Epiglottitis: Diagnosis and Treatment

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Epiglottitis is a rapid and potentially fatal infection of the supraglottic larynx that causes death by upper airway obstruction and Gram-negative bacteremia with *Haemophilus influenzae* type B (HIB). The majority of patients (80%) are less than 5 years of age, and the remaining 20% are scattered over the remaining life span. Although sporadically recognized since the early 16th century, the disease has only become well-described since the late 1940s. Initial mortality rates approaching 100% have steadily declined to about 2% with the use of artificial airways and antibiotics.

The key to obtaining a good outcome with available effective therapy in this disease is prompt recognition and rapid initiation of therapy. Delays of even a few minutes can be disastrous. Advanced planning for the handling of such patients in the office, clinic, or emergency room has been effective in avoiding death from such delays. Decisive action can make the difference between death or anoxic damage as an outcome, and complete recovery without sequelae.

EDUCATIONAL OBJECTIVES

Evaluate the child with fever and croup and be able to diagnose and manage: (1) acute viral laryngitis; (2) laryngotracheobronchitis; (3) diphtheria; (4) foreign body; (5) retropharyngeal abscess; (6) epiglottitis; and (7) adenoiditis.

CLINICAL PICTURE

The disease typically begins abruptly with the onset of fever greater than 102° F (rectal) and sore throat or difficulty swallowing. These two features of epiglottic inflammation signal the onset of this illness and occur simultaneously or within a few hours of each other. If seen at this stage, the diagnosis is not simple, but is suggested by absent or minimal pharyngeal findings and toxicity. About 20% of patients have a cough and, in some, vomiting is present. Respiratory distress is absent initially and this makes the diagnosis more obscure. Some patients are reluctant to move their neck and this may mimic nuchal rigidity, but Kernig's sign is absent. In contrast to patients with meningitis, these patients may resist neck rotation as well as flexion.

A toxic child with fever and signs of pharyngeal tenderness with a normal throat inspection may well have epiglottitis. A soft tissue lateral neck x-ray will usually demonstrate characteristic radiographic features even at this stage.

As the swelling increases, respiratory distress gradually develops over the next few hours. Slight tachypnea with flaring of the alae nasi, retractions of the chest wall, and use of the accessory muscles of respiration progresses with the appearance of characteristic raspy, coarse inspiratory breath sounds quite distinct from the almost musical tone heard in patients with croup. The voice may be noticeably muffled as if the child has a mouthful of mashed potatoes, but this may be a subtle sign, apparent only to the mother. At this stage the child assumes the classic posture of epiglottitis; sitting upright, leaning forward, open-mouthed, and finally drooling as the swollen epiglottis becomes too painful even for swallowing saliva. Although some children at this stage are obviously air-hungry and anxious, others are remarkably still and seem intent on holding their position to maximize ventilation. With the onset of respiratory distress, the diagnosis can virtually always be made from the history and observation of the position of the patient (Table).

From this point onward, the child should have supplemental oxygen (humidified, if possible), proper airway equipment, and experienced cardiopulmonary resuscitation personnel at his side until an endotracheal tube or tracheostomy can be placed. Once tachypnea begins, the child begins to tire and respiratory and cardiac arrest will follow, within minutes to a few hours.

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TABLE. Diagnosis of Epiglottitis

Early symptoms
Fever
Sore throat with minimal or absent pharyngeal findings
Swallowing difficulty
Toxicity
Later symptoms—signs
Limited neck mobility, both flexion and rotation
Muffled voice
Tachypnea, tachycardia
Flaring of alae nasi
Retractions of the chest wall
Raspy inspiratory sound
Late signs
Drooling
Air hunger
Sitting forward

TRANSPORTATION

Once the diagnosis is made or suspected, transportation to a hospital or to a place within a hospital where definitive care can be rendered must be accomplished rapidly and safely. The child should be closely monitored and constantly attended by someone trained in cardiopulmonary resuscitation (CPR). Administration of *humidified* oxygen will temporarily benefit the child while in transit. The child should be disturbed as little as possible (eg, no venipuncture) and usually is most calm if held in an upright position in his mother's arms.

