Case Report

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Website: www.afrjpaedsurg.org
DOI: 10.4103/0189-6725.86066
PMID: 22005369
Quick Response Code:

Female epispadias

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ABSTRACT

Isolated female epispadias without bladder exstrophy is an extremely rare congenital anomaly. The symptoms of female epispadias are primary urinary incontinence and abnormal anatomical features. A 7-year-old girl presented with partial incontinence of urine. On physical examination, bifid clitoris and labia minora were seen. The vagina and hymen were normal. Voiding cystourethrogram showed no reflux. With the diagnosis of isolated female epispadias, single stage reconstruction of the urethra, labia minora and clitoris was performed.

Key words: Bifid clitoris, epispadias, urinary incontinence

INTRODUCTION

Female epispadias is a very rare congenital anomaly representing a mild form of the spectrum of exstrophy of the bladder. The patient presents with characteristic appearance of external genitalia with or without urinary incontinence. Diagnosis might be missed unless carefully examined. Radiographic finding of diastasis of the pubic bones is often associated with it. The anomaly can be treated by surgical reconstruction of urethra, bladder neck and external genitalia.

CASE REPORT

A 7-year-old female child presented with partial incontinence of urine and abnormal appearing genitalia since birth. She was voiding good quantity of urine. There were no other urological complaints. Examination of external genitalia revealed widely separated two halves of the clitoris with labia minora. There was complete dorsal split in the urethra between 9 o'clock and 3 o'clock positions not involving the bladder neck [Figure 1]. The smooth mucosa of the urethra was blending cranially with the thin hairless skin over the mons. Vagina and hymen were normal. On evaluation, haemogram, urine routine and culture, renal function tests were within normal limits. Ultrasonography showed normal upper tracts with a bladder capacity of 150 ml. Radiograph of the pelvis showed pubic diastases. Voiding cystourethrogram revealed good bladder capacity on filling bladder with no vesico ureteric reflux on both sides.

Cystourethroscopic assessment revealed normal appearing ureteric orifices and intact bladder neck. A vertical incision was given over thin skin of mons, which was split inferiorly at the base of the mons to excise full thickness of the thinned out tissue of roof of the urethra to a level near the bladder neck [Figure 2]. Urethra was mobilized on either side till the proximal split part without disturbing the urethral floor, which was kept intact. Urethral plate was not undermined or disrupted in order to avoid vascular compromise. Urethra was reconstructed over a 10 Fr Foley catheter with inverting sutures starting near the bladder neck. The medial aspect of the hemiclitoris and labia minora were denuded and clitoral reconstruction was done [Figure 3]. Mons pubis area was reconstructed by undermining fibro fatty tissue and closing it in two layers after lateral mobilization to obliterate the dead space [Figure 4]. Urethral catheter was removed after 7 days. There was satisfactory cosmetic result after the surgery [Figure 5]. She was continent during day and incontinence occurred sometimes during night. This was managed conservatively by pelvic floor exercises and oxybutynin with significant improvement after six weeks. Review ultrasonography after three months showed a bladder capacity of 300 ml and there was no incontinence of urine.

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Figure 1: Bifid clitoris, patulous urethral meatus with deficient dorsal wall, depressed mons.

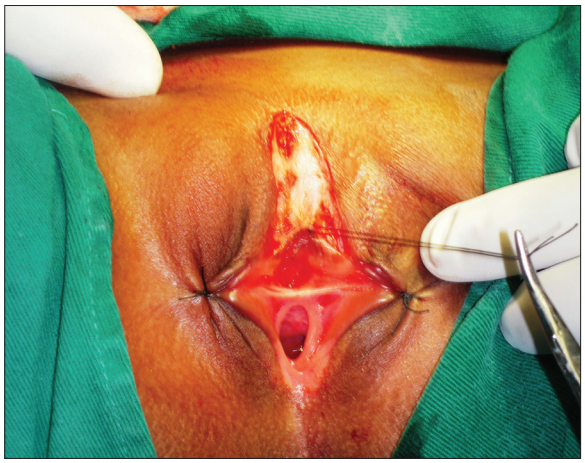


Figure 2: Excision of the glabrous skin of the mons.

