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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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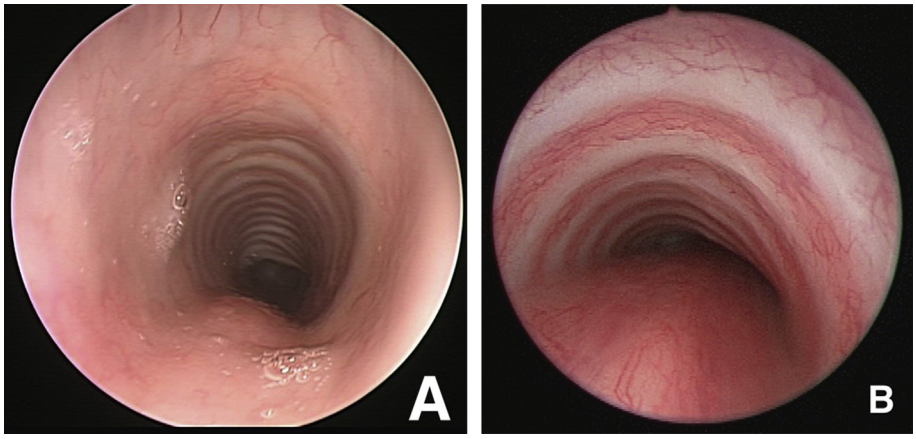


**Fig. 1.** Bronchoscopy images showing normal trachea (A) and tracheomalacia (B).

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-

dimensional (3D)-printed, bioresorbable splints have been described and surgically placed for life-threatening tracheobronchomalacia in children [4]. A custom-printed external splint was created and then used to reverse the tracheobronchial collapse by suturing the wall of the airway to the externally placed splint.

*2.1.2. Acquired/secondary tracheomalacia*

Acquired or secondary tracheomalacia is due to compression upon the airway, which may be from a variety of sources including cardiovascular, gastrointestinal, musculoskeletal, or neoplastic etiologies. This compression results in both a direct narrowing of the lumen as well as a weakening of the tracheal cartilage. A frequently occurring etiology is prolonged endotracheal intubation and associated insults of increased airway pressure, oxygen toxicity, and recurrent infections [2]. In premature infants with respiratory distress syndrome, this is compounded by an inherent immaturity of the tracheal cartilage itself. Another cause is tracheostomy placement. The surgical appliance may weaken the suprastomal trachea and the trachea adjacent to a cuff on the tracheostomy tube. Cardiovascular compression of the airway may be due to double aortic arch, innominate artery compression, vascular rings, or pulmonary sling. Vascular rings typically but not exclusively involve a right aortic arch. Additionally, compression may be due to a cervical or thoracic tumor, cyst, or abscess. Skeletal pathology, such as thoracic dysplasia, may also result in secondary tracheomalacia. The treatment of secondary tracheomalacia relies on alleviating the underlying cause of the external compression. It is notable that even after addressing the underlying compressive etiology, the tracheomalacia may continue due to the weakening and dysmorphia of the cartilage.

*2.1.2.1. Congenital cardiovascular abnormalities.* During embryogenesis, six paired aortic arches help to form the eventual mature aortic arch system. Developmental abnormalities, such as persistence of some of the arches may result in congenital vascular anomalies. Moreover, airway compression may also occur due to underlying congenital heart disease and is often unrecognized. Importantly, in addition to previously described airway symptoms, there may be symptoms of feeding difficulties and dysphagia with cardiovascular-related compression. As a general rule, neonates present with airway issues, whereas dysphagia tends to occur more widely in older children and adults. Evaluation includes AP and lateral chest radiographs, barium esophagram, echocardiogram, CT angiography or cardiac MRI (CMR), and direct microlaryngoscopy and bronchoscopy. When fine resolution of the airway is not necessary, CMR is preferred to CT angiography as it avoids ionizing radiation, which has been shown to cause malignancies in children [5,6]. A normal frontal and lateral chest radiograph significantly decreases the likelihood of finding a vascular ring in a symptomatic patient as the vast majority of vascular rings are associated with a right aortic arch, which can be identified on chest X-ray [7]. Microlaryngoscopy and bronchoscopy can also help identify synchronous primary airway lesions that may be present, as well as help document vocal fold motion prior to cardiovascular surgery. Intraoperative bronchoscopy done at the time of repair of the cardiovascular lesion can help show airway relief in real time. Loose vascular rings may be diagnosed incidentally in patients who undergo a chest X-ray, or CT examination for other reasons. These patients may have little if any symptoms and therefore would not be recommended for surgery.

