Title: Total Artificial Heart Implantation in a Young Marfan Syndrome Patient

Running Head: Total Artificial Heart Marfan Syndrome

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Abstract

Introduction: Cardiovascular complications represent the leading cause of morbidity and mortality in patients with Marfan Syndrome (MFS). Here we describe a unique case where a total artificial heart was implanted in a young MFS woman.

Methods: A 22 year-old postpartum African American female with MFS developed multiple severe valve dysfunction and biventricular failure that was refractory to medical management. She previously had a Bentall procedure for Type A aortic dissection and repair of a Type B dissection.

Results: We implanted a total artificial heart with a good outcome.

Conclusions: TAH is a durable option for severe biventricular failure and multiple valvular dysfunction as a bridge to transplant in a young patient with MFS.

Keywords: Total Artificial Heart, Marfan Syndrome, Bridge to Transplant

Background

Cardiovascular complications represent the leading cause of morbidity and mortality in patients with Marfan Syndrome (MFS). Women with MFS have an increased risk of accelerated aortic dilation and rupture in pregnancy [1]. Other cardiac complications associated with MFS include aortic aneurysms, mitral valve disease, aortic valve insufficiency (AI), arrhythmias and cardiomyopathy. Here we describe a unique case of implanting a total artificial heart (TAH, Syncardia ™, Tucson, AZ) in a patient with MFS.

Case Presentation

A 22 year old African American female with MFS presented to the emergency department with one week of congestive symptoms consistent with heart failure. Following the birth of her first child ten months previously, she had developed a Type A aortic dissection and severe aortic insufficiency that was repaired with a Bentall procedure using a Gelweave Valsalva™ aortic root graft (Vascutek™,Inchinnan, Scotland) and a 25mm Edwards pericardial tissue valve. She then developed a Type B dissection requiring repair with a 20mm Hemashield tube graft (Maquet™, Rastatt,Germany) as well as cardiac arrest secondary to ventricular tachycardia with subsequent EF of 10-15% requiring an implantable cardioverter-defibrillator. On this hospital admission, she became hypotensive and was started on norepinephrine and dobutamine for decompensated biventricular heart failure. Ten days later, she was inotrope dependent and had refractory liver congestion not responsive to diuresis. Echocardiography showed severe biventricular dilation with a LVEDD of 92mm, EF of 10-15%, severely reduced right ventricular function, a severely dilated right atrium, severe mitral and tricuspid regurgitation. Right heart catheterization while on milrinone showed elevated left and right heart filling pressures, and mild pulmonary hypertension (pulmonary artery pressure 40/28mmHg; pulmonary capillary wedge pressure 24mmHg; pulmonary vascular resistance 1.47 Wood units). Given that she was a good transplant candidate with severe valve incompetence, biventricular failure and had a previous median sternotomy, a TAH was preferred over implanting an LVAD with a temporary RVAD. Additionally, she had a high PRA (Class 1 96%, Class 2 17%), so it was felt that a durable biventricular device was best option as a bridge to transplant. Based on the fit test, we knew that a 70cc TAH (Figure 1) would be appropriate.

Treatment

We performed a re-do median sternotomy, initiated cardiopulmonary bypass and crossclamped the aorta. We proceeded to a partial cardiectomy and fashioned atrial cuffs 3 mm below the mitral and tricuspid valves. After opening the aortic root, we explanted the old bioprosthetic valve and sewed the inflow and outflow grafts of the TAH to the left atrium, right atrium, pulmonary artery and aorta respectively. The aortic outflow graft was beveled and sewn to the Dacron graft of the prior Bentall conduit in the ascending aorta (Figure 2). We covered the TAH with an expanded polytetrafluoroethylene membrane. The overall bypass and cross clamp times were 241 min and 209 min respectively.

Outcome and follow-up

On post-operative day (POD) one she developed tonic-clonic seizures. She was diagnosed with status epilepticus and loaded with phenytoin, lacosamide, and levetiracetam. A CT head demonstrated an evolving posterior temporal lobe infarct, which was due to de-airing of the TAH. Seizures were controlled by POD 8. On POD 10 she developed low filling pressures, elevated central venous pressure and increasing pressor requirement. A half-liter of serosanguinous fluid was collected in the right chest tube over 45 minutes, and the patient became severely acidotic, with hemoglobin of 3.5 u/L. We performed a repeat median sterntomy and found her in tamponade from a mediastinal hematoma. The source of bleeding was found to be a branch of the right coronary artery in the ventricular muscle which was promptly ligated. Following this, her CVP, flow rates, lactate, and systolic blood pressure all improved.

The patient's neurological status also improved, and she was able to ambulate with physical therapy by POD 31, and was discharged home on POD 60. She is doing well one year post-implant, and awaiting a heart on the transplant list.

Discussion and Conclusions

Aortic root dilatation with aortic insufficiency and aortic dissection often precedes multiple valvular disease presentations in MFS patients. Concomitant with aortic insufficiency, mitral and tricuspid regurgitation are often observed in MFS patients [2]. Orthotopic heart transplantation (OHT) in MFS patients results in good long term survival similar to those without MFS [3]. Aortic rupture following OHT is a common complication, and some surgeons advocate aggressive treatment with complete aortic arch replacement during OHT. Our patient had an ascending and descending aortic repair before, therefore further aortic intervention was not necessary. Due to the scarcity of organs, OHT is not always possible. A single report shows successful LVAD placement as a bridge to transplant in a MFS patient [4]. Here, we report successful TAH implantation in a patient with MFS. We believe this is a good, durable option in the setting of severe, refractory biventricular CHF and multiple valvular dysfunction as a bridge to transplant in a young patient.

List of Abbreviations

TAH: Total artificial heart

MFS: Marfan Syndrome

AI: Aortic insufficiency

LVAD: Left ventricular assist device

POD: Post-operative day

OHT: Orthotropic heart transplant

Declarations

Ethics approval and consent to participate

Single case report, ethics approval not applicable

Consent for publication

Consent to publish obtained from patient

Availability of data and material

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing interests

Sources of funding

No sources of funding to declare

Conflicts of Interest

No conflicts of interest to declare for any author

Authors' contributions

All authors contributed substantially to the work. P.R performed critical research and wrote portions of the manuscript. J.K. interpreted data, researched, and wrote portions of the manuscript. T.R. interpreted data and wrote portions of the manuscript. S.K. wrote portions of the manuscript, revised, and interpreted data. R.S. contributed to the figures and interpretation of the fit models. O.A contributed to the revision of the manuscript. Z.K. revised and contributed critically important intellectual content. All authors discussed the results and implications and commented on the manuscript at all stages.

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Figure 1

Virtual fit imaging done with a mock 70cc TAH prior to surgery showing the left and right ventricular components. Transverse (a), Coronal (b), and Sagittal (c) views show adequate space thin the ventricular cavities.

Figure 2

Preoperative 3D CT reconstruction (a) and postoperative A/P and Lateral chest x-ray with TAH placement (b,c).