Review Article

Hoarseness in Children

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ABSTRACT

Background: The prevalence of pediatric dysphonia ranges from 6-23%. Chronic dysphonia can negatively affect the lives of children physically, socially, and emotionally. The body of literature continues to grow regarding the pathophysiology and management of dysphonic children.

Methods: This article presents a relevant literature review of vocal fold pathology leading to hoarseness and recent advances in diagnosis and management. Articles were retrieved using a selective search in PubMed employing the terms such as “hoarseness in children,” “pediatric dysphonia.”

Results: 42 articles from the past decade were reviewed that include information regarding the etiology, assessment, and treatment of children with dysphonia.

Conclusion: The care of a child with a voice disorder can be complex and requires a multi-disciplinary approach. Current technological, pharmaceutical, and therapeutic advances have improved the treatment of children with dysphonia.

Keywords: Hoarseness in children, Pediatric dysphonia, Pediatric hoarseness.


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INTRODUCTION

“Hoarseness” and “dysphonia” are terms used to describe changes in voice quality. In the pediatric population, the prevalence of hoarseness ranges from 6 to 23% of 4- to 12-year-old children. The etiology of hoarseness can be classified as infectious, inflammatory, iatrogenic, traumatic, congenital, and functional. Chronic dysphonia can negatively affect the lives of children physically, socially, and emotionally. In contrast to the commonly held belief that children do not appear aware or concerned about the sound of their voices, children as young as 6 years express concerns about dysphonia. This article presents a relevant literature review of the current thinking in vocal fold pathology leading to hoarseness and recent advances in diagnosis and management.

ASSESSMENT OF DYSPHONIA

Perceptual analysis is a key component of voice evaluation in children, and several tools are currently available for assessing these qualities. Perceptual analysis of voice by the patient, the parent, the clinician and speech language pathologist is important in the comprehensive assessment of pediatric hoarseness. Parental questionnaires such as the Pediatric Voice outcome Survey, the Pediatric Voice-Related Quality of Life questionnaire, and the Pediatric Voice Handicap Index are available. In these surveys, the child’s self-evaluation is not considered. Verduyckt et al developed a questionnaire to analyze the discriminatory capacity of 5- to 13-year-old dysphonic children. They demonstrated the need for a standardized questionnaire permitting self-evaluation of the child’s voice.

Visualization of the larynx is critical for accurate diagnosis and management of pediatric dysphonia. In most children, this can be done in the clinic and does not require examination under anesthesia. Videolaryngostroboscopy (VLS) with a rigid endoscope is a standard technique used in adult patients with dysphonia. Videolaryngostroboscopy elucidates subtle features of different disease processes and clarifies differences between benign mucosal disorders and inflammatory processes that contribute to dysphonia. Compared with adults, children have shorter phonation times, shorter attention spans, difficulty completing vocal tasks, and small larynges, all making visualization difficult. Therefore, flexible fiberoptic endoscopy is more frequently used in children. Current technology allows VLS with a flexible fiberoptic nasopharyngoscope. Furthermore, many digital endoscopy systems allow the storing, replaying, and comparing of archived images, which provides a more objective evaluation. In one study of 150 children aged 2.5 to 14 years, VLS was successfully performed in all but one child. High-speed imaging of the larynx as well as laser projection systems have been described to enhance measurements of pediatric vocal fold vibratory patterns, length, and amplitude. Wang et al used ultrasonography to diagnose pediatric vocal fold paralysis and found it to be a reliable assessment tool in unilateral paralysis. Bisetti et al used ultrasound to compare the true vocal fold with flexible endoscopic images in 16 children with dysphonia and found the vast majority...
of cases were in agreement, although ultrasound was unable to differentiate the type of lesion.

BENIGN MUCOSAL LESIONS AND OTHER BENIGN TUMORS OF THE LARYNX

Mass lesions that involve the membranous vocal folds and cause dysphonia in children include nodules, cysts (mucoid or epidermoid), polyps, webs, and sulcus vocalis. In children, the vast majority of vocal changes are secondary to phonotrauma leading to vocal nodules that can be managed conservatively with education (vocal hygiene) and voice therapy. Vocal nodules develop from repeated trauma to the vocal cords, resulting in an inflammatory reaction and damage to the epithelium and lamina propria. They are often bilateral and commonly located at the junction of the anterior one-third and posterior two-thirds of the vocal fold where pressure of contact between the vocal cords is greatest. Respiratory symptoms, such as nasal obstruction leading to chronic mouth breathing and gastroesophageal reflux can be related to the development of vocal nodules.

