BLOATED, BRADYCARDIC, AND BLUE

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History and physical:

1-month-old ex-31 week female with complex medical history including congenital complete heart block, hydrops at 23 weeks, multiple muscular VSDs, dysplastic pulmonary valve with progressive pulmonary stenosis, significant anasarca requiring ongoing peritoneal drainage, respiratory failure requiring prolonged mechanical ventilation on HFOV, recent NEC, and large PDA refractory to ibuprofen x 2. She had worsening anasarca despite octreotide. After multidisciplinary conference, the decision was made to proceed with cardiac catheterization for balloon pulmonary valvuloplasty and possible device closure of PDA in an attempt to stabilize her physiology. She was not an ECMO or surgical candidate due to severity of her illness.

Physical exam showed critically-ill infant, sedated and paralyzed on conventional ventilator with profound anasarca, diminished breath sounds bilaterally, bradycardia with III/VI systolic murmur along the LSB, 1+ distal pulses in all extremities, tense, edematous, discolored, shiny abdomen that was not compressible.

Indication for Intervention:

Severe pulmonary stenosis with hemodynamically significant PDA

Intervention:

Baseline hemodynamics showed elevated CVP with RV EDP 17, suprasystemic RV pressures (130% systemic pressure) with an 85 mmHg gradient across the pulmonary valve. Right ventricular angiography showed a dysplastic pulmonary valve that measured 7.6 mm x 9.1 mm. Balloon pulmonary valvuloplasty was performed using a 9, 10, and 11 mm x 2 cm Tyshak mini balloons. During pulmonary valvuloplasty the patient had worsened compliance on the ventilator and decreased cerebral and flank NIRs to the 30s. There was a residual 55 mm Hg gradient across the pulmonary valve. Angiograms were performed in the descending aorta using the Glidecath that showed a very large type C patent ductus arteriosus that measured 7.4 mm on the pulmonary artery end and 5.1 mm on the aortic end. The length of the duct was approximately 12.4 mm. The PDA was temporarily occluded with an MVP-7Q to see if this improved her hemodynamic state. There was rapid, significant improvement in her hemodynamic status with ductal occlusion. An angiogram was performed through the Glidecath in the MPA to assess branch pulmonary artery flow in relation to the PDA device. The device was not stable to release due to the size and compliance of the PDA. An MVP-9Q device was advanced through a 5-Fr Glidecath and positioned in the patent ductus arteriosus. Transthoracic echocardiogram was performed that showed no significant obstruction to aortic flow. The patient's lung compliance improved with the device in position and the NIRS improved by greater than 20 points. An angiogram was performed in the main pulmonary artery using the Glidecath that showed no obstruction of flow to the branch pulmonary arteries. The device was released and remained in stable position. Repeat transthoracic echocardiogram showed

stable device position. The patient was taken back to the NICU for stabilization with plan to reattempt pulmonary valvuloplasty after device was secure. Despite initial improvement the first 24-48 hr post-procedure, she again developed worsened ascites and the decision was made by the family to withdraw care 5 days post-procedure.

Learning Points of the Procedure:

Timing of intervention for critically-ill infants with multisystem disease, use of unconventional devices for unconventional PDAs, managing complex physiology