

# Anaesthesia for emergency caesarean section in a patient with Gitelman syndrome

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## ABSTRACT

Gitelman syndrome is a rare autosomal recessive salt-losing disorder of renal tubules, which results in hypokalemic metabolic alkalosis. Associated hypomagnesaemia and hypocalcaemia are also seen. Gitelman syndrome is a variant of renal Bartter's syndrome. Anaesthetic management of these obstetric patients is challenging and requires a careful multidisciplinary approach. Close monitoring of potassium and magnesium levels and their supplementation is required to avoid complications to mother and baby.

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**Key words:** Anaesthesia, Gitelman syndrome, hypokalemia, obstetrics

## INTRODUCTION

Gitelman syndrome is a rare autosomal recessive salt-wasting disorder of renal tubules with an incidence of 1 in 40,000.<sup>[1]</sup> It is associated with hypomagnesaemia, hypocalcaemia and hypokalemic metabolic alkalosis. The defect is in thiazide-sensitive Na<sup>+</sup> Cl<sup>-</sup> co-transporter located in the distal convoluted tubules.<sup>[2]</sup> Gitelman syndrome is a variant of renal Bartter's syndrome. Bartter's syndrome results from mutation of transporters of ascending loop of Henle,<sup>[3]</sup> whereas Gitelman syndrome occurs due to a mutation in the solute carrier family 12, member 3, SLC12A3 gene, which encodes the co-transporter.<sup>[2]</sup>

## CASE HISTORY

23-year-old G<sub>2</sub>A<sub>1</sub>, parturient at 35 weeks of gestation, a known case of Gitelman syndrome came to Obstetric out-patient department with a history of raised blood pressure. Two years back she was diagnosed with Gitelman syndrome by a consultant

nephrologist. Since then she was on oral electrolyte supplementation and eplerenone 25 mg twice daily. Her investigations on admission showed potassium of 3.1 and magnesium of 1.2 meq L<sup>-1</sup> respectively. After consulting with nephrologist oral potassium and magnesium correction was given. Five days after admission she started developing labour pain. Her investigations showed normal potassium and magnesium. Electrocardiogram (ECG) did not show any QT prolongation. She was shifted to labour room and on examination was found to have cord prolapse. She was immediately taken up for caesarean section. As it was an emergency, a brief history and

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examination was done and general anaesthesia with rapid sequence induction was planned. On arrival in the operating room, monitoring was done with a 5-lead ECG, non-invasive blood pressure, and pulse oximetry. After securing IV access, rapid sequence induction was performed with 5 mg kg<sup>-1</sup> thiopentone and 0.6 mg kg<sup>-1</sup> of succinyl choline. She was intubated with 7 mm size endotracheal tube. Anaesthesia was maintained with oxygen, air and isoflurane and atracurium. Intraoperatively an arterial blood gas was sent, which showed pH of 7.35 with normal gas exchange, sodium of 132 mmol L<sup>-1</sup>, potassium of 3.4 mmolL<sup>-1</sup>, chloride of 106 mmol/L and calcium of 0.95 mmolL<sup>-1</sup>. Live baby was extracted, oxytocin 3U IV bolus followed by 3U/h infusion was started as per our institutional protocol. IV fentanyl 2µgkg<sup>-1</sup> and anti-emetic ondansetron 4 mg was given in the operation theatre. Procedure was uneventful and patient was reversed and extubated on table. Postoperatively she was monitored for 6 h in the recovery room. She had an uneventful postoperative period and was discharged on third postoperative day.

## DISCUSSION

Gitelman syndrome is a rare autosomal recessive renal tubular disease characterized by hypokalaemia, hypomagnesaemia, metabolic alkalosis and low urinary calcium excretion.<sup>[4]</sup> Presentation is usually seen in late childhood with paresthesia and generalized weakness.<sup>[2]</sup> There are case reports of chondrocalcinosis developing in these patients from severe hypomagnesaemia.<sup>[3]</sup> Other presenting symptoms include ataxia, vertigo, and blurred vision.<sup>[1,2]</sup> Electrolyte disturbances make these patients prone to ventricular arrhythmias. QT prolongation is seen in approximately 50% of these patients.<sup>[2,3]</sup> Even though function of renin-angiotensin-aldosterone is affected, blood pressure usually remains normal.<sup>[5]</sup>

