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## HeartWare ventricular assist device placement in a patient with congenitally corrected transposition of the great arteries

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Congenitally corrected transposition of the great arteries (CC-TGA) is a rare anomaly that is seen in fewer than 0.5% of patients with clinically evident congenital heart disease. Failure of the systemic ventricle (morphologically right ventricle [RV]) is the main cause of heart failure.<sup>1</sup> Heart transplant is the treatment of choice for most patients with CC-TGA who are in end-stage heart failure. Ventricular assist devices (VADs) may become necessary to deal with progressive decline of end-organ function during the wait for an appropriate donor. Patients with CC-TGA present unique challenges for VAD implantation and management.

### CLINICAL SUMMARY

A 66-year-old man with a diagnosis of CC-TGA was seen at an outside hospital for worsening shortness of breath. He had shown slow progression to New York Heart Association functional class IV symptoms during the previous 8 years. His medical history was also significant for chronic atrial fibrillation and dyslipidemia. He was started on a continuous milrinone infusion of 0.3  $\mu\text{g}/(\text{kg} \cdot \text{min})$  and transferred to our hospital for possible VAD placement.

The patient's preoperative hemodynamic values with milrinone infusion were as follows: blood pressure, 110/65 mm Hg; heart rate, 90 beats/min; cardiac index,

1.65 L/(min  $\cdot$  m<sup>2</sup>), and pulmonary vascular resistance, 129 dynes/(s  $\cdot$  cm<sup>-5</sup>). After induction of general anesthesia, transesophageal echocardiography (TEE) was performed. The moderator band and the inferior insertion of the atrioventricular valve leaflets helped to identify the left-positioned ventricle as the morphologically RV and the associated valve as the tricuspid valve. The tricuspid valve was trileaflet, with a large prolapsed septal leaflet, a small anterior leaflet, and a cleft posterior leaflet. The tricuspid annulus was dilated to 49 mm, and color flow Doppler ultrasonography demonstrated severe tricuspid regurgitation. The ejection fraction of the morphologically RV was only 20%, with RV end-diastolic diameter of 8.7 cm. The pulmonary ventricle (morphologically left ventricle) function was nearly normal, with an ejection fraction of 50%.

A standard sternotomy was performed. The systemic aorta and right atrium were cannulated, and cardiopulmonary bypass was established. Because of concerns that the moderator band and multiple papillary muscles in the RV might obstruct the VAD inflow cannula, an 18-gauge angiocatheter was inserted into the RV, and agitated saline solution was injected to determine the optimal angle for the VAD inflow cannula. When the tip of the angiocatheter was found to be in the mid RV cavity without obvious obstruction, the sewing ring was sutured and ventriculotomy was then performed with the punch. On visual inspection, there were no significant crossing fibers. A HeartWare HVAD (HeartWare, Inc, Miramar, Fla) was then inserted with its center in the predetermined site. The outflow cannula was then anastomosed to the systemic aorta with a partially occluding aortic clamp. TEE verified that the VAD inflow cannula was positioned parallel to the interventricular septum without significant obstruction. Initially, a pump flow of 3.5 L/min was achieved after weaning off cardiopulmonary bypass. The device was positioned in the mediastinum, and the outflow graft was placed in the pericardial well (Figures 1 and 2).

Postoperatively, the patient did well hemodynamically and had good pump performance. On postoperative day

