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CASE REPORT

Anesthetic management of child with Gitelman Syndrome: case report

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Abstract Gitelman syndrome is a rare autosomal recessive inherited disease that affects the thiazidesensitive sodium-chloride cotransport channels and the magnesium channels in the distal convoluted tubule, leading to hypokalemic metabolic alkalosis, hypomagnesemia and hypocalciuria. There is no cure for this condition and supportive treatment relies on ionic supplementation and symptom management. Literature regarding the anesthetic approach is scarce. This case report presents the anesthetic management of a child with Gitelman syndrome and its difficult electrolyte optimization.

Introduction

Gitelman syndrome is a rare autosomal recessive inherited disease that affects the thiazidesensitive sodium-chloride cotransport channels and the magnesium channels in the distal convoluted tubule leading to hypokalemic metabolic alkalosis, hypomagnesemia, hypokalemia and hypocalciuria.[1–4] There is no cure for this syndrome and supportive treatment relies on ionic supplementation and symptom management.[1–4]

We describe perioperative management of a 10-year-old girl admitted for tonsillectomy. She presented severe hypokalemia despite maximal oral reposition. Published literature doesn't report pediatric or adult Gitelman syndrome cases with clinically relevant hypokalemia proposed for surgery under general anesthesia.[1–4]

Case report

A 10-year-old girl, weighing 35 kg, with Gitelman syndrome and no other pathological conditions, was proposed for tonsillectomy after being postponed several times due to moderate to severe uncorrected hypokalemia. Her regular oral medication was potassium chloride 2.4 $g.day^{-1}$, spironolactone 25 $mg.day^{-1}$ and magnesium sulphate 5 $g.day^{-1}$.

She was followed by her pediatrician on a regular basis and the potassium replacement doses were increased for the perioperative period; however, with poor ionic control

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in spite of maximal dosage. The maximum potassium levels obtained were approximately 2.9 $mEq.L^{-1}$, with magnesium levels that were also corrected and in the lower limit of normal. Despite the frequent and resistant hypokalemia, the symptoms were minimal, with occasional cramps and myalgia and her preoperative ECG showed QTc in the upper limit of normal.[5]

Hereupon, we were presented with a patient with resistant hypokalemia and baseline QTc prolongation, known risk factors for drug-induced torsade de pointes (TdP) and other dysrhythmias, as well as for cardiac arrest.[1–5]

Considering that the patient had recurrent tonsillitis and a past episode of a peritonsillar abscess treated conservatively with antibiotics, it was decided to proceed with surgery, choosing an anesthetic technique with the least possible electrolyte disturbance and rhythm interference.

On the day of the surgery, potassium levels were 2.9 $mEq.L^{-1}$, the highest value obtained up until that moment, and magnesium was in the lower limit of normal. An intravenous (IV) infusion of KCl $4mEq.kg^{-1}.h^{-1}$ was started, under continuous electrocardiographic monitoring, 10 minutes prior to anesthetic induction. Standard ASA monitoring, pediatric BIS® and acceleromyography for neuromuscular block monitoring were adopted.

Induction of anesthesia was achieved with fentanyl 3 $mcg.kg^{-1}$ IV, propofol 3 $mg.kg^{-1}$ IV and rocuronium 0.6 $mg.kg^{-1}$ IV. Sevoflurane was used for anesthetic maintenance. For postoperative analgesia paracetamol 15 $mg.kg^{-1}$ IV, ketorolac 0.5 $mg.kg^{-1}$ IV, morphine 0.1 $mg.kg^{-1}$ IV and hydrocortisone 2 $mg.kg^{-1}$ IV were administered. Postoperative nausea and vomiting were prevented with dexamethasone 0.1 $mg.kg^{-1}$ IV and metoclopramide 0.1 $mg.kg^{-1}$ IV.

