[NM-149] A Patient with Pulmonary Atresia –VSD & Eisenmenger Physiology for Scoliosis Surgery- How sick is too sick?

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INTRODUCTION: As long term survival after cardiac surgery improves, more palliated patients present for surgery. Spinal fusion is a high-risk procedure that may stabilize cardiopulmonary function. To our knowledge, this is the first report of a patient with Eisenmenger physiology successfully undergoing spinal fusion.

REPORT: AR is a 14 yo with DiGeorge sequence, pulmonary atresia and fenestrated VSD scheduled for T4-L5 posterior fusion for scoliosis (Cobb angle 800). Her cardiac status includes suprasystemic RV pressure, severe pulmonary HTN and SVT. Meds include digoxin and sildenafil. Functionally, she is a NYHA Class II. PFTs showed moderate restrictive disease. Standard monitoring was established. BIS and NIRS were also utilized. After IV access, an epinephrine infusion was established. Induction included ketamine, etomidate, midazolam and sufentanil. Airway was topicalized before intubation. A CVL and arterial line were placed. Maintenance included isoflurane, dexmedetomidine and midazolam, with sufentanil and methadone analgesia. Milrinone, iNO, and tranexamic acid were empirically initiated. Neuromonitoring was utilized. Additional cares included SCD's, and warming blanket. A baseline TEE was obtained, and the probe left in place. Surgical duration was 4.5 hours, with 400cc EBL. The pt transferred to the ICU intubated . The iNO was weaned and patient extubated on POD #2. Milrinone and epinephrine were weaned by POD #3. Additional therapies included continuation of SCD's, aspirin, NG administration of home medications . TPN was given until oral intake resumed. Discharge home was on POD 12. There were no medical or surgical morbidities at 3 month followup.

DISCUSSION:
Scoliosis is found in 2-3% of the general population; the incidence increases to 9% -34% in patients with CHD. 1-2. Surgical correction reduces spine curvature, and may stabilize cardiopulmonary function. Functional improvement is modest at best. Acutely, there can be significant deterioration of pulmonary function. Patients with pulmonary hypertension are known to be at particularly high risk of perioperative morbidity; however surgical and medical colleagues are not always aware of this data. Taggert et al recently reported a 56% complication rate in patients with pulmonary hypertension undergoing PSF. These authors further commented that Eisenmenger physiology is likely associated with even higher risk, and cited one death in an adult who underwent PSF. In our case, AR had been stable medically, except for scoliosis progression. Her cardiologist estimated another decade of life, provided there was no further deterioration in pulmonary status. However, he was not initially aware of the surgical risk. It is critical as we care for these complex patients that there be care conferences to evaluate therapy options. Anesthesiologists are uniquely poised to assist in evaluating the perioperative risk. Successful outcomes in cases such as this should be disseminated, as they may inform decision making for others.

1.Taggart NW et al. Outcomes of Spinal Fusion in Children With Congenital Heart Disease. J Pediatr Orthop 2010; 30: 670-675.
2.Reckles LN et al. The association of scoliosis and congenital heart defects. J Bone Joint Surg Am. 1975; 57:449–455.
