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Transient Nephrogenic Diabetes Insipidus in a Neonate

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Nephrogenic diabetes inspidus (NDI) is a rare disorder which manifests with polyuria, passage of dilute urine concomitant with serum hyperosmolality, with no response to exogenously administered vasopressin. The disease can be inherited or can be secondary to various insults to

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the distal tubules which impair the concentrating capacity of the kidney(1). We report here a case of transient NDI in a neonate after an episode of intravascular hemolysis; so far unreported in literature.

Case Report

A boy of 34 weeks gestation was born by spontaneous vaginal delivery to a 34 year old ninth gravida mother. He weighed 2200 gm at birth and had Apgar scores of 1 and 4 at 1 and 5 minutes, respectively. The child developed hyaline membrane disease which required respiratory support in the form of continuous mandatory ventilation for 48 hours.

The baby had rhesus isoimmunization for which he received two double volume exchange transfusions. Ninety minutes after the second exchange transfusion (on day 1 of life), the baby started passing cola-colored urine. The blood used for the second exchange transfusion was later found to be deficient in glucose-6-phosphate dehydrogenase (G6PD). Two more exchange transfusions were done for rising bilirubin on day 2. Polyuria was detected on day 4 and did not correspond to the improvement of the respiratory distress. Daily urine output and specific gravity were as shown in *Table I*. Serum osmolality done on day 4 and 5 were 332 and 334 mosm/L, respectively. Concomitant urine osmolality was 196 mosm/L on day 5. *Table II* outlines the other relevant investigations done during the period of polyuria.

In view of polyuria with serum hyperosmolality with hyposthenuria, a diagnosis of diabetic insipidus was entertained. A dose of 1 unit of l-AVP was administered intravenously on day 5 and urine osmolality monitored (*Fig.*). Urine remained dilute despite vasopressin administration, thus confirming the diagnosis of nephrogenic diabetes insipidus.

TABLE I—*Urinary Output and Specific Gravity in Relation to Age of Child*

Age (day of life)	Urine	
	Output (ml/kg/h)	Specific gravity
1	1.64	1010
2	1.42	1010
3	3.9	1005
4	5.3	1005
5	6.0	1010
6	3.0	1008
7	2.3	1008
8	1.85	1005
9	3.3	1005
10	3.2	1005

The cola-colored urine started clearing on day 4 of life; polyuria became passive on day 6. Azotemia (*Table II*) noted on days 4 and 5 subsided on day 6. Ultrasound examination of the cranium revealed a left germinal matrix bleed; the kidneys were essentially normal. The subsequent hospital course of the baby was uneventful. He was discharged at the age of 20 days.

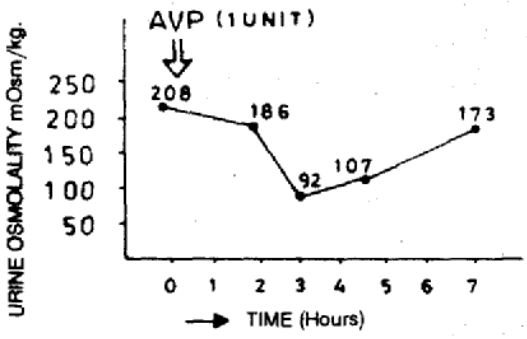

Fig. Graph showing response of urine osmolality to vasopressin administration.

TABLE II—*Other Relevant Investigations*

Age (day of life)	Sodium (mEq/L)	Potassium (mEq/L)	Urea (mg/dl)	Creatinine (mg/dl)	Calcium (mg/dl)
Serum					
1	132	4.0	33.0	—	—
2	140	3.4	26.0	1.0	8.8
3	130	3.2	30.0	1.0	8.1
4	150	5.6	74.5	2.0	8.4
5	138	5.7	52.3	1.0	—
6	130	4.8	28.0	1.0	—
Urine					
5	71	8.0	22.4	8.0	—

Urine—Albumin: Nil, Sugar: Nil, Microscopy: No cells, no casts
Renal failure index 9.0 Fractional sodium excretion 16.0%
Plasma hemoglobin 25 mg/dl Urine hemoglobin 41 mg/dl

Discussion

NDI is a rare disorder characterized by polyuria, hyposthenuria and serum hyperosmolality, with no response to exogenously administered arginine vasopressin (AVP). It can be a congenital condition or can be acquired as a result of damage to the concentrating mechanisms of the kidney. Various causes of acquired NDI have been mentioned(2).

NDI has been described in the diuretic phase of non-oliguric renal failure. This patient showed a rise of blood urea and serum creatinine 24 hours after the onset of intravascular hemolysis. This would suggest that intravascular hemolysis rather than asphyxia was the cause of renal impairment. The azotemia was transient and was seen only on the 5th and 6th days of life in this baby (*Table II*).

Hemolysis and hyperbilirubinemia resulting from the use of G6PD deficient donor blood for exchange transfusion has been described(3). Intravascular hemolysis also accompanies severe rhesus hemolytic disease and causes raised plasma hemoglobin(4). We suspect either of these could have caused the intravascular hemolysis, nonoliguric renal failure and NDI.

Cranial diabetes insipidus following germinal matrix intracranial hemorrhage has been reported(5) and was possible in this patient too. However, the arginine vasopressin test rule it out.

We conclude that transient NDI can occur in newborns following damage to the distal tubules of the kidney following intravascular hemolysis. In our patient, NDI proved to be transient and responded well to management of fluids and electrolytes. Routine screening for G6PD of blood used for exchange transfusions in neonates can help in prevention of such complications.

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Peripheral Precocious Puberty with Hypertension

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Peripheral precocious puberty in boys is most commonly due to adrenogenital syndrome of which congenital adrenal hyperplasia (CAH) accounts for about 50% of cases(1). Deficiency of 11-beta hydroxy-lase (11-OH) accounts for 5-8% of all cases of CAH(2). So far about 100 cases have been reported(2). Hypertension is a common but not an invariable finding, usually absent in the first few years of life. The mean age of onset of precocity is 3.5 years in boys with a range of 1-8 years(3). We report here a case of peripheral precocity with hypertension due to 11-OH deficiency presenting in a 3 ye old boy.

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Case Report

A 3-year-old boy was admitted in Niloufer Hospital in August 1990 for respiratory infection of one week duration. During his hospital stay parents revealed that the child had excessive and progressive growth of the body since the age of one year. Six months later they also noticed the enlargement of external genitalia characterized by increase in the size and color of the scrotum associated with increasing size of penis, along with growth of pubic hair. From the age of 2½ years, there was increasing pigmentation of body including oral mucosa, change in voice resembling an adult voice and papular eruptions on the face. History of early morning erections was present but there were no nocturnal emissions. There were no symptoms of central nervous system involvement, visual disturbances or symptoms related to hypothalamic disturbances or hypothyroidism.

The child was seventh in birth order born to non-consanguinous parents. Three elder brothers had died between the ages of 2-4 years suddenly due to unknown reasons within hours of illness and before hospitalization. Three sisters are healthy and elder sister had attained menarche at normal age.