Congenital Airway Malformations

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Richard G. Azizkhan

Contents

Abstract

Congenital airway malformations comprise a broad array of anomalies extending from the larynx to the distal airway, presenting either acutely postnatally or remaining asymptomatic and therefore undiagnosed for years. Clinical symptoms vary widely, depending on the level

R. G. Azizkhan (✉)
Children's Hospital and Medical Center, University of Nebraska College of Medicine, Omaha, NE, USA
e-mail: Razizkhan@childrensomaha.org

P. Puri (ed.), *Pediatric Surgery*,
https://doi.org/10.1007/978-3-662-43588-5_52

where the obstruction occurs and its severity. Due to the distinct anatomy of the pediatric airway and the possibility of airway symptoms rapidly progressing into life-threatening conditions, early detection, diagnosis, and treatment are imperative.

Keywords
Congenital airway malformations · Laryngomalacia · Subglottic stenosis · Subglottic hemangiomas · Vocal fold paralysis · Laryngeal cleft · Congenital high airway obstruction syndrome · Tracheal stenosis · Tracheal rings · Bronchogenic cysts

Introduction

Congenital airway malformations comprise a broad array of anomalies extending from the larynx to the distal airway. Presentation varies widely and is influenced by the level at which obstruction occurs as well as the severity of obstruction. Given the distinct anatomy of the pediatric airway and the possibility of airway symptoms rapidly progressing to life-threatening airway compromise, early detection, diagnosis, and treatment are imperative. The aim of this chapter is to provide an overview of these anomalies, briefly discussing symptomatology, patient assessment, and current management strategies.

Patient Assessment

Medical History

Assessment of respiratory compromise should begin with a meticulous review of the child's history of airway symptoms. Clinicians should explore circumstances that may elicit the onset of symptoms and should question parents regarding the duration of symptoms and symptom progression. They should also explore possible swallowing or feeding problems, the nature of the child's cry, and the possibility of foreign-body aspiration. Additionally, any history of endotracheal intubation, trauma, or previous cardiac surgery should be carefully reviewed. All of this information may provide important clues as to the underlying etiology and may impact or determine the overall management strategy.

Signs and Symptoms

Patients with mild airway compromise may present with subtle symptoms such as irritability, restlessness, and feeding difficulties. Those with more severe obstruction frequently present with severe suprasternal and intercostal retractions, tachypnea, lethargy, and cyanosis. Stridor, defined as a harsh sound caused by turbulent airflow through a partially obstructed airway, can manifest during the expiratory or inspiratory phases of the respiratory cycle or can be biphasic. Inspiratory stridor usually indicates an airway obstruction in the extrathoracic airway, whereas expiratory stridor generally indicates a problem in the intrathoracic airway. Biphasic stridor typically signifies a fixed glottic or subglottic lesion.

The pitch of stridor, as well as its relationship to the respiratory cycle, is generally helpful in establishing a differential diagnosis and determining priorities for patient assessment. Clinicians should, however, be mindful of the fact that that the degree of stridor does not necessarily reflect the severity of airway obstruction. Even minimal stridor can reflect the lack of airway movement across a critical airway.

Diagnostic Studies

The most critical component of the airway assessment after physical examination is endoscopic evaluation. Depending on the type of suspected airway lesion, either flexible or rigid bronchoscopy or both are performed. To adequately assess the dynamic aspects of some suspected airway lesions (e.g., tracheomalacia), endoscopy should be performed with the patient spontaneously breathing. Because 17% of patients have a synchronous airway lesion, evaluation of the entire airway is essential. Given that up to 45% of

children with congenital airway obstruction also have significant non-airway anomalies, all patients require a thorough overall evaluation.

Imaging studies are helpful in diagnosis as well as patient management (Javia et al. 2016). Computed tomography (CT) and magnetic resonance imaging (MRI) provide a rapid and precise way of assessing and measuring the extent and length of airway narrowing or displacement. These investigations are also helpful in detecting associated mediastinal and pulmonary anomalies. Magnetic resonance angiography (MRA) is valuable in assessing the relationship of mediastinal great vessel anomalies (e.g., vascular rings, pulmonary artery slings) to the airway. Computer software now allows for three-dimensional image reconstruction and is helpful in planning surgical procedures. Echocardiography is valuable in identifying intracardiac defects and most associated great vessel anomalies. Contrast swallow studies are valuable in assessing esophageal motility, aspiration, and some mediastinal lesions that affect the airway. Fiber-optic endoscopic evaluation of swallowing (FEES) is performed to evaluate structural and functional disorders of swallowing and to identify functional problems of the larynx, pharynx, epiglottis, and proximal esophagus.

Congenital Laryngeal Anomalies

Laryngomalacia

Laryngomalacia is the most common congenital laryngeal anomaly and the most common cause of stridor in newborns. This condition is characterized by laxity of both the glottic and supraglottic tissues, which causes the epiglottis, arytenoids, and aryepiglottic folds to collapse and partially obstruct during inspiration. The reported incidence of secondary airway lesions in infants with laryngomalacia varies, with some authors documenting rates as high as 50% (Dickson et al. 2009) and 64% (Cohen et al. 1977).

Inspiratory stridor is the hallmark symptom; it typically presents soon after birth or within the first few days of life. Although stridor is generally mild, it can be exacerbated by feeding, crying, or lying in a supine position. Symptoms generally worsen at 4–8 months, improve between 8 and 12 months, and resolve by 12–18 months, with the majority of affected patients amenable to nonoperative management (Richter and Thompson 2008). When the disorder is severe, however, children may exhibit apnea, cyanosis, severe retractions, and failure to thrive, thus requiring surgical intervention (Rutter 2006). In extremely severe cases, cor pulmonale can develop. Clinical studies indicate that secondary airway lesions lead to an increased incidence of surgical intervention and gastroesophageal reflux disease (GERD) (Dickson et al. 2009).

Flexible transnasal fiber-optic laryngoscopy is used to confirm the diagnosis. Pathognomonic findings include short aryepiglottic folds, with prolapse of the cuneiform cartilages. Collapse of the supraglottic structures is seen on inspiration, and inflammation indicative of reflux laryngitis is also frequently seen (Fig. 1). In some patients, a tightly curled (omega-shaped) epiglottis is observed.

