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CASE REPORT

# Ambulatory management of a patient with bartter syndrome under general anesthesia

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ABSTRACT

A rare inherited renal tubulopathy, Bartter Syndrome is characterized by salt-wasting in the kidneys, resulting in the effects resembling those of loop diuretics: hypokalemia, hypochloremia, metabolic alkalosis, and volume contraction leading to low to normal blood pressure. The marked electrolyte and hemodynamic instability that is often seen in these patients can sometimes result in catastrophic consequences. Because of the relative rarity of this condition, there are only sparse reports on anesthetic management that typically involve preoperative testing carried out prior to the day of surgery. We herein describe a case of a 54-year-old patient with Bartter Syndrome who presented to the hospital for an outpatient dental surgery under general anesthesia. Preoperative consultation with a nephrologist helped to establish our strategy in maintaining the patient's electrolyte balance. Point-of-care blood gas monitoring was carried out at regular intervals and guided the perioperative potassium supplementation. Patient remained stable for the entire course of the surgery and was discharged home the same day after one hour in the recovery unit.

Keywords: Anesthesia, point-of-care systems, Bartter Syndrome, ambulatory surgery, electrolyte management.

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## INTRODUCTION

Bartter Syndrome is a rare inherited renal tubulopathy characterized by salt-wasting in the loop of Henle. It thus mimics the effects of loop diuretics resulting in hypokalemia, hypochloremia, metabolic alkalosis, and volume contraction effecting low to normal blood pressure [1]. Unsurprisingly, patients who carry this diagnosis often encounter marked electrolyte and hemodynamic instability, sometimes resulting in catastrophic consequences. Because of the relative rarity of this condition, there is not a wealth of literature on anesthetic management, and patients who require surgical procedures may encounter providers who are not familiar with the implications of their diagnosis. The few case reports discussing anesthetic management of these patients typically describe the need for preoperative labwork and evaluation followed by significant intraoperative testing and monitoring [2-4]. Herein we describe an ambulatory management of an adult patient with Bartter Syndrome under general anesthesia. We report how we approached testing, consultation, and monitoring for this same-day case.

The syndrome was first described in 1962 by Frederic Bartter as a constellation of hypokalemic alkalosis, hyperaldosteronism, and hyperplasia of the juxtaglomerular complex [5]. The term is now used to cover a set of genetically heterogeneous diseases that show significant clinical variability, making overarching characterization and guidelines complex [1]. Five different forms have been identified based on molecular genetics, though these forms have much phenotypic overlap. Often, the condition is diagnosed in childhood, presenting with polyuria and failure

to thrive, though others are not detected until adulthood. It is estimated that current prevalence [6] is approximately 1 in 1,000,000.

The genetic mutations in the Bartter Syndrome primarily lead to impaired sodium reabsorption in the thick ascending limb of the loop of Henle, leading to its increased resorption in the distal convoluted tubule and concomitant increase of potassium loss. Activation of the renin-angiotensin-aldosterone (RAA) system along with inappropriate prostaglandin synthesis creates a feedback loop that exacerbates the abnormalities. Hypomagnesemia and hypocalcemia may also occur. Patients with the hypercalciuria/hypocalcemia phenotype may present with frequent nephrolithiasis, as well as bone resorption from the jaw and dental mobility and dislocation [7].

Treatment includes lifelong supplementation of deficient electrolytes, chiefly potassium, but magnesium and calcium may also be required. Potassium-sparing diuretics such as spironolactone, as well as non-steroidal anti-inflammatory drugs (NSAIDs) have been employed as therapy [8].

Patients with Bartter Syndrome may require surgery and/or anesthetic care at some point in their lives. The complexity of preoperative workup and perioperative management may depend on the duration and level of invasiveness of the procedure, and whether general anesthesia is called for. Attention should still be paid to correcting any gross electrolyte abnormalities to ensure safe conduct of anesthesia.

## CASE PRESENTATION

A 54 year-old woman presented to the county hospital for extraction of two decayed and necrotic teeth, a pneumatised sinus requiring sinus lift with bone grafting along with platelet rich plasma placement, and an osseointegrated implant placement. Due to her severe anxiety, she could not tolerate the surgery in an office setting and was scheduled for general anesthesia. She presented without preoperative workup, but proved to have keen insight into her condition. During the pre-anesthetic assessment, she reported that she had been diagnosed with Bartter Syndrome at the age of 37 after suffering from a cardiac arrest triggered by hypokalemia. She had since undergone pacemaker/ICD placement and has had no further cardiac or hemodynamic abnormalities. Her medications included potassium chloride 160 mg daily and spironolactone. She did not require magnesium or calcium supplementation and did not take NSAIDs on a regular basis.

After discussion with the surgical team, the case was briefly delayed to obtain a consult from the in-hospital nephrology service. The nephrologist recommended to check a preoperative potassium and supplement intravenously to maintain near her baseline. Since serial intraoperative potassium checks were anticipated, an iStat point of care testing (POCT) device was used, utilizing the Chem8+ cartridge, thereby facilitating timely assays in the operating room. Her initial potassium was 2.5 mEq/L. The patient stated she felt she was "low," describing mild fatigue. Administration of 20 mEq KCl was achieved through a peripheral IV over two hours. When rechecked, the patient's potassium had surprisingly risen to 3.5 mEq/L, though this was likely elevated from hemolysis, as the $pCO_2$ and bicarbonate were elevated and pH was lower (Table 1). The patient stated she felt better, and a repeat sample was deferred until the start of the case.

