Anesthetic Management of a Neonate with Mixed Persistent Pulmonary Hypertension and Severe Pulmonic Valve Stenosis

G DiSilvio, L Donatelli, C Mesia, N Johansson, P Taneja, R Schwartz
Department of Anesthesiology, St. Christopher’s Hospital for Children

Introduction:
- Persistent pulmonary hypertension of the newborn (PPHN) is a complex syndrome characterized by sustained elevation of pulmonary vascular resistance leading to extrapulmonary R → L shunting across persistent fetal channels and hypoxemia.
- The Incidence is ~ 2/1000 live born infants.
- Echocardiography remains the gold standard for diagnosis and is essential to rule out cyanotic congenital heart disease prior to the initiation of treatment, but it is not without limitations.
- Here we describe a case of a unique physiologic combination in which severe PPHN delayed the diagnosis of concurrent severe PV stenosis by standard 2D echo.

Case Presentation:
- Full term male neonate was transferred to our facility at 4 hrs of life due to hypoxemia with marked cyanosis, minimally responsive to 100% O₂ via ET w/ mechanical ventilation. ABG on arrival: 7.31/47/39/23/1.1 and SpO₂ of 71% on SIMV 40, 30/8, 100%.
- Echo demonstrated: Dilated RV, mod TR that estimated a RV systolic pressure of ~ 98 mmHg (UA 45/32 mmHg). PV appeared thickened w/o evidence of obstruction, large PDA w/ R → L shunting, a stretched PFO w/ R → L shunting.
- Medical management included: HFOV, NO, milrinone, vasopressin, and multiple fluid/ blood product boluses.
- DOL 5, pt’s oxygenation and hemodynamics began to stabilize but he was developing progressive anasarca and low UO.
- Repeat echo revealed: Persistent RV dilation with mod RVH, stable RV systolic pressure at ~95 mmHg (UA 50/25 mmHg).
- Progressively increased obstruction across a thickened PV with peak instantaneous systolic gradients increasing up to 56 mmHg. R → L shunting across the PFO and now L → R shunting across the PDA. At this time, the patient was diagnosed with a mixed physiology of improving PPHN and severe valvar PS based on clinical and echo data.
- DOL 6, pt went to cardiac cath for balloon pulmonary valvuloplasty. A TIVA w/ fentanyl (2-4 mcg/kg/hr), lorazepam (0.05-.1 mcg/kg/hr) and pancuronium (1-2 mg/kg) was chosen to provide the most stable hemodynamic conditions. NO was cont’d at 20 ppm.
- Initial RV pressure 95/0 mmHg (descending aorta 32/30 mmHg) w/ a 50 mmHg peak gradient across the PV. Post balloon pulmonary valvuloplasty, RV pressure decreased to 60/0 mmHg (descending aorta 58/34 mmHg) w/ a 15 mmHg residual gradient across the PV.
- Pt remained hemodynamically stable throughout the procedure and he was taken to the CCU where he made a quick recovery.
- Pt was D/C’d ~2 weeks after the procedure. F/U echo on POD 22 revealed slight RV dilatation, BIV hypertrophy and a PDA.
- F/U echo at 33 weeks showed: PV doming w/o significant stenosis, 9.3 mmHg peak gradient and mod PR

Discussion:
- Severe PPHN may mask the diagnosis of valvar PS in the neonate presenting with hypoxemia.
- Here, significantly decreased flow across the PV due to extreme pulmonary pressures made it impossible to accurately measure a peak gradient across the valve.
- Unaware of such an unusual combination, elevated RV pressures with TR was readily attributed to the PPHN alone.
- Severe valvar PS was only identifiable once pulmonary pressures were controlled and R → L shunting across PFO persisted despite shunt reversal across PDA and PV peak gradients were measurable.
- A fentanyl, lorazepam and pancuronium TIVA provided optimal hemodynamic stability for pulmonary valvuloplasty in a neonate with both severe PPHN and PS.

Reference: