Congenital Airway Malformations

33

Patricio Varela and Richard Azizkhan

33.1 Introduction

As an initial step in evaluating an infant with respiratory compromise, a clinician must thoroughly review the history of the child's airway symptoms. This review may provide information that helps in identifying the underlying etiology, which may determine or have an important impact on management strategy. Close attention should be paid to circumstances that trigger the onset of symptoms, the duration of symptoms, and symptom progression over time. Questioning parents about a history of dysphagia or feeding problems, the nature of their child's cry, and the possibility of foreign body aspiration can also yield useful information. Additionally, any previous history of endotracheal intubation, trauma, or cardiopulmonary abnormalities should be carefully reviewed.

Airway obstruction may range from subtle to severe. Less severe airway compromise frequently manifests in subtle symptoms such as irritability, whereas more severe obstruction is likely to manifest in severe suprasternal and intercostal retractions, tachypnea, lethargy, and cyanosis. Stridor, a harsh sound caused by turbulent airflow through a partial obstruction of the airway, is the most important symptom of upper airway obstruction. Depending on the location of the obstruction in the upper airway, this symptom can be present during either the expiratory or inspiratory phase of the respiratory cycle or during both of these phases. The characteristics of stridor as well as its relationship to the respiratory cycle are generally helpful in establishing a differential diagnosis and in setting the priorities for diagnostic evaluation.

33.2 Diagnostic Evaluation

Conducting a complete and thorough endoscopic evaluation is the most critical part of airway assessment. This evaluation generally incorporates both flexible and rigid bronchoscopy. For the evaluation of certain types of lesions (e.g., tracheomalacia), airway assessment is performed with the patient awake or lightly sedated and spontaneously breathing. Clinicians should be aware of the fact that 17% of patients have a second airway lesion. Evaluation of the entire airway is thus essential. As up to 45% of children with congenital airway obstruction also have other significant nonairway anomalies, these

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patients require meticulous and complete overall investigation.

Complementary imaging studies are useful in both diagnosis and patient management. Computed tomography (CT) and magnetic resonance (MR) imaging studies provide rapid and precise methods of measuring the extent and length of airway narrowing or displacement. Additionally, they assist in identifying associated mediastinal and pulmonary anomalies. In particular, MR angiography is useful in assessing the relationship of mediastinal great vessel anomalies (e.g., vascular rings, pulmonary artery slings) to the airway. Newer computer software allows for three-dimensional image reconstruction and is helpful in planning surgical procedures. Echocardiography is primarily used to assess the presence of intracardiac defects and can detect the majority of associated mediastinal vascular anomalies. Contrast swallow studies are valuable in assessing esophageal motility, aspiration, and some mediastinal lesions that impact 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.

33.3 Congenital Laryngeal Anomalies

33.3.1 Laryngomalacia

33.3.1.1 Pathogenesis

Laryngomalacia is characterized by laxity of both the glottic and supraglottic tissues, causing the epiglottis, arytenoids, and aryepiglottic folds to collapse and partially obstruct during inspiration. This malformation is the most common congenital laryngeal anomaly. It accounts for 60–75% of laryngeal problems in the neonate and is also the most common cause of stridor in the neonate. Stridor caused by laryngomalacia usually is evident soon after birth or within the first few days of life. It is generally mild but can be exacerbated by feeding, crying, and lying in a supine position. Fifty percent of children with laryngomalacia experience a worsening of stridor during the first 6 months of life. Children with severe laryngomalacia may have apnea, cyanosis, severe retractions, and failure to thrive. Cor pulmonale has been reported in cases that are extremely severe (Backer et al. 1997; Hysinger and Panitch 2016; Ngerncham et al. 2015).

33.3.1.2 Diagnosis

Diagnosis is confirmed by flexible transnasal fiber-optic laryngoscopy, which reveals 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. 33.1). Although symptoms spontaneously resolve by the age of 1 year, the infant with severe laryngomalacia may die of asphyxiation.

The need for surgical intervention and the type of surgery required are primarily based on the severity of symptoms. For severe symptoms, supraglottoplasty (also termed epiglottoplasty) is the operative procedure of choice. Redundant mucosa of the aryepiglottic folds is excised and one or both cuneiform cartilages also may be removed. If the aryepiglottic folds alone are divided, postoperative intubation is generally not required. Reflux management is useful in helping to minimize laryngeal edema. Patients are observed overnight in the intensive care unit.

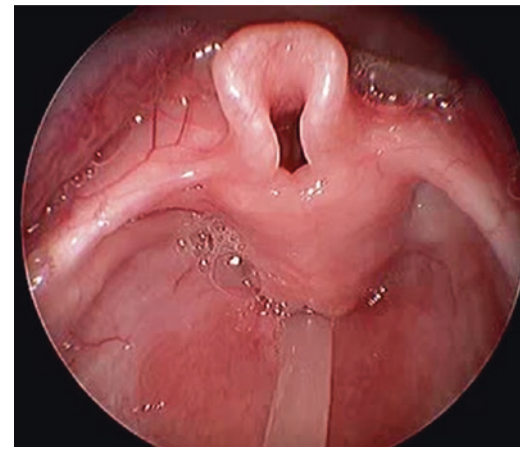

Fig. 33.1 Endoscopic view of laryngomalacia in an infant showing partial collapse of the supraglottic structures during inspiration

Occasionally, the infant's obstructive symptoms continue despite an adequate appearance of the larynx after surgery. Such cases are sometimes related to underlying neurologic problems and have a high likelihood of requiring tracheotomy placement (Austin and Ali 2003; Azizkhan 2005; Rutter et al. 2003).

33.3.2 Subglottic Stenosis

Subglottic stenosis (SGS) involves a narrowing of the subglottic lumen. This malformation may be either congenital or acquired, with the latter more commonly seen and generally a sequela of prolonged intubation during the neonatal period.

33.3.2.1 Pathogenesis

It is thought to be caused by the failure of the laryngeal lumen to recanalize during embryogenesis. Incomplete recanalization results in various degrees of stenosis that range from mild to severe; levels of severity are graded according to the Myer-Cotton grading system (Table 33.1). Mild SGS (no obstruction to 50% obstruction) may manifest in recurrent upper respiratory infections, which frequently are diagnosed as croup. More severe cases (71–99% obstruction) may present with acute airway compromise and require endotracheal intubation or tracheostomy placement at the time of delivery. Many infants with severe obstruction may, however, remain asymptomatic for weeks or months. When stridor is present, it is initially inspiratory. As the severity of obstruction increases, stridor becomes biphasic (Herrera et al. 2007; Ngerncham et al. 2015).

33.3.2.2 Diagnosis

In the full-term neonate, SGS is defined as a lumen of 4 mm or less in diameter at the level of the cricoid; in the premature infant, it is considered present when this lumen measures 3 mm or less. Congenital SGS (Fig. 33.2) may arise as an isolated anomaly or may be associated with other congenital head and neck lesions and syndromes such as a small larynx in a patient with Down syndrome.

