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Chapter

Laryngomalacia

Vaishali Waindeskar, Pooja Thaware, Garima Chamania, Anuj Jain and Ashutosh Kaushal

Abstract

Laryngomalacia is the most common cause of stridor in neonates and infants. In laryngomalacia, there is a supraglottic collapse of the larynx during inspiration leading to obstruction and thus resulting in stridor. The exact etiology of laryngomalacia is still unknown. The neurological basis is one of the leading theories explaining the etiology. Laryngomalacia in most of the patients resolves with conservative management by two years of age. In severe cases of laryngomalacia or when symptoms are persistent beyond two years of age, such cases need surgical management in the form of supraglottoplasty. Flexible fibreoptic laryngoscopy is the gold standard for the diagnosis of laryngomalacia. Various classifications have been proposed to classify laryngomalacia, although considering dynamic airway changes might be the most acceptable basis for classification. Supraglottoplasty has higher success and a low complication rate.

Keywords: flexible fiberoptic laryngoscopy, Laryngomalacia, larynx, stridor, Supraglottoplasty

1. Introduction

Laryngomalacia is the most common cause of stridor in neonates and infants. In fact, laryngomalacia is the common cause of stridor in 60–70% of newborns and infants which makes laryngomalacia the most common congenital laryngeal anomaly [1]. Supraglottic collapse produced by certain anatomic variants causes airway obstruction in Laryngomalacia, which is most severe during inspiration. Flexible fiberoptic laryngoscopy is used to diagnose laryngomalacia. The general course is benign, with stridor progressing for 6 months until gradually disappearing by 12–24 months of age. The majority of cases resolve with minimal or no treatment. Of all the laryngomalacia patients, only ten to fifteen percent patients will have significant upper airway obstruction symptoms, including increased breathing effort, feeding difficulties, and failure to thrive. Supraglottoplasty is recommended in such severe situations.

The larynx is subdivided into three parts supraglottis, glottis, and subglottis. The supraglottis is the part between the inferior boundary of the hyoid bone and the vestibular folds. Therefore, the structures present in the supraglottis area are the epiglottis, arytenoid cartilages, and aryepiglottic folds. Laryngomalacia affects these supraglottic structures.

The pediatric airway is different from that of adults in many ways. The relatively large tongue in relation to the oropharynx, therefore pediatric patients are more likely to sustain airway obstruction under anesthesia due to relatively large tongue. The larynx is located more cephalic in the neck in neonates and infants. This high cephalic position helps to facilitate spontaneous breathing right after birth and prevents aspiration. The epiglottis is short, stubby and omega-shaped, angled over the vocal cords. Also, the infantile laryngeal cartilage is more flexible than older children and adults, which is a probable cause of collapsible characteristics of the neonatal and infantile larynx.

2. Signs and symptoms

The most common symptom in infants with laryngomalacia is positional stridor. Stridor commonly appears in the first few days of life, although children may not seek medical help until they are many months old. Stridor is usually worse in the supine position, particularly during crying or eating, because such activities necessitate increased respiratory efforts. The severity of symptoms increases during several months of infancy but usually resolves by two years of age and often earlier [2].

Although stridor is a common symptom of laryngomalacia, it is not often the cause of presentation to the medical facility. Sometimes patients with severe disease present with other respiratory complaints such as respiratory distress, use of accessory respiratory muscles and hypoxemia. Laryngomalacia can also present with atypical symptoms like snoring, obstructive sleep apnea and difficulty in swallowing. Although incidence and distribution of these atypical presentations are yet to be established [3]. Difficulty in swallowing causes decreased intake and respiratory distress increases metabolic demand thus can lead to failure to thrive in infants with laryngomalacia.

The natural history of laryngomalacia and that it resolves during the second year of life is supported by the low level of evidence and lacks endoscopic evidence. The time range within which laryngomalacia resolves and the proportion of patients in which laryngomalacia do get spontaneously resolved is yet to be answered. Atypical presentations are to be further explored. Therefore, prospective longitudinal trials are required to better understand the natural history of laryngomalacia [4].

Laryngomalacia in older children presents with obstructive sleep apnea syndrome. Other reasons in older children for seeking medical attention are exercise-induced stridor and dysphagia [5].

