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Management of congenital tracheal anomalies and laryngotracheoesophageal clefts

Daniel P. Ryan, MD*, Daniel P. Doody, MD

Department of Pediatric Surgery, Massachusetts General Hospital, Harvard Medical School, 55 Fruit St, Boston, Massachusetts 02114

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ABSTRACT

Congenital obstructions and anomalies of the pediatric airway are rare problems that may be associated with mild symptoms or critical stenoses that may be life threatening in the first few days of life. This review provides an overview of the embryologic development of the airway, different congenital anomalies associated with airway development, and surgical correction that may be associated with good long-term outcome.

Pediatric airway abnormalities may be acquired or congenital. This review primarily addresses the congenital anomalies, which may be mild and associated with stridor to severe and life threatening. The embryology of airway development is reviewed although many of the clinically important pediatric airway anomalies are poorly explained by normal embryologic development. Nonetheless, awareness and early identification of these defects can lead to appropriate surgical correction with good to excellent long-term outcomes.

Embryology of the airway

The development of the larynx and the trachea begins very early in gestation. At about three weeks or 3 mm (stage 10), the laryngotracheal groove or sulcus appears in the proximal foregut. The lung buds are developing just caudal to this structure. After the first appearance of the laryngotracheal groove, the groove extends caudally forming the primordium of the trachea with the embryonic lung mesenchyme coming off of the developing airway. This embryonic airway lies ventral and parallel to the dorsal portion of the foregut, which will become the esophagus. The lateral ridges separating the trachea from the esophagus advance cranially while the esophagus is elongating. The trachea gradually elongates off the larynx and the separation between the airway and esophagus occurs. The lung buds seem to induce separation of the foregut and airway with separation starting at the level of the larynx. As the larynx develops superiorly, the trachea develops inferior to the sulcus, and the esophagus develops dorsal to the trachea and the lung buds. The esophagus (foregut) elongates as the development of the airway continues. With current theories, other than at the level of the larynx, there is no continuity between the foregut and the trachea at any stage. While the epiglottis develops its cartilage at the fourth week of gestation, the cartilaginous development of the upper airway begins around five weeks in the thyroid and cricoid area.[1,2]

It is very difficult to identify the causal embryologic elements with the known common anomalies seen clinically. The common esophageal atresia and tracheoesophageal fistula, for example, do not have an obvious correlation in embryology, since normally the lower trachea is always separated from the esophagus. The anomalies of the larynx that are seen with clefts are quite complicated, and it is not easily seen how they develop from the embryologic specimens. There may be apoptosis between the arytenoid cartilage areas leading to the cleft. The complete laryngotracheoesophageal cleft would suggest that there is an incomplete separation starting as the airway begins to develop. Whether this is an issue with apoptosis, failure of adequate separation, or failure of ingrowth of mesoderm to fill the space between the airway and the foregut is unclear.[3] One interesting factor in the embryology of this area is that the esophagus in the early stages has ciliated epithelium that only in later stages is replaced with the normal stratified squamous epithelium.[1]

Many anomalies that develop in the airway and larynx result in a fatal outcome at birth. Certainly, the fetus can be supported on the placenta even if the airway is not growing. Laryngeal atresia blocks the efflux of tracheal lung fluid leading to overdevelopment of the airways and lungs due to the loss of egress of the fluid. Agenesis of the trachea has been seen occasionally with the lung

* Corresponding author.
E-mail address: dryan@partners.org (D.P. Ryan).

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buds coming off of bronchi that are attached to the esophagus. There are rare case reports of survivors of this seemingly fatal congenital anomaly with the airway reconstructed with the esophagus and an intestinal interposition for swallowing.[4–7]

Laryngeal anomalies

Laryngeal problems will be touched on only as they are usually dealt with by our otolaryngology colleagues. Laryngeal atresia was mentioned above.

Webs of the larynx often present with stridor at birth and have persistent symptoms. The webbed tissue is often anteriorly located. These webs are easily diagnosed with direct laryngoscopy or suspension laryngoscopy. Endoscopic techniques are usually available to deal with these problems if they are significant in the newborn.[8] The airway may need to be secured with a tracheostomy tube below the repair while healing occurs.

Stenosis of the larynx down to the cricoid cartilage is not uncommon. Endoscopic techniques to identify these problems include suspension laryngoscopy, utilizing a magnifying telescope to get a good view of the area and to calibrate the size of the airway with endotracheal tubes. Laryngotracheal reconstruction with cartilage grafts is a common procedure to deal with laryngeal stenosis lesions.[9–11] Some of these stenoses are congenital, but many more are acquired due to intubation, particularly in the premature infant. The neonatal larynx has more of a cone-shaped anatomy, rather than the cylinder that is seen in adults, and the area of the cricoid cartilage represents the narrowest part of the airway.[12–14] Therefore, this subglottic area is susceptible to inflammation and trauma from prolonged intubation. Occasionally, the cricoid split operation can be used if the problem is identified early and the problem is with reversible edema and not dense and organized scar in the area of the cricoid.[15] This operation can help open the airway and avoid tracheostomy.

