to our paediatric intensive care unit. Left ventricular assist device parameters were set to a speed of 4800 rpm, flow of 3.2 L/min, and power of 3.8 W, with satisfactory

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hemodynamic stability. Although the echocardiogram showed an effective ejection fraction of 40%, clinical assessment indicated a lower functional ejection fraction due to the patient's NYHA classification of 4 and modified Paediatric Acute Congestive Heart Failure score of 2. Given the significant dilation of left ventricular end diastole and left atrial dimensions, it was anticipated that with left ventricular assist device placement, the left ventricular end diastole diameter would reduce, resulting in narrowing of the mitral annulus and improvement of mitral insufficiency findings. It was concluded that the patient's chronic atrial fibrillation and haemoptysis were associated with the significantly dilated left atrium. Postoperative echocardiography (August 2024) revealed an left ventricular assist device diameter of 6.1 cm (**3.3 SDS)** and an ejection fraction of 40% Figure 5. The IVS remained intact, and there was moderate coaptation defect and mitral insufficiency observed on the mitral valve Figure 6. Notable reduction in left atrial dilation was recorded (approximately-2.5 cm). Moderate tricuspid insufficiency and right ventricular systolic pressure were at 30 mmHg. TAPSE was 8 mm; the inferior vena cava was 16 mm and was collapsing. No patent ductus arteriosus or coarctation of the aorta was identified, and no pericardial effusion was observed. The patient's tachypnoea, orthopnea, and dyspnoea complaints completely resolved. The chronic atrial fibrillation persisted, but the patient received anticoagulation therapy for three weeks during the paediatric ICU course. Though attempts at cardioversion were made during left ventricular assist device placement, sinus rhythm was not restored. However, the use of beta-blockers achieved ventricular rate control, resolving the tachycardia and eliminating the sensation of palpitations. The hyperdynamic precordium returned to normal, and the patient with previously low exercise capacity (NYHA 4, Paediatric Acute Congestive Heart Failure Score 2) became fully mobilised post-left ventricular assist device and reported no complaints during daily exertion.

Discussion

There is limited knowledge regarding the effectiveness of mitral valve surgery in children with dilated cardiomyopathy. Furthermore, a clear guideline for determining the timing of mitral valve surgery or heart transplantation in paediatric patients with MI and left ventricular dysfunction is lacking. Literature indicates that the presence of mitral insufficiency adversely affects survival rates in adults with dilated cardiomyopathy.[8] Historically, heart transplantation has been considered the definitive treatment for end-stage heart failure associated with significant mitral insufficiency.[18] Concerns regarding perioperative mortality (reported as 2.1 to 11%) exist in adults with significant ventricular dysfunction requiring mitral valve surgery.[19] However, one-year survival rates are comparable to those of heart transplantation.[20] Consequently, surgical correction of mitral insufficiency in the presence of ventricular dysfunction has increased in the past decade.[21,22] Breinholt JP et al. conducted a significant paediatric series that assessed the effectiveness of mitral valve surgery in children with dilated cardiomyopathy and severe mitral insufficiency, reporting some positive outcomes, including clinical improvement and postponement of the transplantation process, despite a lack of improvement in ejection fraction.[17] Traditionally, in paediatric populations, similar to adults, the common practice has been to transition to heart transplantation after medical treatment failure. Recently, however, there have been reports suggesting that mitral valve surgery may serve as a bridge to transplantation in symptomatic mitral insufficiency cases.[15,16,23,24] In a report by Hsu et al., two children with dilated cardiomyopathy underwent emergency mitral valve surgery, both achieving clinical responses, though they experienced recurrent heart failure within six months post-operation and subsequently needed heart transplantation.[15,16]

Mitral insufficiency is a significant morbidity source in children with dilated cardiomyopathy. Severe mitral insufficiency can lead to reduced cardiac output and exacerbation of heart failure, as seen in our case, independent of the measured ejection fraction.[17] With the deterioration of ventricular systolic function, treatment options for mitral insufficiency decline, and patients may lose the opportunity for surgical intervention, making heart transplantation the sole viable option.[18]

Given that mitral valve insufficiency in children with dilated cardiomyopathy can lead to decompensated heart failure, close monitoring is essential. The application of left ventricular assist device in patients who have lost surgical opportunity significantly improves the decompensatory clinical state and aids in the reduction of left ventricular dimensions, contributing to the narrowing of the mitral annulus, thus potentially improving severe mitral insufficiency findings. In countries like ours, where paediatric organ donors are limited, left ventricular assist device should be considered as a valuable option for decompensated dilated cardiomyopathy patients due to severe mitral insufficiency who require bridging to transplantation. Hsu et al. (2012) demonstrated that the application of left ventricular assist device as a bridge to heart transplantation in paediatric patients is both effective and safe. Their study showed that left ventricular assist device significantly improved clinical outcomes and provided stability for patients while awaiting a suitable donor heart.[25] Additionally, recent studies by Doğan et al. support these findings. Their 2024 study from Turkey highlights the positive effects of the HeartMate 3 left ventricular assist device in paediatric patients with end-stage heart failure. The study demonstrates the effectiveness of left ventricular assist device in improving survival rates and overall clinical condition in paediatric patients awaiting heart transplantation.[25] These studies further strengthen the growing body of evidence supporting left ventricular assist device as a critical and reliable therapy for this vulnerable patient group. More studies are necessary to determine the optimal timing for surgery in paediatric patients who have not missed opportunities for valve surgery due to decreased ejection fraction.

References

1. Hsu DT, Pearson GD. Heart failure in children: part I: history, etiology, and pathophysiology. Circ Heart Fail 2009; 2: 63–70.
2. Nandi D, Rossano JW. Epidemiology and cost of heart failure in children. Cardiol Young 2015; 25: 1460–1468.
3. Nugent AW, Daubeney PEF, Chondros P, et al. The epidemiology of childhood cardiomyopathy in Australia. N Engl J Med 2003; 348: 1639–1646.
4. Lipshultz SE, Sleeper LA, Towbin JA, et al. The incidence of pediatric cardiomyopathy in two regions of the United States. N Engl J Med 2003; 348 : 1647–1655.
5. Wilkinson JD, Landy DC, Colan SD, et al. The pediatric cardiomyopathy registry and heart failure: key results from the first 15 years. Heart Fail Clin 2010; 6: 401–413.
6. Blondheim DS, Jacobs LE, Kotler MN, Costacurta GA, Parry WR. Dilated cardiomyopathy with mitral regurgitation: decreased survival despite a low frequency of left ventricular thrombus. Am Heart J 1991; 122: 763–771.

https://doi.org/10.1017/S1047951125100607 Published online by Cambridge University Press

7. Carabello BA. The pathophysiology of mitral regurgita- tion. J Heart Valve Dis 2000; 9: 600–608.
8. Junker A, Thayssen P, Nielsen B, Andersen PE. The he- modynamic and prognostic significance of echo-doppler-proven mitral regurgitation in patients with dilated cardiomyopathy. Cardiology 1993; 83: 14–20.
9. Anguita M, Arizon JM, Valles F, et al. Influence of heart transplantation on the natural history of patients with severe congestive heart failure. J Heart Lung Transplant 1993; 12: 974–982.
10. Geha AS, El-Zein C, Massad MG. Mitral valve surgery in patients with ischemic and nonischemic dilated cardiomyopathy. Cardiology 2004; 101: 15–20.
11. Rickenbacher PR, Haywood G, Fowler MB. Selecting candidates for cardiac transplantation. How to assess exclusion criteria and predict who will benefit. J Crit Illn 1995; 10: 199–206.
12. Bolling SF, Pagani FD, Deeb GM, Bach DS. Intermediate- term outcome of mitral reconstruction in cardiomyopathy. J Thorac Cardiovasc Surg 1998; 115: 381–386.
13. Krishnan US, Gersony WM, Berman-Rosenzweig E, Apfel HD. Late left ventricular function after surgery for children with chronic symptomatic mitral regurgitation. Circulation 1997; 96: 4280–4285.
14. Tsirka AE, Trinkaus K, Chen SC, et al. Improved outcomes of pediatric dilated cardiomyopathy with utilization of heart transplantation. J Am Coll Cardiol 2004; 44: 391–397.
15. Hsu RB, Chen RJ, Wu MH, Wang JK, Wang SS, Chu SH. Non-transplant cardiac surgery for end-stage dilated cardiomyopathy in small children. J Heart Lung Transplant 2003; 22: 94–97.
16. Hsu RB, Chien CY, Wang SS, Chu SH. Nontransplant cardiac surgery as a bridge to heart transplantation in pediatric dilated cardiomyopathy. Tex Heart Inst J 2002; 29: 213–215.
17. Breinholt JP, Fraser CD, Dreyer WJ, et al. The efficacy of mitral valve surgery in children with dilated cardiomyopathy and severe mitral regurgitation. Pediatr Cardiol 2008; 29: 13–18.
18. Wang SS, Chu SH, Ko WJ. Clinical outcome of heart transplantation: experience at the National Taiwan University Hospital. Transplant Proc 1996; 28: 1733–1734.
19. Boucek MM, Edwards LB, Keck BM, Trulock EP, Taylor DO, Hertz MI. Registry of the international society for heart and lung transplantation: eighth official pediatric report—2005. J Heart Lung Transplant 2005; 24: 968–982.
20. Luciani GB, Montalbano G, Casali G, Mazzucco A. Pre- dicting long-term functional results after myocardial revascularization in ischemic cardio-myopathy. J Thorac Cardio- vasc Surg 2000; 120: 478–489.
21. Badhwar V, Bolling SF. Mitral valve surgery in the patient with left ventricular dysfunction. Semin Thorac Cardiovasc Surg 2002; 14: 133–136.
22. Badhwar V, Bolling SF. Mitral valve surgery: when is it appropriate? Congest Heart Fail 2002; 8: 210–213.
23. Wang SS, Hsu RB, Chen YS, et al. Heart transplantation and mitral valve repair in pediatric patients with refractory heart failure. Transplant Proc 2003; 35 : 463–465.
24. Hsu KH, Huang SC, Chou NH, et al. Ventricular assist device application as a bridge to pediatric heart transplantation: a single center's experience. Transplant Proc 2012; 44: 883–885.
25. Doğan E, Ulger Tutar Z, Tuncer ON, et al. Outcomes of HeartMate 3 in pediatric patients with end-stage heart failure: a single-center preliminary experience from Turkey. Front Cardiovasc Med 2024; 11: 1472663.

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