Complete Female Epispadias with Urinary Incontinence: A Single-Stage Perineal Urethroplasty and Cystoscopic-Guided Bladder Neck Plication

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Abstract

Isolated female epispadias without bladder exstrophy is an extremely rare congenital anomaly. The presenting features of female epispadias are urinary incontinence and abnormal anatomical features. A 4-month-old girl presented with primary urinary incontinence since birth. On physical examination, she had a bifid clitoris and labia minora along with an anteriorly placed introitus and wide-open bladder neck. The vagina and hymen were normal. With the diagnosis of isolated complete female epispadias, a single-stage reconstruction of the urethra, labia minora and clitoris was performed along with cystoscopic-guided bladder neck plication. This index case highlights the surgical technique and the necessity to operate on these patients at a younger age in infancy with a single-stage perineal repair to achieve better functional and cosmetic outcomes.

Keywords: Cystoscopy, epispadias, female, perineal urethragenitoplasty

Introduction

Isolated female epispadias without bladder exstrophy is a rare congenital anomaly affecting 1 in 480,000 females.[1] It represents one of the milder forms of the spectrum of bladder exstrophy. In general, the patients present with abnormal external genitalia and a wide anterior gap between the labia majora and bifid clitoris. The urethral plate extends between the clitoral halves. In most cases, the bladder neck is located at the level of the hymenal ring. The urethra and sphincteric mechanisms are absent, resulting in complete incontinence, which is often accompanied by a bladder capacity that is relatively small for age.[2] The vagina and internal genitalia are usually normal. A one-stage urethral reconstruction, intra-abdominal positioning of the bladder neck and proximal urethra and external genital reconstruction appears to be a promising approach for treating female epispadias.

Case Presentation and Operative Technique

A 4-month-old female child with 6 kg body weight presented to the outpatient department with continuous urinary incontinence since birth. On examination, along with epispadias, there was a bifid clitoris, visible open bladder neck, anteriorly placed vestibule, and anal opening [Figure 1]. Further, on radiological examination, there was an underlying wide pubic diastasis of about 5 cm. Ultrasonography of the abdomen was normal. The child underwent cystoscopy which revealed a small capacity bladder (<10 ml on pre-operative cystogram), wide-open bladder neck and normally placed ureteric opening. A perineal urethroplasty was done over 8 Fr. Silicone Foley's catheter using an inverted U-shaped random pattern flap with around the opened bladder neck of size 10 cm in length and 4 cm in width along with genitoplasty as shown in Figure 2a-d. This inverted U-shaped skin flap was taken to create neourethra long enough so that the bladder neck could be easily placed deep inside the pelvis and the neourethra could be brought to its normal location in the vestibule. An infrasymphyseal bladder neck repair or plication was completed by approximating the

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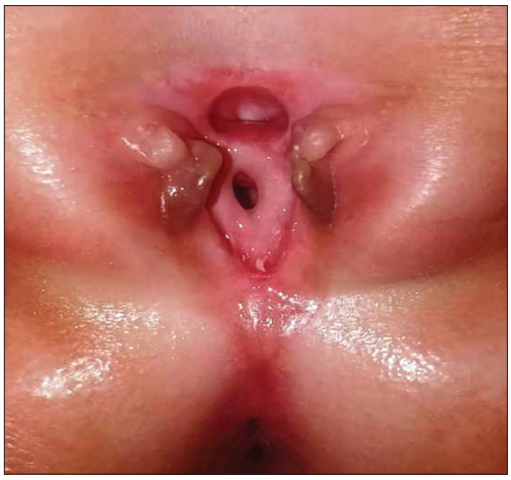


Figure 1: Complete female epispadias with wide-open bladder neck

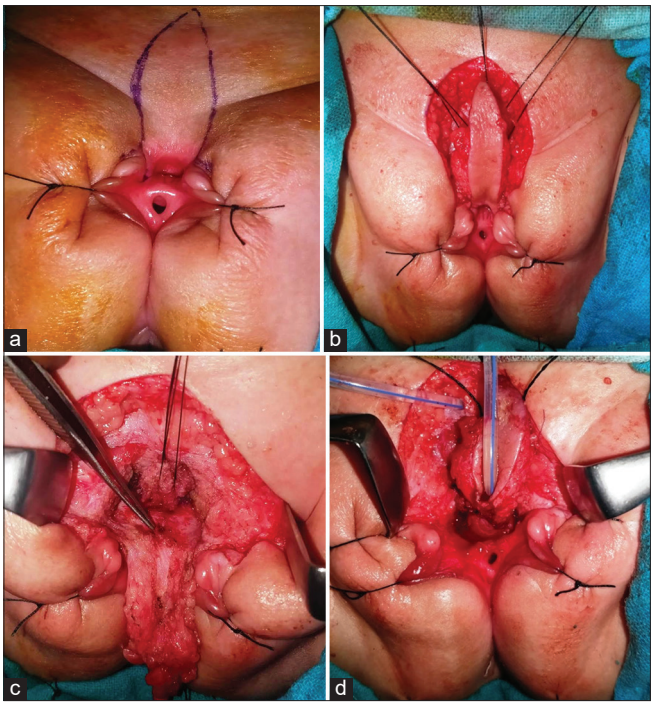


Figure 2: (a-d) Steps of surgery: Perineal urethrogenitoplasty (a) Marking of inverted U-shaped skin flap over exposed bladder neck with inverted 'Y' incisions over bifid clitoris on both sides, (b) flap mobilised completely and bladder neck is pushed inside the pelvis, (c) defattening of flap done (marked with silk suture) along with further bladder neck release, (d) ventral long urethroplasty is done with 6-0 Vicryl suture followed by genitoplasty to achieve cosmesis