Tracheal stenosis

Congenital tracheal stenosis is a very unusual problem. The exact embryology of the problem is uncertain. The theories and observations of development of the trachea and the tracheal cartilages are difficult to reconcile with the clinical problems. The cartilage of the airway starts to develop much later than the elongation of the trachea, and the development of complete tracheal rings is hard to understand from the normal embryology. With tracheal stenosis, there may be a VACTERL association, especially imperforate anus, as well as various anomalies of the great vessels or intra-cardiac malformations. Tracheoesophageal fistula, lung sequestration and pulmonary agenesis (usually right-sided) have been seen as well.[16–19]

Although commonly asymptomatic at birth, these infants develop symptoms with respiratory distress and audible stridor very early in life, typically within the first few months. The evaluation of an infant with these symptoms includes a direct evaluation of the airway in order to identify the problem and the exact location, typically with magnifying telescopes and rigid bronchoscopy. A flexible bronchoscopy may be used, but the rigid instrumentation usually provides a more stable and better view to assess the size and position of the narrowing. Adjunctive evaluation using cross-sectional imaging with CT scanning with vascular contrast is necessary to look at the nearby great vessels and helps to establish a plan for surgical reconstruction. Echocardiography is indicated to evaluate the great vessels and any concurrent intra-cardiac lesions that may be associated with tracheal stenosis.[17,18,20–24]

The types of tracheal obstruction that are seen vary widely and are usually associated with a normal larynx. These obstructions are identified during the evaluation for stridor and can have a segmental tracheal narrowing anywhere from the middle to the lower trachea. When external compression from the innominate artery or a vascular ring is present, one must evaluate the airway anatomy to ensure that intrinsic stenosis is not present. The most common airway finding with vascular compression is tracheomalacia, with the trachealis muscle intact, or even slightly enlarged, indicating that relief of the compression will address the symptoms. Patients with mild tracheomalacia and narrowing from vascular compression will improve with somatic growth. These children may not need surgical intervention. However, if there are complete tracheal rings associated with vascular compression, relief of the extrinsic pressure alone does not help the abnormally narrowed airway.[9,25]

With tracheomalacia, the infants present with a characteristic barking cough. The most severe cases develop cyanotic and/or apneic episodes, often described as blue spells, death spells, dying spells or reflex apnea. Besides vascular compression, there is an association between tracheomalacia and esophageal atresia, most commonly with associated tracheoesophageal fistula. The indications for operative treatment of tracheomalacia usually require severe symptoms (recurrent blue spells), because time allows most infants to outgrow this cartilaginous weakness without surgery. In those severe cases, an aortopexy, placing pledgeted sutures from the aortic arch and the innominate artery anteriorly up toward the sternum, lifts the anterior trachea along with the great vessels, enlarging the overall tracheal lumen and lessening the collapse that occurs with the malacic tracheal cartilages.

One of the unusual cardiac anomalies associated with tracheal narrowing is a left pulmonary artery sling, in which the left pulmonary artery comes off the right pulmonary artery and circles the distal trachea and sometimes the right main stem bronchus. Although one would think that re-routing of the blood vessel would help the problem, there is often an intrinsic stenosis to the airway that needs to be addressed at the same time. Identifying the configuration of the tracheal rings is essential to plan this corrective surgery.[18,26,27]

The treatment for short segment stenosis, if it involves no more than about 25% of the length of the airway, is often a resection and re-anastomosis. Long segment tracheal stenosis is a much more difficult problem. There is a long history of attempts to use grafts of cartilage, pericardium, or periosteum in order to enlarge the airway by using a longitudinal incision and then sewing in the graft.[28] A case of tracheal replacement with a tissue-engineered graft has been reported.[29] Some of those methods have been successful in relieving the immediate and critical problem, but many times there are early failures and long-term problems. The grafts can displace, induce severe granulation, or not grow well over time.

The most successful treatment is the development of the slide tracheoplasty, which allows the airway to be shortened and enlarged in circumference at the same time.[30–33] Over time this reconstruction grows, maintaining a stable airway and allowing the child to function normally.[34]

The slide tracheoplasty is an extension into the pediatric population of methods described for complex problematic airways in adults.[35] The trachea is approached via a cervical incision, along with an extension through the upper sternum, if necessary. A full sternotomy is used if there is a concurrent cardiac procedure to be performed. The front of the airway is exposed, and the thymus and vessels in the area are mobilized. After the complete length of the trachea to the carina is exposed, the external anatomy can be marked to see the center of the stenotic segment. Bronchoscopy is