

**TITLE: Current Concepts in Diagnosis and Management of Laryngomalacia**

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## **Introduction**

Laryngomalacia is the leading cause of stridor in infancy. It is the most common laryngeal anomaly of the newborn followed by subglottic stenosis and bilateral vocal fold paralysis. It is characterized by flaccidity of the supraglottic structures, which prolapse inward during inspiration leading to upper airway obstruction.

## **Etiology**

Review of literature suggests that there are three basic theories that attempt to explain the development of laryngomalacia. These are

- 1) Cartilage Immaturity Theory
- 2) Theory of Anatomic Abnormality and
- 3) Neuromuscular Immaturity Theory.

## **Cartilage Immaturity Theory**

Cartilage Immaturity Theory was first proposed by Sutherland and Lack in the late 19<sup>th</sup> century. According to this theory laryngeal flaccidity develops due to delayed maturation of the cartilagenous support of the larynx. This theory has since been disproved because there is no evidence of chondropathy in these patients. According to the Theory of Anatomic Abnormality, laryngomalacia is an exaggeration of the normal anatomic features of an infant larynx. The infant larynx is softer, more pliable, and easily prone to mucosal edema. There can often be an omega-shaped epiglottis. Exaggeration in one or more of these anatomic features may lead to laryngomalacia. The most accepted theory is that of neuromuscular immaturity which suggests that laryngeal hypotonia occurs due to delayed neuromuscular control of the larynx. The prevalence of other neurological disorders in infants with laryngomalacia supports this hypothesis.

## Association with Reflux

It has been found that there is a strong association of GERD with laryngomalacia and studies have found that 80-100% of infants with laryngomalacia will also have reflux. It is unclear however, whether GERD is a cause or an effect of the respiratory obstruction. It has been proposed that the fixed respiratory obstruction in laryngomalacia leads to a large negative intrathoracic pressure, which in turn leads to reflux and worsening laryngeal edema and obstruction. In addition, the vagal tone to the lower esophageal sphincter may be reduced contributing to the reflux. These observations underscore the importance of empiric reflux therapy in patients with laryngomalacia.

## Risk Factors

Risk factors for severe laryngomalacia include:

- 1) Prematurity- Although premature infants do not necessarily have a higher incidence of laryngomalacia, they tend to develop the more severe form of this condition.
- 2) Neuromuscular disorders- As mentioned earlier, there is a higher incidence of neuromuscular disorders in patients with laryngomalacia and the condition tends to be severe in such patients.
- 3) Synchronous airway lesions- These can be seen in 10-20% of patients with laryngomalacia. Tracheomalacia and subglottic stenosis have been reported to be the commonest lesions, though bronchomalacia, pharyngomalacia and an associated vallecular cyst may also occur. The presence of synchronous airway lesions potentiates GERD and also increases the risk of surgical failure.

## Classification of Laryngomalacia

There are various types of laryngomalacia and various classification schemes have been attempted to correlate type with severity and treatment. For the sake of our presentation, we have classified laryngomalacia into mild, moderate or severe depending upon history, physical exam and flexible laryngoscopy findings. One can describe the location of the laryngomalacia as **anterior** (the epiglottis prolapses posteriorly into the airway), **lateral** (AE folds and the cuneiform or corniculate cartilages prolapse medially into the airway) and **posterior** (the redundant mucosa over the arytenoids prolapses anteriorly into the airway). Laryngomalacia may present as a combination of these subtypes.

## Making the Diagnosis

The majority of the infants are diagnosed in the first few weeks of life. High pitched inspiratory stridor is the hallmark of this condition and the stridor is worsened with activities such as crying, feeding or being placed in the supine position. Reflux and feeding symptoms such as choking, recurrent emesis, prolonged feeding time, cough and weight loss may also be seen. 10-20% of patients with laryngomalacia may present with complications such as life threatening airway obstruction, severe weight loss, failure to thrive, severe apnea, cyanosis, pulmonary hypertension, developmental delay and cardiac failure.

Diagnosis of this condition is primarily clinical. A combination of history, physical examination and flexible laryngoscopy is sufficient to make a diagnosis in the majority of cases. Flexible laryngoscopy should be carried out in an awake child to assess the type and severity of laryngomalacia, look for synchronous airway lesions, hunt for signs consistent with GERD, and also to assess vocal fold mobility. It is important to realize that flexible laryngoscopy does not allow adequate visualization of the subglottis or trachea. Complementary studies may be done in some cases. Chest radiographs can aid in evaluation of potential lower respiratory anomalies such as tracheomalacia, innominate artery compression, vascular rings, and signs of aspiration. An esophagram may be done to document the extent and degree of reflux but can also help rule out a concomitant motility disorder or extrinsic compression from vascular structures. Sleep studies should be done if the presenting symptoms are sleep apnea or if symptoms fail to improve after surgery. A pH probe study should be done prior to consideration of a Nissen's fundoplication for reflux.

