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"SUBGLOTTIC STENOSIS" EMBRYOLOGICAL BASIS AND ITS CLINICAL IMPORTANCE

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ABSTRACT

Subglottic stenosis (SGS) is a partial or complete narrowing of the airway under the vocal cords and above trachea and it is third leading congenital abnormality of larynx. It is because of incomplete development of lumen of laryngotracheal tube in 3rd month of gestation. But it appears in first few months after birth. Larynx develops from 4th and 6th branchial arches. The incidences related to congenital SGS had a significant increment in the late 1960s. Patient with SGS may or may not have some significant respiratory distress. Subglottic stenosis can be membranous or cartilaginous.

Introduction

There are many reasons of noisy and painful breathing, any part of the upper air pathway can be obstructed and it can cause noisy breathing which is also known as dyspnea. These include nasal causes like choanal atresia or nasal stenosis. Pharyngeal causes include lingual thyroid. Laryngeal causes are laryngomalacia and subglottic stenosis. And Tracheo-bronchial cause is tracheal stenosis. Lesions in the laryngotracheal tree will cause stridor. Subglottic stenosis (SGS) is a narrowing of the airway under the vocal cords and above trachea. Subglottic stenosis is the third leading cause of congenital airway abnormalities (after laryngomalacia and vocal cord paralysis). Most cases are self-limited, but some require intervention and that requires a multidisciplinary approach, with a head and neck surgeon. Early in the 20th century, SGS was usually related to trauma or infection from tuberculosis, diphtheria, syphilis, and or typhoid fever. Often, the treatment for SGS at this time, a tracheostomy, led to further damage to the airway. SGS considered as congenital if there is no history of previous incubation. Amendment of this abnormality requires expansion of the lumen of the cricoid area of larynx to increase airflow and decrease obstruction in breathing. The sub glottis is defined as the area extending from the just beneath of the true vocal cords down towards the top of trachea. In adults this corresponds to approximately 10 mm lower to the anterior commissure and 5 mm lower to the posterior commissure. The subglottic airway is the narrowest area of the airway because it is enclosed in a complete, nonexpendable, and non-pliable ring.[2]

Incidence

The frequency of congenital subglottic stenosis (SGS) is unknown. Congenital subglottic stenosis is the third very common congenital anomaly of the larynx, includes 15% of all cases. This disorder is the most common laryngeal anomaly that involves tracheotomy in infants. Males are affected twice as frequently as females.[2]

All studies available after 1983 an incidence reported of neonatal subglottic stenosis is less than 4.0%, and all studies available after 1990 reported an incidence of neonatal subglottic stenosis is less than 0.63%. After studying the literature, we think a sliding trend exists in the incidence of neonatal subglottic stenosis in the late 1990s. The recent incidence of neonatal subglottic stenosis is expected between 0.0% and 2.0%.

The incidence of subglottic stenosis (SGS) has highly decreased over the last 40 years. In the late 1960s, when endotracheal intubation and long-term ventilation for premature newborns began, the incidence of subglottic stenosis (SGS) was high as 24% in patients demanding such care. In the 1970s and 1980s, estimations of the incidence of subglottic stenosis (SGS) were 1-8%. In 1998, Choi described that the incidence of subglottic stenosis (SGS) had remained persistent at the Children's National Medical Center in Washington, DC; it was around 1-2% in children who had been treated in the neonatal ICU. Recently, Walner reported that, among 504 neonates who were admitted to the level III ICU at the University of Chicago in 1997, 281 were intubated for nearly 11 days, and in those patient no one got subglottic stenosis (SGS) develop over for a 3-year period. In 1996, a report from France also said no incidence of subglottic stenosis (SGS) in the neonatal population who went through intubation with very minor endotracheal tubes (i.e., 2.5-mm internal diameter) in efforts to prevent trauma to the airway.[7]

Ontogenesis for the normal development of larynx

The larynx develops from the ectodermal lining of the cranial end of the laryngotracheal tube and surrounding mesenchyme tissue (from branchial arches 4th, 5th, and 6th).

