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Introduction

Congenital abnormalities of the larynx present the otolaryngologist with a wide range of problems ranging from mild to severe. Such anomalies may present at the moment of birth with symptoms requiring immediate attention or may be more insidious in presentation. Thus, it is essential that all otolaryngologists treating children be aware of and have a firm understanding of these conditions which arise from aberrations in embryologic development.

Normal Anatomy

The larynx is a complex, evolutionary structure that permits the trachea to be joined to the pharynx as a common aerodigestive pathway. The larynx is essential in several important functions: 1) ventilation of the lungs, 2) protection of the lungs during swallowing, 3) clearance of secretions by cough, and 4) production of sound. The survival of the infant is predicated on the structural and neurologic integrity of the larynx and, if altered, prompt diagnostic and surgical intervention may be necessary.

The complex structures of the human airway vary in anatomy and physiology from birth to adulthood. The infant larynx and trachea are significantly smaller than that of an adult. At birth, the infant larynx is approximately one third the size of an adult. The glottis of the neonate measures approximately 7 mm in the sagittal plane and 4 mm in the coronal plane. The vocal cords of the newborn infant are 6-8 mm long and the vocal processes of the arytenoids extend one half of that length. In fact, the vocal process of the arytenoid takes up half the length of the vocal cord in the infant larynx, while it only takes up about ¼ of the length of the vocal cord in the adult. The posterior glottis' transverse length is approximately 4 mm. The subglottis has a diameter of between 4.5 and 5.5 mm. These dimensions leave little margin for obstruction in the infant, unlike the adult. The narrowest portion of the airway in the older child and adult is the glottic aperture, while the narrowest part of the airway in the infant is the subglottis. A diameter

of 4.0 mm is considered the lower limit of normal in a full term infant and 3.5 mm in a premature infant. Indeed, an infant with one millimeter of glottic edema will experience a 35% obstruction of the airway. In the subglottis, one millimeter of circumferential edema leads to over 60% narrowing.

The infant larynx is positioned higher in the neck than the adult larynx. The superior border of the larynx of the infant is located at about the level of the first cervical vertebrae with the cricoid positioned at about the fourth cervical vertebrae. In comparison, the adult cricoid rests about the level of the sixth cervical vertebrae. The structures of the infant larynx are more pliable and less fibrous making the infant airway more susceptible to narrowing from edema and less easily palpable.

The epiglottis is proportionally narrower than that of an adult and assumes either a tubular form or the shape of the Greek letter omega.

Embryology

Richter recorded the identification of specific congenital laryngeal anomalies as early as 1792. In 1885, His described the appearance of the respiratory primordium from an outpouching of the cephalic portion of the developing pharynx by the third week of gestation. Indeed, the respiratory system is an outgrowth of the primitive pharynx.   The development of the lower respiratory system begins at 26 days after conception as the laryngotracheal groove (also known as the respiratory primordium) at the ventral aspect of the foregut.

The laryngotracheal diverticulum becomes separated from the foregut by the tracheoesophageal folds, which fuse to become the tracheoesophageal septum. This septum divides the foregut into a ventral laryngotracheal tube and a dorsal esophagus. Failure of the tracheoesophageal folds to fuse during the fourth and fifth weeks can lead to a tracheoesophageal fistula.

The larynx develops from the fourth and fifth branchial arches. The laryngotracheal opening lies between these two arches. This primitive laryngeal aditus is altered to become a T-shaped opening by the growth of three tissue masses. One is the hypobranchial eminence. This mesodermal structure eventually becomes the epiglottis. The second and third growths are two arytenoid masses. As these masses grow between the fifth and seventh weeks, the laryngeal lumen is obliterated. Recanalization occurs by the tenth week. Failure to recanalize may result in atresia, stenosis or web formation in the larynx. The arytenoid masses are separated by an interarytenoid notch, which eventually becomes obliterated. If obliteration does not occur, a posterior laryngeal cleft can result leading to severe aspiration in the newborn.

Clinical Manifestations and Diagnosis

The clinical manifestations associated with congenital anomalies of the larynx include: 1) respiratory obstruction, 2) stridor, 3) a weakened or abnormal cry, 4) dyspnea, 5) tachypnea, 6) aspiration, or 7) episodes of cyanosis, or 8) sudden death. The clinical presentation of each lesion varies from the site being in the supraglottis, glottis, or subglottis. Although some

congenital laryngeal lesions will 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. The varied presentations of particular lesions will be discussed by region.

A child with a suspected congenital laryngeal lesion should undergo a complete history and physical exam. In the infant or child, a thorough history should be obtained which includes: history of prematurity and associated medical problems and intubation records. A premature infant who has been intubated for variable periods of time may develop acquired lesions such as subglottis stenosis, subglottic cysts, or intubation granulomas. Important characteristics of the intubation include the date of first intubation, duration, size of the endotracheal tube, number of intubations, and if any intubations were traumatic. The birth record should be reviewed to assess for any birth trauma. A history of noisy breathing and difficulty feeding should lead to suspicion of airway problems. Growth curves should be reviewed and followed to determine if the child has failure to thrive. Particularly the relationship of airway symptoms to feeding is important to elicit in the history. Suspected foreign body aspirations should be elicited and the details of changes in the baby's cry should be discussed.

