International Journal of Life Sciences, Biotechnology and Pharma Research Vol. 13, No. 10, October 2024 Online ISSN: 2250-3137
Print ISSN: 2977-0122

DOI: 10.69605/ijlbpr_13.10.2024.76

ORIGINAL RESEARCH

Surgical experience in a series of 5 cases of female epispadias, Sardar Patel Medical College, Bikaner

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Received Date: 23 August, 2024 Accepted Date: 29 September, 2024

ABSTRACT

Background: Epispadias is an uncommon urogenital anomaly characterized by the incomplete formation of the urethral tube. The aims of surgical intervention encompass the attainment of urinary continence while safeguarding the integrity of the upper urinary tracts, alongside the reconstruction of genitalia that is both functional and aesthetically pleasing. **Materials & methods:** The present study was conducted fordescribing surgical experience in a series of 5 cases of female epispadias reported at Sardar Patel Medical College, Bikaner. Complete demographic and clinical details of all the patients was obtained. Prospective inclusion of incontinent girls with female epispadias was done. A total of 5 patients were enrolled who were managed surgically. Follow-up was at 1, 3, 6, and 12 months postoperatively, then annually, including physical examination, renal ultrasound at each visit, continence status. **Results:** All the patients belonged to age range of 5 to 10 years. Urinalysis indicated an average of over 13.8 red blood cells and 8 white blood cells per high power field. No abnormalities were observed in the intravenous urogram or ultrasound examinations. Urethrocystoscopy revealed a short and wide urethra in all cases examined. A combined Young-Dees and urethroplasty procedure was subsequently performed. Key aspects of the procedure included the tubularization of the urethral plate and the relocation of the bladder neck and proximal urethra to an intra-abdominal position. Continence was achieved by creating a posterior strip of mucosa measuring 15 mm in width and 30 mm in length, extending from the midtrigone to the posterior urethra. The surgical intervention concluded with a cosmetic reconstruction of the external genitalia, and the pubic symphysis was approximated. A suprapubic tube and a urethral tube were inserted, with the suprapubic tube being removed on the 21st postoperative day and the urethral tube on the 10th postoperative day. At the three-month follow-up, there were no reported instances of urinary incontinence. **Conclusion:** Epispadias in females is an uncommon congenital defect that is frequently subject to misdiagnosis. The objectives of surgical intervention include attaining urinary continence, restoring the anatomy and function of the urethra and genitalia, as well as achieving aesthetic improvement.

Key words: Stroke, Thrombolytic, Renal

INTRODUCTION

Epispadias is an uncommon urogenital anomaly characterized by the incomplete formation of the urethral tube. In contrast to hypospadias, where the urethra is intact but opens proximally on the ventral side of the penis, epispadias presents with an open urethral plate located on the dorsal surface of the penis. This condition is included in the exstrophy-epispadias complex (EEC), which encompasses a spectrum of congenital genitourinary deformities, ranging from mild epispadias to severe cases of bladder or cloacal exstrophy.[1-3] In males, epispadias is associated with distinct anatomical features, such as a short phallus that points upwards, with the urethral opening situated variably along the dorsal side, accompanied by ventral hooding of the prepuce. Female epispadias is significantly rarer, with reported incidences between 1 in 160,000 and 1 in 480,000 live births. Young females with this condition typically exhibit a bifid clitoris, a wide urethral opening, an anteriorly positioned vaginal orifice, and a poorly developed or absent mons pubis. Isolated epispadias is a rare spontaneous defect that predominantly affects males but can also occur in females, necessitating surgical intervention to achieve urinary continence and improve cosmetic appearance.[4, 5]The aims of surgical intervention encompass the attainment of urinary continence while safeguarding the integrity of the upper urinary tracts, alongside the reconstruction of genitalia that is both functional and aesthetically

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pleasing. In cases of severe severity, a range of surgical techniques has been documented to manage continence, including transvaginal urethral and bladder neck placation, muscle grafting, urethral rotation, bladder flap procedures, and Marshall Marchetti vesicourethral suspension. These interventions serve to enhance urethral resistance. Additionally, the elongation of the urethral conduit during the reconstruction process itself contributes to the amelioration of incontinence.[6, 7]Hence; the present study was conducted fordescribing surgical experience in a series of 5 cases of female epispadias reported at Sardar Patel Medical College, Bikaner.

