

TITLE: Treatment of Laryngomalacia

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Stridor is a harsh, high-pitched musical sound that results from turbulent airflow through the upper airway. Evaluation of stridor is dependent upon the clinical situation, as the underlying etiology may range from mild illness to a severe and life-threatening situation. When considering a pediatric patient, it should be noted that eighty-five percent of children under the age of 2.5 years presenting with stridor have a congenital etiology. Congenital stridor often times is not present at birth, but will typically present before the age of four months. The remaining cases are accounted for by inflammation, trauma, or foreign bodies. Pediatric stridor has a variable age of onset. The typical patient usually presents with a sudden onset of symptoms. As a basic guideline, acquired stridor is more likely than congenital stridor to require airway intervention.

When assessing stridor, the location of the turbulent airflow may be determined by the respiratory phase in which the sound is noted. There are three zones of the airway that are responsible for these sounds- supraglottic, glottic and subglottic, and intrathoracic. Supraglottic obstruction results in a high-pitched, inspiratory stridor. Obstruction of the extrathoracic trachea, including the glottis and subglottis, is characterized by biphasic stridor with an intermediate pitch. Obstruction of the intrathoracic trachea (including first and second order bronchi) results in expiratory stridor, or wheezing. This last area of obstruction is associated with retraction of the sternum, costal cartilage, and suprasternal tissue.

The differential diagnosis of noisy breathing in children can be grouped into the anatomic location of the lesion or defect. Underlying causes can be classified as congenital, inflammatory, neoplastic, neuromuscular, and traumatic. The following is a list of diagnoses depending on the location of the lesion or defect:

- Nose and nasopharynx-
 - Congenital: choanal atresia or stenosis, pyriform aperture stenosis, craniofacial anomalies
 - Inflammatory: nasal polyps, rhinitis, retropharyngeal abscess, adenoid hypertrophy
 - Neoplastic: encephalocele, dermoid, glioma
 - Traumatic: foreign body
- Oropharynx/hypopharynx-

- Congenital: glossoptosis/macroglossia, lingual thyroid, vallecular cyst, craniofacial anomalies
- Inflammatory: tonsil hypertrophy, retropharyngeal abscess
- Neoplastic: dermoid, hemangioma, lymphangioma
- Neuromuscular: hypotonia, neurologic disease
- Traumatic: foreign body
- Supraglottic larynx-
 - Congenital: laryngomalacia, laryngocele/saccular cyst
 - Inflammatory: epiglottitis, angioneurotic edema
 - Neoplastic: hemangioma, lymphangioma, papilloma
 - Traumatic: foreign body
- Glottic larynx-
 - Congenital: web/atresia, laryngeal cleft, stenosis
 - Inflammatory: laryngitis, spasm, stenosis
 - Neoplastic: hemangioma, lymphangioma, papilloma, granuloma
 - Neuromuscular: vocal cord paralysis
 - Traumatic: hematoma, fracture, foreign body, stenosis
- Subglottic larynx-
 - Congenital: stenosis, cysts
 - Inflammatory: croup, stenosis
 - Neoplastic: hemangioma, papilloma
 - Traumatic: chondritis, stenosis, fracture, foreign body
- Tracheobronchial-
 - Congenital: stenosis/web, tracheomalacia, vascular ring/sling/complete tracheal rings, foregut cysts, tracheoesophageal fistula
 - Inflammatory: membranous tracheitis, bronchitis, asthma
 - Neoplastic: mediastinal tumors, thyroid, thymus, papilloma
 - Traumatic: stenosis, foreign body (tracheal or esophageal)

Laryngomalacia is the most common cause of stridor in infancy. Furthermore, it is the most common congenital laryngeal anomaly. Males are affected twice as often as females. Laryngomalacia arises from a continued immaturity of the larynx. Symptoms of laryngomalacia are often not present at birth. In fact, onset of symptoms typically occurs days to weeks after birth (most commonly within the first two weeks of life) and they will resolve at twelve to eighteen months of age without intervention. The diagnosis is suspected when inspiratory stridor is auscultated and is confirmed by flexible nasopharyngoscopy. Typical stridor associated with laryngomalacia is low in pitch with a fluttering quality secondary to the circumferential rimming of the supraglottic airway and aryepiglottic folds. It is often associated with general noisy respiration. The stridor is most prominent when the child is in the supine position or when the child is agitated. More forceable inspiration tends to result in a louder stridor quality due to greater prolapse and thus greater obstruction.

