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Continent cutaneous diversion for bladder exstrophy in adults

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KEYWORDS

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Abstract

Objectives: Bladder exstrophy is a rare and devastating congenital anomaly. These patients are usually seen immediately after birth where correction can be attempted. Presentation in adulthood where bony fusion has occurred and bladder capacity is very low presents a major surgical challenge. Such patients require urinary diversion. This is a review of our initial experience with continent cutaneous urinary diversion in patient with classic bladder exstrophy who presented in adulthood.

Patients and methods: We reviewed the social and clinical characteristics, operative reports, and postoperative course of two female and five male patients who have been living with bladder exstrophy since birth. They underwent augmentation cystoplasty or simple cystectomy, bladder neck transection, repair of epispadias followed by a continent cutaneous diversion. The continent catheterizable outlet was constructed using the serous lined extramural tunnel technique as a continence mechanism. Postoperative continence, renal function, and changes in social life were documented.

Results: Follow-up has ranged from 17 months to 5 years in these initial patients. Thus far, all these patients have achieved complete daytime and night time continence. Complications consisted of pouch cutaneous fistula which was successfully repaired with interposition of a rectus abdominis flap and one case of urosepsis. All upper urinary tract imaging studies have shown stable function. All the patients are doing well with improved social interaction.

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Conclusion: Bladder neck closure in conjunction with continent cutaneous diversion is a reliable and safe method for achieving continence in adults presenting with bladder exstrophy. Total continence can be achieved without resorting to multiple complex and expensive surgeries.

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Introduction

Bladder exstrophy is a rare congenital anomaly that occurs with a reported prevalence of 3.52 per 100,000 [1]. The condition involves an open bladder and urethra, wide diastases of the symphysis pubis, a bifid clitoris in girls and a short penis with upward deviation in boys. The visible bladder mucosa with urine dripping from the ureteral orifices is so striking that it cannot go unnoticed in the neonatal period. Surgical management in the neonatal has been shown to provide the best functional and cosmetic outcome [2]. Delayed presentation till adolescence and adulthood is extremely rare. However, due to lack of awareness, lack of access to health care and poor socioeconomic conditions in developing countries some patients may not receive treatment during infancy. We present our experience with seven adult patients with bladder exstrophy treated by bladder neck transection, closure and augmentation in conjunction with a continent cutaneous diversion.

Patients and methods

This study involves seven grown-up patients who presented to the urology unit of a tertiary hospital in Ghana (Korle Bu Teaching Hospital) with features of bladder exstrophy between 2006 and 2010. There were two females and five males. The patients' ages at presentation ranged from 16 to 23 with a mean age of 18 years. Apart from one patient who has had multiple failed attempts at closure during childhood, there has not been any attempt at surgical correction in the others.

These unfortunate patients have major social problems as manifested by the fact that three of them have never been to school. Three of the patients were abandoned by their fathers at birth. One patient was brought to our unit by catholic nuns after being abandoned at age 9 by both parents. Physical examination revealed features of classic bladder exstrophy. Because of the prolonged exposure of the bladder mucosa to the environment there was extensive keratinization of the edges (Figs. 1 and 2). Laboratory investigations showed normal renal function. All patients were adequately educated on continent cutaneous diversion especially the fact that they will have a stoma and have to self-catheterize.

All patients were operated on in one setting by a single surgeon (JEM). In six cases the keratinized edges of the bladder plate were excised and patch left augmented with 30 cm of ileum to increase bladder capacity. In one case a cystectomy was performed because gross examination of the bladder plate was suspicious of a neoplastic change. Histopathological report revealed squamous metaplasia with no evidence of malignancy. Ileal segment was used for augmentation or for the construction of the neobladder in all cases. The continent external conduit was fashioned using the extra mural tunnels as described by Abol-Enein and Ghoneim [3]. The appendix was used in five cases and a tapered ileal segment in two cases. Transection and closure of the bladder neck was performed in all cases.



Fig. 1 Adult female with classic bladder exstrophy.



Fig. 2 Adult male with classic bladder exstrophy.

In the male patients, the urethral plate was transected just distal to the bladder neck and the bladder plate completely separated from the urethra (Fig. 3). Bladder neck transection above the level of the ejaculatory ducts helps to preserve emission of seminal fluid. Epis-padias repair was performed using the partial penile disassembly



Fig. 3 Bladder plate separated from the urethra at the level of the bladder neck.

