Introduction:

This talk will focus on the general topics of understanding causes of obstruction in the pediatric airway. This talk will briefly highlight some general thoughts when evaluating a child with airway obstruction but will primarily concentrate on laryngeal causes of obstruction. With certain key diagnoses, discussion and updates on management will presented.

General Airway Concerns

Children can present with trouble breathing that may range anywhere from the nose to the lungs. It is important to remember that neonates are obligate nasal breathers. To that end, evaluating the nasal cavity is important. Neonates with nasal obstruction demonstrate the classic phenomenon of cyclical cyanosis – cyanosis relieved when the child cries which is then relieved by the ability to breathe then subsequently returning to attempts to breathe through its nose causing cyanosis. Children with nasal obstruction tend to have grunting, nasal flaring, mouth breathing, and snoring/audible breathing.

The table below from Bailey’s demonstrates typical signs and symptoms seen in airway obstruction from the oropharynx to tracheobronchi. Laryngeal obstruction typically demonstrates a hoarse voice in comparison to tracheobronchial obstruction where the voice is fine. Oropharyngeal obstruction can lead to deep or full voice. Stridor is typically inspiratory for oropharyngeal and supraglottic causes of obstruction while glottic and subglottic obstruction progress from inspiratory to biphasic stridor. Tracheobronchial obstruction manifests as expiratory stridor and wheezing. Finally, subglottic obstruction causes barking cough while tracheobronchial obstruction causes brassy cough.
Of final importance to discuss, a child’s airway is quite narrow. Historically, the subglottis is thought to be the narrowest aspect of the airway in a child but there is thought that the glottis may in fact be the narrowest area of the airway regardless of age. Either way, it is important to realize that just a small amount of inflammation/edema can cause drastic decreases in the airway area. In the subglottis, a typical radius is 3 mm for an area of 28.3 mm², but with just 1 mm of inflammation, the area drops to 12.6 mm² – a 56% decrease.

**General Laryngeal Signs**

Laryngeal obstruction is the most common site of stridor. Typical symptoms include difficulty with respiration, phonation, and swallowing. These children may have episodes of aspiration. With advancing obstruction, these children typically demonstrate suprasternal and intercostal retraction. Glottic and subglottic obstruction show some slightly different signs despite both causing biphasic stridor. Glottic obstruction often alters the voice causing an abnormal cry, feeding difficulties, and aspiration. Subglottic obstruction causes a hoarse cry with barking cough and patients often demonstrate xiphoid retractions and alar flaring. Some general differential diagnoses with supraglottic obstruction include laryngomalacia, vallecular cyst, saccular cyst, and supraglottic webs. A rough differential diagnosis list for glottic obstruction includes vocal cord paralysis, laryngeal webs, laryngeal stenosis, laryngeal cleft, neoplasms, and laryngospasm.
Infectious Causes

Infectious diseases present a key differential diagnosis in airway obstruction. Of the many infectious possibilities, we will concentrate on four – croup, epiglottitis, bacterial tracheitis, and recurrent respiratory papillomatosis.

Croup

Croup is the most common cause of upper airway obstruction in children from 6 months to 6 years of age. The causative agent is parainfluenza virus. Children typically present with hoarseness, barking cough, and stridor. Croup typically has a self-limited course and is most often treated by only supportive measures. One area of importance to discuss though with croup is deciding when it may be triggered by underlying anatomical issues. While children may develop croup more than once, having greater than 3 infections typically suggests an underlying anatomical disease. A recent study by Rankin et al demonstrated significant findings in the airway in children who had greater than 3 episodes of croup. Of 90 children, 53 demonstrated some abnormal finding – of which laryngopharyngeal reflux (25 children) was the most common. The remaining 28 children all had some anatomical issue. Another important point brought to light by their article was that a significant portion of these children suffered from allergies which was demonstrated on IgE and RAST testing. This article helps to provide some criteria in thinking that greater than 3 episodes of croup should raise a red flag for possible anatomical obstruction.

Epiglottitis

Epiglottitis is another disease causing airway obstruction. While once more prevalent, it has greatly decreased in frequency due to vaccination for H. influenza. Of note, the prevalence of other bacterial causes of epiglottitis have been increasing – these include S. pneumoniae and M. catarrhalis. If possible, laryngoscopy can be attempted to definitely diagnose epiglottitis. These children typically present with fever, sore throat, and mild respiratory distress which can progress to odynophagia and stridor. These episodes are easily treated with antibiotics but keeping the airway secure is of utmost importance.

