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Upper airway obstruction in children

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Introduction

Upper airway obstruction in children is one of the most challenging acute emergencies, which requires urgent management in order to prevent fatal outcome. Severe upper airway obstruction accounts for 3.3% of all admission to paediatric intensive care unit (PICU).[1] There are many causes of upper airway obstruction that can be further classified as acute or chronic causes (Table 1).

In this review, the characters of children who are admitted to PICU with severe upper airway obstruction will be discussed further. Difficulties of intubation, indications and complications of tracheotomy will be discussed afterwards. Angioedema, bacterial tracheitis, acute epiglottitis and croup will be discussed with details separately. Laryngomalacia will also be discussed because it is one of the most common causes of stridor in infant. Patients with other anatomical abnormalities of the upper airway will not be discussed in this review, as most of them are under cared by the surgeons rather than paediatricians.

PICU admission with severe upper airway obstruction

There is marked heterogeneity in the causes of upper airway obstruction that requires PICU admission. According to studies conducted in Malaysia[1] and London,[2] congenital causes account for 6-23%, which include laryngomalacia, vascular ring, subglottic haemangioma, laryngeal cyst and web of pharynx. Acquired causes account for 77-94%, which include infection and anatomical problems. Viral croup is the most common diagnosis and it accounts for about 30% to 50% of all PICU admissions. Acute epiglottitis is extremely rare in Asia and also not common in Western countries. Bacterial tracheitis and subglottic stenosis are the most likely diagnosis requiring ventilation, as these patients are usually more sick and ill. Anatomical causes include tracheal compression, subglottic granuloma, subglottic stenosis and foreign body are other causes of PICU admission.

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Table 1. Acute and chronic causes of upper airway obstruction

Cause of upper airway obstruction	
• **Acute**	• **Chronic**
• Acute laryngotracheitis	• Laryngeal: Laryngomalacia, Subglottic stenosis/haemangioma, Vocal cord paralysis, Laryngeal web, Cyst (posterior tongue, aryepiglottic, subglottic, larygoceles, laryngeal cleft), laryngeal papillomas
• Acute epiglottitis	
• Suppurative tracheitis	
• Laryngeal foreign body	
• Diphtheria	
• Acute Angioneurotic oedema	
• Retropharyngeal abscess	• Trachea: Vascular ring, Tracheal stenosis, trachemalacia

Difficulty in intubation will be encountered in 43%.[1] However, tracheostomy is not common among children who required PICU admission. Regarding the prognosis, non-survivors have a higher Paediatric Risk of Mortality (PRISM) II score although the outcome is generally favorable. The conditions are similar in Hong Kong according to our experience in a teaching hospital (unpublished data).

Intubation

Difficulty in intubation will be encountered in half of the cases during intubations of severe upper airway obstruction patients. There are some characteristics of patients that we should pay special attention during intubation in order to facilitate the procedure. Delay in intubation in those critical ill patients could have very serious complication and even fatal.

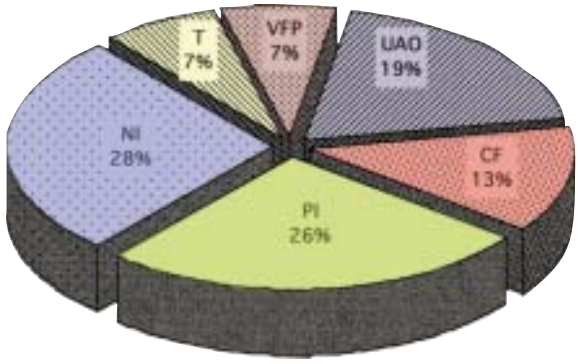
Pierre-Robin syndrome, Treacher-Collins syndrome, Goldenhar syndrome and mucopolysaccharidosis (Hurler, Hunter, Maroteaux-Lamy) are commonly associated with significant cranio-facial abnormalities, and these patients may have problems during intubation. They have micrognathia, relative macroglossia, hypoplasia of the facial bone, macrostomia and even short immobile neck. Mucopolysaccharidosis has excessive intra-lysosomal accumulation of glycosaminoglycans that causes generalised thickening of soft tissues. All these features make the procedure of intubation more difficult.

