Case Report

When Electrolytes Shocks You: Classical Bartter Syndrome (Type III) Presenting with Refractory Shock and Recurrent Extubation Failure in a 9-Month-Old Infant.

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ABSTRACT

Bartter syndrome (BS) is a rare inherited renal tubular disorder characterized by hypokalemia, metabolic alkalosis, renal salt wasting, and secondary hyperreninemic hyperaldosteronism with normal or low blood pressure. The classical form (type III) is commonly present in infancy or early childhood but may demonstrate variable clinical features, leading to delayed diagnosis. A 9-month-old male infant was admitted with severe pneumonia and septic shock requiring mechanical ventilation. The child developed persistent hypokalemia, hyponatremia, and metabolic alkalosis despite appropriate correction, resulting in four episodes of extubation failure. Persistent dyselectrolytemia in the presence of normotension and inappropriate renal electrolyte losses raised suspicion of an underlying renal tubular disorder. Elevated plasma renin and aldosterone levels supported the diagnosis of classical BS. Stabilization of serum electrolytes resulted in successful extubation and clinical improvement. This case highlights the importance of considering BS in infants with refractory electrolyte imbalance and unexplained ventilator dependence, particularly when renal salt wasting is evident.

Key words: Bartter Syndrome, Hypokalemia, Metabolic Alkalosis, Infant, Renal Tubular Transport Disorders, Respiratory Insufficiency

Bartter syndrome (BS) is a rare autosomal recessive renal tubular disorder first described by Bartter et al. in 1962 as a condition characterized by hypokalemic metabolic alkalosis and hyperplasia of the juxtaglomerular apparatus [1]. The estimated incidence is approximately 1 in 1,000,000 live births, though the true prevalence may be higher due to underdiagnosis [2]. BS comprises a heterogeneous group of disorders caused by defects in ion transporters in the thick ascending limb of the loop of Henle. Among its subtypes, classical BS (type III), caused by mutations in the CLCNKB gene encoding the basolateral chloride channel ClC-Kb, commonly presents in infancy or early childhood [3]. The pathophysiology involves impaired sodium, potassium, and chloride reabsorption, resulting in renal salt wasting, volume depletion, and activation of the renin–angiotensin–aldosterone system (RAAS) [4]. Despite elevated renin and aldosterone levels, patients remain normotensive due to renal prostaglandin overproduction and vascular hyporesponsiveness [5].

Clinically, infants may present with polyuria, polydipsia, failure to thrive, vomiting, dehydration, and growth retardation. Laboratory findings typically include hypokalemia, hypochloremia, metabolic alkalosis, elevated renin and aldosterone levels, and normal or increased urinary calcium excretion [6]. Diagnosis is based on clinical suspicion supported by biochemical evaluation and confirmed by genetic testing when available [7]. In critically ill infants, electrolyte abnormalities are frequently attributed to sepsis, medications, or nutritional deficiencies, which may delay recognition of an underlying tubulopathy. We report a case of classical Bartter syndrome presenting as refractory shock with recurrent extubation failure in the pediatric intensive care unit (PICU).

CASE PRESENTATION

A 9-month-old male infant, born to non-consanguineous parents, was admitted with fever, cough, respiratory distress, and poor feeding for five days. The child was born in term via normal vaginal delivery with a birth weight of 2.6 kg. There was no history of polyhydramnios, perinatal asphyxia, or neonatal intensive care admission. Immunizations were age-appropriate. There was no family history of renal disease,

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electrolyte disorders, or sibling deaths. On admission, the infant was lethargic, weighing 3.5 kg (<3rd percentile for age), tachypneic (respiratory rate 68/min), tachycardic (heart rate 172/min), with prolonged capillary refill time and hypotension (BP 70/40 mmHg). Oxygen saturation was 82% on room air. Anthropometry revealed severe failure to thrive. Systemic examination revealed bilateral crepitations on chest auscultation. No dysmorphic features or signs of rickets were noted (Figure 1).

Figure 1. Appearance of the child (After successful extubation).

The child was intubated and mechanically ventilated for severe pneumonia with septic shock. Initial laboratory investigations showed: serum sodium 124 mEq/L, potassium 2.3 mEq/L, chloride 82 mEq/L, bicarbonate 32 mEq/L, arterial pH 7.52, pCO_2 48 mmHg. Serum creatinine and calcium were within normal limits. Urinary sodium and potassium were inappropriately elevated. Blood cultures grew Klebsiella pneumoniae. Chest radiograph showed bilateral infiltrates consistent with pneumonia.

The infant received fluid resuscitation with isotonic saline (20 mL/kg boluses ×3), followed by maintenance fluids with appropriate electrolyte supplementation. Broad-spectrum antibiotics (intravenous meropenem and vancomycin, adjusted per sensitivity) were administered. Inotropic support with dopamine (10 µg/kg/min) was required for 48 hours. Potassium chloride was administered intravenously (0.5–1 mEq/kg/hr under monitoring), followed by oral supplementation.