Bacterial Tracheitis

Bacterial Tracheitis is a typically slowly progressive disease where patients often have a history of URI over several days. The main causative agent is S. aureus although S. pneumoniae, H. influenza, and Moraxella Branhamella catarrhalis. Patients often present with high fever and may deteriorate rapidly to having biphasic stridor. A recent study by Shargorodsky et al espoused the value of having a lateral neck X-ray which can help demonstrate tracheal narrowing. Subsequently, fiberoptic laryngoscopy can be attempted if the patient remains stable. If not, securing the airway should be the first priority. There is also significant benefit in performing a direct laryngoscopy and direct bronchoscopy to evaluate the trachea and obtain cultures. While starting with broad-spectrum intravenous antibiotics, oral antibiotics can be started once culture results are noted.
Respiratory Papillomatosis

Recurrent Respiratory Papillomatosis is a complex but benign disease characterized by episodes of disease progression requiring treatment in order to maintain an airway. While the full complexities of this disease would require its own lecture, one should have awareness of this disease as a cause of hoarseness, audible breath sounds, possible stridor, chronic cough, dyspnea, and dysphagia in children. It is caused by the human papilloma virus – primarily strains 6 and 11 (which account for at minimum 50% of cases) although numerous others have been cited. Children typically present around age 4-5 with complaints of hoarseness. It is also important to remember that there is about 1% chance of malignant transformation. One final thing to note is that a tracheotomy should be delayed as much as possible as the tracheotomy can serve to help seed and spread the disease. With these infectious causes in mind, we will move on to some supraglottic concerns.

Supraglottic Obstruction

While multiple causes exist, we will discuss the more common causes which first begins with laryngomalacia. A typical patient is usually young infant (<10 months of age) who is referred for a weak cry, noisy breathing, or for poor feeding. Laryngomalacia is the most common cause of stridor in infancy (<6 months of age). It typically peaks in the first weeks to months of life before resolution around 12-18 months. Laryngomalacia is typically classified from mild to severe. Mild forms only present with inconsequential stridor. Moderate versions demonstrate inspiratory stridor which do cause feeding symptoms. Children with severe forms demonstrate severe stridor not responsive to medical therapy and may be causing failure to thrive. In laryngomalacia, the exact etiology is not known as there has been theories regarding the neurologic problems. It is thought that the coordination of the suck-swallow-breathe sequence may be the crux of the problem. Other theories include abnormally pliable tissues within the cartilage or an anatomical excess of flaccid tissue.

Treatment for laryngomalacia centers on three levels of management – observation, medication, and surgery. Observation may be encouraged in older infants as they may begin to outgrow the disease by 8 months of age. If further symptoms persist but are mild, then medication can be started. A typical regimen may be zantac 3 mg/kg divided over three times per day or use prevacid solutabs at nighttime. They may be treated for several months if receiving benefit. A recent article brings to light a new topic of whether supraglottoplasty is underutilized. There may be a significant population of infants and toddlers who may in fact be having laryngomalacia as the cause of their breathing issues rather than adenotonsillar hypertrophy. These children may benefit from supraglottoplasty rather than adenotonsillectomy.

Further differential for supraglottic obstruction should also include saccular and vallecular cysts. They are incredibly rare and reported as infrequently as 1-2 children in 100000. Saccular cysts occur secondary to distension of laryngeal ventricle mucosa. They can be either lateral or anterior. Anterior cysts typically lead to obstruction sooner as they grow towards the airway. Posterior saccular cysts typically enlarge and may pierce through the thyrohyoid membrane, presenting as a neck mass. Vallecular cysts occur secondary to obstruction of a
submucosal salivary duct. While these children may have airway obstruction, they typically present with difficulty feeding. After diagnosis, cysts are treated with surgery.

**Glottic Obstruction**

While there are many causes of glottic obstruction, we will discuss a select few today in depth. Laryngeal webs may be present congenitally or acquired. Congenital forms occur secondary to incomplete recanalization of the larynx. The acquired forms typically occur due to intubation, infection, or trauma. Children typically present with a range of symptoms depending on the extent of the web. They may have a weak cry or be stridorous. Respiratory distress may also ensue as well as presenting with a history of recurrent croup.

