

PRELIMINARY RESULTS OF DELAYED PRIMARY BLADDER EXSTROPHY RECONSTRUCTION IN FEMALE PATIENT: A CASE REPORT

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ABSTRACT

Objective: Reporting a rare case and provide a view of bladder exstrophy management. **Case(s) Presentation:** A 12-year-old female complained of an open abdominal wall below the umbilicus accompanied by continuous incontinence of urine since birth. The MRI showed diastasis symphysis 5.1 cm and a soft tissue defect in the suprapubic region, suspicious of bladder exstrophy. Bladder mucosa biopsy showed squamous metaplasia. The patient underwent pelvic osteotomy, bladder wall closure, bladder neck reconstruction, abdominal closure with double keystone flap with proceed mesh, and external fixation. A cystostomy catheter was placed until one week after surgery, and the patient was able to hold the urge to urinate and started to move around while sitting. Two months after the reconstruction, the primary closure and keystone removal on skin defect were carried. The patient was already able to walk and continence the urine. **Discussion:** Bladder exstrophy is a rare condition. Currently delayed reconstruction on bladder growth is the subject of debate and speculation. The patient's condition in this presented case improved, this is in accordance with the literature that states that delayed reconstruction can still be considered in special conditions that do not allow early reconstruction. **Conclusion:** Delayed bladder reconstruction and bladder neck reconstruction can improve the anatomy of the bladder and quality of life on certain cases.

Keywords: Bladder exstrophy, bladder reconstruction, female bladder exstrophy.

ABSTRAK

Tujuan: Melaporkan kasus langka dan memberikan pandangan baru mengenai manajemen ekstrofi kandung kemih. **Presentasi kasus:** Seorang anak perempuan berusia 12 tahun mengeluhkan dinding perut terbuka di bawah umbilikus disertai inkontinensia urin terus menerus sejak lahir. MRI menunjukkan diastasis simfisis 5.1 cm dan defek jaringan lunak di daerah suprapubik, yang mengarah kepada ekstrofi kandung kemih. Biopsi mukosa kandung kemih menunjukkan metaplasia skuamosa. Pasien menjalani osteotomi panggul, penutupan dinding kandung kemih, rekonstruksi leher kandung kemih, penutupan perut dengan double keystone flap dengan prosee mesh, dan fiksasi eksternal. Kateter sistostomi dipasang hingga satu minggu setelah operasi, dan pasien mampu menahan keinginan untuk buang air kecil dan mulai bergerak sambil duduk. Dua bulan setelah rekonstruksi, dilakukan penutupan primer dan pengangkatan keystone pada defek kulit. Pasien sudah bisa berjalan dan buang air kecil. **Diskusi:** Ekstrofi kandung kemih merupakan kondisi yang jarang terjadi. Rekonstruksi pertumbuhan kandung kemih yang tertunda saat ini masih menjadi bahan perdebatan dan bahan spekulasi. Kondisi pasien pada kasus yang disajikan ini membaik, hal ini sesuai dengan literatur yang menyatakan bahwa rekonstruksi tertunda masih dapat dipertimbangkan pada kondisi khusus yang tidak memungkinkan dilakukannya rekonstruksi dini. **Simpulan:** Rekonstruksi kandung kemih yang tertunda dan rekonstruksi leher kandung kemih dapat memperbaiki anatomi kandung kemih dan kualitas hidup pada kasus tertentu.

Kata kunci: Ekstrofi kandung kemih, rekonstruksi kandung kemih, ekstrofi kandung kemih wanita.

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INTRODUCTION

Bladder exstrophy (BE) is a rare congenital disorder with important repercussions in the patient's life. Cross-disciplinary postnatal management is required to successfully close the bladder, as this has

an impact on the capacity and ability of future continence. Therefore, the goal of treatment is urinary continence and adequate sexual function, without causing kidney damage.¹ BE is a rare pathology (1/10.000-50.000 live births). It is more common in male patients and has complex

treatment. Long-term results are not entirely satisfactory regardless of the technique used.¹

Gradual repair (Gearhart-Jeffs), complete primary closure (Mitchell), and total mobilization (Kelly) have become the most widely used techniques in the modern era of bladder exstrophy treatment. However, although the morbidity of patients has decreased, urinary continence has not yet reached the level of success initially expected. 70% of patients achieve urinary continence after various surgical procedures. Therefore, the treatment of bladder exstrophy remains a challenge for pediatric urologists.¹

The purpose of this case report is to report the rare cases and provide an outlook on the management of bladder exstrophy.¹

CASE(S) PRESENTATION

A 12-year-old female patient came with the chief complaint of the opened abdominal wall below the navel, accompanied by the discharge of urine dripping continuously since birth. The patient has a history of spontaneous birth at 32-33 gestational weeks with meconal amniotic and hasn't menstruated until now. A history of similar abnormalities in the family is denied.

