CASE REPORT

Female Epispadias

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Abstract

Female epispadias is a rare congenital anomaly occuring in 1 in 484,0000 female population. Clinical course, management is discussed in the present case report to stress upon careful diagnosis and to decrease chances of mismanagement in highly morbid condition "urinary incontinence".

Key Words

Epispadias, Congenital anomaly.

Introduction

Female Epispadias is a rare congenital anomaly occurring in one in 484,000 female population (1). The patient presents with characteristic appearance of external genitalia with or without urinary incontinence. Diagnosis might be missed unless the labia are separated and examined. Vagina and internal genitalia are usually normal. The anomaly can be treated by surgical reconstruction of bladder neck, urethra and external genitalia.

Case Report

A 10-year-old girl presented to us with total urinary incontinence. History did not reveal any evidence of urinary tract infection, trauma or operative interventions. The child was a product of nonconsanguinous marriage, born through a normal vaginal delivery and had normal developmental milestones. No other female family members had similar problem.

Neurological examination was unremarkable. Inspection of external genitalia (Fig. 1) revealed a bifid clitoris and depressed mons covered by smooth and glabrous skin. The labia minora were ill developed and terminated anteriorly to corresponding half of bifid clitoris. Meatus was wide and patulous. Roof of the distal urethra was splayed with dribbling urine continuously.

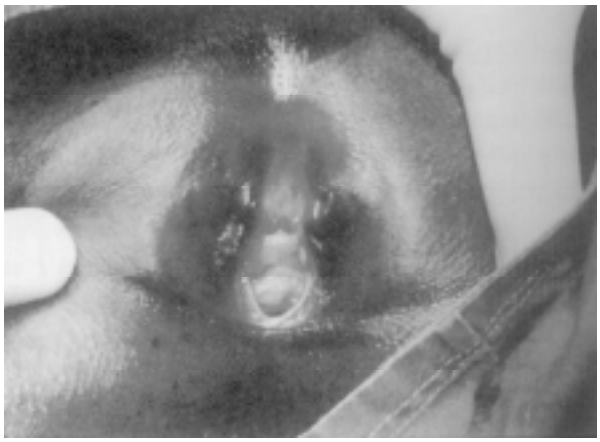

Fig. 1 Showing depressed mons, absent prepuce, bifid clitoris, patulous urethral meatus with deficient dorsal wall and normal vaginal opening.

Urine culture was sterile. Serum creatinine was normal. Ultrasound and intravenous urography

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were normal. Voiding cystourethrogram (Fig. 2) revealed a normal capacity bladder with wide bladder neck and short wide urethra. No Vesico ureteric reflux was seen. Urodynamic study revealed a bladder capacity of 150 ml with abdominal leak point pressure of 7 cm of H_2O. Cystoscopy demonstrated a short wide urethra of 1cm length with splaying of the roof of the urethra and widely opened bladder neck. The ureteric orifices were situated 2 cm from the bladder neck and were of normal caliber. Rest of the bladder was unremarkable.

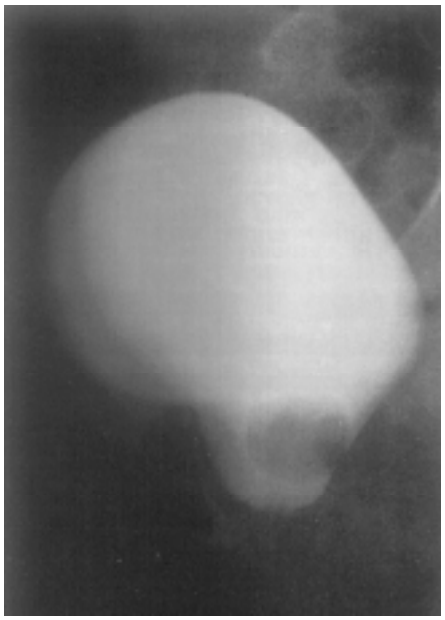

Fig. 2. Cystogram showing normal capacity bladder with wide bladder neck and dilated urethra. Foley's catheter balloon is in the dilated urethra.

A combined abdominal and perineal surgical reconstruction was performed. The thinned out tissue of the roof of urethra was excised and urethra was tubularised to 10Fr by approximation of the lateral halves of urethra and periurethral tissues. The genitalia were reconstructed by denuding the two halves of clitoris on the medial surface and approximation (Fig. 3). Bladder neck was reconstructed trans-vesically by Young - Dees - Lead better technique with Cohen's transtrigonal reimplantation of ureters. Supra pubic and urethral catheter were placed. Urethral catheter was removed on 8th postoperative days. Voiding trial was given at 3 weeks. Post operatively voiding cystourethrogram (Fig. 4) revealed normal bladder neck and lengthened urethra and no reflux. Residual urine was nil and the girl achieved continence in the day for more than 4 hours and no bed wetting in the night at 5weeks post operatively. The postoperative cosmetic appearance was acceptable.

