



Epiglottitis, Acute Laryngitis, and Croup

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Introduction

Epiglottitis, acute laryngitis, and croup are infections of the upper airway, affecting the epiglottis, larynx, and larynx and trachea, respectively. Croup may also involve the bronchi. Epiglottitis is a bacterial infection, while acute laryngitis and croup are primarily viral infections.

Epiglottitis was known to the ancient world. The infection was probably described by Hippocrates 2400 years ago when he wrote: “There is fever, chill, pain in the head, the underpart of the jaw is swollen: the patient swallows with difficulties his saliva. The patient cannot spit, he cannot tolerate to lie down, and if he stays in this position he chokes” [1]. In 1791, the first clear description of acute epiglottitis was published, and the term “epiglottitis” was coined in 1830 [2]. Invention of the laryngoscope in the 1850s allowed direct visualization of the swollen epiglottis, and soon thereafter the mirror examination of the hypopharynx and larynx became widely practiced. In the early twentieth century, the term “acute epiglottitis” became part of standard medical terminology and the role of bacteria

in the pathogenesis of the disease became clear. The prognosis of the disease improved in some cases with tracheotomy, tracheostomy, or intubation. George Washington, the first president of the United States, died in 1799 of an acute infection that was probably epiglottitis [3]. A tracheotomy (a new procedure then) was proposed by one of his physicians but not performed. The major advance in treatment of epiglottitis occurred following the introduction of antibiotics in the mid-twentieth century. The introduction of the *Haemophilus influenzae* type b (Hib) vaccine in 1985 dramatically decreased the incidence of epiglottitis, especially in the pediatric population. The vaccine was improved (conjugated vaccine) in 1987 and 1990 so that efficacy extended to children younger than 18 months.

Croup, an old term meaning “to cry hoarsely,” was originally applied to cases with croup-like symptoms (inspiratory stridor, hoarseness, and a barking cough) due to diphtheria. However, diphtheritic “croup” was subsequently distinguished from viral croup by the twentieth century. Croup now refers to acute viral laryngotracheobronchitis or laryngotracheitis.

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Acute Epiglottitis (Supraglottitis)

Epiglottitis, also called supraglottitis, is an acute infection of epiglottis and surrounding supraglottic tissues that can rapidly lead to life-threatening

airway obstruction. Supraglottitis is a more accurate term because in many cases, several supraglottic structures are involved (e.g., aryepiglottic folds, arytenoids). Prior to the introduction of the Hib vaccine 30 years ago, *H. influenzae* type b caused most cases of epiglottitis and the disease primarily affected children younger than 5 years old. Now, invasive *H. influenzae* type b infections are very rare in children and the incidence of epiglottitis is higher in adults than in children.

Epidemiology

Before the Hib vaccine, the incidence of epiglottitis in children under age 5 years was as high as 15 cases per 100,000 population. The peak incidence occurred in children under age 3. Since the introduction of the Hib vaccine in 1985, the incidence in children has decreased to 0.5–0.7 cases per 100,000. In Finland, 50–60 cases per year of epiglottitis were seen throughout the country in 1985–1986, and this decreased to only two cases in 1992, reflecting the widespread use of the Hib vaccine [4]. In Sweden, the incidence of epiglottitis in children under age 5 decreased from 21 to 0.9 cases per 100,000 following Hib vaccination [5]. The median age of children affected by epiglottitis has also increased in the Hib vaccine era. At one center, the median age of children with epiglottitis before 1989 was 36 months and 81 months (nearly 7 years) after 1990 [6].

The incidence of epiglottitis in adults has remained stable or has increased since the introduction of the Hib vaccine [7, 8]. In Finland, the annual incidence of epiglottitis in adults increased from 1.88 (1990–1999) to 4.7 (2000–2009) per 100,000 [8]. In Denmark, the annual incidence in adults has been stable at 1.9 cases per 100,000 [9]. Most cases of epiglottitis now occur in adults. A study from Australia found that 84% of patients hospitalized for epiglottitis in the vaccine era have been adults [10]. The average age of adults with epiglottitis is mid-40s, but the range in one series was wide (age 18–92 years) [8].

