The goal of this literature review is to provide the anesthesia practitioner with the skill set to detect and prepare for a difficult pediatric airway. The authors have reviewed and compiled information on some of the most common conditions that can predispose pediatric patients to a difficulty airway, such as macroglossia, mandibular hypoplasia, micrognathia, cervical instability, limited cervical movement, maxillary and midfacial hypoplasia, and cleft palate. This article provides an overview of preoperative assessment techniques, normal pediatric airway anatomy, and respiratory physiology. An emphasis is placed on some common syndromes and their related anatomical abnormalities that can compromise the airway, as well as anesthetic approaches recommended to successfully secure a potentially difficult airway.

Keywords: Congenital, difficult airway, malformation, pediatric.

Airway management is a mainstay of anesthesia practice. Practitioners are required to have a firm grasp on airway physiology, assessment, and management, and regular exposure to this aspect of anesthesia is vital to maintaining these skills. Unfortunately, for many anesthesia practitioners, only limited opportunities to manage the pediatric airway exist.

Statistics compiled in 1993 from the American Society of Anesthesiologists Closed Claims Database have demonstrated that “respiratory events” were more common in the pediatric population (ie, 5 years of age and younger) than in the adult population. Specific respiratory events relative to this review (eg, airway obstruction, difficult intubation, inadequate ventilation) were also shown to occur at a greater frequency in children (34%) vs adults (23%). The database also revealed that of 238 pediatric claims reviewed, 67 (28%) occurred in children under the age of 1 year. Since then, the rate of these specific respiratory events for all pediatric patients has decreased to approximately 7%. This decline in adverse respiratory events may be attributed to many factors, including better monitoring equipment, such as pulse oximetry and end-tidal gas analysis, as well as safer medication profiles.

Although most pediatric airways may be easily secured by the experienced practitioner, the potential still exists for discovering a difficult airway (DA) after induction, possibly leading to a cannot-intubate-cannot-ventilate situation. One retrospective study involving 8,434 patients under the age of 18 years demonstrated the overall incidence of a difficult laryngoscopy to be 1.35%. This study also noted that those under the age of 1 year (n = 1,759) had the greatest risk of presenting with a DA (4.7%).

One primary objective of the authors’ assessment of the pediatric airway literature was to enhance the anesthetist’s ability to detect and prepare for a difficult pediatric airway, thereby improving overall airway management skills. For the purposes of this review, the authors have defined a DA as one in which adequate gas exchange is not readily accomplished through mask ventilation, airway adjuncts such as oral and nasal airways, or a combination of the two. A difficult intubation is said to occur when an experienced provider requires 3 or more attempts to place an endotracheal (ET) tube, with or without rescue adjuncts, such as retrograde wires, light wands, video laryngoscopes, or gum elastic bougies.

Review of the Literature

Using multiple online resources, such as EBSCOhost.com, ScienceDirect.com, and PubMed.gov, the authors searched for publications relevant to pediatric airway assessment and management, pediatric genetic malformations with airway management implications, and acute or chronic injuries that may potentially alter the normal pediatric airway, thereby making it difficult to secure. Search words used to locate relevant articles included airway, pediatric, malformation, Down syndrome, cleft palate, difficult, management, and assessment.

Unfortunately, many journal articles reviewed by the authors did not operationalize the term pediatric, making assessment and critique of such literature difficult. For the purposes of this article, the term pediatric is subcategorized as follows: premature, born less than 37 weeks’ gestation; neonate, birth to 1 month; infant, 1 month to 1 year; toddler, 1 to 3 years; child, 3 to 12 years; and adolescent, 13 to 18 years.
Normal Pediatric Airway Anatomy and Physiology

The pediatric patient is both anatomically and physiologically different from the adult patient in many ways. These differences are most important under 2 years of age and decrease as the child matures, with most differences disappearing around 6 to 8 years of age. One difference that is readily noticeable on assessment of the infant and toddler is the proportionately larger head, specifically the occipital area. This difference is clinically significant in that the pediatric patient may potentially benefit from a pillow or roll being placed under the shoulders, as opposed to the head, to assist with neck flexion. In contrast, other researchers have suggested that direct laryngoscopy is most effective when the shoulders and head are on a flat surface and the neck is fully extended.