The decision as to whether to intervene surgically is based more so on symptom severity than on the endoscopic appearance of the larynx. For patients with severe symptoms,

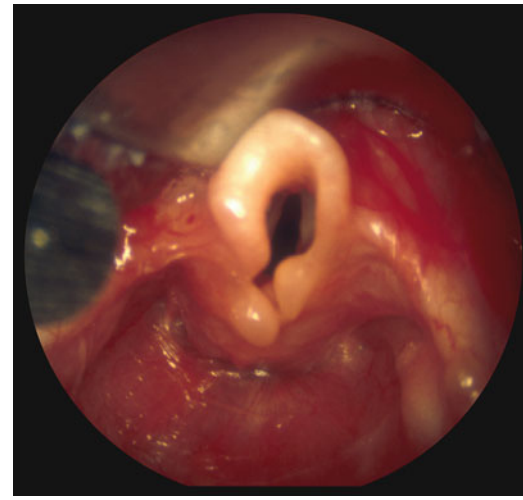

Fig. 1 Endoscopic view of laryngomalacia in an infant showing partial collapse of the supraglottic structures during inspiration

Table 1 Myer-Cotton grading system for subglottic stenosis

Classification	Level of airway obstruction	
	From	To
Grade I	No obstruction	50% obstruction
Grade II	51% obstruction	70% obstruction
Grade III	71% obstruction	99% obstruction
Grade IV	No detectable lumen	

supraglottoplasty (also termed epiglottoplasty) is the preferred operative procedure, with a reported surgical success as high as 94% (Richter and Thompson 2008; Loke et al. 2001; Martin et al. 2005). Both aryepiglottic folds are divided, and one or both cuneiform cartilages may be removed. If the aryepiglottic folds alone are divided, postoperative intubation is usually not required. Patients should be observed overnight in the intensive care unit. Antireflux management is advisable for helping to minimize laryngeal edema. This is especially important in patients with a synchronous airway lesion, who, as mentioned earlier, have an increased incidence of GERD (Dickson et al. 2009; Richter and Thompson 2008). Synchronous airway lesions as well as neurologic conditions and preexisting laryngeal edema can adversely affect operative outcomes (Schroeder et al. 2009). Occasionally, an infant's obstructive symptoms continue despite an adequate postoperative appearance of the larynx. These infants may have an underlying neurologic problem that may become more evident over time, and they are therefore more likely to require tracheotomy placement.

Subglottic Stenosis

Subglottic stenosis (SGS) is an anomaly that involves a narrowing of the subglottic lumen. Although it can be either congenital or acquired, the latter is far more common and is generally a sequela of prolonged intubation of the neonate. Congenital SGS is thought to be caused by failure of the laryngeal lumen to recanalize during embryogenesis. It may occur as an isolated anomaly or may be associated with other congenital head and neck lesions and chromosomal anomalies such as a small larynx in patients with Down syndrome (Azizkhan 2005). In the premature infant, SGS is considered present when this lumen measures 3.0 mm or less in diameter at the level of the cricoid, whereas in the full-term neonate, SGS is defined as a lumen of 4.0 mm or less in diameter at this level.

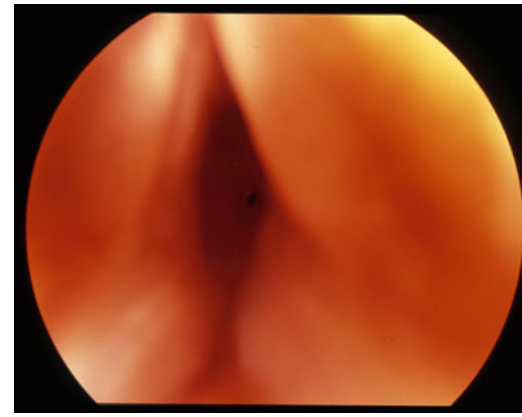

Fig. 2 High-grade subglottic stenosis in a symptomatic neonate

Levels of SGS severity range from mild to severe and are graded based on the Myer-Cotton grading system (Table 1). Patients with mild SGS (no obstruction to 50% obstruction) may present with recurrent upper respiratory infections, often misdiagnosed as croup. In a young child, the greatest obstruction is usually 2–3 mm below the true vocal folds (Rutter 2006). Patients with more severe narrowing may present with acute airway compromise and require endotracheal intubation or tracheotomy placement at delivery (Fig. 2). However, many of these infants, even those with grade III SGS (71–99%), may not be symptomatic for weeks or months. When stridor is present, it initially occurs during the inspiratory phase of respiration. As SGS severity increases, stridor becomes biphasic.

Radiologic evaluation of the non-intubated airway can provide information regarding the site of the stenosis and its extent. Chest X-ray, inspiratory and expiratory lateral soft tissue neck films, and fluoroscopy are helpful in revealing the dynamics of the trachea and larynx. High-kilovoltage airway films identify the classic steeplelike configuration seen in patients with SGS as well as possible tracheal stenosis and are therefore of utmost importance. Flexible endoscopy and rigid endoscopy are used in a complementary fashion for airway evaluation and are both essential. Flexible endoscopy provides critical information regarding the structural dynamics of airflow in the hypopharyngeal and laryngeal airways. Rigid endoscopy provides an assessment of the entire laryngotracheobronchial airway.

In children with mild-to-moderate disease (grade I or II), congenital SGS improves with age. Children with a minor degree of SGS who experience mild symptoms may, nevertheless, benefit from endoscopic intervention. Endoscopic options include radial laser incisions through the stenosis and laryngeal dilatation (Monnier et al. 2005). Outcomes are improved when mitomycin C is used concomitantly with this approach (Smith and Elstad 2009). Less than 50% of these patients require tracheotomy placement to maintain their airway. Children with more severe disease are best managed with open subglottic airway reconstruction. Costal cartilage grafts can be placed through either the anterior or posterior lamina of the cricoid cartilage or both. This may be carried out as a single-stage laryngotracheoplasty (White et al. 2009; de Alarcon and Rutter 2008) or as a two-stage procedure, requiring stenting and placement of a temporary tracheostomy (Monnier 2007). For severe SGS, good results have been achieved by performing partial or complete cricotracheal resection and reconstruction; however, this is a demanding procedure with considerable risks. Successful outcome depends on the management of comorbidities such as GERD, eosinophilic esophagitis, and low-grade tracheal infection.