3. Etiology

The exact etiology and pathophysiology of laryngomalacia are still unknown. Multiple causal theories of laryngomalacia have been proposed. The neurological basis is one of the leading theories, which states that the abnormal integration of the laryngeal nerves leads to altered laryngeal tone. This theory has been supported by a pathologic study that has shown increased diameter of supraglottic nerve in patients with severe laryngomalacia. Another theory proposes an imbalance of demand and supply during inspiration as a cause of congenital laryngomalacia. This theory of imbalance needs further study. Acid reflux disease has not been established as a cause of laryngomalacia, although almost 60% of infants who present with laryngomalacia

also have accompanying acid reflux disease. Acid reflux disease causes irritation and thus edema of the upper airway which further worsens laryngeal obstruction.

In the scenario of debated etiologies of laryngomalacia, a neurological basis is considered as the most probable etiology of laryngomalacia among all the theories mentioned before. Another case report supporting the neurological basis is a unique case report where a child who was diagnosed case of moyamoya disease suffered acquired laryngomalacia following a neurologic insult. The child suffered a cerebrovascular accident following which she developed laryngomalacia presenting with severe stridor and chest retractions and a nocturnal oxygen requirement, and severe laryngomalacia being noted on laryngoscopy. Prior to the cerebrovascular accident, she had no symptoms of laryngomalacia and had undergone several laryngoscopies, both awake and anesthetized, which showed no evidence of laryngomalacia [6].

Children with laryngomalacia showed vitamin D deficiency and increased proinflammatory cytokine IL-6, which may result from dysregulation of the immune responses. Laryngomalacia could be an inflammatory disease secondary to maternal deficiency of 25(OH)-vitamin D with subsequent Vitamin D deficiency in exclusively breast-fed infants during neonatal and infantile periods [7].

4. Factors that influence disease severity

The patient factors that influence disease severity are Apgar score at birth and during the first several minutes after birth, resting oxyhemoglobin saturation level at the time of presentation, and the presence of a secondary airway anomaly. Additional co-morbidities in children with laryngomalacia increases disease severity and also affect the prognosis of surgical outcome. Such co-morbidities can be gastroesophageal reflux disease, laryngopharyngeal reflux, neurologic disease, congenital heart disease, genetic syndrome, or anomaly.

Patients with severe laryngomalacia will require surgery. Patients who have gastroesophageal reflux disease or laryngopharyngeal reflux and one additional co-morbidity are more likely to require revision supraglottoplasty. Those with three medical co-morbidities are more likely to require tracheostomy [8].

5. Classification of laryngomalacia

There are a number of classifications of laryngomalacia. A simple and well-detailed classification of laryngomalacia by Olney et al. describing type 1, type 2, and type 3 laryngomalacia as prolapsing arytenoids, shortened aryepiglottic folds, and prolapsing epiglottis [9]. The classification of Olney et al. is simple, but covers only about two-third of laryngomalacia cases and also mixes static and dynamic findings.

Van der Heijden et al. after studying various laryngomalacia classifications have proposed a Groningen Laryngomalacia Classification System (GLCS) which is based on the photo and video documentation of eighty five patients diagnosed in a tertiary referral centre combined with a review of the literature [10].

This simplified system is supposed to ease communication among professionals and provide a base for treatment algorithms. In laryngomalacia, there is a collapse of the supraglottic airway during inspiration causing obstruction. This is a dynamic change of the airway happening during inspiration. Some previous classifications

were based on static findings. Anatomical findings such as omega-shaped epiglottis, short aryepiglottic folds and acutely angled epiglottis over laryngeal inlet are static findings. In laryngomalacia obstruction is due to dynamic change in the airway, and static findings do not completely explain these dynamic changes and therefore are not the excellent choice for the classification of laryngomalacia. McSwiney et al. were the first to introduce a system in order to classify laryngomalacia and it was based on static findings [11]. McSwiney et al. further combined omega-shaped epiglottis and posterior displacement in one single type of laryngomalacia, suggesting omega shaped epiglottis is exclusively associated with posterior displacement of the epiglottis. Although, omega-shaped epiglottis can also present in conjunction with medial displacement of aryepiglottic folds during inspiration. Holinger et al. divided laryngomalacia into six different types, with static and dynamic findings described as separate entities. Shah et al. exclusively described dynamic changes but the definitions of the different types of laryngomalacia were insufficiently described [12]. Kay et al. use a system in which Type 1 is defined as static finding and Type 2 as a dynamic finding and type 3 is a collection of "all other etiologies" including neuromuscular disease, which renders the classification less reliable [13]. None of these systems discussed here are widely accepted. Classifications require simplification in such a way that they not only provide a genuine classification based on dynamic findings but also allow making a righteous decision for the intervention required.

