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Introduction: Vascular rings are a group of congenital anomalies of the aortic arch and great vessels. These rings can either be complete or incomplete structures that surround the trachea and esophagus resulting in respiratory and gastrointestinal compromise (1). Complete vascular rings include double aortic arch and right aortic arch with an irregular left subclavian artery and left ductus arteriosus. Incomplete vascular rings include pulmonary artery sling, innominate artery compression, and irregular right subclavian artery (2). Clinical symptoms can vary greatly from being asymptomatic to critical airway compromise depending on the type of ring. Children with vascular rings commonly have associated anomalies usually cardiac in nature, such as a VSD, TOF, aortic coarctation, and PDA. Common non-cardiac anomalies include TEF, cleft lip-palate, subglottic stenosis, DiGeorge syndrome, Down syndrome, and CHARGE syndrome(3).

Case: A 6 week old, Ex-40 week, 3.4kg, critically-ill infant presented for cardiac catheterization and PDA transcatheter closure. History included a fever of unknown origin, acute renal failure, a large PDA and increasing oxygen requirements. Echocardiogram revealed significant pulmonary hypertension with an estimated right ventricular systolic pressure of 69 mmHg. Past surgery included G-tube placement at an outside hospital and parents reported multiple intubation attempts with post-operative stridor. Airway exam was otherwise normal. Difficult airway equipment was made immediately available including a videolaryngoscope and appropriate sized LMA's. An IV induction was performed and with his history of pulmonary hypertension a deep plane of anesthesia was achieved prior to airway manipulation. Direct laryngoscopy revealed a grade 2 view and both a 3.0 cuffed and a 2.5 cuffed ETT were unable to be passed. A 2.5 uncuffed tube was then placed successfully but without a leak. The ETT was left in place to avoid further airway manipulation in a remote location. Catheterization revealed aortic coarctation and an extremely large PDA, unsuitable for transcatheter closure. Subsequently, during open PDA ligation a variant right subclavian artery creating a vascular ring was found explaining his difficult intubation. This aberrancy was not apparent during catheterization due to the other cardiac anomalies that impeded catheterization views.

Discussion: As illustrated by this case, vascular rings are challenging to diagnose. Patients with this vascular malformation may go undiagnosed and are at risk for significant airway compromise (3). Difficulties with both ventilation and intubation are possible and airway management should be similar to that of an anterior mediastinal mass. In conclusion, vascular rings present challenges in airway management for the pediatric anesthesiologist, therefore it is important to have a high index of clinical suspicion for vascular rings.

1) Shah R.K, et al. International J Ped Otorhinolaryngology 2007; 71: 57-62

2) Davies M, Br J Rad 2003; 76:491

3) Humphrey C, Pediatrics 2006: 117:903-908
