



“FEMALE EPISPADIAS WITHOUT EXSTROPHY: A RARE CLINICAL REPORT”

Dr. Deepak Kumar	Senior resident, Department of Pediatric Surgery, Institute of Medical Sciences-BHU, Varanasi.
Dr. Sarita Choudhary	Associate professor, Department of Pediatric Surgery, Institute of Medical Sciences-BHU, Varanasi.
Dr. Shyamendra Pratap Sharma	MCH SR, SGPGI Lucknow.
Dr. Preeti Sagar*	PG Final Year, Department of Prosthodontics, Crown & Bridge and Implantology, K D Dental College & Hospital, Mathura. *Corresponding Author

ABSTRACT

Female epispadias case without exstrophy are quite rare. A 8-year old girl presented with complaint of primary total incontinence. Physical examination revealed skin excoriation in the external genitalia due to prolonged urine contact, non-fused labia minora, and very short urethra. Bladder capacity was found to be sufficient in intravenous urogram. Our approach was to use single stage reconstructive procedure that enables correction of the urinary incontinence and enhance good aesthetic appearance.

KEYWORDS : Female epispadias, single stage reconstructive procedure, Urinary incontinence

INTRODUCTION

Female epispadias without exstrophy is an extremely uncommon anomaly reported in females. The bladder and genitourethral repair are two main components of epispadias surgery, which are frequently overlooked on first inspection but should be detected at birth.¹

Case Report

An eight-year-old girl presented with complaint of primary total incontinence. Physical examination revealed skin excoriation in the external genitalia due to prolonged urine contact, non-fused labia minora and very short urethra (Figure 1).. No significant finding was found in laboratory and X-ray examinations except diastasis of the pubic bones in the plain film of abdomen. Intravenous urogram revealed adequate bladder capacity. The short, wide open urethra and the wide, incompetent bladder neck were verified by cystourethroscopy.



Figure 1: Preoperative appearance of female epispadias without bladder exstrophy

During the single-stage reconstructive operation, the urethral plate, bladder collar, and bladder were carefully dissected free and mobilised in a natural plane near to the pubic bones releasing all of the sphincteric muscle that was accessible laterally and anteriorly. The usable 'sphincteric muscle' were approximated in the midline. Urethral plate and bladder neck were tubularized with 8 Fr silicone catheter & disrupted with 5.0 vicryl absorbable sutures wrapped around the bladder neck. (Figure 2 & 3).



Figure 2 & 3 : Surgical procedure carried out

The hemiclitori (glans and corpora) were medially freshened and approximated in the midline above the urethral orifice at the normal location. The procedure was finished with a cosmetic reconstruction of the external genitalia (Figure 4). A suprapubic cystostomy was placed and covered with antibiotics for three weeks. There was no pelvic osteotomy performed.



Figure 4: Final appearance after surgery with sutures placed

No post-surgical complications were noted in the child during recovery period. The passage of a 6 and 8 F Foley catheter through a decent length patent urethra and bladder neck with no stenosis or dehiscence was confirmed under anesthesia 2 weeks after surgery. Her continence had progressed at 6-month follow-up, and she could be dry for at least 120 minutes.

DISCUSSION

The cause of this urethral and bladder neck developmental abnormality is unknown, but it is thought to be the product of a combination of genetic and environmental factors. Despite the fact that epispadias is on the milder end of the exstrophy-epispadias spectrum, it is a profound and complicated developmental anomaly that is often associated with full urinary incontinence. Male epispadias is more common, and female epispadias is extremely rare, with an occurrence of 1:480,000 girls.²

The majority of the associated anomalies are restricted to the urinary tract, with vesico-ureteral reflux accounting for 30 percent to 75 percent of the cases. The pelvis, the pelvic floor, and the abdominal wall are all important to evaluate, but the spine and the anus are also important.³

Female epispadias has distinct physical manifestations that can be detected during a regular informed clinical genital

examination at birth, resulting in inability to potty train and constant wetting at a later age. Early detection allows for early possibility of a scheduled surgical reconstructive operation.

Surgical restoration aims to restore urinary continence while preserving the upper urinary tracts, as well as reconstructing healthy and aesthetically appropriate genitalia.⁴ Various surgical procedures have been identified to manage continence in extreme cases for example, muscle transplantsations, urethra twisting, bladder flap ,transvaginal placion of the urethra and bladder neck and Marshall Marchetti vesicourethral suspension.⁵

The above case is being documented because it is an unusual case of female epispadias without exstrophy, managed by single stage reconstruction. Even the literature supports Single-stage reconstruction to be more healthy and aesthetically acceptable, with a 60 percent to 87.5 percent of urinary continence.^{3,5}

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