

Surgical Management of Extrophy-Epispadias Complex: Early Results from a Tertiary Level Hospital in Bangladesh

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Original Research Article

Abstract:

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The exstrophy-epispadias complex (EEC) presents with diverse clinical manifestations, affecting various bodily systems, including the urinary tract, pelvis, abdominal wall, genitalia, and spine. The prevalence is approximately 1 in 10,000 births, with a higher occurrence in males. This study aimed to assess the efficacy of surgical repair for EEC. The observational study analyzed medical records from the Department of Urology at Rangpur Medical College Hospital, a tertiary level facility in Bangladesh, covering the period from June 2018 to January 2021. Eight EEC cases were managed during this four-year duration. Following careful evaluation and pre-anaesthetic assessments, all patients underwent surgical reconstruction of the urinary bladder and urethra. In suspicious cases, biopsies from the bladder wall were taken. The results showed that all patients successfully underwent reconstruction procedures. Biopsies were obtained in three patients due to suspected abnormalities. One patient with bladder exstrophy experienced disruption of the anterior abdominal wall, requiring later secondary closure. Three patients developed urethra cutaneous fistula, which were subsequently managed through closure procedures. The average follow-up period was 2 years, but one patient was lost to follow-up. The suggested modified surgical approach demonstrated favorable outcomes, leading to substantial improvements in patients' social and psychological well-being. Despite the rarity of EEC, the surgical repair proved to be effective in addressing the complexities associated with the condition.

Key Words: EEC, EC, Pelvic Osteotomy.

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INTRODUCTION

Exstrophy-epispadias complex is an uncommon congenital abnormality that can present in various ways. It's among the trickiest things that pediatric surgeons and urologists have to deal with. The estimated occurrence of this condition is between 1 in 10,000 and 1 in 50,000 live births, yet, it has a major impact on patients and their family's physical, functional, social, sexual, and psychological well-being [1].

Patients often present to the urologist as individuals, especially in the Indian subcontinent, even though this is a congenital abnormality with extremely visible deformities. This is attributable to several societal problems, including ignorance, poverty, and a lack of resources [2]. Initiating reconstructive treatment for exstrophy-epispadias complex as soon as possible after delivery has been shown to improve outcomes. The major goal is to achieve a full epispadias by converting the bladder exstrophy. The resulting incontinence, characterized by a moderate posterior outlet resistance, is beneficial since it both protects the renal and stimulates bladder development [3].

As a result, epispadias correction is often done when the patient is between four and six months old. A child's bladder neck is often repaired between the ages of four and five. Having reached this point, the child's bladder capacity is sufficient, and more significantly, they are ready to begin a post-operative voiding regimen. Repair techniques such as the Erlangen, Kelly, and complete primary repair of exstrophy (CPRE) have been advocated to restore bladder function without requiring a bladder neck repair to achieve continence. However, recent studies have shown that this is not the case for most patients [4,5] Exstrophic bladders have a greater risk of cancer if left untreated over extended periods because of their vulnerability to environmental factors [6]. There is a shortage of data on how often bladder exstrophy manifests in children and adults. We present a series of eight patients, including children and adolescents, who were diagnosed with and successfully treated for bladder exstrophy-epispadias complex at our facility [7].

