

TITLE: Management of the Stridulous Child

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Introduction

Stridor is a clinical sign that is routinely encountered by pediatricians, primary care and ER physicians. Children with stridor are often referred to the "airway specialists" and it is the otolaryngologist's task to identify the etiology of this "noisy breathing" that easily and understandably causes alarm for parents and other physicians. This chapter and the accompanying power point presentation will provide a methodical approach and framework for tackling this physical exam finding as well as a brief overview of some of the more prevalent causes of stridor.

Definitions

Stridor is a harsh sound produced by turbulent airflow through a partial obstruction. The nature of the sounds may be soft and tuneful; musical in quality. Important to remember is that stridor is characteristic of certain pathology but never diagnostic. Stertor, another term used to describe upper airway sounds is a snoring type of noise often made by nasopharyngeal or oropharyngeal obstruction, but may occasionally be created by the supraglottic larynx. Bearing these definitions in mind, it is clear to see that there is not a true separation between these two definitions therefore a wide differential diagnosis must be kept in mind.

Pathophysiology

In terms of physics, stridor can be explained as a combination of the Bernoulli principle and the law of conservation of energy. This, in essence, is how the Venturi principle is derived. This law can be applied to fluids and gases. The Venturi effect is the reduction in fluid pressure that results when a fluid flows through a constricted section of pipe. To satisfy the law of conservation of energy, the liquid/gas velocity must increase upon reaching the constriction while the pressure decreases. The decrease in pressure causes the narrowed, flexible airway of the child to close momentarily to obstruct airflow and cause stridor. In clinical terms, the "pipe" is the pediatric airway, while the "gas" is oxygen. An alternative way to comprehend this is the

fact that gases normally produce equal pressure in all directions when no movement is in effect. However, as gas moves forward (linear) in a tube, it produces pressure in the forward vector while decreasing lateral pressure. When a narrowed section is encountered, the pressure in the forward vector increases while precipitously dropping in the lateral direction. Again, this drop in pressure causes the pliable pediatric airway to intermittently close yielding airway obstruction/stridor.

Anatomy

There are a number of anatomical differences between the pediatric and adult airway that render them vulnerable to compromise. In the child, the larynx is situated high in the neck with the epiglottis located behind the soft palate. The pharyngeal structures are in closer proximity compared to the adult and the hyoid bone is higher. In infants, the subglottis is the narrowest portion of the airway, thus creating a conical shape in contrast to the tubular shape in adults. This is significant because the slightest trauma or inflammation can greatly reduce airway patency. Just 1mm of edema in the pediatric tracheal airway can reduce the cross sectional area to 44% of normal! Likewise, 1mm of edema at the triangular laryngeal inlet can reduce cross sectional area to 35% of normal!

Functionally, anatomic differences associated with the infant airway create a separation between the airway and digestive tract with air movement being predominantly transnasal. As child grows the larynx descends, the pharynx becomes larger to facilitate speech production and produces a common conduit for food and air passage. In turn, this increases risk for foreign bodies, food, and gastric contents to enter the airway.

Evaluation

It is easy to become overwhelmed when presented with stridor, therefore the following mnemonic may prove useful as an initial starting point to gather important details from the **history**:

SPECS-R

- Severity
- Progression
- Eating difficulties
- Cyanosis
- Sleep disturbance
- Radiologic findings
- Inquire about birth history, maternal STD, and history of intubation.

In terms of **physical assessment**, first assess the need for emergent airway intervention (ABC's). Initial evaluation should be noninvasive as the pediatric airway can be quite tenuous and compromise must be avoided. Indicators of severity include respiratory rate, level of consciousness/mental status, and accessory muscle use. Be cognizant of the fact that a child who stops using accessory muscles to breathe (or is not using them when you arrive at bedside) does not signify that all is well. On the contrary, this could be an ominous sign that the patient's condition is about to plummet. During auscultation, pay attention to not only the lungs, but the

neck, mouth, and nasal airway. If cyanosis is encountered in the absence of stridor, further investigation is warranted as cardiovascular, CNS, pulmonary or gastrointestinal factors may be culprits. (A brief algorithm to approaching an infant in respiratory distress is located in the power point presentation of this chapter.)

