Outcomes of Complete Primary Repair of Bladder Exstrophy Epispadias in Adult Patients in a Tertiary Care Centre

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Abstract

Introduction: Classic bladder exstrophy is a rare malformation of the Genito-urinary tract. The surgical reconstruction of the BE-epispadias complex is challenging. Complete primary repair of bladder exstrophy (CPRE) has offer the promise of improved bladder functional outcomes. We present, out come on five patients who have undergone complete primary repair of bladder exstrophy in terms of closure of the bladder and abdominal wall with eventual continence, preservation of renal function, and cosmetic and functional reconstruction of the genitalia. **Materials and methods:** The records of patients areanalysed retrospectively. The sample size is five patients; all adult patients age between 17 to 22 years. All are treated in single stage. Bladder augmentation is done in every case, bladder neck repair is done using paraurethral tissue, primary abdominal closure is done with rotation-advancement abdominal flap, epispadias repair is done using modified Cantwell-Ransley procedure and suprapubic ramus transverse minimal osteotomy lateral to insertion of rectus abdominis and adductor longusis done. All the patients are followed regularly. Minimum period of follow-up is one year. **Results:** All patients are continent. One patient has developed aurethro-cutaneous fistula at the base of the penis. In two patients there are grade II VUR which are treated conservatively. All the patients are on clean intermittent self-catheterization. **Conclusion:** CPRE offers a viable alternative to MSRE, with favourable continence and renal outcomes. Proper surgical execution in the first attempt is critical for long-term success, emphasizing the importance of good surgical technique. Further long-term studies are necessary to solidify CPRE's role as the primary surgical approach for bladder exstrophy epispadias complex.

Keywords: Augmentation, continence, exstrophy bladder, minimal osteotomy, abdominal wall closure.

Introduction

Bladder exstrophy is a relatively rare combination of complex anatomical anomalies that occurs during early fetal development. The disorder usually involves several organ systems in the body, including the urinary tract, the reproductive tract, the digestive system, the muscles and the skin of the lower abdominal wall and the muscles and bones of the pelvis. Some bones of the pelvis are shortened compared to normal pelvic bones. Bladder exstrophy is seen in approximately 1 in every 40,000 births [1].

For individuals born with bladder exstrophy:

- The inner surface of the urinary bladder is open on the lower abdominal wall.
- The bladder is not closed and not covered as it would be normally by muscle and skin, but rather the bladder is essentially inside out with the inner surface exposed. The inner lining of the urethra is exposed on the top surface of the penis in boys or between the separated right and left halves of the clitoris in girls.
- In the absence of normal closure of the bladder and urethra, there is no ability for the bladder to store urine, and urine constantly trickles onto the exposed inner surface of the bladder and surrounding skin. Following birth, this may cause irritation of the nearby skin prior to repair.

The most important goals in the care of a male or female with bladder exstrophy are to:

- preserve normal kidney function
- develop adequate bladder function and promote urinary continence
- provide acceptable appearance and function of the external genitalia

The repair of bladder exstrophy (BE) remains one of the foremost surgical challenges. The surgical management of bladder exstrophy has evolved during the last years, with the standard treatment until the late 1950s being urinary diversion with ureterosigmoidostomy. Afterwards, in 1970s it evolved to a staged repair, with early pelvic ring approximation and abdominal wall, bladder and posterior urethral closure performed as a first stage, followed by second stage neo urethroplasty (modified Cantwell-Ransley technique) and finally a bladder neck surgical reinforcement such as Yong-Dees-Leadbetter procedure [2]. The modern staged repair (MSRE) technique involves bladder closure shortly after birth, followed by epispadias repair at age 6-12 months and bladder neck reconstruction at age 4-5 years when it is thought that the child can cooperate with attempting continence.

In 1990s, Mitchell introduced the concept of one-stage reconstruction of exstrophy, where all the aforementioned procedures were performed as a single surgery comprehensive approach [3]. The concept of this approach was to decrease the number of surgical procedures required to achieve continence as well as achieve early bladder neck resistance and bladder cycling [4]. This technique presented good results,

although it has some drawbacks such as risk of penile tissue loss and necessity of osteotomies in older children or after failed repair.

Placing the posterior urethra and bladder deep into the pelvis in combination with a tension-free closure and adequate postoperative management prevent complications and are now consensus among urologists [5]. The two well-described techniques: MSRE [6] and one-stage reconstruction of bladder exstrophy (CPRE) advocates the observance of these fundamental principles [4].

The literature on the incidence of adolescent and adult patients with bladder exstrophy is scarce. Herein we report a case series of five adult patients with a bladder exstrophy-epispadias complex who have been treated at our institute.

