New 15-mL ventricular assist device in children with restrictive physiology of the left ventricle

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The Berlin Heart Pediatric EXCOR (Berlin Heart GmbH, Berlin, Germany) ventricular assist device (VAD) has been successfully used as a bridge to orthotopic heart transplant (OHT) in infants and children.¹⁻³ The 10- and 25-mL volume pumps have improved outcomes in children weighing less than 10 kg,^{2,3} although their risk of mortality remains higher relative to larger children.³ The risk of VAD-related complications may also be higher when the chamber volume is not appropriately matched to the patient size.⁴

Restrictive left ventricular (LV) physiology presents a unique challenge for left VAD (LVAD) implantation in children because of potential problems with drainage with conventional LV apical cannulation. We describe the use of a novel 15-mL volume Berlin Heart Pediatric EXCOR VAD in 2 children with restrictive LV.

CLINICAL SUMMARIES

Patient 1 was a female patient, diagnosed with LV non-compaction cardiomyopathy with restrictive features in infancy. By 2 years, she had significant left atrial (LA) and LV dilatation (z scores of +9.78 and +5.36, respectively) with severe mitral regurgitation (Figure 1). Cardiac catheterization confirmed a mean LA pressure of 44 mm Hg and elevated pulmonary vascular resistance index of 9 Wood units/m², decreasing to 3.2 Wood units/m² with inhaled nitric oxide and oxygen. At 33 months of age, after lack of response to pharmacologic therapy on intensive care, she was placed on extracorporeal membrane oxygenation support. After 6 days on extracorporeal membrane oxygenation support, at a weight of 9.6 kg and body surface area of 0.48 m², the patient underwent LVAD implantation as bridge to OHT. A 15-mL Pediatric EXCOR pump was implanted with LA-to-aorta cannulation because of the significant LA enlargement. The initial pump rate was

80 to 100 beats/min, providing a cardiac index (CI) between 2.5 and 3.1 L/min/m². She was successfully extubated on day 23 after LVAD implantation and remained free of complications. After 3 months on support, the patient's weight had increased to 13 kg (pump rate of 120 beats/min for a CI of 3.0 L/min/m²), and she started having sweaty episodes on exertion. On the basis of echocardiographic evidence of elevated LV end-diastolic volume, the pump was upsized to 25 mL (pump rate of 80 beats/min for a CI of 3.3 L/min/m²). After a total of 131 days of LVAD support, the patient underwent uncomplicated OHT. She was successfully extubated on day 3 after transplant and discharged from intensive care on day 6.

Patient 2 was a male patient with a diagnosis of restrictive cardiomyopathy with biventricular involvement at the age of 11 months (weight of 10 kg and body surface area of 0.43 m²). His condition worsened during the course of several weeks, necessitating admission to intensive care. Echocardiography demonstrated biatrial enlargement (z score of +4.29) with high LV end-diastolic pressure (17 mm Hg) on cardiac catheterization. The patient was listed for OHT, and a 15-mL LVAD and a 15-mL right VAD were inserted after 12 days of intensive care. LVAD drainage was through the LA. His chest was closed on postoperative day 6, and he was successfully extubated on day 13. The patient has been on VAD support for 5 months, with pump rates ranging from 85 to 115 beats/min and CIs ranging from 2.6 to 3.5 L/min/m², and is currently awaiting a donor organ.

DISCUSSION

Worldwide, more than half of the children supported with the EXCOR VAD weigh less than 10 kg.¹⁻³ The waiting time for OHT may be longer³ and the risk of mortality higher² for these smaller children. Children at the upper end of this weight range need an estimated 1000 to 1500 mL/min of cardiac output to decompress the LV, meet metabolic demands, and thrive. Until recently, they have been supported either with a 10-mL VAD, resulting in small stroke volumes at higher pump rates, or with a 25-mL VAD, delivering larger stroke volumes at lower pump rates but often accompanied by lower diastolic pressure. The consequences of such physiologic anomalies are unclear; however, mismatch between chamber volume and body size may contribute to VAD-related complications.⁴

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FIGURE 1. Echocardiographic studies of patient 1 with left ventricular (LV) noncompaction cardiomyopathy before left ventricular assist device implantation. A, Apical 4-chamber demonstrating severe left atrial (LA) dilation and pulmonary vein (PV) congestion. B, Mitral inflow Doppler trace with short deceleration time. RA, Right atrium; RV, right ventricle; MV, mitral valve; E Vel, peak E wave velocity; DecT, deceleration time; A Vel, peak A wave velocity; E/A ratio, ratio of peak E wave velocity to peak A wave velocity.

We describe the successful use of a new 15-mL EXCOR pump in 2 children whose requirements fell between the ideal fill and flow characteristics of the existing pumps. In children with a body surface area ranging from 0.33 to 0.55 m² and weight ranging from 7 to 12 kg, the 15-mL chamber complements the EXCOR infant range. It can achieve a CI of 2.7 to 3.3 L/min/m² at pump rates of 60 to 120 beats/min.

Restrictive and noncompaction cardiomyopathies are rare but contribute disproportionately to mortality in children with cardiomyopathy.^{2,3,5} Conventional VAD cannulation of the noncompliant LV often results in insufficient drainage, with concomitant poor pump performance, and residual LA hypertension, with subsequent pulmonary congestion. In these patients, the LA/LV disproportion with preservation of systolic function makes LA cannulation an attractive option, perhaps with less risk of ventricular thrombus formation than in dilated cardiomyopathies.

Optimal pump sizing could aid an uninterrupted drainage, and these patients could benefit most from the versatility offered by the additional chamber size.

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Avoidance of completion pneumonectomy by sparing the right middle lobe

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Metachronous primary lung cancers (MPLCs) are defined as separate, unrelated lung cancers occurring at different times. Surgical resection remains a viable treatment option for patients with MPLCs. In patients with a history of lobectomy for the earlier ipsilateral MPLC, an additional lobectomy creates the perceived need for a completion pneumonectomy. On the right side, sparing the right middle lobe by itself is considered inappropriate by most. We challenged this common principle in treating a relatively high-risk patient.