Hybrid procedure with pulsatile ventricular assist device for hypoplastic left heart syndrome awaiting transplantation

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VAD implantation scheme.

▶ Video clip is available online.

Central Message

This report presents a novel technique of supporting a neonate with HLHS, moderate RV dysfunction, and severe tricuspid regurgitation by using a single Berlin EXCOR (Berlin Heart, Inc, The Woodlands, Tex) VAD and hybrid palliation as a bridge to orthotopic heart transplantation.

See Commentaries on pages e63 and e65.

In patients with hypoplastic left heart syndrome (HLHS) who are poor candidates for single ventricle (SV) palliation, the only recourse is orthotopic heart transplantation, sometimes with strategies to mechanically support the failing SV.^{1,2} Weinstein and colleagues,³ using the Berlin EXCOR device (Berlin Heart, Inc, The Woodlands, Tex), noted that patients with SV anatomy had the worst survival before cavopulmonary anastomosis when compared with postbidirectional or total cavopulmonary shunt (11% vs 58% vs 60%). Conway and colleagues,⁴ in an analysis of the Berlin EXCOR experience with the 10-mL pumps, noted that outcomes for patients weighing less than 5 kg are significantly worse, with 27.3% survival, and that congenital heart disease was a risk factor for mortality, with 92% mortality in the subset weighing less than 5 kg with congenital heart disease.⁴ At our institution, we developed a novel strategy for a neonate with HLHS (weight, 3.75 kg; body surface area, 0.24 m²) of combining ventricular support using the Berlin EXCOR device and balance of circulation using hybrid palliation.

CASE REPORT

In a 39-week-old female patient, weighing 3.75 kg and prenatally diagnosed with HLHS (mitral/aortic atresia), an echocardiogram demonstrated a severely hypoplastic left ventricle with mitral and aortic atresia, a severely hypoplastic ascending aorta (0.28 cm), unrestrictive atrial communication, a moderate-sized patent ductus arteriosus, severe tricuspid regurgitation, and deteriorating right ventricular (RV) function. She was deemed a poor candidate for

palliation and listed for orthotopic heart transplantation. Despite maximal medical therapy, end-organ function continued to worsen and a decision was made to support the patient with combined ventricular assist device (VAD)

VIDEO 1. Suggested indications and procedure for hybrid procedure with Berlin EXCOR implantation in HLHS. Video available at: [https://www.jtcvs.org/article/S0022-5223\(18\)33464-0/fulltext](https://www.jtcvs.org/article/S0022-5223(18)33464-0/fulltext).

FIGURE 1. Berlin EXCOR (Berlin Heart, Inc, The Woodlands, Tex) implantation schematic in HLHS with hybrid procedure. PDA, Patent ductus arteriosus; PA, pulmonary artery.

and stage 1 hybrid palliation to stabilize the systemic circulation and protect the pulmonary vasculature.

A median sternotomy was performed, followed by pericardiotomy and evaluation of the cardiac structures. Cannulation of the innominate artery, superior vena cava, and inferior vena cava was performed in preparation for cardiopulmonary bypass. The right and left pulmonary arteries were banded using a cut ring of a 3.5-mm polytetrafluoroethylene graft. Cardiopulmonary bypass was then established. Subsequently, the right atrium was opened and atrial septectomy was performed. The inflow cannula of the VAD was inserted in the atriotomy incision purse-strings and secured with a Prolene 5-0 continuous suture.

The ductus was then snared with a vessel loop, and the main pulmonary artery was opened. The outflow cannula of the VAD was connected to the main pulmonary artery by attaching the Dacron graft extension at an angle optimal to flow. The cannulas were connected to the VAD after adequate deairing, and the transition was made from cardiopulmonary bypass to the VAD. Heparin was reversed, and the cannulas were removed. Under fluoroscopy, the RV was cannulated with a 6F 11-cm sheath, and a guidewire was passed into the descending aorta. After appropriate measurements, a 10 × 30-mm self-expanding stent was deployed in the ductus (Video 1). Postprocedure angiograms confirmed appropriate stent position and pulmonary artery bands. Sternal closure was performed the following day (Figure 1).

The VAD rate was initially set at 70 beats/min with a systolic pressure of 200 mm Hg, diastolic pressure of −20 mm Hg, and cardiac index of 3 L/min/m². Bivalirudin was initiated on postoperative day 1 with a target partial thromboplastin time of 60 to 90 seconds. The VAD rate was gradually increased to provide systemic index (confidence interval, 3.5-4.5 L/min/m²). The patient was extubated 1 week postprocedure and supported for 40 days with appropriate weight gain and end-organ function before transplantation. Target parameters were not unlike other stage 1 procedures.

DISCUSSION

Pulsatile VAD and stage 1 hybrid palliation in a neonate has not been previously reported. We have performed this in 5 patients with HLHS (2.5-3.5 kg), with 1 currently awaiting transplant. Patients' 30-day survival is 100%, with 1 patient currently 1 week from implant. The other patient underwent transplantation after 162 days. Of the other 2 patients, 1 died of severe sepsis (138 days of support) and 1 had support withdrawn (56 days of support) because of liver failure from neonatal hemochromatosis (diagnosed postimplant). Reasons for this strategy were severe tricuspid regurgitation and RV dysfunction in 3 patients and RV-dependent coronary fistulization in 2 patients. All patients were anticoagulated with bivalirudin. There were no major VAD-related complications during mechanical support. However, the patient continued to be at risk of cerebral ischemia from the hybrid circulation.

TABLE 1. Combined hybrid procedure Berlin EXCOR (Berlin Heart, Inc, The Woodlands, Tex) ventricular assist device implantation experience

Patient ID	Diagnoses	Age at implant (wk)	Weight at implant (kg)	Days on support	Cardiac index (L/min/m ²)	Outcomes
1	HLHS/severe TR	3	3.95	64	3.8	Transplantation
2	HLHS/severe TR	18	3.9	138	4	Died
3	HLHS (MS/AA) with coronary fistulae	2	2.4	162	4.5	Transplantation
4	HLHS (MS/AA) with coronary fistulae	2	3.4	56	4.3	Support withdrawn
5	HLHS/severe TR	3	2.9	Ongoing	Ongoing	Currently on support

HLHS, Hypoplastic left heart syndrome; TR, tricuspid regurgitation; MS, mitral stenosis; AA, aortic atresia.

Other strategies include nonpulsatile/continuous-flow ventricular support.⁵ However, in the largest series to date, risk of bleeding (29%), neurologic complications (24%), and only a 60% favorable outcome among patients with SV were noted.⁶ The likely reasons for our success are the anticoagulation strategies and the combination of the hybrid physiology with a targeted higher cardiac index providing a balanced, adequate systemic circulation.

CONCLUSIONS

Our experience with 5 patients suggests that a pulsatile VAD strategy along with the established hybrid palliation can be a successful long-term option to transplant without significant complications (Table 1). Further studies would help this critical population.

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