## **Management Strategies**

For the sake of this presentation, management of a child with laryngomalacia is divided into medical and surgical treatment. Medical management includes empiric reflux therapy, feeding modifications and posture repositioning. Empiric reflux therapy may be started with H2 receptor antagonists (ranitidine 3mg/kg three times daily) or proton pump inhibitors (1mg/kg daily). In more severe cases, 6mg/kg of ranitidine at night along with 1mg/kg of a PPI daily may be required. Feeding modifications include pacing, thickening formula feeds, upright feeding position, and giving the infant small, frequent meals. All parents are instructed on following reflux precautions. These help the baby feed adequately and maintain weight.

Laryngomalacia is a self limiting condition in 85-90% of patients. Absolute indications for surgery include cor pulmonale, pulmonary hypertension, hypoxia, life threatening airway obstruction, apnea, recurrent cyanosis, failure to thrive, pectus excavatum, and stridor with significant retractions. Relative indications include aspiration, difficulty feeding a child who has failed medical intervention and weight loss with feeding difficulty. One simple measure is to routinely plot these children on the growth curve to ensure they are gaining weight appropriately. There are no absolute contraindications for surgery. Weight and age are not contraindications for surgery. The surgeon should proceed with caution in patients with laryngomalacia and other neurological disorders or patients who have multiple levels of obstruction because these conditions are associated with a higher failure rate. Preoperative counseling is a must and the parents should be advised regarding improvement in feeding and stridor. Though the feeding improvement is early and often dramatic, the stridor may take a longer time to resolve. Parents are also advised about the risks and benefits of surgery, including the possibility of postoperative intubation, overnight ICU stay, abnormal scarring and the risk of revision surgery. Parents and primary care physicians must be reminded about the importance of continuing reflux precautions and reflux medications post-operatively.

Supraglottoplasty is the most common procedure for laryngomalacia practiced today. This surgery is designed to trim the AE folds and remove the excess mucosa over the arytenoids. The procedure is carried out under general anesthesia with the child breathing spontaneously. The surgical set up is the same as that for suspension microlaryngoscopy. It is important to assess the tracheobronchial tree and vocal fold mobility prior to the procedure. The surgery may

be performed with cold instruments, CO2 laser, micordebrider or a combination of these. While incising the AE folds, it is important not to extend the incision beyond the anterior edge of the arytenoids and the incision should not involve the pharyngoepiglottic fold. While removing the mucosa over the arytenoid cartilages, one should be gentle and diligent to preserve the interarytenoid mucosa to prevent supraglottic stenosis. Any bleeding during the procedure is easily controlled with afrin pledgets.

Epiglottopexy is a relatively new procedure indicated if the level of obstruction is at the level of the epiglottis. Some authors have presented this as the treatment for any patient with laryngomalacia with good surgical outcomes. In this procedure, the mucosa over the lingual surface of the epiglottis is denuded either with cold instruments or more recently with CO2 laser in a linear fashion. Important complications associated with this surgery include aspiration and supraglottic stenosis.

Tracheostomy is rarely performed today for laryngomalacia alone. Specific indications for tracheostomy include presence of > 3 comorbidities, severe sleep apnea documented on a sleep study and persistence of symptoms after revision supraglottoplasty.

Post-operative care includes overnight stay in the hospital, monitoring of airway symptoms and feeding improvement. The child may feed as soon as he is awake. Postoperative steroids may be given and reflux therapy is continued. The parents are asked to follow up in 2-4 weeks time and monitor the child for any worsening symptoms.

Complications after surgery are relatively uncommon (8%). Site-specific complications include bleeding, infection, web formation, and granulation tissue.

The most feared complication is supraglottic stenosis which is difficult to treat. This can be avoided by meticulous and very conservative surgery. In some cases, it may prove useful to operate on only one side at a time.

## **Summary**

Laryngomalacia is the commonest cause of “stridor” in the newborn. It is a self-limiting condition in the vast majority of cases, and patients with this condition expect symptom improvement and resolution of stridor by 2 years of age. It is vital for parents to know that the condition may worsen around 4-6 months of age before it gets better. High pitched inspiratory stridor is the hallmark clinical presentation. Feeding difficulties and GERD are seen in 80-100% of patients with laryngomalacia. Diagnosis is usually established by history, physical examination and flexible laryngoscopy findings. Empiric reflux therapy is indicated for many patients and the more severe cases may benefit from a combination of medical treatment and surgery. Surgical success in selected patients with severe laryngomalacia is >90% in several studies. In unusual or worsening cases of “laryngomalacia”, the otolaryngologist should have a low threshold for recommending a formal airway evaluation in the operating room.

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