

# Upper Airway Obstruction

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## Educational Gap

The differential diagnosis of upper airway obstruction in children includes infectious and noninfectious causes (Table). When evaluating a child with stridor, the clinician must know how to differentiate between various anatomical anomalies (laryngomalacia, tracheomalacia, and subglottic stenosis) and infectious conditions (croup, epiglottitis, and bacterial tracheitis) to promptly implement appropriate management.

## Objectives After completing this article, readers should be able to

1. Know that upper respiratory tract infections and airway obstruction in young infants can lead to respiratory distress.
2. Know the clinical presentation of laryngomalacia.
3. Know the risks of examination of patients with suspected epiglottitis.
4. Know how to treat a child with epiglottitis.
5. Know the clinical manifestations of laryngotracheitis (croup).
6. Know the appropriate management of croup.
7. Differentiate the clinical and radiographic findings of viral croup from those of epiglottitis and bacterial tracheitis.
8. Distinguish between viral and noninfectious croup.
9. Recognize the signs and symptoms of bacterial tracheitis.
10. Know the typical clinical course of bacterial tracheitis, including biphasic illness, precipitous worsening, requirement for intubation, and relatively prolonged intubation.
11. Know the treatment of bacterial tracheitis.
12. Know the microbiology of bacterial tracheitis.
13. Know that tracheomalacia can occur as a complication of long-term mechanical ventilation in children.
14. Know that tracheoesophageal fistula may result in tracheomalacia.
15. Know the clinical manifestations of tracheomalacia.

**AUTHOR DISCLOSURE** Drs Virbalas and Smith have disclosed no financial relationships relevant to this article. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

### ABBREVIATIONS

BT bacterial tracheitis  
CT computed tomography  
HIB *Haemophilus influenzae* type B

TABLE. Common Causes of Upper Airway Obstruction

| VARIABLE         | LARYNGOMALACIA                       | SUPRAGLOTTITIS (EPIGLOTTITIS)  | LARYNGOTRACHEITIS (CROUP) | BACTERIAL TRACHEITIS  |
|------------------|--------------------------------------|--|---------------------------|---|
| Affected site    | Supraglottis                         | Supraglottis   | Subglottis                | Trachea   |
| Common ages      | 2–4 weeks, resolves around 18 months | 2–6 years  | 6–36 months               | 3 months to 6 years   |
| Onset            | Slow                                 | Rapid  | Slow                      | Rapid   |
| Stridor          | Inspiratory                          | Inspiratory, biphasic  | Biphasic                  | Biphasic  |
| Toxic appearance | Uncommon                             | Yes  | Uncommon                  | Yes   |
| Drooling         | No                                   | Yes  | No                        | No  |
| Hoarseness       | No                                   | Uncommon   | Yes                       | Possible  |
| Cough            | No                                   | Possible   | Yes                       | Yes   |
| Microbiology     | None                                 | Bacterial: <i>Haemophilus influenzae</i> , <i>Streptococcus pneumoniae</i> , $\beta$ -hemolytic streptococci, <i>Staphylococcus aureus</i> | Viral: parainfluenza      | Bacterial: <i>S aureus</i> , <i>S pneumoniae</i> , <i>H influenzae</i> , <i>Moraxella catarrhalis</i> |

## LARYNGOMALACIA

### Definitions

Laryngomalacia refers to the prolapse of supraglottic structures into the laryngeal airway on inspiration, which usually manifests as a primarily inspiratory stridor in young children.

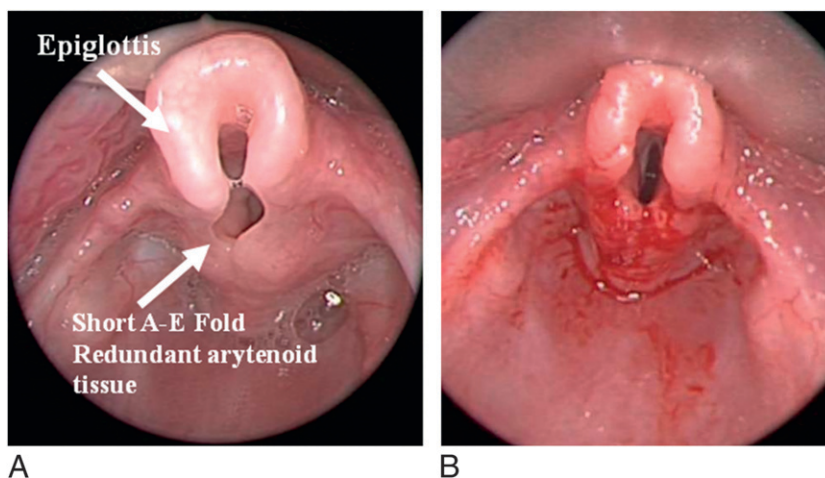
### Introduction

Laryngomalacia is the most common congenital laryngeal anomaly and is the most frequent congenital cause of stridor in infants. Often symptoms are not present at birth, and affected children typically develop stridor in the first 2 weeks of life, with symptoms becoming most pronounced in the first 2 to 4 months. This condition is often self-limited and typically resolves between the ages of 12 and 24 months.

The etiology of laryngomalacia is a matter of some debate. Historically, abnormalities in the shape and resilience of the epiglottis and other supraglottic structures were thought to be the cause of laryngomalacia. Increasingly, poor neuromuscular tone due to an incomplete integration of laryngeal sensation with brainstem-mediated reflexes (the laryngeal adductor reflex) has been implicated. This reflex, which increases laryngeal tone in response to sensation in the laryngeal vestibule, is expected to mature as the child grows. (1)

Although not universally present, the anatomical hallmarks of this condition include an omega-shaped epiglottis, which may be retroflexed; foreshortened aryepiglottic folds; and redundant arytenoid mucosa (Figure 1). The most common finding when evaluating the larynx of a patient with

Figure 1. A. Epiglottis with foreshortened aryepiglottic folds and redundant arytenoid mucosa. B. Supraglottic larynx after a supraglottoplasty in which the aryepiglottic folds were divided and the arytenoid mucosa was debulked.



laryngomalacia is prolapse of redundant tissue overlying the arytenoid cartilage.

