Case Report

Continuous-Flow Left Ventricular Assist Device Therapy in Adults with Transposition of the Great Vessels

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An increasing number of children with congenital heart disease are surviving into adulthood and subsequently developing end-stage heart failure. Two example populations are adults who have been previously operated on for congenitally corrected transposition of the great arteries (CCTGA) and transposition of the great arteries (TGA). Implantation of a continuous flow left ventricular assist device (CF-LVAD) in these patients can present unusual anatomical and physiologic challenges. In this report, we describe outcomes of CF-LVAD implantation in three such patients. These cases demonstrate the feasibility of implanting a CF-LVAD in patients who have undergone surgery for CCTGA and/or TGA.

Keywords: heart failure, mechanical circulatory support, congenital heart disease

Introduction

As the outcomes of continuous-flow left ventricular assist device (CF-LVAD) implantation have improved,1,2 these devices have been used to treat a wider array of patients with end-stage heart failure, including those with congenital heart disease (CHD). Two such patient populations that could be candidates for CF-LVAD implantation are those with transposition of the great arteries (CCTGA) and transposition of the great arteries (TGA). CCTGA is a rare CHD featuring both atrioventricular and ventriculoarterial discordance; thus, the circulatory pathways are in series. In this condition, the right atrium is connected to the left ventricle and the left atrium is connected to the right ventricle; furthermore, the aorta arises from the right ventricle and the pulmonary artery arises from the left ventricle. In patients with CCTGA, as in those with TGA who have undergone the atrial switch procedure, the right ventricle is the systemic ventricle, and adults with this condition can develop systemic ventricular dysfunction.

TGA is a congenital cardiac anomaly in which the aorta arises from the right ventricle and the pulmonary artery arises from the left ventricle, resulting in ventriculoarterial discordance. Surgical repair is necessary during infancy for survival. Before the arterial switch operation (a definitive repair) became the standard therapy, TGA was treated by performing an atrial switch procedure (Mustard or Senning operation), which leaves the anatomic right ventricle as the systemic ventricle. When patients who have undergone this procedure reach adulthood, systemic ventricular dysfunction can be a serious problem.

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Recently, more patients with CCTGA and those with TGA who have undergone an atrial switch procedure are surviving into adulthood and subsequently developing end-stage heart failure. Implanting a CF-LVAD in such patients can be challenging because of the patients’ unique anatomic and physiologic abnormalities. Herein, we describe the outcomes of CF-LVAD implantation in three such patients.

Case Reports

Case 1

This 44-year-old man had a medical history that included CCTGA, dextrocardia, interrupted inferior vena cava, partial anomalous pulmonary venous return, atrial septal defect (ASD), and ventricular septal defect (VSD). He underwent an intracardiac repair operation during childhood. He developed progressive advanced heart failure and was admitted several times for decompensation episodes. After he underwent a complete workup by all services at our institution, we believed the patient was not a candidate for heart and lung transplantation. Thus, we implanted a HeartMate II (St. Jude Inc., St. Paul, MN, USA) LVAD as destination therapy. Cardiopulmonary bypass was established via the left groin. The device was placed below the diaphragm; the inflow cannula was inserted through the right-sided diaphragm because of his situs position and was attached to the apex of the systemic ventricle. The outflow graft was extended over toward the ascending aorta (which was to the left) and anastomosed. The unusual positioning of the HeartMate II is shown in Fig. 1. Although the patient had an ASD, we did not repair it. Postoperatively, we noted decreased oxygen saturation levels of 60%–70% because of right-to-left shunting, which was tolerated by the patient. If pulmonary circulation flow is reduced, oxygen saturation will decrease. We performed serial speed-change echocardiography and decided the pump speed according to the septal position, mean arterial pressure, central venous pressure, and oxygen saturation level. The patient had a long, complicated postoperative course that included a wound infection and respiratory failure and was discharged 6 months after the procedure. He died 8 months after surgery from unrelated causes.

