Paparella: Volume III: Head and Neck

Section 2: Disorders of the Head and Neck

Part 5: The Larynx, Trachea, and Esophagus

Chapter 26: Congenital Abnormalities of the Larynx and Trachea and Management of Congenital Malformations

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Anatomic abnormalities of the larynx and trachea in infants and neonates are frequently characterized by stridor. Stridor is a frightening symptoms for parents and a diagnostic challenge for the clinician. It is a physical sign produced by turbulent air flow through a partial obstruction and may be inspiratory or expiratory, or both. The presence of stridor merits immediate investigation, for it may be indicative of a pathologic narrowing of the airway that could result in significant respiratory compromise and even death. In all cases, an evaluation must be undertaken to determine the underlying cause.

The time available to make the diagnosis may be only a matter of minutes if the onset of stridor is accompanied by severe respiratory obstruction. Without significant respiratory distress, a prompt but more thorough investigation can be made before arriving at a diagnosis and embarking upon appropriate medical or surgical therapy. When the child is not in extremis, radiographic evaluation of the airway may be a valuable adjunct to endoscopic examination.

Although the obstructive process that results in the production of stridor is generally intrinsic to the airway, it may be caused by extrinsic compression of the soft, collapsible pediatric airway. Because of a child's small airway dimensions, even a slight obstruction can be accompanied by significant stridor and respiratory distress.

Pathophysiology of Stridor

A basic knowledge of the Venturi principle is necessary to understand how stridor is produced. The Venturi principle states that the pressure exerted by gas is equal in all directions except when there is linear movement. When this occurs, additional pressure in the forward vector produces a corresponding fall in the lateral pressure. As a result, a partially collapsible tube like the trachea may close when the lateral pressure suddenly drops. Following closure, pressure is released and the lumen opens again. This pattern of intermittent flow creates vibratory patterns, with the production of audible sounds that are interpreted as stridor, wheezes, rhonchi, or rales. When there is an anatomically fixed lesion, similar sounds are generally created with each respiratory cycle.

Regardless of the location or cause, respiratory obstruction can produce hypoventilation with an increase in PCO_2 and a fall in PO_2. Recognition of the signs and symptoms of airway obstruction promotes early diagnosis. When stridor is secondary to a laryngeal or tracheal abnormality, there is frequently a characteristic sound produced, which can be analyzed in regard to its pitch, frequency, and the time of the respiratory cycle during

which it appears. Patients with supraglottic laryngeal obstruction frequently present with a muffled or throaty voice. They often snore during sleep and produce coarse inspiratory sounds at rest. Inspiratory stridor results because the inspiratory pressure and the Venturi principle cause the loose tissues of the pharynx and supraglottis to collapse inward and produce obstruction. During expiration, this area of the airway is generally enlarged.

Patients with glottic obstruction usually have inspiratory stridor initially, which becomes expiratory as the obstruction increases. When the obstruction is severe, the stridor is characteristically to and fro. In a manner similar to that in patients with glottic obstruction, patients who present with subglottic obstruction usually have inspiratory stridor early in their course, which becomes expiratory as the obstruction increases. With severe obstruction, the stridor may be both inspiratory and expiratory. The glottis and subglottis are fixed in size and are unaffected by inspiratory or expiratory air pressure. Because flow depends on lumen size, narrowing in either of these regions can produce biphasic stridor. In an infant or child with biphasic stridor, a congenital anatomic abnormality must be suspected, and prompt evaluation is indicated.

In patients with tracheobronchial obstruction, stridor is characteristically expiratory with a wheezing component. With severe obstruction, the stridor may be both inspiratory and expiratory. During inspiration, the pressure developed within the chest generally enlarges the thoracic trachea and larger bronchi. However, on expiration, there is a collapse of these airways, thus accentuating stridor caused by any lesion in this region (Richardson and Cotton, 1984; Myer and Cotton, 1988).

History

The history is one of the most important steps in evaluating stridor in childhood. Although emergent treatment of a child in respiratory distress may be necessary, an accurate history should be obtained whenever possible. Specific points that must be elicited during the history include (1) the time of onset of stridor and whether it is gradual, progressive, or sudden; (2) the duration of stridor; (3) the nature of stridor (inspiratory, expiratory, biphasic); (4) the characteristics of cry or voice; (5) the relationship of stridor to feeding; (6) the association with aspiration or reflux; (7) cyanosis; (8) association with agitation or positioning; (9) previous trauma, including prior intubation; (10) foreign body aspiration or ingestion; and (11) other associated congenital abnormalities (Richardson and Cotton, 1984).

Physical Examination

Although the physical examination is important in a child with stridor, any child in respiratory distress should not be moved or agitated if one feels respiratory arrest is imminent. Further evaluation should be undertaken only when equipment for resuscitation is available, even if this is in the operating room. When acute distress is not present, one should always observe the child with stridor breathe at rest in order to determine the degree of effort necessary to maintain adequate oxygenation. Auscultation by ear and stethoscope may be used to further quantify and analyze the sounds present. The stridor should be evaluated in terms of its pitch, duration, and timing during the respiratory cycle. The respiratory rate should be noted, and the child should be observed closely for tachypnea or onset of fatigue. In addition, one should look carefully for chest retractions, flaring of the nasal ala, or other signs of

increased respiratory effort.

The influence of positional changes and agitation on stridor should be evaluated as well. The child should be observed both feeding and drinking, and some evaluation of the voice should be performed by having the child cry, cough, or speak. Careful examination of the cervical region should be performed to look for mass lesions that might cause extrinsic compression. If the child does not have severe respiratory distress, a flexible laryngoscope may be used to assess the supraglottis and glottis (Richardson and Cotton, 1984).

In the evaluation of patients with supraglottic laryngeal obstruction, the voice is commonly muffled and throaty. The stridor is noted to be inspiratory and may even be described as snoring or snorting. Retractions are generally not present unless the obstruction is quite severe. Feeding may be difficult secondary to obstruction of the upper aerodigestive tract by the supraglottic lesion. Patients commonly breathe with the mouth open and the jaw held forward.

