Croup (Laryngotracheobronchitis)

Viruses most commonly cause croup, The parainfluenza viruses (types 1, 2, and 3) account for ~75% of cases; other viruses associated with croup include influenza A and B, adenovirus, respiratory syncytial virus (RSV), and measles. Influenza A has been associated with severe laryngotracheobronchitis, the most common form of acute upper respiratory obstruction.

Most patients have an upper respiratory tract infection with some combination of rhinorrhea, pharyngitis, mild cough, and low-grade fever for 1-3 days before the signs and symptoms of upper airway obstruction become apparent. The child then develops the characteristic “barking” cough, hoarseness, and inspiratory stridor. The low-grade fever can persist, although temperatures can reach 39-40°C (102.2-104°F); some children are afebrile. Symptoms are characteristically worse at night and often
recur with decreasing intensity for several days and resolve completely within a week. Agitation and crying greatly aggravate the symptoms and signs. Most young patients with croup progress only as far as stridor and slight dyspnea before they start to recover.

Physical examination can reveal a hoarse voice, coryza, normal to moderately inflamed pharynx, and a slightly increased respiratory rate. Patients vary substantially in their degrees of respiratory distress. Rarely, the upper airway obstruction progresses and is accompanied by an increasing respiratory rate; nasal flaring; suprasternal, infrasternal, and intercostal retractions; and continuous stridor. Croup is a disease of the upper airway, and alveolar gas exchange is usually normal. Hypoxia and low oxygen saturation are seen only when complete airway obstruction is imminent. The child who is hypoxic, cyanotic, pale, or obtunded needs immediate airway management.
Croup is a clinical diagnosis and does not require a radiograph of the neck. Radiographs of the neck can show the typical subglottic narrowing, or steeple sign, of croup on the posteroanterior view. However, the steeple sign may be absent in patients with croup. The radiographs do not correlate well with disease severity. Radiographs should be considered only after airway stabilization in children who have an atypical presentation or clinical course. Radiographs may be helpful in distinguishing between severe laryngotracheobronchitis and epiglottitis, but airway management should always take priority.

**Acute Epiglottitis (Supraglottitis)**
In the past, Haemophilus influenzae type b was the most commonly identified etiology of acute epiglottitis. Since the widespread use of the HiB vaccine,
invasive disease due to H. influenzae type b in pediatric patients has been reduced by 80-90%. Therefore, other agents, such as Streptococcus pyogenes, Streptococcus pneumoniae, and Staphylococcus aureus, now represent a larger portion of pediatric cases of epiglottitis in vaccinated children.

This dramatic, potentially lethal condition is characterized by an acute rapidly progressive and potentially fulminating course of high fever, sore throat, dyspnea, and rapidly progressing respiratory obstruction. Often, the otherwise healthy child suddenly develops a sore throat and fever. Within a matter of hours, the patient appears toxic, swallowing is difficult, and breathing is labored. Drooling is usually present and the neck is hyperextended in an attempt to maintain the airway. A brief period of air hunger with restlessness may be followed by rapidly increasing cyanosis and coma. Stridor is a late finding and suggests near-complete airway
obstruction. Complete obstruction of the airway and death can ensue unless adequate treatment is provided. The barking cough typical of *croup* is absent.

The diagnosis requires visualization of a large, cherry red, swollen epiglottis by laryngoscopy. In a patient in whom the diagnosis is certain or probable based on clinical grounds, laryngoscopy should be performed expeditiously in a controlled environment such as an operating room or intensive care unit. Anxiety-provoking interventions such as phlebotomy, intravenous line placement, placing the child supine, or direct inspection of the oral cavity should be avoided until the airway is secure. If epiglottitis is thought to be possible but not certain in a patient with acute upper airway obstruction, the patient can undergo lateral radiographs of the upper airway first. Classic radiographs of a child who has epiglottitis show the thumb sign. If the concern for epiglottitis still exists after the
radiographs, direct visualization should be performed. A physician skilled in airway management and use of intubation equipment should accompany patients with suspected epiglottitis at all times. An older cooperative child might voluntarily open the mouth wide enough for a direct view of the inflamed epiglottis.

Establishing an airway by nasotracheal intubation or, less often, by tracheostomy is indicated in all patients with epiglottitis, regardless of the degree of apparent respiratory distress, because as many as 6% of children with epiglottitis without an artificial airway die, compared with <1% of those with an artificial airway. In general, children with acute epiglottitis are intubated for 2-3 days, because the response to antibiotics is usually rapid.

**Acute Infectious Laryngitis**

Laryngitis is a common illness. Viruses cause most cases; diphtheria is an exception but is extremely rare in
developed countries. The onset is usually characterized by an upper respiratory tract infection during which sore throat, cough, and hoarseness appear. The illness is generally mild; respiratory distress is unusual except in the young infant. Hoarseness and loss of voice may be out of proportion to systemic signs and symptoms. The physical examination is usually not remarkable except for evidence of pharyngeal inflammation.

**Spasmodic Croup**

- Spasmodic croup occurs most often in children 1-3 yr. of age and is clinically similar to acute laryngotracheobronchitis, except that the history of a viral prodromal and fever in the patient and family are often absent. The cause is usually allergic but viral cause may occur in some cases.

