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Introduction

There are many causes of noisy breathing in the neonate. Any site in the upper airway can become obstructed and cause noisy breathing as well as dyspnea. These include nasal causes such as choanal atresia or nasal stenosis, pharyngeal causes including lingual thyroid, laryngeal causes such as laryngomalacia and subglottic stenosis, and tracheobronchial causes such as tracheal stenosis. Lesions in the oropharynx may cause stertor, while lesions in the laryngotracheal tree will cause stridor. Subglottic stenosis is the third leading cause of congenital stridor in the neonate. Most cases are self limited, but some require intervention that require a multidisciplinary approach, with the head and neck surgeon taking a primary role.

Anatomy

The subglottis is defined as the area extending from the lower surface of the true vocal cords to the lower surface of the cricoid cartilage. In adults this corresponds to approximately 10 mm inferior to the anterior commissure and 5 mm inferior to the posterior commissure.

The infant larynx differs significantly in size and position when compared to the adult larynx. At birth, the infant larynx is approximately one third the size of the adult larynx, however, the infant larynx is proportionately larger than the adult larynx compared with the remainder of the tracheobronchial system. The vocal process of the arytenoid takes up half the length of the vocal cord in the infant larynx, while it only takes up about ¼ of the length of the vocal cord in the adult. The narrowest portion of the airway in the older child and adult is the glottic aperture, while the narrowest part of the airway in the infant is the subglottis. The subglottis in infants measures approximately 4.5 by 7mm. A diameter of 4.0 mm is considered the lower limit of normal in a full term infant and 3.5 mm in a premature infant. One millimeter of edema circumferentially in the subglottis reduces the cross-sectional area by 60%.

The infant larynx is positioned higher in the neck than the adult larynx. The superior border of the larynx of the infant is located at about the level of the first cervical vertebrae with the cricoid positioned at about the fourth cervical vertebrae. In comparison, the adult cricoid

rests at about the level of the sixth cervical vertebrae. The structures of the infant larynx are more pliable and less fibrous making the infant airway more susceptible to narrowing from edema and less easily palpable.

Embryology

The anlagen of the larynx, trachea, bronchi and lungs arise from the ventromedial diverticulum of the foregut called the tracheobronchial groove at day 25. The lining of the larynx and trachea are from endoderm, while the muscles and cartilage are derived from splanchnic mesenchyme. At day 33 the laryngeal primordia appears, and the laryngeal aditus is formed by the growth of three tissue masses, the hypobranchial eminence from arches III and IV (epiglottis), and the paired ventral ends of arch IV (arytenoids), giving it a T-shape. This slit, extends to the first tracheal ring in the fifth and sixth week, and by week seven the cricoid ring is complete, and the cartilaginous hyoid is visible below the epiglottis. By the end of the embryonic period, the larynx and trachea are well formed.

During the fetal period, the structures are refined, and neurological reflexes are developed. In the third month, the thyroid laminae fuse, cartilaginous vocal processes of the arytenoids are seen, and the ventricle and saccule are seen. The fourth month reveals laryngeal tissue with goblet cells, and tracheal tissue with cilia. In the fifth and sixth months, the cuneiform and corniculate cartilages develop, and the epiglottis has fibroelastic cartilage. Throughout the remaining fetal course, breathing becomes a more mature and coordinated event.

Etiology

I. Congenital SGS

- A. Membranous
 - 1. increased fibrous connective tissue
 - 2. hyperplastic submucous glands
 - 3. granulation tissue
- B. Cartilaginous
 - 1. cricoid cartilage deformity
 - a. small cricoid
 - b. elliptical cricoid
 - c. large anterior lamina
 - d. large posterior lamina
 - e. generalized thickening
 - f. submucous cleft
 - 2. trapped first tracheal ring

II. Acquired SGS

- A. Intubation
- B. Laryngeal trauma
 - a. previous airway surgery
 - high tracheotomy
 - cricothyroidotomy
 - prior surgery for respiratory papillomatosis
 - prior laser surgery for SGS

- b. accidental
 - 1. inhalational (thermal or caustic)
 - 2. trauma (blunt or penetrating)
 - C. Autoimmune
 - D. Infection
 - E. Gastroesophageal reflux (GER)
 - F. Inflammatory diseases
 - a. Anti-neutrophil Cytoplasmic Autoantibodies (C-ANCA)
 - b. sarcoidosis
 - c. Systemic lupus erythematosus
 - G. Neoplasms
- III. *Idiopathic SGS***

Congenital Subglottic Stenosis

Subglottic stenosis is the third most common cause of stridor in the neonate behind laryngomalacia and vocal cord paralysis. Involves narrowing of the subglottic lumen in the absence of trauma (intubation). Normal diameter of newborn trachea is 5mm, lumen of 4mm in full-term newborn (3mm in premature infant) represents subglottic stenosis. Proposed mechanism is incomplete recanalization during embryogenesis.

