

Original Research Article

The impact of classic bladder exstrophy-epispadias repair in adult patients without continence

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ABSTRACT

Background: Bladder exstrophy-epispadias complex is a rare congenital anomaly. Very few patients grow with the condition intact in their adulthood which lead to compromise in the results of repair. We herein report our experience of correction of the defects in the adult patients in single stage without doing osteotomy and urinary diversion.

Methods: The sample size was 13 patients over ten years. All were treated in single stage. Bladder neck repair was done using paraurethral tissue, primary abdominal closure was done without flap, epispadias repair was done using modified Cantwell-Ransley procedure and hernial repair was done without mesh. No osteotomy was done. All the patients were followed regularly. Minimum period of follow-up was one year. Urinary continence, change in habits and subjective assessment of well-being were also noted.

Results: 86.4% patients were totally incontinent. Bladder and anterior abdominal wall were closed in all the patients. Complete epispadias was possible in 15.3% patients and rest were converted into coronal hypospadias. Hernia was repaired in 23% patients. All the patients were satisfied with their cosmetic results and they were able to interact socially.

Conclusions: Surgical correction of the exstrophy-epispadias complex is possible in adulthood in the hands of experts in single stage without osteotomy. Regular cystoscopic examination and urine cytology yearly are recommended to diagnose the malignant changes in the native bladder as early as possible. Psychological, social and self-level of satisfaction is high among these patients after the surgical correction.

Keywords: Adult bladder exstrophy, Exstrophy-epispadias complex, Epispadias repair without osteotomy

INTRODUCTION

Bladder exstrophy-epispadias complex is a rare congenital anomaly. It comprises of an absent anterior abdominal wall leading to exposed posterior bladder wall to the environment, the divergent rectus muscles going on either side of the bladder and attached to the pubic rami on either side, which are also divergent evident as pubic diastasis. In males, the penis is short and epispadiac with dorsal plate and splayed glans. In females, the mons pubis, both clitoris and the labia are widely separated, the vaginal orifice is anteriorly displaced. Inguinal hernias are also common, the reason being the wide internal and

external rings and absence of obliquity of the inguinal canal. The incidence of this entity ranges from one per thirty thousand to fifty thousand. The male to female ratio ranges from 1.5 to 5:1.¹⁻⁴ Nelson et al in their study reported that males and females were equally affected by exstrophy.⁵ The male to female ratio did not differ by race, region, insurance, or status. Usually, the correction of the abnormalities is done in the childhood usually because the anxious parents rush to their physician for the correction of the anomaly. Presentation in adulthood is further rare considering the rarity of the anomaly itself.⁶ In spite of the obvious defects of this anomaly, few patients present in adulthood may be due to lack of

awareness, economic and social problems or lack of nearby healthcare facility.⁷ In the adulthood, the bladder capacity is reduced, bony fusion has already occurred, the gait is changed, the phallus also becomes short and thick and there are increased chances of malignancy in the exposed bladder. All these factors lead to compromise in the results as compared to when repair is done in the childhood. We herein present our experience of correction of the defects in the adult patients in single stage without doing osteotomy and urinary diversion. Renal parameters, ultrasonography and diagnostic cystoscopy were done at regular intervals.

METHODS

We conducted a retrospective study after obtaining the Institutional ethical committee permission and reviewed the operated adult male patients admitted in SMBT Institute of medical sciences and research centre, Nashik, Maharashtra, India from August 2010 to July 2020, with classic exstrophy-epispadias complex, where posterior wall of the bladder was exposed, there was complete epispadias with or without inguinal hernia. The study included 13 patients. All data were collected in retrospective manner. For statistical analysis, SPSS 19.0 software was used. The satisfaction with life scale was used to measure the level of satisfaction of patient.⁸

At presentation in our hospital, 100% patients were male and were in their 20's. They did not receive any treatment in the past. The age range was 20 to 29 years. They were advised blood investigations in the form of complete blood count, renal function tests and ultrasound of the kidney, ureter and bladder region and X-ray of the pelvis. All patients were informed about the details of the operative procedure. They were counselled about attainment of continence and the need for continuous catheterisation or clean intermittent self-catheterization at regular intervals. They were also explained about the possible complications of the planned procedure.