Materials and Methods

This retrospective review included the medical records of five patients with an exstrophy-epispadias complex managed between June 2023 and August 2024 at our institute. Our study group consisted of 4 female and 1 male patients with a mean age at presentation of 19 (range 18–22) years. They all are from a rural background. Financial constraints presented the main reason for not seeking medical help earlier. They all attended our hospital with the financial help of villagers. The institution provided the entire treatment free of charge.

The patients were subjected to laboratory investigations (e.g., complete hemogram, renal function test) and upper tract imaging in the form of ultrasound scan and X-ray of the pelvis. All patients were informed about the details of the operative procedure and also counselled about the need for clean intermittent self-catheterization (CISC) at regular intervals.

Prior to the definitive procedure, all patients underwent biopsy from the bladder plate to rule out malignancy or premalignant changes. None of the patients underwent cystectomy.

All five patients undergo single-stage complete repair (CPRE) of the exstrophy-epispadias complex[7]. Bladder cytometric capacity expected to age (BcapE) is determined by Koff's formula, [(age in years + 2) x 30] ml.Using ileocystoplasty for augmentation of bladder without re-implantation of the ureter. For bladder augmentation 45 cm small gut was taken, 15cm proximal to ileocecal junction. 40cm for augmentation and 5cm for creation cystostomy stoma. Used para-urethral soft tissue and inter-symphyseal band for bladder neck reconstruction and the modified Cantwell-Ransley[9] method for epispadias repair.Transverse split osteotomy of superior pubic ramus done just lateral to insertion of rectus abdominis and adductor longus bilaterally for approximation of rectus abdominis muscle in midline to reconstruct neo-symphysis over reconstructed bladder neck, that is an important factor for continence. In all patients, the procedure was concluded by tension free primary abdominal closure, this is done by rotation advancement abdominal flap bilaterally.

The patients were discharged after removal of the abdominal drain, urethral stent and suprapubic catheter. All patients were instructed to perform CISC at fixed intervals and after each episode of voiding, to empty the bladder completely.

Regular clinic visits and periodic kidney, urinary tract and bladder ultrasonography, urine analysis and ACR, and blood for urea /creatininewere performed every 6 months to 1 year. Voiding cystourethrography is performed only in cases of urinary tract infections, upper urinary tract dilatation and/or urinary incontinence.

Demographic and baseline characteristics of our subjects:

Case no.	Age/sex	Associated anomalies	Bladder capacity (ml)	Augmentation	Osteotomy	Duration of surgery
1	20 Y/Fe	none	40	Yes	Yes	5 hrs 30 min
2	17 Y/Fe	none	35	Yes	Yes	5 hrs
3	22 Y/M	none	50	Yes	Yes	6 hrs 30 min
4	19Y/Fe	none	40	Yes	Yes	5 hrs
5	18Y/Fe	none	50	Yes	Yes	5 hrs

Results

The mean follows up of the patients has been one year. Follow up investigations include abdominal ultrasound and the evaluation of post-void residual urine. A micturating cysto-urethrogram (MCU) is not routinely performed. Two patients developed reflux of grade II after bladder closure which are managed conservatively;does not lead to permanent renal damage or scarring. The patients are advised to perform regular CISC through stoma and remain dry in between CISC.

At present, the patients' bladder capacity ranges from 400 to 450 ml. All patients are continent. Maintained their normal albumin-to-creatinine ratio (ACR) and creatinine clearance at follow up. One patient who has developed a urethro-cutaneous fistula at the base of the penis is awaiting repair. The scar of the anterior abdominal wall is healthy in all patients. The male patient gives history of adequate erection and ejaculation of semen. The female patients are satisfied with the appearance of the external genitalia. All patients are doing well socially and psychologically.

