

# Recent Advances in Diagnosis and Management of Croup

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Croup is a syndrome of inspiratory stridor, cough, and hoarseness, due to varying degrees of laryngeal obstruction. It is a viral disease and must be differentiated from epiglottitis. In addition to a careful clinical assessment, neck films are the most valuable diagnostic tool in differentiating these two. The principle modes of therapy for croup include provision of adequate hydration, ensuring maximum available humidification, sedation, and administration of intermittent positive pressure breathing (IPPB) with nebulized racemic epinephrine. The latter mode of therapy will provide symptomatic relief and may eliminate the need for hospitalization and tracheostomy. Steroids offer little benefit in treating this disease, and antibiotics offer none.

Croup is a syndrome involving inspiratory stridor, cough, and hoarseness, due to varying degrees of laryngeal obstruction. More specifically, it is acute infectious laryngotracheobronchitis, which includes the disorders referred to as nondiphtheric croup, false croup, pseudocroup, and spasmodic croup. This common and potentially life-threatening illness requires careful evaluation and specific therapy. The purpose of this paper is to describe recent advances in the diagnosis and management of this illness in order to assure the best possible prognosis.

## Epidemiology

Croup is a disease of viral origin (see Table 1). The agents usually associated with croup are the parainfluenza viruses types 1, 2, and 3, respiratory syncytial virus, influenza virus A<sub>2</sub> and B, and occasionally adenoviruses, rhinoviruses, and enteroviruses.<sup>1,2</sup> In a study of lower respiratory illness seen

in a pediatric practice in Chapel Hill, 1963 to 1969, viruses thought to be the etiologic agents were isolated in 47.2 percent of the cases of croup.<sup>1</sup> In this study, 71 percent of the croup isolates were parainfluenza viruses; over half of these were parainfluenza virus type 1. Another 13.0 percent were respiratory syncytial virus, 4.6 percent were influenza virus A<sub>2</sub>, and the rest were distributed among a variety of other viruses. Croup is only rarely associated with simultaneous bacterial infection.<sup>3</sup> It has been well demonstrated that the viruses which cause croup can also infect other parts of the respiratory tract, and that the propensity to infect one part of the respiratory tract more than another is only relative.<sup>4</sup> There is, however, a striking correlation between infection with parainfluenza type 1 and the appearance of the croup syndrome.

There is a predictable seasonal occurrence of croup-associated viruses. Respiratory syncytial virus infections occur yearly between November and May, but predominate between January and March. Parainfluenza type 1 infections have occurred biennially, with a peak incidence in the autumn of even-numbered years. Parainfluenza type 2 has appeared in the autumn of odd-numbered years. Parainfluenza

type 3 is endemic almost year round, with a slight tendency to be less prevalent during the summer months. Influenza A<sub>2</sub> and B viruses have appeared in two to five-year cycles. Others are seen in small numbers without a seasonal pattern. Therefore, on an annual basis, croup is likely to occur in two cycles — during the winter months, especially January through March, and in the autumn.

## Diagnosis and Assessment

Differentiating subglottic viral croup from supraglottic epiglottitis is crucial. The important features that help to differentiate croup and epiglottitis are summarized in Table 2. Croup symptoms are somewhat gradual in onset, and they are generally of moderate severity. On the other hand, the onset of symptoms in epiglottitis is very rapid and the symptoms, particularly dysphagia, hoarseness, and dyspnea, are much more severe. The typical croup symptoms include coryza, fever, inspiratory stridor, expiratory barking cough, hoarseness, and varying degrees of respiratory distress. In the more severe cases, one may see tachypnea, tachycardia, decreased air entry, hypoxemia, cyanosis, and marked retractions. All of these symptoms are related to the acute narrowing of the subglottic airway.