Prior to the planned procedure, all patients underwent random biopsy from the exposed bladder to rule out malignancy or premalignant changes. Multiple random bladder biopsies were done and suggested nonspecific inflammatory changes. The surgical technique includes the technique of component separation used for incisional hernia (Figure 2). After separation of the components, the bladder was closed with absorbable sutures in two layers, the sheath was closed with non-absorbable sutures and skin with non-absorbable sutures. None of the patients underwent cystectomy. All patients were treated in single stage. Bladder neck repair was done using paraurethral tissue, primary abdominal closure was done without flap, epispadias repair was done using modified Cantwell-Ransley procedure and hernial repair was done without mesh (Figure 1-5). No osteotomy was done. They were followed after 1 week, 3-week, 1 month, 3 months, 6 months, one year and then yearly. Minimum follow-up period was one year in the study. All complications were

noted. Urinary continence, change in habits and subjective assessment of well-being were also noted. For surveillance of bladder malignancy, urine cytology was done at 6 months interval and cystoscopy was done at one year follow-up.



Figure 1: Exstrophy epispadias complex.



Figure 2: technique of bladder and abdominal wall closure.

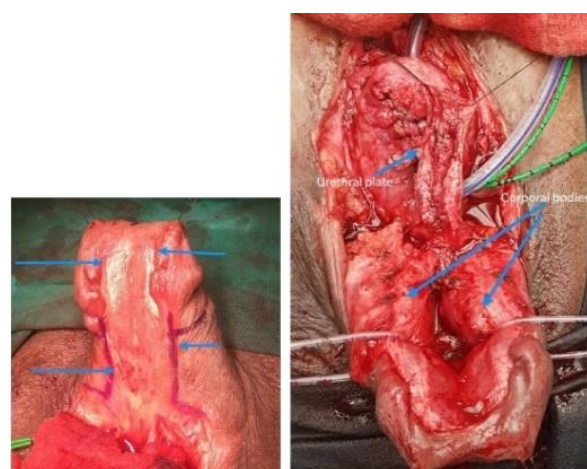


Figure 3: Urethral plate marking and penile disassembly.

RESULTS

The mean age of the patients in the study was 24.5 years (range 20-29 years) (Table 1). All patients were incontinent till the last follow up. 15.3% patients were using self-intermittent catheterisation and rest were on continuous per urethral catheter. 15.3% patient developed partial continence and had stress urinary incontinence for which they were kept on regular pelvic floor muscle therapy. The renal parameters of all the patients were normal till the last follow up. Diagnostic cystoscopy and bladder biopsy were done in all patients and all revealed normal bladder mucosal findings. Bladder and anterior abdominal wall were closed in all the patients but in 86.4% patients epispadias was converted into coronal hypospadias and in 15.3% patients complete epispadias repair was possible. Hernia present in 23% cases was repaired. All the patients were satisfied with their cosmetic results, whether they were on continuous or intermittent catheterisation. They were able to interact socially. Infact, they were able to move alone and wear clothes properly. The self-esteem, confidence and the satisfaction levels were high. Complications were minimal and only 7.7% patients had mild superficial wound infection which was managed accordingly (Table 2).

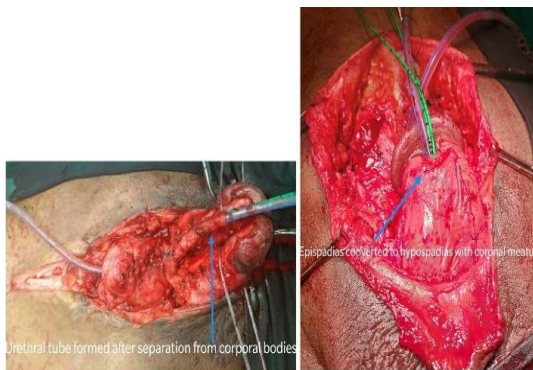


Figure 4: Formation of urethral tube separately and converting it into hypospadias.



Figure 5: Final look after complete repair with normal and hypospadiac meatus.

Table 1: Demographic features.

Parameters	Observations
Total number of patients	13
Mean age of patient (years)	24.5
Education level (%)	Low (100)
Socio-economic status N (%)	Lower class 12 (92.3) Middle class 1 (7.6)
Family status N (%)	Combined 10 (76.9) Nuclear 3 (23)

Table 2: Operative findings, surgical results and complications.