The Groningen laryngomalacia classification system is a newly proposed classification system exclusively based on dynamic laryngeal changes. In Groningen's laryngomalacia classification, laryngomalacia is divided into three types; Type 1 is inward collapse of arytenoid cartilages, Type 2 is medial displacement of aryepiglottic folds, and Type 3 is posterocaudal displacement of epiglottis against the posterior pharyngeal wall.

The GLCS has also proposed the probable surgical intervention for each category required. They suggested that the decision between surgical management and conservative strategy should be based on the severity of laryngomalacia, and when surgical management is planned, the GLCS can suggest the surgical intervention required. The surgical intervention suggested for Type 1 laryngomalacia of GLCS is the removal of redundant mucosa over arytenoids with or without the removal of cuneiform or corniculate cartilage. In Type 2 laryngomalacia suggested intervention is incision or excision of a wedge of aryepiglottic folds. In Type 3, epiglottopexy would be the surgical intervention required. However, these treatment options should be clinically individualized for each patient.

Kay et al. also provided a classification-based treatment algorithm. They discerned three types in which type I is recommended to be treated by dissection of the aryepiglottic folds, type II by resection of redundant mucosa over arytenoid cartilage, but cases with "all other etiology" were supposed to receive tracheostomy.

6. Diagnosis

A thorough physical examination of the infant should be performed, with special attention to the oral cavity, nose, and neck. A complete birth history is required, including any surgical history or intubations performed on the patient. Parents should inform about any breathing problems children may have at home, with a

focus on noisy breathing or apnea episodes. Laryngomalacia is characterized by noisy breathing that worsens with meals or while lying supine. The clinician should investigate the patient's eating habits and keep track of any weight loss or failure to thrive.

It's important to ensure choanal patency and rule out piriform aperture stenosis. A complete oral cavity examination is required to rule out cleft lip or cleft palate, glossoptosis, Pierre-Robin sequence, or micrognathia, all of which can cause breathing and feeding difficulties. A thorough examination of the neck is also required to rule out any tumors or vascular abnormalities. Hemangiomas with a beard-like distribution should be given special attention, as these infants are more prone to have hemangiomas in the airway. In order to properly evaluate a patient with suspected laryngomalacia, a flexible laryngoscopy examination of the supraglottic airway in an awake newborn is required. The infant should be transferred to the operating room for a diagnostic bronchoscopy if the examiner notices serious symptoms.

Flexible fiberoptic laryngoscopy is the mainstay in the diagnosis of infant stridor. It permits the real-time visualization of the aerodigestive tract during spontaneous ventilation. It allows complete visualization of the oropharynx, hypopharynx, supraglottis, glottis and subglottis. Due to the simplicity and ability to thoroughly examine the dynamic collapse of the supraglottic airway during awake respiration, flexible fiberoptic laryngoscopy is presently the gold standard for the diagnosis of laryngomalacia.

Direct laryngoscopy and diagnostic bronchoscopy in the operation theater give the clinician a complete evaluation of the upper aerodigestive tract to the level of carina and the mainstem bronchi. It is a valuable procedure principally in patients with severe symptoms or in patients who have concomitant secondary airway anomalies. Surgical intervention is also possible with direct laryngoscopy when warranted.

Radiologic studies might be helpful in the diagnosis of swallowing difficulty. A modified barium swallow examination is preferred in infants with laryngomalacia since aspiration may be silent and not detectable clinically.

A polysomnogram is beneficial in determining the presence and severity of obstructive sleep apnea, particularly in older children. To improve the apnea-hypopnea index in such children surgical intervention like supraglottoplasty might be beneficial.

Airway fluoroscopy due to low sensitivity and exposure to ionizing radiation is not advocated in the assessment of infant stridor.

