Croup

Synonyms: acute laryngotracheitis, acute laryngotracheobronchitis

Croup is a common childhood illness causing symptoms which may involve a harsh barking cough, hoarse voice and (inspiratory) stridor. It is usually caused by inflammation of the upper respiratory tract (predominantly the larynx and trachea but it may affect the bronchi) as a result of viral infection.

Some consider that the term croup should not be used to describe illness affecting the bronchi, as there is a higher likelihood of secondary bacterial infection in such cases.

Croup tends to be relatively mild and self-limiting but the distressing symptoms may prompt parents to bring their child to their GP or local emergency department. Severe cases may compromise the upper airway and so the condition of the child needs to be assessed carefully and other causes of upper airway obstruction (such as inhaled foreign body and epiglottitis) must be considered and excluded.

Pathophysiology

- Viral upper respiratory tract infection (URTI) causes nasopharyngeal inflammation that may spread to the larynx and trachea, causing subglottal inflammation, oedema and compromise of the airway at its narrowest portion.
- The movement of the vocal cords is impaired leading to the characteristic cough. Occasionally, fibrinous exudation with pseudomembrane formation may occur, causing further airway compromise.
- It is thought that some children who experience recurrent bouts of spasmodic croup have a primarily allergic rather than infective aetiology for subglottal oedema. [1]

Causative organisms

- Parainfluenza virus types I, II, III and IV (thought to be responsible for about 80% of cases - type I causing 50-70% of severe cases). [2, 3]
- Respiratory syncytial virus.
- Adenoviruses.
- Rhinoviruses.
- Enteroviruses.
- Measles.
- Metapneumovirus.
- Influenza A and B (type A is associated with severe disease).
- Mycoplasma pneumoniae (rare cause).
- Studies suggest a specific association between croup and the novel coronavirus NL63. [4]

Epidemiology

- Croup most often affects children aged 6 months to 3 years, with a peak incidence during the second year of life. However, children as young as 3 months of age, or adolescents and, very rarely, adults can be affected. [5]
- The annual incidence is variable depending on the cyclical prevalence of viral infection (particularly parainfluenza virus) in children.
- The incidence is about 60 per 1,000 child years in those aged between 1 and 2 years. [6]

Risk factors

- Male:female preponderance is about 1.43:1 in children younger than 6 months and 1.73:1 in children 6-12 months. [7]
- It is most prevalent in autumn and spring.
- Genetic studies suggest that the C/C variant of the CD14 C-159T gene had a significantly lower prevalence of croup. [8]

Presentation

- Croup normally starts with nonspecific symptoms of viral URTI, such as runny nose, sore throat, fever and cough.
- This progresses over the course of a couple of days to include the characteristic barking cough and hoarseness. These symptoms tend to be worse at night.
- There is a high degree of variability in clinical findings. There may be a mild-to-moderate fever. Check vital signs (including temperature, pulse and blood pressure).
- A barking cough and hoarse cry are nearly always present.
- Stridor (harsh, low-pitched noise heard during inspiration) may be heard at rest or only when the child is agitated or active.
- Chest sounds are usually normal but can be decreased in volume where there is severe airflow limitation.
- Respiratory distress with marked tachypnoea and intercostal recession may be noted. It should be recognised that a child whose stridor appears to be improving and in whom intercostal recession has disappeared may in fact be deteriorating with worsening airways obstruction. Such a child may be at high risk of complete airway occlusion. [8]
Drowsiness, lethargy, and cyanosis despite increasing respiratory distress should be considered as red flags for impending respiratory failure.

The illness tends to last for about 3-7 days but can persist for up to two weeks.

Differential diagnosis

- Epiglottitis.
- Inhaled foreign body.
- Inhaled noxious substance.
- Acute anaphylaxis.
- Bacterial tracheitis.
- Diphtheria.
- Laryngomalacia or another congenital cause of upper airway stenosis (eg, aortic arch abnormality causing external airway compression).
- Peritonsillar abscess (quinsy).
- Retropharyngeal abscess.
- Angioneurotic oedema.
- Laryngeal mucosal lesions such as laryngeal web, papillomata and haemangioma.
- Vocal cord paralysis.

Assessment of severity

There are many clinical scoring systems for croup. The most commonly cited is the Westley clinical scoring system which classifies cases into mild, moderate or severe.\(^9\)

It appears to be useful as a research tool to assess the efficacy of interventions but there is little proof of its clinical usefulness.\(^10\) The classification of symptoms it uses can be subjective and open to inter- and intra-observer variation.

An overall assessment of the patient's condition, taking into account the red flags listed above, SaO\(_2\) readings and the degree of respiratory distress, may be an equivalent and simpler guide to deciding what therapy is required.

The modified Westley clinical scoring system for croup

- **Inspiratory stridor:**
  - Not present - 0 points.
  - When agitated/active - 1 point.
  - At rest - 2 points.

- **Intercostal recession:**
  - Not present - 0 points.
  - Mild - 1 point.
  - Moderate - 2 points.
  - Severe - 3 points.

- **Air entry:**
  - Normal - 0 points.
  - Mildly decreased - 1 point.
  - Severely decreased - 2 points.

- **Cyanosis:**
  - None - 0 points.
  - With agitation/activity - 4 points.
  - At rest - 5 points.

- **Level of consciousness:**
  - Normal - 0 points.
  - Altered - 5 points.

Possible score 0-17: 0-3 = mild croup, 4-6 = moderate croup, >6 =severe croup.