In patients with glottic obstruction, the voice may be hoarse or aphonic. The stridor is generally inspiratory but becomes expiratory as obstruction increases, and it may be described as to and fro with severe obstruction. Retractions are commonly present both in the xiphoid region and, possibly, in the suprasternal or supraclavicular areas. Feeding is usually normal except with severe airway obstruction. Patients commonly breathe with the mouth closed but with the nasal ala flared. There is usually no cough associated with glottic obstruction. Patients with subglottic obstruction have a very similar picture to those with glottic obstruction. However, a barking cough is frequently seen in such patients.

Patients with tracheobronchial obstruction usually have normal voices. The stridor that is present is usually expiratory with wheezing, but it becomes to and fro with increasing obstruction. There are usually no retractions present unless there is severe obstruction. When present, retractions are usually in the xiphoid or sternal region. Patients with obstruction in the tracheobronchial tree are usually able to feed normally except when obstruction is severe or when the obstruction is due to extrinsic pressure and involves the esophagus. Patients usually breathe with the mouth closed and with the nasal ala flared. When a cough is present, it is commonly described as brassy (Myer and Cotton, 1988).

Radiographic Evaluation

Once a thorough history and physical examination have been obtained, radiographic examination of the airway is valuable in evaluating a child with stridor. Generally, this evaluation consists of anteroposterior and lateral plain films of the soft tissues of the neck and chest. If feeding abnormalities have been noted during the history or physical examination, a barium esophagram is helpful in determining if there is a laryngeal cleft, a tracheoesophageal fistula, or a vascular abnormality extrinsically compressing the airway.

Higher-kilovoltage anteroposterior and lateral radiographs of the neck and chest are the basic radiographic studies obtained in all infants with stridor. The studies of these two regions complement each other, but correct technique is very important and difficult to obtain in small infants. The radiographs must be taken in full inspiration with the head in extension. The airway in children is quite mobile, and if proper technique is not used, misinterpretation

is common. For example, if the head is not extended properly, the trachea frequently appears compressed. The mobility of the airway in infants and their rapid respiratory rate presents special problems for radiology technicians. Satisfactory inspiratory radiographs may be impossible to obtain and, under these circumstances, fluoroscopy is important and demonstrates airway dynamics to the clinician. Although use of a flexible endoscope in the infant is superior to radiology in the diagnosis of lesions in the glottis and supraglottis, fluoroscopy adds a dimension to the study of the trachea that may not be obtained even with a rigid endoscope.

Esophageal disease in infants may be associated with stridor because of the softness and elasticity of the cartilaginous framework of the airway, and a barium esophagram is often an important diagnostic tool. Because vascular rings frequently indent the esophagus, as well as compress the trachea, a barium esophagram frequently allows diagnosis of such a lesion. In addition, aspiration secondary to laryngeal paralysis, a laryngeal cleft, or pharyngeal disease may be demonstrated by such a study (Richardson and Cotton, 1984).

Other radiologic investigations may be of benefit in selected circumstances. Computed tomographic (CT) scanning of the upper aerodigestive tract is of limited value in the evaluation of congenital laryngotracheal disease. However, magnetic resonance imaging (MRI) has proved to be an excellent alternative to angiography in identifying anatomic features of suspected vascular rings. It is also beneficial in the evaluation of mass lesions in the mediastinum that cause tracheal compression (Bisset et al, 1987).

Physiologic Techniques

Pulmonary function testing may be of some benefit in the assessment of the respiratory status of some older infants. Though a measure of cooperation is required in order to obtain optimal results, in some children the measurement of inspiratory and expiratory air flow can produce flow-volume loops that are helpful in the diagnosis of the site of anatomic obstruction (Richardson and Cotton, 1984).

Endoscopy

Endoscopic evaluation of the larynx, trachea, and sometimes the esophagus is necessary to definitively make a diagnosis regarding the site of anatomic obstruction. Although the history, physical examination, radiographic examination, and physiologic testing are all important adjuncts in diagnosis, there is not substitute for a thorough examination of the pediatric airway. Photographic documentation should be obtained whenever possible (Richardson and Cotton, 1984).

Laryngeal Anomalies

The most common causes of congenital airway obstruction are laryngeal anomalies. Although obstruction is frequently present at birth, symptoms are often not apparent until the lesion has grown to a critical size or until a superimposed inflammatory process is present. The most frequent congenital laryngeal anomalies causing respiratory obstruction are laryngomalacia, laryngismus stridulus, reflux laryngospasm, laryngeal cleft, vocal cord paralysis, laryngeal web, laryngotracheoesophageal cleft, subglottic stenosis, and subglottic

hemangioma. Congenital tracheobronchial anomalies that may cause stridor include tracheomalacia, vascular rings and slings, innominate artery compression of the trachea, tracheoesophageal fistula, and bronchogenic cysts (Myer and Cotton, 1988; Smith and Catlin, 1984).

Laryngomalacia

Laryngomalacia is a relatively benign, self-limiting condition frequently referred to as congenital laryngeal stridor. It is generally felt to be the most common congenital laryngeal anomaly.

There is no consensus regarding the pathophysiology of laryngomalacia. Some investigators feel the process is secondary to an anatomic abnormality of the epiglottis, whereas others suggest the problem is related to increased flaccidity of the supraglottic airway. McSwiney and colleagues (1977) described three anatomic abnormalities of the epiglottis that predisposed patients to laryngomalacia. The abnormalities included (1) an elongated epiglottis that was curled upon itself (omega-shaped epiglottis) and prolapsed posteriorly on inspiration, (2) foreshortened aryepiglottic folds, and (3) arytenoids that were bulkier than normal and tended to prolapse with inspiration. Though any of these abnormalities may certainly be found in a child with laryngomalacia, they are frequently noted in children without stridor who have no symptoms of airway obstruction. Thus, these abnormalities do not appear to be specific for laryngomalacia.

Other investigators have related the increased flaccidity of the epiglottis in patients with laryngomalacia to cartilaginous immaturity of the epiglottis. However, there have been no consistent gross or microscopic abnormalities seen in the evaluation of the epiglottis in patients with laryngomalacia. Furthermore, the theory of cartilaginous immaturity with increased malleability is questionable, since premature infants do not exhibit laryngomalacia consistently (Zalzal et al, 1987).

Another possible cause of laryngomalacia may relate to poor neuromuscular control, resulting in inadequate muscular support of the cartilaginous framework of the epiglottis with increased compliance of the supraglottic tissues. Not all patients with laryngomalacia have generalized neuromuscular insufficiency, and this concept is certainly difficult to prove. Thus, both mechanisms - that is, poor neuromuscular control, secondary to either hypomaturity or dysfunction, and poor cartilage support - may be responsible for collapse of the supraglottic structures in patients with laryngomalacia (Belmont and Grundfast, 1984).

