JOHNS HOPKINS ALL CHILDREN'S HOSPITAL

Croup Clinical Pathway



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This pathway is intended as a guide for physicians, physician assistants, nurse practitioners and other healthcare providers. It should be adapted to the care of specific patient based on the patient's individualized circumstances and the practitioner's professional judgment.

Johns Hopkins All Children's Hospital **Croup Clinical Pathway**

Rationale

This pathway was developed by a consensus group of JHACH emergency medicine providers, pulmonologists, hospitalists, and critical care providers to standardize the management of children presenting with croup. It addresses the following clinical questions or problems:

- 1. When to treat patients for croup
- 2. Proper medications and dosing for croup treatment
- 3. When to consider admission for further observation and treatment
- 4. When to consult ENT
- 5. When to consider alternative diagnoses

Background

Croup is a common manifestation of upper airway obstruction resulting from swelling of the larynx, trachea, and bronchi. Croup is most commonly caused by acute viral infection. Croup is the most common cause of upper airway obstruction in children 3 months to 6 years of age and affects 3% of children in this age range in the United States. Parainfluenza virus accounts for up to 75% of cases. This prevalence is thought to be secondary to the ability of this virus to activate chlorine secretion and inhibit sodium absorption resulting in an edematous reaction. Croup has also been identified in children with respiratory syncytial virus, influenza, rhinovirus, enterovirus, coronavirus, bocavirus, and metapneumovirus. Croup can also be seen as a result of measles or diphtheria infection in unvaccinated children.

The symptoms of croup typically appear suddenly at night although they can be preceded by a prodromal phase with rhinorrhea, pharyngitis, and/or fevers. The most common symptom is a dry barking cough. Children usually have a clear voice but can present with hoarseness. The most common location of croup related upper airway obstruction is the subglottic region at the level of the cricoid cartilage as this is the most narrow part of the airway in young children. This leads to increased resistance of airflow causing stridor, a harsh, high-tone sound heard mainly on inspiration. Occasionally, children with croup will also have expiratory wheezing from swelling in structures below the glottis including the trachea and bronchi. This is seen in children with laryngotracheitis or laryngotracheobronchitis. Depending on the amount of upper airway obstruction present, patients can present with increased respiratory rate and effort as well as hypoxia and respiratory distress. The most common auscultatory finding in patients with croup is inspiratory stridor heard over the neck. Mild illness typically lasts two to seven days, but symptoms can last up to two weeks.

There is not a universally accepted classification method for severity of laryngeal obstruction, but the Westley croup score is a validated option seen below in Table 1. In mild obstruction

(Westley croup score 0-2), the patient is well appearing, active, and is able to take in oral fluids. The symptoms include a mild barking cough with or without hoarseness. There is no stridor at rest but patients may have stridor with crying. Respiratory retractions are either non-existent or minor. With moderate obstruction (Westley croup score 3-5 points), patients present with an intense barking cough, inspiratory stridor at rest, and respiratory retractions that worsen when the patient is agitated. In severe obstruction (Westley croup score 6-11), the cough is intensive with loud and occasionally biphasic stridor. There are respiratory retractions with increased expiratory phase. In patients with life threatening laryngeal obstruction (Westley croup score 12 or higher), there is tachycardia with cough of variable intensity. Cough may decrease due to respiratory muscle fatigue. Sternal retractions may be observed. The patient will present with fatigue and exhaustion, cyanosis or pallor, and depression level of consciousness. In most children, croup is mild and symptoms may subside without intervention.

Level of Consciousness	Score
Normal	0
Disoriented	5
Cyanosis	Score
None	0
With agitation	4
At rest	5
Stridor	Score
None	0
When agitated	1
At rest	2
Air Entry	Score
Normal	0
Decreased	1
Markedly decreased	2
Retractions	Score
None	0
Mild	1
Moderate	2
Severe	3
Total Score	Croup Severity
= 2</td <td>Mild</td>	Mild
3 to 7	Moderate
8 to 11	Severe
>/= 12	Impending respiratory failure

Table 1: Westley Croup Score

Diagnostic Testing

The diagnosis of croup should be made clinically. Radiography is not routinely indicated. Laboratory studies are indicated only in cases where another diagnosis is suspected. If there is concern for epiglottitis, a lateral neck radiography can be performed. Typical findings in a patient with croup include marked narrowing in the subglottic region (steeple sign). Rapid viral PCR testing should not be obtained routinely in children presenting with croup.

