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Airway Management and Intubation Techniques in Children with Craniofacial Anomalies

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Introduction

Patients with craniofacial abnormalities pose a significant challenge for those specialists charged with management of their airway. Congenital craniofacial anomalies often make laryngeal exposure difficult using conventional techniques. Special equipment, such as flexible fiber optic scopes, the video laryngoscope, rigid telescopes and bronchoscopes often must be utilized. Preoperatively careful planning is imperative. This involves having an adequate knowledge of the patient's condition and a thorough exam of the neck and airway including a flexible laryngoscopy. Well defined back up plans are key in achieving successful and safe airway management in patients with craniofacial anomalies.

Acute respiratory emergencies in patients with craniofacial anomalies have resulted in various pediatric institutions to designate these patients based on the difficulty of their airway exposure as possible "specialist intubation only patients "and mobilize special equipment and personnel to the bedside in case of an airway emergency. In our institution this system is called CART (Critical Airway Response Team). A CART activation brings a pediatric otolaryngologist, pediatric anesthesiologist, and operating room personnel to the bedside with an especially adapted airway cart carrying a variety of pediatric laryngoscopes, bronchoscopes, endotracheal tubes and fiber optic. In cases where the patient is stable enough that they can be transferred to the operating room this option is preferred because it provides a more control environment as well as the ability to use anesthetic gases.

It is advisable to delay intravenous anesthesia with muscle relaxant until endotracheal intubation is achieved. Until that time the patient should be kept spontaneously ventilating but sedated enough to undergo intubation using an inhaled gas anesthetic. A cooperative effort between the pediatric otolaryngologist, pediatric anesthesia is needed to establish and maintain a secure airway in these patients

Cleft Lip and Cleft Palate

Cleft lip and cleft palate are relatively common congenital anomalies that may confer an elevated level of difficulty with laryngoscopy and intubation. A large review by Xue et. al found that bilateral cleft lip and palate posed the greatest challenge in laryngoscopy compared to patients with unilateral cleft lip or simple cleft palate [1] . It has also been found that laryngoscopy is often more difficult in left sided cleft lip compared to right sided [1]. This is likely due to the right-handedness of most anesthesiologists. In patients with bilateral cleft palate, or left sided cleft lip and alveolus, the cleft may prevent appropriate placement of a laryngoscope. To optimize intubation of these patients it is recommended

that a senior anesthesiologist, with experience in intubating patients with cleft lip and palate, be involved. A shoulder roll should placed to put the cervical spine in gentle extension and the operating room nurse should be prepared to provide anterior laryngeal pressure. Wet gauze padding may be placed within the cleft palate to support advancement of the laryngoscope [1]. With these maneuvers the incidence of failed intubation should be <1%.

Hemifacial Microsomia

Also known as craniofacial micosomia, in these patients malformation of the first and second pharyngeal arches results in a hypoplasia of the craniofacial skeleton and soft tissues. It is the second most common craniofacial malformation after cleft lip and palate [2,3]. Patients may exhibit of spectrum of physical exam findings including varying degrees of microopthalmia, microtia, mandibular hypoplasia and malar hypoplasia. Abnormalities are usually unilateral but bilateral malformations are also possible [2,3]. Mandibular hypoplasia is the factor that creates the most significant challenge in intubation and airway management in these patients. When affected bilaterally an approach similar to that used in Robin Sequence patients should be utilized. Most unilaterally affected patients may be intubated over a rigid telescope [3].

To use this technique an appropriately sized endotracheal tube (ETT) is loaded over a rigid telescope. A shoulder roll should be place to put the patient in gentle cervical extension. A laryngoscope is used to attempt to visualize the larynx. The rigid telescope is passed the through the glottis into the trachea, it should be advanced until the carina is visualized. At this point the ETT may be advanced over the bronchoscope into the trachea. As the scope is removed the position of the ETT within the trachea should be confirmed. Otolaryngologist and anesthesiologist should be mindful that after these patients have corrective surgery on their mandible the difficulty of intubation may increase, due to scarring and fibrosis, and techniques used previously may no longer be effective [3].

Robin Sequence

Pediatric patients with Robin Sequence (RS) are notoriously difficult to intubate due to their congenital anatomic anomalies. RS is characterized by micrognathia, glossoptosis, and u-shaped cleft palate. Surgical intervention, in the form of mandibular distraction osteogenesis, is becoming increasingly popular. As such, a safe and effective method for perioperative airway management and intubation is needed. Nasotracheal intubation is preferred to remove the ETT from the surgical field. At the authors' institution fiberoptic transnasal intubation is routinely performed for RS patients. Successful intubation is aided by placement of a 2-0 silk suture through tongue to facilitate anterior retraction. At the same time, a videolaryngoscope is used by a second anesthesiologist to aid in elevation of the larynx. The videolaryngoscope also permits visualization of the airway during intubation by all members of the airway team. Another technique, described by Portnoy and Tatum, involves initially intubating the patient transorally through a laryngeal mask airway [4]. A second small ETT is passed through the nasopharynx and out the mouth. The smaller ETT is inserted into the larger ETT and secured with a securer. Aided by the cleft palate, the smaller ETT is then pulled back out

through the now which in turn moves the ETT with which the patient is currently intubated from transoral to transnasal. Caution must be taken during this step as to not dislodge the ETT from the airway. The anterior comissure scope provides good exposure of the larynx in patients with retrognathia and should always be available as part of the armamentarium. Sharma suggests that a central venous pressure guidewire be placed through the ETT to facilitate reintubation should accidental extubation occur during this step[5]. Although we do not have experience with this technique it stands to reason that it would be most helpful when the patient's anatomy prevents fiberoptic transnasal intubation. Robin sequence may occur in isolation or as part of a syndrome. Stickler Syndrome, Velocardiofacial Syndrome, and Treacher Collins all may exhibit this developmental sequence and as such the same difficulties with airway management should be expected.

