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A rare case of isolated female epispadias

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Abstract

Isolated female epispadias without bladder exstrophy is an extremely rare entity. It usually presents with primary urinary incontinence and abnormal external genitalia. We describe this rare congenital anomaly in a 7-year-old girl who presented with urinary incontinence and abnormal external genitalia. She had a bifid clitoris and widely separated labia minora. The vagina was normal. Voiding Cysto-urethrography (VCUG) showed small capacity bladder with no reflux. Surgical reconstruction of the urethra, labia minora and clitoris was performed.

Keywords

Bifid clitori; epispadias; female; incontinence; pubic diastasis.

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Introduction

Isolated epispadias without exstrophy is a rare congenital malformation occurring in 1 in 117,000 male and 484,000 female population [1,2]. It usually presents as urinary incontinence since birth and abnormal external genitalia. The diagnosis might be missed unless a careful examination performed [3]. A thorough local examination should be performed ideally at birth so as to make an early diagnosis and prevent the later psychosocial and psychosexual problems. Surgical reconstruction of urethra, bladder neck and external genitalia is usually curative [3].

In this case report, we present a 7-year-old girl with isolated female epispadias who underwent simple one stage repair to correct this anomaly.

Case Reports

A 7 year old female child presented with complaints of continuous dribbling of urine since birth. There was no history of fever,

pyuria and hematuria. The urethral opening was patulous, deep seated and extending to bladder neck. The clitoris was bifid and the labia minora widely separated. She had a normal vaginal opening [Fig. 1].

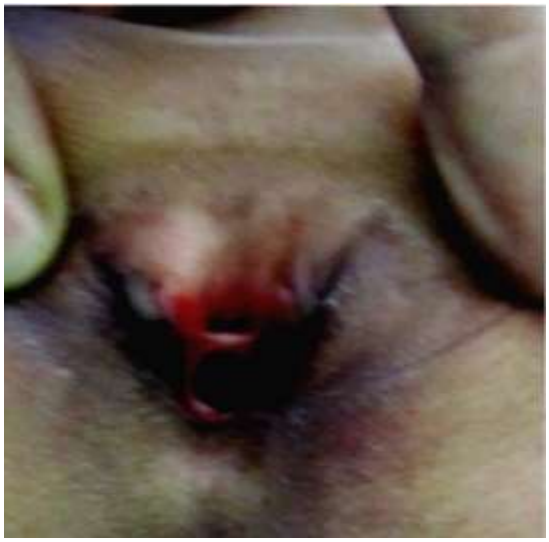

Fig. 1. Local examination showing bifid clitoris, patulous and deep seated urethral opening reaching upto bladder neck, widely separated labia and normal vaginal opening.

On evaluation, hemogram, urine routine microscopy and culture, renal profile and Ultrasound abdomen were normal. Voiding Cysto-urethrography (VCUG) showed a small bladder capacity with no evidence of reflux. Pubic diastasis was present.

On Cystoscopy, the bladder was of small capacity and there were no trabeculations, bilateral ureteric orifices could not be visualized. Epispadias repair was done. A diamond shaped incision was taken around the urethral orifice and extended into mons pubis. The urethra was mobilized and urethroplasty was done [Fig. 2]. The hemi-clitoris were approximated in the midline [Fig. 3].

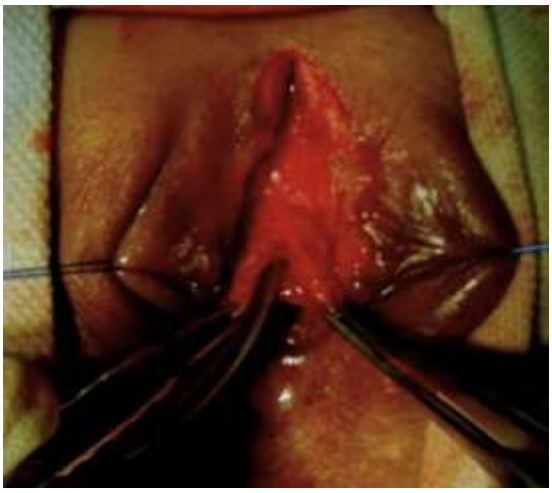

Fig. 2. Intra-operative image showing the mobilized urethra.

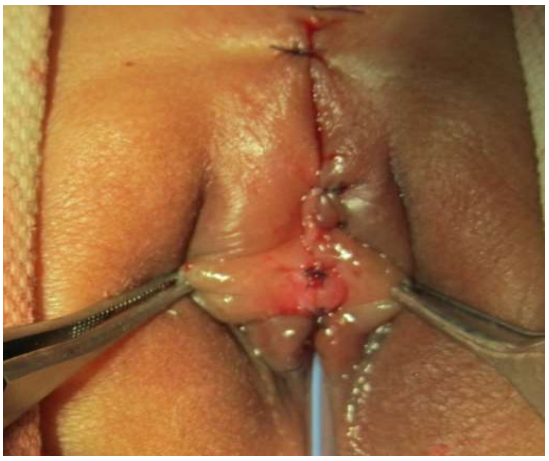

Fig. 3. Completed Epispadias repair.

