Two-year outcome of ventricular assist device via a modified left atrium to aorta approach in cardiac amyloidosis

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Abstract

Cardiac amyloidosis is a debilitating disease associated with poor long-term survival. Medical or palliative treatment is the usual course of therapy, but patients are often intolerant of conventional heart failure treatment. The current standard of care of sequential heart and bone marrow transplant is usually not feasible for ill or frail patients or in countries with limited organ donors or without transplant programmes. Left ventricular assist devices (LVAD) are not usually offered to these patients due to high peri-operative risks and risks of suction events with the LVAD in a small left ventricle. We report the 2 year outcome and discuss the challenges faced in the management of our patient with end-stage heart failure due to cardiac amyloidosis, who was successfully supported with an LVAD using a modified left atrium to aorta implantation technique.

Keywords LVAD; Ventricular assist device; Cardiac amyloidosis; Amyloid cardiomyopathy; Daratumumab; Heart failure

Case report

A 46-year-old female presented with fluid overload. Echocardiogram showed moderate biventricular hypertrophy, moderately impaired left ventricular ejection fraction (LVEF) and small left ventricular (LV) cavity size of 3.5 cm in diastole. Tricuspid annular plane excursion (TAPSE) was 0.8 cm. She also had severe tricuspid regurgitation. Serum high-sensitive troponin T was 279 pg/mL and NT-proBNP 24 800 pg/mL. Work-up confirmed the diagnosis of Cardiac Amyloidosis caused by AL amyloidosis. Her expected survival based on Revised Mayo Clinic criteria was 6 months.1

She responded poorly to medical therapy. She experienced recurrent heart failure with right pleural effusion and bilateral leg oedema and developed acute kidney injury and symptomatic hypotension even with low doses of diuretics. She was not a heart transplant candidate due to her poor pre-morbid frailty and active systemic amyloidosis. She was counselled on palliative therapy but opted to undergo a high-risk left ventricular assist device (LVAD) implantation.

Due to the small left ventricular cavity size, we implanted the LVAD to provide support via the left atrium using a modified technique. Through a right atriotomy, a conduit fashioned out of a 20 mm Gore-Tex® (WL Gore & Associates, Arizona, United States of America [USA]) interposition graft was implanted with one end sewn into an atrial septal defect that was created, and the other end sewn to the anterior right atrial wall with the sewing ring of the LVAD. The HeartWare™ HVAD™ (HeartWare International, Massachusetts, USA) inflow cannula was then inserted into the Gore-Tex® graft on the right atrial wall and secured to the sewing ring (Figure 1). The outflow graft was trimmed and anastomosed to the ascending aorta. The posterior tricuspid leaflet was closed off by stitching between the adjacent commissures. The left atrial appendage was not closed. At the end of the procedure, the HVAD was anchored to the right side of the rib cage before the chest was closed (Figures 2 and 3). Post-operatively, the patient was nursed in the intensive care unit (ICU) and extubated on the second post-operative day (POD). She was transferred out of the ICU and high dependency unit on
POD7 and discharged from hospital on POD 21. Her short-term outcome at 3 months was reported in 2019.²

**Discussion**

We are reporting the 2 year outcome of our patient. She is New York Heart Association functional class I and has been on LVAD support for more than 750 days. She works as a schoolteacher and did not experience any heart failure hospitalization since LVAD implantation.

A major concern with implanting LVADs in patients with end-stage RCM is the risk of “suck-down” events due to a small LV. This modified approach of trans-septal cannulation technique to decompress the left atrium where the atrial septum is cannulated via the right atrium, previously described by Maeda et al in paediatric and adult patients.
with hypertrophic and restrictive cardiomyopathy, was useful in helping us avoid suction events in our patient with cardiac amyloidosis.\textsuperscript{3,4}

The HVAD pump was initially set at 2600 rpm with a pump power of 3.2 Watts and pump flow of 3.1 L/min. The waveform on the HVAD monitor showed an atrial waveform (Figure 4). However, 6 months later, pump flow reduced to 2.8 L/min. After evaluation for causes of low flow such as dehydration, inflow cannula and outflow graft obstruction, pump speed was increased to progressively to 2840 rpm. Despite increasing her daily oral fluid intake, pump speed continued to drop steadily over the next 12 months despite...
an increase in pump speed up to 3100 rpm. Pump power remained stable at 3.2 W. The cause of the progressive drop in pump flow was attributed to the competing flow from the recovering left ventricle. Left heart function had improved after LVAD implantation, from LVEF of 28% immediately post-LVAD implantation to 55% at 12 months. The aortic valve remained closed post-operatively. With the improvement in left ventricular function and reduction in pump flow, the aortic valve eventually opened with every beat. Pump flow subsequently dropped to and stabilized at 1.7 L/min at 2 years. The patient remained asymptomatic and physically active throughout. Her 6 min walk test was 440 m at 2 years.

