Successful use of a ventricular assist device in a neonate with hypoplastic left heart syndrome with right ventricular dysfunction

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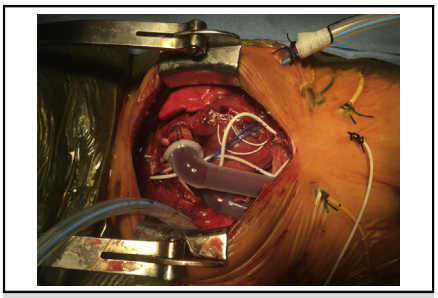

Right atrial to pulmonary trunk VAD cannulation with Berlin Heart EXCOR cannulas.

Central Message

Right atrial to pulmonary trunk temporary VAD support with Thoratec PediMag in combination with bilateral PA banding can be a viable option in children with HLHS born with depressed cardiac function.

See Editorial Commentary page XXX.

Neonates with hypoplastic left heart syndrome (HLHS) and severe ventricular dysfunction are difficult to bridge to heart transplant (HT) with current mechanical support devices.[1,2] In 2017, Gazit and colleagues[3] described a novel approach in which the PediMag device (Thoratec Corporation, Pleasanton, Calif) and Berlin Heart cannulas (Berlin Heart GmbH, Berlin, Germany) were used to support a neonate with HLHS. Although the patient was supported for 35 days, the patient did not survive to transplant because of progressive pulmonary vein disease. We present the case of a neonate with HLHS and severe ventricular dysfunction supported successfully to HT and discuss the lessons that we learned from our experience.

CASE

A term 3.5-kg male infant had a postnatal diagnosis of HLHS and severe right ventricular dysfunction after presenting in cardiogenic shock treated with a prostaglandin infusion, multiple inotropes, and mechanical ventilation. Echocardiogram showed mitral stenosis and aortic stenosis, a dilated right ventricle with severe dysfunction, and mild to moderate tricuspid regurgitation. On day of life 3, the patient underwent bilateral pulmonary artery banding. His cardiac function remained poor, precluding weaning of support, so a decision was made to implant the Thoratec PediMag temporary circulatory support device as a bridge to HT.

PediMag Placement Off Pump

After reopening the chest, and without cardiopulmonary bypass, a side-biting clamp was applied on the proximal pulmonary trunk just above the sinotubular junction. An 8-mm Hemashield Platinum Dacron graft (MAQUET Cardiovascular, LLC, Wayne, NJ) was sewn onto it with 7-0 Prolene running sutures (Ethicon Inc, Somerville, NJ). The graft was then connected into a 6-mm Berlin Heart EXCOR outflow cannula, which was tunneled out from the upper abdomen. For the inflow cannula, a 6-mm Berlin Heart atrial cannula was used (Figure 1). To prevent clot formation and ensure adequate flow, multiple additional

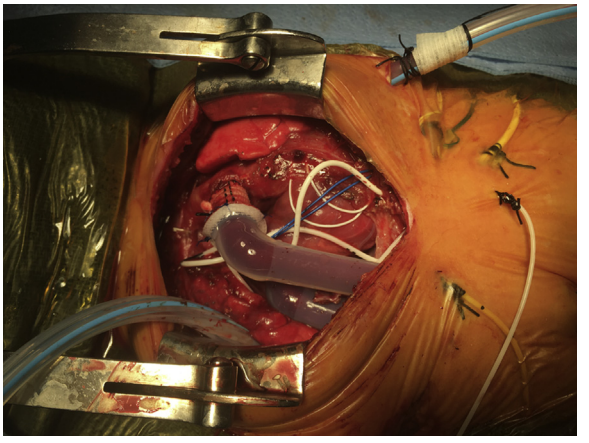

FIGURE 1. Right atrial to pulmonary trunk ventricular assist device cannulation with Berlin heart EXCOR cannulas (Berlin Heart GmbH, Berlin, Germany).