Table 33.1 Myer-Cotton grading system for subglottic stenosis level of airway obstruction

Classification	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	

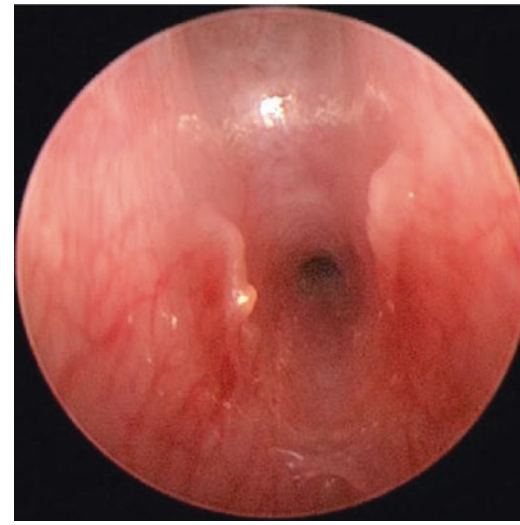

Fig. 33.2 Very high grade of congenital subglottic stenosis in a symptomatic neonate

Radiologic evaluation of the nonintubated airway can provide information regarding the location and extent of the stenosis. Useful studies include a chest X-ray, inspiratory and expiratory lateral soft tissue neck films, and fluoroscopy to demonstrate the dynamics of the trachea and larynx. High-kilovoltage airway films are particularly important as they identify the characteristic steeple-like configuration seen in patients with SGS as well as possible tracheal stenosis. Flexible and rigid endoscopies are the gold standard of evaluating the airway. Flexible endoscopy documents the dynamics of the hypopharyngeal and laryngeal airways, whereas rigid endoscopy provides a clear image of the entire laryngotracheobronchial airway.

In patients with mild to moderate disease, congenital SGS improves with age. Less than 50% of these patients require tracheotomy placement to maintain their airway. Children with significant airway obstruction are best managed with open airway reconstruction. Costal cartilage grafts are placed through either the anterior or posterior lamina of the cricoid cartilage or both.

Stenting and placement of a temporary tracheostomy may be necessary. Recently, superior results for the management of severe SGS have been obtained by performing cricotracheal resection. Successful management depends on the presence of comorbidities such as gastroesophageal reflux (GER), eosinophilic esophagitis, and low-grade tracheal infection.

33.3.3 Vocal Cord Paralysis

Vocal cord paralysis is the third most common cause of neonatal stridor. This condition can be either 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, whereas bilateral vocal cord paralysis is usually evident at birth.

33.3.3.1 Pathogenesis

Bilateral paralysis is generally idiopathic, it is frequently seen with central nervous system problems such as hydrocephalus and Chiari malformation of the brainstem. Most children with bilateral paralysis present with significant airway compromise, though they do not aspirate (Ngerncham et al. 2015).

33.3.3.2 Diagnosis

Diagnosis is made by awake flexible laryngoscopy. Subsequent investigation for the underlying cause should then be carried out. Stabilization can be achieved with intubation, continuous positive airway pressure (CPAP), or high-flow nasal cannula as an alternative temporizing measure. Most infants (90%) affected bilaterally require tracheotomy placement. Up to 50% of children with congenital idiopathic bilateral vocal cord paralysis experience spontaneous resolution of their paralysis by the age of 1 year. As such, decannulation is almost always delayed to allow time for this to occur.

A number of surgical procedures have been used, in the past, for congenital bilateral paralysis, including laser cordotomy and partial or complete arytenoidectomy. Other procedures have also been proposed: vocal process lateralization (open or endoscopically guided), endoscopic posterior graft placement, or anterior posterior cricoid split. Each of these options aims to achieving an adequate decannulated airway while maintaining voice and preventing aspiration.

33.3.4 Posterior Laryngeal Cleft

33.3.4.1 Pathogenesis

Posterior laryngeal cleft is a rare congenital malformation that results from embryologic failure of the laryngotracheal groove to fuse.

33.3.4.2 Classification

This malformation comprises six anatomic subtypes that differ with respect to involvement of the larynx and trachea (Fig. 33.3). Other associations, many of which affect the airway, are common. Such anomalies include tracheomalacia (always present in varying levels of severity), tracheoesophageal fistula (20%), laryngomalacia, vocal cord paralysis, SGS, and innominate artery

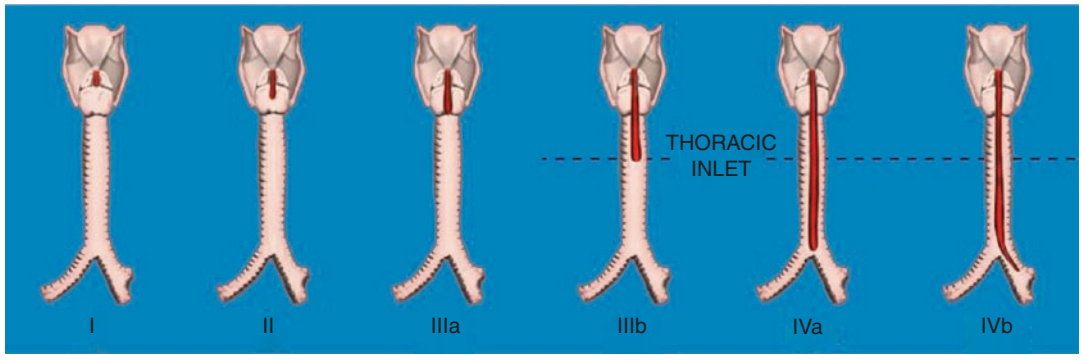

Fig. 33.3 Posterior Laryngeal Cleft Classification

compression (Bennett and Holinger 2003; Davies and Cywes 1978; Eriksen et al. 1990; Evans 1985; Grillo et al. 2002; Kluth et al. 1987; Rahbar et al. 2006, 2009 ; Sandu and Monnier 2006).

Associated conditions that do not involve the airway include GER, which is present in most children, cleft lip and palate, congenital heart defects, and hypospadias. The most common associated syndrome is Opitz-Frias syndrome, which is characterized by hypertelorism, anorectal malformations, genital urinary anomalies, and laryngeal clefting.

33.3.4.3 Diagnosis

Diagnosis can be extremely challenging and elusive, as presenting symptoms vary greatly and are not specifically diagnostic. Symptoms are often subtle and may mimic those of other disorders (e.g., GER). Some patients present early with feeding problems, choking, chronic coughing, wheezing, cyanotic spells, and apnea. There may be associated stridor either due to redundant mucosa on the edge of the cleft or a small cricoid ring. Severe tracheomalacia may also significantly compromise the airway, especially in children with an associated tracheoesophageal fistula. Although contrast swallow studies may demonstrate aspiration, definitive diagnosis requires rigid laryngotracheal bronchoscopy, with the interarytenoid area being specifically probed to determine if a posterior laryngeal cleft is present (Fig. 33.4).

