

Croup (Acute Laryngotracheobronchitis): An Update

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Croup (*Acute laryngotracheobronchitis*) is the most common cause of upper airway obstruction of under 5 children. The word “croup” is derived from the Anglo-Saxon word *kropan* which means “to cry aloud”.¹ It is caused by various viral agents and is characterized by varying degrees of inspiratory stridor, barking cough, and hoarseness as a result of laryngeal and/or tracheal obstruction.² Although most children are deemed to have a mild and short-lived illness, many a times the presentations are frightening and worsen during the early hours of morning. Historically, before the advent of corticosteroids and racemic epinephrine for treatment of severe croup, intubation, tracheotomy, and death were the typical outcomes.^{2,3}

Epidemiology

It is primarily a disease of infants and toddlers, with a peak incidence from age 6 months to 36 months. Incidence peaks in the second year of life, at 5-6 cases per 100 children.³ The disease is most common in late fall and early winter but may be seen at any time of year. The incidence in boys is about 1.4 times that in girls.²

Aetiology

Parainfluenza viruses (types 1, 2, 3) are responsible for as many as 80% of croups and type 1 accounts for about 66% of cases and majority of hospitalizations.³ Less commonly, Adenovirus, RSV, Enterovirus, Coronavirus, Rhinovirus, Echovirus, Reovirus, Metapneumovirus, Influenza A and B are involved. Rarely, Measles, herpes simplex, varicella viruses and *Mycoplasma pneumoniae* has been found to be involved with croup.²⁻⁵

Pathogenesis

After entering through nose and nasopharynx, viruses eventually settle and cause inflammation in subglottic

larynx (*the narrowest part of the airway in children*) and trachea. Inflammation ultimately gives rise to edematous swelling of airway wall, narrowing of airway lumen and airflow limitation as well as decreased mobility of the vocal cords.⁶ This results in seal-like barking cough, turbulent airflow & stridor, chest retractions and hoarseness. In severe cases, fibrinous exudates and pseudomembrane may develop, causing even greater airway obstruction and all these events culminate into poor air entry, impaired alveolar ventilation, ventilation-perfusion mismatch and hypoxaemia.⁷

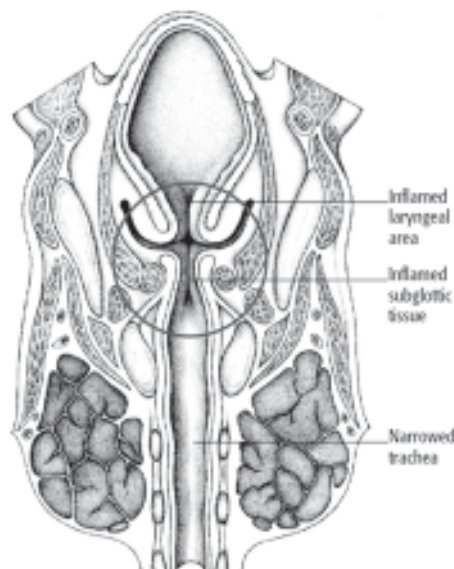


Fig-1: Viral croup causing inflammation and edema of the upper respiratory tract

Clinical features

The onset of the disease is sudden and the affected children usually presents with low grade fever, characteristic barking cough, inspiratory stridor, hoarseness of voice, respiratory distress, suprasternal recession and may be cyanosis.^{2,6-9} Sometimes, similar manifestations may be present in *acute epiglottitis*, *bacterial tracheitis*, *foreign body aspiration*, *retropharyngeal abscess*, *laryngeal diphtheria* etc. and most of these conditions can be diagnosed through their unique presentation as follows.¹⁰⁻¹³

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Acute epiglottitis is caused by *H. influenzae* B and the disease is characterized by sudden onset of high fever, drooling, dysphagia, anxiety and a preference to sit upright and in the so-called sniffing position (i.e. sitting forward with their head extended) to open the airway is very characteristic.

