A variety of congenital anomalies may present within the laryngotracheal airway, with symptoms ranging from mild stridor to severe respiratory distress requiring emergent airway management immediately after birth. Some of these lesions may be relatively asymptomatic in the neonatal period and present later in life, whereas the most severe cases may not be incompatible with life.²

The most common causes of stridor in infants are laryngeal in origin. There are numerous tracheal anomalies, however, that may present with stridor. In addition to stridor, symptomatology often may include aspiration or other feeding disorders. Hoarseness or aphonia also may be involved with congenital laryngeal anomalies, and often accompanies symptoms of airway obstruction. Any child with this symptomatology complex presenting soon after birth should be evaluated thoroughly for the possible existence of a congenital laryngotracheal anomaly. Some of the more common congenital laryngeal and tracheal anomalies are discussed with respect to their diagnostic evaluation, clinical presentation, and management.

DIAGNOSTIC EVALUATION

Although some congenital laryngotracheal anomalies may present later in life, the majority present with symptomatology in the neonatal
period or during infancy. Laryngeal lesions typically present with stridor, hoarseness, aphonia, and possibly feeding disorders. The stridor is usually inspiratory or possibly biphasic in nature, and should be differentiated from stertor, which primarily is caused by airway obstruction in the nasal or pharyngeal regions. Choanal atresia may present with severe airway obstruction and a breathing pattern that may be confused with stridor. To rule out choanal atresia, suction catheters can be passed through the nasal cavity to demonstrate patency. Flexible fiberoptic nasopharyngoscopy can be performed to directly examine the posterior choanae to rule out atresia or stenosis. Patients with tracheal anomalies typically present with expiratory stridor.

A child with stridor and a suspected laryngeal lesion should undergo a complete history and physical examination. Premature infants who have been intubated for variable periods of time may develop acquired obstructive lesions such as subglottic stenosis, subglottic cysts, or intubation granulomas. Flexible fiberoptic laryngoscopy should be performed in all cases to carefully examine the hypopharynx and larynx to rule out glottic and supraglottic problems such as laryngomalacia, saccular and vallecula cysts, vocal cord paralysis, or other glottic or supraglottic problems. Anterior-posterior and lateral radiographs of the cervical airway should be obtained to help rule out any stenotic lesions or other airway problems. A barium swallow can be used in cases of swallowing difficulties to delineate conditions, such as posterior laryngeal clefts, tracheoesophageal fistulae, or vascular rings, which may compress the middle to lower trachea. Airway fluoroscopy also can be used at the time of barium swallow to assess the dynamic condition of the trachea, such as tracheomalacia or innominate artery compression of the trachea. If a conclusive diagnosis has not been reached after examination and tests have been performed, direct laryngoscopy and bronchoscopy using rigid rod-lens optical telescopes can be performed in the operating room under general anesthesia. Definitive treatment may be performed at this time if the lesion is amenable to endoscopic therapy using the CO₂ laser or other endoscopic techniques.

**LARYNGOMALACIA**

Laryngomalacia is the most common congenital anomaly of the larynx and is the most frequent cause of stridor in infants. It accounts for 60% of laryngeal problems in infants. Laryngomalacia also is known as congenital laryngeal stridor. Although this condition is self-limiting and usually resolves within 12 to 18 months, the symptomatology may become severe enough to warrant surgical intervention. Stridor associated with laryngomalacia typically develops within the first few weeks of life and progresses in severity over a period of a few months. Stridor typically is described as high-pitched and fluttering in nature. Severe cases of stridor may be associated with sternal retraction, respiratory distress, and feeding difficulties. The condition usually is exacerbated by exertion, crying, feed-
CONGENITAL ANOMALIES OF THE LARYNX AND TRACHEA

ing, agitation, or supine positioning. Placing the infant in the prone position or on his or her side and extending the neck may relieve the stridor. Approximately 80% of cases are associated with gastroesophageal reflux (GER). 28 This condition also may be associated with failure to thrive. Pectus excavatum is a common associated finding that tends to improve when the airway obstruction resolves. Resolution usually occurs by 18 months of age; some patients, however, may remain symptomatic until age 5. Other synchronous laryngotracheal anomalies associated with laryngomalacia have been reported, including subglottic stenosis and vocal cord paralysis. 47

The cause of laryngomalacia is unclear, although altered embryologic development of the larynx is the most likely cause of the abnormal anatomic findings. Histologic studies have failed to demonstrate any inherent cartilaginous abnormality leading to the increased flaccidity and abnormal collapse of the supraglottic larynx. 40, 59 Abnormal neurologic function of the larynx also has been suggested as a factor.

