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# Salt losing nephropathy simulating congenital adrenal hyperplasia in infants with obstructive uropathy and/or vesicoureteral reflux – value of ultrasonography in diagnosis

**T. L. Levin[1], S. J. Abramson[1], K. A. Burbige[2], J. P. Connor[3], C. Ruzal-Shapiro[1] and W. E. Berdon[1]**

[1] Department of Pediatric Radiology,
[2] Department of Pediatric Urology, Columbia Presbyterian Medical Center, Babies Hospital, and
[3] Department of Urology, Memorial Sloan-Kettering Cancer Center, New York, USA

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**Abstract.** Salt losing nephropathy, occurring predominantly in male infants, has been reported in association with a spectrum of urologic diseases including obstructive uropathy and massive, infected vesicoureteral reflux (VUR). This has been called pseudo-hypoaldosteronism (PHA) or alternatively, pseudo salt-losing congenital adrenal hyperplasia (CAH), and is thought to reflect a tubular unresponsiveness to aldosterone. We report our experience with six cases, discuss one case in detail and review the 39 cases previously reported. A one month old male infant presented with a left upper quadrant mass. Signs and symptoms included vomiting, dehydration, hyponatremia and hyperkalemia. This suggested the diagnosis of CAH for which therapy was instituted. Ultrasonographic examination subsequently revealed the mass to be a urinoma in an infant with posterior urethral valve (PUV) and obstructive hydronephrosis.

Pseudo-hypoaldosteronism (PHA), as originally described in 1958 refers to a salt losing condition seen in infants with vomiting, dehydration, hyponatremia and hyperkalemia. These infants have normal adrenal function and normal renal morphology [1, 2]. A subsequent group of infants have been reported using the same term PHA, or alternatively pseudo salt-losing CAH, in whom a variety of obstructive and non-obstructive renal lesions have been identified [3–6]. Treatment of the urological abnormality often leads to resolution of the salt wasting syndrome. Clinical presentation in these infants is confusing and may lead to an incorrect diagnosis of CAH. We present our experience with six such cases, one of which is described in detail, as well as a review of the literature.

## Case report

The findings in Case 1 are discussed in detail. Pertinent information regarding the remaining cases is presented in Table 1.

T. J., a one month old previously well male infant, presented to an outside institution with vomiting and dehydration and was admitted to rule out hypertrophic pyloric stenosis. On admission, a left upper quadrant abdominal mass was palpated. Ultrasonography, limited to the upper abdomen, demonstrated a cystic structure in the left upper quadrant and follwing a pediatric surgical consultation, the diagnosis of gastric duplication was made. Admission laboratory results revealed hyponatremia, hyperkalemia and metabolic acidosis, which led to a diagnosis of salt-losing CAH.

The infant was transferred to Babies Hospital for endocrinologic treatment of CAH and for surgical correction of the gastric duplication. Upon transfer, the infant was seen by an endocrinologist and received intramuscular cortisone acetate. A repeat ultrasonogram was obtained to evaluate the abdominal mass, and in the setting of possible CAH, to visualize both adrenal glands, and to confirm the presence or absence of a uterus. The ultrasonogram demonstrated a thick-walled bladder, dilated posterior urethra (Fig. 1), bilateral hydronephrosis, and a left perinephric fluid collection consistent with urinoma (Fig. 2). The urinoma was the source of the initial ultrasonographic diagnosis of gastric duplication. Voiding cystourethrography confirmed posterior urethral valve, and demonstrated bilateral VUR and filling of a left urinoma (Fig. 3).

The patient underwent drainage of the kidney and left urinoma, and transurethral resection of the posterior urethral valve. The patient's electrolytes subsequently returned to normal and he has done well.

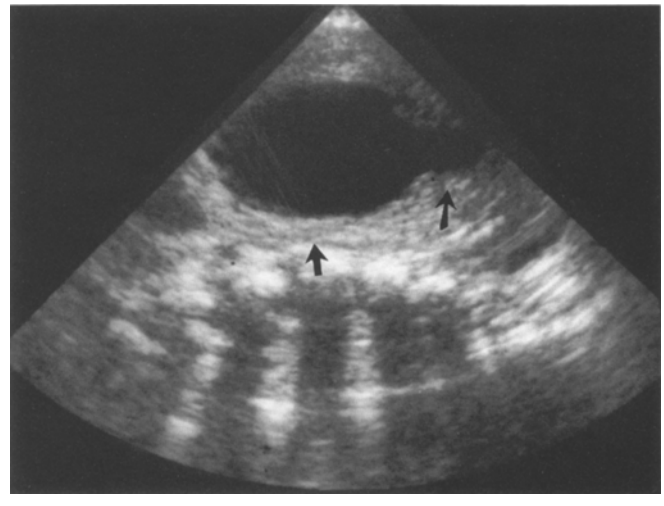

**Fig. 1.** Longitudinal sonogram of the bladder demonstrates a thickened bladder wall *(straight black arrow)* and a dilated posterior urethra *(curved black arrow)*

