



Difficult Airway Management of Neonates, Infants, and Children with Syndromes Involving the Airway

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What Does a Pediatric Difficult Airway Entail?

The difficult pediatric airway is a challenge even for the experienced pediatric anesthesiologist. Children have a higher rate of oxygen consumption than adults leading to a shorter apnea time [1]. There is no standard definition of a difficult airway; however, there are key characteristics of difficult airways found across all practice guidelines [2]. A difficult airway is the situation in which an anesthesiologist has difficulty with face mask ventilation, tracheal intubation, or both. With recent advances in airway technology, descriptions of difficult airways are more detailed, including whether a patient will be difficult to ventilate with a supraglottic airway (SGA), difficult laryngoscopy, difficult tracheal intubation, or difficult front of neck access (FONA). The focus of this chapter will be on anticipated or known difficult airways. Some of the principles

discussed may apply to unanticipated difficult or emergency airway management.

Respiratory critical events remain a significant cause of morbidity and mortality in children. A recent study from 261 participating centers across 33 European countries reported an incidence of respiratory critical events of 3.1% (2.9–3.3) in 30,874 children undergoing anesthesia with a mean age of 6.35 years [3]. Airway management in children with difficult airways is associated with an even higher incidence of complications and respiratory critical events. The Pediatric Difficult Intubation (PeDI) multicenter registry is a prospectively collected dataset of difficult tracheal intubation in children from institutions all over the world. A recent analysis of the PeDI registry data showed that out of 1018 children with difficult airways, 204 (20%) had at least one complication, the most severe being cardiac arrest which occurred in 15 (2%) children. The most common overall complication of hypoxemia (oxygen saturation <85%) occurred in 94 (9%) children [4]. Risk factors for complications included more than two tracheal intubation attempts, weight less than 10 kg, a short thyromental distance, and three direct laryngoscopy attempts prior to an indirect technique. One of the key take-home messages from these data is that intubation attempts should be considered critical events and clinicians should do their best to minimize laryngoscopies in this population. Furthermore, indirect laryngoscopy should be

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considered early in the airway management plan. Additionally, infants are a particularly vulnerable group as they had almost three times the complications of older children. Valois-Gomez et al. looked at risk factors for difficult bag-mask ventilation (DBMV) in children ages 0–8 years old. They found that there is an inverse relationship with age and DBMV [5].

Risk factors for difficult intubation in children include congenital craniofacial pathology, cervical spine instability, neck immobility, presence of a mass compressing the neck micrognathia, macroglossia, facial asymmetry (especially abnormalities of the ears), limited TMJ mobility, and severe subglottic or tracheal stenosis.

Despite advances in difficult airway management, failure to predict difficult oxygenation or ventilation remains a major cause of failed airway management [6]. The National Audit Project of The Royal College of Anaesthetists and the Difficult Airway Society (DAS) in the United Kingdom looked at major complications in airway management and found that human factors contributed to a majority of serious adverse outcomes [7]. Human factors included deficits in communication, judgment, planning, and fixation errors, such as perseverance with intubation attempts despite patient desaturation, perseverance with the same intubation technique despite failure of that technique, and perseverance of intubation despite multiple failed attempts [8]. Although rarely discussed, cognitive biases can play a significant role in outcomes of difficult airway management. These biases include anchoring, loss aversion, overconfidence, and framing effects. The first step to countering these biases is to understand when they are occurring. Loss aversion is the tendency to view a loss as having more impact than an equivalent gain. An example of this would be a clinician who fails to perform an awake fiber-optic intubation because of concerns of appearing incompetent if they fail. The loss of reputation is seen as being worse than the gain of a successful intubation. Anchoring occurs when one is fixated on an initial condition. If a patient becomes difficult to ventilate, the clinician may persevere because they are anchored to the prior condition of easy ventilation and

believe the airway obstruction will resolve without needing a surgical airway since it was easy before. Overconfidence occurs when a clinician has an inaccurately high self-assessment – this could lead them to proceed to secure a tenuous airway under general anesthesia rather than with a sedated or awake fiber-optic intubation. Framing a clinical situation changes the perception of the situation without altering the facts. For example, a difficult airway patient in extremis can be thought of as needing a life-saving tracheostomy, in contrast to the situation being thought of as being bad and dire and likely to result in a lawsuit if surgical access is performed. The first frame encourages the right action, while the second does not [9].