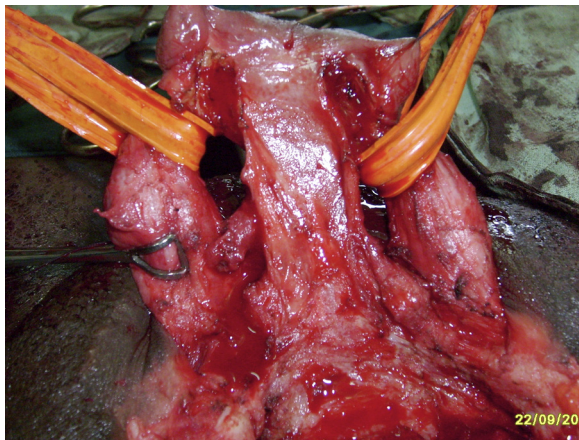


Fig. 4 Penile disassembly.

technique [4] which involves keeping the urethra attached distally at the glans (Fig. 4) and transposition of the urethra from dorsal to ventral position using corporeal rotation. The urethral plate was then tubularized from the prostatic urethra distally to form a neourethra.

There was no attempt at genital reconstruction in the female patients.

The abdominal wall was successfully closed in all cases without flaps or osteotomy. Postoperatively, all patients had an abdominal drain, 10Fr Nelaton catheter as a ureteral stent, and suprapubic catheter for urinary drainage. A gentle bladder wash with normal saline was started on postoperative day 6 to prevent catheter blockage from mucus. Broad-spectrum antibiotics were administered for 7 days. The stent was removed on postoperative day 14 and self-catheterization started. The suprapubic tube was removed when the patient can confidently self-catheterize the abdominal stoma using a 16Fr Nelaton or red rubber catheter. The follow-up included urinalysis, ultrasound scan for the upper tract and serum creatinine.

Results

After the reconstruction, all patients were regularly reviewed at the teaching hospital by the urological team every fortnight for 2 months and then every month for 6 months. They were then assigned to an interested physician in their district hospital who has a direct telephone contact with urological team at the teaching hospital. This arrangement saves transportation cost to the teaching hospital and

ensures that only complications which cannot be managed locally are referred. Follow-up period has ranged from 17 months to 5 years. All patients are completely dry day and night. Four patients required self-catheterization at 4-h intervals and 1 patient who is still in school however prefers to leave the catheter indwelling and spigotted. The mean bladder capacity was 400 mL (range 300–550). Complications included two patients who had persistent postoperative suprapubic urinary leakage that required a rectus flap for closure. One patient has been admitted on two different at his district hospital with suspected pyelonephritis. All these episodes were successfully managed at the district level. There has however been no deterioration of renal function seen in any of these patients during follow-up.

Direct personal interaction with the patients indicates a high degree of satisfaction with the cosmetic results of the procedures, however, one male patient whose penis was very small before reconstruction is not happy with the size of the penis. All report increased social interaction and self-confidence. Both girls have completed vocational training and are gainfully employed. Two of the male patients are currently in school are doing very well academically.

Discussion

The current primary objectives of the surgical management of the classic bladder exstrophy are to close the abdomen and bladder, establish urinary continence with the preservation of renal function and provide acceptable external genitalia [5,6]. This is best started in the neonatal period, where results of primary reconstruction have been good [7,6]. The single most important step to achieve urinary continence is a successful primary anatomical closure of bladder, posterior urethra, abdominal wall and pelvic bone. When necessary, pelvic osteotomy is done to allow a tension-free abdominal closure which has been shown to consistently reduce the chance of breakdown [8]. Presentation in adulthood where bony fusion has occurred and the exposed bladder plate is contracted therefore presents a major surgical challenge.

Different types of surgical options have been reported to achieve continence and provide an acceptable appearance of the genitalia. Often several operations are needed to achieve acceptable results. The first reported case of adult bladder exstrophy was in 1987 by Matsuda et al. [9]. He treated two adult female patients with cystectomy, construction of a Kock's continent ileal reservoir, and closure of the abdominal fascial defect using alloplastic material. Gulati et al. [10] reported on management of two adult females presenting with untreated bladder exstrophy. They underwent cystectomy and modified Mainz pouch with primary closure of the abdominal wall defect. Ozdiler and Sarica [11] reported a 50-year-old woman, Quattara and Daffe [12] a 39-year-old man, and Andankar et al. [13] reported five cases of which 3 underwent staged reconstruction and 2 complete primary repair. Recently Ansari et al. [14] have shown that it is possible to reduce the pelvic diastases in adult patients. They reported on a technique of pelvic osteotomy in adult female patients with bladder exstrophy. Their procedure involved bilateral innominate and vertical iliac osteotomies from an anterior approach and the placement of an external fixator. The fixator and pelvic bones were gradually cranked together over a period of 2–3 weeks until the diastasis was less than 4 cm. The latest report on adult bladder exstrophy was by Shoukry and Shoukry [15] who reported on five patients. Three patients had bladder preservation