Radiographic studies can suggest the diagnosis of laryngomalacia, however the mainstay of diagnosis is flexible nasopharyngoscopy. For completeness, we will include radiographic findings. An inspiratory film with neck extension can show medial and inferiorly displaced arytenoids and epiglottis. Fluoroscopy can demonstrate the collapse of the supraglottic structures with inspiration. Chest radiographs can aid in evaluation of potential lower respiratory tract anomalies such as innominate artery compression, tracheomalacia, and vascular rings.

Flexible nasopharyngoscopy is best performed in an unanesthetized child in an upright position with a 1.9mm nasopharyngoscope. The scope should be passed through both nasal passages when examining the patient. Classic findings with flexible nasopharyngoscopy are a cyclical collapse of the supraglottic larynx with inspiration. The aryepiglottic folds are short and often times there is an omega shaped epiglottis. Short aryepiglottic folds draw the cuneiform and corniculate cartilages forward over the laryngeal inlet resulting in prolapse during inspiration. These findings will be reviewed when discussing the five types of laryngomalacia. It should be noted that the vocal cords are mobile in laryngomalacia. Although laryngomalacia can often make visualization of the vocal cords difficult, they still must be examined.

Multiple factors may contribute to the development of laryngomalacia. The main contributors are thought to be anatomic, neurologic, and inflammatory. The underlying anatomic pathologies are shortening of the aryepiglottic folds and anterior collapse of the cuneiform cartilages. Immature neuromuscular control and movement has also been thought to impact laryngomalacia. Additionally, there is an association of gastroesophageal reflux and laryngomalacia. Reflux can induce posterior supraglottic edema and secondarily laryngomalacia.

A recent article published in April 2005 by Dr. SC Manning et al. evaluated the anatomical impact on laryngomalacia. The specific objective of the article was to compare the aryepiglottic length in pediatric patients who have severe laryngomalacia and are undergoing aryepiglottoplasty with the aryepiglottic length of a sample of control patients without laryngomalacia. The study design was a prospective case-control. Measurements were compared by creating a ratio of the aryepiglottic fold length (distance from the most anterior arytenoids cartilage to the closest posterior lateral edge of the epiglottic cartilage) to the glottic length (distance from the anterior border of the interarytenoid muscle to the anterior commissure). The mean ratio of patients with severe laryngomalacia was calculated to be 0.380, while the mean ratio of control patients was calculated to be 0.535.

There are five types of laryngomalacia. Type I is an inward collapse of the aryepiglottic folds (mainly the cuneiform and corniculate cartilages). These cartilages are drawn inward during inspiration and open passively during expiration. Type II is a long, tubular epiglottis which curls on itself and contributes to obstruction during inspiration. This often occurs in association with Type I laryngomalacia. Type III is anterior, medial collapse of the cuneiform and corniculate cartilages to occlude the laryngeal inlet during inspiration. Type IV is posterior inspiratory displacement of the epiglottis against the posterior pharyngeal wall or inferior collapse to the vocal folds. Type V is short aryepiglottic folds.

The requirement of surgical intervention in patients with laryngomalacia is rare, as it is generally a self-limiting condition. Severe symptoms such as inability to feed orally, cor pulmonale, failure to thrive, and life threatening airway obstruction may necessitate surgical intervention. Prior to the 1980s, tracheotomy was used in this setting. The rationale for this intervention was that a tracheotomy bypassed the area of obstruction until the supraglottic pathology spontaneously resolved. Today, this strategy is only employed in the severely affected infant. Instead, the strategy has changed to address the area of obstruction directly with a supraglottoplasty. This can be performed by making use of microlaryngeal instruments, a carbon dioxide laser, or a microdebrider to excise redundant mucosa and/or cuneiform cartilages as well as releasing the shortened aryepiglottic folds and trimming the lateral edges of the epiglottis. It is important to note that there is more than one obstructing mechanism in laryngomalacia and

when performing supraglottoplasty, all potential mechanisms should be addressed. When removing tissue, the surgeon should be conservative, as supraglottic stenosis can result from excessive tissue removal. Unilateral supraglottoplasty should be considered and the second side should be operated on only if symptoms do not resolve. Also, direct laryngoscopy and bronchoscopy must be performed prior to supraglottoplasty to rule out concomitant pathology contributing to the airway obstruction. The direct laryngoscopy should be performed when the patient is breathing spontaneously under general anesthesia. Post-operatively, the patients are usually left intubated over night with extubation the following morning. Antibiotics should be given at least five days post-operatively to prevent infection. Antireflux precautions including both positioning and medication are recommended to minimize the raw mucosal surfaces exposure to gastric secretions. Below is a review of the literature

The original description of endoscopic removal of supraglottic tissue with a nasal snare for laryngomalacia was first described in 1922 by Dr. S Iglauer. In 1984, Dr. RW Lane et al. described a single report of the removal of the corniculate cartilages and redundant arytenoids mucosa in a child with laryngomalacia. In 1985, Dr. AB Seid et al. described the use of the carbon dioxide laser for treatment of laryngomalacia in a series of three patients.