Although laryngomalacia typically is described as a congenital anomaly, there have been cases where symptoms associated with laryngomalacia do not develop until several months after birth. 34 In addition, children with severe neurologic impairment and pharyngeal and hypopharyngeal hypotonia may develop a chronic acquired flaccidity of the supraglottic larynx with similar symptomatology. 40

The gold standard in diagnosis for laryngomalacia is flexible laryngoscopy. With the patient awake, the larynx is examined under dynamic conditions. Obviously, history and physical examination is also important to reach a correct diagnosis. Although the diagnosis of laryngomalacia can be made using radiographic studies, this modality is best suited as an adjunct to evaluation for associated laryngotracheal anomalies. In severe cases for which operative intervention is considered, rigid endoscopy in the operating room is warranted.

Typically, with flexible laryngoscopy, the larynx demonstrates an omega-shaped epiglottis (that may occur in 30% to 50% of normal asymptomatic infants), 51 redundant aryepiglottic (AE) folds, and excessive tissue in the supra-arytenoid area that may prolapse into the laryngeal inlet upon inspiration (Fig. 1A). A deep interarytenoid cleft, or possibly a posterior laryngeal cleft, may be seen on flexible endoscopy, but is best assessed using rigid endoscopy. Vocal cord paralysis also may be ruled out simultaneously by this technique. Laryngomalacia also may be site specific. There may be evidence of primarily posterior laryngomalacia with prolapse of the supra-arytenoid tissue noted, or possibly prolapse of the anterior aspect of the AE folds and epiglottis into the airway (anterior laryngomalacia).

In the majority of patients, laryngomalacia resolves and no surgical intervention is required. Encouraging the family to have the child sleep on his or her side and not in the supine position helps relieve symptomatology and is safer for the child. Surgical intervention should be considered in patients with severe respiratory distress, failure to thrive, severe obstructive apnea, or other severe symptomatology. In the past, trache-
Figure 1. A, Laryngomalacia with omega-shaped epiglottis and excessively taught aryepiglottic folds. B, Appearance of supraglottic larynx after bilateral supraglottoplasty with conservative excision of tissue of the aryepiglottic fold, lateral epiglottis, and supra-arytenoid tissue.

Otomotomy had been advocated to relieve airway obstruction. More recently, however, epiglottoplasty or supraglottoplasty has been considered the surgical procedure of choice to treat laryngomalacia. This procedure is performed using suspension laryngoscopy. Excessive supraglottic tissue causing airway obstruction (AE folds, lateral epiglottis, and supra-arytenoid tissue) is resected conservatively using the carbon dioxide laser or sharp dissection instrumentation (see Fig. 1B). Typically, a bilateral approach to surgical resection is performed. Gray et al, however, have advocated unilateral resection in cases of laryngomalacia.\textsuperscript{36} Conservative resection minimizes the likelihood of complications, such as supraglottic stenosis or aspiration. A secondary procedure may be performed if symptoms are not adequately relieved.

Recently a new condition has been described known as discoordinate pharyngolaryngomalacia.\textsuperscript{23} This is a condition that has been noted in patients originally diagnosed with laryngomalacia who did not respond well to surgical intervention. This condition is associated with severe laryngomalacia manifested by complete supraglottic collapse during inspiration without shortened AE folds or redundant mucosa and with accompanying pharyngomalacia. Many of these patients require further surgical intervention, including tracheotomy; some may respond however, to bi-level positive airway pressure (BiPAP) management to avoid tracheotomy.

