Presentations of bladder exstrophy in a resource-limited setting and the role of Mainz II continent diversion for late referrals or failed primary closures: a multicentric report

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Key words: bladder exstrophy; urinary diversion; low resource contest.

Conflict of interest: the authors declare no potential conflict of interest, and all authors confirm accuracy.

Ethics approval: not applicable. The study was retrospective and conducted on anonymous hospital files. The study is conformed with the Helsinki Declaration of 1964, as revised in 2013, concerning human and animal rights.

Informed consent: all patients participating in this study signed a written informed consent before treatment according to the rules of the Country.

Patient consent for publication: not applicable. The study was retrospective and conducted on anonymous hospital files.

Availability of data and materials: all data generated or analyzed during this study are included in this published article.

Received: 11 May 2023.
Accepted: 2 August 2023.

La Pediatria Medica e Chirurgica 2023; 45:323
doi:10.4081/pmc.2023.323

Abstract

Primary closure techniques that have been updated and long-term follow-up for CBE (classic bladder exstrophy) may be out of reach for many patients living in resource-limited settings. Late referrals to medical care and primary closures that lack the necessary skills and facilities for comprehensive treatment are still common. Alternative and long-term surgical solutions may improve the lives of these unfortunate patients. During surgical outreach missions, patients with CBE, either non-operated or with a previous unsuccessful bladder closure, who were referred from vast under-resourced rural areas to three Eastern African hospitals, were studied. The following information is provided: mode of presentation, clinical history, diagnostic workout, management, and outcome. There were 25 cases (M/F ratio 17/8) ranging in age from two days to twenty years. Five of the seventeen patients who were not treated (35%) were under 120 days old and eligible for primary closure in a qualified tertiary center when one was available in the country. There were twelve late referred cases (ranging from 120 days to 20 years). Between the ages of ten months and twelve years, eight children arrived following a failed primary closure. In all of them, the bladder plate was too altered to allow closure. Following a preoperative diagnostic workout, a Mainz II continent internal diversion was proposed to fourteen patients with acceptable bowel control and postponed in the other three. Three cases were lost before treatment because parents refused the procedure. Twelve cases ranging in age from three to twenty years (mean seven years) were operated on. Eight people were followed for a total of 53.87 months (range: 36-120). Except for three people who complained of occasional night soiling, day and night continence were good. The average voiding frequency during the day was four and 1.3 at night. There was no evidence of a metabolic imbalance, urinary infection, or significant upper urinary tract dilatation. Two fatalities could not be linked to urinary diversion. Four patients were not followed up on. Due to the limited number of specialist surgical facilities, CBE late referral or failed closure is to be expected in a resource-limited context. In lieu of the primary closure, a continent internal diversion will be proposed and encouraged even at the level of a non-specialist hospital to improve the quality of life of these unfortunate patients. It is

recommended that patients be warned about the procedure's potential long-term risks, which will necessitate a limited but regular follow-up.

Introduction

Bladder exstrophy and epispadias complex (BEEC) is an anterior midline defect with a variable grade expression that involves the infra-umbilical abdominal wall, including the pelvis, urinary tract, and external genitalia.[1] The prevalence of EEC at birth is 1/10,000. Epispadias (E), classic bladder exstrophy (CEB), and cloacal exstrophy (EC) are clinical variants within the same spectrum. Most studies report a male-to-female ratio of around 2.4-1, but a ratio as high as 6-1 has also been reported. CEB is evident from birth, with the bladder mucosa visible in the lower abdomen and urine dripping from the ureteric orifices on the bladder surface. Diastasis of pubic bones and recti muscles with inguinal hernias are associated. In males, the penis is short and broad with a dorsal chordee. The urethral plate covers the dorsum of the penis from the bladder to the glans, and both corpora cavernosa lie beneath it. Ejaculatory duct openings are visible in the colliculus seminalis, and the prostate is located dorsally. Female patients have a bifid clitoris next to an open urethral plate. The vagina and the anus are anteriorly displaced, and the perineum is shortened. Spinal anomalies are associated with about 7% of cases.

