Laryngomalacia

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KEYWORDS

• Stridor • Laryngomalacia • Upper airway obstruction • Supraglottoplasty
• Aryepiglottoplasty

KEY POINTS

- Laryngomalacia is the most common congenital laryngeal anomaly, accounting for up to 70% of patients who present with stridor. Most cases are mild and self-resolve, but severe symptoms require investigation and treatment.
- Laryngomalacia presents as a spectrum of disease, from mild intermittent stridor to life-threatening airway compromise.
- There is a strong association of laryngomalacia with gastroesophageal reflux disease (GERD), which warrants medical treatment of GERD in many cases.
- In children with severe laryngomalacia, supraglottoplasty is the preferred surgical option, which can achieve improvement in both airway and feeding symptoms.
- Laryngomalacia can play a role in sleep-disordered breathing and obstructive sleep apnea.

Videos of flexible fiberoptic laryngoscopy and supraglottoplasty accompany this article at http://www.pediatric.theclinics.com/

INTRODUCTION: NATURE OF THE PROBLEM

Laryngomalacia is the most common congenital laryngeal anomaly and accounts for 60% to 70% of cases of stridor in neonates and infants.[1] The physical finding of stridor is a manifestation of upper airway obstruction caused by collapse of the supraglottic tissue because of excess mucosa, and abnormal and/or reduced laryngeal tone. Symptoms generally become apparent after the first 2 weeks of life, and, in most cases, resolve between 12 and 18 months of age. Most cases resolve with minimal or no treatment; approximately 10% of cases require surgical intervention.

The term laryngomalacia, or soft larynx in Latin, replaced the more antiquated term congenital laryngeal stridor, which had previously been used to describe the

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condition. First coined by Jackson and Jackson[2] in 1942, the term differentiated the condition from other causes of stridor and more clearly depicted the flaccidity of the larynx.[3]

THE INFANT LARYNX: ANATOMIC CONSIDERATIONS

A review of laryngeal anatomy aids in the understanding of pathophysiology in laryngomalacia. The structure of the larynx is divided into 3 areas: the supraglottis, glottis, and subglottis. Laryngomalacia affects the supraglottic structures, which include the portions of the larynx above the level of the vocal cords. Important supraglottic structures that can be involved in laryngomalacia include the epiglottis, arytenoid cartilages, and aryepiglottic folds (which connect the epiglottis to the arytenoids).

Neonatal and infantile larynges have several important differences from those of older children and adults. At birth, the position of the larynx is higher than in older children and adults.[4] The growth of the larynx is accelerated during the first 3 years of postnatal life and gradually achieves its final shape. Postnatal descent of the hyoid and larynx is unique to humans. The high position of the larynx at the time of birth facilitates transition to spontaneous breathing and prevention of aspiration, and this also accounts for obligate nasal breathing in neonates. The descent of the larynx is crucial for appropriate development of speech. Major postnatal changes to the larynx occur in the first year of life.

The infantile epiglottis is longer than the laryngeal length in older children, which may predispose it to posterior displacement. The cartilage of the infantile larynx is also more pliable than that of the larynx later in life, a property that has been proposed to play a role in the collapsibility of the laryngeal airway.

CLINICAL PRESENTATION

Inspiratory stridor is the primary feature of laryngomalacia. Characterized by a harsh, high-pitched sound, stridor as it presents in laryngomalacia often worsens while the infant is supine, feeding, or crying. Feeding difficulties also often accompany the presence of stridor in patients with laryngomalacia, because the delicate balance between the suck-swallow sequence and respiration is often disrupted.[5] Coughing, choking, regurgitation with feedings, and slow oral intake are all common symptoms. There is thought to be a close relationship between laryngomalacia and gastroesophageal/laryngopharyngeal reflux, although the exact mechanism has yet to be fully elucidated.

Other causes of stridor must also be considered when evaluating a patient with these symptoms (**Table 1**). One of the most useful ways to differentiate between causes of noisy breathing is to identify in which phase of the respiratory cycle the sound is heard. Different causes of airway obstruction can lead to stridor during different phases of respiration: inspiration, expiration, or both (biphasic).

Stridor present during inspiration is usually caused by partial obstruction at the level of the supraglottic tissues. Variable extrathoracic obstruction results in primarily inspiratory stridor. During inspiration, atmospheric pressure is greater than extrathoracic intraluminal airway pressure, leading to collapse of supraglottic structures. During the expiratory phase, the exhalatory breath increases extrathoracic airway pressure such that it overcomes the collapse. Stridor primarily present during expiration is usually caused by obstruction in the lower tracheal airway. Negative intrathoracic pressure during inspiration allows air movement into the lungs, but the increase in intrathoracic pressure during expiration causes affected portions of the tracheal airway to collapse, leading to expiratory stridor.