Epiglottitis usually occurs in previously healthy children or adults. There is no clear seasonal variation. Slightly more males than females are affected, with a ratio of 1.4:1–1.7:1 in most series [8].

Pathophysiology

The epiglottis is a leaf shaped elastic cartilage with overlying loose connective tissue and a thin mucosa. The laryngeal airway in children is narrow. A comparatively minor swelling of the mucosa may cause significant airway narrowing. Acute epiglottitis usually starts with swelling of the lingual surface of the epiglottis, and then spreads to the laryngeal surface and the aryepiglottic folds. Bacterial invasion of the mucosa leads to fulminant infection. Especially in children, the epiglottis is the primary focus of infection and other parts of larynx may not be swollen. An epiglottic abscess can develop in either adults or children.

Microbiology

As noted above, *H. influenzae* type b caused most cases of epiglottitis in the pre-vaccine era. This was determined from blood cultures (positive in many children with *H. influenzae* epiglottitis), and some throat cultures. The true etiology of epiglottitis is now difficult to determine in most cases, because adults comprise the majority of cases and only 10% are bacteremic [8]. Throat cultures are contaminated by oral flora so may not demonstrate the true pathogen. In a series of 34 adults, only one had a positive blood culture (*H. influenzae*), while eight with throat cultures grew oral flora organisms in seven and *H. influenzae* in one [11].

Streptococci such as *Streptococcus pneumoniae* and Group A *Streptococcus* are the predominant causes of epiglottitis in most series in the vaccine era [8, 9, 12]. *Staphylococcus aureus* causes some cases, and cases due to methicillin-resistant *S. aureus* (MRSA) have been described [13]. A large series of epiglottitis in adults

reported positive blood cultures in 16 (10% of those in whom blood cultures were obtained) [8]. Of these 16 cases, cultures grew streptococci (including *S. pneumoniae*, Group A *Streptococcus*, *Streptococcus milleri*) in 11 (69%), *S. aureus* in two (13%), and Gram-negative bacilli in two (13%) including a case due to *Pseudomonas* [8]. Only one patient in that study had a culture positive for *H. influenzae*, and that grew on a throat culture.

The microbiology of epiglottitis in children is similar to that in adults. The Hib vaccine effectiveness is approximately 98%, so cases of epiglottitis due to *H. influenzae type b* are rare but do still occur. In one series of 19 children admitted in the vaccine era, *H. influenzae type b* was the etiology of six cases; streptococci accounted for another six cases (*S. pneumoniae*, Group A *Streptococcus*, other beta-hemolytic streptococci) [14]. In immunocompromised patients, *Pseudomonas* and *Candida* have caused some cases of epiglottitis [15–17].

Clinical Presentation

Children

Children with epiglottitis typically present with a high fever, inspiratory stridor, restlessness, and drooling. Breathing difficulties may cause a toxic and anxious appearance. The child usually presents sitting forward in a “tripod” or “sniffing” position and does not want to lie down. The child’s head is hyperextended to maintain a patent airway. The child has pain with swallowing and may have a sore throat. The voice is often muffled, described as a “hot potato voice.” Symptoms arise very quickly, in most cases within 12–24 h including symptoms of any antecedent upper respiratory tract infection. Unlike croup, cough is uncommon. Children with epiglottitis are often systemically unwell and the situation is considered an otolaryngologic emergency.

The differential diagnosis in children may include croup (acute laryngotracheobronchitis), discussed later. However, children with croup

have a barking cough and are less likely to have drooling or to insist on sitting forward in the “sniffing” position. Children with croup usually appear less toxic than children with epiglottitis.

