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Review

Rings, slings, and other tracheal disorders in the neonate

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SUMMARY

Tracheal and bronchial pathologies in the neonate can be rapidly fatal if prenatal or quick postnatal diagnosis and intervention is not performed. Close multidisciplinary collaboration between multiple medical and surgical specialties is vital to the effective diagnosis and treatment of these pathologies. The fetal and neonatal airway may be affected or compromised by more prevalent pathologies such as tracheomalacia and tracheo-esophageal fistula with esophageal atresia. However, it is imperative that we also consider other potential sources that may perhaps be less familiar such as congenital cardiovascular abnormalities, tracheal stenosis, complete tracheal rings, tracheal sleeve, and foregut duplication cysts. Modern imaging studies and surgical techniques are allowing us to better serve these children.

1. Introduction

Congenital tracheal and bronchial anomalies can present acutely within the first few days of life with potentially rapidly fatal respiratory distress or remain hidden, asymptomatic, and undiagnosed for many years. Immediate diagnosis and appropriate intervention can prevent morbidity and mortality.

Neonates may present in respiratory distress, or alternatively with the symptom of noisy breathing. Stridor is most often due to laryngomalacia. However, a normal laryngoscopic examination should warrant further investigation of potential tracheal or bronchial anomalies. Delineation of the lower airways and cardiovascular anatomy with microlaryngoscopy and bronchoscopy and radiologic imaging (computed tomography angiography, CTA; or magnetic resonance angiography, MRA) is critical for accurate diagnosis and development of a management plan.

Ultimately, congenital anomalies of the trachea and bronchi result from either an intrinsic abnormality of the cartilage or extrinsic compression of the airway from cardiovascular or gastrointestinal malformations. A multidisciplinary approach with the close co-operation of anesthesiologists, cardiologists, cardiothoracic surgeons, general surgeons, neonatal and pediatric intensivists, and pediatric otolaryngologists specializing in airway reconstruction is necessary for effective management of congenital airway disease. Tracheomalacia (both primary and secondary), congenital cardiovascular abnormalities, tracheo-esophageal fistula with esophageal atresia, tracheal stenosis, and foregut duplication cysts can all lead to airway compromise.

2. Anomalies of the trachea

2.1. Tracheomalacia

Tracheomalacia is the most prevalent pathology affecting the trachea for both full term and premature neonates. Tracheomalacia is a dynamic narrowing of the lumen of the trachea during breathing due to a weakness of the trachea wall (Fig. 1). The trachea is composed of 16–20 C-shaped cartilaginous rings anteriorly and a soft membranous trachealis muscle posteriorly. The tracheal lumen normally undergoes dynamic changes during the respiratory cycle; however, the degree of airway collapse is excessive in patients with tracheomalacia and results in symptoms [1]. Most instances of tracheomalacia are intrathoracic with tracheal narrowing seen with forced expiration or cough. Extrathoracic tracheomalacia occurs during inspiration when negative intrapleural pressures are transmitted to the extrathoracic trachea [2].

Pathologic tracheomalacia is typically seen when the tracheal lumen narrows by >50%. The normal tracheal ratio of the cartilaginous ring to the posterior membranous wall ranges from 4:1 to 5:1 and changes to between 2:1 and 3:1 with pathologic tracheomalacia resulting in the development of symptoms. Symptoms of

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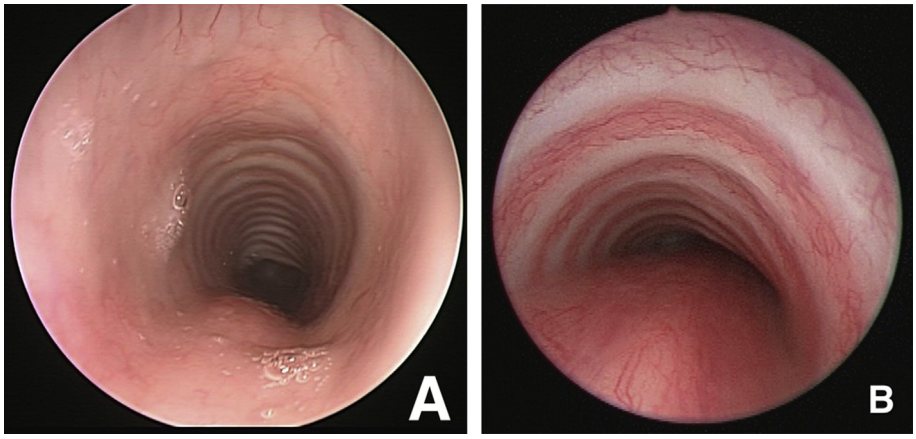

Fig. 1. Bronchoscopy images showing normal trachea (A) and tracheomalacia (B).

tracheomalacia may include cough (83%), recurrent lower airways infection (63%), dyspnea (59%), recurrent wheeze (49%), recurrent rattling (48%), reduced exercise tolerance (35%), symptoms of reflux (26%), retractions (19%), and stridor (28%) [3]. These findings are often more prevalent with increased activity or agitation. Patients with tracheomalacia may also present with recurrent respiratory infections due to impaired clearance of secretions with luminal collapse. Patients with intrathoracic tracheomalacia present with a wheeze on expiration, whereas patients with extrathoracic tracheomalacia present with stridor on inspiration [1]. Infants with extrinsic vascular compression may also present with feeding difficulties such as dysphagia, regurgitation, and coughing and cyanosis with feeding.

