

## **PBLD – Table #5**

### **A 9 month old with tracheal obstruction requiring ECMO cannulation**

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#### **Objectives:**

1. The Learner will be able to discuss the anatomy, presentation, and preoperative assessment of vascular rings
2. The Learner will be able to describe the anesthetic and surgical management of patients with vascular rings
3. The Learner will be able to describe the perioperative issues that can arise from repair of vascular rings
4. The Learner will be able to describe how these complications can impact future anesthetic encounters

#### **Case history:**

A 9 month old, 7 kg male born with a left pulmonary artery sling and a resultant long segment tracheal stenosis. Both were repaired at 6 months of age, but due to recurrent tracheal obstruction and dehiscence, he had multiple revision procedures on his trachea. He now presents for airway evaluation of severe recurrent tracheal obstruction and potential ECMO cannulation.

#### **Questions:**

What are the different types of vascular rings? What is long-segment tracheal stenosis? What are the typical clinical presentations in children with vascular rings? What are the critical aspects of their evaluation and diagnosis? What is a slide tracheoplasty?

#### **Case history and physical examination (continued):**

In the weeks following his initial slide tracheoplasty, he had multiple revision procedures secondary to his tracheal dehiscence. He also had multiple respiratory arrests secondary to recurrent airway obstruction requiring emergent microlaryngoscopy, bronchoscopy, and laser fulguration.

For this procedure, the infant arrives to the operating room with a 24 gauge intravenous line in place and with a saturation of 95% on one-liter high flow nasal

cannula. His respiratory rate is 34 breaths/min and he has an audible inspiratory stridor and significant retractions.

**Questions:**

How do you prepare the operating room for the patient's arrival? What are some of the key elements of the discussion that should be had with the Otolaryngologist? Cardiac surgeon?

**Case history (continued):**

It is decided that the best approach is to electively place the infant on ECMO as a bridge to a definitive tracheal repair in a few days. The otolaryngologist would like to perform a rigid bronchoscopy to evaluate the vocal cords, trachea and to secure the airway prior to proceeding with ECMO cannulation.

**Questions:**

What are your options for induction of anesthesia in this patient? What are your options for maintaining anesthesia while rigid bronchoscopy is being performed? In a patient who has had multiple respiratory arrests in the preceding week, would it be wiser to perform ECMO cannulation prior to manipulating the airway? How feasible is it to place an infant on ECMO without securing an airway first?

**Intraoperative Care:**

A titrated induction of anesthesia is begun with a combination of intravenous ketamine and propofol infusions with the intent to maintain spontaneous ventilation during rigid bronchoscopy. Rigid bronchoscopy shows poor visualization of the carina and trachea due to extensive granulation tissue and continued dehiscence of the prior tracheoplasty. Soon after inserting the rigid bronchoscope into the trachea, inability to ventilate through the side port of the rigid bronchoscope with a subsequent hypoxic and bradycardic event ensue. The bronchoscope is removed and mask ventilation is resumed with an oral airway which results in an improvement of vital signs. Rigid bronchoscopy is again attempted but again results in extremely poor ventilation (both spontaneous and controlled).

**Questions:**

Why do you think there is such difficulty ventilating through the side port of the bronchoscope? The otolaryngologist has difficulty visualizing the trachea due to severe stenosis and does not think it is safe to pass an endotracheal tube through the stenosis/possible dehiscence. What are your options at this point?

**Intraoperative Care:**

Due to the challenges faced with rigid bronchoscopy and the potential for loss of the infant's airway with repeated airway instrumentation, the decision was made to place a laryngeal mask airway and proceed with ECMO cannulation.

**Questions:**

Which vessels should be cannulated for ECMO in a 9 month old infant? What is the difference between VV ECMO and VA ECMO? When is VV ECMO indicated? VA ECMO?

**Intraoperative Care:**

Veno-arterial ECMO cannulation is performed through the right neck without complications while the patient is maintained with assisted spontaneous ventilation via the LMA. Following cannulation, a 4.0 uncuffed endotracheal tube was placed just below the vocal cords but above the level of the stenosis by the otolaryngologist and the infant was transported to the pediatric intensive care unit.

**Discussion:**

A vascular ring is a congenital defect where the aorta or its branches forms a complete ring around the trachea and/or the esophagus. The two most common forms of vascular rings are the double aortic arch and a right aortic arch with an anomalous origin of the left subclavian artery. Symptoms of airway and/or esophageal obstruction can result depending upon the exact anatomy and degree of compression. For example, in double aortic arch, where the ring is comprised of the persistence of the right aortic arch, the left arch, and the ligamentum arteriosum (or the ductus itself if still patent), infants usually present with respiratory distress, stridor, and swallowing difficulties within the first six months of life. Recurrent respiratory difficulty or dysphagia in a young infant should raise the question of the presence of a vascular ring. Partial vascular rings may be relatively asymptomatic if there is little tracheoesophageal compression. Older infants and adults with undiagnosed vascular rings have presented with acute esophageal foreign body impaction, refractory "asthma", and progressive dyspnea on exertion. The presence of a right aortic arch on CXR increases the likelihood of symptoms being due to a vascular ring (conversely, a left aortic arch markedly reduces the odds). Although a pulmonary artery sling is among the most infrequent forms of vascular ring, it is most likely to be associated with complete tracheal rings.

Patients with vascular rings who are scheduled for surgery need to have a thorough evaluation to determine the precise anatomy of the ring, as well as the location, degree, and cause (e.g. extrinsic compression vs. complete tracheal rings) of tracheal narrowing. Cardiac MRI, CT angiography, echocardiography, and bronchoscopy can provide important information to this assessment. Choices regarding induction methods, monitoring, and airway management (e.g. ETT size and placement) should be based upon this understanding of the anatomy, tracheal pathology, and planned procedure. A bronchoscopic evaluation of the trachea is frequently performed at the end of a repair to confirm relief of tracheal narrowing. Most patients with vascular rings can be extubated successfully at the end of the procedure. Varying degrees of residual tracheomalacia can be expected when significant extrinsic airway compression was present. In the case of a repair of complete tracheal rings, opinions and practice vary regarding the necessity and duration of postoperative endotracheal intubation +/- muscle relaxation.

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