**Table 1.** Clinical and laboratory findings

| Patient | Age | Sex | Clinical features | Serum values | | Urine Na (m Eq/L) (range 43–217) | Diagnosis | Therapy |
|---|---|---|---|---|---|---|---|---|
| | | | | Na (m Eq/L) normal (135–145 m Eq/L) | K (m Eq/L) normal (3.2–4.6 m Eq/L) | | | |
| 1. TJ | 1 mo | M | Poor feeding Vomiting Abdominal mass | 121 | 9.7 | 85 | PUV Left urinoma Bilateral VUR (Grade 4) | Inappropriate cortisone acetate injection. Open drainage of urinoma. Transurethral resection of valve. |
| 2. JR | 1 mo | M | Fever Vomiting Irritability | 119 | 9.7 | – | PUV Sepsis Bilateral VUR (Grade 3) | Inappropriate cortisone acetate injection. Transurethral resection of valve. Bilateral urethral reimplantation. |
| 3. JS | 3 wks | M | Vomiting | 117 | 7.2 | 75 | PUV Right urinoma (Fig 4) | Transurethral resection of valve. |
| 4. DB | 6 mo | M | Dehydration | 124 | 5.9 | – | Left ectopic ureterocele Bilateral renal duplication Bilateral VUR (Grade 4) | Left ureterocele excision. Bilateral ureteral reimplantation. |
| 5. KG | 3 wks | M | Fever Vomiting Left flank mass | 111 | 6.5 | 52 | Bilateral renal duplication. Staph. aureus sepsis Bilateral VUR (Grade 4) | Bilateral ureteral reimplantation. |
| 6. SW | 2 mo | F | Poor feeding Dehydration | 116 | 6 | – | Bilateral VUR (Grade 3) | Bilateral ureteral reimplantation. |

## Discussion

Fetal hydronephrosis, whether due to obstruction or VUR, is recognized with increasing frequency due to the wide use of prenatal ultrasound. Once the finding is made, post natal confirmation of the diagnosis includes ultrasonography, voiding cystourethrography, intravenous pyelography or renal scintigraphy.

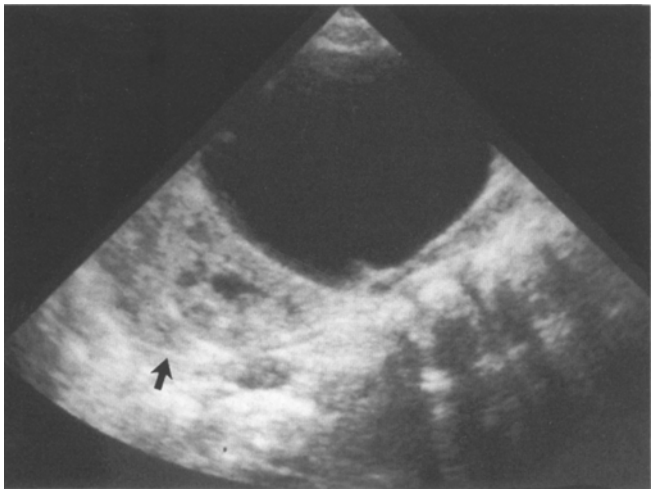

**Fig. 2.** Longitudinal sonogram of the left flank shows a large cystic collection compressing the left kidney *(arrow)*. This proved to be a urinoma. Mild dilatation of the left renal collecting system is also shown

There are a group of infants in whom the diagnosis of a primary urologic lesion has not been made prenatally. These infants present days to weeks postnatally with a severe salt-losing renal dysfunction termed PHA or pseudo salt losing CAH. This is characterized by hyponatremia, hyperkalemia, dehydration and metabolic acidosis. Although adrenal function is normal in these infants, their presentation has suggested CAH and has led to inappropriate therapy.

A wide variety of urologic lesions has been reported in association with this salt wasting syndrome [3–6]. These include both obstructive and non-obstructive lesions (Table 2). The obstructive lesions that have been described include PUV, ectopic ureterocele and ureteropelvic junction obstruction. Non-obstructive lesions consist of massive VUR, usually accompanied by urinary tract infection. Reflux may be unilateral or bilateral, or may be segmental as occurs with reflux into the lower pole of a renal duplication.