Adults

Symptoms of acute epiglottitis usually develop more slowly in adults than in children. The most common symptom, seen in nearly all the cases, is sore throat. In a study from Finland of 308 adults admitted with epiglottitis 1989–2009, the mean duration of symptoms prior to presentation was 3.7 days, and 94% of patients complained of sore throat [8]. Other symptoms included odynophagia or dysphagia (80%), fever (60%), dyspnea (38%), hoarseness (30%), muffled voice (12%), drooling (11%), cough (8%), and stridor (7%) [8]. Patients with epiglottitis find breathing in the supine position difficult or impossible and prefer to sit, leaning forward with a hyperextended neck. The patient may cough although infrequently, to clear secretions in the hypopharynx. An epiglottic abscess may be present and this is a risk factor for requiring airway intervention [7]. The differential diagnosis of epiglottitis in adults includes other causes of acute sore throat and severe odynophagia. In epiglottitis, however, the sore throat is more severe than expected based on the findings of the oropharynx examination.

Diagnosis and Evaluation

Children

The extent of physical examination performed prior to securing the airway should be individualized, depending on the severity of the child’s illness and the likelihood that their illness is epiglottitis. Rare cases of cardiopulmonary arrest have occurred with attempts at visualization of the epiglottis. Similarly, asking the child to lie supine may be dangerous and respiratory arrest has been reported in this position. When acute epiglottitis is suspected, arrangements to secure the airway should be made immediately. The help of a pediatric otolaryngologist or anesthesiologist is often necessary.

Supplemental humidified oxygen should be given. In children who are not in any respiratory distress, clinical examination may be cautiously performed as long as airway rescue is readily available. The classic red epiglottitis may be visualized by gently depressing the anterior tongue with a tongue depressor. Similarly, plain neck x-rays can be obtained in the non-acute setting as the patient can remain in the seated position. The lateral view will indicate thickening of the epiglottis, similarly the anterior view will evaluate for subglottic finding which may be associated with other diagnoses in the differential such as croup. All children suspected of having epiglottitis who are undergoing x-ray evaluation should be closely monitored and observed during the entire period while these films are obtained. In more severe cases, the child should be transported to the operating room where the airway can be secured, by controlled intubation or tracheotomy, and diagnostic endoscopy can be performed.

Diagnostic tests such as blood cultures and routine laboratory tests (e.g., complete blood count with differential) should be performed only after the airway is secured. Throat or epiglottic cultures should be obtained only after the airway is secured. If a deep neck infection or epiglottic abscess is suspected, computed tomography (CT) with contrast can be considered after the airway is secured.

Adults

If acute epiglottitis is suspected, patients should be kept in the sitting position and not asked to lie down. In severe cases with impending airway obstruction, the airway should be secured prior to any attempt at examination. In most cases of adult epiglottitis, there is less risk to performing a direct examination than in children. However, a specialist such as an anesthesiologist or otolaryngologist should be on hand, as the patient can deteriorate rapidly. Indirect laryngoscopy with mirror examination can usually be performed in adults to diagnose epiglottitis, although nasolaryngeal endoscopy with a flexible fiberoptic

may be safer and more easily done. The diagnosis of epiglottitis is made by noting a swollen and red epiglottis, although in adults the primary site of swelling may be in other supraglottic structures. One study found swelling of the epiglottis in 73% of adults with epiglottitis but swelling of the epiglottis alone occurred in only 17% [8].

After confirming the diagnosis, the airway should be secured if necessary. An intervention (intubation or tracheotomy) may not be necessary in adults with epiglottitis. In a study of 61 patients in the vaccine era (60 adults, 1 child), only 21% required airway intervention (11 intubations, 2 tracheotomies) [12]. In a study of 308 adults with epiglottitis, 15% required airway intervention (two-thirds intubation, one-third tracheotomy) [8]. Factors associated with the need for airway intervention include a rapid onset of symptoms, stridor, drooling, and tachypnea [8, 12]. Diagnostic tests should be done once the airway is secured, or once it has been determined that the airway is sufficiently patent so as to not require immediate intervention. If there is a suspicion of deep neck infection or an epiglottic abscess, a CT with contrast should be considered once the airway is secure.