The most prevalent vascular anomalies that result in airway or esophageal compression are (in decreasing order of frequency): (i) double aortic arch, (ii) right aortic arch with aberrant left subclavian artery originating from a retro-esophageal diverticulum (diverticulum of Kommerell), (iii) innominate artery compression, (iv) left aortic arch with aberrant right subclavian artery, (v) and pulmonary artery sling [7]. Left aortic arch with aberrant right subclavian artery is highly prevalent in the general population but rarely causes symptoms and is often considered a normal variant in the context of people without symptoms. Furthermore, though right aortic arch with mirror image branching pattern is considered normal for right arches, there still could be a vascular ring if the patient also has a left ligamentum arteriosum to complete the ring. This circumstance can be challenging to diagnose as there is no obvious retro-esophageal diverticulum, but rather the only evidence of the ring may be a small leftward-directed dimple off the aorta.

*2.1.2.1.1. Vascular rings.* With true vascular rings, the trachea and esophagus are anatomically completely surrounded and compressed. Sometimes the mainstem bronchi may be narrowed. Complete vascular rings include double aortic arch and right aortic arch with aberrant left subclavian artery originating from a retroesophageal diverticulum and associated left ligamentum arteriosum. Symptoms involve biphasic stridor, wheezing, cyanosis with feeding, recurrent pneumonias or lower airway infections, and dysphagia. Initial diagnostic testing includes barium esophagram and airway fluoroscopy. This often shows a posterior indentation of the esophagus on lateral views. Cardiac MRI/MRA will define the vascular and tracheal anatomy and help with definitive diagnosis. A pulsatile tracheomalacia is often seen on bronchoscopy.

Double aortic arches account for 50–60% of vascular rings and are the most frequent cause of vascular compression of the airway in children (Fig. 2) [8,9]. As they are complete and tight, they are often symptomatic early in infancy and most often require surgical correction by dividing the smaller, non-dominant arch. Approximately 30% of patients undergoing surgical correction will still exhibit symptoms after surgery due to weakness of the cartilaginous rings from longstanding compression [9]. If severe symptoms do not resolve over time as the cartilage matures and stiffens, then a tracheotomy may need to be placed. A right aortic arch with left ligamentum arteriosum and aberrant left subclavian artery also encircles both the trachea and esophagus. This is not usually as constricting and thus often is asymptomatic or presents with symptoms later in life, sometimes when solids are introduced. This entity is the second most frequently occurring vascular ring and accounts for 12–25% of cases [8]. Surgical correction involves division of the left ligamentum arteriosum. It is also important to retract or excise Kommerell's diverticulum if it is causing significant compression. Kommerell's diverticulum is the outpouching of the descending aorta which embryologically derives from ductus arteriosus tissue. The fibrous left ligamentum segment which is in continuity with the diverticulum (but not observed on MR or CT angiograms) completes the ring and is tethered inferiorly to the left pulmonary artery. The left subclavian artery originates from the retroesophageal diverticulum. If the ring were divided and the retroesophageal diverticulum left intact, this situation can result in late compression of the trachea or esophagus [9].

*2.1.2.1.2. Innominate artery compression.* The innominate artery can cause tracheal compression as it travels (from its more distal origin from the aortic arch) anterior to the trachea towards the right. This may manifest as a focal anterior tracheal compression at the level just proximal to the carina. Bronchoscopy will exhibit focal, pulsatile and asymmetric airway compression just proximal to the carina and of the right mainstem bronchus. Definitive diagnosis can be made with dark blood MRI with tracheal imaging or CT angiography. Deformity of the tracheal rings and resultant cartilaginous weakness may compound the problem and result in persistent airway symptoms despite surgical intervention. In mildly affected patients, conservative monitoring is all that is needed as patients can improve as the airway grows. However, with severely affected children, surgical correction in the form of aortopexy with