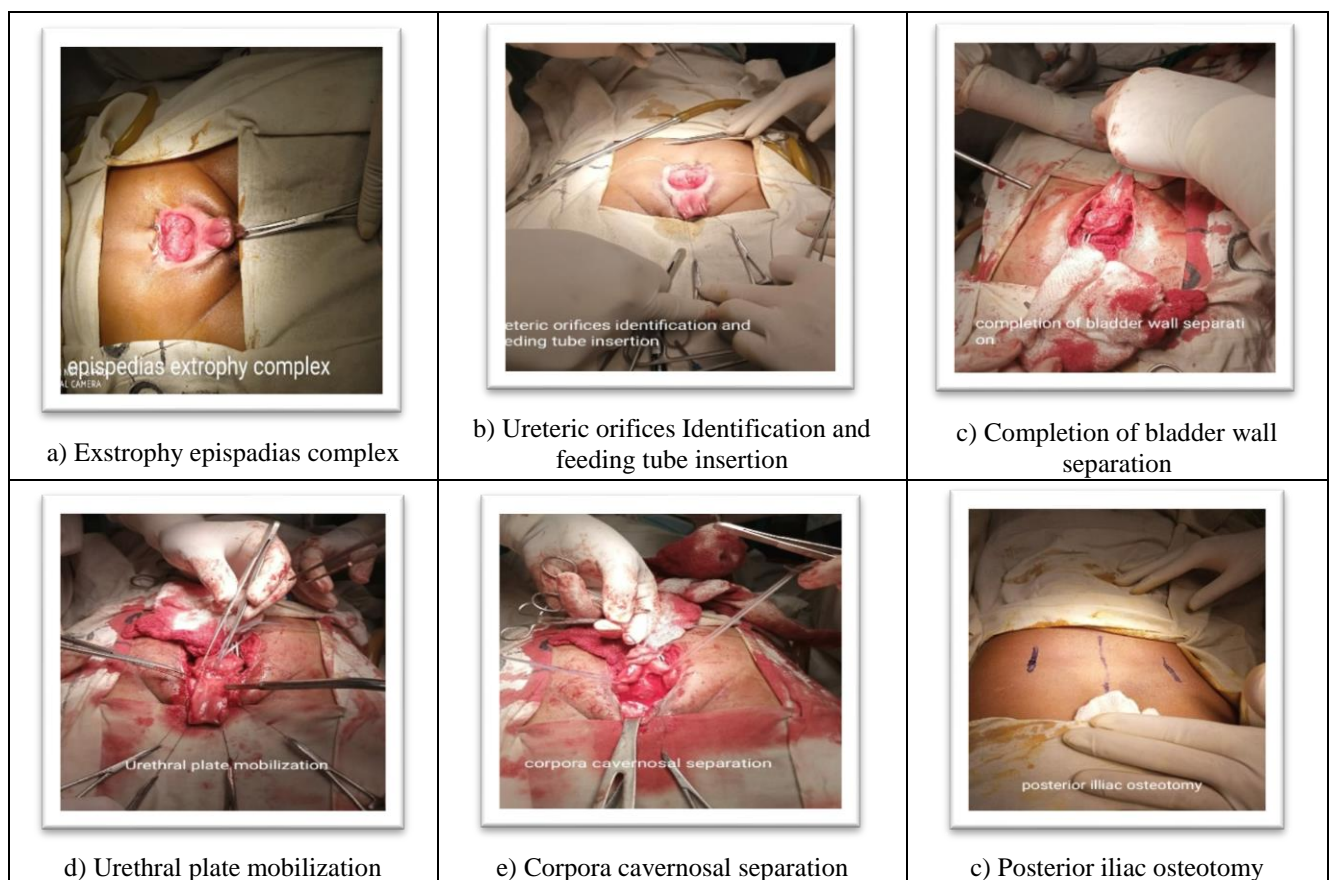
MATERIALS AND METHODS

This retrospective study reviewed medical records of 8 male patients with exstrophy-epispadias complex managed at Rangpur Medical College Hospital, Bangladesh, from June 2018 to January 2021. Patients aged below 5 years were included. The surgical approach involved bladder mobilization, urethral reconstruction, and hip spica immobilization. Suprapubic catheterization was performed in all cases. Biopsies were taken from the bladder plate before surgery. Complete repair was performed in seven patients, using Ile cystoplasty, para-urethral tissue reconstruction, and modified Cantwell-Ransley method for epispadias repair. After surgery, patients practiced clean intermittent self-catheterization. Regular follow-ups, urine analysis, and interviews assessed progress and social well-being. We analyzed the patients' urine at regular intervals and performed abdominal ultrasounds to monitor their progress. As for their mental and social health, as well as their desire, they were also interviewed [8].

RESULTS

There has been a mean of three years between visits with these individuals. Abdominal ultrasonography and post-void residual urine analysis are examples of follow-up tests. For the first six months, the patients took preventive antibiotics. After that, antibiotics have only been continued in patients symptomatic for urinary tract infection (UTI). A micturating cystourethrogram (MCU) was not routinely performed. All our patients developed reflux after bladder closure. There may be an increased incidence of recurrent infection due to reflux, which can be treated with antibiotics.

During the first six months, patients were encouraged to engage consistently in CISC. Afterwards, the intestinal epithelium undergoes alterations that result in less mucus production. So, CISC is needed by patients less frequently in the long run. Except for one who did not have epispadias surgery, all patients can control their bladders. One patient who developed a urethral-cutaneous fistula at the base of the penis is awaiting repair, and both were managed by minimally invasive methods (one patient with percutaneous cystolitholapaxy) PCCL and the other with LASER cyst lithotripsy). The scar of the anterior abdominal wall is healthy in all patients. Four patients with a history of adequate fractionation complain about some amount of lateral chordee. All patients are doing well socially and psychologically.