Vocal Fold Paralysis

Vocal fold paralysis can be congenital or acquired and can occur either unilaterally or bilaterally. Unilateral paralysis is usually an acquired condition caused by damage to the recurrent laryngeal nerve (RLN). Because of the length and course of the left RLN, this is far more likely to be damaged than the right RLN. Acquired paralysis thus generally affects the left vocal fold. Unlike unilateral vocal fold paralysis, bilateral paralysis is usually evident at birth. It is generally idiopathic but often is seen with central nervous system conditions such as hydrocephalus and Chiari malformation of the brainstem.

The diagnosis is made by flexible laryngoscopy with the patient awake. Investigation aimed at finding the underlying cause is then carried out. Stabilization can be achieved with intubation, continuous positive airway pressure (CPAP), or high-flow nasal cannula as an alternative temporizing measure. Almost all infants with bilateral paralysis require tracheotomy placement to ensure a safe and adequate airway. However, up to 50% of children with congenital idiopathic bilateral paralysis experience spontaneous resolution of their paralysis by age 1 (Miyamoto et al. 2005). In view of possible resolution, decannulation is almost always delayed to allow time for this to occur. Children with acquired bilateral paralysis may also experience spontaneous recovery, provided that the RLN was stretched or crushed but otherwise intact.

Because no single surgical approach offers a universally acceptable outcome, a number of surgical approaches have been used for children with bilateral paralysis. These approaches include laser cordotomy, partial or complete arytenoidectomy, and vocal cord medialization or lateralization (open or endoscopically guided) (Sipp et al. 2007; Chen and Inglis 2008). Recently, endoscopic percutaneous suture lateralization has been reported to be a safe and effective nondestructive primary treatment modality for neonatal bilateral vocal fold immobility (Montague et al. 2018). The aim of each of these procedures is to achieve an adequate decannulated airway while maintaining voice and preventing aspiration.

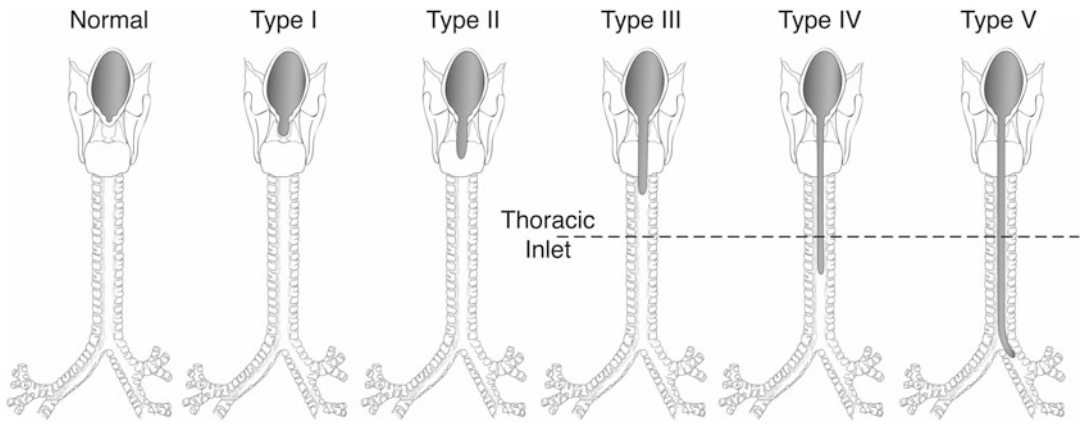

Fig. 3 Posterior laryngeal cleft classification

Posterior Laryngeal Cleft

Posterior laryngeal cleft is a rare congenital anomaly that results from failure of the laryngotracheal groove to fuse during embryogenesis. This anomaly comprises four anatomic subtypes that differ with respect to involvement of the larynx and/or trachea (Fig. 3). Patients frequently have coexisting anomalies, many of which affect the airway. Associated airway anomalies include tracheomalacia (almost always present in varying levels of severity), tracheoesophageal fistula (TEF) formation (20%), laryngomalacia, vocal fold paralysis, SGS, and innominate artery compression. Associated non-airway conditions include anogenital anomalies, cleft lip and palate, congenital heart defects, and GERD, which affects most children. The most common associated syndrome is Opitz-Frias syndrome, characterized by hypertelorism, anogenital anomalies, and posterior laryngeal clefting.

Diagnosis can be extremely difficult, as presenting symptoms vary greatly. In type I and type II clefts, symptoms are often subtle and may mimic those of other disorders such as GERD. Nonetheless, aspiration is a hallmark clinical feature of this spectrum of disease. With more severe clefts, gross aspiration may occur with associated apnea, cyanosis, and even pneumonia. For milder clefts, the symptoms are those of microaspiration, with choking episodes, transient cyanosis, and recurrent chest infections (Rutter 2006; Rutter et al. 2003a). Airway obstruction manifested by stridor may also be present and is caused either by redundant mucosa on the edge of the cleft or a small cricoid ring. In patients with severe tracheomalacia, especially those with an associated TEF, the airway may be significantly compromised. Contrast swallow studies may demonstrate aspiration; however, rigid laryngoscopy and bronchoscopy are essential for definitive diagnosis. The interarytenoid area is specifically probed to determine if a posterior laryngeal cleft is present.

In children who are symptomatic and do not have other more severe anomalies, repair of the posterior laryngeal cleft should be performed as soon as possible to prevent chronic microaspiration with long-term pulmonary sequelae. Depending on the extent of the airway anomaly, tracheotomy and gastrostomy tube placement may be required before definitive surgical repair of the airway. Because of the high incidence of GERD, fundoplication is often required and is preferably performed prior to surgical repair. Most type I and some type II clefts are amenable to endoscopic surgical repair, whereas clefts that extend into the cervical or thoracic trachea require open repair. A transtracheal approach is advised, as it provides optimal exposure of the cleft while protecting the recurrent laryngeal nerves. A two-layer closure is recommended, with the option of performing an interposition

graft if warranted. Type IV long clefts, which extend to the carina or beyond and are often associated with multiple congenital anomalies, are exceedingly difficult to repair and are prone to anastomotic breakdown (Rutter et al. 2003a). Success rates for cleft repair vary significantly (50–90%) depending on both the severity of the cleft and the presence of comorbidities.