Patients with laryngomalacia generally present with inspiratory stridor that is intermittent and of variable intensity; it decreases with extension of the neck and prone positioning but becomes augmented with agitation. Affected patients will have normal cry without cyanosis, and there is rarely dyspnea or difficulty with swallowing. The process usually appears during the first 10 days of life, and other coexistent anomalies of the larynx and trachea may be found. Although use of a rigid endoscope is not required in all patients with a diagnosis of laryngomalacia, one should be used whenever the patient does not improve spontaneously over time or when there is significant respiratory distress.

In making the diagnosis of laryngomalacia, other congenital anomalies of the larynx and trachea should be considered in the differential diagnosis. In children with bilateral vocal cord paralysis, the cry is frequently weak, and there is severe stridor and respiratory distress. Conversely, unilateral vocal cord paralysis usually presents with cough and feeding difficulties. Patients frequently have positional stridor, which improves during sleep. However, when the patient is awake, the stridor appears louder than that commonly seen in patients with laryngomalacia. Patients with subglottic hemangioma usually present later in life than do those with laryngomalacia, and almost one half of the patients with subglottic hemangioma will have cutaneous hemangiomas as well.

Routine radiographic studies obtained in the evaluation of stridor may be consistent with laryngomalacia. One is often able to demonstrate bowing with downward and inferior displacement of the aryepiglottic folds. Both the epiglottis and the hyoid bone may appear to be positioned lower than normal secondary to imbalance between the suprahyoid and infrahyoid muscles. The diagnosis is made definitively, however, by the use of a flexible laryngoscope, which demonstrates a small, long, narrow epiglottis with long, floppy aryepiglottic folds, prominent arytenoids, and a deep interarytenoid cleft. During inspiration, the structures collapse and leave a slitlike opening.

Symptoms are often relieved when the infant is placed in the prone position or when the head is flexed so that the supraglottic structures are removed from the glottic inlet. As mentioned previously, the condition is usually self-limited, and most patients improve before the age of 2 years. In patients who do not improve spontaneously or in those who exhibit symptoms of obstructive sleep apnea, cor pulmonale, or failure to thrive, surgical intervention may be necessary.

Formerly, tracheotomy was done in almost all severe cases. Though it is certainly an effective treatment for severe cases, patients with a tracheotomy require constant monitoring and attention. Hyomandibulopexy is an alternative form of therapy that has been used successfully in a limited number of patients. In this procedure, a cervical incision is made between the hyoid bone and the mandible. Sutures are used to suspend the hyoid bone as close as possible to the mandible in order to pull the epiglottis and aryepiglottic folds anteriorly and superiorly, thus providing relief from supraglottic obstruction.

Another method of therapy for patients with laryngomalacia is supraglottic tissue excision, or epiglottoplasty. In this procedure. scissors are used to trim the lateral edges of the epiglottis and the aryepiglottic folds in addition to the mucosa overlying the arytenoid and corniculate cartilages. Prior to the performance of this procedure, direct laryngoscopy and bronchoscopy should be performed to rule out concomitant causes of airway obstruction. Caution should be exercised to limit the amount of supraglottic tissue removed in order to minimize possible complications such as aspiration or supraglottic stenosis (Zalzal et al, 1987).

In selected individuals, cricopharyngeal incompetency and gastroesophageal reflux may exacerbate otherwise relatively mild laryngomalacia. Esophageal dysfunction should be controlled prior to any surgical treatment for laryngomalacia.

Vocal Cord Paralysis

The second most common congenital laryngeal abnormality is vocal cord paralysis, which accounts for approximately 10 per cent of all congenital laryngeal anomalies. Vocal cord paralysis may be unilateral or bilateral, corresponding generally to peripheral or central neurologic deficits, respectively.

Unilateral vocal cord paralysis is a problem that is more common than bilateral vocal cord paralysis, and affected patients generally do quite well. Patients frequently have a weak or muffled cry, but airway obstruction is usually minimal except for stridor during periods of stress or aggravation. Respiratory distress may occur during feeding on occasion, and careful nursing may be necessary. Unilateral vocal cord paralysis is most often caused by a peripheral lesion, and one must investigate the entire path of the recurrent laryngeal nerve for a pathologic condition. Frequently, the investigation will reveal cardiac or great vessels abnormalities. Recurrent laryngeal nerve injury during birth must always be considered in cases of unilateral paralysis and, less frequently, with bilateral paralysis. The diagnosis can usually be made by use of the flexible endoscope and no therapy is necessary in most patients. In some individuals, symptoms can be minimized by placing the child on the side of the paralysis so that the paralyzed vocal cord is passively moved away from the midline. In patients who tend to aspirate, the affected vocal cord may be moved closer to the midline by polytetrafluoroethylene (Teflon) injection of the vocal cord or by an external medialization procedure. Microlaryngoscopy and bronchoscopy of the entire airway is indicated prior to surgical intervention in any child with unilateral vocal cord paralysis. Since most patients with unilateral vocal cord paralysis require no airway support, an additional pathologic condition must be suspected in patients who do require support. In general, surgical intervention for unilateral true vocal cord paralysis should be delayed at least 9 months to allow for spontaneous recovery.

Patients with bilateral vocal cord paralysis usually present with acute airway distress, which necessitates intubation and, subsequently, tracheotomy. The vocal cords are usually located in the paramedian position and fail to abduct on inspiration. The patient's cry may be normal, but there is characteristically a high-pitched inspiratory stridor that worsens during periods of agitation. Recurrent pneumonia may be a problem secondary to frequent aspiration and an inability to cough effectively. Patients with bilateral vocal cord paralysis may have significant problems with dysphagia secondary to pharyngeal incoordination. This is most likely secondary to dysfunction of multiple cranial nerves. Though a thorough investigation may fail to yield a cause for bilateral vocal cord paralysis, central nervous system anomalies such as hydrocephalus and the Arnold-Chiari malformation with brain stem herniation must be considered as potential causes. Other possible central nervous system causes of bilateral vocal cord paralysis include intracerebral bleeding during birth, meningocele, encephalocele, cerebral agenesis, and dysgenesis of the nucleus ambiguus. Spontaneous resolution often takes place during the first several years of life, and definitive therapy should be deferred for at least this period of time once an airway is established. If, however, vocal cord movement is not noted after several years, an arytenoidectomy may be appropriate to augment the airway (Richardson and Cotton, 1984).

