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Renal sonographic patterns in Bartter's syndrome

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Abstract The renal sonographic findings in ten cases of Bartter's syndrome investigated at the King Khalid University Hospital, Riyadh, Saudi Arabia are described. There were various sonographic abnormalities other than those of hyperechoic pyramids as previously described. These were diffuse increased renal echogenicity and hyperechoic echogenicity in the kidneys with the exception of the pyramids. This condition can be suspected early if nephrocalcinosis is present in a child with a history of polyhydramnios and premature delivery.

Introduction

In 1962, Bartter and colleagues [1] described the syndrome of hypokalemia, alkalosis, hyperaldosteronism, hyperreninism, hypertrophy of the juxtaglomerular apparatus, and normotension. The basic aetiology of this syndrome is the presence of defects in the tubular transport mechanism [2]. There are a few reports of Bartter's syndrome showing only medullary nephrocalcinosis on sonographic examination [3, 4, 5]. This is a review of ten cases with documentation of the various renal sonographic echopatterns, including those which have not been previously described.

Materials and methods

The renal sonographic appearances of ten known cases of documented Bartter's syndrome occurring in King Khalid University Hospital, Riyadh, Saudi Arabia, from April 1982 to April 1994 were reviewed. The diagnoses of the ten cases were based on combined clinical presentations and laboratory investigations, with the patients fulfilling the criteria for the diagnosis of Bartter's syndrome of hypokalemic metabolic alkalosis with normal blood pressure.

Results

Clinical findings

The clinical findings are shown in Table 1. There were four girls and six boys; in six cases there was parental consanguinity, with two patients (cases 7 and 8) being siblings. The symptom complex consisted of failure to thrive (ten cases), developmental delay (nine cases), polyuria with polydipsia (seven cases), and constipation (five cases). A history of preterm delivery with polyhydramnios during intrauterine life was obtained in seven cases.

The follow-up ranged from 1 month to 9 years. All the patients maintained normal serum creatinine levels and corrected creatinine clearance except patient 9 who presented at the age of 7 years. He developed extensive nephrocalcinosis and chronic renal failure and is currently on conservative medical therapy. Treatment with indomethacin was tried but the patient developed duodenal ulceration necessitating the withdrawal of the medication. Patients 7 and 8 were also treated with indomethacin for a 1-year period but showed no significant improvement. Repeated bouts of vomiting led to the withdrawal of the drug.

Renal sonograms were obtained with a real-time sector or linear scanner with either a 5- or 3.5-MHz trans-

Table 1 Clinical presentation of ten patients with Bartter's syndrome

Case	Sex	Gestational age (weeks)	Consanguinity	Polyhydramnios	Polyuria/ polydipsia	Failure to thrive	Constipation	Developmental delay
1	F	29	–	+	+	+	+	+
2	M	28	+	+	+	+	+	+
3	F	38	–	–	+	+	–	+
4	M	34	+	–	–	+	–	+
5	M	34	–	+	–	+	–	+
6	F	40	+	–	–	+	–	–
7	M	34	+	+	+	+	–	+
8	F	32	+	+	+	+	–	+
9	M	39	–	–	+	+	–	+
10	M	39	+	+	+	+	–	+

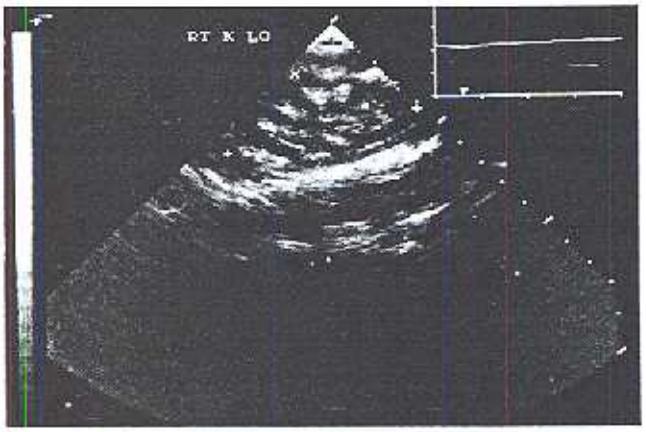

Fig 1. Longitudinal *(LO)* right *(RT)* renal sonogram shows only hyperechoic pyramids. *K.* Kidney