The diagnosis of croup generally can be made on the basis of these presenting clinical features. However, careful examination of the posterior pharynx in an attempt to observe the epiglottis and appropriate x-rays of the neck and chest can be most helpful. Anterior-posterior and lateral x-ray projections of the neck during inspiration are necessary to evaluate croup and epiglottitis.<sup>5-7</sup> Figure 1 shows a lateral neck film taken in a case of epiglottitis, demonstrating the typical findings of swelling of the epiglottis and aryepiglottic folds, ballooning of

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the hypopharynx above this swelling, and a normal appearing larynx and subglottic trachea. Figure 2 shows an anterior-posterior neck film in a case of croup. While the radiologic features of croup vary with the severity of the illness, the typical picture as demonstrated here consists of concave medial swelling of the subglottic trachea for a distance of approximately 1.0 to 1.5 cm, with the trachea above the level of the true cords appearing normal. Occasionally, moderate ballooning of the

hypopharynx can be seen because of the relative obstruction during inspiration.

The utility of other laboratory tests in the diagnosis of croup is low. The white cell count is of little value. The frequency of occurrence of a white cell count in excess of 10,000/mm<sup>3</sup>, or a neutrophilia in excess of 70 percent or a combination of the two is no greater with bacterial isolates from lower respiratory tract infections than it is with viral isolates.<sup>3</sup> Cultures, even if they include both bacterial and viral cultures of the nasopharynx and throat, are rarely helpful in making an immediate decision about management. Acute and convalescent serum antibody titers can be used to help identify the offending viral agent after the illness has resolved for epidemiologic purposes.

Once the diagnosis of croup is well established, an assessment of the severity of the child's case should be made. The following clinical parameters should be assessed when examining a

child with croup: respiratory rate, pulse, presence and nature of cough, presence and degree of retractions, use of accessory respiratory muscles, increasing inspiratory to expiratory ratio, decreased air entry on auscultation, and presence and degree of cyanosis. Table 3 summarizes criteria that can be applied to evaluate the severity of each of these. As another means of assessing severity, in all but the mildest cases, blood gases should be obtained. In a series of 35 cases of croup, none of whom were cyanotic, 29 were found to be hypoxemic.<sup>8</sup> In that study, the respiratory rate was the best clinical correlate of hypoxemia, but no correlation could be found between the degree of hypoxemia and air entry, stridor, or other symptoms.

### Therapy

The principle modes of therapy to be considered in croup include provision of adequate hydration, humidification, oxygen, sedation, IPPB with nebulized racemic epinephrine, ste-

**Table 1. Causative Agents in Viral Croup in Order of Importance**

1. Parainfluenza virus 1, 2, 3
2. Respiratory syncytial virus
3. Influenza virus A<sub>2</sub>, B
4. Mycoplasma pneumonia
5. Enteroviruses
6. Adenoviruses
7. Rhinoviruses

**Table 2. Important Differentiating Features Between Croup and Epiglottitis**

Characteristics	Laryngotracheobronchitis	Epiglottitis
Age	usually under 3 years	usually over 3 years
Sex	boys > girls	boys > girls
Onset	gradual, usually over several days	rapid, often in a matter of hours
Antecedent symptoms	usually coryza	occasional coryza
Presenting symptoms		
fever	often none	present, often severe
hoarseness	may be present	usually severe
dysphagia	absent or mild	usually severe
stridor	moderate to severe	usually severe
toxic appearance	mild	severe
Presenting signs		
nasopharynx	hyperemia, edema	hyperemia, edema, excessive salivation
epiglottis	normal	swollen, cherry red
breath sounds	occasional rhonchi, decreased	no rhonchi, decreased
chest	retractions	marked retractions
Treatment		
antibiotics	not necessary	ampicillin or chloramphenicol
steroids	not effective	not effective
IPPB, racemic epinephrine	effective	not effective
tracheostomy	rarely necessary	usually necessary, recommended in all cases
mist	moderate benefit	moderate benefit
oxygen	often necessary	benefit only temporary, may give sense of improvement
Epidemiology	viral, multiple agents	H. influenza, type B
X-rays	normal supraglottic structures, concave medial swelling of hypopharynx	epiglottis swollen, filling hypopharynx, ballooning of hypopharynx, normal larynx and trachea

roids, antibiotics, and tracheostomy.