Syndromes That Involve the Pediatric Airway

Although specific syndromes have been associated with a difficult airway, the common characteristics in these patients are anatomically based. Conditions that affect the oropharyngeal space, the anterior mandibular space, the maxilla, the temporomandibular joint, and the vertebral column are associated with difficult intubation.

Oropharyngeal Space Patients with tumors of the airway and lymphatic malformations may have very little pharyngeal space making intubation challenging. Beckwith-Wiedemann syndrome is associated with an enlarged tongue which rarely presents challenges during intubation. The laryngoscopist may need an assistant to retract the tongue to facilitate tracheal intubation. Ventilation may be difficult and should be considered during the airway management plan. Children with mucopolysaccharide disorders such as Hunter and Hurler syndromes have poor pharyngeal and laryngeal tissue compliance making these patients some of the most difficult to intubate. Patients who have had radiation treatment to the head and neck may also present with poorly compliant airway structures and can be extremely difficult to intubate.

Anterior Mandibular Space Pierre Robin sequence, Goldenhar syndrome, trisomy 13, trisomy 18, and cri-du-chat syndrome all present with micrognathia which limits the anterior mandibular space available to displace the tongue.

Temporomandibular Joint Temporomandibular joint ankylosis may severely limit mouth opening making intubation difficult.

Maxilla/Midface Apert syndrome, Crouzon syndrome, and Pfeiffer syndrome all have midface hypoplasia. Many of these disorders are associated with small nasal passages and a high arched palate making them prone to upper airway obstruction. Patients with Treacher Collins syndrome have both maxillary and mandibular hypoplasia and are prone to severe upper airway obstruction.

Vertebral Column Patients with Goldenhar syndrome may have fused *vertebrae* or hemivertebrae, limiting their cervical neck range of motion. Klippel-Feil syndrome is associated with severe restriction in neck movement due to fusion of cervical vertebrae and atlanto-occipital abnormalities. VACTERL association includes vertebral anomalies that may make tracheal intubation difficult [10].

Best Location for the Management of These Children

Airway management of children with difficult airways outside the operating room (OR) is associated with worse outcomes. Emergency intubation in children by pediatric anesthesiologists in the emergency department (ED) or inpatient units are associated with a significantly higher rate of difficult intubation and intubation-related adverse events, compared to those intubated in the OR [11, 12]. In general, children in extremis should be managed at their bedside location, though moving the child may be appropriate if the necessary equipment or personnel support are not available at bedside. This decision should not be made lightly as many of these patients often have

comorbidities that are unrelated to the airway that need special consideration. If transport to the OR is deemed necessary for airway management, a member of the child's primary care team should accompany the airway managing team to the OR. The child's primary team can be invaluable in managing non-airway-related medical issues. Children who are stable should be managed wherever the care team feels is safest. Clinicians should be mindful that the most important goal in airway management is to maintain adequate oxygenation and ventilation.

Team Approaches

As with any high-risk situation, teamwork is imperative in pediatric difficult airway management. Schmutz et al. found through a systematic review that training teams in process behaviors were associated with an improvement in clinical performance [13]. Qualities of high-functioning teams include clear closed-loop communication, understanding of roles, mutual trust among members, a culture of no blame, collaboration, and having a shared goal.

For anticipated difficult airways, it is important to have a preinduction time-out to introduce team members, discuss the plan and potential challenges, and identify key roles. Each member needs to trust and respect the other members. We have found that an a priori plan of who will make the first, second, and third tracheal intubation attempts is important for successful airway management with the least number of attempts.

Another important aspect of teamwork is that all team members must feel empowered to speak up when they see something they disagree with and not be afraid of the authority gradient or decision-making power hierarchy. Protocols such as Co-PILOT (Co, confirm failure; P, propose other equipment; I, immediate senior anesthesiologist assistance to be called; L, laryngeal mask airway – second generation; O, oxygenate; and T, tracheal access) have been developed to help groups improve teamwork. These protocols include the participation of an anesthesia assistant and employ decision-making tools such

PACE (Probe, Alert, Challenge, Emergency) that involves increasing levels of assertiveness for team members as the urgency of a situation increases [14].