The mesenchyme tissue proliferates and produces a paired arytenoid swelling, which gives the primitive glottis which has a T-shaped appearance and reducing the laryngeal lumen to a cut. The laryngeal cartilages are developed within the arytenoid swellings from the cartilage bars of the branchial arches. The epiglottis grows from the caudal half of the hypobranchial eminence, which is a derivative of branchial arches 3rd and 4th [5].

The orifice to the larynx ends dimly, between the 7th to 10th weeks. It is because of the fusion of the epithelium, but as the epithelium dissolve, the laryngeal inlet/aditus expands and recanalizes. A pair of lateral recesses, and the laryngeal ventricles, forms which are bounded cranially by anteroposterior folds of mucous membrane, and caudally by the future vestibular (false) and vocal (true) folds. The laryngeal muscles are developed from muscle elements in branchial arches 4th to 6th and are innervated by laryngeal branches of the 10th cranial nerve (vagus).

The initial event of laryngeal development happens when the embryo is just 2 mm in length, with the appearance of a median pharyngeal groove as the precursor to the primitive foregut. By day 25, a ventromedial diverticulum originates from the upper foregut that rapidly forms the tracheobronchial tree and lung buds. Lateral processes commence to form that will fuse in the midline, separating the trachea and oesophagus. At 33 days, the primitive epiglottis and arytenoid cartilage precursors form from the mesenchymal parts of pharyngeal arches 3/4 and 6, respectively. The cricoid ring gets complete about 7 weeks of development, and formation of the remaining laryngeal cartilages is well under way at this point. The trachea, oesophagus, and larynx are fully developed organs by 12–13 weeks, with evidence of the primeval function follow canalization by resorption and enlargement of endodermal tissues. [8]

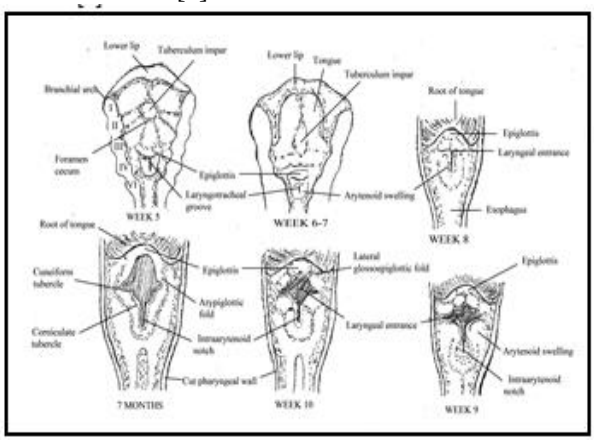

Fig 1: Schematic representation shows the normal development of Larynx.

In laryngeal development, the fetal phase is marked by fetal "breathing," in which the amniotic fluid is circulated into the airway, in addition to by swallowing through the esophagus. Both are significant for the development of distal structures and for the modification of laryngeal function. Therefore, failure of canalization leads to not only in airway blockage from laryngeal atresia but also in alimentary and pulmonary underdevelopment. This fetal phase also shows further differentiation of the epithelium of several portions of the larynx into respiratory and the formation of goblet cells and squamous epithelium.

At the time of birth, the larynx is very much different in form and function as compared to the adult larynx. The cricoid cartilage originally presents at the level of C2 but fall away to C5 at 2 years of age and to C6 by 5 years of age. The adult cricoid cartilage is completely descended at C6/C7 at around 15 years of age.

The newborn thyroid cartilage is correspondingly shorter and wider than in the adult larynx, and the cricoid cartilage is comparatively smaller. This leads to a funnel shape laryngeal airway. In addition, the subglottic submucosa has a larger amount of glandular soft tissue than the rest of the airway, making it the narrowest part of the newborn airway as opposed to the glottis in the adult. Other differences comprise increased compliance of the thyroid cartilage of the young larynx and its undeveloped neuromuscular reflexes, which influence neonates to certain levels of aspiration, and particularly of reflux fluid.

