

Management of Recurrent Pelvic Fluid Collections in Adult Male Bladder Exstrophy Patients With Maintenance of Erectile Function



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OBJECTIVE	“To describe management options for pelvic fluid collections in adult patients with classic bladder exstrophy.”
METHODS	A single institution retrospective chart review was performed of patients who presented between 1998 and 2016 with a history of bladder exstrophy and pelvic fluid collections and 3 patients were identified. Patients had been followed for a mean of 9.0 years (1-23).
RESULTS	All 3 patients required urinary diversions at various intervals following their exstrophy repair as newborns. All initially presented with symptomatic fluid collections located inferior to the bladder visualized by cross-sectional imaging. Mean age at presentation was 32.3 years (26-38 years). Two patients underwent drainage and sclerosing of cystic fluid collections with durable symptomatic relief for 1 patient. The other had recurrence of the fluid collections so he underwent marsupialization of the fluid collection which failed to sufficiently alleviate his symptoms. Ultimately, he along with the last patient, underwent open excision of the presumed hypoplastic prostate leading to resolution of pain symptoms, though the last patient did have some persistence of the fluid collection. All patients maintained their erectile function subsequent to these interventions.
CONCLUSION	Adult patients with bladder exstrophy can present with painful cystic fluid collections potentially due to secretions from presumed hypoplastic prostate tissue. Sclerosing of the cyst can be successful in a subset of these patients, though some may require removal of the presumed prostatic tissue, which is curative and can be achieved with preservation of erectile function. UROLOGY 125: 256–259, 2019. © 2018 Elsevier Inc.

The exstrophy-epispadias complex is a spectrum of urogenital defects that ranges from epispadias associated with mild pubic diastasis and a dorsally located urethral meatus to classic bladder exstrophy (CBE) in which there is wide pubic diastasis, and an abdominal wall defect exposing an open bladder and urethra with an epispadiac meatus. It is a rare condition affecting 3-5 of every 100,000 births.¹⁻³

Given the complexity of defects associated with classic bladder exstrophy, these patients often require multiple reconstructive surgeries to achieve acceptable continence and cosmesis. There are many long-term complications associated with these interventions which challenge not only pediatric urologic management but also their transition into

adult care. However, long-term complications, while essential to managing transitional care, are not well-described. In this case series, we report the diagnostic approach and management of 3 adult patients with a history of CBE and various repair procedures who presented with recurrent pelvic pain attributed to cystic fluid collections inferior to the bladder. We believe these fluid collections result from presumed hypoplastic prostate secretions. Prostates in individuals with a history of CBE are classically hypoplastic and difficult to identify, both by tissue planes and histology.^{4,5} Despite lack of tissue diagnosis, we believe that the source of the fluid collections is residual prostatic tissue given the appearance of the tissue on cross-sectional imaging and also during resection, the prostate gland's exocrine function and the location and large size of the fluid collections.

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METHODS

A single institution retrospective chart review was performed of patients who presented between 1998 and 2016 with a history of bladder exstrophy and pelvic fluid collections and 3 patients

were identified. Patients had been followed for a mean of 9.0 years (1-23). Their status on presentation and subsequent clinical course, treatments received, and outcomes were noted.

RESULTS

Case 1

A 46-year-old man born with CBE underwent ureterosigmoidostomy at birth and closure of the abdominal wall using the bladder muscle as a bolster. He had subsequent operations including revision of the abdominal wall closure with mesh and cystectomy leaving the prostate in situ. He maintained erectile and ejaculatory function. At the age of 38, he was found to have a painful collection in a posterior urethral cavity. He underwent urethroscopy and had the cavity drained endoscopically. Five years later, the painful pelvic collection reaccumulated. Urethroscopy and retrograde urethrogram demonstrated an occluded urethra without connection to the collection cavity. The fluid collection was drained percutaneously and sclerosis was performed by interventional radiology (IR). Since that intervention, over 17 years of follow-up, the fluid collection has not recurred and he has maintained erectile function.

