

Mitral valve replacement for inflow obstruction of left ventricular assist device in a child with restrictive cardiomyopathy

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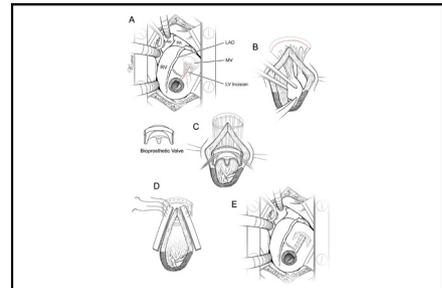
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Restrictive cardiomyopathy (RCM) poses a significant challenge in ventricular assist device (VAD) support. An associated stiff and small left ventricular (LV) cavity can result in obstruction to VAD inflow.¹

CLINICAL SUMMARY

A 2-year-old male patient weighing 10.9 kg was admitted with a new diagnosis of RCM. The echocardiogram showed severely dilated atria and venae cavae (Figure 1, A). Cardiogenic shock developed, requiring emergency peripheral cannulation for extracorporeal membrane oxygenation. Development of pulmonary edema prompted conversion to central cannulation with the addition of a left atrial (LA) vent. After approval for a heart transplant, the patient was bridged to a Berlin Heart EXCOR LVAD (25-mL pump; Berlin Heart, The Woodland, Tex). Intraoperatively, the small LV barely accommodated a 9-mm LV inflow cannula. Cardiopulmonary bypass was

weaned with epinephrine and inhaled nitric oxide. Intraoperative transesophageal echocardiography showed a small LV cavity, with hypertrophied papillary muscles encroaching on the inflow cannula (Figure 1, B). The sternum was closed with preserved pump filling. Overnight, severe inflow cannula obstruction developed, and he was taken back to the operating room. After establishment of



Approach to mitral valve replacement to enlarge LV cavity with LVAD placement. Printed with permission of Texas Children's Hospital.

Central Message

A novel surgical approach to left ventricular assist device management was used in a child with restrictive cardiomyopathy.

See Editorial Commentary page e15.

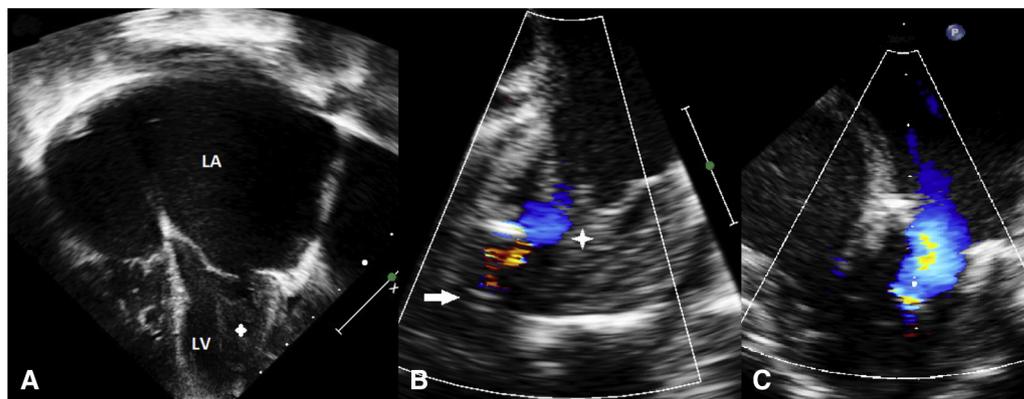


FIGURE 1. A, Transthoracic echocardiogram in the apical 4-chamber view showing a severely dilated left atrium, diminished left ventricular (LV) cavity size, and hypertrophied papillary muscles (denoted by *asterisk*). B, Intraoperative transesophageal echocardiogram with left ventricular assist device placement. Cannula (denoted by *arrow*) is seen in the left ventricular cavity. Mitral valve and papillary muscle (denoted by *asterisk*) are encroaching on the inflow cannula in diastole. C, Intraoperative transesophageal echocardiogram after papillary muscle resection and prosthetic mitral valve replacement demonstrating laminar flow through the mitral valve into the left ventricular inflow cannula. LA, Left atrium.