In 1987, GH Zalzal et al. presented epiglottoplasty as a new procedure. His data included a case series of ten patients. He described a laryngoscopic approach in which redundant mucosa was excised from the lateral edges of the epiglottis, aryepiglottic folds, arytenoids, and corniculate cartilages. This was done with the patient in the supine position while visualizing the larynx through a laryngoscope. The tissue to be excised was grasped with cup forceps and trimmed with Bellucci scissors. Dr. Zalzal reported that “all patients who underwent epiglottoplasty achieved complete relief from their symptoms with the exception of one patient in whom mucosa from the aryepiglottic folds and arytenoids area had not been adequately excised. Further excision resulted in complete relief.” His indications for operating on a patient with laryngomalacia were severe stridor not resolved with time that may be associated with failure to thrive, cor pulmonale, feeding difficulties, and apnea. Additionally, inability to view the vocal cords with a flexible nasopharyngoscope during inspiration because of laryngeal inlet collapse is a clear indication for surgery.

In 1995, G Roger, et al. published a retrospective study of 115 cases of resection of the aryepiglottic folds with or without a carbon dioxide laser. The success rate was determined to be 98% with an average follow-up period of thirty months. Failure of the procedure was assigned to two children who needed tracheotomies. Seven patients required a revision surgery. In this paper, Dr. Roger established criteria for the definition of severe laryngomalacia:

1. dyspnea at rest and/or severe dyspnea during effort
2. feeding difficulties
3. height and weight growth rate stagnation
4. sleep apnea or obstructive hypoventilation
5. uncontrollable gastroesophageal reflux
6. history of intubation for obstructive dyspnea
7. effort hypoxia (10% higher than the normal values for the same age group)
8. effort hypercapnia (10% higher than the normal values for the same age group)
9. abnormal polysomnography with an increased apnea/obstructive hypoventilation index

The presence of at least three of these criteria was deemed to be a formal indication for

endoscopic surgery.

Dr. SM Kelly et al. evaluated the effectiveness of unilateral endoscopic supraglottoplasty for treatment of severe laryngomalacia in 1995. The study design was a retrospective review of eighteen patients with severe laryngomalacia that had undergone unilateral carbon dioxide laser removal of redundant supraglottic tissue. Three of the eighteen patients required a contralateral supraglottoplasty. Relief of obstructive symptoms was achieved in 94% of the patients. The one patient without obstructive relief had tracheomalacia secondary to previous tracheotomy for severe laryngomalacia.

In 1999, DR Olney et al. performed a retrospective chart review aimed at determining the airway outcome of infants with laryngomalacia who do not undergo routine direct laryngoscopy and bronchoscopy, the age at which laryngomalacia resolves, and the outcome of supraglottoplasty as a function of the type of laryngomalacia and the presence of concomitant disease. Dr. Olney found that direct laryngoscopy and bronchoscopy as part of the routine evaluation of laryngomalacia is not warranted and should only be performed when there is clinical and physical evidence of a concomitant airway lesion. The median time to resolution of isolated laryngomalacia was 36 weeks, and by 72 weeks, 75% of infants were free of stridor. This finding did not vary significantly in infants with severe neurological compromise or in infants with other congenital anomalies. Supraglottoplasty was determined to be necessary in approximately 15-20% of affected infants which is attributed to episodes of apnea and failure to thrive.

A chart review was performed in 2001 by CW Senders et al. to evaluate different carbon dioxide laser procedures on children with various types of laryngomalacia and determine the role of associated anomalies on the outcome. Twenty-three children were included in a retrospective chart review that underwent carbon dioxide laser vaporization of redundant supraglottic mucosa of the aryepiglottic fold, arytenoids, and the epiglottis. Patients without associated anomalies did well, with 78% immediately resolving their respiratory symptoms and 100% resolving their respiratory symptoms within a week. Unfavorable immediate results, as well as long term surgical failures all had associated anomalies including Arnold-Chiari, Cerebral Palsy, CHARGE association, and Rieger syndrome. Conclusions determined by Dr. Senders were that laser supraglottoplasty is a safe and effective treatment for all types of laryngomalacia, but children with associated neurological or anatomic anomalies will have a more complicated immediate and short term course, as well as a significant incidence of failure.