Case 2

A 27-year-old male presented with history of CBE and multiple reconstructive surgical repairs including augmentation cystoplasty, bladder neck closure, and catheterizable appendicovesicostomy. He presented for evaluation at our institution at the age of 26 with recurrent pain at the dorsal base of his penis. His urethra was not patent and he managed his bladder with clean intermittent catheterization through his appendicovesicostomy. He had intact erectile function.

Prior to transferring his care to our adult hospital, he presented on several occasions to our children's hospital with similar complaints where he was found to have a fluid collection at the base of his penis. It was thought that this was the likely focus of his pain, and he underwent several percutaneous drainage procedures by IR with sclerosing of the cavity using doxycycline or a combination of doxycycline and ethanol. These interventions did little to alleviate his symptoms.

On initial evaluation, magnetic resonance imaging (MRI) showed a 2.2 by 1.4 cm collection dorsal to the posterior penile urethra, between the cavernosal crura, anterior to the expected location of a prostate, and inferior to the bladder. There was no patent connection between the bladder and urethra. Urethroscopy and retrograde urethrogram showed an obliterated urethra distal to the collection, and no connection between the collection and the urinary tract. The fluid collection was drained by IR.

The patient presented once again less than 2 months later with recurrent pain at the base of his penis identical to his previous presentations. A repeat MRI once again showed an identical fluid collection at the base of his penis, which is now 4.1 by 2.9 by 2.8 cm. Again, the collection was drained by IR.

In the subsequent months, the patient continued to experience recurrent pain at the base of his penis. He returned for 3 ultrasound-guided percutaneous aspirations and 2 attempts at ethanol sclerosis of the cystic space, which unfortunately were ineffective in preventing recurrence of the collection or his pain.

Given that this cyst was refractory to recurrent drainage attempts, and because the patient was continuing to experience severe pain requiring hospital admissions for narcotic pain

control, the patient elected to escalate surgical therapy with marsupialization of the cyst. Intraoperatively, ultrasound was used to identify the location of the suprapubic cyst, which was then aspirated. While an attempt was made to anastomose the cyst to the urethra proximal to the ablated segment to allow cyst drainage, there was insufficient mobility of either the urethral stump or the cyst to allow this, so the cyst wall was marsupialized to the suprapubic skin. A follow-up MRI 2 months later demonstrated almost complete remission of the cystic fluid collection with only a residual 0.7 cm collection remaining. Despite these radiographic findings, the patient continued to feel pressure in his urethra and pain at the base of his penis. Given his escalating symptoms, an MRI was repeated 3 months postmarsupialization demonstrating a stable residual cyst size of 0.8 by 0.9 cm. In the setting of his continued pain and the presumed prostatic origin of these minimal secretions, the patient elected to pursue prostatectomy as a final escalation of surgical therapy. Intraoperatively, a cystic structure that was not contiguous with the prior marsupialization was identified and circumferentially excised. This cyst abutted on what appeared to be remnant prostatic tissue which was removed. His postoperative course was uncomplicated and he reports remission of his pelvic pain at 7 months follow-up. He also has maintained erectile function.

Case 3

A case of 38-year-old man with a history of CBE was presented who underwent closure of the bladder exstrophy and epispadias repair as an infant. He subsequently required multiple bladder reconstructions and 2 bladder augmentations. Upon initial presentation to our institution, he managed his bladder with urethral self-catheterizations. He had preserved sexual function and fertility.

He presented to our institution having difficulty with self-catheterization and recurrent urinary tract infections (UTIs), characterized predominantly by penile pain and purulent urethral discharge. He underwent cystoscopy and retrograde urethrogram showing an abnormal and tortuous urethra with a pan-urethral stricture and hourglass deformity of the bladder. We hypothesized that these anatomic abnormalities were preventing him from completely emptying his bladder with intermittent self-catheterization, and thus predisposing him to recurrent UTIs and pyocystis. After discussing various treatment options with the patient, we opted to fashion a Monti abdominal catheterizable channel allowing him to resume self-catheterization with complete bladder emptying. One year later, despite a decrease in symptomatic UTIs, the patient was still bothered by stomal incontinence. Based on further discussion and this patient's preferences he then opted to undergo a simple cystectomy with Indiana pouch creation to provide him with a permanent form of urinary diversion. Given his erectile and fertility status, no attempt was made to perform a cystoprostatectomy at the time, and a very small bladder remnant was left.