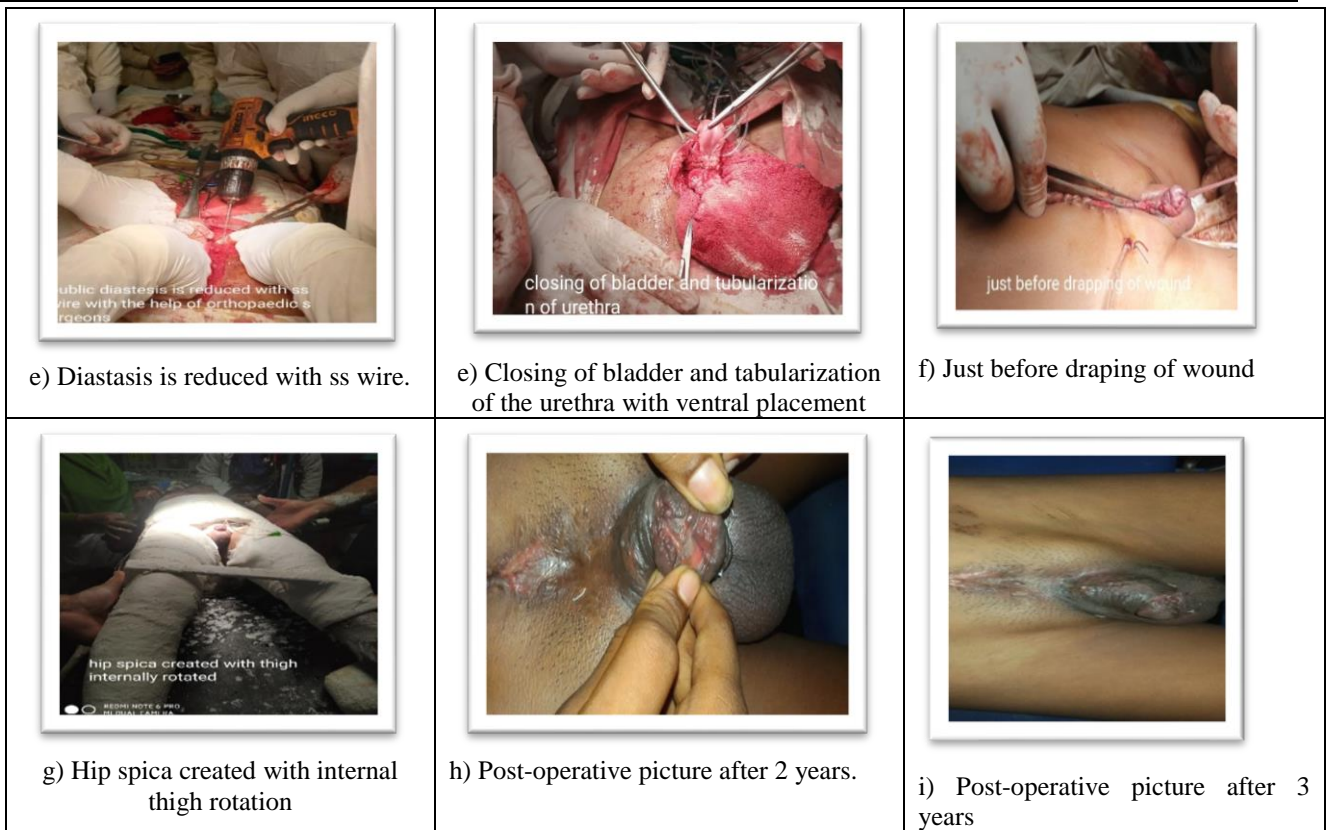


Figure 1: patients' complete repair of the exstrophy-epispadias complex

The recovery time for these surgeries can vary depending on the stage of the surgery and the patient's condition. It is generally a long process that requires close follow-up and monitoring by a team of specialists.