Increasing the water content of inspired air results in an increase in deposition of water in the airways and an addition of water to secretions and exudates, thereby thinning them and facilitating their removal by cilia and cough. In lower respiratory tract diseases such as croup, large amounts of nebulized water need to be provided for liquefaction of exudates in the lower airways.<sup>9</sup> This means that, in the home, a standard vaporizer or humidifier is of little benefit if no means is provided for limiting the volume for dispersion of the mist and, thus, concentrating it. The most effective method for providing sufficient mist concentration is either turning the bathroom into a steam room by running a hot water shower or constructing an enclosed "tent," such as one made by draping a sheet over the top and open sides of a crib. In addition, ultrasonic nebulization is significantly better than the home style vaporizers, so that in some cases it may be necessary to admit the child to the hospital in order to provide sufficient mist therapy. For hypoxemia, oxygen should be administered. Interestingly, inhalation of cool air, such as cool night air, also may relieve symptoms.

The child with croup is usually irritable, and his irritability frequently causes increased stridor and dyspnea. As much as possible, the child should be placed in a familiar, quiet, controlled environment in an effort to provide maximum sedation. If pharmacologic sedation is needed, either phenobarbital (2 to 3 mg/kg per dose) or chloral hydrate (5 mg/kg per dose) is effective.

The use of corticosteroids is of little apparent benefit in the management of croup. In previously reported studies, adrenocorticotrophic hormone (ACTH), hydrocortisone, prednisone, prednisolone, methyl prednisolone, and dexamethasone all have been used. Eight controlled studies have been reported. Six of these studies demonstrated no difference between control groups and corticosteroid treated groups with respect to clinical response (decrease in stridor, dyspnea, retractions, cyanosis), length of hospital stay, or number of tracheostomies performed.<sup>10-15</sup> In one study, patients treated with corticosteroids improved more rapidly than did a control

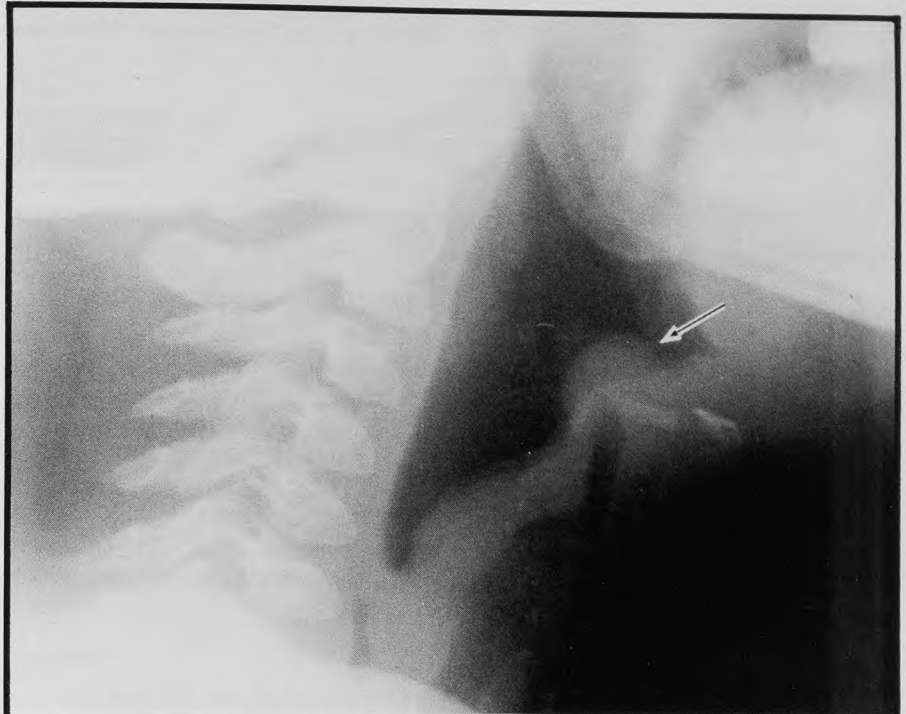


Figure 1. Lateral neck film showing swollen epiglottis, ballooning of hypopharynx, and normal subglottic airway in a case of epiglottitis