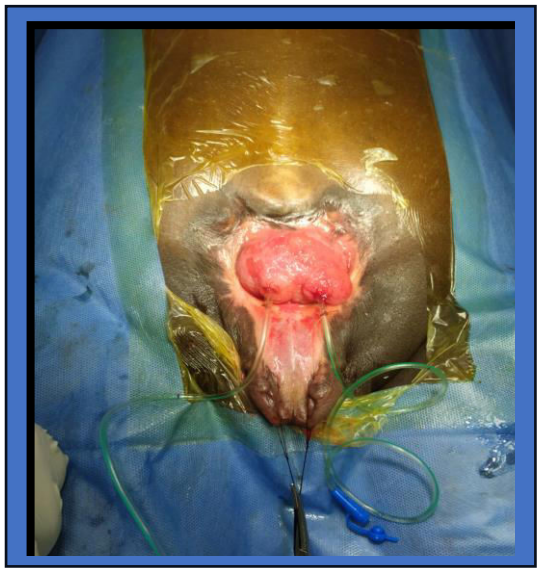

Figure1: Pre-operative picture of CBE

Figure2: Pre-operative picture of CBE

Figure 3: X-Ray pelvis of CBE

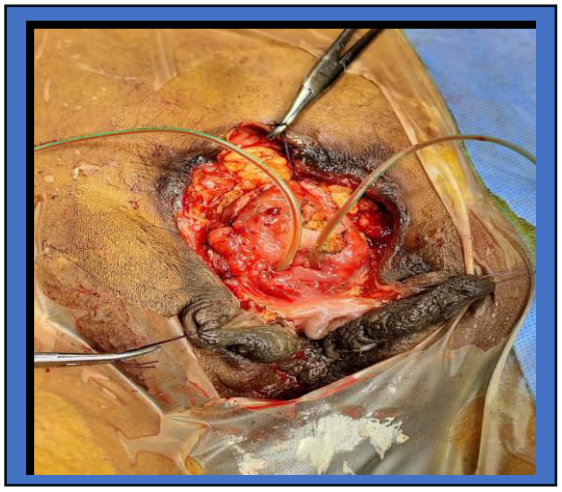

Figure 4: Picture of incision line

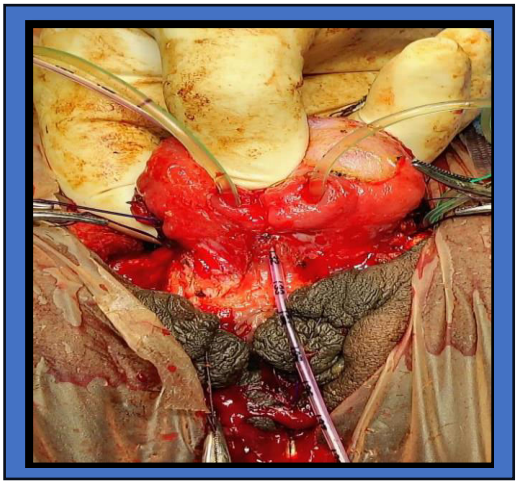

Figure 5: Bladder templet

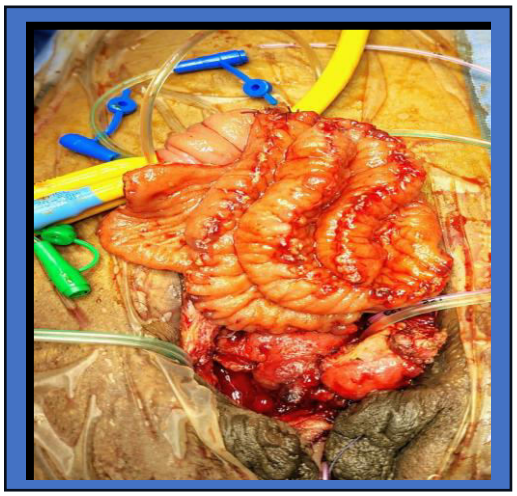

Figure 6: part of small gut for augmentation

Figure 7: augmentation cystoplasty

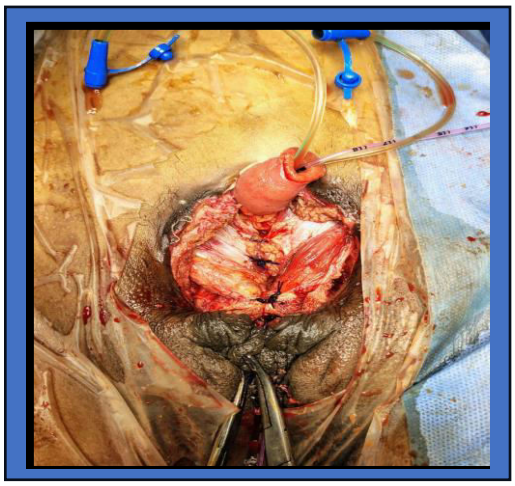

Figure 8: midline approximation RA

Figure 9: post op picture in female

Figure 10: follow-up picture in female

Figure 11: post-op picture in male

Figure 12: follow up picture in male

Discussion

Bladder exstrophy is a rare, but severe, life-altering congenital deformity of the genitourinary tract. The primary goals of treatment of classical bladder exstrophy are preservation of renal function, continence, cosmetically acceptable genitalia and ability to sexually cohabitate and reproduce. Initial management of the exstrophy-epispadias complex is usually started at birth with an assessment of the general condition of the neonate. This early closure also provides better pelvic approximation due to greater pliability of the neonatal pelvic bones. Adult bladder exstrophy is extremely rare in developed countries as most of the repairs are carried out during childhood. However, because of the factors such as poor awareness, limited healthcare access, social embarrassment and poverty, patients may carry their condition up to adolescence or adulthood.

