

Complete primary repair of adult bladder exstrophy-epispadias complex without iliac osteotomy: a case report and literature review

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Bladder exstrophy-epispadias complex is a rare anomaly of the genitourinary system. Adult presentation is uncommon because most patients would have completed their treatment in early childhood. We report a 32-year-old man with classic bladder exstrophy who underwent complete primary repair with no iliac osteotomy to highlight the possibility of complete repair of adult bladder exstrophy without performing an iliac osteotomy despite a wide pubic symphyseal diastasis. The patient presented with a lower abdominal wall defect and an abnormal penis since birth. On examination, he had a lower abdominal wall defect, exposing the bladder plate with a flat and spade-like phallus. Pelvic X-ray showed a wide pubic symphyseal diastasis. He underwent complete repair of exstrophy without an iliac osteotomy. Horizontally oriented double V-Y flaps were raised to cover the abdominal wall defect.

Keywords: bladder exstrophy, bladder exstrophy-epispadias complex, complete primary repair of bladder exstrophy, epispadias, urethrocutaneous fistula

Case report

A 32-year-old man presented to the urology outpatient clinic of the Federal Teaching Hospital Gombe with complaints of a lower abdominal wall defect, urine leakage, and an abnormal penis since birth. On examination, there was an 8 × 6 cm infraumbilical abdominal wall defect with an exposed, grossly normal bladder plate (Figure 1A). No apparent bladder mucosal changes were seen, such as vesicles or polyps, and the ureteric orifices were jetting out urine. There was minimal dermatitis of the surrounding skin. The dorsally curved phallus was flat and spade-like, with an exposed urethral plate. The corpora cavernosa were ventrally located to the urethral plate, and the testicles were high-riding within the scrotum.

A diagnosis of bladder exstrophy-epispadias complex was made. Pelvic X-ray showed a pubic symphyseal diastasis of 6.8 cm (Figure 1B). An abdominal ultrasound scan showed normal kidneys with preserved corticomedullary differentiation and no pelvicalyceal dilation. His full blood count (FBC), electrolytes, urea, and creatinine (E/U/Cr) were normal, while the culture of urine collected by catheterising the ureters yielded *Pseudomonas aeruginosa* sensitive to amikacin.

The patient was treated with intramuscular amikacin for five days and subsequently underwent complete primary repair of the exstrophy without iliac osteotomy. An incision line was marked around the bladder plate, the bladder was dissected off the rectus abdominis muscles, and a plane was created between the bladder dome and peritoneum. The bladder was further mobilised by dividing the bladder ligaments and intersymphyseal bands to facilitate tension-free placement of the bladder deep within the pelvis. After completely freeing the bladder neck, it was gently tapered to create outlet resistance. Feeding tubes were used to stent the ureteric orifices before closing the bladder. Closure of the posterior urethra continued without further tapering to minimise the

risk of urethral stenosis. An additional muscle strip from the pelvic floor was wrapped around the bladder neck to increase the outlet resistance further.

The epispadias was repaired using the modified Cantwell–Ransley technique. The estimated bladder capacity was above 200 ml, which is expected to increase with bladder cycling and training. Primary bladder closure was possible because of its adequate capacity, absence of disease, and the ability to appropriately mobilise it deep within the pelvis. The rectus muscles were approximated to the midline to reduce the risk of ventral hernia. However, the abdominal

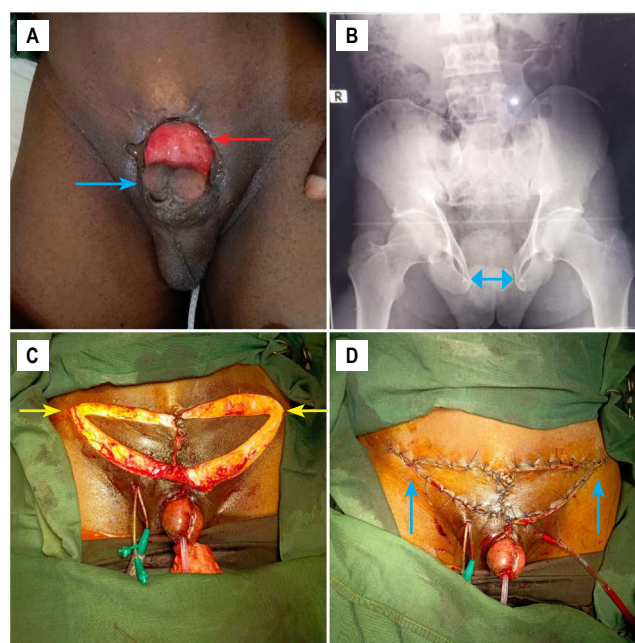


Figure 1: Adult bladder exstrophy-epispadias complex presentation and treatment A – Preoperative image of the exposed bladder plate (red arrow) with dorsally curved epispadias phallus (blue arrow), B – pelvic X-ray showing the wide pubic symphysis diastasis, C – double V flaps (yellow arrows), D – double V flaps closed as double Y (blue arrows)