**BIFID EPIGLOTTIS**

Congenital bifid epiglottis is a rare congenital laryngeal anomaly that may present as laryngomalacia with inspiratory stridor and airway ob-
struction. This condition was described first in 1874. Since that time, numerous other reports have been published. Bifid epiglottis may be associated with congenital syndromes, most notably Pallister-Hall syndrome, and the familial inheritance pattern of bifid epiglottis with polydactyly described by McClay et al. Patients with bifid epiglottis should undergo an endocrine evaluation because of the possibility of associated hypothyroidism or hypothalamic abnormalities. In cases of severe airway obstruction, surgical management is necessary. Endoscopic removal of the malacic portion of the bifid epiglottis should be excised using sharp instrumentation or CO₂ laser to relieve the airway obstruction.

**SACCULAR CYSTS**

Congenital laryngeal saccular cysts are unusual laryngeal anomalies that are similar in their embryologic development to laryngocele (Fig. 2). These lesions arise from the vestigial laryngeal structure known as the saccule. Whereas the laryngocele is an abnormal dilatation of the larynx that is filled with air, a saccular cyst is filled with thick, mucoid fluid. Saccular cysts typically do not communicate with the internal laryngeal lumen. It is likely that the cause of this problem is a congenital obstruction of the laryngeal opening of the saccule. DeSanto classified laryngeal cysts into superior cysts that extend medially and posteriorly, extending into the laryngeal lumen in the region of the ventricle, and posterior cysts, which extend into the region of the false vocal cord and aryepiglottic fold. In cases of severe airway obstruction at birth, immediate intervention is warranted requiring endotracheal intubation or emergency tracheotomy. Surgical management of saccular cysts can be performed either endoscopically using CO₂ laser, with sharp dissection techniques to marsupialize

*Figure 2. Right congenital laryngeal saccular cyst is demonstrated with significant obstruction of the supraglottic airway.*
the cyst, or with open surgical resection. In many cases, endoscopic management is successful. The high rate of recurrence after endoscopic drainage, however, is well-documented in numerous reports.\textsuperscript{10} Ward et al described an open surgical approach for surgical excision of this lesion with excellent postoperative results.\textsuperscript{54} Surgical resection is warranted in cases of recurrent saccular cysts or may be necessary as an initial surgical procedure in cases with severe airway obstruction.

ANOMALIES OF THE SUPRAGLOTTIS

Supraglottic Webs

Other unusual anomalies may occur in the supraglottis. Supraglottic webs are very rare and account for less than 2\% of all congenital laryngeal webs.\textsuperscript{12} Webs usually arise anteriorly and may have minimal symptoms if the airway is not obstructed significantly. Thick webs obstructing a large region of the airway, however, may present with respiratory symptomatology. Hoarseness also may be a presenting symptom. The treatment of choice is endoscopic division of the web, either using sharp instrumentation or CO\textsubscript{2} laser.

Anomalous Cuneiform Cartilage

Anomalous cuneiform cartilage may cause severe airway obstruction in infants and require immediate surgical intervention. Infants may present with symptoms similar to laryngomalacia and may have other associated anomalies, such as ankyloglossia and microglossia. Templer et al reported a case of anomalous cuneiform cartilage with severe airway obstruction requiring tracheotomy and subsequent supraglottic laryngectomy.\textsuperscript{52}

ANOMALIES OF THE GLOTTIS

Vocal Cord Paralysis

Vocal cord paralysis accounts for 10\% of all congenital laryngeal anomalies and is the second most common cause next to laryngomalacia.\textsuperscript{18} Birth trauma may induce vocal cord paralysis, possibly by stretching of the recurrent laryngeal nerve associated with difficult deliveries. Central nervous system abnormalities may present with bilateral vocal cord paralysis. Peripheral causes, such as cardiovascular and mediastinal problems, may present as unilateral vocal cord paralysis (the left side being more common). Bilateral vocal cord paralysis typically presents with high-pitched inspiratory stridor with normal cry or possibly a mildly hoarse cry. Unilateral paralysis typically presents with a weak, breathy cry and
feeding difficulties. This may manifest as aspiration, resulting from the inability of the vocal cords to approximate to protect the airway. Airway obstruction from bilateral vocal cord paralysis presents immediately after birth and may be severe, requiring emergent airway intervention such as intubation and possibly tracheotomy.