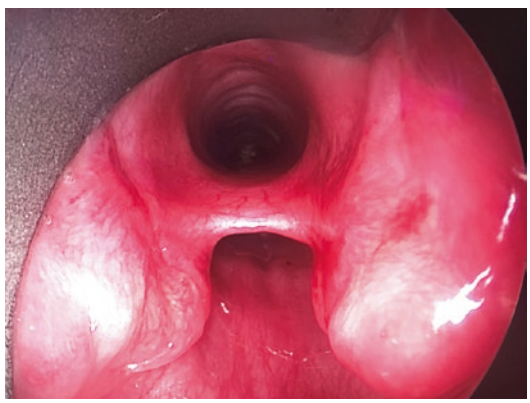

Fig. 33.4 Type III laryngotracheal cleft. View under 0 degrees rigid endoscopy

In children who are symptomatic and do not have other more severe anomalies, repair of the posterior laryngeal cleft should be carried out as soon as possible to prevent chronic microaspiration with long-term pulmonary sequelae. Prior to repair, consideration should be given to whether the infant requires tracheotomy placement, gastrostomy tube placement, and Nissen fundoplication. 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 unparalleled exposure of the cleft while protecting the recurrent laryngeal nerves. A two-layer closure is recommended, with the option of performing an interposition graft (perichondrium or periosteum) if warranted. Type IV clefts are often associated with multiple congenital anomalies. These long clefts are exceedingly difficult to repair and are prone to anastomotic breakdown. Success rates for cleft repair vary significantly (50–90%) depending on both the severity of the cleft and the presence of coexisting congenital anomalies and comorbidities.

33.3.5 Laryngeal Atresia

33.3.5.1 Congenital High Airway Obstruction Syndrome (CHAOS)

CHAOS is a life-threatening, prenatally diagnosed condition caused by complete or near-complete obstruction of the fetal airway. This obstruction may be due to laryngeal atresia (Fig. 33.5a) or tracheal agenesis. Atresia is sometimes an isolated anomaly but is often seen associated with a spectrum of other anomalies, including hydrocephalus malformation of the aqueduct of Sylvius, bronchotracheal fistula, esophageal atresia, tracheoesophageal fistula, syndactyly, and genitourinary, vertebral, and cardiac anomalies (Lim et al. 2003). Prenatal ultrasound findings indicative of CHAOS include bilaterally enlarged echogenic lungs, dilated airways, and flattened or everted diaphragms with associated fetal ascites and nonimmune hydrops

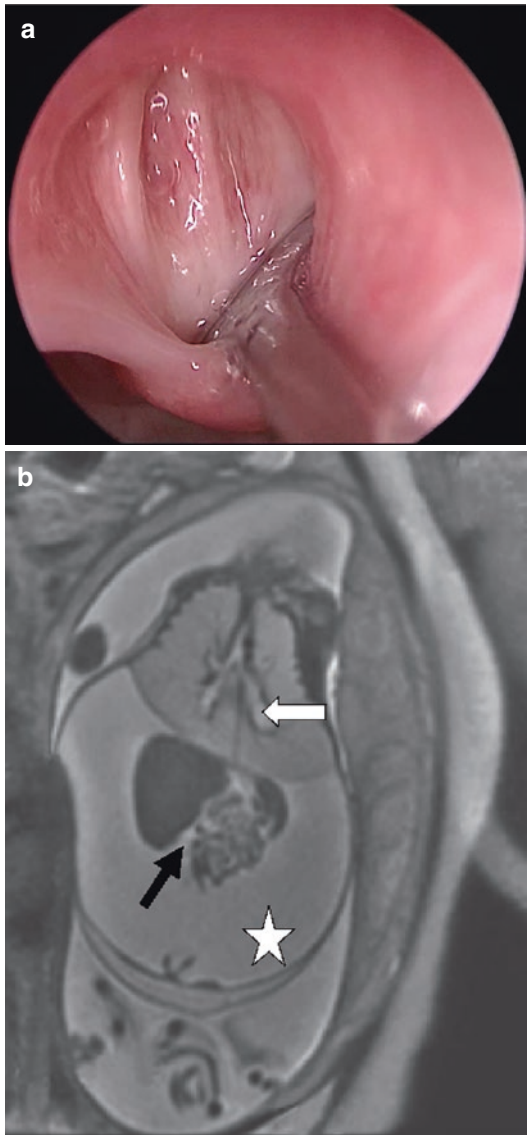

Fig. 33.5 (**a**) Laryngeal atresia. No lumen in the subglottic space. (**b**) Fetal ultrasonography demonstrating findings consistent with the diagnosis of congenital high airway obstruction: enlarged echogenic lungs, dilated airway (white arrow), flattened or everted diaphragms, and fetal ascites (white star) and hydrops. Fetal liver and intestines are marked with a black arrow (courtesy of Timothy Crombleholme MD, Cincinnati, Ohio)

(Fig. 33.5b). A fetus identified with these sonographic features is at significant risk of intrauterine death and faces a high mortality rate during the rest of pregnancy to delivery.

Diagnosis in the middle of the second trimester generally correlates with a poor outcome. A fetus presenting in the third trimester with CHAOS, in the absence of associated anomalies or hydrops, is likely to have incomplete obstruction. As such, this fetus is more likely to do well until delivery by the ex utero intrapartum technique (EXIT) procedure. This procedure maintains placental circulation to the fetus, while securing the airway at the time of delivery. Instrumentation of the airway, including tracheostomy, may be accomplished at this time.

For the newborn diagnosed with CHAOS, securing and maintaining the airway are of utmost importance. 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. Nevertheless, optimal timing for reconstruction has not yet been determined. Although an adequate airway can be constructed, adequate speech may not be feasible.

33.4 Anomalies of the Trachea and Bronchi

33.4.1 Tracheal Agenesis

Tracheal agenesis is a rare developmental abnormality that is almost always incompatible with life. Severe respiratory distress is present at birth, with the neonate attempting ventilation through bronchoesophageal communications. Although temporary ventilation may be possible with intubation of the esophagus, this generally cannot be sustained and results in neonatal demise. When a bronchoesophageal communication is not present, the fetus will have CHAOS (Lim et al. 2003; Ngerncham et al. 2015; Tazuke et al. 2015).

33.4.2 Tracheal Webs and Stenosis

A broad spectrum of rare tracheal anomalies is classified as tracheal stenosis. Affected segments differ in the degree and extent of stenosis, ranging from gossamer thin webs to more severe long segments of stenosis that may involve the entire airway.

