CRNA Symposium PBLD Table #: 1

Hypokalemic Periodic Paralysis at the Ambulatory Surgery Center

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Objectives:

1. Identify the different types of familial periodic paralysis
2. Discuss the anesthetic management for a patient with HOKPP
3. Discuss the suitability of such case and other high risk patients at an ASC
4. Review the genetic disorders associated with malignant hyperthermia

Case History:

A 5 year old male was scheduled for a right orchiopexy and inguinal hernia repair. Preoperative screening revealed a strong family history of hypokalemic periodic paralysis. The patient's father, sister, and paternal uncle were all diagnosed with HOKPP and were symptomatic. Genetic testing was done on the entire family prior to the patient's surgery and 4 out of 5 of the children tested positive. The genetic test confirmed the patient was positive for the familial CACNA1S R528H mutation.

Questions:

What is Hypokalemic Periodic Paralysis? What are the other Familial Periodic Paralyses? What are some differential diagnoses of HPP? What are the causes of hypokalemia? What are the manifestations of HOKPP? At what age are patients usually symptomatic? What are preoperative considerations for such a case?

Case History:

The patient was scheduled in our ASC.

Questions:

Is this case appropriate for an ASC? What are your concerns? Are there any risk factors or predictors associated with hospital admission in pediatric ambulatory surgery? What are the factors that determine suitability of high risk pediatric patients for an ASC? Do you need any work up before you proceed?

Case History (preoperative):

Preoperatively, a 12 lead EKG and serum potassium level were obtained which were both within normal limits. The patient was premedicated with 0.5 mg/kg oral midazolam with good effect.

Questions:

What are the triggers of HOKPP? What can you do to reduce the potential triggers? What are the risks and possible complications that should be discussed with the parents? What questions would you like to ask the parents in addition to the normal preoperative interview? Is there an association with HOKPP and malignant hyperthermia? What other conditions have a definitive association with MH? How can you prepare your new generation anesthesia work station for MH susceptible patients? Is there any difference from old generation? Is there any difference between different brands of anesthesia machines?

Case History (intraoperative):

The patient was brought to the OR with no IV in place.

Questions:

How would you induce this patient? How can you maintain the anesthesia during this case? If you decide to intubate, would you use a neuromuscular blocking agent? What are the potential issues with a depolarizing neuromuscular blocker versus a non-depolarizing?

Case History (intraoperative):

The patient was induced with nitrous oxide and oxygen in the operating room and a peripheral intravenous line was placed. Propofol 2.5 mg/kg was given, and the airway was secured by inserting a laryngeal mask airway. A caudal block was then placed for pain management. Anesthesia was maintained with a propofol infusion at a rate of 250 mcg/kg/min. Lactated ringers was used as maintenance fluid. A forced air warmer was used to avoid hypothermia.

Questions:

Is there a contraindication for local anesthesia in HOKPP patients? What is the potential benefit of regional anesthesia in patients with HOKPP? Would you use LA with epinephrine or without?

Case History (intraoperative):

You decide to check your potassium level intraoperatively so you send a venous blood gas and the K=2.8 mmol/dl

Questions:

What are the causes of hypokalemia intraoperatively? What are other manifestations of intraoperative hypokalemia? Would you treat hypokalemia? How would you mange intraoperative hypokalemia?

Case History:

Surgery and the intraoperative course proceeded uneventfully, and the patient was extubated deep and transferred to the postanesthesia care unit.

Questions:

How long do you want to observe this patient postoperatively? Do you want to admit patient overnight? What are the potential postoperative complications in a patient with HOKPP? What laboratory data would you order and when? What monitors would you want to remain on if kept as an inpatient?

Case History (postoperative):

The patient was observed for one hour in the postoperative care unit. A serum potassium was drawn postoperatively and the morning after surgery with no change in the potassium level from baseline. The patient did well and was discharged home the day after surgery.

Discussion:

Familial hypokalemic periodic paralysis (HOKPP) is an autosomal dominant disorder first described in 1882.[1] Anesthetic considerations for a patient with HOKPP focus on avoiding triggers and medications that cause a shift of potassium. Epidural/caudal analgesia has been shown to minimize fluctuations in serum potassium levels by reducing hyperventilation related to pain and lowering serum catecholamine levels which can be helpful in this patient population.[2] While there are no definitive reports of malignant hyperthermia (MH) developing in patients with HOKPP, the mutation for the two defects can be located on the same gene and recommendations leave it to the discretion of the anesthesia provider whether or not to avoid MH triggers.[3]

References:

1. Ellis, FR. Inherited Muscle Disease. BJA 1980;52: 153-164.
2. Viscomi, CM, Ptacek, LJ, Dudley, D. Anesthetic Management of Familial Hypokalemic Periodic Paralysis During Parturition. AA 1999;88:1081-82.
3. Parness, J, Bandschapp, O, Girard, T. The Myotonias and Susceptibility to Malignant Hyperthermia. AA 2009;109:1054-64.