As many as 80% of children with laryngomalacia have esophageal and laryngopharyngeal reflux. Reflux may be a consequence of laryngomalacia that results from the negative intrathoracic pressure that accompanies upper airway obstruction. The inflammation and edema that result from contact of laryngeal mucosa with stomach contents worsen airway obstruction and dull the laryngeal adductor reflex. This cycle of airway obstruction and worsening laryngopharyngeal reflux may allow symptoms of laryngomalacia to persist and worsen even as the child ages.

### Clinical Presentation

Children with laryngomalacia may have stridor at birth or may present during the first few weeks of life. Stridor is usually high pitched and inspiratory. The stridor may change with position; classically, it improves when the child is prone and worsens when the child is feeding, is lying supine, and during periods of agitation. Depending on the nature of the anatomical obstruction, some children will have less typical responses to positioning maneuvers. Some children develop respiratory distress while feeding and may not be able to tolerate an oral diet. Children may present with failure to thrive from the inability to tolerate oral diet or from burning too many calories with increased work of breathing. Hoarseness is not a symptom of laryngomalacia, and this finding may suggest a vocal cord abnormality.

### Diagnosis

The clinical presentation of an afebrile neonate with progressive stridor that is worse when feeding, supine, or agitated suggests a diagnosis of laryngomalacia. The differential diagnosis for congenital stridor includes vocal cord paresis or paralysis, glottic or subglottic stenosis, tracheal stenosis, vascular anomalies, and subglottic hemangioma. A child with a history of surgery in the neck or mediastinum, including ligation of a patent ductus arteriosus, should be evaluated for a parietic vocal cord. A history of intubation, particularly prolonged intubation as a neonate, suggests a fixed obstructive subglottic stenosis. Although subglottic hemangiomas often present without a cutaneous lesion, a child with stridor and a cutaneous hemangioma should undergo bronchoscopy to evaluate for subglottic hemangioma.

Diagnosis of laryngomalacia can be easily confirmed by flexible fiberoptic laryngoscopy to assess for prolapse of supraglottic structures on inspiration. This procedure can be performed at bedside or in the office. This examination can also evaluate the child's vocal cord mobility.

Plain radiography of the neck can provide information regarding other potential causes of stridor. Airway radiography

can identify narrowing of the subglottis that may suggest a hemangioma or scarring consistent with a subglottic stenosis. In a child with recent onset of stridor, upper airway radiography may identify inflammatory or infectious causes of upper airway obstruction, such as croup, epiglottitis, and bacterial tracheitis (BT). A barium swallow can also be helpful in identifying compressive vascular rings formed by an anomalous formation of the great vessels, which can cause stridor in infancy.

### Management

Most children with laryngomalacia experience spontaneous resolution of their stridor between ages 12 and 18 months. Children with intermittent, mild inspiratory stridor who are feeding well and gaining weight without any cyanotic episodes or acute life-threatening events usually do not require surgical intervention. If symptoms are mild in character and improving over time, the child may not require a referral to an otolaryngologist for evaluation. These children with mild disease should be followed up to monitor progression of the disease and ensure adequate weight gain. Children with atypical symptoms, significant dyspnea, or worsening stridor should be evaluated by an otolaryngologist. Children with failure to thrive, episodes of apnea or cyanosis, progressive or severe stridor, or inability to tolerate an oral diet are likely candidates for surgical intervention and require an urgent referral to an otolaryngologist or inpatient admission for evaluation.

Children with laryngomalacia and symptoms of gastroesophageal reflux disease may benefit from treatment with histamine<sub>2</sub>-blockers or proton pump inhibitors. (1) These patients may also benefit from some simple lifestyle changes to decrease reflux, such as keeping them upright for 30 minutes after feeds, feeding slowly with frequent breaks for burping, and elevating the head of the mattress 20° while sleeping. Breastfeeding is usually best for infants with reflux because it is more hypoallergenic than formula and is digested twice as fast as formula. If breastfeeding is not chosen or is not possible, changing formula can help some infants. If the infant has a milk intolerance, giving the infant formula that is milk based can make reflux symptoms worse. Mothers who are breastfeeding can try to eliminate foods that can make reflux worse. Dairy products are a big offender, as are caffeine, fatty foods, spicy foods, and citrus fruits. If eliminating these products seems to help, parents can slowly start to reintroduce one thing at a time into the diet and watch the infant's reactions. Appropriate medical treatment of reflux disease can reduce glottic inflammation and may significantly improve symptoms of laryngomalacia.

In children with severe laryngomalacia, surgical intervention can provide significant improvement in respiratory and feeding symptoms. Generally, the indications for surgical

intervention include severe respiratory distress, failure to thrive, cyanotic episodes, acute life-threatening events, or inability to tolerate an oral diet. Approximately 10% of children referred to an otolaryngologist for laryngomalacia undergo surgical intervention. Surgery involves a supraglottoplasty. In this surgery, usually the aryepiglottic folds are divided, and any redundant tissue overlying the arytenoids complexes is removed. Sometimes the epiglottis itself is also addressed (Figure 1). In those children with symptoms severe enough to warrant supraglottoplasty, an endoscopic evaluation of the lower airway should also be performed to evaluate for concurrent tracheobronchial abnormalities. Up to 20% of children with significant laryngomalacia will have a synchronous airway lesion identified on endoscopy.

## EPIGLOTTITIS

### Definitions

Epiglottitis describes inflammation of the epiglottis. The term *epiglottitis* typically refers to a disease process that involves the epiglottis and adjacent supraglottic structures and may more accurately be termed *supraglottitis*.

### Introduction

Supraglottitis is an important infectious cause of airway obstruction in children and adults. In affected children, edema narrows the supraglottic airway and alters the shape of the epiglottis, typically causing that structure to curl posteriorly and inferiorly into the airway (Figure 2). Because of this obstruction, the child may be unable to clear his or her secretions, further compromising the patient's airway.

The epidemiology of this disease is changing rapidly because of routine vaccination against *Haemophilus influenzae* type B (HIB). Since introduction of the vaccine, the incidence of supraglottitis has decreased markedly in young children. However, HIB remains the most common pathogen associated with epiglottitis in children. This bacteria causes epiglottitis even among vaccinated children. Other causes include other types of *H influenzae* (A, F, and non-typeable), *Haemophilus parainfluenzae*, *Streptococcus pneumoniae*, *Staphylococcus aureus*, other streptococci, *Klebsiella*, and *Pseudomonas*. Multiple viruses have also been implicated in the pathogenesis of supraglottitis.

### Clinical Presentation

Epiglottitis presents more commonly in children ages 2 to 6 years, although it can affect children and adults of any age. Older children and adults may experience severe throat pain. Young children, however, typically present with rapid onset of respiratory distress, high temperatures, and a muffled

voice. Drooling and dysphagia are also common. Stridor is frequently noted, but unlike croup, a cough and hoarseness are not characteristically present. Children commonly sit still in the hallmark "tripod" position, leaning forward on outstretched hands with the neck extended. This position is believed to increase the caliber of the supraglottic airway. These patients appear very uncomfortable and are often anxious.

### Diagnosis

The examination and diagnostic techniques used in a patient with suspected supraglottitis vary, depending on the severity of the illness, the age of the patient, and the clinical suspicion. A clinical diagnosis of supraglottitis may be presumed in children with classic symptoms of supraglottic airway obstruction, including stridor, tripod positioning, muffled voice, and absence of cough. Although supraglottitis and croup share some common characteristics, the subtle differences in the typical presentation often suggest one diagnosis over the other.

In a patient with mild or intermittent stridor in whom you suspect supraglottitis, physical examination should be performed with care not to provoke anxiety, which may precipitate respiratory distress. In older children who are cooperative, examination of the pharynx without a tongue blade may reveal an enlarged epiglottis. The oropharynx, however, is typically normal with pooled secretions. Gentle manipulation of the laryngotracheal complex, particularly the hyoid, may be painful. Flexible fiberoptic laryngoscopy or rigid laryngoscopy with a 70° endoscope may be used by an experienced practitioner to establish a diagnosis of epiglottitis. This examination should be reserved for patients with mild symptoms who are not in respiratory distress.

Neck radiography can help differentiate epiglottitis from other causes of stridor but is not necessary if the clinical suspicion of epiglottitis is high. In a young child or a child in whom a pharyngeal examination would provoke anxiety, one may consider lateral neck radiography to assist with the diagnosis only if that child's diagnosis is unclear and the child is medically stable to undergo these studies. Neck radiography should be avoided if it causes the child anxiety or delays definitive management of presumed supraglottitis. If performed, lateral neck radiography can reveal an enlarged and rounded epiglottis projecting into the hypopharyngeal airway. This finding is commonly referred to as the thumbprint sign (Figure 3). Thickened aryepiglottic folds with obliteration of the vallecular space are also suggestive of epiglottitis.

### Management

Examination of a child with severe obstructive symptoms should be performed cautiously. Any intervention that causes discomfort, forces the child to move from his or her adopted

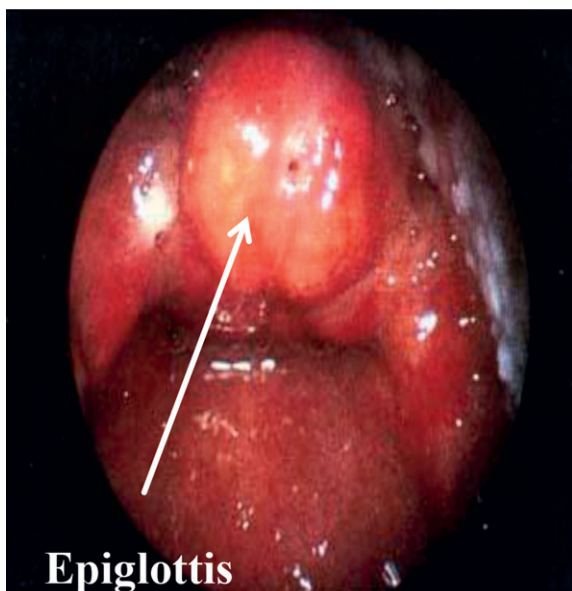


Figure 2. An inflamed epiglottis in a patient with supraglottitis.

posture, or elicits crying can worsen airway obstruction. Exacerbation of the child's anxiety can provoke tachypnea with epiglottic prolapse, laryngospasm, and cardiopulmonary arrest. In cases of severe obstruction, characterized by stridor with retractions, immediate otolaryngology and/or anesthesiology consultation should be obtained for airway evaluation and management. Diagnosis may be confirmed during the establishment of an airway in a controlled setting, such as the operating room. Attempts to visualize the epiglottis should not be performed in an environment that is not equipped to manage an obstructed airway.