Congenital Subglottic Stenosis

The third most common congenital laryngeal anomaly is congenital subglottic stenosis. This entity results from inadequate recanalization of the laryngeal lumen following the completion of normal epithelial fusion at the end of the third month of gestation. The magnitude of stenosis depends upon the degree of recanalization and may range from complete laryngeal atresia through varying degrees of stenosis and webs. In fact, the mildest form of congenital stenosis is one in which the cricoid ring merely has a smaller than normal diameter or is eccentrically shaped.

Children with mild to moderate congenital subglottic stenosis are often asymptomatic until an upper respiratory tract infection causes additional narrowing of an already compromised airway. Progressive respiratory difficulty will develop as the prime symptom of airway obstruction associated with biphasic stridor, dyspnea, air hunger, and vigorous efforts of breathing. Suprasternal, intercostal, and diaphragmatic retractions are common. Cry is normally not affected, but dysphagia and feeding abnormalities may be present when there is severe obstruction.

A complete physical examination of the upper aerodigestive tract is required to make the diagnosis and to rule out associated congenital anomalies or acquired injuries. The radiographic studies previously described are again useful in the evaluation of a child with suspected congenital subglottic stenosis. Indirect laryngoscopy cannot be relied upon for diagnosis, and visualization of the larynx is essential to carefully examine the stenosis, assess its thickness and length, monitor active mobility of the vocal cords, obtain diagnostic biopsy specimens to exclude lesions other than fibrosis, and to look for other pathologic airway conditions. A flexible fiberoptic laryngoscope should always be used in patients with stridor to assess the dynamics of vocal cord function. An impression of subglottic stenosis may be obtained, but for a detailed assessment of this area, the child must be anesthetized and the larynx carefully examined using a microlaryngoscope. A rigid Storz-Hopkins optical telescope is used to examine the area and, if possible, it is passed through the region of narrowing to assess the trachea. An accurate impression of the size of the lumen can be obtained by passing a bronchoscope or endotracheal tube with a known outside diameter.

The diameter of the normal subglottic lumen in a full-term neonate is 4.5 to 5.5 mm. In premature infants, it is approximately 3.5 mm. When the subglottic diameter is less than 4 mm in a full-term neonate, or less than 3 mm in a premature neonate, the subglottis is considered to be narrowed. Subglottic stenosis is considered to be congenital when there is no prior history of endotracheal intubation or other apparent acquired cause of stenosis. The diagnosis may be difficult to substantiate, and it is not known how many intubated premature infants who fail extubation actually have an underlying congenital stenosis that is aggravated by intubation.

Congenital subglottic stenosis may be either membranous or cartilaginous, or both. The membranous type is a fibrous soft tissue thickening of the subglottic area that is due to either increased fibrous connective tissue or hyperplastic dilated mucous glands without an inflammatory reaction. It is usually circumferential, and the narrowest area is approximately 2.5 mm below the vocal cords, sometimes extending upward to include the true vocal cords. The cartilaginous types of congenital subglottic stenosis are more variable, but the most

common is a thickening or deformity of the cricoid cartilage, which results in a shelflike plate of cartilage that partially fills the concave inner surface of the cricoid ring anteriorly and extends posteriorly as a solid rigid sheet, leaving only a small posterior opening. Alteratively, the cricoid cartilage may be of normal configuration but small size. This is frequently difficult to diagnose endoscopically because of the lack of a precise means of measuring endolaryngeal dimensions endoscopically. Another diagnostic dilemma is the trapped first tracheal ring. In this situation, the first tracheal ring fails to descend normally and is trapped inside the lower cricoid cartilage. This entity is difficult to distinguish endoscopically from a congenitally small cricoid cartilage (Richardson and Cotton, 1984; Cotton, 1987).

As previously mentioned, many patients with congenital subglottic stenosis are asymptomatic except during periods of upper respiratory infection. In some individuals, there may actually be a history of recurrent croup. Airway support is indicated in those patients with severe upper respiratory obstruction, but a previous report by Holinger and colleagues (1976) stated that only 44 per cent of their patients with congenital subglottic stenosis needed a tracheotomy.

Most congenital subglottic stenoses improve as laryngeal growth occurs, and a less vigorous surgical approach is indicated for congenital subglottic stenosis than is advocated for acquired subglottic stenosis. However, an anterior cricoid split procedure may be appropriate in selected cases when there is a congenital malformation of the cricoid cartilage with a shelflike plate anteriorly and a posterior airway. If the stenosis is more extensive, laryngotracheal reconstruction when the child has reached approximately 10 kg in weight is appropriate.

One should always remember that congenital subglottic stenosis may be associated with other congenital lesions and syndromes, such as Down's syndrome. Patients with this condition have a larynx that is smaller than that of other patients of the same size and age. Though the child with Down's syndrome may be asymptomatic with the small larynx, endotracheal intubation in these patients should always be done with an endotracheal tube that is smaller than that used normally. The duration of intubation should be minimized as well (Richardson and Cotton, 1984).

Laryngeal Web

Laryngeal webs are uncommon anomalies that may occur in the supraglottis, glottis, or subglottis. They are representative of the spectrum of disorders associated with abnormal recanalization of the embryonic larynx, with laryngeal atresia as the most extreme form.

Laryngeal webs are often symptomic from birth, as patients present with signs and symptoms of airway obstruction, a weak cry, and, occasionally, aphonia. If the web is quite extensive, emergency intubation or tracheotomy in the delivery room may be necessary.

The diagnosis of a laryngeal web is made during endoscopy. Approximately 75 per cent of webs are located at the level of the glottis and are usually anterior with a concave posterior glottic chink of varying size. The thickness of the web is quite variable and may range from a membrane that is easily ruptured to a thick, fibrous band that extends into the subglottis anteriorly. Webs of similar consistency may be found in equal distribution between

the supraglottis and the subglottis.

Treatment must be modified depending upon the thickness of the web. Although thin webs may respond well to endoscopic lysis using either a knife or the CO_2 laser, thicker webs more commonly will require tracheotomy in addition to an open laryngeal procedure. In such circumstances, lysis of the web is followed by placement of a keel for a short time. In all circumstances, several procedures, including laryngeal dilatation, may be necessary to prevent re-formation of the web (Richardson and Cotton, 1984).