Bacterial tracheitis presents with worsening respiratory distress, a “croupy” cough, and high fever. They have a toxic appearance and do not respond favorably to treatment with nebulized epinephrine.² The most frequently isolated pathogen is *Staphylococcus aureus*.⁶

Peritonsillar or retropharyngeal abscess presents with low-grade fever, dysphagia, drooling, stridor, dyspnoea, tachypnoea, a muffled “hot potato” voice, neck stiffness, unilateral neck pain and unilateral cervical lymphadenopathy. It is associated with the presence of trismus, which results from irritation of the internal pterygoid muscle.^{2, 3}

Laryngeal diphtheria is characterized by low-grade fever, hoarseness and barking cough along with dysphagia and inspiratory stridor, and the characteristic pseudomembrane is seen on throat examination.^{2, 3, 6}

Severity of croup

Croup may be of any severity ranging from mild to a state of respiratory failure as follows¹⁴:

- Mild - Occasional barking cough, no audible stridor at rest, and either no or mild suprasternal and/or intercostal retractions
- Moderate - Frequent barking cough, easily audible stridor at rest, and suprasternal and sternal wall retractions at rest, with no or minimal agitation
- Severe- Frequent barking cough, prominent inspiratory (and occasionally expiratory) stridor, marked sternal wall retractions, significant agitation and distress
- Impending respiratory failure - Barking cough (often not prominent), audible stridor at rest, sternal wall retractions may not be marked, lethargy or decreased consciousness, and often dusky appearance without supplemental oxygen support.¹⁴

Scoring system

To assess the degree of respiratory compromise, croup scores have been developed. The most commonly cited is the Westley scoring system¹⁵. The score evaluates

the severity of croup by assessing the following 5 parameters, with a score range of 0 to 17:

Parameters	0 point	1 point	2 points	3 points	4 points	5 points
Inspiratory	None	Upon agitation	At rest			stridor
Retractions	None	Mild	Moderate	Severe		
Air entry	Normal	Mild decrease	Moderate decrease			
Cyanosis	None				Upon agitation	At rest
Level of consciousness	Normal, including sleep	Normal, including sleep				

A score of < 3 represents mild disease; 3-6 represents moderate disease; and a score > 6 represents severe disease.¹⁵

Diagnosis

Croup is basically a clinical diagnosis. However, X ray neck AP view shows steeple sign (*narrowing of air column*) which is characteristic of croup.



Fig.-2: X ray neck AP view showing steeple sign

Complete blood count may be done but there is no characteristic finding.

Treatment

Treatment of croup is mainly supportive with a view to minimize respiratory distress, ensure proper oxygenation, and ventilation.^{6, 8}

- The child should be kept as comfortable as possible, allowing him to remain on the arms of parent. Unnecessary painful interventions should be avoided as these may cause agitation, worsen airway obstruction and increased oxygen requirements by the child.

- Along with that child may need IV fluid if he/she is unable to take oral feeds.
- If patient is kept on bed he/she should be placed in a neck extended position to keep the airway open. Clearing of air passage through oropharyngeal and nasopharyngeal suction may be needed.
- Monitoring of vital parameters, e.g. pulse, blood pressure, oxygen saturation and other parameters should be done routinely.⁸

Oxygen inhalation

Oxygen inhalation by nasal cannula (2L/min) or by face mask (3-5 L/min) if SPO₂ in room air is <92%.⁹ Oxygen can also be given via a plastic hose with the opening held within a few cm of the nose and mouth (blow-by oxygen) which will render minimum irritation to the patient.^{2,9}

Corticosteroid

The effectiveness of oral corticosteroids in croup is well established. They reduce subglottic oedema through their anti-inflammatory action and significant relief is obtained by 6 hours of administration.¹⁶ Steroids either IV or oral form is adequate to control mild and most cases of moderate croup. In mild croup, steroids reduce the rate of hospitalization, hospital stay and most importantly reduce the need for subsequent intervention such as epinephrine administration¹⁶. In severe croup, steroids significantly reduce the rate of intubation and also the duration of intubation.