From physical examination in suprapubic region showed a fixed mass with size of 10 cm x 7 cm, appears external meatus urethra. Her renal function within normal limits. Pelvic X-Ray showed groundglass appearance on the projection of the

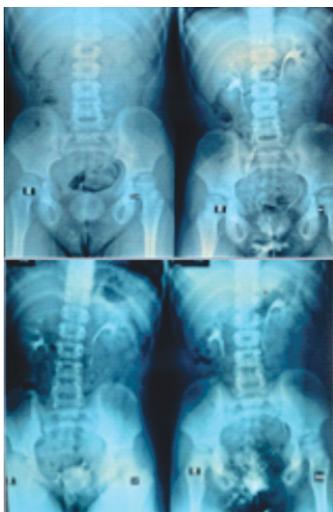


Figure 1. The results of BNO-IVU soft tissue opacity ec bladder extropia accompanied by distasis os pubis.

pelvic cavity on the inferior side, accompanied by pubic diastasis consistent with bladder extropia. The Intravenous Urography (IVU) showed a soft tissue opacity et causa bladder extropia accompanied by distasis os pubis.

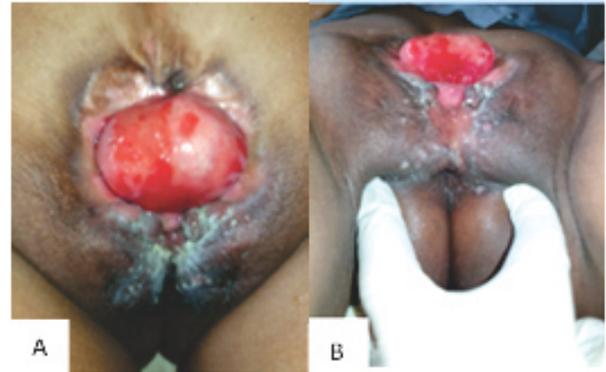


Figure 2. A female with bladder exstrophy (BE). (A) In preoperative, there was a mass in the suprapubic region. (B) The hole underneath was a vagina (B).

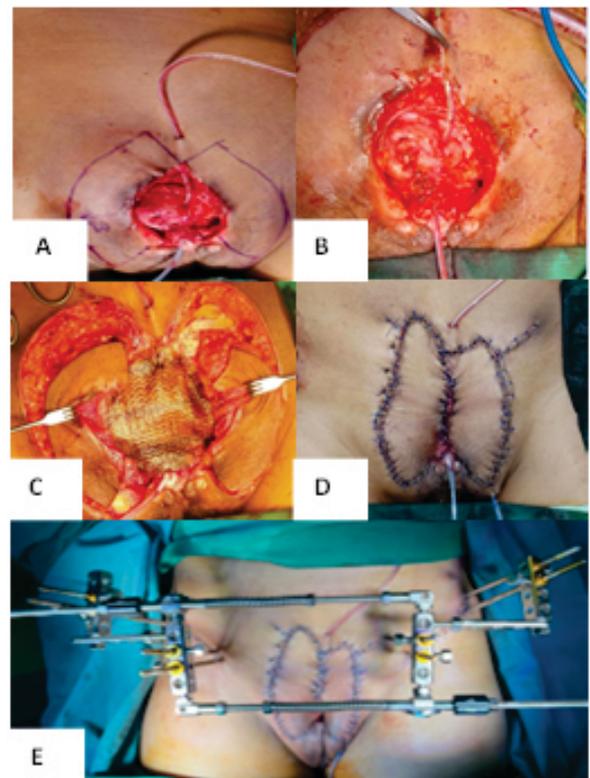


Figure 3. (A) Incision Design of Pelvic Osteotomy (B) Bladder Neck Reconstruction (C) Double Keystone Flap with Proceed Mesh (D) Immediate result after surgery (E) and External Fixation.

A bladder biopsy showed bladder exstrophy tissue with a picture of squamous metaplasia, cystitis glandularis. Some patients have reactive atypia.

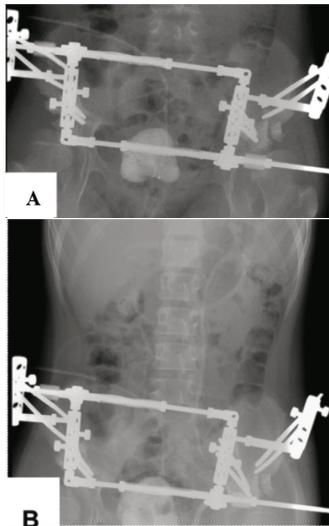


Figure 4. (A) Cystography showed no bladder extravasation (B) and left VUR grade III and right VUR grade II.

