Ventricular Assist Device Implantation in a Patient Congenitally Corrected Transposition of the Great Arteries With I, D, D

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Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital heart anomaly that often leads to systemic heart failure, necessitating mechanical circulatory support or transplantation. We report a case of a 54 year old male diagnosed with dextrocardia and ccTGA (I, D, D), who had been suffering from congestive heart failure for over 7 years. Despite receiving intensive treatment, his condition deteriorated. Preoperative evaluation revealed significant systemic right ventricular dysfunction with severe valvular regurgitation and pulmonary hypertension. The surgical approach included tricuspid and aortic bioprosthetic valve replacement, mitral valve annuloplasty, and right thoracic ventricular assist device (VAD) implantation. The patient recovered well postoperatively and was discharged on postoperative day 39 with New York Heart Association (NYHA) class I. This case demonstrates the feasibility of using VAD in right heart ccTGA as a bridge to transplantation or destination therapy, emphasizing the importance of meticulous preoperative planning and intraoperative management for successful outcomes.
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Key Words: atrial situs inversus, congenitally corrected transposition of the great arteries, dextrocardia, ventricular assist device

Congenitally corrected transposition of the great arteries (ccTGA) is a rare anomaly that is seen in fewer than 0.5% of patients with clinically evident congenital heart disease.[1] Patients with ccTGA have inconsistent arterial connections to both ventricles, with the left ventricle connected to the pulmonary artery and the right ventricle connected to the aorta. Patients also had discordance atrioventricular connections, with the right atrium connected to the left ventricle through the mitral valve and the left atrium connected to the right ventricle through the tricuspid valve. Most patients are diagnosed late in their life and undergo surgical repairs, eventually leading to systemic ventricular failure and needing heart transplant or mechanical circulatory assistance.[2] Patients with ccTGA present unique challenges for ventricular assist device (VAD) implantation and management. The Corheart 6 is a newly developed magnetically levitated left VAD with small size, low weight, and low power consumption. It has good hemodynamic performance and blood compatibility when implanted in the human body.[3] In this case, we demonstrate the feasibility of the use of Corheart 6 in a ccTGA with atrial situs inversus (I, D, D) patient.

Case Report

A 54 year old man with dextrocardia and ccTGA (I, D, D) experienced refractory congestive heart failure for over 7 years despite intensive treatment with β-blockers, angiotensin-converting enzyme (ACE) inhibitors, and diuretics. He presented with severe fatigue, leg edema, and reduced exercise tolerance. A continuous dobutamine infusion was initiated on admission. N-terminal pro-B-type natriuretic peptide (NT-proBNP) exceeded 35,000 ng/L. Echocardiography and magnetic resonance imaging revealed a systemic morphologic right ventricle (mRV) dilated >90 mm with an ejection fraction (EF) of 0.12, severe atrioventricular and aortic regurgitation, and a morphologic left ventricle (mLV) EF of 0.35. No other structural defects were observed.

For such patients, our center generally provides three treatment plans before surgery: 1) valve replacement/repair; 2) valve treatment + VAD implantation; 3) medical therapy while awaiting transplantation. After multidisciplinary consultation and patient consent, VAD implantation with aortic and tricuspid valve replacements and mitral valve repair was chosen. Preoperative imaging confirmed the heart's structure (Figure 1). Right heart catheterization showed pulmonary artery pressure of 68/29 (42) mm Hg, right atrial pressure of 7/0 (2) mm Hg, wedge pressure of 31/21 (26) mm Hg, and cardiac index of 1.6 L/min/m^2. Thermodilution measured cardiac output at 4.2 L/min. Coronary angiography showed no stenosis.

During surgery, after general anesthesia and median sternotomy, the tricuspid valve (systemic atrioventricular valve) and aortic valves were replaced with Hancock II bioprostheses (Medtronic, Minneapolis, MN), and mitral valve annuloplasty was performed using a Carpentier-Edwards Physio ring (Edwards Lifesciences, Irvine, CA). A Corheart 6 ventricular assist device (Shenzhen Core Medical Technology Co., Ltd, Shenzhen, Guangdong Province, China) (Figure 2) was implanted. A Teflon ring was secured to the systemic ventricle using 12 pledgeted Polyester (Ethibond Excel) 2-0 sutures, and the myocardium within the ring lumen was excised with a coring knife. The inflow cannula was inserted, and the outflow graft, oriented toward the right pleural space, was anastomosed to

