Check for updates

Airway Management

Monica S. Ganatra

Introduction

Proper airway management is vitally important to ensure adequate oxygenation and ventilation in a patient; without it, death, brain injury, and cardiopulmonary compromise can quickly ensue. Whether a patient receives general anesthesia—in which normal airway reflexes are deliberately suppressed in a controlled fashion—or intravenous sedatives that depress normal respirations in a dose-dependent manner, it is paramount to understand the anatomic abnormalities that can affect management of the airway. This chapter will examine features of both the normal and abnormal airway; highlight the nature of airway anomalies found in common pediatric and adult syndromes; discuss components of a comprehensive airway examination; and describe the airway management for these patients. A thorough understanding of proper airway assessment can go a long way in minimizing the morbidity and mortality that can result from inadequate oxygenation and ventilation.

Normal Airway Anatomy

Airway management refers to the ability to maintain adequate oxygenation and ventilation. In spontaneously ventilating patients, the primary goal is to avoid airway obstruction; and in cases of general anesthesia or unconsciousness from underlying pathology, the aim is to support or control these functions via various invasive devices.

There are several key considerations of a normal airway, including: the relative size of the tongue versus the pharynx in the oral cavity; space in the anterior



M. S. Ganatra (🖂)

Department of Anesthesiology, Yale University School of Medicine, New Haven, CT, USA e-mail: Monica.Ganatra@yale.edu

[©] Springer Nature Switzerland AG 2021

D. Narayan et al. (eds.), Surgical and Perioperative Management of Patients with Anatomic Anomalies, https://doi.org/10.1007/978-3-030-55660-0_3

mandible; integrity of the vertebral column; mobility of the temporomandibular joint (TMJ); and development of the maxilla [1, 2].

When the goal is to intubate a patient for a surgical procedure, there must be sufficient space for the laryngoscope/intubating device, endotracheal tube (ETT), and adequate visualization of the larynx in order to successfully and atraumatically intubate. This is why the ratio of tongue size to pharynx is so important. Similarly, mouth opening is also important; the inability to widen the mouth (which could be due to a congenital anomaly, pain, infection, or scar tissue from radiation) can make it difficult to fit in all the necessary equipment [3].

The oral cavity can be considered to be a box that is composed of the bones of the maxilla and the mandible. The partial content of this box, then, is the soft tissue of the tongue. Any enlargement of the tongue or decrease in box size (due to hypoplasia of the maxilla and/or mandible) will alter this ratio and predispose the patient to airway obstruction [2].

The anterior mandibular space refers to the space in the mandible into which the tongue can be displaced during laryngoscopy. Any decrease in this area will make intubation more difficult because there is literally no room to displace the tongue during laryngoscopy. Mandibular hypoplasia can decrease the anterior mandibular space, and an anterior larynx will also create a similar problem during laryngoscopy [2]. Similarly, hypoplasia of the maxilla will alter the framework of the oral cavity and create a smaller space (Fig. 3.1).

When referring to vertebral column integrity, the two most important elements are the cervical vertebral bones and the atlanto-axial joint, as they play a crucial role in neck extension and flexion. This is necessary to get the best view for intubation. The presence of hemivertebrae, arthritic changes, or vertebral fusion can make intubation more difficult. In the presence of loose ligaments (e.g., atlanto-axial joint laxity in Down syndrome), excess force can cause vertebral column instability and damage to the spinal cord from impingement [2].

The temporomandibular joint is an important bilateral joint. There are articular discs interspersed between the mandible and the temporal bones, splitting the

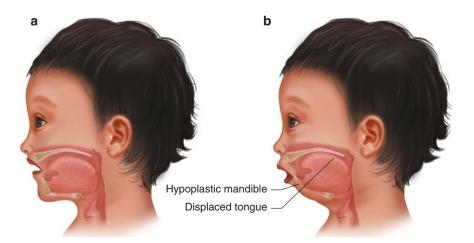


Fig. 3.1 Mandibular hypoplasia and anteriorly displaced tongue

Pediatric Airway	Adult Airway
Long, narrow, and stiff epiglottis	Short and floppy epiglottis
Larynx located at level of C3-C4	Larynx located at level of C4-C5
Larger tongue in proportion to oral cavity	
Narrowest portion at level of cricoid cartilage	Narrowest portion at level of vocal cords
Relatively larger head, especially the occiput	-

Table 3.1 Pediatric versus Adult Airway

articular space into upper and lower compartments. The upper compartment is responsible for gliding movements, while the lower compartment serves as a hinge joint. Mouth opening, which is important so that a laryngoscope can safely be inserted with adequate space, relies on two distinct motions. The first is a hinge action that enables the mouth to be opened about halfway. In the second action, the forward gliding of the articular surface on the articular tubercle is necessary [4].

To best visualize the vocal cords, optimal alignment between the oral and pharyngeal long axes is necessary. The "sniffing position" describes head and neck positioning such that the oral and pharyngeal axes are most advantageously aligned to provide the best visualization of the vocal cords, such that the view from the lips to the glottic opening is most nearly a straight line. Typically, the head should be extended by 15 degrees and the neck flexed 35 degrees. The patient's head is elevated by approximately 8 cm with the use of foam pads or blankets [1, 5].

Previously, it had been thought that the "sniffing position" was required to optimize visualization of the larynx; however, more recent studies have shown that head extension is the more important factor for the majority of patients. Such patients must have normal dentition, adequate submental distance (for tongue decompression), normal glottic structure, and larynx position. For obese patients, the sniffing position is still considered highly beneficial [3].

Table 3.1 highlights the important anatomical differences between the normal pediatric and the adult airway.

Difficult Airway

The definition of a difficult airway, according to the ASA Task Force on Management of the Difficult Airway, is: "the clinical situation in which a conventionally trained anesthesiologist experiences difficulty with facemask ventilation of the upper airway, difficulty with tracheal intubation, or both" [6]. Proper assessment of the airway and knowledge of airway anomalies can help minimize creating a situation where the provider cannot ventilate or intubate a patient.

Congenital Airway Anomalies

There are several characteristics of the pediatric airway that make intubation more difficult. A congenital airway anomaly can make airway management particularly challenging. This section will examine common pediatric syndromes, describe the airway anomaly, and discuss airway management.

