Biventricular assist device use in non-dilated hypertrophic cardiomyopathy

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Received 19 February 2014; received in revised form 24 April 2014; accepted 2 May 2014

Abstract

Advanced heart failure is a rare but important complication of hypertrophic cardiomyopathy (HCM). The only definitive treatment is heart transplantation and the role of ventricular assist devices remains uncertain. We describe the use of implantable biventricular assist devices in the treatment of a patient with ‘end-stage’ non-dilated HCM.

Keywords: Hypertrophic cardiomyopathy • Circulatory support devices

INTRODUCTION

Heart failure in hypertrophic cardiomyopathy (HCM) is characterized by diastolic dysfunction. This may progress to systolic dysfunction, culminating in ‘end-stage’ HCM. Heterogeneous remodelling patterns mean that some cases exhibit a non-dilated ventricular morphology and persistence of marked hypertrophy [1]. We describe such a case treated with implantable biventricular assist device (BiVAD).

PATIENTS AND METHODS

A 24-year old man with non-dilated HCM presented with decompensated heart failure after 18 months on the transplant waiting list without a suitable donor. Transthoracic echocardiography showed gross biatrial dilatation and hypertrophied ventricles with small cavities (Fig. 1). Right heart catheterization revealed elevated right atrial and pulmonary capillary wedge pressures (28 and 20 mmHg, respectively) with reduced cardiac index (1.4 l·min⁻¹·m⁻²). Urgent listing yielded no suitable donor, and he deteriorated to INTERMACS Profile 2 despite intravenous inotropic and diuretic therapy. Therefore, the decision was made to bridge him to transplantation using mechanical circulatory support (MCS).

Following median sternotomy and normothermic cardiopulmonary bypass (CPB), two HeartWare HVADs (HeartWare, Inc., Framingham, MA, USA) were implanted in a BiVAD configuration. Both atrial appendages were grossly enlarged and tense with thrombus; these were stapled off and excised.

The inflow sewing ring was secured to the left ventricular (LV) apex. A core of myocardium >2 cm thick was excised with the supplied coring knife, revealing an extremely restricted LV cavity. An aortic cross-clamp was applied, and the heart was arrested with 1.0 litre of antegrade cold blood cardioplegia. A circumferential endocardial myomectomy was carried out from the LV apex coring site towards the base of the papillary muscles in order to enlarge the LV cavity (Fig. 1). The inflow cannula was inserted after extensive endocardial myomectomy, the heart and left-sided HVAD (LVAD) were de-aired, and the aortic cross-clamp was removed. The outflow graft was anastomosed to the ascending aorta. Given the tiny right ventricular (RV) cavity, the right atrium was chosen for right-sided HVAD (RVAD) inflow placement. The pleura was widely opened and the inflow sewing ring was secured to a...
‘sandwich’ of pericardium and right atrium just anterior to the phrenic nerve. A crucifix incision was made across the composite tissue ‘sandwich’ inside the sewing ring, the RVAD inflow cannula was inserted half-length and locked in position. The device was de-aired, and the outflow graft was anastomosed to the pulmonary artery. The RVAD was separated from the lung with an ePTFE membrane. The patient was weaned off CPB, and the HVADs were activated sequentially. Initial flows of 4.0–5.0 l·min⁻¹ were achieved. CPB time was 221 min. Coagulopathy necessitated delayed sternal closure.

Initial progress was good but complicated by left-sided haemothorax on Day 18 requiring thoracotomy for evacuation. The patient was discharged home on Day 29. BiVAD support of >6.0 l·min⁻¹ was achieved, with reduction of RAP to 6–8 mmHg and no suck-down events.

DISCUSSION

Although transplantation remains the best therapy for medically refractory end-stage biventricular heart failure, MCS therapy offers a bridge-to-transplant possibility. Established durable MCS options include the SynCardia TAH (SynCardia Systems, Inc., Tucson, AZ, USA), and BiVAD with Thoratec PVAD or IVAD (Thoratec Corporation, Pleasanton, CA, USA). However, these pulsatile devices have relatively large portable drivers that restrict patient activity. Newer third-generation implantable continuous-flow pumps have been used as BiVADs, and have the advantages of silent operation, no pump pocket requirement and wearable battery packs [2].

The nature of HCM in our case required careful MCS selection. The small ventricular cavities and predominantly non-compliant RV meant that an LVAD alone would probably have been insufficient, being limited by poor RV diastolic filling, and hence the need for BiVAD support.

The TAH was deemed unsuitable given the patient’s small native ventricles and their excision would not have provided sufficient space to accommodate the artificial ventricles of a TAH. In HCM, VADs are usually contraindicated because of the high risk of inflow cannula obstruction through sudden collapse of the ventricle (suck-down events), impairing pump function and increasing the risk of ventricular arrhythmia. Therefore, in order to safely accommodate the LVAD inflow cannula, extensive LV myomectomy was required [3].

Use of the HeartWare HVAD for RV support is currently off-label, with no published experience in non-dilated HCM. Although there are reports describing RV inflow cannulation for RVAD, this approach was unsuitable in the described case, given the exceptionally hypertrophied RV with virtually no cavity. Instead, the grossly dilated right atrium was chosen for the RVAD inflow (Fig. 2) [2]. The potential pitfall of right atrial inflow placement is that of displacement due to collapse of the atrial wall under the weight of the RVAD. By placing the RVAD extra-pericardially just anterior to the lung hilum, the pericardium provided support for the weight of the RVAD, thereby ensuring stable inflow cannula orientation. Furthermore, the inflow cannula was only inserted halfway into the atrial cavity to minimize the risk of contact with and occlusion by the interatrial septum. The composite pericardium/atrial ‘sandwich’ in the centre of the sewing ring was incised, but not excised, thereby leaving a gasket around the inflow cannula.

Figure 2: Postoperative chest radiograph.

We believe that it is unnecessary to restrict the diameter of the RVAD outflow graft since the VAD output is inflow-dependent. With RVAD and LVAD speeds of 2400 and 3200 rpm, respectively, resultant BiVAD flows ranged 5.8–6.8 l·min⁻¹. The LVAD was deliberately set at a much higher rpm than that of the RVAD, partly because of the higher impedance of the systemic circulation, and partly to ensure that the LVAD had adequate flow reserve to offload the left-sided cardiac chambers for whatever flow rate was delivered by the RVAD.

CONCLUSION

We report a novel procedure for the implantation of a BiVAD in a patient with small biventricular cavities. This case demonstrates the feasibility of BiVAD support in the normally contraindicated setting of non-dilated HCM.

ACKNOWLEDGEMENTS

We thank Catherine Sudarshan for her expert assistance during the implant operation.

Conflict of interest: Steven S.L. Tsui is an investigator, a consultant and a proctor for HeartWare, Inc.

REFERENCES