It is important to note that when a patient enters the can't intubate, can't oxygenate (CICO) emergency scenario, front of neck access (FONA) should be attempted. In this chapter, FONA refers to all cannula and scalpel emergency airway rescue techniques via the cricothyroid membrane or anterior tracheal wall. Preparation for CICO is crucial since it is rarely performed by anesthesiologists. Outcomes are very poor in children, and therefore a surgical tracheostomy by an ear, nose, and throat (ENT) surgeon is preferred. In the setting that an ENT surgeon is not immediately available, the clinician can perform a cannula or scalpel FONA, although there is not enough literature to support one as the first-line technique over the other. The Association of Paediatric Anaesthetists of Great Britain and Ireland (APAGBI) recommends percutaneous cannula cricothyroidotomy as the first FONA to be attempted in children 1–8 years old as it can be performed quickly and is relatively noninvasive [15]. Holm-Knudsen showed that scalpel FONA was more successful than cannula FONA using piglets as a pediatric model; however it was associated with significant rates of posterior tracheal wall damage [16]. With current evidence, it is likely more important that an institution chooses one technique and provides adequate training and familiarity with that technique and equipment than which technique is chosen. Once FONA has been obtained, adequate oxygenation can be provided using an Enk Flow Regulator (Cook Medical, Bloomington, IN, USA) [17]. The Ventrain (Ventinova Medical B.V., Eindhoven, the Netherlands) is a new device designed to ventilate through a small lumen tube in patients with difficult airways. It is unique in that it uses suction in the expiratory phase, thereby making expiration an active rather than a passive process as it is with other devices. This may reduce the risk of barotrauma seen with jet ventilators and other devices [18]. A team at the Royal Children's Hospital in Melbourne, Australia, developed an institutional approach

to the CICO scenario that includes a CICO pack with two separate pouches containing equipment needed for either a cannula or scalpel FONA technique [19]. Other considerations for a CICO bundle include human factors such as the person performing FONA should be different from the person who failed to intubate or oxygenate the patient, to avoid task fixation [20].

How to Best Sedate/Induce a Child with an Anticipated Difficult Airway

Awake intubations are rarely performed in children because of difficulty with patient cooperation and will not be discussed in this chapter. A controversial topic in management of the difficult pediatric airway is whether to use muscle relaxants or to maintain spontaneous ventilation. In infants without difficult airways, it has been shown that the addition of muscle relaxants during intubation to sevoflurane induction improved intubating conditions when compared to placebo or alfentanil and was associated with fewer adverse respiratory events [21]. Further work needs to be done to determine the effect of muscle relaxants in difficult pediatric airway management.

Spontaneous ventilation should be maintained in patients with difficult mask ventilation. This can be achieved either by volatile anesthetics or via intravenous hypnotics. Airway adjuncts such as nasopharyngeal or oropharyngeal airways, two-handed and two-person face mask ventilation, or supraglottic airways may improve mask ventilation.

The challenge of keeping a patient spontaneously ventilating during airway management is the ability to instrument the airway without including coughing, bucking, laryngospasm, bronchospasm, or vomiting. Sevoflurane alone may not provide adequate conditions for intubation of a pediatric difficult airway in a spontaneously ventilating patient. Erb et al. showed in a study that defensive airway reflexes including cough, expiration reflex, and spasmodic panting were almost completely suppressed with high concentrations of sevoflurane (sevoflurane

4.7% = $MAC_{ED95Intubation}$) as compared with sevoflurane 2.5% (= 1 MAC). However, the incidence of laryngospasm is only partially reduced even with high concentrations of sevoflurane [22]. We have found a 5 s jaw thrust to be a reliable test of adequate depth of anesthesia for airway instrumentation in a spontaneously ventilating patient. The clinician should look for increases in respiratory rate and heart rate and body movement as signs of inadequate depth of anesthesia.

Total intravenous anesthesia with propofol with or without remifentanyl is a common technique in a spontaneously ventilating patient. Propofol acts via the inhibitory neurotransmitter GABA and causes muscle relaxation and suppresses respiratory drive. Ketamine has also been used as an adjunct for a spontaneously breathing patient and works on blocking *N*-methyl-D-aspartate (NMDA) receptors and preserves ventilation. However, ketamine activates cholinergic systems and leads to hypersalivation – an antisialagogue may help reduce this side effect.