There are two types of congenital subglottic stenosis: membranous and cartilaginous. Membranous is usually circumferential, soft and dilatable, in contrast, the cartilaginous has a more variable appearance. Mild-normal shape with narrowed lumen, or can have abnormal shape of cricoid cartilage with prominent lateral shelves giving an elliptical appearance to the lumen.

Symptoms

Symptoms of upper airway obstruction dominate, with inspiratory stridor, progressing to biphasic as obstruction worsens. **Stridor** is an important symptom of SGS as well as many other causes of neonatal obstruction. The characteristics of stridor can be a clue as to where the obstruction is occurring. Three distinct zones have been identified, the supraglottic/supralaryngeal zone, the extrathoracic tracheal zone which includes the glottis and subglottis, and the intrathoracic trachea. Supraglottic stridor is high-pitched, and inspiratory. An example of this is laryngomalacia, where the tissues are floppy and are sucked in during inspiration causing narrowing of the airway. Glottic and subglottic stridor is usually biphasic in nature because these structures are rigid, and there is no collapse of tissue during either phase of respiration, there is a fixed obstruction. Intrathoracic stridor has a expiratory noise, this is due to the positive pressure exerted on the bronchial tissues by contraction of the thorax.

Causes of stridor in children according to site of obstruction

- I. Pharynx
 - a. Congenital
 - i. Lingual thyroid
 - ii. Choanal atresia
 - iii. Craniofacial anomalies

- iv. Cysts
 - b. Inflammatory
 - i. Abscess
 - ii. Allergic polyps
 - c. Neoplasm
 - d. Adenotonsillar hypertrophy
 - e. Foreign body
- II. Larynx
 - a. Congenital
 - i. Laryngomalacia
 - ii. Webs, cysts, laryngocele
 - iii. Subglottic stenosis**
 - iv. Laryngeal cleft
 - b. Inflammatory
 - i. Croup, epiglottitis
 - ii. Tb, rare infections
 - c. Neoplasm
 - i. Subglottic hemangioma
 - ii. Laryngeal papilloma
 - iii. Cystic hygroma
 - iv. Malignant (chondrosarcoma, rhabdomyosarcoma)
 - d. Trauma
 - i. Intubation
 - ii. Foreign body
 - iii. Neck trauma
 - e. Vocal cord paralysis
 - f. laryngospasm
- III. Tracheobronchial tree
 - a. Congenital
 - i. Vascular anomalies
 - ii. Webs, cysts
 - iii. Tracheal stenosis
 - b. Neoplasm
 - c. Foreign body
 - d. Trauma
 - i. Intubation
 - ii. Tracheotomy

In addition to stridor, may show intercostal or xyphoid retractions and have a barking cough. Mild-moderate SGS is exacerbated by URI, and these children tend to have recurrent croup.

Diagnosis

When these children present, it is important to perform a thorough history and physical exam. Birth injury, or intubation, as well as prematurity are important to note. The timing, onset, and duration of stridor, voice/cry quality, feeding abnormalities or failure to thrive,

cyanosis, and possible foreign body aspiration are important to document. Also, recurrent croup or hospitalizations for respiratory illnesses. The physical exam should include a thorough head and neck exam, as well as careful characterization of stridor, and signs of respiratory distress. A flexible laryngoscopic exam should also be performed. At that time, an assessment of laryngomalacia, vocal cord paralysis, laryngopharyngeal reflux, or other laryngeal pathology can be elicited.

