SPA Spring 2013 PBLD

**Title:** Opening Pandora’s Box: A 2-day-old neonate with duodenal atresia, found to have a critical airway anomaly during induction!

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**Goals:** At the end of the PBLD, participants will be able to:

- Discuss the differential diagnosis of neonatal airway abnormalities, including structural and vascular anomalies associated with airway compression/compromise.
- Describe the various anesthetic management styles regarding a patient with poor vascular access and requiring urgent/emergent surgery on a full stomach.
- Review the ASA difficult airway algorithm regarding management of a difficult airway.
- Discuss/debate the need for an airway evaluation and work-up on a neonate with unknown airway anomalies, prior to proceeding with a general anesthetic for an urgent procedure.
- Time permitting; the group will discuss the repair of complete tracheal rings.

**Case:**

Your surgery colleagues call you; they have just posted a case of a 2-day-old neonate for a duodenal atresia repair. The baby will arrive in our NICU shortly and will be transferred back to the outside hospital POD #2 if everything goes well. The infant was born full term via C-section and per the outside hospital the prenatal ultrasound was negative for any major anomalies. The baby lost its first IV and after multiple unsuccessful attempts, the outside hospital deferred placing the IV with the expectation that anesthesia could place it intra-operatively.

*What is duodenal atresia? Are there any other anomalies that are associated with duodenal atresia? Can this surgery be postponed until the infant is older? What things / problems would we expect to commonly be found on the prenatal ultrasound? Do you have any concerns because the outside hospital has been unable to establish a PIV? How would you induce this baby without an IV? (Find an IV, mask induction, etc.)*

The baby arrives in the NICU and you send your trainee to assess the infant. He reports that the baby is slightly tachypnic and tachycardic and is on ½ L nasal cannula O₂. The baby has mild retrognathia and is making some mild inspiratory
noises that is not relieved by repositioning. The baby’s extremities are slightly cooler and with slightly decrease capillary refill.

What do you attribute the baby’s need for oxygen to? What is your potential differential diagnosis for the mild inspiratory noises? What do you attribute the cool extremities and decreased perfusion to?

The baby arrives in the OR crying and is gagging on copious secretions. You attempt to suction the baby and are unable to pass a suction catheter down either nares. However, when you remove the nasal cannula from the nares the sats drop to the mid 80’s. You successfully suction the oral airway and set to work finding a PIV. After establishing a 24g PIV in the left foot, you take the time to give the baby a 10 ml/kg fluid bolus of LR and the baby settles under the warming lights.

Does the inability to pass your suction catheter down the nares raise any concerns? What is your differential diagnosis concerning the airway? Is the infant appropriately resuscitated? Would you like any laboratory values? Do you need a T&C?

The infant’s extremities are now warm and with appropriate capillary refill times. The infant is now asleep on the operating table under the heat lights with only slight inspiratory noises noted and the following vital signs:

HR 120, BP 69/43, RR 45, T. 36.8 Axillary, and SpO₂ of 99 on ½ L NC., Wt. 3.3 Kg

How would you induce this patient?

You elect to perform a modified rapid sequence induction. After switching to oxygen via your mask, the nurse positions herself to give cricoid. You give small incremental boluses of Propofol for a total of 10 mg and instruct your trainee to adjust the airway and give small breaths to supplement the baby’s breathing.

Would you give a paralytic now? Would you consider a Propofol only induction? Are there any problems associated with high dose Propofol? Which drug would you choose to paralyze with?

You are able to ventilate the infant well and elect to give Rocuronium 2mg. After waiting the appropriate time, your trainee intubates the infant with a 3.0 cuffed ETT but reports significant resistance and can not push the ETT past the vocal cords. You then have the trainee reintubate the patient with a 3.0 uncuffed ETT. He reports the ETT passed the vocal cords but meets some resistance past the vocal cords. You ask him to step aside and slightly withdraw the ETT and try passing it yourself. You agree that there is more resistance than expected but are able to get EtCO₂ and chest rise from ventilating the endotracheal tube. The infant remains well perfused and the saturations are 99-100%.
What is in your differential diagnosis with regard to the airway? What would be your next anticipated move? What does the ASA difficult airway algorithm suggest? Do you ask for any help from your colleagues? Do you request an ENT consult? What do you tell your surgeon?