Preoperative vital signs were: blood pressure 106/84 mmHg, heart rate 97 bpm, $SpO_2$ 100% on room air, respiratory rate 16 breaths/min, temperature 36.7°C. Patient stated that the above blood pressure was normal. To account for fasting, 700 mL of lactated Ringer's solution were administered in the preoperative area with little change of her vital signs.

The patient experienced a stable operative course. She was premedicated with intravenous midazolam 2 mg and fentanyl 100 µg. Induction consisted of lidocaine 100 mg, propofol 200 mg, and rocuronium 50 mg, with no hypotension noted. Succinylcholine was specifically avoided to decrease the risk of sudden potassium shifts.

Intraoperatively, repeat potassium was drawn after induction and was found again to be 2.5 mEq/L. An additional 10 mEq was infused over the next hour. A second 10 mEq bag of KCl was started, however a repeat check revealed a potassium of 3.0 mEq/L and this was stopped, as overcorrection is as dangerous as undercorrection in patients with chronic severe hypokalemia.

 

**Table 1.** Values of Successive iStat Chem8+ Blood Tests

| Tests | Times Samples Resulted | | | |
|---|---|---|---|---|
| | 09:47 | 11:34 | 14:35 | 15:27 |
| Sodium (mEq/L) | 133 | 136 | 136 | 133 |
| Potassium (mEq/L) | 2.5 | 3.5 | 2.5 | 3.0 |
| Glucose (mg/dL) | 113 | 103 | 97 | 124 |
| Hgb (g/dL) | 15.6 | 13.6 | 12.2 | 12.6 |
| Hct (%) | 46 | 40 | 36 | 37 |
| pH | 7.42 | 7.33 | 7.41 | 7.47 |
| $pCO_2$ (mmHg) | 41 | 60 | 39 | 34 |
| $pO_2$ (mmHg) | 30 | 18 | 81 | 64 |
| $HCO_{3-}$ (mEq/L) | 27 | 32 | 25 | 25 |
| Total $CO_2$ (mmHg) | 28 | 34 | 26 | 26 |
| $O_2$ Saturation (%) | 58 | 24 | 96 | 95 |

Symbols: mEq/L – milliequivalents/liter; mg/dL – milligrams/deciliter; g/dL – grams/deciliter; mmHg – millimeters mercury.

The patient remained hemodynamically stable throughout the case, with blood pressures at or higher than her baseline. She did exhibit occasional premature atrial contractions (PAC's) on telemetry, and occasionally her pacemaker initiated some beats when her heart rate fell below 70 bpm. We elected not to place a magnet on the pacemaker as she was not pacer-dependent, and we wished to keep the ICD function intact in case of life-threatening arrhythmias. In addition, the patient return electrode pad was placed on the right thigh with the surgery taking place on the right maxilla, decreasing the possibility of an electrical arc passing through the pacemaker's left chest location.

Emergence was uneventful, and the patient recovered in the post-anesthesia care unit for approximately 1 hour prior to discharge home. She gave her consent to report this case. In a follow-up phone call the next day the patient reported faring well apart from some soreness at the surgical site.

## DISCUSSION

We present this case to establish the feasibility of management of a patient with Bartter Syndrome in an ambulatory setting while managing the type of evaluation, workup, and monitoring such environments afford. While some patients undergoing more extensive procedures would benefit from more robust preoperative preparation, there are many others like our patient who present for relatively brief and minimally invasive procedures for which anesthesia is required. Anesthesia providers faced with these patients must decide their level of comfort with the degree of testing and monitoring available to them.

Our patient's anxiety and duration of the procedure dictated the choice of general anesthesia. While monitored anesthesia care (MAC) was also an option, we elected to have a controlled airway that general anesthesia afforded. We were fortunate in our patient's knowledge of her disease (she reported her phenotype does not suffer from hypocalcemia and hypomagnesemia so as to require regular supplementation), the timely response of our in-hospital nephrologist, and a facility that could accommodate a several hour delay in the case. While we did encounter some inconsistency in our iStat POCT monitoring (a likely hemolyzed sample in preop and a more robust than expected response to 10 mEq KCl intraoperatively), our discussion with the nephrologist reassured us that a gentle replacement was a safer course than attempting to achieve exact blood levels of potassium. The patient was also able to give us feedback when she was awake, stating that the preoperative replacement was helpful symptomatically. We erred on the side of cautious replacement rather than disrupting the delicate electrolyte balance of chronic hypokalemia to which her body had become accustomed.

We discussed with the surgeons whether to admit the patient to observation postoperatively. Some providers may have chosen this option out of an abundance of caution. However, given that the

patient required minimal potassium correction, was hemodynamically stable throughout the perioperative period, and had a remotely-monitored pacemaker/ICD, we felt comfortable releasing her home. We did advise her to contact her primary care physician, be particularly vigilant to any new symptoms, and gave her strict return precautions. On her follow-up phone call, the patient reported a high level of satisfaction with her experience.

## Author contribution

Study conception and design: EK; data collection: EK; analysis and interpretation of results: EK and RK; draft manuscript preparation EK and RK. All authors reviewed the results and approved the final version of the manuscript.

## Ethical approval

Ethical Approval is not required for this article. Patient provided us with a written consent to submit her case for publication.

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## Conflict of interest

The authors declare that there is no conflict of interest.

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