Intubation of a pediatric trachea is anatomically more challenging than intubating an adult trachea. First, children have a long, thin, and stiff epiglottis. The infant epiglottis is angled especially posteriorly during laryngoscopy, making it difficult to visualize the vocal cords as the epiglottis blocks direct vision. Straight-tipped blades for intubation (such as the Miller) are more commonly used in neonates and infants because the narrower tip of the Miller (as opposed to the curved tip of the Mac blade) makes it easier to lift the epiglottis, a maneuver known to significantly improve the laryngeal view for intubation.

Because the pediatric larynx is located more cephalad at C3–C4 than the adult C4–C5, the tongue is closer to the palate and causes upper airway obstruction more easily than in adults. Additionally, the pediatric tongue in proportion to the mouth is larger than that of an adult. This also means there is less space to maneuver the laryngoscope to obtain an adequate view for intubation. Additionally, the more cephalad location of the larynx creates a more acute angulation during laryngoscopy, which increases the difficulty of the intubation procedure.

The upper airway obstruction in children (due to the relatively larger tongue) is made worse with sedation, inhalational induction of anesthesia, and emergence from anesthesia [14]. When patients are in a deeper plane of anesthesia, the airway muscles relax and predispose the patient to upper airway obstruction.

A shoulder roll placed under the occiput can help improve views for laryngoscopy.

The narrowest portion of the airway in children is at the level of the cricoid cartilage, which is located below the glottic opening. Therefore, while it may seem that a larger endotracheal tube may be passed through the vocal cords, it will not fit. Diligence must be taken to place correctly sized endotracheal tubes in children so that no damage is incurred from traumatizing the airway by trying to place too large a tube. Such complications can include tracheal stenosis, airway perforation, edema, and scarring.

Historically, cuffed endotracheal tubes were discouraged in children less than 8–10 years of age, out of concern that the inflated cuff would cause subglottic injury. Uncuffed tubes were also thought to be beneficial because a tube with greater internal diameter size could be used. (When cuffed endotracheal tubes are placed, the internal diameter is decreased by 0.5 mm.) However, this belief has been disproven, as modern endotracheal tubes are now equipped with high volume, low pressure cuffs [21]. Nonetheless, uncuffed endotracheal tubes are still preferred in neonates, owing to the use of an endotracheal tube with a larger internal diameter and concern for causing tracheal injury.

Intubated patients are at risk for post-extubation croup and stridor, especially infants and small children. Edema in the upper airway at the narrowest portion (which is the cricoid cartilage in pediatric patients) can cause severe resistance to airflow. Poiseuille's Law states that airflow resistance is dependent on the radius to the fourth power. Consequently, any decrease in radius that occurs from edema will decrease airflow to the fourth power. Infants and small children already have small airways, and any swelling can cause severe restrictions to breathing. The edema is a result of excess pressure on the tracheal submucosa, which can occur with use of too large an endotracheal tube. Venous congestion (and with high enough pressures,

even arterial blood flow compromise) can occur [7]. The edema can be symptomatic, and not surprisingly it is more easily manifest in those with smaller airways (neonates, infants, and small children). It is therefore essential that the proper endotracheal tube size is used. Cuffed tubes are safe to use in infants >30 days of age, provided that a leak is checked so that there is not excessive pressure on the tracheal mucosa.

A simple formula for determining uncuffed endotracheal tube size is:

$$(Agein years / 4) + 4$$

Note that age is in years and the formula works for patients 1 year and up. To obtain a cuffed endotracheal tube size, subtract the obtained number by 0.5.

To estimate depth of endotracheal tube insertion, one can simply triple the endotracheal tube in size; however, it is important to always auscultate for equal, bilateral breath sounds and to inspect for equal, bilateral chest rise. Table 3.2 summarizes age-appropriate endotracheal tube sizes (both uncuffed and cuffed) along with approximate depth of insertion. Of course, clinical judgment should be used to alter these recommendations based on each individual patient.

Laryngotracheomalacia

Laryngotracheomalacia is the most common laryngeal disease of infancy. In this congenital disease, the supraglottic structures collapse with the inspiratory phase of respiration, resulting in high-pitched stridor. The epiglottis is long and narrow, and the aryepiglottic folds are floppy [8]. Most patients (70–90%) have a mild form of the

	Endotracheal tube size: uncuffed	Endotracheal tube size: cuffed	Depth of endotracheal
Age of Patient	(internal diameter, mm)	(internal diameter, mm)	tube at lips, <i>cm</i>
Premature neonate	2.5		8
Term neonate	3.0		9
Infant	3.5	3.0	10
<6 months			
Infant	4.0	3.5	11
>6 months			
1 year	4.0	3.5	12
2 years	4.5	4.0	13
4 years	5.0	4.5	14
6 years	5.5	5.0	15
8 yrs	6	5.5	16
10 yrs	6.5	6.0	18
12 yrs	7	6.5	19
Adult female	7–8	7.0–7.5	21
Adult male	8-8.5	7.5-8.0	23

 Table 3.2
 Appropriate Endotracheal Tube Size Selection and Depth Insertion Based on Age of the Patient

disease, with occasional stridor as their only symptom (which is exacerbated with crying or upper respiratory infections), and resolution usually occurs by 12–24 months of age. Symptoms tend to be worse in the supine position. In more severe cases, the child may demonstrate dyspnea with retractions, obstructive sleep apnea (OSA), dysphagia, failure to thrive (poor weight gain), and episodes of choking while feeding [9]. For the small proportion of patients who have the severe form, associated laryngotracheal lesions (subglottic stenosis, tracheomalacia, vocal cord paralysis, and laryngeal dyskinesia) are more frequently present [9]. Laryngotracheomalacia is commonly associated with gastro-esophageal reflux disease (GERD).

There is serious concern that the child may have total airway obstruction. It is best to maintain spontaneous ventilation, with continuous positive airway pressure (CPAP) as needed. It is best to minimize coughing, and topical lidocaine on the vocal cords can help accomplish this [8]. Prone positioning may have some benefit.

Non-invasive ventilation is useful as it can increase alveolar ventilation and decrease the patient's work of breathing. CPAP, bilevel positive airway pressure (BiPAP), and positive end-expiratory pressure (PEEP) are recommended for improving ventilation in these patients. Maintaining spontaneous ventilation is preferred. More severe cases require surgical evaluation, to assess for other lesions and possible therapeutic interventions. A supraglottoplasty is a procedure designed to remove the excess tissue that contributes to the airway collapse. Severe cases may require tracheostomy [9].