Parameters	N	%
Number of incontinent patients	11	86.4
Number of partial continence patients	2	15.3
Number of patients with stress urinary incontinence	2	15.3
Modified Cantwell-Ransley procedure	13	100
Meatus at tip of the penis after epispadias repair	2	15.3
Hypospadiac meatus after epispadias repair	11	86.4
Number of patients with concomitant hernia	3	23
Number of patients on continuous per urethral catheter with intermittent clamping	11	86.4
Number of patients on clean intermittent catheterisation	2	15.3
Superficial wound infection	1	7.7

DISCUSSION

The management of exstrophy-epispadias complex starts in the early childhood. The anxious parents bring their child/children to the paediatric surgeon. Very few patients are spared of the repair in their childhood. It could be due to unavailability of the health facility or the surgeon, or the poor financial condition or the different social problems. Whatever the condition, these patients reach the urologists very late. The treatment of this condition in adulthood is a bit difficult. Reason being the fused bones and their epiphyses, the altered gait and so on. The basic steps of exstrophy-epispadias repair are: osteotomy, urinary bladder closure, bladder neck reconstruction for continence, epispadias closure for proper sexual needs and closure of the abdominal wall. All these steps remain same in adults except the osteotomy which becomes very difficult in adulthood. The repair is done in stages in children but in adults it can be done in a single stage. There are many reports of single stage repair in adults.^{9,10} We also treated our all patients in a single stage repair.

In the older times these patients were managed by either urinary diversion or excision of the residual bladder to avoid the risk of malignancy or a staged repair. Perlin et

al treated adult exstrophy-epispadias patient with cystectomy and formation of ileal continent reservoir. Mensah et al used continent cutaneous diversion for bladder exstrophy patients in adults.¹¹ Pathak et al treated 3 patients of exstrophy-epispadias complex in two stages.¹² But now a days, repair can be done in single stage. This could be due to high patients' expectations or increased expertise of the urologists. Mansour et al treated adult exstrophy-epispadias complex patients in single stage.⁹ Pathak et al treated one such patient in one stage. We also did single stage repair in all the patients.¹²

When repair is done in childhood, osteotomy is must. In adults, the repair of the exstrophy-epispadias complex can be done in single stage and osteotomy is not compulsory. Mansour et al in all his patients did single stage repair without osteotomy while Jana et al did single stage repair but used superior pubotomy for complete closure of the defect. Shoukary et al in their study also did not do osteotomy in any of their patients.^{9,10,13} In our all cases we did not do any osteotomy and closure was possible in single stage. Continence is of utmost importance in exstrophy-epispadias complex patients. Mensah et al achieved total continence by doing bladder neck closure and continence cutaneous diversion.⁷ Jana et al had 100% continence in their patients after single stage repair.¹⁰ Our study also showed only partial continence in 15.3% patients only with the presence of stress incontinence. These patients were on regular pelvic muscle floor therapy and improved further.¹³

As there are increased chances of malignancy in the native bladder, the patients are kept under surveillance by doing cystoscopic examination at regular interval. Smeulders and Woodhouse in 2001 showed that there is 700-fold increase in the chances of malignancy of bladder in exstrophy patients, inspite of the fact that primary surgery was done in early age and whether the bladder was closed primarily or diversion was done.¹⁴ Therefore, it is recommended to do regular cystoscopic examination of the bladder to identify and diagnose the malignant changes as early as possible. In our study we did urine cytology six monthly and cystoscopic examination on yearly basis to rule out the malignancy.

Post-operatively after the repair is done and patients are discharged, all the patients showed high level of satisfaction with respect to their body image, social interaction and the urinary control. Infact, they were able to move alone and wear clothes properly. They started going in social gatherings. Diaper usage was almost stopped in all the patients. Their overall satisfaction level was high. Similar level of satisfaction was shown by Mansour et al, Mensah et al and Nerli R B et al.^{7,9,15} This is one of its kind of study where satisfaction of the patients was high inspite of the fact that they were incontinent and were on continuous or intermittent catheterisation.

Limitations

Limitations of current study were the retrospective nature and smaller study cohort. Further randomised controlled trials or studies with larger number of patients will add further to future research. Still our study adds at least a pinch to the larger sea.

CONCLUSION

Bladder exstrophy-epispadias complex is a rare congenital defect. Few patients become adult with the defect persisting. Surgical correction of the deformity is possible in adulthood in the hands of experts in single stage with or without osteotomy. Regular cystoscopic examination and urine cytology yearly are recommended to diagnose the malignant changes in the native bladder as early as possible. Psychological, social and self-level of satisfaction is high among these patients after the surgical correction.

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