

Anaesthesia for a patient with Gitelman's syndrome undergoing abdominal hernia repair

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INTRODUCTION

Anaesthetic management for patients with Gitelman's syndrome hasn't been reported yet. Gitelman's syndrome is due to a defect in sodium chloride transport in the distal tubule (the site of action of thiazide-type diuretics). Acute exacerbation of the hypokalemia and the metabolic alkalosis may interfere with surgery and general anaesthesia. We hereby describe the anaesthetic course of a 76 year old patient, diagnosed with Gitelman's syndrome, undergoing abdominal surgery.

CASE REPORT

A 76 year old female patient was scheduled for elective umbilical hernia repair.

The patient (weight 58 kilograms, height 143 centimeters) had a proven metabolic alkalosis and chronic hypokalemia (average 2,5-3,5 mg/dl) in spite of a normal bicarbonate.

Other medical antecedents were arterial hypertension, ischemic heart disease with negative coronarography ten years ago, hypercholesterolemia, reflux esophagitis with a hernia diafragmatica, colitis ulcerosa and a fracture of the arm.

At the time of surgery the patient complained of gastrointestinal obstipation ; she was known with anorexia but without further weight loss.

Furthermore, this patient was known with stable angina pectoris described as retrosternal pain, radiating to the throat and especially present at efforts. Occasionally she had edema at her right ankle.

At last she complained of vertigo, suggestive for orthostatic hypotension.

Her present therapy consisted of ranitidine 300 mg/day, Chloropotassuril 54 mEq/day,

Magnesium gluconate 3 g/day and spironolactone 100 mg/day.

This patient got an umbilical hernia repair with a Polypropylene mesh, using the Rives-Stoppa technique.

The week before surgery she underwent cardiologic and respiratory investigations and also checking her neurologic and nefrologic status. A transthoracic echocardiography was normal. The blood pressure taken in supine and upright position couldn't confirm a clear orthostatic hypotension. There were no arguments for venous trombosis or flebitis or abdominal abnormalities. The day before surgery, she received 120 mEq kalium chloride, IV over 24 hours.

Induction of general anaesthesia was done with propofol (130 mg), sufentanil (15 µg) and cisatracurium (12 mg). The patient already had a 20 G peripheral venous catheter but a second 18 G catheter was inserted at the left arm. A cuffed endotracheal tube n° 8 was inserted and mechanical ventilation was started. We used the AS 3, Datex® mechanical ventilator to perform a volume controlled ventilation at constant flow and a mixture of oxygen / air (FiO₂ : 0.5) with sevoflurane (1,5% MAC) was chosen.

Inspiratory time was 33% of total time with an end inspiratory pause of 25% and a respiratory rate of 10 breaths/min. The ventilatory settings were kept constant throughout the intervention. No external positive end-expiratory pressure was given.

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Monitoring consisted of non-invasive arterial blood pressure (cuff on the right arm), a five leads ECG, pulse-oximetry and capnography. The surgery was in supine position with protective cushions placed under the arms and legs of the patient. The procedure lasted about 2 hours and was uneventful.

The preoperative blood analysis showed a plasma potassium of 2,74 mg/dl, a sodium of 146 mg/dl, a magnesium level of 1,83 mg/dl and a creatinine of 0,67 mg/dl.

A bloodgas analysis done during the surgery showed a pH of 7,53, a carbon dioxide partial pressure (pCO₂) of 33 mmHg, an oxygen partial pressure (pO₂) of 174 mmHg, an oxygen saturation (sat O₂) of 100%, Na⁺ of 131 mg/dl, K⁺ of 2,3 mg/dl, Ca⁺⁺ of 1,05 mg/dl, hematocrite of 33% and hemoglobin level of 10,2 g/dl, HCO₃⁻ of 27,6 mg/dl.

During surgery she received 1 liter of crystalloids (Glucose 5% in Ringer lactate solution).

For pain relief 1 gram of acetaminophen and 100 mg of tramadol was administered.

About 10 minutes after skin closure the patient began spontaneously breathing and we successfully managed to extubate the trachea in the operating room. The patient was then transferred to the recovery room for postoperative observation.

During her stay in the recovery, potassium was checked every 4 hours but she didn't need supplemental potassium.

Haemodynamic and respiratory parameters remained stable with supplemental oxygen administered via a face mask at a flow rate of 6 L/min. Recovery was uneventful and the patient was discharged the same day to the ward.

DISCUSSION

The diagnosis of Gitelman's syndrome in this patient was mainly based on the combination of a proven hypokalemia and metabolic alkalosis. Although the patient received supplemental potassium daily, we decided not to give during anaesthesia any potassium considering that Ringer Lactate was used, without any important blood loss.

Gitelman's syndrome, an autosomal recessive disorder, is often not diagnosed until late childhood or even adulthood (1). The syndrome is usually symptomatic and can be associated with serious clinical manifestations (2) such as cramps (especially arms and legs), severe fatigue (3), polyuria and nocturia. Affected patients may also present

with tetany (approximately 10% of individuals), particularly in association with decreased intestinal absorption of magnesium (e.g. vomiting, diarrhea) (4).

These patients have mutations in the gene coding for the thiazide-sensitive sodium-chloride cotransporter in the distal tubule, which most commonly result in impaired cellular routing of the cotransporter (5, 6). A defect in this transport can account for both the magnesium wasting and the often marked decrease in calcium excretion which is similar to that induced by thiazide therapy and the opposite of the hypercalcemia seen in classic Bartter's syndrome (1).

Vomiting and diuretic use are the two major causes of unexplained hypokalemia and metabolic alkalosis in normotensive patients (7). Patients with Gitelman's syndrome tend to be clinically euvolemic with chloride excretion being equal to intake. The net effect is a urine chloride concentration that is usually above 40 mEq/l.

The combination of supplemental potassium and a potassium sparing diuretic such as spironolactone, can raise the plasma concentration of potassium toward normal, can reverse the metabolic alkalosis and potentially can correct the hypomagnesemia also.

Diuresis can be followed, but knowing that the tubular defect in Gitelman's syndrome cannot be corrected, no special treatment was done in this case perioperatively.

In conclusion, it is important to keep an eye on the electrolytes and pH's during and after surgery but it is not necessary to correct them immediately except when there are important differences in plasma potassium etc., depending on the type and magnitude of surgery.

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