Diagnostic work-up should include laryngoscopy, performed either with a flexible endoscope while the patient is awake, or with rigid endoscopy under light anesthesia. To assess vocal cord mobility, it is best to perform the examination while the patient is undergoing respirations and crying, if possible. Bronchoscopy and esophagoscopy also should be performed as part of the evaluation. MR imaging of the head should be obtained to rule out any central nervous system abnormalities. In cases of severe airway obstruction, tracheotomy should be performed in patients with bilateral vocal cord paralysis. Tracheotomy also may be required in patients with unilateral vocal cord paralysis if there is significant aspiration. Multiple cranial nerve deficits are common in neonates with congenital vocal cord paralysis, resulting in a high incidence of dysphagia and chronic aspiration. With Arnold-Chiari malformation or other central nervous system problems, tracheotomy can be obviated if an intracranial shunt procedure has been performed. Tracheotomy should be performed, however, if the paralysis and airway obstruction persists. Recurrence of vocal cord paralysis may herald an intracranial shunt malfunction. With bilateral vocal cord paralysis where airway obstruction is not severe, tracheotomy may be deferred and the patient followed closely as an outpatient with an apnea monitor if the family has good access to a health care facility. Bilateral vocal cord paralysis may resolve within 6 months to 1 year. In cases where the paralysis does not resolve with time, a vocal cord medullization technique, such as arytenoidectomy performed endoscopically or through an external approach, may be performed.

Laryngeal Web

Laryngeal webs form when there is failure of recanalization of the larynx during embryologic development; this may cause respiratory distress at birth (Fig. 3). Webs occur primarily at the level of the glottis. There rarely have been reported supraglottic webs. These may be very thin and limited to the level of the anterior glottis. Most are thick and fibrous with significant subglottic extension, however. Thin webs limited to the glottis may present with minimal airway obstruction, demonstrating exclusively with hoarseness. More extensive webs involving the glottis, however, may be associated with aphonia, accompanied by significant airway obstruction. Laryngeal webs always should be considered in children with a congenital history of hoarseness and recurrent croup presenting before 6 months of age.

Surgical management includes endoscopic management versus open laryngotracheal reconstruction, depending on the extent and thickness of the web and the degree of associated congenital cricoid malformation.
Figure 3. Congenital laryngeal web that was managed by endoscopic division.

Small limited webs may be divided endoscopically without significant sequela. Extensive webs with subglottic extension presenting with severe airway obstruction require tracheotomy at a later age with subsequent laryngotracheal reconstruction and possible costal cartilage graft and stents. Supraglottic webs may require placement of a laryngeal keel to allow the optimum healing of the anterior commissure after surgical repair, allowing the best possible voice results postoperatively.

OTHER GLOTTIC ANOMALIES

Laryngeal atresia is a rare laryngeal anomaly that is incompatible with life. These infants usually survive only if there is an associated tracheoesophageal fistula or if a tracheotomy is performed immediately after birth. Anterior midline clefts of the larynx have been reported, and these are often asymptomatic.¹² Cri du chat syndrome is a congenital chromosomal abnormality involving chromosome 5 (B Group) and associated with an unusual form of congenital laryngeal stridor, which is a characteristic high-pitch similar to the mew of a cat. Classically, upon inspiration, the vocal cords of these patients have a narrow diamond-shaped appearance.

CONGENITAL ANOMALIES OF THE SUBGLOTTIS

Subglottic Hemangioma

Subglottic hemangioma is a congenital vascular lesion with symptoms ranging from minimal airway obstruction to severe, life-threatening respiratory distress (Fig. 4). Hemangiomas are only present at birth 30%
of the time, with the majority of cases presenting within the first few weeks or months of life. An infant with congenital stridor or progressively worsening stridor within the first few months of life should be evaluated for the possibility of subglottic hemangioma.

The natural history of these congenital vascular tumors is similar to hemangiomas found elsewhere in the body. Typically, there is a rapid growth period that is initiated within the first few weeks or months of life and continues for 12 to 18 months. There is a phase where the lesion is stable, and then a subsequent period of involution, which may take months or years to complete. Most hemangiomas involute completely by 5 years of age, although some lesions may take much longer or may never involute. Subglottic hemangiomas have a 2:1 female to male preponderance. Symptomatology other than airway obstruction includes cough, cyanosis, hoarseness, dysphagia, and hemoptysis. Cutaneous hemangiomas occur in approximately 50% of children with subglottic hemangiomas. A complete examination of the child’s skin should be included in an evaluation for stridor.