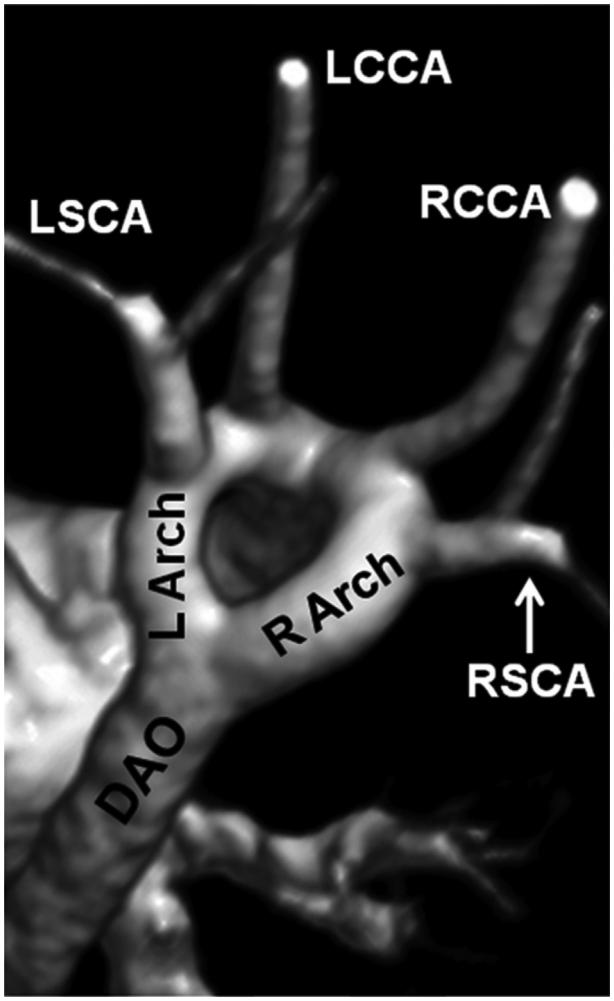


**Fig. 2.** Magnetic resonance angiography demonstrating double aortic arch. DAO, descending aorta; L Arch, left-sided arch; LCCA, left common carotid artery; LSCA, left subclavian artery; R Arch, right-sided arch; RCCA, right common carotid artery; RSCA, right subclavian artery. (Reprinted with permission from Lioy J, Sobol S, editors. Disorders of the neonatal airway: fundamentals for practice, p. 115. © Springer, 2015.)

suspension of the innominate artery to the undersurface of the sternum may be needed. Reimplantation or transection may be another surgical option [10,11].

*2.1.2.1.3. Pulmonary artery sling.* This abnormality involves the anomalous course of the left pulmonary artery, which originates from the posterior aspect of the right pulmonary artery instead of from the common pulmonary artery trunk (Fig. 3). In contrast to vascular rings where the ring includes the trachea and the esophagous, in pulmonary sling the anomalous left pulmonary artery passes in between the trachea and esophagus as it is directed toward the left hilum, compressing the distal trachea and right mainstem bronchus as well as the anterior aspect of the esophagus. Although a rare entity, pulmonary artery slings are often associated with other severe abnormalities: about 50% have congenital heart disease, 58–83% have congenital anomalies, 50% have complete cartilaginous tracheal rings, and others have tracheomalacia, abnormal pulmonary lobulations, or bronchus suis (right upper lobe bronchus originates directly from the trachea) [8]. Others caution that the incidence of complete tracheal rings and resultant tracheal stenosis is much more than 50% and should be carefully evaluated [9]. Importantly, the workup of these children should not only include CTA or cardiac MRI/MRA imaging, it should also include a bronchoscopy to evaluate the airway prior to cardiac surgery. Repairing the tracheal stenosis and the pulmonary artery sling at the same time is associated with better prognosis [9]. Mild cases can be monitored; however, if significant respiratory symptoms are present, then surgical translocation and reimplantation of the left pulmonary artery may be needed. If the tracheal stenosis is limited, a tracheal resection can be done at the same time. However, if there is a long segment tracheal stenosis present, then a slide tracheoplasty (described in Section 2.2.2) may be performed with the repair of the pulmonary artery sling.

*2.1.2.1.4. Other vascular abnormalities.* An aberrant right subclavian artery results in the posterior compression of the esophagus when, in the setting of a left aortic arch, there is an abnormal origin of the right subclavian artery as the last branch from the aorta. Dysphagia can be a presenting symptom, but most often this entity is asymptomatic [7].

A cervical aortic arch is a rare entity and occurs when the aortic arch abnormally courses high into the neck superior to the clavicles (potentially as high as C2). Although typically people with isolated

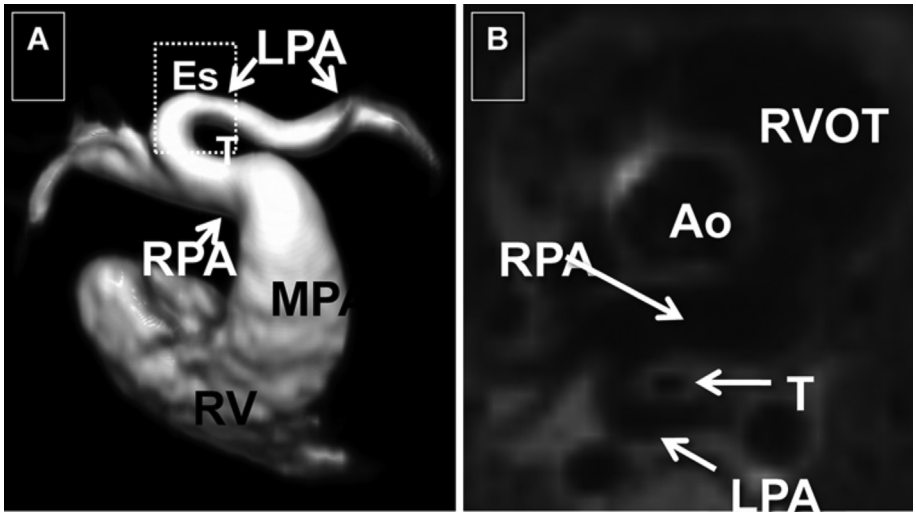


**Fig. 3.** (A) Magnetic resonance (MR) angiography showing pulmonary artery sling. The left pulmonary artery can be seen originating from the right pulmonary artery and then swinging around the trachea towards the left. (B) MR spin echo dark blood image showing narrowing of trachea at the level of the left pulmonary artery sling. aorto (Ao), esophagus (Es), left pulmonary artery (LPA), right pulmonary artery (RPA), right ventricular outflow tract (RVOT), trachea (T). (Reprinted with permission from Lioy J, Sobol S, editors. Disorders of the neonatal airway: fundamentals for practice, p. 117. © Springer, 2015.)

cervical aortic arch are asymptomatic, symptoms of pulsatile cervical mass, respiratory symptoms, and/or dysphagia may be present and ultimately depend upon the variant present.