Once you've determined the patient is not facing imminent respiratory compromise, a more detailed and comprehensive examination may ensue. This exam should include a general assessment (weight, growth percentile, development), the nasal cavity, oral cavity and oropharynx. A heart and lung exam should always be performed. Finally, flexible fiberoptic laryngoscopy can be performed. This simple test is often the most helpful in trying to narrow down the differential diagnosis.

A formal airway evaluation in the OR is not necessary in every case. For example, in children with a history and exam (including fiberoptic), consistent with mild laryngomalacia, watchful waiting is probably all that is required. However, it is important to realize that children can have more than 1 airway issue. The flexible scope rarely allows an adequate exam of the subglottic region. In unusual or difficult cases, the ENT surgeon should have a low threshold for recommending a formal airway evaluation.

In the OR, all of the equipment (laryngoscopes, Hopkins rod-lens telescopes, and bronchoscopes) must be checked before the patient arrives to ensure they are in working condition. As a rule of thumb, a tracheostomy tray should be in room just in case an emergent surgical airway is needed. Most importantly, good communication and rapport between endoscopist and anesthesiologist is a must.

Various Etiologies of Stridor

Now, a brief review of some common causes of pediatric stridor will be examined. This approach will begin at the nose and end in the trachea. This is by no means an all inclusive list and details regarding surgical procedures of these various conditions are beyond the scope of this chapter.

Choanal Atresia (CA)

This rare disorder occurs 1 in 10,000 births with females affected more than males. There is roughly a 50/50 split between unilateral and bilateral occurrences. There are theoretically 2 types: membranous or bony; however the literature states that there are no purely membranous cases. Approximately 29% are bony and 71% mixed bony-membranous (Brown et al, Laryngoscope 1996). The pathogenesis is controversial.

Clinical Signs/Symptoms include respiratory distress/paradoxical cyanosis (i.e. cyanosis and respiratory distress that is relieved with crying), feeding difficulty and association with CHARGE syndrome:

- C- Coloboma
- H- Heart anomaly
- A- Atresia of choana

- R- Retarded growth
- G- Genital hypoplasia
- E- Ear anomalies and/or deafness

Clues to diagnosis include inability to pass 8 french catheter beyond 3.5 cm from nasal vestibule, and a mirror under nares that fails to fog on expiration. The flexible scope can also be quite helpful. Axial CT confirms diagnosis but in order to get the best radiographic picture it is helpful to decongest the nose and suction the secretions just before the scan.

Initially, management is conservative with the use of an oral airway or a McGovern nipple. Surgical approaches are utilized once the conservative measures fail. Each method has its pros and cons. The transpalatal approach gives better visualization, and a high success rate although this can damage the palate growth plate resulting in cross bite deformities. The transnasal method has less blood loss, and requires less procedure time; however, there is increased CSF leak and meningitis risk. Lastly, the laser (CO₂, KTP, Holmium:YAG) is being utilized with good success in combination with endoscopic techniques. The operating microscope with the CO₂ laser is also being employed.

Congenital Nasal Pyriform Aperture Stenosis (CNPAS)

This condition is caused by premature fusion and overgrowth of the medial nasal processes. Some believe this could represent a microform of holoprosencephaly. Associated abnormalities include a central megaincisor (60% of cases) and concomitant malfunction of pituitary/adrenal axis.

The clinical picture is very similar to CA: respiratory distress, feeding difficulty, cyclical cyanosis. The exam reveals bony obstruction of the vestibule and inability to pass a catheter/scope into the nose. A thin cut CT with emphasis on the pyriform aperture is the image modality of choice.

As with CA, management is initially conservative with use of the McGovern nipple, topical decongestants, and corticosteroids. Once conservative measures have failed, surgery is the next option. Briefly, the aperture is widened via a superior gingivolabial incision/premaxillary degloving approach to preserve the mucosa. Nasal stents are left in place 1-4 weeks.

In terms of prognosis, mild cases may resolve as the child grows but if conservative measures fail excellent long term results are usually achieved with surgery.