A complete head and neck exam should be performed on all patients, if possible. It begins with observing the patient for any apparent airway symptoms, which may include irritability and restlessness in an infant or dyspnea, tachypnea, cyanosis, and stridor in the infant. Voice quality in a child and crying quality in an infant should be evaluated for weakness, hoarseness, breathiness, or complete absence. Flexible fiberoptic nasopharyngolaryngoscopy may be performed at the bedside or in clinic on infants and some cooperative children if the patient is stable. Vocal cord immobility, reflux changes, immediate subglottic abnormalities, supraglottic sensation, and other glottic and supraglottic abnormalities may be detected.

Radiographic evaluation includes AP and lateral views of the neck and chest. Such films are particularly useful to help rule out any stenotic lesions of the airway. A narrowed subglottic air column suggests a diagnosis of subglottic stenosis. If inspiratory and expiratory films are difficult to obtain, airway fluoroscopy can be beneficial to view the dynamic properties of the trachea. A barium swallow can be used in cases of swallowing difficulty to delineate conditions such as posterior laryngeal clefts, tracheoesophageal fistulae, or vascular rings which may compress the trachea. The use of computerized tomography or MRI has not been proven to be beneficial in assessing the majority of congenital laryngeal etiologies, but may be useful in select cases.

If the diagnosis remains uncertain, the gold standard for diagnosis of any congenital laryngeal abnormality remains direct laryngoscopy and tracheobronchoscopy under general anesthesia. This should be performed in the operating room with an experienced anesthesiologist. It is imperative to have all of your equipment arranged and checked prior to the patient coming into the operating room. Be sure to have a wide range of bronchoscope sizes in case the airway is much smaller than you anticipated! The potential need for tracheotomy should be discussed with the patient's family prior to endoscopy. A rigid bronchoscope or a rod lens

telescope may be used to assess the airway. The important things to document during endoscopy are as follows: (1) the outer diameter of the largest bronchoscope or endotracheal tube that can be passed through a stenotic segment, if present, (2) the location/subsites (glottis, subglottis, trachea) and length of the stenosis, if present, (3) other separate sites of stenosis, (4) other airway anomalies in infants (clefts, webs, cricoarytenoid joint fixation, neoplasms, etc.), and (5) reflux changes.

Supraglottic Anomalies

Laryngomalacia

Laryngomalacia is the most common congenital laryngeal anomaly. This entity accounts for approximately 60 percent of laryngeal problems in the newborn. Boys are affected twice as often as girls. It is usually a self-limiting condition, but when severe may produce life-threatening obstructive apnea, cor pulmonale, and failure to thrive. Fatal outcomes have been described. Severe cases may require intubation or tracheotomy to secure the airway. In a series of 56 infants wit congenital airway anomalies requiring tracheotomy, 21 cases had an underlying laryngomalacia.

This condition arises from a continued immaturity of the larynx, as if the fetal stage of laryngeal development has persisted. The abnormality appears to be flaccidity or incoordination of the supralaryngeal cartilages, especially the arytenoids that is expressed when the infant is stressed by excitation with an increased respiratory rate. Stridor is typically noted in the first few weeks of life and is characterized by fluttering, high-pitched inspiratory sounds. The supraglottic structures are pulled into the lumen around a vertical axis with inspiration. The epiglottis is commonly omega shaped and the aryepiglottic folds are short. Eating difficulties and respiratory distress are rare. Sternal retractions are seen frequently with labored respiratory effort.

The diagnosis can only be made by clinical observation of the larynx during respiration. Inspection of the remainder of the respiratory tract is sometimes necessary to rule out associated secondary lesions such as innominate artery compression of the trachea or subglottic stenosis. Radiologic assessment can sometimes be a helpful adjunct if it captures the characteristic medial and inferior displacement of the arytenoid cartilage or epiglottis.

Therapy consists of confirming the diagnosis by flexible laryngoscopy and reassuring the parents that the prognosis for the child is favorable. Position changes of the infant may help alleviate the stridor as it typically worsens in the supine position. Continued vigilance by the pediatrician to be certain that the child continues to grow, feed, and breath well is important.

In the past, tracheotomy was the surgical procedure of choice for severe cases. Variot was the first to describe incising the aryepiglottic folds in severe cases of laryngomalacia. However, the first effective surgical procedure was reported in 1922, when Iglauer performed a partial epiglotticetomy. Lane was the first to publish supraglottic trimming for the treatment of laryngomalacia in 1984. A year later, Seid described using the laser to divide the AE folds.

Several series since have popularized this supraglottoplasty technique with some investigators advocating the use of associated epiglottiopexy in selected cases.

Supraglottoplasty has proven successful for the correction of supraglottic obstruction and is now the surgical procedure of choice. Direct laryngoscopy and bronchoscopy should be done prior to the supraglottoplasty to ensure that no other concomitant pathology is present. The precise mechanism of obstruction should be confirmed with flexible laryngoscopy and surgical maneuvers should be directed at those sites using sharp dissection or the CO_2 laser. It is important to be conservative to prevent supraglottis stenosis. Unilateral supraglottoplasty should be considered and the second side operated on only if symptoms continue. Polonovski describes a suction test where an aspiration cannula is introduced in the supraglottic inlet producing negative pressure, thereby reproducing the areas of collapse. The test is repeated after surgical excision to confirm improvement.

