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Ventricular Assist Device Use In Pediatric Restrictive And Hypertrophic Cardiomyopathy: An Advanced Cardiac Therapies Improving Outcomes Network (ACTION) Analysis

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Abstract:
Purpose Use of ventricular assist devices (VAD) in patients with restrictive (RCM) and hypertrophic cardiomyopathy (HCM) remains low compared to use in dilated cardiomyopathy. We describe the outcomes of patients with RCM and HCM supported by VAD and characteristics associated with successful VAD support.
Methods We examined the Advanced Cardiac Therapies Improving Outcomes Network (ACTION) registry for outcomes of patients with RCM, HCM and mixed HCM/RCM who underwent VAD implantation from March 2012 - July 2021, including major adverse events, and survival.
Results Fourteen patients were identified and represent 2.2% of the ACTION registry at data collection. There were 9 initial LVAD (with 2 subsequent RVAD) and 5 initial BIVAD (1 total artificial heart). Of 12 LVAD patients with data, 4 had LA and 8 had LV cannulation. Median age was 3.4 years (range 0.3-16.9), weight 15 kg (5.2-57.2), and BSA 0.64 m2 (0.29-1.67). Diagnoses included RCM (50%), HCM (35.7%), and mixed RCM/HCM (14.3%). Illness severity at implant was high with 29% Intermacs profile 1, 50% on ECMO, 57.1% on ventilator support, 85.7% on 1 or more inotrope, and 64.3% on TPN. Twenty devices were used in 14 patients: 10 (50%) paracorporeal pulsatile, 6 (30%) paracorporeal continuous, 3 (15%) implantable continuous, and 1 (5%) intracorporeal pulsatile. Median duration of support was 80 days (8-412). Four patients died on device, 9 transplanted and 1 alive on device. Though not statistically significant, patients who died had higher incidence of pre-VAD: ECMO, TPN, Intermacs profile 1, HCM, initial BIVAD and major VAD-related adverse events. Cause of death was inadequate support (1, HCM with Rotaflow BiVAD), infection (2), and multi-organ failure (1).
Conclusion Patients with restrictive and hypertrophic cardiomyopathy have high pre-implant illness severity, however, most patients had successful VAD outcomes. VAD support is a reasonable strategy for select RCM and HCM patients.

	Transplanted + Alive on Device *(n = 10)*	Died *(n = 4)*	p-value
Diagnosis			
RCM (plus HCM/RCM)	8 (80%)	1 (25%)	*p >0.05*
HCM	2 (20%)	3 (75%)	
Device days, median [range]	89 [22 – 412]	63.5 [8 -83]	*p >0.05*
INTERMACS Profile			
1	2 (20%)	2 (50%)	*p >0.05*
2	6 (6%)	2 (50%)	
ECMO (pre VAD)	4 (40%)	3 (75%)	*p >0.05*
Mechanical Ventilation (Pre VAD)	6 (60%)	2 (50%)	*p >0.05*
TPN (Pre VAD)	5 (50%)	4 (100%)	*p >0.05*
Inotropes (Pre VAD)	8 (80%)	4 (100%)	*p >0.05*
Multiple Inotropes (Pre VAD)	2 (20%)	1 (25%)	*p >0.05*
BiVAD	4 (40%)	2 (50%)	*p >0.05*
All Devices			
Intracorporeal	2 (15.3%)	2 (40%)	*p >0.05*
Paracorporeal	11 (84.6%)	3 (60%)	
Continuous	4 (36.3%)	3 (60%)	*p >0.05*
Pulsatile	7 (63.6%)	2 (40%)	
Adverse Event Rate (%)			
Major Bleeding	20%	50%	*p >0.05*
Major Infection - Sepsis	30%	50%	*p >0.05*
Neurologic Dysfunction			
Extra-axial Hemorrhage	0%	50%	*p >0.05*
Ischemic Stroke	10%	0%	*p >0.05*
Renal Dysfunction	10%	25%	*p >0.05*

Author Disclosure Information:

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