Retropharyngeal Abscess (RPA)

Retropharyngeal abscesses secondary to their oropharyngeal location can present as an emergent situation causing stridor. Expedient diagnosis and management are necessary.

Anatomically, the retropharyngeal space has its superior border at the skull base and extends inferiorly to as far as T6. Its posterior and anterior boundaries are the prevertebral fascia and the buccopharyngeal fascia, pharyngobasilar fascia and the esophagus respectively.

Laterally, the space is bordered by the carotid sheath. Importantly, there is an anterolateral communication with the parapharyngeal space.

This entity is more prevalent in childhood with 70% of cases being in patients 6 years old or younger. The retropharyngeal space has two paramedian chains that drain the adenoids, nasopharynx, oropharynx, paranasal sinuses and possibly the middle ear. These lymph nodes are prominent in childhood but atrophy as the child approaches adolescence. When these lymph nodes suppurate, a retropharyngeal abscess may develop.

Fever, sore throat, progressive dysphagia, and drooling are all symptoms representative of retropharyngeal abscess. Patients may also present with neck stiffness and mild torticollis. In younger children, stridor may be present, the degree of which can be correlated with the size of the abscess. On physical exam, asymmetrical posterolateral pharyngeal swelling is present and may be accompanied by cervical adenopathy.

When retropharyngeal abscess is suspected CBC with differential and lateral neck films should be ordered. Lateral neck films can be up to 90% sensitive for RPA. The gold standard, however, is CT scan with contrast as this can make clarification between retropharyngeal cellulitis or abscess.

When imaging suggests cellulitis, a trial of IV antibiotics, preferably clindamycin or ampicillin-sulbactam, is a reasonable option. However, if an abscess is initially revealed on CT scan or after repeat scanning 48 hrs after antibiotics has been initiated, incision and drainage should be performed in the OR.

Laryngomalacia

This entity is the most common cause of congenital stridor. It may manifest days/weeks after birth but symptoms usually resolve by 12-18months. The stridor in laryngomalacia is believed to be caused by prolapse of supraglottic structures into laryngeal inlet.

Signs and symptoms typical of laryngomalacia include low pitched, fluttering inspiratory stridor that peaks at 6-9months of age, has positional variations, and can be exacerbated by activity (i.e. feeding, exertion). Cyanosis is rarely produced by laryngomalacia and if it is observed, suspicion for other pathology should be high.

Physical exam with awake fiberoptic laryngoscopy is needed to confirm the diagnosis. Direct laryngoscopy/bronchoscopy is sometimes needed to rule out synchronous lesions.

Management is conservative and based on the fact that this condition is self-limited. Surgical treatment (~10% of cases) comes in the form of a supraglottoplasty which is indicated for cases with severe stridor, failure to thrive, apneas, cor pulmonale, or pulmonary HTN.

Laryngeal Cysts

Laryngeal cysts are a rare form of stridor in infants. Typical symptoms include stridor, feeding difficulty, and cyanosis.

There are two types of laryngeal cysts: ductal and saccular. Ductal cysts are the most common type. The etiology of this type is obstruction of submucous glands and they can be located anywhere in larynx but most commonly in supraglottis. Saccular cysts are the least common. They are usually congenital in infants and located in the laryngeal ventricle without communication with the laryngeal lumen.

Management of laryngeal cysts is comprised of endoscopic excision or unroofing.

Congenital Laryngeal Web

Congenital laryngeal webs arise from failure of recanalization of the larynx in the embryo. They are predominantly in the anterior glottis and associated with subglottic stenosis in cases of severe webbing. Common presenting symptoms include abnormal cry and stridor.

Diagnostic endoscopy is required for diagnosis as well as ruling out other abnormalities. There appears to be an association between anterior glottic webs and velocardiofacial syndrome. There is ample evidence to support investigating for a 22q11 deletion in any child found to have a laryngeal web. Treatment ranges from simple incision for small webs to laryngofissure with stenting for severe webbing. Endoscopic laser treatment is also an option.

Posterior Laryngeal Cleft

Laryngeal clefts arise from failure of the posterior larynx to fuse (may involve trachea). Patients are usually void of stridor but present with aspiration and hoarseness. There are 4 types of laryngeal clefts and the classification correlates with the severity:

Type I-Interarytenoid cleft; superior to the glottis

Type II-Partial cricoid cleft; extends inferior to the glottis and partially through the posterior lamina of the cricoid.