Results of supraglottoplasty have been impressive. Roger published the largest series comprising 115 cases. Complete regression of symptoms occurred in 53% of cases. Stridor or effort dyspnea prevailed in 36%. Seven children improved after a second procedure and 2 required eventual tracheotomy. Smaller studies have documented success rates from 77-100%. Failures may be the result of concomitant airway anomalies including pharyngomalacia. Tracheotomy may be necessary in these in these cases, however some may respond to BiPAP, obviating the need for tracheotomy.

Complications are rare and include supraglottic stenosis and swallowing problems such as aspiration. Hemorrhage, granuloma formation, posterior glottic stenosis, and cricoarytenoid joint fixation has been described.

The relationship of laryngomalacia and GERD has been described but a direct causal relationship has not been proven. Belmont found an 80% radiological prevalence of GERD in infants with LM. Phelan suggested that GERD might be a result of the high intraesophageal pressures generated. However, GERD may exacerbate LM and some authors recommend treating this condition before eventual surgery. Interestingly, however, Polonovski did not find improved respiratory symptoms with antireflux therapy.

Supraglottic Webs

Congenital webs are diaphragmatic growths of differing thickness that partially occlude the supraglottic lumen. Supraglottic webs represent less than 2 percent of congenital laryngeal webs with symptoms depending on the size and position of the web. Symptoms can include voice changes and dyspnea. Ten percent of children have other associated anomalies.

Treatment consists of surgical lysis using either the laser or sharp instrumentation, followed by dilatation. Tracheotomy should be considered if the web is large and supraglottic swelling is anticipated postoperatively.

Bifid Epiglottis

Congenital bifid epiglottis is a rare congenital laryngeal anomaly that may present with similar symptoms as laryngomalacia with inspiratory stridor and airway obstruction. The patient's history also frequently includes episodes of cyanosis associated with feeding and occasional airway obstruction. The midline split in the epiglottis often renders the epiglottis incompetent in protecting the airway during feeding and is often drawn into the airway during inspiration. Bifid epiglottis is associated with other congenital syndromes including Pallister-Hall syndrome and polydactaly (40% of cases). Patients should undergo an endocrine evaluation because of the possible associations with hypothyroidism and hypothalamic abnormalities. Surgical management of the airway with tracheotomy may be necessary in severe cases.

Saccular Cysts

Congenital saccular cysts are unusual laryngeal anomalies that are similar in their embryologic development to laryngoceles. These lesions arise from the vestigial laryngeal structure known as the saccule. Whereas the laryngocele is filled with air and is connected to the airway, a saccular cyst is fluid filled. These cysts typically do not connect to the internal laryngeal lumen.

Desanto has classified laryngeal cysts as superior cysts (extend medially and posteriorly in the region of the ventricle) or posterior cysts (extend into the region of the false cord and AE fold). In cases of severe airway obstruction at birth, immediate intervention is warranted with either tracheotomy or intubation. Surgical management of saccular cysts can be performed endoscopically using CO2 laser and with sharp microlaryngeal instruments to marsupialize the cyst. Open surgical resection with an external laryngofissure has also been described. A high rate of recurrence is reported in several reports using endoscopic techniques and many authors stress the importance of removing the entire cyst lining.

Laryngocele

The laryngocele is a sac like structure with an internal lumen that is dilated and filled with air. It represents a dilation of the ventricular sinus of Morgagni beyond the confines of the laryngeal cartilage. They are classified as internal if they remain within the laryngeal cartilage or external if they extend through the thyrohyoid membrane. Combined lesions may also occur. Fortunately, these are rare in infants and can cause intermittent hoarseness and dyspnea that increases with crying. Diagnosis may be difficult as these can contract and may not be visualized under anesthesia. A CT may be valuable in these instances. Endoscopic and open procedures have been advocated depending on the size and location of the laryngocele.

Lymphatic and Vascular Malformations

Hemangiomas are a type of hamartoma that results in an anomalous development of blood vessels in a particular region. Hemangiomas of the supraglottic structures are rare. They can present with dyspnea, stridor, or feeding difficulties. As with hemangiomas of the subglottis, up to 30% are present at birth and most grow over the first 6-18 months. Most children, however, present in the first six months of life. Evaluation consists of careful endoscopic

examination with possible CO2 laser excision, observation, tracheotomy, or open excision. As with other sites, gradual regression is expected during the first five years of life.

Lymphatic malformations of the supraglottis are cystic malformations that result from abnormal development of lymphatic vessels. Lymphangiomas of the valleculla may compress the epiglottis and cause airway distress. Symptoms may include bleeding, changes in speech, dyspnea, and dysphagia. These lesions do not regress with age and treatment typically consists of careful endoscopic confirmation followed by reduction of the lesion with the CO2 laser or YAG laser. Complete excision of these lesions is often compromised by surrounding vital structures that should not be sacrificed.

Anomalous Cuneiform Cartilages

This embryonic abnormality is the result of a malformation of the lateral masses, which are derived from a portion of the sixth branchial arch. Symptoms mimic laryngomalacia with stridor, airway distress, and dyspnea on exertion. Associated anomalies including ankyloglossia and macroglossia may occur. Treatment should be tailored to symptoms. Should severe airway obstruction be present, tracheotomy and possible supraglottic laryngectomy may be necessary.