Cleft Lip and Palate

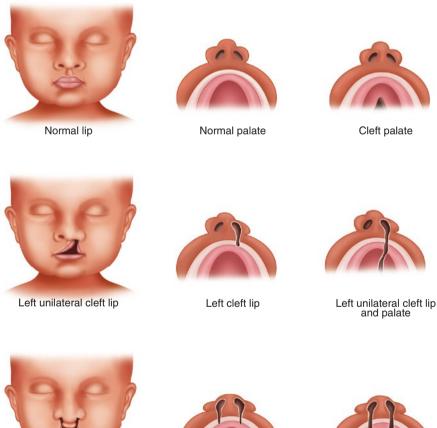
Cleft lip, with or without associated cleft palate, is the most common congenital anomaly of the head and neck. The clefts may be unilateral or bilateral. For complete cleft lips, there is extension through the lip and into the nasal sill, whereas an incomplete cleft lip extends through the orbicularis oris and skin, but intact lip tissue is present [10].

For cleft palates, the most commonly used classification system is the Veau system. In this system, there are four groups. Group 1 defects involve clefts of the soft palate only. Group 2 defects extend into the hard palate. For Group 3 defects, there is unilateral extension into the entire palate and alveolus. And with Group 4 defects, this is bilateral [10].

The greater the severity of the cleft, the greater the chance that there are other associated anomalies, such as those of the limbs, vertebral column, and cardiovascular system. The cleft lip/palate may be associated with other syndromes that have special airway considerations, including, for example, Pierre–Robin, Goldenhar, and Treacher–Collins syndrome. Additionally, children with cleft lip/palate commonly have chronic nasal and sinus infections, which can increase their risk of respiratory complications (such as laryngospasm and bronchospasm) while under general anesthesia [11].

Intubation of patients with clefts may be difficult; therefore, personnel and equipment for managing a difficult airway must be readily available. Packing the cleft with moist gauze can help prevent damage to surrounding tissue. For patients

in whom a difficult airway is anticipated, administering glycopyrrolate prior to airway manipulation can help decrease the amount of secretions and decrease the risk of laryngospasm [38]. Inhalational induction and maintenance of spontaneous ventilation is preferred if there is reason to suspect that laryngoscopy may be difficult. Paralytics should be administered only when it has been ascertained that the patient can be easily mask-ventilated. Care should be taken to select a facemask that fits the patient well and does not place undue pressure on the clefts. While it should be noted that those cleft patients who are less than 6 months old or have associated anomalies are at higher risk of difficult intubation [11], it should be acknowledged that patients with isolated cleft palate tend to become easier to intubate as they get older (Fig. 3.2) [12].



Bilateral cleft lip and palate



Bilateral cleft lip



Bilateral cleft lip and palate

63

Fig. 3.2 Normal lip and palate and cleft lip and palate

Juvenile RA

In juvenile rheumatoid arthritis, a systemic autoimmune disease, deposits of rheumatoid factor accumulate in the joints and cause damage. The temporomandibular joint may be involved, causing limited opening of the oral aperture. There may also be restricted mobility of the cervical vertebrae, causing limited extension and flexion of the neck. This can make intubation of the trachea challenging and require use of video-assisted laryngoscopy or a fiberoptic scope. Other anatomical anomalies include involvement of the cricoarytenoid joints, narrowing of the glottic opening, and laryngeal deviation caused by chronic fibrotic changes [7].

Mucopolysaccharidoses

The mucopolysaccharidoses refer to a group of genetic disorders known as glycogen storage diseases, where absence of key enzymes leads to deposition of glycosaminoglycans [13]. Specifically, there is absent or aberrant function of lysosomal enzymes which are normally required to break down the glycosaminoglycans. The accumulation of these long chain sugar carbohydrates can be seen in bone, skeletal structures, connective tissue, and organs. These deposits increase with time, and it should be expected that airway management will be more difficult as these patients grow older. Bone and joint disease may be present, including instability of the cervical spine. Cervical canal stenosis may also be present, placing the patient at risk for spinal cord compression. Extreme care should be taken when manipulating the airway, and in-line stabilization should be employed. Further work-up with an MRI may be needed if the patient has signs of a cervical myelopathy.

Always assume a patient with a mucopolysaccharidoses is a difficult airway. This is because the patient may have deposits of glycosaminoglycans in the airway, a short and often unstable neck, poor joint mobility affecting not just the c-spine but the temporomandibular joint as well, micrognathia, and macroglossia. Furthermore, OSA may be present. As there is no way for the body to break down these carbohydrates, the airway worsens with time.

Because of the abnormal facies, facemasks may not be a good fit. In such cases, children will often obstruct upon induction of anesthesia. Placement of an oropharyngeal airway may exacerbate the situation by pushing the enlarged epiglottis posteriorly and cause laryngeal obstruction. Due to nasopharyngeal deposits, it may be difficult to place a nasopharyngeal airway. It has been suggested that forward traction of the tongue may be beneficial in relieving obstruction. Laryngeal mask airways (LMAs) have proven to be beneficial as a way to ventilate the patient and as a conduit for intubation. Because of the glycosaminoglycan deposits, endotracheal tubes that are smaller than expected for age should be used [14].

Trisomy 21

Trisomy 21, more commonly known as Down syndrome, has several features that can make airway management more challenging. These patients have macroglossia and hypotonia, placing them at high risk for airway obstruction. Their oral and nasal passages are characteristically narrow [15]. They also have short necks, mid-facial and mandibular hypoplasia, atlanto-axial instability with vertebral ligamentous abnormalities, and a higher incidence of congenital subglottic or tracheal stenosis [13]. Additionally, these patients often have adenotonsillar hypertrophy and obstructive sleep apnea. Use of an oral or nasal airway will help relieve the obstruction. To move the tongue out of the way for intubation, a Macintosh blade is preferable for intubation.

Patients with Down syndrome are at risk of having atlanto-axial instability. Asymptomatic patients with plain cervical flexion and extension radiographs demonstrating an atlanto-dens interval of less than 4.5 mm and a neural canal width of more than 14 mm should be able to proceed to surgery. If the patient has abnormal x-rays and/or symptoms, consultation with a neurosurgeon is advisable in the elective setting. Lateral films may not rule out atlanto-axial instability with any precision, and are often not routinely performed at many centers. Since ongoing mineralization precludes accurate radiological imaging in children under the age of 3 years old, the safest course is to treat all Down syndrome patients as if they do have laxity of the atlanto-axial joint. C-spine neutrality should be maintained at all times, using foam pillows and gel-cushioned pads to support the head. During LMA placement and intubation, maintain in-line stabilization by having one provider hold the neck neutral while the other person intubates without placing force on the neck. Any undue flexion or extension of the head can cause damage to the cervical spinal cord. When intubating, endotracheal tubes that are 0.5 to 1 mm smaller than expected are often used because of the known tracheal narrowing and also to avoid subglottic trauma. Intubation is not generally known to be difficult, as the mouth opening is normal and the large tongue is usually easy to displace [13].