Type III- Total cricoid cleft, with or without extension into the cervical tracheoesophageal wall.

Type IV- Laryngotracheoesophageal cleft extending beyond the thoracic inlet.

Diagnosis is made by demonstration of laryngeal penetration on contrast swallow, but the confirmation, as with most laryngeal pathology, is made at endoscopy.

Surgical intervention may be avoided with mild clefts and the only intervention needed may be to thicken feeds. If aspiration continues despite conservative measures then consideration of endoscopic closure is reasonable. Surgical closure utilizing lateral pharyngotomy or laryngofissure approach may be necessary in cases of extensive clefts.

Vocal Cord Paralysis

Vocal Cord Paralysis comprises 10% of congenital laryngeal lesions. It may be congenital or acquired but most often the cause is idiopathic. Various etiologies are as follows:

- Traumatic/Iatrogenic
- Obstetric/birth trauma

- Cardiac surgery
- Esophageal surgery
- Other congenital abnormalities
- Cardiac anomalies
- CNS origin (*Chiari malformation*)

Vocal cord paralysis can exist unilaterally or bilaterally. The key differences are briefly summarized below:

Unilateral

- Breathy voice/cry
- Mild stridor and/or dyspnea
- Aspiration
- Treatment: speech therapy. If tracheotomy needed, decannulation is usually possible as the child develops

Bilateral

- Severe stridor
- Aspiration
- Treatment: tracheotomy usually required, serial endoscopies, surgery after at least 1 year status post tracheostomy w/o improvement

Evaluation of vocal cord paralysis can be seen with fiberoptic laryngoscopy while pt is awake. Despite this, laryngotracheobronchoscopy must be performed to palpate the arytenoids and rule out congenital arytenoid fixation and exclude synchronous lesions. As further work-up, an MRI of the brain, brain stem, neck and chest are reasonable if the cause is not obvious (delineate course of vagus). FEES/MBS may be utilized in cases of aspiration.

When considering management options it is important to know that vocal cord paralysis in infants usually resolves in 6-18mos, so scheduled monitoring is reasonable for the first 2 yrs. During this time, a temporary tracheotomy may be necessary. If the cord paralysis does not resolve various surgical methods are being employed such as CO2 transverse partial cordotomy, costal cartilage grafting, and arytenoidopexy w/wo arytenoidectomy (CO2 laser or external approach).

Recurrent Respiratory Papillomatosis

Recurrent respiratory papillomatosis although rare overall, is the most common neoplasm of the larynx in children. The incidence of newly diagnosed RRP in children <15yo is 4.3/100,000. The childhood onset is often diagnosed between 2-4 yrs old with males being affected more than females, however there is no gender/ethnic difference regarding surgical frequency. The childhood onset is more aggressive compared to the adult onset with 19.7 surgeries per child (~4.4 per year).

The etiology is linked to HPV types 6 & 11 via maternal-fetal transmission.

Commonly seen is the hallmark triad of progressive hoarseness, stridor, and respiratory distress. Patients most often present with dysphonia and stridor is usually the 2nd symptom to manifest as the condition worsens.

A list of current surgical and adjuvant treatments for RRP are below:

Surgical

- Microlaryngoscopy with cups forceps removal
- Microdebrider
- CO2 laser
- Phono-Microsurgical
- KTP/Nd:YAG laser
- Flash scan lasers

Adjuvant

- Interferon
- Indole-3-carbinol
- Photodynamic therapy
- Cidofovir
- Acyclovir
- Ribavirin
- Retinoic acid
- Mumps vaccine
- Methotrexate
- Hsp E7

Subglottic stenosis

Subglottic stenosis may be congenital or acquired:

- Congenital
 - Diagnosis made in absence of factors causing acquired stenosis
 - Moderate-severe stenosis=Stridor at birth.
 - Mild stenosis= Intermittent stridor
- Acquired
 - More common than congenital
 - Usually more severe and difficult to manage
 - Endotracheal intubation trauma=most common cause

In subglottic stenosis, the degree of stenosis dictates the symptoms. During cases of severe stenosis, an infant may have stridor at birth whereas mild stenosis may not manifest until a URI takes place. In acquired SGS, a clue in neonates may be a failed extubation trial. Older children may successfully extubate but present later with progressive worsening respiratory distress.

