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Acute Management of Croup

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Intro

Hi everyone, I’m Dr. Alia Sunderji, a third-year pediatrics resident at the Hospital for Sick Children in Toronto. This podcast is produced by PedsCases and the Canadian Pediatric Society to summarize the recently published 2016 Canadian Pediatric Society (CPS) statement on the management of croup in young childrenⁱ. This podcast was developed with the lead author of the statement, Dr. Oliva Ortiz Alvarez, MD, MSc FRCPC, she is an Assistant Professor at Dalhousie University and a paediatric consultant at Saint Martha’s Regional Hospital. The complete CPS statement can be found at www.cps.ca and a written transcript of this podcast can be found at www.pedscases.com.

Let’s start with a commonly encountered case. You are working in the Emergency Department when your preceptor asks you to see Omar, an 18 month old boy who presents with 2 days of fever, runny nose and cough. Before you enter the room you take a look at Omar’s vitals his heart rate is 100 bpm, BP: 80/50, RR: 60 breaths per minute, T: 38.7 C tympanic his oxygen saturation is 98% on room air. You take a brief history from Omar’s mother who describes his cough as barking and “seal-like”, which worsened over the evening. His immunizations are up to date. On examination, he is febrile, alert, and non-toxic appearing. He is making this sound (*insert inspiratory stridor*)ⁱⁱ when breathing in. He has subcostal retractions but is alert and playful with his parents. You hear an intermittent cough. There are no signs of cyanosis or fatigue.

As you prepare to review with your preceptor, you wonder how to describe Omar’s cough and the sound he makes when breathing in. At the top of your differential, you wonder whether his cough sounds like the “barking cough” commonly associated with croup.

Objectives

The objectives of this podcast are to review the new CPS guidelines on the management of croup. In particular,

1. To discuss the clinical manifestations of croup and its underlying pathophysiology;
2. To review the differential diagnosis for stridor;
3. To outline the treatment of croup; and
4. To review considerations for determining the disposition of children with croup.

Let's start by discussing the definition, pathophysiology and clinical manifestations of croup.

The word croup, is derived from the word “kropan” meaning to “cry out loud”ⁱⁱⁱ. Croup is often seen in infants aged 6 months-3 years with a mean age of diagnosis of 18 months. Children with croup can present with a barking cough, which is typically worse at night. This barking cough can be accompanied by inspiratory stridor, hoarseness and respiratory distress.

Causative Organisms and Pathophysiology

The sound you heard Omar make upon inspiration, (*insert inspiratory stridor sound*) is described as *stridor*. Stridor is the sound produced by increased air turbulence, which can arise from a narrowing in the airway. In the case of croup, stridor arises from a narrowing of the subglottic tissues and trachea due to increased inflammation and edema caused by a virus. In infants, the narrowest part of the trachea lies below the larynx, and inflammation of this area can restrict airflow considerably! This inflammation and edema is often caused by a viral infection. In 65 percent of cases, croup is caused by parainfluenza virus 1, 2 and 3^{iv}. Other viruses implicated in croup include influenza A, influenza B, adenovirus, respiratory syncytial virus (RSV) and metapneumovirus.

Since croup arises from a viral infection, symptoms are generally self-limiting, lasting from 3 to 7 days. In the majority of patients, the barking cough generally disappears after 48 hours. Less than 5% of children have symptoms that last longer than 5 days. A child who presents with worsening symptoms without improvement should be evaluated for a congenital narrowing of their airway.

In summary, a diagnosis of croup should be suspected in children between 6 months-3 years of age who present with a rapid onset of a barking cough, which is typically worse at night accompanied by inspiratory stridor, hoarseness and respiratory distress. Parainfluenza types 1,2 account for the majority of cases of croup. As a result, symptoms are generally short lived, with the barking cough resolving in 48 hours.

Differential Diagnosis of Stridor

A diagnosis of croup can be established clinically, with a detailed history and thorough physical exam. Other causes of stridor should be excluded. The differential for stridor in infants includes severe entities such as epiglottitis, anaphylaxis, foreign body aspiration, retropharyngeal abscess and bacterial tracheitis. When obtaining a history, it is

important to rule out these diagnoses by inquiring about the patient's immunization status, history of choking and possibility of aspiration. Features associated with anaphylaxis should be reviewed including exposure to a potential allergen, urticaria, angioedema, and a history of allergic reactions. Finally, a history suggestive of retropharyngeal abscess including a high fever, neck pain, sore throat, dysphagia and torticollis should be elicited. If a child appears very unwell with a fever and stridor and doesn't respond to the typical treatment for croup, always remember to consider bacterial tracheitis.