Beckwith–Wiedemann Syndrome

Beckwith–Wiedemann Syndrome is characterized by macroglossia, visceral organomegaly, and gigantism. OSA may be present. Maxillary hypoplasia and macroglossia can cause upper airway obstruction and lead to difficult direct laryngoscopy. Abdominal organ visceromegaly may push the diaphragm upward, shortening the distance from the lips to the carina and making endobronchial intubation more likely.

The macroglossia can cause upper airway obstruction. Oral and nasopharyngeal airways can help relieve this obstruction, but if this is not successful, the patient may be placed in a lateral or prone position for relief. Having someone pull the tongue forward (e.g., with McGill forceps) can help facilitate direct laryngoscopy. The tracheal diameter in these patients tends to be wider than expected for age; cuffed endotracheal tubes are recommended. It is advisable to have a fiberoptic bronchoscope available for intubation [13].

Pierre–Robin Syndrome

Pierre-Robin Syndrome consists of micrognathia, glossoptosis, and cleft palate. The micrognathia (which is also known as mandibular hypoplasia) forces the tongue to be relatively posterior within the oropharynx, such that intubation via direct laryngoscopy is extremely difficult, if not impossible, because the vocal cords cannot be visualized. The symptoms can be so severe that the newborn in a supine position can have complete obstruction. Placing the patient in a prone position can help relieve the obstruction by displacing the tongue. In some cases, a glossolabiopexy may be necessary, in which the tongue is kept anterior and prevented from prolapsing posteriorly by placing a stitch that connects the tongue to the lower lip [7]. During anesthesia, if the tongue has not already been sutured to the lip, use of McGill forceps to pull out the tongue can help relieve airway obstruction. The lateral position can also be beneficial because it moves the tongue out of the way and prevents obstruction of the epiglottis [4]. In addition to maintaining spontaneous ventilation, a flexible fiberoptic scope should be readily available for intubation as direct laryngoscopy is often unsuccessful. An otolaryngologist should also be readily available should fiberoptic intubation be difficult or unsuccessful. The LMA is an important tool for Pierre–Robin patients, because it can move the tongue out of the way and thereby minimize or prevent airway obstruction that will occur from the prolapsing tongue [7]. Fortunately, as infants grow, so do their mandibles, making it easier to manage the airway. Severe cases of Pierre-Robin may require a surgical airway (Fig. 3.3).

Treacher–Collins (Mandibulo-Facial Dysostosis)

In Treacher–Collins Syndrome, there is maxillary, zygomatic, and mandibular hypoplasia. Downward sloping palpebral fissures, notched lower eyelids, small mouth opening, and a high arched palate are characteristic features, with cleft palate and velopharyngeal incompetence as secondary concerns. There may be associated TMJ abnormalities as well. Mask ventilation and intubation is extremely difficult and nearly impossible if there are associated TMJ abnormalities. As these children grow, airway management can become even more challenging as the basilar kyphosis of the cranial base may increase. Therefore, although it may be useful to examine previous anesthetic records for airway management, it is important to recognize that in patients with mandibulo-facial dysostosis, the airway management may subsequently be even more difficult (Fig. 3.4) [2, 16].

Glossoptosis -

Definition: A tongue that is positioned higher and further back in the mouth than usual. The tongue is displaced toward the roof of the mouth.

Although the actual tongue size is normal, the abnormal tongue position causes airway obstruction.

Cleft palate

Definition: Embryonic structures known as palatal shelves fail to fuse together properly, leaving an opening in the roof of the mouth. This creates an abnormal connection between the nasal passages and the mouth.

60-90% of patients with Robin Sequence will have a cleft palate.

Micrognathia -

Definition: An underdeveloped or abnormally small lower jaw (mandible).

Infants with micrognathia have jaws that are much smaller and more set back compared to the upper jaw and face.

Fig. 3.3 Features of Pierre–Robin syndrome

Airway obstruction

Definition: Difficulty breathing due to blockage of the airway.

While airway obstruction can have many different causes, in babies with Robin sequence the breathing problem is due to the abnormal tongue position.

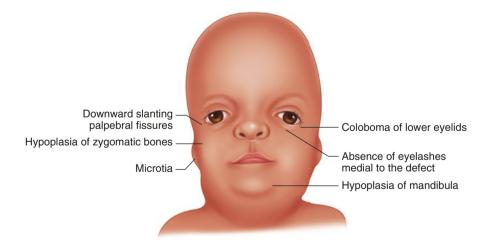


Fig. 3.4 Treacher–Collins syndrome

Goldenhar Syndrome (Hemifacial Microsomia)

The developmental disorder Goldenhar Syndrome, also known as oculo-auriculovertebral syndrome, features mandibular hypoplasia, malformations of the external and middle ear, eye abnormalities (microphthalmos and epibulbar dermoids), and vertebral anomalies that include scoliosis and cervical spine malformations. There may be various phenotypic presentations of Goldenhar, ranging from mild facial asymmetry to severe deformities with mandibular hypoplasia [15]. Fiberoptic intubation or video-assisted laryngoscopy is frequently needed for tracheal intubation.

Apert's Syndrome

The disorder known as Apert's Syndrome is characterized by a flat, elongated forehead; obtuse nasofrontal angle; and maxillary retrusion/mandibular prominence. There is also acrocephaly (congenital abnormality of the skull in which the top of the head assumes a conical or pointed shape) due to synostosis of the coronal suture. A cleft soft palate is also featured, with reports of closure causing obvious airway obstruction. Cervical spine fusion is often present, most commonly at the C5–C6 level. Laryngomalacia, bronchomalacia, and a cartilaginous tracheal sleeve may also be seen. Obstructive sleep apnea is also associated with this syndrome. Choanal stenosis or atresia may be present, and a definitive airway (tracheostomy) may be necessary. And finally, it is important to note that midface hypoplasia may make it more difficult to mask ventilate [15].

Crouzon's Syndrome

Crouzon's Syndrome is a type of craniosynostosis characterized by premature fusion of the bicoronal sutures, maxillary hypoplasia, and shallow orbits that lead to ocular proptosis and a characteristic beaked nose. Due to midfacial deformities, nasopharyngeal obstruction is common and can lead to sleep apnea and debilitated nasal breathing [15]. Intervertebral fusion, including that of the c-spine, may be present. A difficult airway should be anticipated.