A retrospective review was performed in 2001 by SC Toynton et al. looking at one hundred patients that had endoscopic aryepiglottoplasty performed for severe laryngomalacia. Surgery was used for treatment of patients that had oxygen saturation below 92% and feeding difficulties causing failure to thrive. Ninety-four percent of the patients had improvement of their stridor after one month, with 55% completely without stridor. Patients with slower progression of improvement were found to have a serious neurological condition. Seventy-two percent of patients with preoperative feeding difficulties improved their feeding. Dr. Toynton concluded that endoscopic aryepiglottoplasty is the operation of choice for severe laryngomalacia. However, underlying neurological disease decreases the success rate of operative intervention.

In 2001, Dr. DK Reddy et al. performed a retrospective review to evaluate the efficacy of unilateral supraglottoplasty compared to bilateral supraglottoplasty. This study included 106

patients- 59 patients undergoing bilateral supraglottoplasty and 47 patients undergoing unilateral supraglottoplasty as the initial surgery. Surgical success was defined as the resolution of clinically significant laryngomalacia and was reported to approach 96% in this study. Fifteen percent of the patients that had unilateral supraglottoplasty performed needed an additional contralateral procedure performed for resolution of symptoms. Two patients (3%) that underwent bilateral supraglottoplasty initially developed supraglottic stenosis. No patients undergoing unilateral supraglottoplasty developed supraglottic stenosis. Dr. Reddy recommended that unilateral supraglottoplasty is a reasonable initial surgical management of pediatric patients with severe laryngomalacia.

Dr. D Loke et al. performed a retrospective review in 2001 that examined the outcome of 32 cases of severe laryngomalacia that underwent a simple division of the aryepiglottic folds. Approximately 69% showed complete resolution of stridor and associated complications of laryngomalacia, while 22% showed partial resolution of stridor with no further surgical intervention required. Six percent required one additional procedure which included more extensive excision of redundant mucosa. One patient required a tracheotomy. It was concluded that endoscopic excision of the aryepiglottic folds is highly effective and should be the first-line treatment for severe laryngomalacia.

In 2002, isolated posterior displacement of the epiglottis was addressed by Dr. JA Werner et al. by performing an epiglottopexy for the treatment of severe laryngomalacia. Six patients underwent epiglottopexy- 4 of which were solely epiglottopexy and 2 of which were combined epiglottopexy with transaction of the aryepiglottic folds. The procedure was performed transorally with a carbon dioxide laser. All six children demonstrated significant airway improvement without any further stridor. It was also noted that deglutition was not affected by this procedure.

The failures and complications of supraglottoplasty were analyzed in 2003 by Dr. F Denoyelle et al. The study design was a retrospective review that included 136 patients, 102 of which had isolated laryngomalacia and 34 of which had additional congenital anomalies (Pierre Robin, psychomotor retardation, CHARGE association, Down syndrome, miscellaneous). Outcome measures included persistence of dyspnea, sleep apnea, and/or failure to thrive; need for additional treatment; presence of granuloma, edema or web; and supraglottic stenosis. Failure or only partial improvement of symptoms was only seen in patients with additional congenital anomalies (8.8%). The need for revision surgery was 4.4% of patients, minor complications (granuloma, edema or web) occurred in 3.7% of patients and supraglottic stenosis occurred in 4.4% of patients. Dr. Denoyelle concluded that supraglottoplasty failed only in patients that had laryngomalacia and additional congenital anomalies. The complication rate was found to be similar between isolated laryngomalacia and patients that had laryngomalacia with additional congenital anomalies.

This year (2005), Dr. GH Zalzal et al. presented a new approach to supraglottoplasty by making use of the microdebrider. He reported on a series of five patients that had been diagnosed with severe laryngomalacia and underwent a microdebrider-assisted supraglottoplasty. The technique involved initially dividing the aryepiglottic fold with microlaryngeal scissors. Subsequently, the folds are resected with the microdebrider with tissue removal extended anteriorly to the lateral edge of the epiglottis and posteriorly to the arytenoids cartilage. If redundant supra-arytenoid mucosa was identified, it was removed with the microdebrider. All five patients had resolution of their stridor postoperatively. No complications including

supraglottic stenosis have been identified and no revision surgeries have been performed.

