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Emergent perioperative care of a patient with Apert's syndrome, increased intracranial pressure and a difficult airway

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Key points

1. During resuscitation and emergency care, airway management with endotracheal intubation remains a primary therapy to provide effective oxygenation and ventilation that is necessary for resuscitative efforts to be successful.
2. Following traumatic brain injury, significant secondary injury may result from the release of inflammatory and excitotoxic medicators or physiological perturbations including hypoxemia, hypercarbia, hypocarbia, and hypotension.
3. In the clinical scenario of intracranial hypertension with midline shift on neuroimaging, appropriate hemodynamic conditions must be provided during rapid sequence intubation since direct laryngoscopy is a potent stimulus that may result in significant increases in ICP.
4. Although the data are conflicting, succinylcholine has been reported to increase intracranial pressure in the setting of TBI and may therefore be relatively contraindicated.

Abstract

Increased intracranial pressure (ICP) is a medical emergency requiring immediate therapy which often includes airway control and endotracheal intubation. Given the need to prevent further increases in ICP, sedation and neuromuscular blockade is frequently used to facilitate endotracheal intubation. However, concerns regarding the ease of endotracheal intubation may complicate the decision process regarding the pharmacologic management of endotracheal intubation. The authors present a 22-year old patient with Apert's syndrome and increased ICP who required an emergent craniotomy for the treatment of an expanding intracerebral hematoma. Airway management was complicated by a history of a previous difficult intubation. The potential options for anesthetic induction and endotracheal intubation are discussed in the context of competing concerns in this patient with an intracranial mass lesion leading to increased ICP, a history of a difficult airway, and the potential aspiration risk of a full stomach in the emergent setting.

Keywords: Traumatic brain injury, Apert's syndrome, difficult airway, increased intracranial pressure, rapid sequence intubation, cricoid pressure.

Introduction

During resuscitation and emergency care, airway management with endotracheal intubation remains a primary

therapy to provide effective oxygenation and ventilation that is necessary for resuscitative efforts to be successful.[1,2] In the trauma setting, various factors may complicate airway management including pre-existing conditions that may make routine airway management difficult as well as acquired conditions related to the traumatic event such as head trauma with the potential for increased intracranial pressure (ICP). Furthermore, several factors including a depressed level of consciousness and emergency surgery with a full stomach may increase the risk of aspiration, requiring consideration of a rapid sequence intubation (RSI).[3,4] The authors present a 22-year old patient with Apert's syndrome with a history of a previous difficult endotracheal intubation who required an emergent craniotomy for the treatment of an expanding intracerebral hematoma. The decision process regarding the technique and pharmacologic management of airway management was complicated by increased ICP in the setting of emergency surgery with a full stomach. The potential options for anesthetic induction and airway management are discussed.

