

Acute epiglottitis in children : a review following an atypical case

S. DEPUYDT (*), M. NAUWYNCK (**), M. BOURGEOIS (**), and J.-P. MULIER (**)

INTRODUCTION

Acute epiglottitis (AE) is a short-lived disease that usually presents characteristic signs and symptoms (1). Since the generalized use of *Haemophilus influenzae* type B (Hib) vaccine in children, presentations of this disorder have decreased dramatically (2). Presentations of this, and other acute epiglottic swelling, vary remarkably and may easily be misdiagnosed by physicians who have little or no experience with the acutely obstructed airway. Early suspicion and a proper evaluation is mandatory to prevent a life-threatening crisis. Anesthesiologists should be aware that this disease will still be seen and not to think of it as a totally extinct entity. We report a case of atypical AE in a child.

CASE REPORT

A 5 year old male child was admitted during the night to the emergency ward of a general hospital. The evening before the admission, the child presented "barking cough", acute difficulty of swallowing and high fever (39°C). The symptoms have developed over the night, in some hours. At admission, the child was lethargic, sitting forward, with neck extended and mouth open, presenting paradoxal ventilatory movements, with sternal retraction.

Oxygen was administered by mask. Therapeutic approach with aerosolized racemic epinephrine and inhalational corticosteroids (Pulmicort™) didn't change the symptomatology. A 22G catheter was placed in a peripheral vein. A first arterial blood gas (ABG) sample (under oxygen therapy by mask) showed a partial pressure of oxygen (paO₂) 102 mmHg, a partial pressure of carbon dioxide (pCO₂) 49 mmHg and bicarbonate 5,9.

A diagnosis problem was risen to differentiate between laryngotracheobronchitis and AE.

A chest X-Ray showed a right lower lobe infiltrate. On the lateral X-ray of the neck the typically thumb print sign was observed, suggesting

AE. The child was admitted to the ICU. Deterioration of consciousness occurred, from lethargy to restlessness and with paO₂ : 83 mmHg, pCO₂ : 63 mmHg and pH : 7,19. Orotracheal intubation was decided. Anesthesia for intubation was achieved with propofol (2 mg/kg) and fentanyl (25 mcg), while the patient was ventilated with an FiO₂ of 1.0. A 4 mm endotracheal tube (ETT) without tracheal cuff (Mallinckrodt™, Mallinckrodt Laboratories, Athlone, Ireland) was used. During laryngoscopy, the epiglottis was described as edematous, cherry-red colored, but easy to pass through. The child was mechanically ventilated (Siemens Servo 900 C, Siemens® Elema™, Solna Sweden). The baseline ventilatory settings used were : minute volume of 4 liters, respiratory rate 18/min., with an external PEEP of 5 cmH₂O. No audible airway leak was noted. The child was sedated for ventilation with continuous infusion of midazolam, fentanyl and propofol. A radial artery catheter was placed for easier blood gas sampling.

The FiO₂ was rapidly (24 hours) lowered to 0.25 and the PEEP to 3 cm H₂O, however it was not possible to discontinue mechanical ventilation. On the chest X-ray the lung-image was normal. Still no leaks around the tracheal tube were noted, when the peak inspiratory pressures were about 20 cmH₂O.

Intravenous antibiotics (Ceftriaxome) were started ; the hemoculture never positivized. Intravenous corticosteroids were also administered from admission, but stopped on day 3 after admission. At day 3, there was an audible leak around the tracheal tube and a respiratory weaning was initiated, followed by successful removal of the ET. At this stage, arterial blood gases were normal.

S. DEPUYDT, M.D. ; M. NAUWYNCK, M.D. ; M. BOURGEOIS, M.D. ; J.-P. MULIER, M.D.

(*) Departments of Anesthesiology, Gent University Hospital.

(**) Department of Anesthesiology and Intensive Care, Brugge General Hospital.

Address correspondence : Dr. Sarah Depuydt, Department of Anesthesiology, Gent University Hospital, 185 De Pintelaan, 9000 Gent, Belgium.

Initially, the extubation was followed by stridor, with progressive amelioration. Light sore throat was still present. The day after, the patient was released from the ICU in optimal conditions (spontaneous respiration, without stridor, with the following ABGs : paO_2 : 112 mmHg, pCO_2 : 37 mmHg and pH 7.41, with 3 liters/minute oxygen administered by face mask).

