The use of ventricular assist devices in pediatric patients with univentricular hearts

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Improved survival outcomes for pediatric patients with ventricular assist devices (VADs) has expanded their application to complex univentricular cardiac lesions, including patients with systemic-to-pulmonary shunts, bidirectional cavopulmonary anastomoses (BCPAs), and total cavopulmonary Fontan circulations. Although it is generally recognized that complex cardiac anatomy is likely a significant risk factor affecting VAD survival,^{1,2} there remains a paucity of data examining the pediatric VAD experience with univentricular hearts.

CLINICAL SUMMARY

A 3-year-old 11.5-kg boy with mitral valve atresia, hypoplastic left ventricle, coarctation, and transposition of the great vessels underwent an atrial septectomy with pulmonary artery banding followed by a BCPA. An extracardiac fenestrated Fontan procedure was performed at 3 years of age. Postoperatively, he had increasing diastolic dysfunction and thrombus formation in the extracardiac conduit necessitating emergency revision back to a BCPA with extracorporeal membrane oxygenation. A 30-mL Berlin Heart Excor Pediatric device was implanted with systemic right ventricular and aortic cannulation on postoperative day 10. His post-VAD implantation course was complicated by a left subdural hemorrhage requiring decompressive craniotomy and evacuation on day 15 despite adherence to a standard anticoagulation protocol (the Edmonton anticoagulation protocol). On days 38, 89, and 111, fibrin collections in the outflow valve necessitated pump exchanges. The remainder of his VAD course was uneventful for bleeding or thromboembolic events. At 174 days after VAD implantation, he underwent successful transplantation and remains well with no neurological sequelae.

Ten pediatric patients with VADs with single-ventricle anatomy have been described in the literature since

1987: 6 patients with Fontan circulations, 2 patients with BCPAs, and 2 patients with a systemic-to-pulmonary shunt (Table 1).³⁻¹²

DISCUSSION

The paucity of published reports on VAD support in patients with single-ventricle anatomy precludes firm recommendations regarding a preferred surgical technique and postoperative management strategy. However, clinical experience to date can be used to formulate suggestions based on the specific cardiac anatomy and physiology of the clinical scenario.

Patients with shunt-dependent circulation can be adequately supported with systemic ventricular inflow and aortic outflow cannulation. However, the parallel circulations usually require increased pump flow rates ($\geq 200 \text{ mL} \cdot \text{kg}^{-1} \cdot \text{min}^{-1}$) to accommodate the need for both pulmonary and systemic perfusion and to maintain adequate oxygenation. Inadequate systemic oxygenation caused by impaired lung function can be addressed by placing a low-resistance oxygenator within the outflow circuit.

Patients with BCPAs, as in this case report, can be supported with a systemic ventricular inflow and aortic outflow cannulation technique. This management strategy has the potential advantage of maintaining systemic cardiac output in the presence of increased pulmonary vascular resistance at the expense of lower systemic oxygen saturations. For this reason, as with shunt-dependent circulation, a larger pump size might be needed to meet end-organ oxygen demand. BCPA physiology inherently unloads the hepatic, renal, and splanchnic circulations by reducing venous hypertension and thereby decreasing the risk of multiorgan failure.

The failing Fontan circulation poses several surgical options for VAD support that can be influenced by patient specific circumstances. Ventricular dysfunction as the primary cause for failing Fontan circulation can be addressed with single-pump VAD support of the systemic ventricle. However, increased pulmonary vascular resistance might require an additional pump within the pulmonary circulation to address systemic venous hypertension. In this situation implantation of a pulmonary VAD requires revision of the Fontan pathway for separation of the systemic venous and pulmonary circulations. Notwithstanding the extensive nature of the operation, there is an increased risk associated with the creation of a totally VAD-dependent pulmonary

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TABLE 1. Ventricular assist devices in pediatric patients with single-ventricle anatomy

Case reports	Age (y)	Weight (kg)	Anatomy	Surgical procedure	Device	Duration of support (d)	Complication	Outcome
Matsuda et al, 1988 ³	10	16.0	Single-ventricle PS	Fontan	Toyobo	7	MOF Cardiac tamponade Re-exploration × 2	Death day 7
	3	10.0	Single-ventricle PS	Systemic-to-pulmonary shunt (BT shunt)	Toyobo	5	Respiratory failure	Death day 5
Sadeghi et al, 2000 ⁴	8	22.0	HLHS MA	Fontan	BVS 5000	8	Pump change for fibrin	Transplantation
Nathan et al, 2006 ⁵	4	12.0	HLHS	Fontan	Berlin Heart	28	None	Transplantation Death caused by graft failure
Chu et al, 2007 ⁶	4	14.6	HLHS	BCPA	Berlin Heart	13	MOF Bowel necrosis Mesenteric, hepatic, and renal thrombi	Death day 13
Calvaruso et al, 2007 ⁷	10	32.0	Single RV MA, PA	Fontan (extracardiac TCPC)	Berlin Heart	7	None	Transplantation
Frazier et al, 2005 ⁸	14	63.0	TA	Fontan	Centrifugal pump and HeartMate I	45	ARF, HIT	Transplantation
Russo et al, 2008 ⁹			HRHS					
Pearse et al, 2009 ¹⁰	1.3	NS	DORV, MA d-TGA	Systemic-to-pulmonary shunt	Berlin Heart	49	None	Transplantation
Cardarelli et al, 2009 ¹¹	1.5	NA	HLHS	Fontan (extracardiac TCPC)	Berlin Heart	179	Neuroimpairment Change in pump size	Decannulation
Irving et al, 2009 ¹²	2.9	13	HLHS	BCPA	Berlin Heart	7	None	Transplantation

