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Our initial experience with the technique of complete primary repair for bladder exstrophy

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Abstract *Objective:* We reviewed our initial results with complete primary repair of exstrophy in regard to continence status and the need for subsequent continence procedures.

Patients and methods: We performed a retrospective review of our surgical records from 1996 to 2008 to identify all patients with bladder exstrophy managed at our center.

Results: Sixteen children were closed successfully. Six patients (37.5%) experienced complications: umbilical hernias in two, transient penopubic fistula in three, and subcoronal fistula due to meatal stenosis in one. Of the 12 males, seven (58.3%) were left with a hypospadias at the time of primary closure. Two (22.2%) children required a formal bladder neck reconstruction to achieve continence. Bladder augmentation and continent catheterizable stoma was performed in four cases (44.4%), and bladder neck injection in one case (11.1%). Bladder neck closure was also performed in another child following primary closure. Three of these children are continent and void spontaneously (33.3%). The remaining six require clean intermittent catheterization four to six times a day, resulting in four (44.4%) being continent. The number of continence procedures and mean number per patient were 15 and 1.66, respectively.

Conclusion: Our early experience with this technique has been encouraging, with few major complications, a highly successful closure rate and a cosmetically normal result.

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Attainment of urinary continence and preservation of renal function are the primary objectives of complex reconstructive surgeries in patients born with bladder exstrophy. Improvements in urinary continence rates in such children have been achieved through the development of two well

described techniques: modern staged repair (MSR) [1] and complete primary repair of bladder exstrophy (CPRE) [2]. Controversy regarding the optimal treatment of exstrophy begins in the newborn period and centers on whether initial surgery should be limited to the bladder, posterior urethra and abdominal wall closure (MSR), or bladder closure and epispadias repair by total penile disassembly in one stage (CPRE).

In the last few years there has been considerable interest in CPRE with various series published regarding

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surgical outcomes. Postoperative issues recently evaluated in the literature include the need for ureteral reimplantation in the first year of life, the number of patients requiring hypospadias repair after this procedure, and the complication rates of this operation [3,4]. However, little has been published about the need for subsequent continence procedures in this group of patients. We reviewed our initial results with CPRE in regard to continence status and the need for subsequent continence procedures. Pertinent clinical features and continence outcomes are discussed.

Patients and methods

We performed an IRB-approved retrospective review of our surgical records from 1996 to 2008 to identify all patients with bladder exstrophy managed at our center. Clinical records were reviewed to determine patient characteristics and immediate surgical outcome of exstrophy closure, and results of subsequent secondary procedures to achieve continence following CPRE were carefully detailed. The techniques for CPRE have been described in detail elsewhere [5]. Bladder neck reconstruction (BNR) was performed using the Young–Dees–Leadbetter technique [6].

Continence was determined by patient and/or caregiver interviews, as documented in charts by the health-care provider, to assess the presence or absence of leakage. We defined continence as a dry interval of 4 h and no nighttime leakage. Patients were considered incontinent if they experienced any urinary leakage between catheterizations from either the catheterizable channel or the urethra.

Results

All 16 patients were closed by one fellowship-trained pediatric urologist (BK) at our center. Of the 16 patients, 11 underwent this operation without osteotomies within the first 72 h of life. Five children underwent repair with anterior iliac osteotomies between the ages of 1 week and 17 months. All of the children were immobilized in modified Bryan's traction for 1 week after the operation and then placed in a spica cast.

All 16 patients were successfully closed. Six patients (37.5%) experienced complications including: umbilical hernias in two, transient penopubic fistula in three (one with concomitant urethral stricture) and subcoronal fistula due to meatal stenosis in one child. The hernias and the urethral stricture were repaired successfully and the penopubic fistulae resolved spontaneously.