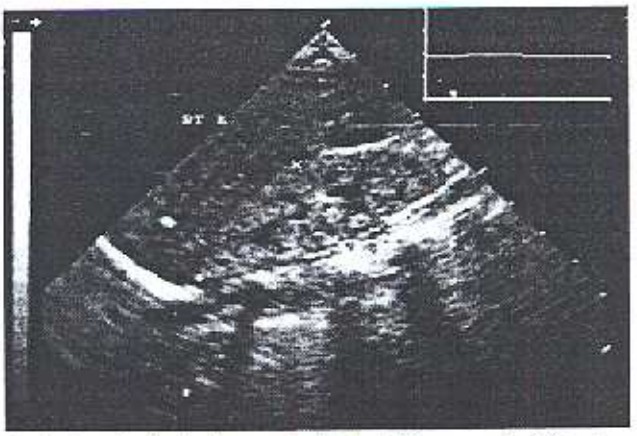

Fig. 2 Longitudinal ultrasound of right kidney reveals diffuse hyperechoic parenchyma

ducer. Renal echopatterns were recorded in the following groups:

A. Normal: the echo intensity of the cortex of the right kidney is less than that of the liver; the renal medulla is hypoechoic compared to the renal cortex

B. Hyperechoic renal medullary pyramids (partial or complete; Fig. 1)

C. Diffuse increased renal cortical and medullary echogenicity (Fig. 2)

D. Diffuse increased renal echogenicity except in the medullary pyramids (Fig. 3)

The renal sonographic findings are shown in Table 2. Five cases were normal at the onset of the study and two of these (3 and 7) remained normal throughout, while three others developed either hyperechoic pyramids (cases 1 and 8) or diffuse increased parenchymal echogenicity (case 9). Case 2 showed hyperechoic medulla at an early stage but progressed to diffuse parenchymal increased echogenicity.

Laboratory findings

The laboratory findings are shown in Table 3. All patients had abnormally high serum renin and high serum aldosterone levels ranging from 18.3 to 70 ng/ml/h and 35 to 179 mg/dl, respectively, with a normal range of 0.68–1.36 ng/ml/h and 6–22 mg/dl, respectively. The results shown in Table 3 confirm hypokalemic metabolic alkalosis. Two patients (1 and 7) had no hypercalciuria (hypercalciuria was defined by the findings of a random urinary calcium/creatinine molar ratio of more than 0.7).

Discussion

Despite the common use of ultrasound in renal disorders, the sonographic findings in Bartter's syndrome are rarely demonstrated due to paucity of such cases [3–6]. Few cases have been reported in the literature, the largest series being of five cases [4, 5]. Nephrocalcinosis is the commonest sonographic finding, but it is of-

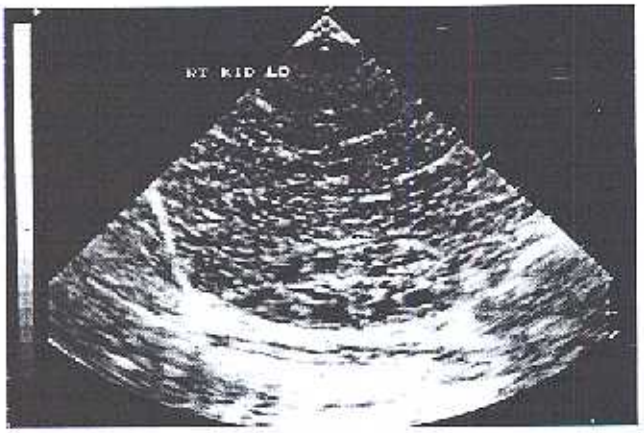

Fig. 3 Longitudinal *(LO)* ultrasound of right kidney *(RT KID)* shows hyperechoic parenchyma with the exception of the renal pyramids