The histology of hemangioma reveals hyperplasia of endothelial cells, mast cells, fibroblasts, and macrophages. A biopsy may be performed to make a definitive diagnosis.

The diagnosis of subglottic hemangioma, in most cases, can be made on clinical history, physical examination, and endoscopic appearance. Rigid endoscopy should be performed to make a definitive diagnosis. A biopsy of the lesion is not necessary if the endoscopic appearance and clinical history are consistent with the diagnosis. If there is any doubt about the diagnosis, however, biopsy should be performed.

Subglottic hemangiomas also have a characteristic radiographic appearance, demonstrating asymmetric subglottic narrowing on an anterior-posterior cervical airway radiograph (Fig. 5). Other tumors or subglottic cysts, however, also may present with the same radiographic characteristics.
Numerous management options exist for subglottic hemangiomas. The treatment modalities that have been described in the literature include laser ablation using CO₂ or potassium-titanyl-phosphate (KTP) laser, tracheotomy, external beam radiation, radioactive gold-grain implantation, cryotherapy, sclerosing agents, corticosteroid steroid therapy (systemic or intraleisional injection), and open surgical excision. In cases where airway obstruction is minimal, conservative therapy with no intervention may be warranted and the lesion may involute with complete resolution of symptoms. The most common intervention is CO₂ laser ablation. Used conservatively, this is an appropriate treatment modality. There is a risk, however, of subglottic stenosis developing as the result of aggressive laser ablation. This most likely is caused by over aggressive, circumferential lasering of large lesions. A 20% subglottic stenosis rate following laser excision has been reported in a large series of cases. Corticosteroid therapy also may be used for subglottic hemangiomas. Large lesions may respond to high-dose therapy (1 to 2 mg/kg per day) using prednisone. Significant systemic side effects may arise from long-term use of corticosteroids, such as cushingoid features, growth retardation, and possible development of sepsis. Open surgical excision also has been advocated. Children with large subglottic hemangiomas causing severe airway distress and respiratory obstruction, necessitating tracheotomy, may benefit from open surgical resection. This may be performed as a single-stage procedure requiring postoperative endotracheal intubation for 3 to
7 days. Also, a staged procedure may be performed with delayed tracheotomy decannulation after subglottic healing is complete.

POSTERIOR LARYNGEAL CLEFT

Posterior laryngeal cleft, or laryngotracheal esophageal cleft, is an uncommon congenital laryngeal anomaly that typically presents with aspiration, stridor, and weak cry at birth. Mild cases may be asymptomatic or have minimal symptomatology. Extensive cases, however, may be life-threatening or incompatible to quality of life. Posterior laryngeal cleft (PLC) may be an isolated laryngeal anomaly or associated with other congenital anomalies.

- Tracheoesophageal fistula
- Esophageal atresia
- Cleft lip and palate
- Congenital heart defects (transposition of great vessels)
- Gastrointestinal anomalies (imperforate anus, rectal stenosis, Meckel's diverticulum)
- Genitourinary anomalies (hypospadias, hypoplastic kidneys)
- Subglottic stenosis
- Hamartomas

Posterior laryngeal cleft also may be associated with congenital syndromes, such as G syndrome (Opritz-Frias syndrome), Pallister-Hall syndrome, or BBB syndrome (hypertelorism-hypospadias syndrome). The embryologic defect that causes an abnormal opening in the posterior larynx or posterior trachea results from a failure of fusion of the tracheoesophageal septum and of the dorsal laminae of the cricoid cartilage. The failure of fusion may result in an abnormal communication limited just to the larynx itself, or extending into the cervical and, possibly, thoracic trachea (Fig. 6). The most minor manifestation of this fusion defect is a deep interarytenoid cleft (Fig. 7). The mortality rate for laryngotracheal esophageal clefts is high; this is related, however, to the type of cleft involved. A 93% mortality rate is associated with laryngotracheal esophageal clefts with significant intrathoracic involvement. A 43% mortality rate, however, is associated with clefts limited to the laryngeal region. With the advent of more advanced surgical techniques, the mortality rate is possibly much less than this for clefts limited to the larynx.

Numerous classification systems exist for laryngeal clefts, including the system described by Benjamin and Inglis shown below.

