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Peer-Reviewed Case Report

Orthotopic heart transplant following implantation of a systemic ventricular assist device in a patient with congenitally corrected transposition of the great arteries and dextrocardia situs inversus totalis

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Abstract

Congenitally corrected transposition of the great arteries (CCTGA) is a rare congenital heart disease that commonly results in congestive heart failure due to exposure of the anatomic right ventricle to systemic pressures. Additional anatomic abnormalities, such as situs inversus, in which the viscera are mirror images of normal anatomy, or dextrocardia in which the heart apex is located on the patient's right, once were contraindications to transplant in the treatment of advanced heart failure. However, recent advancements now allow for the management of heart failure through mechanical circulatory support and subsequent transplant. This case presents a 52 year-old man with advanced heart failure secondary to CCTGA and dextrocardia situs inversalis totalis that was managed with a ventricular assist device (VAD) after maximal medical treatment, and subsequently received an orthotopic heart transplant. This case will also review CCTGA in relation to heart failure and the surgical challenges faced in terms of ventricular assist device implantation and orthotopic heart transplant.

Keywords

Heart transplant, VAD, dextrocardia, situs inversus, congenitally corrected transposition of the great arteries, heart failure

Introduction

Congenitally corrected transposition of the great arteries (CCTGA), or Levo-Transposition of the Great Arteries (L-TGA) is a rare form of congenital heart disease that results from abnormal leftward looping of the primitive heart tube yielding reversal of the ventricles. Patients with this condition exhibit both atrioventricular and ventriculoarterial discordance. As an isolated condition, patients are asymptomatic during early life. Nearly 90% of cases are associated with additional cardiac abnormalities, which may account for any early symptoms observed.[1] The most common of these conditions are ventricular septal defects, pulmonary outflow obstructions, as well as tricuspid and mitral valve abnormalities.

Dextrocardia can be an isolated occurrence, but is commonly associated with situs inversus totalis.[2] Typically, this condition is asymptomatic, although it may be associated with occurrence of congenital conditions such as Kartagener's Syndrome (Primary Ciliary Dyskinesia), congenital transposition of the great arteries, and various other conditions, all in varying combinations.[3] In the case of NYHA stage IV Heart Failure in these patients, situs inversus totalis was once considered a contraindication to mechanical circulatory support and heart transplantation.. However, recent advancements have made it possible to overcome this limitation.[4] Special considerations may include anastomosis of donor and recipient IVC and creation of a conduit out of the recipient's right atrium.[2-7] Prior to heart transplantion in this patient population, correction of extracardiac congenital conditions may be required.[8]

Many patients with CCTGA and situs inversus develop congestive heart failure (CHF) later in life. In these patients, mechanical circulatory support is one option, which presents increased technical difficulty with respect to surgical intervention. Southard et al have described one such case of ventricular assist device (VAD) placement in a patient with both dextrocardia and CCTGA.[9] Other cases have been reported in the literature regarding VAD placement or transplantation in either of these two conditions.[8,10,11] We present a case of systemic VAD placement in a patient with CCTGA and dextrocardia situs inversus to address the challenges of heart failure management in this particular patient population.

Clinical Summary

A 52 year-old man with a known history of CCTGA and dextrocardia situs inversus totalis presented in January 2013 with worsening shortness of breath, dyspnea on exertion, and lower extremity edema. Ejection fraction was 15%. His past medical history was significant for chronic atrial flutter, deep vein and atrial

thromboses, chronic kidney disease, and pulmonary hypertension. Outpatient medical management consisted of a beta blocker, angiotensin receptor blocker, loop diuretic, digoxin, and sildenafil.

He was in atrial fibrillation on admission. After optimization of hemodynamics with milrinone 0.5 mcg/kg/min and diuresis, catheterization demonstrated right atrial pressure 7mmHg, pulmonary artery pressure 48/26 with a mean of 34mmHg and pulmonary capillary wedge pressure 9mmHg. Cardiac index remained reduced at1.89 L/min/m2 by thermodilution and 1.78 L/min/m2 by Fick's method. Pulmonary vascular resistance was 6.6 Wood units. Coronary arteries were normal. It was determined that pulmonary ventricular function (morphological LV) was sufficient to allow use of a systemic ventricular assist device.