Glottic Anomalies

Vocal Cord Paralysis

Vocal fold paralysis has long been recognized as a significant cause of stridor and hoarseness in infants and children. It is the second most common cause of stridor in the newborn behind laryngomalacia. The frequency of vocal cord paralysis varies. Narcy quotes it as representing 23% of congenital laryngeal pathology, whereas Holinger and Fearon quote lower figures of 10% and 6.5% respectively. Some authors report unilateral paralysis to be more common and others report bilateral paralysis to be more frequent. Laryngeal paralysis may be present at birth or may manifest itself in the first month or two of life. The neurologic impairment reflects an injury to the vagus nerve. The lesion can occur anywhere from the brain through the neck into the chest and into the larynx. Many paralyses are idiopathic (cause unknown) in up to 47% of cases, however the most common causative factors include entities such as Arnold Chiari malformations, hydrocephalus, neonatal hypotonia, and multiple peripheral paralysis (myasthenia gravis). Other causes include birth trauma and cardiac anomalies. Associated laryngeal lesions such as clefts and stenosis are also commonly often found.

Any or all of the normal laryngeal functions (voice, respiration, deglutition) may be abnormal in the pediatric patient with laryngeal paralysis. The most common symptom is stridor. Ineffective cough, aspiration, recurrent pneumonia, and feeding difficulties are also commonly reported. Consistent stridor, cyanosis, and apnea are frequent. Voice and cry, however, may be normal particularly in cases of bilateral vocal cord paralysis. Hoarseness and dysphonia are common in cases of unilateral vocal fold paralysis.

The initial concern in any child with suspected laryngeal paralysis is airway stability. Extensive diagnostic evaluations are deferred until the airway is stabilized and secured. This is

best achieved under the controlled setting of an operating room with appropriate airway instrumentation available. In those instances in which respiratory distress is not a primary concern, a thorough and unhurried evaluation is warranted. A careful history and physical exam is necessary. A complete past medical history including a difficult delivery, prior surgical procedures, or other congenital anomalies can provide important clues to this diagnosis.

Various methods have been used to document laryngeal paralysis. Neck films, fluoroscopy, and ultrasound have all been evaluated and are unsatisfactory in diagnosing this condition for a number of varied reasons. Chest films however are useful in diagnosing associated cardiac or pulmonary anomalies, both of which are common in cases of unilateral paralysis. Flexible laryngoscopy in the awake patient, however, is the gold standard for the diagnosis of this condition. Once the diagnosis is made, a search for the underlying cause is warranted. The entire course of the vagus nerves should be imaged. This should include CT or MR imaging. A barium swallow can provide evidence of subtle neurologic abnormalities, abnormal sensation to the larynx, and can document associated mediastinal anomalies such as vascular rings. While flexible laryngoscopy is invaluable in this diagnosis, rigid laryngoscopy and bronchoscopy is important for the identification of associated anomalies or when the cause remains unknown after a noninvasive workup has been complete. Paralysis must be differentiated from cricoarytenoid joint fixation or posterior glottic stenosis, both which can lead to a similar presentation.

Management strategies depend on the child's underlying condition. Children with bilateral vocal fold paralysis frequently require surgical intervention. The airway is often markedly compromised and in over 50% of cases a tracheotomy is required. In cases of mild airway symptomatology with bilateral vocal fold paralysis, expectant close follow up is possible. In cases of Arnold Chiari malformation and bilateral vocal fold paralysis, many authors recommend that prior to an invasive airway procedure (tracheotomy), a VP shunt or posterior fossa decompression be performed. Some authors contend that in these cases, nasotracheal intubation for four weeks should be considered, as the potential for vocal cord return is good. Others recommend immediate tracheotomy to secure the airway prior to further procedures and testing. Once a tracheotomy is performed, serial endoscopy is performed to detect a return of function. Most authors recommend waiting at least one year prior to any irreversible lateralization procedures. EMG may provide prognostic information during this time frame. Spontaneous resolution of vocal cord paralysis is thought it occur in 48-64% of cases, with rapid improvement in many patients after correction of their hydrocephalus and ACM. Multiple lateralization procedures have been proposed for those patients who do not resolve. Surgical widening of the glottis must balance voice and airway patency issues. Older techniques such as the Woodman procedure have been abandoned. CO2 laser cordotomy and open arytenoidectomy, artenoidpexy, arytenoid separation with cartilage grafting or laser arytenoidectomy and cordectomy have been proposed as more efficacious alternatives. Decannulation rates of over 60% can be expected. Reanimation of the larynx is another form of vocal cord rehabilitation for patients with bilateral vocal cord paralysis. Phrenic to recurrent laryngeal nerve anastomosis, phrenic to PCA muscle, and omohyoid nerve muscle pedicles have all been utilized. Success rates in the pediatric population are reported to be 50%. Electrical stimulators are also being explored.

The management of unilateral vocal cord paralysis in children is usually less urgent than that of bilateral paralysis. Children adjust well to persistent unilateral vocal cord paralysis with few sequelae. A weakened cry may result but an adequate airway is the typically maintained. Lateral augmentation procedures and thyroplasty techniques are not recommended in children, as speech therapy is the mainstay in this population. Thyroplasty may play a role in older children that fail conservative measures with significant dysphonia.

Laryngeal Webs

Laryngeal webs form when there is a failure of recanalization of the larynx during embryonic development, and if it persists at birth, may cause respiratory distress. Seventy-five percent of webs occur at the level of the glottis as a membrane of differing thickness that partially occludes the lumen. The web is generally located anteriorly with a concave posterior glottic opening. Most webs are thick and fibrous with a subglottic extension.

