**Introduction**

Age old philosophy of exstrophy bladder repair is to make patients continent as well as to preserve renal functions. Goal of treatment is to make patients continent via naturalis or by clean intermittent catheterization (CIC) to offer a productive life with normal body image. Literature has witnessed great endeavors with different surgical procedures in making exstrophy bladder continent and to avoid diversion.[1]

The purpose of this retrospective study is to evaluate “critical factors” those were responsible in achieving urinary continence in our patients. We analyzed files of 18 patients to explore the rationale behind their achieving of continence.

**Patients and Methods**

Since 2004 till 2019, we operated on 52 patients. Out of 52 patients, we selected files of 18, age ranged from 5 months to 14 years. As per records, these 18 patients were treated in between 2011 and 2019. Institute ethical board approved and institute ethical committee (IEC) consent taken. Patients were divided into two groups.

- **Group A**
  - Eleven out of 18 patients were presented from beginning and 7 were referred after around 8 to 14 years, as incontinent bladder following good repair of bladder neck and posterior urethra. Eleven were operated with complete primary repair of exstrophy along with pubic osteotomy minimal and were kept on cystostomy track (CT) till augmentation to vent out vesical pressure. In seven patients, we did reduction of caliber of posterior urethra and bladder neck along with CT followed by augmentation after 6 months.

- **Group B**
  - All 18 patients are maintaining dry period for 24 h. Two patients had enuresis but are manageable with partial fluid restriction from evening. Seventeen out of 18 patients are maintaining their renal functions.

**Conclusion**

No tension abdominal wall closure with rectus muscle apposition is essential to preserve repaired bladder exstrophy. Osteotomy prevents lateral drag to overcome failure of whole reconstruction. Increased “systolic” vesical pressure from contraction of small bladder might destroy the mechanism of continence and renal functions. Hence, venting of vesical pressure through CT is obligatory till augmentation which is of necessity to be done as early as possible to create a low-pressure continent system.

**Keywords:** Augmentation, continence, cystostomy, exstrophy bladder, posterior urethra, pubic osteotomy
committee approved file study. All 18 patients had homogeneity in the philosophy of treatment offered. They were fully continent, all were communicable, had necessary investigations, and had minimum 2 years of follow-up [Flow Chart 1]. Eleven patients out of 18 were presented without prior intervention (Group A) and rest 7 were referred (Group B) after 8–14 years as incontinent bladder following good repair of bladder and epispadias. The incidence of exstrophy bladder is very low. That is why we got scope to study only on 18 patients in this series. Probably for the same reason, other authors[2–5] reported similar number of patients of exstrophy bladder in their article.

All 18 patients got high bowel wash for 2 days before operative procedures. Patients of Group A were operated with complete primary repair of exstrophy (CPRE) as per Grady and Mitchell[2] along with pubic osteotomy minimal (POM). In POM procedure,[3] insertions of rectus abdominis (RA) and adductor longus (AL) muscles were identified and osteotomy was done at the lateral border of the insertions of RA and AL on both sides. Medial half of bodies of pubic bones attached with RA and AL (like patella of knee joint) were separated from lateral halves. Two medial halves of pubic bones were then pulled in midline and were tied with thick Prolene suture to reconstruct “neo-symphysis” over reconstructed bladder neck [Figure 1]. All patients of reconstructed bladder of Group A were kept on cystostomy track (CT) to vent out bladder contraction (“systolic”) pressure. For reconstruction of CT, we marked a tongue-shaped skin attached with the cephalad margin of exstrophy bladder [Figure 2a] to get a 15 mm wide skin flap [Figure 2b] from anterior abdominal wall to reconstruct a tube named as CT. Reconstructed CT took the shape of tube and looked like Mitrofanoff’s channel [Figure 2c] and got mature [Figure 2d] with Foley’s catheter. Afterward, CIC was advised through CT to keep the track patent and to vent high “systolic” pressure from small bladder till augmentation which was done after 6 months.