Occurring most commonly in the evening or nighttime, spasmodic croup begins
with a sudden onset that may be preceded by mild to moderate coryza and hoarseness. The child awakens with a characteristic barking, metallic cough, noisy inspiration, and respiratory distress and appears anxious and frightened. The patient is usually afebrile. Usually, the severity of the symptoms diminishes within several hr, and the following day, the patient often appears well except for slight hoarseness and cough. Similar, but usually less severe, attacks without extreme respiratory distress can occur for another night or 2. Such episodes often recur several times. Spasmodic croup might represent more of an allergic reaction to viral antigens than direct infection, although the pathogenesis is unknown.

Differential Diagnosis

These 4 syndromes must be differentiated from one another and from a variety of other entities that can present as upper airway obstruction. Bacterial tracheitis is the most important differential diagnostic
consideration and has a high risk of airway obstruction. Diphtheritic croup is extremely rare in North America. Early symptoms of diphtheria include malaise, sore throat, anorexia, and low-grade fever. Within 2-3 days, pharyngeal examination reveals the typical gray-white membrane, which can vary in size from covering a small patch on the tonsils to covering most of the soft palate. The membrane is adherent to the tissue, and forcible attempts to remove it cause bleeding. The course is usually insidious, but respiratory obstruction can occur suddenly. Measles croup almost always coincides with the full manifestations of systemic disease and the course may be fulminant).

Sudden onset of respiratory obstruction can be caused by aspiration of a foreign body. The child is usually 6 mo-3 yr of age. Choking and coughing occur suddenly, usually without prodromal signs of infection. A retropharyngeal or peritonsillar abscess
can mimic respiratory obstruction. CT scans of the upper airway are helpful in evaluating the possibility of a retropharyngeal abscess. A peritonsillar abscess is often a clinical diagnosis.

Upper airway obstruction is occasionally associated with angioedema of the subglottic areas as part of anaphylaxis and generalized allergic reactions, edema after endotracheal intubation for general anesthesia or respiratory failure, hypocalcemic tetany, infectious mononucleosis, trauma, and tumors or malformations of the larynx. A croupy cough may be an early sign of asthma. Epiglottitis, with the characteristic manifestations of drooling or dysphagia and stridor, can also result from the accidental ingestion of very hot liquid.

Complications

Complications occur in \(~15\%\) of patients with viral croup. The most common is extension of the infectious process to involve other regions of the respiratory tract, such as the middle ear, the terminal
bronchioles, or the pulmonary parenchyma. Bacterial tracheitis may be a complication of viral croup rather than a distinct disease. Bacterial tracheitis can produce a two-phased illness, with the second phase associated with high fever, toxicity, and airway obstruction. Alternatively, the onset of tracheitis occurs without a second phase and appears as continued but higher fever and worsening respiratory distress rather than the usual recovery after 2-3 days of viral croup. Mediastinal emphysema and pneumothorax are the most common complications of tracheotomy.

**Treatment**

The mainstay of treatment for children with croup is airway management and treatment of hypoxia. Treatment of the respiratory distress should take priority over any testing. Most children with either acute spasmodic croup or infectious croup can be managed safely at home. Nebulized racemic epinephrine is an accepted treatment for moderate or
severe croup. The mechanism of action is believed to be constriction of the precapillary arterioles through the β-adrenergic receptors, causing fluid resorption from the interstitial space and a decrease in the laryngeal mucosal edema. Traditionally, racemic epinephrine, a 1:1 mixture of the D- and L-isomers of epinephrine, has been administered. A dose of 0.25-0.5 mL of 2.25% racemic epinephrine in 3 mL of normal saline can be used as often as every 20 min. Racemic epinephrine was initially chosen over the more active and more readily available L-epinephrine to minimize anticipated cardiovascular side effects such as tachycardia and hypertension. There is evidence that L-epinephrine (5 mL of 1:1,000 solution) is equally effective as racemic epinephrine and does not carry the risk of additional adverse effects. This information is both practical and important, because racemic epinephrine is not available outside the USA.
The duration of activity of racemic epinephrine is <2 hr. Therefore, observation is mandated. The symptoms of croup might reappear, but racemic epinephrine does not cause rebound worsening of the obstruction.

The effectiveness of oral corticosteroids in viral croup is well established. Corticosteroids decrease the edema in the laryngeal mucosa through their anti-inflammatory action. Most studies that demonstrated the efficacy of oral dexamethasone used a single dose of 0.6 mg/kg; a dose as low as 0.15 mg/kg may be just as effective. Oral dosing of dexamethasone is as effective as intramuscular administration. A single dose of oral prednisolone is less effective.

Antibiotics are not indicated in croup.

Epiglottitis is a medical emergency and warrants immediate treatment with an artificial airway placed under controlled conditions, either in an operating room or intensive care unit. All
patients should receive oxygen en route unless the mask causes excessive agitation. Racemic epinephrine and corticosteroids are ineffective. Cultures of blood, epiglottic surface, and, in selected cases, cerebrospinal fluid should be collected after the airway is stabilized. *Ceftriaxone, cefotaxime, or meropenem* should be given parenterally, pending culture and susceptibility reports, because 10-40% of *H. influenzae* type b cases are resistant to ampicillin. After insertion of the artificial airway, the patient should improve immediately, and respiratory distress and cyanosis should disappear. Epiglottitis resolves after a few days of antibiotics, and the patient may be extubated; antibiotics should be continued for 7-10 days.

Endotracheal intubation or tracheotomy is required for most patients with bacterial tracheitis and all young patients with epiglottitis.