Obstructive Sleep Apnea

Obstructive sleep apnea (OSA) occurs when there is periodic obstruction of the upper airway, and the obstruction may be partial or complete. This leads to repetitive arousal during sleep, and the child may exhibit signs of aggressive behavior or easy distraction. Additionally, as in adults, children may also have daytime hypersomnolence. Clinically, patients will have oxygen desaturation while sleeping and

exquisite sensitivity to opioids, benzodiazepines, and inhaled anesthetics. They may also have hypercarbia and cardiovascular dysfunction [17]. Even asymptomatic patients will show signs of upper airway obstruction when given sedatives and anesthetics.

In the adult population, obesity is the single most important physical characteristic associated with OSA. Pharyngeal airway tissue enlargement leads to upper airway obstruction. In children, OSA is mostly due to either craniofacial anomalies or tonsillar hypertrophy, although the incidence of obesity in children is rising [18].

Almost half of all patients with craniofacial dysostosis develop OSA and require airway intervention at some point in time. Having a reduced cranial base angle pulls the pharyngeal wall forward and results in anteroposterior shortening of the nasal and oral airway. Furthermore, the temporomandibular joint may be drawn posteriorly, such that the mandible is now positioned retrusively. With a mandible that is small and retrognathic (e.g., patients with Pierre–Robin), the tongue is positioned posteriorly and encroaches on the oropharynx and hypopharynx. Upper airway obstruction may be exacerbated in the presence of other anomalies, such as choanal atresia, septal deviation, and turbinate hypertrophy [15].

Sher and colleagues [19] used flexible fiberoptic nasopharyngoscopy to identify the ways in which pharyngeal obstruction occurs in patients with OSA. The four mechanisms are outlined as follows:

The tongue moves posteriorly against the posterior pharyngeal wall.

The tongue moves posteriorly and compresses the soft palate or cleft palatal tags posteriorly against the back pharyngeal wall. The tongue, velum, and posterior pharyngeal wall meet in the upper oropharynx.

The lateral pharyngeal walls move medially and appose each other.

The pharynx constricts in a circular or sphincteral manner.

Drugs that act centrally affect OSA patients by decreasing the action of the pharyngeal dilator muscles, encouraging pharyngeal collapse. Furthermore, volatile anesthetics and intravenous sedatives cause respiratory depression, which can exacerbate the hypoxemia and hypercarbia seen in patients with OSA [18].

Patients who have OSA will especially benefit from oxygen supplementation prior to administration of any sedative drugs. Continuous positive airway pressure may be required. Placing the patient in the lateral, recumbent, or prone position may increase the functional residual capacity (FRC) and help to relieve the obstruction. Oral or nasopharyngeal airways are beneficial in maintaining airway patency, and are much better tolerated in heavily sedated and anesthetized patients.

OSA patients can be difficult to intubate, whether or not they have an associated craniofacial anomaly. LMAs are useful as they can push the tongue out of the way and help relieve airway obstruction. In some cases, an awake fiberoptic intubation may be needed, although the advent of advanced airway videolaryngoscopy has decreased this need (Fig. 3.5).

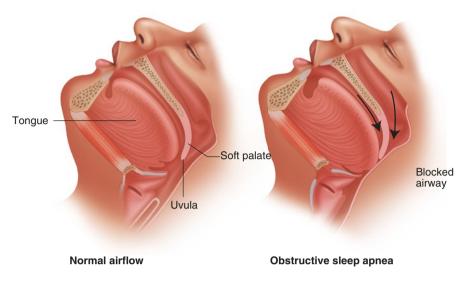


Fig. 3.5 Airflow in a normal patient and in a patient with obstructive sleep apnea

Klippel–Feil Syndrome

Klippel–Feil Syndrome is characterized by congenital fusion of the cervical vertebrae, severe shortness of the neck, and a low posterior hairline. Intubation may be difficult because of limited neck flexion and extension. Intubation may require use of a fiberoptic scope or other advanced airway management device [7].

Subglottic Stenosis

The subglottic area is defined as the area extending from the lower surface of the true vocal cords to the lower surface of the cricoid cartilage [20]. A partial or complete narrowing of the subglottic area may be congenital or acquired. If there is no history of intubation or other acquired causes (such as trauma, infection, foreign body, inflammation, or chemical irritation), then the stenosis is classified as congenital. Severe cases of congenital stenosis are diagnosed in childhood, with misdiagnoses of asthma and bronchitis. Symptoms of subglottic stenosis include dyspnea, stridor, hoarseness, recurrent pneumonitis, cyanosis, and a brassy cough.

Subglottic stenosis involves narrowing of the cricoid cartilage, the only complete cartilage that forms a circumferential ring around the trachea. If the patient requires intubation, care must be taken to not place an endotracheal tube that is larger than the narrowest portion of the airway, as this can cause further inflammation and damage to the surrounding tissue. If the otolaryngologist has already evaluated the patient's airway, sizing of the patient's airway should be documented, and this can

be used as a guide for what size endotracheal tube can be placed. If the patient is coming in for evaluation of the stenosis, then the surgeon and anesthesiologist will work together to evaluate and maintain the patient's airway during the procedure. The surgeon will use a rigid bronchoscope to evaluate movement of the patient's vocal cords, assess the extent of the stenosis, and possibly perform an intervention to decrease the diameter of the stenotic lesion. This may be accomplished by a laser or balloon dilation. More severe cases of airway stenosis may require laryngotracheal reconstruction, where portions of the stenotic airway are removed and replaced with cartilage from the ribs. Initial assessment of the stenosis can occur in the otolaryngologist's office, with the use of a flexible nasal fiberoptic scope. However, children may not cooperate with this procedure, in which case the patient will require anesthesia.

The anesthetic goals during assessment of airway stenosis are challenging. The patient must have enough anesthesia on board to be still during the airway exam and simultaneously be spontaneously breathing so that the surgeon can assess movement of the vocal cords. Two popular techniques are total intravenous anesthesia with propofol and inhalational anesthesia with sevoflurane [21]. In the first case, an IV infusion of propofol is incrementally titrated to achieve the goals of adequate sedation with spontaneous ventilation. Once this is achieved, and the surgeon has a view of the glottis, topical lidocaine is applied to the vocal cords to minimize the risk of laryngospasm. In the second method, the patient's ventilation is intermittently assisted via the use of bag and mask to deliver an inhalational agent. Sevoflurane is most commonly used as it is considered to be the least irritating volatile agent. Occasionally, an endotracheal tube must be placed intra-operatively to help with ventilation. Dexamethasone (up to 1 mg/kg, max 20 mg) is often administered intra-operatively to help decrease airway edema. Table 3.3 summarizes the Meyer–Cotton grading system for classifying the severity of the subglottic stenosis.

