

Hybrid Use of Pulmonary Artery Pressure with Simultaneous 3-Dimensional Echocardiography to Evaluate Right Ventricular Function in Pediatric Pulmonary Hypertension

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Background:

Pulmonary hypertension (PH) is a severe, progressive disease requiring multiple treatments. We used simultaneous CardioMEMS PA pressure and 3DE of RV to assess ventricular arterial coupling in pediatric PH patients over time.

Methods:

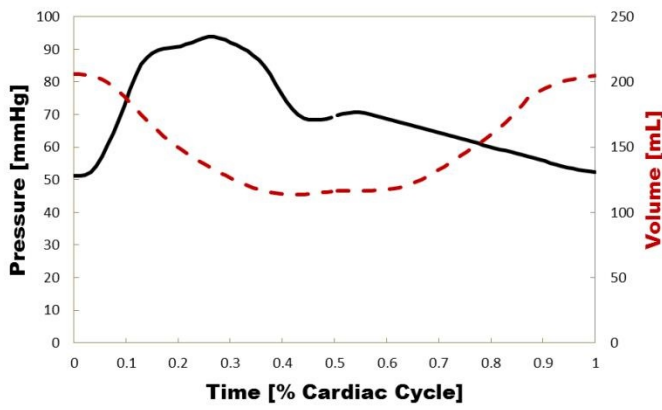
We present a case series of two patients with PH after getting a CardioMEMS device. Patient-1 has idiopathic PH. Patient-2 has PH associated with d-transposition of the great arteries after arterial switch. Both patients had group 1 PH and were managed on tadalafil, ambrisentan, and treprostinil. Lung transplantation was not an alternative for either patient. Patient-1 had six datasets of 3DE with PA pressure; Patient-2 had three datasets. RV volumes were analyzed by 4D RV Function (TomTec). Custom MATLAB code (MathWorks) was used to merge with a RV systolic pressure-volume (PV) loop. The arterial elastance (Ea) and end-systolic elastance (Ees) were generated from the PV loop (**Figure**). Ventricular arterial coupling was measured by Ees/Ea ratio and compared to a reference of 0.68, lower limit of RV energetic reserve.

Results:

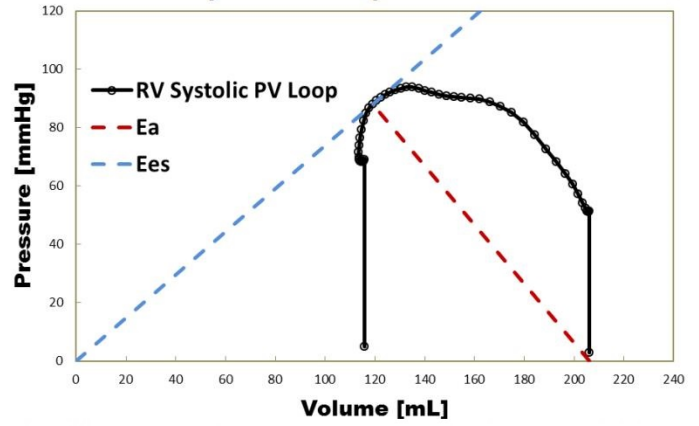
In these patients, PA pressure was used to guide the reverse Potts shunt (RPS) placement. In Patient-1, RV contractility and PA afterload decreased after the RPS, but returned near baseline after. Ees/Ea was above the threshold during the observation, indicating RV energetic reserve. In Patient-2, Ees/Ea was below the threshold during the observation, showing RV dysfunction and RV-PA uncoupling.

Conclusions: Our cases reveal the successful use of CardioMEMS for real-time PA pressure readings without repeat invasive cardiac catheterization. Measuring ventricular arterial coupling may offer insight pathophysiology of PH and guide medical management.

Avg. Pressure & Avg. Volume Waveform



RV Systolic PV loop with Ea and Ees



Patient-1	HR bpm	EDP mmHg	ESP mmHg	MPA mmHg	EDV ml	ESV ml	SV ml	EF %	Ea mmHg/ml	Ees mmHg/ml	Ees/Ea
Pre-placement	65.4	46.1	87.4	62.3	215	100	115	53.5	0.760	0.874	1.150
Post-placement within 5 days	75.5	51.0	93.1	68.5	206.1	113.5	92.5	44.9	1.006	0.820	0.815
Post-placement 1 month	69.6	54.1	101.4	73.6	179.7	88.4	90.9	50.6	1.115	1.141	1.023
Post-placement 6 month	57.8	52.2	101.4	71.6	172.2	84.8	87.3	50.7	1.161	1.195	1.030
Post-placement 1 year	76.6	60.8	111.4	80.8	155.7	78.3	77.4	49.7	1.439	1.423	0.990
Patient-2											
Pre-placement	69.8	39.6	103.8	66.8	433	269.1	163.9	37.9	0.633	0.386	0.609
Post-placement within 5 days	77.8	42.2	97.7	65.0	398.6	244.7	153.9	38.6	0.635	0.399	0.629
Post-placement 1 month	67.5	51.2	120.8	75.5	434.6	263.7	170.9	39.3	0.707	0.458	0.648