Laryngeal Atresia

Laryngeal atresia is a very unusual condition that is incompatible with life unless it is immediately recognized and treated within the first few minutes of delivery. An exception occurs when there is a concomitant tracheoesophageal fistula that is large enough to support ventilation. Tracheostomy is necessary to prevent death.

Patients with laryngeal atresia make active respiratory efforts without success, and cyanosis rapidly develops following umbilical cord clamping. Attempts at intubation are unsuccessful and, once the diagnosis is suspected, laryngoscopy and bronchoscopy should be performed in order to obtain diagnosis. On occasion, the atretic region may be relatively thin and may allow passage of a bronchoscope in order to sustain ventilation. If this is not possible, however, an alternative airway must be provided in the form of a tracheotomy, a cricothyrotomy, or the insertion of a large-bore intravenous catheter or needle into the trachea distal to the obstruction. Patients with laryngeal atresia may have other associated congenital anomalies that are themselves lethal (Richardson and Cotton, 1984).

Subglottic Hemangioma

Although hemangioma is the most common head and neck tumor in infancy and early childhood, infantile subglottic hemangioma is rare. Approximately 50 per cent of children who have subglottic hemangioma have associated cutaneous lesions. In a child who presents with signs and symptoms of upper airway obstruction, such a cutaneous lesion suggests the diagnosis of subglottic hemangioma (Batsakis, 1979). Though a subglottic hemangioma may be present at birth, patients frequently are not symptomatic until they are at least 3 to 6 months of age. This increase in symptoms is secondary to the growth of the lesion, which usually occurs during the first year of life. Stridor in patients with subglottic hemangioma is characteristically inspiratory but may become biphasic. There is usually no associated hoarseness or change in cry. Agitation or upper respiratory infection can increase airway symptomatology by an increase in venous pressure (Shikhani et al, 1986).

Radiographs of the airway have a characteristic appearance if a subglottic hemangioma is present. An anteroposterior neck film demonstrates asymmetric narrowing in the subglottis, whereas a lateral radiograph demonstrates a posterior subglottic mass. When there are supraglottic or glottic manifestations of a hemangioma, use of the flexible endoscope may allow the diagnosis to be made. Without these signs, however, the diagnosis may be more evasive, because a flexible endoscope is of limited value in the diagnosis of a subglottic pathologic condition. If the child presents with marked respiratory distress, use of the flexible endoscope should be bypassed in lieu of direct endoscopy.

Direct endoscopy is the definitive diagnostic procedure for subglottic hemangioma. The lesion usually can be recognized endoscopically as a unilateral, sessile, compressible mass arising between the undersurface of the true vocal cord and the lower border of the cricoid ring. Occasionally, a circumflex mass can be noted. In most cases, the mass is reddish blue and the diagnosis is easily made, but the diagnosis may be more difficult if the hemangioma is colorless. The advisability of biopsy is controversial because of the potential for hemorrhage, but there appears to be a very low incidence of bleeding following biopsy (Batsakis, 1979; Shikhani et al, 1986).

The management of subglottic hemangioma is directed by the degree of airway obstruction, and this is determined by the clinical examination, blood gas evaluation, airway radiography, and endoscopic examination. Because the natural history of the lesion is to involute spontaneously, observation is a rational therapeutic approach for a patient who has minimal distress.

More severe symptoms, however, almost always require definitive intervention. Management options have included (1) tracheotomy and observation until spontaneous regression occurs, (2) radiation therapy, (3) implantation of radioactive gold seeds, (4) injection of sclerosing agents, (5) systemic corticosteroid therapy, (6) surgical excision, (7) cryotherapy, and (8) CO_2 laser excision (Healy et al, 1984). For the patient who does not have life-threatening airway obstruction but does require intervention, dexamethasone sodium phosphate (1 mg/kg/day for approximately 1 week) and the prednisone (3 mg/kg/qid) until the child is approximately 1 year of age may be administered to diminish the size of the hemangioma. Higher dose steroids may be necessary in selected situations. At the time the child is approximately 1 year of age, weaning from the steroids should be attempted, since spontaneous regression may have begun. Reducing the steroid dose may be necessary if the child gains insufficient weight. As a therapeutic alternative, the CO_2 laser is often useful in the treatment of small, well-circumscribed lesions. This mode of therapy should be avoided for extensive lesions because of the potential for creating subglottic stenosis. Similarly, open surgical excision is avoided if possible. A patient who presents with severe upper airway obstruction secondary to a hemangioma should undergo a tracheotomy (Cotton and Tewfik, 1985; Hawkins et al, 1984; Healy et al, 1984; Mizono and Dedo, 1984; Narcy et al, 1985).

Generally, a physician may await resolution of a hemangioma after a tracheotomy is performed. However, some patients with subglottic hemangiomas will have an associated hemangioma in the mediastinum. Continued growth of the mediastinal hemangioma can increase the degree of respiratory distress even though a tracheotomy has been performed to bypass the subglottic obstruction. When there is a significant mediastinal component, patients frequently have increased amounts of secretions that are difficult to clear and there is a propensity to pulmonary infection. Additionally, there may be dysphagia and poor weight gain secondary to both esophageal compression and airway obstruction. The physician should maintain a high index of suspicion of mediastinal involvement in patients with subglottic hemangioma. Bronchoscopy can confirm significant anterior tracheal compression, and a CT or MRI scan of the mediastinum can clearly demonstrate a mass in this region. A chest radiograph may demonstrate anterior tracheal compression from mediastinal involvement, but a CT or MRI scan is necessary for definitive diagnosis.

When there is symptomatic airway obstruction from a mediastinal hemangioma, oral steroids should be given in the doses previously mentioned. If steroids do not shrink such a lesion and symptomatology persists, irradiation may be considered (Shikhani et al, 1986; Healy et al, 1984).

In summary, the natural history of subglottic hemangioma in children is favorable. Medical and surgical intervention should be limited unless there is significant airway obstruction. Above all, any therapeutic maneuvers that significantly risk complications should be avoided, since the natural history of hemangiomas is spontaneous regression.