Differential diagnosis of laryngotracheal stenosis

I. Congenital

- A. Tracheomalacia
- B. Laryngomalacia
- C. Vocal cord paralysis
- D. Laryngeal cleft
- E. Congenital cysts
- D. External compression from congenital abnormality or lesion
 - 1. Vascular compression
 - a. innominate artery compression (most common)
 - b. right-sided aortic arch with persistent ductus arteriosus
 - c. aberrant left pulmonary artery
 - 2. Mass
 - a. teratoma
 - b. cystic hygroma
 - c. hemangioma

II. Infectious/inflammatory

- A. Viral laryngotracheobronchitis (croup)
- B. Retropharyngeal abscess
- C. GER
- D. Tracheitis

III. Neoplastic

- A. Subglottic hemangioma
- B. Recurrent respiratory papillomatosis

IV. Traumatic

- External compression
- Foreign body

The gold standard for diagnosis of any laryngotracheal abnormalities is direct laryngoscopy and tracheobronchoscopy under general anesthesia. This should be performed in the operating room with an experienced anesthesiologist. It is important to delay endoscopy for at least two weeks following an acute episode of croup to minimize the risk of postoperative airway obstruction. The potential need for tracheotomy should be discussed with the patient (adults) or patient's family (children) prior to endoscopy. A rigid bronchoscope or a rod lens

telescope may be used to assess the airway. The important things to document during endoscopy are as follows: (1) the outer diameter of the largest bronchoscope or endotracheal tube that can be passed through the stenotic segment, (2) the location/subsites (glottis, subglottis, trachea) and length of the stenosis, (3) other separate sites of stenosis, (4) other airway anomalies in infants (clefts, webs, cricoarytenoid joint fixation, neoplasms, etc.), and (5) reflux changes. After removing the sizing endotracheal tube or bronchoscope it is important to observe the stenotic segment for edema which may result in the need for tracheostomy

There are two widely excepted staging systems for classifying subglottic stenosis: Myer-Cotton grading system and the McCaffrey system. Other systems have been described as well, however, none are universally applicable or useful. At this time, no staging system exists that allows comparison of patients treated at different institutions.

The Myer-Cotton staging system is useful for mature, firm, circumferential stenosis confined to the subglottis. It describes the stenosis based on the percent relative reduction in cross-sectional area of the subglottis which is determined by differing sized endotracheal tubes. Four grades of stenosis are described with this system: **grade I** lesions have less than 50% obstruction, **grade II** lesions have 51% to 70% obstruction, **grade III** lesions have 71% to 99% obstruction, and **grade IV** lesions have no detectable lumen or complete stenosis.

The McCaffrey system classifies laryngotracheal stenosis based on the subsites involved and the length of the stenosis. Four stages are described: **stage I** lesions are confined to the subglottis or trachea and are less than 1cm long, **stage II** lesions are isolated to the subglottis and are greater then 1 cm long, **stage III** are subglottic/tracheal lesions not involving the glottis, and **stage IV** lesions involve the glottis.

Management

The management of congenital SGS ranges from observation with supportive care in times of exacerbations, to complicated surgical reconstructions of the patients airway. Most grade I and II lesions are managed with observation. If a grade II lesion becomes symptomatic, causing a decrease in exercise tolerance or respiratory distress, then endoscopic repair, dilation, or an expansion procedure can be undertaken. **KTP and CO2 laser** has also been used for lesions that are thin, and circumferential. Failures tend to occur with thick, circumferential cicatricial scarring greater then 1 cm in vertical dimension, and posterior commissure involvement. Complications of laser include chondritis, and perichondritis, as well as re-stenosis. With high grade stenosis, grade III-IV will need an open surgical procedure.

Surgical treatment options for high grade subglottic stenosis:

I. Tracheostomy

II. Open procedure

- A. Expansion procedure (one-stage or with stent placement)
 - 1. Anterior cricoid split with or without cartilage graft
 - 2. Posterior cricoid split with or without cartilage graft
 - 3. Anterior and posterior cricoid split with cartilage graft

4. Four quadrant LTR
- B. Segmental resection (cricotracheal resection - CTR)
 1. Primary CTR
 2. Salvage CTR
 3. Extended CTR – CTR with and expansion procedure, arytenoid lateralization, or arytenoidectomy

The goal of surgery is to have patients with adequate airway to allow for normal activity without the need for tracheostomy. Secondary goal to have single-stage procedure, minimal postoperative morbidity, and minimal hospital stay. Most surgeries are performed in spring and summer, when chance of developing RSV is lower.