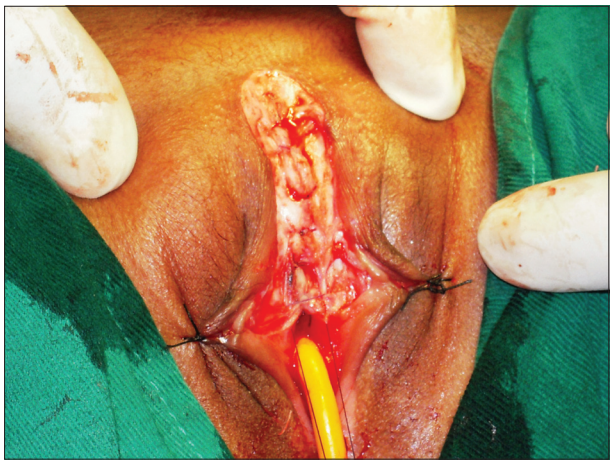


Figure 3: Neourethra reconstructed, medial aspect of labia minora and hemiclitoris denuded.



Figure 4: Final appearance after complete reconstruction.



Figure 5: Appearance of external genitalia after four weeks of surgery.

DISCUSSION

The paper is a "case report" on an uncommon congenital abnormality of the lower urogenital tract, i.e., female epispadias. Female epispadias is often associated with urinary incontinence as in the presented case. The incontinence is varying from continuous dribbling of urine without bladder filling to single episodes of daytime stress-incontinence. Often the bladder capacity is reduced as consequence of the lack of filling ("Never functioning or poorly functioning bladder"). This rare congenital anomaly occurs with an incidence of one in 484,000 female patients.[1] External genitalia have varied appearance as classified by Davis ranging from lesser degrees with patulous urethral 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 and bladder neck rendering the sphincteric mechanism incompetent.[2] Milder forms of epispadias are extremely rare.[3] This may be because they go unnoticed due to absence of incontinence in these children. Genital defects include bifid clitoris and poorly developed labia minora, which end anteriorly at the corresponding half of bifid clitoris.[4] The mons is depressed and covered by a smooth, glabrous area of

skin while symphysis pubis is usually closed but may be represented by a narrow fibrous band. Diagnosis may be missed if genitals are not examined carefully by separating the labia majora. The vagina and internal genitalia are usually normal. These external appearances are most characteristic. The bladder is often small with poorly developed bladder neck and incompetent sphincteric mechanism.

About 30% to 75% of the epispadias are associated with reflux as the ureterovesical junction is inherently deficient in these cases and ureteric openings are often laterally placed in the bladder with a straight course so that reflux occurs.[5] Complete radiological evaluation is required in all cases to identify reflux and rule out other causes of incontinence. Cystourethroscopy is done to assess bladder capacity and the position of the ureteric orifices.

The objectives of surgical repair include achievement of urinary continence with preservation of the upper urinary tracts and the reconstruction of functional and cosmetically acceptable genitalia.[1] In severe degree cases, various surgical procedures have been reported to control continence including transvaginal placation of the urethra and bladder neck, muscle transplantations, urethra twisting, bladder flap and Marshall Marchetti vesicourethral suspension.[1,3] These procedures increase urethral resistance. Lengthening of urethral tube during reconstruction by itself improves incontinence.

This case is reported because it was a rare case of intermediate degree epispadias having dorsal splitting of urethra with partial incontinence of urine. Although her presenting complaint was partial urinary incontinence, on cystourethroscopy the bladder neck was intact. Cause of partial incontinence in this case could be because of reduced urethral resistance. Our technique of urethroplasty increased the urethral resistance, which is one of the factors important for continence apart from good bladder capacity and intact bladder neck.[6] Repair of urethra, clitoris and labia minora gave her good and acceptable appearance of external genitalia and urinary continence was achieved. Need for careful genital examination of an incontinent child is emphasized in order to avoid misdiagnosis.

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Cite this article as: Krishna Shetty MV, Bhaskaran A, Sen TK. Female epispadias. Afr J Paediatr Surg 2011;8:215-7.

Source of Support: Nil. **Conflict of Interest:** None declared.