The anatomy of the pediatric airway is composed of the same anatomical components as seen in adults, although the size and position of several structures are noticeably different. The most notable differences can be seen when the adult upper airway is compared with that of the neonate and infant upper airway. Compared with the adult, the neonate and infant nose is softer, with more mucous and lymphoid tissues present. Because of the smaller diameter of the nasal passages, it takes much less swelling or secretions to decrease total airflow, which can easily lead to obstruction since it has been suggested that infants are obligate nasal breathers until they reach 2 to 6 months of age. Being obligate nasal breathers is also important since an improperly placed face mask can occlude the nares, creating an obstructed airway. One airway structure that is proportionally larger in the pediatric patient than in the adult is the tongue. The pediatric tongue, especially during the neonatal and infant stages, is located closer to the palate by a more superiorly located larynx, which potentiates airway obstructions. The epiglottis also sits at a greater angle to the anterior pharyngeal wall, making visualization of the vocal cords more difficult. Like the epiglottis, most of the cartilaginous structures of the pediatric airway are softer (less ossified) and more pliable than the adults, increasing the chances of compression and obstruction of the airway when pressure is applied, such as with the Sellick maneuver.

The last important anatomical difference of the upper airway is the diameter of the cricoid ring. In the adult, the narrowest portion of the upper airway is the vocal cords, as opposed to the cricoid cartilage in the neonate, infant, and young child. This difference makes the larynx funnel shaped, as opposed to the cylindrical shape of the adult and adolescent larynx. This feature allows the uncuffed ET tube to create a relative subglottic seal; the narrowing of the airway below the vocal cords presses against the sides of the tube.

There are several anatomical differences in the pediatric patient that are important to the anesthesia provider, although they are not part of the airway. The costae are more horizontal and contribute less to inspiratory and expiratory efforts, forcing the diaphragm to do most of the work in neonates. The infants’ respiratory muscles are primarily composed of type II (fast-twitch) fibers, and have lower stores of glycogen and fat, allowing them to become easily fatigued after short periods of exertion or labored breathing. This physiologic characteristic further emphasizes the importance of rapidly establishing and securing the pediatric airway.

One of the most clinically significant physiologic factors unique to neonates, infants, and toddlers is their elevated metabolic rate, leading to a rate of oxygen consumption more than double that of the adults (7-9 mL/kg/min compared with 3 mL/kg/min in adults). Another contributing factor to their more rapid rate of desaturation is their lower functional residual capacity, 22 mL/kg on average for toddlers vs 34 mL/kg for the adult. The newborn has just half the number of alveoli and only 1/20th the surface area available for gas exchange compared with the adult, limiting oxygen absorption. The neonate’s and infant’s nervous system is predominantly influenced by the parasympathetic system (because of the incomplete maturation of the sympathetic nervous system), causing bradycardia in response to hypoxia and further decreasing oxygen delivery. Because of these factors, when infants begin to experience an airway obstruction or become relatively tachypneic, they compensate more rapidly than do adults.

Preoperative Assessment

Adult airway assessment systems are not always applicable to children. For example, the Mallampati classification system and various measurements used to assess the adult airway, such as thyromental and incisor distance, do not consistently correlate with the pediatric patient population. However, the Mallampati classification system has been shown to be applicable for use in those 4 to 8 years of age. Because of this, the anesthesia provider will commonly rely on his or her physical assessment and the surgeon’s history and physical examination for predicting the DA in children.

As with all patients, the assessment process should begin with a thorough chart review, including the sur-
One prominent reason for a DA in the pediatric patient is an anatomical malformation. Although many congenital, genetic, and acquired malformations and abnormalities may present as a cluster, many may also present in isolation (eg, cleft palate). Malformations presenting as a group may also define a syndrome (eg, Pierre Robin). Although most anesthesiology practitioners may never encounter a rare malformation, other abnormalities may be seen infrequently depending on the provider’s practice setting; for example, Down syndrome has a reported incidence of 11.8 cases per 10,000 live births.²⁰ Although it is outside the scope of this article to cover all acquired and congenital malformations, as well as all types of airway devices, some of the more familiar anatomical abnormalities known to affect the pediatric airway are listed in the Table.²⁰-²⁴