You proceed to personally place a 2.5 uncuffed ETT in this term infant that moves freely and has a leak noted at 22 cm of H₂O pressure. You discuss with your surgeon that you have now found two airway anomalies in this infant and would like to have ENT evaluate the airway before proceeding with the laparotomy. The surgeon is worried you may cancel the case but agrees to discuss the matter after ENT comes into the room for an evaluation.

The ENT surgeon had been working in the next room and arrives quickly. You discuss with him your findings and he wishes to proceed with a rigid bronchoscopy. During the rigid bronchoscopy, the neonate is noted to have complete tracheal rings along the entire trachea. The ENT surgeon also reports a tortuous airway with unusual take offs.

What does this mean for the infant? Do you elect to proceed with the laparotomy? If there is a pulsatile wall/ bulge noted by the ENT, does that change your differential diagnosis? Would you proceed with the duodenal atresia repair now?

The 2.5 uncuffed ETT is replaced. You continue to be able to ventilate the patient well. After discussing the risks and benefits with your surgeons, you allow the surgeon to proceed with the duodenal repair. At the end of the surgery, the patient is transferred back the NICU intubated.

Considering the airway anomalies and recent surgery, would you extubate? What further evaluations do you expect this infant to undergo? How do we correct complete tracheal rings? What about vascular rings?
Discussion:

Duodenal Atresia

Congenital duodenal atresia and stenosis is a common cause of intestinal obstruction. Embryologically, it results from the failure of recanalization of the solid core of duodenum. The incidence of this congenital anomaly is about 1 per 5000 to 10,000 live births and affects boys more than girls (Holcomb & Murphy). More than 50% of those patients have associated congenital defects including pancreatic defects, intestinal malrotation with congenital bands, esophageal atresia, Tracheoesophageal fistulas, Meckel’s diverticulum, imperforate anus, congenital heart disease, central nervous system defects, renal anomalies, and biliary tract anomalies. Trisomy 21 is strongly associated with duodenal atresia/stenosis/web and is present in approximately 25% to 50% of cases (5).

The diagnosis of duodenal atresia is commonly made prenatally when ultrasonography demonstrates gastric and proximal duodenal dilation and polyhydramnios. The non-contrast radiographs of the infant with duodenal atresia classically demonstrate the presence of air in the stomach and in the first portion of the duodenum, the “double –bubble” sign and absence of air beyond the second bubble. The infant with duodenal atresia is often born prematurely and clinically presents with early feeding intolerance characterized by vomiting and upper abdominal distention. Emesis is usually bilious because most lesions occur distal to the entry of the bile duct into the duodenum. Nonbilious emesis may be present in 15% to 20% of cases due to a more proximal obstruction (5).

Duodenal atresia is a serious condition of neonates that has high mortality rate secondary to multiple comorbidities including prematurity, chromosomal and other associated anomalies. The pediatric anesthesiologists face a series of challenges including dehydration leading to metabolic and electrolyte disturbances and shock with end organ damage, difficult peripheral IV access and potentially challenging airway management due to airway anomalies. There is a significant risk of aspiration of gastric contents during or after induction (3). Commonly infants with duodenal atresia will arrive to operating room for surgical intervention within a few days to a few weeks of life. But prior to the OR trip these infants usually have multisystem evaluations including a genetic evaluation and a search for other associated anomalies. This evaluation is especially important in infants diagnosed postnatally.

Congenital airway anomalies

Congenital lesions of the airway include congenital tracheal lesions and cardiovascular causes of airway compression. Congenital tracheal lesions are rare;
hence experience in their management is limited and dispersed. On the other hand, vascular compression of the pediatric airway is relatively common and an often unrecognized complication of a variety of cardiovascular diseases. The intimate anatomic relationship between the cardiovascular and airway structures plays an important role in this phenomenon (1, 7).