Nine children have reached the age of continence. Two of them (22.2%) required a subsequent formal BNR to become continent and neither required redo BNR. Bilateral ureteral reimplantation was performed following primary closure in five children (55.5%). The reasons for reimplantation were encountering breakthrough UTI despite suppressive antibiotic therapy after closure in one case, and/or performing this operation at the time of augmentation or BNR procedure as a part of these operations after closure in four cases. Subsequent bladder augmentations and catheterizable stoma were performed in the same four (44.4%) children. One child (11.1%) underwent bladder neck

injection and another (11.1%) bladder neck closure to become continent (Table 1). The reason for bladder neck closure was urethral failure, because significant scarring and fibrosis around the bladder neck provided little intrinsic urethral resistance.

Of the 12 males, seven (58.3%) were left with a hypospadias following penile complete penile disassembly at the time of primary closure. Four children, all with penoscrotal hypospadias, have undergone successful hypospadias repair. Three additional boys with penoscrotal hypospadias are awaiting definitive repair.

To determine the continence rate we included only the nine children older than 5 years. Three of these children are continent and void spontaneously (33.3%). These three void at least at 4-h intervals and are dry day and night. The remaining six require CIC four to six times a day. They use CIC per urethra or continent stomas because they were augmented. Four out of six are continent using CIC (44.4%). The other two are incontinent (22.2%) with dry intervals for 1–2 h and urine leakage between CICs (Table 2). A total of 15 continence procedures were performed in nine patients undergoing CPRE with a mean number of 1.66 per patient (Table 3).

Six patients are currently younger than 5 years and are therefore not evaluated for continence. Eventual urinary diversion in the form of a vesicostomy was required in two patients secondary to the hydronephrosis. These children had very small bladder templates at the time of closure, and small and high-pressure bladders developed subsequently which required vesicostomy. The other four void spontaneously into a diaper and their families have confirmed the presence of a strong urinary stream during spontaneous voids. These children are maintained on CIC for observation of upper urinary tract dilatation. One child underwent successful CPRE but was lost to follow up (Table 1).

Discussion

The management of children with bladder exstrophy remains controversial. Today successful initial closure without immediate severe complications like bladder prolapse or dehiscence can be expected in most patients treated in specialized centers. Fundamental principles of placing the posterior urethra and bladder deep into the pelvis in combination with a tension-free closure and adequate postoperative management should prevent these devastating early complications [5]. But the achievement of later urinary continence is still challenging and the success rates vary widely between institutions and surgical techniques used.

The inability to obtain universally dependable, reproducible results, and psychological trauma of multiple failed attempts in children with bladder exstrophy led some authors to explore alternative techniques for bladder closure [4,5,7]. The same principles of MSR, such as proper positioning of the bladder neck in the pelvis and the proximal urethra within the pelvic diaphragm, are applied in CPRE. But instead of leaving the bladder neck open to drainage, the bladder neck is tightened and the urethra reconstructed. Historically, initial attempts at CPRE were fraught with upper urinary tract deterioration due to

Table 1 Patient characteristics and results of operation with CPRE.

Case no.	Gender	DOB	Ostet.	BNR	Reimpl.	Augment.	CS	BNC	HS	Results	
Patients completing CPRE											
1	M	2002			+	+	+		+	Continent (CIC)	
2	F	2002	+	+	+	+	+			Continent (CIC)	
3	F	2001			+	+	+			Continent (CIC)	
4	M	1998	+							Continent (VS)	
5	M	1998	+							Continent (VS)	
6	F	1996			+	+	+	+		Continent (CIC)	
7	F	2001								Incontinent	
8	M	2002							+	Incontinent	
9	M	2002		+	+				+	Continent (VS)	
Patients with successful CPRE but under 5 years											
10	M	2007							+	Continent (VS, D)	
11	M	2006							AHS	Continent (VS, D)	
12	M	2008								Continent (VS, D)	
13	M	2006			+					Continent (VS, D)	
Patients with successful CPRE but lost to follow up											
14	M	1999	+						AHS	Continent (CIC)	
Patients awaiting completion of CPRE											
15	M	2003	+	with vesicostomy, meatus in penoscrotal junction							
16	M	2004		with vesicostomy							