Table 2 Renal sonographic findings

Case	Normal	Hyperechoic pyramids	Hyperechoic renal echogenicity except pyramids	Diffuse increased renal echogenicity
1	–	+	–	–
2	–	–	–	+
3	+	–	–	–
4	–	–	–	+
5	–	–	–	+
6	–	–	+	–
7	+	–	–	–
8	–	+	–	–
9	–	–	–	+
10	–	–	–	+

Table 3 Laboratory findings: serum mmol/l

Case	Na	K	Cl	HCO_3	pH
1	132	2.9	90	30	7.5
2	135	2.4	86	39	7.5
3	138	2.9	97	28	7.5
4	127	2.7	65	42	7.6
5	132	3	85	30	7.7
6	131	1.8	70	30	7.6
7	138	2.3	90	29	7.49
8	139	2.2	89	30	7.47
9	130	2.1	85	32	7.49
10	132	2.5	94	28	7.47

ten microscopic and acoustic shadowing is not always present [3, 7].

Children under 3 months of age often show renal cortical echogenicity similar to the adjacent liver parenchyma, and this is a normal finding at birth. In children above 3 months of age, the normal renal cortical echogenicity is less than that of the adjacent liver parenchyma [8]. All our patients were above 6 months of age at the time of study. The quoted incidence of nephrocalcinosis in Bartter's syndrome shown by sonography is 66.7% compared to 80% in the present series [4].

Two of our patients (1 and 8) showed only increased echogenicity in the medullary pyramids similar to previous reports [3, 5]. One showed peripheral medullary hyperechogenicity similar to that described by Garel et al. [4]. Two other patterns not previously described were seen. Firstly, cortical hyperechogenicity (that is of the kidney other than the renal pyramid) occurred in one case, and is rare (Fig. 3). Secondly, diffuse renal hyperechoic parenchyma of cortices and medulla was shown in five cases (Fig. 2).

Nephrocalcinosis was explained on the basis of an increase in the level of prostaglandin-E_2, which stimulates conversion of the inactive metabolite of vitamin D to the active form. This, in turn, causes an increase in the absorption of calcium from the gastrointestinal tract, resulting in an increase in the urinary excretion of calcium. Precipitation of calcium occurs in the collecting tubules of the medullary pyramids [5], where the urinary concentration of calcium is highest due to the effect of the antidiuretic hormone (nephrocalcinosis). However, extension into the tubules within the cortex is expected in severe and chronic cases. This is proven by our study, which showed progression from normal appearance to hyperechoic pyramids or diffuse parenchymal echogenicity with the passage of time. This view is further strengthened by the fact that the follow-up of our cases ranged from 1 month to 9 years, compared to the series of Garel et al. which ranged from 0.4 to 4 years with an average of 1.44 years. Moreover, the fact that one of our two patients with no hypercalciuria showed normal sonographic appearances (case 7) while the other had only hyperechoic pyramids (case 1) proves the progression and extension of nephrocalcinosis with time. Our study also showed that those with increased renal echogenicity had marked hypercalciuria, which is at variance with previous observations [4].

The differential diagnosis of nephrocalcinosis in children includes renal tubular acidosis, chronic therapy with frusemide, primary and secondary hyperparathyroidism, vitamin A intoxication, Cushing's syndrome and Fanconi's syndrome. Bartter's syndrome with hypercalciuria is not widely recognised as a cause of nephrocalcinosis. Polyhydramnios and prematurity have been described as associations of the syndrome, and were present in seven of our patients [9]. Bartter's syndrome should always be considered if nephrocalcinosis is present in a preterm infant with a history of polyhydramnios. Early diagnosis is important because, with treatment, there is correction of the hypercalciuria, as well as a decrease in nephrocalcinosis, with preservation of renal function.

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