Establishment of an artificial airway is determined based on the severity of the obstruction, the age of the child, and

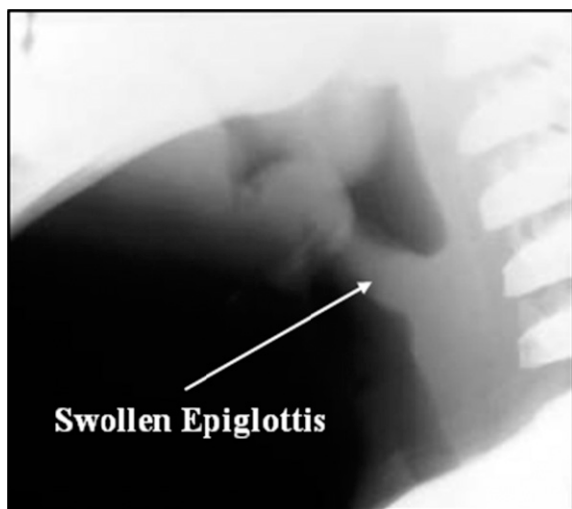


Figure 3. The arrow is pointing to swollen aryepiglottic folds; the swollen epiglottis is located anterosuperior to the aryepiglottic folds.

the degree of suspicion for epiglottitis. Any patient with presumed epiglottitis and symptoms of severe obstruction should be intubated in a controlled setting, ideally the operating room. Older children with rapid onset of symptoms and those who are diabetic or otherwise immunocompromised may also be candidates for intubation. The threshold for intubating a young child with epiglottitis should be low. Because of the smaller airway and tendency toward rapid disease progression, there is a consensus that children younger than 5 years with epiglottitis should be intubated whether or not they have respiratory distress. A systematic review of 749 children with epiglottitis found a significant reduction in mortality in children who are intubated rather than observed (6.1% vs 0.92%). (2) Some studies describe management of selected children with epiglottitis without the establishment of an artificial airway. This may become increasingly plausible with the decreasing rate of HIB epiglottitis because evidence suggests that HIB epiglottitis is associated with a more severe disease course when compared with other bacterial pathogens

Parenteral antibiotics should be administered to cover HIB and respiratory flora, including *Pneumococcus*,  $\beta$ -hemolytic streptococci, and *S aureus* (including methicillin-resistant *S aureus*). Although resistance patterns vary by community, a commonly used empiric regimen includes a third-generation cephalosporin (ceftriaxone or cefotaxime) and appropriate methicillin-resistant *S aureus* coverage (vancomycin or clindamycin). The antibiotic regimen should be narrowed when culture results become available. Often a 7- to 10-day antibiotic course is adequate, although clinical response to treatment may vary.

The role of corticosteroids in the management of epiglottitis is not clearly established. Retrospective studies have not found efficacy of corticosteroids in reducing length of hospital stay, length of intensive care unit observation, or duration of intubation. No randomized studies have been performed to examine corticosteroid use in the management of epiglottitis.

Intubation is often necessary for approximately 48 hours to allow appropriate response to intravenous antibiotics. Extubation can often be performed safely when an air leak is present around the endotracheal tube. Visualization of the larynx, either by flexible fiberoptic laryngoscopy or direct laryngoscopy in the intensive care unit, should be performed to verify resolution of obstructive edema in the supraglottis.

## LARYNGOTRACHEITIS

### Definitions

Laryngotracheitis, or croup, refers to inflammation of the glottis and subglottis. The inflammatory edema produces stridor and a characteristic barking cough.

## Introduction

Viral laryngotracheitis is the most common infectious cause of upper airway obstruction in children. Inflammation and the concomitant narrowing of the subglottis are the primary sources of airway obstruction. Inflammation and edema in the subglottis cannot expand the cartilaginous cricoid ring, which consequently projects into the airway lumen. This narrowing produces both stridor and the characteristic barking cough. Obstruction at this level may also generate a large, negative intrathoracic pressure, resulting in collapse of the cervical trachea, which further compromises an affected child's respiratory status.

Croup is the most common cause of stridor in the febrile child. It is approximately 1,000 times more common than epiglottitis and is most common in children ages 6 to 36 months. Croup occurs more frequently in the late fall and winter months, with peak frequency coinciding with parainfluenza type 1 infections.

Parainfluenza types 1 and 2 are the predominant microbes responsible for croup, with type 1 being most common. Other viruses, including respiratory syncytial virus, influenza A and B, human coronavirus NL63, and herpes simplex virus type 1, have all been implicated. In regions where measles persists, that virus may produce particularly severe croup symptoms. Viral croup can be transmitted through contact with infected respiratory secretions. For parainfluenza virus type 1, the child may begin to show symptoms between 2 and 6 days after exposure to the virus.

Bacteria have also been implicated in laryngotracheitis, although this is frequently a secondary infection of glottic and subglottic mucosa previously compromised by a viral infection. The most common bacterial isolates include *S aureus*, *S pyogenes*, and *S pneumoniae*. (3) *Mycoplasma pneumoniae* is thought to cause mild symptoms of croup.

## Clinical Presentation

This condition typically presents in children ages 6 to 36 months. Younger children typically present with hoarseness and a barking cough, whereas signs of lower airway disease are rarely seen. The characteristic cough is uniformly present, and lack of this symptom should suggest a different diagnosis. In older children, hoarseness may be significant and may be the predominant symptom. Stridor may manifest only after significant disease progression narrows the glottic and subglottic airway significantly. Children with croup rarely exhibit the marked respiratory failure, fever, and anxiety seen in epiglottitis and BT.

Many children experience a prodrome similar to other viral upper respiratory tract infections. Symptoms may

include low-grade fevers, nasal congestion, and rhinorrhea. Hoarseness, the hallmark barking cough, and stridor typically develop in a short period, frequently only 12 to 24 hours.

## Diagnosis

The diagnosis of croup is often made clinically based on the characteristic presentation of a child with a barking cough and a preceding viral prodrome. Although a complete blood cell count may reveal leukocytosis, the white blood cell count is commonly normal.