PS, Pulmonary stenosis; MOF, multiorgan failure; BT, Blalock–Taussig; HLHS, hypoplastic left heart syndrome; MA, mitral atresia; BCPA, bidirectional cavopulmonary anastomoses; RV, right ventricle; PA, pulmonary artery; TCPC, total cavopulmonary; TA, tricuspid atresia; HRHS, hypoplastic right heart syndrome; ARF, acute renal failure; HIT, heparin-induced thrombocytopenia; NA, not applicable; DORV, double-outlet right ventricle; d-TGA, transposition of the great arteries.

circulation. In the event of pump failure or a necessary pump change, complete cessation of pulmonary flow and cardiac output would occur.

For all types of palliated univentricular anatomy, strict adherence to anticoagulation protocols with frequent monitoring of the thromboelastogram, as opposed to reliance on activated clotting times, has improved the rate of thrombotic and hemorrhagic complications.^{13,14} Patients with single-ventricle circulations are at increased risk of thromboembolic events caused by liver dysfunction and coagulation factor alterations, such as protein C deficiency. Vigilant surveillance of neurological status is pivotal.

Improvement in survival to transplantation has been demonstrated in recent years for all types of palliated single-ventricle anatomy. This is likely attributable to refinement in patient selection, surgical technique, and anticoagulation management. Further experience and refinement of the preimplantation and postimplantation management is necessary to improve overall outcomes in this challenging group of patients.

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Pulmonary valve replacement in repaired tetralogy of Fallot by left thoracotomy avoid ascending aorta injury

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We report the follow-up of a patient born with tetralogy of Fallot (TOF), which was corrected in childhood, who required when he was 28-years-old a pulmonary valve replacement (PVR) through a left anterolateral thoracotomy. This case demonstrates the advantages of this approach of simplicity and low risks of aortic lesions in particular.

CLINICAL SUMMARY

In the neonatal period, this male patient underwent a Blalock anastomosis through a left posterolateral thoracotomy. When the patient was 2 years old, a second systemic–pulmonary shunt through a right posterolateral thoracotomy was performed. The total repair was performed when the patient was 4 years old through a median sternotomy with extracorporeal circulation (ECC) and aortic clamping, with transanular enlargement of an hypoplastic pulmonary arterial trunk (PAT) with autologous pericardium. The outcome was quite favorable, with normal growth and without any functional symptoms, so medical monitoring was more deliberately spaced.

At the age of 28 years, during an ordinary medical check-up, the patient was asymptomatic. Furthermore, the Holter electrocardiogram showed sinus rhythm, a complete right

bundle branch block, and QRS prolonged to 0.2 seconds. Echocardiography showed significant pulmonary regurgitation with dilated hypokinetic right ventricle. A computed tomographic scan showed dilatation of the PAT to 55 mm. Magnetic resonance imaging confirmed the infundibular dilatation, right ventricular ejection fraction of 25%, and end-diastolic and end-systolic volumes of 172 mL/m² and 128 mL/m², respectively. The ascending aorta was dilated to 44 mm and completely adherent to the posterior of the sternum (Figure 1, A).

PVR was indicated. To avoid the sternotomy-associated risk of damage to the ascending aorta, the preferred route of left anterolateral thoracotomy was chosen, especially because the PAT was just subcostal (Figure 1, B).

With the patient under general anesthesia, left femorofemoral ECC was initiated. A left anterolateral thoracotomy was performed (Figure 2, A). After opening of the 4th intercostal space with preservation of the left internal thoracic artery, the pericardium was exposed after release of some of the lung adhesions. The PAT was easily exposed because it was decompressed under ECC. A longitudinal incision and resection of the pulmonary infundibular excess allowed good exposure of the pulmonary ring. A No. 25 Mitroflow bioprosthetic valve (Sorin SpA, Saluggia, Italy) was fixed with a 3-0 Prolene suture (Ethicon Inc, Somerville, NJ; Figure 2, B). Direct closure of the PAT, weaning from ECC, and closure of the thoracotomy were conventional.

The patient was transferred to the intensive care unit, extubated at 6 postoperative hours, and transferred to the cardiology department. Echocardiography showed a well-functioning bioprosthetic valve and absence of pericardial effusion. The patient returned to his home 8 days after the operation without functional symptoms.

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