

# ADRENOGENITAL SYNDROME

## (A Rare Case Report)

by

K. SAXENA

R. RIZVI

D. MADAN

and

V. S. LOCHAB

### Introduction

Adrenogenital syndrome is the commonest cause of virilism in the female child but is very rare. The virilism is because of a deficit in 21-c-hydroxylase enzyme. The common causes of adrenogenital syndrome are adrenal hyperplasia, virilizing tumours of ovary in a child, exogenous androgens, post-natal adrenogenital syndrome, virilizing adrenal tumours and Cushing's syndrome. A case is being reported for clinical interest.

### Case Report

Miss P a young girl, aged 5 years was admitted on 20-1-82, for the change in the external genitalia which she noticed few days back. She had only one sister who was normal. Family history was negative.

She had normal feminine appearance, weighed 27 kgms. No lump was palpable in abdomen. Local examination revealed few pubic hair. Labia majora were normal but labia minora were not well defined. The clitoris was hypertrophied, 3 cms x 1.5 cms. The urethral

opening was underneath the clitoris as shown in Fig. 1. The vagina was 6 cms. deep and was made out by passing a catheter in vagina and another in urethra.

Rectal examination was done under anaesthesia. Small uterus could be felt and there were no adnexal masses.

### Investigations

Buccal smear was positive for Barr bodies and the peripheral smear was positive for Drumstick pattern. X-ray skull was normal. X-ray chest was also normal. The urinary 17-ketosteroid excretion was increased and it was 8.8 mgm per 24 hours, while normal excretion in young children is 0.1 to 0.3 mgms/24 hours. Intravenous pyelography did not show any abnormality. A vaginogram was done which showed the presence of normal vagina. The uteurs and tubes were also visualised normally as shown in Fig. 2.

### Management

The patient was given corticosteroids orally. 40 mgms were given initially and the dose was reduced gradually. It was then maintained on 5 mgms. orally daily. The clitoris was excised on 26-4-82 under general anaesthesia. The 17-ketosteroids excretion levels came down after giving cortisone therapy to the patient. It was 1 mgm/24 hours at the time of discharge on 22-5-82.

From: J. N. Medical College, A.M.U., Aligarh-202001.

Accepted for publication on 24-2-83.

See Figs. on Art Paper VI