MH, difficult airway, now a pneumothorax- should we cancel the case??

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Objectives:

1. Review the induction options in syndromic infants with very difficult airways and a family history of malignant hyperthermia.
2. Identify the risk factors of intraoperative tension pneumothoraces in infants; discuss clinical signs and symptoms that lead to the diagnosis and the management.
3. Discuss at what point a semi-elective case should be postponed for further stabilization after a difficult and long induction period resulting in complications versus proceeding with surgery.
4. Discuss the location options and safety considerations when asked by the PICU to assist in extubations in difficult airway patients.

Case History:

A 3 month old with a history of Emanuel Syndrome, cardiac atrial septal defect, cleft palate, micrognathia, conductive hearing lose, gastroesophageal reflex, Dandy Walker variant, and mild hydrocephalus was transferred from an outside hospital for definite management of mandibular reconstruction. She had a history of severe obstructive sleep apnea with apnea hypopnea index of 35, hypoventilation and desaturations to the 60s.

Questions:

What is Emanuel Syndrome? Are there any anesthetic implications of Emanuel Syndrome? What are your main concerns regarding her history? Is there anything else you would like to know? What is the safest way to induce general anesthesia? Are you anticipating a difficult airway?

Case History, cont.

Mother states a history of malignant hyperthermia in her brother. The baby has peripheral intravenous access in the PICU.
Questions:

What is malignant hyperthermia? Does this change your anesthetic plan? Would you investigate the family history further? Would you pursue a muscle biopsy for definitive diagnosis on this infant before proceeding with the scheduled surgery?

You decide to proceed with the scheduled surgery.

How will you induce? How important is it to keep the baby spontaneously breathing? What agents would you use? Would you try to intubate this baby awake? Fiberoptic? Nasal? How can you oxygenate a baby during a difficult fiberoptic intubation?

Case History, cont.

The baby arrives in the OR and we flush the IV and it is determined to be non-functional. After significant struggle, PIV access is obtained and a dexmedetomidine bolus is given followed by infusion. Small doses of propfol were also administered to facilitate nasal fiberoptic intubation, which was eventually successful. After nasal endotracheal tube was secured, additional venous access was attempted because existing PIV was very positional and tenious. During this time baby became apneic and difficult to ventilate through nasal ETT. Baby desaturated and began to become bradycardic.

Questions:

What could be causing the difficulty in ventilation? What would be your next step? If PIV was working? If PIV was not working? Give an IM medication? Would you take out the nasal ETT?

Case History, cont.

IM ketamine was given and nasal ETT removed and LMA placed, and ventilation became possible. Once baby was stabilized, intubation was once again attempted with fiberoptic scope, this time oral endotracheal tube was placed through the LMA and then LMA removed. Because of the nature of the surgery (mandibular distraction), oral ETT was then exchanged over a cook tube exchanger for oral rae ETT and sutured by surgeons. At the same time, left subclavian access was obtained. During the time, breath sounds were noted to be greater on the left versus right side. Baby was also hypotensive.

Questions:

Was it necessary to exchange the regular ETT for an oral rae for this procedure? What could be some causes of unequal breath sounds? The hypotension? How would you treat it? Could her ASD be contributing?
Case History, cont.

Glucose, albumin, and small doses of epinephrine were given. A chest xray was then obtained in the OR because of the newly placed subclavian line which revealed a right sided tension pneumothorax. Right sided needle decompression was performed and pediatric surgery consulted for chest tube placement.

Questions:

Would you have done a needle decompression before obtaining a chest xray if breath sounds were unequal and the baby was hypotensive? What could have caused the tension pneumothorax? Is this common in babies? What are the clinical signs and symptoms? How do you perform a needed decompression? Now that the induction period has been prolonged and complicated, would you post pone this case until baby stabilized in PICU? Is this an urgent case? Would you leave the oral rae ETT in or exchange it again for a regular ETT for the PICU? What would you tell the parents?

Case History, cont.

Decision was made with the surgical team to postpone the case. Parents were informed. An arterial line was placed and the baby was kept intubated and taken to PICU with a sutured oral rae ETT. Baby was kept intubated until two days later when the procedure was completed uneventfully.

Questions:

It is now a few days later and your PICU colleagues ask for your assistance in extubating this baby. Where would you do this? PICU? OR? What is the safest technique? Would you request surgical presence? Use an airway exchange catheter? What are your concerns? How would you assess if the baby is ready for extubation?

Case History, cont.

Baby was brought to OR from PICU for extubation on dexmedetomidine infusion. Infusion was stopped, secretions were suctioned, nares were sprayed with oxymetazoline. The oral RAE ett was then exchanged for a 3.0 ETT over an 8 french cook tube exchanger, and once it was determined the baby could cry around the ETT, patient was extubated. Racemic epinephrine treatment was given.
Discussion Outline:

Emanuel Syndrome is a rare anomaly associated with multiple systemic malformations including facial dysmorphism, micrognathia, congenital heart disease, renal anomalies, hypotonia, failure to thrive, GERD, and mental retardation. It is caused by the presence of a supernumerary derivative consisting of portions of chromosome 11 and 22. The anesthetic management of these patients is challenging because of micrognathia causing upper airway obstruction and difficult intubation.

Inhalation induction using sevoflurane and oxygen is usually most desirable in pediatric patients with anticipated difficult airways; however, a family history of malignant hyperthermia presents a challenge in this case. Malignant hyperthermia is a rare disorder of calcium metabolism in skeletal muscle. It is characterized by evidence of a hypermetabolic state and can result in death if untreated. It may be difficult to obtain more history or a definite diagnosis with further testing in urgent situations when a case cannot be delayed long enough to go to a specific MH testing center in the country. In addition, for a child a diagnostic muscle biopsy may also require anesthesia.

Although securing the difficult pediatric airway after the induction of general anesthesia is an acceptable technique and safe option for most patients, there can be disadvantages, especially if unable to ventilate the child because of airway collapse and obstruction. “Non-asleep” airway management has also been described using topicalization, antisyialogogue, and a combination of midazolam and ketamine. Syndromes with craniofacial abnormalities can be a real challenge in terms of airway management and effective preparation, including a surgical airway team, is of utmost importance.

An intraoperative tension pneumothorax can quickly cause cardiovascular collapse. Clinical signs include hypotension, bradycardia, desaturation, increased airway pressures, and unequal breath sounds. Iatrogenic causes include resuscitation measures, including bag ventilation, mechanical ventilation, and traumatic intubation. Pneumothorax can also be an underappreciated complication with an airway exchange catheter, almost 11% in literature. Trauma can be caused by the device to the tracheobronchial tree, the risk which is increased with smaller airway exchange catheters. Needle decompression should be performed emergently until a chest tube can be placed.

Extubations of difficult pediatric airways require just as much attention and preparation as intubations. These are usually safest performed in the operating room after optimizing the child for extubation in the PICU. The presence of an ENT surgeon skilled in performing surgical airways and all re-intubation equipment is essential.
References:


