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Mitral valve replacement in dilated cardiomyopathy: A valid option?

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Abstract

Background: Dilated cardiomyopathy (DCM) is characterized by left ventricular dilation and dysfunction. The association with significant mitral regurgitation worsens the prognosis.

Case report: A 2-year-old girl presented with DCM and severe mitral regurgitation. She had a history of viral myocarditis at the age of 4 months, necessitating recurrent hospital admissions for management of intractable heart failure, pneumonia, and failure to thrive. The decision was taken to proceed for mitral valve surgery, which ended with mitral valve replacement. Over 3 years of follow-up after surgery, there was significant improvement in her weight gain and she improved clinically. There were still recurrent admissions, but mostly for adjustment of her deranged anticoagulation medications.

Conclusion: Mitral valve surgery might be indicated in selected patients with DCM.

Keywords: Children, Dilated cardiomyopathy, Mitral regurgitation, Mitral valve surgery

1. Introduction

Dilated cardiomyopathy (DCM) is characterized by left ventricular dilation and dysfunction. The association with significant mitral regurgitation (MR) worsens the prognosis [1,2].

In case cardiac function does not respond to anti-failure treatments, heart transplant remains the ultimate treatment for these patients [3].

As heart transplant is not easily available for children, some authors introduced pulmonary artery banding [4], but this management did not get enough recognition. Other authors suggested, especially in patients with associated severe mitral valve regurgitation, mitral surgery as a bridge to cardiac transplant [2,3,5].

We present a 2-year-old girl with DCM and severe MR who improved clinically after mitral valve replacement.

2. Case report

A 2-year-old girl referred to our hospital with DCM and severe MR for further management. She had a history of viral myocarditis at the age of 4 months that was complicated by DCM. Since then, she had recurrent admissions with chest infection.

On first presentation, her weight was 9 kg, below the 3rd percentile for age, and had a full blown picture of congestive heart failure. Her heart rate was 130 beats/minute (90th centile), gallop rhythm, blood pressure 86/48 mmHg, respiratory rate 35–40 breaths/minute (90th centile), hepatomegaly, bilateral basal fine crepitations, and brain natriuretic peptide (BNP) elevated with 2000 pg/mL.

She was on high doses of anti-failure medications (Furosemide 3.3 mg/kg/day, ACE Inhibitor 1.5 mg/kg/day, spironolactone 3 mg/kg/day, and digoxin 10 mcg/kg/day).

Electrocardiogram showed left ventricular hypertrophy. Chest X-ray on admission revealed

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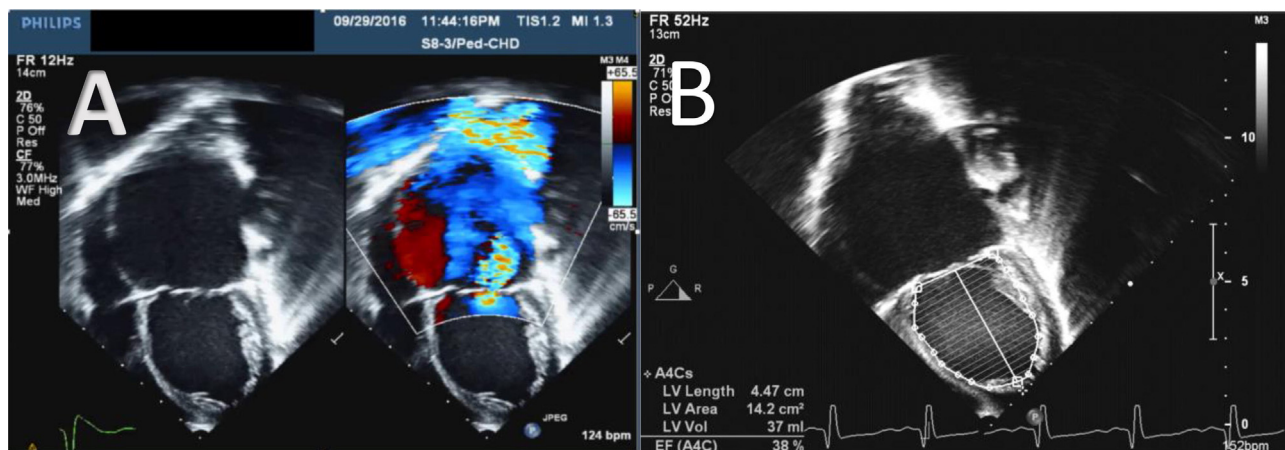


Fig. 1. (A) Apical four-chamber view showing dilated left atrium and left ventricle with severe mitral regurgitation; (B) depressed left ventricular function with ejection fraction of 38%. EF = ejection fraction; LV = left ventricle.

cardiomegaly and lung congestion. Echocardiography revealed dilated left atrium, left ventricle with severe MR, and depressed left ventricular function; ejection fraction (EF) was 38% (Fig. 1).

In the following 2 months, she was admitted three times with intractable heart failure; in the last admission, the patient required admission to the intensive care unit for inotropic support. After group discussion and reviewing the literature [5–7], mitral valve surgery was decided. At surgery and after two failed trials of mitral valve repair (Alfieri procedure), mitral valve replacement was performed with 23-mm St Jude Medical mechanical valve (Minneapolis, Minn, USA). The patient had a smooth postoperative course with mild bleeding, which was managed by blood transfusion, and transient supraventricular tachycardia, which was managed by cardioversion and intravenous magnesium sulfate. Anticoagulation was started in the form of Coumadin. The same doses of anti-failure medications were resumed.

During 3 years of follow-up, she was admitted 12 times, mostly for adjustment of anticoagulation; eight times for international normalized ratio (INR) adjustments, twice because of gastroenteritis, and twice for treatment of pneumonia.

However, from a cardiac standpoint, she was clinically stable. There was significant improvement in her weight, heart rate, and respiratory rate. Her weight increased from 9 kg (3rd centile) to 15 kg (50th centile). Her respiratory rate decreased from 50 breaths/minute (99th centile) to 30 breaths/minute (75th centile). Her heart rate decreased from 150 beats/minute (95th centile) to 110 beats/minute (75th centile). Chest X ray also showed decrease in cardiothoracic ratio compared with the initial condition (Fig. 2). On presentation, the Z score of left ventricular end-diastolic diameter (LVEDD) was 5, left ventricular end-systolic diameter (LVESD) was 5, and EF was 38%.

One month later, the Z score of LVEDD was 3.6, LVESD was 5.7, and EF was 42%. Six months later,

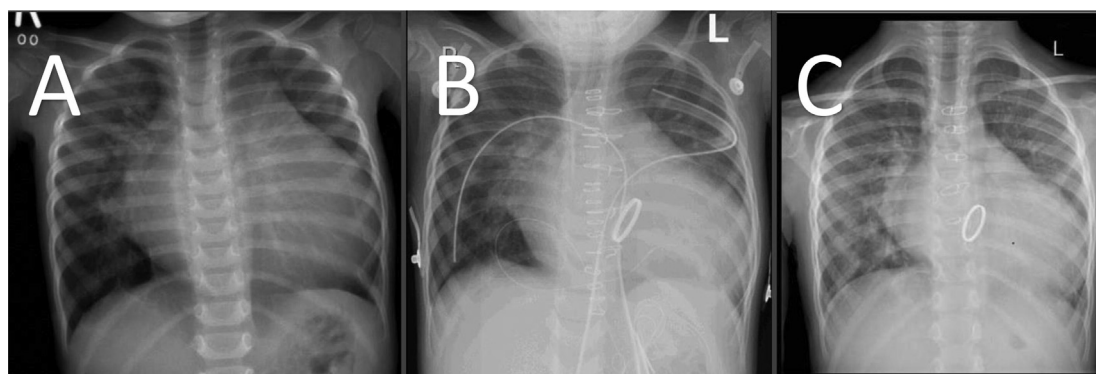


Fig. 2. Serial improvement in cardiothoracic ratio in chest X-ray (A) on presentation, (B) 6 months after surgery, and (C) last follow-up 3 years after surgery.

the Z score of LVEDD was 3.6, LVESD was 5.9, and EF was 25%. At the last follow-up, 3 years after surgery, the Z score of LVEDD was 4.1, LVESD was 6.3, and EF was 28%. This LVEDD decreased, however EF did not improve.

The girl is following-up in the clinic and is playful, asymptomatic, and her parents are having a more stable life. At this stage, there is no need for further invasive management (heart transplant).

3. Discussion

The case we presented shows that mitral valve surgery is a feasible and a beneficial option. Until now, it not only improved quality of life but also decreased hospitalization frequency due to heart failure. Unfortunately, adjustment of INR, as typical for this age group, remained a challenge.

In the literature, there are not many studies discussing mitral valve surgery in DCM in childhood [5–7]. Summary of the few studies available showed that mitral valve surgery in DCM associated with at least moderate MR and EF not less than 25% is beneficial in postponing the need for cardiac transplant and decreasing hospitalization frequency, with clinical improvement in most patients. This is at the expense of no improvement in cardiac function as well as complications of anticoagulation [5], especially in the young age group.

In one case, a 6-year-old patient with this entity survived over 10 years without the need for any other management or intervention [3]. Our patient was fitting in the suggested criteria proposed by Sugiyama et al [5], namely (1) more than moderate MR with a severely dilated left atrium on echocardiography; (2) fraction shortening >10% or EF >25% on echocardiography; (3) BNP >1000 pg/mL; and (4) previous mitral repair (Alfieri procedure) or coexisting mitral stenosis.

4. Conclusion

Mitral valve surgery improves quality of life in selected children with DCM associated with at least moderate MR. Adjustment of INR remains a challenge especially in the young age group. This management should be contemplated especially in areas where cardiac transplant is not readily available.

Further studies are needed to determine which patient might benefit from this management and the proper time for this intervention.

Conflict of interest

The authors have no conflict of interest.

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