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Urology Section

# Adult Bladder Exstrophy Managed by Complete Primary Repair: A Case Report

DEEPAK MANE<sup>1</sup>, SHIVAM SINGH<sup>2</sup>, VIKRAM P SATAV<sup>3</sup>, VILAS P SABALE<sup>4</sup>

# ABSTRACT

Bladder Exstrophic diseases, referred to collectively as Bladder Exstrophy-Epispadiasis complex, are a spectrum of embryological anomalies with a rare prevalence of approximately 3.3 per 1,00,000 live births with a male gender predominance. An 18-year-old man presented to Urology Outpatient Department (OPD) with subumbilical abdominal wall defect, absent anterior bladder wall with epispadiasis and was diagnosed as a case of adult classic bladder exstrophy with Epispadiasis complex. There was no history of any fever, haematuria, dysuria or abdominal pain. His ultrasound abdomen and intravenous pyelography reported normal kidney function. He had a low Body Mass Index (BMI) (17.5 kg/m<sup>2</sup>) with serum creatinine of 0.6 mg/dL. The patient underwent a complete primary repair, including bladder template closure and bladder neck reconstruction using Young-Dees-Leadbetter technique. Epispadias was repaired with the Modified Cantwell-Ransley approach. Abdominal wall defect closure was done with the help of rectus abdominis muscle flap which was later grafted using Split Thickness Skin Graft (STSG). Postoperatively, a small vesicocutaneous fistula developed which healed spontaneously over two weeks period. On follow-up patient was on clean intermittent catheterisation with some incontinence episodes during night-time. Bladder exstrophy in adult patients is rare, as the condition is usually identified in the neonatal period and can be surgically corrected early on. Due to the complexity of the surgery, these cases require thorough multidisciplinary evaluation and careful planning for successful correction.

# **CASE REPORT**

An 18-year-old man, presented to urology OPD with chief complaints of defect over lower abdomen with leakage of urine from the defect and small sized, upward deviated penis present since birth. He had no history of any fever, haematuria or abdominal pain and no history of any other deformity. There was no history of any co-morbidity or any surgeries in past with negative history of any similar complaints in the family. Patient belonged to a low-socioeconomic status.

On general physical examination his vitals were normal with a low BMI of 17.5 kg/m<sup>2</sup>. There was no obvious gait deformity. On local examination there was a subumbilical abdominal wall defect of size approximately 14 cm with posterior wall of bladder visible. Bilateral ureteric orifices were present as umbilical defects in the bladder with urine leakage. Epispadiasis was also seen with short upward deviated penis [Table/Fig-1a]. His blood parameters were normal with a serum creatinine of 0.6 mg/dL (normal- 0.74 to 1.35 mg/dL). An ultrasound of the abdomen and intravenous pyelography showed bilateral normal sized and functioning kidneys with no other obvious congenital anomaly. Wide pubic diastasis was seen on X-ray hip [Table/Fig-1b]. Multiple random bladder biopsies were suggestive of



[Table/Fig-1]: a) Clinical presentation of classic bladder exstrophy with epispadiasis complex - absent lower abdominal wall (yellow arrow), deficit anterior bladder wall (black arrow), bilateral ureteric orifices (blue arrow) and complete epispadiasis (red arrow); b) X-ray Hip showing wide public diastasis (yellow arrow).

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focal areas of keratinisation with non specific inflammatory changes. Psychiatric evaluation stated a patient with adequate IQ.

After thorough work-up, orthopaedic and plastic surgery consultation, patient was offered the surgical treatment options of either a bladder template excision followed by ileal conduit urinary diversion or a complete primary repair of exstrophy and followed by a second staged procedure for bladder neck reconstruction with ileocsytoplasty, if required. Patient and his family were counselled regarding the complexity of the procedure and the need for clean intermittent catheterisation or a urinary stoma in the postoperative period.

Intraoperatively, two ureteral stents were placed and liberated the entire bladder plate [Table/Fig-2a-c], preserving the anterior and inferior pedicles. Decision was taken for complete primary repair of the bladder along with placement of 14 fr Suprapubic Catheter (SPC) due to its adequate volume. Bladder neck reconstruction was done using Young-Dees-Leadbetter technique [1].