When evaluating an infant that presents with stridor, laryngomalacia tops the differential diagnosis. It is established that the majority of symptoms of laryngomalacia resolve without intervention by the ages of twelve to eighteen months. The otolaryngologist must decide if the associated symptoms justify operative intervention. Furthermore, it must be decided if there are associated congenital anomalies that could impact the success of the surgery. If surgical intervention is pursued, all potential mechanisms contributing to laryngomalacia should be addressed.

Bibliography

Bailey BJ. Head and Neck Surgery – Otolaryngology 3rd Edition. 2001: 902-3.

Cotton RT. Practical Pediatric Otolaryngology. 1999: 497-501.

Denoyelle F, Mondain M, Gresillon N, Roger G, Chaudre F, Garabedian EN. Failures and complications of supraglottoplasty in children. Archives of Otolaryngology Head and Neck Surgery. 2003 Oct;129 (10): 1077-80.

Hadfield PJ, Albert DM, Bailey CM, Lindley K, Pierro A. The effect of aryepiglottoplasty for laryngomalacia on gastro-oesophageal reflux. International Journal of Pediatric Otorhinolaryngology. 2003 Jan; 67 (1): 11-4.

Iglauer S. Epiglottidectomy for the relief of congenital laryngeal stridor, with report of a case. Laryngoscope. 1922; 32: 56-59.

Lane RW, Weider DJ, Steinem C, Marin-Padella M. Laryngomalacia: a review and case report of surgical treatment with resolution of pectus excavatum. Archives of Otolaryngology Head and Neck Surgery. 1984; 110: 546-51.

Loke D, Ghosh S, Panarese A, Bull PD. Endoscopic division of the ary-epiglottic folds in severe laryngomalacia. International Journal of Pediatric Otorhinolaryngology. 2001 Jul 30; 60 (1): 59-63.

Mancuso RF, Choi SS, Zalzal GH, Grundfast KM. Laryngomalacia: The search for the second lesion. Archives of Otolaryngology Head and Neck Surgery. 1996 Mar; 122 (3): 302-6.

Manning SC, Inglis AF, Mouzakes J, Carron J, Perkins JA. Laryngeal anatomic differences in pediatric patients with severe laryngomalacia. Archives of Otolaryngology Head and Neck Surgery. 2005 Apr; 131 (4): 340-3.

Olney DR, Greinwald JH Jr, Smith RJ, Bauman NM. Laryngomalacia and its treatment. Laryngoscope. 1999 Nov; 109 (11): 1770-5.

Reddy DK, Matt BH. Unilateral vs. bilateral supraglottoplasty for severe laryngomalacia in children. Archives of Otolaryngology Head and Neck Surgery. 2001 Jun; 127 (6): 694-9.

Roger G, Denoyelle F, Triglia JM, Garabedian EN. Severe laryngomalacia: surgical indications and results in 115 patients. Laryngoscope. 1995 Oct;105: 1111-7.

Seid AB, Park SM, Kearns MJ, Gugeheim S. Laser division of the aryepiglottic folds for severe laryngomalacia. International Journal of Pediatric Otorhinolaryngology. 1985; 10: 153-8.

Senders CW, Navarrete EG. Laser supraglottoplasty for laryngomalacia: are specific anatomical defects more influential than associated anomalies on outcome? International Journal of Pediatric Otorhinolaryngology. 2001 Mar; 57 (3): 235-44.

Toynton SC, Saunders MW, Bailey CM. Aryepiglottoplasty for laryngomalacia: 100 consecutive

cases. *Journal of Laryngology and Otology*. 2001 Jan; 115 (1): 35-8.

Venkatarthikeyan C, Thakar A, Lodha R. Endoscopic correction of severe laryngomalacia. *Indian Journal of Pediatrics*. 2005 Feb; 72 (2): 165-8.

Werner JA, Lippert BM, Dunne AA, Ankermann T, Folz BJ, Seyberth H. Epiglottopexy for the treatment of severe laryngomalacia. *European Archive of Otorhinolaryngology*. 2002 Oct; 259: 459-64.

Zalzal GH, Anon JB, Cotton RT. Epiglottoplasty for the treatment of laryngomalacia. *Annals of Otology, Rhinology, and Laryngology*. 1987; 96: 72-6.

Zalzal GH, Collins WO. Microdebrider-assisted supraglottoplasty. *International Journal of Pediatric Otorhinolaryngology*. 2005 Mar; 69 (3): 305-9.