

## Anesthesia in a patient with Gitelman syndrome

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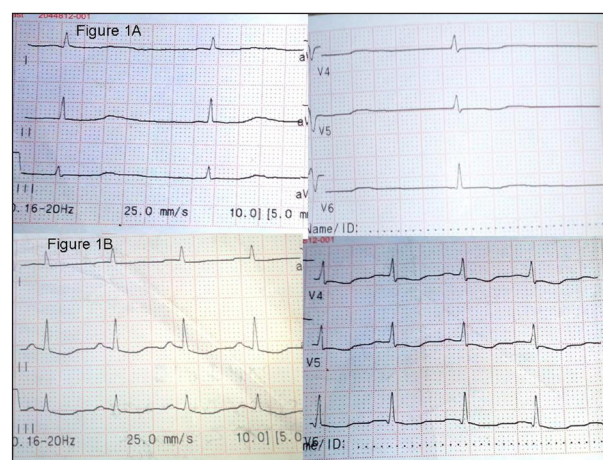
Gitelman syndrome (GS) is an autosomal recessive renal disorder caused by mutation of genes encoding the thiazide-sensitive sodium chloride co-transporters and magnesium (Mg) channels in the distal convoluted tubule leading to hypokalemic metabolic alkalosis along with hypomagnesemia and hypocalcinuria.<sup>[1]</sup> Hereby, we describe perioperative management of a 27-year-old female with GS presented for temporary ileostomy with “borderline” QT interval prolongation. Our search of literature did not reveal any patient with GS having prolonged QT interval being anesthetized for ileostomy.<sup>[2]</sup>

A 27-year-old female (45 kg weight and 145 cm height) was posted for temporary ileostomy for acute on chronic colitis. She had a recent history of generalized seizures/tetany and was on phenytoin sodium. On clinical examination, she

was severely lethargic. Electrocardiography (ECG) showed sinus bradycardia with heart rate 44/min, QT interval 534 ms and corrected QT interval (QTc) 457 ms (“borderline” QT prolongation) [Figure 1a]. Her laboratory values were significant for hemoglobin-7.6 g% (12-16), serum potassium (K) 2.21 mEq/L (3.5-5.5), serum Mg 1.28 mg/dl (1.8-2.5) with normal serum sodium (Na), serum chloride (Cl), serum calcium (Ca) and serum creatinine. 24 h urinary electrolytes (Ca-117 mg/24 h [female <250, male <300] with normal Na and K) and arterial blood gas analysis (pH-7.542, pO<sub>2</sub>-79.9 mmHg, PaCO<sub>2</sub>-42.4 mmHg, HCO<sub>3</sub>-35.6 mmol/L, base excess: +11.9 mmol/L) confirmed the diagnosis of GS. For the next 3 days, hypokalemia and hypomagnesemia were corrected preoperatively by oral syrup potassium chloride and injection Mg sulfate intravenously (IV), respectively, along with tablet spironolactone. One unit of packed red cells was infused, also.

On the day of surgery, her blood laboratory values were within normal limits and normal sinus rhythm with heart rate 100/min, QT interval 320 ms and QTc 413 ms recorded on ECG [Figure 1b]. Her general condition improved remarkably. Injection ceftriaxone 1 g IV was given as antimicrobial prophylaxis.

After premedication with fentanyl 100 µg IV, the patient was given total IV anesthesia (TIVA). Induction was done with propofol 100 mg and atracurium 30 mg was given to facilitate endotracheal intubation and was maintained with propofol infusion 50-150 µg/kg/min along with oxygen-air at a 1:1 ratio; additional doses of fentanyl and atracurium were administered as appropriate. Standard monitors were applied intraoperatively. Surgery lasted for 70 min and 1 L of lactated Ringer’s fluid was infused intraoperatively. At the end of surgery, residual neuromuscular blockade was reversed



**Figure 1:** Electrocardiography. (a) Before electrolyte correction. QT interval (QT)-534 ms, corrected QT interval (QTc)-457 ms, (b) After electrolyte correction. QT-320 ms, QTc-413 ms

with neostigmine 2.0 mg and glycopyrrolate 0.4 mg and trachea was extubated. She was shifted to the recovery area and monitored for 24 h uneventfully. Tramadol was used for control of postoperative pain.

Anesthetic management in our patient aimed at preoperative correction of electrolytes and acid-base disturbances and avoidance of drugs and events that prolong QT interval.

Hypokalemia, hypomagnesemia, and baseline QTc prolongation, corrected preoperatively in our patient, are all risk factors for drug-induced torsades de pointes (TdP) and sudden death.<sup>[3]</sup> Certain drugs, such as macrolide antibiotics, antidysrhythmic drugs, antihistaminics and antipsychotic, are known to cause TdP and should be avoided perioperatively.<sup>[3]</sup> Halogenated volatile anesthetics are known to cause QT prolongation; therefore, we used TIVA with propofol infusion for maintenance of anesthesia.<sup>[4]</sup> Laryngoscopy and endotracheal intubation, noxious surgical stimulation and stormy extubation may aggravate QT prolongation. These events were managed in our patient by maintaining adequate depth of anesthesia. Respiratory alkalosis from hyperventilation may acutely exacerbate hypokalemia and should be avoided. Though reversal of neuromuscular blockade should be avoided in patients with prolonged QT interval, it was done in our patient in view of high chances of respiratory insufficiency due to muscle weakness.

To conclude, though GS is mild disorder, severity of symptoms may be dramatic. Perioperative management relevant to the pathophysiology of the syndrome ensures safe outcome of patients with GS.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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### References

1. Knoers NV, Levchenko EN. Gitelman syndrome. *Orphanet J Rare Dis* 2008;3:22.
2. Farmer JD, Vasdev GM, Martin DP. Perioperative considerations in patients with Gitelman syndrome: A case series. *J Clin Anesth* 2012;24:14-8.
3. Ritter JM. Cardiac safety, drug-induced QT prolongation and torsade de pointes (TdP). *Br J Clin Pharmacol* 2012;73:331-4.
4. Fazio G, Vernuccio F, Grutta G, Re GL. Drugs to be avoided in patients with long QT syndrome: Focus on the anaesthesiological management. *World J Cardiol* 2013;5:87-93.

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