### Table. Congenital Syndromes and Their Related Anatomical Abnormalities

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Incidence rate</th>
<th>Common clinical features</th>
<th>Potential related anatomical abnormalities</th>
</tr>
</thead>
<tbody>
<tr>
<td>Down</td>
<td>11.8:10,000 live births²⁰</td>
<td>Short stature, mental retardation, hypotonia, epicanthal fold, microcephaly, CHD</td>
<td>Macroglossia,²¹ cervical instability,²²,²³ mandibular hypoplasia/micrognathia,²¹,²⁴ maxillary/midfacial hypoplasia²¹,²⁴,²⁵</td>
</tr>
<tr>
<td>MPS (eg, Hurler, Hunter, Morquio)</td>
<td>1:25,000 live births²⁶</td>
<td>Cognitive deficit, hearing/vision deficit, dwarfism, skeletal dysplasia, CHD, coarse facial features, myelopathy</td>
<td>Macroglossia,²⁷,²⁸ cervical instability,²⁷,²⁸ limited cervical movement²⁷,²⁸</td>
</tr>
<tr>
<td>Pierre Robin</td>
<td>1:8,500 live births²⁹</td>
<td>Respiratory compromise, feeding difficulty, macroglossia, glossoptosis, gastroesophageal reflux, CHD</td>
<td>Macroglossia (relative),³⁰ mandibular hypoplasia/micrognathia,³⁰,³¹ cleft palate³⁰,³¹</td>
</tr>
<tr>
<td>Treacher Collins</td>
<td>1:50,000 live births²²</td>
<td>Aplastic zygomas, microstomia, CHD</td>
<td>Mandibular hypoplasia/micrognathia,³³ maxillary/midfacial hypoplasia,³³,³⁴ cleft palate³²,³⁴</td>
</tr>
</tbody>
</table>

Anatomical Abnormalities of Congenital, Genetic, and Acquired Malformations

One prominent reason for a DA in the pediatric patient is an anatomical malformation. Although many congenital, genetic, and acquired malformations and abnormalities may present as a cluster, many may also present in isolation (eg, cleft palate). Malformations presenting as a group may also define a syndrome (eg, Pierre Robin). Although most anesthesiology practitioners may never encounter a rare malformation, other abnormalities may be seen infrequently depending on the provider’s practice setting; for example, Down syndrome has a reported incidence of 11.8 cases per 10,000 live births.²⁰ Although it is outside the scope of this article to cover all acquired and congenital malformations, as well as all types of airway devices, some of the more familiar anatomical abnormalities known to affect the pediatric airway are listed in the Table.²⁰-²⁴

- **Macroglossia.** Macroglossia, defined as a proportionately large tongue, has the potential to create a DA through posterior migration of the tongue, either during or after induction. This can cause an airway obstruction and make it difficult to mask ventilate and obtain an adequate view of the vocal cords during laryngoscopy. One congenital defect that commonly presents with macroglossia is mucopolysaccharidosis (MPS).²⁷ This lysosomal storage disease consists of 7 different syndromes (Hurler, Hunter, Sanfilippo, Morquio, Maroteaux-Lamy, Sly, and Natowicz); with Hurler syndrome being associated with the highest incidence of difficult intubation (27.6%-54%).²⁷,³³

Although macroglossia typically presents as a result of...
a congenital malformation, it may also occur secondary to acute trauma or an allergic response. This could include angioedema of the tongue; angiotensin-converting enzyme inhibitors, which are regularly prescribed to pediatric patients with congenital heart defects (CHDs), have been known to produce lingual subcutaneous edema.

Macroglossia is one of the more easily treated malformations, potentially requiring an oral airway while performing mask ventilation and use of video laryngoscopy during ET tube placement. Pulling the tongue forward with padded forceps can aid in visualization of the vocal cords. The straight blade, such as the Wisconsin or Miller blade, has been recommended by many authors as being the ideal blade of choice in neonates and infants with macroglossia.

- **Cervical Spine Abnormalities.** Cervical spine abnormalities fall into 2 categories: those that cause instability and those that limit mobility. Cervical spine instability is a hallmark finding in some of the more common congenital malformations, with Down syndrome being the most widely known. Cervical spine instability also can have an acute presentation, such as in patients with injuries sustained from a motor vehicle accident or other trauma. Patients with cervical instability are at risk of compression of the cervical spinal cord, especially due to manipulation of the cervical vertebrae during laryngoscopy. Depending on the severity of the malformation or trauma, preoperative lateral flexion and extension images of the cervical spine may be beneficial. In-line stabilization (not traction) is of utmost importance, and the neck and head should remain in a neutral position as possible throughout the entire procedure.

The fiberoptic bronchoscope, Bullard laryngoscope, bougie, and light wand, may be useful in patients with limited or unstable cervical vertebrae since these devices do not require the neck to be mobilized during attempted visualization of airway structures. The Bullard laryngoscope has been shown in one prospective study involving 18 “fresh” cadavers intubated by one anesthesiologist, to provide the best view of the vocal cords with the least amount of cervical manipulation compared with the GlideScope (Verathon) video laryngoscope, and the Viewmax (Rusch) or Macintosh laryngoscope blades. The GlideScope also has been shown to improve the laryngoscopic view in the pediatric patient with a DA. In contrast, in normal pediatric airways, one prospective randomized study involving 134 patients (birth to age 10 years) found the Glidescope and Truview PCD (Truphatek) video laryngoscopes were inferior to direct laryngoscopy. In patients not at risk of aspiration, a laryngeal mask airway (LMA) is an excellent choice for airway management. It is also recognized as being a rescue airway device for failed intubations. Contraindications to the use of an LMA include a full stomach, recent trauma, and the need for high (> 20-25 cm H2O) positive pressure ventilation.

