Case Report

Female Epispadias

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Abstract

Female epispadias without exstrophy is a rare malformation occurring in 1 in 117,000 male and 480,000 female population. It is imperative that a thorough local examination be performed ideally at birth so the diagnosis and the later psychosocial and psychosexual problems could be prevented. Although, on occasions when the apparent malformation is not that predominant, epispadias can be missed at birth and is diagnosed when the child (usually female) remains wet after toilet training. Reconstructive surgery of the urethra and bladder neck improves the outcome in case of early intervention.

Introduction

Female Epispadias is a rare congenital anomaly occurring in one in 480,000 female population.[1] The usual complaint is abnormality of the external genitalia which may or may not be associated with urinary incontinence or sexual dysfunction. The diagnosis can only be made after a detailed local examination by separating the labia. In most instances the vagina and internal genitalia are found to be normal. The anomaly can be corrected by surgical reconstruction of bladder neck, urethra and external genitalia.

A case is presented here because of its rarity and to emphasize upon careful examination of an incontinent child, to make and early diagnosis as it is amenable to surgery and to reduce the psychosocial and psychosexual problems and prevent the patient from a prolonged misery of urinary incontinence.

Case Report

A 4-year-old girl presented to us with urinary incontinence with a mild bladder control and voiding of small quantity of urine twice or thrice in the toilet. There was no past significant history of any urinary tract infection, trauma or operative interventions but the parents complained of a very foul smell of the urine. The child was a product of nonconsanguineous marriage, born through a normal vaginal delivery and had normal developmental milestones. No other female family members had a similar problem. Neurological examination was unremarkable. Inspection of external genitalia (Fig. 1) revealed a bifid clitoris and depressed mons. The labia minora were ill developed and terminated anteriorly to corresponding half of bifid clitoris. Roof of the distal urethra was splayed with dribbling of urine intermittently on applying pressure on the bladder per abdomen.

Urine DR showed numerous leucocytes with nitrites. Urine culture was sterile. Serum creatinine was normal. Ultrasound and intravenous urography were normal. Voiding cystourethrogram (Fig 2) revealed a normal capacity bladder with a prominent urethral and vaginal openings and short wide urethra. Mild dribbling of urine through the vagina was seen. There was no evidence of vesico ureteric reflux. Pubic diastasis was found. The above physical findings were compatible with epispadias without exstrophy.

The child was referred to Sind Institute of Urology and Transplantation (SIUT) for surgical reconstruction. The urethra was reconstructed. The clitoris was atrophic so it wasn't reunited. Bladder control was planned to be tested three weeks later.

Discussion

Female Epispadias is a rare congenital malformation of the urogenital system consisting of the defect of the dorsal wall of the urethra.The rarity of the case can be judged by a 10 years data collection (1991-2000) in which out of 13 patients diagnosed as epispadias only 4 were females.[2] Epispadias occurs more commonly in males than in females, with a prevalence of 1 case in 10,000-50,000 persons.[3] The male-to-female ratio is 2.3:1.Usually it occurs sporadically but in some cases there is a strong genetic component.[4] A survey was conducted to investigate the inheritance pattern of exostrophy epispadias complex by paediatric urologists and it was found that bladder exostrophy recurred in 9 out of 2500 families (1 in 275), and 215 offspring were identified with exostrophy or epispadias. In these the inheritance was in 3 offsprings i.e. (1 in 70).[5]

Epispadias usually presents with continuous dribbling of urine since birth or may come to notice because of abnormal appearance of the genitalia. The severity of the defect can vary from a mild glandular defect to complete exostrophy of the bladder. There may be a mild variant showing a 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. Most females present with a characteristic bifid clitoris, depressed mons, ill developed labia, patulous wide meatus and occasional symphyseal separation. Even the most minor cases have a notch in the symphysis pubis and oblong

