IAPO-Interamerican Association of
Pediatric Otorhinolaryngology

A Comprehensive Approach to Evaluating the Pediatric Larynx

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Introduction

Pediatric laryngeal evaluation includes an effort to diagnose and manage complex issues in respiration, swallowing and voice production. Examination of the larynx should address all these functional aspects. Developments in fiber optics and anesthetic management have improved the Otolaryngologists ability to perform a dynamic assessment of the pediatric larynx. The purpose of this chapter is to describe a modern approach to evaluation of the pediatric larynx.

Evaluation of Stridor

The most common reason for prompt airway evaluation is respiratory distress or stridor [1-3]. Stridor or noisy breathing is a nonspecific term and in itself is not pathognomonic or diagnostic of any disease process. Abnormal sounds are caused by turbulence when air flows through a narrowed area in the airway. Even a slight degree of airway narrowing in an infant or child may result in significant increase in airway resistance. With narrowing in the supraglottic airway inspiration will increase negative pressure and cause the supraglottic structures to collapse. Expiration will force these structures open and allow unobstructed airflow. The resultant stridor with supraglottic obstruction will occur only with inspiration. This is typically seen with laryngomalacia.

The subglottic space is encompassed by the circumferential cricoid ring. Narrowing in this area will be present on expiration and inspiration, or biphasic stridor. This is seen in viral croup and subglottic stenosis. Stridor associated with pathology of the lower trachea will be present solely on expiration. The relative positive pressure associated with expiration will collapse the already narrowed tracheal lumen, and cause a low-pitched expiratory wheeze. Inspiration will be unobstructed. Therefore by determining if the stridor is present on inspiration (supraglottic), expiration (tracheal), or both (subglottic), one should be able to make an approximation of the anatomic level of the obstruction [4].

The most common causes of acute respiratory distress are inflammatory diseases of the pediatric airway, but initial consideration in the neonate should also include cardiac or neurologic difficulties. The degree of respiratory distress can be estimated by assessing the patient's color, retractions, respiratory rate, air entry, and state of consciousness. The level of anatomic involvement can easily be accurately predicted by listening to the stridor and the phase of respiration. The majority of cases are evaluated with flexible fiber optic laryngoscopy. In cases

of severe distress examination in the operating room may be necessary for a safe airway evaluation. Obviously at times, one must secure the airway immediately and it is not possible to get to the operating room. Extensive investigations should wait until the patient is stabilized.

The operating room provides a controlled environment to asses and establish an airway. A successful trip to the operating room depends on excellent communication among members of the operating team. The plan should be discussed in detail with the anesthesiologist and the nursing staff. The otolaryngologist is responsible to select the appropriate sized bronchoscopes. Although the choice of anesthetic technique is up to the anesthesiologists, maintaining spontaneous ventilation is ideal. This allows for excellent ventilation and provides a dynamic view of the airway.

Topical lidocaine is applied to the vocal cords at concentrations of 2% or 4% depending on the patient's age. It is critical to position the laryngoscope in the vallecula in order to give excellent exposure of the larynx and avoid pinning a vocal cord which might give the false impression of vocal cord paralysis. The use of a rigid pediatric bronchoscope and a rod lens telescope allows for excellent visualization of the airway while oxygenating through its side arm. The rigid bronchoscope also provides an adequate amount of working space to permit the use of foreign body forceps and suction. Tracheoscopy using the magnifying telescope alone, during spontaneous ventilation is another way to visualize the lower airway without risk of iatrogenic edema.

Patients with chronic stridor are able to tolerate a more leisurely workup and examination. Chronic stridor may present as mild respiratory distress from birth, with exacerbations during periods of upper respiratory infection [3]. A complete history should include questions regarding birth history, previous intubations, mode and age of onset, and presence of any associated symptoms will be most helpful. Careful observation of the patient in a variety of positions, awake, asleep, and during feeding will provide the astute physician with a great deal of information. Symptoms, such as phase of the stridor, feeding difficulties, quality of the cry and voice, and presence or absence of retractions will help identify the anatomic level of obstruction.

The most common cause of chronic stridor is laryngomalacia. In laryngomalacia, the elongated, tubular-curved omega shape of the infant epiglottis, the redundant arytenoids and short aryepiglottic folds partially obstruct the airway on inspiration. This is a benign, self-correcting condition of unknown etiology. Laryngomalacia usually resolves between the ages of 12 and 18 months, most commonly without any intervention. In laryngomalacia the inspiratory stridor is not present at birth but it develops shortly thereafter, days or weeks after birth. Cyanosis is extremely uncommon in laryngomalacia and should always raise suspicion for the presence of another lesion.