Laryngeal Webs

Laryngeal webs account for 5% of congenital anomalies of the larynx. Webs occur when the laryngeal lumen fails to recanalize during embryogenesis. This can result in simply a thin strand of tissue between two areas of the larynx to complete airway infiltration with thick tissue. Seventy-five percent of the webs occur at a glottic level [23]. Patients with only a thin membrane of tissue in the larynx may

Table 3.3	Meyer-Cotton Grading
System for	Subglottic Stenosis

Stenosis grade	Degree of obstruction
Ι	0-50% obstruction
II	51-70% obstruction
III	71–99%
IV	Absence of a detectable lumen

Data from Hartnik and Cotton [22]

be asymptomatic or present with a weak cry or mild hoarseness. Those with bulky webs, webs that extend into the posterior glottis, or those that involve the subglottis will have biphasic stridor and either a weak voice or aphonia [22]. Flexible laryngoscopy is usually performed by an otolaryngologist for diagnosis; in the operating room, rigid bronchoscopy is performed and treatment may involve ablation with laser, excision of the web, and/or serial dilations. Shared airway procedures with the surgeon often require use of intravenous anesthesia and spontaneous ventilation.

Pyriform Aperture Stenosis

Congenital nasal pyriform aperture stenosis (CNPAS) is a rare disorder where narrowing of the anterior-most nasal process of the maxilla is caused by bony overgrowth in utero. Clinical signs and symptoms are similar to bilateral choanal atresia—tachypnea, feeding difficulties, apnea, or cyanosis—that resolves with crying or mouth-breathing, and difficulty passing a suction catheter through the nose [23]. These patients are often managed with conservative treatment that includes nasal decongestants, frequent suctioning, intranasal steroid drops, humidification, and use of an oral airway. Usually, as the neonate grows and develops, the nasal passage is no longer stenotic enough to cause symptoms. However, surgical repair is warranted if conservative management fails and the child needs persistent ventilation or has failure to thrive. When managing the airway of a patient with CNPAS, care must be taken to not traumatize the nasal passages. Oral airways will help relieve the obstruction. This is a rare neonatal disease, and if surgery is indicated, it usually occurs within the first month of life [24].

Laryngeal Clefts

Another rare pediatric congenital airway anomaly is laryngeal clefts. These are posterior airway defects, where there is an abnormal connection between the larynx and hypopharynx or esophagus, and it is caused by failure of the interarytenoid tissues, cricoid cartilage, or tracheoesophageal septum to fuse [23]. The Benjamin and Inglis classification is often used to describe the laryngeal clefts based on its depth. Type 1 clefts involve the interarytenoid region superior to the level of the vocal cords; Type 2 clefts partially extend into the cricoid lamina; Type III clefts involve the entire cricoid cartilage; and Type IV clefts extend into the intrathoracic aorta [25]. The clinical presentation can vary, depending on type of cleft as well as any co-morbidities the patient may have. The symptoms may range from none to mild feeding difficulties to aspiration, cyanosis, stridor after feeding, and recurrent pneumonia or cough.

The goals of airway management are to maintain spontaneous ventilation and avoid positive pressure ventilation so as to not obscure the signs of airway collapse. Use of a fiberoptic scope can help to assess the airway [26].

Syndrome	Airway pathology	Airway management
Freedman– Sheldon	Microstomia Micrognathia Microglossia A high arched palate Midfacial hypoplasia characteristically known as whistling mouth syndrome	Abnormal facial features make direct laryngoscopy and intubation extremely challenging maintain spontaneous ventilation and use LMA [27]
Cri Du chat	Microcephaly Micrognathia Long curved, floppy epiglottis Narrow, diamond-shaped vocal cords Laryngomalacia Stridor [28]	
Cretinism	Macroglossia Goiter that may compress trachea Laryngeal or tracheal deviation	Obtain CT imaging to determine extent of tracheal compression Maintain spontaneous ventilation with LMA
Cherubism	Tumorous lesion of mandible and maxillae with intraoral masses	
Meckel syndrome	Microcephaly Micrognathia Cleft epiglottis	
Ankylosing spondylitis	Ankylosis of c-spine Lack of c-spine mobility Ankylosis of temporomandibular joints	
Acromegaly	Macroglossia Prognathism	

Table 3.4 Airway Pathology and Management for Additional Congenital Syndromes

Based on data from sources cited and Rosenblatt and Sukhupragar [5]

It is impossible to describe all the anatomical anomalies and implications for airway management. In addition to the anomalies described above in detail, Table 3.4 summarizes other syndromes and describes the airway considerations in bullet form.

Pre-Operative Assessment of the Airway

When assessing a patient's airway, it is prudent to look at prior anesthetic records to review previous airway management, specifically how easy or difficult it was to mask ventilate the patient and the ease or difficulty with which an LMA or endotracheal tube was placed. However, it is important to note that the airway anatomy changes over time, and it may be easier or more difficult to manage the airway than had been previously shown. Therefore, it is always best to have back up plans/ equipment/personnel in case it is the latter.

Children often do not have formal sleep studies for OSA. Therefore, it is important to ask families if their child snores, and if so, use narcotics judiciously as OSA

Clinical Feature	How to Assess
Anterior mandibular space	Thyromental distance should be greater than 3 fingerbreadths <i>of the patient</i>
Cervical spine mobility	Assess degree of neck flexion and extension
Tongue versus pharyngeal	Mallampati classification
size	

Table 3.5 Physical Examination to Predict Ease of Tracheal Intubation

Table 3.6 Mallampati Classification

Grade I	Soft palate, anterior and posterior tonsillar pillars, fauces, and uvula visible
Grade II	Tonsillar pillars and base of uvula hidden by posterior tongue
Grade III	Soft palate and base of uvula is visible
Grade IV	Hard palate only visible

may be suspected. Airway obstruction should be anticipated and properly sized oral or nasopharyngeal airways readily available.