Laryngotracheoesophageal Cleft

The laryngotracheoesophageal (LTE) cleft is a rare congenital anomaly that is associated with high mortality. The mortality rate is reflection not only of the severity of the anomaly itself but also of the consequence of delayed diagnosis and treatment and other coexistent major congenital anomalies. Early diagnosis and management is the key factor in reducing the high mortality of this disorder. The LTE cleft results from the lack of fusion of the posterior cricoid lamina, creating an abnormal communication between the larynx and the hypopharynx. The defect may be confined to the cricoid itself or it may extend down the entire common wall between the trachea and the esophagus. The defect is the result of failure of the rostral development of the tracheoesophageal septum at approximately 7 weeks of gestational age. Maternal polyhydramnions is felt to be a predisposing factor (Richardson and Cotton, 1984).

Patients with an LTE cleft commonly present with feeding problems and respiratory distress secondary to repeated episodes of aspiration. Coughing, choking, and cyanosis are characteristically exacerbated by attempts at feeding. Stridor and voice abnormalities are present in many patients. The symptomatic severity if frequently proportionate to the extent of the cleft, and small clefts may remain asymptomatic for a long time. Approximately 20 per cent of the patients may have an associated tracheoesophageal fistula, and other congenital abnormalities are seen in smaller proportions.

In a patient in whom an LTE cleft is suspected, a radiographic evaluation is appropriate prior to the endoscopic examination. The chest radiograph is not diagnostic but may show an infiltrate characteristic of an aspiration pneumonia. A barium esophagram may show spillover of contrast medium into the trachea but may appear normal unless carefully performed. The radiologist should be alerted to the possibility of the diagnosis so that he or she may carefully examine the patient for evidence of tracheoesophageal fistula in addition to the LTE cleft.

Endoscopic examination is necessary to make the definitive diagnosis of an LTE cleft. A high index of suspicion is necessary by the endoscopist so that the lesion is not missed. If the lesion is considered, one part of the endoscopic examination should consist of the placement of an endotracheal tube in order to stent the edges of the cleft apart so that the cleft becomes obvious. Other differential diagnostic considerations include other anatomic defects, central nervous system lesions, esophageal disorders, and a variety of neuromuscular disorders.

Key elements in the management of a patient with an LTE cleft include providing respiratory support, preventing aspiration, and performing an appropriate surgical repair. Initial airway maintenance should be obtained either by placement of an endotracheal tube or a tracheotomy, but a tracheotomy will be required at some point for almost all patients. It should be noted, however, that if the cleft is extensive, the tracheotomy tube frequently slips over the superior edge of the defect into the esophageal lumen. Because of this, early reconstructive surgery is necessary in severe cases.

Following the establishment of an airway, gastric decompression and alimentation are required unless the cleft is quite limited. Gastric decompression is essential to prevent repeated aspiration that would lead to pneumonia and possibly death. Though a nasogastric tube may be used initially, two problems prevent its long-term use: (1) the presence of a nasogastric tube may contribute to reflux and aspiration and (2) the tube may dislodge anteriorly into the airway and cause obstruction. Though a gastrostomy has been advocated by some, this does not eliminate the danger of aspiration and pulmonary sequelae. An acceptable alternative appears to be a high gastric diversion with a double gastrostomy until such time that successful surgical repair of the cleft takes place and there is restoration of adequate esophageal physiology. In this way, aspiration is minimized.

Several approaches have been advocated for the repair of LTE clefts. For extensive clefts, a lateral pharyngotomy is advocated, as it offers excellent visualization of the laryngeal and esophageal defects. The repair is performed in layers, using laterally based flaps. The recurrent laryngeal nerve is rarely found during the surgical procedure, even with a careful surgical dissection. With clefts limited to the cricoid lamina, an anterior laryngofissure provides excellent exposure. Potential complications of surgical repair include complete or partial separation of the repair, posterior glottic stenosis, and esophageal stricture (Richardson and Cotton, 1984; Cotton and Schreibner, 1981).

Laryngeal Cysts and Laryngoceles

The clinical presentation of laryngeal cysts and laryngoceles is quite similar, since both involve a cystic dilation of a portion of the larynx. A laryngocele arises as a dilation of the saccule of the laryngeal ventricle. The saccule itself arises from the anterior end of the ventricle and courses upward between the thyroid cartilage and the ventricular fold, often ending at the upper border of the thyroid cartilage. A laryngocele generally results from an increase in intralaryngeal pressure, causing expansion of the saccule. Laryngoceles have been classified as internal, external, and combined, depending on their location in relation to the thyroid cartilage. The lesions may be unilateral or bilateral and may become infected during periods of upper respiratory infection. Endoscopic excision is appropriate with small lesions, but an external approach is required for most external laryngoceles and larger internal laryngoceles. A tracheotomy may be necessary in selected circumstances. Congenital laryngeal saccular cysts of the newborn are usually located in the supraglottis. They commonly arise within the aryepiglottic folds and may extend inferiorly to the laryngeal ventricle. Airway obstruction and feeding abnormalities are common because the cysts prolapse into the laryngeal lumen medially and into the pyriform sinus laterally.

Patients generally present with stridor and airway obstruction, which often necessitates early airway support. Use of the flexible endoscope may provide a diagnosis, but atraumatic

technique is essential, since complete airway obstruction may develop from laryngeal edema or hemorrhage into the cyst. The cyst usually appears as a large, blue, fluid-filled cyst occupying one or the other of the aryepiglottic folds with partial obstruction of the pyriform sinus. A lateral neck radiograph demonstrates a large, smooth supraglottic swelling.

There is no consensus regarding the origin of these cysts, and some feel that they may be of branchial origin. Davidson (1943) postulated that there was a failure of the epithelial cord connecting the ventricle to the laryngeal lumen to become hollowed out, which resulted in the formation of a cyst in the 3-month-old embryo. Others have stated that a laryngeal cyst is caused by the pinching off of some of the cells that normally form the appendix of the laryngeal ventricle. Holinger and Steinmann (1947), in contrast, state that congenital laryngeal cysts are simply secondary disturbances in development and are not true malformations of the larynx.

If a child develops distress immediately after birth, airway support is essential. Endotracheal intubation or tracheotomy may be necessary, but needle aspiration or incision and drainage of the cyst may allow satisfactory ventilation. Decompression of the cyst may provide satisfactory emergency therapy, but it is usually insufficient for definitive treatment. Removal of the cyst roof and lining of the cyst is required. Endoscopic excision of the cyst and its lining, using the CO_2 laser, is an effective method for definitive treatment of this problem. In refractory cases, a tracheotomy may be necessary and an external approach used after some months of growth (Richardson and Cotton, 1984; Abramson and Zielinski, 1984).