with primary repair, including bladder closure, bladder neck reconstruction and epispadias repair; two of them also had augmentation ileocystoplasty. The remaining two patients underwent ureterosigmoidostomy, cystectomy and epispadias repair.

The main concern of our patients at the time of presentation was urinary incontinence and social isolation. Most adult exstrophy patients consider dryness a more important criterion of a good QOL than bladder reconstruction or urinary diversion [16]. Reports from a similar third world environment also indicated that urinary incontinence was the main complaint, which greatly affected the patients schooling and social life [17]. All these patients in our series came from very deprived areas and their management was funded by religious bodies and philanthropist who have limits on how much they can contribute. We therefore decided to adopt a surgical strategy that is straightforward, acceptable to the patient and inexpensive to perform without the need for repeated anti-incontinence procedures. Several types of urinary diversions were therefore discussed with the patients and they encouraged to express their own views on the procedures.

Ileal conduit although easy to perform was not acceptable to any of the patients because the urostomy bags are not easily available or affordable in a third world environment, additionally the patients found the idea of a bag on the abdomen will affect their social interaction.

For many years ureterosigmoidostomy was the established form of urinary diversion for patients with bladder exstrophy when bladder reconstruction is not feasible or has failed. It provided a normal body image with perfect social integration for the patient because they do not have to wear a bag and there is no stoma on the anterior abdominal wall [18]. Although the initial series was associated with multiple metabolic problems, results improved markedly with newer modification such as the Mainz pouch II. Even though these modifications could reduce the perioperative and postoperative complications of the procedure, ureterosigmoidostomy is still fraught with long-term problems including ascension of bacteria to the upper urinary tract, hypokalemic acidosis, rectal incontinence, ureteral obstruction, and delayed development of malignancy [18–20].

Bladder neck closure in conjunction with a continent cutaneous diversion procedure is a highly effective method of achieving continence [21,22]. Several reports on the functional outcome of continent catheterizable outlet using the serous lined extramural tunnel technique have shown a continence rate exceeding 90% [23]. The down side of bladder neck closure is that, the patient is obligated to continue lifelong intermittent catheterization and must be committed to this regimen, because non-compliance can lead to renal deterioration.

There has been some debate in the literature regarding malignant transformation in extrophied bladder especially when the mucosa has been exposed to the environment till adulthood. Some authors recommend cystectomy with urinary diversion for all adults with exstrophy because of the malignant potential [10,24]. However, Engel [25] after examining the histological changes in exstrophic bladders and analyzing the clinical course of treated patients concluded that histologic changes were present in all bladders, including those of neonates, and that malignant transformation was not prevented by closure of the exposed bladder plate even during the neonatal period. These patients, therefore, require regular follow-up

to detect malignant transformation at an early stage. We maintained the bladder plate in six out of the seven cases because its offers significant advantages as suggested by Gearhart [26] who wrote that the preservation of the bladder offers two major advantages, First, using bladder template meant taking less bowel for augmentation and second, if ureteral reimplantation was necessary, the bladder template was a much better substrate for reimplantation than a bowel wall.

The penis in exstrophy patients is short and broad before and after reconstruction [27]. This is a major source of worry to the patient. Long-term follow-up of exstrophy patient has revealed that at least 36% of the patients are severely restricted in their sexual lives [18]. The most commonly cited reasons for sexual dissatisfaction were short penis, curved erection and erection too close to the abdominal wall. It is therefore important that these patient are counselled appropriately about how much can be achieved surgically. Four out of the five male patients in our series are nevertheless satisfied with the length of the penis. Despite these limitations all the patients have become well adjusted psychosexually and psychologically primarily because they are dry.

Conclusion

Management of bladder exstrophy in adults is a major surgical challenge especially in a resource poor environment. The results of our small series indicate that, continence can be achieved with bladder neck closure and augmentation in conjunction with continent cutaneous diversion.

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