A few months later, the patient represented with pelvic pain. A computed tomography (CT) scan was performed showing a 1.2 by 1.6 cm fluid collection in the subcutaneous tissue as well as a 1.4 by 0.6 cm fluid collection along the anterior margin of the bladder remnant. A repeat CT 2 weeks later showed an additional new fluid collection measuring 1.6 by 0.7 cm deep to the midline ventral abdominal wall. Furthermore, there was a new thin fluid collection measuring 4.6 by 1.0 cm alongside the left lateral aspect of the bladder remnant. All the collections were drained percutaneously with temporary relief of his symptoms.

About a month later, he returned to the hospital with abdominal pain. A CT scan showed resolution of the 2 small fluid collections and reduction in size of the fluid collection seen along the left lateral aspect of the bladder remnant, but appearance of 2 new large fluid collections measuring 7.1 cm in greatest dimension in the right hemipelvis and 3.3 cm in the left hemipelvis. IR drainage of the pelvic fluid collections was deferred as he otherwise appeared clinically well. Follow-up CT about a month later showed spontaneous resolution of the fluid collections.

Six months later, he returned to clinic with purulent daily urethral discharge and some bulging, erythema, and discomfort in the suprapubic region. Cystoprostatectomy was recommended at this point given that he had failed all forms of conservative management. The patient, however, decided not to proceed with this option at the time. The urethral discharge and suprapubic pain persisted for several years and he experienced recurrent UTIs which required hospital admission. An MRI of the pelvis in 2017 showed a 4.9 by 1.1 cm fluid collection near the bladder remnant consistent with the fluid collection seen on CT in 2013. A transrectal aspiration of the fluid collection was attempted but was unsuccessful. He finally decided to undergo cystoprostatectomy after extensive counseling. The procedure was successful with resolution of pelvic pain and urethral discharge and preservation of sexual function at 1 month follow-up. A follow-up surveillance CT scan 4 months postoperatively showed a small persistent fluid collection of unclear origin. However, he remained asymptomatic and has not sought further treatment or follow-up.

DISCUSSION

This is a case series of 3 adult patients with a history of bladder exstrophy repair as infants who presented with painful cystic fluid collections in the pelvis. We believe the fluid to be, at least in part, secretions from prostatic tissue remnants. A serum prostate-specific antigen may have been helpful although the significance of any detected level in guiding treatment decisions is unclear. There is no definitive treatment strategy outlined in the literature for such cystic fluid collections. In this article we present, to our knowledge, the first case series on the subject and propose a treatment paradigm progressing from least to most invasive treatment that we have found to be effective and can preserve erectile function, as illustrated in Figure 1.

A reasonable first step in managing these patients is sclerosis of the fluid collections. Sclerosis is minimally invasive, and as seen in our first case, can result in long-term cure. These fluid collections, however, can also recur despite multiple treatments as noted in the second case. It is unclear what factors are associated with success or failure of sclerosis but if the fluid collections persist, proceeding to marsupialization of the cyst wall to the external skin to allow it to drain freely can be the next step. This procedure is ideal for superficial fluid collections but may be unsuitable for more deeply located fluid collections given the difficulty of accessing and marsupializing those regions. If marsupialization is ineffective, removal of the suspected prostatic and surrounding tissue can be entertained. However, this treatment choice must be