33.4.2.1 Tracheal Webs

Tracheal webs are rare and involve an intraluminal soft tissue stenosis of the trachea. These webs may be membranous or composed of thick, inelastic tissue. Symptoms include biphasic stridor or expiratory wheezing, with severity dependent on the degree of tracheal narrowing. Thin webs can be readily managed by hydrostatic balloon dilatation alone. Children with a web greater than 1 cm in length, or in whom the airway cartilage is suspected to be deficient or structurally abnormal, are best managed by segmental tracheal resection (Backer et al. 1997; Ngerncham et al. 2015).

33.4.2.2 Cartilaginous Ring Aplasia

Cartilaginous ring aplasia is an exceedingly rare anomaly in which a short segment of the trachea lacks cartilage. This creates a region that is both malacic and stenotic. The remainder of the trachea is normal and children generally do not have other congenital anomalies. Segmental resection of the trachea is usually curative (Ngerncham et al. 2015; Puri and Höllwarth 2006).

33.4.2.3 Tracheal Cartilaginous Sleeve

Tracheal cartilaginous sleeve is an anomaly in which there are no discrete cartilaginous rings but rather a fused cartilaginous cylinder, with or without a membranous portion. This anomaly is often associated with craniosynostosis syndromes such as Apert, Pfeiffer, Crouzon, and Goldenhar. Patients present during the neonatal period with respiratory illness or in early infancy with acute respiratory symptoms, which may include biphasic stridor with respiratory distress, cough, and recurrent respiratory infections. Also, tracheal rigidity may cause difficulty in clearing secretions. Bronchoscopy shows a smooth anterior tracheal wall without the normal appearance of tracheal rings. The membranous posterior tracheal wall may be normal, reduced, or absent. CT and MR imaging may assist in delineating the extent of the lesion. Most of the patients require resection and repair.

33.4.2.4 Complete Tracheal Rings

Complete tracheal rings are an anomaly in which the trachea alone or both the trachea and bronchi are narrowed. In more than 50% of infants, a segmental stenosis is found. In such patients, the tracheal cartilage is abnormally shaped and forms complete rings (Figs. 33.6a, b). The clinical manifestations of complete tracheal rings vary from life-threatening respiratory distress at birth to subtle symptoms of airway compromise in older children. Many infants present with worsening 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. Over 80% of children with complete tracheal rings have other congenital anomalies, the most common of which are esophageal, cardiac, skeletal, and genitourinary. Fifty percent of children with complete tracheal rings also have a left pulmonary artery sling or vascular ring (Backer et al. 2001; Bando et al. 1996; Benjamin and Inglis 1989; Cotton 2003; DeMarcantonio et al. 2017; Fiore et al. 2005; Furman et al. 1999; Gorostidi et al. 2016; Gustafson et al. 2000; Hewitt et al. 2016; Hysinger 2018; Rutter 2006; Speggiorin et al. 2012).

In some patients, placement of an endotracheal tube may exacerbate respiratory distress by causing acute swelling and inflammation. Partially

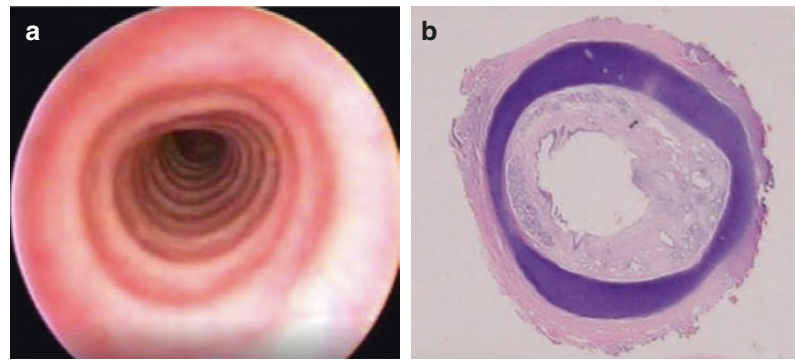

Fig. 33.6 (**a**, **b**) Congenital tracheal stenosis. (**a**) Endoscopic view demonstrating complete tracheal rings. (**b**) Histology of tracheal segment demonstrating virtually complete tracheal cartilaginous ring

obstructing tracheal lesions may also 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 decreased by one-third to one-half of its normal diameter with as little as 1 mm of edema. This accounts for the rapid progression of symptoms in some children who have acute inflammatory conditions superimposed on existent tracheal narrowing (Backer et al. 2001; Bando et al. 1996; Benjamin and Inglis 1989; Cotton 2003; DeMarcantonio et al. 2017; Fiore et al. 2005; Furman et al. 1999; Gorostidi et al. 2016; Gustafson et al. 2000; Hewitt et al. 2016; Hysinger 2018; Rutter 2006; Speggiorin et al. 2012).

33.4.2.5 Diagnosis

Expeditious diagnostic evaluation to define aberrant and normal tracheobronchial anatomy is required. Although an initial high-kilovolt airway film may show tracheal narrowing, the precise location and extent of the narrowing is best achieved by endoscopic techniques. CT scans provide a rapid and precise method of measuring the extent and length of airway narrowing or displacement. Visualization of the anatomic relationship between the airways and surrounding structures can be enhanced with intravascular contrast. Newer computer software allows for three-dimensional image reconstruction and is helpful in planning surgical procedures (Fig. 33.7). MR imaging is useful in assessing the relationship of the mediastinal great vessels to the airway. Echocardiography is primarily used to assess the presence of intracardiac defects and can detect the majority of associated mediastinal vascular anomalies.

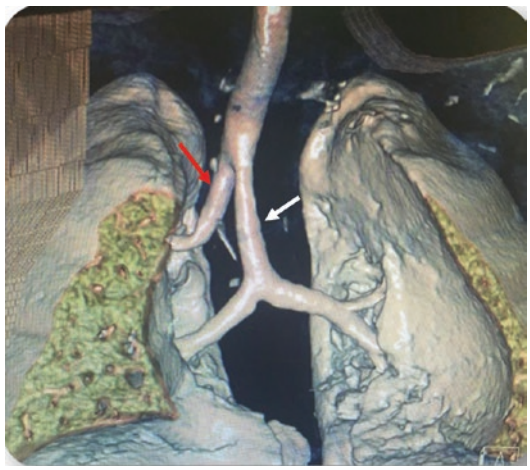

Fig. 33.7 CT scan with three-dimensional reconstruction to demonstrate anatomy of the trachea in a patient with congenital tracheal stenosis involving the distal trachea. This patient had an aberrant tracheal right bronchus

Most children with complete tracheal rings require tracheal reconstruction. If a pulmonary artery sling or vascular ring is present, repair of such an anomaly should be undertaken concurrent with the tracheal repair. Segmental tracheal resection with end-to-end anastomosis is considered the treatment of choice for short-segment tracheal stenosis. Slide tracheoplasty is currently the procedure of choice for long segments of tracheal involvement, having replaced patch tracheoplasty (Fig. 33.8). This approach yields significantly less morbidity than other tracheal reconstruction techniques and is applicable to virtually all anatomic variants 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 upper half of the trachea and the posterior wall of the lower trachea are incised. These segments are then slid over each other and anastomosed with 5–0 monofilament and absorbable sutures. Following surgery, the airway has four times the cross-sectional area and one-half the length of its previous dimension. Airflow is increased 16-fold with this method of airway reconstruction.