Type I Supraglottic, interarytenoid cleft
Type II Partial cricoid cleft
Type III Complete cricoid cleft with or without extension into part of the tracheal esophageal wall
Type IV Laryngotracheal esophageal cleft
Figure 6. Type III posterior laryngeal cleft with extensive redundant mucosa noted in the interarytenoid area extending into the subglottis and upper trachea.

Surgical repair must be undertaken in all cases of symptomatic posterior laryngeal clefts or laryngotracheal esophageal clefts. For clefts limited to the larynx or upper trachea, an anterior laryngofissure approach may be used. This provides excellent exposure for the surgical repair.\textsuperscript{25,43} This may be performed as a single-stage operation with extubation taking place 3 to 7 days postoperatively. In cases where a tracheotomy is placed, a staged approach may be performed with decannulation taking place after healing is complete. For cases of more extensive clefts, a posterior lateral pharyngoesophagotomy approach may be used.\textsuperscript{38} Thoracotomy also may be necessary to obtain exposure for extensive clefts. Garabedian et al has reported the use of tibial periosteum as an interposition graft to

Figure 7. Type I posterior laryngeal cleft, presenting as deep interarytenoid cleft.
improve surgical results for repair of posterior laryngeal clefts. In addition to surgical repair of the cleft because of the significant aspiration problems of these children, a gastrostomy should be placed and fundoplication performed for alimentation requirements that need to be addressed perioperatively. The larynx may remain dysfunctional for a prolonged period of time following surgical repair, requiring long-term gastrostomy tube feedings.

CONGENITAL SUBGLOTTIC STENOSIS

Subglottic stenosis may be considered congenital or acquired. Since the advent of prolonged mechanical ventilation in cases of premature neonates with immature pulmonary development, the incidence of acquired subglottic stenosis is higher than the congenital form. In addition, the congenital form of stenosis tends to be less severe. Subglottic stenosis is defined as the presence of an abnormally small subglottic lumen (less than 3.5 mm diameter in newborn infant). In the absence of a history of endotracheal intubation, external neck trauma, or other cause of acquired subglottic stenosis, congenital subglottic stenosis accounts for 19% of all congenital laryngeal anomalies and is the most common cause of airway obstruction in children under the age of 1 year who necessitate tracheotomy. Subglottic stenosis may be associated with other congenital anomalies, such as vocal cord paralysis or congenital syndromes, such as Down syndrome.

Patients with congenital subglottic stenosis present with various degrees of symptomatology ranging from minimal airway obstruction to severe airway obstruction at birth. Occasionally, children may be diagnosed with mild congenital subglottic stenosis incidentally, for example, when a smaller than normal endotracheal tube is needed to intubate a patient at the time of another surgical procedure. Recurrent croup is another common presentation for congenital subglottic stenosis. Children who develop frequent episodes of croup with upper respiratory tract infections during the first few years of life should be evaluated for the possibility of congenital subglottic stenosis. Symptomatology usually consists of stridor, which is typically biphasic in nature, and a weak cry from the decrease in airflow through the airway obstruction.

Congenital subglottic stenosis may be demonstrated by radiographic techniques. Patients have a classic subglottic hourglass narrowing apparent on an anterior-posterior cervical airway radiograph. Flexible fiberoptic laryngoscopy should be performed in all cases to rule out other glottic or supraglottic pathology, such as vocal cord paralysis or laryngomalacia. The definitive diagnosis is made in the operating room using rigid endoscopy. The subglottic airway can be sized objectively in the operating room using a bronchoscope or an endotracheal tube and measuring the air leak around the endotracheal tube with positive pressure ventilation. Typically, the larynx should admit an endotracheal tube that allows an air leak of 20 cm of water pressure or less around the tube. The majority of
patients with congenital subglottic stenosis outgrow the condition within the first few years of life. Severe cases, however, may require tracheotomy or laryngotracheal reconstruction using costal cartilage grafting for expansion of the subglottic airway.\textsuperscript{15}