- **Mandibular Hypoplasia and Micrognathia.** Mandibular hypoplasia and micrognathia are considered by some authors as synonyms for an undersized mandible. One syndrome commonly associated with this condition is Pierre Robin syndrome. The clinical triad of this syndrome consists of micrognathia, cleft palate, and glossoptosis, or a downward displacement of the tongue. When this clinical triad is accompanied by additional malformations or syndromes, it is referred to as Pierre Robin sequence. Although the jaw in the patient with Pierre Robin syndrome is hypoplastic, it is also retrognathic, meaning the attachment location of the mandibular condyle to the skull is in a more posterior position, further increasing the difficulty of intubation.

Micrognathia causes the proportionately large tongue to be forced to a more posterior position in the oropharynx, making it difficult to obtain an adequate view of the vocal cords with direct laryngoscopy. Because of this, the posterior portion of the tongue and glottis is more acutely angled. The Glidescope, Airtraq optical laryngoscope (Prodol Meditec), DCl video laryngoscope (Karl Storz), Truvieview PCD, and flexible fiberoptic bronchoscope are well suited for attempting to intubate the patient with a DA. Manual external tracheal manipulation is also helpful in decreasing the angle of the base of the tongue and glottis, aiding in the visualization of the vocal cords. The proportionately large tongue is also a hindrance during direct laryngoscopy because there is no room to sweep the tongue when attempting to align the axes of the head and neck.

- **Maxillary/Midfacial Hypoplasia.** Maxillary hypoplasia, which is a primary component of “midfacial hypoplasia,” is seen with several congenital malformations (see the Table), and intubating conditions may be similar to those with micrognathia. Midfacial hypoplasia typically consists of a hypoplastic nasal bone, maxilla, and zygomas. One common problem encountered with midfacial hypoplasia is OSA. The underdeveloped facial bones force the tongue to a more posterior position, making airway obstruction a common problem. Airway management modalities are similar to those used for micrognathia.

- **Cleft Palate.** One anatomical malformation that may complicate the management of the pediatric airway is the cleft palate. The incidence of cleft palate, with or without cleft lip, is reported to be between 9.1 and 15.6 cases per 10,000 live births worldwide, which makes it one of the most common birth defects. Epidemiologic studies have shown that clefts are an isolated malformation in 71% of occurrences, and 29% to 33% of cases are associated with a syndrome, chromosomal abnormality, or multiple congenital anomalies of unknown etiology. Patients with Pierre Robin syndrome have a 50% to 80% chance of presenting with cleft palate. Male occurrence is also greater, with an odds ratio of 1.7.
During assessment of a patient with a cleft palate, there are several other closely related maladies that should be considered. Up to 29% of patients presenting with cleft palate (with or without cleft lip) will have some form of CHD, such as patent foramen ovale, patent ductus arteriosus, or coarctation of aorta. Depending on the severity of the cleft palate, a complete airway obstruction may occur if the tongue enters the cleft, effectively occluding both the oral and nasal cavities simultaneously. Although the general pediatric population has OSA at an occurrence of 2% to 3%, patients with cleft palate experience OSA at a significantly higher rate (31%), and patients with Down syndrome are at an even greater risk of OSA (45%).

Airway management of patients with an isolated cleft palate (ie, cleft palate with no cleft lip) is usually accomplished with minimal difficulty. One retrospective study involving infants and toddlers (1 month to 3 years of age) reported 4.77% of cases being classified as a difficult laryngoscopy and 1.93% as a difficult intubation. In that study, 985 subjects were scheduled for repair of cleft lip and palate. The researchers also found that the incidence of difficult laryngoscopy in infants (1-6 months of age) with cleft lip/palate was 7.06%.

For patients with bilateral cleft palates, the premaxilla (the most anterior portion of the maxilla) is angled forward, causing the upper incisors to hamper blade movement and obstruct the line of sight during direct laryngoscopy. During a direct laryngoscopy in a patient with a unilateral left or bilateral cleft palate, the laryngoscope blade tends to drop into the left cleft palate, further obstructing the view. This can be remedied by gently packing the cleft palate with gauze before intubation or by using a plastic tooth guard to span the cleft. An oral airway placed immediately after induction typically prevents the tongue from obstructing the oropharyngeal and nasal cavities, allowing for easy mask ventilation.