The subglottis is the part of the upper airway tract, bounded by the meeting point of two epithelium simple squamous and pseudostratified ciliated columnar epithelium superiorly. The subglottis is located a few millimeters below the free edge of the true vocal fold and is bounded inferiorly by the inferior edge of the cricoid cartilage. It is surrounded from all sides by the cricoid ring, which itself forms the only completely enclosed ring in the complete airway and is a very important contributor to the development of obstructive lesions. The tracheal and thyroid cartilages do not form complete rings and therefore they have the potential for physiologic expansion, at least tentatively. The complete ring of thick cricoid cartilage is challenging in the face of airway compromise, further intensifying the urgency of airway considerations when dealing with lesions of the subglottic.[9]

Ontogenesis for the congenital subglottic stenosis

The imperfect recanalization of the laryngotracheal tube during the 3rd month of gestation leads to different types of congenital subglottic stenosis and with complete laryngeal atresia being its extreme form.

The appearances of congenital subglottic stenosis generally appear in the first few months of life. The stenosis is characteristically not evident until the small kid develops an acute inflammatory process that further compromises the subglottic stenosis. [3]

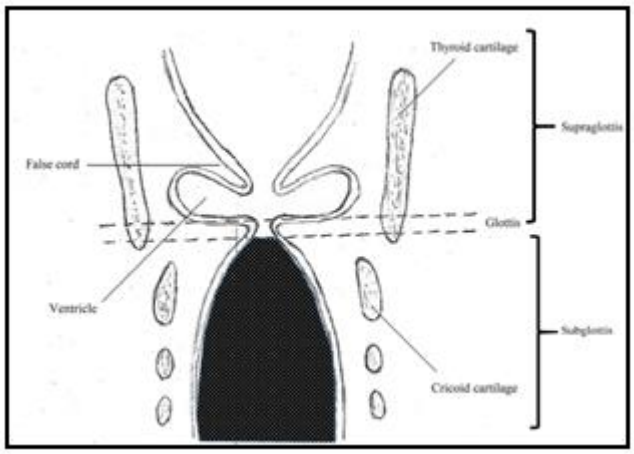

Fig 2: Schematic representation shows the congenital subglottic stenosis of the larynx.

Discussion

The stenosis is typically not apparent until the child develops an acute inflammatory course, which further compromises to the sub glottis. The scientific presentation of a child throughout these periods does not differ from that of infectious laryngo-tracheal bronchitis also known as croup. Biphasic stridor with or without symptoms of extreme respiratory pain is the most common presenting symptom. The child may have a woofing/barking cough, but the cry of child is usually normal.

We can Suspect congenital subglottic stenosis when these symptoms are frequent or if they are continued beyond the normal period of infectious croup.

Asymptomatic children who are hard to extubate, intubate, or decannulate exist another clinical scenario that stimulates suspicion of congenital stenosis. Children who have Down syndrome have more risk of having congenital subglottic stenosis and may existing in this fashion.

On examination, the children with congenital subglottic stenosis can or cannot have significant respiratory distress (Cyanosis, supraclavicular or intercostal indrawing, nasal flaring). In Head and neck examination results are usually normal. The Flexible endoscopy does not effectively assess the sub glottis but it is important to include the diagnoses of vocal cord paralysis and other glottic or supraglottic abnormalities. Do not pass the scope beyond the vocal cords since this may precipitate the airway obstruction in a patient with a compromised sub glottis.

A history of repeated croup usually suggests congenital subglottic stenosis. Perform a inflexible bronchoscopy to check the diagnosis and to assess the airway for any other anomaly. Estimate the stenosis in terms of its diameter and length. By Passing a endotracheal tube or scope through the stenosis may effectively assess the length and diameter. The largest tube or scope which passes through the airway gives decent measurement of the lumen diameter. Congenital subglottic stenosis is detected when the diameter of lumen is less than 4 mm in a term infant and less than 3 mm in preterm infant. The conclusions at endoscopy that support a diagnosis of congenital subglottic stenosis are characteristically not much severe than in children with acquired subglottic stenosis. The radiographic evaluation can help in measure the subglottic airway before bronchoscopy or when the diagnosis is uncertain. Plain lateral or anteroposterior radiographs will show a characteristic narrowing at the level of the sub glottis.
Maximum cases of congenital subglottic stenosis are resolve naturally with growth of the child. Tracheotomy and Endotracheal intubation may be required in patients who have significant airway obstruction. Most of the children who require tracheotomy can be decannulated by the age of 3-4 years when the subglottic space expands.