Patients of Group B were earlier operated either with iliac osteotomies or rectus muscle flap[4] for closure of abdominal wall [Table 1]. All had good abdominal, bladder, and urethral closure but lost coaptation of posterior urethra and bladder neck on clinical and endoscopic examinations. One of seven patients of Group B, aged 14 years, presented with bilateral dilated refluxing ureters, bladder capacity 150 ml, and creatinine 1.5 along with incontinence. We augmented bladder with left megaureter of 8 cm in length following transureteroureterostomy and did reduction of caliber of posterior urethra and neck. In another six patients of Group B, reduction of caliber of posterior urethra and bladder neck was done along with CT to vent out bladder contraction pressure. Similarly, augmentations were done after 6 months in five patients of Group B. Some patients, particularly female [Figure 2a], had wider urethral plate. As well as in some patients of Group B had diluted urethra due to effacement. We excised excess urethral mucosa from lateral margins of urethral plate to get 6–10 f urethral tube (as per age) for better coaptation of urethral sphincters.

We regularly checked albumin-to-creatinine ratio (ACR) to ascertain whether kidneys were on strain or not.
Similarly, regular creatinine clearance (CCr) was checked 3 monthly to judge the status of chronic kidney disease (CKD) on follow-up. Urodynamic study (UDS) was done particularly in those patients who used to void via naturalis.

**Results**

Our study period was ~ 8 years similar to other authors.\(^2\)\(^-\)\(^5\) During this period of study, we found that all 18 patients have got adequate bladder capacity as per body weight to maintain convenient dry period for 24 h like normal individual. One patient, now on CIC from Group B, presented at 14 years of age; achieved voiding capacity to 380 ml from 150 ml following ureteral augmentation. However, creatinine increased from 1.5 (glomerular filtration rate [GFR]: 44, CKD stage 3) to 2.1 (GFR: 34, CKD stage 3) after 11 years, at the age of 25 years.

All other patients maintained their normal ACR and CCr at follow-up. Two patients had enuresis and they were managed with partial fluid restriction from evening [Table 1]. One patient having higher Pdet.Qmax in UDS was advised for CIC and urethral dilation. After 16 months, UDS parameters became acceptable.

**Discussion**

Trendelenburg emphasized on approximation of pubic bones\(^6\) possibly to approximate RA muscles to eliminate lateral drag on repaired abdominal wall as well as to hold the soft tissues around reconstructed bladder neck and urethra. We also experienced disruption of CPRE following disruption abdominal wall closure in our previous patients. Hence, any type of osteotomy or even minimal osteotomy\(^3\) is necessary along with CPRE\(^2\) to stay away from lateral drag and to keep repaired abdominal wall, bladder neck, and posterior urethra intact. Following successful repair of bladder neck and posterior urethra, another problem pops up. That is harmful high “systolic” pressure from contraction of very small bladder. As per Laplace’s principle, pressure in any cavity following contraction is inversely proportional to its volume. That translates; “systolic” pressure of small bladder will exert more pressure compared to full-size bladder. Hence, following reconstruction, small bladder with increased “systolic” pressure for longer duration might widen repaired bladder neck and posterior urethra (similar to “effacement” of cervix following contraction uterus) resulting in failure of continence mechanism. Similarly, “systolic” pressure might also set off vesicoureteric reflux and might damage kidney functions. Hence, we maintained CT [Figure 2c] to vent out “systolic” pressure of bladder contractions, and without much delay, we created capacious bladder by augmentation to avoid those injurious effects of high pressure originating from “systole” of small thick bladder. We did not expect required growth from subnormal bladder\(^7\) to achieve near normal capacity. In this series, only 1 patient out of 18 and 3 out of 52 patients had near normal bladder capacity as per age. Hence, we detain their bladder augmentation, which might be done if necessary. ACR and CCr were checked at follow-up, to monitor renal strain and renal functions, respectively. We checked CCr/GFR, not serum creatinine alone, to monitor status of CKD.

