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Mini-symposium: Upper Airway Abnormalities

Laryngomalacia: Review and Summary of Current Clinical Practice in 2015

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EDUCATIONAL AIMS

The reader will be able:

- To discuss the etiology, prevalence and clinical presentation of laryngomalacia.
- To discuss the use of conservative treatment for children with mild-moderate laryngomalacia.
- To review surgical indications for supraglottoplasty and discuss surgical goals, outcomes and peri-operative care.
- To discuss treatment alternative treatment options for children who fail supraglottoplasty or are not appropriate surgical candidates.

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SUMMARY

Laryngomalacia is the most common cause of stridor in neonates and infants. Associated feeding difficulties are present in approximately half of the children. A definitive diagnosis can generally be made with flexible fiberoptic laryngoscopy. The disorder is most often self-limited with resolution of symptoms within the first 24 months of life, and the majority of children can thus be managed conservatively. The approximately 5%-20% of children with severe or refractory disease may require more aggressive intervention, most commonly in the form of trans-oral supraglottoplasty [1,2]. High success rates and a low rate of complications have been reported for this procedure in otherwise healthy children. Children with syndromes or medical comorbidities are more likely to have complications or persistent symptoms after supraglottoplasty and may require additional interventions.

DEFINITION AND PRESENTATION

Laryngomalacia is defined as collapse of supraglottic structures during inspiration, resulting in intermittent airflow impedance and associated stridor. It is the most common cause of stridor in neonates and children, accounting for 60-70% of cases [3].

The characteristic high pitched inspiratory stridor associated with laryngomalacia is not always present at birth but generally becomes apparent by several weeks of age. Symptoms may worsen over the first 4-8 months of life. The stridor is often exacerbated by agitation, crying, feeding, upper respiratory tract infections or supine positioning. The stridor commonly diminishes or resolves during sleep in mild to moderate cases. In approximately 5%-20% of children, respiratory concerns such as obstructive sleep apnea, tachypnea, dyspnea, respiratory distress or hypoxemia can occur. In the most severe cases, progression to pulmonary hypertension and cor-pulmonale can occur without appropriate treatment [4]. Feeding difficulties are present in approximately half of children with mild to moderate laryngomalacia and virtually all children with more severe disease, and can include coughing and

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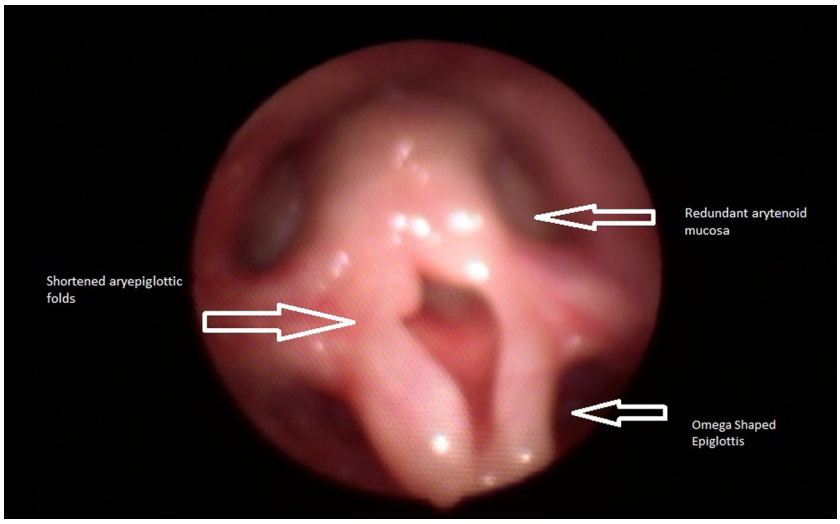

Figure 1. Fiberoptic view of child with laryngomalacia during expiration. Note the typical findings including omega-shaped epiglottis, shortened AE folds and redundant arytenoid mucosa obstructing view of vocal folds.

choking, cyanotic episodes, regurgitation, emesis or slow feeding. In more severe disease, recurrent aspiration pneumonia or failure to thrive from decreased caloric consumption and heightened metabolic demand from increased work of breathing occurs [5].

ETIOLOGY

Historically, it was thought that laryngomalacia represented an anatomic abnormality of the laryngeal cartilage. This theory was supported by a prospective study showing a lower aryepiglottic (AE) fold to glottic length ratio in patients with severe laryngomalacia compared with unaffected children [6], but failed to explain why some children with similar laryngeal examinations were asymptomatic. The theory of immature and abnormally collapsible cartilage was further discredited by histologic examinations demonstrating normal fibro-elastic cartilage tissue in children with symptomatic disease [7]. Recently, attention has focused on a neuromuscular etiology, consisting of immaturity or abnormal integration of the peripheral nerves, brainstem nuclei and pathways responsible for swallowing and maintenance of airway patency. This is supported by physiologic studies in infants with laryngomalacia demonstrating increased stimulus threshold requirements for elicitation of normal motor responses correlating with disease severity [4]. Additional corroboration is provided by histologic studies showing significant size differences in the superior laryngeal nerve branches of patients with severe laryngomalacia compared to age matched controls [8]. Subsequent neurological and central nervous system maturation would provide a reasonable explanation for the spontaneous resolution generally seen in the disease.

DIAGNOSIS

History and Examination

A presumptive clinical diagnosis of laryngomalacia can be made based on the classic symptoms of inspiratory stridor worsened by feeding, agitation, supine positioning or crying. Pertinent history should include birth circumstances (including gestational age and endotracheal intubation), congenital or genetic abnormalities, respiratory symptoms with aggravating or temporizing factors, and feeding concerns including retarded growth, choking or gagging, reflux symptoms, or recurrent pneumonia. Physical examination should include height and weight, respiratory sounds including timing in the respiratory cycle, chest movement to determine the presence of retractions or pectus excavatum, and auscultation of the lung fields.

A definitive diagnosis of laryngomalacia can be made accurately by flexible fiberoptic laryngoscopy alone in the vast majority of cases (88%), regardless of the experience level of the examiner [9]. The procedure can generally be performed on awake children in the arms of their caregiver without the need for sedation. The flexible fiberoptic laryngoscope is passed along the nasal floor and positioned above the larynx during several cycles of spontaneous respiration. Topical anesthetics should be avoided when possible as they may exacerbate airway collapse and alter the examination [10]. Characteristic findings include inspiratory supraglottic collapse with poor visualization of vocal cords due to shortened AE folds, collapse of arytenoids into airway, edema of the posterior glottis or a curled (omega-shaped) or retroflexed epiglottis. (Figures 1 and 2) Of note, the severity of stridor or symptoms does not reliably correlate closely with the extent of collapse on flexible fiberoptic examination.

Several anatomical staging systems for laryngomalacia have been proposed which focus on defining the site of collapse including:

1. posterior collapse (from redundant arytenoid mucosa or cuneiform cartilage)
2. lateral collapse (from shortened AE folds)
3. anterior collapse (from a retroflexed epiglottis)
4. combined collapse (involving multiple areas of anatomic collapse)

Although none of these systems has achieved universal acceptance to date, consistent and detailed documentation of the type and severity of collapse seen on flexible laryngoscopy provides crucial information when contemplating surgical manipulation.

Alternately, iterations of symptom based grading systems are frequently used in clinical practice to stratify disease severity and help inform the timing or need for more aggressive intervention (Table 1) [11].

Additional Diagnostic Evaluations

Associated conditions, most notably reflux disease or synchronous airway lesions (SALs), have been commonly reported in