Vocal cord nodules most commonly occur in males due to vocal abuse caused by behavioral and structural differences compared with females. However, vocal nodules in females are more likely to persist with age due to differences in glottic growth. During puberty, the vocal cords have greater elongation and the thyroid cartilage becomes more acute in males. Speech therapy is the most widely accepted treatment for vocal fold nodules in males and females. In one prospective study, the effect of voice therapy in a cohort of 39 children with vocal cord nodules was examined via VLS and revealed significant improvements. Microlaryngeal surgery may be indicated for highly dysfunctional or persistent nodules. When surgical intervention is performed, vocal nodules can recur in as little as 3 days if poor vocal habits are not improved with speech therapy. When a child with “nodules” does not respond to treatment, it is important to consider alternative diagnosis.

Vocal cysts are classified into epidermic or mucus retention cysts. Epidermic cysts are diagnosed by the presence of vascular dilation on their surface and described as pearly lesions that decrease vibratory wave on videostroboscopy. Epidermic cysts have dense keratinized epithelium, making them more resistant to rupture with surgical manipulation. Mucus retention cysts develop from obstruction of glandular ducts and are cavitory lesions covered with cylindrical epithelium, and are more likely to rupture with manipulation. Cysts are caused by laryngeal inflammation associated with gastroesophageal reflux disease (GERD), upper airway infections, or vocal abuse. In a study performed by Martins et al, 304 children aged 7 to 9 were evaluated endoscopically. Epidermal cysts were the second most common mucosal lesions diagnosed. A total of 57% of lesions were nodules, while 15.4% had epidermal cysts. Both lesions were associated with other findings such as mucosal bridges and sulci. While acquired benign lesions are often caused by vocal trauma or abuse, it is still unclear why some children develop nodules vs cysts. The type of vocal habit may play a role. For example, children with vocal nodules often have hard glottal attacks and significant strain when phonating.

Vocal cord polyps are rarely seen in children. Vocal cord polyps occur in the superficial layer of the lamina propria; they are usually unilateral and located at the junction of the anterior one-third and posterior two-thirds of the vocal cord. Traditionally, surgery has been the mainstay of treatment for vocal cord polyps, although a Korean study evaluated 248 patients with vocal cord polyps and 94 were treated conservatively with vocal hygiene and laryngopharyngeal reflux (LPR) treatment measures. In 43% of polyps, there was a reduction in size and 36% resolved completely without surgery. The majority of patients were female with small, recent-onset polyps.

Sulcus vocalis is a rare lesion affecting one or both of the mucosal surfaces of the vocal folds. It is characterized by a linear groove on the mucosal surface of the vocal fold with varying depths, at times approaching or touching the vocal ligament. Several studies have identified a genetic origin. Many times a sulcus is missed in the initial exam and found at the time of surgery under microscopy, although better imaging techniques currently allow for visualization in the office. In many cases, surgical management offers improvement in dysphonia in these children.

Vocal cord granulomas are benign inflammatory lesions and typically located in the posterior glottis and associated with the arytenoid cartilages. They are often iatrogenic and caused by prolonged or traumatic intubation. Chronic inflammation secondary to LPR, habitual throat clearing, and vocal abuse are also factors involved in the development of vocal cord granulomas. Treatment is similar to vocal cord nodules.

Benign tumors of the larynx include hemangiomas, papillomas, cystic hygromas, and neurofibromas. The most common of these is recurrent respiratory papillomatosis (RRP), an infection of the upper airway by the human papilloma virus (HPV). Viral transmission is usually acquired from birth. Approximately 75% of cases present before 5 years of age and 25% occur...
between 5 and 12 years of age, which is thought to be caused by latent virus remaining in the laryngeal tissues. The symptoms can progress from mild hoarseness to dyspnea and stridor as the papillomas progress and obstruct the glottic airway. The pediatric form, juvenile-onset RRP (JORRP), is the more aggressive subtype.\textsuperscript{21} Laryngeal mucosal RRP is affected most often by the low-risk subtypes, HPV-6 and HPV-11, which accounts for approximately 90\% of JORRP. Patients with HPV-11 subtype often demonstrate more aggressive disease than those with HPV-6, with the following features: younger age of presentation, more frequent recurrence with need for more surgical procedures, higher rate of subglottic or bronchogenic spread, and longer periods of disease activity. High-risk subtypes HPV-16 and 18 have also been identified in JORRP, but account for only 1\% and are more often associated with malignant transformation, or “carcionoma ex-papilloma.”\textsuperscript{22} The mainstay treatment modality for this condition is surgical removal of lesions with microlaryngoscopy using a powered microdebrider or laser. Children who require more than four surgical procedures have rapid regrowth, or airway compromise should be considered for adjunct medical therapy.\textsuperscript{23} Alpha interferon was the first widely employed of these therapies and now available as pegylated interferon. The cyclooxygenase-2 enzyme is a growth stimulator and overexpressed in RRP, and Celecoxib, a nonsteroidal anti-inflammatory drug, may inhibit the papilloma growth.\textsuperscript{23} Cidofovir is an antiviral and the most widely used adjunct therapy for RRP, although there is insufficient evidence from randomized controlled trials to support the efficacy of antiviral agents.\textsuperscript{24} Cidofovir reduces the efficiency of deoxyribonucleic acid (DNA) transcription on the growing DNA chain. Dose ranges of 2.5 to 15 mg/mL have been reported, and a review of the literature recently indicated that higher infiltrated doses result in longer relapse free times. Furthermore, previous concerns of use of cidofovir and increased malignant transformation have been refuted by studies demonstrating no statistical difference in cidofovir and non-cidofovir groups.\textsuperscript{22} Bevacizumab is a chemotherapeutic agent used to inhibit growth of papillomas.\textsuperscript{25} Bevacizumab is recombinant humanized monoclonal immunoglobulin G1 antibody that binds to and inhibits the biologic activity of human vascular endothelial growth factor \textit{in vitro} and \textit{in vivo}. There is evidence that vascular endothelial growth factor is an important factor in the development of RRP, and there have been some recent reports of promising results achieved with relatively high dosing (45–50 mg) of bevacizumab in all patients treated. The dose approved by the US Food and Drug Administration is 12.5 mg.\textsuperscript{26} The HPV vaccination includes the viral subtypes responsible for laryngeal papillomatosis, and will hopefully decrease transmission and result in future eradication of this disease in children.\textsuperscript{5}

