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Glottic stenosis

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ABSTRACT

Glottic stenosis is a fixed, focal narrowing at the level of the laryngeal inlet, the true vocal cords. It may be either congenital or acquired and be related to a wide range of etiologies. The stenosis may be either anterior, posterior, or in rare cases, complete. Isolated glottic stenosis is rare; lesions often involve adjacent regions, namely the subglottis. A diagnosis is made from careful history and examination, including evaluation by microlaryngoscopy and bronchoscopy. The management of glottic stenosis is challenging and should be tailored to each individual case. A secure and adequate airway is the treatment priority alongside optimization of voice and laryngeal competence. Endoscopic and open techniques in either single or multiple stages have been described.

Introduction

Glottic stenosis describes a fixed, focal narrowing of the upper airway at the level of the true vocal cords. It may result from a wide range of etiologies and be either congenital or acquired. It may occur in isolation or in combination with either supra- or subglottic stenosis.

Posterior glottic stenosis is most commonly associated with trauma related to endotracheal intubation while anterior glottic stenosis is often either congenital or post-traumatic. The management of glottic stenosis is complex and may present a significant surgical challenge. The three key treatment concerns are airway, voice, and laryngeal competence.

Relevant anatomy and embryology

The superior boundary of the glottis is the junction between the true and false cords, the level of the laryngeal ventricle. The caudal boundary of the glottis is less well defined. It may be defined as the horizontal plane 1 cm below the lateral margin or apex of the laryngeal ventricle; however, a revised definition of a horizontal plane 1 cm caudal to the superior aspect of the free edge the true vocal cord has been suggested given difficulty in discerning the anatomical apex of the ventricle.[1,2]

The rima glottidis is the opening between the true vocal cords and the arytenoid cartilages. It can be subdivided into 2 parts: the posterior intercartilaginous portion between the arytenoid cartilages (respiratory glottis) and the anterior intermembranous portion between the true vocal folds (glottis vocalis).

The anatomical structure of the larynx forms by recanalization of the lumen of the developing fetal airway. This occurs around the end of the third month of gestation after completion of normal epithelial fusion. If the laryngeal lumen is not recanalized, laryngeal atresia results. Partial and incomplete recanalization may result in a spectrum of stenosis or webbing.[3] It is of note that abnormal development of the cricoid cartilage may coexist.[4]

Etiology

Congenital

Congenital laryngeal stenosis is rare and reflects a failure of the laryngeal lumen to recanalize during fetal development. Varying degrees of abnormality range from thin anterior glottic webs to complete stenosis in very rare cases.

Laryngeal atresia

Laryngeal atresia may be also described as congenital high airway obstruction syndrome (CHAOS). A prenatal diagnosis of laryngeal atresia may be suspected due a pattern of ultrasonographic findings that result from fetal upper airway obstruction. These include dilation of the trachea, bilateral hypoechoic enlargement of the fetal lung fields, flattening or eversion of the diaphragm, and ascites alongside polyhydramnios. Cardiac compression may occur in severe cases. These features are not, however, universal and may not occur when an associated tracheoesophageal fistula is present.

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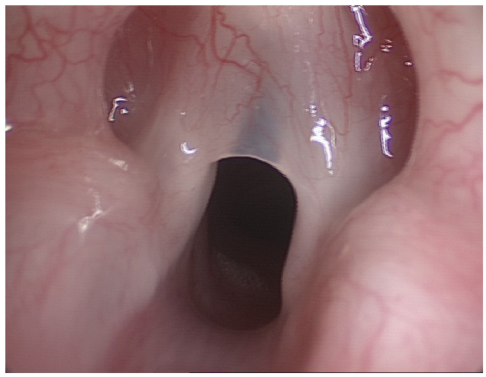

Fig. 1. Endoscopic photograph of congenital laryngeal web.

Laryngeal webs

Congenital laryngeal webs have been estimated to represent 5% of congenital laryngeal anomalies.[5] They are most commonly found anteriorly and may vary significantly in size and severity (Figure 1). Small, membrane-like webs are rarely seen while more severe webs extend into the subglottis in a wedge shape. It should be noted, however, that a subglottic stenosis can also occur independently of the web due to abnormality at the level of the cricoid. Cohen has classified the extent of anterior glottic stenosis from grade 1 to 4 (Table 1).[6]

Glottic webs may be associated with deletions of chromosome 22q11.2. This genetic abnormality is linked to range of phenotypes including velo-cardio-facial (Shprintzen) syndrome and DiGeorge syndrome. In addition, Fraser syndrome is a rare genetic disorder characterized by cryptophthalmos, syndactyly, and laryngeal atresia or webs. Investigation of other congenital abnormalities should be considered in the pediatric patient presenting with either a laryngeal web or atresia.