If the child appears in extremis and the transport time is prolonged (greater than 30 minutes), emergency intubation or tracheostomy prior to transporting should be considered seriously. Alternatively, meticulous and forceful mouth-to-mouth or bag and mask ventilation should be the initial therapeutic approach to sudden airway obstruction, since these techniques are generally available and familiar to pediatricians.

Ambu bags, Hope resuscitation bags, and similar self-inflating bags are equipped with pressure-release valves which open when ventilation pressures exceed approximately 50 mm Hg. Patients with epiglottitis whose airway is obstructed may require pressures greater than 50 mm Hg to achieve ventilation. Thus, ventilation may be impossible using a bag with a pressure-release or "pop-off" valve. Many chambers that are used to humidify oxygen are equipped with a similar pressure-release valve. If ventilation is not achieved when using equipment with pressure-release valves, change to either mouth-to-mouth ventilation or use a limp bag that inflates by incoming gas pressure connected directly to the oxygen outlet without a humidifying chamber. Both techniques can produce the higher pressures needed to overcome the upper airway obstruction.

Transport, even within the hospital, should follow these guidelines. If transporting from the office or clinic to a hospital, specific information (age, heart rate, respiratory rate, estimated arrival time) should be transmitted early on to ensure maximal preparedness at the center.

DIAGNOSIS

Confirmation of the diagnosis involves two considerations: (1) Is it epiglottitis? and (2) What is the causative organism and its antibiotic sensitivities?

If the patient presents the typical picture as described, the diagnosis is virtually certain. If significant respiratory distress is present, as shown by more than a 50% increase above the normal respiratory rate, further diagnostic efforts, especially throat culture, should be deferred until someone with expertise suitable to secure the airway is present. If the patient is not having respiratory distress, a soft tissue lateral neck x-ray is most helpful. The patient should be attended by CPR personnel, and humidified oxygen should be provided continuously while obtaining the roentgenograms. Although only anecdotal information may be available, most authorities consider examination of the posterior pharynx with a throat stick or mirror to be too dangerous since even minor alterations of the child's position may produce total obstruction. In a modest number of instances, the child can voluntarily open his mouth wide enough to enable the examiner to see the swollen cherry-red epiglottis without using a stick or changing the child's position.

The lateral neck radiograph must be reviewed promptly since the decision to perform a tracheotomy or to intubate the patient may be influenced by the interpretation. Fig 1 is a lateral neck film typical of those seen with epiglottitis. There are five hallmarks: (1) an open and/or protruding jaw, (2) a dilated hypopharynx, (3) encroachment of the lingual aspect of the epiglottis on the vallecula, (4) a doughy, thickened appearance of the enlarged epiglottis, arytenoids, and aryepiglottic folds which resemble the shadow of an adult thumb, and (5) a straight or kyphotic cervical column as opposed to the usual lordosis. The appearance of the supraglottic structures ("thumb sign") is most critical, while the other findings (1, 2, 3, and 5) may be variable depending on the degree of obstruction. For comparison, a normal lateral neck film is shown in Fig 2, and a case with croup (laryngotracheobronchitis) is shown in Fig 3 where the subglottic haze and narrowing are apparent. An anterior-posterior film done at the same time as the lateral view allows for the best discrimination between epiglottitis and croup.

Identification of a bacterial etiology for epiglottitis is best done by blood culture, after the airway is secured. If drawn shortly after intubation or tracheostomy, more than 80% of such cultures will be positive for HIB. Other pathogens are isolated from the blood in less than 1% of the reported cases. Typically, three or four days are necessary for adequate growth in the laboratory, and thus decisions about antibiotics must be made before the results are available. If counter-immunoelectrophoresis or latex fixation antigen studies on the blood are available, added certainty of the presence of HIB can be available immediately. These studies are often positive

even in patients who fail to demonstrate the organism in blood cultures.

Nose, throat, or epiglottal surface swabs yield HIB in about half the cases, but are also often positive for *Streptococcus*, *Neisseria* and other pathogens from the oropharynx which do not correlate positively with the blood cultures. Thus, these cultures cannot be assumed to be diagnostic. They should be obtained only with airway immediately available.