open bladder neck in continuation of neourethra. Moreover, the bladder neck had to be further mobilised laterally on both the sides while doing repair to place it deep inside the pelvis. The dead space between the pubis above and bladder neck below was then covered with the residual subcutaneous tissue remained after defattening of the skin flap done after the creation of neourethra. Further, in this technique, during bladder neck repair, the on-table tightening of the bladder neck by the surgeon is assessed simultaneously by urethrocystoscopy done by the assistant. This procedure is done to assess the effectiveness and completion of bladder neck repair and also prevent excessive tightening. A cystoscope of 8.5 Fr could pass through the neck snugly but easily and on-table bladder filling and emptying was also done to measure its capacity (~20 ml). For pubic symphyseal diastasis, the approximation of the anterior pubis was done using no. 1 Ethibond suture and further supported by a hip spica [Figure 3a and b]. The post-operative period was uneventful. The urinary catheter was removed after 10 days; however, the suprapubic catheter and hip spica were removed on day 21. The wound site healed well with a good cosmesis [Figure 4a and b]. The urinary incontinence improved significantly, as the child had a dry period of 2 h from the 1st week of catheter removal. Post-operative cystogram at 6 months showed improved bladder size (50 ml) with a good bladder neck repair without any evidence of vesicoureteric reflux. On follow-up at 1 year, she had 10 kg body weight and presented with improved urinary continence with occasional daytime stress incontinence on coughing or crying and dry periods of more than 3 h.

Discussion

Female epispadias is an uncommon congenital abnormality of the lower urogenital tract, which often presents with urinary incontinence, as in the index case. Usually, it occurs sporadically, but in some cases, there is a strong genetic component.[3] The incontinence varies from continuous incontinence of urine without bladder filling to episodes of daytime stress incontinence. Often the bladder capacity is reduced as a consequence of the lack of filling.[4] External genitalia can have varied appearance as classified by Davis ranging from lesser degrees with the 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 the urethra and bladder neck, rendering the sphincteric mechanism incompetent.[5] Milder forms of epispadias are extremely rare.[2] The objectives of surgical repair include the achievement of urinary continence with preservation of the upper urinary tracts and the reconstruction of functional and cosmetically acceptable genitalia. Further, to achieve continence, an intra-abdominal position of the bladder neck and proximal urethra is essential.[6] A single-stage perineal approach with bladder neck plication has the advantage to attain good bladder size and urinary continence. Similarly, as seen in our case, a one-stage perineal urethrogenitoplasty along with cystoscopic-guided bladder neck plication was performed at the age of 4 months to attain early and better functional and cosmetic results. Even though we had good results with our technique, more experience is needed to confirm the superiority of this technique for this rare anomaly. Therefore, we recommend this single-stage random flap-based repair for urethral reconstruction and cystoscopic-guided bladder neck plication technique, as it is a simple, reproducible and effective procedure for continence as well as cosmesis in patients with isolated female epispadias.

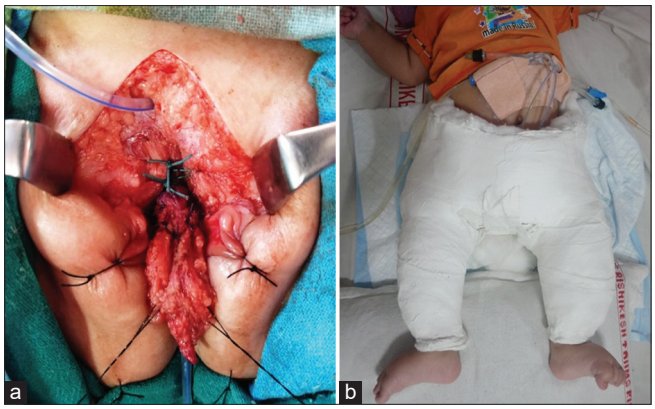


Figure 3: (a and b) Pubic diastasis suture correction and hip spica application post-correction

Conclusions

Complete female epispadias is a rare urogenital anomaly. An early single-stage perineal urethrogenitoplasty and a simultaneous cystoscopic-guided bladder neck repair or plication are recommended to achieve urinary continence and excellent cosmesis. The surgical repair should include proper skin flap and incision planning, cutaneous flap liberation to create a neourethra, complete bladder neck release to allow a controlled plication and use of simultaneous intraoperative cystoscopy to accomplish ideal bladder neck repair or plication to achieve better urinary continence.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

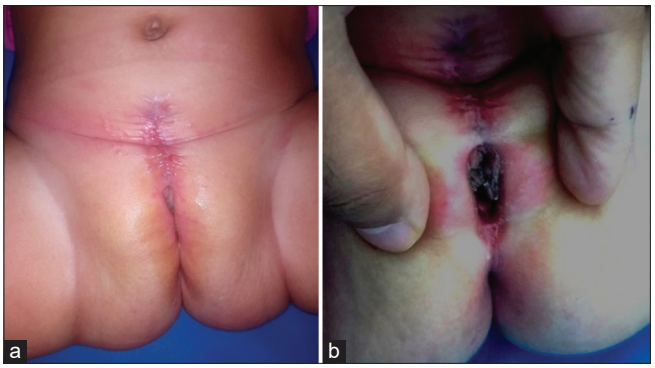


Figure 4: (a and b) Post-operative appearance and perineal cosmesis

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Conflicts of interest

There are no conflicts of interest.

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