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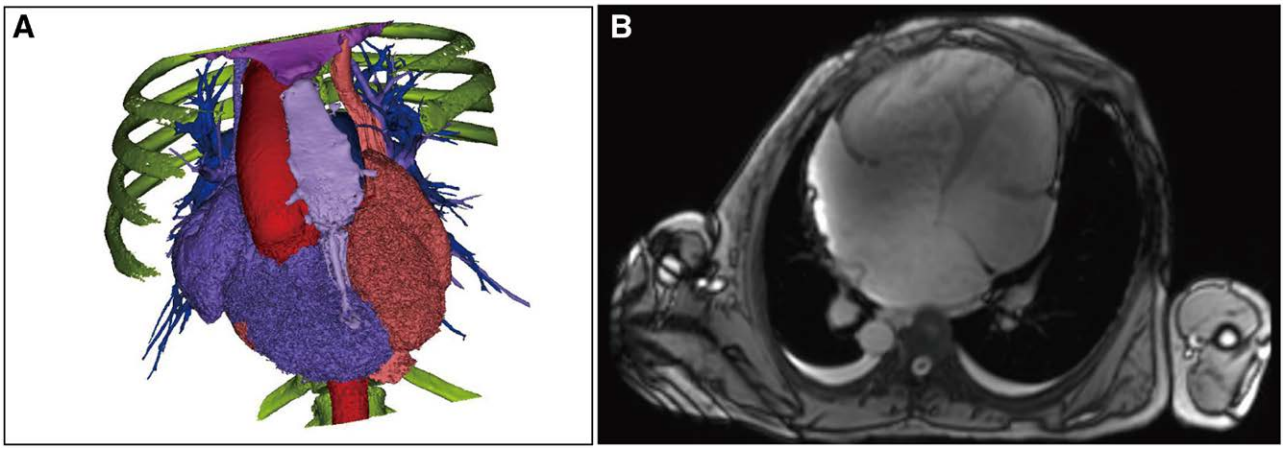

Figure 1. Preoperative imaging examination of the patient's heart. Three-dimensional cardiac reconstruction reveals the relative position of the heart within the thoracic cavity (**A**). Cardiac magnetic resonance imaging displays the sizes and positions of the heart's atrial and ventricular chambers (**B**).

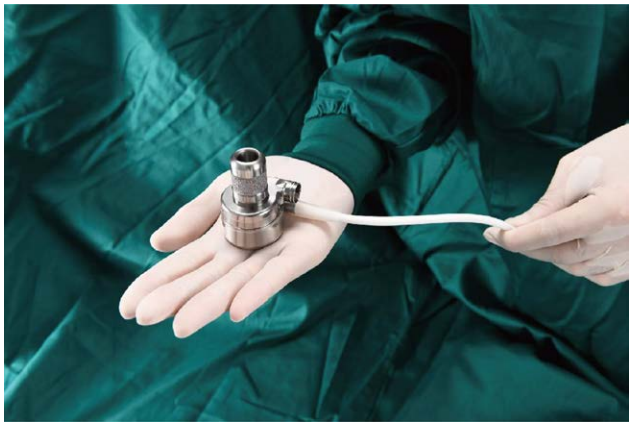

Figure 2. Photo of the Corheart 6 used in the operation.

the ascending aorta using a side-biting clamp and 5-0 running polypropylene sutures. Postoperatively, the patient required 48 hours of ventilatory support and stayed in the Intensive Care Unit (ICU) for 14 days. All inotropic drugs were discontinued within 20 days. Nitric oxide inhalation prevented right heart dysfunction. Continuous Renal Replacement Therapy (CRRT) improved renal function, and warfarin maintained an International Normalized Ratio (INR) of 2.0–2.5. Chest X-rays confirmed favorable thoracic conditions (Figure 3). There were no major complications, and the patient was discharged on day 39 with NYHA class I status.

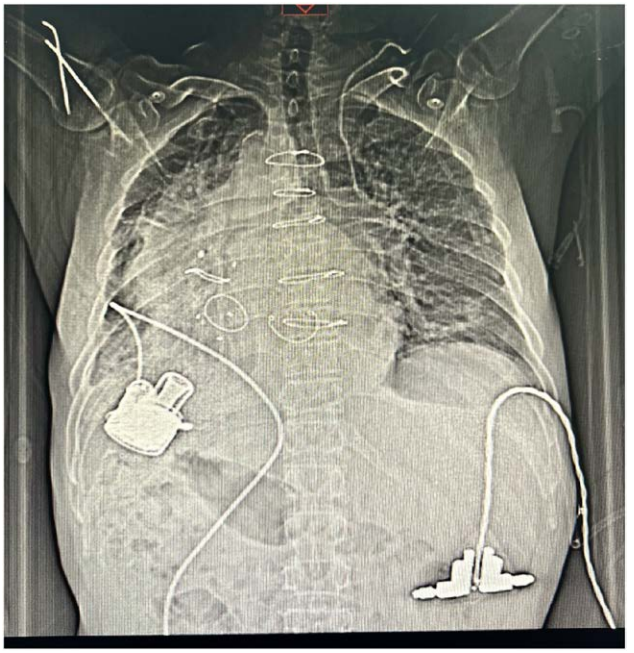

Figure 3. Postoperative chest X-rays showed the VAD pump head located in the right thoracic cavity, along with the replaced aortic valve, tricuspid valve, and the annuloplasty ring for the mitral valve. VAD, ventricular assist device.