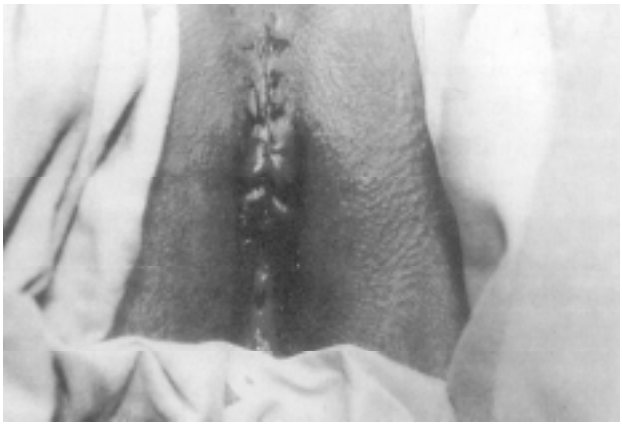

Fig. 3 Immediate post reconstruction appearance of external genitalia .

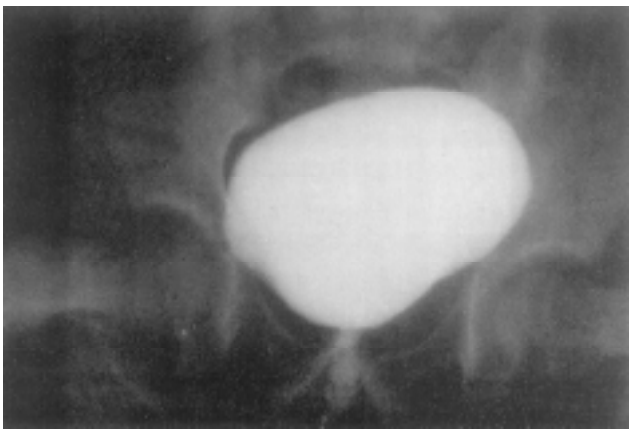

Fig. 4 Micturating cystourethrogram at 3 weeks post surgery, showing normal caliber urethra and no vesico ureteric reflux.

Discussion

Female epispadias, just like its male counterpart, presents with continuous dribbling of urine since birth or may come to notice because of abnormal genitalia. There may be history of recurrent urinary tract infection. External genitalia have varied appearance as described by Davis ranging from lesser degrees with patulous orifice to intermediate cases with urethra dorsally split along most of its length to the most severe cases which involve the entire length of urethra rendering the sphincteric mechanism incompetent (2). Most present characteristically with bifid clitoris, depressed mons, ill developed labia, patulous wide meatus and occasional symphyseal separation. Diagnosis may be missed if genitals are not examined carefully by separating the labia majora. Vagina and internal genitalia are usually normal. The bladder is often small with poorly developed bladder neck and incompetent sphincteric mechanism. The ureters open laterally in more or less straight course, which are refluxing in 33% to 75% of cases (3). Complete radiological evaluation is required in all cases to identify reflux and rule out other causes of incontinence. Urodynamic study helps in confirming the poor out let resistance. Cystoscopy is required to assess bladder capacity and the position of the ureteric orifices. The condition requires surgical reconstruction of bladder, urethra and external genitalia. More than 50 years ago Young recognized the need to revise radically the urethra and bladder neck to achieve continence in these cases(4). Various modifications have since occurred. Hendren's combined abdominal-perineal single stage reconstruction, is one of the currently followed procedures associated with best continence results (5). Cohen's cross trigonal ureteric reimplantation is simultaneously performed to prevent vesicoureteric reflux (6). The same procedure was adopted in our case also, with satisfactory result. Alternatively, staged procedures with primary genital and urethral reconstruction at 1 to 1½ years and delayed bladder neck reconstruction at 4-5years have also been described,. The continence rates and duration to achieve continence are similar in both single and staged procedures, but staged procedure requires multiple sessions of surgery, anesthesia and is associated with higher morbidity.

The case is presented here because of its rarity and to stress upon careful examination of an incontinent child, reduce chances of misdiagnosis and decreases mismanagement of a highly morbid condition "Urinary Incontinence".

References

1. Gearhart JP. Exstrophy, Epispadias and other bladder anomalies. In Walsh PC, *et al* (Eds.). Campbell's Urology, 7th ed. Philadelphia, WB Saunders, 1998. pp. 2136.
2. Davis DM. Epispadias in females and surgical treatment. *Surg Gynaecol Obstet* 1928; 47: 600
3. Kramer SA, Kelalis PP. Surgical correction of female epispadias. *Eur Urol* 1982b; 8:321.
4. Young HH. A new operation for cure of incontinence associated with epispadias. *J Urol* 1922; 7: 1-32.
5. Hendren WH. Congenital female epispadias with incontinence. *J Urol* 1981; 125: 58.
6. Cohen SH. Ureteroneocystostomy: A new technique for reflux prevention. *Aktuel Urol* 1975; 6: 1.