After induction, an arterial blood sample revealed 2.5 $mEq.L^{-1}$ potassium level. There was a slight prolongation of the QTc interval (458 msec) but no severe arrhythmia was recorded. Potassium infusion rate was doubled to 8 $mEq.kg^{-1}\cdot h^{-1}$. Magnesium levels remained stable and no supplementation was needed. Surgery was uneventful, including no excessive bleeding, and lasted approximately 1 hour. Suggammadex2 $mg.kg^{-1}$ IV was used to reverse neuromuscular block.

After the anesthetic emergence, the patient had minimal cramps and myalgias. Previous electrocardiographic changes were resolved. Continuous electrocardiographic monitoring and surveillance was maintained throughout anesthesia recovery. Potassium infusion rate was decreased to 4 $mEq.kg^{-1}.h^{-1}$ and maintained during late recovery until the patient started oral intake. Until hospital discharge potassium and magnesium levels were monitored by her pediatrician.

Discussion

The aim of this case report is to describe the anesthetic management of a patient with Gitelman syndrome since literature findings about this issue are scarce.

The authors performed a literature review in PubMed with the key words: Gitelman syndrome and Anesthesia. The literature review revealed very rare reports of anesthetic management of patients with this syndrome, particularly in children. All patients described in the literature had normal or near normal levels of potassium corrected prior to surgery, unlike this patient.[1–3]

There is a report of a 3-year-old child with Gitelman syndrome admitted for adenotonsillectomy. This child was being treated with oral potassium, magnesium and a potassium-sparing diuretic,[1] just like the patient we report. However, in that report, preoperative serum levels of potassium were 3.2 $mmol.L^{-1}$.[1] Since our patient had severe hypokalemia, a potassium infusion was needed to keep ionic balance. Nevertheless, in both cases there were no adverse events during the perioperative period.[1]

Complete correction of hypokalemia and hypomagnesemia may be difficult to achieve due to side effects, but near normal values should be sought.[1,4] We intend to show that although these patients may have poor ionic control, the chosen anesthetic approach should be based on the essential physiologic, pharmacokinetic, and pharmacodynamic principles in order to reduce the risk associated with general anesthesia under these conditions.[1,4] Drugs and therapies that may prolong the QTc interval or worsen electrolyte abnormalities should be avoided if possible.[1,4]

In this case, only drugs with little or none electrolyte and rhythm interference were chosen. Droperidol, known to prolong the QT interval,[5] was excluded for nausea and vomiting prophylaxis.

Painful stimuli may worsen QT prolongation and trigger potentially fatal arrhythmia.[1,5] Therefore, noxious stimulation from laryngoscopy and endotracheal intubation, surgical stimulation and stormy extubation were prevented by maintaining adequate analgesia and anesthesia depth. Pediatric BIS® was a helpful monitor.

Fentanyl, morphine, and rocuronium elimination is affected by abnormal kidney function, but drug elimination is not affected by this ion channel defect;[5] this patient did not present abnormal kidney function.

Succinylcholine is a depolarizing muscle relaxant and causes potassium release from muscle cells as an initial side effect.[5] We decided not to use it as potassium increase would be shortlasting and it would interfere with the ionic supplementation perfusion. Also, no rapid induction was needed. There are, notwithstanding, reports of succinylcholine use with no adverse events related to it.[1–3]

The risk of *torsades des points* is higher with QTc prolongation greater than 500 msec, however it still exists with QTc prolongation under 500 msec.[4,5] In this case, no severe arrhythmias occurred and no new symptoms were experienced in the immediate postoperative period. Even so, the potassium infusion was maintained until discharge from the post-anesthetic care unit.

Although this case had a good outcome and the procedure was uneventful, further studies and case descriptions are needed to guide a safe anesthetic approach of these patients, especially if more complex surgeries are required.

Conclusion

Gitelman syndrome is a mild disorder, however severity of symptoms may be dramatic. Understanding the syndrome's pathophysiology and its perioperative implications is the key to ensure safe outcome of patients.

Ethical approval

Parental written consent was given for the publication of this case report.

Conflicts of interest

The authors declare no conflicts of interest.

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