Postoperatively, endotracheal intubation is required for 1 day to several weeks, though most patients are extubated within 48 h. To minimize the risk of damage to the newly reconstructed airway, unnecessary movements of the endotracheal tube or unplanned extubation must be avoided. Nasotracheal intubation is used preferentially because the endotracheal tube can be stabilized in position more securely. Patients require continuous monitoring, careful pulmonary toilet, and endoscopic removal of any obstructing granulation tissue. Just prior to and to ensure a safe extubation, the integrity and patency of the reconstructed airway are assessed by flexible fiber-optic endoscopy through the endotracheal tube. Airway configuration following slide tracheoplasty may resemble figure of 8 trachea (Fig. 33.9) but is not associated with an obstruc-

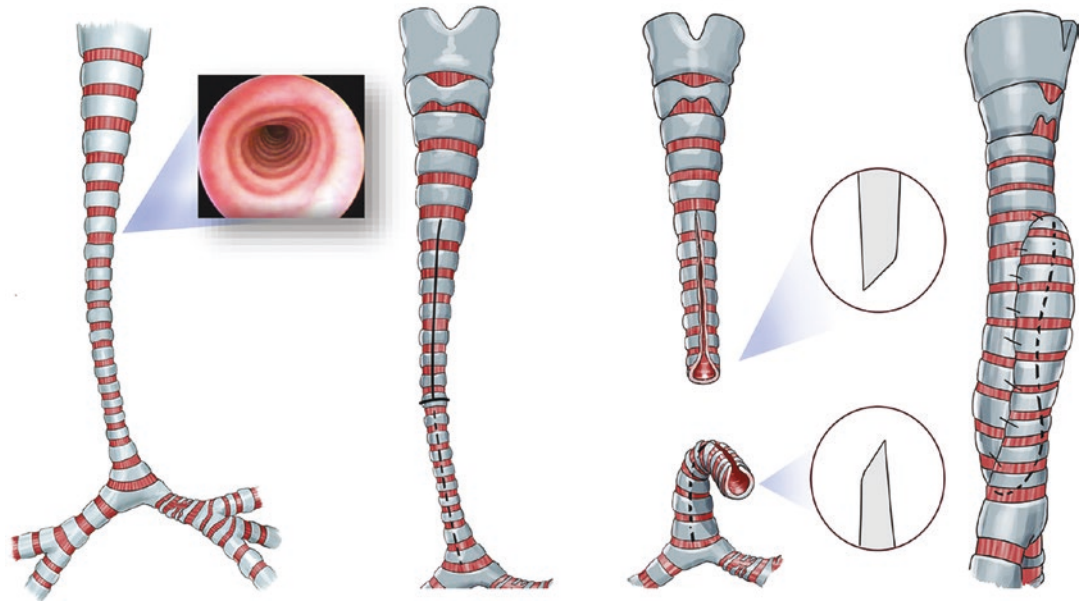

Fig. 33.8 Slide tracheoplasty procedure: the trachea is transversely divided at the midpoint of the tracheal stenosis. After proximal and distal tracheal mobilization, the anterior portion of the cephalic trachea segment and the posterior portion of the caudal tracheal segment are incised. The two tracheal segments are then overlapped and obliquely sutured together (*courtesy Gaston Bellia Md, Buenos Aires, Argentina)

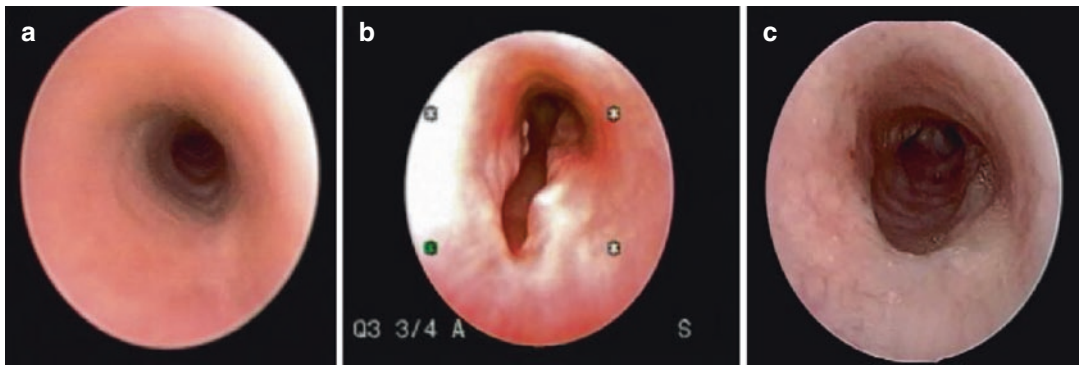

Fig. 33.9 Figure of 8 trachea following repair of complete tracheal rings: (**a**) preoperative endoscopy documents complete tracheal rings; (**b**) postoperative endoscopy at 6 weeks demonstrates the figure of 8 trachea, and the patient had no airway symptoms; (**c**) endoscopy at 2 years demonstrates significant remodeling of airway

tive airway. In most cases, the trachea remodels to a normal oval shape within 1 year of reconstruction. Long-term survival following this procedure is currently 80–90% in main airway centers. Mortality is usually associated with severe comorbidities such as cardiac disease rather than airway complications.

33.4.3 Tracheal Diverticulum and Tracheal Bronchus

Tracheal diverticulum and tracheal bronchus are relatively common abnormalities of tracheal budding that occur during the third and fourth gestational weeks, when the trachea bifurcates and

differentiates. Tracheal diverticula resemble a bronchus, though they originate from the trachea and end blindly or communicate with a rudimentary lung. Tracheal bronchi most often affect the right upper lobe bronchus and may connect to an isolated intrathoracic lung segment or the apical segment of an upper lobe. Both anomalies are often seen in children with other congenital defects. Diagnosis is established by airway endoscopy (Ngerncham et al. 2015). Most children are asymptomatic and do not require treatment. Those who are symptomatic experience symptoms such as pneumonia and respiratory distress during the neonatal period. Additionally, they have an associated stenosis of the bronchus or other lung anomalies. Resection of involved lobe and bronchus in these patients is generally curative.

33.4.4 Tracheomalacia and Bronchomalacia

33.4.4.1 Pathogenesis

Tracheomalacia and bronchomalacia are conditions in which the structural integrity of the trachea or bronchi is diminished and the cartilaginous rings of the airway lack the necessary rigidity to prevent airway collapse during expiration. Malacia may occur in localized segments or diffusely throughout the airway. Tracheomalacia is the most common congenital tracheal anomaly.