Laryngeal Atresia

Congenital High Airway Obstruction Syndrome

Congenital high airway obstruction syndrome (CHAOS) is a rare, life-threatening, prenatally diagnosed condition caused by complete or near-complete obstruction of the larynx and trachea. Fetal lung fluid becomes trapped, causing the lungs to become abnormally distended. This creates massive lung expansion that characteristically everts the hemidiaphragms.

This type of fetal airway obstruction may be caused by multiple etiologies, including laryngeal atresia, laryngeal web, tracheal atresia, and laryngeal cyst (Marwan and Crombleholme 2006). Prenatal findings on ultrasonography include diffuse and enhanced echogenicity of the lungs, dilated airways, and flattened or everted diaphragms with associated fetal ascites and nonimmune hydrops (Fig. 4). A fetus identified with these sonographic features is at significant risk of intrauterine death and faces a high likelihood of mortality should the pregnancy progress to delivery. Although US provides a good initial assessment of CHAOS, MRI is clearly superior in identifying severity and the level of airway obstruction and optimizes planning for airway management at delivery (Mong et al. 2008). These patients all require delivery by the ex utero intrapartum technique (EXIT) procedure (Laje et al. 2016). This procedure maintains placental circulation to the fetus while securing the airway at the time of delivery (Marwan and Crombleholme 2006). Securing the airway may include a full endoscopic diagnostic assessment and tracheostomy.

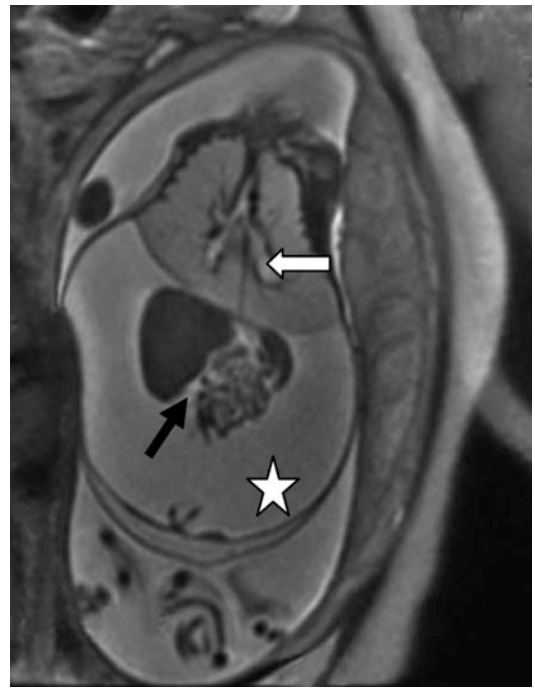

Fig. 4 Fetal ultrasonography at 27 weeks gestation demonstrating findings consistent with the diagnosis of congenital high airway obstruction: enlarged echogenic lungs, dilated airway (*white arrow*), flattened or everted diaphragms, and fetal ascites (*white star*) and hydrops. Fetal liver and intestines marked with a *black arrow*

For the newborn diagnosed with CHAOS, securing and maintaining the airway are the highest priority. These patients are almost always extremely ill and require a prolonged period of critical care and ventilatory support. Once the infant's cardiorespiratory status is stable and other critical or potentially life-threatening anomalies are ruled out, careful endoscopic evaluation of the airway precedes elective laryngotracheal reconstruction; however, consensus has not been reached as to optimal timing of airway reconstruction. Although a functional airway can be constructed, patients do not always attain intelligible speech capabilities.

Diagnosis in the middle of the second trimester generally correlates with a poor perinatal outcome. A fetus presenting in the third trimester with CHAOS in the absence of associated anomalies or hydrops is likely to have incomplete obstruction and is therefore more likely to survive.

Subglottic Hemangioma

Hemangiomas of infancy (also referred to as infantile hemangiomas) are the most common vascular tumors, affecting one in ten white infants in North America (Mulliken et al. 2000) and occurring with a threefold female preponderance. These benign lesions usually follow a predetermined phase of growth (proliferation) and later tumor regression (involution). The involutive phase occurs at 12–18 months and is generally complete by the first decade of life.

Hemangiomas generally present cutaneously but can occur in any organ or anatomic site. Airway hemangiomas most frequently occur in the subglottis; however, they are also seen in the glottic and supraglottic regions. Their natural history generally mirrors that of cutaneous lesions. More than 50% of patients with a subglottic hemangioma also have cutaneous hemangiomas, which provide an indication for the possible presence of a synchronous subglottic lesion. Patients at the highest risk for a subglottic hemangioma include those with a hemangioma occurring in a beard distribution (Orlow et al. 1997) (Fig. 5a) and those with PHACE syndrome, which is characterized by *p*osterior fossa abnormalities, *h*emangiomas of the cervicofacial region that are usually plaque-like and segmental, *a*rterial defects, *c*ardiac and aortic arch defects, and *e*ye abnormalities (Perkins et al. 2009; O-Lee and Messner 2008).

As a subglottic hemangioma undergoes proliferation, progressive worsening of the airway usually occurs. Presenting symptoms include biphasic stridor with retractions. The degree of obstruction varies and can be exacerbated by certain positions or crying, both of which increase venous pressure and lead to vascular engorgement. When airway narrowing is severe, apnea, cyanosis, and "dying spells" may result.

The diagnosis is based on medical history and findings on airway endoscopy. Lesions are typically asymmetric and may be covered by a normal smooth mucosa (Fig. 5b). Because of the risk of hemorrhage, biopsy is not advised. Most patients require treatment, and combining various treatment modalities is often essential. At Cincinnati Children's Hospital Medical Center, symptomatic patients with significant stridor are currently managed with systemic steroids combined with propranolol, a nonselective beta-blocker used to treat infants with cardiovascular conditions. In recent years, numerous publications have documented dramatic results with the use of propranolol for severe subglottic hemangiomas (Truong et al. 2010; Jephson et al. 2009; Léaute-Labrèze et al. 2008; Denoyelle et al. 2009; Gunturi et al. 2013; Leboulanger et al. 2010), thereby changing the paradigm of