The mainstay of the physical exam in a child with suspected croup is to avoid agitating them. Allow the child to sit on their parents' lap as you enter the room. Begin by reviewing the vitals. Children with croup can present with tachypnea, which the World Health Organization (WHO) defines as a respiratory rate of greater than 50 in infants aged 2-12 months, and greater than 40 in children aged 1-5 years. Start by simply examining the child at rest. If a child appears cyanotic, lethargic or anxious and agitated – this may be a sign of impending respiratory failure or a more severe underlying disease. If this is the case, get help immediately.

Beware of the drooling child who prefers to keep their chin pushed forward and refuses to lie down, in what we describe as the “sniffing position”. A diagnosis of epiglottitis must be considered in children who appear toxic and present with drooling, dysphagia. Alternatively, children who present with torticollis, drooling and stridor, may have features more consistent with a retropharyngeal abscess.

Children with croup can be divided into mild, moderate and severe presentations depending on the extent to which they exhibit the characteristic barking cough, inspiratory stridor, subcostal indrawing and perioral cyanosis.

In mild presentations, children can present with an occasional cough, minimal stridor, minimal indrawing with no lethargy or respiratory distress.

In moderate croup, children have a frequent barking cough, stridor at rest, indrawing of intercostal/subcostal muscles. In severe cases, the child exhibits distress, agitation, lethargy or cyanosis along with severe indrawing and prominent stridor.

Investigations

Let's return to our case. You review Omar's case with your preceptor, who asks you if there are any additional tests that can help confirm your suspected diagnosis of croup.

The diagnosis of croup can be established clinically. Radiographs are not routinely recommended to assist with the diagnosis. The characteristic “steeple” sign demonstrating a narrowing of the air column in the area below the glottis is only present in 50% of cases with croup. The “steeple sign” can also be present in children without croup depending on their phase of respiration at the time of radiograph. Since croup results from narrowing of the subglottic tissues in the upper airway, alveolar gas

exchange is generally not compromised. A venous blood gas does not usually assist in making the diagnosis of croup.

Management

Now, let's discuss the management of children with croup.

Corticosteroids are the mainstay of treatment in children with croup and should be offered to all children with croup regardless of the severity of presentation. Children show improvement within 2-3 hours after a single dose of oral dexamethasone, the effects of which can last up to 48 hours after the initial dose. Children who receive corticosteroids have a reduced length of stay in the ED (12 hours!) and have fewer return visits to hospital. A Cochrane systematic review demonstrated that dexamethasone was the most frequently tested steroid in children with croup at a dose of 0.6 mg/kg/dose. It remains unclear whether smaller doses 0.15-0.3 mg/kg are equally effective.

In children with moderate to severe presentations, nebulized epinephrine has been shown to reduce the need for intubation and more invasive methods of ventilation such as tracheotomy. Nebulized epinephrine should take effect quickly - within 10-30 minutes of after its initiation. Its duration is short, lasting up to 2 hours maximum. If a child's respiratory distress does not improve within this time, an alternative diagnosis should be considered.

Although the initial trials to probe the usefulness of epinephrine in treating croup were done using racemic epinephrine, which is a mixture of epinephrine L and D isomers, racemic epinephrine has not been available in Canada for more than 5 years. Nebulized epinephrine has proven to be equally effective while treating croup. Epinephrine is believed to decrease laryngeal mucosal edema by decreasing capillary fluid leakage via alpha adrenergic vasoconstriction. The most common side effects of nebulized epinephrine are hypertension and tachycardia.

Given the short duration of epinephrine, children treated with a combination of dexamethasone and epinephrine should be observed for at least 2-4 hours after the last nebulized epi dose. If symptoms do not recur within this time period, they can be discharged safely from the ED.

Children with mild croup, characterized by an occasional barking cough, minimal stridor without any lethargy, respiratory distress can be given 1 dose of oral dexamethasone and discharged safely from the ED.

Children with stridor audible at rest, a frequent barking cough, visible subcostal and intercostal indrawing are characterized as having moderate croup. After receiving oral dexamethasone, these children should be observed for at least 2 hours. If the indrawing and stridor have not improved, they can be observed another 2 hours to allow the dexamethasone take effect. At four hours, if they do not show signs of improvement,

consider giving them nebulized L-epinephrine (1:1000). Given the short effect of epinephrine, be sure to observe them for an additional 2 hours in order to ensure that the symptoms do not worsen once the epinephrine wears off. If their symptoms worsen during this time a repeat dose of epinephrine can be administered.