The presentation of laryngeal webs varies with the severity and type of the web. Type I laryngeal webs involve 35% or less of the glottis. The true vocal cords are visible through the web and there is little or no subglottic extension. Symptoms include a mildly abnormal cry with some hoarseness. Respiratory distress is usually not a feature. Type II webs are anterior webs involving 35-50% of the glottis. Subglottis involvement stems more from thick anterior webbing than from cricoid abnormalities. Airway symptoms are uncommon except during infection or after intubation trauma. The voice is typically weak. Type III webs involve 50-75% of the glottis. The web is thick anteriorly and the true vocal cords may not be visualized. There may be associated cricoid anomalies. Airway symptoms are often severe and marked vocal dysfunction may occur. Type IV webs occlude 75-90% or more of the glottis. It is uniformly thick and the true vocal cord is not identifiable. The patient is aphonic and immediate airway management is required at birth.

The diagnosis of this condition is usually clear on flexible laryngoscopy, however airway films may aid in the diagnosis if subglottic or cricoid pathology is also present. Treatment of this anomaly is dependent of the type of web and symptomatology. Thin membranous type I webs which produce only minimal symptoms can be observed until age 3-4 years and then divided with either the CO2 laser or cold knife. Mitomycin-C may also be applied. Some authors employ local flaps to prevent recurrence. Type II webs can be managed by incising the web along one vocal cord and then proceeding with staged dilations or by incising the web on the opposite cord two weeks later. Keel placement through an open or endoscopic approach may be necessary and a tracheotomy is generally advised if a keel is placed. Treatment of type III and IV webs is usually delayed until the child is 3-4 years old. Most infants will have a tracheotomy in place for airway control until definitive surgery is undertaken. The standard treatment for these conditions entails a tracheotomy, laryngotomy, and keel insertion. An alternative treatment is an early single stage laryngotracheal reconstruction with submucosal resection of the abnormal cricoid, mucosal flap elevation, and rotation to the anteromedial aspect of the TVC's. A tracheotomy may be avoided with this approach.

Posterior glottic webbing is rare but usually consists of a thin membranous sheet between the posterior TVC's. Minor webs may respond to simple division and dilation, however

interarytenoid webs with significant posterior glottic stenosis may require a laryngofissure, a posteriorly placed costal cartilage graft, and stenting.

Laryngeal Atresia

Laryngeal atresia is a rare congenital anomaly that is incompatible with life unless emergency measures are undertaken at birth. This condition represents the most severe end of a spectrum of diseases arising from failed recanalization of the larynx during embryogenesis. These infants only survive if there is an associated tracheoesophageal fistula or if a tracheotomy is performed immediately after birth. If a tracheoesophageal fistula is present, intubation of the esophagus may allow ventilation while airway access is being obtained. Repair of the atresia requires a formal laryngotracheal reconstruction at a later stage. This disorder is usually accompanied by a host of another anomalies including tracheoesophageal fistulae, esophageal atresia, urinary tract anomalies, and limb defects.

Congenital High Upper Airway Obstruction (CHAOS)

Congenital high airway obstruction (CHAOS) was defined by Hedrick in 1994 as upper airway obstruction that is diagnosed in utero by ultrasound, with concomitant findings of large echogenic lungs, flattened diaphragms, dilated airways distal to the obstruction, and fetal ascites or hydrops. There have been several reports of survival in infants with CHAOS who undergo the EXIT procedure (ex utero intrapartum treatment). The EXIT procedure was originally designed to treat children with large cervical masses that where diagnosed in utero and caused airway obstruction. CHAOS may not be diagnosed if a TEF is present as the fetal lung fluid is able to pass from the pulmonary system.

The EXIT procedure requires a multidisciplinary team approach. A cesarean section is performed with partial deliverance of the fetus while maintaining fetoplacental circulation. Bronchoscopy and tracheotomy can be performed expeditiously with successful outcomes reported.

Subglottic Anomalies

Subglottic Hemangiomas

Subglottic hemangiomas are congenital vascular lesions that present with symptoms ranging from minimal airway obstruction to severe, life threatening respiratory distress. Hemangiomas are present at birth only 30% of the time, with the majority of cases presenting within the first few months of life. The natural history of these lesions is similar for 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 12-18 months. There is a phase where the lesion is stable and then a subsequent period of involution. Most hemangiomas involute completely by five years of age though some may not involute completely. Subglottic hemangiomas have a 2:1 female predominance.

Typically infants with subglottic hemangiomas are asymptomatic for the first few months of life and become symptomatic by the age of 3 months. Almost all are symptomatic by the age

of 6 months. Stridor may initially be inspiratory but quickly becomes biphasic. The cry may be altered and the infant may have a barking cough, hoarseness, croupy symptoms, and occasional hemoptysis. Cutaneous hemangiomas occur in approximately 50% of children with subglottic hemangiomas.   Thus, a child's skin must be thoroughly examined in all cases of infant stridor.

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 the definitive diagnosis, however biopsy is not always necessary. The lesion is typically a compressible symmetric bluish or reddish submucosal mass most often found in the posterior lateral subglottis. Asymmetric subglottic narrowing seen on neck films is almost pathognomic of a subglottic hemangioma as this finding is rarely seen in croup, subglottic cysts, subglottic stenosis, or RRP. MRI or CT with contrast can better delineate the lesion and assess for neck or mediastinal extension.