Tracheal Anomalies

Another potential cause of stridor in the newborn is the tracheal abnormality. Though a less common than laryngeal anomalies, these lesions should be sought in any child with unexplained stridor. Tracheal anomalies usually present with stridor during the expiratory phase of respiration, though there may an inspiratory component, especially if the tracheal abnormality is a fixed intrinsic lesion. In addition to the airway symptomatology, lesions within the trachea may be associated with difficulties in feeding, since a bolus of food may produce external compression of the posterior wall of the trachea (Richardson and Cotton, 1984; Myer and Cotton, 1988).

Tracheomalacia

Tracheomalacia is a term that, unfortunately, is often ascribed to any airway noise in a neonate. In fact, tracheomalacia is a specific lesion that produces expiratory stridor of a wheezing nature secondary to collapse of the trachea on expiration. Though the anatomic defect is not certain, the tracheal rings are often of an abnormal shape and consistency, which permits the accumulation of secretions in the distal portion of the neonatal airway because of tracheal collapse.

The voice is generally normal in these patients, and they usually have a normal inspiratory phase of respiration. A barking cough is characteristically seen in patients with this abnormality. They may have some feeding difficulties, but these are generally mild. In some patients, there is an associated tracheoesophageal fistula. In patients with fistula, there may be isolated areas of tracheomalacia following surgical repair. Radiographic examination may

reveal narrowing of the trachea, but this must be differentiated from extrinsic compression caused by mediastinal masses or vascular abnormalities. Fluoroscopic examination may demonstrate collapse of the trachea on expiration because of its ability to demonstrate dynamic movement of the trachea.

In a manner similar to that in patients with laryngomalacia, tracheomalacia is usually a self-limited process in which affected children improve spontaneously by approximately 2 years of age. In some situations, continuous positive airway pressure is necessary until the child improves. A tracheotomy is rarely necessary for this process (Richardson and Cotton, 1984).

Tracheal Stenosis

Tracheal stenosis is a congenital abnormality associated with both inspiratory and expiratory stridor, with the expiratory phase being more pronounced. Patients may be asymptomatic when the tracheal stenosis is mild, but when it is severe, patients have severe respiratory insufficiency that progressively worsens as the child grows and the respiratory demands increase. Radiographic examination may demonstrate tracheal stenosis. It is frequently difficult to identify, however, when the stenosis is mild, and radiographic diagnosis may be simpler when there is an isolated area of tracheal stenosis. In addition to the stenosis, the radiographs may identify other congenital airway abnormalities.

Patients with tracheal stenosis usually have increased secretions distal to the stenosis and present with respiratory distress that is worsened by agitation. These episodes are often confused with croup or inflammatory disease of the trachea because of the increased secretions and the infiltrates that are frequently seen on the radiographs. Racemic epinephrine may provide temporary relief for these patients, but improvement is usually transient and definitive therapy is necessary.

The definitive diagnosis must be made on endoscopy. Depending on the severity of the stenosis, surgical repair may include dilatation, segmental excision of the stenotic area, or tracheoplasty (Richardson and Cotton, 1984). Tracheoplasty may involve anterior and posterior division of the trachea from the region of the cricoid to the carina, with the patient on cardiopulmonary bypass during the operative procedure. Though a tracheotomy may benefit some patients, in those with severe distal lesions, tracheotomy may not be possible. The experience of surgical repair of extensive cases of tracheal stenosis is not great, and current techniques are undergoing an evolutionary process. Improved survival with decreased morbidity will occur as experience increases.

Innominate Artery Compression

There has been a great deal of controversy regarding innominate artery compression of the trachea since Gross and Neuhauser (1948) first reported this entity. Though no one has doubted the existence of this vascular malformation, there have been questions raised about the symptoms produced by such a lesion and the necessity of surgical intervention. Fearon and Shortreed (1963) initially documented endoscopically tracheal compression secondary to a congenital vascular anomaly. They also coined the term *reflex apnea,* which they defined as a reflex respiratory arrest initiated by irritation of the area of tracheal compression either

by a bolus of food or mechanical stimulation. Subsequent to this, Mustard and associates (1969) reported 285 patients, ranging in age from newborn to 3 years, with innominate artery compression of the trachea. Approximately 86 per cent of these patients were managed conservatively, whereas the remainder had surgery to relieve symptoms of reflex apnea or recurrent pulmonary infections. In all cases, surgical therapy involved suspension of the innominate artery to the sternum. Moes and co-workers (1975) updated the experience from the Hospital for Sick Children in Toronto with a report of 60 surgically corrected patients. In this series, arteriopexy was successful in 54 children, whereas 6 children required division of the innominate artery. Strife and colleagues (1981) evaluated the normal radiographic anatomy of the innominate artery in children. They found that the innominate artery normally originates either completely or partially to the left of the trachea and normally crosses in front of it. Welz and co-workers (1984) reported a series of 16 patients from West Germany with innominate artery compression of the trachea. Their study confirmed the previous indications for surgical intervention and also stated that the decision for surgery should never be based on the endoscopic degree of tracheal narrowing alone. Wenig and Abramson (1984) reported a small series of patients with innominate artery compression and introduced the use of tidal breathing flow-volume loops as a diagnostic technique to evaluate this problem.

Patients with innominate artery compression of the trachea commonly present with expiratory stridor and recurrent cough. They frequently have a history of recurrent bronchopulmonary infections as well. Less frequently, parents describe periods of apnea. Several mechanisms have been related to the production of symptoms, including tracheal compression, ciliary immobility, and the inability to clear secretions, which may have increased viscosity (Mustard et al, 1969; Welz et al, 1984; Wenig and Abramson, 1984).

Since it has been shown that the innominate artery normally crosses the anterior portion of the trachea questions arise regarding the true cause of the syndrome. Several theories have been postulated, including thymic pressure, crowding of the anterior portion of the mediastinum and a short, taut innominate artery (Moes et al, 1975).