*2.1.2.2. Congenital cardiac malformations.* Acyanotic or cyanotic congenital heart disease can result in direct compression of the airways or lungs with resultant respiratory symptoms. This category may be underappreciated. The trachea, carina, and left mainstem bronchus are closely associated with various cardiac structures such as the left atrium, left pulmonary artery, and left pulmonary veins. It therefore appears that various cardiac pathologies resulting in dilation/enlargement of cardiac structures or massive cardiomegaly may lead to airway compression and symptoms. Congenital heart disease resulting in left-to-right shunting (such as with ventricular septal defects, patent ductus arteriosus, or atrioventricular canal), tetralogy of Fallot with absent pulmonary valve leaflets syndrome or pulmonary atresia, mitral regurgitation, truncus arteriosus, or dilated cardiomyopathy may all be potential sources of airway compression. Increased intracardiac filling pressures may lead to enlarged bronchial vessels and lymphatics, and thus ultimately to intraluminal bronchial edema and obstruction [8].

In addition to cardiac MR, echocardiography, and bronchoscopy, some patients may benefit from cardiac catheterization to delineate cardiac pathology and potential effects upon the airway. If possible, the underlying cardiac defect should be addressed to improve the airway issues. Furthermore, patients with complex structural heart disease who undergo interventional catheterization for balloon dilation and stent implantation of a branch pulmonary artery may experience resultant ipsilateral mainstem bronchus compression [12].

*2.1.2.3. Tracheo-esophageal fistula with esophageal atresia.* Tracheo-esophageal fistula with esophageal atresia (EA) involve a defect in esophageal continuity that may occur with or without tracheal communication. EA occurs relatively widely with an incidence of one in 2500–3000 live births. The majority of instances are sporadic/non-syndromic, with <1% of the total being due to familial/syndromic cases of EA [13,14]. TEF and EA have a classification scheme that is based by most clinicians on descriptions by Vogt or Gross (Table 1).

A high degree of suspicion is needed to look for concurrent anomalies, as >50% of cases occur with other congenital issues. Pathologies within the cardiovascular (29%), anorectal (14%), genitourinary (14%), gastrointestinal (13%), vertebral/skeletal (10%), and respiratory (6%) systems may be present [13]. Combined anomalies from vertebral, anorectal, tracheo-esophageal and renal or radial abnormalities should raise concerns for VATER association. Additionally, EA may also be seen in children with CHARGE association.

TEF and EA should be suspected prenatally with the presence of a small or absent fetal stomach bubble on ultrasonography, especially with concomitant polyhydramnios. It is suggested that in babies born to mothers with polyhydramnios, a nasogastric tube should be passed after birth to evaluate for EA. Neonates with TEF and EA may not accomodate a nasogastric tube, may have excessive salivation, regurgitation, coughing or choking with feeding, or respiratory distress or cyanosis immediately after birth. A complete evaluation of the neonate with EA and TEF is essential for good surgical planning. EA with distal TEF is suggested by an air-filled abdomen on radiography or aspiration of gastric reflux resulting in pneumonias. A radiologic examination with a water-soluble contrast may be used to visualize an H-type. Endoscopy is essential to visualizing the airway anatomy, fistula, and vocal fold mobility. Echocardiography allows for the evaluation of possible contemporaneous cardiovascular abnormalities.

Surgical intervention involves thoracotomy or thoracoscopy with division of the TEF and primary anastomosis of the EA. Spitz classification uses birth weight and presence of cardiac anomalies to help stratify patient survival. Group I patients have a birth weight of >1500 g without major cardiac anomalies; Group II patients have a birth weight <1500 g or major cardiac anomalies; Group III patients have a birth weight <1500 g and major cardiac anomalies. Survival is reported as 98%, 82%, and 22–50%, respectively for the three groups of patients [13]. Surgical complications may include esophageal stenosis/webs, anastomotic leaks, recurrent fistulae, tracheomalacia and respiratory distress, vocal fold paralysis, dysphagia and feeding issues, gastro-esophageal reflux disease, and respiratory infections. Tracheomalacia is present in 75% of patients undergoing surgery for EA/TEF; however, it is only clinically relevant in 10–20% of patients [15]. It is important to mention that even after repair of EA and TEF, tracheomalacia may persist and in some patients remain clinically significant. If persistent tracheomalacia is significant enough, positive pressure ventilation and/or tracheotomy may be necessary. A deep residual pouch after repair may trap secretions and lead to recurrent pulmonary infections.