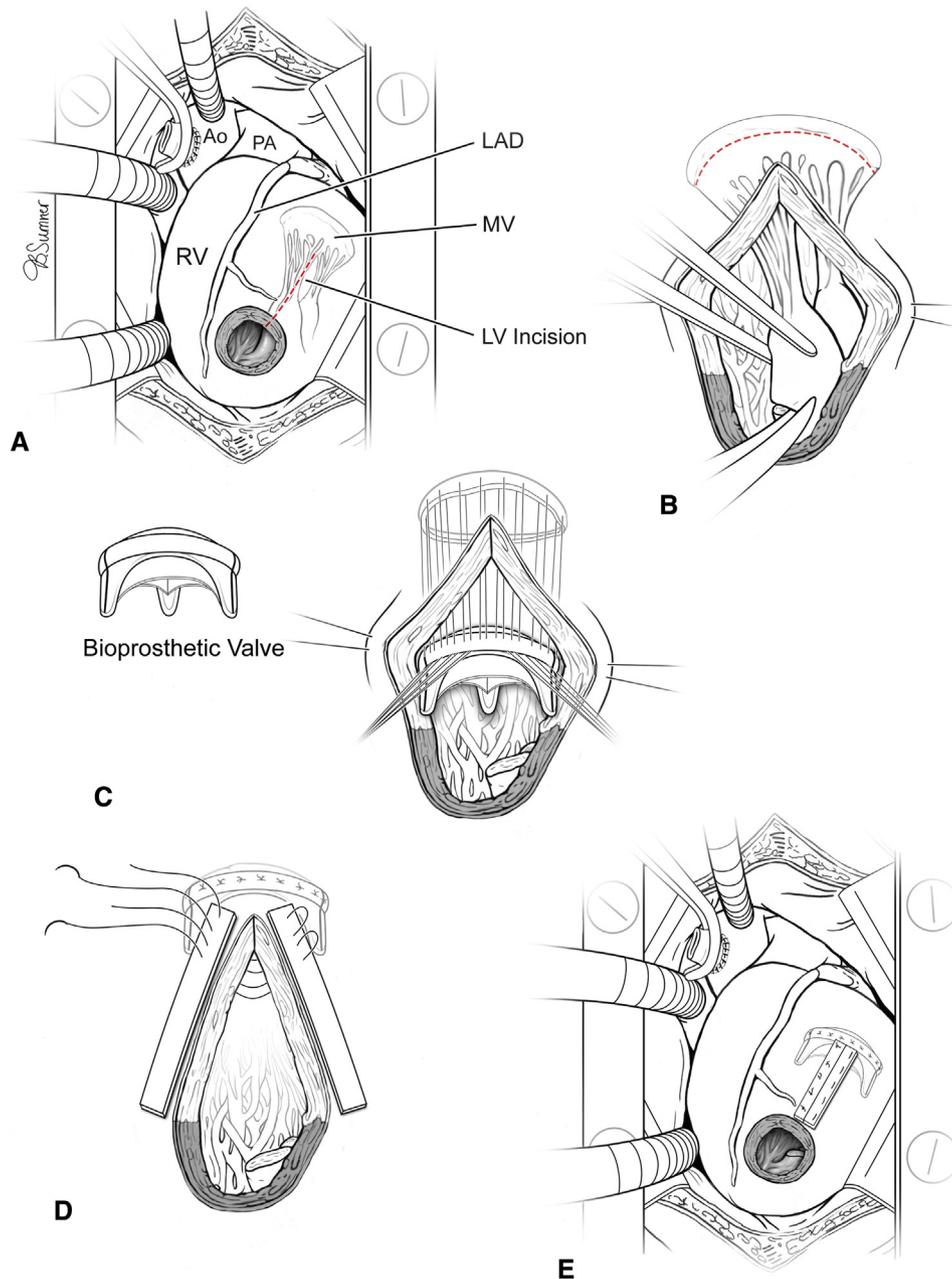


FIGURE 2. A, After establishment of cardiopulmonary bypass, the Berlin inflow and outflow cannulas are detached. The left ventricular (LV) incision is indicated with a red dotted line. B and C, The mitral valve (MV) and its apparatus are excised, and a bioprosthesis valve is placed. D and E, The ventriculotomy is reapproximated with interrupted horizontal mattress sutures over felt strips, leaving an opening for reinsertion of the Berlin inflow cannula. Ao, Aorta; PA, pulmonary artery; LAD, left anterior descending coronary artery; RV, right ventricle. Printed with permission of Texas Children’s Hospital.

cardiopulmonary bypass, cardioplegia was given to arrest the heart, and the LV inflow cannula was detached from the LV apex. The resultant apical opening was extended toward the mitral valve parallel to the left anterior descending coronary artery (Figure 2). Through the created ventriculotomy, the hypertrophied papillary muscles and

mitral valve apparatus were excised. A 19-mm porcine bioprosthesis valve (Epic; St Jude Medical, St Paul, Minn) was implanted with interrupted horizontal mattress pledgeted stitches. The ventriculotomy was reapproximated with interrupted horizontal mattress sutures over felt strips, leaving an opening for inflow cannula insertion

at the apex. Left VAD support was reinstated, with much improved filling of the pump. Echocardiography showed laminar inflow into the LV apical cannula with good function of the prosthetic mitral valve (Figure 1, C). Two weeks later, a large pericardial effusion developed, manifesting as poor left VAD filling. We then placed a right VAD to exclude right heart failure as a potential factor contributing to compromised filling. The patient was successfully bridged to orthotopic heart transplant after 5 months of biventricular assist device support without any thromboembolic or bleeding events. Anticoagulation was according to our standard protocol for the Berlin EXCOR. Inspection of the explanted heart revealed a well-positioned mitral bioprosthetic valve without any thrombus formation.

DISCUSSION