The pictures above demonstrate Cohen’s classification for laryngeal webs which is based on glottic opening. Grade I as shown is mild with only 0-35% obstruction. Grade II spans from 35-50% while Grade III includes 51-75%. Grade IV is from 75-99%. Benjamin also has a classification which uses similar percentages but includes location of the web in the classification – noting if the web is truly glottic, subglottic, supraglottic, or interarytenoid. Small laryngeal webs are often anterior and may be clinically observed. They may cause some mild voice changes or audible breathing but never anything more severe. If the web is larger, it is important to assess if the web is thick or thin. Thin webs may be addressed with use of KTP Laser and application of mitomycin C topically to prevent scaring. Larger webs or thicker webs may not be as amenable to surgical treatment. These webs are best treated by an open approach or endoscopic approach. Laser may still be used for thicker or larger webs; however, a surgical flap should be made to help prevent scarring or web re-formation. Some authors suggest placement of a temporary stent. In severe cases, a laryngotracheal resection may need to be
performed with an anterior cartilage graft. This approach is typically reserved for anterior webs with subglottic extension.

**Laryngeal Clefts**

Another cause of laryngeal obstruction are laryngeal clefts. They arise due to incomplete fusion of the tracheobronchial groove and present at birth. The grooves develop and fuse in the midline typically by week 6 of life while the cricoid cartilage starts to form around week 5 and completes chondrification by week 6 also. When either fusion or chondrification does not occur, a child may develop a laryngeal cleft or tracheoesophageal fistula. Clefts are typically more common in males, seen in premature children and in cases of polyhydramnios, and in children with VACTERL syndrome.

This figure demonstrates grading of laryngeal clefts – ranging from Type I (least severe) to Type IV (most severe). Type I clefts are limited to the interarytenoid region, above level of true vocal folds. These clefts are often asymptomatic or may cause mild hoarseness. Type II clefts involve the cricoid lamina and have extension below the level of the true vocal folds. These children typically have hoarseness and mild aspiration. Type III clefts involve the entire cricoid and may contain extension into the cervical trachea. These children tend to have moderate to severe aspiration and pulmonary infections as well as regurgitation. Type IV clefts extend into the posterior wall of the thoracic trachea and can extend to the carina. These children have severe aspiration, constant pulmonary infections, and regurgitation. Large clefts create this
excessive pulmonary complications and require early evaluation and treatment. While causing airway concerns, these children infrequently have stridor.

Laryngeal clefts are diagnosed on endoscopy following suspicion of a cleft based on history. A Barium swallow can also help provide key information to suggest a laryngeal cleft. Type I and mild Type II clefts may be managed with anti-reflux medications and thickened feeds. If able to thrive, many children outgrow the detrimental effects of the cleft. If unresponsive to medication, a laser may be used to ablate the site. If needed, the raw edges may be sutured together endoscopically to heal the site of the cleft. More complex Type II clefts, type III clefts, and type IV clefts mandate an open approach. An anterior approach via a laryngofissure or a lateral approach via lateral pharyngotomy may be used for exposure.

**Vocal Cord Paralysis**

Another cause of laryngeal obstruction is vocal cord paralysis. Unilateral vocal cord paralysis presents more of a management discussion but a quick word on bilateral vocal cord paralysis. It typically is quite rare, and patients present soon after onset. Bilateral vocal cord paralysis results in immediate airway obstruction and audible stridor. These patients need to be managed aggressively by securing the airway via intubation or a tracheotomy.

Unilateral vocal cord paralysis is slightly less rare with children presenting noisy breathing, feeding difficulty, and hoarseness. Presentation of unilateral vocal cord paralysis changes with age. Neonates and infants tend to present with poor feeding, weak cry, and aspiration. They rarely present with cyanosis. Older children and adolescents typically compensate for the paralysis and may only demonstrate mild hoarseness or poor vocal projection. The causes of unilateral vocal cord paralysis are numerous and include the following:

- Traumatic delivery – Need for Forceps
- CNS malformation – Arnold-Chiari
- Cardiovocal Syndrome = Thoracic Vascular or Cardiac Anomalies + Congenital Left VCP
- Iatrogenic - PDA ligation, TEF repair, Esophageal Atresia Repair
- Infectious - Lyme Disease, EBV, Polio
- Neoplasms and Chemotherapy
- Idiopathic – nearly 50% of children with UVCP