\section*{CONGENITAL ANOMALIES OF THE TRACHEA}

\subsection*{Tracheomalacia}

Tracheomalacia is a condition that is characterized by abnormal flaccidity of the trachea during the respiratory cycle leading to abnormal collapse on expiration. This condition also may involve the bronchi, where it is known as bronchomalacia. Patients with tracheomalacia may present with minimal symptomatology or severe life-threatening airway obstruction. Tracheomalacia may occur with laryngomalacia, but often occurs independently from laryngomalacia. The classic expiratory stridor of tracheomalacia may be present at birth. Often it is not apparent, however, until the infant becomes more active or during periods of respiratory tract infections. The tracheobronchial tree is more collapsible in a child than in an adult. During routine rigid or flexible bronchoscopy, a degree of collapse of the tracheobronchial tree may be noted. Tracheomalacia is defined as collapse on expiration of the trachea, which results in greater than 10\% to 20\% obstruction of the airway.\textsuperscript{4,6,37}

Tracheomalacia can be categorized into primary or intrinsic tracheomalacia, which is caused by an inherent weakness of the tracheal cartilaginous rings. In secondary tracheomalacia, the collapse on expiration may be because of extrinsic compression of the trachea by a mass or vascular structure, or a condition causing an intrinsic weakness of the tracheobronchial tree. Secondary tracheomalacia may occur with conditions such as tracheoesophageal fistula, cardiac abnormalities and vascular compression, localized tracheomalacia associated with tracheotomy, compression by mediastinal masses, or laryngotracheal esophageal cleft.\textsuperscript{57}

The assessment of a child with tracheomalacia should include airway fluoroscopy and a barium swallow to assess the airway under dynamic conditions radiographically and to demonstrate evidence of vascular compression. Rigid bronchoscopy should be performed to completely assess the airway, not only to evaluate the degree of the prolapse of the tracheobronchial tree, but also to assess the presence of other associated congenital airway lesions. Endoscopy should be performed under spontaneous ventilation technique so that the airway dynamics can be visualized endoscopically. In cases of suspected vascular compression or cardiac anomalies contributing to tracheomalacia, MR imaging and echocardiography also may be warranted. In patients with associated gastroesophageal reflux, upper gastrointestinal series and a pH probe study also may be warranted, especially in children who have secondary conditions, such as tracheoesophageal fistula.
The treatment of tracheomalacia depends on the severity of symptomatology. Mild cases of tracheomalacia often resolve within the first 1 to 2 years of age, necessitating no further intervention. In cases of severe airway obstruction and tracheal collapse, tracheotomy with long-term positive airway pressure ventilation may be required. In cases of vascular compression of the trachea, surgical decompression of the trachea may lead to significant improvement.

Surgical procedures specifically used to correct diffuse tracheomalacia have had various degrees of success. Surgical procedures that have been considered include placement of internal stents, segmental resection, cartilage grafting of the trachea, and external tracheal stents.6,37,46

**TRACHEAL STENOSIS**

Various degrees of tracheal stenosis may occur in infants, causing mild to severe airway symptomatology. Less severe, short-segment stenosis of the trachea may cause minimal symptomatology and resolve with time or be amenable to surgical repair, whereas long-segment diffuse tracheostenosis may cause severe symptomatology and have a very high mortality rate.20 Three types of tracheal stenosis have been described, including: (1) generalized tracheal hypoplasia; (2) funnel-like stenosis, tapering down toward the carina; and (3) short segmental stenosis. In addition, congenital complete tracheal rings are a condition in which the tracheal rings are fused posteriorly with no evidence of a posterior membranous tracheal wall. This may result in tracheal stenosis when diffusely located throughout the trachea, leading to a stove-pipe trachea or microtrachea.35 The cause of tracheal stenosis is usually congenital, although acquired stenosis may occur from prolonged intubation, inhalation burns, trauma, or primary tumors. Congenital airway anomalies may be associated with congenital tracheal stenosis, including congenital heart disease, tracheoesophageal fistula, skeletal abnormalities, subglottic stenosis, tracheomalacia, hypoplastic lungs, and bronchostenosis.3