Evaluation by fiberoptic laryngoscopy may reveal signs of inflammation in the soft tissues of the glottis and subglottis with possible restricted vocal cord mobility. In severe cases of laryngotracheitis, the subglottic airway may be appreciably narrowed due to mucosal edema. Mucous, fibrinous exudates or pseudomembranes can further narrow or entirely obstruct an inflamed subglottic airway. Fiber optic examination is not necessary to establish the diagnosis, which is normally made based on clinical suspicion with or without radiography.

Radiographic imaging is not routinely necessary but may help identify the site of upper airway obstruction if the diagnosis is unclear. In a child with moderate to severe disease, subglottic narrowing can be seen on anteroposterior and lateral airway radiographs. In a normal anteroposterior radiograph of the subglottis, the radiolucent tracheal airway reaches the vocal folds superiorly to form a convex outline in the shape of shoulders. The anteroposterior radiograph of a narrowed subglottis will show the tracheal air column narrowing progressively toward the subglottis due to the mucosal edema beneath the vocal folds. This narrowing is commonly referred to as a steple sign (Figure 4).

## Management

Croup is commonly mild and self-limited; however, severe episodes may involve significant upper airway obstruction with respiratory failure. Careful history and physical examination can identify those children with severe airway obstruction. Most children with croup can be managed as outpatients, with less than 15% of children requiring hospital admission and only 1% to 5% requiring intubation during hospitalization. (4) It is critical to differentiate between milder versions of the disease and children who are at risk for airway obstruction. Signs of a more aggressive illness include biphasic stridor, rapid respiration, retractions, and agitation. Oxygen desaturation and cyanosis are late signs of severe obstruction.

In mild cases of croup, the child presents with a barking cough and mild hoarseness along with other upper respiratory tract symptoms, such as nasal congestion, rhinorrhea, or sore



**Figure 4.** Anteroposterior radiograph of a narrowed subglottis showing the tracheal air column narrowing progressively toward the subglottis due to the mucosal edema beneath the vocal folds.

throat. The child with mild croup may have no stridor or retractions at rest but may develop these signs when agitated. In the clinic or emergency department, children with mild croup may be treated with humidified air and a single dose of oral corticosteroids. If more concerning symptoms of stridor, tachypnea, retractions, or agitation are not present, these children can typically be safely discharged after the caregiver has been educated regarding the signs of disease progression. At home, children can be treated supportively with humidification and antipyretics. Although studies have not found any benefit to humidified air, significant anecdotal evidence has made this element of supportive care standard in the management of croup regardless of severity. At home, a child with mild croup may experience some symptom relief and improved expectoration of secretions by sitting in a closed bathroom while steam from a hot shower fills the room.

A child with moderate croup has a barking cough and stridor with mild to moderate retractions at rest. The child, however, exhibits little or no agitation and has normal air entry. These children should be managed in the emergency department or an office that is appropriately equipped to support children with upper airway obstruction. Oral or intravenous corticosteroids should be administered to control the progression of edema. One study suggests that the efficacy of this medical management has reduced the rate of hospital admissions for croup by up to 86%. (5) Nebulized

racemic epinephrine or l-epinephrine should be considered in children with moderate croup. The local vasoconstriction in the subglottic mucosa produced by epinephrine can generate rapid improvement in symptoms. Corticosteroids are necessary in any patient treated with racemic epinephrine to limit the effect of rebound edema that may occur 2 to 3 hours after the use of epinephrine. Racemic epinephrine may be administered in the outpatient setting as long as the child is observed beyond the 3-hour window within which rebound mucosal edema is expected. Often children with mild to moderate croup can be managed at home with oral corticosteroids and supportive care. Any child younger than 6 months and children with supplemental oxygen requirement, increased work of breathing, lethargy, or poor oral intake should be admitted for inpatient observation.

Children with severe croup may present with pronounced stridor at rest, although this symptom may appear to improve as obstruction worsens and air entry decreases. The patient may exhibit severe retractions and significant agitation, although this, too, can subside as the child tires and becomes lethargic. Only in near-complete subglottic obstruction will reduced oxygen desaturation and cyanosis be evident. It is critical to recognize the symptoms of severe obstruction in the context of rapidly progressing disease and to treat the child quickly and aggressively. Decreasing stridor may indicate worsening disease. Clinicians should be sure that the child with decreasing stridor is exchanging an appropriate volume of air with each breath.

As with moderate croup, children with severe symptoms will require corticosteroids, nebulized epinephrine, and humidified oxygen. In the acute management of severe croup, nebulized epinephrine can be repeated as often as every 15 to 20 minutes. Repeated administration will require that the patient be cared for in a monitored setting due to the cardiac effects of the epinephrine. Children with severe obstruction may also benefit from the administration of heliox. This mixture of helium and oxygen is significantly less dense than air, which facilitates the laminar flow of the gas through a narrowed airway.

Agitation and anxiety may exacerbate airway obstruction, so a child with severe symptoms must be kept comfortable and managed with caution. In these children, the clinician must consider the risks of additional interventions, such as placing an intravenous catheter, examining the pharynx, and even undergoing neck radiography without a caregiver.

Bag mask ventilation and endotracheal tube intubation are rarely necessary. Approximately 1% of children seen in the emergency department for croup will require

intubation. In the event that a child with progressive respiratory failure requires intubation, an uncuffed endotracheal tube that is at least 0.5 mm smaller than the standard age-appropriate tube should be used. Intubation should be considered only after other means of noninvasive support have been attempted. Intubating a child with croup who has significant inflammation of the subglottis may predispose the child to developing subglottic stenosis after extubation.

## RECURRENT OR ATYPICAL CROUP

Spasmodic croup, also called frequently recurrent croup or nocturnal croup, is a noninfectious variant of croup. This condition presents with symptoms similar to viral laryngotracheitis, including the hallmark barking cough and inspiratory stridor. This disease entity is characterized by nighttime symptoms, which begin suddenly and resolve rapidly. The symptoms frequently recur, often within hours or on successive nights. Although spasmodic croup is frequently associated with a mild viral prodrome, it is believed that the subglottic edema in spasmodic croup may be allergic in origin. Some studies suggest that gastroesophageal reflux disease may play a role in this entity.