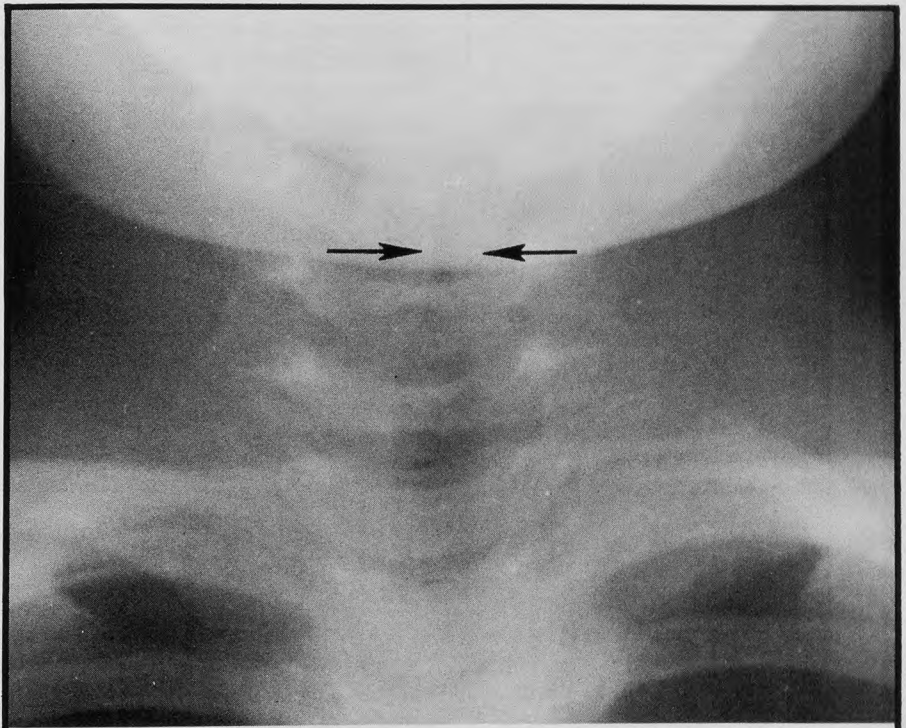


Figure 2. Anterior-posterior neck film showing concave medial swelling of subglottic airway in a case of croup

Table 3. Assessing the Severity of Viral Croup

Clinical Parameter	Degree of Severity			
	Normal	Mild	Moderate	Severe
Cyanosis	No cyanosis or hypoxemia with exertion	Hypoxemia or slight perioral cyanosis on exertion	Cyanosis on exertion	Cyanosis present at rest
Respiratory rate	Less than 30	30-39	40-59	60 or greater
Cardiac rate	Less than 110	110-139	140-159	160 or greater
Cough	None	Expiratory "bark" without inspiratory stridor	Expiratory "bark" with noticeable inspiratory stridor	Expiratory "bark" with predominant inspiratory stridor
Air entry	Normal	Air entry diminished	Air entry diminished, with a prolonged inspiratory phase	Little, if any, air movement is heard on auscultation
Retractions	None	Suprasternal only	Suprasternal and infrasternal	Suprasternal, infrasternal, and intercostal
Use of accessory respiratory muscles	None	+	++	+++
Inspiration expiration ratio	2:1	3:1	4:1	> 4:1

Table 4. Summary of Therapeutic Management of Croup

1. Encourage maximum available humidification.
2. Provide sedation when necessary.
3. Encourage adequate hydration.
4. Administer nebulized racemic epinephrine by intermittent positive pressure breathing for moderate to severe distress.
5. Administer oxygen for significant hypoxemia.
6. Tracheostomy should rarely, if ever, be needed.
7. Steroids are of no benefit.
8. Antibiotics are of no benefit.