Inhalation injury makes visualisation of the normal airway anatomy more difficult. Epiglottitis is another

potential life-threatening cause for difficult intubation. Trauma can distort the normal anatomy of the upper airway and make the intubation more complicated. For these patients, an anesthetic approach should be adopted. Atropine pre-medication should be administered to dry up secretions, and oxygen should be given. It is better to use ketamine for sedation for these patients. Muscle relaxants should be withheld until the airway is secured. Intubation should be performed under deep inhalational anaesthesia. Surgical airway should be performed rapidly if the above methods fail.

Tracheotomy

During 1970s, infection such as laryngo-tracheobronchitis and epiglottitis were the common causes for tracheotomy in children. With the popular use of endotracheal intubation, fewer tracheotomies and decannulations were performed. Figures 1 and 2 show the common indications for tracheotomy and the associated decannulation rate. According to Carron et al, the overall decannulation rate is 41%. The overall complication of tracheotomy is 44%, with granuloma formation being the most common.[3] Table 2 shows the common complications associated with tracheostomy. In the same study, the overall mortality rate is 19% (Figure 3), with the vast majority of deaths are due to the child's primary illness. Only 3.6% was known to be directly due to tracheotomy. Most of them have plugging of the tracheotomy tube with resultant respiratory arrests occurring between 0.3 and 30.8 months after the tracheotomy. Misplacement of the tube after operation has been reported.



Primary indications for tracheotomy by grouping. PI=prolonged intubation; NI=neurologically impaired; T=trauma; VFP=vocal fold paralysis; UAO=upper airway obstruction; CF=craniofacial abnormality.

Figure 1. Primary indications for tracheotomy (adapted from reference [3]).

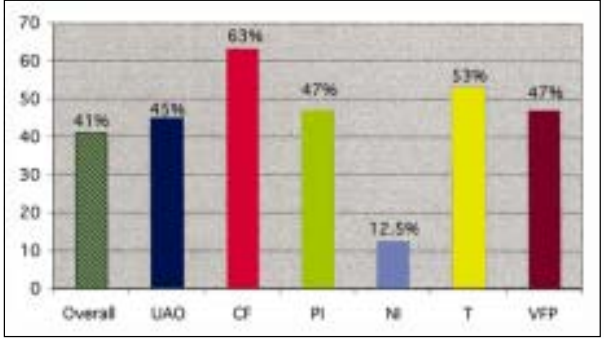


Figure 2. Decannulations rates by group (adapted from reference [3]).

Angioedema

Angioedema is an anatomically limited non-pitting edema that may result in life-threatening airway obstruction.[4] Facial swelling is present in 80% of cases. Other common symptoms include tenderness, dyspnea, dysphagia or hoarseness. Food allergy accounts for about 40% of the causes, and insect bites, infection and drugs are the other common causes. These patients seldom require PICU admission, as the symptoms of these patients usually resolve soon after pharmacological treatment with adrenaline, antihistamine or steroid.

Bacterial tracheitis

Bacterial tracheitis is not a common disease but can be very serious. The clinical presentation is similar to that of severe viral croup, epiglottitis, or foreign-body aspiration. It can range from mild stridor to even

Table 2. Complications of tracheotomy

- Stomal granuloma 20%
- Tracheo-cutaneous fistula 13%
- Tracheal stenosis 2%
- Tube plugging with respiratory arrest 2%
- Accidental decannulation 1.5%
- False passage creation 1%
- Stomal keloid formation 1%

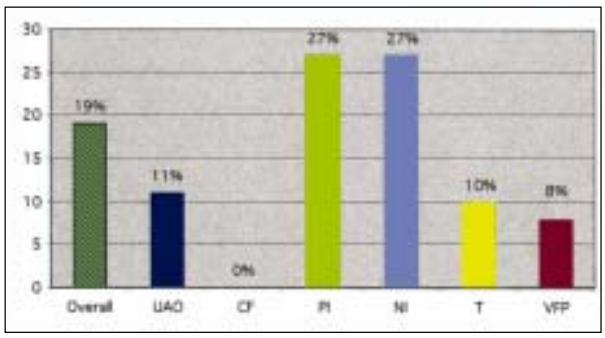


Figure 3. Mortality rates by group (adapted from reference [3]).

cardio-respiratory arrest. The clinical course is generally less acute than that of epiglottitis and also with prodromal symptoms. Patients exhibit high fever, appear more ill, and are less responsive to nebulised epinephrine and other supportive measures than children with viral croup. Drooling and neck hyperextension are uncommon in bacterial tracheitis. Tracheal secretions are copious, thick, and tenacious which persist for 3 to 5 days and even up to 3 weeks. Once these patients are admitted to PICU, 57% of them will be intubated.[5] Intubated patients are usually younger than the non-intubated patients.

