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Pseudohypoaldosteronism in a Family with Variable Presentation

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Pseudohypoaldosteronism is a condition of infancy characterized by salt wasting and failure to thrive(l). Renal salt loss with elevation of plasma aldosterone and renin is thought to reflect an unresponsiveness of renal tubules to mineralocorticoid hormones(2). A family with three children of pseudohypoaldosteronism is presented.

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Case Report

The index patient was a male infant born to Saudi parents by non-consanguineous marriage. He was asymptomatic, but investigations were done due to positive family history of salt wasting and concern of parents to rule out an endocrine disease. Investigations revealed the following: serum

sodium-139 mmol/L; serum potassium-4.4 mmol/L; serum chloride-110 mmol/L; serum bicarbonate-22 mmol/L; plasma urea-19 mg/dl; random sugar-78 mg/dl; 24 hour urinary sodium-40 mmol/L; plasma renin-88.1 pg/ml; plasma aldosterone-1740.8 pg/ml; and serum cortisol-45.1 nmol/L. The baby on follow up at one and six months was doing well without salt wasting symptoms and had normal serum electrolytes. He received no treatment.

Case 2: A male baby, second cousin of index patient was born to consanguineous parents. On day six, there was sudden onset of vomiting and shock. Investigations revealed serum sodium-120 mmol/1; serum potassium-10 mmol/1; serum cortisol-normal, plasma aldosterone-1634 pg/ml; and plasma renin-50 pg/ml. The baby was given hydrocortisone and mineralocorticoid but there was no response. A diagnosis of pseudohypoaldosteronism was considered and intravenous and later oral sodium was given. The infant improved clinically and biochemically. The child as now 4 years old and growing well with oral salt supplement of 400 mmol per day.

Case 3: A second cousin sister of index case was a product of consanguineous marriage with uneventive pregnancy and delivery. In the light of family history, plasma electrolyte level and body weight were closely monitored. By day 5, the baby had lost 10% of her weight and hyperkalemia (sodium-135 mmol/L, and potassium 8 mmol/L) was noted. High doses of intravenous saline returned the plasma electrolytes to normal. Her plasma aldosterone and renin levels were high. At present, the child is 2 years old; she is maintaining normal electrolytes on oral sodium intake of 300 mmol/day and her growth is appropriate for her age.

Discussion

The cases described in the present paper show features typical of pseudohypoaldosteronism(2,3). This disorder is characterized by salt wasting in infancy that is responsive to supplementary sodium but not to mineralocorticoids and recently it has been subcategorized as pseudohypoaldoste-ronism type 1 and type 2(4). Excess aldosterone and renin are always present in these children as was seen in the reported cases. Salt supplementation can often be discontinued after infancy without adverse effects. The family pedigree of our cases revealed death of 5 babies within 2 weeks of birth indicating a severe defect. Babies with moderate defect required sodium supplement while the index patient did not require any salt supplement as he«w^s probably suffering from only a biochemical defect.

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