Transesophageal echocardiography (TEE) demonstrated biventricular systolic failure with an ejection fraction (EF) of 15%, severe regurgitation of the systemic atrioventricular valve (morphologic tricuspid valve) with a markedly dilated systemic atrium. A small membranous VSD with continuous flow was noted. Flow was not measured.

Despite optimal medical management, the patient was admitted with rapidly deteriorating NYHA Class IV, Stage D heart failure and was subsequently evaluated by an interdisciplinary team for the placement of a VAD. A cardiac MRI (Fig. 1) confirmed atrial and visceral situs inversus with L-transposition, biventricular hypokinesis with severe dilation of the systemic ventricle (systemic ejection fraction 23%), systemic atrio-ventricular valvular insufficiency, and pulmonary artery dilation to 5.7cm. The anatomy of the pulmonary veins and systemic thoracic veins was conventional. Based on hemodynamic and anatomic data, he was referred for mechanical circulatory support.

Implantation of the HeartWare HVAD ventricular assist device was performed through a median sternotomy. The patient's anatomy was identified carefully, with the superior vena cava (SVC) and inferior vena cava (IVC) clearly on the left side, and the systemic ventricle was obviously a morphologic right ventricle. The aorta and the pulmonary artery were relatively normally oriented, although the pulmonary artery was quite large, consistent with known pulmonary hypertension. An appropriate site on the ventricular free wall towards the right side of the left anterior descending coronary artery was identified. The HVAD ring was placed on the lateral side of the apex of the systemic (morphologic right) ventricle to approximate the permanent position of the VAD, allowing for stitches to be placed through the ventricular wall and the VAD ring. The outflow graft was anatamosed to the ascending aorta. Upon removal of the ventricular core, a papillary muscle had been injured, leading to a flail systemic atrioventricular valve leaflet. This was confirmed by intraoperative TEE, and the valve was replaced with a 31 mm pericardial tissue valve by approaching the mitral valve through the dome of the left atrium. The patient was successfully weaned from cardiopulmonary bypass with excellent VAD function (Fig 2).