Urethral mucosal coaptation, i.e., passive closure of the urethral lumen at rest, is considered to be an important component of urinary continence. Surrounding muscles

| Number of patients | Remarks |
|--------------------|---------|
| Patients in Group A | Primary presentation |
| Patients in Group B | Secondary; after primary repair |
| Male | |
| Female | |
| POM done in | |
| Patients in Group B: Prior oper. with muscle flap closure | |
| Patients in Group B: Prior oper. with iliococcygeal osteotomy | |
| Augmentation with ileum | Sufficient length (~20 cm) is not always available |
| Augmentation with sigmoid done in | We got ~8 cm of dilated ureter |
| Augmentation with ureter (patient with stage 3 CKD) | |
| No augmentation | He was counseled for future augmentation if necessary |
| Narrowing of wide posterior urethra and neck | |
| Dry period 24 h | |
| Bed wet on excessive fluid intake | |
| Natural void | |
| CIC + natural void | They are on dilation to achieve normal void |
| Only CIC | Pondering on necessity of limited, repeat bladder neck incision to make them natural voider |

POM: Pubic osteotomy minimal, CKD: Chronic kidney disease, CIC: Clean intermittent catheterization
and tissue form a cushion-like support and can improve urethral coaptation and hence restore continence. Embryologically, posterior urethra in males and whole of urethra in females are of the same origin and their sphincters are responsible for urinary continence. Bladder neck sphincter in male is intended to be for the continence of seminal fluid, not for urinary continence. On the other hand, hypertrophy and spasm of bladder neck set off resistance to urinary outflow. Similarly, some authors mention that the various techniques of bladder neck reconstruction (BNR) have been described in literature to create “urinary outflow resistance.” Hence, it appears BNR actually is an “iatrogenic resistance.” It might set off urinary outflow hindrance or obstruction. Nevertheless, this “iatrogenic obstruction” following BNR helps in CIC. On the other hand, BNR might increase leak point pressure and might preclude natural void due to “rafted resistance” or “iatrogenic obstruction.” Accordingly, we ceased to do BNR and started reduction of wide posterior urethral and wide bladder neck caliber for better coaptation and expected natural void [Table 1]. In 11 patients, we did CPRE, POM, and CT in 1st stage followed by augmentation in 2nd stage after 6 months. CT vents out “systolic” pressure of bladder and preserved bladder neck and posterior urethral caliber, and prevented renal injury till augmentation. That is why we kept CT to vent vesical “systolic” pressure till augmentation of bladder. We did ureterocystoplasty in one patient from Group B [Table 1] to avoid metabolic complications. By tradition, complete continence has been defined as the dry period, i.e., ability to retain urine for more than 3 h without leakage. Traditional “dry period” in published literature has not disclosed amount of fluid intake and bladder capacity following repair. In our series, all patients had normal bladder capacity and fluid intake was ad libitum. They used to sense fullness of bladder to carry out CIC or natural void. Hence, they enjoy “dry period” for 24 h. However, the frequency of urination varied depending on intake of fluid and “voiding capacity of bladder,” similar to normal individual.

**CONCLUSION**

As per evaluation of “critical factors” in this retrospective study, we have found that abdominal wall closure with no tension in rectus muscle apposition is essential to preserve CPRE. In this respect, POM procedure seems easier to perform as well as effective.

Preservation of coaptation of bladder neck and posterior urethra following CPRE is mandatory for continence to achieve and it is possible with CT [Figure 2]. Otherwise, effacement of bladder neck and dilation posterior urethral caliber might take place following increased, longstanding “systolic” pressure from bladder contraction of small bladder as per Laplace’s principle. Harmful “systolic” pressure might wipe out continence mechanism and might deteriorate renal functions. Hence, venting of vesical “systolic” pressure is necessary and we put forward CT till augmentation. Augmentation increases bladder volume to create a low pressure system as per Laplace’s principle. Hence, we suggest augmentation to be done as early as possible to conserve continence and renal functions.

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**Conflicts of interest**

There are no conflicts of interest.

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