“LARYNGOMALACIA”

EMBRYOLOGICAL BASIS AND ITS CLINICAL SIGNIFICANCE

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ABSTRACT
Laryngomalacia is the most common cause of stridor in infants. Stridor results from upper airway obstruction due to supraglottic tissue collapse. Most causes of laryngomalacia are mild and self solve investigation and intervention required in severe cases. There is a strong association with gastroesophageal reflux disease in patients with laryngomalacia and thus medical treatment with antireflux medications surgical treatment is preferred, while it indicate supraglottoplasty, severe cases are reserved. Proper identification of those patients when requires medical and surgical invention is key to providing with successful outcomes.

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Introduction
Laryngomalacia is a congenital disorder of larynx. The term “malacia” means softness of larynx. It is noted in the young children’s in the month of four to eight. This condition cause noisy breathing in infancy, due to the tissue to fall over the airway and partially block it, when laryngeal structure malformed and floppy. Laryngomalacia may also affects the epiglottis or the arytenoid cartilages, sometime the both. When epiglottis involved it is often elongated, and the lesion has been referred to as an omega shaped epiglottis. If the arytenoids cartilages are involved, they appear enlarged [1].

The inspiratory obstruction causes an inspiratory noise, which may be high-pitched sounds frequently heard with the other causes of stridor coarse sounds resembling nasal congestion and low pitched stertorous noises. Infants with laryngomalacia have a higher incidence of gastro-esophageal reflux. It presumably results in negative intrathoracic pressure for necessary to overcome the inspiratory obstruction [2]. Occasional inflammatory changes are observed in the larynx, which is referred as reflux laryngitis. Laryngomalacia may presents with the three degrees like mild, moderate and severe. About 99 percent of infant with laryngomalacia have mild and moderate laryngomalacia [3].

The first indication of the future lower respiratory system appears in the 4 mm embryo, the primitive pharyngeal floor just behind the pharyngeal pouches, earlier in 4th week of embryo, as a longitudinal groove, the laryngotracheal groove externally it is seen as ridge. The endodermal lining of the laryngotracheal groove forms the epithelium and glands of the larynx, trachea, bronchi, and pulmonary lining epithelium [4]. The splanchnic mesenchyme ventral to the foregut will give rise to the connective tissue, the cartilage, and the smooth muscles accompanying these structure. When appearance of groove, optic vesicle and auditory placode are already present, pharyngeal pouches are formed, oropharyngeal membrane (stomodeum and foregut) is disintegrating and the vessels that form heart fused in the single tube.

The laryngotracheal groove deepens and, with development, the external ridge grows caudally below the pharynx to become a diverticulum, the tubular lung bud.

1. As the diverticulum grows from pharyngeal floor, it is investing by splanchnic mesenchyme. The cranial part of the tube becomes the laryngeal epithelium; the caudal part forms the epithelium of the lower respiratory system.

2. As the diverticulum grows, it becomes separated from the pharynx by a partition the tracheoesophageal septum, which divides the foregut into the laryngotracheal tube and the esophagus. The laryngotracheal tube and surrounding splanchnic mesenchyme give origin to the larynx, the trachea, the bronchi and the lungs.

3. When the tubular lung bud forms, it develops two knob like enlargements at its distal end, the so called bronchial buds[5].

The larynx develops from the endodermal lining of cranial end of the laryngotracheal tube and surrounding mesenchyme. The mesenchyme proliferates to produce paired arytenoid swellings, giving the primitive glottis a T shaped appearance and reducing the laryngeal lumen to a slit.

- The laryngeal cartilages develop within the arytenoid swellings from the cartilages bars of the branchial arches.
- The epiglottis develops from the caudal half of the hypobranchial eminence derivative of branchial arches 3 and 4 [6].

The laryngeal inlet ends blindly, between weeks 7 to 10, because of the fusion of epithelium, but as the epithelium breaks down, the laryngeal aditus enlarges and recanalizes. A pair of lateral recessed the laryngeal ventricles form which are bound cranially and caudally by anteroposterior folds of mucus membrane, the future vestibular (false) and vocal (true) folds membrane, respectively [7].
Ontogenesis of normal development of larynx and trachea

Knowing the development of the larynx is of prime importance to understand how the congenital anomalies appear clinically and how they should be managed. The development of larynx can be divided into the prenatal and postnatal stages. At birth, the larynx is located high in the neck between the C1 and C4 vertebrae, allowing concurrent breathing or vocalization and deglutition [9]. By age 2 years, the larynx descents inferiorly by age 6 years, it reaches the adult position between C4 and C7 vertebrae. This new position provides a greater range of phonation (because of the wider supraglottic pharynx) at the expense of losing third separation of function, deglutition and breathing [9].