In cases of severe laryngomalacia, epiglottoplasty is indicated. A variety of procedures using the CO_2 laser, or cold instruments, are used to incise the aryepiglottic folds, in order to release a tethered epiglottis, or reduce of redundant arytenoid mucosa to prevent prolapse into the airway.

Although laryngomalacia is an extremely common diagnosis, it is important to remember that all stridor is not laryngomalacia. Other causes of chronic congenital stridor include subglottic stenosis, vocal cord paralysis, cysts, webs, cricoid anomalies and laryngeal clefts. Extrinsic compression of the airway may occur with vascular ring formation, mediastinal or thyroid tumors. Aortic arch abnormalities usually present shortly following birth, although a final diagnosis may not be established for several months. Symptoms frequently include recurrent bronchopneumonia, dysphagia, and wheezing. In a review of 132 patients who underwent evaluation for stridor, both acute and chronic, on endoscopy Holinger found that the majority had congenital abnormalities. Laryngomalacia was the most common diagnosis with inflammatory illnesses of the airway, and aspirated foreign bodies being second and third [1].

In patients with less severe distress, fiberoptic nasolaryngoscopy can be safely performed and is often diagnostic. Flexible fiberoptic can provide a complete evaluation of the airway including nasal cavity and nasopharynx down to the level of the glottis, but should not be passed below the vocal cords in the awake child. Flexible laryngoscopy is able to give a dynamic view of the larynx. Flexible laryngoscopes are available ranging from 1.9 mm to 4 mm in diameter.

Flexible laryngoscopy is usually well tolerated and can be performed in all ages including newborns. Use of decongestant drops and topical anesthetic facilitate the exam. In pediatric patients assistance holding the patient's head is usually necessary to obtain an adequate exam.

There are advantages and limitations of flexible laryngoscopy in comparison to the rigid examination. The most significant advantage of flexible endoscopy is the ability to have a dynamic view the larynx or trachea during spontaneous respiration without an obstructed view due to an endotracheal tube. This is a particular help when evaluating vocal cord function or tracheomalacia.

Flexible fiberoptic examination, however. is not recommended when the airway is not controlled in patients with a history of possible foreign body aspiration. Without a secured airway the duration of the examination is limited making it difficult to access many of the more complex, uncommon or subtle situations. The sizes of pediatric flexible endoscopes allow limited ability for suctioning secretions, and for removal of foreign bodies.

Radiographic studies to assess patients with mild to moderate acute or chronic stridor are usually more useful than blood tests or other laboratory studies. However no radiographic studies should be attempted in patients with severe respiratory distress. Fluoroscopy of the airway will reveal the dynamic changes associated with respiration. Since many foreign bodies are not radiolucent; the physiologic changes caused by the obstruction will often be apparent on fluoroscopy. Evidence of heart and mediastinal shift during expiration consistent with of air trapping are the most common radiologic findings with foreign body aspiration. The lack of abnormal findings on airway fluoroscopy however, does not mean that there is no possibility of a small foreign body**.** A barium swallow is a straightforward diagnostic test to look for an extratracheal cause for obstruction, e.g., a vascular ring for this reason a swallow study with airway fluoroscopy

should be a part of the workup of neonates in respiratory distress with negative flexible laryngoscopy findings [5].
The final diagnosis is most often established by laryngoscopy and bronchoscopy. An experienced endoscopist should perform pediatric endoscopy with the necessary instruments available in a variety of sizes [6] optimally with experienced nurses and anesthesiologists. The rigid ventilating bronchoscope allows for the relatively leisurely inspection of the entire length of the larynx, trachea, and upper bronchi. Use of the magnifying telescope allows evaluation of the neonate and premature infant. Tracheoscopy with the magnifying telescope alone without a rigid bronchoscopy can be performed with the patient anesthetized with spontaneous ventilation. This anesthetic technique is an excellent choice as it allows one to access the dynamic changes of the trachea during normal breathing. Since an esophageal foreign body may present primarily with respiratory symptoms, esophagoscopy should be performed in order to have a complete. evaluation