An episode of right heart failure immediately post-operatively was treated with intravenous levosimendan, diuretics and judicious fluid management. After hospital discharge, she did not experience right heart failure for the 2 years on LVAD support. There was only trivial tricuspid regurgitation post-LVAD implantation. TAPSE remained at 0.8 cm post-operatively. In addition to an improvement in the LV systolic function, there was also a significant reduction in LV wall thickness from 1.3 cm before LVAD implantation to 0.9 cm at 2 years post-LVAD implantation. LV diameter was unchanged at 3.5 cm prior to LVAD implantation, 2.7 cm immediately after LVAD implantation, and 3.4 cm at 2 years. The indexed left atrium volume was 33 mL/m² before LVAD implantation and 30 mL/m² at 2 years post LVAD. NT-proBNP and high-sensitive troponin T levels dropped from 24 800 and 279 pg/mL respectively before LVAD implantation to 55% at 2 years post LVAD implantation. Blood culture was positive for Candida. CT scan did not show any collection or abscess. She was treated with intravenous micafungin for 2 weeks before she was changed to long term suppressive therapy with oral fluconazole. She did not have any other LVAD-related infections.

The current standard of care for patients with cardiac amyloidosis secondary to AL amyloidosis is sequential heart and autologous bone marrow transplantation. However, our patient was ineligible for multi-organ transplant in view of her frailty and renal impairment prior to the LVAD. She continued novel targeted therapy (bortezomib and daratumumab) for AL amyloidosis and eventually achieved complete and sustained haematological remission after 1 year on LVAD support. She was maintained on daratumumab once a month with the aim of completing a 2 year maintenance regime. Her kidney function normalized post-LVAD implantation and eGFR was 80 mL/min/1.73 m² at 2 years.

She has declined heart transplantation listing despite medical advice as she felt well with LVAD support and was reluctant to undergo another major surgery. Despite being counselled that myocardial recovery was unlikely and not previously reported, she remained hopeful that there might be sufficient heart function recovery to eventually allow for the LVAD to be explanted. Nevertheless, with a long national heart transplant waiting list, the chances of getting a donor heart would be low. The favourable long-term outcome of our patient on LVAD support offers another therapeutic option in the treatment of patients with advanced heart failure from cardiac amyloidosis.

To our knowledge, this is the first reported long-term follow-up of a left atrium-to-aorta configuration LVAD support in amyloid cardiomyopathy with a small left ventricle. This case demonstrates that LVAD is a feasible and potentially life-saving therapy for patients with end-stage cardiac amyloidosis.

**Conflict of interest**

None declared.

**Table 1** The list of cardiac medications that our patient was prescribed before and after LVAD implantation

| Before LVAD implantation | Medications on discharge | Medications at 1 year | Medications at 2 years |
|--------------------------|--------------------------|-----------------------|-----------------------|
| Ivabradine 5 mg BD       | Spironolactone 25 mg OD  | Spironolactone 25 mg OD | Spironolactone 25 mg OD |
| Spironolactone 25 mg OD  |                         |                       | Sidenafl 25 mg TDS     |
| Frusemide 40 mg OD       | Frusemide 40 mg BD       |                       | Sidenafl 25 mg TDS     |
| Frusemide 40 mg (titrated according to symptoms and renal function) | Potassium Chloride 1000 mg BD |                       | Valsartan 40 mg BD     |
|                          | Aspirin 100 mg OD        |                       | Warfarin 1 mg OD       |
|                          | Valsartan 40 mg OD       |                       |                       |
|                          | Warfarin 1 mg OD         |                       |                       |

BD, twice daily; OD, once daily; TDS, three times daily.
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