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Review

Advances in the diagnosis, management, and treatment of neonates with laryngeal disorders

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SUMMARY

Neonatal airway obstruction due to laryngeal pathology may cause significant morbidity and even mortality. The most frequently occurring etiologies anatomically from superiorly to inferiorly include: laryngomalacia, saccular cyst, vocal fold paralysis, anterior glottic web, laryngeal atresia, laryngeal cleft, subglottic stenosis, and subglottic hemangioma. The pathophysiology, presentation, and treatment options for each of these entities are discussed with a focus on a multidisciplinary, evidence-based approach.

1. Introduction

Respiratory issues secondary to laryngeal abnormalities in neonates may cause significant morbidity and be life-threatening. This makes timely recognition and treatment of neonatal laryngeal pathology essential. This article reviews the pathophysiology, diagnostic considerations, and management options for the most frequently occurring laryngeal disorders encountered during the neonatal period.

2. Laryngomalacia

Laryngomalacia is the most frequent cause of stridor in newborns. Short aryepiglottic folds narrow the soft cartilage of the infantile epiglottis so that it resembles the Greek letter omega (Ω) (Fig. 1). During inspiration, the epiglottis is pulled inward to intermittently obstruct the glottis, causing stridor. Respiratory effort against a closed glottis is believed to draw acid reflux up the esophagus, which then leads to edema of the soft tissue overlying the arytenoid cartilages. This edematous tissue also becomes drawn into the glottis to worsen airway obstruction. Immature neuromuscular control may additionally contribute to supraglottic collapse. This condition should not be confused with laryngeal anomalies in various syndromes such as CHARGE (Coloboma, Heart defects, Atresia choanae, Retarded growth, Genitourinary anomalies, Ear deformities), whereby a long tubular epiglottis and large arytenoid cartilages also lead to airway obstruction [1].

Newborns with laryngomalacia typically present within the first six weeks of life with stridor that is worse when supine or agitated and slightly improved while prone or in an upright position. A substantially increased work of breathing may cause the caloric demand to outstrip supply, resulting in failure to thrive. In severe cases, respiratory distress, evidenced by pronounced retractions, apnea and cyanosis, may occur. Neurologically impaired children may have progressive symptoms.

The diagnosis of laryngomalacia must be confirmed by laryngoscopy, which is most usually performed using a flexible nasopharyngoscope. Neonates should be scoped in the position that recreates their stridor. Careful attention must be paid to the timing of the stridor: stridor that coincides with supraglottic indrawing is likely to be laryngomalacia, whereas stridor that does not coincide with supraglottic indrawing is likely due to a more distal tracheal anomaly. Approximately 80% of neonates with severe laryngomalacia have been found to have secondary airway lesions, the most frequent being subglottic stenosis and tracheomalacia [2]. As such, atypical and severe cases warrant consideration of rigid bronchoscopy.

Observation is appropriate for mild to moderate laryngomalacia because the natural history is resolution of symptoms by 12–24 months. Early on, close follow-up with weekly weights plotted on a normalized growth curve is essential (without a diaper and using the same scale each time). For neonates with cyanotic episodes or

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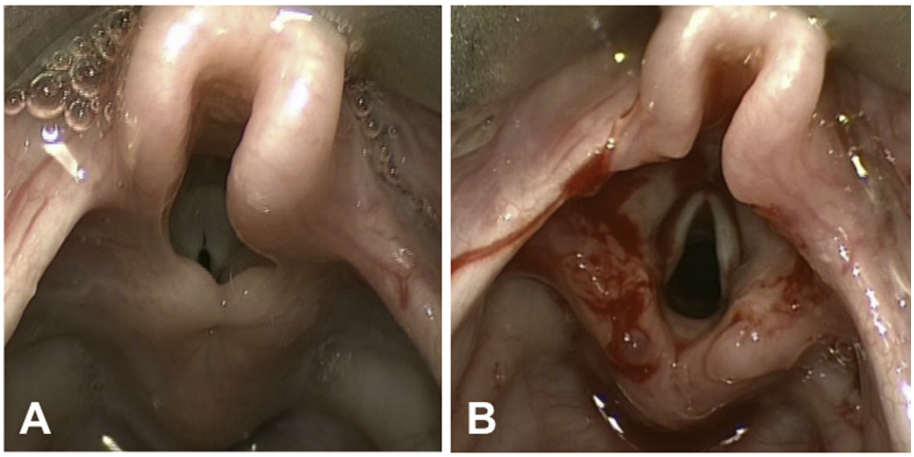

