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Short-term outcomes of the multi-institutional bladder exstrophy consortium: Successes and complications in the first two years of collaboration☆

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Summary

Introduction/background
Bladder exstrophy is a rare diagnosis that presents major reconstructive challenges. To increase experience and proficiency in the care of bladder exstrophy (BE), the Multi-Institutional BE Consortium (MIBEC) was formed, with a focus on refining technical aspects of complete primary repair of bladder exstrophy (CPRE) and subsequent care.

Objective
Outcome measures included successful CPRE (absence of dehiscence), complications, and integrated points of technique and care over the short-term.

Study design
Boston Children's Hospital, Children's Hospital of Philadelphia and Children's Hospital of Wisconsin alternately served as the host, with observation, commentary and critique by visiting collaborating surgeons. CPRE with bilateral iliac osteotomy was performed at 1–3 months of age. High-definition video capture of the surgery allowed local and distant broadcast to facilitate real-time observation and teaching, and recording of all procedures.

Results
From February 2013 to February 2015, MIBEC participating surgeons performed CPRE on 27 consecutive patients (22 classic BE, five epispadias). There were no dehiscences in 27 patients (0%, 95% CI 0–12.5%). Thirteen girls and 14 boys underwent CPRE at a median age of 2.3 months (range 0.1–51.6). One boy had a hypospadiac urethral meatus at CPRE completion. Hydronephrosis of mild or moderate grade was present postoperatively in eight girls and two boys. Additional results, per gender, are presented in the Summary table below.

Discussion
Absence of dehiscence in this cohort was comparable or compared favorably with the literature. However, several girls had significant obstructive complications following CPRE. The rate of bladder outlet obstruction (BOO) in girls was increased compared with published reports. A low complication rate was noted in the boys following CPRE, which was comparable to reports in the literature, and early signs of continence and spontaneous voiding were noted in some boys and girls. Limitations included variation in patient age at presentation, thereby introducing a wide age range at CPRE. Outcome data were limited by short follow-up regarding voiding with continence.

Conclusion
This collaborative effort proved beneficial regarding significantly increased surgeon exposure to CPRE, refinement of CPRE technique, surgeon learning and expertise. Technical refinement of CPRE is ongoing.

Summary Table Diagnosis, timing of CPRE and complications per gender.

Gender	Girls	Boys
Classic BE/epispadias	10/3	12/2
Median age at CPRE (range)	1.9 months (0.1–51.6)	2.9 months (0.4–28.8)
Successes	No dehiscence 7 without complications	No dehiscence 11 without complication
Complications	6 girls total 5 pyelonephritis 4 urinary retention (BOO) 2 temporary CIC 1 vesicostomy 1 bladder rupture	3 boys total 1 pyelonephritis 2 urethrocutaneous fistula

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Introduction

Bladder exstrophy is a rare diagnosis that presents complex anatomical and reconstructive challenges. Goals of surgery are to recreate normal anatomical and physiologic bladder, bladder neck and urethra for purposes of maintaining upper urinary tract health, urinary continence, and sexual function and cosmesis. Grady and Mitchell first described the complete primary repair of bladder exstrophy (CPRE) procedure in 1999 [1]. Outcomes following CPRE have been reported by several authors [2—6].

Bladder capacity following CPRE in the newborn has been reported as comparable to that after staged repair [7], and in comparison of patients undergoing neonatal bladder closure versus closure between 1 and 9 months of age, there is no statistically significant difference in eventual bladder capacity [8].

Although it has been well established that CPRE is safe, efficacious, and the standard of care at many institutions, there is still substantial variability in care and associated costs. Practice patterns and resource utilization across the United States has been reported [9]. Nelson and colleagues described improved clinical outcomes, lower hospitalization costs, and lower morbidity and mortality rates when patients were treated at hospitals that treat more than five exstrophy cases annually [10].

In an effort to refine and optimize CPRE technique, and overall patient care and outcomes, the Multi-Institutional BE Consortium (MIBEC) was formed. Preliminary findings demonstrated feasibility and increased exposure afforded to participating surgeons [11].