7. Management

Most of the children of laryngomalacia can be managed conservatively as the symptoms usually disappear by the end of the second year of life. In the majority of cases, laryngomalacia is a self-limiting condition. Only 5–20% of children with severe laryngomalacia undergo surgical intervention. As mentioned before transoral supraglottoplasty has a low complication in otherwise healthy children [14]. To reduce inspiratory obstruction in laryngomalacia, redundant tissue in the upper airway is cut and/or the aryepiglottic folds are loosened in bilateral supraglottoplasty. While bilateral supraglottoplasty is generally well tolerated, about 10% of individuals experience side effects such as laryngeal edema, new-onset aspiration, or supraglottic stenosis. Supraglottic stenosis is a life-threatening condition that is difficult to treat surgically.

A few clinicians have reported performing unilateral supraglottoplasty to lessen the risk of problems associated with bilateral surgery.

For the treatment of severe laryngomalacia, Walner et al. advocated a staged approach to bilateral supraglottoplasty [15]. Staged supraglottoplasty implies a unilateral supraglottoplasty on the most affected side, followed by, if necessary, an opposite-side operation week to months later. For the surgery, they used cold steel instruments, with or without the use of a microdebrider.

In the staged approach first stage of surgery involves the removal of the redundant tissue on the most affected side using either straight or curved microscissors. Then to release the ipsilateral aryepiglottic fold a small wedge of tissue was removed. Microdebrider can also be used to remove redundant tissue overlying the arytenoid cartilage on the most affected side, and then a small wedge of tissue from the aryepiglottic fold on the same side can be removed after using microscissors. Afrin-soaked pledgets were used on the cut surfaces to reduce bleeding. Walner et al. further describe that after surgery all patients were extubated and monitored for 24 hours prior [15]. Patients were reevaluated for symptoms after 4 to 6 weeks. If the problems of breathing or feeding continued, the second step of surgery was employed to provide further relief. An opposite-side supraglottoplasty was performed, with redundant arytenoid mucosa excised and the aryepiglottic fold released on the side opposite to the original surgery. If aryepiglottic fold on the side previously operated appears to be too tight it was re-released. According to Walner et al., 73% of individuals who underwent the first stage of surgery had considerable improvement or resolution of stridor, while 100% of those who underwent the second stage had significant improvement or resolution of stridor. There were no complications in any of the patients.

Low-temperature plasma radiofrequency ablation (LTP-RFA) is another surgical therapy option. According to the severity of laryngomalacia, Hongming Xu et al. presented the first prospective four-arm randomized trial to compare the efficacy and short outcomes of patients with moderate and severe laryngomalacia who were randomly treated with LTP-RFA, traditional surgical supraglottoplasty, or wait-and-see policy [16]. When compared to typical surgical supraglottoplasty, LTP-RFA treatment dramatically reduced operating time, length of hospital stay, and amount of intraoperative hemorrhage in children with severe laryngomalacia, but treatment efficacy was equivalent. In addition, when compared to the control group, LTP-RFA treatment dramatically alleviated laryngomalacia symptoms in children with moderate laryngomalacia. Post-operative pneumonia was the most common consequence, affecting 11% of patients.

8. Anesthesia

Choice of ventilation strategy is the main concern for the anesthesia team in the case of laryngomalacia. The following are the ventilation strategy that can be utilized [17]:

a. Spontaneous breathing is the choice of ventilation with experienced anesthesia and surgeon teams.

b. Controlled mechanical ventilation using a small internal diameter endotracheal tube is nowadays used rarely, as the tube interferes in the vicinity of the surgical site.

c. Intermittent apnea technique is another choice for ventilation under anesthesia but surgery needs to be interrupted in between for manual ventilation when the patient desaturates and thus surgeon gets limited time duration in between two ventilations.

d. Jet ventilation can sometimes be used for this surgery.

9. Conclusions

Laryngomalacia despite the fact that it is a self limiting disease, caregivers must recognize severe cases. A child's growth may be hindered in severe cases of laryngomalacia due to breathing and feeding difficulty. Laryngomalacia is resolved in more than 90% of cases after supraglottoplasty, which enhances the child's quality of life. Only in a few cases a second surgery is required to resolve residual symptoms.

Conflict of interest

The authors declare there are no conflict of interest.