DOB: date of birth, Ostet.: osteotomy, BNR: bladder neck reconstruction, Reimpl.: reimplantation, Augment.: augmentation, CS: catheterizable stoma, BNC: bladder neck closure, HS: hypospadias surgery, AHS: awaiting hypospadias surgery, CIC: clean intermittent catheterization, VS: voids spontaneously, VS, D: voids spontaneously into a diaper.

bladder neck obstruction [5]. Grady and Mitchell modified the CPRE technique to exclude formal BNR at the time of initial closure; rather, the bladder neck is approximated and placed deep in the pelvis, with anterior closure of the pelvic diaphragm and epispadias repair [7]. There are theoretical benefits of performing epispadias repair at the time of exstrophy closure, because urethroplasty increases urethral resistance and restores bladder cycling, which results in the expansion of even very small exstrophic bladder plates [8,9].

Using this technique, the incidence of hypospadias is 30–70% and many of these patients will require later surgical correction [2,4,8]. Hypospadias results because the inherently short urethra found in exstrophy does not reach the glans once the bladder, bladder neck and urethra are moved deep into the pelvis. Numerous modifications have been applied to prevent the occurrence of hypospadias, including suturing of the urethral plate with interrupted sutures, augmenting the urethral plate using preputial or paramental skin flaps, and modification of the total penile disassembly technique by use of either a cavernostomy or limited penile disassembly to achieve a glandular meatus [10,11]. Others have used standard hypospadias repair techniques to correct this condition. In this study, complete penile disassembly resulted in hypospadias in 7/12 boys (58.3%).

Of more concern than the frequent presence of hypospadias are the reports of ischemic loss of the glans or corporal bodies. The exact incidence of ischemic changes is currently unknown; however, in one study ischemia of the glans and penile skin was reported in 12% of patients, and sloughing or loss of a hemiglans in 5% [12,13]. The etiology

of glandular or corporal loss may be part of the learning curve for the procedure. In our series we did not encounter such complications.

Urethrocuteaneous fistula formation occurs most commonly at the penopubic junction. In delayed cases or after initial failure the CPRE technique has resulted in fistula formation in up to 50% of cases, of which all closed spontaneously [4]. We saw penopubic fistula in three cases (18.7%) which resolved spontaneously.

While increased outlet resistance may allow for an increase in bladder capacity, a recognized consequence is elevated bladder pressure which may lead to upper tract changes. The incidence of progressive or severe hydronephrosis and/or renal scarring has been reported to range from 0% to 30% after CPRE [7]. Later surgical repair of the VUR is reported to be necessary in 0–50% of patients [7]. In our series, we encountered two children with progressive hydronephrosis (13.3%) who required a vesicostomy to alleviate high intravesical pressures.

The incidence of urinary continence using this surgical approach is not available from a large number of institutions. In the published series, the reported initial success

Table 2 Continence rate of nine children more than 5 years old.

Operation	Type of voiding	Continent
CPRE alone	Spontaneous void	2 (22.2%)
CPRE + BNR	Spontaneous void	1 (11.1%)
CPRE + adjuvant procedure	CIC	4 (44.4%)

Table 3 Adjuvant procedures performed in nine patients completing CPRE.

BNR	2 (22.2%)
Augmentation	4 (44.4%)
Catheterizable stoma	4 (44.4%)
Bladder neck injection	1 (11.1%)
Bladder neck closure	1 (11.1%)
Hypospadias surgery	3 (33.3%)

rate (defined as volitional continence without the aid of a modified BNR or augmentation) ranges widely from 22% to 65% [8,10,12]. Complete urinary continence will eventually be obtained in 70–90% of patients. However, a modified BNR is reported to be necessary in 15–90% of patients, and 5–10% will require a bladder augmentation [7]. In the current series nine children who underwent CPR were older than 5 years and available for evaluation of continence. Three of these children are continent and void spontaneously (33.3%). The remaining six require CIC, and two of them are still incontinent (22.2%).

In conclusion, our early experience with modest patient numbers has been encouraging, with few major complications, a highly successful closure rate and achievement of a cosmetically normal result with this technique.

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