

Long-term Right Ventricular Assist Device Support for Congenitally Corrected Transposition of the Great Arteries

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ABSTRACT

With congenitally corrected transposition of the great arteries, the right ventricle (RV) supports the systemic circulation. These patients have an increased risk of developing heart failure. Implantation of a ventricular assist device may be the only therapeutic option for patients who are not transplantation candidates. The technical aspects of implanting a mechanical device into the RV have not been well described, however. We describe our experience with one such case and describe our operative strategy in obtaining optimal placement of the inflow cannula.

INTRODUCTION

Patients in whom the right ventricle (RV) supports the systemic circulation have an increased risk of developing heart failure [Graham 2000]; the RV has inherent anatomic limitations in its ability to serve as the systemic ventricle. The extensive trabeculation of the RV muscle and the relatively inefficient mechanism of the tricuspid/atrioventricular (AV) valve make the RV prone to fail when subjected to higher systemic afterload over a prolonged period of time [Graham 2000].

In congenitally corrected transposition of the great arteries (CCTGA), a rare cardiac anomaly consisting of <1% of all forms of congenital heart disease, there is AV and ventriculoarterial discordance—the so-called double discordance [Warnes 2006]. Blood flows in the normal direction with parallel systemic and pulmonary circulations; however, venous blood returning to the right atrium via the vena cavae crosses the mitral valve into the morphological left ventricle (LV) and then is pumped to the lungs via the pulmonary artery. Oxygenated blood returns to the left atrium via the pulmonary veins, crosses the tricuspid valve into the morphological right ventricle (RV), and is pumped into the aorta, where it travels to the periphery. The tricuspid valve, located between the

left atrium and the morphological RV, is more appropriately termed the systemic AV valve since it services the systemic circulation. The mitral valve, between the right atrium and morphological left ventricle, is the pulmonary AV valve.

Some patients with CCTGA are asymptomatic until adulthood and do not receive a diagnosis for this anomaly until relatively late in life [Piran 2002]. By 45 years of age, congestive heart failure develops in 25% of patients who have CCTGA without additional anomalies and in 67% of patients who have CCTGA with additional cardiac anomalies, such as a ventricular septal defect, AV valve insufficiency, or pulmonic stenosis [Graham 2000]. Heart transplantation is ultimately required in 13% of patients with CCTGA [Graham 2000].

Implantation of an LV assist device (LVAD) may be the only therapeutic option for patients who are not transplantation candidates [Beauchesne 2002]; however, the technical aspects of implanting a mechanical device into the RV have not been well described. Anatomic challenges associated with LVAD implantation in CCTGA include identification of the appropriate inflow site and division of the moderator band and increased trabeculae that might cause inflow obstruction. In this report, we describe our experience with implanting a long-term mechanical support device into the RV of a patient with CCTGA. In particular, we focus on our operative strategy in obtaining optimal placement of the inflow cannula.

CASE REPORT

A 66-year-old man with CCTGA diagnosed at the age of 50 years presented with nonischemic dilated cardiomyopathy, severe pulmonary hypertension, and progressive worsening of heart failure symptoms. His echocardiogram was remarkable for significant RV dilatation (Figure 1) with an RV ejection fraction of 20% and moderate-to-severe systemic AV (tricuspid) valve regurgitation. The patient also had a moderately decreased LV (pulmonary ventricle) function, mild-to-moderate mitral valve regurgitation, and mild-to-moderate aortic valve insufficiency. Because of his severe pulmonary hypertension, the patient was not a transplantation candidate and therefore underwent implantation of a HeartMate II LVAD (Thoratec Corporation, Pleasanton, CA, USA). Cardiac transplantation may be reconsidered in the future if his pulmonary hypertension resolves.

