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Case Report

Successful Bridge to Heart Transplantation through Ventricular Assist Device Implantation and Concomitant Fontan Completion in a Patient with Glenn Physiology: A Case Report

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A 3-year-old boy with Glenn physiology exhibited refractory heart failure with reduced ejection fraction. To improve the patient's oxygen saturation, he underwent ventricular assist device (VAD) implantation with concomitant Fontan completion. The extracardiac conduit Fontan operation was performed with a 4-mm fenestration. For VAD implantation, Berlin Heart cannulas were positioned at the left ventricular apex and the neo-aorta. Following weaning from cardiopulmonary bypass, a temporary continuous-flow VAD, equipped with an oxygenator, was utilized for support. After a stabilization period of 1 week, the continuous-flow VAD was replaced with a durable pulsatile-flow device. Following 3 months of support, the patient underwent transplantation without complications. The completion of the Fontan procedure at the time of VAD implantation, along with the use of a temporary continuous-flow device with an oxygenator, may aid in stabilizing postoperative hemodynamics. This approach could contribute to a safe transition to a durable pulsatile VAD in patients with Glenn physiology.

Keywords: Ventricular assist device, Glenn, Heart transplantation, Case report

Case report

A 3-year-old boy, weighing 13.2 kg and with a body surface area of 0.59 m^2, who had been diagnosed with tricuspid atresia type IIC was referred to Severance Cardiovascular Hospital for heart failure with reduced ejection fraction. He had undergone a Norwood operation at 2 months of age and received a bidirectional Glenn shunt at 7 months of age at our institution. Because the patient was living abroad, he returned to his home country after recovering from the bidirectional Glenn shunt operation. Two years later, he presented to the local hospital with severe heart failure symptoms and cyanosis. Echocardiography revealed severely depressed ventricular function, and he was urgently transported to our hospital. The patient's left ventricular ejection fraction was measured as 8%–15% on echocardiography, with a markedly dilated ventricle. A computed tomography scan similarly revealed a markedly dilated ventricle, along with severe stenosis of the left pulmonary artery, left pulmonary vein stenosis due to compression by a large neo-aorta, and multiple aortopulmonary collaterals. Despite being supported with dobutamine and milrinone, the patient's ejection fraction did not improve. Cardiac catheterization was performed, revealing a superior vena cava pressure of 18 mm Hg. Although many of the aortopulmonary collaterals were embolized, several remained. With no improvement in cardiac function, the decision was made to pursue ventricular assist device (VAD) implantation. Given that his oxygen saturation was below 80% even with oxygen supply, concomitant Fontan completion was planned. The heart was exposed via repeat median sternotomy, and cardiopulmonary bypass was established with distal ascending aorta and bicaval cannulation. A 16-mm expanded polytetrafluoroethylene graft (Gore Medical, Newark, DE, USA) was used to construct an extracardiac Fontan pathway. Due to the small size of the pulmonary artery, a 4-mm fenestration was made. For outflow cannula anastomosis, a 12-mm Hemashield graft

(Maquet, Rastatt, Germany) was anastomosed to the neo-aorta. An apical hole was made at the left ventricular apex, and stitches reinforced with a bovine pericardial strip were placed around the hole. A 12/9-mm apical cannula was fixed to the apical hole and tunneled out to the left upper abdominal quadrant. The graft adapter outflow cannula was tunneled and anastomosed with the previously placed Hemashield graft connected to the neo-aorta. The cannulas were then connected to a temporary continuous-flow device (Rotaflow; Maquet, Wayne, NJ, USA) with an oxygenator. As bypass was weaned, VAD support was initiated. The patient was separated from cardiopulmonary bypass with good hemodynamics. His mean blood pressure was 75–80 mm Hg, and his oxygen saturation was 100%. The VAD flow rate was 1.95 L/min at a pump speed of 1,900 revolutions/min (rpm). After the intravenous volume status and blood pressure were optimized, the required pump speed was decreased to 1,700 rpm to maintain the same VAD flow. The patient was extubated on the first postoperative day. On postoperative day 7, the sweep gas of the oxygenator was turned off, and the patient demonstrated acceptable oxygenation. The continuous VAD was discontinued and transitioned to a pulsatile Berlin Heart EXCOR 25-mL pump (Berlin Heart GmbH, Berlin, Germany). The pump rate was set at 70 beats/min, and the VAD flow was maintained at 1.75 L/min. After the pump exchange, the patient's blood pressure remained stable, and his oxygen saturation was 85%. One month later, the patient met the conditions for heart transplantation for foreigners and was listed for a heart transplant. He thrived on support and experienced no thromboembolic events. However, the patient was highly sensitized (panel-reactive antibodies: class 1, 97%; class 2, 92%), so 3 rounds of plasmapheresis were performed for desensitization. After a 3-month waiting period, he received a transplant from a 12-year-old donor. During transplantation, both pulmonary arteries were enlarged with a patch, and the enlarged aorta was reduced in size to prevent compression of the pulmonary veins after cardiectomy. Extremely high pulmonary venous return and excessive backflow from both pulmonary arteries were evident, suggesting numerous aortopulmonary collaterals. Despite no evidence of acute rejection, 4 additional plasmapheresis procedures were performed. The patient was extubated on the first postoperative day. On postoperative day 3, a second aortopulmonary collateral embolization was performed to reduce volume overloading of the transplanted heart. The patient provided written informed consent for publication of the case details and clinical images.

