

Feminizing Genitoplasty in a Teenage Girl

C. James, S. George, H. Fernandes, N. Zachariah, M. S. Seshadri, K. George

Dept. of Reproductive Medicine, Pediatric Surgery and Endocrinology, Christian Medical College and Hospital, Vellore 632004

An eighteen year old girl presented to us with complaints of delayed menarche and marked hirsutism. Her parents had noticed ambiguous genitalia during infancy for which strangely no treatment was sought. Hypotensive episodes were not encountered in childhood. She was short statured with a height of 146cm and height standard deviation score of minus 2.5 and an android muscular build with marked hirsutism and a BP of 130/80 mm Hg. Breast development was Tanner stage III. External genitalia revealed an enlarged clitoris with labio scrotal fusion extending upto the urethral orifice. Gonads were not palpable. Ultrasound examination of the pelvis confirmed normal internal female organs. Laboratory studies revealed an elevated serum 17 hydroxy progesterone > 200 ng/ml, testosterone 3 ng%, DHEAS, a diagnosis of congenital adrenal hyperplasia was made, probably 21 α hydroxylase enzyme deficiency.

She was started on glucocorticoid replacement therapy and Tab. Aldactone for her hirsutism. Six months later she underwent a feminising genitoplasty which consisted of clitoral reduction and cut back vaginoplasty under stress level steroidal supplementation. Principle utilized for reduction clitoroplasty was subcutaneous amputation of the shaft of the clitoris but sparing the glans with its neurovascular bundle, followed by suturing the glans to the stump of the corpora in order to preserve clitoral sensation. Since a low vagina was visualized, a simple cut back incision from the perineum into the vagina was sufficient to create an adequate vaginal introitus. Three months later she commenced spontaneous onset of menstrual flow and her hirsutism had decreased significantly. For psychological and practical reasons surgical reconstruction should be performed at an early age. Reconstruction should achieve two goals. 1. Reduction of clitoromegaly 2. Creation of an adequate vaginal introitus



Fig. 1: Presurgery



Fig. 2: Postsurgery

We report this case as it is rare for a girl with ambiguous genitalia due to congenital adrenal hyperplasia to present at teenage without prior treatment.