# [NM-254] Anesthestic Management of a Pediatric Patient with Familial Hypokalemic Periodic Paralysis in an Ambulatory Surgery Center

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Introduction:
Familial hypokalemic periodic paralysis (HOKPP) is an autosomal dominant disorder first described in 1882.1 It is characterized by recurrent attacks of skeletal muscle weakness and flaccid paralysis with associated hypokalemia. Attacks are precipitated by multiple factors including stress, hypothermia, and carbohydrate load.
Case Report:
We present a case report of a 5 year old male who was scheduled for a right orchiopexy and inguinal hernia repair in our Ambulatory Surgery Center. Preoperative screening revealed a family history of hypokalemic periodic paralysis. The patient's father, sister, and paternal uncle were all diagnosed with HOKPP. The genetic test confirmed the patient was positive for the familial CACNA1S R528H mutation. Although the patient had never experienced any symptoms related to his diagnosis, a non-triggering anesthetic plan was chosen to be given at our Ambulatory Surgery Center (ASC).
Preoperatively, a 12 lead EKG and serum potassium level were obtained which were both within normal limits. The patient was scheduled as the first case of the day in our ASC. The patient was premedicated with 0.5 mg/kg oral midazolam with good effect. 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. Surgery and the intraoperative course proceeded uneventfully, and the patient was extubated deep and transferred to the postanesthesia care unit.
Discussion:
Anesthetic considerations for a patient with HOKPP focus on avoiding triggers and medications that cause a shift of potassium. Minimizing stress, maintaining body temperature, and avoiding glucose containing intravenous solutions and long acting neuromuscular blockers are all important. 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 However, the sympathomimetic effect of epinephrine containing local anesthetics may also precipitate hypokalemia. 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 Knowledge about HOKPP and its anesthetic implications allowed us to manage this patient safely in an ambulatory surgical setting.
References:
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3. Parness, J, Bandschapp, O, Girard, T. The Myotonias and Susceptibility to Malignant Hyperthermia. AA 2009;109:1054-64.

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