Management of Maternal Bartter Syndrome during Pregnancy: A Case Report

Savannah Vogel and Sana Waheed*
Department of Medicine, University of Wisconsin School of Medicine and Public Health, USA

Abstract

Bartter Syndrome type 3 is a rare renal tubular disorder characterized by metabolic disturbances, most notably hypokalemia. Pregnancy presents a unique challenge to the management of this disorder as the mainstays of treatment are contraindicated due to risk of fetal side effects. Here we report a case of the successful management of maternal Bartter Syndrome with 20mg amiloride BID and increased potassium supplementation up to 400mEq/day.

INTRODUCTION

Bartter syndrome type 3 is a rare autosomal recessive renal tubular disorder caused by a mutation in the CIC-Kb chloride channel in the ascending loop of Henle and is characterized by a post-natal presentation of hypokalemia, metabolic alkalosis, hypomagnesemia and failure to thrive. Here, we report a case of maternal Bartter Syndrome during pregnancy and the challenges faced in management, as many of the medications routinely used for treatment are contraindicated in pregnancy [1,2].

CASE PRESENTATION

A 22 year old G1P0 female presented to us at 19w0d gestation upon referral from OB/GYN for management of her previously diagnosed Bartter Syndrome. The patient was diagnosed with Bartter Syndrome Type 3 at six months of age after presenting with decreased feeding, weight loss and failure to thrive. Prior to her pregnancy, the patient was treated by pediatric nephrology with the non-steroidal anti-inflammatory drug (NSAID) indomethacin 50mg once daily (QD), spironolactone 25mg twice per day (BID) and potassium chloride (KCl) 30mEq BID. Spironolactone was discontinued at the start of the pregnancy out of concern for under-virilization of a male fetus. We discontinued indomethacin due to concern for premature closure of the ductus arteriosus. Potassium levels were expected to fall once the NSAID was discontinued so KCl was increased to 30mEq TID and the patient’s potassium levels were followed twice weekly labs. Three days later, the patient presented to the ED with fatigue and muscle cramps and was found to be hypokalemic with a serum potassium level of 2.9mEq/L. While in the ED, the patient received 40mEq IV KCl over four hours. Following discharge, she was started on amiloride 10mg QD and potassium supplementation was uptitrated. Potassium levels were continuously monitored every three days and supplementation further increased over an eight-week period. During this time, the patient was hospitalized twice with symptomatic hypokalemia, with symptoms of fatigue and muscle cramping, and received four KCl infusions in total. Potassium levels stabilized after an eight-week titration period on 10mg of amiloride BID and a total of 400mEq per day of potassium (Figure 1). Her potassium levels remained stable between 3.2-3.7 mEq/L for the remainder of the pregnancy.

The patient delivered a healthy male infant with Apgar scores of 9 at 1 and 5 minutes by spontaneous vaginal delivery at 38w5d. Amiloride was discontinued at the start of labor and the patient was given potassium and magnesium supplementation throughout labor, delivery and her postpartum hospital stay. Following delivery, as the patient was breastfeeding, indomethacin was restarted at 50 mg QD and potassium supplementation was titrated down to 30mEq BID. Postpartum potassium levels stabilized in the range of 3.4-3.6mEq/L.

DISCUSSION

Patients with Bartter Syndrome often experience severe metabolic disturbances without treatment. NSAIDs, aldosterone antagonists and angiotensin converting enzyme (ACE) inhibitors are considered the mainstay of treatment, however, these drugs have documented fetal side effects and thus cannot be safely used during pregnancy. Specifically, indomethacin can cause premature closure of the ductus arteriosus during the third trimester, aldosterone antagonists can lead to under-virilization of a male fetus and ACE inhibitors have been documented to cause adverse fetal effects including oligohydramnios and fetal renal failure [3, 4]. In addition to these contraindications to

Keywords

• Bartter syndrome
• Amiloride
• Pregnancy
• Hypokalemia

*Corresponding author
Sana Waheed, Department of Medicine, Division of Nephrology, University of Wisconsin School of Medicine and Public Health, 1685 Highland Ave, Madison, WI, USA, 53792, Tel: 513-238-1717; Email: sWaheed@medicine.wisc.edu
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normal therapy, management of Bartter Syndrome in pregnancy is further complicated by increased potassium needs due to increased volume of distribution. In our patient, we successfully used amiloride, a class B drug during pregnancy, along with increased KCl supplementation to maintain serum potassium levels. Of note, the patient tolerated 400meq per day of KCl during pregnancy without issues.

After delivery, it was challenging to decide on the best course of management as there are some concerns regarding safety of amiloride while breastfeeding. Theoretically, certain ACE-inhibitors can be used in lactating mothers, however, the patient was unwilling to start them at that time. Therefore, she was restarted on indomethacin and her KCl supplements were tapered. Her potassium level remained stable on indomethacin and 240mEq of potassium per day.

Previous case reports have documented the management of Bartter Syndrome during pregnancy. Many of these were cases of Bartter Syndrome which presented in pregnancy and were successfully managed with potassium supplementation alone [5-7]. Other cases have illustrated the successful use of amiloride in addition to potassium supplementation [8]. Our patient was unable to tolerate potassium supplementation alone.

Although the management of Bartter Syndrome during pregnancy represents a challenge as the mainstays of treatment are considered unsafe, amiloride seems to be a safe and effective medication.

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**REFERENCES**