Case report

Institutional Review Board approval for single case reports is not required by Nationwide Children's Hospital (Columbus, Ohio). The patient was a 22-year old, 62 kilogram male with a history of Apert's syndrome. His surgical history included multiple craniofacial surgical procedures including midface advancement. Additional co-morbid conditions included attention deficit disorder, developmental delay, and an unspecified seizure disorder. The patient was transported to the emergency department after he fell from his bike and struck his head on a concrete surface. He was not wearing a helmet. No loss of consciousness was noted, but there was one episode of emesis after the event. On evaluation by the emergency medicine physician, the patient was noted to be lethargic but arousable. He complained of head pain, but denied neck pain, shortness of breath, abdominal pain, nausea, or extremity pain. The patient had a Glasgow Coma Scale (GCS) of 15. On physical examination, there was a head contusion, left hemotympanum, and unequal pupils. Imaging revealed a large left epidural hematoma measuring 5.6 x 5.1 x 3.6 cm, a left parietal-occipital subdural hematoma, left temporal/parietal/occipital non-depressed fracture, and a 13 mm left to right midline shift. The emergency medicine physician contacted the pediatric anesthesiologist and otolaryngologist to alert the team to the potential for difficult airway management after obtaining a history from the parents of a previous need for fiberoptic bronchoscopy for endotracheal intubation. The patient's level of consciousness deteriorated and periodic episodes of bradycardia were noted in the ED. His respirations were irregular, he was hypertensive to 148/109, he had a new complaint of left ear pain, and his GCS remained at 15 though his right pupil was 3mm and brisk while his left pupil was 4mm and sluggish. The decision was made to perform an emergent left craniotomy for evacuation of hematoma. He was transported to the operating room where a difficult airway cart had already been placed. Routine American Society of Anesthesiologists' (ASA) monitors were applied while the patient was placed in a 20-30^{o} head-up tilt. An otolaryngologist was present in the operating room with rigid bronchoscopy and emergency tracheostomy equipment available. The patient was pre-oxygenation with 100% oxygen. After the administration of lidocaine (1 mg/kg), anesthesia was induced with intravenous propofol (2 mg/kg) and fentanyl (2 µg/kg) which were administered through a pre-existing intravenous cannula. Endotracheal intubation was facilitated with rocuronium (1 mg/kg) and an indirect videolaryngoscope (Glidescope®, Verathon Inc, Bothwell, WA) was used to place an endotracheal tube. Cricoid pressure was not applied. Oxygen saturation remained at 99-100% throughout the endotracheal intubation process. There was no change in his blood pressure. Following anesthetic induction and endotracheal intubation, an arterial cannula was placed. Maintenance anesthesia consisted of inhaled isoflurane (exhaled concentration of 0.8-1.25%). Neuromuscular blockade was

achieved with periodic doses of rocuronium titrated utilizing to train-of-four monitoring. During the procedure, phenylephrine (total of 2 µg/kg) and 5% albumin (250 mL) were administered to maintain the systolic blood pressure (sBP) ≥ 90 mmHg. The surgical procedure lasted 63 minutes. Estimated blood loss was 400 mL and intraoperative fluids included 1000 mL of normal saline. At the completion of the procedure, a propofol infusion (150 µg/kg/min) was initiated and the patient was transported for a CT scan of the head to confirm satisfactory decompression of the hematoma. Following the completion of imaging, the patient was transported to the Pediatric ICU for further management. He was extubated to room air on postoperative day 1. Extubation was tolerated without difficulty and performed with ENT and ICU teams at the bedside. At the time of extubation the patient was hemodynamic stable as an arterial line measured blood pressure at 139/60, pulse was 60, respiratory rate was 16, and oxygen saturation was initially 93% but rose to 100% over the next hour. The postoperative course was remarkable for a continued complaint of blurry vision. He was discharged home on postoperative day 4. At the patient's first postoperative visit, approximately two weeks later, the patient and his mother reported that his post-concussive symptoms were improving but the patient continued to experience daily headaches. Two months postoperatively the patient still complained of a few mild headaches that he attributes to his new glasses, but feels that he is otherwise back to baseline.

Discussion

Various factors may complicate the perioperative care of a patient with traumatic brain injury (TBI).[5] In addition to the primary injury from the impact, there may be significant secondary injury caused by the release of inflammatory and excitotoxic mediators, which result in increasing edema, alterations in ICP, and a further reduction of cerebral perfusion pressure (CPP).[6,7] Secondary injury may also result from physiological perturbations including hypoxemia, hypercarbia, hypocarbia, and hypotension, further emphasizing the importance of swift and effective airway control throughout the perioperative period.[8]

Epidural hematomas usually occur as a result of a tear to the middle meningeal artery, often associated with a skull fracture. As noted in our patient, there may be a period of lucidity following the initial traumatic event followed by a rapid deterioration of mental status. Venous hemorrhage may also be an etiology when shearing forces tear bridging veins. Epidural hematomas increase ICP through mass effect by increasing intracranial volume. This may be accentuated by cerebral edema that occurs in the surrounding tissue related to the initial traumatic event and the release of inflammatory mediators. These processes may occur rapidly and cause herniation leading to compression of neural and vascular structures resulting in focal or global ischemia. In our patient, the clinical scenario of a rapid decline in mental status with an expanding epidural hematoma necessitated emergent transport to the operating room for surgical treatment. The clinical factors that impacted the clinical decision making process included increased ICP, a history of a previous difficult intubation, and a full stomach.