In both infectious and noninfectious croup, alternative diagnoses should be considered in children with recurrent croup symptoms or symptoms that fail to respond to appropriate medical management. Such signs may indicate a fixed or evolving subglottic pathologic origin. In children ages 2 to 6 months who present with stridor without a history of endotracheal intubation, the diagnosis of subglottic hemangioma should be considered. Patients with subglottic hemangioma may also have a cutaneous hemangioma, although the absence of a cutaneous hemangioma does not rule out a subglottic hemangioma because these lesions occur in isolation in most cases. In children with a history of endotracheal intubation, suspicion should be raised for subglottic scarring and a resultant stenosis. In cases of intransigent or atypical presentations of croup, the child should undergo an evaluation for other less typical causes of croup. Direct laryngoscopy and bronchoscopy to evaluate airway narrowing should be considered. Radiographic evaluation of the subglottis with magnified airway radiographs and/or esophagrams can also be helpful in this setting. Typical infectious croup should happen 1 to 2 times in a child's life. Children with a prolonged course, multiple recurrent episodes, severe symptoms, age outside the typical range (6–36 months), or previous history of endotracheal intubation should receive consultation with an otolaryngologist for evaluation of the airway.

## BACTERIAL TRACHEITIS

### Definitions

BT, also called bacterial croup or membranous croup, is a bacterial infection of the subglottic airway. Mucosal inflammation as well as thick, purulent exudates and pseudomembranes can cause severe airway obstruction.

### Introduction

BT is a rare and potentially life-threatening airway infection that almost always occurs as a secondary infection of the subglottic mucosa superimposed on a viral laryngotracheitis. As a result of the preceding viral infection, the subglottic and tracheal mucosa becomes inflamed and susceptible to bacterial infection. The secondary infection may come from concomitant bacterial infections in the upper airway or from species colonizing the laryngotracheal region. Unlike other infectious causes of upper airway obstruction, in BT the lower airways are frequently involved.

There is significant variability in the severity of BT. In its severest form, BT can result in complete obstruction of the subglottic or tracheal airway. Early in the disease course, inflammation produces mucosal edema with a narrowing of the tracheal lumen. As the disease progresses, the child may exhibit a cough productive of copious purulent exudates. These secretions, however, can become increasingly inspissated and difficult to expectorate.

In severe BT, pseudomembranes form along the trachea. As this membranous disease sloughs off, it may collect in the airway, worsening the child's airway obstruction. Mucosal ulcerations form in the denuded trachea, and erosion into the tracheal cartilage is possible.

Blood cultures will rarely yield bacteria, but tracheal cultures typically yield results. BT is commonly polymicrobial, with *S aureus* being the most frequently isolated species. *S pneumoniae*, *S pyogenes*, *H influenzae*, and *Moraxella catarrhalis* also appear in most case series. Influenza A is the virus that most commonly renders a patient susceptible to BT. Influenza B, respiratory syncytial virus, parainfluenza virus, measles, and enterovirus have all also been associated with BT.

### Clinical Presentation

BT is usually seen in otherwise healthy children between the ages of 3 months and 6 years. Children younger than 3 years may have more severe symptoms, in part due to smaller airway diameter.

Children with BT may present to the emergency department with recent onset of cough and progressive stridor suggestive of upper airway obstruction. Caregivers will often describe preceding signs and symptoms of a viral upper



respiratory tract infection followed by a marked and rapid escalation of symptoms, including high temperature and worsening respiratory status. Although significantly less common, BT can represent a primary infection with no preceding upper respiratory tract infection. Case reports also describe children in whom aspiration of bacteria from a bacterial sinusitis or pharyngitis was thought to cause the tracheal infection. Children with BT typically have concomitant lower airway involvement (tracheobronchitis), and rales or wheezes may be evident. Approximately 50% of children with BT develop a concurrent pneumonia. The airway obstruction in this disease is often profound. Most children with BT (53%–91%) require intubation. (6)(7) Children with preexisting tracheostomy tubes have a higher incidence of BT. The mucosal injury that occurs because of the mechanical manipulation of the tracheal mucosa by the artificial airway renders these patients more susceptible to bacterial superinfection. In addition, the tracheostomy tube and the ventilation circuits are frequently colonized by bacteria, including *S aureus*, *S pneumoniae*, *H influenzae*, *M catarrhalis*, *Acinetobacter* species, *Klebsiella pneumoniae*, *Pseudomonas aeruginosa*, and *Escherichia coli*.

### Diagnosis

The cough and biphasic stridor exhibited by children with BT suggest upper airway obstruction, and differentiation of the various causes of infectious obstruction is typically made on a clinical basis. If a child with suspected croup experiences no relief after the administration of corticosteroids and nebulized epinephrine, the diagnosis of BT should be considered. A child with epiglottitis may also appear toxic with an acute onset of high temperature and stridor; however, children with BT are typically able to swallow their secretions, and a complete blood cell count is more likely to reveal leukocytosis. Rather than the classic tripod seen in patients with epiglottitis, children with BT often prefer to lie flat.

Neck radiography in a stable child may help elucidate the diagnosis; however, there are no pathognomonic signs of BT on plain radiography. Anteroposterior neck radiographs characteristically show the subglottic narrowing seen in laryngotracheitis (Figure 2). In BT, the edges of the tracheal air column may be irregular or poorly defined due to the swelling, pseudomembranes, and exudates found on the tracheal wall. These findings may be most prominent on lateral radiographs. The pseudomembranes and exudates may also appear as opacifications within the tracheal lumen. Chest radiographs may show pulmonary infiltrates, air trapping, or atelectasis.