It may be idiopathic or associated with a number of conditions, including esophageal atresia or tracheoesophageal fistula, aberrant innominate artery, mediastinal masses, prolonged intubation for interstitial lung disease, or bronchopulmonary dysplasia. Premature neonates with bronchopulmonary dysplasia or children with chronic indwelling cuffed endotracheal or tracheostomy tubes are at particular risk for developing combined severe tracheal and bronchial malacia. Several types of tracheal collapse have been described (Fig. 33.10a–d).

Presenting symptoms vary and depend on the severity, duration, and location of the malacia. Most children are either asymptomatic or minimally symptomatic and most cases involve posterior malacia of the trachealis, with associated broad tracheal rings. Other associated abnormalities include laryngeal clefts and tracheoesophageal fistulae. 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 rigid or flexible bronchoscopy, with the patient breathing spontaneously; this demonstrates dynamic distortion and compression of the trachea. Children who are minimally symptomatic are watched expectantly. Their symptoms often resolve by age 3. Children who experience a worsening of symptoms require more intensive medical or surgical intervention. Respiratory monitoring with nasal CPAP may be beneficial in some patients.

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. More diffuse malacia may require tracheotomy placement with positive pressure

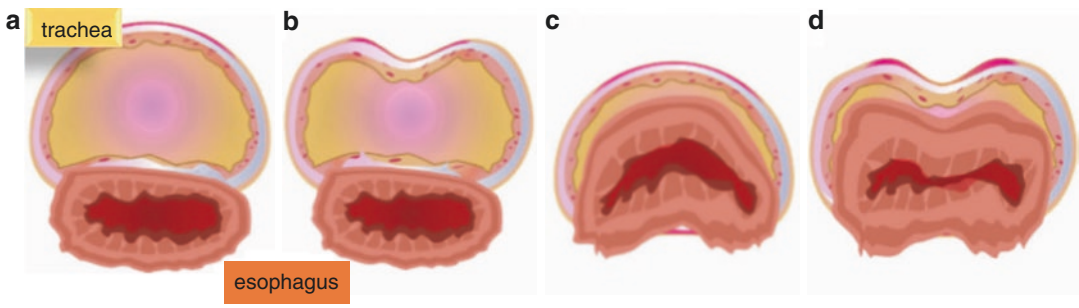

Fig. 33.10 (**a**–**d**) Types of tracheal collapse (**a**) normal anatomy, (**b**) anterior collapse, (**c**) posterior intrusion, (**d**) combined (*courtesy Diana Romero MD, Bogotá, Colombia)

ventilation over a long period of time. The placement of intratracheal stents (Fig. 33.11) is used selectively in patients with severely problematic tracheomalacia or bronchomalacia that is unresponsive to nonoperative therapy or not suitable for surgical treatment (Gerber and Holinger 2003; Grillo 1994; Hysinger and Panitch 2016; Monnier 2011; Serio et al. 2014; Shieh et al. 2017; Sztano et al. 2016; Torre et al. 2012; Valerie et al. 2005; Varela et al. 2018; Vinograd et al. 1987).

Major complications associated with this approach can occur, including stent collapse, dislodgement, or rarely, stent erosion into the great vessels. Additionally, stent removal can cause tracheal tearing or major hemorrhage.

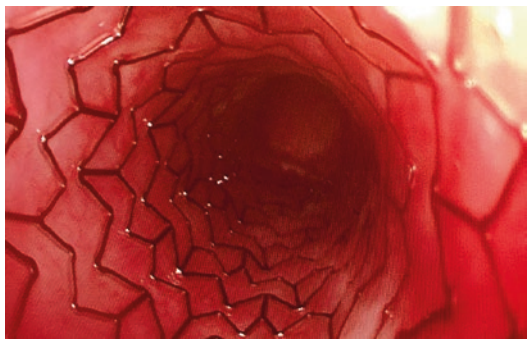

Fig. 33.11 Balloon-expandable metallic stent placed in the trachea with a severe tracheomalacia associated with a type III esophageal atresia

A posterior tracheopexy is proposed for severe tracheomalacia. During the procedure, the esophagus is mobilized to the right, and the posterior wall of the trachea is attached to the spine with multiples stitches under endoscopic visualization (Fig. 33.12a–c).

Regarding severe bronchomalacia, a three-dimensional external airway splint (Fig. 33.13a, b) has been proposed to reinforce the bronchial wall, avoiding collapse due to the aorta or pulmonary artery.

33.4.5 Esophageal Lung

Isolated bronchial connection between the esophagus and the airway is extremely rare and occurs more frequently in females (2:1) (Fig. 33.14). Associated cardiac, genitourinary, vertebral, and diaphragmatic anomalies are common. This malformation is thought to develop from a supernumerary lung bud arising from the esophagus. Most commonly, a lower lobe is aerated by this ectopic bronchus, but an entire main bronchus and lung may be involved. As in pulmonary sequestration anomalies, the pulmonary vasculature in this anomaly may be abnormal, with the arterial supply coming off the aorta and venous drainage going into either the systemic or pulmonary veins.

Because of inadequate bronchial drainage, children usually have recurrent pulmonary infec-

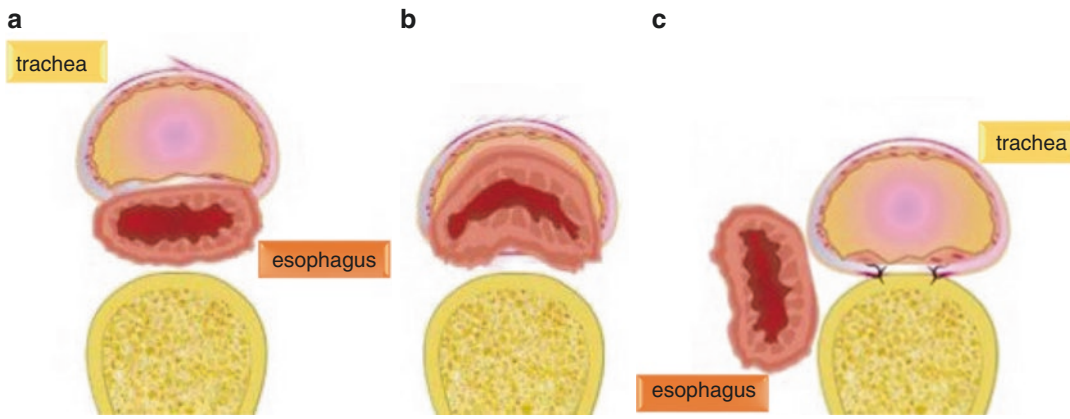

Fig. 33.12 (**a**–**c**) Posterior tracheopexy. Esophagus is mobilized to the right and the posterior tracheal wall is stitched to the spine. (**a**) Normal anatomy, (**b**) severe posterior tracheal collapse, (**c**) posterior tracheopexy (courtesy Diana Romero MD, Colombia)