Figure 2: Three-month postoperative image of the healed abdominal wall and epispadias repair

wall could not be closed primarily due to the large defect. Therefore, horizontally oriented double V-Y flaps were raised to cover the defect (Figure 1C and D).

Postoperatively, the patient had a retropubic drain, urethral stent, and bilateral, external ureteral catheters. He also received analgesics, cefuroxime, and metronidazole. However, on the 14th postoperative day, he developed a 2 × 2 mm urethrocutaneous fistula at the junction of the urethra and the bladder. He was managed non-operatively with continuous bladder drainage and oral levofloxacin as the postoperative urine culture yielded *Pseudomonas aeruginosa*. The fistula resolved completely after three weeks of non-operative management. The patient was subsequently discharged home to continue with follow-up visits.

On follow-up visits, the patient reported voiding per urethra and was mostly dry. Figure 2 shows the postoperative repair at three months. Similarly, a follow-up renal ultrasound scan at three months postoperative was normal. We intend to repeat the renal scan at six months and a year postoperative.

Discussion

Bladder exstrophy-epispadias complex is a rare congenital anomaly. The condition's spectrum varies from the isolated epispadias to classic bladder exstrophy and the most severe and rarest form, cloacal exstrophy.¹ The estimated incidence of exstrophy-epispadias complex is approximately 1 per 35 000 live births, and patients with the condition are 700 times more likely to develop bladder cancer than their age-matched counterparts.² The male-to-female ratio is estimated to be 2.5:1.³ Bladder exstrophy treatment usually starts in the neonatal period. It is recommended that such fetuses be delivered in centres with expertise in the surgical management of bladder exstrophy.

Adult bladder exstrophy is extremely rare in developed countries as most of the repairs are carried out during childhood. However, because of factors such as poor awareness, limited healthcare access, social embarrassment, and poverty in developing countries such as Nigeria, patients may carry their condition up to

adolescence or adulthood or when they are about to get married. Our index patient did not undergo treatment during early childhood because his parents were poor, and he was embarrassed by his situation when he became an adult. However, he eventually sought treatment due to pressure from his spouse and colleagues.

The best results are obtained with early exstrophy repair. This is because prolonged exposure of the bladder plate to the external environment may predispose the patient to dermatitis, recurrent urinary tract infection, bladder atrophy, metaplasia, and bladder malignancy. Surprisingly, despite having an exposed bladder for three decades, the patient maintained a grossly healthy-looking and good-capacity bladder. He only had a few episodes of symptomatic urinary tract infection and minimal dermatitis of the surrounding skin. He maintained the bladder by irrigating it twice daily with clean, lukewarm water. Additionally, he applied petroleum jelly to the surrounding skin and covered the exposed bladder with a clean, non-adherent nylon bag and a clean, absorbent cloth, which was changed regularly. He learned this procedure from his parents in early childhood. Upon discharge from the hospital, he was counselled and educated on the long-term complications of bladder exstrophy, including bladder malignancy and the need to adhere to follow-up.

Numerous theories have been proposed to explain the pathogenesis of bladder exstrophy. Kulkarni et al.⁴ proposed that an abnormal origin of the primordial genital tubercle in a more cephalad position than normal leads to a wedge effect that interferes with the medial migration of the mesoderm and the midline approximation of the mesodermal structures in the lower abdominal wall. The diagnosis of bladder exstrophy can be made prenatally with ultrasonography between the 15th and 32nd gestational weeks.⁵ The findings that suggest bladder exstrophy include non-visualisation of the urinary bladder despite a normal amniotic fluid volume and a solid bulging mass in the lower abdominal wall. Other minor findings include low insertion of the umbilical cord, normal kidneys, small penis or bifid clitoris, and widening of the iliac crests.⁶ Prenatal diagnosis allows for parental counselling and arranging the delivery to be performed in a specialised institution, so immediate repair can be performed by a multidisciplinary team with experience in managing the disorder.⁵ Notwithstanding, up to 75% of patients born with exstrophy-epispadias complex are diagnosed postnatally.⁵

When a child is born with exstrophy, a complete neonatal evaluation with thorough pulmonary and cardiac assessments should be made.⁵ Also, routine laboratory investigations should be performed, such as FBC and E/U/Cr. Renal ultrasonography should also be performed to rule out any concomitant anomaly. The bladder should be covered with non-adherent dressings, such as plastic wrap, and kept clean with saline washes at every diaper change.⁵ This will help prevent infection and sticking of the bladder to diapers or clothing.

