Evaluation and Management of Congenital Laryngeal Anomalies

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ANATOMICAL CONSIDERATIONS

The infant larynx differs from that of the adult in more ways than size alone. Its position in the neck is more cephalad than in the adult. The thyroid notch positioned high, posterior to the hyoid bone, the bone obscuring the thyroid notch as a landmark for tracheotomy. The epiglottis has an omega shape and may be somewhat tubular. The cuneiform cartilages, arytenoids, and posterior supraglottic soft tissue are of relatively large size. Of particular interest, as in the adult, the cephalad portion of the cricoid cartilage is V-shaped, not circular.

SYMPTOMS

1. Upper Airway Obstruction

   The infant or child who has symptoms of airway obstruction during sleep is most likely to have pathology in the pharynx, while the infant with laryngeal, tracheal or bronchial pathology will be more symptomatic awake, particularly during periods of stress such as crying and feeding. Congenital laryngeal anomalies (e.g., laryngomalacia, bilateral vocal fold paralysis) typically produce inspiratory stridor whereas intrathoracic lesions (e.g., bronchomalacia, tracheomalacia) produce expiratory stridor which mimicks the wheezing of asthma. Fixed lesions (such as subglottic stenosis) are most likely to have biphasic stridor.

2. Abnormalities of Phonation

   An abnormal or absent cry suggests a lesion of the glottis. A weak, breathy cry suggests unilateral vocal fold paralysis. A muffled sound is associated with supraglottic obstruction. A high-pitched or absent cry may occur with a laryngeal web or congenital cyst.

3. Dysphagia

   Aspiration of secretions or feedings produces coughing, choking, and gagging episodes. Esophageal atresia, laryngeal cleft, tracheoesophageal fistula, and neurogenic dysfunction of the larynx or pharynx produce these symptoms which typically include the...
constant presence of secretions in the hypopharynx, larynx, and trachea. Aspiration pneumonia may result.

DIAGNOSTIC EVALUATION OF THE AIRWAY

When anxious parents bring their stridous infant for evaluation, the mnemonic “SPECS-R” can help determine the need for endoscopy.

- **S**: severity of the airway obstruction, parents' impression.
- **P**: progression of the obstruction.
- **E**: eating or feeding difficulties, failure to thrive.
- **C**: cyanotic episodes.
- **S**: sleep-obstructive sleep apnea, particularly with evidence of cor pulmonale.
- **R**: radiology-specific abnormalities detected by radiogram may be indications for endoscopy.

Soft tissue posteroanterior and lateral films of the neck and chest are routine. With moderate to severe stridor or a history of dysphagia, preoperative barium esophagram is also obtained.

Flexible fiberoptic laryngoscopy is carried out in the office when the infant has mild symptoms. Complete direct laryngoscopy is accomplished in the operating room under general anesthesia when symptoms are moderate to severe. Preoperative medication is given with 0.02 mg/kg of atropine intramuscularly. Monitoring devices are placed: precordial stethoscope, blood pressure cuff, temperature probe, pulse oximeter, and ECG. Care is taken that the room is adequately warmed.

To induce anesthesia, nitrous oxide and oxygen, than halothane are administered by mask. If not already done, an intravenous line is placed. The child is hyperventilated before topical anesthesia (typically Xylocaïne 1% to 4%) is sprayed into the larynx and upper trachea. Spontaneous respiration is allowed to return. It is important for the endoscopist and anesthesiologist to communicate well, discuss strategy in advance, and cooperate throughout the procedure.

In most diagnostic cases, particularly when a neurologic lesion is considered, flexible fiberoptic laryngoscopy is carried out prior to the induction of general anesthesia. The small flexible fiberoptic laryngoscope is lubricated and, with the child gently restrained, passed through one nostril and into the pharynx for visualization of vocal fold mobility. Occasionally, it is necessary to conduct the awake laryngoscopy with a standard laryngoscope.

A side-slide laryngoscope is placed gently in the right side of the mouth, displacing the tongue to the left. No instrument is ever forced. The laryngoscope is lifted, avoiding a prying motion which uses the superior alveolar ridge as a fulcrum. If the insufflation technique of anesthesia is to be used, a catheter is placed through one nostril and taped with the catheter tip at the level of the epiglottis. Three to four percent halothane (and oxygen) is insufflated through the catheter with the child breathing spontaneously, entraining room air. The surgeon's right hand may be used to manipulate the larynx to gain a better view or different perspective of the laryngeal structures.

A magnified, well-lighted view of the pharynx, larynx, trachea, and bronchus is obtained using a 0-degree rod-lens telescope. This instrument is used without a bronchoscope, particularly if the diameter of the newborn larynx is substantially compromised. A complete view of the larynx, trachea, and main bronchus can be obtained with minimum trauma and in an exceedingly short period of time.

