CASE STUDY

# Case Report: Long Term Follow Up of Bladder Exstrophy Repaired in Stages with Complete Urinary Continence

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## Abstract
Bladder exstrophy is a rare congenital anomaly displaying symptoms such as the urinary bladder turned inside out, an exposed dorsal urethra, foreshortened penis, anteriorly displaced anus and widely separated symphysis pubis. This abnormality is managed by many methods, one of which includes a functional reconstruction. The major goals of surgical intervention are: Preservation of the renal function, achieving urinary continence with adequate sexual function and acceptable cosmetic appearance. In this report we present a long term follow up (25 years) of a male patient with classical type bladder exstrophy, managed at our hospital in stages, he is fully continent and voids spontaneously without clean intermittent self-catheterization, with normal renal function, adequate penile erection and has ejaculation

## Keywords
Bladder exstrophy; Staged repair; Urinary continence

## Introduction

Bladder exstrophy is a rare anomaly that involves the complex urogenital system, commonly occurring in males[1]. The incidence of bladder exstrophy is 3.3 per 100,000 births[2]. Even though the exact etiology is yet unknown, recent evidence implicates on poor mesenchymal ingrowth with subsequent deleterious effects on the cloacal membrane rupture in its timing and position[3]. Preliminary diagnosis may be suspected during routine prenatal ultrasound; however, the definite diagnosis is conducted after birth[1]. Assyrian tablets from 2000 BC are thought to be the first description found of bladder exstrophy. In 1597, von Grafenberg was the first to describe exstrophy as a medical condition, but Chaussier established the term exstrophy in 1780. However, it was not until 1748 where Mowat provided the first complete description of bladder exstrophy in his work[2]. Bladder exstrophy is an abnormality where the lower abdominal wall fails to fuse during embryonic development, leaving the bladder exposed inside out on the lower abdomena with a flattened pelvis and a wide diastasis of the symphysis pubis[4]. Many descriptions have been found on this anomaly, one of which describes this condition as if a scissors was passed through the urethra of a normal person and was used to cut through the skin, abdominal wall, anterior wall of the bladder and urethra, and the symphysis pubis; with the cut edges folded laterally, such as opining the pages of a book[5].

Bladder exstrophy is a severe defect requiring extensive surgical demands, starting mostly during the first days of life and usually extending through adolescence[3]. Bladder exstrophy is managed surgically in single or multiple stages with a wide range of operations, the aims are; closure of the bladder and abdominal wall defect, preserve renal function, achieve urinary continence and maintain an adequate sexual function with cosmetic appearance; this can be achieved by either urinary diversion; or by anatomic reconstruction, which is favored[5].

## Case Report

AA, who is now a 25 year old man, presented to us at the age of 18 months with classical type bladder exstrophy (Fig. 1).

The patient was managed in stages. The first stage consisted of primary closure of the bladder with bilateral iliac osteotomies (Fig. 2).

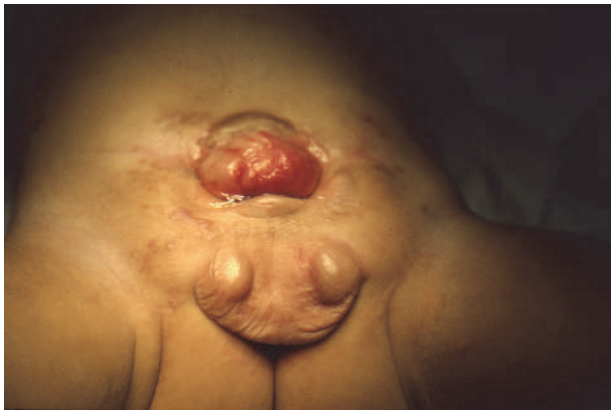

**Figure 1.** Patient at initial presentation age 18 months.

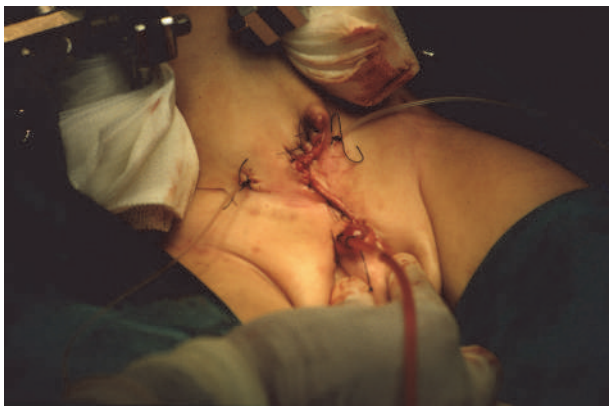

**Figure 2.** Appearance after initial repair with bilateral iliac osteotomies.

A year later, bilateral re-implantation of the ureters with bladder neck tightening (Young-Dees-Leadbetter (YDL) technique) was done. At the age of 4 years old, he had penile elongation and repair of the epispadias (Fig. 3). Because of inadequate bladder capacity and high intravesical pressure, bladder augmentation with removal of newly formed two bladder stones was done two years later.

