

# Mechanical Support in Children with Restrictive Left Ventricle – Single Centre Experience

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## Background

- Patients with restrictive left ventricle (rLV) are generally considered poor candidates for mechanical circulatory support.
- There are currently limited data on the efficacy of a ventricular assist device (VAD) in paediatric patients with rLV.

## Aim

- To examine outcomes of children with rLV supported by EXCOR Berlin Heart (BH) VAD.

## Methods

- All children with rLV who received EXCOR BH at a single centre between 2008 and 2017 were included in the study.
- rLV was defined as a non-compliant left ventricle (LV) with atrial enlargement, encompassing LV non-compaction (n=3), mixed (n=3) and restrictive (n=6) cardiomyopathies. Pre-implantation, all patients were INTERMACS profile 1 or 2.
- Patient demographics, echocardiogram and cardiac catheterisation data, EXCOR BH type and duration, cannulation strategy, and clinical outcome were obtained from our database.
- Data points were represented as a median value, and qualitative data were expressed as percentages.

## Disclosures

- The authors have no conflict of interest to disclose.

## Results

- A total of 12 patients were included (Table 1).
- Median age at VAD implantation was 3.8 years, and median weight was 13.3kg.
- Left ventricular assist device (LVAD) was used in 7 patients, with the remaining 5 receiving biventricular assist device (BiVAD) support.
- The first 4 consecutive patients were cannulated to LV apex with 50% survival to transplant.
- Left atrium (LA) cannulation was used more recently; the survival of patients who were cannulated to LA was 67%.
- Overall 4 patients died, and the cause of death was pump malfunction leading to cerebral thromboembolism in all.
- 5 patients had elevated pulmonary vascular resistance index (PVRI) >6WU pre-VAD implantation (median PVRI 8.9WU, range 6.1-10.6WU) and in these, VAD was used to optimise their pulmonary vasculature pre-transplant.
- In 1 patient who was deemed non-transplantable (PVRI 10.6WU), LVAD served as bridge to candidacy; PVRI reduced to 4.9WU after 8 weeks on LVAD, allowing successful transplantation (Figure 1).

Table 1: Summary of patient characteristics, device and cannulation, EXCOR BH course and outcome.

Patient	Age (y)	Sex	Weight (kg)	Diagnosis	VAD Type	Cannulation Strategy	VAD Support (days)	Outcome
1	3.9	F	13.5	LV non-compaction	LVAD	LV	225	Transplant
2	6.8	F	15.1	Restrictive cardiomyopathy	BiVAD	LV	38	Death
3	6.8	M	14.8	Mixed cardiomyopathy	BiVAD	LV	53	Transplant
4	2.6	F	10.0	Restrictive cardiomyopathy	BiVAD	LV	6	Death
5	2.8	F	9.6	LV non-compaction	LVAD	LA	130	Transplant
6	3.2	F	10.0	Restrictive cardiomyopathy	LVAD	LA	147	Transplant
7	4.8	F	13.0	Restrictive cardiomyopathy	BiVAD	LA	2	Death
8	3.8	F	9.7	Mixed cardiomyopathy	LVAD	LV	97	Transplant
9	2.0	F	8.8	LV non-compaction	LVAD	LA	230	Transplant
10	4.1	M	13.9	Mixed cardiomyopathy	LVAD	LV	66	Transplant
11	3.4	M	14.5	Restrictive cardiomyopathy	BiVAD	LA	140	Transplant
12	10.5	F	34.0	Restrictive cardiomyopathy	LVAD	LA	16	Death

Figure 1: EXCOR BH as bridge to candidacy in a child with rLV and severe pulmonary hypertension.

### Case Report

- A 2-year-old girl with a brief history of breathing difficulties and sweating was diagnosed with end-stage heart failure due to LV non-compaction.
- Of note, her mother also had LV non-compaction.
- Pre-transplant investigations including hybrid magnetic resonance imaging (MRI)/cardiac catheterisation revealed elevated pulmonary pressure with baseline PVRI 10.6WU, dropping to 4.9WU with inhaled nitric oxide and 100% oxygen.
- Additionally, MRI identified left main bronchus compression by enlarged left atria.
- She was stabilised on diuretics, milrinone and high-flow oxygen, and referred to our unit for LVAD as bridge to candidacy.
- LVAD was implanted on 12/12/2016 and her post-implantation course was complicated by transient right heart dysfunction, which was controlled with inotropes.
- Repeat cardiac catheterisation was performed after 8 weeks of LVAD support, demonstrating a reduction of baseline PVRI to 4.9WU and 3.4WU, respectively.
- She was listed for transplantation and was successfully transplanted after a total of 230 days on LVAD.

## Conclusions

- Children with rLV constitute a high-risk population for mechanical circulatory support.
- LA cannulation may yield improved outcomes in comparison to LV apical cannulation in rLV.
- Left heart decompression on VAD can halt progression of pulmonary hypertension, allowing bridge to either recovery or transplant.