The Acutely Altered Airway

There are numerous conditions that may cause the pediatric airway to become acutely difficult to secure. Some examples include acute epiglottitis, hemangiomas, papillomatosis, venous lymphatic malformations, burns, laryngoceles, angioedema, or arteriovenous malformations; fortunately, most of these are rare.

One potential alteration with an infectious etiology is acute epiglottitis. In 1989, acute epiglottitis had an occurrence of 15 cases per 100,000 live births, but after the development and subsequent mass administration of the Haemophilus influenzae type B vaccination in 1990, the rate dropped 73% to 4 cases per 100,000 live births. Epiglottitis occurs rapidly within 24 hours, is supraglottic, and typically affects 2- to 7-year-olds, although it may occur in adults. Elective early intubation in the operating room (OR) is recommended for every patient presenting with confirmed acute epiglottitis, effectively avoiding the risks associated with an emergent intubation.

For many of the acute airway alterations listed above, inflammation of some part of the airway is a large contributing factor to the problem. One method of minimizing this inflammation is with oral or intravenous (IV) dexamethasone (0.2-0.5 mg/kg orally or IV). Dexamethasone is also useful in reducing inflammation from surgery and airway manipulation during direct laryngoscopy. Airway management methods of acute presentations are similar to those used to treat congenital ones, as described previously and in the following section.

Considerations for Management of the Difficult Airway

There are several recommendations the authors would like to make that may assist anesthesia providers in managing the difficult pediatric airway. For example, during transport of patients with obstructive abnormalities to the OR, the prone position may help to prevent airway obstruction. This would include patients with macroGLOSSIA, acute airway masses, or epiglottitis. Once the patient is in the OR, preoxygenation should begin as soon as possible. An awake flexible fiberoptic intubation can be highly effective in securing a documented and compromised DA. This may be accomplished with prudent administration of sedation (midazolam, 0.3-0.5 mg/kg orally or per rectal, or ketamine, 1-2 mg/kg intramuscularly [IM] or IV), antisialagogues (atropine, 0.02 mg/kg IM, or glycopyrrolate, 0.01 mg/kg IM), and subsequent local anesthesia of the airway (lidocaine, maximum of 4 mg/kg), administered either by aerosolization or local injection. Maintenance of adequate spontaneous ventilation and effectively maintenance of normal airway tone are essential components of this technique.

The use of IV medications at times may also be a desired method of induction, although preserving spontaneous respirations may be difficult without judicious titration. Propofol can be an ideal drug for this technique because of its short time of onset and duration of action. One caveat to an IV induction is that a peripheral IV catheter is needed, which may be difficult to obtain in an uncooperative child. It is important to note that airway obstruction may occur with all forms of induction, and methods of achieving effective ventilation, such as with oral and nasal airways, LMAs, and jet ventilation, should be readily available.

Following induction of general anesthesia for a surgical procedure requiring a protected airway, the anesthesiologist may at his or her discretion intubate with or without paralytics. Any concerns for substantial airway inflammation due to a difficult intubation should be managed before extubation. Exubation criteria for a child who had a difficult intubation should include full reversal of paralytics, evidence of an adequate spontaneous minute volume, and being completely awake. If the patient's...
postoperative pain is predicted to require the use of high-dose narcotics, a prolonged intubation should be considered because of the respiratory depressant effects of opioids. After the patient has been extubated and transferred to the postanesthesia care unit, all healthcare providers and parents should be informed of any difficulties with perioperative airway management.

Conclusion
Although many pediatric patients commonly present with an uncommonly airway, it is prudent for the anesthetist to be vigilant for the occult challenging airway. Being cognizant of clinical markers and diagnoses that are associated with difficulties in securing an airway is of value. This would include having a cardinal understanding of the anatomical and physiologic differences between a pediatric and adult patient's airway.

Furthermore, a working knowledge of how the airway of neonates, infants, toddlers, children, and adolescents may become altered because of congenital or acquired malformations will allow the anesthetist to establish a patent airway before or during induction of anesthesia. For example, children born with Down syndrome are known to have anatomical abnormalities such as macroglossia, cervical instability, and mandibular and/or maxillary hypoplasia.

Because of the existence of many diverse congenital and acquired malformations that can afflict pediatric patients, it is difficult to establish recommendations for airway management that would be universally applicable to each abnormality. Thus, the authors’ intent with this review was to compile the more prevalent syndromes that anesthetists may encounter and their common clinical features that can impose problems with ventilation, in addition to describing airway management techniques and strategies that may minimize airway mishaps.

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