The patient was kept on clean intermittent self-catheterization (CIC) and followed up for a year, then disappeared from follow up. He presented this time (after 18 years) for evaluation and marriage counseling. He mentioned that he went back home and he was well

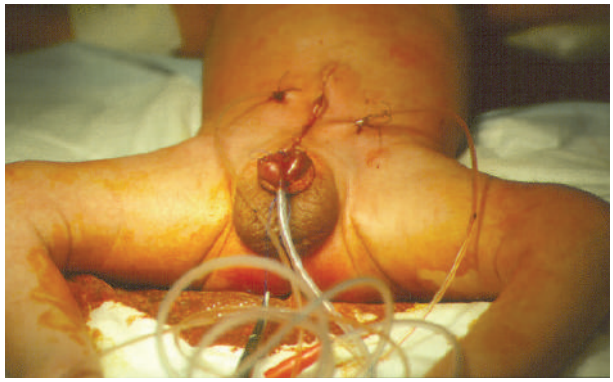

**Figure 3.** Appearance after penile reconstruction and epispadias repair.

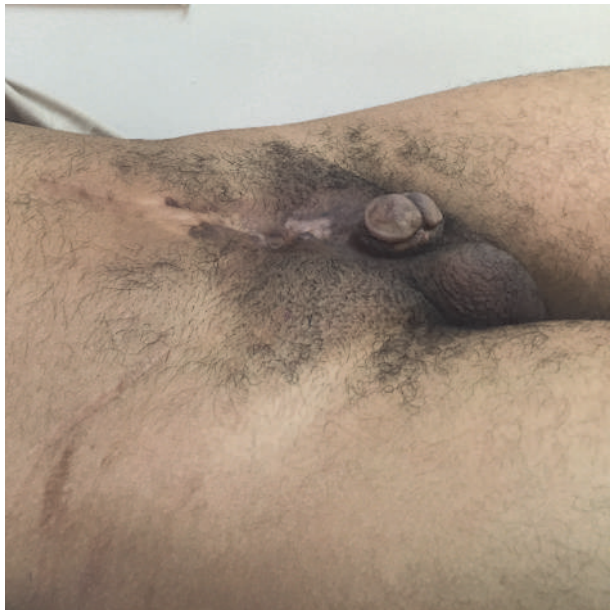

**Figure 4.** Patient at recent presentation with abdominal scars of previous surgeries.

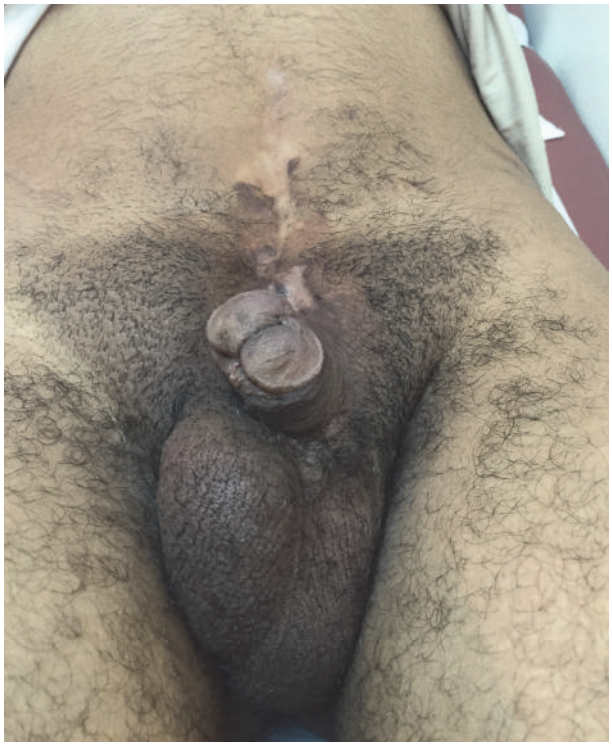

**Figure 5.** Another view at recent presentation revealed penile tilt and bilateral normally descended testes.

during that long time, apart from a single admission at age of 15 years old for an operation. A medical report indicated that he had a redo re-implantation of the left ureter. At the time of presentation general examination revealed multiple abdominal scars of previous surgeries, his penis is short (6 cm) and mildly tilted to the right side with both testes down in the bottom of the scrotum (Figs. 4, 5) and he is completely dry. He mentioned that he is capable of voiding urine spontaneously with normal fecal continence. Biochemical assessment revealed normal renal function. CT with IV contrast showed mild hydroureter and hydronephrosis in the right side, with small diverticulum in the posterior aspect of the bladder, VCUG revealed thick walled augmented bladder and wide neck of diverticulum with no VU reflux. Urological assessment by uroflorometry showed normal voiding parameters with mild residual post voiding urine.