Antibiotics should be administered intravenously immediately after the blood cultures have been drawn as detailed below.

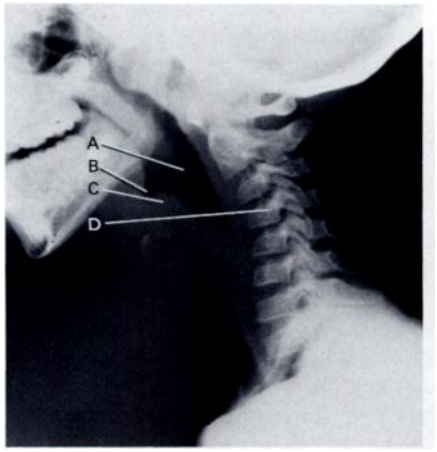

Fig 1. Lateral radiograph of the neck of a patient with epiglottitis: A, dilated hypopharynx; B, reduced size of vallecula due to encroaching swollen epiglottis; C, rounded, thickened epiglottis (``thumb sign'') with swollen aryepiglottic folds extending posteriorly and inferiorly; D, reversed cervical curvature. This radiograph does not demonstrate an open mouth and protruded mandible characteristic of more advanced obstruction.

Fig 2. Lateral radiograph of the neck of a normal child. The vallecula is spacious and the supraglottic structures are delicate.

Fig 3. Lateral radiograph of the neck of a child with laryngotracheobronchitis (croup). The hypopharynx is dilated, but the vallecula and epiglottis are normal. Note the haziness and blurring of the air column at the level of the fourth cervical vertebra indicating subglottic swelling.

TREATMENT

The primary therapy before and en route to securing the airway is humidified oxygen administered continuously. Children often sense the anxiety of their parents and staff and become frightened by the oxygen mask or funnel. Generally, the child has better acceptance of oxygen therapy when it is administered by the mother.

The choice of method for securing the airway depends primarily on the resources available. If appropriate intubation expertise, nursing skills, and prompt replacement capability in the event of accidental extubation are all present, then intubation is the preferred method, having slightly lower morbidity and the advantage of avoiding a surgical procedure. However, if the available skills favor tracheostomy, it is entirely appropriate. The intubated child with epiglottitis requires experienced nursing attention to prevent plugging of the tube by secretions, and accidental or self-extubation. The ever-present hazard of accidental extubation also requires the availability of skilled personnel to re-intubate on short notice. Accidental decannulation of a tracheostomy tube with soft tissue collapse into the stoma can likewise create an emergency condition, but it is technically easier to pass an endotracheal tube into the stoma than to re-intubate transorally or transnasally, particularly if retention sutures are in place to assist in elevation of the trachea.

Endotracheal tubes should be suctioned hourly, and regular chest physical therapy should be employed to mobilize secretions. Intubated patients are usually placed in humidified tents to be sure that all inspired air is moist, whether inhaled through the tube or around it, since these patients seem to have appreciable leakage of air around the tube.

Oral endotracheal tubes are treacherous and should be avoided if at all possible in favor of nasal endotracheal tubes. Orally placed tubes are irritating to the patient, easily dislodged by tongue and jaw motions, and may be associated with life-threatening aspiration should the patient vomit, since the oral airway (placed to prevent biting through the endotracheal tube) occludes the oropharynx. The placement of a nasogastric tube helps to reduce this risk during the period of intubation, even when nasal endotracheal tubes are used.

The airway tube is left in place between 24 and 72 hours, and after extubation, the patient should be closely monitored for several hours. An additional 24 hours of humidification following extubation is advisable.

Antibiotics are essential in therapy, but their initiation should not

delay securing the airway. Since more than 99% of cases are due to HIB, the choice of antibiotics depends on the local sensitivity patterns of HIB. If ampicillin-resistant HIB are present locally, chloramphenicol, 25 mg/kg IV every six hours (100 mg/kg/24 hours), is preferred. This dose is maximal, and close monitoring of blood counts, and/or blood levels, if available, is needed, especially for patients under 2 years of age. Once sensitivities are available, it may be possible to change from chloramphenicol to ampicillin. If no ampicillin-resistant HIB have been demonstrated locally, ampicillin, 100 mg/kg IV every six hours (400 mg/kg/24 hours), may be used initially, although many clinicians prefer to use chloramphenicol in all cases until sensitivity results are reported.