Palpation of the laryngeal structures is important to rule out arytenoid fixation (in
cases of vocal fold immobility) and to evaluate the nature of the subglottic stenosis (hard or soft). An anterior commissure laryngoscope is used gently to separate the vocal folds and examine the posterior glottis and subglottic larynx in greater detail. An elliptical cricoid cartilage (producing a subglottic stenosis) and a cleft larynx are overlooked very easily without this maneuver. Since it is difficult to estimate the size of the subglottic space accurately with the naked eye or telescope, a bronchoscope or endotracheal tube may be used to calibrate the diameter precisely.

LARYNGOMALACIA

Laryngomalacia is both the most common congenital laryngeal anomaly and the most frequent cause of stridor in children. 59.8% of congenital laryngeal anomalies in children who presented with stridor were due to laryngomalacia. Intermittent low-pitched inspiratory stridor is the hallmark of laryngomalacia. Symptoms usually appear within the first two weeks of life, although rarely, presentation may occur months after birth. This delay is quite interesting, since laryngomalacia is presumed to be congenital. An increase in the severity of stridor over the initial several months is generally followed by a gradual improvement. Symptoms are usually most severe by age 6 months, when they plateau and begin to resolve. Although most patients are symptom free by 18-24 months of age, stridor may persist for years.

Stridor is typically exacerbated by any exertion, crying, agitation, feeding, or supine positioning. If extreme cases go untreated, the infant may asphyxiate and die. Severe cases may be complicated by dysphagia, gastroesophageal reflux, failure to thrive, cyanosis, intermittent complete obstruction, and/or cardiac failure (as seen in obstructive sleep apnea).

The precise pathophysiologic abnormality that causes laryngomalacia remains elusive. Anatomic, histologic, and neurologic factors may all contribute to greater or lesser degrees in any one patient. Anatomic abnormalities regularly identified include the following:

Type 1 – Inward collapse of the cuneiform cartilages (which are often enlarged). Obstruction occurs as the cuneiforms are drawn inward during inspiration. Like a one-way valve, these cartilages open passively during expiration.

Type 2 – A long, tubular epiglottis (a pathologic exaggeration of the so-called omega-shaped epiglottis), which curls upon itself and contributes to obstruction during inspiration. This often occurs in association with type 1 laryngomalacia.

Type 3 – Anterior, medial collapse of the arytenoid cartilages to occlude the laryngeal inlet during inspiration.

Type 4 – Posterior inspiratory displacement of the epiglottis against the posterior pharyngeal wall or vocal folds. Type 4 may be associated with short aryepiglottic folds (type 5) or an overly acute angle of the epiglottis at the laryngeal inlet (type 6). Although two or more of these mechanisms often occur simultaneously, one feature usually dominates.

Expectant observation is suitable in the great majority of cases, as most patients’ symptoms resolve spontaneously. However, a small percentage of patients have such severe symptomatology that surgical intervention is unavoidable. Tracheotomy has been the standard means used to manage this group of patients, bypassing the pathology of the disease until the child outgrows the problem.

The term “supraglottoplasty” is used to describe surgical procedures that modify or resect flaccid obstructing supraglottic tissues. No two procedures are quite the same,
because anatomy and pathophysiology vary from one patient to the next. The location and extent of resection, therefore, is adapted to fit the individual mechanical problems. We have employed the CO₂ laser which provides a high degree of precision while maintaining hemostasis and minimizing postoperative edema, but its use is not mandatory. In some patients (such as those in whom laryngomalacia is reported to be due to short aryepiglottic folds) the use of microscopic scissors may be preferable.

In our experience, obstruction in the most severe cases of laryngomalacia is due to anteromedial collapse of the cuneiform cartilages (type 1) and/or anterior collapse of the arytenoid cartilages and overlying mucosa (type 3). The cuneiform cartilages are often disproportionately large.

Concerning surgical technique, conservative excision minimizes the likelihood of postoperative complications such as stenosis, aspiration, edema, or dysphagia. If the excision proves inadequate, the child can be returned to the operating room for more aggressive removal of tissue.

**SUBGLOTTIC STENOSIS**

Congenital subglottic stenosis typically presents with symptoms of stridor (biphasic or inspiratory), recurrent episodes of croup, or prolonged croup. The experienced pediatric otolaryngologist will realize "there is no such thing as croup under one year of age" and thoroughly investigate the airway of any infant with croup.

Rational therapy for subglottic stenosis is based upon an accurate determination of the histopathology and the precise location of the stenosis. The histopathologic classification of subglottic stenosis is presented below.