DISCUSSION

AE continues to cause life-threatening airway obstruction in children. AE is an inflammation of the epiglottis, secondary to an infectious process. It can involve other supraglottic structures such as the arytenoid, false cords, and posterior tongue, resulting in airway obstruction (2, 3, 4). This is the reason some prefer the designation of this disease as supraglottitis rather than epiglottitis (1). The causative agent is usually *Haemophilus influenzae* type B (Hib), but *Streptococcus pneumoniae* has also been infrequently associated with AE (5, 6). Since the description of acute epiglottitis in children in 1941 by SINCLAIR (5), a major advance in the treatment of the disease was the introduction of the Hib vaccine. Although vaccination failures have been reported (there is evidence that 27% of AE in children in the USA are due to vaccine failure) (7), the introduction of new vaccines has had a major impact in virtually wiping out the infection. Vaccination programs have been introduced in several countries over the world (7, 8). A decrease from 3.47 per 100,000 in 1980 to 0.63 per 100,000 in 1990 has been ascribed to the routine use of Hib vaccination (9).

The incidence of the AE is highest in the 2- to 6-year age group, but cases have been reported from infancy to adulthood (1, 4). Epiglottitis occurs throughout the year, with the highest incidence during October and December (7), but seasonal attacks of the disease have been reported in the winter months (10), during the spring and summer (11) or in the late spring and fall (12). AE contributes to approximately 7-8 per cent of the pediatric infectious respiratory obstructions admitted to hospitals (13). The onset is sudden, only a few hours before signs of severe obstruction occurs. The children present with a history of acute difficulty in swallowing, as well as high fever and inspiratory stridor. There may be also excessive drooling, a muffled voice, and the characteristic position of sitting upright and leaning forward. This change in posture may cause more airway obstruction.

AE is a clinical diagnosis and a true emergency, radiographs are not necessary to confirm it (4, 14). However, with other diagnostic considerations, such as laryngotracheobronchitis, foreign body in the upper airway, retropharyngeal abscess or congenital anomalies, the child may be escorted to the X-rays. The time taken for the X-rays should obviously be avoided in the critically ill child (because of the possibility of sudden complete airway obstruction). A lateral radiography of the neck may demonstrate the classic thumb sign, which is an aptly named shape seen with epiglottic enlargement (14). Recently, magnetic resonance imaging has demonstrated the standard measurements for the hypopharyngeal space in various age groups, and it has correlated this measurement with the change produced by epiglottic inflammation (15).

Laboratory findings are non-specific, although a high white count with neutrophilia may be present.

The definitive diagnosis of AE is made during direct laryngoscopy. However, in the doubt of diagnosis and without life threatening obstruction, a fiberoptic pharyngoscopy performed gently by a skilled examiner can immediately confirm or eliminate the diagnosis (16).

Some differential diagnosis problems, as in the presented case, should be considered (Table 1). Laryngotracheobronchitis (croup) is a viral infection with more common incidence (accounts for more than 80% of children with stridor, while AE for less than 5%), present in the age group under 3 years, with a gradual onset, with signs of upper airway infection (rhinorrhea) and low grade fever. In contrast to AE, the progression is slower (days); the patient can lay down; dysphagia and drooling are absent, and there is a characteristic "barking cough" and a hoarse voice. The presence of the "barking cough" in our case has risen the differential diagnosis problem. Normally the evolution is not severe, and the administration of oxygen and aerosolized racemic epinephrine and corticosteroids may ameliorate the symptoms.

With the presence of a radiographic right lower lobe infiltrate, we also considered the possibility of foreign body aspiration or the presence of a pulmonary infection. However, there was no evidence of pulmonary shunting (paO_2 was normal) and in case of recent foreign body aspiration, fever is not present.

The child with suspected AE must be admitted to the hospital. Oxygen is administered by mask continuously from the time diagnosis is suspected. Because of the potential for sudden complete

Table 1

Differentiation between epiglottitis and laryngotracheobronchitis

	Epiglottitis	Laryngotracheobronchitis
Incidence	Accounts for 5% of children with stridor	Accounts for more than 80% of children with stridor
Site of obstruction	Supraglottic	Subglottic
Etiology	Bacterial	Viral
Age	2-6 years	< 2 years
Onset	Sudden (hours)	Gradual
Progression	Rapid	Slow, variable
Position	Insists on sitting up and leaning forward	Can lie down
Dysphagia	Severe	Absent
Drooling	Present	Absent
Cough	Suppressed	Barking
Voice	Muffled	Hoarse
Fever	Often > 39°C	Rarely > 39° C
Respiratory pattern	Tachypnea	Rapid, struggling

