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HAVE 'GUTS' TO SAY IT IS 'KIDNEY'-ANTENATAL BARTTER SYNDROME

Paediatrics

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ABSTRACT

Bartter syndrome is a rare renal tubular disorder characterized by hypokalemia, hypochloremic metabolic alkalosis, normal blood pressure with hyperreninemia and increased urinary loss of sodium, potassium and chloride1. We report a baby girl born preterm at 31 weeks with severe polyhydramnios NEC at 7 HOL, polyuria, hypochloremic, hypokalemia, significant weight loss, metabolic alkalosis consistent with renal tubulopathy – bartter syndrome. She was treated with aggressive fluid and electrolyte management and NSAID – Indomethacin.

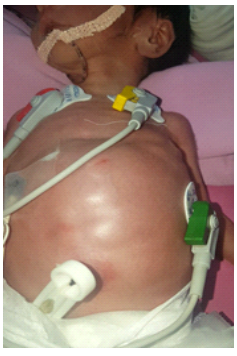
KEYWORDS

INTRODUCTION

Bartter syndrome is a group of disorders characterized by hypokalemic metabolic alkalosis with hypercalciuria and salt wasting[2]. It is a rare autosomal recessive disease with incidence of 1 in 1,000,000 according to framingham heart study[3]**. Antenatal** Bartter syndrome (types I, II, and IV; also called **hyperprostaglandin E syndrome**) typically manifests in infancy and has a more-severe phenotype than **classic** Bartter syndrome (type III). Genetic testing can confirm the diagnosis and specific mutation type; however, the diagnosis is primarily made clinically[2].

Case Report

A baby girl was born second in order to second degree consanguineous marriage delivered at 31 weeks of gestation through Emergency LSCS with birth weight of 1350 grams. Antenatal USG detected polyhydromnios at 21 weeks (AFI - 21) with increasing severity by 28 weeks (AFI – 36). Mother recieved Indomethacin (For polydromnios), Antibiotics and steroids (for lung maturity). Baby was managed in NNU from birth with regular vitals, Blood sampling & Urine assessment throughout the course of management. Baby developed Necrotizing entercollitis by 7 HOL triggered by Indomethacin given to mother antenatally.

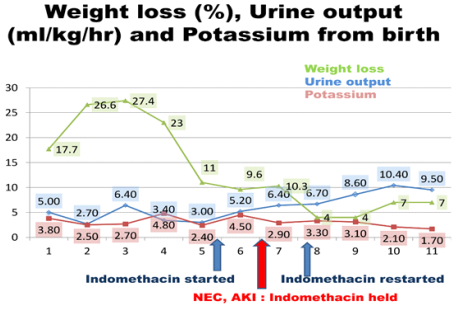


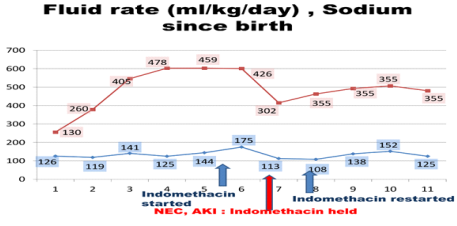
Persistent hyponatremia(126), hypochloremia(83), increased blood urea(58) Metabolic Alkalosis with weight loss of 19 percent were seen on DOL 5,that was managed as Pseudo-Bartter secondary to NEC.

Persisting Metabolic alkalosis and Low BP : Hypokalemic Hypochloremic Metabolic Alkalosis of Renal origin (Urine K^+ > 15 mmol/day, Urine Chloride > 20 mmol/day, Raised TransTubular Potassium Gradient, Raised Renin Activity and Hypercalciuria) established by Day 15 of life. Baby was nursed intensively, from trough weight of 900 gm (Br.Wt 1.35 kg) till 1.25 kg by correcting electrolyte/hydration, Renal tubulopathy / hyperaldosteronism is considered due to persisting dyselectrolytemia with ongoing polyuria and profound weight loss. Ongoing aggressive fluid and electrolyte management was given. Suspecting Renal Tubulopathy, Renin activity has been sent and Indomethacin was started at 0.5 mg/kg/day. After starting indomethacin baby improved hemodynamically. As volume status and weight improved blood pressure was recorded normal for age. Improvement in the biochemical parameters and fluid requirement is also noted after starting indomethacin.

	19-6-18	12-7-18 Before starting Indomethacin	23-7-18 After starting indomethacin
TTKG	10.3	14.4	12.55
FeK	24.45	15.52	16.6
FeNA	22	17.8	10.8
FeCa	26.52	5.69	
WEIGHT loss from birth	27% 950 grams	14% 1160 grams	9.6% 1220 grams

	Peak requirement	After Indomethacin
Sodium (mmol/ kg/ day)	37	11
Potassium (mmol/kg/day)	29	14
Fluid rate (ml /kg /day)	475 to 500	350





Review of Literature

Bartter syndrome is a rare renal tubulopathy first described by Frederic Bartter in 1962[4]. It is characterized by severe volume depletion, hypokalemia, metabolic alkalosis, hyperreninemia, and hyperaldosteronism due to mutations in genes encoding transporter proteins in channels of sodium, chloride, calcium, and potassium. There are two distinct presentations of Bartter syndrome (antenatal and classic)[5].

Antenatal Bartter syndrome[5]:

patients presenting with this form of the syndrome exhibit significant

symptoms from birth

Usual history of:

- Polyhydramnios (from fetal polyuria) starting between 24 and 30 weeks of gestation
- Preterm delivery
- Rapid weight loss immediately after birth
- Postnatal polyuria 6
- Polyuria that persists for 4 to 6 weeks after birth
- Chronic dehydration (marked by polydipsia) associated with varying degrees of: 7
- Recurrent vomiting
- Growth restriction
- Failure to thrive in infancy
- Poor feeding and lethargy may be noted if diagnosis is delayed.
- Hyposthenuria (urine of low specific gravity) is an important finding noted after birth
- Blood pressure is typically normal
- Distinctive facial features are common (eg, large eyes, triangular face, pointed ears, prominent forehead, drooping mouth/pouting expression)

The neonatal form differs from the classic Bartter syndrome by the age of onset, presence of nephrocalcinosis and very high urinary loss of sodium, calcium and chloride[6]. Other differential diagnoses are Gitelman's syndrome (characterized by hypomagnesemia, hypocalciuria), pseudohyperaldosteronism (hypertension with no evidence of increased secretion of mineralocorticoids) and pseudo-Bartter syndrome due to administration of high doses of prostaglandin E1.

The clinical diagnosis of Bartter or Gitelman syndrome is largely one of exclusion. Although plasma renin and aldosterone levels are not required for the diagnosis, they should both be elevated. Although genetic testing is increasingly available for many of the suspected gene mutations, there are likely many unrecognized mutations. The practicality of widespread genetic testing is limited[3].

Treatment:

Treatment of Bartter syndrome is directed at preventing dehydration, maintaining nutritional status, and correcting hypokalemia. Potassium supplementation, often at very high doses, is required. In addition, patients with Bartter syndrome are usually treated with nonsteroidal anti-inflammatory drugs (NSAIDs).

The long-term prognosis is guarded; lack of satisfactory control may lead to morbidity, growth failure and renal insufficiency.

CONCLUSION

1. High index of suspension is needed to diagnose Antenatal Bartter Syndrome
2. Managing hydration in the context of polyuria is challenging
3. Episodes of NEC on Indomethacin makes managing hydration more difficult
4. Indomethacin related NEC/AKI warrants close monitoring
5. Genetic diagnosis is possible and important for prognosis but is limited by availability and resources

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