

BARTTER SYNDROME

Asmat Ara Khattak, Irshad Ahmad and Mohammad Uzair

*Department of Paediatrics,
Postgraduate Medical Institute,
Lady Reading Hospital Peshawar.*

INTRODUCTION

Bartter syndrome is an uncommon renal tubular disorder.

The first case was reported in 1960 with subsequent three cases in 1962 and thereafter several cases have been reported. Bartter syndrome is characterized by hypokalemic, hypochloremic alkalosis, normal blood pressure despite elevated serum levels of angiotensin and marked hyperplasia of juxtaglomerular apparatus.

There is failure to thrive, polyuria, polydipsia, constipation, muscle weakness, salt craving, tetany and sometimes-renal osteodystrophy and renal insufficiency. Biochemical changes include hypokalemia, hyponatremia, hypomagnesemia and hypochloremic metabolic alkalosis. Urinary potassium and chloride excretion is increased inspite of low serum levels. Endocrine changes include very high plasma renin and angiotensin levels and increased aldosterone secretion rates with a normal urinary aldosterone excretion.¹ The diagnosis is suspected by finding characteristic biochemical changes already described and confirmed by histologic demonstration of hyperplasia of juxta-glomerular apparatus.²

CASE REPORT

A three months old male baby was referred to us from a peripheral hospital with the history of fever, fits, loose motions, vomiting and irritability. On examination he was found to be unwell, irritable, febrile, dehydrated with marked carpopedal spasm. His B.P was 75/50 mm Hg. Our initial diagnosis was sepsis / meningitis with electrolyte imbalance (hypocalcemia). His initial investigations showed serum sodium (Na^+) 135 mmol/L, Potassium (K^+) 2.2 mmol/L, Calcium (Ca^{++}) 10.5 mg%, Chloride (Cl^-) 90 mmol/L, blood urea 30 mg%, creatinine 0.6 mg% and urine specific gravity 1.080. His cerebrospinal fluid (C.S.F) analysis showed a clear fluid with glucose 93 mg%, Protein 20 mg %, white cell count (W.B.C) 12/cmm with a differential count of 4% polymorphs and 96% lymphocytes. C.S.F culture yielded no growth. The patient was resuscitated with intravenous fluids and electrolytes along with parenteral ceftriaxone and calcium gluconate infusion but the baby was still dehydrated, febrile and lethargic with carpopedal spasm inspite of the overnight fluids and calcium gluconate infusion. Repeat investigations revealed serum Na^+ 148mmol/L, K^+ 2.9 mmol/L, Ca^{++} 11.5mg% and Cl^- 90.6mmol/L. Calcium gluconate infusion was stopped. Arterial blood gases (A.B.G.s) were done and

- light and electron microscopic observations: *Pediatrics* 1971; 47(2): 254.
5. Laragh JH, Cannon PJ, Ames RP. Interaction between aldosterone secretion, sodium and potassium balance and angiotensin activity in man: studies in hypertension and cirrhosis: *Can. Med. Assoc. J.* 1964; 90: 248.
 6. Laragh JH, Kelly WG: Aldosterone: Its biochemistry and physiology: *Adv. Metab. Dis:* 1964; 1: 217.
 7. Cannon PJ, Lemming JM, Sommers SC, Winters RW, Laragh JH. Juxtaglomerular hyperplasia and secondary hyperaldosteronism (Bartter's syndrome): A reevaluation of the pathophysiology: *Medicine* 1968; 47:107.
 8. De Wardner HE, Mills IH, Clapham WF, Hayter CJ. Studies on the efferent mechanism of the sodium diuresis which follows the administration of intravenous saline in the dog: *Clin. Sci.* 1961; 21: 249.
 9. Mills IH, Wardner HE, Hayter CJ, Clapham WE. Studies on the afferent mechanism of the sodium chloride diuresis which follows intravenous saline in the dog. *Clin. Sci.* 1961; 21: 259.
 10. Shoemaker LR, Bergstrom W, Ragosta K, Welch TR. Humoral factor in children with neonatal Bartter syndrome reduces bone calcium uptake in vitro: *Pediatric. Nephrol.* June 1998; 12(5): 371.
 11. Mourani CC, Sanjad SA, Akatcherian-CY. Bartter syndrome in a neonate: early treatment with indomethacin: *Pediatr. Nephrol.* Febr 2000; 14(2): 143.
 12. Hussain W, Mahmood R, Maqbool S, Bartter syndrome: *Pakistan Paediatric Journal* Mar 1994; 18(1): 33.
 13. Rodriguez Soriano J. Bartter and related syndromes: the puzzle is almost solved: *Pediatric. Nephrol.* May 1998; 12(4): 315-27.
 14. Kikuchi M, Sato-M, Chiba A, Chiba Y, Nagao K, Suzuki T, Fujigaki-Y. Studies on the site of renal tubular defect in Bartter's syndrome: *Acta-Paediatr-Jpn.* June 1997; 39(3): 358.
 15. Calcagno PL, Hollerman CE. Renal alkalosis with hyperplasia of the Juxtaglomerular apparatus, hyperaldosteronism and normotension (Bartter syndrome): In Rubin MI, and Barrett TM (ed): *Pediatric Nephrology: William and Wilkins Baltimore* 1975; 671.
 16. Nakagawa Y, Toya K, Natsume H, Nasuda K, Takenchi H, Kubota A, Ogawa H and Igarashi Y: Longterm follow up of a girl with the neonatal form of Bartter syndrome: *Endocr J.* Apr. 1997; 44(2): 275.
 17. Reinalter S, Devleiger H, Proesmans W. Neonatal Bartter syndrome: spontaneous resolution of all signs and symptoms: *Pediatric. Nephrol.* Apr. 1998; 12(3): 186-8.
 18. Abdel-al YK, Badawi MH, Yaesh SA, Habib YQ, Khuffash FA, Ghanim MN, Najidi AK. Bartter syndrome in Arabic children: review of 13 cases: *Pediatric. Int.* Jun. 1999; 41(3): 299.