Anterior Cricoid Split

The anterior cricoid split (ACS) procedure was originally described for a neonate who has had multiple failed extubations instead of performing a tracheostomy (Cotton and Seid, 1980). This procedure is also used for older infants and those who have already been tracheotomized. Indications were later expanded to patients with congenital subglottic stenosis. The lesion responsive to this procedure is a mild anterior subglottic narrowing with extensive fibrosis but a normal cricoid. ACS may also be used to decompress subglottic cysts. Strict criteria for ACS have been established by Cotton and include: extubation failure on two occasions or more due to laryngeal pathology, weight >1500g, no assisted ventilation for 10 days prior to evaluation, O₂ requirements <30%, no CHF for one month prior to evaluation, no acute respiratory tract infection, no antihypertensive medications ten days prior to evaluation. The procedure is performed after direct laryngoscopic and bronchoscopic confirmation of the diagnosis. All other airway pathology must be ruled-out.

A vertical midline incision is made through the cricoid cartilage and first two tracheal rings as well as the lower thyroid cartilage. This allows the cartilages to spring open and allow edematous mucosa to drain, increasing airway size. Prolene stay sutures are placed on either side of the cricoid cartilage and the skin is re-approximated after placement of a drain. The child is then left intubated, sedated and paralyzed in the ICU for 7-14 days. Cotton has guidelines for endotracheal tube sizes for stenting and for duration of stenting based on the infant's weight.

Laryngotracheal Expansion Surgery

Laryngotracheal expansion surgery involves scar division with distraction of the edges by interposition of graft material (augmentation) to widen the airway lumen. It is important to avoid removing scar which results in a large surface area of denuded mucosa and leads to restenosis. Cotton recommends augmenting the airway with grafts when the distraction of the laryngotracheal framework must be greater than approximately 3mm. There are several techniques depending on the location and severity of the stenosis. Laryngotracheoplasty can be performed with a tracheostomy and formal stenting or by using the endotracheal tube as a stent, the latter known as a single-stage LTP (SS-LTP). Gustafson et al performed a retrospective chart review of 200 patients undergoing SS-LTP. There was a 96% successful decannulation rate, with 29% requiring reintubation. Those undergoing both posterior and anterior grafting had

more complications than those undergoing either anterior or posterior grafting alone. Younis et al, performed a retrospective chart review of 46 patients undergoing anterior/posterior SS-LTP and had a 83% decannulation rate. Thus, this procedure should be used for either anterior or posterior stenosis, but other procedures should be considered if there is anterior and posterior stenosis.

Several stents have been used in the past, but the aboulker stent (teflon) is the most used. It has the benefit of being rigid, and having a lumen with a sufficient diameter for aggressive pulmonary toilet. The duration of stenting depends on the length of the prior stenosis, and the grade of stenosis. Stenting can last from four weeks to greater than two months. (Cable et al 2004)

Graft material has included rib cartilage, auricular cartilage, thyroid cartilage hyoid bone, and irradiated cartilage. Autogenous cartilage is the material of choice for grafting, most commonly costal or auricular cartilage. Cartilage is better material because it has a lower rate of resorption, is easy to carve, and is viable without a vascular pedicle. They also retain bulk even without functional use. Hyoid bone requires vascular pedicle, can resorb, and is difficult to carve.

Anterior laryngofissure with anterior lumen augmentation

This technique is good for anterior subglottic stenosis or anterior tracheal wall collapse. The lesion should not involve the glottis. Other procedures should be considered if there the cricoid cartilage is deformed or weak. Anterior grafts are made considerably larger and thicker than grafts placed posteriorly. The perichondrium is oriented to the luminal side to allow for epithelialization. The perichondrium is also a good barrier against infection. A large external flange is created to prevent the graft from prolapsing into the airway.

Laryngofissure with division of posterior cricoid lamina

This is indicated for patients with posterior subglottic stenosis, posterior glottic stenosis that extends to the glottis, complete or circumferential stenosis, or if there is significant cricoid deformity. Division of the anterior and posterior cricoid must be carried out for this procedure. If possible, one should avoid a complete laryngofissure to avoid damaging the anterior commissure, however this is often needed for posterior glottic involvement for access. The posterior cricoid cartilage is incised in a manner that is vertically oriented to the cartilage to allow maximal purchase for the graft. The incision is extended superiorly to the interarytenoid area and inferiorly 5 to 10 mm into the membranous trachea. The graft is elliptical in shape. It should not be too thick as it can cause swallowing difficulties and can lead to aspiration. The width of the graft is determined by the desired distraction of the cut edges of the incised posterior cricoid cartilage. 0.05 to 1.00 mm of distraction can be obtained for each year of age, up to 1 cm. It is sutured in place with absorbable suture on a small cutting needle. The knots should be buried so that they remain extraluminal to prevent development of granulation tissue. Long-term stenting is usually necessary (3-6 months).

