Failed repair of isolated female epispadias: Insights and outcome of single stage repair

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Abstract
Female epispadias without extrophy is a very rare entity and occurs one in 480,000 female population. Early surgical reconstruction of the bladder neck, urethra and external genitalia is important in establishing urinary continence and to reduce psychological stress on the parents and child. In this case report we present a 25 year unmarried female presented with failure to achieve continence after bladder neck reconstruction at age 4 year by urologist. Due to previous surgical failure, Patient developed psychological problems with poor esteem and wants a procedure which makes her life socially acceptable. A continent cutaneous urinary diversion was done at a single procedure with a follow up of 02 year. Patient is continent with pouch capacity about 400ml and emptying her pouch every 4hrly and actively involving in social activities, enjoying an excellent lifestyle, accepted body image and good personal satisfaction.

Introduction
Female Epispiadias is a rare congenital anomaly occurring in one in 480,000 female population [1]. The condition is often missed at first examination but should be diagnosed immediately at birth. Diagnosis can only be made after separating the labia. Early diagnosis and surgical reconstruction of the bladder neck, urethra and external genitalia are relevant to improve the chance for urinary continence [2]. In most instances the vagina and internal genitalia are normal. Lack of labial anterior commissure, bifid clitoris, short and wide urethra and immature bladder neck are main properties of this pathology.

Case report
A 25-year old unmarried female presented to our outpatient department because of total urinary urinary incontinence both day and night. She had been previously evaluated and surgically treated at age of 04 years as young dees procedure only. She was consulted by many urologist and multiple cystoscopies and no proper diagnosis was made. On evaluation of records only bladder neck reconstruction as young dees procedure was done. urethral and external genitalia reconstruction was not addressed. Due to failure of treatment, the patient developed psychological problems with poor esteem and wants a procedure which makes her life socially acceptable. A continent cutaneous urinary diversion was done at a single procedure with a follow up of 02 year. Patient is continent with pouch capacity about 400ml and emptying her pouch every 4hrly and actively involving in social activities, enjoying an excellent lifestyle, accepted body image and good personal satisfaction.
closing bladder neck by foley’s catheter. Urodynamics was tried twice but was inconclusive as the patient was not able to hold urine because of severe epispadias. Because of earlier failed reconstruction and never attained continence with the reason hardly much bladder template for reconstruction and even though requires anti incontinence procedure for continence after augmentation. Pyocystitis is unlikely as there is type 3 female epispadias and bladder neck is visible on genital examination. For closure of bladder neck with augmentation cystoplasty chance of fistula was risk and even though she needs mitrofanoff for intermittent procedure All pros and cons about surgical reconstructive procedures were explained in detail. Patient was insisting one time durable procedure. Patient consented for continent catheterizable reservoir made from ileum (Figure 2). Patient is on regular follow up for the last 02 year with a pouch capacity about 400ml and is continent actively involved in indoor and outdoor activities.

Discussion

The aetiology of female epispadias is unclear and is considered possibly to be the result of combination of several genetic and environmental factors [3]. Associated anomalies are commonly confined to the urinary tract with an incidence of vesicoureteric reflux at 30–75%. Incontinence is not only a common complaint in female epispadias but is also associated with noticeable anatomical features like an absent or bifid clitoris [4] depressed mons and ill developed labia. The bladder is often small with poorly developed bladder neck and incompetent sphincteric mechanism. Early diagnosis allows early parenteral counseling and a planned surgical reconstructive procedure at 4 to 5 years of age when the bladder capacity reaches approximately 80 to 85ml. In our case report of adult female epispadias with type 3 variant and early reconstructive surgery as young dees procedure at age of 04 years, three options were given to the patient for the female epispadias. First option using bladder template with augmentation of bowel, urethral reconstruction, genitoplasty and optional sling procedure for incontinence and continuous indwelling catheterization if fails to void. Second option was closure of bladder neck with augmentation of bowel and continent catheterizable pouch. In second option risk of bladder fistula was explained in view of previous bladder neck surgery. Third option was leaving bladder and make an continent catheterizable reservoir from ileum and incorporate ureters into the reservoir. Patient agreed for third option and the issues were bladder in situ. As the patient was never continent after toilet training, so no feeling of incomplete emptying and spasm will occur. Moreover if need arises cystoscopy is possible for the native bladder. In our case it was an isolated type 3 epispadias, no adenocarcinoma of retained bladder will occur as is the case with exstrophy of bladder. However pyocystitis may or may not develop on follow up. A 60 cm ileum was used and divided into three segments. 40 cms for reservoir and 10cms each for afferent and efferent limbs and stoma was brought through umbilicus (Figure 3). On follow up after 06 months upper tracts were evaluated and reservoir was reconstructed (Figure 4) using radiocontrast studies. Patient is continent both day and night.
with catheterization interval of 4hrs and enjoying an excellent lifestyle, accepted body image and good personal satisfaction. As bowel is used for neobladder needs long term follow up for metabolic complications and even malignancy.

**Conclusion**

The case is presented here because of its rarity and to emphasize upon careful examination and proper reconstruction of an incontinent child, providing them proper treatment, to reduce psychosocial problems. Moreover a cutaneous continent catheterizable stoma is a viable option in patients with early failed reconstruction in female epispadias.

**References**


**Highlights**

- Signatory publisher of ORCID
- Signatory Publisher of DORA (San Francisco Declaration on Research Assessment)
- Articles archived in worlds’ renowned service providers such as Portico, CNKI, AGRIS, TDNet, Base (Bielefeld University Library), CrossRef, Scilit, J-Gate etc.
- Journals indexed in SCIE, SHERPA/ROAR, Google Scholar etc.
- OAI-PMH (Open Archives Initiative Protocol for Metadata Harvesting)
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