Impediments to adequate LV filling in patients with a left VAD include a small LV cavity, obstruction to mitral valve inflow, and transient or persistent pulmonary hypertension. Children with RCM often have a small LV cavity size and pulmonary hypertension that can limit LV filling, resulting in VAD inflow cannula obstruction.¹ In this patient, hypertrophied papillary muscles compounded the problem of a small LV cavity, resulting in an obstructive mitral valve apparatus and cannula obstruction with “suck-down” events.

To our knowledge, replacement of the mitral valve to address left VAD inflow obstruction has not been previously described. Mitral valvectomy alone may be considered sufficient to enlarge the LV cavity. We believe, however, that competency of the mitral valve is critical to avoid pulmonary edema, particularly with a pulsatile VAD, where the LV is not decompressed when the VAD is in systole (typically 40% of each pump cardiac cycle). Although the presence of a prosthetic mitral valve could

potentially increase the risk of thromboembolic complications, data from a high-volume adult center suggest this was only a theoretic concern.² Our patient did not have any hematologic complications with a standard anticoagulation regimen.

We recognize that this was an aggressive approach relative to an alternative option of LA cannulation. Although VAD support with LA cannulation was associated with a worse outcome, primarily as a result of thrombotic complications, in the previous adult experience,³ this may not be the case in children with RCM. According to the Berlin Heart registry as of 2011, there have been 30 EXCOR implantations worldwide in children with RCM, 40% (n = 12) with LA cannulation and the remaining 60% with LV cannulation. Mortality in the LV cannulation group was high at 39% (7 of 18). In contrast, there was a lower mortality in the LA cannulation group at 17% (2 of 12). This finding undoubtedly highlights the difficulty of VAD support in this group of patients. On the basis of this limited data set, even though it is orally reported to Berlin Heart’s clinical affairs team and not audited, it may be reasonable to state that LA cannulation may be a safer approach in children with RCM relative to standard LV cannulation. With novel modifications of the LV cannulation technique, however, such as our method, the outcome of LV cannulation in RCM may become more favorable.

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