The larynx develops from the endodermal lining and the adjacent mesenchymal of the foregut is first identifiable with a funnel shape to a win-

The thyroid cartilage develops from bilateral chrondrificational centers of the fourth branchial arch, and the cuneiform cartilages develop from the sixth branchial arch. The superior laryngeal nerve emantes from the fourth branchial arch, becoming evident by day 33. By day 37; the recurrent laryngeal nerve derived from the sixth branchial arch becomes evident [13].

Beginning at age 18-24 months, the larynx starts descents down to the neck to achieve its final position at the cricoid cartilages changing from interstitial to perichondrial growth [15].

The main changes occurring in the larynx postnatally are a change in the axis luminal shape, length, and proportionally growth of the laryngeal elements. The larynx grows rapidly during the first 3 years of life, while the arytenoids remain approximately the same size. The arytenoids in the adult larynx are thus proportionately smaller than in the child larynx [16].

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Ontogenesis of abnormal development of larynx and trachea

Laryngomalacia also known as congenital laryngeal stridor, although this condition is self-limiting and usually resolves within 12 to 18 month, the symptomatology may become severe enough to warrant surgical interventions. Stridor associated with laryngomalacia typically develops within the first few weeks of life and progresses in severity over a period of a few months. Stridor typically is described as highly pitched and fluttering in nature.
Severe cases of stridor may be associated with sternal restriction, respiratory distress, and feeding difficulties[26]. The condition usually is exacerbated by exertion, crying feeding agitation or supine positioning, placing the infant in the prone position or on his or her side and extending the neck may relieve the stridor .approximately 80% of cases are associated with gastroesophageal reflux .this condition also may be associated with failure to thrive. Pectus excavatum is the common on associated finding that tends to improve when the airway obstruction resolves .resolution usually occurs by 18 months of age. Some patients however, may remain symptomatic until age 5 .other synchronous laryngotracheal anomalies associated with laryngomalacia have been reported, including subglottic stenosis and vocal cord paralysis [25].

The causes of laryngomalacia is unclear ,although altered embryologic development of the larynx is the most likely cause of the abnormal anatomic findings .histologic studies have failed to demonstrate any inherent cartilaginous abnormality leading to the increase flaccidity and abnormal collapse of the supraglottic larynx .abnormal neurologic function also has been suggested as a factor[24].

Although laryngomalacia typically is described as a congenital anomaly, there have been cases where symptoms associated with laryngomalacia do not develop until several months after birth, in addition, children with severe neurologic impairment and pharyngeal and hypopharyngeal hypotonia may develop a chronic acquired flaccidity of the supraglottic larynx with similar symptomatology.

The gold standard in diagnosis for laryngomalacia is flexible laryngoscopy[23] .with the patient awake ,under dynamic condition history and physical examination is also important to reach a correct diagnosis of laryngomalacia can be made using radiographic studies, this modality is best suited as an adjunct to evaluation for associated laryngomalacia anomalies. In the severe cases for which operative intervention is considered, rigid endoscopy in operating room is warranted.

Typically, with flexible laryngoscopy the larynx demonstrated an omega shaped epiglottis (that may be occur in 30% to 50% of normal asymptomatic infants .redundant aryepiglottic (AE) folds ad excessive tissue in the supraraytenoid area that may be prolapsed into the laryngeal inlet upon inspiration. A deep interarytenoid cleft ,or possibly a posterior laryngeal cleft, may be seen on flexible endoscopy, but is best as accessed using rigid endoscopy. Vocal cord paralysis also may be ruled out simultaneously by this technique. Laryngomalacia also may be site specific. There may be evidence of primarily posterior laryngomalacia with prolapsed of supra arytenoids tissue noted, or possibly prolapsed of anterior aspect of the AE folds and epiglottis into the airway (anterior laryngomalacia) [17].

In the majority of patients, laryngomalacia resolves and no surgical intervention is required .encouraging the family to have the child sleep on his or her side and not in the supine position helps relieve symptomatology and is safer for the child [18]. Surgical intervention should be considered in patients with severe respiratory distress, failure to thrive, severe obstructive apnea, or other severe symptomatology [19].