Case 2

This 34-year-old man had a history of TGA, for which he had undergone a Mustard procedure during infancy. He was a status 1A candidate for cardiac transplantation and was receiving milrinone therapy (continuous intravenous infusion) at home while awaiting a donor organ. He was admitted to our hospital for progressive systemic right ventricular failure. Although he was receiving aggressive medical therapy, his condition continued to worsen. A HeartMate II LVAD was implanted as a bridge to heart transplantation. A preoperative computerized tomography scan showed a thickened, trabeculated systemic right ventricular wall. The femoral artery and vein were exposed, and a redo median sternotomy was performed. Cardiopulmonary bypass was instituted via the groin. We then placed the LVAD pump inside the peritoneal cavity, resting it on an omental flap anterior to the
We passed the inflow cannula through the diaphragm and placed it on the diaphragmatic surface of the anterior systemic ventricle. After coring the myocardium and resecting all crisscrossing trabeculations and the moderator bands, we created a wide, unobstructed opening to the ventricular cavity. The outflow graft was anastomosed to the ascending aorta (Fig. 2). His postoperative course was uneventful, and he was discharged on postoperative day 25. He underwent LV AD exchange 2 years after the initial implantation procedure because of a driveline infection and then underwent a heart transplantation 6 years after the initial LV AD implantation. He is now receiving follow-up care from an outpatient clinic.

**Case 3**

This 49-year-old man had a medical history that included CCTGA, situs inversus, ASD, VSD, pulmonic stenosis, and pulmonary hypertension. He underwent an uneventful intracardiac repair at 13 years of age. Heart failure began to develop when he was 47-year old, and at age 48 years, he was admitted for acute on chronic heart failure with cardiogenic shock requiring inotropic and vasopressor support and an intra-aortic balloon pump. An HVAD (Medtronic, Minneapolis, MN, USA) was implanted as a bridge to heart transplantation. After the femoral artery and vein were exposed, a redo median sternotomy was performed. Cardiopulmonary bypass was instituted via the groin. The inflow cannula was placed. The outflow graft was laid over on the right side of the heart and anastomosed to the ascending aorta. Because of his situs inversus, the HVAD was implanted in the normal pericardial position, but in the right chest. The unusual positioning of the HVAD is shown in Fig. 3. He was discharged home on postoperative day 49. He was supported with the HVAD for 4 years with no device-related complications, but he died of complications related to a stroke after 4 years of support.

**Discussion**

Adults with CCTGA and those with TGA who have undergone an atrial switch procedure can develop severe systemic ventricular (anatomic right ventricular) dysfunction, which can lead to heart failure. CF-LVADs may be able to provide good systemic right ventricular support in such patients; however, only a few reports have described successful CF-LVAD implantation in these patients. This may be due to the fact that these operations can be performed only in high-volume LVAD centers by experienced surgeons because of the unique anatomic and physiologic abnormalities of these patients.

Pigula et al. showed that the results of heart transplantation in adults with a CHD were comparable to those of adults without a CHD. When patients with a CHD present with end-stage heart failure and a suitable donor heart is not available, systemic ventricular assistance should be considered. Here, we describe the cases of three patients with CHD who received CF-LVAD support, one of whom successfully underwent heart transplantation after CF-LVAD implantation. Similarly, Hanke et al. described five cases in which patients with TGA who had undergone a Mustard operation later underwent HeartMate II implantation; three of those patients subsequently underwent a successful heart transplantation.

For all CF-LVADs, the inflow cannula should be directed away from the lateral wall and interventricular septum of the ventricle. This can be accomplished by either implanting the device into the true apex toward the mitral valve or implanting the device on the diaphragmatic surface of the ventricle. With all three approved devices, the HeatMate II, HeartMate III, and HVAD, the speed should be adjusted to allow the ventricle to eject through the native valve to prevent leaflet fusion and valvular regurgitation. Additionally, when adjusting the device speed, much attention should be given to not cause a shift of the interventricular septum from midline and cause failure of the other ventricle. The most
common speed range that allows for achievement of these goals is generally 8600-9000 RPM for HMII, 2200-2800 for the HVAD, and 5200-5800 RPM for the HM3. Periodic echocardiograms should be obtained to serially monitor these parameters and goals.

Conclusion

CF-LVAD implantation seems to be feasible in patients with TGA and end-stage heart failure, and may be a viable option for providing systemic right ventricular support in such patients.

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