Corticosteroids recommended are single dose of either i) dexamethasone: 0.6 mg/kg (same efficacy if administered intravenously, intramuscularly, or orally)^{8,10,16-24} or ii) Nebulized Budesonide: 2 mg in 4 ml of water (expensive in comparison to dexamethasone or prednisolone)^{25,26}. A single dose of oral prednisolone (1 mg/kg) may be given but it is less effective and is associated with more return to hospitals. Prednisolone is less potent to reduce inflammation and shortened half-life (18-36 hrs) than that of dexamethasone (36-54 hrs).^{27,28} There are no controlled studies examining the effectiveness of multiple doses of corticosteroids¹⁶.

Adrenaline/epinephrine

Nebulized racemic epinephrine is an accepted treatment for moderate-to-severe croup and this option of treatment substantially reduces the number of intubation or tracheotomy.^{2,6,8-10} Its effect is immediate and lasts for 90-120 minutes. Epinephrine causes constriction of the precapillary arterioles, thereby decreasing capillary hydrostatic pressure through beta adrenergic receptors. This in turn leads to fluid resorption from the interstitium

and improvement in the laryngeal mucosal edema. It also causes bronchial smooth muscle relaxation and bronchodilation.

Racemic epinephrine at a dose of 0.25 to 0.5 ml to be diluted in 2.5-3 ml of normal saline can be used as often as every 20 minutes¹⁶. Alternatively, L-epinephrine (1:1000 dilution) 5 ml is equally effective and does not carry the risk of cardiovascular side-effects^{8,13-16,29}. The duration of activity of racemic epinephrine is about 2 hours¹⁶. Therefore, patients who received epinephrine should be observed for at least 3 hours because of concerns for a rebound phenomenon of bronchospasm, worsening respiratory distress, and/or persistent tachycardia. Patients can be discharged home only if they demonstrate healthy color, good air entry, baseline consciousness, and no stridor at rest and have received a dose of corticosteroids.²⁹

Analgesics and antipyretics

The use of analgesics or antipyretics is reasonable for the benefit of reduction of fever or discomfort in children with croup.³⁰⁻³⁵

Antibiotics

Since laryngotracheitis and spasmodic croup are viral illnesses, there is no reason to treat them with antibiotics unless clinical manifestations or laboratory values suggest secondary bacterial infection. Moreover superinfections, such as bacterial tracheitis and pneumonia, are described. However their rare frequency (<1 per 1000 cases of croup) makes use of prophylactic antibiotics unreasonable.³⁰⁻³⁶

Humidified air

Throughout the 19th and most of the 20th century, cool mist administration was the mainstay of treatment of croup. Hospitals had "croup rooms" filled with cool mist. Theoretically, mist moistens airway secretions, decreases their viscosity, and soothes the inflamed mucosa.³⁷

Despite the observation of beneficial effect of cool mist, Cochrane review has found no evidence supporting its use in croup¹⁶. Moreover, mist tents can disperse fungus and molds if not properly cleaned and, more importantly, separates the child from the parent, causing anxiety and agitation, worsening the patient's symptoms.^{37,38} Hot humidified air can cause scald injuries.³⁹

Heliox

Helium is an inert low-density gas with no inherent pharmacological or biological effects. Administration of helium-oxygen mixture (heliox) to children with severe respiratory distress can reduce their degree of

distress since the lower density helium gas decreases airflow turbulence through a narrow airway⁴⁰⁻⁴². Helium decreases the force necessary and facilitates the movement of oxygen through the airways and decreases the mechanical work of respiratory muscles. This clinical response reduces respiratory distress^{41, 43, 44}.