In the clinical scenario of intracranial hypertension with evidence of midline shift on CT scan, appropriate hemodynamic conditions must be provided during RSI since direct laryngoscopy is a potent stimulus that may result in significant increases in ICP.[9] While avoiding hypertension is essential, it is equally important to avoid hypotension as a decrease in mean arterial pressure especially in the setting of increased ICP may compromise cerebral perfusion pressure (CPP) leading to ischemia, particularly in the penumbra zone surrounding the hematoma. Another concern in this scenario is the effect of the anesthetic agent on cerebral metabolic rate for oxygen ($CMRO_2$), cerebral blood volume (CBV), and ICP. Therefore, the choice of anesthetic induction agents and neuromuscular blocking agents is of the utmost importance.

Agents commonly used to induce anesthesia and provide sedation during endotracheal intubation include propofol, thiopental, etomidate, and ketamine. Propofol activates $GABA_A$ receptors, inhibits the NMDA receptor, and has been shown to significantly reduce neuronal damage induced by ischemia.[10] More importantly, as it decreases, it results in reflex cerebral vasoconstriction, a decrease in CBV and a concomitant decrease in ICP.[11] A similar protective effect during periods of ischemia and a decrease of ICP occurs following administration of the short-acting barbiturate, thiopental.[12,13] However, this agent is no longer available for clinical use in the United States. Although these agents have beneficial effects on CNS dynamics, they may decrease MAP through direct effects on myocardial contractility, a decrease in systemic vascular resistance, and vasodilatation.[14,15] This response may be exaggerated in the setting of hypovolemia or with co-morbid cardiovascular disorders. In those setting, alternative agents should be considered as thiopental and propofol may not be appropriate given their vasodilatory and negative inotropic effects.[14,15]

Ketamine and etomidate are alternative induction agents that generally have minimal hemodynamic effects, making them useful in patients with trauma or co-morbid cardiovascular disorders. However, there are additional issues that should be considered, particularly in patients with traumatic brain injury. Etomidate has a limited effect on MAP and decreases cerebral metabolic rate for oxygen ($CMRO_2$), decreasing ICP, and thereby maintaining CPP.[16] Etomidate affects the endogenous production of corticosteroids through an inhibitory effect on the enzyme 11β-hydroxylase. The clinical impact of this effect remains debatable and our center continues to use etomidate in various clinical scenarios.[17,18]

Ketamine also remains a popular anesthetic induction agent in the trauma setting as it generally maintains or increases heart rate and blood pressure due to the release of endogenous catecholamines.[19] Concerns about ketamine increasing ICP and questions regarding its contraindication in patients with TBI are based on older studies with few subjects and multiple confounding factors.[20] More recent evidence has demonstrated that ketamine may actually decrease ICP while maintaining MAP and therefore CPP.[21] There may be theoretical concerns regarding an exaggerated increase in MAP in patients with TBI and altered cerebral autoregulation. Although ketamine generally increases MAP, this is an indirect response through the release of endogenous catecholamines. As ketamine has direct negative inotropic effects, cardiovascular collapse has rarely been reported following its administration to patients who have depleted their endogenous catecholamine stores.[22] Given our patients stable hemodynamic status and the need for an agent that would be helpful in controlling $CMRO_2$ and ICP, we chose to induce anesthesia with propofol.

Neuromuscular blockade provides optimal conditions for laryngoscopy and endotracheal intubation, which must be accomplished quickly for protection from aspiration. The most common neuromuscular blocking agents employed for RSI are succinylcholine and rocuronium.[23] Succinylcholine, a depolarizing neuromuscular blocking agent, is popular because of its rapid onset of action and short duration of effect. It is associated with a number of side effects ranging from myalgias to life-threatening hyperkalemia or malignant hyperthermia.[24,25] Although the data are conflicting, succinylcholine has been reported to increase intracranial pressure in the setting of TBI.[26,27] Rocuronium, a non-depolarizing neuromuscular blocking agent, has a comparable onset time when compared to succinylcholine with minimal hemodynamic effects and no effect on ICP.[28] When compared to succinylcholine, the conditions for endotracheal intubation are also comparable.[29] Given the above mentioned issues, we chose to use rocuronium for RSI in our patient.