TABLE 1. Lessons learned from problems encountered and Pedimacs-defined adverse events

Problem	Description	Lessons learned
Severe hemolysis early after implantation	Frank hemoglobinuria was present, with LDH peak of 3740 U/L and total bilirubin peak of 21.4 mg/dL within 72 h of implantation.	Although pump cylinder appears to fit snugly within pump housing, it is possible for it to sit slightly off-axis. Pump was replaced on postoperative day 2 with proper positioning verified, with rapid resolution of hemolysis.
Recurrent bloody stools suspect for NEC	Despite excellent pump flows and evidence of good systemic perfusion, hematochezia (in absence of radiographic evidence of NEC) persisted, permitting only intermittent trophic feeds while on support.	It can be difficult to be differentiate hematochezia and NEC in a neonate receiving anticoagulation during VAD support. Because we were unsure, and recurrent bleeding could lead to stopping heparin (leaving the pump unprotected), we elected to feed parenterally for much of the patient's course. Despite this, the patient gained weight.
Splenic infarct	Noted incidentally on CT of abdomen obtained to evaluate NEC.	It was difficult to determine whether this was related to PediMag device because of the lack of previous CT; however, it would be attributed to PediMag by most adjudication processes.
Culture-negative sepsis	Fever, hypotension, and AKI were present in absence of positive blood culture.	Treated conservatively with broad-spectrum antibiotics, antifungals, and line replacement because of presence of hardware.
Limited mobility and confinement to intensive care	We were unable to discharge the patient to floor or engage in aggressive physical therapy because of extracorporeal pump configuration.	Patient was able to sit in parent's lap and feed while awake and extubated.
Persistent venous congestion complicated by AKI despite VAD support	VAD rotational speed was increased to escalate flows to as high as 1.5 L/min (7 L/min/m^2), and dopamine was used to maintain an adequate renal perfusion pressure to clear free water.	This is a common problem in patients with single ventricle, who tend to be volume loaded. Additional side holes in atrial cannula permitted escalation of flows as needed to overcome.
Pulmonary edema	Pulmonary edema despite low central venous pressure prompted cardiac catheterization and stent placement in atrial septum.	Patient may require repeated cardiac catheterizations to maintain stability during wait for a donor heart. Weekly echocardiographic surveillance of hybrid circulation is required to allow early intervention for evolving anatomic problems.
Labile saturations and pulmonary blood flows	Labile saturations and pulmonary blood flows, attributed to variable pulmonary to systemic blood flow ratio on PGE, improved with ductal stent.	Patient may require repeated cardiac catheterizations to maintain stability during wait for a donor heart; this also mirrors our general experience with single-ventricle hybrid circulations.

LDH, Lactate dehydrogenase; *NEC*, necrotizing enterocolitis; *VAD*, ventricular assist device; *CT*, computed tomography; *AKI*, acute kidney injury; *PGE*, prostaglandin E.

holes were created with a 6-mm punch on the ''basket'' portion of the cannula. Purse-string sutures were placed on the right atrium, and this modified cannula was then placed and tunneled out from the right upper abdomen. The cannulas were then connected into the Thoratec PediMag through a ¼-inch connector and tubing. The PediMag pump was started and ramped up to 2400 rpm, generating 1.4 L/min of flow (6.5 L/min/m^2), to achieve a central venous pressure of 5 mm Hg. The chest was closed primarily without impairing VAD flow.

Postoperative Course

Postoperatively, the speed was decreased to 2100 rpm (flow of 1.1 L/min, or 5.2 L/min/m^2), where it remained for the rest of his course (range, 0.4-1.5 L/min). Heparin was started after bleeding stopped and switched to enoxaparin sodium (anti-Xa level 0.6-1.0), aspirin (20 mg/kg), and clopidogrel (0.8 mg/kg). The patient was extubated on postoperative day (POD) 10. His atrial septum required stenting on POD 26 as a result of progressive restrictive physiology, and his patent ductus arteriosus was stented on POD 29 for labile saturations. Pedimacs-defined adverse and problems encountered are summarized in Table 1. The patient gained weight despite being unable to tolerate enteral feeds consistently. After 4.5 months of support, he underwent a successful transplant. He was discharged home on posttransplant day 90 and is now doing well 6 months posttransplant.

DISCUSSION

In this report, we document our first successful use of the PediMag device to bridge to HT a neonate with HLHS and