Treatment

Medical Treatment

Empiric antibiotic treatment of acute epiglottitis in children or adults should cover *H. influenzae*, *S. pneumoniae* (including penicillin-resistant strains), Group A *Streptococcus*, and *S. aureus* (including MRSA). Intravenous vancomycin plus ceftriaxone will cover these organisms. Even in regions where MRSA is rare, the addition of vancomycin to ceftriaxone is usually recommended because some *S. pneumoniae* isolates are not susceptible to ceftriaxone, and vancomycin will provide additional staphylococcal coverage. In immunocompromised patients, an antibiotic regimen that will also treat *Pseudomonas* should be considered (e.g., intravenous vancomycin plus either meropenem or imipenem). Antibiotics can be tailored once results of blood and throat cultures are known.

Airway Management

Managing the airway is the utmost importance in epiglottitis. Even in patients who appear stable, clinical deterioration can occur at any point. Acute loss of the airway is the primary cause of death in epiglottitis.

Securing the airway by intubation usually can be performed safely, but requires experienced and expert anesthesiologists or otolaryngologists. Preparations for obtaining a surgical airway, such as tracheotomy, should always coincide with any attempt at intubation, as acute airway obstruction can occur with a failed intubation attempt as discussed later. Awake fiberoptic intubation can be attempted in the cooperative patient as an initial attempt to secure the airway. Direct and video-assisted laryngoscopy may be attempted, but the necessary sedation requires rapid access to airway and associated swelling and secretions can limit visualization. If an emergency surgical airway is required, the type depends on the age of the patient. Needle cricothyrotomy can be performed on patients of any age, but this is the preferred surgical technique in children under age 12. Needle cricothyrotomy is easier to perform and less likely to damage the larynx in young children than surgical cricothyrotomy. Needle cricothyrotomy provides stabilization of the patient allowing a formal surgical airway such as a cricothyrotomy or tracheotomy to be performed in a controlled manner. Should immediate airway access be required, then emergent cricothyrotomy such be undertaken, with potential conversion to a standard tracheotomy if warranted.

In adults with epiglottitis, the airway can be maintained without intubation in most patients (80 to 85%) [8, 12]. Severe airway distress is possible in adults but less common than in children, since adults have larger airways. Humidified oxygen and close monitoring (usually in an intensive care unit) may be sufficient for airway management in many adults. Intravenous corticosteroids are often used in an effort to reduce supraglottic inflammation, although randomized controlled trials have not been performed to assess benefit. For adults with moderate to severe respiratory distress, more active intervention is necessary.

Tracheotomy is not the first choice but should be chosen if expertise in difficult intubations is limited. Endotracheal intubation should be attempted only by an experienced anesthesiologist (or otolaryngologist) and always with readiness for immediate cricothyroidotomy. A failed endotracheal intubation attempt can result in immediate and total airway distress. There is no time to waste in providing an artificial airway in these cases. In cases with severe airway distress, awake tracheotomy with local anesthesia is the most secure and safe way to provide an airway. The transtracheal intubation should be also left to the most experienced airway specialists.

Epiglottic Abscess

In 10–20% of adults with epiglottitis, there is a concomitant abscess in the epiglottis. The abscess is associated with more severe symptoms and airway distress [7]. In these cases, surgical drainage of the abscess is indicated after the airway is secured. A tracheostomy is often indicated and opening of the abscess is carried out under direct laryngoscopy.

Acute Laryngitis

Clinical Presentation

Acute laryngitis is a common inflammatory disorder of the larynx. Typical symptoms include hoarseness, loss of voice, sore throat, and other nonspecific symptoms of upper respiratory tract infection. Fever and tender cervical lymphadenopathy may be present. Nearly all cases are due to upper respiratory viruses and are self-limited. Patients with laryngitis caused by infection may also have fever and swollen lymph nodes. The symptoms last less than 2 weeks.

