Anesthetic Management of Pediatric Patient with Familial Periodic Paralysis
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INTRODUCTION

Hypokalemic periodic paralysis (HPP) is an autosomal dominant disorder characterized by recurrent episodes of reversible flaccid paralysis with concomitant hypokalemia. It is the most common form of primary periodic paralysis with symptoms beginning in the first or second decades of life. We present a case report of an 11 year old Marshallese boy with HPP who was admitted at our institute for laparoscopic appendectomy under general anesthesia.

CASE REPORTS

An 11 year old, 36 Kg Marshallese boy presented to an outside hospital with right lower quadrant pain and diarrhea. Patient had known history of HPP and a computerized tomography at outside hospital revealed acute inflammation of his appendix. He was transferred to Arkansas Children’s Hospital for further medical management. His medications on transfer included acetazolamide 125mg orally twice daily and potassium 80 millequivalents orally three times a day. His admitting serum potassium was 3.4meq. Six hours later he was brought to operating room for laparoscopic appendectomy. Prior to induction of anesthesia serum potassium was checked in the operating room and a lab value of serum potassium was 2.2meq reported. Patient did not have any symptoms of muscle weakness or respiratory distress but decision was made to postpone the surgery and correct his serum potassium. He was admitted to pediatric intensive care unit and aggressive but cautious intravascular (IV) potassium replacement was initiated. Following twelve hours of ICU stay with IV potassium chloride replacement and serial serum potassium measurements, his serum potassium stabilized and he was rescheduled for the surgery. On arrival to operating room standard American society of anesthesiology monitors were applied. His serum potassium pre induction this time was 3.4meq. Propofol 200milligrams with Fentanyl 35micrograms intravenously were used for induction of anesthesia and an endotracheal tube 5.5mm was placed to secure the airway. Target controlled propofol and remifentanil infusions were used for the maintenance of anesthesia during the surgery. No muscle relaxants were administered at any time for induction of anesthesia or during the surgery. At the end of the appendectomy, propofol and remifentanil infusions were stopped. Patient regained complete strength and was extubated in the operating room uneventfully. On post-operative day three the patient was discharged home in stable condition.

DISCUSSION

HPP is a rare genetic disorder with an autosomal dominant inheritance. Careful peri-operative care of patients with HPP, include the close control of plasma potassium levels, adequate potassium replacement , avoidance of triggers for HPP (reduction of anxiety, avoidance of large glucose and salt loads, carbohydrate-poor diet, maintenance of body temperature and acid–base balance) and careful use or avoidance of neuromuscular blocking drugs. In conclusion, we report the anesthetic management of a patient with HPP using total intravenous anesthesia. Maintenance of adequate levels of serum potassium peri-operatively and throughout the hospital stay remains one of the most critical issues in the management of patients with Hypokalemic Periodic Paralysis.

REFERENCES


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