[Table/Fig-2]: a) Ureteral catheterisation (white arrow) with marking of bladder template and urethral plate (black arrow) for mobilisation; b) Bladder mobilisation and primary repair of bladder template (arrow). c) Illustrative image showing modern stage I (MRSE) repair technique for Complete Primary Repair of Exstrophy (CPRE).

[Table/Fig-2c] showed an illustration of Modern Stage I (MRSE) repair technique for Complete Primary Repair of Exstrophy (CPRE) [2]. Using Modified Cantwell-Ransley repair technique by Gearhart JP et al., repair of the epispadiasis was done, involving disassembly of the corporal bodies along with ventral transposition of the corpus spongiosum and neourethra which was tubularised over 14 french foleys catheter [Table/Fig-3a-c] [3].



[Table/Fig-3c] showed Modified Cantwell-Ransley repair [2]. An inferior epigastric artery-based rectus abdominis muscle flap was utilised for abdominal wall closure. The rectus sheath was incised on the lower medial aspect for mobilisation of rectus muscle. Upper end of the muscle was cut and fixed to the opposite side rectus sheath, covering the abdominal defect [Table/Fig-4a,b]. Similar approach has been utilised by Fahiem-Ul-Hassan M et al., in a comparative study, over 30 paediatric patients and found that use of rectus muscle flap lowered the incidence of wound dehiscence and also helped in achieving continence [4].



**[Table/Fig-4]:** a) Abdominal wall defect post bladder repair (arrow); b) Rectus muscle flap (arrow) based on inferior epigastric artery to cover the defect.

Postoperatively, on day 2, patient did develop a small wound dehiscence of the flap for which secondary suturing was done. On postoperative day 5 (pod 5), STSG was taken from left thigh which was placed over the rectus abdominis muscle flap bed.

On POD 8, patient developed a small vesicocutaneous fistula which healed spontaneously over period of time. Skin Graft uptake was adequate and patient was discharged on POD 15 after suture removal with foleys catheter in situ and clamped SPC. Patient counselled for regular follow-up for check cystoscopies and ultrasound of bladder. SPC was removed three weeks postoperatively [Table/Fig-5].



[Table/Fig-5]: Adequate graft uptake (arrow) on postoperative day 15.

# DISCUSSION

Bladder exstrophy, cloacal exstrophy and epispadias are variants of the exstrophy-epispadias complex, ranging from epispadiasis in milder form to cloacal exstrophy in severity of the spectrum of embryological anomaly disease. It has a rare prevalence of approximately 3.3 per 1,00,000 live births, with a male gender predominance [5]. It has been linked to maternal smoking and irradiation during the first trimester [6]. Major defects involve absent subumbilical anterior abdominal wall, with loss of ventral surface of bladder, bony pelvis defect with pubic diastasis and 'open book configuration' having short anterior bony segments and outward rotated both anterior and posterior segments [7]. Anterior wall of bladder is deficient but with normal neurophysiological composition [8]. The diagnosis of bladder exstrophy is clinical and only requires routine investigations as a preoperative work-up and for evaluation of upper urinary tracts [9]. The theory of embryonic maldevelopment in exstrophy held by Marshall VF and Muecke EC is that the basic defect is an abnormal overdevelopment of the cloacal membrane during the fourth week of gestation [10].

This patient underwent primary repair, which included bladder closure, bladder neck reconstruction and epispadias repair without osteotomy. In a study by Shoukry AI and Shoukry I, involving five adult patients with bladder exstrophy, three patients were treated similarly, using fasciocutaneous M-plasty for abdominal wall closure [11]. [Table/Fig-6] outlines previously reported cases of bladder exstrophy with epispadias in adulthood, highlighting the management approaches and the surgical armamentarium utilised along-with the postoperative complications faced [11-16].

In present case, both cosmetic appeal and closure of the bladder and abdominal wall were attained, resulting in a more anatomically naturallooking penis. Patient was rendered free of ammoniacal smell along with preservation of upper tracts; however, the patient currently has difficulty in maintaining continence during night-time and is on clean intermittent catheterisation. Adequate penile length was not achieved due to the persistence of pubic diastasis and short corporal bodies length. On repeat check cystoscopy at three months after surgery, bladder capacity was estimated to be around 400 mL with no signs of inflammation. Patient was advised for regular follow-up for evaluation of upper urinary tracts and any signs of urinary tract infection.

