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Long-term follow-up of composite bladder augmentation incorporating stomach in a multi-institutional cohort of patients with cloacal exstrophy

J.T. Casey [a], K.H. Chan [a], Y. Hasegawa [b], T. Large [a], B. Judge [a], M. Kaefer [a], R. Misseri [a], R.C. Rink [a], K. Ueoka [b], M.P. Cain [a]

[a]Division of Pediatric Urology, Riley Hospital for Children, Indiana University School of Medicine, Indianapolis, IN, USA

[b]Division of Urology, Department of Surgical Subspecialties, National Center for Child Health and Development, Tokyo, Japan

Correspondence to: K. H. Chan, Division of Pediatric Urology, Riley Hospital for Children, Indiana University School of Medicine, 705 Riley Hospital Drive, Suite 4230, Indianapolis, IN 46202, USA, Tel.: +1 317 944 8896

kchubert@iupui.edu (K.H. Chan)

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Summary

Introduction
Composite bladder augmentation, incorporating gastric and bowel segments, has the theoretical advantage of metabolic neutrality while potentially avoiding the morbidities of gastrocystoplasty, such as hematuria-dysuria syndrome. The most common indication for this operation is a paucity of bowel, such as in cloacal exstrophy. Despite several early descriptive studies of this technique, there are no reports, to date, of long-term follow-up in this population.

Objective
To describe the outcomes of composite bladder augmentation utilizing stomach in a cohort of cloacal exstrophy patients.

Materials and Methods
A retrospective review of cloacal exstrophy patients who underwent composite bladder augmentation from 1984 to 2006 at two institutions was performed. The incidence of mortality and morbidities related to augmentation was evaluated.

Results
Eleven patients with cloacal exstrophy underwent composite bladder augmentation. Median age at initial augmentation was 6.4 years (interquartile range (IQR) 4.4–9.1). Median follow-up was 13.2 years (IQR 11.2–24.6). The Summary table describes the types of composite bladder augmentations. Of the three patients with pre-operative metabolic acidosis, two improved with composite bladder augmentation and one developed metabolic alkalosis. Three developed hematuria-dysuria syndrome: one improved with staged ileocystoplasty, and two had persistent symptoms successfully treated with H2 receptor blockers. Two of 11 developed symptomatic bladder stones. There were no reported bladder perforations, bladder malignancies, conversions to incontinent urinary diversions, or deaths.

Conclusion
With long-term follow-up, very few patients developed metabolic acidosis/alkalosis after composite bladder augmentation. The composite bladder augmentation will continue to be used in patients with cloacal exstrophy, in order to minimize the impact on the pre-existing short gut in these patients.

Summary table Description of the types of composite bladder augmentations.

Type of composite bladder augmentation	Number of patients (%)
Gastrocystoplasty onto hindgut incorporated during initial bladder closure	2 (18.2%)
Gastrocystoplasty with subsequent ileocystoplasty	2 (18.2%)
Initial composite bladder augmentation with both stomach and ileum	5 (45.5%)
Multiple augmentations with a final composition of stomach and ileum	2 (18.2%)

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Introduction

The use of gastric segments for bladder reconstruction was first described in an animal model in 1956 [1]. In the 1990s, gastrocystoplasty became an alternative to the use of small or large intestine for bladder augmentation, especially in patients with renal failure and associated acidosis or insufficient bowel for reconstruction [2]. Since the initial reports, long-term studies have reported complications unique to primary gastrocystoplasty such as metabolic alkalosis and hematuria-dysuria syndrome (HDS) [3–6]. Composite bladder augmentation, incorporating gastric and bowel segments, has the theoretical advantage of metabolic neutrality while avoiding some of the complications of isolated gastrocystoplasty such as HDS [7–11].

Cloacal exstrophy patients have a spectrum of congenital anomalies that affect many organ systems, including the genitourinary and gastrointestinal tracts. Patients may need bladder reconstruction in order to obtain continence, which often consists of augmentation in addition to bladder neck reconstruction and continent catheterizable channel creation after the initial approximation of the bladder halves [12–14]. Additionally, short bowel syndrome has been observed in 25% of cases, even in the presence of normal bowel length, indicating that preservation of bowel is paramount in this population [15,16].