The most common pathogens in bacterial tracheitis are *Staphylococcus aureus* and *Haemophilus influenzae*. However, there are reports of bacterial tracheitis caused by *Streptococcus pyogenes*, *Streptococcus pneumoniae*, *Streptococcus viridans*, *Pseudomonas aeruginosa*, *Escherichia coli*, *Moraxella catarrhalis*, non-group A streptococci, and Neisseria spp.

Patients may be extubated safely when their body temperature returns to normal, detection of air leaking around the nasotracheal tube, and when the amount of secretions markedly decreases.

Acute epiglottitis

Paediatric epiglottitis is a serious, potentially life-threatening condition although it is extremely rare in Asia. After Haemophilis influenza (Hib) vaccination in Western countries, the annual incidence of children below 5 years of age decreases from 20.9 per 100,000 in 1987 to 0.9 per 100,000 in 1996.[6] In adults a tendency toward a decrease in incidence is also evidence initially but then rises from a low in 1998 to reach pre-vaccine levels in 2003.[7] Immunisation programme in children may have unexpected effects on the epidemiology of disease.

The presentations of the patients remained static but a decrease in Hib cases (especially the number of ampicillin-resistant organism) are seen. Affected children are, on average, much older after the implementation of universal vaccination programme. Vaccine failures still occur but are rare. Acute epiglottitis should still need to be considered when a child presents with severe upper airway obstruction even after Hib vaccination.

Laryngomalacia

Congenital stridor was first reported in 1853 by two French physicians, Rilliet and Barthez.[8] Sutherland and Lack published the first review of this condition in 1897.[9] In 1942, Jackson first used the term laryngomalacia, coming from the Greek malakia with the meaning of morbid softening of part of an organ. Jackson clearly defined it as softness, flabbiness, or loss of consistency of the laryngeal tissues.[10]

In affected patients, inspiratory stridor is usually present since birth. In some cases, stridor may become apparent few weeks or even months later. Stridor may be exacerbated in some cases by an upper respiratory infection. It is often intermittent and is aggravated when the child is active and crying. On the other hand, symptoms may also be precipitated by supination and head flexion and are relieved by pronation and head extension.

For most of the cases, the symptoms will disappear with time. In 10% of cases, however, upper airway obstruction is so severe that patients develop apnoea or failure to thrive.[11] In these situations, surgery or tracheotomy may be needed.

Croup

Acute respiratory illness caused by inflammation and narrowing of the subglottic region of the larynx is defined as croup.[12] Barking cough, hoarseness, stridor and respiratory distress are the usual presentations. Croup is usually self-limiting and symptoms usually last for 3-7 days. Yet, it still can cause severe upper airway obstructions that result in intubation or even mortality. For those patients with very severe croup, other causes of upper airway obstructions, especially epiglottitis, should be ruled out. If the diagnosis is not certain, patients must be treated empirically as epiglottitis, and intubation should be performed by experienced anaesthetists under deep inhalation induction. Surgical airway should be performed rapidly if the above methods fail. Neck radiograph is usually not necessary for the diagnosis but if taken, it would show the steeple sign (narrowing of the subglottic area) in patients with croup.

The incidence of croup is 1.5-6 per 100 children per year. The admission rate for croup varies from 1.5% to 30%. This wide range of admission rate reflects different criteria for admission. In the United States, croup has been estimated to cause 41,000 hospitalisations annually.[13] The intubation rate is about 0.5-1.5% for those patients admitted to hospital. The re-intubation rate is 5-16%.