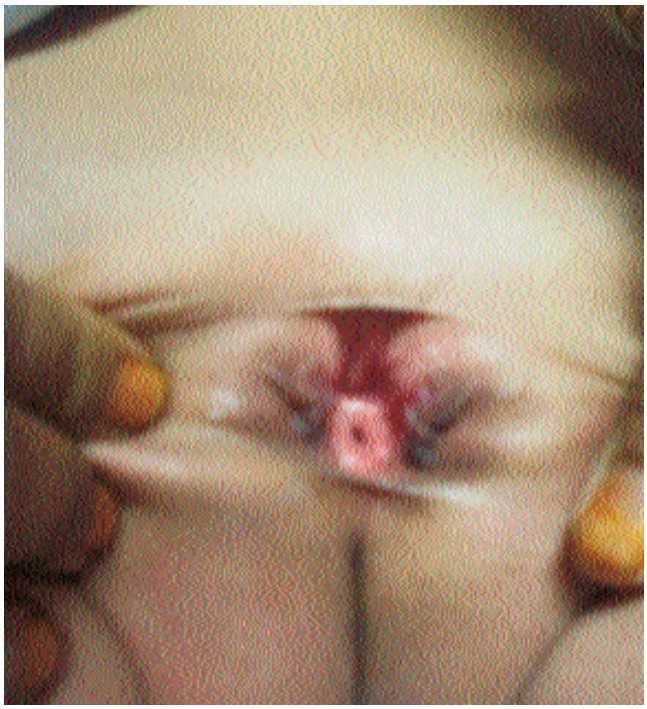

Fig 1: Showing depressed mons, bifid clitoris, with deficient dorsal wall and normal vaginal opening.

external urethral meatus.

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. Patient may complain of recurrence of UTI.

Serious psychological consequences have occurred due to the misdiagnosis of this disease. Females have had social

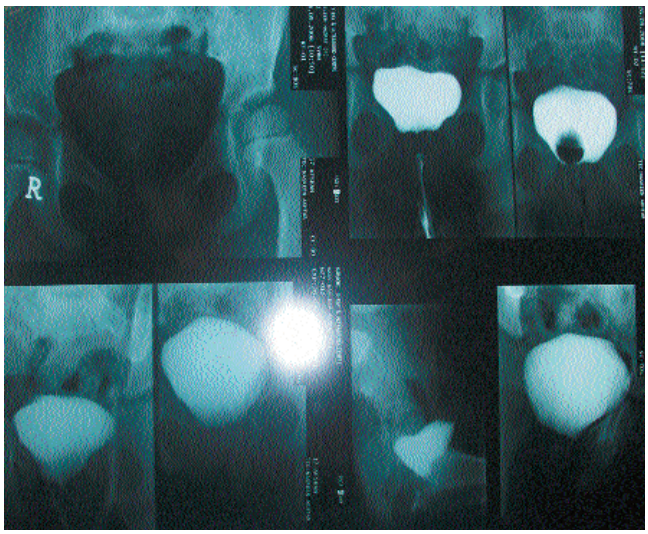

Fig 2: Micturating cystourethrogram showing pubic diastasis normal caliber urethra and no vesico ureteric reflux.

disruption consequences and sexual dysfunction Ineffective treatment and wrong diagnosis has lead to anxiety and depression in young women.[6]

Complete radiological evaluation is required in all cases to test reflux and rule out other causes of incontinence. Urodynamic studies reveal 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. A combined abdominal and perineal surgical reconstruction is performed. The genitalia are reconstructed by denuding the two halves of clitoris on the medial surface and approximation. Bladder neck is reconstructed transvesically. The pubic symphysis is approximated.

The continence rates and duration to achieve continence are similar in both single and staged procedures, but staged procedure requires multiple sessions of surgery, anaesthesia and is associated with higher morbidity.

Years ago Young recognized the need to revise radically the urethra and bladder neck to achieve continence in these cases.[7] A non endoscopic device using a periurethral injection has been used for control of urinary incontinence.

Zuidex is a device used to increase the bladder outlet resistence that has been tried above 5 years of age. It helps to increase the continent period.[8]

To correct the genital aspect of the anomaly the basic principles of Ransley's technique have been used.[9] In addition, the island flap technique are applied to the severe form.[10]

The results of different techniques have been variable. Most children are treated especially if the diagnosis has been made early. In the other conditions there persists some degree of incontinence in the females.[11]

Conclusion

The case is presented here to emphasize upon the fact of detailed genital examination of patients presenting with incontinence and avoiding misdiagnosis, providing them with early treatment to get better psychological and surgical outcome.

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