She had undergone pelvic osteotomy, Bladder Reconstruction, Bladder Neck Reconstruction, Double Keystone Flap with Proceed Mesh and External Fixation, Bladder Augmentation is planned and surveillance is carried out every 6 months with urine cytology and serial imaging.



Figure 5. Eleventh month follow up post operative.

Post operative wounds results in good surgical wounds, no pus or slough. Cystography was performed 14 days postoperatively with results normal bladder wall appears, 80cc bottle filling appears contrast reflux filling the left ureter up to the left renal pelvis with cupping calyx, mild dilatation of the ureter appears; contrast reflux filling the right ureter up to the right renal pelvis, right ureter dilatation does not appear.

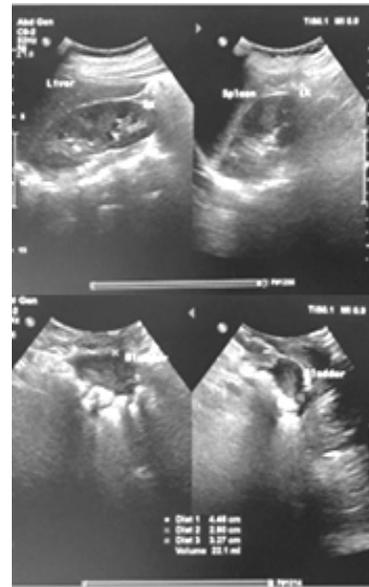


Figure 6. Ultrasound evaluation of kidney showed no abnormality of kidney and intact bladder.

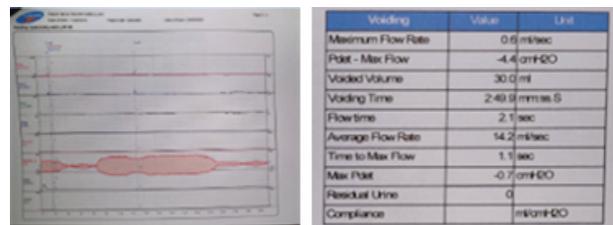


Figure 7. Urodynamics is done at a speed of 10ml/min, urine is obtained without the sensation of wanting a urinating.

Then, the patient complained of bedwetting five month after surgery Complaints are felt continuously, especially when the patient coughs and walks. Patients have undergone bladder training and regular medication of Solifenacin orally once daily. At this time the patient has independent mobilization. Furthermore, patient is planned bladder augmentation procedure.

DISCUSSION

Bladder exstrophy (BE) is a rare and serious congenital disorder, also often said to be the most severe congenital disorder that does not affect the central nervous system. This is because of its implications for long-term bladder function, the appearance of the genitals, sexual function and its associated psychological impact. Figure 8 shows examples of images of BE in women and men.²⁻³

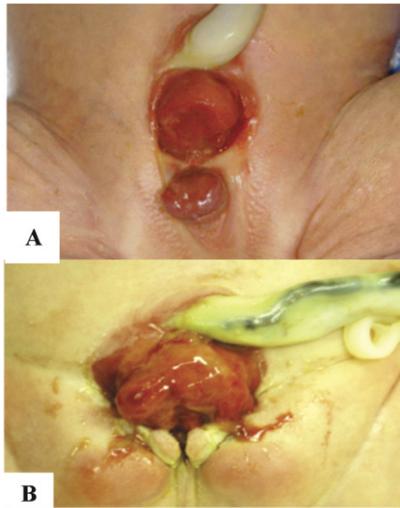


Figure 8. Features of classic bladder exstrophy (A) male (B) female, low umbilicus, small bladder and open bladder neck, pubic diastasis, separation of the rectus and pelvic floor, anterior ectopia of the anus and/or vagina and bifid epispadia or clitoris.