The factors that lead to virus activation have not been identified; however, GERD and LPR have been suggested. When chronically exposed to pepsin and gastric contents, the inflammation induced by chronic acid exposure may result in the expression of HPV in the ciliated respiratory epithelium of the larynx, possibly contributing to more advanced presentations of the disease or a more frequent need for surgical debridement. Some of the concerning findings in disease progression of RRP, such as laryngeal webs, may be diminished or even prevented if these children receive antireflux therapy.\textsuperscript{27,28} This emphasizes the importance of initiating antireflux medication in children who have evidence of RRP.

\section*{CONGENITAL ABNORMALITIES}

An infant that presents with an unusual cry or respiratory distress should be evaluated for congenital abnormalities of the larynx, including laryngeal webs and hemangiomas. Patients with concomitant cardiac anomalies have higher incidence of congenital laryngeal anomalies.\textsuperscript{29} Failure of resorption of the epithelial layer that normally obliterates during the development of the glottis results in laryngeal webbing, or incomplete separation of the vocal cords. These can occur anywhere along the vocal cords. Atresia, or complete webbing of the larynx, is incompatible with life and requires tracheostomy at birth. Laryngeal webs are commonly associated with other airway abnormalities, DiGeorge and velocardiofacial syndromes.\textsuperscript{26,30} Presentation varies depending on the degree of airway obstruction, and treatment correlates with the severity. Small anterior webs can be treated with sharp dissection or lasers; larger webs may require laryngeal stents, reconstruction, or tracheostomies.\textsuperscript{31}

\section*{PARALYSIS}

A common cause of infantile and neonatal dysphonia is unilateral or bilateral vocal cord paralysis. Unilateral vocal cord paralysis may present with symptoms of chronic aspiration, weak or absent cry. In bilateral vocal cord paralysis, the vocal cords assume a midline position, thus protecting the airway from aspiration and permitting a normal cry. Children will often compensate well early in life and may not be symptomatic until they become more active or suffer an acute respiratory infection. Diagnosis relies on laryngoscopy, but other adjuncts such as ultrasound and laryngeal electromyography may also be helpful in making the diagnosis and formulating a treatment plan.\textsuperscript{32} Etiologies include neurologic, iatrogenic, birth-related trauma, or
idiopathic. Damage to the recurrent laryngeal nerve can occur during cardiothoracic surgery, thyroid surgery, or tracheoesophageal fistula repair. The most common surgery that puts the nerve at risk is ligation of patent ductus arteriosus. Neurologic causes include hydrocephalus, Arnold–Chiari malformation, posterior fossa tumors, and meningomyelecele. Children with vocal cord paralysis should have prompt airway and swallow evaluations, as well as imaging of the chest and brain. In unilateral vocal cord paralysis, surgical intervention is indicated for children who are unable to compensate with the nonaffected vocal fold after speech therapy. Procedures such as vocal cord injections are temporary and can provide medialization in situations where the nerve is expected to recover. Medialization thyroplasty is a more permanent surgical technique that allows an implant to be placed through a window in the thyroid cartilage resulting in the medial movement of the vocal cord. This has limited applications in children due to concerns about laryngeal and overall physical growth. If concerns for vocal fold atrophy are present with symptoms lasting for greater than 1 year, or there is a known transected nerve, ansa cervicalis to recurrent laryngeal nerve graft can be used to provide bulk and tone to the affected vocal fold. This procedure has been shown to improve expert and self-perceptual ratings, voice handicap indices, maximum phonation times, and pitch range values. In contrast to unilateral vocal cord paralysis, surgical intervention is often needed for bilateral paralysis. Procedures to lateralize the vocal cords can be conducted via an endoscopic approach and include unilateral laser arytenoidectomy, cordotomy, or vocal cord lateralization. Tracheostomy is often required prior to procedures, such as cordotomy and arytenoidectomy.