Dexmedetomidine has many properties that are advantageous for anesthesia for a spontaneously ventilating patient, including anxiolysis, analgesia, and sympatholysis. Dexmedetomidine is an α_2 -adrenoceptor agonist and causes sedation by acting in the locus coeruleus. It has also historically been viewed to cause minimal respiratory depression. However, Lodenius et al. showed that sedation with dexmedetomidine significantly reduced hypoxic and hypercapnic ventilation in healthy male volunteers, to a similar extent as sedation with propofol [23]. They induced sedation in ten patients with a bolus over 10 min of dexmedetomidine up to 1.1 $\mu\text{g}/\text{kg}$ or propofol 750 $\mu\text{g}/\text{kg}$, followed by an infusion of dexmedetomidine 0–1 $\mu\text{g}/\text{kg}/\text{hr}$. or propofol 0–75 $\mu\text{g}/\text{kg}/\text{min}$. Conversely, other case reports demonstrate dexmedetomidine attenuates airway reflexes while maintaining stable respiratory profiles in spontaneously ventilating children [24], particularly when combined with another intravenous anesthetic, such as propofol [25]. It is likely that combining two intravenous anesthetics decreases the doses of both drugs needed to provide adequate intubating conditions while preserving spontaneous ventilation.

When dexmedetomidine/propofol (DP)-total intravenous anesthesia (TIVA) was compared to remifentanyl/propofol (RP)-TIVA for rigid bronchoscopy for foreign body removal in 77 children, both groups had similar incidence of desaturation, coughing, and breath-holding rates. However, DP-TIVA patients had respiratory rates closer to baseline and an Et_{CO_2} closer to baseline, suggesting it didn't impair respiratory drive as much as RP-TIVA [26].

Dexmedetomidine has a biphasic effect on blood pressure. There is an initial hypertensive response caused by peripheral vasoconstriction, followed by a sympatholytic effect leading to a decrease in blood pressure. At lower bolus doses (0.5 $\mu\text{g}/\text{kg}$), the hypertensive response is minimized [27]. Another hemodynamic effect of dexmedetomidine is bradycardia – up to a 30% decrease from baseline. Though at a lower bolus dose of 0.49 $\mu\text{g}/\text{kg}$ over 5 s, there isn't significant hemodynamic compromise. Mason et al. report extreme hypertension in response to the administration of glycopyrrolate to treat dexmedetomidine-associated bradycardia and caution treating bradycardia in a normotensive patient [28]. We have achieved very good intubating conditions by combining dexmedetomidine with inhaled sevoflurane in spontaneously ventilating patients with difficult airways. Other combinations have been described including dexmedetomidine with ketamine.

There is increasing awareness that passive oxygenation during intubation is useful for reducing complications and increasing the desaturation free time available to secure the airway. This can be done using a nasal cannula, supraglottic airway, modified nasopharyngeal airway, and modified oral RAE endotracheal tube or through the working channel of a fiber-optic scope. It is important to note the risk of delivering *Oxygen Through the Working Channel* (OTWC) of a fiber-optic scope, including the risk of tension pneumothorax if it is delivered without allowing gas egress. We recommend using this technique only when the scope is outside the trachea and suggest oxygen flows of 2 L/min in infants and 3 L/min for all other ages. Oxygen insufflation can be helpful to blow away secretions and blood. If used in the trachea, it is

important to not wedge the scope, as this can lead to pneumothorax [29]. A prospective randomized controlled trial in 48 healthy children compared passive oxygenation using transnasal humidified rapid-insufflation ventilatory exchange (THRIVE) to standard practice in paralyzed children and demonstrated that THRIVE prolongs the apnea time in children [30]. Steiner et al. examined the use of deep laryngeal oxygen insufflation during laryngoscopy. In one condition they used a laryngoscope with an oxygen cannula attached to the blade and concluded that laryngeal oxygen insufflation increases the time to 1% desaturation and reduces the overall rate of desaturation during lower endoscopy in children [31]. Oxygenation during intubation of the child with a difficult airway is beneficial and should be performed whenever possible.

The application of topical lidocaine to the larynx is a common practice in pediatric anesthesia. Some studies have shown that it decreases perioperative respiratory adverse events, while other studies have shown an increased incidence of complications such as laryngospasm and bronchospasm [32–35].