Numerous management options exist for subglottic hemangiomas. The decision of what therapeutic measure to take is directed at maintaining an airway while minimizing the potential long-term sequelae of the treatment itself. The treatment modalities that have been described include laser ablation using the CO_2 or KTP laser, tracheotomy, external beam radiation, radioactive gold grain implantation, cryotherapy, sclerosing agents, corticosteroid therapy (systemic or intralesional), and open surgical excision. The most common intervention is CO_2 laser ablation. When used conservatively, this is an appropriate treatment modality. There is, however, a risk of subsequent subglottic stenosis cause by overaggressive, circumferential lasering of large lesions.   A recent report documented a 20% rate of subglottic stenosis after this treatment modality. Laser therapy is often coupled with intravenous and injectable corticosteroid therapy. Large lesions may respond solely to high dose intravenous corticosteroid therapy (1-2 mg/kg/day). Long-term side effects from this therapy may occur (growth retardation, cushingnoid appearance, and sepsis). Children with large subglottic hemangiomas causing severe airway obstruction, necessitating tracheotomy, may benefit from an open procedure. This may be performed in a single stage procedure requiring postoperative intubation for 7-10 days. A delayed procedure with delayed tracheotomy decannulation after subglottic healing may also be performed. Many authors feel the standard of care is tracheotomy with observation for involution. This method is the method by which all others should be compared.

Posterior Laryngeal Cleft

The laryngeal cleft arises at approximately 35 days of gestation from failure of rostral development of the tracheoesophageal septum. Failure of the interarytenoid tissue or cricoid cartilage to fuse in the posterior midline will result in a laryngeal cleft.

The incidence is less than 0.1% and the majority of cases are sporadic. There is a strong association with other anomalies such as tracheoesophageal fistulae. It is important to remember that 6% of patients with a TEF will have a concomitant laryngeal cleft producing continued aspirations symptoms. Laryngeal clefts are also seen in Pallister-Hall syndrome and G-syndrome. Respiratory distress is usually precipitated by feeding and is often associated with cyanosis. Voice abnormalities are often present and GERD is a major contributor to overall

pulmonary compromise. Infants with these clefts often have recurrent aspiration, leading to pneumonia and death. The severity of the symptoms depends on the extent of the laryngeal cleft.

Patients suspected of having a laryngeal cleft require radiographic evaluation. Chest radiographs frequently show aspiration pneumonitis that is severe. A barium swallow is the most important diagnostic tool for showing spillover of contrast material into the trachea. Endoscopy is required to make the definitive diagnosis of a laryngeal cleft. Great care must be taken to document the relationship of the cleft to the level of the vocal cords.

Numerous classification systems exist for laryngeal clefts. Benjamin and Inglis describe type I clefts as a supraglottic, interarytenoid clefts. Type II clefts are a partial cricoid cleft. Type III clefts are a complete cricoid cleft with or without extension into the esophagus and type IV cleft are full laryngotrachealesophageal clefts.

Surgical repair must be taken in all cases of symptomatic clefts. Type I clefts can sometimes be managed nonsurgically with speech and feeding therapy aimed towards decreasing aspiration. GERD must be controlled. When conservative measures fail to prevent aspiration, endoscopic or open repair of the cleft may be possible. Type II and III clefts can be approached via an anterior laryngofissure or a lateral pharyngotomy. A tracheotomy and two-layer closure are then performed. Type IV clefts require a lateral pharyngotomy and right thoracotomy or a midline anterior approach with a median sternotomy with the patient on cardiac bypass.

The overall mortality rate of laryngeal clefts is 43%. Type IV clefts have a reported mortality rate of 93%, however the mortality rate is decreasing with advances in surgical technique.

Subglottic Stenosis

Subglottic stenosis (SGS) may be classified as either acquired or congenital. Although congenital subglottic stenosis is uncommon, accounting for 5% of all cases, it is the third most common congenital airway problem (after laryngomalacia and vocal cord paralysis). Congenital SGS is thought to be secondary to failure of the laryngeal lumen to recanalize properly during embryogenesis. SGS is considered congenital if there is no history of endotracheal intubation or other forms of laryngeal trauma.

Congenital SGS is divided histopathologically into membranous and cartilaginous types. Membranous SGS is usually circumferential and consists of fibrous soft-tissue thickening caused by increased fibrous connective tissue or hyperplastic submucous glands. It may involve the vocal folds as well. The cartilaginous type usually results from a thickened or deformed cricoid cartilage that forms an anterior subglottic shelf that extends posteriorly allowing only a small posterior opening. Other malformations can occur such as an elliptical cricoid leaving a slit-like opening or a trapped first tracheal ring. Membranous SGS is usually less severe than the cartilaginous type.

Congenital SGS is often associated with other congenital malformations. A thorough search for associated anomalies is necessary.

Subglottic stenosis is defined as a subglottic lumen 4.0 mm in diameter or less at the level of the cricoid in a full term infant. The normal newborn subglottic diameter is 4.5 – 5.5 mm and in premature neonates around 3.5 mm. A subglottic diameter of less than 3.5 mm in a premature infant is stenotic.

The severity of congenital subglottic stenosis depends on the degree of subglottic narrowing. The symptoms can range from mild with a picture of recurrent croup to severe with respiratory distress at delivery. Children with subglottic stenosis usually present with stridor and/or respiratory distress.   Symptoms include irritability, restlessness, dyspnea, tachypnea and cyanosis. The stridor is typically biphasic (inspiratory and expiratory components) due to turbulent airflow through the partially obstructed airway. Frequently, children with mild congenital SGS have no symptoms until they develop an upper respiratory tract infection or under physical exertion. Any child under age one with recurrent croup should undergo endoscopy to rule out congenital SGS.