# CONCLUSION(S)

Adult patients with bladder exstrophy are usually of low socioeconomic status, socially neglected and with low self-esteem. Patients undergoing cystectomy with creation of a continent pouch or bladder reconstruction tend to have improved self-confidence and social interaction after surgery. In patients of exstrophy bladder, reconstructive management is best started in the neonatal period with good results. Adult bladder exstrophy patients are a surgical challenge, requiring a multidisciplinary approach for the correction of the skeletal and abdominal wall deformity along with urological correction. Good knowledge of different methods of anterior abdominal wall reconstruction following cystectomy or cystoplasty is mandatory along with proper planning.

Authors name and year of publication	Place	Number of patients/ Age/Gender	Findings	Management	Follow-up
Mokadem S et al., [12] (2022)	Tunisia	1/26 years/F	Inflammatory bladder plate with two clitoral halves and a fourth-grade uterine prolapse	lliac osteotomy, bladder enlargement using the ileum a Monti-type continent urinary diversion and a Promentofixation.	Vesicocutaneous fistula developed after surgery hence, cystectomy was done with continent diversion.
Peteinaris A et al., [13] (2022)	Patras, Greece	25 years/F	Classic bladder exstrophy with acute right pyelonephritis	Managed with i.v. antibiotics however no surgical reconstruction was done.	
Kiran PS et al., [14] (2020)	New Delhi, India	25 patients/Mean age 25 years	Classic bladder exstrophy only	18 patients underwent continent catheterisable pouch (Pen Pouch), 3 patients – complete primary repair, 4 patients – Ileal Conduit. No osteotomy was done.	At 6.5 years follow-up – patients with continent pouches were continent, except one patient of Complete Primary Repair of Exstrophy (CPRE) other two were continent.
Shoukry Al and Shoukry I, [11] (2013)	Cairo, Egypt	5 male patients/17- 30 years	All patients had classic Exstrophy-Epispadiasis complex	Three patients underwent primary repair, with bladder closure, bladder neck reconstruction and epispadias repair while two had augmentation ileocystoplasty. Remaining two patients underwent ureterosigmoidostomy, cystectomy and epispadias repair. Fasciocutaneous M-plasty was done for abdominal wall closure. No osteotomy was done in any case.	
Nerli RB et al., [15] 2008	Kamataka, India	7 adult male patients/Mean age 19 years	Classic bladder exstrophy with epispadiasis	5 patients underwent cystectomy with continent pouch (Mainz) and two had bladder reconstruction (lleocystoplasty with young dees bladder neck reconstruction). Epispadiasis repair with modified Cantwell-Ransley method.	On follow-up all patients were continent during daytime and one patient had night-time leak.
Pathak HR et al., [16], 2001	Mumbai, India	4 adult male patients	Classic bladder exstrophy with epispadiasis	lleocystoplasty, bladder neck reconstruction with epispadiasis repair in two stages.	At 48 months follow-up, three patients were satisfied with the cosmetic results while one complained of small sized penis.
Present case, 2024	Pune, India	18 years/M	Classic bladder exstrophy with epispadiasis	Complete primary repair of bladder template, with bladder neck repair using Young-Dees- Leadbetter approach. Epispadiasis repair was done with Cantwell-Ransley method. Rectus muscle flap was used for abdominal wall repair. No osteotomy.	No long-term incidence of wound dehiscence. Patient on clean intermittent catheterisation with some night incontinenece. Short penile length.

[Table/Fig-6]: Similar cases on review of literature [11-16]

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### PARTICULARS OF CONTRIBUTORS:

- Associate Professor, Department of Urology, Dr. D. Y. Patil Medical College, Pimpri, Chinchwad, Pune, Maharashtra, India.
- 2. Resident, Department of Urology, Dr. D. Y. Patil Medical College, Pimpri, Chinchwad, Pune, Maharashtra, India.
- Professor, Department of Urology, Dr. D. Y. Patil Medical College, Pimpri, Chinchwad, Pune, Maharashtra, India. З.
  - Head, Department of Urology, Dr. D. Y. Patil Medical College, Pimpri, Chinchwad, Pune, Maharashtra, India.

# NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Shivam Singh, Urology Department (OPD No. 14), Hi-tech Building, Dr. D. Y. Patil Medical College,

Sant Tukaram Nagar, Pimpri, Pune-411018, Maharashtra, India. E-mail: droptoshivam@gmail.com

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