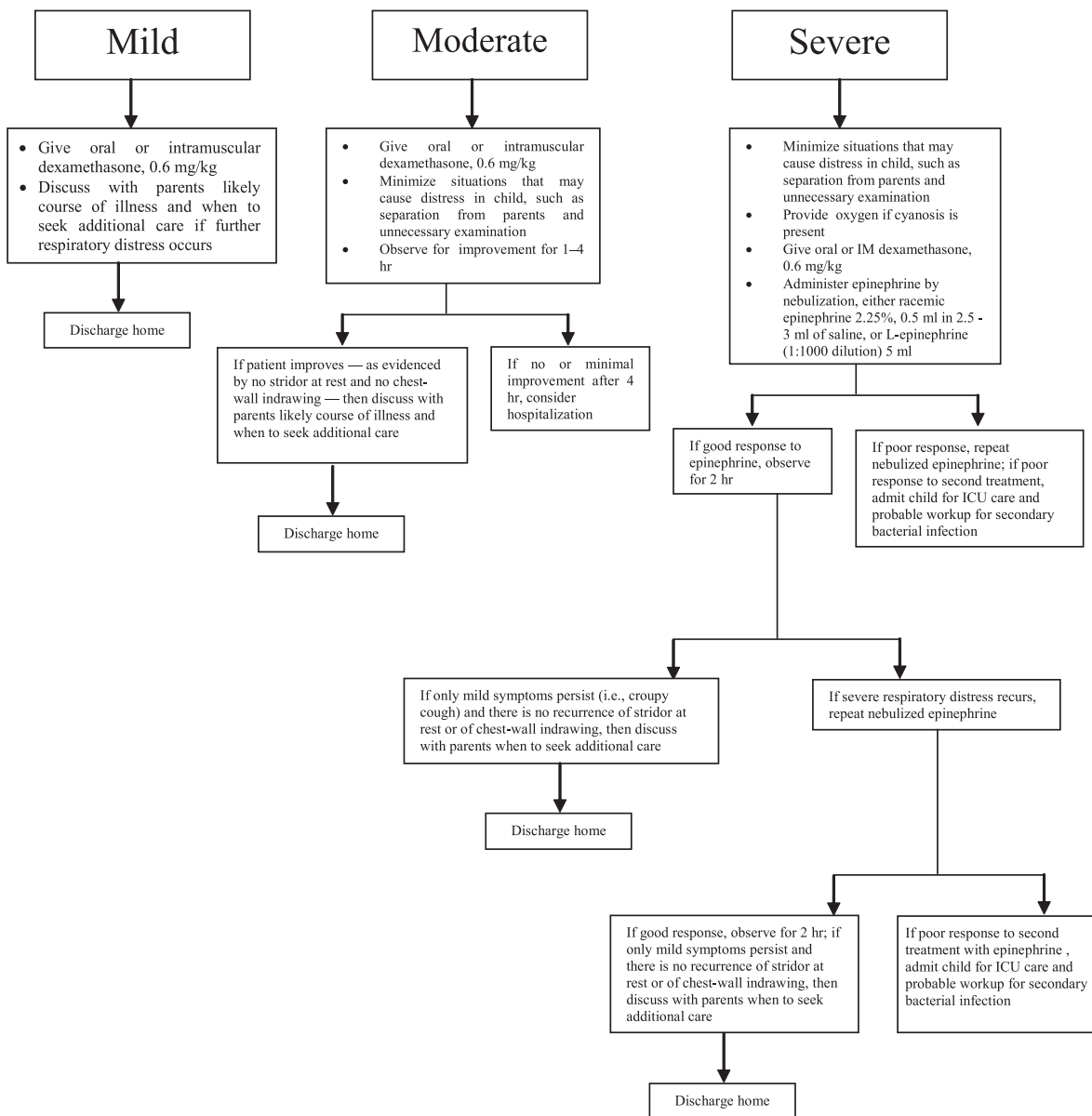
Both heliox and racemic epinephrine were associated with similar improvements in croup score over time. However, since heliox has yet to be shown to offer greater improvements than standard treatments and can

be difficult to use in unskilled hands, there is insufficient reason to recommend its general use in children with severe croup.⁴⁰⁻⁴⁷

Cough and cold medications: Use of antitussive and decongestants are ineffective and not indicated.^{2, 3, 30}

Bronchodilators: In view of the pathophysiology of croup as an upper-airway disease, there is no reason to use short-acting β_2 agonists for treatment of the disease.^{2, 9}

Stepwise management of croup according to severity³⁰



Prognosis

The prognosis for croup is excellent, and recovery is usually complete. The majority of patients are managed successfully as outpatients, without the need for inpatient hospital care.²

Conclusion:

Although croup is an important cause of severe respiratory distress from upper airway obstruction of young children, it is often overlooked. If there is suspicion in clinician's mind, diagnosis is easy through its unique clinical presentations. However, outcome of the disease is rewarding through prompt intervention with corticosteroid, epinephrine and other supports.

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