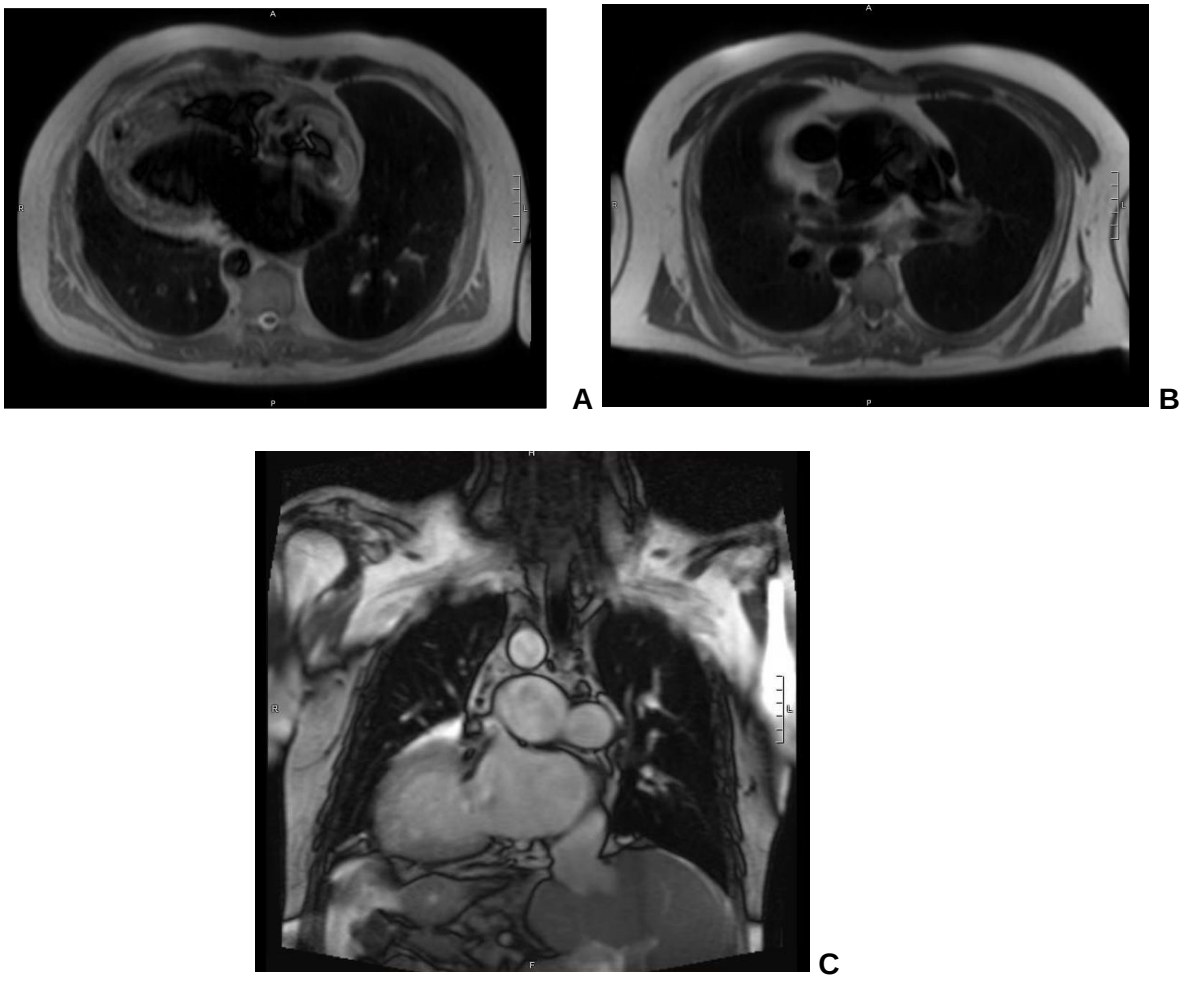

Figure 1. Preoperative MRI revealing dextrocardia and cardiomegaly (A), marked dilation of the pulmonary trunk (B), and situs inversus totalis (C).

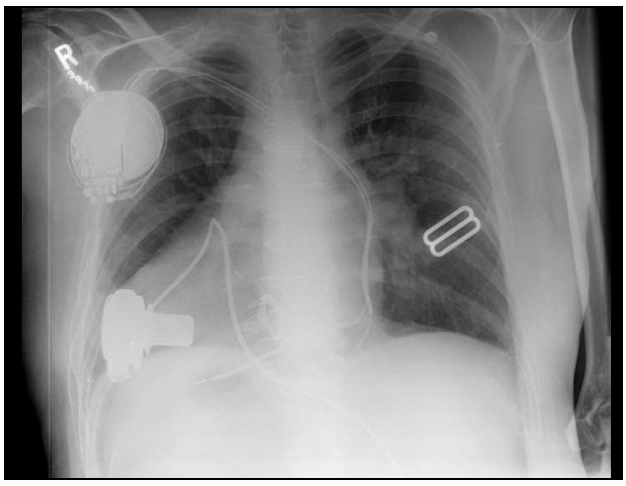

Figure 2. Postoperative chest x-ray showing right-sided placement of VAD

While recovering in the ICU, the patient was weaned from pressors and placed on sildenafil. Post op day 1, the patient developed complete atrioventricular block and a dual-chamber implantable cardioverter-defibrillator was implanted on post-operative day 2. The patient was extubated on post-operative day 9. Catheterization on post-operative day 22 showed excellent flow and filling pressures, with improved cardiac index (2.53 L/min/m2 by thermodilution, and 3.51 by Fick's method) and improvement of pulmonary hypertension (3.3 Wood units by thermodilution, 2.1 Wood units by Fick's method). Post-operative complications included a transient ischemic attack of right sided weakness that resolved right before discharge on post-operative day 28. The patient was found to have aspirin resistance via thromboelastogram, and he was placed on clopidogrel 75mg.

With the exception of difficulties related to anticoagulation that resulted in a subarachnoid hemorrhage without residual deficits, the patient was relatively stable with outpatient VAD management until a donor heart was located.