MATERIALS & METHODS

The present study was conducted fordescribing surgical experience in a series of 5 cases of female epispadias reported at Sardar Patel Medical College, Bikaner. Complete demographic and clinical details of all the patients was obtained. Prospective inclusion of incontinent girls with female epispadias was done. A total of 5 patients were enrolled who were managed surgically. Follow-up was at 1, 3, 6, and 12 months postoperatively, then annually, including physical examination, renal ultrasound at each visit, continence status. All the results were recorded in Microsoft excel sheet and were subjected to statistical analysis using SPSS software.

RESULTS

Table 1: Age-wise distribution

Value	Age
Case 1	5 years
Case 2	7 years
Case 3	6 years
Case 4	6 years
Case 5	10 years

Table 2: Urine analysis

Value	Red blood cells per high power field	White blood cells per high power field
Case 1	12	8
Case 2	14	7
Case 3	14	9
Case 4	16	8
Case 5	13	8

All the patients belonged to age range of 5 to 10 years. Urinalysis indicated an average of over 13.8 red blood cells and 8 white blood cells per high power field. No abnormalities were observed in the intravenous urogram or ultrasound examinations. Urethrocystoscopy revealed a short and wide urethra in all cases examined. A combined Young-Dees and urethroplasty procedure was subsequently performed. Key aspects of the procedure included the tubularization of the urethral plate and the relocation of the bladder neck and proximal urethra to an intra-abdominal position. Continence was achieved by creating a posterior strip of mucosa measuring 15 mm in width and 30 mm in length, extending from the midtrigone to the posterior urethra. The surgical intervention concluded with a cosmetic reconstruction of the external genitalia, and the pubic symphysis was approximated. A suprapubic tube and a urethral tube were inserted, with the suprapubic tube being removed on the 21st postoperative day and the urethral tube on the 10th postoperative day. At the three-month follow-up, there were no reported instances of urinary incontinence.

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. The incontinence varies from continuous dribbling of urine without bladder filling to episodes of day-time stress-incontinence. Often the bladder capacity is reduced as a consequence of the lack of filling. External genitalia can 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. Milder forms of epispadias are extremely rare. 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.[8- 10]Hence; the present study was conducted fordescribing surgical experience in a series of 5 cases of female epispadias reported at Sardar Patel Medical College, Bikaner.

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et al presented a case of unrecognized female epispadias. A 16-year-old female with epispadias, history of mild urinary incontinence, auditory neuropathy and functional hyperandrogenism. The patient was referred for evaluation of excessive weight gain, secondary amenorrhea and abnormal external genitalia. Examination under anesthesia revealed bilateral labia minora hypertrophy, bifid clitoris and a patulous urethra, consistent with female epispadias. Hormonal evaluation showed functional hyperandrogenism while peripheral blood array-based comparative genomic hybridization (array CGH) showed no chromosomal deletions or duplications.Female epispadias is a rare abnormality, not commonly recognized by most practitioners. The diagnosis is supported by a history of urinary incontinence and physical findings of bifid clitoris and patulous urethra. The condition can have serious physical and psychological consequences leading to a gross disruption of social function.[11]

Leclair MD et al assessed the results of a surgical management using perineal approach in girls with normal bladder capacity, and Kelly radical soft-tissue mobilization (RSTM) in patients with inadequate bladder, based on the assumption that bladder capacity (BC) is a reliable marker of epispadias severity.16 consecutive children were prospectively included in this study, at a median age of 39 months (5-102 months). Seven girls were included in group 1 and underwent PUCP; at the last follow-up, five out of seven were dry by day (4/5 day and night), although three out of five required bladder-neck injection after perineal reconstruction due to stress incontinence. Two patients with persistent incontinence and absence of BC increase after PUCP subsequently underwent RSTM. Eleven patients with low BC were included in group 2. Among the eight evaluable patients, eight out of eight achieved diurnal continence, and 3/8 were fully continent. One girl with obstructive micturition required clean intermittent catheterization.The traditional approach of female epispadias based on staged reconstruction (urethroplasty followed by bladder-neck reconstruction) raised concerns regarding the risk of non-physiological obstructive micturition.[12]

CONCLUSION

Epispadias in females is an uncommon congenital defect that is frequently subject to misdiagnosis. The objectives of surgical intervention include attaining urinary continence, restoring the anatomy and function of the urethra and genitalia, as well as achieving aesthetic improvement.

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