

rendered by Dr. R. Muralidharan of the Department of Endocrinology, PGIMER, Chandigarh in managing this patient.

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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

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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 year old boy.

## 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-consanguineous 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.

On examination he was a tall and healthy child with acneiform eruptions on the face and generalised hyperpigmentation of the body including oral mucosa and tongue. No neurocutaneous markers or skeletal deformities were observed except for mild genu varum. Blood pressure was persistently raised above 150/90 mm of Hg. Anthropometry revealed a weight of 17 kg, height 107 cm and head circumference 52 cm, which were more than 90th percentile for age. Pubic hair was sparse, curly and pigmented. Penis was 8.5 cm in length when stretched and 3.5 cm in circumference. Scrotum was about 5 cm in length, darkly pigmented with marked rugosity, corresponding to Tanner Stage III(5), Testicular size was 1.5 to 2 cm in length and 2-3 ml in volume corresponding to Tanner Stage I. Axillary hair growth was absent. Systemic examination was normal.

On investigation urine specific gravity, serum electrolytes, blood urea, serum creatinine, serum calcium and serum alkaline phosphates were in normal range. X-rays of chest and skull were normal. X-ray hands revealed a bone age of 7-8 years. CT scan of cranium was normal. CT abdomen showed that both the adrenals were uniformly large for age but no mass could be

detected. Karyotyping was normal male (XY). Hormonal changes are given in the Table.

### Discussion

Excessive and in orderly growth of penis with pubic hair, increased height and weight, advanced bone age and abnormally high levels of adrenal steroids in a 3 year old boy is described. There was a marked increase in the 24 hour urinary 17-ketosteroids, serum ACTH, serum testosterone, serum 17-hydroxyprogesterone and serum 11-deoxycortisol and the dexamethasone suppression test was positive. The child had hypertension due to the high serum levels of 11-deoxycortisol. The serum electrolytes were normal on more than one occasion. The CT scan abdomen suggested increased size of adrenals. All these findings confirm the diagnosis of CAH due to 11-OH deficiency. Only 5-8% cases of CAH have the rare 11-OH deficiency(2) Khandekar *et al.*(4) from Chandigarh reported 31 cases of precocious puberty in 1990, but no case was reported with hypertension or 11-OH deficiency. Prasanna Kumar *et al.*(3) from Delhi reported 15 children with precocious puberty in 8 boys

TABLE—Hormonal Evaluation

Test	Level	Normal Range
1. 24 h urinary 17-ketosteroids	8.66 mg/day	0.8-1.8 mg/day
2. 24 h urinary pregnanetriol	1.1 mg/day	<0.5 mg/day
3. Serum ACTH (8.00 a.m.)	400 pg/ml	<90 pg/ml
4. Serum testosterone	3.6 ng/ml	0-0.45 ng/ml
5. Serum 17 hydroxy-progesterone	3.8 ng/ml	0.1-0.8 ng/ml
6. Serum 11 deoxycortisol	2.0 mcg/dl	<1.0 mcg/dl

and 7 girls and CAH in 50% of the boys but none with hypertension.

The child is under treatment with hydralazine and prednisolone. On nine months follow up, the hypertension is under control, the skin pigmentation has disappeared and the linear and genital growth is static.

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## Choledochal Cyst in a Neonate

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Choledochal cyst is a relatively rare condition. A review of the literature reveals

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six patients of choledochal cyst who presented in the neonatal period. Four of these six patients were diagnosed antenatally(1,3). We present a child with choledochal cyst in the neonatal period.

## Case Report

A two-day-old boy weighing 3.9 kg presented at Park Children's Centre for Treatment and Research, Calcutta with history of distension of abdomen since birth and nonbilious vomiting for last 24 hours. He was passing high colored urine and had passed meconium on the second day of life. On examination he was mildly icteric. Examination of the abdomen revealed a large cystic lump extending from the right hypochondrium to the right iliac fossa and crossing the midline at the level of the umbilicus. It had slight side to side mobility and was dull on percussion. Plain X-ray of the abdomen showed a soft tissue shadow occupying the whole of the right half of the abdomen with intestinal gas shadows pushed to the left (*Fig. 1*). Ultrasonography showed a cystic lesion occupying the whole of the right half of the abdomen (*Fig. 2*). Intrahepatic biliary channels were dilated. A diagnosis of choledochal cyst was suggested. His hematological and biochemical parameters were within normal limits for his age except a raised total bilirubin level of 3.7 mg/dl (unconjugated 1.4, conjugated 2.3).

Peroperative preparation included administration of injection vitamin K, intravenous glucose infusion and decompression of the bowel by nasogastric suction and bowel washes. Laparotomy was performed on the 5th day of life. Abdomen was opened through right transverse supraumbilical incision. A unilocular cystic lump measuring 15 cm × 10 cm was seen