Different surgical techniques have been proposed to close the abdominal and bladder wall, restore a continent reservoir and preserve sexual and reproductive function.[2] Today neonates with CBE may expect a better chance of achieving dryness, good cosmetic appearance, and an acceptable quality of life, even under the sexual aspect, after an early (0–3 days) or delayed (4–120 days) closure by a single or staged procedure, associated or not to pelvic osteotomy.[3] Primary closures far beyond this deadline have been published, but it is hard to conclude for a practical benefit.[4] A higher complication rate associated with delayed closure has also been reported.[5] Best outcomes are still reserved for centers having a large caseload, a specific experience, and a lifelong care program, including additional surgery to optimize bladder storage and emptying or improve genital and reproductive function. Monitoring of upper urinary tract and bladder function, psychosocial and psychosexual support, and follow-up by a multidisciplinary team is strictly needed from birth into adulthood.[5] The risk of bladder wall dehiscence, symphysis, and neck reopening[6-8] is high whenever a primary closure is attempted in under-resourced health facilities. Patients may also be lost from follow-up and cannot benefit from secondary procedures to increase bladder compliance, restore continence, or protect against upper tract deterioration from a small, high-pressure bladder. Data about functional outcomes and long-term sequelae may be missed.[10-12]

In rural Sub-Saharan areas,[6,7] many deliveries still occur outside health centers, and patients with a CBE may be referred late to medical attention, sometimes in adult life. Successful primary bladder closure reports are increasing from African tertiary centers, although the scarcity of facilities limits access to appropriate surgery compared to the great demand for specialist Healthcare. Nevertheless, even after a successful primary closure, complementary procedures (augmentation, bladder neck reconstruction, stomas, catheterization) may not be affordable or not culturally acceptable for people living in distant rural areas.[13]

Children experimenting with unsuccessful primary closure present a shrunken and profoundly altered bladder plate. The only option for them and the late referrals is removing the bladder plate and confection of a continent urinary diversion.[14,15] Quality of life is improved, although the impact of these procedures on their lives, including long-term risks, cannot be underestimated.[16,17]

The present report concerns CBE cases observed in three Health Centres in Eastern Africa, each with a large rural catchment area, during regular outreach missions aiming to upgrade the standard of local pediatric surgical services. The experience with the "Mainz II rectosigmoid Pouch" continent urinary diversion[18,19] as an alternative to primary closure for late referrals and surgical failures is discussed.

Materials and Methods

Study setting

The study involved three referral Hospitals with a large rural catchment area: the Orotta National Referral Hospital (ONRH) of Asmara, Eritrea, the Consolata Regional Referral Hospital Ikonda (CHI), Njombe, in the Southern Highlands of Tanzania, and the Gezira National Centre for Pediatric Surgery (GNCPS) of Wad Medani, Sudan.

Only the GNCPS has an established specialist Pediatric Surgical capacity, but no neonatal intensive care facilities. Referrals to a higher level of specialist care in the Country for CBE are granted from CHI and GNCPS only.

Study population

The present study was based on patients with a CBE who had either not been treated since birth or had been submitted to an unsuccessful primary bladder closure in the same institution. They were observed during 43 weeks of outreach visits to the ONRH (2009-2012), 27 weeks to the CHI (2016 to date), and 36 weeks to the GNCPS (2017 to date).

Data collection

The presentation mode, gender, age on admission, associated abnormalities, previous surgery, local conditions, and management were recorded.

Diagnostic workout

All patients had a complete renal and metabolic assessment on admission (BUN, Creatinine, Electrolytes, Urinalysis) and Ultrasound (US) investigation of the upper urinary tract.

Surgical management

Internal urinary diversion (Mainz II Pouch) was decided according to age at referral, previous surgery, size and aspect of bladder plate, presence of associated abnormalities, acquired bowel control, and family compliance to surgery. The repair of male external genitalia by the penile disassembly technique, according to Mitchell and Bagley, was always considered a second-step procedure.

Follow-up protocol

Families of operated cases were made aware of the need for periodic control after surgery (1 month, six months, and every year after discharge) and referral to the Hospital in case of high temperature, persistent vomiting, and eye and ankle swelling. Follow-up records included metabolic and renal function assessment, quality and amount of urine discharge, and US imaging of the upper urinary tract.

Results

Case description and epidemiology

Twenty-five patients with a CBE were observed during a total of not consecutive 103 weeks of outreach missions. Table 1 reports the mode of presentation, male-female ratios, mean age at referral, and associated abnormalities of our cases.

Management

Referral to a tertiary center of the Country with neonatal intensive facilities for early primary reconstructions and comprehensive management was proposed, when possible, to parents of five cases seen within the first 120 days of life (29%; Table 2). In addition, bladder reconstruction was excluded for twelve late referred and unoperated cases (ages between seven months and 20 years) and eight surgical failures (ages between 3 and 8 years). A small and shrunken bladder plate was observed in all late referrals. Concerning failures, associated orthopedic procedures and immobilization recommended for reclosures[20] were not compatible with the local context. The high risk of eventual incontinence also made diversion the preferred option.

After a preoperative assessment, Mainz II internal continent diversion was confectioned in twelve cases (eight males and four females) ranging from three to twenty years of age (mean 92.72 months, median 84, SD 52.703). The procedures were all performed under the 1964 Helsinki Declaration and later amendments or comparable ethical standards by local surgeons, assisted by the outreach team.

An internal continent diversion was postponed in three patients still under one year of age (one male and two females) until complete bowel control was achieved. Families of three male patients (between five and eight years of age) refused an internal diversion, and two (one eight years and one 12 twelve years males) were lost before surgery.

Surgical procedure

The diversion was made as described by Fisch[18] following bowel preparation and antibiotic prophylaxis.

After stenting and isolating both ureters, the bladder mucosa was meticulously excised from the detrusor muscle, which was left to fill the gap between the recti muscles. The abdomen was entered through a transverse incision two cm above the bladder plate edge. The distal sigmoid and the rectum were approached and fixed each other with stay sutures, and an inverse U shape 15-20 cm incision was made on their antimesenteric side. The posterior flaps of the incision were sutured. Both ureters were mobilized and reimplanted on the posterior pouch wall through a submucosal tunnel. The ureter was trimmed and spatulated before being fixed to the bowel mucosa with 5/0 polyglactin-interrupted sutures. Two 6-9 Fr. Stents were positioned into the ureters, fixed to the bowel mucosa by absorbable sutures, and passed through the anus to be anchored separately to the skin of the inner face of the thigh and removed after ten days. A double-layer suture of the anterior flap of the rectosigmoid incision completed the closure of the pouch. The abdominal wall was sutured in layers, and rotation skin flaps covered the demucosed bladder plate's muscular layer.

Follow-up and outcome

All patients were discharged after a postoperative stay between 15 and 20 days (Table 3). Surgical complications were limited to two wound infections and one minor wound dehiscence, which needed to be sutured. A transient mild acidosis was found only in a few cases, but no bicarbonate supplementation was requested. The US of the upper urinary tract excluded post-reimplant significant dilatations. A regular follow-up schedule on an annual base was attended by four patients only. Four other cases had to be contacted and came irregularly even after a 2-3 years delay. Four patients were lost from follow-up (33%) since discharge from the Hospital. The mean follow-up time, when available, was 53.87

Table 1. Mode of presentation and demographics of patients con CBE.

Cases with CBE	No.	m/f	Age range on admission	
Total	25	17/8	2 days - 20 years mean 60.4 months, median 60, SD 55.994	
Not operated before	17 (68%)	11/6	2 days - 20 years mean 59 months, median 60, SD 61.301	Birth -120 days: 5 (29%) >120 day - 4 yrs: 4 (24%) 6 yrs - 9 yrs: 6 (35%) 10 yrs - 20 yrs: 2 (12%)
Previous primary closure (failed)	8 (32%)	6/2	10 months - 12 years mean 63.5 months, median 60, SD 42.411	

Table 2. Management of patients with CBE.

Cases with CBE	No	Management
Previous primary closure (failed)	8	4 Mainz II Pouch 1 Diversion postponed achieving bowel control 2 Treatment refused by parents 1 Lost from F.U. before surgery
<120 days of life Not operated	5	Referral to a tertiary centre for primary closure proposed
>120 days of life Not operated	12	8 Mainz II Pouch 3 Diversions postponed achieving bowel control 1 Treatment refused by parents 1 Lost from F.U. before surgery

months (range 36-120, median 44, SD 28.0532). Two deaths were registered at three and four years after surgery. In both cases, the Hospital was informed by the parents after they were called to schedule control. The cause of death was malaria in one and unexplained in the other. Quality of life was pretty acceptable for all our cases that could be followed. All acquired daily continence in one-two months after diversion, and micturition frequency varied from five to three (mean four). Night continence was good in five, but three patients experienced occasional soiling. The mean night micturition frequency was 1.3.

Discussion

Management of CBE is a real challenge for pediatric surgeons. Open questions remain about detrusor development and function after primary closure and continence. There is still a debate if an early or deferred approach must be preferred and about the type of pelvic osteotomy if required.[21] Results from different published series[22,23] are sometimes hard to compare. All these procedures require a meticulous surgical technique, postoperative short and long-term care, and close follow-up looking at the need for additional surgery.[24] A comprehensive follow-up into adult life, radiological and urodynamic assessment, and psychological support should be included.[25] Primary closure is unethical whenever all specialist facilities are not available. Trying to close the bladder without all the required skills and support exposes one to a high risk of dehiscence and failure and a poor functional outcome even after a successful closure.[26] The perspectives for an infant with a CBE born in rural areas of some LIC may be so far dramatic.[27]

Reports from Sub-Saharan African tertiary centers about a successful surgical approach to CBE by primary closure[28-33] still represent a minority considering the large number of children from LIC. Many do not easily access specialist pediatric surgical care.[34] Frustration is manifested in the number of late referred patients, high failure rate, and frequently missed long-term follow. These children also need reconstructive surgery of external genitalia, by experienced hands, as a second step. Length of corpora and dorsal chordee is a real challenge in giving male genitalia a normal appearance and guaranteeing sexuality and paternity. Upgrading the CBE surgical standard of care in LIC toward successful early primary closure and providing universal access to health facilities for multidisciplinary postoperative care and follow-up may be a long-term achievement. However, sporadic self-contained international volunteer missions to treat a few CBE patients are not a solution. They often do not transfer knowledge and expertise, and the lack of long-term follow-up often limits their results.[35,36]

Therefore, a continent urinary diversion must be proposed for the many late referred cases[37,38] or when primary closure fails. The Mainz II sigmoid pouch[18] is the most frequently adopted approach. Good continence is achieved in more than 90%. Metabolic acidosis is a possible complication in 30%, to be corrected by oral Bicarbonate. Stenosis of the implanted ureter occurs in 2% and pyelonephritis in 14%.[39-42] Long-term risks of colonic cancer[16,17] are rated at 9.8% in a recent review[43] 15 to 48 years after the diversion (median 34 years) and may require a periodic rectal endoscopic assessment. Mainz II pouch is a frequently adopted alternative for primary closure of CBE in most African Centres.[44-46] Children must achieve bowel control before surgery, and the procedure must be explained to families. By the time patients get accustomed to passing urine and stools through the rectum without problems. Male patients live this experience with more problems than females, which may explain the refusal of the proposed procedure by the families of three of our male patients. Our series confirmed the favorable postoperative course in diverted cases. Night continence was a problem for three of them (two were under four years of age). We have no evidence that two late fatal cases registered were related to the urinary diversion. Cases lost from follow-up were a third of our series, which can reasonably be expected in vast rural areas where poverty and lack of transport make it difficult to comply with medical prescriptions. The same reasons were at the origin of the irregular attendance to follow-up by some patients.

Conclusions

According to our experience, the Mainz II pouch requires few resources and can be successfully performed by trained general surgeons outside a specialist tertiary center. Diversion produces a better quality of life for several CBE patients not eligible for pri-

Table 3. Follow-up and outcome of twelve CBE after Main II Pouch continent diversion.

	Gender	Previous surgery	Age at diversion	Complications	Follow up (months)	Night continence	Night micturition	Day continence	Day micturition	US upper urinary tract	Renal function	Outcome
1	F	Yes	3			Lost from follow up						
2	F		3		38	Occasional soiling	2	Good	5	Not dilated	Normal	Dead (Malaria)
3	M	Yes	4		38	Occasional soiling	2	Good	5	Not dilated	Normal	
4	F		5	Wound dehiscence, (suture)	120	Good	1	Good	5	Mild dilatation	Normal	
5	F	Yes	5		38	Good	1	Good	3	Not dilated	Normal	
6	M		6		60	Good	1	Good	4	Not dilated	Normal	
7	M		7		51	Good	1	Good	4	Not dilated	Normal	Dead (?)
8	M		8			Lost from follow up						
9	M	Yes	8		50	Occasional soiling	2	Good	3	Not dilated	Normal	
10	M		9	Wound infection		Lost from follow up						
11	M		10	Wound infection		Lost from follow up						
12	M		20		36	Good	1	Good	3	Not dilated	Normal	

mary closure. Nevertheless, early referral for appropriate primary treatment is recommended for cases of CBE seen in the first four months of life, but exclusively when access to a tertiary qualified specialist Centre is granted. The recourse to a continent internal diversion in low resources contexts, even at a non-specialist hospital level, must be recommended in all other cases. Nevertheless, local surgeons must be aware of the risks related to this procedure. Periodic laboratory monitoring of metabolic acidosis, urinary tract infections, and pyelonephritis, the US of the upper urinary tract for distention, and endoscopy for secondary rectal cancer must never be omitted.

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