Johns Hopkins All Children's Hospital Emergency Center Croup Clinical Pathway



Treatment

Patients presenting with signs and symptoms of croup should be treated with a one-time dose of dexamethasone 0.6 mg/kg PO (preferred), IM, or IV (*Level of Evidence A*). Treatment with dexamethasone has been shown to expedite resolution of symptoms and reduce risk of repeat visits to the ED within 7-10 days of the initial visit when compared to prednisolone (*Level of Evidence B*). Oral and intramuscular dexamethasone have comparable efficacy in improvement of croup symptoms. If the patient is able to tolerate oral intake, the dose should be given orally. Repeat doses have not been found to be beneficial in improving the duration and severity of croup symptoms.

Children presenting with hypoxemia should be given oxygen. Patients presenting with stridor at rest and any other clinical factor including retractions, tachypnea, restlessness, or agitation should be treated with racemic epinephrine (*Level of Evidence A*). Epinephrine is thought to be helpful in these patients as it causes vasoconstriction in the upper airway mucosa, helping to decrease edema. Epinephrine has a quick onset of action with a short half-life. It is therefore recommended to combine racemic epinephrine with dexamethasone in treating croup patients, as dexamethasone has a longer onset of action as well as a longer half-life. The recommended dose for racemic epinephrine is 0.5 mL/kg of 1:1000 epinephrine in 2.5 mL of normal saline via nebulizer with a maximum dose of 5 mL (*Level of Evidence B*). This should be repeated every two hours as needed. Racemic epinephrine should be used cautiously in patients with congenital heart disease. The effects of epinephrine wane after 2 hours, so patients should be monitored for at least two hours after administration before they are discharged.

For patients with impending respiratory failure, endotracheal intubation may be required until laryngeal edema resolves. Laryngoscopy is indicated in patients where foreign body aspiration is suspected as a cause of airway obstruction or when the possibility of superimposed bacterial tracheitis is a concern. Consider ENT consult for patients with multiple episodes of croup with age less than 36 months, history of intubation, or prolonged/severe disease.

Heliox should be considered in patients who continue to have increased respiratory effort despite being treatment with racemic epinephrine or in patients who are not candidates for treatment with racemic epinephrine. Heliox is a combination of helium and oxygen. This mixture has a much lower density than atmospheric air which allows for decreased turbulence when entering the airway. This allows for increased flow rates and lower airway resistance. When given at a ratio of 70/30, heliox can provide a short term benefit including decreased work of breathing and improvement in gas exchange in patients with moderate to severe croup. However, if the patient requires greater than 30% humidified oxygen, heliox is not indicated as it will not theoretically provide any additional benefit. (*Level Evidence D*). Patients receiving heliox should be admitted to the PICU for observation and treatment.

Admission

Criteria for admission includes:

- 1. Severe presentation with altered level of consciousness and impending complete upper airway obstruction requiring multiple doses of racemic epinephrine
- 2. Hypoxemia or increased work of breathing requiring oxygen administration
- 3. Persistent or deteriorating work of breathing during the observation period in the EC despite administration of dexamethasone and racemic epinephrine.
- 4. Inability of the patient to tolerate oral intake or concern for dehydration

Johns Hopkins All Children's Hospital Inpatient Croup Clinical Pathway



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Outcome measures

- Length of stay in the EC
- Rate of x-ray use
- Rate of viral testing
- Time to racemic epinephrine administration
- Time to dexamethasone administration
- Rate of EC return visits in less than 48 hours

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Disclaimer

Clinical Pathways are intended to assist physicians, physician assistants, nurse practitioners and other health care providers in clinical decision-making by describing a range of generally acceptable approaches for the diagnosis, management, or prevention of specific diseases or conditions. The ultimate judgment regarding care of a particular patient must be made by the physician in light of the individual circumstances presented by the patient.

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