Treatment with these dosages of ampicillin or chloramphenicol given intravenously for seven days has been completely effective and the patient can be discharged without any additional medication.

Children with epiglottitis should not be sedated before the airway is secured by nasotracheal intubation or tracheostomy. After the airway is secured, the intubated child may require sedation while the endotracheal tube is in place. Chloral hydrate given by the nasogastric tube or rectally at a dosage of 25 to 40 mg/kg/day divided into four equal doses is commonly used. Even in the child with an endotracheal tube or tracheostomy, the possibility of hypoxic agitation must be constantly considered before ordering sedation since the airway appliance may become blocked with secretions at any time.

Although steroids have been advocated in the treatment of epiglottitis to reduce the swelling associated with the inflammation, there is no persuasive evidence that they are of benefit. Racemic epinephrine, perhaps of some value in croup, is not recommended in epiglottitis. Most clinicians feel that the increased oxygen consumption induced by the drug and the risks of manipulating a child with a tenuous airway are sufficient reasons not to use racemic epinephrine.

Other foci of HIB infection have been reported in epiglottitis patients, but their incidence is low. Once the airway is secured and antibiotics begun, a careful examination for evidence of pneumonia, meningitis, septic arthritis, and pericarditis is in order. Although there is increasing concern that HIB infections may be associated significantly with secondary cases, prophylaxis is not currently recommended for siblings and other contacts. However, should siblings or contacts develop fever (greater than 102 F) they should be evaluated by their physician in light of their exposure to a child with HIB epiglottitis.

In more than 500 reported cases in the literature, HIB epiglottitis is known to have recurred only once.

CONCLUSION

Epiglottitis is a life-threatening infectious disease characterized by the abrupt onset of fever and dysphagia followed shortly by respiratory distress. Initial management focuses on maintaining oxygenation and placing an artificial airway as quickly as possible. Soft tissue radiographs of the neck can help establish the diagnosis in questionable cases. Intravenous antibiotics to cover HIB are instituted after the airway is secured and blood cultures obtained. An excellent outcome can be achieved if patients are given oxygen promptly and attended closely from presentation to placement of an airway.

RECOMMENDED READING

Berenberg W, Kevy S: Acute epiglottitis in childhood: A serious emergency, readily recognizable at the bedside. *N Engl J Med* 258:870, 1958

Cantrell RW, Bell RA, Morioka WT: Acute epiglottitis: Intubation versus tracheostomy. *Laryngoscope* 88:944, 1978

Daum RS, Bates JR, Smith AL: Epiglottitis (supraglottitis), in Feigan RD, Cherry JD (eds): *Textbook of Pediatric Infectious Diseases.* Philadelphia, WB Saunders, in press 1980

Myers M: More on treatment of epiglottitis, letter. *J Pediatr* 83:168, 1973

Rapkin RH: The diagnosis of epiglottitis: Simplicity and reliability of radiographs in the neck in the differential diagnosis of the croup syndrome. *J Pediatr* 80:96, 1972

Abstract

Nebulized Racemic Epinephrine by IPPB for the Treatment of Croup. Estley CR, et al. *Am J Dis Child* 132:484, 1978.

In a well controlled study of viral croup, patients who received racemic epinephrine by intermittent positive-pressure breathing (IPPB) were helped compared to controls. The help was short-lived; ie, there was only a short response to therapy and then return to distress. Therefore, therapy will need to be repeated and the patient should be hospitalized. Whether repeat therapy remains effective has not been studied; whether IPPB with epinephrine shortens hospitalization has not been studied.

Comment: If a child with croup is hospitalized and if he does not have epiglottitis and if he looks like he may need to be intubated (rising Pco_2s) then I would try IPPB with epinephrine (racemic or 1% aqueous are equivalent). If there was a response I would repeat as often as necessary to prevent intubation. If no response I would intubate. (*R.H.R.*)

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