The two most widely used approaches in the definitive management of bladder exstrophy-epispadias complex are the CPREand the modern stage repair of bladder exstrophy MSRE [7]. Lack of long-term follow-up has been the major drawback in the assessment of superiority of one technique over the other. The current emphasis in the management of exstrophy bladder is on limiting the frequency and morbidity of surgical procedures. we have performed exstrophy and epispadias repairs in a single stage. The procedure [8] incorporates the three goals of staged reconstruction performed in a single-stage surgery: Bladder closure, epispadias repair and achievement of urinary continence. This technique can avoid hypospadias, but may result in reduced penile length and persistence of penoscrotal dissociation. We did superior pubic ramus transverse splitosteotomy distal to insertion of rectus abdominis and adductor longus in all cases resulted in easy midline tension-free approximation of rectus abdominis muscles.This can provide acceptable continence without the need for multiple complex and expensive surgeries.

Pathak et al.[9] presented their experience of vesical preservation in a small series of adult patients. They treated the patients using ileocystoplasty, bladder neck reconstruction and abdominal wall closure with flaps. According to Gearhart[10], using the bladder template has a dual advantage: firstly, the requirement of bowel segment is less and secondly, if ureteric re-implantation is required, the native bladder template is a better substrate than the bowel wall.

We did not excise the bladder template. Our patients underwent a single-stage procedure, and the bladder template was used for ileocystoplasty. As we use a bowel segment for bladder augmentation, normally a low-pressure system is generated. Also, asymptomatic reflux in adults is not really significant. The bladder capacity of the augmented bladder is higher than that of the non-augmented bladder. The most important factor for continence in this group of patients is the bladder capacity. Our patients are all continent as they have an average bladder capacity around 400 ml.

Possible malignancy in the preserved bladder plate is one concern. Early closure does not necessarily prevent malignancy at a later time; it takes a longer time to develop malignancy. Our patients are on regular follow up to detect any malignancy at an early

stage. Our study showed that the preservation of the bladder template, limited osteotomy, para-urethral tissue support to the bladder neck and abdominal closure are a possible method for the management of the exstrophy-epispadias complex in children and adults.

Reports about sexual function are controversial in the literature. Some authors report an almost normal sexual function after genital reconstruction, but psychological assistance is recommended to overcome anxieties about genital appearance and sexual performance. Female genitalia reconstruction shows acceptable cosmetic results in the majority of cases, whereas male dissatisfaction with penile length due to the anatomical shortness and broadness of the corpora clearly exists.

The limitations of our study are that the experience with one-stage reconstruction was relatively small. However, we do believe it has advantages over the traditional approaches to bladder exstrophy. Based in our experience, one--staged reconstruction is feasible at any age, even after previous failed procedures, reducing the surgical steps and facilitating closure of the structures. It enables concomitant abdominoplasty, with good cosmetic results. In our patients treated with primary repair, the need for bladder augmentation is significant, but complications are less frequent than in the staged procedures. Pelvic osteotomy has a role in the surgical management of the exstrophy-epispadias complex, as it decreases tension across the abdominal wall, reduces the pubic diastasis, and helps restore the pelvic ring and floor to the normal anatomical configuration.

Another advantage of our technique in comparison to Mitchell's one-stage reconstruction is that we do not perform complete penile disassembly, reducing risks of penile ischemia and loss. In our series, ischemic loss of the glans or corporal bodies was not observed.

Conclusions

Most patients with bladder exstrophy will require multiple operations to achieve continence and provide cosmetically acceptable and functional genitalia. Onestage reconstruction (CPRE) minimizes the number of surgical procedures required to achieve the treatment goals (urinary continence, normal genital cosmesis and preservation of upper urinary tract). A modified surgical technique yields encouraging results providing improved social and psychological conditions for most patients. Preservation of upper tracts with achievement of continence is the ultimate goal.

Ethical Committee Approval: It is a retrospective study and analysis of the patient's medical record and followed up of the patients, the authors did not seek for ethical committee clearance.

Conflict of Interest: The authors have no conflict of interest to declare.

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Authors' Contributions: Manas Sasmal, in association with Uday Sankar Chatterjee, Dawood Khan, and Tapan Kumar Mandal, conceptualized and designed the study, as well as wrote the manuscript. In addition, he conducted the clinical and experimental studies and analysed the data.

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