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Bridge to transplantation with long-term mechanical assist device in adults after the Mustard procedure

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KEYWORDS:
congenital heart disease;
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bridge to transplant;
VAD;
systemic right ventricle

BACKGROUND: There is limited clinical experience with bridging to transplant with a left ventricular assist device (VAD) in patients with previously palliated transposition of great arteries.
METHODS: Five adult patients presenting with systemic right ventricular failure 30 years after a Mustard operation were implanted with a HeartMate II VAD. The implant was completed using standard procedures with only minor modifications to accommodate right ventricular cannulation.
RESULTS: All 5 patients were men, with a mean age of 31.5 ± 1.8 years and a median time since Mustard operation of 30 (range 28 to 32) years. All patients had sternal closure on Post-operative Day (POD) 1, and 2 patients required additional re-operation for bleeding. One patient required temporary support of the non-systemic ventricle. The mean duration of VAD support was 284 ± 177 days; 3 patients underwent heart transplant and 2 died on PODs 502 and 34, respectively. Both deaths were due to progressive heart failure and pump thrombosis. Comorbidities, anatomy and mediastinal scarring did not preclude implantation and heart failure symptoms improved in all patients.
CONCLUSIONS: With the increased prevalence of late post-Mustard heart failure, bridge to transplant with a VAD may be a suitable treatment option for patients who are severely ill.
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Transposition of the great arteries (TGA) is a congenital cardiac malformation in which the aorta arises from the right ventricle and the pulmonary artery arises from the left ventricle, resulting in ventriculoarterial discordance. Corrective cardiac surgery is usually necessary in infancy to assure survival beyond the first year of life; almost 90% of these patients survive to adulthood.[1] Before to the arterial switch operation became standard care in the mid-1980s, corrective surgery was often completed by an atrial switch procedure (Mustard or Senning operation), which leaves the morphologic right ventricle to provide the systemic circulation. As patients age, the systemic right ventricular function deteriorates until symptomatic heart failure ensues. Approximately one third of all patients with a systemic right ventricle will develop heart failure by 3 to 4 decades after their atrial switch operation and, for those who are symptomatic, the 1-year mortality rate is nearly 50%.[2,3] Accordingly, as this population of patients enters the era of highest risk for heart failure, there are increasing numbers of patients presenting with severe symptoms requiring heart transplantation or mechanical circulatory support. Due to the

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congenital cardiac defects and previous surgery decades earlier, these patients present significant management challenges.

Clinical experience and the number of published reports on the application of mechanical circulatory support in adults with systemic right ventricles are limited.[4–7] We report 5 cases of adult patients with TGA 3 decades after their Mustard procedure who developed severe heart failure requiring heart transplant and mechanical circulatory support.

Methods

Adult patients with the initial diagnosis of TGA treated by atrial switch operation (Mustard procedure) who presented with severe heart failure necessitating mechanical circulatory support as a bridge to transplant were included in the analysis. Five patients meeting these criteria were identified and had received VAD implantation between August 7, 2008 and July 18, 2012. Medical records were retrospectively reviewed to obtain demographic and peri-operative data. Specific data regarding pre-operative status, operative data (including cardiopulmonary bypass time, blood loss and other procedures), post-operative complications, length of hospitalization and outcome were recorded.

Device implantation

The HeartMate II left ventricular assist device (VAD; Thoratec Corporation, Pleasanton, CA) was implanted in all 5 patients through a median sternotomy and using femoral vessel cannulation for cardiopulmonary bypass. The right ventricle was cored with a coring knife on the anterior surface in a location that would centralize the inflow cannula within the ventricle. Trabeculations and the moderator band were excised to avoid obstruction of the inflow cannula. The apical cuff was sewn over the opening in the ventricle, the inflow was inserted and secured, and the pump was positioned medially within the abdomen. The outflow graft was sewn to the ascending aorta and the drive-line was externalized in the usual manner. The operation was completed and typical post-operative care was provided according to standard practices.

Results

The indication for VAD support was bridge to transplant for all 5 patients. The demographics and history for the group are given in Table 1. All 5 patients were men and their mean age was 31.5 ± 1.8 years. The median time since the Mustard operation was 30 (range 28 to 32) years. Two patients had a history of neurologic events: Patient 1 had a stroke 3 years before implant and Patient 2 had a transient ischemic attack (TIA) 2 years earlier. The 2 patients with renal insufficiency (Patients 2 and 3) had received dialysis, but renal function was adequate at the time of implant. Three patients were Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS) Profile II, and 1 patient each presented with Profiles III and I; all patients were receiving inotropic support (Table 2).

The implant operation was completed without major technical difficulties, with adequate pump flow and hemodynamics at VAD start-up (Table 3). The final pump position was similar to that in implants in patients without TGA, with the inflow through the left hemidiaphragm and the outflow graft just to the right of the mid-line (Figures 1 and 2). All patients had extensive mediastinal adhesions, and 2 (Patients 4 and 5) underwent second re-operations in the early post-operative period. The average blood loss during implant for the 5 patients was 5,946 ml (range 1,660 to 8,810 ml). One patient required temporary support of the non-systemic ventricle. The mean duration of VAD support was 284 ± 177 days; 3 patients underwent heart transplantation and 2 died on Days 502 and 34, respectively.

Case summaries

Patient 1

The first patient was a 29-year-old male who presented in cardiogenic shock (INTERMACS Profile I) and was not responsive to medical therapy. After VAD implantation, the patient could not be weaned from cardiopulmonary bypass due to poor non-systemic (left) ventricular function. Temporary ventricular support was provided with a CentriMag ventricular assist device (Thoratec). Cannulation for CentriMag support was through the inferior vena cava for the pump inflow, and the outflow via a cannula placed in the main pulmonary artery. After 2 days of support, the CentriMag was weaned and removed. He was discharged from the hospital on Post-operative Day (POD) 36. Seven months after VAD implantation, the patient had a drive-line infection that was treated with antibiotics, surgical debridement and application of a vacuum-assisted wound-closure system (V.A.C. Therapy; Kinetic Concepts, Inc., San Antonio, TX). He then underwent successful heart transplant after 341 days of VAD support.

Table 1 Patients' Demographics and History

Patient number	Age (years)	Gender	BSA (m^2)	Time since Mustard (years)	Diabetes	Hypertension	Renal insufficiency	Stroke
1	29	Male	1.71	28	N	N	N	Y
2	31	Male	2.17	29	N	N	Y	Y (TIA)
3	33	Male	1.81	32	N	Y	Y	N
4	31	Male	2.01	30	N	N	N	N
5	33	Male	1.99	32	N	N	N	N

BSA, body surface area; TIA, transient ischemic attack.