Rapid sequence induction (RSI) is a technique employed to achieve endotracheal intubation in a manner that minimizes the risk of pulmonary aspiration. The key components of RSI include preoxygenation, the in-

travenous induction of anesthesia, and the administration of a neuromuscular blocking agent that achieves rapid paralysis. In the classic teaching RSI generally does not include the provision of bag-valve-mask ventilation although this is sometimes included using the descriptive term "modified RSI".[3] Additionally, cricoid pressure is generally included in RSI. The use of cricoid pressure to control the passive regurgitation of gastric contents during anesthetic induction was first suggested by Dr. Brian Sellick. The cricoid cartilage is the only circular, cartilaginous structure in the upper airway, which permits occlusion of the esophagus.[30] The purpose behind properly performing cricoid pressure is to theoretically prevent the passive regurgitation of stomach content, but there is no evidence-based medicine to support its efficacy during RSI.[31] Moreover, it has been demonstrated that, even when Sellick's maneuver is performed correctly, the use of too much force can distort the airway.[32] Furthermore, the literature has demonstrated that in pediatric patients, the application of cricoid pressure is frequently not correctly performed.[33] The theoretical risk being that improper placement may result in jugular venous occlusion thereby increasing ICP. For these reasons, we chose to perform RSI without cricoid pressure during the induction sequence in our patient with elevated ICP and a potentially difficult airway.

The perioperative care was further complicated by the history of difficulties with airway management during a previous anesthetic encounter. Apert's syndrome is notable for craniosynostosis, midface hypoplasia, and syndactyly. Syndromic features that may be obstacles to successful airway management include reduced nasopharyngeal dimensions and choanal atresia due to maxillary hypoplasia, fusion of the cervical spine (usually at C5-C6), and subglottic stenosis due to fused tracheal rings (bamboo trachea). In a known or potentially difficult airway there are two decisions to be made: route of intubation and level of consciousness.[34-36] Pediatric patients and patients with an altered level of consciousness typically are not cooperative, which makes them poor candidates for awake or sedated intubation. Furthermore, such techniques are generally contraindicated with altered intracranial compliance. One approach is to induce general anesthesia and maintain spontaneous ventilation with the incremental administration of sevoflurane. However, given its effects on ICP and the need to rapidly control the airway in our patient with altered mental status, this was not an acceptable option. RSI is the alternative choice in this situation, but the administration of a neuromuscular blocking agent will rapidly result in hypoxemia if ventilation cannot be established. The ability to properly secure a difficult airway using advanced airway devices minimizes the likelihood of the cannot intubate-cannot ventilate (CICV) emergency scenario.[37,38] Some of these devices include indirect videolaryngoscopy (e.g., GlideScope, C-MAC), laryngeal mark airway as a conduit for fiberoptic intubation, nasal or oral fiberoptic bronchoscopic intubation, and optical stylets (Bonfils). In addition to these devices, we would suggest having the needed support in the operating room including other pediatric anesthesiologists and a pediatric otorhinolaryngologist in the event that a surgical airway is needed. The possibility of such was discussed with our patient's parents prior to transport to the operating room. Fortunately, the airway was easily managed with an indirect laryngoscopy and endotracheal intubation accomplished on the first attempt.

Conclusions

This case illustrates several important considerations in the management of the patient with TBI and a history of a difficult airway.

1. Effective interdisciplinary communication among emergency medicine, otolaryngology, and anesthesiology personnel was essential to prepare personnel and equipment for the care of this patient.
2. Consideration of the multiple anesthetic issues including TBI with increased ICP, full stomach and aspiration risk, potential difficult airway in a patient

with known risk factors (Apert's syndrome), allowed prioritization of management goals.

3. There was the selection of appropriate pharmacologic agents to minimize hemodynamic perturbations, maximize neuroprotective effects with lowering of ICP while permitting rapid airway control.
4. There was emergency equipment with airway adjuncts and surgical personnel if airway instrumentation could not be achieved by videolaryngoscopy.

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