Laser ablation has a limited role in the treatment of congenital subglottic stenosis and is usually held in reserve for soft lesions less than 5 mm in thickness. Laryngotracheoplasty is normally unnecessary but may be required to recreate the airway in patients, who could not be decannulated. Laryngotracheoplasty is reserve for some severe cases of subglottic stenosis.

Types of congenital subglottic stenosis

Congenital subglottic stenosis can be divided into 2 categories: membranous and cartilaginous. Membranous SGS includes increment in fibrous connective tissue, hyperplastic submucous glands and granulation tissue. Membranous stenosis is more frequently seen than cartilaginous stenosis. Usually, membranous stenosis is symmetric and circumferential and may extend upward till the true vocal cords. Cartilaginous stenosis defined as deformity of the tracheal ring or cricoid cartilage projecting into the lumen. It can be symmetric in which small but regular shaped cricoid or a no distensible complete first tracheal ring stuck inside the cricoid. Or asymmetric which has oval-shaped or elliptic cricoid and isolated expansion of either the anterior or posterior part of cricoid lamina, or laryngeal cleft.

Classification of subglottic stenosis

The Myer-Cotton staging (MCS) system is very much useful for matured, resistant, circumferential stenosis restricted to the subglottis. The MCS system describes the stenosis based upon the percent relative lessening in cross-sectional area of the subglottic region which is insistent by differing size of endotracheal tubes. Four grades of subglottic stenosis are described with this system: grade I lesions have 50% or lesser obstruction in lumen, grade II lesions confined 51% to 70% obstruction, grade III lesions includes 71% to 99% obstruction, and grade IV lesions there is no detectable lumen or called complete stenosis.

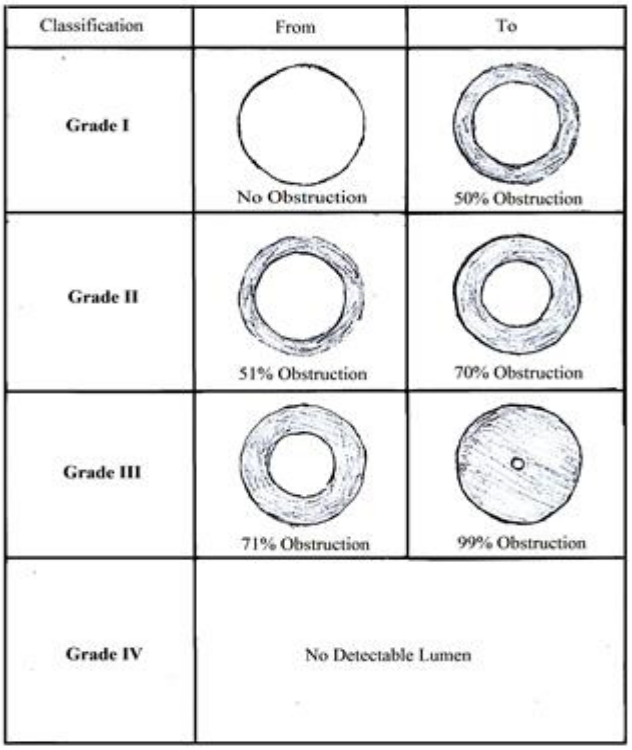

Fig 3. Schematic representation shows the different grades of classifications in the congenital subglottic stenosis.