Epidemiology

Acute laryngitis is the most common disease of the larynx. The true prevalence in the adult population is difficult to determine, because most

patients do not seek medical attention. A study conducted by the Royal College of General Practitioners in the United Kingdom estimated an incidence of nearly seven cases per 100,000 population per week [18].

Pathophysiology

Inflammation of the larynx may involve any area of the larynx, including the supraglottic, glottic, and subglottic areas [19]. Edema of the vocal cords leads to dysphonia.

An upper respiratory tract infection is the most common cause of acute laryngitis and nearly all cases are viral. The viruses involved are presumed to be those that cause other common upper respiratory infections, such as rhinovirus, influenza, parainfluenza, and adenovirus [19]. Rare cases of acute laryngitis are due to bacteria, or are initially viral but develop bacterial superinfection. Bacterial causes include the usual respiratory bacterial pathogens (Group A *Streptococcus*, *S. pneumoniae*, *H. influenzae*, *M. catarrhalis*) [19]. A rare case of MRSA membranous laryngitis has been reported in a child with concurrent influenza [20]. Evidence of a bacterial infection elsewhere (e.g., pneumonia, streptococcal pharyngitis) supports a bacterial etiology.

Noninfectious causes of laryngitis include gastroesophageal reflux, fungal infections of the larynx due to inhaled corticosteroids, and irritation from inhalers used for asthma. In most cases, symptoms develop subacutely rather than acutely, and are not accompanied by other symptoms of upper respiratory infection. In many cases of laryngitis due to noninfectious etiologies, the symptoms are chronic by the time the patient presents to the otolaryngologist.

In patients whose duration of symptoms is greater than three weeks, direct visualization of the larynx should be undertaken with the consideration of a possible malignant etiology. This is especially true in patients with a significant smoking history.

Treatment

Because most acute laryngitis cases are due to viruses, treatment is conservative and includes voice rest. A brief course of inhaled or oral corticosteroids can help reduce vocal cord inflammation, but this treatment should only be used in patients who have an urgent need to use their voice. One study found that inhaled versus oral corticosteroids reduced laryngeal hyperemia and edema to a similar degree [21]. Antibiotics should not be used unless there is evidence of bacterial superinfection. A Cochrane review of the literature found no benefit to the use of antibiotics for acute laryngitis [22].

Croup (Acute Laryngotracheobronchitis)

Croup is a clinical diagnosis referring to symptoms of acute viral laryngotracheobronchitis or laryngotracheitis, the latter description preferred by some authors [23]. The clinical presentation is due to acute laryngeal and subglottic swelling and is characterized by the abrupt onset of a barking cough, typically accompanied by hoarseness, inspiratory stridor, and respiratory distress [24]. The term “spasmodic croup” is sometimes used to refer to afebrile episodes of croup that may be recurrent [25].

Epidemiology

Croup is one of the most common causes of respiratory distress in young children, affecting approximately 5% of children during the second year of life [23]. Children between the ages of 6 months and 3 years old are most often affected, although some cases occur in children as young as 3 months and rare cases occur in adolescents [24]. Boys are affected approximately 1.4 times more often than girls [24].

Croup occurs most often in autumn (September to December) in temperate climates [24], but

cases may occur at any point throughout the year. Parainfluenza virus type 1 is the most common cause of croup and this virus produces epidemics of respiratory illness, including croup, in the fall every other year [25–27]. In North America, odd-numbered years have an increased incidence of croup compared with even-numbered years, as a consequence [24].

Microbiology

In a study of 144 children presenting to an emergency department in Helsinki with respiratory stridor, parainfluenza viruses accounted for over 40% of cases with parainfluenza virus type 1 the most common virus identified [28]. Other major causes of croup include parainfluenza virus types 2 and 3, while less common etiologies include influenza A and B and respiratory syncytial virus [25]. Human metapneumovirus, adenovirus, and coronavirus cause some cases.