Assessment of the urologic abnormalities in the reported cases is limited by the lack of imaging studies in the majority of the previously reports [3–6]. These papers have defined "obstructive uropathy" in a very broad sense; included in these cases of obstructive uropathies were a significant number of infants described as having

**Table 2.** Review of previously reported and current cases

| | # Pts. | Male | PUV infection | | UPJ | UVJ | Uretero-cele | Bilat vur Infection | | Uni vur Infection | | Other |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | with | w. o. | | | | with | w. o. | with | w. o. | |
| Soriano [3] | 6 | 5 | 0 | 1 | 0 | 2[b] | 1 | 1 | 0 | 2 | 0 | |
| Heijden [4] | 11 | 11 | 2 | 2 | 0 | 5[b] | 0 | 0 | 1 | 0 | 0 | 1[c] |
| Marra [5] | 18 | 12 | 0 | 1 | 13[a] | 1 | 0 | 0 | 1 | 0 | 1 | |
| Vaid [6] | 3 | 3 | 0 | 0 | 0 | 1[b] | 0 | 3 | 0 | 0 | 0 | |
| Levin | 6 | 5 | 1 | 2 | 0 | 0 | 1 | 1 | 1 | 0 | 0 | |

[a] In 5 infants, UPJ obstruction was bilateral, [b] Associated with infection, [c] Infant with neurogenic bladder

ureterovesical junction (UVJ) stenosis [3–5]. Since there were no published images of these cases, it is unclear whether the cases reported as UVJ stenosis truly represent an obstructive uropathy, or rather, represent hydroureteronephrosis on a different basis such as atonic ureteral dilatation in association with infection. Vesicoureteral reflux was included as "obstructive uropathy" since it too raised intrapelvic pressure [4].

It is, therefore, difficult to invoke a single explanation for the development of salt wasting in these infants with such varied renal abnormalities. A transient unresponsiveness of the distal renal tubules to aldosterone is believed to occur [3]. What leads to this unresponsiveness is uncertain, however, increased intrarenal pressure whether due to obstruction or reflux may, in part, be responsible for the tubular dysfunction. It remains unclear why most infants with severe VUR or true obstructive lesions do not develop this entity and why others with only unilateral or segmental renal abnormalities do.

We believe ultrasonography (US) is the most useful tool in the evaluation of these infants with signs and symptoms of salt wasting. In cases where CAH is considered, US can confirm the presence or absence of a uterus, and assess the adrenal glands which may be enlarged in CAH [7]. As evidenced by the case presented, US is also useful in diagnosing the associated renal abnormalities that may be present in an infant with PHA, and which may mimic CAH. In the case discussed, US demonstrated a urinoma which had been incorrectly diagnosed as a gastric duplication cyst. US was able to diagnose the cause of the urinoma as being due to PUV, by demonstrating a thick walled bladder and enlarged posterior urethra. At the time of diagnosis, the patient had already started inappropriate therapy for presumed salt-losing CAH. While urinoma is a known complication of PUV, it has not previously been reported in association with salt losing nephropathy.

Salt losing nephropathy in association with a primary urologic abnormality is not a common entity. It is not found in the standard pediatric endocrinology tests, nor is it mentioned in the standard pediatric or pediatric radiology texts. Insofar as its presenting signs simulate salt losing CAH, US is warranted as the initial study to exclude a renal lesion as the cause of the electrolyte disturbances.

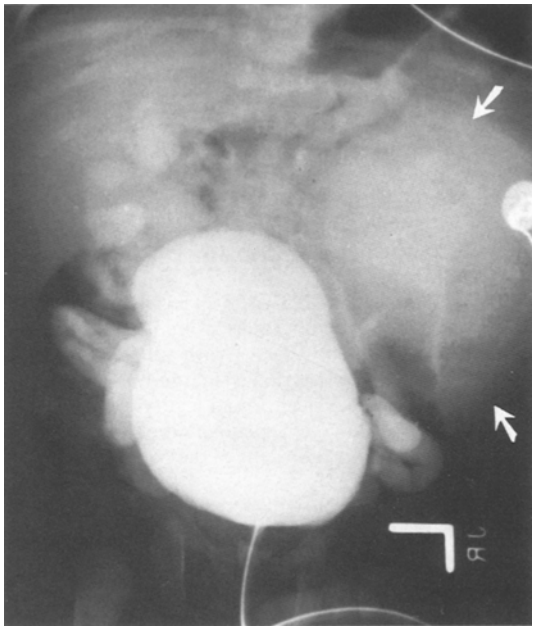

**Fig. 3.** Image from a voiding cystourethrogram shows bilateral VUR and contrast filling the left urinoma *(arrows)* which has displaced the left kidney and ureter medially

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Dr. S. J. Abramson
Department of Radiology
Babies Hospital
3959 Broadway
New York, N. Y. 10032, USA