Treacher Collins Syndrome

The craniofacial abnormalities that may be present in patients with Treacher Collins Syndrome include malar hypoplasia, mandibular hypoplasia and/or Robin sequence, downsloping palpebral fissures, coloboma of the lower lid, microstomia, microtia/atresia and hearing loss [6]. They often suffer from upper airway obstruction which may require tracheostomy. Anatomic abnormalities make mask ventilation and intubation difficult in these patients. A method which has proven effective in Treacher Collins patients is fiberoptic intubation through a laryngeal mask airway (LMA) [7]. For this technique first, an LMA is placed. A fiberoptic bronchoscope is loaded with an appropriately sized endotracheal tube. The fiberoptic scope is passed though the LMA, the glottis is visualized and the flexiblebronchoscope is passed through the true vocal fold into the subglottis. The bronchoscope should be advanced until the carina is visualized. Next the ETT is advanced over the flexible bronchoscope, through the LMA, into the trachea. As the bronchoscope is withdrawn to the tip of the ETT to confirm its position within trachea. Once confirmed, the bronchoscope, and then the LMA, may be carefully removed.

Down Syndrome

Patient's affected by Down Syndrome, also known as trisomy 21, often have macroglossia, a narrowed nasopharynx, and are prone to atlantoaxial subluxation [7]. As such, significant manipulation of the cervical spine during intubation is to be avoided. Obtaining preoperative x-rays may be obtained to better assess risk. Additionally, use of an endotracheal two sizes smaller than would be predicted based on age should be considered as Down Syndrome typically have a narrowed subglottis and are prone to developing acquired subglottic stenosis [8].

Apert's Syndrome, Crouzon Syndrome and Pfeiffer's Syndrome

Apert's, Crouzon and Pfeiffer's syndromes have in common maxillary hypoplasia, high arched palate, narrowed nasopharynx, and craniofacial synostosis, which refers to premature closure of one or more cranial suture [2,6]. It may occur in isolation or as part of a syndrome the most popular of which are Apert's, Crouzon, and Pfeiffer's [2,6]. Challenges in airway management in these patient begins with mask ventilation. The hypoplastic midface causes a poor mask fit making it difficult to obtain a good seal [2]. Mask ventilation is aided by holding

the mouth open and or placing an oral airway, as the abnormal anatomy in these patients creates a significant amount of upper airway obstruction when the mouth is closed [2]. Typically, laryngeal exposure and intubation usually does not pose a significant problem. [2] It is important to remember that if a tracheotomy is required in these patients there is the possibility of a tracheal sleeve which may complicate identifying the tracheal and laryngeal landmarks.

Beckwith-Wiedemann Syndrome

Beckwith-Wiedemann Syndrome is characterized by macroglossia, exomphalos, gigantism, and hyperplasia of the kidney's and pancreas [9]. Macroglossia may cause significant upper airway obstruction and poses a challenge during intubation. At times these patients are unable to be mask ventilated as closure of the mouth causes near total upper airway obstruction [2].Fiberoptic bronchoscope guided nasotracheal intubation should be utilized in these patients who are undergoing surgical intervention for macoglossia. Alternatively, use of a videolaryngoscope can aid laryngeal visualization around the enlarged tongue. [10]

Klippel-Feil Syndrome

The distinguishing feature of patients with Klippel-Feil Syndrome (KFS) is fusion of two or more cervical vertebrae [11].Other characteristics include a short neck, low posterior hairline, scoliosis, and decreased neck range of motion [2, 11]. Although many KFS patients will be intubatable with conventional laryngoscopy techniques manipulation of the cervical spine should be avoided to prevent spinal cord injury [2,12].

Mucopolysaccharidosis

Mucopolysaccharisosis is a spectrum of disorders in which absence of a metabolic enzyme results in deposition of mucopolysacchrides in multiple organ systems including the upper airway [13]. Progressive deposition of glycosaminoglycans in the soft tissue of the throat and trachea is thought to be responsible for the airway dysfunction and obstruction, which characterize the syndrome. Other physical characteristics, including rib anomalies, abdominal organ enlargement, short neck and immobile jaw, further contribute to the respiratory problems [13]. Options for intubation for those in whom conventional laryngosocopy fails include fiberoptic intubation through a laryngeal mask airway, use of a videolaryngoscope or intubation over a rigid bronchoscope [14]. When the patients present in their teen years with significant facial and body deformities these can be the most challenging of airways and thourough planning is required to achieve an airway. Due to their short neck, and chest , facial deformities and tracheal mucopolysacchrides deposits an emergency tracheotomy is not a straight forward endeavor.

Freeman-Sheldon Syndrome

Freeman-Sheldon Syndrome is also known as whistling baby syndrome and craniocarpotarsal dysplasia. It is characterized by significant microstomia, muscular contractures, camptodactyly with ulnar deviation and talipes equinovarus [9]. These patients are prone to malignant hyperthermia thus succinycholine and inhaled anesthetics are contraindicated. Limited neck range of motion due to contractures,

along with microstomia, make laryngosocpy and intubation difficult. Severe microstomia may also limit placement of laryngeal mask airway[15]. Success using awake, transnasal fiberoptic intubation has been reported [15].

Conclusion

Patients with craniofacial abnormalities often pose a significant challenge in airway management. Preoperative planning is paramount and it is important to have multiple options available for airway intervention. A team approach between pediatric otolaryngologists and pediatric anesthesiologists is needed. Finally, one should always be prepared to perform tracheostomy as a life saving measure with other non-invasive techniques fail to secure an airway. Parents should be counseled accordingly preoperatively.

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