CHILDREN'S HOSPITAL
PEDIATRIC REVIEW
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# Pediatric Stridor

*By **Sohit Paul Kanotra, MD,** pediatric otolaryngologist at Children's Hospital and assistant professor of Otolaryngology-Head & Neck Surgery at LSU Health New Orleans School of Medicine. This issue of Pediatric Review is intended for pediatricians, family physicians and all other interested medical professionals.*

*For CME purposes, the author has no relevant financial relationships to disclose.*

## Objectives

1. Discuss the various causes of stridor in the pediatric population
2. Discuss the evaluation of stridor in children
3. Describe the management of pediatric airway disorders

Stridor is a high-pitched sound generated due to turbulent flow of air through a narrowed airway (supraglottis, glottis, subglottic and proximal trachea). Stridor is different from snoring/stertor, which is a low-pitched inspiratory noise due to nasal or nasopharyngeal obstruction, and wheezing, which is an expiratory sound produced by turbulent airflow through narrowed distal airways (bronchioles).

## Pathophysiology of Stridor

Stridor can be explained on two fundamental principles of physics:

1. Poiseulle's Law, according to which $Q = \pi Pr4/8nl$

Where Q is the flow rate through a tube, P is the pressure, r is the radius and L is the length of the tube.

2. Bernoulli Phenomenon, according to which, as the velocity of airflow increases, the pressure exerted by the airflow decreases, leading to a negative airway pressure. This results in airway collapse, producing an alteration of the laminar flow causing stridor.

## Causes

Stridor can be classified based on the phase of respiration as:

1. **Inspiratory:** due to obstruction at the level of the supraglottis and the vocal cords. The most common cause of inspiratory stridor is laryngomalacia.

2. **Expiratory:** due to obstruction in the trachea and bronchi. Examples of expiratory stridor include tracheomalacia and extrinsic compression of the trachea due to a vascular ring.

3. **Biphasic**: due to a fixed obstruction at the level of the glottis or the subglottis. Examples of biphasic stridor include subglottic stenosis and bilateral vocal cord paralysis.

**TABLE 1: CAUSES OF STRIDOR IN CHILDREN**

| SITE OF OBSTRUCTION | CAUSE |
|---|---|
| **Supraglottis** | |
| | Laryngomalacia |
| | Laryngocele |
| | Epiglottitis |
| **Glottis** | |
| | Bilateral Vocal Cord Paralysis |
| | Recurrent Respiratory Papillomatosis |
| **Subglottis** | |
| | Croup |
| | Subglottic Stenosis |
| | Subglottic Hemangioma |
| **Trachea** | |
| | Tracheomalacia |
| | Vascular Ring |
| | Complete Tracheal Rings |
| | Foreign Body |

## Evaluation of a Stridor in a Pediatric Patient

### History

The evaluation of stridor differs in the clinical setting where the patient presents. Regardless, the first step in the evaluation of a child with stridor is to assess the severity of the respiratory obstruction and the need for an emergent airway intervention. Commonly, a child presents in the office with a chronic stridor and a stable airway. A thorough history can effectively diagnose up to 90 percent of cases of stridor. A common acronym, which is used for evaluation of stridor is **SPECS (Table 2) .**

**TABLE 2: SPECS**

| | |
|---|---|
| S | Severity of the obstruction |
| P | Progression of the obstruction over time |
| E | Eating or feeding difficulties, aspiration, FTT |
| C | Cyanotic episodes, acute life-threatening episodes |
| S | Sleep disturbance |

Important components in the history include the timing and duration of the symptoms. Stridor associated with laryngomalacia presents within the first two weeks of life and progressively increases, reaching a peak around two months of life. The stridor of subglottic hemangioma, on the other hand, begins around two months of life. Stridor present at birth can be due to bilateral vocal cord paralysis, congenital subglottic stenosis or a laryngeal web. The presence of associated feeding difficulties point toward laryngomalacia, gastroesophageal reflux, a laryngeal cleft or a tracheoesophageal fistula. Past medical history significant for intubation is characteristic of subglottic stenosis. A history of previous thoracic procedures point to a vocal cord paralysis.