Discussion

Heart failure, accompanied by Glenn physiology secondary to ventricular dysfunction, can lead to systemic venous congestion, impaired pulmonary blood flow with notable cyanosis, and a maladaptive cycle of aortopulmonary collateral formation [1]. While practitioners have become more experienced with mechanical circulatory support for a single ventricle, the outcomes of ventricular support in cases involving Glenn circulation remain suboptimal [2]. We present a case of successful bridging to transplantation in a child with Glenn circulation and heart failure, achieved through a combination of Fontan completion and VAD implantation. Mechanical circulatory support for Glenn physiology presents unique challenges. The distinctive physiological state in Glenn circulation, characterized by a difference in upper and lower body venous pressure, has been identified as a major factor in reducing the efficiency of VAD support in Glenn circulation. Because the direct decompressive effect of the VAD is limited to the systemic venous system of the lower body in Glenn circulation, hypoxia may be exacerbated by the development of veno-venous collaterals from the upper to the lower systemic venous system [3]. Persistent cyanosis may result in exercise intolerance and the formation of excessive aortopulmonary collaterals. From this standpoint, upgrading to Fontan circulation at the time of VAD implantation is preferable to maintaining Glenn physiology. Moon et al. [3] described this strategy as "mechanically assisted Fontan completion." The present patient exhibited low oxygen saturation in Glenn circulation, necessitating Fontan completion to alleviate cyanosis. However, the patient had hypoplastic pulmonary arteries and an obstructive pulmonary vein, with poor pulmonary vasculature to accommodate Fontan circulation. Fenestration was required, although the benefit of improved oxygen saturation can be slightly offset by this procedure. Glenn physiology represents a volume-unloaded condition compared to shunted patients. However, excessive cardiac output is required due to the presence of aortopulmonary collateral circulation [1]. When using a Berlin Heart device, potential flow variation from collaterals should be considered when determining the pump size and rate. It is also challenging to identify an appropriate support rate for the pulsatile device in univentricular patients, as accurately estimating the required cardiac output can be difficult. In such cases, patients may benefit from a temporary continuous device [4]. Short-term support with temporary continuous-flow devices connected to Berlin Heart cannulas may offer easier titration of support in the

initial postoperative setting. Additionally, the ability to incorporate an oxygenator into the circuit is advantageous in the postoperative period. Following cardiopulmonary bypass and complex surgery, pulmonary vascular resistance and pulmonary parenchymal condition can deteriorate. Temporary continuous VAD support with an oxygenator may help patients with poor pulmonary vasculature, such as the patient described in this report, navigate this reactive period. A smooth transition to a pulsatile VAD, following stabilization of cardiac output and pulmonary condition with a temporary device, was safely achieved in our patient.

In summary, VAD implantation with concomitant Fontan completion can be a safe and beneficial strategy for patients with Glenn physiology and advanced heart failure. The application of temporary continuous-flow VAD with an oxygenator may aid in stabilizing postoperative hemodynamics, thereby facilitating a safe transition to a durable pulsatile VAD.

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Conflict of interest

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