### 2.2. Tracheal stenosis

#### 2.2.1. Tracheal webs

Membranous tracheal webs are rare and considerably less frequent than airway narrowing from cartilaginous defects or laryngeal webs [16]. They occur in the neonatal and juvenile airways at the level of the cricoid and are usually treated by endoscopic means. These can be incised sharply with an instrument or with a laser followed by balloon dilation. More extensive webs may need treatment with tracheal resection.

#### 2.2.2. Congenital tracheal stenosis

Congenital tracheal stenosis (CTS) involves at least 50% narrowing of the tracheal lumen and may encompass a few rings or even the entire trachea (Fig. 4). If the tracheal rings grow out of proportion to the posterior membranous trachea, complete or near-complete tracheal rings may be formed. In contrast to the normal "C-shaped" rings of a normal trachea, complete tracheal rings are narrower and are believed to be restricted in growth (Fig. 5A).

Most children with CTS and complete tracheal rings present during infancy. Symptoms include biphasic stridor, expiratory stridor, retractions, wet-sounding biphasic stridor/wheeze, cyanosis, apnea, chest congestion, and respiratory distress. Patients may also be identified at time of a difficult intubation or subsequently from difficult intubations, which result in airway edema and granulation tissue that can acutely worsen an already narrow

**Table 1**
Vogt and Gross classification schemes of esophageal atresia (EA) and tracheo-esophageal fistula (TEF) [13,14].

| Vogt | Gross | Incidence | Description |
|---|---|---|---|
| 1 | – | – | Absent esophagus |
| 2 | A | 7–8.5% | Isolated EA without TEF |
| 3a | B | 1–2% | EA with proximal TEF |
| 3b | C | 85–86% | Proximal EA (blind pouch) with a distal TEF attaching posteriorly to trachea |
| 3c | D | <1–1.5% | EA with proximal and distal TEF; double fistula |
| 4 | E | 4% | TEF present between intact trachea and esophagus, "H-type," may have multiple fistulas |

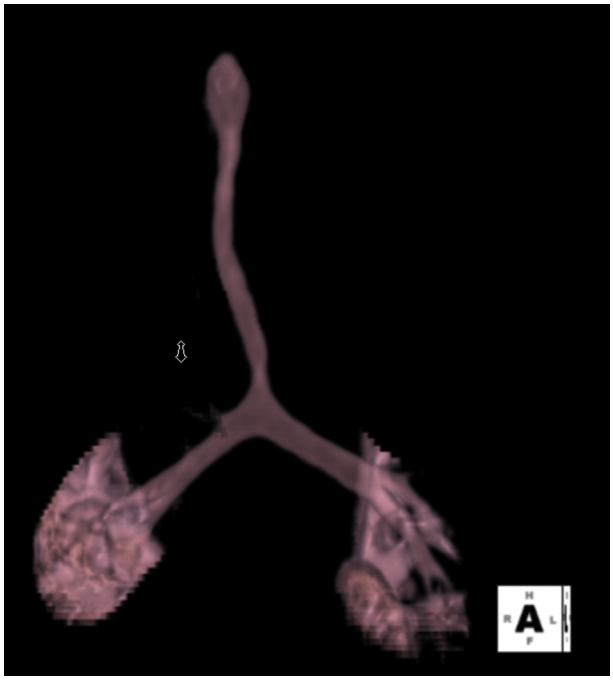

**Fig. 4.** Three-dimensional reconstruction of computed tomography images demonstrating long segment tracheal stenosis in a patient with complete tracheal rings. Note that most of the tracheal length is narrowed.

and marginalized airway. Patients may not become symptomatic until after a few months of age, as infants' respiratory needs "outgrow" their restricted airway. Moreover, a respiratory illness might unmask CTS as symptoms sometimes progress rapidly.

CTS and complete tracheal rings are usually associated with cardiovascular anomalies and thus require full evaluation with preoperative imaging with CTA or cardiac MR/MRA. In one large series, 71% of patients presented with cardiovascular anomalies; left pulmonary artery sling was the most frequent entity (48%). It is important to identify these cardiovascular anomalies as they can be corrected contemporaneously with tracheal repair [17]. Besides associated cardiovascular anomalies, infants with CTS may have pulmonary anomalies, Down syndrome, and Pfeiffer syndrome.