The evaluation of SGS includes a complete history and physical examination. The standard for diagnosis is rigid endoscopy under general anesthesia however airway films can demonstrate subglottic narrowing, particularly at multiple levels. These should be taken prior to undergoing evaluation in the operating room.

Historically, classification of subglottic stenosis has been a problem. Measurements were done either subjectively or by using various instruments including rigid bronchoscopes, laryngeal forceps and angioplasty catheters.

Today, there is still no universally accepted staging system for subglottic stenosis. The most commonly used system was developed by Cotton in 1984 then revised in 1989. The percentage of obstruction and anatomic location of the lesion were determined endoscopically and assigned a grade I-IV based on perceived percentage of obstruction. Although this system was successful at relating the severity of the obstruction with the prognosis for decannulation, it remained imprecise and dependent on skilled judgment. For these reasons, Myer, Conner and Cotton proposed a grading system based on endotracheal tube sizes.

The Myer-Cotton staging system is useful for mature, firm, circumferential stenosis confined to the subglottis. It describes the stenosis based on the percent relative reduction in cross-sectional area of the subglottis which is determined by differing sized endotracheal tubes. Four grades of stenosis are described with this system: grade I lesions have less than 50% obstruction, grade II lesions have 51% to 70% obstruction, grade III lesions have 71% to 99% obstruction, and grade IV lesions have no detectable lumen or complete stenosis.

The McCaffrey system classifies laryngotracheal stenosis based on the subsites involved and the length of the stenosis. Four stages are described: stage I lesions are confined to the subglottis or trachea and are less than 1cm long, stage II lesions are isolated to the subglottis and are greater then 1 cm long, stage III are subglottic/tracheal lesions not involving the glottis, and stage IV lesions involve the glottis.

Treatment of congenital SGS is tailored to the symptoms and grade of the stenosis. Symptoms are typically less severe in congenital SGS than in the acquired form. Congenital SGS also improves as the child grows, and less than half of children with this disorder will require a tracheotomy. For those children who do require surgical intervention, several options are available.

Mild stenosis (Cotton-Myer grades I and II) can usually be treated conservatively with observation. In cases that do require surgery, endoscopic techniques such as CO_2 laser resection of a membranous web can be performed. Dilation has nothing to offer in the management of cartilaginous subglottic stenosis. Factors associated with failure of these endoscopic techniques include: previous attempts at endoscopic repair, circumferential scarring, loss of cartilaginous support, exposure of cartilage during laser excision leading to chondritis, severe bacterial infection, posterior inlet scarring with arytenoid fixation, combined laryngeal or tracheal stenosis or vertical scar length >1cm. Endoscopic dilation has had disappointing results and should be abandoned for congenital SGS.

Grade III or IV stenosis may require some form of open surgical procedure, as these typically are the result of a cartilaginous stenosis. Several techniques have been described.

The anterior cricoid split (ACS) procedure was originally described for a neonate who has had multiple failed extubations instead of performing a tracheotomy (Cotton and Seid, 1980). This procedure is also used for older infants and those who are have already been tracheotomized. Indications were later expanded to patients with congenital subglottic stenosis. Strict criteria for ACS have been established by Cotton and include: extubation failure on two occasions or more due to laryngeal pathology, weight >1500g, no assisted ventilation for 10 days prior to evaluation, O_2 requirements <30%, no CHF for one month prior to evaluation, no acute respiratory tract infection, no antihypertensive medications ten days prior to evaluation. The procedure is performed after direct laryngoscopic and bronchoscopic confirmation of the diagnosis. All other airway pathology must be ruled-out.

A vertical midline incision is made through the cricoid cartilage and first two tracheal rings as well as the lower thyroid cartilage. This allows the cartilages to spring open and allow edematous mucosa to drain, increasing airway size. Prolene stay sutures are placed on either side of the cricoid cartilage and the skin is re-approximated after placement of a drain. The child is then left intubated, sedated and paralyzed in the ICU for 7-14 days. Cotton has guidelines for endotracheal tube sizes for stenting and for duration of stenting based on the infant's weight.

Laryngotracheal expansion surgery involves scar division with distraction of the edges by interposition of graft material (augmentation) to widen the airway lumen. It is important to avoid removing scar, which results in a large surface area of denuded mucosa and leads to restenosis. Cotton recommends augmenting the airway with grafts when the distraction of the laryngotracheal framework must be greater than approximately 3mm. There are several techniques depending on the location and severity of the stenosis. Laryngotracheoplasty can be performed with a tracheostomy and formal stenting or by using the endotracheal tube as a stent, the latter known as a single-stage LTP (SS-LTP). There are a several stents that can be used for LTS including: endotracheal tubes, Silastic sheet rolls, Montgomery T-tubes, and laryngeal

stents. Laryngeal stents include: teflon stents [Aboulker stent (short or long), ETS Poirot, Paris], and silastic stents (Montgomery stents: Boston Medical Products, Boston. The primary consideration when deciding on the type of reconstruction and stent material is to provide a safe airway and adequate support for the graft. Success of LTR among, other things, is determined by the surgical procedure, including possible need for stenting; choice of type and length of stent; and duration of stenting. Choosing the appropriate method for stenting requires considering consistency of stenosis, altered anatomy, size, location and stability of grafts when used for surgical repair and host tissue healing factors (Zalzal, 1988)

Autogenous costal cartilage is the material of choice for grafting. Many other materials have been used for grafting including auricular, hyoid and thyroid cartilage and bone. Cartilage has much less resorption over time compared to bone. Although bone provides good structural support, grafts in the airway do no bear a lot of stress or weight.