Despite clinical improvement in pulmonary status by day 5, four attempts at extubation over the next week failed due to respiratory muscle weakness and recurrent hypokalemia (lowest recorded 2.1 mEq/L). Persistent hyponatremia and metabolic alkalosis were noted despite aggressive correction. Urine output remained high (3–4 mL/kg/hr) with normotension. There was no history of vomiting, diarrhea, or diuretic exposure.

Ultrasonography of the abdomen revealed normal-sized kidneys with preserved corticomedullary differentiation and no nephrocalcinosis. Serum magnesium levels were normal. Plasma renin activity and aldosterone levels were elevated. Based on persistent renal salt wasting, metabolic alkalosis, normotension, and exclusion of extrarenal causes, classical BS was diagnosed clinically. Genetic examination confirmed CLCNKB gene mutation, confirming the diagnosis of classical BS (Type III) (Figure 2).

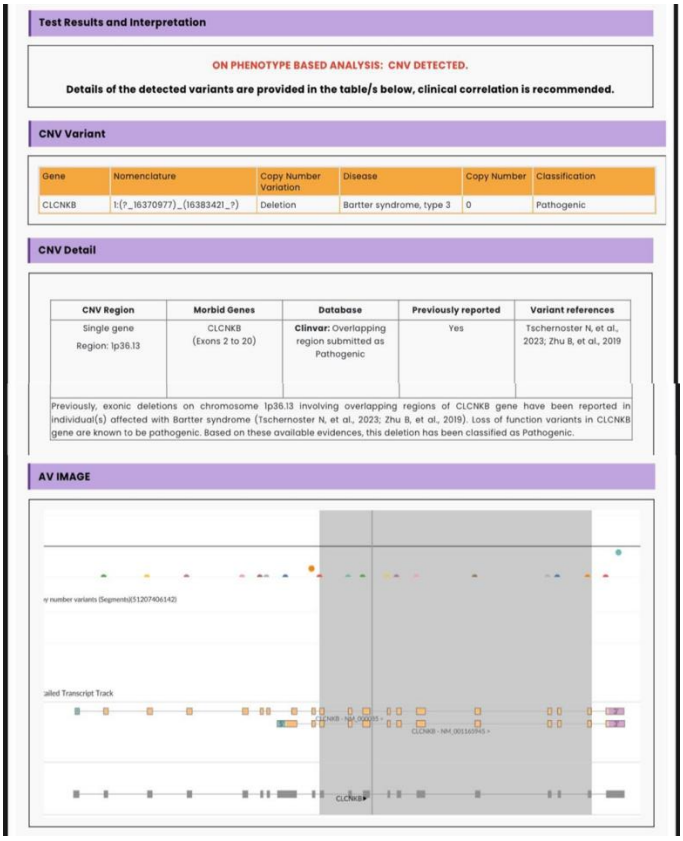

Figure 2. Genetic test results of the patient showing the CLCNKB gene mutation.

The child required mechanical ventilation for 12 days. After stabilization of electrolytes with oral potassium (3 mEq/kg/day) and sodium supplementation, successful extubation was achieved. The infant was shifted to the pediatric ward on day 15 and discharged on day 21 with oral potassium supplements and close nephrology follow-up. After a 3-month follow-up, weight gain and electrolyte levels showed improvement.

DISCUSSION

This case illustrates an atypical and severe presentation of classical Bartter syndrome manifesting as refractory shock and recurrent extubation failure. Persistent hypokalemia was the key factor contributing to respiratory muscle weakness and ventilator dependence. Hypokalemia impairs skeletal and diaphragmatic muscle function, predisposing to weaning failure in critically ill patients [8].

Classical Bartter syndrome (type III) results from CLCNKB mutations leading to defective basolateral chloride transport in the thick ascending limb [9]. Compared to

antenatal forms, type III may present later and with variable severity [10]. Studies have shown that growth failure and recurrent dehydration are common presenting features in infancy [11]. However, presentation during critical illness may obscure diagnosis, as electrolyte disturbances are often attributed to sepsis or medications.

Differentiation from Gitelman syndrome is essential. Gitelman syndrome typically presents in adolescence or adulthood and is characterized by hypomagnesemia and hypocalciuria [12]. In our case, normal magnesium levels and early infancy onset supported Bartter syndrome. Early diagnosis allows targeted management with electrolyte supplementation and, in some cases, prostaglandin synthesis inhibitors such as indomethacin [13].

The prognosis of classical Bartter syndrome is generally favorable with early recognition and treatment, although growth retardation and chronic kidney disease may occur in some cases [14]. The limitation of this case report is the absence of genetic confirmation due to resource constraints. Future recommendations include early genetic testing for definitive diagnosis and long-term follow-up to monitor growth and renal function.

CONCLUSION

Classical BS should be considered in infants with persistent hypokalemia, metabolic alkalosis, renal salt wasting, and unexplained ventilator dependence, even in the context of sepsis. Early recognition and appropriate electrolyte management are crucial to improving clinical outcomes and preventing recurrent complications.

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