Recently a new condition has been described known a disco ordinate pharyngolaryngomalacia. This is a condition that has been noted in patients originally diagnosed with laryngomalacia who did not respond well to surgical intervention[20] .this condition is associated with severe laryngomalacia manifested by complete supraglottic collapse during inspiration without shortened AE folds or redundant mucosa and with accompanying pharyngomalacia. Many of these patients require further surgical intervention, including tracheotomy [21]. Some may respond however, to bi-level positive airway pressure (BiPAP) management to avoid tracheotomy [22].

**Fig 2. Omega shaped epiglottis affected condition of larynx (laryngomalacia).**

**Discussion**

Laryngomalacia is a common cause of stridor in infants. There is inspiratory collapse of lax supraglottic tissue into the airway causing respiratory obstruction. The stridor worsens with feeding, crying, supine position, and agitation [16]. There may be regurgitation, coughing, choking or failure to thrive. Those with signs and symptoms of severe airway obstruction require urgent airway examination to determine the cause and secure the airway.hollinger’s classification is used for the various types of laryngomalacia --type 1 is anterior prolapsed of the arytenoid and corniculate cartilages; type 2 is tubular epiglottis which cues on itself, often associated with type 1 ; type 2 is anteromedial collapse of the arytenoids; type 4 is posterior prolapse of the epiglottis ; and type 5 is short aryepiglottic folds[15].

Zero-degree laryngoscopy, flexible laryngoscopy, or bronchoscopy is recommended for diagnosis for direct visualization of airway anatomy and dynamics. This should be done by team OF pediatric otolaryngologist, intensive care specialty, and anesthetist in an ICU or operation theater setup. It is important to maintain spontaneous ventilation while anesthetizing these children to allow complete dynamic assessment of the airway to confirm diagnosis. Proper identification of patients who require medical and surgical intervention is key to providing successful treatment. The criteria for severe laryngomalacia with surgical indication include that haing clinical evidence of severe respiratory obstruction, and/ or pectum excavatum, and those having swallowing disorders children fulfilling these criteria and requiring airway support with weaning failure are ideal candidates for surgical intervention without delay [27].

Earlier, tracheostomy was the only treatment available to avoid fatal events. At present, better surgical techniques such as tracheal supraglottoplasty and epiglottopexy have evolved.
Current procedures depend on individual anatomic and functional alteration on a patient to patient basis.

Supraglottoplasty were first described in 1987 by zal zal et al. in a case series of ten patients having severe laryngomalacia. It has now become the mainstay of surgical management and involves laryngoscopic resection of excess and tax mucosa from epiglottis, aryepiglottic folds, arynoids, and corniculate cartilages, thus laryngeal inlet during inspiration. Carbon dioxide laser and microbebrider assisted supraglottoplasty introduced later were the effective first line treatment for severe type 1, and 3 laryngomalacia bilateral supraglottoplasty may also be performed. Failures and complications of supraglottoplasties include need for revision surgery vocal cord granuloma, edema, webs, and supraglottic stenosis [28].

Glossoepiglottoplasty is another relatively new surgical strategy which is simple, nonexpensive and effective. It is useful in type 4 laryngomalacia with isolated posterior displacement of epiglottis where the obstruction is due to epiglottis inhalation. Two nonabsorbable sutures are taken bilaterally from the base of the tongue to epiglottis one on each side[29]. Thus, this prevents collapse of epiglottis over the glottis opening, however, swallowing problems, aspiration, or reflux may occur postoperatively[30].

The success of these procedures is clinically measured by (postoperative) reduced requirement for airway support, resolution of feeding difficulties, weight, and stature gain[31].

Thus though laryngomalacia usually resolves spontaneously withinth the 1 st year of life (median time to resolution in isolated laryngomalacia being approximately 36 weeks), surgical intervention may be needed in 15-20% of infants with severe, life threatening laryngomalacia. These cases should be evaluated by flexible fiberoptic bronchoscopy or zero degreee laryngoscopy to plan further management. With proper selection, glossoepiglottopotomy may prove to be a good, simple, and effective management option, whereby tracheostomy can be avoided[32].

Conclusion

Laryngomalacia is a common disease of infancy where the diagnosis is suspected by primary care providers based on history [33]. Those with mild disease can be managed expectantly. Continued monitoring of the symptoms is necessary as symptoms can progress over the natural course of the disease. Recognizing patient factors and symptoms associated with moderate and severe disease helps determine which infants will benefit from otolaryngology consultation. Identification patient factors that influence disease severity and outcomes is an important aspect of counseling care givers and providing care to infants with laryngomalacia [34].

References