Criteria for Admission

Children who continue to exhibit signs of moderate respiratory distress after receiving oral dexamethasone 4 hours prior should be considered for admission. Signs of moderate respiratory distress include: stridor at rest, chest wall indrawing, or tachypnea.

Children with severe work of breathing, prominent inspiratory and expiratory stridor and a frequent barking cough should be closely monitored for signs of lethargy, decreased level of consciousness and impending respiratory failure. They should be given dexamethasone and nebulized epinephrine. In severe cases, heliox, a helium-oxygen mixture can be considered to reduce respiratory distress in children with severe croup. The low density of helium is believed to facilitate its movement through narrow airways. Although heliox has been occasionally used to avoid intubation in children with croup, it has not been proven to improve croup symptoms and should not be used routinely.

Now that we have all of that information, let's return to our case:

Omar, an 18 month old boy who presents with 2 days of fever, runny nose and cough. He developed an intermittent barking cough. His immunizations are up to date. On examination, he is febrile, alert, and non-toxic appearing. He is tachypneic with inspiratory stridor. He has subcostal retractions and intercostal retractions but is alert with no signs of cyanosis or fatigue.

You believe that Omar's clinical presentation is in keeping with a moderately severe presentation of croup. You order 0.6mg/kg/dose of oral dexamethasone. His clinical exam is unchanged after 2 hours. You recommend one dose of nebulized epinephrine 1:1000. You observe Omar for an additional 2 hours for a rebound effect. When you reassess him, you find that he is no longer tachypneic and does not have any signs of subcostal or intercostal indrawing. He is not lethargic, nor does he have any signs of cyanosis. Apart from an intermittent barking cough, he no longer has any signs of stridor and exhibits equal air entry bilaterally. You determine that it is safe to discharge Omar and ensure that his parents are aware of signs and symptoms to warrant a return.

Omar's parents ask you whether the steam of a warm shower or taking him for a drive with window open to the cold air will help reduce his cough. Although very popular, there is no evidence for humidified air or mist tents in the treatment of croup.

They also ask you if Omar needs antibiotics. You reassure them that his symptoms are generally caused by viruses and that there is no indication for antibiotic therapy. There is a low incidence of bacterial infection with croup, around 1:1000 cases.

Your preceptor asks you how to determine when patients who have received both dexamethasone and epinephrine are safe to be discharged. If symptoms have not recurred after 2-4 hours after dexamethasone and epinephrine, and the patient does not exhibit any stridor, increased work of breathing or decreased LOC, they can be discharged home safely.

Summary

Let's summarize our discussion with 3 take home points in 3 different categories:

Category 1: Clinical Presentation

Here are three features of croup to remember:

1. Croup occurs in children 6 months to 3 years old with a mean age of diagnosis of 18 months.
2. Children with croup have a barky cough, which is often worse at night.
3. They can have inspiratory stridor, caused by subglottic airway narrowing.

Category 2: Treatment

Again here are three things to remember

1. Dexamethasone 0.6 mg/kg – you're going to give this to everyone who presents with croup, regardless of severity. The maximum dose of dexamethasone is 10mg per dose.
2. Consider nebulized epinephrine (1:1000) for moderate severity
3. Consider heliox in severe cases

Category 3: Disposition

1. Children with mild croup can be discharged after dexamethasone.
2. Children with moderate croup severity should be given dexamethasone and reassessed in 2 hours. If the child improves, they can be discharged home. If not, give them nebulized epinephrine. Nebulized epinephrine has a short half life, reassess these kids after another 2 hours – if they've improved, discharge, if not – admit them!
3. Children with severe croup should receive dexamethasone, nebulized epinephrine, and be considered for admission.

That concludes our podcast on croup, brought to you by PedsCases and the Canadian Pediatric Society. Thanks for listening!

ⁱ Ortiz-Alvarez, Oliva. Canadian Paediatric Society, Acute Care Committee. Acute management of croup in the emergency department. January 6, 2017.

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ⁱⁱⁱ Malhotra, A., & Krilov, L. R. (2001). Viral croup. *Pediatrics in Review*, 22(1), 5-12.

^{iv} Malhotra, A., & Krilov, L. R. (2001). Viral croup. *Pediatrics in Review*, 22(1), 5-12.

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