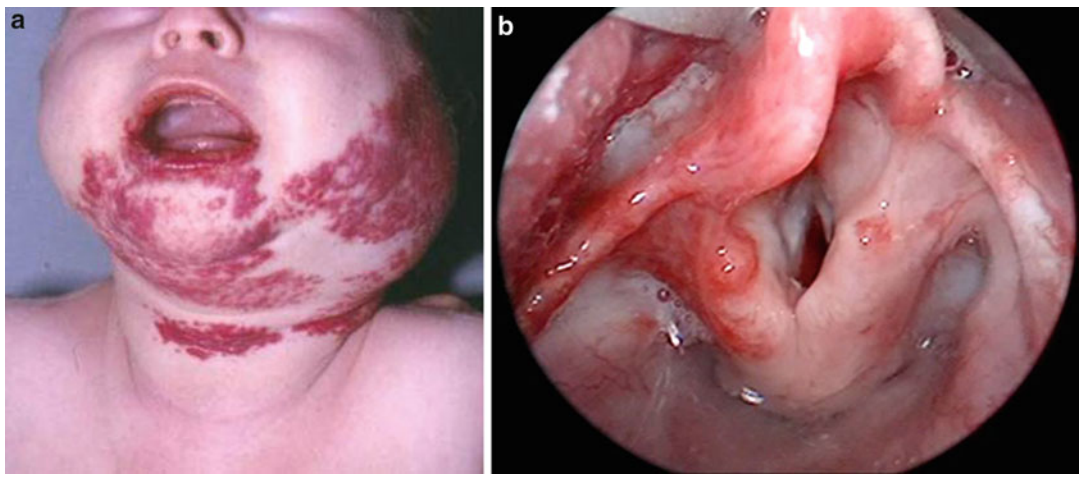

Fig. 5 (**a**) Patient with multiple cutaneous lesions in a beard distribution. (**b**) Endoscopic view of a subglottic hemangioma

both pharmacologic and surgical treatment. A systematic review of 41 propranolol studies published between June 2008 and June 2012 that collectively included more than 1200 patients (mean age, 6.6 months) with a spectrum of potentially life- or function-threatening hemangiomas at various anatomic sites showed an impressively high efficacy rate (98%) and a low rate of serious adverse advents (Marqueling et al. 2013).

For critical airways, some surgeons still advocate laser fulguration or translaryngeal resection, whereas others place a tracheotomy below the lesion, with the expectation of removal following involution of the hemangioma (Perkins et al. 2009; Bajaj et al. 2006).

Anomalies of the Trachea and Bronchi

Tracheal Agenesis and Atresia

Tracheal agenesis is very rare, occurring in 1 in 50,000 to 1 in 100,000 live births and usually results in fatal outcome (Boogaard et al. 2005; Varela et al. 2018). The cervical trachea is usually absent. The bronchus or the carina is connected to the esophagus. Floyd proposed an anatomic classification in three types (Floyd et al. 1962):

Type I: representing the 20%. There is agenesis of the upper trachea. Bronchus is normal. There is a tracheoesophageal fistula.

Type II: the most frequent representing 60% of all the cases reported. There is a complete tracheal agenesis. Bronchus is normal, and there is a fistula between the carina and esophagus.

Type III: the bronchus arises from the esophagus separately.

The presence of a tracheoesophageal or bronchoesophageal fistula is important for salvage treatment of a critically ill neonate with tracheal agenesis, because it allows esophageal intubation and mechanical ventilation.

Tracheal Stenosis and Webs

Tracheal stenosis encompasses a broad spectrum of rare tracheal anomalies. Affected segments of the trachea differ in the degree and extent of stenosis, which can range from extremely thin webs to more severe long segments of stenosis affecting the entire airway.

Tracheal Webs

Tracheal webs involve an intraluminal soft tissue stenosis of the trachea. These webs may be membranous or consist of thick, relatively rigid tissue. Patients typically present with biphasic stridor or expiratory wheezing, and the severity of these symptoms depends on the degree of the stenosis. Thin webs can be easily managed by hydrostatic balloon dilatation (Ganzer 1987). For thicker webs that are not associated with underlying cartilage deformity, laser ablation is often used (Azizkhan et al. 1990). The carbon dioxide (CO_2) or potassium-titanyl-phosphate (KTP) laser is beneficial for treating lesions in the proximal trachea. Lesions in the distal airway are best managed with the KTP laser, which can be used through small fiber-optic cables. For children with a web greater than 1 cm in length or those in whom the airway cartilage is thought to be structurally deficient or anomalous, operative treatment with segmental tracheal resection or slide tracheoplasty is usually carried out.

Cartilaginous Ring Aplasia

Cartilaginous ring aplasia is an extremely rare anomaly in which only a small region of the trachea lacks cartilage, creating a distinct anatomic area that is both malacic and stenotic. The remainder of the trachea is unaffected, and most children do not have coexisting congenital anomalies. Management entails segmental resection of the trachea, which successfully restores the airway.

Tracheal Cartilaginous Sleeve

Tracheal cartilaginous sleeve is also extremely rare. In children with this anomaly, discrete cartilaginous rings are replaced by a fused cartilaginous cylinder, with or without a membranous portion. It is typically seen in children with craniosynostosis syndromes such as Pfeiffer, Apert, Crouzon, and Goldenhar (Davis et al. 1992; Hockstein et al. 2004; Elloy et al. 2006).

Neonates usually exhibit respiratory illness. Patients presenting in early infancy often experience acute respiratory symptoms, which may include biphasic stridor with respiratory distress, cough, and frequent respiratory infections. Because of tracheal rigidity, the mechanism for clearing secretions is impaired.

On endoscopy, the anterior tracheal wall appears smooth, though the membranous posterior tracheal wall may be normal, stenotic, or absent. CT and MRI are sometimes useful in determining the extent of the lesion. Tracheotomy placement may be used as a temporizing measure; however resection and repair are imperative to achieve normal function.

Complete Tracheal Rings

Although rare, complete tracheal rings are the most common congenital tracheal stenosis (Windsor et al. 2016). With this spectrum of potentially life-threatening anomalies, either the trachea alone or both the trachea and bronchi are significantly narrowed. The tracheal cartilage in these patients is abnormally shaped and forms complete rings (Fig. 6). More than 50% of infants have a segmental stenosis. The clinical manifestations of complete tracheal rings vary from life-threatening respiratory distress during the perinatal period to subtle symptoms of airway compromise in older children. Most symptomatic infants exhibit deterioration of respiratory function over the first few months of life. Symptoms include stridor, retractions, cough, and alterations of cry. Atypical and persistent wheezing and rhonchi and sudden death can also occur. More than 80% of children with complete tracheal rings have other and often multiple congenital anomalies; 50% have congenital heart disease with or without great vessel anomalies (Rutter 2006).

