Use of a HeartWare Ventricular Assist Device in a Patient With Failed Fontan Circulation

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We present a successful case of the use of a HeartWare ventricular assist device as a bridge to transplantation in an 11-year-old with a hypoplastic left heart and failed Fontan circulation.


Efforts to achieve effective and durable mechanical circulatory support (MCS) in patients with Fontan failure confront important challenges. The combination of a high-risk patient population and the need to modify available applications may account for the limited published experience with MCS in this patient population [1–7]. We report, to our knowledge, the first successful long-term application of the HeartWare Ventricular Assist Device (HVAD, HeartWare, Framingham, MA) in an 11-year-old with hypoplastic left heart and failed Fontan circulation.

An 11-year-old boy (body surface area 1.1 m²) with hypoplastic left heart syndrome had undergone completed staged palliation with a fenestrated Fontan at age 2 years. He was listed for cardiac transplantation (35% class I and 22% class II human lymphocyte antigen [HLA] antibodies) at 11 years of age because of significant systemic right ventricular dysfunction and medically refractory symptoms of failure. Worsening organ failure prompted the decision to proceed with an HVAD.

The strategy for device cannulation was based on detailed measurements from cardiac magnetic resonance imaging. Left hemisternal retraction resulted in ventricular fibrillation, leading to prompt cardiopulmonary bypass through the right groin vessels. After conversion to central aorto-bicaval cannulation and with the heart filled and ejecting, transesophageal echocardiography and fine needle localization confirmed the plan of cannulating the anterior half of the diaphragmatic surface of the right ventricle. The inflow ring was buttressed with three doughnut-shaped felt “washers” to position the tip of the inflow cannula past the tricuspid subvalve apparatus and toward the right ventricular outflow tract. The position of the pump within the chest and cardiac silhouette is shown in Figure 1.

With the patient under cardioplegic arrest, the fenestration was closed, and the inflow ring and cannula were secured and seated; subsequent extensive resection of ventricular trabeculations was performed through the inflow ring. The remainder of the implantation was completed according to the usual technique. No restriction was placed on the outflow graft. The sternum was closed on postoperative day 1, and extubation occurred on postoperative day 3.

Anticoagulation with heparin began on postoperative day 2 and was converted to aspirin and warfarin as the patient tolerated oral medications. The target INR level was maintained at 2.5 to 3.5. Aspirin effectiveness was monitored by thromboelastography with platelet mapping. The final aspirin dose was 121.5 mg daily. Afterload reduction with milrinone and nitropresside was transitioned to enalapril and sildenafil. Invasive mean arterial pressure correlated with a palmar flash test with a sphygmomanometer on the wrist. The mean arterial pressure ranged from 64 to 78 mm Hg on follow-up. The HVAD was titrated from an initial revolutions per minute of 2500 up to 2900 over the first week according to displayed waveforms and frequent echocardiographic interrogations to assess decompression of the ventricle. No flow was seen across the native neoaortic valve on follow-up echocardiograms. The flow estimation on follow-up was 3.2 to 4.8 L/min, with an average of 3.5 L/min.

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Repeated HLA antibody testing revealed a significant increase in allosensitization with 100% class I and II antibodies. Luminex single-antigen bead testing was performed to characterize antibody strength and specificities. Desensitization therapy was initiated with bortezomib, rituximab, and intravenous immunoglobulin without resolution of sensitization.

After multidisciplinary rehabilitation and arrangement for outpatient follow-up, the patient was discharged 93 days after implantation. He was followed up as the first outpatient receiving MCS within the institution. He was readmitted four times (twice for infusion of desensitization therapy, once for fever without identification of a source, and once for transient slurred speech at home, which was not present on admission; no further investigation was pursued). The patient was supported on the HVAD with no hemodynamic concerns or complications other than allosensitization. Psychosocially he had difficulty with insomnia, anxiety, depression, and attending school despite close follow-up with psychology service.

Successful cardiac transplantation occurred 148 days after implantation. Despite persistent sensitization with 100% class I and II antibodies, the donor had a favorable HLA antigen profile, providing for a negative virtual crossmatch and a subsequent negative retrospective crossmatch by flow cytometry. The patient’s post-operative course was uncomplicated, and he was discharged 11 days after transplantation. He continues to thrive 4 months after transplantation with diminished but ongoing anxiety about the function of his new heart.

Comment

Despite the recognition of an increasing population of patients with failing Fontan circulations who would potentially benefit from MCS, reports of success are uncommon. A recent report of the Berlin Heart EXCOR (Berlin Heart Inc, The Woodlands, TX) in 204 pediatric patients included 19 single-ventricle patients with a mortality of 42% [8]. Successful uses of other devices have also been described in isolated case reports [1-3, 6, 7].

To our knowledge, this is the first report of successful long-term HVAD in a patient with a Fontan circulation. Certain factors played key roles in making this successful. The mechanism of failure was predominantly systolic ventricular dysfunction, which is theoretically more amenable to isolated VAD support of the systemic circulation. With preserved systolic function but systemic venous hypertension, a bi-VAD or total artificial heart may be necessary for effective support. However, this is not a straightforward issue, and little is presently known regarding MCS for the spectrum of Fontan failure.

Another important factor was multidisciplinary collaboration and good communication among the pediatric cardiothoracic surgery, cardiology, and intensive care teams and our adult cardiology/cardiothoracic surgery colleagues. The portable nature of the HVAD system and a newly formed multidisciplinary outpatient management team composed of a small number of staff made the transition to outpatient care possible.

As the medical aspects of the patient’s care stabilized, the most complex aspect became his psychosocial recovery. He perseverated on the possibility of device failure at night, resulting in insomnia. Outpatient follow-up with his primary care team of physicians and psychology service was integral to his mental health and his family’s well-being. We encouraged his gradual increase in school attendance accompanied by his mother, who was trained on the device. School administration, educators, and classmates were provided information regarding the HVAD through an in-person session led by our nurse practitioner and the patient.

The only complications of his support were the development of allosensitization. The use of the virtual crossmatch strategy to risk stratify donor-recipient pairs informed our assessment of risk versus benefit to decline or proceed with a given donor offer. Although a suitable donor was eventually found, the sensitization significantly prolonged his waiting period.

This case demonstrates the success that can be achieved with a high-risk Fontan patient by minor modification of available VAD technology and intense multidisciplinary effort. We emphasize the need for collaboration among institutions and expertise in the VAD industry to advance our understanding and care of patients with Fontan failure.

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References