Over the years, there has been significant improvement in the outcome of patients with exstrophy-epispadias complex due to advancements in diagnosis and genitourinary reconstruction. However, repair of bladder exstrophy is still challenging because of the rarity of the disorder and its complexity, with few centres offering specialised care for the condition. The definitive management of classic bladder exstrophy involves multiple procedures such as

bladder closure, epispadias repair, bladder neck reconstruction, bladder augmentation, and ureteroneocystostomy. Other anomalies that may be tackled alongside the exstrophy repair are abnormalities of the bony pelvis.

The two most widely used approaches in the definitive management of bladder exstrophy-epispadias complex are the complete primary repair of exstrophy (CPRE) and the modern staged repair of bladder exstrophy (MSRE). In MSRE, the bladder is repaired, and the posterior urethra closed in the newborn period, the epispadias repair is performed at 6–12 months of age, and the bladder neck reconstruction is performed when the child can cooperate in a good voiding programme.⁷

One critical aspect of the first stage of repair is ensuring that the bladder and posterior urethra are placed deep in the pelvis to simulate normal morphology. This is achieved by adequate dissection of the pelvic diaphragm to ensure the deep location of the bladder and posterior urethra.⁷ When the bladder is too small, or there are significant polypoid changes in the bladder at birth, bladder closure may be delayed for 4–6 months, following which the bladder is re-evaluated.⁷

Patients with classic bladder exstrophy are born with pubic diastasis, which increases with age from a mean of 4 cm to about 8 cm at 10 years, compared with a mean normal width of 0.6 cm across all ages.⁸ Our index patient had a wide pubic diastasis of about 6.8 cm. Osteotomy is usually needed in older children and aims to relax the tension on the bladder and abdominal wall repair during wound healing.⁸ The different osteotomy techniques include posterior iliac osteotomy, anterior osteotomy of the superior pubic rami, anterior diagonal iliac osteotomy, and anterior innominate osteotomy with optional vertical posterior iliac osteotomy.⁸ However, osteotomy may not be necessary if repair is carried out within the first 72 hours of life due to limited amounts of pubic diastasis or malleability of the pelvis.

Despite the wide pubic diastasis in our index patient, bladder and abdominal wall repair without osteotomy was possible, thereby avoiding complications related to osteotomy, such as pain, malunion, non-union, infection, and iatrogenic fracture.⁹ This was because of the adequate bladder mobilisation within the pelvis and the use of flaps to cover the abdominal wall defect. The technique used most often for epispadias repair is the Cantwell–Ransley technique, or its modification, which was equally employed in our patient.⁷ Following epispadias repair, there may be a gradual improvement in the bladder capacity due to increased outlet resistance.

One of the most common complications after epispadias repair is early fistula formation, which occurs in up to 16% of patients who undergo epispadias repair, with spontaneous resolution in 4% by four months.⁷ Patients with combined bladder closure and epispadias repair have a higher fistula rate; our index patient developed a fistula after combined repair. However, the fistula resolved spontaneously in less than one month of non-operative management.

Bladder neck reconstruction is mostly performed at 4–5 years, with an excellent continence rate.⁷ The important determinants of success are the preoperative bladder capacity and motivation of the

patient and parents.⁷ Conversely, in CPRE, the urinary bladder and urethra are treated as a single unit and moved posteriorly into the pelvis. This is based on the concept that the primary defect of the bladder and cloaca is anterior herniation.¹⁰ This technique's urinary continence rate is comparable to staged repair and minimises the number of operative procedures required to achieve urinary continence. It also potentiates the bladder neck function and allows bladder cycling in the first year of life.¹⁰

Conclusion

With adequate bladder dissection and mobilisation deep within the pelvis, CPRE is feasible in adult exstrophy repair without the need for iliac osteotomy. Flaps can further aid in achieving abdominal wall closure. However, careful patient selection and surgeon experience are important determinants of a good outcome.

Conflict of interest

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