The evaluation of patients with innominate artery compression of the trachea is straightforward. All patients with stridor that is unexplained by plain radiographs of the neck and chest and use of the flexible laryngoscope should undergo diagnostic microlaryngoscopy and bronchoscopy. Many radiographic studies have been described for the evaluation of this problem, including the use of plain films, fluoroscopy, tracheography, contrast esophagram, angiography, digital vascular imaging (DVI), and CT scanning. At the present time, MRI appears to be the most helpful. Lastly, the use of pulmonary function testing with measurements obtained both before and after exercise can demonstrate quite graphically the degree of symptomatology produced by innominate artery compression of the trachea.

In patients who are mildly symptomatic, conservative medical therapy is indicated. This includes the aggressive management of upper and lower respiratory tract infections that might compromise an already narrowed airway. Supplemental oxygen may be necessary in some patients for a brief time. Surgical intervention is necessary in a selected group of individuals. The absolute criteria for surgical therapy include reflex apnea, failure of medical management after 48 hours (intubation, administration of steroids and antimicrobial agents) and repeated episodes of tracheobronchitis or bronchopneumonia, or both. Relative criteria for surgical intervention include a previously repaired tracheoesophageal fistula with repeated

episodes of respiratory distress, concomitant subglottic stenosis, asthma, cystic fibrosis, tracheomalacia or laryngomalacia, and exercise intolerance (Mustard et al, 1969; Welz et al, 1984).

The standard therapy for innominate artery compression of the trachea has been suspension of the innominate artery from the sternum. In a few individuals, ligation and division of the innominate artery has been performed (Mustard et al, 1969). In other selected patients, reimplantation of the innominate artery has been performed. In this last procedure, the artery is moved proximally on the aorta and to the right side so that it does not cross the trachea.

Other Vascular Anomalies

A double aortic arch results from the persistence of both fourth branchial arch vessels. Patients present with stridor, dysphagia, and varying degrees of respiratory distress. There may be episodes of aspiration pneumonia, and reflex apnea has been described. A chest radiograph will demonstrate a wide base of the heart on the anteroposterior view, whereas the lateral film demonstrates a narrow trachea with displacement at the level of the aortic arch. A barium esophagram is frequently diagnostic and reveals bilateral constriction with posterior indentation of the esophagus. If endoscopy is performed, bilateral tracheal compression will be seen. Both areas of compression are pulsatile, but there is no pulse reduction with compression. Angiography will demonstrate the lesion quite well but is usually unnecessary. At the present time, MRI is frequently used as an alternative to angiography when the diagnosis is in question. Therapy consists of ligation and division of the smaller arch, usually the left one.

Another uncommon anomaly is a right aortic arch with an aberrant left subclavian artery and left ductus or ligamentum arteriosum. The developmental defect responsible for this anomaly is persistence of the right fourth branchial arch becoming the aorta. In a manner similar to that of a patient with a double aortic arch, patients present with stridor, respiratory distress, and dysphagia. An anteroposterior chest radiograph will demonstrate tracheal deviation to the left secondary to the right aortic arch. A barium swallow demonstrates bilateral constriction with posterior indentation of the esophagus, which is greater on the right than on the left. The endoscopic examination is very similar to that seen with a double aortic arch, though the right tracheal compression is usually pulsatile. Therapy consists of ligation of the ligamentum arteriosum or the left ductus arteriosum.

A pulmonary sling results from the anomalous origin of the left pulmonary artery from the right pulmonary artery. Patients present with expiratory stridor, respiratory distress, and wheezing. An anteroposterior chest radiograph demonstrates the left hilum to be positioned lower than normal, with emphysema or atelectasis on the right side. The lateral film demonstrates anterior bowing of the right mainstem bronchus and trachea. A barium esophagram will show anterior indentation above the carina between the trachea and the esophagus on the lateral view. The anteroposterior view is normal. Endoscopic examination demonstrates tracheal displacement to the left and compression of the proximal right mainstem bronchus. Either angiography or MRI will be diagnostic. Therapy consists of detachment of the left pulmonary artery and reanastomosis to the main pulmonary artery anterior to the trachea.

The anomalous right subclavian artery arises from the dorsal aspect of the aorta and produces dysphagia without airway symptoms. A chest radiograph is normal, but a barium esophagram demonstrates posterior indentation of the esophagus. Microlaryngoscopy and bronchoscopy are normal, but esophagoscopy demonstrates external posterior compression of the esophagus. Compression of the indentation leads to a decrease in the right radial pulse without a change in the temporal pulse. Angiography is unnecessary in these cases, and no therapy is generally necessary (Myer and Cotton, 1988; Park, 1981).

Tracheoesophageal Fistula

Patients with tracheoesophageal fistulas arouse suspicion in the clinician from birth because of their significant feeding problems and cyanosis. Other symptoms include recurrent pneumonia, choking or coughing while feeding, and retention of secretions, frequently secondary to isolated tracheomalacia. Several different types of fistulas are seen.

In the most common type, which occurs in approximately 87 per cent of cases, the dilated upper esophagus ends in a blind pouch, and the lower esophageal segment is attached to the trachea. In approximately 8 per cent of cases, there are blind upper and lower esophageal segments without a true fistula to the trachea. In those patients, airway distress is still a problem because of aspiration. Less commonly, one finds an "H" deformity in which there is a true fistula without evidence of esophageal atresia. In a very small number of patients, the upper esophageal segment opens directly into the trachea, or the upper and lower esophageal segments open independently into the trachea.

A barium esophagram is generally diagnostic, but small lesions may not be demonstrated. A chest radiograph may show right upper lobe pneumonia secondary to aspiration. An abdominal film may demonstrate marked air filling. An endoscopic examination may reveal the connection between the trachea and the esophagus in some cases. Open surgical repair with the insertion of tissue between the trachea and esophagus is essential to effect permanent separation (Pillsbury and Donovan, 1987).

Extrinsic Compression - Miscellaneous

In addition to the vascular anomalies previously described, extrinsic compression of the trachea secondary to mass lesions constitutes a large group of patients who present with expiratory stridor. Compression may come from the thymus or thyroid gland, mediastinal tumors, bronchogenic cysts, or cystic hygromas. Symptoms are variable depending on the type of compression and the actual site (Myer and Cotton, 1988; Richardson and Cotton, 1984).

Endoscopic examination will demonstrate the tracheal compression, but radiographic examination, employing the CT scan and MRI, is necessary to arrive at the exact diagnosis. Surgical repair is dependent on the causative factor.