Although clinical and radiographic findings can suggest BT, only airway endoscopy can conclusively confirm the

diagnosis. The supraglottis and glottis are typically not involved, distinguishing this from supraglottitis. Visualization of the subglottis and trachea will reveal inflamed and irregular mucosa. Pseudomembranes or thick, purulent secretions were noted on tracheoscopy in 89% of cases in one recent report. (7)

### Management

Narrowing of the subglottic and tracheal airway combined with a patient's inability to expectorate secretions cause severe airway obstruction. While the patient is intubated, aggressive tracheal and pulmonary toilet should be maintained, including frequent saline lavage and suctioning through the endotracheal tube. Flexible or rigid bronchoscopy may be necessary to adequately suction tenacious secretions and remove pseudomembranes that have been sloughed into the airway. In severe cases, it may be necessary to leave the patient intubated for 7 days or longer. Extubation should take place only when the patient is afebrile, an air leak is present, and tracheal secretions appear manageable.

Culture-directed parenteral antibiotics should be administered at presentation and continue after extubation. In an emergency scenario for which a Gram stain is not available, immediate therapy may include vancomycin to target methicillin-resistant *S aureus* and a third-generation cephalosporin, such as ceftriaxone, to treat gram-negative organisms and mixed flora. As the results of Gram stain, culture, and sensitivity testing become available, antibiotics should be appropriately targeted. After the patient has been safely extubated and improvement has been maintained with intravenous antibiotics, a 10- to 14-day course can be completed with oral therapy.

## TRACHEOMALACIA

### Definitions

Tracheomalacia describes an anomaly of tracheal cartilage that allows dynamic collapse of that structure during respiration.

### Introduction

The dynamic collapse of tracheal cartilage can be due to an intrinsic defect in the cartilaginous tracheal rings (primary tracheomalacia) or from extrinsic compression of the trachea (secondary tracheomalacia). In primary tracheomalacia, airway collapse is typically seen on expiration when the intrathoracic pressure is positive relative to the tracheal lumen. Rarely, when the extrinsic lesion is in the cervical trachea rather than intrathoracic, patients can present with inspiratory or biphasic stridor.

Tracheomalacia is categorized in the following manner:

- Type 1 is due to an intrinsic abnormality in tracheal cartilage, which weakens the typically rigid tracheal rings. This defect may arise from a tracheoesophageal fistula or polychondritis of the tracheobronchial tree.
- Type 2 is caused by extrinsic tracheal compression, which may be related to aberrant mediastinal vasculature, neoplasms, or lymphatic malformations.
- Type 3 is acquired from prolonged inflammation or irritation. Most commonly, this results from a tracheotomy or prolonged endotracheal tube intubation. Irritation from laryngopharyngeal reflux or local infection may also produce type 3 tracheomalacia.

### Clinical Presentation

Patients with tracheomalacia may present with stridor, wheezing, recurrent barking cough, or frequent respiratory infections. Rarely, an acute life-threatening event may be due to tracheomalacia. The severity of the presenting symptoms depends on the degree of collapse, and children with significant collapse may exhibit respiratory distress. In primary tracheomalacia, symptoms tend to improve as children age and the diameter of their lower airway increases.

### Diagnosis

Static imaging techniques, such as chest radiography, computed tomography (CT), or magnetic resonance imaging, have little role in the diagnosis of tracheomalacia. Definitive diagnosis is obtained during bronchoscopy with the child breathing spontaneously. This technique allows direct observation of dynamic collapse of the tracheobronchial tree.

A barium esophagram can be used to assess for abnormal mediastinal vasculature that may compress the trachea and esophagus. Magnetic resonance angiography is also a sensitive technique for evaluating such vascular anomalies but is not generally used as a screening tool. Dynamic imaging techniques, such as airway fluoroscopy, can be used to identify tracheomalacia. Newer technologies, such as cine CT and multidetector CT with end-inspiratory and end-expiratory images, may have a role for certain patients.

### Management

Type 1 tracheomalacia rarely requires long-term management. As children grow and the caliber of their trachea increases, symptoms frequently resolve spontaneously. In those cases in which obstruction is severe and disabling, intervention may be necessary. Positive pressure ventilation

at night or during disease exacerbations is frequently effective in supporting the airway.

These children with tracheomalacia are often mistakenly treated as if they have reactive airway disease. Children with reactive airway disease that is refractory to standard medical therapy should be referred to a pediatric pulmonologist for evaluation of tracheomalacia. When the diagnosis of tracheomalacia is established, medical therapy tailored specifically for tracheomalacia can be very effective at improving symptoms.

Surgical intervention may be considered for children with severe obstructive symptoms, failure to thrive, history of acute life-threatening event, or recurrent pulmonary infections. Surgery is often required for cases of type 2 tracheomalacia to relieve the external obstruction. Type 2 tracheomalacia, in contrast to type 1, generally will worsen over time until surgery is performed to relieve the obstruction. Aortopexy, in which the aorta is suspended from the sternum, offers effective relief of tracheal compression by the aorta and the innominate artery. Placement of airway stents for treatment of tracheomalacia is controversial because of the high rates of complication with these devices. Three-dimensional printing techniques have been successfully used to fashion a customized resorbable stent placed outside the airway to open the malacic segment. (8)

## Summary

- Children with laryngomalacia typically develop stridor in the first 2 weeks after birth, with symptoms becoming most pronounced in the first 2 to 4 months. Stridor is frequently worse with feeding, supine positioning, and agitation.
- Many studies reveal an association between acid reflux and laryngomalacia. (1) Most children with laryngomalacia can be managed with simple lifestyle changes or antireflux medication. The indications for surgical intervention include severe respiratory distress, failure to thrive, cyanotic episodes, acute life-threatening events, or inability to tolerate an oral diet.
- A clinical diagnosis of supraglottitis may be presumed in children with classic symptoms of supraglottic airway obstruction, including stridor, tripod positioning, muffled voice, and absence of cough.
- Many children younger than 5 years with supraglottitis may require intubation whether or not they exhibit respiratory distress. A systematic review of 749 children with epiglottitis found a significant reduction in mortality in children who are intubated rather than observed. (2)