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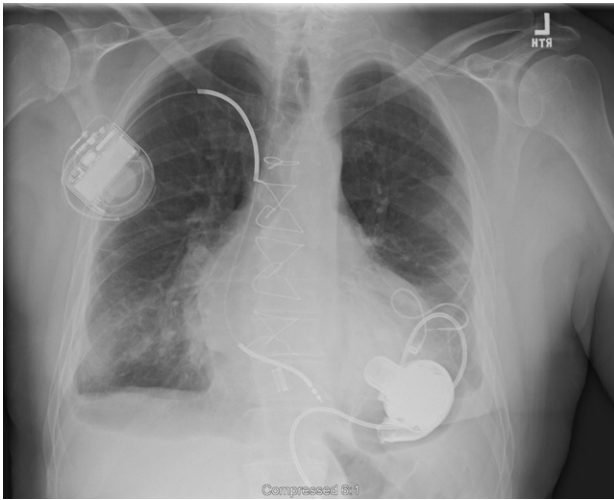
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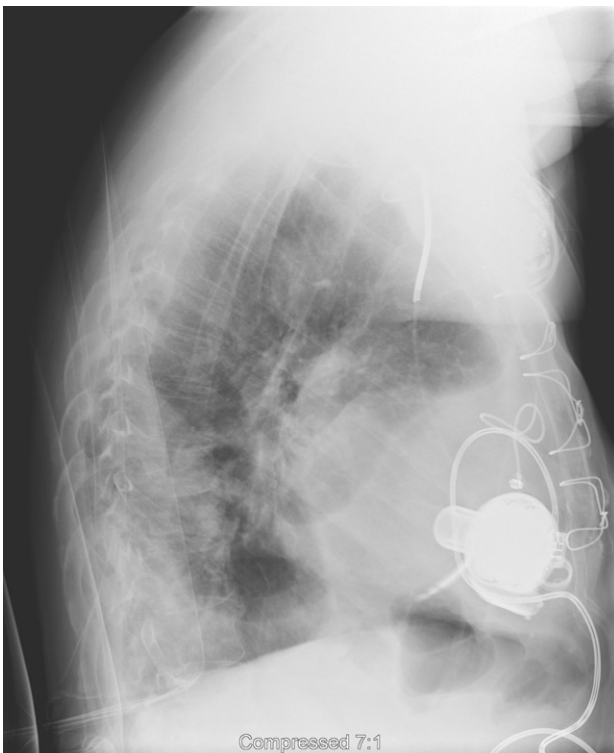
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**FIGURE 1.** Postoperative anteroposterior chest radiograph demonstrates positioning of the HeartWare device leftward in the mediastinum.

7, the patient was noted to have a sudden drop in the platelet count and discoloration of his digits. The patient was started on argatroban (GlaxoSmithKline, Philadelphia, Pa) with the diagnosis of heparin-induced thrombocytopenia. He was treated for several weeks with argatroban and was subsequently discharged to a rehabilitation facility. Ultimately, he did require amputation of several digits. After rehabilitation, the patient was discharged home with no



**FIGURE 2.** Postoperative lateral chest radiograph demonstrates insertion of the HeartWare inflow cannula posterior to the right ventricle.

further complications. Echocardiography 2 years after VAD insertion showed mild tricuspid regurgitation. At the time of this report, 24 months after VAD implantation, the patient remains fully supported by his device and has had no complications secondary to his congenital anatomic variation.

## DISCUSSION

CC-TGA refers to the abnormal looping pattern of the heart tube during embryologic development which results in discordance between the atrioventricular and ventriculoarterial connections. Only a minority of patients with CC-TGA survive into their seventh or eighth decade, and they are almost exclusively those who have no other associated anomalies.<sup>1</sup> Our patient was fortunate not to have any other anomalies other than a cleft tricuspid leaflet, which helps explain the delayed onset of symptoms.

Failure of the systemic ventricle (morphologically RV) is the norm and usually occurs earlier in life. Serial examinations of the ejection fraction of the morphologically RV are mandatory, along with careful evaluation of the tricuspid valve morphology and competency. TEE offers superior imaging resolution and windows to delineate cardiac chamber connections and associated anomalies.