Table 1: Following bladder closure, upper urinary tract dilatation and continence

Upper tract	Dry >1 h	Incontinent	No data about dryness
Dilation	2	1	3
No Dilation	0	2	0
Total	2	3	3

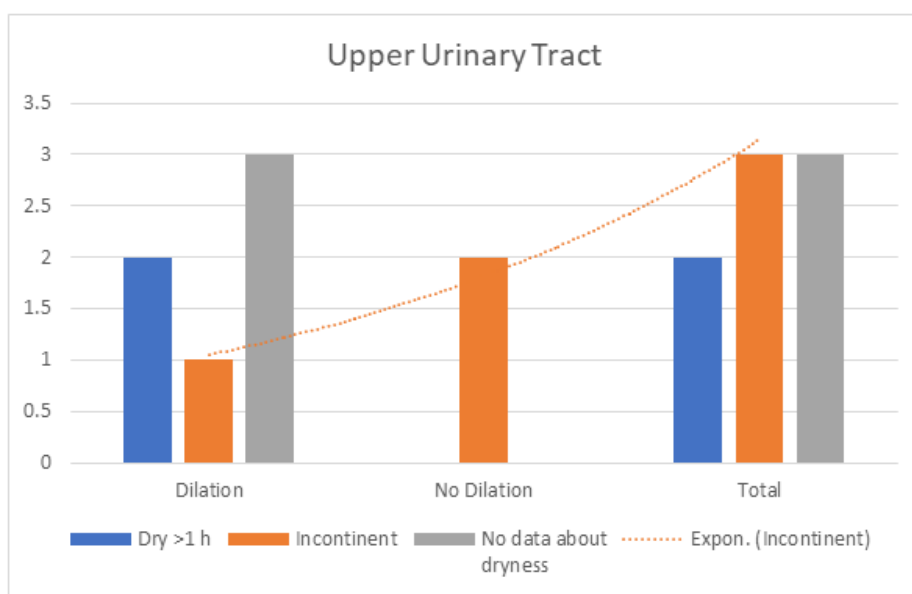


Figure 2: This can occur as a result of various conditions

Upper urinary tract dilation refers to the enlargement of the upper portion of the urinary tract, specifically the renal pelvis and ureters, such as obstruction, infection, or injury. Our early experiences regarding the reconstruction of EEC are satisfactory. However, more frequent procedures are needed for optimum outcomes.

DISCUSSION

In most cases, a newborn's overall health is evaluated as part of the first care for the exstrophy-epispadias complex. The idea of surgical repair goes beyond merely closing the urine bladder and urethra to address the underlying anatomical flaw of the condition. Osteotomy is one of the most important surgery steps for the future continent mechanism [9]. Nowadays, there are two ways of correction: a modern stage repair and a single-stage repair [10]. However, there is no head-to-head comparison of the results of these two procedures. The basic steps are osteotomy, closure of the urinary bladder, bladder neck reconstruction, epispadias repair and secure abdominal closure [11].

All of these techniques have been tried and true in the pediatric population. However, there is a severe lack of information on the adult exstrophy-epispadias correction. Previously, the bladder template was excised, and any diversion was the preferred option. According to earlier reports, Bladder exstrophy was reliably and safely managed with bladder neck closure along with continent cutaneous diversion. This can provide acceptable continence without the need for multiple complex and expensive surgeries [9].

While osteotomy is mandatory in pediatric patients, a repair can be possible without osteotomy in adults. Mansour *et al.* showed that abdominal closure without osteotomy is possible when using the primitive bladder muscle plate and urinary diversion in a single stage. We performed a superior pubiotomy just lateral to the insertion of the rectus muscles to ease abdominal closure [12].

Pathak *et al.* presented their experience of vesical preservation in a small series of patients. They treated the patients using Ile cystoplasty, bladder neck reconstruction and abdominal wall closure with flaps. They also showed that abdominal closure could be possible without transposition flaps [13].

We didn't cut off an area for the bladder. Our patients had a one-step treatment in which the bladder served as a mould for ileal- cystoplasty. Antibiotic prophylaxis is necessary after exstrophy closure since 100% of patients will have reflux. A low-pressure system is usually created when we employ a tube segment for bladder augmentation. Also, asymptomatic reflux in adults is not significant. The bladder capacity of the augmented bladder is higher than that of the non-augmented bladder [14,15]. The most important factor for continence in this group of patients is bladder capacity.

CONCLUSION

The combination of exstrophy and epispadias is extremely rare. Results from a modified surgical procedure are promising, with most patients seeing considerable improvements in their social and psychological well-being. Our early experiences regarding the reconstruction of EEC are satisfactory. However, more frequent procedures are needed for optimum outcomes.

Conflict of interest: None

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