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<tr>
<th>HISTOPATHOLOGIC CLASSIFICATION OF SUBGLOTTIC STENOSIS</th>
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<tbody>
<tr>
<td>1. Cartilagenous Stenosis (Usually Congenital)</td>
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<tr>
<td>a. Cricoid Cartilage Deformity (Stenosis)</td>
</tr>
<tr>
<td>1. -Normal shape, small size</td>
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<tr>
<td>2. -Abnormal shape</td>
</tr>
<tr>
<td>• large anterior or posterior lamina</td>
</tr>
<tr>
<td>• generalized thickening</td>
</tr>
<tr>
<td>• elliptical shape</td>
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<tr>
<td>• cleft : partial, occult (submucosal), complete</td>
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<tr>
<td>• flattened shape</td>
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<tr>
<td>• other</td>
</tr>
<tr>
<td>b. Trapped First Tracheal Ring</td>
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<td>2. Soft Tissue Stenosis (Usually Acquired)</td>
</tr>
<tr>
<td>a. mucous gland hyperplasia</td>
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<tr>
<td>b. ductile cysts</td>
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<tr>
<td>c. fibrosis (fibrous connective tissue)</td>
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<td>d. granulation tissue</td>
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Tracheotomy is to be avoided if possible. Dilation, laser, or use of the resectoscope is rarely effective since most congenital stenoses are cartilaginous. Anterior laryngotracheal decompression (anterior cricoid split) or the single stage laryngotracheal reconstruction with costal cartilage graft may be used to avoid tracheotomy in infants and small children. When tracheotomy is unavoidable, follow up management includes:
1) conservative management, waiting for the patient to outgrow the problem. Occasional
dilation or calibration every 3-4 months is undertaken during this period.
2) laryngotraheal reconstruction, either with costal cartilage grafts or using the castillated incision in certain cases. A stent is placed whenever the posterior lamina of the
cricoid cartilage is divided. Each case is individualized and the management customized for the individual patient.

NEUROLOGIC LESIONS

Neurologic lesions are the third most common congenital laryngeal anomaly producing stridor in infants and children.27 Seventeen of 132 patients with congenital laryngeal anomalies (13%) had vocal fold paralysis. Fourteen were bilateral, 13 unilateral.

The diagnosis of vocal fold paralysis is made by awake laryngoscopy, since general anesthesia affects mobility. Fixation of the vocal folds is ruled out by palpation under general anesthesia. It is important that the child is completely relaxed for this part of the laryngoscopy.

Bilateral vocal fold paralysis typically produces a high-pitched inspiratory stridor, a phonatory sound or inspiratory cry. Approximately half the babies with congenital bilateral paralysis have no associated anomalies. The presence of neurologic, laryngeal, and cardiac anomalies associated with bilateral paralysis is the remaining group necessitates thorough evaluation of the infant including a neurological work-up. Bilateral paralysis is frequently related to myelomeningocele, Arnold-Chiari malformation, or hydrocephalus. Evaluation for increased intracranial pressure is important since appropriate treatment may produce resolution of the paralysis, obviating the need for tracheotomy.

Unilateral vocal fold paralysis produces aspiration, coughing, and choking. The cry is weak and breathy. Stridor and other symptoms of airway obstruction are less common which may account for the less frequent diagnosis of unilateral paralysis. The cause of unilateral paralysis is usually found within the chest. It is most commonly on the left and likely to be associated with a cardiovascular anomaly.

Vocal fold paralysis in infants usually resolves spontaneously. Temporary tracheotomy is often necessary for bilateral paralysis. Operative management is usually deferred until age 4 or 5.

CONGENITAL LARYNGEAL WEBS

Most congenital laryngeal webs are glottic. These cause abnormalities of phonation, typically producing a high-pitched cry or squeak. The infant may be completely aphonic and is likely to have respiratory difficulties as well. Thin, membranous webs are unusual. These thin webs may be treated by dilation alone or by incision and dilation until each side epithelialized and the web no longer reforms. Thick glottic webs with a broad anterior base and extension into the subglottic larynx are more common. Some of these represent a fusion of the vocal folds or a congenital cricoid cartilage abnormality with a thickened anterior lamina. These require laryngotomy and placement of a stent or keel to maintain the laryngeal lumen. Tracheotomy is required during this period. Since the voice is usually serviceable and the airway adequate, surgical intervention may be best deferred until age 5-10, unless the severity of airway obstruction makes tracheotomy necessary at an earlier age.

Supraglottic laryngeal webs are produced by fusion of a portion of the ventricular
folds anteriorly. These infants may have a normal cry and present with airway obstruction.