Pathophysiology

Croup is a viral infection that leads to edema of the larynx and trachea; the bronchi may also be involved. This is most critically manifested by edema within the cricoid ring, which has a fixed circumference as well as being the narrowest region of the pediatric airway. Significant narrowing in this region can lead to life-threatening airway compromise. The narrowed subglottic region leads to the typical barking cough. The subglottic region of a young child is narrower and more pliable than in older individuals, and the narrowing that occurs with inspiration may be exaggerated in a young child with croup [25].

Clinical Features

Croup usually begins with nonspecific upper respiratory tract symptoms (coryza, nasal congestion), but then 12–48 h later there is the abrupt onset of a

barking cough. The onset of this cough is usually late at night. Stridor, hoarseness, and fever are other features of the infection. Fever may be high (39.4–40 °C, or 103–104 °F), especially in cases due to influenza or parainfluenza virus [25]. Respiratory distress occurs in varying degrees, depending on the severity of the airway obstruction. In mild cases of croup, stridor is absent at rest but may be present when the child is upset or crying. Cases classified as moderate to severe croup are associated with stridor at rest and an increasing degree of chest wall retractions (although retractions may decrease in severe croup with impending airway failure). In severe croup, the child is agitated or lethargic.

Lateral and anteroposterior (AP) plain film x-rays can be obtained in stable patients, while maintaining close observation. The AP film can demonstrate subglottic airway narrowing, the classic “steeple sign” supporting the diagnosis of laryngotracheobronchitis. Similarly, the lateral film can assist in evaluating for findings associated with alternative diagnoses, such as supraglottitis.

Treatment

Most cases of croup are mild, and treatment consists of symptomatic treatment plus a single dose of corticosteroid (0.15–0.6 mg/kg, maximum 10 mg) [24]. A single dose of oral dexamethasone was shown to be beneficial in mild croup in a randomized controlled trial [29]. Humidified air has a long history of use in treating croup, but there is evidence that it is not effective and should not be used [24].

Children with moderate to severe croup require evaluation in an emergency department. Care must be taken to keep the child calm (e.g., the child can sit on the parent’s lap), as agitation can worsen symptoms. Treatment of moderate croup is with a single dose of dexamethasone and oxygen as needed [24]. Very brief courses of corticosteroids are well tolerated and safe in children [30].

Treatment of severe croup includes blow-by oxygen (optional unless cyanosis is present), corticosteroids, and nebulized epinephrine. The ben-

efit of nebulized epinephrine in respiratory distress is rapid but short term. Onset of the effect starts within 10 min and lasts from 1 to 2 h [4]. Retreatment with nebulized epinephrine may be required. Children with severe croup may require admission to a pediatric intensive care unit, and intubation is required in some children (<3%). The diagnosis of bacterial tracheitis should be considered in children with a high fever and a toxic appearance; this diagnosis requires treatment with antibiotics.

Conclusion

Epiglottitis, acute laryngitis, and croup (acute laryngotracheobronchitis) are infections of the upper airway, affecting the epiglottis, larynx, and larynx and trachea, respectively. Epiglottitis is a bacterial infection, while viruses cause nearly all cases of acute laryngitis and croup. Acute laryngitis in adults is usually self-limited. Epiglottitis, which used to be prevalent in children under age 5, is now seen more often in adults than in children. This decline in childhood epiglottitis is due to the *Haemophilus influenzae* type b (Hib) vaccine. Streptococci, including *Streptococcus pneumoniae*, are now important causes of epiglottitis. Croup is a viral infection, usually due to parainfluenza virus, that primarily affects children ages 6 months to 3 years old. Epiglottitis and croup can cause sudden and life-threatening loss of the airway, and misdiagnosis or mismanagement can result in fatalities. With appropriate management, however, death from these infections is very rare.

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