As with any airway concern or voice issue, the first step in management is endoscopy. If possible, a flexible laryngoscopy should be performed at bedside. Once the diagnosis is confirmed and no other concurrent pathology is noted, causes of the paralysis should be explored. If no obvious cause is noted from history or recent trauma (including iatrogenic), a MRI can be performed from the brain to the thoracic inlet to identify possible lesions along the course of the vagus and recurrent laryngeal nerve. Laryngeal EMG may also be used as a key predictor and decision point for further medical management. No normal action potentials after 6 months is highly indicative of nerve injury. EMG can also be very useful in ascertaining injury after iatrogenic complications.
Management of unilateral vocal cord paralysis ranges includes speech therapy and varying degrees of surgical intervention. Concomitant lesions or injuries including laryngeal webs, crico-arytenoid fixation, subglottic stenosis, vocal cord granulomas, laryngomalacia, and tracheomalacia are frequently encountered in patients with vocal cord paralysis. Therefore, a thorough airway evaluation is necessary. In an older child where hoarseness is the primary symptom, speech therapy should be attempted first to assess level of benefit. Unlike adults, children tend to demonstrate more compensation of the working vocal cord compared to adults. In cases refractory to speech therapy, surgical intervention including vocal cord injection, medialization thyroplasty, and re-innervation procedures should be considered. Injection medialization may be a very valuable procedure in treating younger children until they reach puberty and are closer to their adult sizes (with regards to the larynx). In adults, recent studies have shown improved benefit in adults younger than 50 with re-innervation rather than medialization thyroplasty. This appears to be similar in children as well as a recent study showed improved outcomes with use of Ansa in re-innervation procedures. Children also demonstrate better ability at adaptation and providing the paralyzed cord with tone is all they require.

**Vocal Cord Granuloma**

A final thing to mention in assessment of the laryngeal causes of obstruction is a vocal cord granuloma. As with adults, these are secondary to laryngopharyngeal reflux and typically resolve with observation and anti-reflux medication. Larger lesions are rarer, but if present, they can cause significant obstruction. They may be treated with a laser and then observed along with anti-reflux medication.

**Subglottic Obstruction**

While there are several causes of subglottic obstruction, this section will concentrate on subglottic hemangioma and subglottic stenosis. Subglottic hemangiomas typically manifest within the first weeks to months of life. They are typically unilateral although may be bilateral. Approximately 50% of patients with subglottic hemangioma also have a cutaneous hemangioma. Subglottic hemangiomas are more common in females. Like all hemangiomas, they progress through a proliferative phase and then eventually an involution phase. Children typically present with poor feeding, biphasic stridor, and a weak cry. The classic thought for subglottic hemangioma is a child with recurrent croup-like infections.

**Subglottic Hemangiomas**

The treatment of subglottic hemangiomas has been greatly improved since the implementation of propranolol. A suggested regimen is starting a patient on 1 mg/kg/day divided in 3 doses. If the patient tolerates this dose, then the dose should be increased to 2 mg/kg/day starting on week 2. While on propranolol, it is important to monitor BP, pulse, and blood sugars. It is also suggested to evaluate these children every 3 months with endoscopy to assess for improvement. Further, 12 months should be a time when the child should either be weaned off of propranolol. If significant involution has not occurred or airway concerns progress, then surgical intervention should be considered.
Subglottic hemangiomas\textsuperscript{12} can be treated with endoscopic laser resection, of which carbon dioxide laser is preferred. Endoscopic laser resection is best utilized for unilateral lesions. With circumferential or bilateral lesions, there is an increased risk (6-25\%) of scarring leading to subglottic stenosis. In children where a large subglottic hemangioma is being removed and concern for scarring is high, concomitant tracheotomy may be necessary. Other options for surgical intervention include intra-lesional steroids although steroids only reduce the size of the lesion rather than speed up involution. If the hemangioma is too large for endoscopic removal, open excision via a cricoid split can be performed in conjunction with a tracheotomy.

**Subglottic Stenosis**

Subglottic stenosis can present as a congenital lesion or acquired lesion. The congenital form is incredibly rare and is due to a malformed cricoid cartilage. The more common cause of subglottic stenosis is acquired from intubation. It occurs in approximately 1-2\% of intubated children. These children often have associated factors including gastro-esophageal reflux disease, malnutrition, infections, or systemic illnesses.