The present study hypothesized that collaboration within the MIBEC would: (1) lead to changes and improvement in surgical technique; (2) streamline postoperative care; and (3) in long-term follow-up would result in comparable or improved outcomes relative to previously published reports.

Materials and methods

The MIBEC consisted of pediatric urologic surgeons with a specific interest and dedication to the care of patients with BE enlisted from Boston Children's Hospital, Children's Hospital of Philadelphia and Children's Hospital of Wisconsin. One of the three institutions alternately served as the host site for scheduled surgery, with observation, commentary and critique by visiting surgeons. Visiting surgeons did not perform any of the surgery at the host site.

Pre-operative evaluation for patients with BE consisted of renal ultrasound (RUS) and plain radiograph of the abdomen to measure pubic diastasis. For patients with epispadias, evaluation included RUS (including bladder), VCUG, cystourethroscopy, and urodynamic study (UDS). Pre-operative preparation for the surgeon(s) included CPRE video review.

The technique of choice, at all institutions, for initial BE reconstruction was CPRE [1,11]. CPRE with bilateral iliac osteotomy was performed at 1—3 months of age in consecutive patients diagnosed with BE at participating institutions. Indications for CPRE in patients with epispadias included female gender, and in boys, a penopublic defect with wide pubic diastasis and patulous bladder neck on VCUG and cystourethroscopy.

All procedures were recorded on high-definition video. The high-definition video was also broadcasted live in both closed circuit and distant live broadcast (when a collaborating attending surgeon could not physically attend the host site surgeries) in order to facilitate real-time observation and teaching. Technical details of the surgery were discussed real time and recorded for analysis, retrospective review, and critique. Videos were edited to produce streamlined playback images of each surgery and these videos were used for immediate pre-operative preparation.

Patients were followed prospectively. Outcome measures included successful CPRE (absence of dehiscence), complications, and integrated points of technique and care over the short term. All patients underwent postoperative RUS and VCUG. The Wilson score interval for dehiscence in 27 subjects was calculated [12]. UDS with simultaneous cystometrography (CMG) and needle electrode electromyography (EMG) of the external urethral sphincter muscle complex was performed in two girls with urinary retention or BOO after CPRE. The standardized technique for UDS has previously been reported [13].

During the course of the MIBEC, a common postoperative protocol for patient management and evaluation was developed. Case report forms (CRF) for pre-operative, intraoperative and postoperative details of care were used to record data. As an example, the intraoperative CRF was used to record measurements of the exstrophied bladder (width, length and depth), urethra and genitalia. All pertinent technical details including, but not limited to, final bladder neck width, and suture type, size and technique (running, interrupted) were documented. Revisions and alteration of major points technique were achieved at several venues, including: (1) on-site meetings prior to each surgery, (2) intraoperative discussion, (3) review of operative video, and (4) biannual dedicated half-day agenda-driven meetings.

Results

From February 2013 to February 2015, MIBEC participants performed CPRE on 27 consecutive patients (22 classic BE, five epispadias). Thirteen girls and 14 boys underwent CPRE at a median age of 2.3 months (range 0.1—51.6). Iliac osteotomy was performed via either anterior or posterior approach, given each institutions extensive orthopedic experience favoring a specific method. Patients were immobilized for 4—8 weeks following bilateral iliac osteotomy and CPRE with either spica cast or modified Bryant's traction. Spica cast immobilization became the preferred technique at all institutions. Median follow-up for the group as a whole was 15.6 months (16.5 months for boys, 14.8 months for girls).

There were no dehiscences in 27 patients (0%, 95% CI 0—12.5%). Seventeen patients were without complications, whereas 10 suffered one or more complications. Pyelonephritis of one or more episodes occurred in five girls and one boy. Eight patients had a DMSA scan after CPRE within the time period reported herein. Three of eight (38%) had cortical defects identified; all were girls, had recurrent