### *Examination*

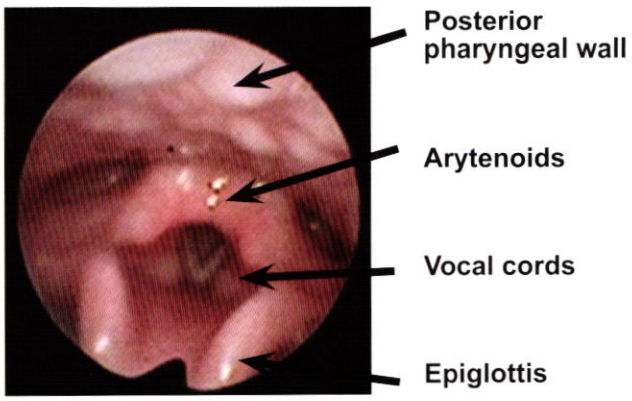
The initial evaluation is to determine the severity of respiratory obstruction. The presence of intercostal and suprasternal retractions are signs of increased respiratory effort. Fatigue or mental status changes point to a need for intubation.

The phase of respiration associated with stridor can give a clue regarding its cause. The presence of dysmorphic features like retrognathia/micrognathia point to a supraglottic collapse as a cause of stridor. The presence of cutaneous hemangiomas, especially in the beard distribution of the face, point to a subglottic hemangioma. The presence of pectus excavatum suggests significant upper airway obstruction and is an indication of surgical intervention.

### *Fiberoptic Laryngoscopy*

Awake fiberoptic laryngoscopy involving passage of a flexible endoscope through the nose to visualize the pharynx and the voice box is the initial investigation of choice for evaluation of a stridulous child.

**Figure 1: Normal larynx on fiberoptic laryngoscopy**




### *Diagnostic Radiography*

A high kilovolt or airway radiograph is a safe and inexpensive way to identify airway lesions. It is used as a screening tool for subglottic or tracheal lesions.

**Figure 2: Classic steeple sign in croup**

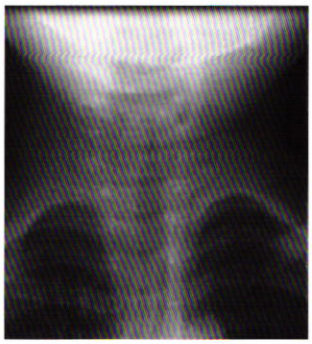
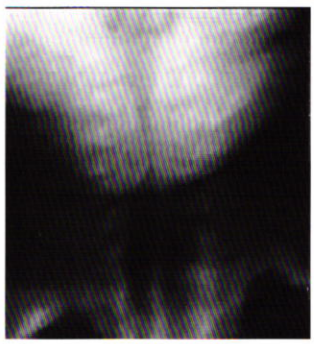


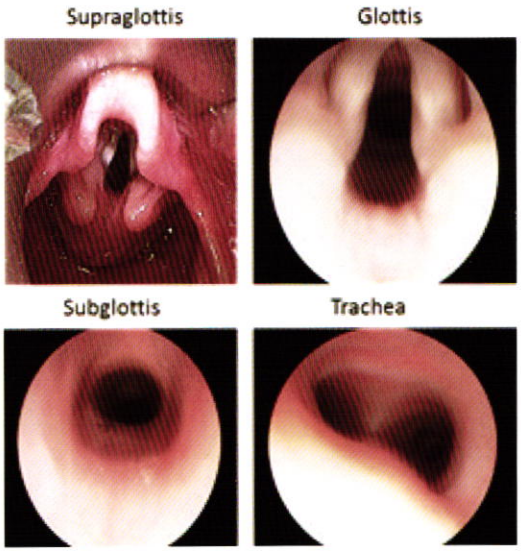
**Figure 3: Bulge in the subglottis suggestive of a hemangioma**



### *Operative direct laryngoscopy with bronchoscopy*

Diagnostic evaluation in the operating room involving a direct laryngoscopy and bronchoscopy is the gold standard for establishing a diagnosis. A systematic approach is adopted, and the airway is evaluated from above the voice box to the carina. Drug-induced sleep endoscopy (DISE) allows the determination of levels of obstruction that may occur during sleep.

**Figure 4: Systematic bronchoscopic examination**



## Common causes of stridor in children

The most common cause of stridor in the neonate is laryngomalacia. The most common cause in older infants and children is croup (laryngotracheobronchitis) and foreign body in the airway.

