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## Achieving urinary continence in cloacal exstrophy: The surgical cost<sup>☆</sup>

Mahir Maruf<sup>a</sup>, Matthew Kasprenski<sup>a</sup>, John Jayman<sup>a</sup>, Seth D. Goldstein<sup>b</sup>, Karl Benz<sup>a</sup>, Timothy Baumgartner<sup>a</sup>, John P. Gearhart<sup>a,\*</sup>

<sup>a</sup> Robert D. Jeffs Division of Pediatric Urology, The James Buchanan Brady Urological Institutions, The Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, USA

<sup>b</sup> Division of General Pediatric Surgery, Department of Surgery, The Johns Hopkins Hospital, Baltimore, MD, USA

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### ABSTRACT

**Introduction:** Cloacal exstrophy (CE) is a severe midline congenital abnormality that requires numerous surgical corrections to achieve an acceptable quality of life. Candidates for urinary continence undergo multiple procedures, most often continent bladder diversions, to become socially dry. Here, the authors investigate the number of genitourinary interventions that patients with CE undergo to attain urinary continence.

**Materials and methods:** A retrospective review of a prospectively maintained database of 1311 exstrophy epispadias complex patients was performed. Patients with CE who have had at least one continence procedure were included. A continence procedure was defined as bladder neck reconstruction with or without augmentation, bladder neck transection with continent urinary diversion, augmentation cystoplasty, or use of injectable bulking agents. Continence was defined as a dry interval greater than 3 hours without leakage at night.

**Results:** In total, 140 CE and CE variant patients have been managed at the authors' institution. Of the 116 CE patients, 59 received at least one continence procedure, 14 were excluded for incontinent diversion or cystectomy, and the remaining 43 patients are awaiting a continence procedure. At the time of analysis, 42 (71%) patients who underwent a continence procedure were dry. The median number of total urologic procedures to reach urinary continence was 4 (range 2–10). This included 1 bladder closure (range 1–3), 2 urinary continence procedures (range 1–4), and 1 (range 0–4) "other" genitourinary procedures. The median time to urinary continence was 11.0 years (95% CI [9.2–14.2]).

**Conclusions:** A majority of CE patients who undergo a diversion procedure can achieve urinary continence. However multiple continence procedures are likely necessary. Of patients who are candidates for a continence procedure, half will be continent by the age of 11.

**Level of Evidence:** Level IV, Case series with no comparison group.

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Cloacal exstrophy (CE) is the most severe form of the exstrophy-epispadias complex (EEC), characterized by an abnormality in cloacal membrane development, resulting in gastrointestinal, genitourinary, nervous, and musculoskeletal defects [1,2]. Historically, management of CE was principally concerned with survival, but with both surgical and nutritional advances in the past 3 decades, survival has reached near 100% [3–5] and the focus has transitioned to quality of life improvements.

Urinary continence in CE has been described as early as the 1980s, in which 3 of a series of 7 patients were able to achieve urinary continence. Subsequent cases were reported on an individual basis [6–8]. With an increased focus on urinary outcomes, reports of continence have

become more frequent [9]. A small series by Mitchell et al. demonstrated that 100% of patients were able to achieve a dry period of >3 hours using a continent stoma and clean intermittent catheterization, and only 50% were dry during the day and night [10]. The ability to achieve continence is dependent on multiple factors, including the timing and type of continence procedure, bladder growth and capacity, as well as parental and social issues [9].

Emotional maturity, physical ability, and desire for continence in the child and guardians are all necessary for a patient to qualify for a continence procedure. As such, successful reconstruction typically employs an individualized approach [11]. Children who lack this emotional maturity or physical ability for volitional voiding or performing intermittent catheterization may be successfully managed with incontinent diversions. Still, those who qualify for continence may require multiple reconstructive procedures for eventual urinary continence [10]. Though subject to variability, the number of procedures and time to reach urinary continence in CE patients are largely unknown. This study aims to investigate the number of urologic interventions, and time from

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\* Corresponding author at: The Johns Hopkins University School of Medicine, James Buchanan Brady Urological Institute, Division of Pediatric Urology, Charlotte Bloomberg Children's Hospital, 1800 Orleans St, Suite 7304, Baltimore, MD 21287.

E-mail address: [jgearha2@jhmi.edu](mailto:jgearha2@jhmi.edu) (J.P. Gearhart).

birth necessary to achieve urinary continence for children with CE. The authors hypothesize that almost all CE patients that achieve urinary continence do so by continent urinary diversion (CUD), often after numerous surgeries.

## 1. Methods

A prospectively maintained Institutional Review Board approved institutional and daily updated database of 1311 EEC patients was reviewed for CE patients with attempted continence procedures. Patients who are awaiting their first continence procedure, or had a cystectomy were not included. A bladder neck reconstruction (BNR) with or without augmentation, bladder neck transection with catheterizable stoma, augmentation cystoplasty, or use of injectable bulking agents was considered an attempted continence procedure. In instances when a catheterizable stoma was performed with or without a bladder neck transection, this was considered a CUD. Continence was defined as a reported dry interval >3 hours with no nighttime leakage.

The number of surgical procedures performed in this patient cohort was calculated by totaling the number of bladder closure events, continence procedures, and other genitourinary procedures. The “other” procedures included surgical interventions in genitourinary tract for the management of cloacal exstrophy, and did not include diagnostic or assessment procedures. The omphalocele closure, other gastrointestinal procedures, neurological procedures, and musculoskeletal procedures were also not included in this calculation as these are not directly related to urinary continence, and are thus outside of the scope of this investigation. Though neurologic procedures in particular may indirectly contribute to continence status, the purpose of such procedures is not solely to provide urinary continence.