Parainfluenza virus type 1 is the main virus causing croup.[14] The parainfluenza virus 1, 2 and 3 can be isolated in all age groups, and altogether account for about 2/3 of all cases. For patients younger than 5 years old, respiratory syncytial virus (RSV) tends to

be isolated more commonly. On the other hand, influenza virus and *Mycoplasma pneumoniae* affect children older than 6 years of age.[15] Moreover, influenza virus tends to cause more serious illness as compared with parainfluenza virus.[16]

Mortality rate for croup is less than 0.5% in intubated patients.[17] According to Sacenkova et al, 92.5% of these cases die of severe pneumonia, 7.5% die of sepsis; 70% of them are 0-2 year old, 75% are boy and nearly all patients had aggravated pre-morbid background.[18] In this study, virus and resistant strains of staphylococci and streptococci (penicillin, ampicillin and cefazoline) could be isolated in the sputum or organ tissues from most of the death cases. The authors concluded that children with croup died of severe pneumonia complications with a low systemic reactivity and high antibacterial resistance.

Treatments of croup include glucocorticoids, nebulised epinephrine, humidification and heliox. Since late 1980s, glucocorticoids have been recognised to provide some clinical benefit for children with croup. Kairys et al published a meta-analysis of clinical trials examining the benefit of glucocorticoids in 1989.[19] The Cochrane Database of Systemic Reviews in 2004 showed that glucocorticoid treatment is effective in improving symptoms of croup in children as early as six hours and for up to at least 12 hours after treatment, and the efficacy was supported by improvement in croup score, decrease in return visit or admission, reduction in length of stay and epinephrine use as an additional intervention.[20] With the introduction of steroid, less croup patients need intubation and intensive care,[21] and the duration of intubation and need for re-intubation also decreased.[22]

Regarding the administration for glucocorticoid, oral route is preferred as this is easily given and the efficacy for oral route is the same as with intramuscular injection. Some children find nebulised therapy distressing. The standard dose for glucocorticoid is dexamethasone at 0.6 mg per kilogram body weight per dose, with a maximum of 10 mg although some authors suggest that lower dose is also effective.

Epinephrine (adrenaline) is a potent agonist of alpha- and beta-adrenergic receptors. Since early 1970s, epinephrine has been used to treat croup. It helps to reduce bronchial and tracheal secretions and oedema. Following nebulised therapy, these effects are noted within 10-30 min and last for about one hour. However, 35% of patients who received epinephrine had a relapse of their symptoms within 2 hours of treatment.[23] Racemic epinephrine, a mixture of equal amounts of dextro(d) and levo(l)-isomer, was traditionally used because this was believed to have fewer side effects and better effectiveness. However, with more published data, racemic epinephrine and levo-epinephrine were found to be the same for their effectiveness and side effect profile.[24] In fact, levo-epinephrine is cheaper and more easily available. Therefore, levo-epinephrine is recommended for treatment of croup instead of racemic epinephrine.

Humidification was a routine therapy in early 1990s. It was believed that this therapy relieved discomfort and prevented the drying of inflamed laryngeal mucosa. However, there is no evidence for these effects. During the therapy, children need to stay inside mist tents. They can become wet and cold. These patients may be quite irritated during isolation. It was also difficult to observe the general conditions of these children in mist tents. With the introduction of steroid therapy, humidification therapy is no longer use as a standard therapy for croup.

Heliox is an experimental treatment. It is a metabolically inert, non-toxic gas that combines helium with oxygen. It has low viscosity and low specific gravity, and these properties allow for greater laminar airflow through the respiratory tract. Heliox is well tolerated, and some studies suggested that it decreased the croup score.

In summary, glucocorticoid reduces croup score, rate and duration of intubation. However, there is no conclusion on the best route and dosage of its administration. Nebulised epinephrine can relieve the symptoms of croup transiently but symptoms will recur following transient improvement. Humidification is no longer a routine treatment for croup. Heliox is still an experimental treatment, and more evidence is needed to support its routine clinical use.

Summary

Severe upper airway obstruction in children requires urgent management. Rapid and effective management can prevent severe complications and even fatality. Tracheotomy is rarely needed. Infections such as croup remain the most common cause of severe upper airway obstruction that requires intensive care management. Acute epiglottitis and bacterial tracheitis are rare, but should be the differential diagnoses in critically ill patients. Patients with angioedema are rarely admitted to PICU, and most of them resolve soon after medical treatment. Laryngomalacia is common but most of them will be resolved without treatment. Steroid is very effective for croup, and much less patients with croup require intubation following the use of steroid treatment.

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