Evaluation of the swallowing function

Children with swallowing difficulties are at higher risk for malnutrition, dehydration and pulmonary problems. Causes of feeding and swallowing problems are varied and the majority of them are not related to laryngeal function. Evaluation of the swallowing function should include a multidisciplinary approach involving the speech pathologist, the pediatrician and the otolaryngology service.
Assessment of swallowing function begins with a full history and physical examination. Patients with neurologic deficit, with a history of recurrent pneumonias or vocal cord paralysis are likely to have decrease laryngeal sensation.
Swallowing function is divided in three different phases oral, pharyngeal and esophageal. The laryngeal adductor reflex allows for coordination of swallowing and breathing by producing glottic closure during swallowing in the pharyngeal phase. This reflex response is elicited by chemical or mechanical stimulation of the supraglottic mucosa. Abnormal laryngeal adductor reflex will lead to an increased laryngeal penetration and aspiration due to inability to sense secretions or food in the laryngopharynx.
Modified barium swallow is a video fluoroscopic procedure in which the child is fed different consistencies while in a seating position. The different phases of swallow and the ability of the child to tolerate different consistencies are recorded.
Bedside evaluation of swallowing function and video fluoroscopic swallowing studies have been complemented by a new generation of test such as the flexible endoscopic evaluation of swallowing (FEES) and sensory testing to evaluate the laryngopharyngeal sensory threshold.
In flexible endoscopic evaluation of swallowing and sensory testing (FEESST) a calibrated puff of air is applied to the aryepiglottic folds eliciting an involuntary reflex. In FEEST a response is described as normal if the laryngeal adductor reflex is induced a <4 mm Hg. A response at 4.0 - 6.0 is consistent with a moderate sensory deficit and > 6 mm Hg indicates a severe deficit [7]. In a recent study of 100 patients ages 1 month to 24 years, patients with a history of recurrent pneumonia

and neurologic deficit had significantly higher sensory thresholds than patients without this conditions [8]. This technique differs from FEES in which colored food of different textures are given to the patient to evaluate for pooling, penetration and aspiration. Not every patient tolerates these test since cooperation is necessary to evaluate the swallowing function.

These new videoendoscopic assessment techniques of swallowing function allow adequate documentation of the anatomy and airway protection without radiation exposure or ingestion of contrast mediums. While these techniques produce a quantitative measurement of the sensory stimulation threshold, they require patient cooperation, training and are not yet widely available.

Voice Evaluation

Over a million children have voice disorders in the United States. Diagnosis and treatment of dysphonia in children is important to identify layrngolical pathology and to determine appropriate treatment. Disorders of phonation can be caused by a variety of abnormalities. Careful history taking and physical exam will help distinguish between laryngeal, velopharyngeal or cognitive delays associated with voice disorders.

Pediatric dysphonia can be classified as congenital or acquired. Congenital causes of dysphonia may manifest as a weak cry, or respiratory distress. Examples of congenital dysphonia include glottic webs and vocal cord paralysis. Acquired dysphonias include infectious, neoplastic, inflammatory, anatomic, neurologic and iatrogenic causes.

Infectious processes are the most common cause of dysphonia in children. Vocal cord edema from viral upper respiratory infections will lead to temporary hoarseness. Other viral conditions such as acute laryngotracheobronchitis may present with hoarseness, low grade fever, biphasic stridor and a barky cough. Epiglottis, which was prevalent prior to the introduction of the *H. influenzae* type B vaccine, has significantly decreased. Laryngeal papillomas, which are caused by human papilloma virus initially present with hoarseness but will progress to airway obstruction over time. Treatment of this condition usually requires multiple surgical procedures often resulting in permanent dysphonia. Current treatment is palliative and aimed at maintaining a patent airway. Research will hopefully provide a cure [9].

Vocal cord nodules represent the most common cause of hoarseness in children after upper respiratory infections [10]. Nodules are usually found on the anterior to middle one third of the vocal cords and are associated with vocal abuse. In a recent study by Shah et al. out of six hundred forty six children evaluated for dysphonia, two hundred and fifty-four patients (40%) were identified as having vocal nodules. Of these, 72% were male aged 3-10 years old. Although rigid stroboscopy in children is challenging, it is useful. The mucosal wave on stroboscopy is diminished in the presence of vocal cord nodules where the mucosal wave is not affected in several other causes of hoarseness such as subepithelial cysts [11].

Gastropharyngeal reflux has been associated with chronic hoarseness and vocal cord nodules in children. Intermittent dysphonia with worsening of symptoms

during the morning. Empiric therapy with proton pump inhibitors may be warranted as a diagnostic trial or referral to a gasteronterologist should be considered.

Congenital causes of dysphonia include glottic webs, unilateral and bilateral vocal cord paralysis. Smith and Caitlin reported that glottic web and atresia account for 5% of laryngeal congenital anomalies [12]. Depending on the extent of the lesion patients may present with dysphonia, aphonia or respiratory distress. Initial evaluation consists of flexible laryngoscopy in the clinic in the stable, cooperative patient. Glottic webs are classified depending on their severity in 4 types. Type 1 is an anterior web involving 35% or less of the glottis. Type 2 involves up to 50% of the glottis and the vocal cords are still visible within the web. Type 3 involves up to 75% of the glottis and extends into the subglottis. Type 4 involves up to 90% of the glottis and the web is uniformly thick with subglottic extension [13].