A recent study sought to better understand the correlation between length of intubation and risk of subglottic stenosis. Manica et al\textsuperscript{13} performed a prospective trial of children under 5 years of age who were intubated for greater than 24 hours. They found that there was a 50\% increase in risk of subglottic stenosis with every 5 days that a child was intubated. Further, they also identified sedation as a cause of subglottic stenosis with each day of sedation correlating to a 12\% increased risk of subglottic stenosis.
Subglottic stenosis is graded via the Cotton-Myer grading system ranging from Grade I to Grade IV. Grade I spans 0-50%, Grade II from 51% to 70%, Grade III from 71-99%, and Grade IV has no detectable lumen. Subglottic stenosis is managed surgically. The options include anterior cricoid split, laryngotracheal reconstruction, cricotracheal resection, balloon dilation, sliding tracheoplasty, and tracheotomy. In grade I and mild grade II cases, observation alone may be sufficient. In moderate grade II cases, endoscopic approaches with use of balloon dilation, laser, and possible application of mitomycin C can be therapeutic. Balloon dilation can also be used with steroid injections as well and should be tried up to three times before changing in favor of open surgical approaches.

Any stenosis unresponsive to endoscopic approaches or with Grade III requires an open approach after a tracheotomy to secure the airway. Laryngotracheal reconstruction with a cartilage graft is typically preferred for grade III although other options may be more beneficial in individual cases. In grade IV cases, a tracheotomy is necessary for life and should be followed
by a cricotracheal resection. In neonates, anterior cricoid split may be used with the indications being multiple failed extubations, off ventilator support for at least 10 days, supplemental oxygen less than 30%, and weight greater than 1500 grams.\textsuperscript{15}

**Conclusion**

The causes of airway obstruction in a child are numerous and too exhaustive for an individual lecture; however, this lecture's aim was to bring to light several of those causes and highlight some recent articles which have shed new light on these pathologies. The table below from Bailey's summarizes neatly the causes of airway obstruction. The key in these children is to remember to secure the airway, and as often is true, make sure to rule out concomitant pathology.
DISCUSSION: Dr. Pine on Dr. Venkatesan’s presentation on Pediatric Airway

Do we treat kids with laryngomalacia with reflux meds? We have a chapter on this in our upcoming book in clinic but I couldn’t get Dr. Underbrink to really stick his neck out (on treatment of laryngomalacia) because he didn’t want pediatricians all over the world to throw these kinds on Prevacid. That being said, I have leaned more and more when I get sent children with decent laryngomalacia I actually put them on some reflux med. I try initially with some H2 blockers and if that doesn’t seem to help especially if they have reflux in addition then I try to gently bump them up on proton pump inhibitors.

The issues with papillomas, of course, fortunately I don’t have a whole lot of them in my practice anymore. We had lots of them at Great Ormond Street, but I think I hold with the recommendation that at least once a year you probably ought to pull off a little biopsy just to make certain there’s not something new developing. I always hate doing that because you always stir up a bunch of bleeding before you do your micromanipulating and it always makes a bit of a mess. More and more I’m trying to do these with spontaneous respiration so as to have a little bit more room to work down there. Then the other thing- and this is more for anesthesia- it’s always to communicate when you’re about to put a kid to sleep for a papilloma case you absolutely have to have that talk about how you’re going to manage the airway and one of the things that was proven to me the hard way you generally want to stay away from paralytics like they sometimes do to put the tube in because you can turn a relatively stable airway into no airway in about five seconds.

For the children that we find with glottis webs we’re always trying to look smart in front of our pediatric colleagues so nowadays if I find one I always recommend a consultation to look for the twentytwo eleven problem; there’s a big association with that and glottis webs and finally I’m glad I finally see somebody coming out and helping me with what to do radiographically with the children we find with unilateral vocal cord paralysis. We find it then I’m like well, you take the history and most of the time you come up with empty hands and I never quite know how aggressive to recommend about scanning and so forth but I like the idea of an MRI from the head all the way down. That comes with a cost certainly, and it certainly comes with the probability of another general anesthesia. So, don’t recommend these things lightly and be able to defend why you’re recommending these things. And then, as always, whether you’re going to the unit to see a child with an airway issue or you’re bringing it out to the operating room for an endoscopic evaluation, it behooves you to think a couple steps ahead in what are you likely to see and what is going to be your plan of action if you see A, B, or C, and that will keep the stress level down in the operating room and will lead to a better outcome.
Bibliography:

15. Cotton RT. Management of Subglottic Stenosis.