In some patients, placement of an endotracheal tube may further exacerbate respiratory distress by causing acute swelling and inflammation of the mucosa. Partially obstructing tracheal lesions also may become life-threatening following the onset of a respiratory infection. In an infant or child with an abnormal trachea, the cross-sectional area of airway can be significantly decreased with as little as 1 mm of edema. This accounts for the rapid worsening of symptoms in some children with acute inflammatory conditions and coexisting tracheal narrowing.

Prompt diagnostic evaluation to define tracheobronchial anatomy is essential. An initial high-kilovolt airway film may indicate stenosis; however, bronchoscopy is required to reveal the precise location and extent of the stenosis.

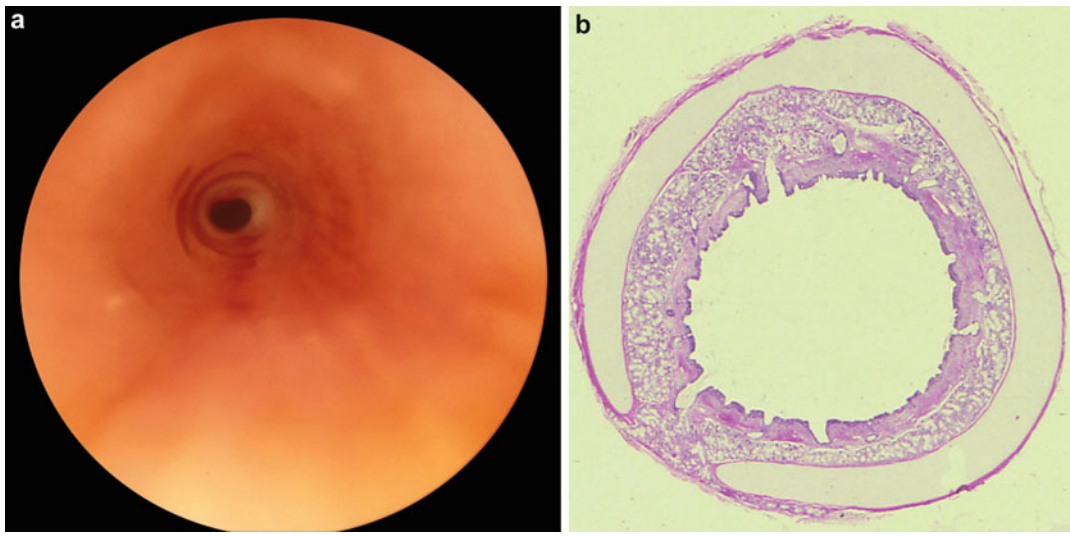

Fig. 6 Congenital tracheal stenosis. (**a**) Endoscopic view demonstrating complete tracheal rings. (**b**) Histology showing cartilaginous rings that are circumferential

Bronchoscopy should be performed with extreme caution, using the smallest possible telescopes, and any airway edema in the region of the stenosis may turn a narrow airway into a critical airway (Rutter 2006). CT scans provide a rapid and precise method of measuring the extent and length of airway narrowing or displacement. Three-dimensional reconstruction of the airway and its relationship to the great vessels aids in operative planning. Furthermore, with new software enhancements, virtual bronchoscopic images can be obtained. These images are particularly useful in assessing the airway distal to the obstruction (Fig. 7). MRI is also valuable in evaluating the relationship of the mediastinal great vessels to the airway. Echocardiography is used mainly to determine whether intracardiac defects are present and can identify most coexisting pulmonary artery slings.

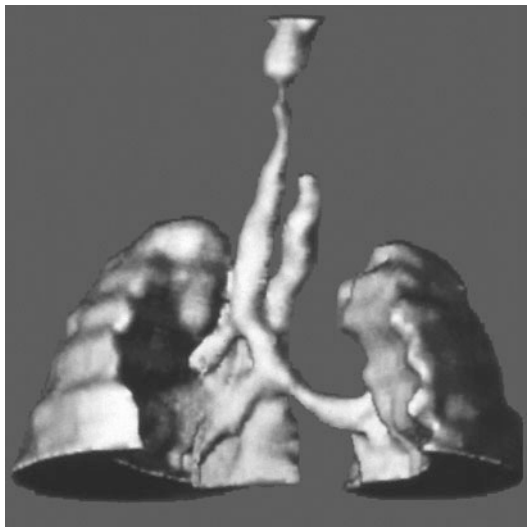

Fig. 7 CT scan with three-dimensional reconstruction to demonstrate anatomy of the trachea in a patient with congenital tracheal stenosis involving a significant portion of the trachea

Approximately 10% of patients with complete tracheal rings are minimally symptomatic and can be managed nonoperatively, though they require ongoing observation. Most children must undergo tracheal reconstruction (Rutter et al. 2004). Repair of coexisting anomalies such as pulmonary artery sling or vascular ring should be carried out concurrent with the tracheal repair. Although patch tracheoplasty was historically the preferred procedure for long segments of narrowing, slide tracheoplasty is now the procedure of choice for both short- and long-segment stenosis (Fig. 8) (Rutter et al. 2003b; de Alarcon and Rutter 2012). This approach results in significantly less morbidity