**Laryngomalacia** is the most common congenital anomaly of the larynx and the most common cause of congenital stridor. The stridor is typically absent at birth, begins within the first few weeks of life, increases over several months and resolves by 18-24 months of life. The stridor is inspiratory and worsens in supine position, feeding or crying. Other presentations include feeding difficulties, coughing, choking, regurgitation with feeds, slow oral intake and gastroesophageal reflux. Initial evaluation includes an in-office flexible laryngoscopy to make the diagnosis and rule out other causes. Typically, findings on flexible laryngoscopy include an omega-shaped epiglottis, tight aryepiglottic folds and excessive redundancy of the arytenoids. Approximately 40 percent of children with laryngomalacia have a secondary airway lesion, most commonly tracheomalacia. The indications of bronchoscopy include: absence of any abnormality in a patient with stridor, symptom severity doesn't correspond to laryngoscopic examination and significant aspiration symptoms suggesting tracheoesophageal fistula or a laryngeal cleft. The treatment depends on the severity of the symptoms. The mild cases can be reassured and, in the presence of reflux, started on antireflux medication and feeding changes with thickening of feeds.

**Figure 5: Typical laryngoscopy findings in laryngomalacia**

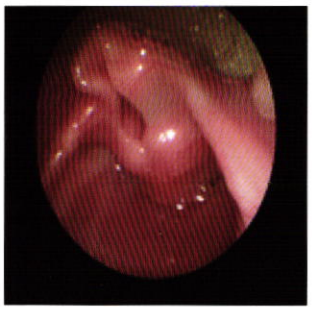


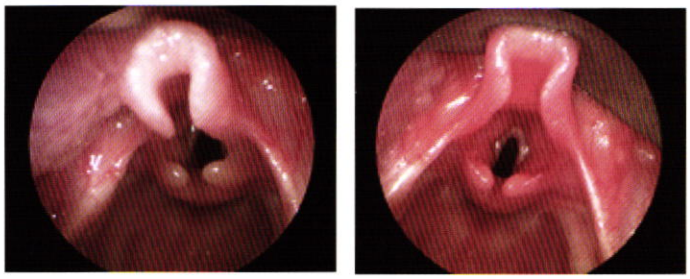
**TABLE 3: INDICATIONS OF SURGICAL MANAGEMENT OF LARYNGOMALACIA**

| |
|---|
| **Cor pulmonale** |
| **Pectus excavatum** |
| **Hypoxia/Hypercapnia** |
| **Respiratory compromise** |
| **Failure to thrive** |
| **Weight loss** |
| **Sleep apnea** |

### *Surgical Management of Laryngomalacia*

The surgical procedure is called supraglottoplasty and involves removal of redundant tissue from the arytenoids or wedge-shaped resection of the aryepiglottic folds. Surgery has been shown to improve feeding issues in children with laryngomalacia as well as improve airway difficulties.

**Figure 6: Preoperative and postoperative supraglottoplasty**



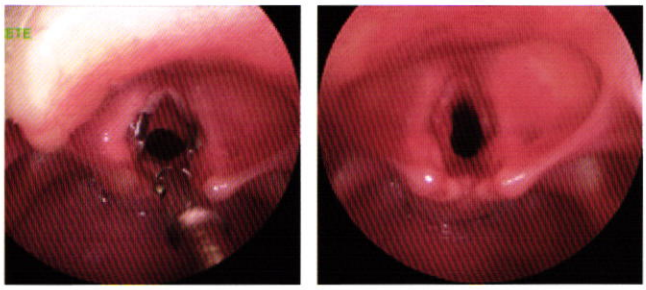
**Subglottic hemangioma** is another cause of stridor in children. The stridor typically presents as biphasic and is usually noticed at 5 weeks of age. The subglottic hemangioma proliferates at 1 year and gradually involutes over 5 years of age. Subglottic hemangioma has a 40-70 percent mortality if left untreated. The diagnosis can be made on a direct laryngoscopy and bronchoscopy. Treatment involves propranolol, which has been shown to accelerate the involution of the hemangioma.

**Bilateral vocal cord paralysis** is the second most common anomaly of the larynx. Bilateral vocal cord paralysis can either be congenital and associated with hydrocephalus, an Arnold Chiari malformation, or can be acquired due to forceps delivery, thyroid surgery or cardiac surgery. The diagnosis is made on flexible laryngoscopy. An MRI, brainstem to chest, is done to rule out various causes. Treatment options include observation, if no respiratory compromise or aspiration; neurosurgical decompression of Chiari malformation; tracheotomy and vocal cord lateralization.