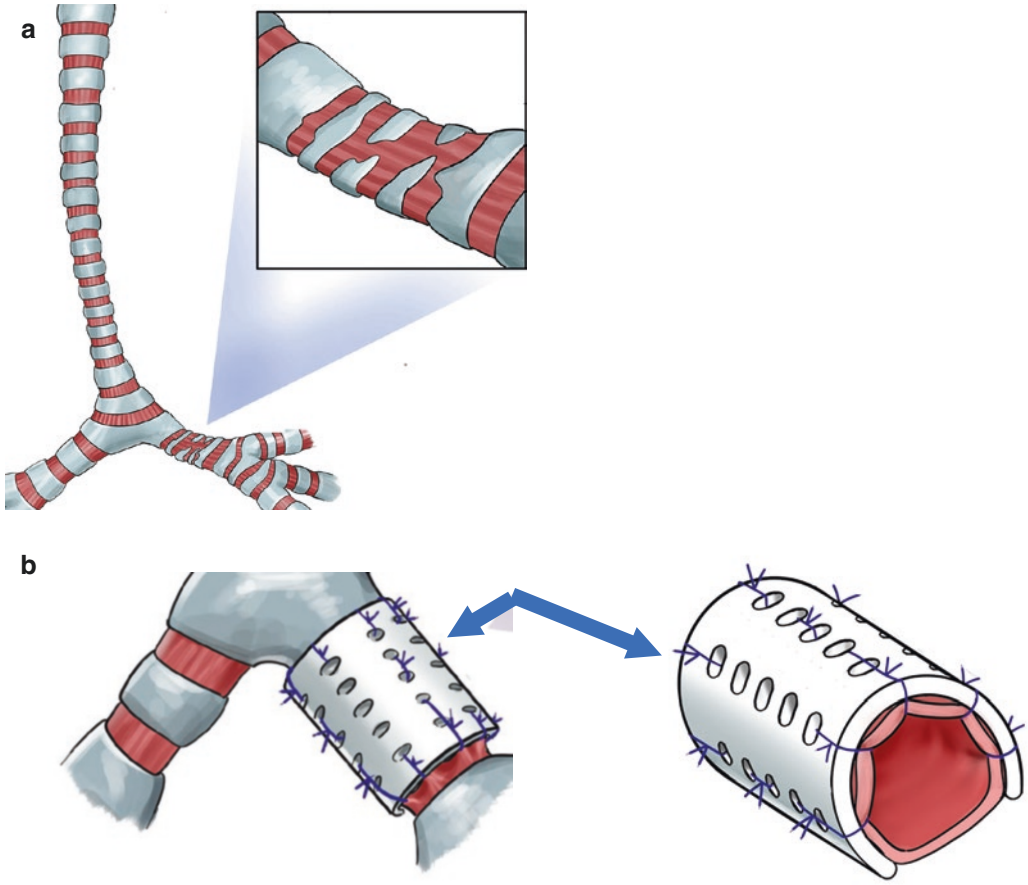

Fig. 33.13 (**a**) Severe left bronchial collapse (courtesy Gaston Bellia MD, Buenos Aires, Argentina). (**b**) 3D airway splint is placed in a severe left bronchial collapse (Courtesy Gaston Bellia MD, Buenos Aires, Argentina)

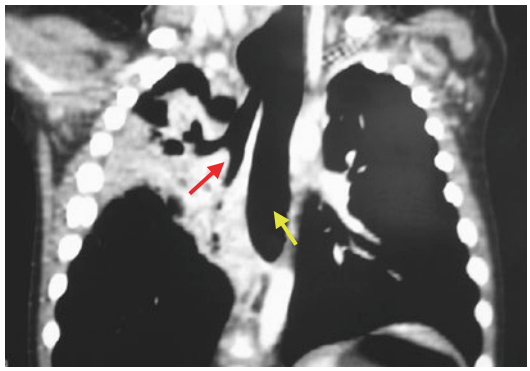

Fig. 33.14 Esophageal lung. A pulmonary lobe (red arrow) is connected to the esophagus (yellow arrow)

tion; however, occasionally esophageal bronchus is not discovered until adolescence or adulthood. Although radiographic findings differ depending on the affected segment of the lung, collapse, consolidation, cavitation, and cyst formation within the pulmonary parenchyma are commonly seen. The diagnosis is confirmed by contrast studies of the esophagus, though occasionally, false-negative results occur. Excision of the abnormal lung and closure of the bronchoesophageal fistula are the treatments of choice. Prognosis depends on early diagnosis and treatment and the severity of associated anomalies.

33.4.6 Tracheobronchial Biliary Fistula

Congenital tracheobronchial biliary fistulae are extremely rare and may arise from the distal trachea or either mainstem bronchus. All children

with this anomaly have significant respiratory problems but the cardinal symptom is bile-stained sputum. The diagnosis is established either by bronchoscopy or endoscopic retrograde cholangiopancreatography (ERCP). Surgical division of the fistulous tract is the only effective therapy for this malformation.

33.4.7 Subglottic Hemangioma

33.4.7.1 Pathogenesis

Hemangiomas of infancy are benign congenital vascular tumors. These tumors are characterized by vascular endothelium that undergoes a phase of growth followed by slow, spontaneous involution that occurs over several years and is generally complete by the first decade of life.

These tumors most commonly present cutaneously but can occur in any anatomic site. No symptoms occur during the first weeks of life. They usually become evident between 2 and 4 months of age. Progressive resolution occurs between 12 and 18 months with a threefold female preponderance.

Almost all lesions that occur within the tracheobronchial tree are in the subglottis. The natural history of subglottic hemangiomas (SGH) generally mirrors that of cutaneous lesions, and more than 50% of patients with a subglottic hemangioma also have cutaneous hemangiomas. As the hemangioma undergoes proliferation, progressive deterioration 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 obstruction is severe, apnea, cyanosis, and "dying spells" may occur. 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. 33.15). Because of the risk of hemorrhage, biopsy is not advised.