Anterior laryngofissure with anterior lumen augmentation is a technique that is good for anterior subglottic stenosis or anterior tracheal wall collapse. The lesion should not involve the glottis. Other procedures should be considered if there the cricoid cartilage is deformed or weak. Anterior grafts are made considerably larger and thicker than grafts placed posteriorly. The perichondrium is oriented to the luminal side to allow for epithelialization. The perichondrium is also a good barrier against infection. A large external flange is created to prevent the graft from prolapsing into the airway.

Laryngofissure with division of posterior cricoid lamina is indicated for patients with posterior subglottic stenosis, posterior glottic stenosis that extends to the glottis, complete or circumferential stenosis, or if there is significant cricoid deformity. Division of the anterior and posterior cricoid must be carried out for this procedure. If possible, one should avoid a complete laryngofissure to avoid damaging the anterior commissure, however this is often needed for posterior glottic involvement for access. The posterior cricoid cartilage is incised in a manner that is vertically oriented to the cartilage to allow maximal purchase for the graft. The incision is extended superiorly to the interarytenoid area and inferiorly 5 to 10 mm into the membranous trachea. The graft is elliptical in shape. It should not be too thick as it can cause swallowing difficulties and can lead to aspiration. The width of the graft is determined by the desired distraction of the cut edges of the incised posterior cricoid cartilage. 0.05 to 1.00 mm of distraction can be obtained for each year of age, up to 1 cm. It is sutured in place with absorbable suture on a small cutting needle. The knots should be buried so that they remain extraluminal to prevent development of granulation tissue. Long-term stenting is usually necessary (3-6 months).

Laryngofissure and division of posterior cricoid lamina with anterior and posterior grafts should be used for patients who have SGS similar to those above but with a significant amount of stenosis posteriorly such that grafting is necessary to maintain the adequate separation.

Once the grafts have been sutured into place in any of the above procedures, the decision must be made on whether it should be single or double-staged. Cotton and Walner (1999) recommend a double-staged procedure for patients with severe stenosis, history of reactive airway, or poor pulmonary function. This should also be considered at institutions with

inadequate intensive care facilities. Double-stage procedure implies placement of stent above the tracheostomy tube instead of using an endotracheal tube as the stent (single-staged procedure). Once this decision is made, the strap muscles are closed to provide blood supply to the outer surface of the anterior graft.

The first cricotracheal resection (CTR) with thyrotracheal anastomosis was performed by Conley in 1953 in a patient undergoing surgery for chondroma of the cricoid cartilage. It was later popularized by Ogura and powers (1964) as a technique for treatment of traumatic stenosis. In the 1970s it became the treatment of choice in adults with acquired subglottic stenosis from long term intubation. Until recently, surgeons were reluctant to perform this procedure in the pediatric patients because of the risk of anastamotic dehiscence and recurrent laryngeal nerve injury, and disturbing the normal growth of the larynx. The first successful CTR performed in a child was occurred in 1978 (Savary). It wasn't until 1993, however, that the first series of 15 pediatric patients treated with CTR for severe LTS was published. Multiple subsequent series have reported using CTR for severe LTS with good outcomes (Monnier et al., 1995, Monnier et al., 1998, Stern and Cotton, 1999).

This technique is indicated if there is severe deformity of the cricoid making grafting very likely to fail. Most say that there must be at least 10 mm of normal airway below the glottis, however Cotton states that the resection can be up to the vocal folds but to expect prolonged edema. This technique is technically difficult due to the close proximity of the vocal cords and recurrent laryngeal nerves. Stenosis less than 4 cm can be resected by laryngeal release and cervical tracheal mobilization. Stenting is not required and the tracheotomy tube can usually be removed at around 4 weeks.

The goal of management of subglottic stenosis is decannulation. Success rates are dependent on the cause of the stenosis, the number of previous failed attempts, the status of the remainder of the airway, especially the glottis and the severity of the stenosis. Cotton has reported an overall pediatric LTR success rate of 92%, 97% for Grade II, 91% for Grade III, and 72% for Grade IV.

Bailey (1988) reported results of 131 pediatric airway reconstructive procedures. He had a 92% success rate with patients who underwent laryngotracheoplasty procedures (no grafting) and 80% success with patients who underwent LTR (with grafting). He did not report on use of any grading system.

Monnier et al. (1999) has reviewed the experience with CTR at the Department of Otolaryngology at the University of Lausanne, Switzerland. 69 CTRs were performed (48 infants and children and 21 in adults). 95% and 100% of the pediatric and adult patients, all of whom had Cotton-Myer grade III or IV stenosis, were successfully decannulated. Stern and Cotton (1999) reported on 38 pediatric patients who underwent CTR for severe LTS (grade III and IV). 33 patients were successfully decannulated. Complications preventing decannulation in this study included one patient with persistent aspiration, three who restenosed, one with arytenoid prolapse, and one with recurrent laryngeal nerve injury. Overall, 94% successful decannulation has been reported in the literature when CTR is used in pediatric patients with severe LTS (Monnier, 1999).

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