## Subglottic Stenosis

Subglottic stenosis can either be congenital or acquired and presents as biphasic stridor or dyspnea on exertion, exercise intolerance and recurrent croup. Acquired subglottic stenosis is a result of intubation injury and presents most commonly within three to six weeks of extubation. Symptoms include stridor, oxygen desaturations and increased work of breathing. Symptomatic relief can be achieved with the use of racemic epinephrine and systemic steroids. Laser resection with balloon dilatation is recommended for minor grades of subglottic stenosis. If the airway cannot be restored with endoscopic techniques, the airway can be expanded using laryngotracheal reconstruction techniques.

**Figure 7: Preoperative view of subglottic stenosis due to intubation trauma. Post-operative after laser incisions and balloon dilatation**



## Recurrent respiratory papillomatosis

Respiratory papillomatosis is the most common benign neoplasm of the larynx in children. It is caused by human papilloma virus and presents typically with airway obstruction and hoarseness in children. The disease has a highly variable course and can be lifelong, respond quickly to excision or resolve by adolescence. The mainstay of treatment is excision of the laryngeal papillomas using microdebrider or $CO^2$ laser. Adjuvant therapies include cidofovir and avastin, which have shown to reduce frequency of excision and severity of recurrence.

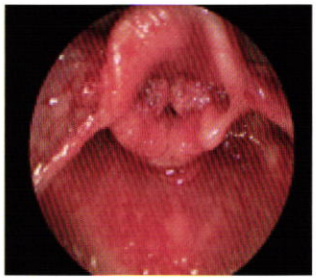


**Figure 8: Extensive laryngeal papillomatosis in a 2 year old**

## Tracheomalacia

Tracheomalacia implies collapse of the trachea, which can be congenital, due the excessive weakness of the tracheal cartilage or acquired, due to external compression by a mass or abnormal vasculature. Most tracheomalacias do not require any intervention, but severe tracheomalacia may require tracheostomy.

## Conclusion

Pediatric stridor is a symptom of upper airway obstruction and requires proper evaluation. Even though laryngomalacia is the most common cause of stridor, other airway lesions need to be excluded. A multidisciplinary approach is often essential in managing children with stridor.

# Pediatric Airway Clinic

**Appointments Phone:** (504) 896-2888
**Office Phone:** (504) 896-9832
**Location:** Children's Hospital, Main Campus

**Physicians:**
Sohit Paul Kanotra, MD
Dean Edell, MD

The Pediatric Airway Cinic is a multidisciplinary outpatient service in which children with disorders of the airway, swallowing and sleep are managed. The pediatric otolaryngologists in collaboration with pulmonologists and with support from speech language pathologists, evaluate children with complex aerodigestive problems. A multidisciplinary approach is adopted in the management of children with complex airway problems to provide a comprehensive workup and management options with inputs from different specialties.

Conditions treated include:

- Tracheostomy
- Airway obstruction
- Subglottic stenosis
- Tracheal stenosis
- Laryngomalacia
- Tracheomalacia
- Laryngeal clefts
- Base of tongue/epiglottic collapse
- Post tonsillectomy and adenoidectomy
- Severe sleep apnea
- Swallowing and aspiration problems

The clinic is held on the third Saturday of every month at the Children's Hospital main campus in New Orleans. The clinic provides a comprehensive treatment plan for children with airway issues who need input from both Otolaryngology and Pulmonology.

"This is a one of a kind clinic in Louisiana," said Sohit Paul Kanotra, MD, Pediatric Otolaryngology. "Children with complex airway issues require a multidisciplinary approach and input from various specialties in order to give them the best possible care."

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Please record your responses to the questions on the form below.
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   a. Chest X-ray
   b. Direct laryngoscopy and rigid bronchoscopy
   c. Chest CT scan
   d. No investigation

2. A 5-month-old infant presents with stridor since 2 weeks of life with coughing and choking episodes while feeding. The child is gaining weight and has no respiratory compromise, but spits up frequently. The initial treatment will be:
   a. No intervention is required
   b. Surgery
   c. Antireflux therapy
   d. Thickening of feeds
   e. Both 3 and 4

3. A 6-month-old child has been having four attacks of croup for the past four months. The child is given steroids which helps him temporarily. He also has a hemangioma on his neck. The child most likely has:
   a. Laryngomalacia
   b. Subglottic stenosis
   c. Subglottic hemangioma
   d. Complete tracheal rings

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