Congenital vocal cord paralysis

Risks factors for development of congenital vocal cord paralysis include traumatic delivery, Arnold–Chiari malformation, and intracranial abnormalities; however, in a large proportion of cases the etiology is not well understood. Neonates may present with evidence of upper airway obstruction including stridor soon after delivery; approximately 50% will require a tracheostomy. Spontaneous recovery may also be seen in around 50% of cases, supporting a theory of underlying delayed neurological development. This recovery is, however, unpredictable and may occur after a period of months to years.[7]

Acquired

Infective causes

The vast majority of laryngeal infections, both viral and bacterial, do not result in laryngeal stenosis. Particular exceptions include tuberculosis (TB) of the larynx and diphtheria, and occur principally in areas of endemic disease. Rare instances of laryngeal stenosis secondary to typhoid, syphilis, leprosy, and scarlet fever have been described. Mycosis of the larynx has also been observed in an immunocompromised child; this may respond well to systemic anti-fungal treatment.

Tuberculosis commonly affects the post-cricoid region, posterior commissure, and posterior aspect of the true vocal cords. Both supraglottic and subglottic involvement may also be seen and in rare cases, widespread laryngeal disease. The clinical appearance may mimic squamous cell carcinoma and may appear as an ulcerated or exophytic lesion at either a single or multiple sites. If is of note that glottic TB may occur in isolation or alongside pulmonary TB. Significant laryngeal destruction and deformation may occur with resultant glottic stenosis despite appropriate medical treatment.

Table 1
Cohen classification of laryngeal webs.

Type	% Of glottic area
1	< 35%
2	35–50%
3	50–75%
4	75–90%

Traumatic causes

The majority of acquired glottic stenoses are due to trauma relating to endotracheal intubation. Mucosal necrosis and ulceration of the posterior glottis may be caused by endotracheal tube pressure. Localized infection and inflammation can follow with perichondritis and chondritis of the posterior laryngeal cartilages; this may be reflected by development of granulation tissue, characteristically over the vocal process of the arytenoid cartilage. Subsequent localized fibrosis can result in contraction, scar formation, and potential fixation of the cricoarytenoid joints.

Several factors are known to increase the likelihood of laryngeal trauma and subsequent scarring in the intubated patient; these include cycles of repeated intubation and extubation, prolonged intubation, extra-esophageal reflux, excessively large endotracheal tube size, and significant movement of the endotracheal tube. Figure 2 shows acute laryngeal changes related to prolonged endotracheal intubation; posterior laryngeal mucosal ulceration and multiple granulations crowding the laryngeal inlet are demonstrated.

Other causes of mucosal and laryngeal framework injury with resultant stenotic scarring include external trauma, both blunt and penetrating, and ingestion of caustic substances. Prompt management of laryngeal fractures with attention to restoration of the laryngeal framework and preservation of mucosal coverage is key to prevention of subsequent stenosis.

Aggressive endolaryngeal surgery, particularly in the region of the anterior commissure or simultaneously involving both vocal cords may result in significant mucosal injury with subsequent fibrosis and scarring between areas of anatomical proximity. Vulnerable areas therefore include the anterior and posterior commissures. Repeated surgery for recurrent respiratory papillomatosis, particularly with laser use is also associated with a high risk of stenosis.

Inflammatory causes

Glottic stenosis of an inflammatory cause is very rarely seen in the pediatric population. Systemic diseases such as granulomatosis with polyangiitis and sarcoidosis are associated with laryngeal stenosis, principally of the subglottis; however, glottic stenosis has been described. Mucosal diseases including epidermolysis bullosa, pemphigoid, and major aphthous ulceration may also cause laryngeal stenosis.

Acquired vocal cord paralysis

The left recurrent laryngeal nerve is more vulnerable to injury in its longer intrathoracic and cervical course. It may be damaged during either neck or cardiothoracic surgery, for example, ligation of a persistent ductus arteriosus (PDA) or tracheoesophageal fistula repair. Unilateral vocal cord paralysis does not typically result in a significant narrowing of the glottic airway but may have marked impact on voice and laryngeal competence.