Multidisciplinary management of adult cloacal extrophy complications: A challenging case of nephrolithiasis and abdominal wall extrusion of a catheterizable ileal pouch

Sarah S. Christianson a, Eric Ghiraldi a, Justin Friedlander a, d, Sameer Patel b, c, Alfred Trang b, Jay Simhan a, d, *

a Department of Urology, Einstein Healthcare Network, Philadelphia, PA, 19141, USA
b Department of Surgery, Einstein Healthcare Network, Philadelphia, PA, 19141, USA
c Division of Plastic and Reconstructive Surgery, Department of Surgical Oncology, Fox Chase Cancer Center, Philadelphia, PA, 19111, USA
d Division of Urologic Oncology and Urology, Fox Chase Cancer Center, Philadelphia, PA, 19111, USA

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ABSTRACT

Cloacal extrophy is a rare congenital syndrome which comprises multiple genitourinary, gastrointestinal and musculoskeletal anomalies. The long-term effects following childhood cloacal extrophy management is poorly characterized in living adults. In this report, a 42-year-old female born with cloacal extrophy presented after numerous prior surgical reconstructions with abdominal extrusion of a catheterizable ileal pouch and bilateral staghorn calculi. We review the steps in surgical management of this uncommon scenario with a goal of improving the patient’s quality of life.

Introduction

While cloacal extrophy (CE) syndrome in neonates is extremely rare, high volume pediatric centers have developed multidisciplinary collaborations with staged reconstructions. Accordingly, CE patients warrant continued long-term follow up into adulthood due to delayed complications.

We report the case of an adult born with CE requiring multidisciplinary management of nephrolithiasis and abdominal wall reconstruction for management of urinary incontinence via a catheterizable pouch extruded through the anterior abdominal wall.

Patient presentation

A 42-year-old-patient born with CE syndrome presented originally to her local urologist with longstanding weakness, electrolyte abnormalities, urinary incontinence and flank pain. The patient’s surgical history was significant for a staged primary closure performed in infancy including bilateral pelvic osteotomies. Additional anatomic abnormalities included absence of the hindgut requiring end ileostomy and a solitary testicle removed prior to puberty. The patient had a complicated course throughout adolescence and required multiple surgeries including a vesicostomy and subsequent cystectomy, creation of continent reservoir with a catheterizable channel, intestinal neovaginal reconstruction, vaginoplasty, repair of vesicocutaneous fistula, and multiple surgeries to treat nephrolithiasis.

On physical examination, the patient had a right-sided end ileostomy, a large midline abdominal wall defect with extruded ileal reservoir and a stenosed non-functional catheterizable channel (Fig. 1). After initial workup the patient was diagnosed with a right staghorn calculus, left-sided ureterolithiasis, and urinary incontinence due to abdominal extrusion of her catheterizable ileal pouch.

Kidney stone management

The patient was taken to the operating room and had a right percutaneous nephrolithotomy, nephroscopogram (Fig. 2), right antegrade ureteroscopy and laser lithotripsy. Intraoperative findings were consistent with a left to right transureteroureterostomy (TUU) with the right distal ureter anastomosed to the neobladder with a widely patent single anastomosis. This allowed for left sided retrograde ureteroscopy across the TUU and stone removal.

Abbreviations: CE, Cloacal Exstrophy; PCNL, Percutaneous Nephrolithotomy; TUU, Transureteroureterostomy.

* Corresponding author. Einstein Healthcare Network, Department of Urology, 1200 Tabor Road 3rd Floor, Philadelphia, PA, 19141, USA.
E-mail address: jsimhan@gmail.com (J. Simhan).
Urinary/Abdominal wall reconstruction

A multidisciplinary surgical plan was conceived involving reconstructive urology, general surgery and plastic reconstructive surgery specialists.

A standard laparotomy incision was utilized to enter the abdominal cavity. Extensive adhesiolysis was performed, incidental enterotomies were repaired and nonviable of bowel was resected. The end ileostomy and extruded reservoir were mobilized and the TUU anastomosis was identified. The reservoir was deemed unsalvageable and ultimately excised. A short, distal ileal segment was isolated to create a new ileal conduit. The distal right ureter was excised and the TUU was incorporated directly into the ileal conduit with a Wallace type anastomosis using 4-0 PDS in running fashion. Bilateral ureteral catheters were placed across the anastomosis. The ileal conduit and ileostomy were matured in standard fashion.

For the abdominal wall closure, soft tissue was elevated off the deep anterior abdominal wall fascia to recruit fascia to the midline. Anterior component separation was not deemed necessary given the ability to approximate the fascia. While the upper abdominal fascia was closed primarily, from the mid abdomen down to the pubis an allograft select thick (2.4mm) biological mesh was placed in an underlay fashion to reinforce the primary closure using interrupted #1 PDS sutures (Fig. 3A). The anterior fascia was then closed in an interrupted fashion above the mesh (Fig. 3B). Scarpa’s fascia and skin was closed primarily and an abdominal binder was placed. The patient required a short course of parenteral nutrition while admitted and IV fluid supplementation on home discharge in order to maintain fluid status but ultimately presented at 3 month follow up with a well healing abdominal wound and good function of both ostomy appliances.

Discussion

Transition of care into adulthood and management of complications represents a substantial obstacle for urologists caring for patients with CE. Our case required multidisciplinary collaboration including
nephrolithiasis management followed by urinary tract reconstruction with urinary diversion, bowel diversion and complex abdominal wall closure.

When caring for CE patients into adulthood, one cannot overlook the importance of obtaining a detailed surgical history. In many such adult patients with a remote surgical history, this can be especially challenging since often, the originally performed reconstructions were done elsewhere many years prior. In this case, we utilized a combination of computed tomography and intraoperative pyelography during kidney stone management to help formulate a reconstructive plan. The pyelogram shown in Fig. 2 was critical in identifying the TUU and understanding the anatomy of our patient’s urinary tract.

In CE patients there is a large incidence of associated gastrointestinal tract anomalies including anatomically short bowel with hindgut remnants of varying sizes. Short gut syndrome may occur despite normal length of bowel suggesting inherent absorptive dysfunction may play a significant role. Given these additive risk factors, efforts should be made in CE patients to preserve as much bowel length as possible in both the index surgery as well as in future reconstructive measures. Therefore, it is vital for general surgeons to be intimately involved in caring for these patients. In the index patient, we also utilized gastroenterology/nutritional specialists post-operatively to tailor a bowel regimen with bulking agents to prevent short gut syndrome.

Paramount in the management of CE is surgical reconstruction and reinforcement of the abdominal wall defect. In this case anterior component separation was not performed given good fascial approximation as well as the concern for further weakening of the fascia and potentially increasing the risk of a parastomal hernia. Henderson and associates reported the use of Alloderm as an adjunct in bladder closure for 43 pediatric extrophy patients undergoing primary closure or reclosure procedures. No patients experienced failure of closure, vesicocutaneous fistula, infrapubic stitch erosion or intrasymphyseal plate erosion. Giordano and colleagues reported primary fascial closure with biological mesh reinforcements resulted in a reduced hernia rate and overall complication rate when compared to bridged mesh repairs (6.2% vs. 33.3%; p < 0.001) and (30% vs. 59%; p = 0.001) respectively. Both of these investigations provided encouraging evidence that utilization of alloderm grafts may mitigate risk of future adverse events in closing large abdominal wall defects in complicated CE patients.

**Conclusion**

Given the complex anatomic and underlying medical comorbidities in adult cloacal extrophy patients, we advocate that late complications be managed with multidisciplinary efforts with great care to optimize quality of life in this rare, but challenging patient population.

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