Fig. 1. Laryngomalacia. (A) Prior to surgery, shortened aryepiglottic folds create an omega-shaped epiglottis. (B) Following supraglottoplasty, released aryepiglottic folds allow the epiglottis to unfurl.

who are failing to thrive, supraglottoplasty under general anesthesia is the treatment of choice (Fig. 1). Roughly 10% of patients with laryngomalacia will develop severe enough symptoms to warrant surgical intervention. The larynx is exposed using suspension microlaryngoscopy and the patient may be kept spontaneously breathing or may be nasotracheally intubated with a small endotracheal tube. Supraglottoplasty may be performed with scissors or a laser. Patients are observed overnight postoperatively in the intensive care unit either with or without an endotracheal tube. Children with associated comorbidities such as neurologic impairment (Down syndrome), cardiac issues, or gastroesophageal reflux disease (GERD) have an increased risk of surgical failure and postoperative aspiration [3]. If the patient is not intubated, thickened oral feeds may be initiated slowly on the first postoperative day. Patients with CHARGE syndrome should not undergo supraglottoplasty because disruption of the aryepiglottic folds causes collapse of the long tubular epiglottis leading to complete airway obstruction [1].

Because supraglottic edema seen in laryngomalacia may be due to acid reflux, many clinicians initiate proton pump inhibitors (PPIs). The intent of treatment is to break the cycle of edema obstructing the glottis, leading to increased negative pressure in the esophagus that worsens acid reflux and subsequently increases edema. Although the use of PPIs in laryngomalacia has not formally been investigated, anecdotal reports suggest that many infants gain weight within one to two weeks of starting medication, thus obviating the need for surgery. PPIs should be given on an empty stomach 1 h before a feed to ensure that they have taken effect before gastric parietal cells become active during the next meal. Second-line medications such as H2 blockers and pro-motility agents may be necessary for more severe acid reflux.

3. Saccular cyst

The laryngeal saccule communicates with the laryngeal ventricle and extends superiorly to occupy the potential space between the false vocal fold and the inner lamina of the thyroid cartilage. Numerous mucous glands line the saccule to keep the larynx lubricated. Ductal obstruction within the saccule may lead to formation of a mucous-filled saccular cyst. Saccular cysts are typically lined with squamous or respiratory epithelium. Rarely, they may also contain mesodermal elements that are suggestive of a laryngeal duplication [4]. Extralaryngeal extension into the neck either through the thyrohyoid or cricothyroid membrane have been reported [5].

Saccular cysts present with stridor and respiratory distress in the neonatal period and may necessitate intubation immediately after birth. Intubation may be challenging because distorted supraglottic anatomy may preclude the endoscopist from obtaining a view of the glottis. Furthermore, complete airway obstruction may occur on induction if paralysis is used and laryngeal tone is lost.

The gold standard for diagnosing saccular cysts is rigid laryngoscopy under general anesthesia (Fig. 2). However, they may be visible using flexible nasopharyngoscopy in an awake, stable neonate. Magnetic resonance imaging (MRI) or computed tomography may be considered to define the extent of the lesion.

Endoscopic marsupialization is the first-line treatment for most saccular cysts. The patient is kept spontaneously breathing or is intubated with a small nasotracheal tube. Microlaryngeal scissors or a laser are used to unroof the cyst so that its contents will drain into the pyriform sinus rather than the larynx. For recurrent cysts causing persistent airway obstruction or cysts with extensive extralaryngeal extension, an open approach with laryngofissure may be required [5].

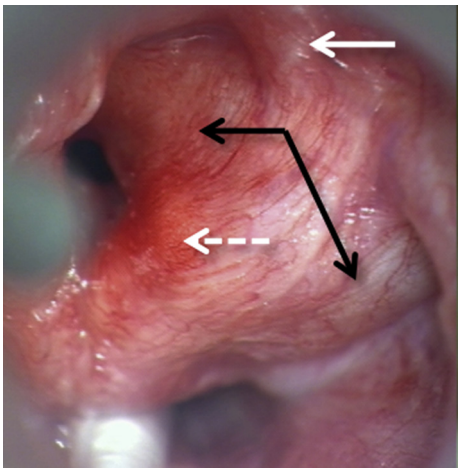

Fig. 2. Right-sided congenital saccular cyst. The cyst is seen as a submucosal fullness (black arrows) deep to the aryepiglottic fold that travels from the epiglottis (solid white arrow) to the arytenoid cartilage (dashed white arrow).