Acronyms and abbreviations

GLCS	Groningen Laryngomalacia Classification System
LTP-RFA	Low-temperature plasma radiofrequency ablation

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References

[1] Dobbie AM, White DR. Laryngomalacia. Pediatric Clinics of North America. 2013;**60**(4):893-902. DOI: 10.1016/j.pcl.2013.04.013

[2] Hysinger EB. Laryngomalacia, tracheomalacia and bronchomalacia. Current Problems in Pediatric and Adolescent Health Care. 2018;**48**(4):113-118. DOI: 10.1016/j.cppeds.2018.03.002

[3] Cooper T, Benoit M, Erickson B, El-Hakim H. Primary presentations of laryngomalacia. JAMA Otolaryngology. Head & Neck Surgery. 2014;**140**(6):521-526. DOI: 10.1001/jamaoto.2014.626

[4] Isaac A, Zhang H, Soon SR, Campbell S, El-Hakim H. A systematic review of the evidence on spontaneous resolution of laryngomalacia and its symptoms. International Journal of Pediatric Otorhinolaryngology. 2016;**83**:78-83. DOI: 10.1016/j.ijporl.2016.01.028

[5] Digoy GP, Burge SD. Laryngomalacia in the older child: Clinical presentations and management. Current Opinion in Otolaryngology & Head and Neck Surgery. 2014;**22**(6):501-505. DOI: 10.1097/MOO.0000000000000111

[6] Born H, Wineland A, Rutter MJ. Neurologically acquired laryngomalacia in a pediatric patient with Moyamoya: A case report and literature review. International Journal of Pediatric Otorhinolaryngology. 2019;**116**:34-37. DOI: 10.1016/j.ijporl.2018.10.001

[7] Hassan MM, Emam AM, Mahmoud AM, Awad AH, Rezk I, Abou-Taleb A, et al. Congenital laryngomalacia: Is it an inflammatory disease? The role of vitamin D. Laryngoscope. 2020;**130**(2):448-453. DOI: 10.1002/lary.27997

[8] Thompson DM. Laryngomalacia: Factors that influence disease severity and outcomes of management. Current Opinion in Otolaryngology & Head and Neck Surgery. 2010;**18**(6):564-570. DOI: 10.1097/MOO.0b013e3283405e48

[9] Olney DR, Greinwald JH Jr, Smith RJ, Bauman NM. Laryngomalacia and its treatment. Laryngoscope. 1999;**109**(11):1770-1775. DOI: 10.1097/00005537-199911000-00009

[10] Van der Heijden M, Dikkers FG, Halmos GB. The Groningen laryngomalacia classification system--based on systematic review and dynamic airway changes: Groningen Laryngomalacia classification system. Pediatric Pulmonology. 2015;**50**(12):1368-1373. DOI: 10.1002/ppul.23186

[11] McSwiney PF, Cavanagh NP, Languth P. Outcome in congenital stridor (laryngomalacia). Archives of Disease in Childhood. 1977;**52**(3):215-218. DOI: 10.1136/adc.52.3.215

[12] Shah UK, Wetmore RF. Laryngomalacia: A proposed classification form. International Journal of Pediatric Otorhinolaryngology. 1998;**46**(1-2):21-26. DOI: 10.1016/s0165-5876(98)00111-6

[13] Kay DJ, Goldsmith AJ. Laryngomalacia: A classification system and surgical treatment strategy. Ear, Nose, & Throat Journal. 2006;**85**(5):328-336. DOI: 10.1177/014556130608500514

[14] Zhang YM, Wang ZN, Xia ZF. Research progress of laryngomalacia in children. Lin Chuang Er Bi Yan Hou Tou Jing Wai Ke Za Zhi. 2017;**31**(2):162-166. DOI: 10.13201/j.issn.1001-1781.2017.02.023

[15] Walner DL, Neumann DB, Hamming KK, Miller RP. Supraglottoplasty in infants: A staged approach: A staged approach. The Annals of Otology, Rhinology, and Laryngology. 2015;**124**(10):803-807

[16] Xu H, Chen F, Zheng Y, Li X. Efficacy and toxicities of low-temperature plasma radiofrequency ablation for the treatment of laryngomalacia in neonates and infants: A prospective randomized controlled trial. Annals of Translational Medicine. 2020;**8**(21):1366

[17] Ayari S, Aubertin G, Girschig H, Van Den Abbeele T, Denoyelle F, Couloignier V, et al. Management of laryngomalacia. European Annals of Otorhinolaryngology, Head and Neck Diseases. 2013;**130**(1): 15-21. DOI: 10.1016/j.anorl.2012.04.003