Posterior supraglottic webs involved the soft tissues posterior, superior to the cricoid cartilage. There may be fusion for some distance superiorly producing the congenital interarytenoid webs described by Benjamin. These are typically associated with subglottic stenosis. Tracheotomy is usually necessary.

**LARYNGOCELES AND SACCULAR CYSTS**

Laryngoceles and saccular cysts are abnormal dilations of the laryngeal saccule. The saccular cyst (congenital cyst of the larynx) is a mucous filled dilation of the saccule that does not communicate with the laryngeal lumen. The lateral saccular cyst extends posterosuperiorly into the ventricular fold and aryepiglottic fold from its origin at the non-patent orifice of the saccule. The anterior saccular cyst extends medially and posteriorly from its origin at the non-patent orifice of the saccule to protrude into the laryngeal lumen from the ventricle between the vocal and ventricular folds. Congenital saccular cysts may form as a result of simple atresia of the saccular orifice, a process commonly encountered as a cause of congenital abnormality.

A laryngocele is an abnormal dilation or herniation of the saccule which communicates with the laryngeal lumen. It is filled with air but on occasion may be temporary distended with mucous. An internal laryngocele is confined to the interior of the larynx and extends posterosuperiorly into the area of the ventricular fold and aryepiglottic fold. An external laryngocele extends cephalad to protrude laterally to the neck through the opening in the thyrohyoid membrane for the superior laryngeal nerve and vessels. When an external laryngocele is combined with a symptomatic dilation of the internal portion, it is termed a combined laryngocele. Laryngoceles are rare in infants and children.

Symptoms of laryngoceles in infants and children include intermittent hoarseness and dyspnea which increases with crying. A weak cry or aphonia has also been recorded. Saccular cysts typically present with respiratory distress, most commonly inspiratory stridor. An inaudible or muffled cry may be present. Dysphagia is occasionally encountered.

The diagnosis of a laryngocele is best made on the basis of a soft tissue lateral film of the neck. Direct laryngoscopy may disclose fullness of the involved ventricular fold and aryepiglottic fold. However, the laryngocele may not be distended with air at the time of endoscopic evaluation and the larynx may appear completely normal.

Saccular cysts are also diagnosed at direct laryngoscopy. Preoperative soft tissue lateral neck films contribute important information. Excision biopsy of anterior saccular cysts with cupped forceps not only confirms the diagnosis but also usually provides complete and permanent treatment. Needle aspiration of lateral saccular cysts confirms the diagnosis. When the cyst recurs it may be carefully dissected out endoscopically. Unroofing or "marsupialization" is rarely effective. Should the cyst recur, an external approach is considered. The airway can usually be managed by a brief period of intubation post-operatively so that tracheotomy is avoided.

The ductal cyst (acquired cyst, mucous retention cyst) is distinctly different from the saccular cyst. These common cysts may be found anywhere within the larynx and result from retention of mucous in the collecting ducts of the submucousal glands. The term ductal cysts is preferred because the cysts are dilated ducts and not distended glands as is commonly thought. In contrast to saccular cysts, ductal cysts are usually less than 1 cm in diameter. They are more common than saccular cysts and are typically found in the
subglottic larynx in patients who have been intubated for extended periods of time. Simple laser excision is usually adequate.

NEOPLASMS

Congenital subglottic hemangiomas typically produce symptoms of airway obstruction during the first six months of life. Approximately half are associated with cutaneous lesions. Lateral neck radiographs show an asymmetric narrowing of the subglottic larynx which may be similar in appearance to papillomas or ductal cysts. The diagnosis is made endoscopically. If the lesion is deep to the mucosa, the mucosa may have a normal appearance, the lumen having an asymmetric shape. The lesion is soft to palpation. Medical management includes systemic steroids. If endoscopic excision by CO₂ laser is considered, a computerized tomographic scan with contrast will determine the limits of the lesion. Over aggressive laser therapy can lead to stenosis. Since these lesions are self limited and will resolve by age 2-3 years, therapy should be conservative. Tracheotomy will maintain an airway in the interim.

Lymphangiomatous malformations rarely occur in the larynx. When they do they are invariably an extension of disease in the head and neck region. The supraglottic larynx is most often involved. Airway obstruction may necessitate tracheotomy. Often the lesion can be debulked using the CO₂ laser to maintain an adequate airway.

Neurofibromatosis rarely involves the larynx in the infant. When children are affected, limited local resection should be undertaken to maintain an airway and optimize the voice. Recurrent respiratory papillomatosis, while of viral etiology, is mentioned because it often presents within the first year of life and has even been reported as the onset of symptoms at birth. Papillomas must therefore be considered in the differential diagnosis of congenital anomalies of the larynx.

REFERENCES