The McCaffrey system categorizes laryngotracheal stenosis on the basis of subsites involved and the extent of the stenosis. Four stages are described as: stage I lesions are confined to the subglottis and or trachea are less than 1cm long, stage II lesions are secluded to the subglottis and are greater than 1 cm long, stage III are tracheal/subglottic lesions not involving the glottis part, and stage IV lesions involve the glottis region.

The incidences related to congenital SGS had a significant increment in the late 1960s, after that McDonald and Stocks introduce long term intubation as a treatment process for children's in need of prolonged ventilation in 1965.The increased incidence of SGS try to focused new attention on the larynx of child, as well as the need for development of novel treatment modalities.

In 1971, Rethi and Rhan proposed a procedure for vertical division of the posterior lamina of the cricoid cartilage with Aboulker stent placement.

In 1974, Evanston and Todd described success with a castellated incision of the anterior cricoid cartilage and upper part of trachea, which was sewn open, and a stent made of a rolled silicone sheet was placed in it for 6 weeks.

In 1974, Fearon and Cotton described the successful use of cartilage grafts to enlarge the subglottic lumen in children's and in African green monkeys with severe laryngotracheal stenosis. In 1980, Cotton and Seid described a method in which tracheotomy is avoided called the anterior cricoid split

(ACS). After some time Cotton reported his understanding with laryngeal expansion with cartilage grafting. His success rate depends on level of stenosis. Cotton use Aboulker stent.

In 1991, Seid explained a form of single-stage laryngotracheal reform in which cartilage was placed anteriorly to expand the subglottis and upper trachea to evade a tracheotomy.

In 1992, Cotton proposed a 4-quadrant cricoid split, in company with anterior and posterior grafting.

In 1993, Zalzal reported nearly 90% decannulation with any level of SGS his first surgical procedure. He customized the rebuilding on an individual basis, and most patients received Aboulker stents for stabilization.

Conclusions

Approximately more than five percent of children which are undergoing treatment procedures will require a smaller endotracheal tube than expected due to minor subglottic stenosis. Most of these children will never went to the otolaryngologist, but sometimes for more severe cases, these children give a challenging problem for the head and neck surgeon. It is important to perform detailed history, physical, and characterization of the level and severity of the stenosis. Rigid endoscopy is crucial for preoperative preparation for any of the clinical procedures that can be used for correction of stenosis. Choice of procedure is mainly dependent on surgeon comfort, postoperative competences, and severity of subglottic stenosis.

In conclusion of results from the literature studied and from some of the present authors' experience confirm that laryngotracheal resection signifies the medicinal treatment of choice for benign subglottic stenosis which allows high success rates and for long term. Most of the patients who experienced foremost postoperative difficulties can be effectively treated by non-operative (generally endoscopic) procedures and accomplished stable results over time. Low compliance patients who got psychiatric and/or neurologic disorders, and patients with ID stenosis won't show increased failure and complication rates after surgical resection.

References

[1] Ajit Kumar, E.Ganesh, T.Malarvani, Manish Kr. Singh. Bilateral supernumerary heads of biceps brachii. Int J Anat Res. 2014; 2(4):650-52.

[2] Ajit Kumar, Ganesh Elumalai, Malarvani Thangamani, Nirmala Palayathan, Manish Kr Singh. A Rare Variation in Facial Artery and Its Implications in Facial Surgery: Case Report. Journal of Surgery.2014; 2(5): 68-71.

[3] Ganesh Elumalai, Sushma Chodisetty. Anomalous "Mutilated Common Trunk" Aortic Arch Embryological Basis and its Clinical Significance. Texila International Journal of Basic Medical Science. 2016; 1(1): 1-9.

[4] Ganesh Elumalai, Emad Abdulrahim Ezzeddin. "The sudden soul reaper" - hypertrophic cardiomyopathy – its embryological basis. Elixir Embryology. 2016; 99: 43284-43288.

[5] Ganesh Elumalai, Muziwandile Bayede Mdletshe. "Arteria lusoria"- aberrant right subclavian artery embryological basis and its clinical significance. Elixir Embryology. 2016; 99: 43289-43292.

