Subglottic Stenosis

Katherine Hanlon, CRNP,[a] R. Paul Boesch, DO,[b] and Ian Jacobs, MD[a]

Subglottic stenosis refers to narrowing of the airway diameter below the vocal folds and may be congenital or acquired. Typical signs and symptoms range from recurrent croup and exertional stridor to complete airflow obstruction requiring tracheotomy. Management of moderate and severe subglottic stenosis often requires intricate surgical techniques. To optimize the success of these surgeries, a thorough assessment of the child's airway, lungs, reflux, and swallow needs to be evaluated. In order to provide concerted and coordinated care between typically otolaryngology (ENT), pulmonary, gastroenterology (GI), speech, swallow and language pathologists (SLP), "aerodigestive" teams have been developed and increasing in prevalence at children's medical hospitals. This article sets out to provide a brief overview of an aerodigestive program and evaluation, review a few of the more common laryngotracheal conditions, and the surgical techniques involved to augment the airway.

Curr Probl Pediatr Adolesc Health Care 2018;48:129-135

Subglottic Stenosis (SGS)

Subglottic stenosis refers to narrowing of the airway diameter below the vocal folds and may be congenital or acquired. Typical signs and symptoms range from recurrent croup and exertional stridor to complete airflow obstruction requiring tracheotomy. The configuration of the stenosis is largely related to the etiology: either **congenital** or **acquired**.

Congenital subglottic stenosis is due to failure of recanalization of the larynx during embryogenesis. Congenital SGS may vary in severity from small glottic webs, mild lateral narrowing, or to complete laryngeal atresia. The more common presentation of congenital SGS is lateral elliptical shelves of cartilage at the cricoid.[1–3] Congenital SGS represents more of a hyaline cartilaginous defect rather than fibrous scar tissue, which is important in determining the surgical repair strategy.

The more common etiology of SGS is **acquired** SGS. This is where there is remodeling of cricoid cartilage[3] (Fig. 1). The etiology of acquired subglottic stenosis is multifactorial: prematurity, long-term intubation, GERD and/or systemic illness.[3] Prolonged irritation leads to inflammation leading to a fibrous scarring. Both congenital and acquired SGS are evaluated and staged via a microlaryngoscopy and bronchoscopy and the Myer-Cotton system. The Myer-Cotton system uses endotracheal tubes that are passed through the stenosis to measure the diameter of the airway. A leak test is performed, when the endotracheal tube's leak is between 10 and 25 cm H_2O, then that tube size is compared to what is the expected normal size for a child of that age.[15]

The more common etiology of SGS is acquired SGS

Clinical manifestations: Mild forms of SGS, grade I-grade II, result in stridor, dyspnea with exertion, and recurrent croup. This may lead to multiple admissions to the hospital emergency room, frequent pediatrician visits and potentially misdiagnosis. The more severe SGS, grade III or IV, can lead to difficult intubations, inability to be mask ventilated, and require a tracheotomy. This poses a higher risk of anoxic brain injury when there is accidental decannulation in cases of an infant or child with a precarious or "critical" airway. An airway may be considered **"critical"** if there is severe narrowing of the airway making mask ventilation difficult or not possible at all and **"difficult"** if intubation is challenging, whether be craniofacial abnormalities or SGS.

From the [a]The Center for Pediatric Airway Disorders, Division of Otolaryngology, The Children's Hospital of Philadelphia, Philadelphia, PA; and [b]Mayo Clinic, Children's Center, Pediatric and Adolescent Medicine, Rochester, MN.

Curr Probl Pediatr Adolesc Health Care 2018;48:129-135
1538-5442/$ - see front matter

https://doi.org/10.1016/j.cppeds.2018.03.007

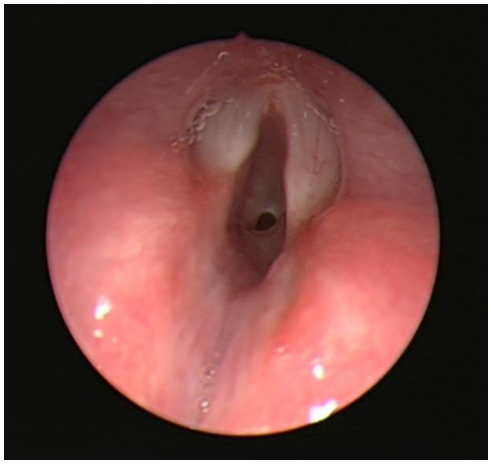

FIG 1. Grade III subglottic stenosis (permission from Ian Jacobs) File: F.Saleem grade 3 subglottic stenosis.

These distinctions and classifications assist providers and caregivers in communicating the status of the child's airway, thus providing safe care when they enter the hospital, imaging center, operating room, or office visits.

Aerodigestive Program and Evaluation

Aerodigestive Programs are a collaboration primarily involving ENT, Pulmonary, GI, and SLP but can include other subspecialties depending upon the patient's medical needs. Due to the intricate nature of laryngotracheal disorders, an aerodigestive team provides the necessary in-depth assessment required to establish a patient specific surgical plan and recovery without causing increased burden to the family or hospital.[4]

The process starts with contact from a caregiver or referring physician. The triage comprises of a review of the patient's medical history, including pertinent medical records specifically involving otolaryngology, GI, Pulmonary evaluations, previous operative reports, and swallow studies/evaluation. The medical summary is presented to the aerodigestive team and discussed. A tentative medical and surgical plan is created and the recommendations discussed with the caregivers. The child and family arrive to the office for a comprehensive history and physical exam by all the services. A feeding evaluation is performed at the time by the SLP, either clinical or objective (video functional swallow study (VFSS)). By having all of the appropriate specialists in one area, in one room in some cases, the team can discuss their findings and collaborate with the family on creating a safe, realistic treatment plan for the child.

A thorough airway diagnostic evaluation is essential to surgical planning for laryngotracheal reconstruction. This evaluation should identify all potential airway obstructive lesions from the nose through segmental bronchi. In addition to defining the anatomic airway lesions, evaluation should be performed with intent to identify infectious, inflammatory, and esophageal pathology that may interfere with successful surgery.[21] The preferred method for this evaluation is the "triple endoscopy." This is defined as the performance of microlaryngoscopy and bronchoscopy (MLB), flexible fiberoptic bronchoscopy (FFB) with lavage, and esophagogastroduodenoscopy (EGD) under a single anesthetic. The benefits of this approach to airway evaluation are several. First, combination under one anesthetic reduces the risk and cost of multiple anesthetics. There are additional benefits from having all three providers simultaneously witness the same findings. These include improved shared understanding of issues, honest ongoing dialogue regarding the relevance and interpretation of findings, and fostering collaborative creativity. It is common in this setting that initial impressions are modified as one procedure progresses through another. Lastly, when the otolaryngologist, pulmonologist and gastroenterologist review all findings together with the family, with a unified consensus plan, a very strong impression is made which engenders confidence.

The preferred method for this evaluation is the "triple endoscopy."

The order of procedures may vary as long as the goals of each procedure are met. The goals of the FFB are to evaluate the upper airway dynamics in a sleep state, evaluate lower airway anatomy in a dynamic state, assess the burden of secretions and mucosal inflammation, and obtain a bronchoalveolar lavage (BAL) from a clinically relevant segment for analysis. Previously unrecognized laryngomalacia, arytenoid prolapse, tracheomalacia or vocal cord dysfunction may result in extubation failure after single stage laryngotracheoplasty.[21,5,6] For the FFB, the level of anesthesia is highly relevant as it will strongly influence findings. The goal should be to utilize a light anesthetic with spontaneous respiration and no