**LARYNGITIS**

Children have smaller airways than adults and are more susceptible to inflammation and edema of the subglottic airway. Laryngitis is usually related to viral respiratory tract infections in children but can occur secondary to acute vocal strain and submucosal vocal cord inflammation or hemorrhage. Laryngotracheobronchitis, or croup, is commonly viral in origin and affects children less than 5 years of age. Management is conservative with steroids reserved for airway compromise. Young infants less than 6 months old are more likely to have viral infection superimposed on a congenital defect of the airway such as laryngomalacia or subglottic stenosis, especially if noisy breathing is present prior to illness. A toxic appearing child is more likely to have bacterial tracheitis, retropharyngeal abscess, or epiglottitis. Chronic laryngitis is less likely to be infectious. If symptoms of laryngitis last for more than 3 months, chronic changes, such as mucosal edema and thickened epithelium, can occur in the larynx. Chronic systemic diseases, medications, dehydration, allergies, and gastroesophageal reflux are common causes of chronic laryngitis. Treatment requires correction of the underlying disorder.

Gastroesophageal reflux disease and LPR disease are common causes of pediatric hoarseness. Laryngopharyngeal reflux is defined as reflux of gastric or duodenal juices beyond the esophagus into the larynx, oropharynx, or nasopharynx. Recent pediatric literature tends to identify LPR as a distinct disease process, not as an extension of GERD. Chronic cough, vomiting, dysphagia, globus sensation, bitter taste, and laryngospasm are common presentations. Children with radiographic evidence of GERD often demonstrate LPR findings as described above. In one study, endoscopic changes found in LPR include lingual tonsil hyperplasia (69%), postglottic edema (83%), arytenoid edema and loss of arytenoid architecture (55%), ventricular obliteration (39%), and true vocal cord edema (67%). A total of 66% showed improvement with speech and pharmacotherapy.

Large interobserver variability exists in endoscopic findings of LPR. For example, vocal cord nodules can be confused with inflammation and edema of vocal cords seen from LPR. In nodules, the vocal cord should be sharply defined anterior and posterior to the nodule. In contrast, the edema seen in LPR has been termed “pseudo nodules”; it is not sharply defined and often found in the anterior one-third of the cord.

Treatment options for LPR include lifestyle modifications, medical therapy, and surgical therapy. Lifestyle modifications include avoiding diet triggers, postural positioning, and altering food composition with thicker feeds. If lifestyle modifications are insufficient, first-line therapies are proton pump inhibitors (PPIs). Esomeprazole has been shown to provide the greatest benefit, but in children lansoprazole and omeprazole are the only PPIs approved by the Food and Drug Administration. Histamine-2 receptor antagonists are second line, and are helpful for weaning off of PPIs. If symptoms are life threatening or the child has failed maximal medical therapy, Nissen fundoplication may be considered.

**FUNCTIONAL VOICE DISORDERS**

Dysphonia in the absence of an underlying structural or neurologic pathology is known as a functional voice disorder. Abnormal contraction of laryngeal musculature compensating for another abnormality can lead to functional dysphonia. Previous literature has reported
an incidence of 4 to 7% of children presenting with dysphonia. Causes of increased laryngeal tone include vocal misuse, maladaptation following respiratory infection, inflammation, compensation for underlying glottic insufficiency, and psychological etiologies. Treatment of functional dysphonia focuses on speech therapy and rebalancing the laryngeal mechanism to produce normal voice. Children with psychogenic dysphonia typically have a normal cry, laugh, and cough, and is a diagnosis of exclusion, with treatment targeting the underlying disorder. Puberphonia is the failure of the male voice to change to lower pitch in adulthood. It can be considered as muscle tension dysphonia of adolescent males and even long-standing cases can often be resolved in a single therapy session.

CONCLUSION

The care of the child with a voice disorder requires a multidisciplinary approach involving otolaryngology, speech language pathology, and other disciplines, all of which contribute to the diagnosis and management. Previous beliefs that pediatric hoarseness would resolve with time is no longer valid. With current technological, pharmaceutical, and therapeutic advances, we can better diagnose and treat the pediatric patient with a hoarse voice. The body of literature continues to grow as health care providers become increasingly knowledgeable about the pathophysiology and management dilemmas that were once troublesome in the pediatric patient.

REFERENCES

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