Children can often be uncooperative and fail to follow directions. Fortunately, external examination is often sufficient (Table 3.5). The thyromental distance is important as it predicts anterior mandibular space. For children, adequate space is anticipated if it is equal to or greater than the width of the child's three fingerbreadths. Inspect for airway abnormalities such as micrognathia, macroglossia, and mandibular hypoplasia. If the patient's chin is posterior to upper lip, expect a difficult airway; if the chin is neutral with upper lip, it is likely the patient has a normal airway [7]. Assess c-spine mobility by asking the patient to extend and flex the neck.

A cursory dental exam is important. Specifically, inquire about loose teeth, as they can be inadvertently dislodged during airway management and aspirated. The absence of upper incisors allows for better laryngoscopic vector alignment; conversely, the presence of long upper incisors makes laryngoscopy and tracheal intubation more difficult [3].

In predicting the ease of tracheal intubation, the importance of each physical component has already been described in detail at the beginning of this chapter.

The Mallampati Classification system (Table 3.6) is used to visually grade the size of the oral cavity relative to the tongue. The examiner asks the patient to maximally open his/her mouth (normal adult opening is 5–6 cm) and protrude the tongue as far as possible. Based on the pharyngeal structures that are visible, the patient is assigned a Mallampati Score. Grade I and II views are technically easy, whereas a grade III or IV view often predicts a technically difficult or impossible intubation [29]. In children, the Mallampati score has less predictive value (Fig. 3.6).

General Principles of Airway Management

It is beyond the scope of this chapter to describe how to intubate, place a laryngeal mask airway, or use advanced airway management devices such as a fiberoptic scope or videolaryngoscopy. Airway management should be performed in the hands of a skilled provider, an anesthesiologist, or an otolaryngologist who is specially trained to manage airways. Which particular method is used will depend

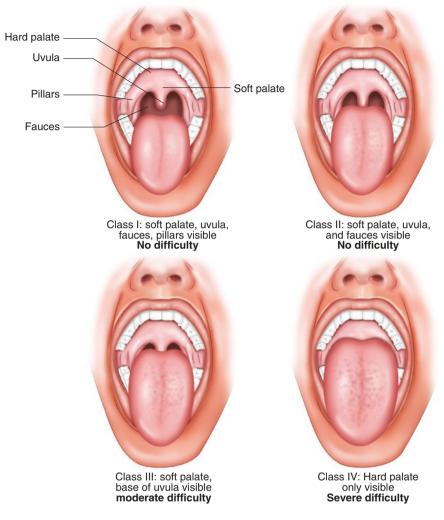


Fig. 3.6 Mallampati Classification

on operator experience, what is available at your institution, and preference of operator. Listed below is a simple explanation of some of the most commonly used airway devices.

Laryngeal Mask Airway (LMA)

The LMA is a supraglottic device that can be rapidly and blindly inserted. Since it is placed above the glottis, there is no risk of placing the LMA in the wrong place (i.e., the esophagus), as there is with intubation. Patients often breathe spontaneously through the LMA, though it is possible to manually ventilate the patient as

well. There are various sizes and types of LMAs (e.g., the LMA Supreme has a port through which an orogastric tube can be placed so that gastric contents and air can be removed). LMAs can also be used as a guiding device for intubation; this requires advanced management skills. The primary disadvantage of the LMA is that it does not protect the patient from aspiration, which can cause severe morbidity, including death.

Endotracheal Tube (ETT)

Intubation is a technique in which an endotracheal tube (with or without a cuff) is placed into the trachea. Direct laryngoscopy is used to visualize the vocal cords and to confirm that the tube is in fact passing through the trachea and not the esophagus. With the advent of modern devices, video laryngoscopy is often used to visualize the vocal cords. There are many anatomical factors that will help determine if the patient is expected to be easy or difficult to intubate, as this chapter has described in detail. The ETT provides a secure, definitive airway, and cuffed tubes minimize the risk of aspiration. However, the process of intubation has several risks, including but not limited to: dental injury; bronchospasm; laryngospasm; mainstem intubation; esophageal intubation; aspiration; trauma to surrounding tissue; and bleeding.

Pre-Oxygenation

Pre-oxygenation is a simple but important task to perform prior to any manipulation of the airway. Prior to the induction of anesthesia, pre-oxygenation by mask maintains higher oxygen saturation values compared with room air controls [30]. Whenever possible, 100% oxygen should be delivered to the patient using an appropriately fitting mask that forms a good seal. The time it takes for a patient to reach desaturation with thresholds of 93–95% oxygen concentration are longer for patients who received 3 minutes of pre-oxygenation compared to those who did not [31–33].

The ability to mask ventilate should not be underestimated—this technique alone can save lives, especially in situations where experts in airway management are not readily available.

In a study involving 53,041 adult patients, Kheterphal et al. reported an incidence of 0.15% for patients that were impossible to mask ventilate [34]. This means that 99.85% of patients were able to be successfully ventilated. Independent risk factors for those that are impossible to mask ventilate included: neck radiation changes, male sex, obstructive sleep apnea, Mallampati score of III or IV, and the presence of a beard [34]. In a study by Langeron et al., "Using a multivariate analysis, five criteria were recognized as independent factors for a difficult mask ventilation (DMV): [age older than 55 yrs., body mass index >26 kg/m², beard, lack of teeth, and history of snoring]; the presence of two indicating high likelihood of DMV (sensitivity, 0.72; specificity, 0.73)" [35].

Summary

Normal airway anatomy, specifically assessment of oral aperture opening, anterior mandibular space, ratio of tongue size to oropharyngeal cavity, and cervical spine mobility are all important features in predicting the ease of airway management. Anatomical abnormalities in one or more of these areas can result in difficulties with ventilating the patient, intubating the patient, or both, all of which can have detrimental effects. The vital organs of the body—namely the brain, heart, and lungs—cannot sustain prolonged periods of hypoxemia, and the resulting damage from cellular hypoxia and organ dysfunction may be irreversible and lead to permanent disability or death. Without proper oxygenation and ventilation at the cellular level, the body's organs will not be able to sustain life. Careful pre-operative evaluation, adequate preparation, and an understanding of how anatomical anomalies can affect airway management and go a long way in ensuring safe airway management.