The trachea in a full-term infant is about 4 cm long, whereas 11 to 13 cm long in an adult. The average diameter of the trachea in a full-term infant is 4-6 mm and 12 to 17 mm in an adult (6, 7). Because of these size differences, the pediatric airway is more susceptible to airway obstruction in the infant.

The diagnosis is of a congenital tracheal anomaly or an obstructive extrinsic compression is based on a high degree of suspicion in infants and children with respiratory distress. This high index of suspicion is especially important in infants and children with recurrent respiratory difficulties, stridor, wheezing, and dysphagia or apnea unexplained by other causes. Sometimes, recurrent or persistent cough, exercise intolerance, and difficulty intubating leads to the diagnosis (1, 4, 7).

If an airway anomaly is suspected, prompt diagnosis and work up is necessary to avoid further damage to the airway and possible death. There are varieties of diagnostic modalities available including plain chest radiography and echocardiogram, diagnostic investigations may include barium esophagograph, MRI, CT scans, cardiac catheterizations, and flexible and rigid bronchoscopy. The echocardiogram is essential for the evaluation of any associated congenital heart disease and is usually able to show the abnormal vascular structures. Recent advancements in CT’s and MRI’s can produce three-dimensional reconstructions of all anatomic elements providing more precise information about the cross-sectional area and the extent of the lesion. Despite all these noninvasive diagnostic modalities, the more invasive flexible and rigid bronchoscopy are often required for congenital acquired tracheal anomalies, e.g., tracheoesophageal fistula, tracheobronchomalacia and congenital tracheal stenosis and complete tracheal stenosis (1, 4, 7).

Vascular Compression of the Airway

Airway compromise secondary to underlying heart/vascular pathology is mainly due to two reasons – 1) the anomalous relationship between the tracheobronchial tree and vascular structures and 2) extrinsic compression by enlarged cardiac or pulmonary vascular structures (1). The term “vascular ring” is an aortic arch anomaly in which the trachea and esophagus are surrounded completely by vascular structures. Rings are formed by the abnormal persistence and/or regression of components of the aortic arch complex, which do not need to be patent but can cause compression of the trachea, bronchi and esophagus. A second vascular structure that can affect the airway is a “vascular sling”. The vascular sling
causes tracheal and esophageal compression without these structures being completely encircled by the vascular structures (1).

Table 1: Causes of vascular compression of the airway in children (4)

I. Anomalies of the aorta
   - Double aortic arch
   - Interrupted aortic arch (post-surgical repair)
   - Right sided aortic arch
     - With aberrant left subclavian artery
     - With mirror image branching and right ligamentum arteriosum
   - Left sided aortic arch
     - With aberrant right subclavian artery and right ligamentum arteriosum
     - Right sided descending aorta
   - Cervical aortic arch

II. Absent pulmonary valve syndrome

III. Aberrant left pulmonary artery (aka pulmonary artery sling)

IV. Acquired cardiovascular disease
   - Dilated cardiomyopathy
   - Aneurysm

Surgery is indicated in all patients with symptomatic vascular rings to prevent cartilage destruction and malacia of the infant airway, which has been observed to appear with in weeks of induced external compression. The goal of the operative repair is to divide the compressive vascular rings or slings to relieve tracheobronchial and esophageal compression and to maintain aortic perfusion (1). The most common surgical approach to vascular rings is via left thoracotomy; rarely necessitates a right thoracotomy. A sternotomy is indicated when the concomitant repair of intracardiac defects is planned or a case of pulmonary artery sling with tracheal reconstruction in which case cardiopulmonary bypass is required. Anesthesia is also complicated for these patients requiring pediatric thoracic and cardiac anesthesia skills (1).