Subglottic stenosis is quantified using the Myer-Cotton grading system:

- 0-50% obstruction= grade I
- 51-70% obstruction = grade II
- 71-99% obstruction = grade III
- No detectable lumen = grade IV

When evaluating SGS, stenotic portions may be visualized on plain films however, direct laryngoscopy/bronchoscopy is needed for confirmation and airway staging.

The best way to address SGS is to avoid the occurrence altogether. Preventative measures currently being used include the use of uncuffed, polyvinylchloride endotracheal tubes, smaller tubes, and nasotracheal intubation. Conservative treatment options have the primary goal of achieving decannulation (if tracheostomy present) or the prevention of a tracheostomy by means of close observation (grades I-II). Definitive surgical options include endoscopic methods employing use of lasers, anterior cricoid split, laryngotracheal reconstruction, cricotracheal resection.

Subglottic Hemangioma

Subglottic hemangiomas comprise 1.5% of all congenital laryngeal anomalies. Female occurrences predominate with a 2:1 female to male ratio. This condition is one of the most common neoplasm of infant airway.

Clinically, subglottic hemangiomas are usually asymptomatic at birth. The majority of patients present with biphasic stridor in first 6 months and cutaneous hemangiomas are present in 50% of patients at the time of diagnosis. The lesions are characterized by rapid growth that ceases at 12 months and may resolve by 5 years of age.

When diagnosing subglottic hemangiomas, biopsy is unnecessary due to the lesion's pathognomonic appearance seen during endoscopy described as a compressible, submucosal mass with a reddish or bluish hue that is asymmetric and located in the posterior left subglottis.

The objective of treatment in dealing with subglottic hemangiomas is to preserve a stable airway while mitigating the long term sequelae of the treatment. Current treatment modalities involve tracheotomy (temporizing measure), steroids, laser excision, surgical excision, and interferon.

Vascular Causes

Congenital vascular anomalies make up 5% of stridor cases. Symptoms are caused by tracheal/bronchial external compression by:

- Innominate artery compression
- Vascular ring (double aortic arch)
- Pulmonary artery sling
- Aberrant right subclavian artery
 - Most common anomaly in mediastinum

A double aortic arch develops as a result of persistence of the fourth branchial arch and dorsal aortic root bilaterally. This is the most common symptomatic vascular ring. On the other hand, pulmonary artery slings are the most symptomatic of the noncircumferential anomalies. The right mainstem bronchus is affected in the majority of cases. Interestingly, they are associated with the presence of complete tracheal rings.

The presentation of patients with the above conditions may be subtle or present with biphasic stridor/expiratory grunting along with chronic cough, recurrent bronchitis, pneumonia, feeding difficulty, and/or failure to thrive.

Diagnostic imaging of choice is CT with contrast or MRI, however, barium esophagram may reveal filling defects characteristic of these anomalies. Plain films are of limited value. Endoscopy allows greater assessment of the degree of compression.

Absolute indications for surgery include reflex apnea, 48hrs of failed medical management, and prolonged intubation. Relative indications include recurrent infections, exercise intolerance, dysphagia causing failure to thrive, concomitant SGS, asthma, and cystic fibrosis.

Tracheomalacia

Tracheomalacia is a congenital deformity of the tracheal rings. Patients usually present with expiratory stridor or respiratory distress. Like many airway lesions, the severity of symptoms depends on the extent of the lesion.

Diagnosis is made by flexible bronchoscopy in the awake patient. During this exam, collapse of anterior tracheal wall against membranous posterior portion of the trachea can be observed.

Treatment is rarely needed as most cases are self-limited although some cases may need temporary tracheotomy. In secondary tracheomalacia, treatment is directed at the underlying cause.