In May 2014, 1 year and 5 months after VAD placement, a suitable donor heart became available and the patient underwent successful orthotopic heart transplantation. Redo median sternotomy was performed, significant scar tissue within the mediastinum was dissected carefully. The patient was heparinized and cannulated. Cardiopulmonary bypass was initiated, the aorta was cross-clamped and the heart with its accompanying VAD was fully excised. The left-sided SVC was addressed by using a 16mm externally supported Gore-Tex graft, directed left to right across the diaphragm. Once the left atrium was anastomosed to the left atrial cuff, the IVC graft was anastomosed to the donor IVC. The recipient pulmonary artery was much larger in diameter than the donor pulmonary artery, requiring complex reconstruction with a bovine pericardial patch. To perform the SVC anastomosis, it was clear that the donor innominate vein did not have enough length to reach the recipient SVC, so a 10mm graft was placed. The patient was weaned successfully from cardiopulmonary bypass. The chest was left open as hemostasis was not adequate for closure and the patient was returned to the ICU. However, over the next two days, the patient developed plethora of the face and neck. The patient was returned to the OR, and a 13mm graft was placed between the recipient SVC and the donor SVC, and the 10mm graft was revised to connect the recipient innominate vein and the ligated donor jugular vein. (Fig. 3, 4). All grafts were reconstructed without interrupting venous return from the upper body. The chest was closed on post-operative day 5.

The postoperative course involved multiple arrhythmias treated with amiodarone, serratia bacteremia, acute kidney injury, thrombocytopenia, prolonged ventilation, pleural effusion requiring thoracentesis, and CO2 retention. As the left ventricle is directly under the sternum, echocardiographic assessment is difficult. Serial measurements of left ventricular function have been made with radionucleotide ventriculography. After a prolonged hospitalization and inpatient rehabilitation, he had remained well with normal functional capacity. Left ventricular function has remained normal, now 18 months after transplantation.

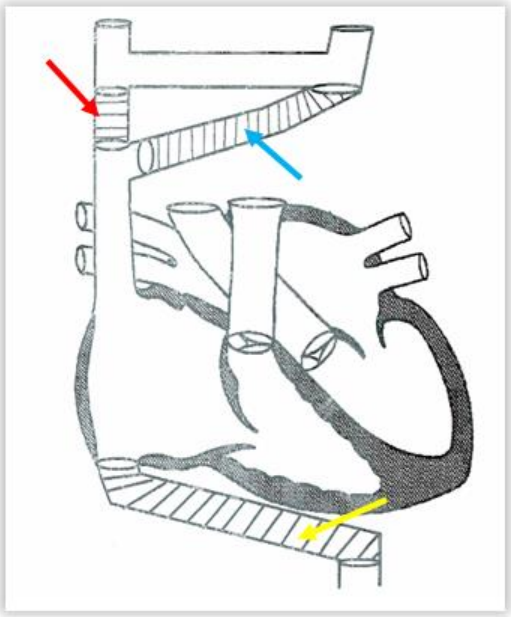

Figure 3. Illustration demonstrating placement of the 10mm (red arrow) recipient innominate vein to donor right jugular vein graft and 13mm (blue arrow) recipient SVC to donor SVC graft, and the 16mm (yellow arrow) IVC graft.