Intrinsic Tracheal Lesions

Intrinsic tracheal lesions include tracheobronchomalacia (TBM), stenosis, complete tracheal rings or aberrant bronchi. In the pediatric population, primary or secondary TBM is the most common cause of significant airway collapse resulting airway obstruction. In most infants and children, TBM is self-limited and resolves within 1 to 2 years of life. Patients with “dying spells”, recurrent pneumonia or those who cannot be weaned from mechanical ventilation require aggressive medical and surgical intervention (7).
Table 2: Secondary causes of tracheobronchomalacia (7)

1. Esophageal atresia with TEF
2. Extrinsic compression
   a. Vascular compression
   b. Cardiac causes – enlarged structures
   c. Cysts – lymphatic malformations, thymic cysts, bronchogenic cysts
   d. Mediastinal neoplasm
   e. Infection
3. Prolonged intubation
4. Chondrodysplasias
5. Post traumatic result or following tracheoplasty

Complete Tracheal Rings

Congenital tracheal stenosis (CTS) in neonates and infants is characterized by a structural tracheal constriction, and is an often under diagnosed, life threatening respiratory anomaly, most commonly due to complete tracheal rings. The absence of the membranous portion of the trachea can create localized or generalized stenosis. More than 50% of the patients with CTS may have accompanying other malformations, including pulmonary, cardiovascular, gastrointestinal malformations or genetic anomalies.

Figure 1: Normal 1-month-old infant’s midtrachea with anterior cartilaginous rings and a membranous posterior (B) Three-month-old who has tracheomalacia; note the flattening of the lumen with a widened trachealis muscle. (C) Complete tracheal rings with absence of posterior membranous portion (Ha)
In the literature tracheal stenosis is defined as a reduction of the anatomic luminal diameter of the trachea by more than 50% compared with the remaining normal trachea (6, 7). Usually this anomaly is severe in nature and is diagnosed in the neonatal period. Sometimes diagnosis is made when patients require general anesthesia requiring an endotracheal tube or develop a respiratory tract infection resulting in further luminal narrowing. There are multiple case reports in the literature of complete tracheal rings being diagnosed months to years after birth. The condition is rare, with an incidence approximately 1 in 64,500 patients and represents 0.3 – 1% of all laryngotracheal stenosis. Mortality in these patients is related only to the severity of the stenosis and to the presence of other comorbid anomalies (6).

The variability in the timing of diagnosis reflects the variability of stenosis. There are three types of complete tracheal rings – 1) generalized hypoplasia of the trachea, 2) a funnel type stenosis, and 3) a segmental stenosis involving 2-3 rings. This anatomic description also correlates with the symptoms. Those having generalized hypoplasia, becomes symptomatic earlier in life with stridor, wheezing, and respiratory distress and whereas those having segmental stenosis may be symptomatic later in life or asymptomatic altogether.

![Figure 2: Type I is generalized hypoplasia, Type II is funnel type stenosis with one normal end and the other one stenotic, and Type III is segmental stenosis with two or three cartilage rings involved](image)

The CTS has been managed via both conservative medical as well as surgical interventions, depending on the presentation and severity of the stenosis. Surgical techniques includes -1) enlargement, using a costal cartilage transplantation, a pericardial patch or tracheal autograft, 2) resection of the affected segment and reanastomosis of the trachea in short segment stenosis, and 3) a slide tracheoplasty in patients with long segment stenosis.
Figure 3: Surgical treatment of CTS. (I) Resection and reconstruction, suitable only for shorter stenosis. (II) Patch tracheoplasty. Incision and tracheal widening with free cartilage or pericardial grafts.

Figure 4: Slide tracheoplasty.

Because tracheal lesions can be due to pure congenital tracheobronchial origin or the result of cardiovascular compression and are frequently associated with other congenital anomalies, patients should have a complete cardiovascular and airway evaluation before proceeding with anesthesia for a non-emergent or urgent procedures. For any urgent/emergent procedures, the risks versus benefits of proceeding with the anesthesia and surgery should be weighted and discussed among the multidisciplinary team involved in the care of the patient.

References:


