

Bladder Exstrophy: Modern Staged Repair Experience in our Institution

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Abstract

Introduction: Bladder exstrophy is a major malformation in paediatric urology. The treatment results are not still completely satisfactory, and their management is an enormous problem in Sub-Saharan Africa. While outlining our challenges, we report our management experience to improve our results. **Subjects and Methods:** We retrospectively reviewed the records of patients undergoing surgical repair of classic bladder exstrophy at our department between January 2010 and December 2019 (10 years). Epidemiological, clinical, therapeutic and evolution data were analysed. **Results:** Twenty-five children with classic bladder exstrophy were treated. Our series included 16 boys and 9 girls with a sex ratio of 1.7. Age ranged from 0 day to 6 years. Twenty-five bladder closures were performed, associated to pelvic osteotomy in 11 cases. Epispadias repair was performed on nine boys. Eight cases of bladder neck reconstruction and three cases of bladder enlargement were performed. We observed six bladder fistulas, four wound dehiscence, of which three partial, two parietal suppurations and six cases of urinary tract infection. Eight children had a continence of 1–2 h. **Conclusion:** The treatment of bladder exstrophy in our context is still limited because of financial difficulties encountered by the population and the insufficient technical platform in our country.

Keywords: Augmentation cystoplasty, bladder exstrophy, continence, primary closure

INTRODUCTION

Bladder exstrophy is a complex and severe congenital anomaly but fortunately rare. It appears once in 30.000–50.000 live births occurring about four times more often in boys than in girls.^[1] In Ivory Coast, the frequency is unknown. Management of bladder exstrophy remains challenging for paediatric surgeons and must resolve two problems: urinary and genital problems. The treatment includes several steps and functional results are better, compatible with a normal life at the cost of multiple surgeries. The purpose of this study is to expose difficulties of management to improve our results.

SUBJECTS AND METHODS

We retrospectively reviewed the records of patients undergoing surgical repair of classic bladder exstrophy at our department between January 2010 and December 2019 (10 years).

Pre-operative assessment was performed in all cases and included laboratory tests, abdominal and renal ultrasound, chest X-ray, spine and pelvis X-ray and echocardiography.

Surgical management was done by the same surgeon, and staged repair of bladder exstrophy was the method used in all cases. In our institution, bladder closure was carried out as soon as possible after the consultation. Osteotomies were realised when the basin was deemed to be rigid. Cantwell-Ransley and Mitchell techniques were used in epispadias repair and Young-Dees-Leadbetter technique in bladder neck reconstruction (BNR).

Post-operative assessment was clinical, radiologic, namely urinary tract ultrasound, retrograde cysto-urethrogram (RCU)

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and uraemia and serum creatinine. Urodynamics examinations were not done in our condition.

Continence, corresponding to the time between urination without urinary leakage, was evaluated. Continence was defined by dry during the day for a minimum of 3 h without stress and nocturnal incontinence. Partial continence is dryness between 1 and 3 h, a stressed urinary incontinence or moderate nocturnal incontinence. Considered as incontinent, those who had dryness for <1 h in a day, an incontinence at the least stress and nocturnal.^[2]

RESULTS

Twenty-five children with classic bladder exstrophy were treated, with a frequency of 2.5 cases/year. Sixteen boys and nine girls consulted at the paediatric surgery department for the first time. Sex ratio was 1:7. Age at consultation ranged from 0 day to 6 years.

None of our patients had an antenatal diagnosis. Fifty-two percent (13 patients) were referred to our institution before the 72nd h of life and 80% (20 patients) before the age of 1 month. Classic bladder exstrophy was found in all cases [Figure 1]. The presence of a polyp on the bladder plate was noted in three patients, one had two and the other three. All patients had pubic diastasis. The size of the bladder plate was specified in twenty cases. The mean size of the bladder plate during the consultation was 3.92 cm ranging from 2.5 cm to 5.5 cm.

Pelvis X-ray was performed in all patients and showed a pubic diastasis in all cases. Diastasis length ranged from 2 cm to 6 cm. Abdominal ultrasound was normal. Echocardiography was performed in 17 patients and was normal in 12 cases, showed a patent oval foramen in 2 cases, persistence of the arterial duct in 2 cases and a case of interventricular communication associated with a permeable oval foramen. However, all our patients had a good systolic ejection function. Various pathologies were associated with bladder exstrophy [Table 1]. Some children had several associated pathologies. Inguinal hernia was bilateral in six cases and unilateral in four cases.



Figure 1: Bladder plate seen at day 0 of life in a female newborn

Therapeutically, no patient was operated in the first 72 h of life. In 11 cases (44%), the bladder plate was closed before the 1st month of life [Figure 2]. Osteotomy was performed in 11 patients (44%) at the same time as the bladder closure. Vertical iliac osteotomy combined with bilateral innominate osteotomy was performed in five cases (45.45% of osteotomies), followed by bilateral innominate osteotomy in four cases and posterior iliac osteotomy in two cases. A bone immobilization with a Bachelor cast was performed systematically and maintained for 3–4 weeks.

Ten patients who presented inguinal hernia were operated, and the evolution was simple.

Epispadias repair was performed in 9 out of 16 male patients (56.25%). It was performed at an average age of 3.8 years with extremes of 1–7 years [Figure 3]. Cantwell-Ransley technique was used in six cases and Mitchell technique in three. The complications were a fistula in two cases which required an operative revision. Neither penile necrosis nor urethral stenosis was noted.

BNR was done in eight cases (32%). We used only the Young-Dees-Leadbetter technique.

Augmentation cystoplasty was performed between 5 and 10 years in four patients. It was an enterocystoplasty.

Mitrofanoff procedure or transappendicular continent cystostomy was performed to obtain continence in two cases.

Table 1: Distribution according to the associated pathologies

Associated pathologies	Number of case, n (%)
Inguinal hernia	10 (40)
Testicular ectopy	3 (18.75)
Omphalocele	2 (8)
Cardiac malformations	5 (20)
Clubfoot	3 (12)
Cleft lip and palate	1 (4)



Figure 2: Bladder closure in a female newborn

Three patients (12%) had vesicoureteral reflux (VUR). It was revealed by the RCU during post-operative follow-up. Bilateral Grade 2, 3 and 4 VUR was noted. Bilateral ureteral reimplantation was performed in those with Grade 3 and 4 VUR.

In the short term, bladder closure was favourable in 14 cases (56%). Complications were fistulae, wound dehiscence, suppuration and urinary tract infection [Table 2].

Several complications were found in the same patient. Fistula, parietal suppuration and urinary tract infections were treated with antibiotic therapy with favourable outcomes in all cases. The most found germ was *Escherichia coli* in 57.14% of cases. Wound dehiscence was repaired successfully, and fistulae resolved spontaneously.

Long-term outcome was enamelled by complications such as repeated urinary tract infections in five patients who each time required an ECBU and then an antibiotherapy with favourable evolution. Bladder stones were observed in two patients during post-operative monitoring by ultrasound.



Figure 3: Post-operative appearance after epispatias repair in a 1-year-old boy

Table 2: Distribution of complications of bladder closure	
Evolution	Number complications, n (%)
Favourable	14 (56)
Unfavourable	11 (44)
Fistula	6 (24)
Wound dehiscence	4 (16)
Wall suppuration	2 (8)
Urinary tract infection	6 (24)
Total	25 (100)

Table 3: Comparison of the complications of the bladder closure					
Series	Total number	Fistula, n (%)	Dehiscence, n (%)	Suppuration, n (%)	Urinary tract infection, n (%)
Schaeffer <i>et al.</i> ^[8]	194	2 (1)	6 (3)	3 (1.5)	6 (3)
Kibar <i>et al.</i> ^[9]	16	3 (18.75)	0	0	0
Our series	25	6 (24)	4 (16)	2 (8)	6 (24)

Eight patients in whom it was realised BNR were partially continent. The other patients were incontinent.

DISCUSSION

Bladder exstrophy is a severe urology malformation. The management must be early as possible and complete to obtain good results.

Associated malformations can be observed. Radiological investigations are intended to look for these malformations. In our study, several cardiac, orthopaedic and abdominal wall malformations were found. Some of these would condition the prognosis according to their gravity. Other malformations concerning the central nervous system (spina bifida, anencephaly and microcephaly) were found by Jayachandran^[3] but not in our study.

According to management, classic stage repair was used in all patients. Closing the plate must be quickly realized, before 72nd h, to give the best chance of success. Most authors are unanimous on the precocity of bladder closure this would allow a bladder closure without osteotomy due to the malleability of the pelvis, due to hormonal impregnation.^[4]

In a study in South Carolina (USA), STEC found an average age of bladder closure of 4.6 days, whereas MUSHTAQ in London (England) had an average age of 3 days with extremes of 1–152 days.^[5,6] In our series, no patient was operated in the first 72 h of life due to delayed consultation or care.

Pelvic osteotomy allows bringing the pubic bone and abdominal structure on the median line, protecting the closed bladder and posterior urethra. The importance of osteotomy associated to good immobilisation in bladder exstrophy treatment was proved by several authors, whereas in our study, it was realized on 44% of patients. Arlen *et al.*^[7] published about 15 patients who underwent bladder closure (10 staged and 5 complete repair) and 14 were immobilised with spica casts. Initial closure was successful in 11 cases (73%). Success rates were higher in patients undergoing osteotomy (6/7, 86%) compared to those without osteotomy (5/8, 63%). Among four patients who had presented a failure at initial closure, two underwent an osteotomy during the second closure and the two operations were successful.

Complications of bladder closure in our series included fistula, wound dehiscence, parietal suppuration and urinary tract infection. Several studies highlighted these complications which are represented in the table below [Table 3].

Our results could be due to poor caring of the bladder plate and the management delay, therefore increasing the risk of bladder plate infection. Bladder closure was performed without

osteotomy in 56% of cases. It leads to a bladder closure under tension that can explain the high percentage of fistula and wound dehiscence.

In fact, 11 patients who underwent osteotomy did not show present wound dehiscence, whereas 4 out of 14 patients who did not have an osteotomy (28.57%), presented it.

In total, osteotomy and effective immobilisation are many important factors in ensuring the best chance of success of bladder closure.^[10]

The second surgical time which is the repair of the epispadias was done at an average age of 3.8 years with 22, 22% of fistula. While BAIRD found 19 months in its study and 16% in 97 patients treated with the Cantwell-Ransley technique.^[10]

BNR is the last stage of classic exstrophy repair after epispadias repair. Results were considered acceptable in eight cases (24%) with partial continence. After BNR, Gargollo^[11] and Shaw^[2] obtained, respectively, 50% and 90% of continence in their series.

Augmentation cystoplasty was not systematic and was performed at the discretion of the surgeon based on inspection of the bladder at the time of reconstruction in our condition. It was realised in four cases associated with Mitrofanoff procedure to obtain continence.

This catheterise stoma of Mitrofanoff would result in a continence rate of 98%. But not used in our context due to a lack of catheter.

Life quality in this malformation is improved with prolonged follow-up and must be realised by multidisciplinary team (paediatric urologist, nephrologist, endocrinologist and psychologist).

In our series, the real problem was incontinence in 68% of cases. Our results are due to the fact that bladder neck reconstitution was done only on eight patients, and osteotomy realised on 56% of the patients, whereas the bladder closure was done after the 72nd h of life.

And financial difficulties did not allow an optimal care of these patients.

Urodynamics is in the learning stage in our country. It will contribute to improve the care of patients.

CONCLUSION

Surgical management of bladder exstrophy is extremely complex and requires multiple interventions. The success of initial bladder and posterior urethra closure is a factor determining the urinary continence and good bladder capacity. In our series, osteotomy, epispadias repair and bladder neck repair were not realized in all cases due to financial difficulties and patients lost to follow-up. This did not allow us to achieve one of the main purposes of the treatment, that is, continence.

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Conflicts of interest

There are no conflicts of interest.

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