The use of composite bladder augmentation in the cloacal exstrophy population allows for less use of small intestine, thereby preserving the gastrointestinal tract for digestion. Despite several early descriptive studies of the outcomes after composite bladder augmentation [7–11], there are only reports of 5-year follow-up in a population with multiple underlying etiologies [9–11]. The present study describes the long-term follow-up of cloacal exstrophy patients after composite bladder augmentation from two institutions.

Methods

A retrospective chart review was performed of cloacal exstrophy patients who underwent composite bladder augmentation (concurrent or metachronous) at an age of $\leq$21 years from 1984–2006 at two institutions. Composite bladder augmentation was defined as use of gastric tissue and another source of intestine (ileum, colon, hindgut) either as a staged procedure or an initial composite, as per the technique initially described by Lockhart et al. [7]. The incidence of mortality and morbidities related to augmentation were evaluated, including: metabolic abnormalities (acidosis or alkalosis), chronic kidney disease defined by estimated glomerular filtration rate (eGFR), HDS, bladder stones, bladder perforation, bladder malignancy, and conversions to incontinent urinary diversion. Additionally, overall bladder functionality was evaluated by documenting recorded capacity, incontinence and progressive hydronephrosis. Both institutions survey these patients with yearly upper tract imaging, yearly cystoscopy after 10 years of augmentation and urodynamics, as needed, for changes in hydronephrosis or symptoms.

The bedside Schwartz formula for eGFR was used in patients $<$18 years [17]. This formula approximates eGFR in children from plasma creatinine and body length, as eGFR (ml/min/1.73 m^2) = (0.413 $\times$ height in cm/creatinine in mg/dl). The four-variable Modification of Diet in Renal Disease (MDRD) formula, which includes demographic and serum variables, was used in patients $\geq$18 years, as eGFR (ml/min/1.73 m^2) = 175 $\times$ (serum creatinine)$^{-1.154}$ $\times$ (age)$^{-0.203}$ $\times$ 0.742 (if female) $\times$ 1.212 (if black race) [18]. End-stage renal disease (ESRD) was defined as chronic kidney disease (CKD) stage 4 or renal transplantation.

Results

Eleven patients with cloacal exstrophy underwent composite bladder augmentation (see Table 1). The median age at initial augmentation was 6.4 years (interquartile range (IQR) 4.4–9.1) with a median follow-up of 13.2 years (IQR 11.2–24.6). Of the nine patients with a 46,XY karyotype, three were raised as female and underwent early genital reconstruction, five were raised as male, and one transitioned from female to male gender at 9 years of age. Both patients with a 46,XX karyotype were raised as female. One of 11 patients was lost to follow up and not been seen by Urology in the past 5 years.

Patients represented both initial concurrent composite bladder augmentations and various combinations of metachronous staged procedures. Two patients underwent gastrocystoplasty onto hindgut incorporated during initial bladder closure, two underwent gastrocystoplasty with subsequent ileocystoplasty, five underwent initial composite bladder augmentation with both stomach and ileum (one with ileum from a prior ileal chimney), and two underwent multiple augmentations with a final composition of stomach and ileum. Many of the patients have no to minimal original bladder mucosa after either cystectomy or multiple and/or aggressive bladder neck reconstructions/ligation, and therefore currently have composite reservoirs.

Three of 11 patients had pre-operative metabolic acidosis (based on a serum bicarbonate of $<$20 mEq/l); of these, two patients improved after composite bladder augmentation construction and one developed mild metabolic alkalosis. One patient had pre-operative CKD stage 2 (defined as eGFR of 60–89 ml/min) that improved to CKD stage 1 postoperatively (eGFR $>$90 ml/min). One patient transitioned from CKD stage 1 to CKD stage 2 postoperatively. The remaining nine patients had eGFR of $>$90 ml/min pre-operatively and postoperatively. There was no progression to ESRD.

Three patients developed HDS and required treatment after gastrocystoplasty. One patient developed HDS (associated with pain with catheterization) after initial primary gastrocystoplasty, and it improved with a metachronous staged ileocystoplasty. Two patients had mild hematuria without pain that persisted after composite augmentation, but was successfully treated with H2 receptor blockers. Therefore, two of the 11 patients with HDS required treatment after composite bladder augmentation.

Two of 11 patients developed symptomatic bladder stones that required operative intervention. Bladder stone compositions were both infectious stones: (1) a mixture of