- light and electron microscopic observations: *Pediatrics* 1971; 47(2): 254.
5. Laragh JH, Cannon PJ, Ames RP. Interaction between aldosterone secretion, sodium and potassium balance and angiotensin activity in man: studies in hypertension and cirrhosis: *Can. Med. Assoc. J.* 1964; 90: 248.
 6. Laragh JH, Kelly WG: Aldosterone: Its biochemistry and physiology: *Adv. Metab. Dis:* 1964; 1: 217.
 7. Cannon PJ, Lemming JM, Sommers SC, Winters RW, Laragh JH. Juxtaglomerular hyperplasia and secondary hyperaldosteronism (Bartter's syndrome): A reevaluation of the pathophysiology: *Medicine* 1968; 47:107.
 8. De Wardner HE, Mills IH, Clapham WF, Hayter CJ. Studies on the efferent mechanism of the sodium diuresis which follows the administration of intravenous saline in the dog: *Clin. Sci.* 1961; 21: 249.
 9. Mills IH, Wardner HE, Hayter CJ, Clapham WE. Studies on the afferent mechanism of the sodium chloride diuresis which follows intravenous saline in the dog. *Clin. Sci.* 1961; 21: 259.
 10. Shoemaker LR, Bergstrom W, Ragosta K, Welch TR. Humoral factor in children with neonatal Bartter syndrome reduces bone calcium uptake in vitro: *Pediatric. Nephrol.* June 1998; 12(5): 371.
 11. Mourani CC, Sanjad SA, Akatcherián-CY. Bartter syndrome in a neonate: early treatment with indomethacin: *Pediatr. Nephrol.* Febr 2000; 14(2): 143.
 12. Hussain W, Mahmood R, Maqbool S. Bartter syndrome: Pakistan Paediatric Journal Mar 1994; 18(1): 33.
 13. Rodriguez Soriano J. Bartter and related syndromes: the puzzle is almost solved: *Pediatric. Nephrol.* May 1998; 12(4): 315-27.
 14. Kikuchi M, Sato-M, Chiba A, Chiba Y, Nagao K, Suzuki T, Fujigaki-Y. Studies on the site of renal tubular defect in Bartter's syndrome: *Acta-Paediatr-Jpn.* June 1997; 39(3): 358.
 15. Calcagno PL, Hollerman CE. Renal alkalosis with hyperplasia of the Juxtaglomerular apparatus, hyperaldosteronism and normotension (Bartter syndrome): In Rubin MI, and Barrett TM (ed): *Pediatric Nephrology*: William and Wilkins Baltimore 1975; 671.
 16. Nakagawa Y, Toya K, Natsume H, Nasuda K, Takenchi H, Kubota A, Ogawa H and Igarashi Y: Longterm follow up of a girl with the neonatal form of Bartter syndrome: *Endocr J.* Apr. 1997; 44(2): 275.
 17. Reinalter S, Devleiger H, Proesmans W. Neonatal Bartter syndrome: spontaneous resolution of all signs and symptoms: *Pediatric. Nephrol.* Apr. 1998; 12(3): 186-8.
 18. Abdel-al YK, Badawi MH, Yaeesh SA, Habib YQ, Khuffash FA, Ghanim MN, Najidi AK. Bartter syndrome in Arabic children: review of 13 cases: *Pediatric. Int.* Jun. 1999; 41(3): 299.