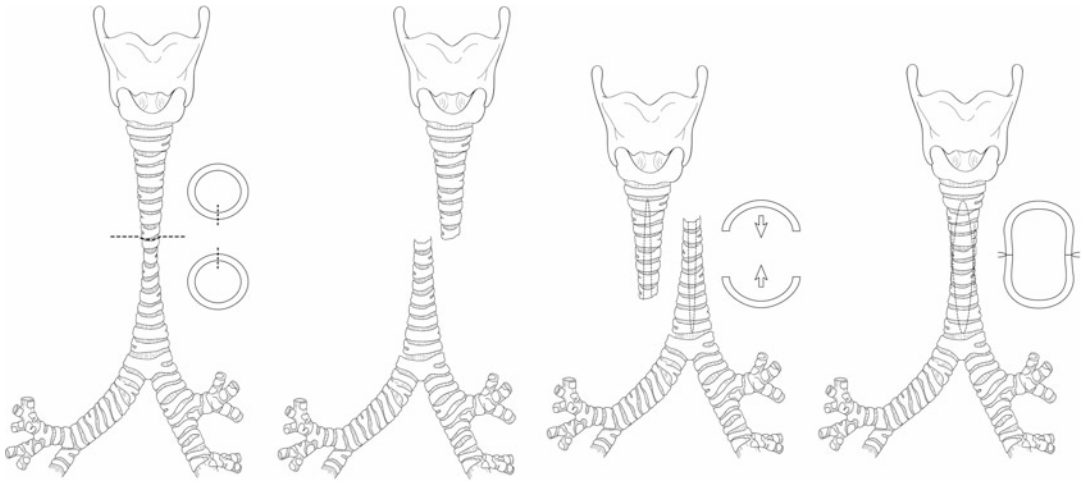

Fig. 8 Slide tracheoplasty. The trachea is transversely divided at the midpoint of the tracheal stenosis. After proximal and distal tracheal mobilization, the posterior portion of the cephalic trachea segment and the anterior portion of the caudal tracheal segment are incised. The two tracheal segments are then overlapped and obliquely sutured together

than other tracheal reconstruction techniques and is adaptable to all anatomic configurations of complete tracheal rings. Slide tracheoplasty uses only autologous tracheal tissue and is performed by transecting the trachea into two equal segments. The anterior wall of the lower half of the trachea and the posterior wall of the upper trachea are incised. These segments are then slid over each other and anastomosed with 5-0 monofilament and absorbable sutures. Postoperatively, the cross-sectional area of the airway has a fourfold increase, and the length of the involved airway decreases by half. Airflow is increased 16-fold.

Postoperatively, endotracheal intubation is generally required for 1–2 days, though some patients with parenchymal pulmonary disease require longer ventilatory support. During the perioperative period, unnecessary movements of the endotracheal tube or unplanned extubation must be avoided so to minimize the risk of damage to the newly reconstructed airway. Nasotracheal intubation is preferred, as the endotracheal tube can be more securely stabilized. Patients require continuous monitoring, careful pulmonary toilet, and endoscopic removal of any obstructing granulation tissue. Immediately prior to extubation, the integrity and patency of the reconstructed airway are assessed by flexible fiberoptic endoscopy through the endotracheal tube, thus ensuring a safe extubation.

Although airway configuration following slide tracheoplasty may resemble a figure eight, this does not indicate airway obstruction. The trachea generally remodels to a normal oval shape within 1 year of reconstruction. Long-term survival is currently 90% in our institution. Mortality is usually associated with severe comorbidities such as cardiac disease rather than airway complications.

Tracheal Diverticulum and Tracheal Bronchus

Tracheal diverticulum and tracheal bronchus are relatively common embryologic abnormalities of early tracheal budding. Tracheal diverticulum resembles a bronchus, though it originates from the trachea and ends blindly or communicates with a rudimentary lung. Tracheal bronchus most often affects the right upper lobe bronchus and may connect to an isolated intrathoracic lung segment or the apical segment of an upper lobe. Both anomalies frequently occur along with other tracheal, esophageal, and pulmonary anomalies. Diagnosis is established by airway endoscopy. Most children are asymptomatic and do not require treatment. Pneumonia and respiratory distress may be the presenting symptoms during the neonatal period. These symptoms are almost always associated with stenosis of a bronchus or other lung anomalies. Resection of involved lobe and bronchus in these patients is generally curative.

The most common cause of tracheal diverticulum is iatrogenic, following division of a TEF where a small remnant of the esophagus is left on the tracheal side. These defects can be readily managed with endoscopic resection (Cheng and Gazali 2008; Shah et al. 2009; Johnson et al. 2007).

Airway Malacia

Airway malacia is a condition in which the structural integrity of either the trachea or bronchi or both is weakened and the cartilaginous rings of the airway lack the rigidity required to avoid airway collapse during expiration. Malacia may be localized or occur diffusely throughout the airway. Tracheomalacia is the most common congenital tracheal anomaly. This condition may occur in isolation or in conjunction with other congenital anomalies. TEF, esophageal atresia, and posterior laryngeal clefts are particularly common associations (McNamara and Crabbe 2004). Premature neonates and children with chronic lung disease are at high risk for developing combined severe tracheobronchomalacia.

Presenting symptoms vary depending upon the severity, duration, and region of airway involvement. Most children are either asymptomatic or minimally symptomatic, and most cases involve posterior malacia of the trachealis, with associated broadening of the tracheal rings. Presenting symptoms may include a honking cough, stridor, wheezing, respiratory distress when agitated, and cyanosis. Some children are

misdiagnosed with allergic asthma and unsuccessfully treated with bronchodilators. Diagnosis is best established by bronchoscopy, with the patient breathing spontaneously; this demonstrates dynamic distortion and compression of the trachea. In children who are minimally symptomatic, symptoms often resolve by age 3. These children are managed with observation alone. Children who experience symptom progression require medical or surgical intervention (McNamara and Crabbe 2004). For some patients, respiratory monitoring with nasal CPAP may be sufficient to effect improvement.

Segmental tracheal involvement is managed with endoscopic or open aortopexy, with thymectomy and anterior suspension of the ascending arch of the aorta to the posterior periosteum of the sternum (Perger et al. 2009). More diffuse malacia may require tracheotomy placement with positive pressure ventilation over a long duration. For patients with severely problematic tracheobronchomalacia that is unresponsive to nonoperative therapy or unsuitable for surgical treatment, intratracheal stents are placed; however, this approach is associated with serious complications such as stent collapse, stent dislodgement, or, rarely, stent erosion into the great vessels (Wallis and McLaren 2018). Additionally, stent removal can cause tracheal tearing or major hemorrhage.

Esophageal Bronchus

Isolated bronchial connection between the esophagus and the airway is extremely rare and occurs more frequently in females (2:1). Associated cardiac, genitourinary, vertebral, and diaphragmatic anomalies are common. Esophageal bronchus is thought to develop from a supernumerary lung bud arising from the esophagus. Most commonly, a lower lobe is aerated by this ectopic bronchus; however, an entire main bronchus and lung may be affected. As in pulmonary sequestration anomalies, the pulmonary vasculature may be abnormal, with the arterial supply coming off the aorta and venous drainage going into either the systemic or pulmonary veins.