obstruction, an artificial airway should be inserted in all patients with AE (4, 17, 18). During years, there has been much debate as to whether one should perform tracheotomy or secure the airway by means of long-term intubation (19). However, a growing number of reports support the view that long-term intubation is superior to tracheotomy (20). An attempt to visualize the epiglottis should not be undertaken until the child is in the operating room (best) and preparations are completed for intubation of the trachea and for possible emergency tracheostomy. Equipment for endotracheal intubation, emergency tracheostomy and resuscitation should be kept with the patient. Blood sampling is best deferred until the child is adequately anesthetized in the operating room (21). The definitive treatment of epiglottitis includes the use of appropriate antibiotics and surely a secured airway, until inflammation of the epiglottis has subsided. An ETT that will pass through the glottis without excessive pressure should be selected. A tube 0.5 to 1 mm smaller in diameter than usual is used, and may still be a snug fit; attention should be given also to the length of the tube, as the smaller tubes commonly needed may be too short for older children. One must note that the ETT will be resting in an already inflamed portion of the trachea and that the tube's presence may initially increase the edema. There are different opinions in how to induce anesthesia in a child with AE, however, when anesthetizing a patient with an AE for intubation, experience is crucial. Some authors (1, 21, 22) propose induction and maintenance of anesthesia with a volatile agent in oxygen, with the child in sitting position. After the onset of drowsiness, the child is placed in supine position and ventilation of the lungs is assisted as necessary. When an adequate depth of anesthesia is achieved, direct

laryngoscopy is performed and a tube is placed in the trachea. After successful intubation of the trachea, a thorough direct laryngoscopy is performed to confirm the diagnosis of AE. A nasotracheal tube is preferred as it is easier to secure. Other authors (23) propose the mandatory insertion of an intravenous line before induction of anesthesia, which can be done this way; eventually administration of atropine (6 to 10 mcg/kg) or glycopyrrolate (3 to 5 mcg/kg) may be useful. Administration of muscle relaxants for intubation is not indicated. Some authors suggest awake intubation with topical anesthesia (1). In such cases there is a possibility of adenoidal bleeding, difficult visualization, agitation, trauma or compromised ventilation.

CRYSDALE and SENDI (24) proposed that children six years of age or older may be considered for observation, but this decision is to be made by the otolaryngologist in consultation with the intensivist only after careful clinical assessment. Such children should be orientated, have good air entry on chest auscultation, and tachycardia should be mild and in keeping with the degree of pyrexia. Dysphagia may be present but drooling must be absent. An intravenous is established, appropriate parenteral antibiotics are administered and the child receives constant observation care. The clinical condition of the child should be improved within two or three hours of inception of this treatment. If there is any doubt about the clinical status of the patient, one should proceed to intubation.

As some patients with AE may have atypical features, they may be thought to have severe acute laryngotracheobronchitis. There are such cases reported (25) where treatment with racemic epinephrine was followed by rapid deterioration. While this deterioration may be an odd reaction to the racemic epinephrine, it is more likely due to the

head and neck manipulation required during this procedure. The administration of any nebulized drug to a child who is already frightened will cause further agitation and hence, further compromise of an already narrowed airway.

After anesthesia and intubation, the child should be kept in the ICU. The child may be allowed to awaken from the anesthetic or, if necessary, sedated and mechanically ventilated like in the case presented above. Usually, intubation of the trachea is required for 48 to 96 hours, although following a report (26), 8 to 12 hours may be sufficient. Initial antibiotherapy should be of broad-spectrum in nature (cephalosporines like ceftriaxone and cefotaxime or ampicillin, eventually associated with vancomycin if resistant gram-positive organisms are a concern). When hemoculture results are available, the choice of antibiotics must be adjusted. Hib is an invasive organism and infection may include pneumonia, septic arthritis or meningitis (1).

As a complication of upper airway relief, pulmonary edema is well-described (27, 28), occurring in about 7% of cases. The mechanism of this edema is complex and not yet fully understood. In addition to hypoxia, profound hemodynamic changes occur during the inspiratory phase of the obstruction; highly negative transpulmonary pressure may lead to an increase in pulmonary blood volume and biventricular dysfunction, and possibly disruption of integrity of the pulmonary endothelium. These hemodynamic changes appear to be counterbalanced by the positive pleural and alveolar pressures and decreased venous return during the expiratory component of the obstruction. Nevertheless, when an artificial airway is inserted, this compensation is disrupted abruptly, resulting in an increase in systemic venous return and thus pulmonary edema. Although this type of edema usually is observed in cases of severe obstruction, it may go unrecognized or misdiagnosed (28).

Extubation may be considered if the patient's body temperature is no longer elevated and if there is a fall in neutrophil count. An important clinical sign of resolution is the presence of some airway leakage around the ETT. Regardless of the clinical and biological evolution, best extubation is done in the OR (1, 4, 21), after direct laryngoscopic visualization of the epiglottis (under general anesthesia) for confirmation that the inflammation of the epiglottis and other supraglottic tissues is resolved.

In conclusion, it should be kept in mind that AE as often life-threatening emergency, and its diagnosis is not always easy. Even with the vac-

nation programs, introduced in the early '90s, AE is still present, and anesthesiologists should be aware that they may be still confronted with it.

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