References

- 1. Miller RD, Pardo M Jr, Stoelting RK. Basics of anesthesia. 6th ed. Philadelphia: Elsevier/ Saunders; 2011.
- 2. Nargozian C. The airway in patients with craniofacial abnormalities. Pediatr Anesth. 2004;14:53–9.
- 3. Ramachandran SK, Kheterpal S. The expected difficult airway. In: The Difficult Airway.
- Block C, Brechner V. Unusual problems in airway management II. The influence of the temporomandibular joint, the mandible, and associated structures on endotracheal intubation. Anesth Analg. 1971;50(1):114–23.
- Rosenblatt W, Sukhupragarn W. Airway management. In: Barash P, Cullen B, Stoelting R, Cahalan M, Stock C, Ortega R, editors. Clinical Anesthesia. 7th ed. Philadelphia: Lippincott Williams & Wilkins; 2013. p. 762–802.
- Apfelbaum JL, Hagberg CA, Caplan RA, Blitt CD, Connis RT, Nickinovich DG, et al. Practice guidelines for management of the difficult airway: an updated report by the American Society of Anesthesiologists Task Force on Management of the Difficult Airway. Anesthesiology. 2013;118(2):251–70.
- Infosino A. Pediatric upper airway and congenital anomalies. Anesthesiol Clin North Am. 2002;20:747–66.
- Hagberg C. Benumof and Hagberg's airway management. 3rd ed. Philadelphia: Elsevier/ Saunders; 2013.
- Ayari S, Aubertin G, Girschig H, Van Den Abbeele T, Denoyelle F, Couloignier V, et al. Management of laryngomalacia. Eur Ann Otorhinolaryngol Head Neck Dis. 2013;130:15–21.
- 10. Crockett D, Goudy S. Cleft lip and palate. Facial Plast Surg Clin North Am. 2014;22(4):573-86.
- 11. Steward DJ. Anesthesia for patients with cleft lip and palate. Semin Anesth. 2007;26(3):126-32.
- 12. Gunawarda RH. Difficult laryngoscopy in cleft lip and palate surgery. Br J Anaesth. 1996;76(6):757–9.
- 13. Raj D, Luginbuehl I. Managing the difficult airway in the syndromic child. Contin Educ Anaesth Crit Care Pain. 2014;15(1):7–13.
- Stuart G, Ahmad N. Perioperative care of children with inherited metabolic disorders. Contin Educ Anaesth Crit Care Pain. 2011;11(2):62–8.
- 15. Gregory GE, Andripoulos DB. Gregory's pediatric anesthesia. 5th ed. Wiley-Blackwell: Chichester, UK; 2012.

- Inagawa G, Miwa T, Hiroki K. The change of difficult intubation with growth in a patient with Treacher Collins syndrome. Anesth Analg. 2004;99:1874.
- 17. Gross JB, Bachenberg KL, Benumof JL, Caplan RA, Connis RT, Coté CJ, et al. Practice guidelines for the perioperative management of patients with obstructive sleep apnea: A report by the American Society of Anesthesiologists Task Force on Perioperative Management of Patients with Obstructive Sleep Apnea. Anesthesiology. 2006;104(5):1081–93. Updated in: American Society of Anesthesiologists Task Force on Perioperative Management of patients with obstructive sleep apnea. Practice guidelines for the perioperative management of patients with obstructive sleep apnea: an updated report by the American Society of Anesthesiologists Task Force on Perioperative Management of patients with obstructive sleep apnea: an updated report by the American Society of Anesthesiologists Task Force on Perioperative Management of patients with obstructive sleep apnea. Anesthesiology. 2014;120(2):268–86
- Benumof JF. Obstructive sleep apnea in the adult obese patient: implications for airway management. J Clin Anesth. 2001;13(2):144–56.
- 19. Sher AE, Shprintzen RJ, Thorpy MJ. Endoscopic observations of obstructive sleep apnea in children with anomalous upper airways: predictive and therapeutic value. Int J Pediatr Otorhinolaryngol. 1986;11(2):135–46.
- Bath AP, Panarese A, Thevasagayam M, Bull PD. Paediatric subglottic stenosis. Clin Otolaryngol Allied Sci. 1999;24:117–21.
- 21. Eid EA. Anesthesia for subglottic stenosis in pediatrics. Saudi J Anaesth. 2009;3(2):77–82. https://doi.org/10.4103/1658-354X.57882.
- Hartnik CJ, Cotton RT. Congenital laryngeal anomalies: laryngeal atresia, stenosis, webs and clefts. Otolaryngol Clin N Am. 2000;33:1293–308.
- 23. Windsor A, Clements C, Jacobs I. Rare upper airway anomalies. Pedi Respir Rev. 2016;17:24-8.
- 24. Al Abri R, Javad H, Kumar S, Bharga D, Koul R, Al Futaisi A, et al. Congenital nasal pyriform aperture stenosis: first case report in Oman. Oman Med J. 2008;23(3):192–4.
- Benjamin B, Inglis A. Minor congenital laryngeal clefts: diagnosis and classification. Ann Otol Rhinol Laryngol. 1989;98:417–20.
- 26. Reza R, Rouillon I, Gilles R, Lin A, Nuss R, Denoyelle F, et al. The presentation and management of laryngeal cleft: a 10-year experience. Arch Otolaryngol Head Neck Surg. 2006;132(12):1335–41.
- Patel K, Gursale A, Chavan D, Sawant P. Anaesthesia challenges in Freeman-Sheldon syndrome. Indian J Anaesth. 2013;57(6):632–3.
- 28. Han I, Kim YS, Kim SW. Anesthetic experience of a patient with cri du chat syndrome. Korean J Anesthesiol. 2013;65(5):482–3.
- 29. Frerk CM. Predicting difficult intubation. Anaesthesia. 1991;46(12):1005-8.
- Haynes SR, Allsop JR, Gillies GW. Arterial oxygen saturation during induction of anesthesia and laryngeal mask insertion: prospective evaluation of four techniques. Br J Anaesth. 1992;68:519–22.
- Baraka AS, Taha SK, Aouad MT, El-Khatib MF, Kawkabani NI. Preoxygenation: comparison of maximal breathing and tidal volume breathing techniques. Anesthesiology. 1999;91:612–6.
- 32. Valentine SJ, Marjot R, Monk CR. Preoxygenation in the elderly: a comparison of the fourmaximal-breath and three-minute techniques. Anesth Analg. 1990;71:516–9.
- Gambee AM, Hertzka RE, Fisher DM. Preoxygenation techniques: comparison of three minutes and four breaths. Anesth Analg. 1987;66:468–70.
- Kheterphal S, Martin L, Shanks AM, Tremper KT. Prediction and outcomes of impossible mask ventilation: a review of 50,000 anesthetics. Anesthesiology. 2009;110:891–7.
- Langeron O, Masso E, Huraux C, Guggiari M, Bianchi A, Coriat P, et al. Prediction of difficult mask ventilation. Anesthesiology. 2000;92:1229–36.