To compare demographic characteristics of patients who attained continence and those that did not, Chi-square analysis was performed for categorical variables and Wilcoxon rank sum test for continuous variables. A two sided  $p$ -value <0.05 was considered statistically significant. Continence probability was calculated using the Kaplan–Meier method. The patients' date of birth was the starting point of analysis and date of successful continence procedure was the event studied. Follow-up time was censored at date of last continence status. Statistical analysis was performed using R version 3.4.0 (R Foundation for Statistical Computing, Vienna, Austria).

## 2. Results

### 2.1. Patient demographics

In total, 140 patients with CE or a variant of CE have been managed at the authors' institution. Of the 116 CE patients, 59 had undergone at least one continence procedure, fourteen patients who underwent incontinent diversion or cystectomy were excluded and the remaining 43 patients are awaiting a continence procedure. Twenty-six (44%) were identified as male while 33 (56%) were female, however of the female patients, 14 were karyotyped as XY while 5 were XX. The remaining 14 were labeled as female based on examination and intraoperative characteristics. The median follow-up time was 13.4 years (range 3.5–53.2).

Demographic and primary closure information is displayed in Table 1. No statistical difference in sex, race, period of primary closure, primary closure type, or age at first continence procedure was observed between continent and incontinent patients. During or before the primary bladder closure, 37 (62.7%) received a pelvic osteotomy. Fifteen (25.4%) of the primary closures ultimately failed, of which 6 (40%) had received a pelvic osteotomy.

### 2.2. Number of surgical interventions

Of the 59 patients who have had a continence procedure, 42 (71.2%) have achieved urinary continence at the time of analysis. Forty-six patients have had only one bladder closure, 9 have had two, and 4 have

**Table 1**  
Patient demographics.

Covariate	Entire Cohort (n = 59)	Continent (n = 40)	Incontinent (n = 19)	p-value
Sex				0.725
Male	26 (44.1%)	17 (42.5%)	9 (47.4%)	
Female	33 (55.9%)	23 (57.5%)	10 (52.6%)	
Race				0.625
Caucasian	45 (76.3%)	29 (72.5%)	16 (84.2%)	
Primary Closure				0.097
Neonate	25 (42.4%)	14 (35.0%)	11 (57.9%)	
Delayed	27 (45.8%)	19 (47.5%)	8 (42.1%)	
Unknown	7 (11.9%)	7 (17.5%)	0 (0%)	
Closure Type				0.224
1-Stage-cloacal	23 (39.0%)	13 (32.5%)	10 (52.6%)	
Staged Cloacal	33 (55.9%)	24 (60.0%)	9 (47.4%)	
Other/Unknown	3 (5.1%)	3 (7.5%)	0 (0%)	
Age at first CP, years [IQR]	5.5 [2.5-7.6]	5.6 [3.5-8.2]	4.7 [1.2-7.0]	0.094

Abbreviations: CP, continence procedure; IQR, interquartile range.

had three. The proportions of those who attained continence after one, two, and three bladder closures were 71.7%, 77.8%, and 50.0% respectively.

The total number of therapeutic urologic procedures performed in the entire cohort was 268. The median number of urologic interventions per patient in this cohort was 4 (range 2–10), which included a median of 1 bladder closure (range 1–3), 2 continence procedures (range 1–4), and a median of 1 “other” genitourinary procedures (range 0–4). The “other” genitourinary procedures were not intended to affect urinary continence and may be completely independent of the continence procedure (epispadias repairs, urethroplasty, or ureteral reimplants, etc.) or a related sequelae of the continence procedure. Twenty-eight percent of these procedures were in the latter group, and consisted primarily of cystolitholapaxy/cystolithotomy, repair fistula after BNT, or stenosis correction.

For those patients who ultimately achieved urinary continence, the total number of urologic procedures was 185 in the entire cohort. Per patient, the median number of surgical procedures necessary for continence was 4 (range 2–10), which included 1 closure (range 1–3), 2 continence procedures (range 1–4), and 1 (range 0–4) “other” genitourinary procedures.

In total, 33/59 (55.9%) patients have undergone more than one continence procedure. The proportions of patients who have had one, two, three or four continence procedures were 26/59 (44.1%), 25/59 (42.4%), 5/59 (8.5%), and 3/59 (5.1%), respectively. After their last procedure, 18/26 (69.2%), 18/25 (72.0%), 4/5 (80.0%) and 2/3 (66.7%) achieved urinary continence.

A CUD was the most commonly performed continence procedure during the first attempt at urinary continence. Of the 15 patients who underwent a BNR with or without a stoma as their first continence procedure, 5 (33%) underwent a bladder neck transection with CUD for further continence management. Fig. 1 displays the frequency of continence procedures for each successive procedure number performed in this cohort. In the entire cohort, 40 patients (68%) have had an augmentation cystoplasty, of which 6 required a repeat augmentation.

### 2.3. Time to continence and method of achieving dryness

A total of 55 patients had complete data for Kaplan–Meier analysis. The 4 excluded patients were missing the date of their last continence procedure. The median time to continence was 11.0 years (95% CI [9.2–14.2]) as displayed in Fig. 2. The 5 and 10 year probabilities of continence (from birth) were 9.2% (95% CI [1.2–16.5]) and 41.7% (95% CI [26.1–54.0]) respectively. By the age of 5 and 10, patients had a median of 6 (range 3–6) and 5 (range 3–10) genitourinary procedures to achieve continence, respectively.

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