History and physical exam findings can be combined with results from pulmonary function tests and radiological imaging studies in order to narrow the differential diagnoses and determine etiology. Pulmonary function testing may show a truncated expiratory flow–volume loop in older children, but are not as helpful in infants due to the need for sedation. Whereas fluoroscopy is not useful to determine cross sectional area of the airway, it can be used to look for an anterior–posterior (AP) luminal decrease in diameter with a specificity of 96–100%. However, this modality is poorly sensitive (23.8–62%) for tracheomalacia as during periods of crying the AP diameter can decrease by up to 50% in a normal infant trachea [1]. Barium esophagography can be used to discern a tracheoesophageal fistula or vascular ring. Computed tomography (CT) and magnetic resonance imaging (MRI) performed with contrast can help evaluate external compression of the trachea, when looking for either masses or vascular compression. MRI/MRA is considered preferable to CT because it does not involve radiation exposure; however, CT may be the ideal study in a more medically fragile patient as it can be performed much quicker and is more sensitive for the airway. Modern CT imaging can help image the airway during the different phases of respiration, thus making it easier to detect dynamic changes in caliber. However, there are a few concerns with this method. First, it requires radiation exposure. Moreover, in infants, this requires the patient to be sedated and intubated, which can distort the airway and change the tracheal dynamics [1]. The best way to evaluate for tracheomalacia remains flexible bronchoscopy under spontaneous ventilation. Dynamic movements of the airway during tracheomalacia can be masked with heavy sedation, use of a paralytic agent, or with positive pressure ventilation, resulting in a false negative result. Currently the challenge remains that the diagnosis of tracheomalacia is largely subjective, determined by the bronchoscopist as there is no standard definition at this time.

Tracheomalacia may be associated with many conditions [2]. Cardiovascular anomalies are associated in 20–58% of patients with tracheomalacia. These anomalies include septal defects of the atrium or ventricle, patent ductus arteriosus, tetralogy of Fallot, abnormalities of the aortic arch, hypoplastic left or right heart, dextrocardia, and valvular stenosis. Bronchopulmonary dysplasia is seen in up to 52% of infants with tracheomalacia. Gastroesophageal reflux has been seen in up to 78% of patients with tracheomalacia that is severe or life-threatening. Secondary airway lesions are also seen in patients with tracheomalacia, including subglottic stenosis, laryngomalacia, and vocal fold paralysis. There is also thought to be a neurologic relationship, since tracheomalacia is associated with 8–48% of patients having neurologic impairment and 26% of patients having severe developmental delay.

2.1.1. Congenital/primary tracheomalacia

Congenital or primary tracheomalacia results from inadequate maturity of the tracheal cartilage itself due either to premature birth, or to an inherent immaturity or abnormality of the cartilage matrix itself. Congenital tracheomalacia may occur in full-term infants, but more usually is seen in premature infants. The overall incidence of primary tracheomalacia has been reported to be one in 2100 children by conservative estimates [3]. Primary tracheomalacia has been associated with numerous conditions including Ehlers–Danlos syndrome, mucopolysaccharidosis, CHARGE, VACTERL/VATER anomaly (Vertebral anomalies, Anal atresia, Cardiac defects, Tracheo-esophageal fistula and/or Esophageal atresia, Renal & Radial anomalies and Limb defects), trisomy 21, Pfeiffer syndrome, DiGeorge syndrome, Pierre Robin sequence, and tracheo-esophageal fistula (TEF). Some may consider tracheomalacia associated with TEF as a secondary form but many feel it is really a primary form of tracheomalacia because the weakness of the trachea is not caused by external compression, rather is due to an innate weakness of the involved tracheal cartilage. Most patients will not require any intervention and will improve over time with maturation of the infant and airway; most resolve by two years of age [3]. In rare instances, a tracheotomy and positive pressure ventilation may be needed for infants with growth derangements, persistent respiratory distress, or feeding difficulties. Some have used endotracheal or endobronchial stents. However, due to the inherent small nature of the pediatric airway, it is the opinion of the authors that these be avoided until problems with extrusion, migration, bleeding, granulation tissue, difficulty with removal, pneumonia, and death can be resolved. Endotracheal or endobronchial stents should be reserved for emergent situations and require close surveillance of the airway. More recently, three-