Successful management of severe pulmonary hypertension in a boy with complex congenital heart disease prior to heart transplant: Two-step approach

Background: Severe pulmonary hypertension (PHT) has been traditionally viewed as an absolute contraindication for a heart transplant, that often necessitates combined heart and lung transplant.

Case description: A 16-year-old boy with severe PHT and cardiogenic shock was referred to the advanced heart failure service at the University of Nebraska Medical Center for consideration of implantable ventricular assist device (VAD) as destination therapy (DT). The patient was diagnosed at birth with Shone's complex, ventricular septal defect, patent foramen ovale and large patent ductus arteriosus. After having 5 heart surgeries between the age of 8 days and 12 years, he developed high-grade AV block requiring epicardial dual-chamber pacemaker during last surgery. Over the following 4 years, he was getting progressively short of breath with drop in his LVEF to about 10% in 8/2019 when he presented with cardiogenic shock. After initial medical stabilization requiring IV milrinone support, he became inotropic dependent. Right heart catheterization (RHC) showed severe PHT (Table 1) with markedly elevated pulmonary vascular resistance (PVR), but with good response to nitroprusside challenge (mean PA pressure improved to 49 mmHg with drop in PVR to 6.1 Wood units). Cardiac CT-derived 3-D printing of his heart was performed to find the DT-VAD type that suits his body size.

HeartWare VAD (HVAD) was then successfully implanted, with significant improvement in PHT (Table 1). He was then listed for heart transplant as status 1A, to receive successful orthotropic heart transplant in 11/2019. Post-transplant residual PHT and RV failure was managed successfully with Sildenafil and Macitentan in addition to diuresis and IV Milrinone to successfully wean epoprostenol on post-op day #7. He was discharged home on post-op day # 17 on Sildenafil 40 mg TID and Macitentan 10 mg daily. On post-op day #34, Macitentan was stopped with complete weaning of Sildenafil over the following month. Repeat RHC at 3 months post-heart transplant continued to show normal PA pressure and PVR, without need for PHT therapies up till now.

Conclusion: Severe PHT can be completely reversed in select cases. We describe a 2-step approach with the use of a VAD followed by medical therapy for residual PHT following heart transplant, with complete resolution of PHT.

Table 1: Right heart catheterization data.

	Pre-LVAD 8/8/2019	Post-LVAD 8/29/19	Post-LVAD-2 11/14/19	Post-HT-1 12/4/19	Post-HT-2 2/24/2020
PA pressure (mean)	80/55 (65)	76/25 (44)	57/17 (33)	40/24 (31)	39/18 (27)
PCWP	26	15	17	22	14
CO / CI (Fick's)	2.9/1.9	2.7/1.8	4.4/2.7	4.6/2.9	5.2/3.2
PVR	13.4	10.7	3.6	1.97	2.5