Laryngofissure and division of posterior cricoid lamina with anterior and posterior grafts

This should be used for patients who have SGS similar to those above but with a significant amount of stenosis posteriorly such that grafting is necessary to maintain the adequate separation.

Once the grafts have been sutured into place in any of the above procedures, the decision must be made on whether it should be single or double-staged. Cotton and Walner (1999) recommend a double-staged procedure for patients with severe stenoses, history of reactive airway, or poor pulmonary function. This should also be considered at institutions with inadequate intensive care facilities. Double-stage procedure implies placement of stent above the tracheostomy tube instead of using an endotracheal tube as the stent (single-staged procedure). Once this decision is made, the strap muscles are closed to provide blood supply to the outer surface of the anterior graft.

Segmental resection/Cricotracheal resection (CTR) with thyrotracheal anastomosis

The first CTR was performed by Conley in 1953 in a patient undergoing surgery for chondroma of the cricoid cartilage. It was later popularized by Ogura and Powers (1964) as a technique for treatment of traumatic stenosis. In the 1970s it became the treatment of choice in adults with acquired subglottic stenosis from long term intubation. Until recently, surgeons were reluctant to perform this procedure in the pediatric patients because of the risk of anastomotic dehiscence and recurrent laryngeal nerve injury, and disturbing the normal growth of the larynx. The first successful CTR performed in a child was occurred in 1978 (Savary). In 2005, White et al performed a retrospective chart review of 96 patients undergoing CTR. 89 of these patients had grade III/IV stenosis, and they achieved a 94% decannulation rate. They found that vocal fold dysfunction was the only significant risk factor for failure to decannulate after one procedure.

This technique is indicated if there is severe deformity of the cricoid making grafting very likely to fail. Most say that there must be at least 10 mm of normal airway below the glottis, however Cotton states that the resection can be up to the vocal folds but to expect prolonged edema. This technique is technically difficult due to the close proximity of the vocal cords and recurrent laryngeal nerves. Stenosis less than 4 cm can be resected by laryngeal release and cervical tracheal mobilization. Stenting is not required and the tracheostomy tube can usually be removed at around 4 weeks.

Complications

These include bleeding, pneumothorax, pneumomediastinum, recurrent laryngeal nerve injury, slipped graft, slipped stent, plugged stent, wound infection, keloid formation, suprastomal/infrastomal collapse, re-stenosis, tracheocutaneous fistula, granulation tissue and death.

Factors contributing to LTR failure (Choi et al 1999)

Preoperative - inadequate assessment of posterior laryngotracheal stenosis

Intraoperative – duration of stent, type of stent, correct assessment with failure to address posterior stenosis, type of graft, and length of stent.

Postoperative – keloid formation, anterior suprastomal collapse, poor follow-up, slipped stent, broken stent, GERD, no discernable factors.

Postoperative Care

Patients should be admitted to intensive care unit, and care must be coordinated with the ICU team, pediatrics, and otolaryngology. Patients require sedation, but the length of sedation will vary on the age of the child, and the procedure performed. In children over 4 years old, there is a better chance of weaning sedation within 48 hours after the procedure. Aggressive and meticulous tracheostomy care and pulmonary toilet needs to be undertaken. Post operative antibiotics, antireflux medications and dexamethasone are also indicated.

Conclusions

Approximately five percent of children undergoing procedures will require a smaller than expected endotracheal tube due to mild subglottic stenosis. Most of these children will never present to the otolaryngologist, but for more severe cases, these children present a challenging problem for the head and neck surgeon. It is imperative to perform detailed history, physical, and characterization of the extent and severity of the stenosis. Rigid endoscopy is essential for preoperative planning for any of the surgical procedures that can be used for correction. Choice of operation is dependent on surgeon comfort, postoperative capabilities, and severity of disease. For high grade stenosis, the single-stage laryngotracheal resection, or the cricotracheal resection are the best options. CTR is also available as a salvage to LTR failures. Remember that the goal of surgery is to allow for an adequate airway for normal activity without the need for tracheostomy.

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