Vocal Fold Immobility (VFI) is a common cause for dysphonia and respiratory distress in the pediatric patient. Flexible laryngoscopy in the awake child has become the keystone for diagnosis of pediatric VFI with the vast majority of children with VFI being accurately diagnosed using this technique [14]. Flexible laryngoscopy, however, can be challenging in the very young child with copious secretions, rapid respiration or associated laryngomalacia all of which can make visualization of the cords difficult. We believe the ability to record and replay the exam in slow motion offers a significant advantage and often allows to make the diagnosis even in difficult cases. There is still a role, however, for direct laryngoscopy which allows to palpate the arytenoids to rule out fixation and in when indicated to perform intraoperative electromyography (EMG) [15]. Ultrasound can also be used to aid with diagnosis in cases where visualization is limited [16].

Vocal fold immobility is divided in unilateral and bilateral, and into congenital or acquired.

The majority of children with unilateral vocal fold immobility adequately compensate for loss of function without the need for surgical intervention. Some patients however, may present with symptoms such as aspiration, recurrent pneumonia, dysphonia, feeding difficulties, poor ability to cough and even stridor associated to unilateral vocal fold immobility (UVFI).

There is not much written about management of unilateral vocal fold immobility in children since most patients compensate adequately and surgeons being aware of the risk that good surgical results at an early age may be lost with subsequent growth. Sipp et al reported in 2007 a series of 15 pediatric patients with unilateral vocal fold immobility requiring medialization techniques including injection laryngoplasty, thyroplasty and ansa cervicalis-recurrent laryngeal nerve reinnervation with good voice and swallowing outcomes [17].

Children with bilateral vocal fold immobility tend to present early in life. Most cases are congenital due to a neurological defect such as Arnold -Chiari malformation or an idiopathic cause. Respiratory distress is more severe in cases of bilateral VCP. A loud biphasic stridor, cyanosis, and apnea are common findings in patients with bilateral VCP. On the other hand, voice and cry may be virtually normal in children with bilateral VCP.

The initial management is to provide a secure airway and to elucidate the etiology of the paralysis. Most physicians agree that tracheotomy is necessary in over 50 percent of these cases [18]. Miyamoto et al recently reported a retrospective study with a mean follow up of 50 months in which 15 of 22 patients (68%) with bilateral vocal cord paralysis required tracheostomy [19].

Paradoxical vocal cord movement can be characterized as an abnormal adduction of the vocal cords during the inspiratory phase of the respiratory cycle. This produces airflow obstruction at the level of the larynx. This condition frequently mimics asthma and due to this misdiagnosis is often treated with high-dose inhaled and/or systemic corticosteroids, bronchodilators. These patients may require multiple emergency room visits and hospitalizations. Diagnosis is made with a flexible nasolaryngoscopy which will reveal vocal cord adduction during the inspiration. Patients with paradoxical vocal cord movement may often be able to talk long sentences without stridor or respiratory distress which may provide a diagnostic clue during examination. These patients benefit from speech therapy and at time psychological counseling. Paradoxical vocal fold movement can also occur in the newborn as recently described by Omland in 2008 [20]. Patients with this condition often present with symptoms similar to bilateral vocal fold immobility and in some cases with failure to thrive and persistent respiratory distress may require a tracheotomy.

Physical examination for the child with voice disorders varies with the age of the patient. An endoscopic flexible nasolaryngoscopy exam can be easily performed in most ages without the need of sedation. Adequate topical anesthesia and vasoconstriction should be applied to the anterior nares prior to performing the examination. This technique allows adequate anatomic and functional evaluation of the larynx [21].

Rigid endoscopy allows an excellent resolution and can be used in cooperative children above the age of 5. Stroboscopy is useful to delineate lesions that affect mucosal wave motion such as vocal cord nodules or as follow up in patients with laryngeal papillomas to evaluate for possible surgical sequelae [21]. Unfortunately rigid stroboscopy is not well tolerated in some pediatric patients. The use of laryngeal electromyography (EMG) for vocal cord paralysis has limited use in the pediatric population and often requires general anesthesia and suspension laryngoscopy in younger patients.

In conclusion, evaluation of the pediatric larynx includes more than assessment of the airway and laryngeal function. Otolaryngology must also address the swallowing and phonation functions of each patient. Advances in diagnostic techniques and implementation of minimally invasive techniques following phonosurgical principles will be exciting options in the future of pediatric laryngology.

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