Inadequate bronchial drainage usually results in recurrent pulmonary infection and parenchymal damage (Tsugawa et al. 2005). Nonetheless, some patients remain undiagnosed until adolescence or adulthood despite recurring pneumonias and persistent radiographic abnormalities. Although radiographic findings vary with the segment of the lung affected by the anomaly, collapse, consolidation, cavitation, and cyst formation within the pulmonary parenchyma are commonly seen. The diagnosis is confirmed by a contrast study of the esophagus, though false-negative results sometimes occur. Excision of the abnormal lung and closure of the bronchoesophageal fistula is the treatment of choice in patients beyond the neonatal period. Prognosis depends on early diagnosis and treatment and the severity of associated anomalies. Bronchotracheal reconstruction has been successfully accomplished in neonates diagnosed with esophageal bronchus (Michel et al. 1997).

Bronchogenic Cyst

Bronchogenic cysts stem from aberrant embryogenesis of the bronchial tree in which a segment of the lung bud develops independently. The walls of the cyst often contain fibrous tissue and cartilaginous remnants, while the internal surface consists of ciliated columnar epithelium (Stocker 2009). Lesions usually expand, causing extramural compression of the airway. The most commonly seen symptom in infants is respiratory distress. Coughing, wheezing, or chest pain also may be present. A plain chest radiograph may suggest the presence of a bronchogenic cyst. A CT scan or MRI is valuable in establishing a definitive diagnosis (Fig. 9a). Patients are successfully managed by open or thoracoscopic resection (Fig. 9b) (Koontz et al. 2005; Hirose et al. 2006).

Bronchial Atresia and Bronchial Lobar Agenesis

Bronchial Atresia

Localized bronchial atresia is a rare abnormality in which the atretic bronchus impedes the flow of

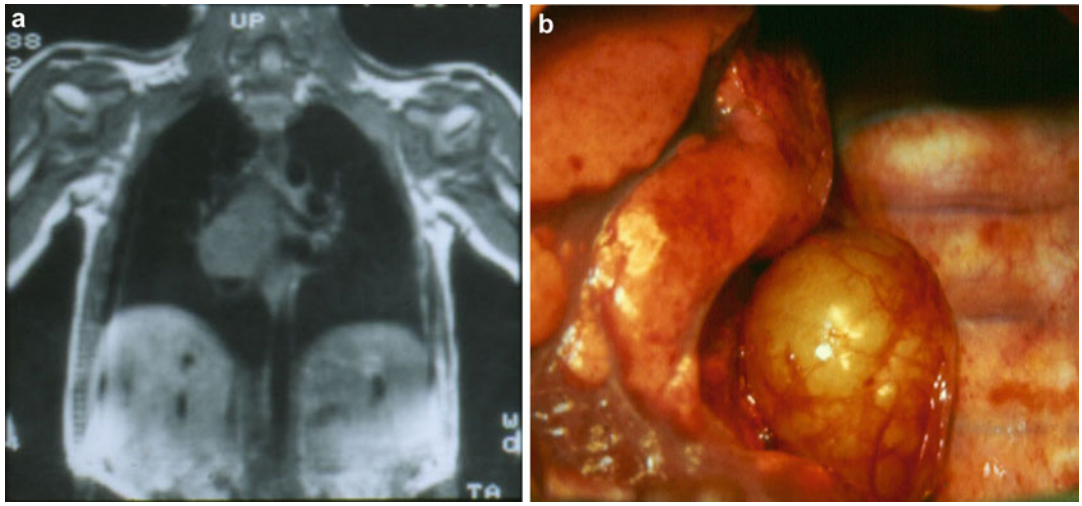

Fig. 9 (**a**) MRI demonstrating a right-sided bronchogenic cyst just below the right main bronchus. (**b**) Thoracotomy view of a bronchogenic cyst. The parietal pleura has been opened in the process of removal

secretions and air from the distal lung to the main tracheobronchial tree. This condition may mimic lobar emphysema or a mediastinal mass (Morikawa et al. 2005). At birth, the affected lung retains fluid. A CT chest scan may show a cystic central mucocele and is valuable in distinguishing bronchial atresia from a bronchogenic cyst or lobar emphysema. Although children may initially be asymptomatic, secretions trapped in the lung may result in serious pulmonary infection. Surgical resection of the affected lobe restores normal lung function.

Bronchial Lobar Agenesis

Bronchial agenesis is more commonly seen than tracheal agenesis, and unlike tracheal agenesis, it is compatible with life. Several anatomic configurations have been described; these include lobar, bronchial, and parenchymal agenesis. In the most severe form, complete agenesis of the lung and its bronchus and blood supply may occur (Morikawa et al. 2005). Also, there may be a rudimentary bronchus and aplasia of the lung. As with most airway malformations, children may have coexistent congenital anomalies. Most commonly, these anomalies involve the skeletal, cardiovascular, gastrointestinal, and genitourinary systems. Diagnosis is established by chest radiographs and airway endoscopy. Most patients can be managed nonoperatively. Bronchial agenesis is important to identify, as it may mimic other airway and cardiovascular anomalies requiring treatment (e.g., bronchial stenosis or extraluminal airway obstruction by tumors or masses).

Bronchial Stenosis

Isolated congenital bronchial stenosis is extremely rare, with causality including compressive vascular, cardiac, and congenital cystic lesions or soft tissue cartilaginous stenoses. Symptoms and treatment vary, depending on both the severity and anatomic location of the lesion. Surgical management includes resection and reconstruction of the bronchus and slide bronchoplasty (Antón-Pacheco et al. 2007; Grillo et al. 2002).

Acquired bronchial stenosis is more common than its congenital manifestation and is a significant cause of morbidity and mortality in infants who have undergone prolonged intubation and respiratory support. Most cases can be managed with endoscopic balloon dilatation or laser resection (Azizkhan et al. 1990).

Conclusion and Future Directions

Congenital airway malformations may be a diagnostic and therapeutical challenge as they include a wide array of anomalies, some of them extremely rare, with a broad spectrum of symptoms. A close collaboration between pediatric surgeons, neonatologists, radiologists, and anesthetists is crucial for an optimal treatment of these potential life-threatening conditions to prevent morbidity and mortality.

Cross-References

- ▶ Embryology of Congenital Malformations
- ▶ Pediatric Respiratory Physiology
- ▶ Specific Risks for the Preterm Infant
- ▶ Stridor in the Newborn
- ▶ Tracheostomy in Infants
- ▶ Vascular Rings

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