- Croup is the most common cause of stridor in the febrile child. It is approximately 1,000 times more common than epiglottitis and is most common in children ages 6 to 36 months.
- Recurrent croup or croup that does not respond appropriately to therapy may indicate the presence of fixed subglottic disease that requires evaluation by an otolaryngologist.
- Children with bacterial tracheitis may present with recent onset of cough and fever followed by rapidly progressive stridor and respiratory distress.
- Retrospective reviews suggest that most children with bacterial tracheitis require intubation for airway protection and to lavage obstructive pseudomembranes and secretions. (6)(7)
- Children with tracheomalacia may present with stridor, wheezing, recurrent barking cough, or frequent respiratory infections. Children with reactive airway disease that is refractory to standard medical therapy should be referred to a pediatric pulmonologist for evaluation of tracheomalacia.

To view Reference list for this article, visit <https://pedsinreview.aappublications.org> and click on the “Upper Airway Obstruction” link.

To view PowerPoint slides that accompany this article, visit <http://pedsinreview.aappublications.org> and click on the Data Supplement for this article.

**Upper Airway Obstruction**

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## PIR Quiz

1. A 3-month-old infant presents with worsening stridor in the past month. The mother notes that it seems better when the child is prone but worsens with feeding, supine positioning, and agitation. Which of the following correctly describes the pathologic findings in this most common cause of congenital stridor in infants?
  - A. Absence of aryepiglottic folds.
  - B. Air trapping on the expiratory phase of respiration due to collapse of the extra-thoracic airway.
  - C. Intrinsic abnormality of the tracheal cartilage causing collapse of the trachea.
  - D. Paralysis of the vocal cords.
  - E. Prolapse of the supraglottic structures into the laryngeal airway on inspiration.
2. A 3-year-old child presents to the emergency department with rapid onset of respiratory distress, high temperature, muffled voice, and drooling. She prefers to sit leaning forward on outstretched hands. Which of the following is true regarding the management of this patient?
  - A. Antibiotics targeting *Haemophilus influenzae* type B are no longer warranted because of widespread vaccine use.
  - B. Children younger than 5 years with this disease should be intubated whether or not they demonstrate respiratory distress.
  - C. Diagnosis should be confirmed with flexible fiberoptic laryngoscopy in the emergency department.
  - D. Retrospective studies have found conclusive benefit from the use of systemic corticosteroids
  - E. The steeple sign is a classic finding on lateral neck radiographs in patients with this disease.
3. A 2-year-old child presents to your office after 2 days of rhinorrhea and mild fever. This morning the mother noted that the child sounds hoarse and the cough has become barky. There is no tachypnea, and the child's oxygen saturation is 99%. This is a typical presentation for which infectious cause of upper airway obstruction in children?
  - A. Bacterial tracheitis.
  - B. Epiglottitis.
  - C. Laryngotracheitis.
  - D. Retropharyngeal abscess.
  - E. Spasmodic croup.
4. A 4-year-old child has had symptoms of an upper respiratory tract infection for 7 days. On day 8, the child develops a high temperature, worsening cough, and biphasic stridor. A trial of nebulized racemic epinephrine and corticosteroids provides no relief. What is the most likely bacterial cause of this child's illness?
  - A. *Haemophilus influenzae* type B.
  - B. *Staphylococcus aureus*.
  - C. *Streptococcus pneumoniae*.
  - D. *Streptococcus pyogenes*.
  - E. *Moraxella catarrhalis*.
5. A 4-month-old child presents to the emergency department with mild stridor. She developed rhinorrhea and cough during the past 24 hours but remained afebrile. The mother reports that the infant had a similar pattern of noisy breathing with a prior upper respiratory tract infection. There is no change in the stridor after administration of nebulized racemic epinephrine. You suspect tracheomalacia. What imaging study is most commonly used to screen for extrinsic compression of the trachea?
  - A. Magnetic resonance angiography.
  - B. Barium esophagography.
  - C. Computed tomography of the neck.
  - D. Lateral neck radiography.
  - E. Magnetic resonance imaging of the neck.

**REQUIREMENTS:** Learners can take *Pediatrics in Review* quizzes and claim credit online only at: <http://pedsinreview.org>.

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**Upper Airway Obstruction**  
Jordan Virbalas and Lee Smith  
*Pediatrics in Review* 2015;36;62  
DOI: 10.1542/pir.36-2-62

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The most likely diagnosis is:

- A. Legg-Calvé-Perthes disease.
- B. Lyme disease.
- C. Osgood-Schlatter disease.
- D. Osteochondroma.
- E. Sever disease.

### Corrections

In the February 2015 article “Upper Airway Obstruction” (Virbalas J, Smith L. *Pediatrics in Review*. 2015;36(2):62–73, doi: 10.1542/pir.36-2-62), the caption on Figure 3 should read: “The arrow is pointing to swollen aryepiglottic folds; the swollen epiglottis is located anterosuperior to the aryepiglottic folds.” The online version of the article has been resupplied.

Also, the discussion of Bacterial Tracheitis contains an image citation error on page 70, column one, under “Diagnosis,” paragraph two, sentence two. The figure cited in that second sentence should be Figure 4. The corrected sentence should read: “Anteroposterior neck radiographs characteristically show the subglottic narrowing seen in laryngotracheitis (Figure 4),” and should link to Figure 4.

The journal regrets the errors.

## Parent Resources from the AAP at HealthyChildren.org

- Sprains and Strains: <http://www.healthychildren.org/English/health-issues/conditions/orthopedic/Pages/Sprains-Strains.aspx>



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