group. However, the difference occurred only in patients with a history of allergic manifestations (urticaria, hay fever, eczema, asthma). If patients did not have an allergic history, there was no difference in response between the steroid treated and control groups.<sup>16</sup> One other study found a variable response in patients who were given steroids when compared to controls. There was no difference in pulse rate, respiratory rate, or length of time the patient was in a croupette, but the steroid treated group fared better with

respect to relief of stridor, retractions, and length of hospital stay. Unfortunately, the data in this study is complicated by their finding that the relief of stridor and the decrease in length of stay was present at a dose of 4 mg dexamethasone given every six hours for 24 hours, but when the dose was increased to 6 mg every six hours for 24 hours, no difference between the treated and untreated groups could be seen. Conversely, the positive benefit of decreased retractions was found only with the higher dose of dexamethasone and not with the lower dose.<sup>17</sup> The results of these studies cast significant doubt as to whether there is any benefit from corticosteroid therapy, with the possible exception of the case of a child with a history of previous definite allergic manifestations.

By definition, viral croup should not be treated with antibiotics. There is no reason to expect any benefit from antibiotics, and the child should not be subjected unnecessarily to the hazard of allergic manifestations or other adverse reactions to these drugs.

One of the newest modes of therapy to be used in the management of croup is intermittent positive pressure breathing using nebulized racemic epinephrine. Adair et al reported on ten year's experience with this mode of therapy in 1971.<sup>18</sup> Initially this meth-

od was used in the most severe cases of croup, but has come to be used in the mild or moderately severe case. The method uses nebulized 2.5 percent racemic epinephrine solution, diluted 1:8 with water, administered with a positive pressure respirator for 15 minutes using a face mask. If the response is not satisfactory or if symptoms recur, additional treatments can be given. Adair et al reported that during a seven-year period, while they were using this mode of therapy, not a single tracheostomy for croup was performed at their institution and not a single death occurred. By contrast, during the three years previous to this, tracheostomies were performed in seven percent of the cases admitted to the hospital. They also reported a decrease in the average length of hospitalization of over 50 percent. While using a similar method, Melnick et al also reported the successful elimination of the need for tracheostomies over a three-year period.<sup>19</sup> On the other hand, another study found no change in the length of hospital stay, frequency of tracheostomies, or deaths, when comparing the period during which racemic epinephrine was used to a period during which it was not.<sup>20</sup> However, these same authors found that 31 out of 35 children evaluated showed clinical improvement following racemic epinephrine

therapy. Whether the critical element in the therapy is the IPPB or racemic epinephrine is not clearly delineated. In this same study, nebulized racemic epinephrine was compared to nebulized normal saline in a controlled, double-blind study of 20 children. The majority of the children in both groups showed a clinical response, but the researchers were unable to demonstrate any difference between the two groups.<sup>20</sup>

The reported studies indicate that intermittent positive pressure breathing treatments with nebulized racemic epinephrine are effective in relieving the symptoms of croup. As a matter of personal experience, we have used it and find it to be an effective way to provide symptomatic relief to the dyspneic child with stridorous, labored breathing. It is not clear whether this mode of therapy is effective in eliminating the need for tracheostomies. Some studies of croup treated without using IPPB during the same years as those reported by Adair et al show a similar record of not having had to perform a single tracheostomy.<sup>13</sup> However, other series associated with epidemics within this period show a very high frequency of tracheostomies.<sup>21</sup>

The use of IPPB also may decrease the need for hospitalizations. Al-

though no clear evidence has been reported to date, it is our experience that children with mild to moderate croup symptoms can be treated effectively in the Emergency Room with IPPB and returned home. While the evidence also is unclear about the impact of this therapy on length of hospital stay, a decrease in the rate of hospitalization would be an important benefit. One caution with IPPB is that the diagnosis of croup must be accurate. IPPB is not effective in epiglottitis and may delay appropriate treatment and tracheostomy in that condition.

Additional therapy in the form of fluids, electrolytes, buffers and antipyretics may be necessary. If managed appropriately, croup should have an excellent prognosis.

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