Hospital assessment\(^5\)

Most children will have mild croup, which can be managed at home. Immediate assessment in hospital is required for:

- Moderate or severe croup, or impending respiratory failure.
- Any suspicion of epiglottitis, bacterial tracheitis, peritonsillar abscess, retropharyngeal abscess, or laryngeal diphtheria.
- Any suspicion of inhaled foreign body, angioneurotic oedema, hypocalcaemic tetany, or ingestion of corrosives.

Admission to hospital may be required if any of the following are present:
• History of severe obstruction, previous severe croup, or known structural upper airways abnormalities (eg, laryngomalacia, tracheomalacia, vascular ring, Down’s syndrome).
• Age less than 6 months.
• Immunocompromised.
• Inadequate fluid intake.
• Poor response to initial treatment.
• Uncertain diagnosis.
• Significant parental anxiety, late evening or night-time presentation, the child’s home is a long way from the hospital, or the parents have no transport.

Investigations
The diagnosis is usually made on clinical grounds but the following investigations may be indicated:

• A low SaO\textsubscript{2} on pulse oximetry (<95%) indicates significant respiratory impairment.
• It is important to weigh the benefits of investigations such as CXRs and blood tests against the risks of distressing the child and making the symptoms worse.\textsuperscript{[11]}
• A rapid influenza A test can be performed if it is considered vital to do so but even this investigation (which requires a throat swab) can distress the child.\textsuperscript{[12]}
• Direct or indirect laryngoscopy is not usually required but may be employed where the course of the illness is atypical or there is reason to suspect a congenital or other alternative cause for upper airway obstruction.

Management\textsuperscript{[5, 9]}

• Do not give antibiotics unless there are sound clinical reasons to suspect secondary bacterial infection.
• Keep the child as calm and as comfortable as possible. Allow the child to remain in a parent's arms and avoid any unnecessary painful interventions. Persistent crying increases oxygen demands and respiratory muscle fatigue and worsens the obstruction.\textsuperscript{[13]}
• Use paracetamol or ibuprofen to control any discomfort from symptoms or fever.
• Ensure an adequate fluid intake.
• Do not advise humidified air (eg, steam inhalation). A systematic review found that the croup score of children managed in an emergency setting with mild-to-moderate croup did not improve greatly with inhalation of humidified air. However, there was insufficient evidence to exclude either a small beneficial effect or a harmful effect. The authors commented that the studies they reviewed all took place in a hospital setting. The use of humidification in the community - frequently recommended in primary care - may at least reduce an anxious situation by giving the parents something to do. However, the benefits of this needed to be weighed against the risk of scalding to the child.\textsuperscript{[6]}
• Inpatient care includes oxygen therapy if required to maintain SaO\textsubscript{2} above 93%.

Steroids
Systematic reviews of the use of systemic dexamethasone and nebulised budesonide have shown that they are effective in easing symptoms within a few hours. Treated patients have fewer re-attendances or hospital admissions, and those who are admitted require shorter inpatient stays.\textsuperscript{[14]}

• Mild croup is largely self-limiting but treatment with a single dose of a corticosteroid (eg, dexamethasone 150 micrograms/kg) by mouth may be of benefit.\textsuperscript{[15]}
• More severe croup (or mild croup that might cause complications) requires hospital admission. A single dose of a corticosteroid (eg, dexamethasone 150 micrograms/kg or prednisolone 1-2 mg/kg by mouth) should be administered before transfer to hospital.\textsuperscript{[13]}
• In hospital, dexamethasone 150 micrograms/kg (by mouth or by injection), prednisolone 1-2 mg/kg by mouth or budesonide 2 mg (by nebuliser) will often reduce symptoms. The dose may need to be repeated after 12 hours if necessary.\textsuperscript{[15]}

Adrenaline (epinephrine)\textsuperscript{[9]}

• Nebulised adrenaline (epinephrine) is usually reserved for patients in moderate-to-severe distress.
• Nebulised adrenaline (epinephrine) solution 1 in 1,000 (1 mg/mL) should be given with close clinical monitoring in a dose of 400 micrograms/kg (maximum 5 mg) repeated after 30 minutes if necessary.\textsuperscript{[15]}
• The effects of nebulised adrenaline (epinephrine) last 2-3 hours and the child needs to be monitored carefully for any recurrence of the obstruction.\textsuperscript{[15]}

Complications\textsuperscript{[9]}

• Bacterial superinfection may result in pneumonia or bacterial tracheitis. The most frequent organism is Staphylococcus aureus, followed by group A streptococcus, Moraxella catarrhalis, Streptococcus pneumoniae, Haemophilus influenzae and anaerobes.
• Pulmonary oedema, pneumothorax, lymphadenitis and otitis media have also been reported.
• Inability to maintain adequate fluid intake may lead to dehydration.

Prognosis

• Complications are generally rare and case series show that 1.5-6% of children require hospitalisation.\textsuperscript{[16]} One study found that only 1 in 4,500 children with croup need to be intubated.\textsuperscript{[17]}
- Secondary bacterial infection leading to pneumonia or tracheitis is rare and pulmonary oedema and pneumothorax are extremely rare.
- Mortality rates in intubated children are less than 0.5%. [9]
Further reading & references

- MD Calc Westley Croup Score Calculator, 2015
- Nottingham Children's Hospital; Croup (Laryngotracheobronchitis), 2009.
- CDC; Guidance for Clinicians on the Use of Rapid Influenza Diagnostic Tests, 2014.
- British National Formulary (BNF); NICE Evidence Services (UK access only)

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