Gitelman syndrome can affect women of child bearing age group.<sup>[5]</sup> Increased renin-angiotensin-aldosterone system activity associated with normal pregnancy causes loss of potassium and magnesium.<sup>[2]</sup> Increased foetal demands, hyper-emesis, diarrhoea during pregnancy may tilt the balance in pregnant patients with Gitelman syndrome.<sup>[4,5]</sup> In a case series of five patients with Gitelman syndrome by Mascetti *et al.*, four of them had uneventful pregnancy but one was complicated by fatigue and tetanic seizures.<sup>[6]</sup> Seizure could be due to hypomagnesemia and metabolic alkalosis.<sup>[7]</sup>

Multidisciplinary approach is required in the management of a parturient with Gitelman syndrome. Serial monitoring of potassium and magnesium levels have to be performed.<sup>[8]</sup> Preoperative ECG is necessary to rule out arrhythmias and QT prolongation. Oral supplementation of electrolytes during pregnancy may be required to ensure that these patients are asymptomatic. In some patients, correction of electrolyte levels cannot be achieved inspite of supplementation.<sup>[5,9]</sup> Normalisation of electrolyte values is not recommended.<sup>[10]</sup> Magnesium depletion alone is less likely to cause arrhythmias but may exacerbate arrhythmias caused by hypokalemia. Intravenous supplementation may be required in labour and during caesarean section.<sup>[5]</sup> Treatment with aldosterone antagonists during pregnancy is controversial but sometimes used. Potassium-sparing drugs such as amiloride and eplerenone is preferred to spironolactone in pregnancy.<sup>[11]</sup>

There are no clear guidelines in the anaesthetic management of Gitelman syndrome. Regional anaesthetic techniques may be preferable over general anaesthesia to avoid polypharmacy and risk of failed intubation and aspiration, and it also provides better post-operative analgesia.<sup>[12]</sup> Epidural or combined spinal epidural anaesthesia could be used. Uneventful spinal anaesthesia for caesarean delivery in pregnant women with Gitelman syndrome have been reported.<sup>[2]</sup> We used general anaesthesia as our patient did not have QT prolongation and there was severe foetal distress. In patients with QT prolongation, anaesthetic drugs have to be selected carefully. Supine hypotension has to be avoided by the use of wedge under hip.<sup>[13]</sup> Thiopentone and propofol are considered safe. Ketamine is better avoided. Halogenated inhalational agents can prolong QT interval. Sefoflurane has to be avoided, but isoflurane has been reported to be safe. Succinyl choline can prolong QT, but this effect could be reversed by using alfentanil along with it. Among muscle relaxants rocuronium, vecuronium and atracurium are safe, but pancuronium needs to be avoided. Antiemetic ondansetron has to be avoided. During positive pressure ventilation high airway pressures and Valsalva maneuver are to be avoided.<sup>[14]</sup> Complications that may occur peri-operatively include laryngeal spasm, stridor, paraesthesia, tetany, convulsions, and ventricular arrhythmias.<sup>[14]</sup> Hypotension during surgery usually responds to fluid resuscitation, electrolyte replacement and vasopressors.<sup>[6,7]</sup> Stable parturient with near normal electrolytes posted for

caesarean section does not require invasive lines; a baseline arterial blood gas (ABG) prior to procedure may be useful. Invasive lines and ICU care should be considered in patients with unstable potassium levels, symptomatic patients and high risk pregnancies. Continuous ECG monitoring during labour could be a safe strategy to follow since up to 50% of these patients are reported to have prolonged QT syndrome.

In conclusion, multidisciplinary team including obstetricians, endocrinologists, nephrologists, anaesthetists and neonatologists are required for good obstetric and neonatal outcome. Anaesthetic management of these parturients requires a detailed preoperative assessment, optimisation, serial monitoring of electrolytes and awareness of potential complications. In uncomplicated pregnancy with asymptomatic patients, near normal level of potassium and magnesium are considered acceptable.

#### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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#### Conflicts of interest

There are no conflicts of interest.

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