Initial management is centered on providing respiratory support. This is often a challenging situation as even the smallest (2.0) endotracheal tube may not bypass the stenosis. Thus, often a "high" nasotracheal intubation is necessary with the endotracheal tube just through the vocal cords. This has to be carefully positioned such that the end of the endotracheal tube does not rub against the proximal, narrowed complete tracheal ring, which may lead to progressive mucosal edema and inflammation or granulation tissue, both leading to potential inability to ventilate. To avoid this situation, efforts should be made to support the airway without intubation. In situations where the infant cannot be safely and effectively ventilated even with intubation, extracorporeal membranous oxygenation (ECMO) may be performed to bridge to a definitive reconstructive surgery. Prior to tracheal reconstructive surgery, bronchoscopy is essential in delineating the tracheal stenosis, measuring the degree of narrowing, and the length of the narrowing. For situations where a severe stenosis precludes the passage of a bronchoscope for full airway visualization, CT imaging of the airway with fine cuts can render a full picture of the airway. A larger caliber airway in a patient who is overall stable may be monitored. However, most patients with CTS will need surgical correction.

In the past, CTS has been repaired with tracheal resection and anastomosis, cartilage graft augmentation, and patch tracheoplasty with a material such as pericardium. However, the gold standard has become the use of slide tracheoplasty. Surgeons do have the option of performing a tracheal resection and anastomosis if only a few rings are involved. This should only be considered an option if there are no more than four or five rings involved. In performing a tracheal resection, 25–30% of the trachea can be removed before excessive anastomotic tension and threat of tracheal dehiscence precludes this as a therapeutic option [18]. However, even such a short span is often now managed with slide tracheoplasty. Slide tracheoplasty offers several advantages over previously described techniques including use of autologous tracheal tissue, earlier extubation, avoidance of a stent, decreased granulation tissue formation, and distribution of tension over a longer anastomotic length. Slide tracheoplasty shortens the trachea by half of the involved stenotic segment instead of the full length as would be required with a traditional tracheal resection approach. A slide tracheoplasty may be performed from cricoid to carina if required (Fig. 5B). Briefly, the trachea is divided at the midpoint of the stenotic section, the trachea is freed up from its deep attachments down to the level of the carina and superiorly towards the larynx to help increase segment mobility. Lateral tracheal attachments are preserved to help decrease any potential damage to the tracheal vascular supply. Next, an anterior longitudinal incision is made in the proximal segment through the stenotic area until normal tracheal cartilage is encountered. A posterior, longitudinal incision is made in the distal segment through the stenotic area until normal trachea or carina is encountered. The corners are trimmed and the tracheal segments are then slid over each other. A running polydioxanone or Prolene suture is used to perform the anastomosis. The

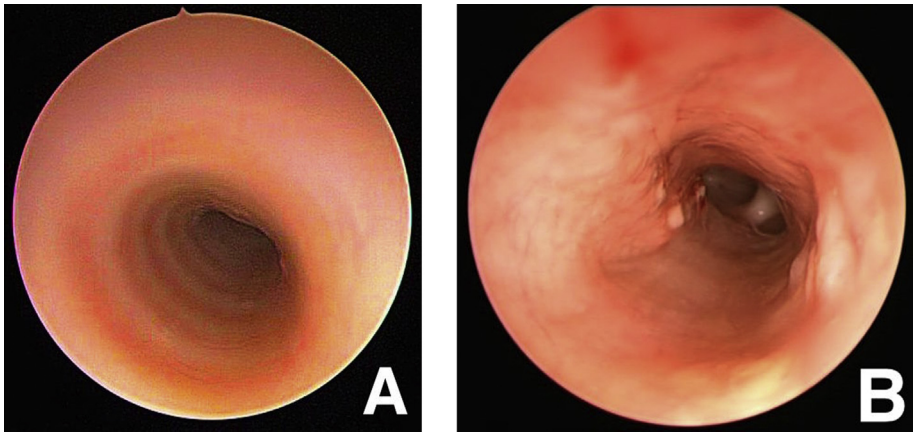


**Fig. 5.** Bronchoscopy images showing complete tracheal rings before reconstruction (A) and enlarged airway after slide tracheoplasty (B) in the same patient.

slide can be extended into the anterior aspect of the cricoid or even into a bronchus if needed. Potential complications include recurrent laryngeal nerve injury, dehiscence and restenosis, and dysphagia. Postoperatively, bronchoscopy is performed to monitor and help mold the healing process through adjuvant techniques such as excision of granulation tissue and balloon dilation.

*2.2.3. Tracheal cartilaginous sleeve*

Tracheal cartilaginous sleeve is a rare tracheal anomaly in which the trachea forms as a long cylindrical sleeve of cartilage rather than with normal tracheal rings. The posterior membranous trachea is absent or greatly diminished. The sleeve may be restricted to the trachea alone or may extend into the bronchial tree. Respiratory distress results from restricted growth of the caliber and length of the airway, and from mucous plugging. Tracheal cartilaginous sleeve is mostly seen in craniosynostosis syndromes such as Apert, Crouzon, and Pfeiffer syndromes. These syndromes may have other airway issues including hypopharyngeal obstruction or obstructive sleep apnea that can delay or mask diagnosis of a tracheal sleeve. The prognosis is poor with one report finding 90% of patients with both tracheal sleeves and craniosynostosis syndromes were dead by two years of age with 58% of the mortalities due to airway pathology. Furthermore, the same report states that the mean age of death is younger than three years of age in essentially all patients with tracheal cartilaginous sleeves [19]. The poor compliance of the cartilaginous sleeve is thought to result in issues with granulation tissue, functional issues, poor airway clearance mechanisms and mucous plugging which can be life-threatening [20]. Tracheotomy is often necessary to bypass upper airway pathology and allow for pulmonary clearance of the airway. However, due to the rigidity and abnormal configuration of the airway, tracheotomy in this population may be challenging and fraught with issues with granulation tissue and plugging, requiring frequent monitoring for this.