Patients with tracheal stenosis usually present with stridor which may be biphasic in nature if located high within the trachea, or primarily expiratory if located lower in the tracheobronchial tree. Other findings, which may be associated with tracheal stenosis, include wheezing, cyanotic episodes, failure to thrive, atypical bronchiolitis, tachypnea, cough, and recurrent croup. A definitive diagnosis of tracheal stenosis usually is made by a combination of radiographic studies and rigid endoscopy. Airway fluoroscopy helps differentiate tracheomalacia and tracheal stenosis. CT scanning or MR imaging also may be beneficial to help assess the size of the airway, the diffuse nature of the underlying condition, and associated surgical and thoracic anomalies affecting the trachea. Rigid endoscopy should be performed only in the presence of an experienced endoscopist and anesthesiology team. Minimal trauma to the stenotic segment may lead to edema and acute airway obstruction requiring immediate
106 WIATRAK

airway intervention and possibly cardiopulmonary bypass to maintain oxygenation. Passage of instrumentation through the stenosis should be avoided unless the surgical team is present and prepared to perform a definitive repair at that time. For mild cases of tracheal stenosis with minimal symptomatology, conservative therapy is warranted. With time, as the patient grows, the stenosis may become less significant and less symptomatic.

Severe cases of tracheal stenosis with airway obstruction require surgical intervention. Numerous surgical modalities have been described for tracheal stenosis. Short segment lesions are amenable to tracheal resection with primary reanastomosis. Longer segmental lesions may be repaired using anteriotracheal split with pericardial patching, possibly combined with a posteriotracheal split. Costal cartilage grafting may also be used for expansion of the tracheal airway. Recently, slide tracheoplasty has been advocated, as well as homograft tracheal transplantation. Postoperative management may be difficult because of recurring granulation tissue at the repair site requiring multiple endoscopic procedures using laser excision of the granulation tissue. Tracheal stenosis involving the carina or bronchi is associated with a poor prognosis and a high mortality rate.

VASCULAR COMPRESSION OF THE TRACHEA

Although a significant portion of the population have anomalies involving the great vessels of the mediastinum (3%), only a small number result in symptomatic airway compression. Airway compression by vascular anomalies may present with minimal symptomatology, it may be asymptomatic and noted as an incidental finding at endoscopy, or it may result in severe compression requiring airway intervention. Vascular anomalies of the trachea may be divided into vascular rings that completely encircle the tracheoesophageal region compressing the airway; or vascular slings that are non-circumferential vascular compressions and may also lead to significant morbidity and occasional mortality. Double aortic arch is the most common vascular ring. The most common vascular sling causing airway compression is innominate artery compression. Patients with vascular rings typically present earlier in life than patients with vascular slings and with more severe airway symptomatology. Patients with a double aortic arch often need early surgical intervention to relieve airway obstruction. In the majority of patients with double aortic arch (Fig. 8), the left arch is smaller and even may be atretic or nonpatent in some cases. This is joined into a common aorta that usually descends on the left side.

Another less constricting vascular ring includes a right aortic arch with a descending right aorta associated with an aberrant left subclavian artery and persistent ligamentum arteriosum. Although this situation usually results in less airway compromise than a true double aortic arch, surgical intervention is required in many cases. Barium swallow is usually
diagnostic for vascular rings. MR imaging, however, also may add important diagnostic information for presurgical evaluation.

Innominate artery compression is the most common vascular sling resulting in airway symptomatology, and first was described by Gross and Neuhauser in 1948. Symptomatology for innominate artery compression may be less severe than vascular rings. Symptomatology warranting surgical intervention however, may occur. Cases usually present within the first year of life; cases have been reported, however, as late as 15 years of age. Symptomatology includes expiratory stridor, recurrent cough, apnea, and recurrent broncopulmonary infection. A condition known as reflex apnea or dying spells have been reported to be associated with innominate artery compression.

Anterior compression of the trachea may be noted on airway fluoroscopy. The definitive diagnosis, however, may be made using rigid bronchoscopy (Fig. 9). Typically, the compression is relatively short-segment, located a few centimeters above the carina. Classically, upward compression of the narrowing with the tip of the bronchoscope obliterates the right radial pulse upon palpation.
Figure 9. Anterior tracheal compression consistent with innominate artery compression.

In severe cases of airway obstruction, surgical intervention is warranted. Innominate artery compression of the trachea can be relieved by arteriopexy where the innominate artery is suspended from the sternum to relieve compression upon the trachea. The innominate artery also may be divided and reimplanted more proximal on the aorta to the right side so that it does not cross and compress the trachea.

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