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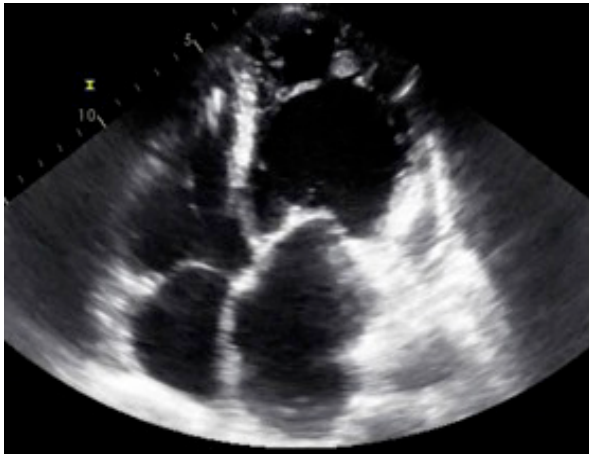


Figure 1. Echocardiogram demonstrating severely enlarged right (systemic) ventricle. MLV indicates morphological left ventricle; MV, mitral valve; RA, right atrium; MRV, morphological right ventricle; TV, tricuspid valve; LA, left atrium.

Description of VAD Implantation into the Systemic Right Ventricle

The aorta was anterior and to the left of the pulmonary artery (Figure 2). Owing to the significant aortic insufficiency, the aortic valve was repaired by closing the central portion of the point of leaflet coaptation. The outflow graft was then sized from the outflow housing to the proximal ascending aorta and cut accordingly. The outflow graft anastomosis was performed in an end-to-side fashion with single-layer 4-0 Prolene suture. The aortic cross-clamp was removed. The outflow graft was deaired. Transesophageal echocardiography was used to identify the AV valve, the interventricular septum, the aortic valve, and the RV apex. An incision was made in the RV apex, followed by insertion of a large Foley catheter and inflation of the balloon. The RV was volume loaded. While tension was maintained on the Foley catheter and the inflated balloon, the RV apex was cored with the circular coring knife in the direction of the AV valve. The inside of the RV was then inspected. The moderator band was excised (Figure 3), as were several additional trabeculae. Pledgeted horizontal mattress sutures were then placed around the ventriculotomy, with pledgets on the external surface. The HeartMate II inflow cannula was then inserted into the RV, directed toward the AV valve, and secured with a Vicryl tie and multiple umbilical tapes (Figure 4).

Intraoperative and Postoperative Course

The patient was weaned off cardiopulmonary bypass without difficulty, had an uneventful postoperative course, and was discharged home on postoperative day 14.

Mid-term Follow-up

At the time of this report, the patient has been on VAD support for approximately 9 months. He is in New York Heart Association functional class I and has not experienced any VAD-related adverse events. His hemodynamics have significantly improved, including a substantial reduction in central venous and pulmonary artery pressures. The patient is currently listed for cardiac transplantation in status IB.

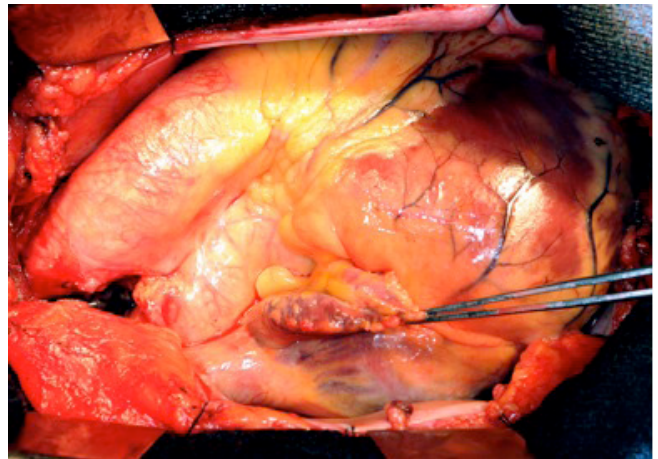


Figure 2. Relationship of aorta to pulmonary artery: Aorta is anterior and to the left of the pulmonary artery.