## Discussion

The goal of surgical reconstructions in bladder exstrophy is to correct the underlying urogenital defect; this is mainly favored by the anatomic reconstruction, also called modern staged reconstruction of exstrophy (MSRE)[5]. To achieve the desired outcome, several procedures should be performed[3]. In MSRE, the bladder closure is achieved by trimming the outer edges of the exstrophic bladder until healthy viable mucosa and muscle is available for closure. Ureteric stents are placed, anchored and brought out on either side of the bladder suture line. The abdominal wall then closed in layers whenever possible[6]. The second stage is bladder neck repair that is preformed according to the Young-Dees-Leadbetter (YDL) technique, which is done by one layer tabularization of the posterior urethra from the bladder neck to the external meatus. Some patients, with relatively small bladder may undergo augmentation cystoplasty, by utilizing part of the colon or loop of small bowel[7]. Young-Dees-Leadbetter (YDL) technique may achieve urinary continence and complete bladder evacuation, however intravesical pressure may be raised; low-pressure reservoir can be achieved by augmentation cystoplasty, so a combination of these procedures is advisable when the residual bladder is small. These patients usually maintain continence on CIC program, however a few will still suffer from continuous wetting, in which case appendic-ovesicostomy (Mitrofanoff) procedure with frequent CIC through the exteriorized appendicular tip is indicated[8]. Finally, in patients who fail to achieve continence following anatomic reconstructions, urinary diversion may be useful[5]. Some of the significant long-term complications that are associated with urinary diversion like uretosigmoidostomy (USO) are; hyperchlormic metabolic acidosis, chronic pyelonephritis, renal failure, urinary calculi, and a higher risk of developing adenocarcinoma at the site of anastomosis[9-11]. Combination of epispadias repair with bladder exstrophy closure in a single stage may reduce the chance of failed repair in staged closure[12,13]. Ureteric re-implantation is done when the ureters are dilated as a result of V-U reflux[7]. Patients who underwent MSRE mostly achieved urinary continence and acceptable dry interval during the day but not at night, through a program of CIC[14,15]. Complete dryness day and night after CIC was reported in those who had bladder augmentation[16,17]. Adequate urinary continence following staged reconstruction without CIC like our case was reported infrequently in the literature. Factors contributing to successful results may include; early bladder closure, surgical interval of at least 6 months between each stage, except for epespadias repair, where it should be done later on in the patient's life, also adequate bladder neck reconstruction[18]. Our patient had the steps of reconstruction with a minimal interval of one year between steps, this may have helped in achieving these good results. Fertility

following BE repair were widely reported in females while in males it was less. Some studies showed that most patients would achieve satisfactory erection, adequate amount of ejaculation, and satisfactory orgasms for both partners, while a few patients were not satisfied because of the small penis[19]. It has been reported that at the time of puberty, patient's concern would shift onto self-esteem, sexuality, fertility and sexual function[19,20]. Our patient, although he has a small penis, has erection and ejaculation, as well as self-satisfaction. The Patient will be followed through his marriage to observe his fertility, and seminal analysis might be done for effective evaluation of his fertility when the patient comes again for follow up.

## Conclusion

Overall the surgical repair of bladder exstrophy is a challenging surgery, requiring repair in a single or multiple stages, with the high risk of developing both early and long-term complications, thus a close follow up is required.

We conclude that full continence with satisfied sexual function can be achieved following staged repair of classical type bladder exstrophy, and recommend all surgeons dealing with bladder exstrophy diseases to study the long term results of their male patients to enrich the available medical library for comparison.

## Conflict of Interest

The authors have no conflict of interests.

## Disclosure

The authors did not receive any type of commercial support either in forms of compensation or financial for this study. The authors have no financial interest in any of the products or devices, or drugs mentioned in this article.

## Ethical Approval

Obtained.

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# تقرير حالة: متابعة حالة المثانة الهاجرة على المدى الطويل الذي تم اصلاحه في مراحل عدة ولديه القدرة على حصر البول كاملة

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***المستخلص.*** المثانة الهاجرة عيب خلقي نادر بحيث المثانة البولية بارزة إلى الخارج، عدم التحام الأعضاء التناسلية الخارجية وصغر حجمه، نذرح الشرج للأمام، وبعد عظام الحوض. هذا العيب يتم علاجه عن طريق إعادة تأهيل وظيفي للأجهزة البولية التناسلية. الأهداف الرئيسية من الجراحة هي؛ الحفاظ على وظائف الكلى، تحقيق القدرة على حصر البول، كفاءة الجنسية الوظيفية وأيضًا الحصول على مظهر تجميلي مقبول.

في هذا التقرير نقدم لكم متابعة حالة على مدى ٢٥ سنه لمريض بالمثانة الهاجرة النوع الكلاسيكي، تم علاجه في مستشفى جامعة الملك عبد العزيز على عدة مراحل، ولديه القدرة الكاملة على حصر البول والقدرة علي التبول بشكل عفوي دون الحاجة الى القسطرة البولية النظيفة المتقطعة لأخلاء المثانة، مع وظائف كلى طبيعية، لديه قدرة كافية على انتصاب القضيب والقدرة على القذف.