Newest Gadgets for Difficult Pediatric Airways

Since its advent in the 1970s, the fiber-optic bronchoscope (FOB) has been the gold standard for intubation in difficult airway management. Despite a plethora of new devices and technologies, it still remains the gold standard. Newer devices include video laryngoscopes and second-generation supraglottic airway devices (SGAs).

Video Laryngoscopes

Video laryngoscopes (VLs) have become very popular in difficult airway management. They can be classified as angulated or non-angulated devices. Although they look similar, angulated and non-angulated VLs are very different. The angulated devices are most helpful for difficult airways, while non-angulated devices are useful for routine

intubations. VLs require dedicated practice to master. Although the glottic view is almost always excellent, the clinician has to learn the new skill of inserting the tracheal tube indirectly. Counterintuitively, a grade II view of the airway makes it easier to insert the tracheal tube than a grade I view as there is usually more length in the oropharynx to advance the tracheal tube. Other maneuvers to improve the ease of insertion of the endotracheal tube include rotating the tube, external laryngeal manipulation, and reverse loading the tube onto the stylet. Reverse loading refers to bending the stylet tube at 60 or 90° against its natural concave curve [36]. VLs are associated with longer intubation times than traditional laryngoscopy because of the indirect manipulation of the tracheal tube. The GlideScope VL is the most commonly used VL in children with difficult airways. Park et al. performed the largest study of children with difficult airways comparing the GlideScope to standard direct laryngoscopy. They found that the GlideScope was successful in 82% of patients vs. 21% with direct laryngoscopy [37]. Other VL options for the child with the difficult airway include the Airtraq, the TruView EVO2, and the C-MAC D-blade. Almost all the evaluations of these devices were done in manikins or children with normal airways [38–40].

VLs fail when there is limited space in the oropharynx, limited mouth opening, macroglossia, or large masses in the oropharynx. Visualization of the airway may also be compromised by fogging, secretions, vomitus, or blood.

Fiber-Optic Intubation

Fiber-optic intubation remains the most versatile technique to secure the airway of a child with difficult intubation. Newer fiber-optic bronchoscopes integrate a camera in the tip, thereby enhancing the quality of the image produced. Fiber-optic intubation can be performed freehand or through a laryngeal mask. Burjek et al. compared fiber-optic intubation through a supraglottic airway in children to videolaryngoscopy and found that first attempt success rates were similar; however, fiber-optic intubation through a

supraglottic airway had a higher first attempt success in infants. Furthermore, ventilating continuously through a SGA was associated with a lower incidence of hypoxemia. The Air-Q is the most commonly used SGA for fiber-optic intubation in children. This is because it is designed with a wide airway tube that facilitates the placement of a cuffed tracheal tube.

Supraglottic Airway Devices

SGAs can be used as the primary airway management technique in children with difficult airways. Technical difficulties with using SGAs are inversely proportional to the age of the child. Newer SGAs incorporate a gastric access channel that allows the stomach to be emptied of gastric contents and helps in confirming the SGA is in the correct position. Clinicians should be prepared to secure the airway in the event of any technical issues. Two simple tests can be used to confirm that the second-generation SGA is in good position. First, a small amount of lubrication is placed on the orifice of the gastric access channel – pressing a finger slightly in the supra-sternal notch should cause the lubricant to move in and out if the tip of the mask is in the upper esophageal sphincter. This maneuver compresses the esophagus and sends a slight amount of air up the drain tube causing the lubricant to move slightly. The second test involves placing a small amount of occlusive lubricant on the gastric drain tube while giving a tidal volume breath through the airway tube of the mask. Disruption of the lubricant from the drain tube suggests inadequate separation of the airway and the esophagus – this may occur if the mask is underinflated, inappropriately sized, or sitting too high. Second-generation SGAs with pediatric sizes include the LMA Supreme, Igel, and the Ambu AuraGain.

Summary

Children with difficult airways are particularly vulnerable. Preparation is the key to caring for them successfully – a simple checklist can help

optimize care. Introduce all team members, and review the induction and intubation plan with the entire team, including identifying the clinician making each intubation attempt. Identify an individual who is available to assist before beginning airway management. The following principles should guide the care of these patients.

1. Limit the number of tracheal intubation attempts.
2. Know your equipment. Be prepared.
3. Always attempt to oxygenate during intubation.
4. Ensure an adequate depth of anesthesia before instrumenting the airway.
5. Be wary of cognitive biases that may delay action.

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