Foreign Body Aspiration

The majority of foreign body aspiration occurs in patients less than 3 years old. Aspiration of various foreign bodies are responsible for approximately 150 pediatric deaths/year in US. Choking accounts for 40% of accidental deaths in children <1yo.

Coins are the most commonly ingested object while food such as nuts and seeds are the most commonly aspirated. In older children fish and chicken bones are likely as well.

As stated previously, foreign bodies can be either ingested or aspirated. The clinical presentation can help the astute clinician determine whether ingestion or aspiration is more likely:

Esophageal

- Drooling

- Dysphagia
- Emesis
- Chest pain

Airway

- Cough
- Stridor
- Cyanosis
- Wheezing
- Asymmetric breath sounds

Plain films are important in FB assessment. PA and lateral CXR are good for radiopaque objects and can still prove useful despite lack of obvious foreign body. Rigid endoscopy is warranted when clinical suspicion is high despite “innocent/negative” films.

Unstable airway foreign bodies should be dealt with at the time of presentation. Of course, not all airway foreign bodies are emergencies that warrant operative intervention. When a patient with an airway foreign body presents in stable condition in the middle of the night or “after hours”, it may behoove the endoscopist to postpone going to the OR until the appropriate team and staff is present during the light of day. These personnel are already familiar with the equipment, pediatric anesthesiology is readily available, and the situation becomes less stressful for everyone involved. In the case of esophageal foreign bodies, it is possible to closely observe in the hospital in hopes of spontaneous passage (mid/distal esophagus). The exception is when an object such as a disc battery is ingested—this requires prompt removal in the OR.

Inflammatory Causes: Croup & Epiglottitis

Below is a chart briefly summarizing the differences between croup and epiglottitis as these are often confused.

	Croup	Epiglottitis
Onset	2yo	1-5yo
Etiology	Parainfluenza virus type 1	H. Influenza Gram + bugs
Symptoms/Signs	Barking cough, inspiratory stridor	Odynophagia, “sniff position” with mouth open

Diagnostic	AP neck film="steep sign"	Lateral neck film="thumb sign"
Treatment	Racemic epi, corticosteroid, humidified O2	Airway established in OR, IV abx

DISCUSSANTS' REMARKS – Drs. Pine and Mukerji

Evaluation of Stridor in Children

Faculty Comments by Dr. Pine and Dr. Mukerji

The vital first step in evaluating any child with stridor is first to decide whether the patient requires urgent airway intervention. This decision is often made quickly and with limited information. A failure in judgment at this point places the patient at risk and puts the otolaryngologist in an untenable situation, having to later obtain an emergent airway in less than ideal circumstances with little or no equipment. If the clinical situation merits, it is safer to bring the patient to the operating room to secure the airway. Diagnostic and sometimes therapeutic endoscopy can be performed at the same time.

Fortunately, most children who present for evaluation of stridor are stable enough to undergo a complete history and physical exam. Additional lab tests and radiographs are ordered as needed. The single best test is probably flexible laryngoscopy. It allows a quick and usually excellent view of the nasal cavity, the nasopharynx, the hypopharynx and the larynx. There are small scopes (2.2mm) which can be easily passed thru an infant's nasal cavity. If such a scope is not available, the standard sized scope can usually be passed thru the mouth to obtain a view of the larynx. I only attempt this in children with no teeth. This procedure is almost always possible at the bedside with few complications. It is important to realize that flexible laryngoscopy is not a substitute for a formal airway evaluation in the operating room. There are certainly cases where the flexible laryngoscopy exam can be normal in a child with serious airway issues. (i.e. vascular compression, tracheomalacia) One must also realize that finding one problem like laryngomalacia does not rule out another problem further down in the airway. In fact, many children have more than one airway problem. Otolaryngologists should have a low threshold for recommending a formal airway evaluation to include laryngoscopy bronchoscopy and possibly esophagoscopy.

There are a host of things that can cause or exacerbate stridor in children. Having a systematic approach to the history and exam can help narrow down the differential diagnosis. Ultimately, direct airway evaluation using both flexible and rigid techniques provide the most useful information and can often guide further testing if necessary. Even in this modern age with all sorts of fancy equipment, a host of possible tests to order, there is still no substitute for good clinical judgment.

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