Post-operatively, the external appearance of the genitalia was satisfactory [Fig. 4]. The patient has a dry period of approximately 2

hours. Her current bladder capacity is approximately 50 cc. She is regularly doing perineal exercises.

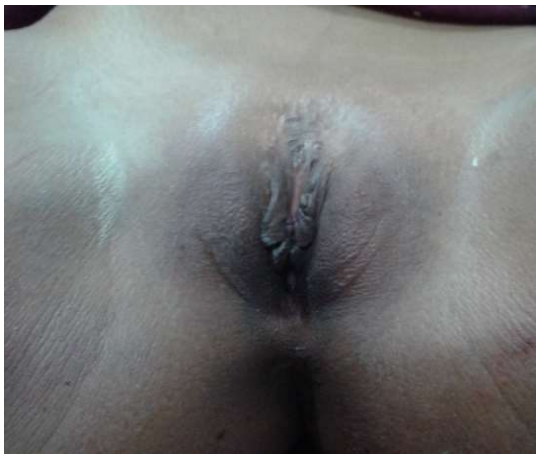

Fig. 4. Local examination at follow up.

Discussion

Female epispadias is a rare congenital anomaly and presents clinically as continuous dribbling of urine since birth and/or recurrent urinary tract infection. Generally, external genitalia are abnormal and characterized by bifid clitoris; the mons is depressed in shape and coated by a smooth glabrous area of skin. The labia minora are usually poorly developed and terminate anteriorly at the corresponding half of the bifid clitoris, where there may be a rudiment of a preputial fold. On minimal separation of the labia, the urethra is seen, which may vary considerably, as mentioned previously. In most cases, the bladder neck is located at the level of the hymenal ring. The symphysis pubis is usually closed but may be represented by a narrow fibrous band. In contrast, the vagina and internal genitalia are usually normal [2,4].

The diagnosis may be missed in infancy and patient may sometimes present in childhood with primary urinary incontinence like our case. The incontinence varies from continuous dribbling of urine without bladder filling to single episodes of daytime stress-incontinence depending upon the degree of bladder neck involvement. The bladder capacity is often small with poorly developed bladder neck and incompetent sphincteric mechanism [3].

Epispadias is classified as per the Davis Classification which describes three degrees of epispadias [2, 5]. In the lesser degree of epispadias, the urethral orifice simply appears patulous; in the intermediate epispadias, the urethra is dorsally split along most of the urethra; in the most severe degree of epispadias, the urethral cleft involves the entire length of the urethra and sphincteric mechanism and the patient is rendered incontinent [5]. Milder forms of epispadias are extremely rare and usually go unnoticed due to absence of incontinence in these children [6]. Internal genitalia are usually normal. The incidence of reflux is reported to be 30% to 75% because of the ureters open laterally in a more-or-less

straight course [7,8]. There is no outlet resistance, the bladder is small and the wall is thin. However, after urethral reconstruction, the mild urethral resistance created allows the bladder to develop an acceptable capacity for potential later bladder neck reconstruction [2]. Urodynamics study helps in confirming the poor outlet resistance. Cystoscopy is required to assess bladder capacity and the position of the ureteric orifices.

Surgical reconstruction requires the repair of the bladder neck, urethra and external genitalia. The objectives for management of female epispadias include achievement of urinary continence, preservation of the upper urinary tracts, and reconstruction of functional and cosmetically acceptable external genitalia [2,4]. More than 50 years ago, Young recognized the need to revise radically the urethra and bladder neck to achieve continence in these cases [8]. Hendren's combined abdominal-perineal single stage reconstruction was one of the best continence results [10]. Cohen's cross trigonal ureteric re-implantation is simultaneously performed to prevent vesicoureteric reflux [11].

The three main female concerns are the appearance of the external genitalia, adequacy of the vaginal opening and uterine prolapse. Although usually corrected at birth, sometimes “touch-up” surgery needs to be performed at puberty. The three cosmetic concerns are the appearance of the lower abdominal wall, the mons pubis and the clitoris [2]. Reconstructive surgery is applied by a simple technique in most of the cases. The single stage repair has good and acceptable appearance of external genitalia with 60–87% urinary continence rate [10,12,13], as applied in the present case. Patients with female epispadias usually have good long-term results regarding quality of life and sexuality, despite having some degree of urinary incontinence [14].

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