Bladder exstrophy is a herniation of the open bladder and posterior urethra through a defect of the lower abdominal wall and is accompanied by a wide pubic diastasis. The most important factor for obtaining a positive result is a successful early reconstruction/closure. However, in practice, there may be situations that prevent reconstruction from being carried out during neonatal, such as small bladder patterns or concomitant defects that must be treated first. Delayed reconstruction/closure in such situations can be carried out with good results in connection with urinary continence. There appears to be an upward trend in delayed reconstruction measures, as seen in the previous studies.⁴⁻⁵ However, if a persistent delay can negatively affect the growth of the bladder, thereby ultimately affecting the patient's ability to achieve urinary continence.³

Currently, the effect of delayed reconstruction on bladder growth is the subject of debate and speculation. A report by Ferrara et al, describes the experience of researchers in the delayed reconstruction of BE.⁶ From 2007 to 2012, 41 babies underwent delayed reconstruction and all were successful.⁶ However, due to concerns about the loss of bladder compliance that occurs after prolonged bladder contact, researchers have studied bladder capacity development. The results of gravity cystogram showed no significant difference in bladder capacity between patients with early reconstruction and delayed reconstruction. But the study was based on a single bladder measurement, rather than a longitudinal study of several measurements over several years. Further research is needed to confirm the results.^{2,6}

In addition to the studies mentioned above, another study has successfully examined 19 male patients with BE who performed surgery from 2001-2018.¹ A total of 13 patients entered the early bladder reconstruction group which reconstructed in the first 24-48 hours of life and as many as 6 patients entered the delayed reconstruction group which was reconstructed in the first month of life.¹ Researchers followed up for 12 months in the early and delayed reconstruction groups and found that there were no significant differences related to complications between early and delayed reconstruction. The results are shown in the following Table 1.

Some of review studies provides results that contradict these findings. In this study, bladder capacity was significantly reduced in patients whose improvement was delayed. Researchers examined three interest groups: (1) reconstruction during neonatal, (2) delayed reconstruction due to a small bladder template and (3) delayed reconstruction with a normal template. In this study, there was a statistically significant decrease in bladder volume, with a decrease of 36 cc and 29 cc in patients with small template and normal template compared to neonatal reconstruction. The results found in this

Table 1. Comparison of complications between early and delayed reconstruction.

	<i>Early reconstruction</i> (n=13)	<i>Delayed reconstruction</i> (n=6)
Failure of primary reconstruction	2 (15%)	0 (0%)
Transient hydronephrosis (< 6 mo)	3 (23%)	2 (33%)
Persistent hydronephrosis (> 6 mo)	1 (8%)	1 (17%)
Recurrent UTIs	5 (38%)	3 (50%)

study confirm the findings in other studies of an overall decrease in the final capacity of the bladder with delayed reconstruction.⁷

Statistical methods that take into account repeated measurements, in a retrospective study showed the result that there is bladder capacity found in cystography is much smaller in the group with delayed reconstruction compared with closure during neonatal (early reconstruction).³ The time points for the most significant decline appear after the 2nd and 4th quartiles, representing 4-6 months and more than 9 months, respectively. From this, the researcher theorizes that the appropriate time to close as early as possible (1st quartile), or if a delay is necessary for the growth of the bladder template, then between 6 and 9 months (3rd quartile). Clinically, bladder capacity is an optimal predictor of urinary outcome.⁷

The patient in this case was a 12-year-old female patient who came with complaints of persistent dripping urine, physical examination obtained appear fixed mass, hyperemic size 10 cm x 7 cm. Then the patient underwent a Pelvic Osteotomy + Delayed Bladder Reconstruction + bladder Neck Reconstruction + Double Keystone Flap with Proceed Mesh + External Fixation on September 7, 2022 when the patient was 12 years old.

The monitoring and evaluation post operative through an outpatient setting, cystography carried out 14 days after the operation showed the bladder capacity and compliance quite good. The results of urodynamics performed at a speed of 10ml /min obtained urine out without the sensation of micturition. Further action is considered in the form of periodic evaluations every month and further consideration is done bladder augmentation or urinary diversion.

Exposure of the case shows that delayed reconstruction in BE can be considered because the patient's outcome is quite good although in March the patient complained of urinary incontinence. This is in accordance with the literature that states that delayed reconstruction can still be considered in special conditions that do not allow early reconstruction. A recent study showed that the results of delayed reconstruction in these situations can be performed with good results in relation to urinary continence.⁸ However, if the delay is persistent it can negatively affect the growth of the bladder, thus ultimately affecting the patient's ability to achieve urinary continence as experienced in patients.⁹

CONCLUSION

Bladder exstrophy (BE) is a rare congenital disorder with important repercussions in the patient's life. Delayed reconstruction can be performed under special conditions that do not allow early reconstruction. Delayed reconstruction proves to be advantageous because it avoids general anesthesia at the time of the still physiologically immature newborn. Long-term follow-up is needed to assess aesthetics and genital function in this delayed reconstruction. However, if the delay is still persistent it can negatively affect the growth of the bladder, thus ultimately affecting the patient's ability to achieve urinary continence.

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