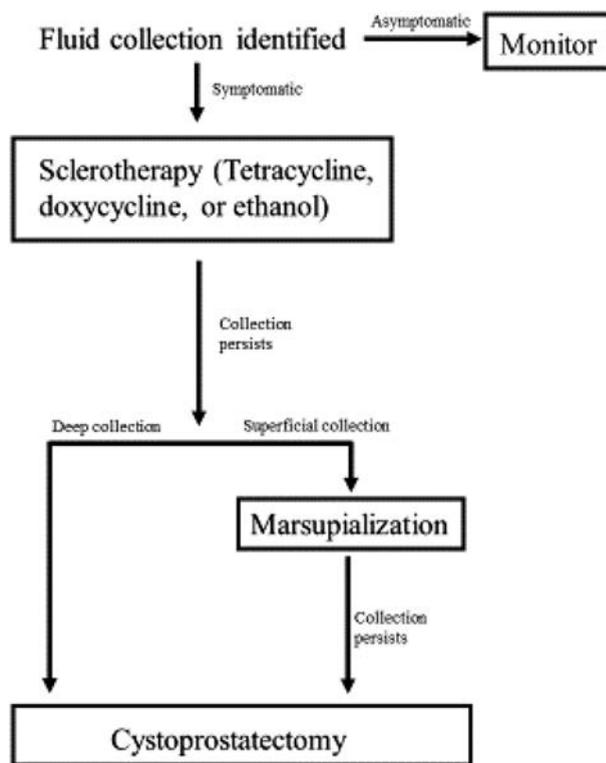


Figure 1. Treatment algorithm for management of recurrent pelvic fluid collections in adult male bladder exstrophy patients. (Color version available online.)

considered carefully as extensive scarring from prior repairs can make the procedure technically difficult. Dissection to access the hypoplastic prostate can also jeopardize the blood supply to the reconstructed bladder, if present, although in our cases we did not observe any impact on the reconstructed bladder.

An additional consideration is the preservation of erectile function and fertility by natural means. These factors drive a lot of the decision making in terms of reconstructive options throughout early life. Maintenance of these functions should continue to inform decision-making in adulthood when faced with problematic pelvic fluid collections. While the concern for erectile dysfunction in particular can dissuade patients and surgeons alike from entertaining more aggressive options, our series shows that erectile function can be maintained, even with more extensive exenterive procedures. We, therefore, suggest tailoring the treatment strategy to each patient specifically considering the patient's unique anatomy and the varying levels of distress experienced from the cystic fluid collection. Possible treatment complications must be carefully balanced against the adverse effects the patient would experience from watchful waiting.

Limitations of our case series include the small number of cases which makes it difficult to analyze success or failures of interventions in a more generalizable manner. This is reinforced by the often unique anatomic presentations of each patient. This being said, to our knowledge

this is the largest series of such patients reported in the literature. Furthermore, we present a treatment strategy that was developed based on our experience encountering these patients. We suggest progressing from least (sclerosing) to most invasive (excision of remnant prostatic tissue), while also considering the patient's anatomical factors and personal preferences.

CONCLUSION

Drainage and sclerosis of the fluid collections is the least invasive option and was successful in one of the two patients. If this strategy fails, patients can attempt marsupialization of the collections, though this is most suitable for superficial fluid collections. Finally, if the patient continues to experience distress from the fluid collections, cystoprostatectomy is a viable option that can potentially provide lasting resolution of the fluid collections and pain symptoms.

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Editorial Comment



The authors present an unusual series of patients with complaints of persistent suprapubic pain. The report details their presumptive explanation for the etiology of the discomfort and a successful treatment regimen. This observation gives urologists a reasonable plan for management of symptomatic suprapubic fluid collections in males with a history of bladder exstrophy and/or epispadias repair.

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Author Reply



Bladder exstrophy is a rare condition yet these patients can be challenging to manage in adulthood especially when presenting with painful abdominal fluid collections which we believe are from remnants of prostatic tissue. As urologists may encounter these patients, even if only rarely, our goal was to provide recommendations for management based on our experience. Even if not universally applicable to every patient with a repaired bladder exstrophy, we believe our algorithm may be a good starting point.

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