Late referral for operative correction of CC-TGA is common among patients with a poor ejection fraction and is the major predictive marker for poor survival after surgery.<sup>2</sup> Options for our patient included heart transplant with home inotropic support, VAD, or total artificial heart. His worsening heart failure symptoms with milrinone infusion made waiting for heart transplant unacceptable. With a nearly normal pulmonary ventricle (morphologically LV), we anticipated that a total artificial heart would not be necessary and that a systemic VAD would be enough to improve the patient's function. Although about 10 patients have been reported in the literature with dextrotransposition of the great arteries and VAD support, only 3 cases have been reported of VAD placement in patients with CC-TGA.<sup>3,4,5</sup> HeartMate (Thoratec Corporation; Pleasanton, Calif) VAD systems were used in 2 cases. Because the systemic RV in CC-TGA lies posterior and inferior to the LV (pulmonary ventricle), although it is less posterior than a usual LV, this orientation might require unconventional positioning of VAD pumps. HeartMate VAD pumps had to be placed close to the midline, which led to abdominal wound dehiscence in 1 patient.<sup>3</sup> Joyce and colleagues<sup>5</sup> recently used the DeBakey VAD (MicroMed Technology, Houston, Tex) in a patient with CC-TGA.<sup>5</sup> They resected significant amounts of RV trabeculae, rotated the pump 180°, and placed the pump in a more posterior position to avoid inflow obstruction.

The HeartWare HVAD system is a small pump with a wide-blade rotor design and a magnetically suspended impeller that generates centrifugal blood flow. This design has

improved durability and performance relative to previous VADs. Our major rationale for choosing the HeartWare HVAD was its relatively small size and ability to be contained within the pericardium, thus eliminating the potential need to create a pocket or rotate the pump configuration. In addition, the device's integrated inflow cannula could decrease the chance of inflow obstruction related to the abnormal position of the ventricles in CC-TGA. TEE guidance ensured successful selection of the optimal insertion site for the VAD inflow cannula.

In summary, we report a case of successful HeartWare HVAD implantation in a patient with CC-TGA to support the function of the RV (systemic ventricle).

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## The role of the total artificial heart in the treatment of post-myocardial infarction ventricular septal defect

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Post-myocardial infarction ventricular septal defect (VSD) can be a fatal complication. Mechanical support to stabilize the patient until repair or transplant may be necessary, because emergency operative repair carries a mortality as high as 60%.<sup>1-3</sup> The CardioWest Total Artificial Heart (TAH-t; SynCardia Systems, Inc, Tucson, Ariz) has been used successfully to replace the failing heart as a bridge to transplant.<sup>4</sup> We report the cases of the first 2 patients treated with TAH-t implantation for unstable cardiogenic shock resulting from post-MI VSD rupture.

### CLINICAL SUMMARIES

#### Patient 1

A 68-year-old man was transferred to our institution in extremis after an anterior MI and left anterior descending stent 2 placement weeks previously. Evaluation by echocardiography showed a large pericardial effusion, a 22-mm apical VSD, and biventricular failure. The patient had deterioration to cardiogenic shock (systolic blood pressures less than 75

mm Hg, pulmonary arterial pressure 50/20 mm Hg, central venous pressure of 16-21 mm Hg, and pulmonary wedge pressure of 31 mm Hg) and was intubated and transferred to our institution on maximal pressor support. At arrival, emergency venoarterial extracorporeal membrane oxygenation (ECMO) was instituted through the femoral vessels, and the patient's condition was allowed to stabilize for 24 hours. The patient underwent surgery for possible repair and device support; however, because of extensive myocardial damage and poor function, the decision was made intraoperatively for TAH-t placement. The patient was extubated within 24 hours and supported for 76 days until heart transplant. He had an uncomplicated hospital course and was discharged 2 weeks after transplant. At 3 years of follow-up, the patient remains in excellent condition.

#### Patient 2

A 65-year-old woman with a medical history significant for diabetes mellitus type 2, hypertension, hyperlipidemia, and scleroderma was admitted to outside hospital for an acute inferior MI. She was taken for emergency cardiac catheterization and had 2 stents placed in the proximal and mid right dominant coronary artery. On the next day, she became dyspneic and hypotensive. Physical evaluation noted a new heart murmur, and echocardiography revealed a large inferior base VSD of 14 mm with biventricular failure. The patient was intubated, an intra-aortic balloon pump was placed, and pressor support was maximized. The patient's condition continued to deteriorate, and she was transferred to our

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