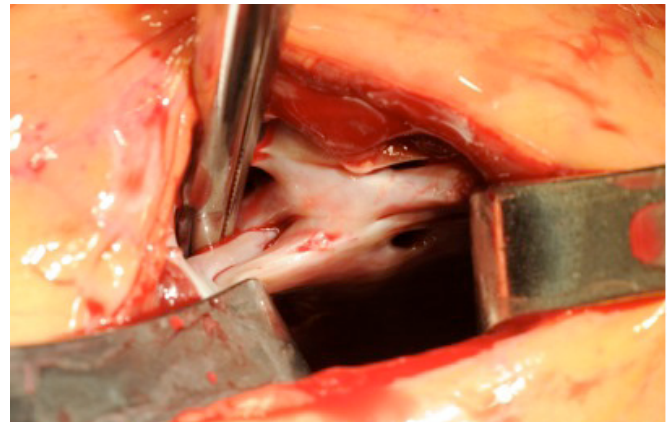


Figure 3. Coring of right ventricle with excision of the moderator band (located within right-angled clamp).

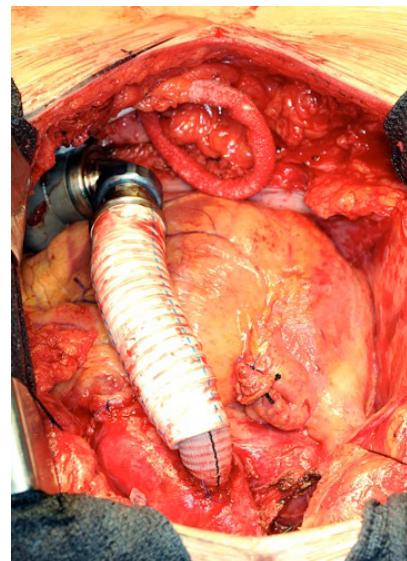


Figure 4. HeartMate II device in place.

DISCUSSION

Implanting a long-term VAD into the RV presents several anatomic challenges, which include appropriate coring of the RV, excision of the moderator band, and orientation of the inflow cannula. Unlike the LV, the RV does not have a true apex to guide the location of core excision and insertion of the cannula. The AV valve and septum should be identified via intraoperative transesophageal echocardiography, and the RV should be cored in a way that allows the inflow cannula to be directed toward the AV valve [Schoy 2009]. It is also essential to excise the moderator band and other prominent RV trabeculae to prevent obstruction of the inflow cannula. The techniques for optimal inflow cannula placement described in this report are also valuable for avoiding inadvertent injury of the interventricular septum and/or the posterior wall of the RV with the coring knife.

We are likely to see an increasing number of patients who will require long-term mechanical circulatory support of the RV. Although arterial switch is now the standard operation for surgical correction of transposition of the greater arteries, a large number of patients have undergone Mustard and Senning atrial switch procedures in the past, and the RV remains the systemic ventricle in these patients. Additionally, many congenital cardiac patients with a single-ventricle physiology who have undergone Fontan procedures also have a systemic RV. Implanting a VAD in the systemic RV presents several unique technical challenges but can be performed safely and effectively with an appropriate understanding of RV anatomy. Careful attention must be paid to cannula placement and orientation, as outlined in this report.

We are aware of 5 cases reported in the literature of patients with transposition of the great vessels who underwent LVAD implantation [Joyce 2010; Huebler 2012]. Inserting devices into the morphological RV requires an understanding

of RV anatomy. Additionally, there needs to be a willingness to aggressively resect moderator bands and trabeculae that may interfere with the inflow cannula. Without aggressive trabeculae resection, there is the possibility of developing inflow cannula-related complications, such as thrombus formation, pump thrombosis, and/or thromboembolic complications. The method for obtaining the optimal position of the inflow cannula in the RV that we have described may serve to decrease the incidence of these complications.

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