[6] Ganesh Elumalai, Sushma Chodisetty, Pavan Kumar D.2016. Ganesh Elumalai et al Classification of Type - I and Type - II "Branching Patterns of the Left Arch Aorta". Imperial Journal of Interdisciplinary Research. 2(9): 161-181.

[7] Ganesh E, Sushma C. The deer horn aortic arches" embryological basis and surgical implications. Anatomy Journal of Africa.2016; 5(2): 746 – 759.

[8] Ganesh Elumalai, Sushma Chodisetty. Teratological Effects of High Dose Progesterone on Neural Tube Development in Chick Embryos. Elixir Gynaecology. 2016; 97: 42085-42089.

[9] Ganesh Elumalai, Sushma Chodisetty. "The True Silent Killers" - Bovine and Truncus Bicaroticus Aortic Arches its Embryological Basis and Surgical Implications. Elixir Physio. & Anatomy. 2016; 97: 42246-42252.

[10] Ganesh Elumalai, Sushma Chodisetty, Bridget Omo Usen and Rozminabanu Daud Patel. "Patent Ductus Caroticus" - Embryological Basis and its Clinical significance. Elixir Physio. & Anatomy. 2016; 98: 42439-42442.

[11] Ganesh Elumalai, Sushma Chodisetty, Eliza Arineta Oudith and Rozminabanu Daud Patel. Common anomalies origin of left vertebral artery and its embryological basis. Elixir Embryology. 2016; 99: 43225-43229.

[12] Ganesh Elumalai, Sushma Chodisetty, Sanjoy Sanyal. Common Nasal Anomalies and Its Implications on Intubation in Head and Neck Surgeries. Journal of Surgery. 2016; 4 (4): 81-84.

[13] Ganesh Elumalai, Malarvani Thangamani, Sanjoy Sanyal, Palani Kanagarajan. Deficient sacral hiatus cause mechanical low back pain: a radiological study. Int J Anat Res. 2016; 4(1):1758-64.

[14] Mark E Gerber, MD Clinical Associate Professor of Surgery, University of Chicago Pritzker School of Medicine, Laryngeal Stenosis.2016 http://emedicine.medscape.com/article/867177-overview

[15] John E McClay, MD Associate Professor of Pediatric Otolaryngology, Department of Otolaryngology-Head and Neck Surgery, Children's Hospital of Dallas, University of Texas Southwestern Medical Center. 2016 http://emedicine.medscape.com/article/995328-overview

[16] Ted L Tewfik, MD Professor of Otolaryngology-Head and Neck Surgery, Professor of Pediatric Surgery, McGill University Faculty of Medicine. ttp://emedicine.medscape.com/article/837630.

[17] The-myer-cotton-staging-system , 2016 https://www.researchgate.net/figure/12245392_fig1_Figure-1-Figure-The-Myer-Cotton-classification-for-subglottic-stenosis

[18] Medical embryology book by Ben Panksy, Ph.D., M.D. 2016 http://discovery.lifemapsc.com/library/review-of-medical-embryology/chapter-58-development-of-the-lower-respiratory-system-larynx-and-trachea

[19] John E McClay, MD Associate Professor of Pediatric Otolaryngology, Department of Otolaryngology-Head and Neck Surgery, Children's Hospital of Dallas, University of Texas Southwestern Medical Center. 2016 http://emedicine.medscape.com/article/864208-overview

[20] John E McClay, MD Associate Professor of Pediatric Otolaryngology, Department of Otolaryngology-Head and Neck Surgery, Children's Hospital of Dallas, University of Texas Southwestern Medical Center. 2016 http://tracheostomy.com/resources/articles/subglottic_stenosis.

[21] Jonathan B. Ida, MD, MA, * J. Lindhe Guarisco, MD, Tulane University School of Medicine, Department of Otolaryngology-Head and Neck Surgery, New Orleans, LA

[22] Kimsey H. Rodriguez, MD, and Ronald G. Amedee, MD, FACS , Ochsner Clinic Foundation, Department of Otolaryngology-Head and Neck Surgery, New Orleans, LA.