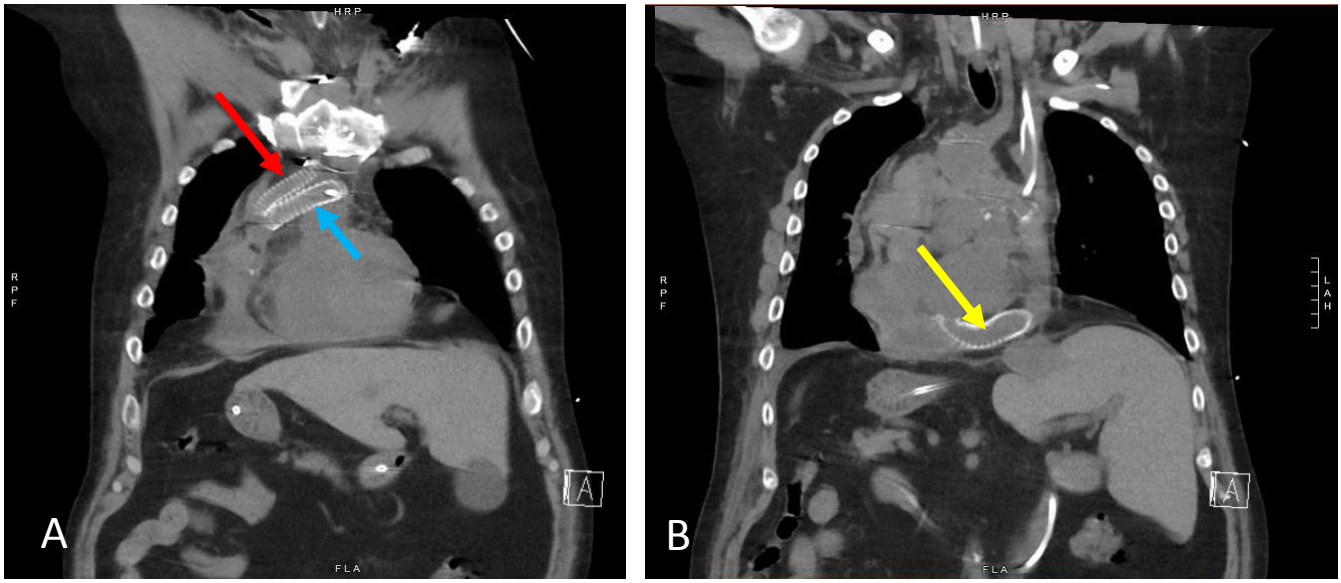

Figure 4. Postoperative computed tomography scan showing the 10mm (red arrow) recipient innominate vein to donor right jugular vein graft and 13mm (blue arrow) recipient SVC to donor SVC graft (A), and the 16mm (yellow arrow) IVC graft (B).

Discussion

CCTGA is a rare congenital anomaly occurring less than 0.5% of patients with clinically evident congenital heart disease. CCTGA presents as an atrioventricular discordance and ventriculoarterial discordance. Among these individuals, 67% with severe associated anomalies and 25% of patients with minor or no associated lesions will develop heart failure by age 45.[1] The double discordance in CCTGA results in a physiologic circulation, and unless there are symptomatic associated anomalies, patients with CCTGA may remain undiagnosed until the 4th or 5th decade, at which time they present often with heart failure. Physiologically, the long term overload of a morphologic RV supplying the systemic circulation, in combination with progressive AV valve dysfunction and arrhythmia, overcomes any compensatory hypertrophy and results in failure of the systemic ventricle. Subsequently, this can lead to pulmonary hypertension, and eventual biventricular failure. Therefore, the timing of referral for operative interventions in these patients is the major predictor of outcome in CCTGA.[12] For patients presenting with ventricular failure, options for intervention may include inotropic support, valve repair, pulmonary banding, anatomic repair via the “double switch” procedure, mechanical circulatory support, and ultimately cardiac transplantation. Although these patients are already high risk due to anatomical considerations and technical difficulty, mechanical circulatory support in patients with CCTGA is a feasible and viable option for patients either as a destination therapy or while awaiting transplantation.

Until recently, mechanical circulatory support was difficult in these patients, even without the concomitant presence of dextrocardia and situs inversus, which is evident by the fact that there have been very few cases of placement of a VAD in patients with CCTGA in the literature.[10,11,13] Additionally, there have been few cases of VAD placement in patients with either situs inversus or dextrocardia. Based on prior case reports and through our experience with this particular patient, it is evident that adjustments have to be made to ensure proper flow through the VAD as these devices were designed to reside in the wall of a morphologic left ventricle. In contrast to the near conical shape of the left ventricle, the right ventricle is more triangular in shape, with possible inflow obstructions such as an additional papillary muscle with accompanying chordae, trabeculations, and the moderator band. Using TEE guidance, these must be considered when determining placement of the VAD to prevent inflow obstruction. With the introduction of smaller devices, placement of VADs in this particular patient population will become more straightforward. The HeartWare HVAD has a displacement volume of 50 cc. and is intended to be implanted in the pericardial space. The outflow graft has a strain relief design that allows for flexible implantation in patients with varied anatomy.

Orthotopic heart transplantation into a space formerly configured to a dextrocardic situs inversus heart presents unique challenges. The main challenge is to reconstruct the systemic venous return pathways to accommodate a normally configured heart. This was first described in a case of heart-lung transplantation by Miralles et al, in which a large single atrium was

created to allow for venous anastomoses.[14] This had its own limitations, such as acting as point of stasis and possible thrombus formation, apart from its other physiological implications. An alternative, similar to the technique used in our case, was described in Deuse et al, consisting of a bi-caval technique with graft interposition depending on the length of the donor innominate vein available.[4] As evidenced by the SVC syndrome requiring graft revision encountered postoperatively in our case, special attention must be given to ensure that there is adequate venous return.

In conclusion, we report a case of successful HeartWare HVAD placement with subsequent orthotopic heart transplantation in a patient with CCTGA and dextrocardia situs inversus.

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