

# Anesthesia for a 14 year old Patient with untreated Tricuspid Atresia, scheduled for coil embolization of a cerebral aneurysm.

**AUTHORS:** Jenny Dolan, MD, Michael Rossi, MD, Norman Halliday, MD, Douglas Miller, MD.

**AFFILIATION:** Department of Anesthesiology, University of Miami, Miami, FL 33136.

**Introduction:** Tricuspid Atresia (TA) is an uncommon congenital heart defect accounting for 1-3% of all congenital heart lesions [1]. TA is characterized by an absent tricuspid valve and a hypoplastic right ventricle, with associated defects necessary for survival such as VSD, ASD or PDA. Over 60% of infants die before age 1 year and only 10% survive to age 10 years without intervention [2]. In developed nations, surgical intervention is usually undertaken at an early age. Here we report our experience with a 14 year old from Bolivia with untreated Tricuspid Atresia for coil embolization of a cerebral aneurysm.

**Case Report:** In addition to TA she also had a Ventricular Septal Defect (VSD). Sixth months before, she had been hospitalized in Bolivia for a left Subarachnoid Hemorrhage, at that time an angiogram revealed a left ophthalmic aneurysm, and she was transferred to our institution for treatment. It was decided to treat the aneurysm before the TA as the anticoagulation necessary for cardiopulmonary bypass may result in a cerebral bleed. On physical examination she was noted to be very small, (25 kg), frail, profoundly cyanotic, SpO<sub>2</sub> 60% at rest and with minimal exertion (e.g. eating or speaking) her SpO<sub>2</sub> would decrease to 40%, unable to ambulate, and constantly in the fetal position. Her Hgb/Hct was 17g/dl/51%.

Auscultation revealed a single S1 with no murmurs.

The evening before coil embolization an esmolol infusion was started to control heart rate. On the morning of the procedure her vital signs were BP 90/60, HR 70 bpm, SpO<sub>2</sub> of 60% (on RA) and RR 18. Following preoxygenation intravenous induction was performed with etomidate 0.3 mg/kg, lidocaine 1mg/kg, fentanyl 0.5 mcg/kg and cisatracurium 0.15mg/kg. The plan was to preserve adequate peripheral vascular resistance and promote adequate pulmonary blood flow. During induction a Phenylephrine infusion was started at 2 mcg/kg /minute and it was titrated to maintain an adequate mean arterial pressure throughout the case, the Esmolol infusion was maintained. Following induction an arterial catheter was placed. Arterial blood gas analysis revealed combined respiratory and metabolic acidosis. Ventilation was optimized, additional IV fluids and sodium bicarbonate were administered. General anesthesia was maintained with sevoflurane, and fentanyl. Despite minimal blood loss her hematocrit decreased to 46%, and her saturation decreased despite 100% FIO<sub>2</sub>. The patient was transfused with the intention of increasing the oxygen carrying capacity to an HCT over 50's with good response. The aneurysm was successfully embolized. After adequate reversal of neuromuscular blockade, she was extubated and transported to the Pediatric Intensive Care Unit, where she arrived with stable vital signs. The post-operative course was uncomplicated.

Approximately two months after the neurological procedure, the patient underwent surgical palliation of her cardiac lesion. She had a superior vena cava to right pulmonary artery shunt placed. Post-operatively, she progressed well, and with physical therapy markedly improved her functional capabilities. Her postoperative SpO<sub>2</sub> was over 80%.

**Discussion:** Unpalliated tricuspid atresia in a patient of this age is an extremely rare condition. Presentation of an arteriovenous malformation at such an early age is also somewhat uncommon. The anesthetic goals for the management of this patient presented important potential areas of conflict. First, in a patient with obstructed pulmonary blood flow, adequate systemic vascular resistance as well as lower pulmonary vascular resistance must be maintained in order to preserve adequate oxygenation. Generally, the intraoperative management of an AVM calls for relative hypotension. This patient clearly

would not have tolerated this strategy. Furthermore patients with cerebral aneurisms sometimes present with cerebral vasospasm, requiring hemodilution. In view of our patient's extreme cyanosis, and need for high hemocrit to maintain sufficient oxygen carrying capacity, this would have been imprudent. Despite the advanced stage of progression of her cardiac lesion, and the rarity of the constellation of her conditions, both were amenable to intervention. Following these procedures, her quality of life was dramatically improved.

**References:**

1. Pediatric Cardiology for Practitioners. Myung K. Park. 4<sup>th</sup> Edition. P 207.
2. Pediatric Cardiac Anesthesia. Carol L. Lake, Peter D. Booker. 4<sup>th</sup> Edition. Tricuspid Atresia Susan Tebich P 436.