Congenital Nephropathy and Chronic Diarrhoea with Hypokalemic Alkalosis

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Abstract. Long-term observation and laboratory investigations of a female infant with chronic diarrhoea, hypokalemia, alkalosis and hypochloremia are presented. Diarrhoea with hypokalemic alkalosis persisted despite large-scale potassium chloride supplementation. An intravenous pyelogram showed duplication of the right pelvis and fetal lobate kidneys. She was normotensive. Plasma renin activity was increased. A permanent loss of electrolytes through urinary and digestive tracts was observed. Renal biopsy revealed the presence of fetal-like glomeruli, hyperplasia of juxtaglomerular apparatus and thickening of arterial walls. Hypertrophy and hyperplasia of the zona glomerulosa of adrenal cortex were present.

It is supposed that a case presenting with co-existence of electrolytes disturbances on digestive and urinary tracts stands between congenital chloride diarrhoea and Bartter's syndrome, being an unknown kind of congenital hypokalemia.

Key words: Congenital nephropathy — Chloride diarrhoea — Bartter's syndrome — Hypokalemic alkalosis.

Congenital chloride diarrhoea is a clinical syndrome characterized by a loss of electrolytes with stools and a presence of secondary hyperaldosteronism [6, 9, 11]. Many authors say that this syndrome results from primary disturbances of active electrolyte transport in bowels. Inhibition of electrolytes reabsorption leads to watery diarrhoea [11, 12]. Aaronson's observations [1] on several patients after jejunal resection who developed watery diarrhoea with chloride losing seem to support this hypothesis.

In 1962, Bartter et al. [2] described a syndrome of normotensive secondary hyperaldosteronism associated with hyperplasia of juxtaglomerular apparatus and a presence of fetal-like glomeruli. The function of digestive tract appears to be normal. Loss of electrolytes takes place through kidneys [4, 8, 14, 16, 17].

Royer et al. [13] include these both syndromes to congenital chronic hypokalemia emphasizing many symptoms as hypokalemic alkalosis.
with hypochloremia, juxtaglomerular hyperplasia, elevated renin activity, normal blood pressure, dwarfism.

There are many unexplained points in these syndromes and this communication describes another case presenting unknown combination of symptoms which may clarify some mechanisms governing electrolytes reabsorption in man.

Methods

Urine and stool were separately collected in 12-hour aliquots. Serum and urine sodium and potassium by flame photometry, pH of constitutional fluids by pH-meter (PHM 71), chloride by mercurio-metric method were determined.

Acid-base balance parameters on ABC 1 Radiometer apparatus using Siggaard-Andersen curve nomogram were determined.

Plasma renin activity was assayed by modification of Boucher's et al. method [10].

Case Report

A six-month-old infant girl was admitted to Department of Pediatrics in Zabrze because of proteinuria and chronic watery diarrhoea appearing from the second week of life.

Pregnancy proceeded normally. Gestation was 37 weeks. Labor and delivery were uncomplicated and her birth weight was 2200 g, length 51 cm.

At admission she was dangerous ill, with pale skin, decreased muscles tonus, significant dehydration, weight 5200 g. She passed numerous, watery stools. Diarrhoea with toxicosis was at first diagnosed. After a 10 days period of intravenous and peroral rehydration under the control of acid-base and electrolytes balance her general state was much improved (Fig. 1). She did not need more intravenous therapy but her stools were all the time watery without blood and mucus.

Blood counts, serum calcium and phosphorus, alkaline phosphatase, SGOT, SGPT, LDH, bilirubin, creatinine, urea, chest and skull X-rays, stools investigations (repeated cultures), rate of digestion, content of lactic acid and reduction substances, pancreatic enzymes in duodenal juice were all normal or negative. Glucose tolerance curve was normal. Urinanalyses revealed specific gravities between 1.016—1.023, pH usually above 7.0, proteinuria to 10% occasionally cylinduria, 17-hydroxy- and 17-keto-steroids excretions were normal. Electrolytes excretion was considerably increased, at the beginning especially sodium and chloride, but after several weeks also potassium (Table 1).

Investigations of acid-base and electrolytes serum balance, in many occasions for all the period of observation, showed significant disturbances in the direction of hypokalemic alkalosis with moderate hypochloremia (Table 1). Serum magnesium levels were 1.3—1.9 mg\% (normal values 2.5—3.2 mg\%).

Intravenous pyelography demonstrated a duplication of the right pelvis with slight dilatation. Abdominal aortography showed normal adrenal and renal arteries and in the phase of nephrogram typical shapes for fetal lobate kidneys.