Discussion

Patients with ccTGA and systemic RV dysfunction often undergo physiologic repair surgery, but their prognosis is poor. If a donor heart is unavailable and the patient's condition deteriorates, VAD implantation becomes the only option. Historically, VADs were used as a bridge to transplantation (BTT), but with recent advancements, they have become an acceptable treatment as destination therapy, especially for patients with pulmonary hypertension. Mechanical assist devices now primarily aim to reduce pulmonary hypertension, preventing late-stage right ventricular dysfunction and lowering pulmonary artery pressure. A study by Etz *et al.*[4] showed that continuous mechanical assistance can reverse pulmonary hypertension, making patients eligible for heart transplantation. In addition, pulmonary artery pressure did not significantly increase within 6 months post-transplant.[4]

Before 2018, about 25% of heart transplant candidates received a left VAD as a BTT, 25% as BTT, and around 50% as destination therapy. However, since the introduction of the new heart allocation system, less than 10% of implants were BTT, while over 70% were destination therapy in 2019, a trend expected to continue.[5] Ventricular assist device implantation in patients with systemic ventricular failure after palliated ccTGA

is supported by clinical experience but lacks strong evidence-based guidelines. The indications for using VAD as BTT or destination therapy in ccTGA patients with systemic ventricular failure are not fully established. The 2024 guidelines from the International Society for Heart and Lung Transplantation do not recommend heart transplantation alone for patients with severe irreversible pulmonary hypertension. However, combined heart-lung transplantation may be considered after a thorough evaluation. In addition, a pulmonary artery systolic pressure (PASP) ≥ 50 mm Hg, transpulmonary gradient (TPG) ≥15 mm Hg, or pulmonary vascular resistance (PVR) ≥3 Wood units, along with systolic blood pressure >85 mm Hg, is considered a relative contraindication to heart transplantation due to increased early mortality risk post-transplant.[6]

In this case, we implanted the VAD into the systemic right ventricle, and the main advantages can be reflected in three aspects: on the one hand, it directly produces ventricular assist in the systemic circulation; second, the anatomic left ventricle, which is stronger in itself, undertakes the pulmonary circulation, which can significantly reduce the likelihood of postoperative right heart failure. Last but not least, we used aggressive replacement surgery for tricuspid valve insufficiency with simultaneous clipping of the tendon cords and part of the papillary muscles, which significantly reduced the risk of sucking and the difficulty of localization that occurs after VAD implantation.

Currently, the surgical treatment of tricuspid regurgitation, whether isolated or concomitant, remains controversial. Tricuspid valve repair is the most commonly performed procedure due to its lower perioperative complication and mortality rates compared to valve replacement.[7] However, in the presence of annular dilatation or right ventricular enlargement, tricuspid valve replacement should be prioritized. Based on single-center experience, in the ccTGA patient population, the tricuspid valve, which carries the systemic circulation, is bound to regurgitate in the long term, leading to right ventricular failure. Therefore, we still chose tricuspid valve replacement in these patients.

Familiarity with ccTGA anatomy, morphology, and variations in the hemodynamic physiology is important when implantation of a VAD is being contemplated. In this operation, we inserted an inflow cannula to the right ventricular apex, and an outflow cannula to the ascending aorta. Transesophageal echocardiography (TEE) guidance ensured the successful selection of the optimal insertion site for the VAD inflow cannula. Any obstructive chordae might need to be sacrificed, but papillary muscles were preserved. The inflow cannula, unlike the usual arrangement, was placed to the right side of the outflow cannula, because the patient had dextrocardia. Therefore, the VAD pump was placed in a flipped-over position. However, this did not cause any problems. Three-dimensional reconstruction of preoperative chest computed tomography is the useful information for treatment strategies. In addition, patients with this condition have difficulty exposing the tricuspid valve, and it is recommended to undergo tricuspid valve replacement surgery through the left atrial approach. Considering the patient's apical rotation and right ventricular dysmorphic disorder, it is recommended that the pump inlet be positioned toward the diaphragmatic surface of the heart. Finally, it is recommended to place the drive cable on the right side of the patient's body.

We describe the use of Corheart 6 in a rare case of dextrocardia, ccTGA with atrial situs inversus (I, D, D) patient thus demonstrating the feasibility of its use as a bridge to heart transplantation or destination therapy. Ventricular assist device may serve this group of patients who are not immediately able to be transplanted due to donor supply shortage or hemodynamic instability. Implantation of VAD in these patients is a complex procedure that improves survival with a low rate of device-related complications and mortality. Preoperative three-dimensional computed tomography images and TEE guidance were useful to determine the inflow and pump positions. Recent advances in durable mechanical circulatory support have integrated advanced imaging and computer-assisted methods, enabling virtual cardiac implantation. This allows the prediction of hemodynamic effects and optimization of personalized perioperative management.[8]

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