Symptoms include respiratory distress, cyanosis, recurrent respiratory infections and croup, severe biphasic stridor, and failure to thrive. Bronchoscopic visualization is necessary for diagnosis and elucidation of the extent of the sleeve. Fine-cut CT imaging of the airway can also help to visualize tracheal configuration. Treatment options include tracheotomy for airway support, tracheal resection for short segments, or slide tracheoplasty for longer segments.

*2.2.4. Agenesis and atresia of the trachea*

This rare tracheal anomaly involves the complete or partial absence of the trachea and is sometimes seen with a tracheo-esophageal fistula. The incidence is one in 50,000 live births with a 2:1 male predominance. Reportedly, 52% of affected individuals are premature, 50–94% are associated with other congenital malformations, and at least 50% involve pregnancies with polyhydramnios [21]. The Floyd classification is the most widely used system for tracheal agenesis. Type I (13%) has an absent proximal trachea with distal tracheo-esophageal fistula. Type II (65%) has a complete absence of the trachea with the carina usually opening into the esophagus, but not always. Type III (22%) has the origin of both mainstem bronchi directly from the esophagus [22].

Immediately after birth, clinical presentation may include cyanosis, severe respiratory distress despite efforts, absence of audible cry, poor air exchange on auscultation, and difficulty with or inability to intubate. If a tracheo-esophageal fistula is present, positive pressure ventilation via bag and mask ventilation or esophageal intubation can temporarily help to ventilate the child. ECMO may be needed to provide support while a more definitive plan is formulated.

Prenatal MRI may assist in diagnosis. Ex-utero intrapartum treatment (EXIT) procedure should be employed if tracheal agenesis is diagnosed prenatally to secure an airway. Further delineation of the airway includes microlaryngoscopy and bronchoscopy, esophagoscopy, and fine-cut CT imaging of the airway. Tracheal agenesis continues to present a challenge and there are not many good surgical options for reconstruction. Tracheotomy can be attempted. Postnatally diagnosed tracheal agenesis is often lethal. The unavailability of suitable grafting material results in frequent mortality.

Congenital high airway obstruction syndrome (CHAOS) deserves special mention. CHAOS results from tracheal agenesis or larnyngotracheal stenosis/obstruction. CHAOS can be diagnosed prenatally when ultrasound and MRI reveal enlarged hyperechogenic lungs, fluid-filled dilated trachea, flattened or inverted diaphragms, compression of the heart, and sometimes visualization of the obstruction [23]. An EXIT procedure can help secure an airway through a fetal tracheotomy or laryngoscopy and bronchoscopy. Laryngotracheal reconstruction can then be performed.

## 3. Foregut duplication cysts

Developmental anomalies of the embryonic foregut may result in foregut duplication cysts. These include bronchogenic cysts, esophageal duplication cysts, and neurenteric cysts, to name a few. Foregut duplication cysts may occur in the head and neck but are most often in the thorax. Symptoms relate to the level of the cyst, the mass effect of the lesion, and specific complications related to the malformation (such as acid-secreting mucosal cells) [24].

Bronchogenic and esophageal cysts are more frequent in the mediastinum. Symptoms include stridor, wheezing, cough, recurrent pneumonia, fever, chest pain, respiratory distress, dysphagia, and vomiting. Many are incidental findings on imaging and asymptomatic. Whereas chest X-rays may detect this pathology, MRI or CT and barium esophagram can better visualize foregut duplication cysts. Surgical resection provides symptom relief and resolution [25,26].

**Practice points**

- The best way to evaluate for tracheomalacia remains flexible bronchoscopy under spontaneous ventilation.
- CT angiography or cardiac MRI are vital for delineating cardiovascular anomalies that can cause airway obstruction.
- Stridor in a neonate or infant with a normal nasopharyngolaryngeal fibroscopic exam should be further evaluated with bronchoscopy to evaluate for complete tracheal rings or other tracheal pathology.
- Slide tracheoplasty is the primary treatment modality for long segment tracheal stenosis due to complete tracheal rings.

**Research directions**

- Tracheal transplantation for tracheal agenesis or other tracheal pathologies not amenable to current techniques.
- Use of resorbable external splints in the treatment of tracheomalacia.
- Development and implementation of custom 3D-printed tracheas.