Most patients require treatment and many treatment modalities are often combined. These include medical treatments, endoscopic resection, and open surgical procedures. Depending on both the severity of the obstruction and the expertise of involved clinicians, early symptoms are managed with intralesional or systemic steroids. The use of systemic steroids is not recommended for longer than 3 weeks if the symptoms don not improve. The use of propranolol seems to be an effective medical treatment that allows

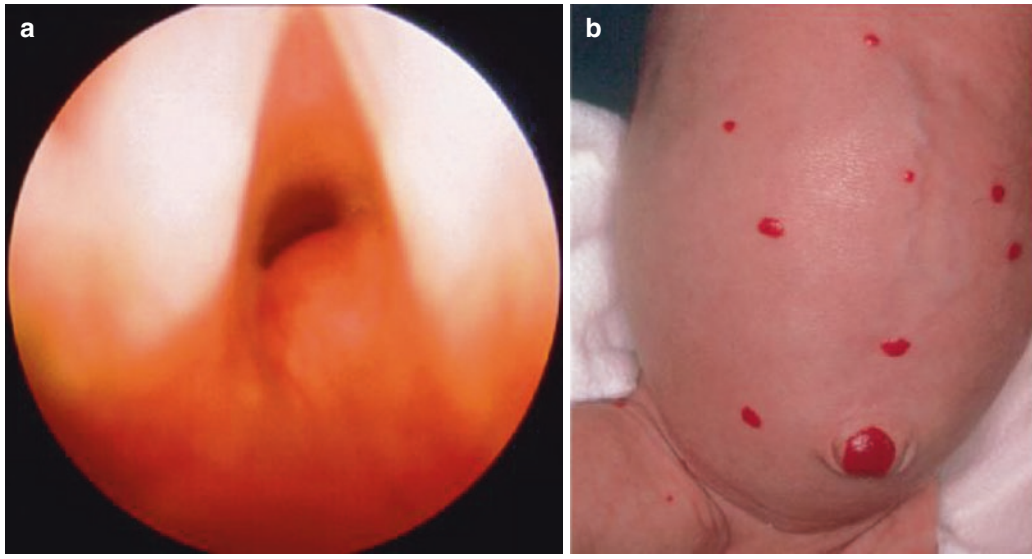

Fig. 33.15 (**a**) Endoscopic view of a subglottic hemangioma in a patient with multiple cutaneous infantile hemangiomas (**b**)

complete and fast resolution with evident improvement of respiratory symptoms. Propanol is our first treatment approach in most of the SGH. Some surgeons advocate endoscopic treatment with laser fulguration or the use of microdebrider. Surgical options consider translaryngeal resection, whereas others place a tracheotomy below the lesion, with the expectation of removal following involution of the hemangioma.

33.4.8 Bronchogenic Cyst

Bronchogenic cysts result from abnormal budding of the bronchial tree in which a portion of the lung bud develops independently. The cyst walls frequently contain cartilage and are lined with ciliated columnar epithelium. These lesions tend to enlarge, thus causing airway obstruction. Infants with bronchogenic cysts most commonly present with respiratory distress. They also may have cough, chest pain, or wheezing. Although a plain chest X-ray may suggest the presence of a bronchogenic cyst, a CT scan and barium esophagram are useful in confirming this diagnosis. Thoracoscopic resection is curative (Fig. 33.16a, b).

33.4.9 Bronchial Atresia

Localized bronchial atresia is a rare anomaly in which the atretic bronchus obstructs the flow of secretions and air from the distal lung to the main tracheobronchial tree. This condition may simulate lobar emphysema or a mediastinal mass. At birth, the obstructed lung retains fluid, but eventually the affected lobe or segment becomes hyperaerated as air enters through the pores of Kohn. Patients accumulate secretions proximal to the atresia and a mucocele forms. Emphysema of the segment may cause compression of the normal lung tissue and may be associated with wheezing and stridor. Plain chest radiographs often demonstrate a hilar mass with radiating solid channels surrounded by hyperaerated lung. A CT chest scan may indicate a cystic central mucocele and can help differentiate bronchial atresia from a bronchogenic cyst or lobar emphysema. Although children with bronchial atresia may be asymptomatic for long intervals, they are at risk of serious pulmonary infection when entrapped secretions become infected. When this anomaly is identified, resection is both indicated and curative (Azizkhan 2005; Puri and Höllwarth 2006).

33.4.10 Bronchial Agenesis

Congenital absence of a bronchus occurs more commonly than tracheal agenesis, and in contrast to tracheal agenesis, this anomaly is compatible with life. Several possible anatomic variants have been described, including lobar, bronchial, and parenchymal agenesis. Specifically, there may be complete agenesis of the lung and its bronchus and blood supply; aplasia, in which there is a

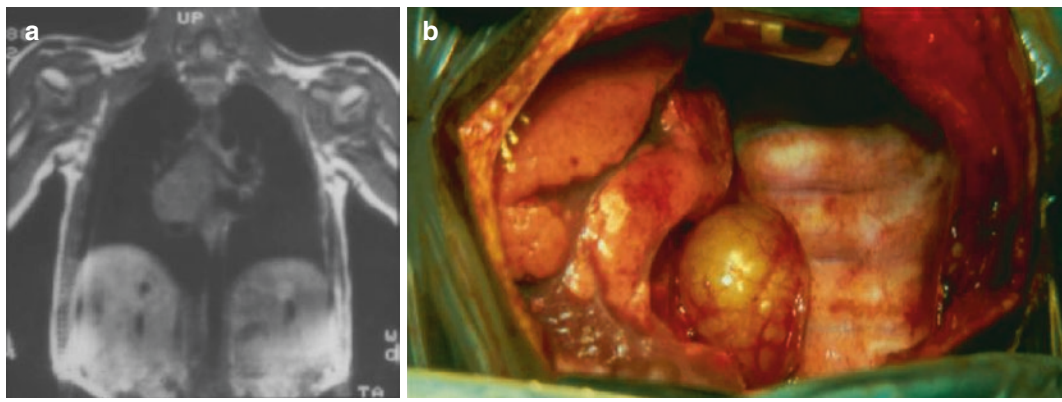

Fig. 33.16 Bronchogenic cyst: (**a**) right hilar lesion seen on CT scan, (**b**) operative photograph demonstrating the bronchogenic cyst in situ

rudimentary bronchus and absent lung; or hypoplasia, in which there is a rudimentary bronchus and hypoplastic lung. As is common in children with airway malformations, children with bronchial agenesis also may have other congenital anomalies of the skeletal, cardiovascular, gastrointestinal, and genitourinary systems. Diagnosis is confirmed by chest radiographs and airway endoscopy. The majority of patients do not require surgical intervention. Nevertheless, these patients are important to identify as bronchial or lobar atresia may mimic other airway anomalies such as bronchial stenosis, extraluminal airway obstruction by tumors or masses, or complete intraluminal obstruction in which there is no aeration distal to the obstruction.

33.4.11 Bronchial Stenosis

Congenital bronchial stenosis is extremely rare, with reported cases caused by compressive vascular, cardiac, and congenital cystic lesions or soft tissue cartilaginous stenoses. Symptoms and treatment depend on the anatomic location of the lesion and its severity. In contrast, acquired bronchial stenosis is more common and is a major cause of morbidity and mortality in infants who require prolonged intubation and respiratory support. Most such cases can be managed endoscopically.

33.5 Conclusions

Congenital airway malformations include a wide number of anomalies with a broad spectrum of symptoms. Treatments of all these conditions are usually performed by an airway team, working in collaboration with critical and intensive care, neonatologists, radiologists, and anesthetists.

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