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## References

[1] Hysinger E, Panitch H. Paediatric tracheomalacia. Paediatr Respir Rev 2016;17:9–15.
[2] Carden K, Boiselle P, Waltz D, Ernst A. Tracheomalacia and tracheobronchomalacia in children and adults: an in-depth review. Chest 2005;127:984–1005.
[3] Boogaard R, Huijsman S, Pijnenburg M, Tiddens H, de Jongste J, Merkus P. Tracheomalacia and bronchomalacia in children. Chest 2005;128:3391–7.
[4] Zopf D, Hollister S, Nelson M, Ohye R, Green G. Bioresorbable airway splint created with a three-dimensional printer. N Engl J Med 2013;368:2043–5.
[5] Brenner D, Elliston C, Hall E, Berdon W. Estimated risks of radiation-induced fatal cancer from computed tomography. Am J Roentgenol 2001;176:289–96.
[6] Hall E. Lessons we have learned from our children: cancer risks from diagnostic radiology. Pediatr Radiol 2002;32:700–6.
[7] Shah R, Mora B, Bacha E, Sena L, Buonomo C, Del Nido P, et al. The presentation and management of vascular rings: an otolaryngology perspective. Int J Pediatr Otorhinolaryngol 2007;71:57–62.
[8] Kussman B, Geva T, McGowan F. Cardiovascular causes of airway compression. Pediatr Anesth 2004;14:60–74.
[9] McLaren C, Elliott M, Roebuck D. Vascular compression of the airway in children. Paediatr Respir Rev 2008;9:85–94.
[10] Tatekawa Y, Tojo T, Hori T, Komuro H, Urita Y, Kudou S, et al. A new technique for treatment of tracheal compression by the innominate artery: external reinforcement with autologous cartilage graft and muscle flap suspension. Pediatr Surg Int 2008;24:431–5.
[11] Tsugawa C, Ono Y, Nishijima E, Takamizawa S, Satoh S, Muraji T. Transection of the innominate artery for tracheomalacia caused by persistent opisthotonus. Pediatr Surg Int 2004;20:55–7.
[12] O'Byrne M, Rome N, Santamaria R, Hallbergson A, Glatz A, Dori Y, et al. Intraprocedural bronchoscopy to prevent bronchial compression during pulmonary artery stent angioplasty. Pediatr Cardiol 2015 Nov 6 [Epub ahead of print].
[13] Spitz L. Oesophageal atresia. Orphanet J Rare Dis 2007;2:24.
[14] Höllwarth M. Esophageal atresia and tracheoesophageal fistula. In: Puri P, Höllwarth M, editors. Pediatric surgery: diagnosis and management. New York: Springer; 2009. p. 329–38.
[15] Achildi O, Grewal H. Congenital anomalies of the esophagus. Otolaryngol Clin North Am 2007;40:219–44.
[16] Sandu K, Monnier P. Congenital tracheal anomalies. Otolaryngol Clin North Am 2007;40:193–217.
[17] Butler C, Speggiorin S, Rijnberg F, Roebuck D, Muthialu N, Hewitt R, et al. Outcomes of slide tracheoplasty in 101 children: a 17-year single-center experience. Cardiovasc Surg 2014;147:1783–90.
[18] Grillo H, Wright C, Vlahakes G, MacGillivray T. Management of congenital tracheal stenosis by means of slide tracheoplasty or resection and reconstruction, with long-term follow-up of growth after slide tracheoplasty. J Thorac Cardiovasc Surg 2002;123:145–52.
[19] Noorily M, Farmer D, Belenky W, Philippart A. Congenital tracheal anomalies in the craniosynostosis syndromes. J Pediatr Surg 1999;34:1036–9.
[20] Scheid S, Spector A, Luft J. Tracheal cartilaginous sleeve in Crouzon syndrome. Int J Pediatr Otorhinolaryngol 2002;65:147–52.
[21] Heimann K, Bartz C, Naami A, Peschgens T, Merz U, Hörnchen H. Three new cases of congenital agenesis of the trachea. Eur J Pediatr 2007;166:79–82.
[22] Floyd J, Campbell D, Dominy D. Agenesis of the trachea. Am Rev Respir Dis 1962;86:557–60.
[23] De Groot-van der Mooren M, Haak M, Lakeman P, Cohen-Overbeek T, van der Voorn J, Bretschneider J, et al. Tracheal agenesis: approach towards this severe diagnosis. Case report and review of the literature. Eur J Pediatr 2012;171:425–31.
[24] Azzie G, Beasley S. Diagnosis and treatment of foregut duplications. Semin Pediatr Surg 2003;12:46–54.
[25] Berrocal T, Madrid C, Novo S, Gutiérrez J, Arjonilla A, Gómez-León N. Congenital anomalies of the tracheobronchial tree, lung, and mediastinum: embryology, radiology, and pathology. Radiographics 2004;24:e17.
[26] Chen J, Holinger L. Congenital tracheal anomalies: pathology study using serial macrosections and review of the literature. Pediatr Pathol 1994;14:513–37.