Variant Presentations of the Exstrophy-Epispadias Complex: A 40-Year Experience

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OBJECTIVE To investigate the diagnosis, surgical management, and outcomes in patients with variant EEC. Variant presentations of the exstrophy-epispadias complex (EEC) span a wide range of abnormalities. The rarity and diversity of EEC variants can lead to challenges in the diagnosis and subsequent management of this population.

METHODS The authors reviewed an institutional database of 1336 EEC patients from 1975 to 2018 for variant presentations of EEC. Variant presentations included those with skin covered bladder exstrophy (BE), duplicate bladders, superior vesical fistula, and epispadias with major bladder prolapse. Surgical management and outcomes were assessed.

RESULTS In total, 44 EEC variants were identified. Nineteen (43%) presented with a skin-covered BE variant. Five patients presented with duplicate BE, while 6 presented with superior vesical fistula. Fourteen patients (32%) presented with epispadias with major bladder prolapse. Overall, 36 (82%) EEC variants underwent primary bladder closure, at a median of 135 days after birth (range 1-2010), with 21 (58%) undergoing pelvic osteotomy. Primary closures were successful in 89% of cases. Continence procedures were performed in 17 patients. This includes 5 patients who underwent bladder augmentation. However even without a continence procedure, continence with volitional voiding was found in 8 patients.

CONCLUSION The most common EEC variant is the skin-covered form of BE. In order to expedite appropriate management, accurate diagnosis upon initial presentation is crucial. Still, successful surgical reconstruction often results in continence that is similar to, or better than, nonvariant EEC presentations. UROLOGY 125: 184–190, 2019. © 2018 Published by Elsevier Inc.

The exstrophy-epispadias complex (EEC) is a group of rare congenital anomalies with a spectrum of musculoskeletal, gastrointestinal, and genitourinary abnormalities. Classic bladder exstrophy (CBE) is the most common form of EEC, with an incidence of 3.3 per 100,000 live births.[1] Rarer presentations of EEC include variant presentations of CBE and epispadias. Data regarding these variants are largely limited to case reports. An early investigation from Marshall and Meucke identified a number of variant presentations.[2] Lowentritt et al, first reported a classification scheme for variants of EEC, and included 19 CBE variants.[3] Herein, the authors report their institutions experience with a large cohort of EEC variants over a 40-year period.

CBE variants fall into 3 broad categories: skin-covered bladder exstrophy, superior vesical fissure (SVF), and duplicate bladder exstrophy. The skin-covered variant was first described by Mackenzie, and presents as an infraumbilical bulge of intact skin with laterally displaced rectus muscles (Fig. 1).[4] The bladder, directly beneath the skin, is typically intact and attached to the subcutaneous fascia. The SVF, presents as an abdominal wall defect communicating with the urinary bladder. The size of the defect has been used to distinguish between the fissure and fistula, but for practical purposes, the authors will refer to this variant as a SVF. Duplicate bladder exstrophy may take 2 broad forms: an anterior-posterior (AP) presentation, and a side-to-side presentation. In the former, a patch of exstrophic mucosa appears on the infraumbilical aspect of the abdomen with a second separate, closed bladder posteriorly.

The purpose of this study was to report the wide range of variant presentations, surgical management, and long-term urinary continence outcomes of this unique group.

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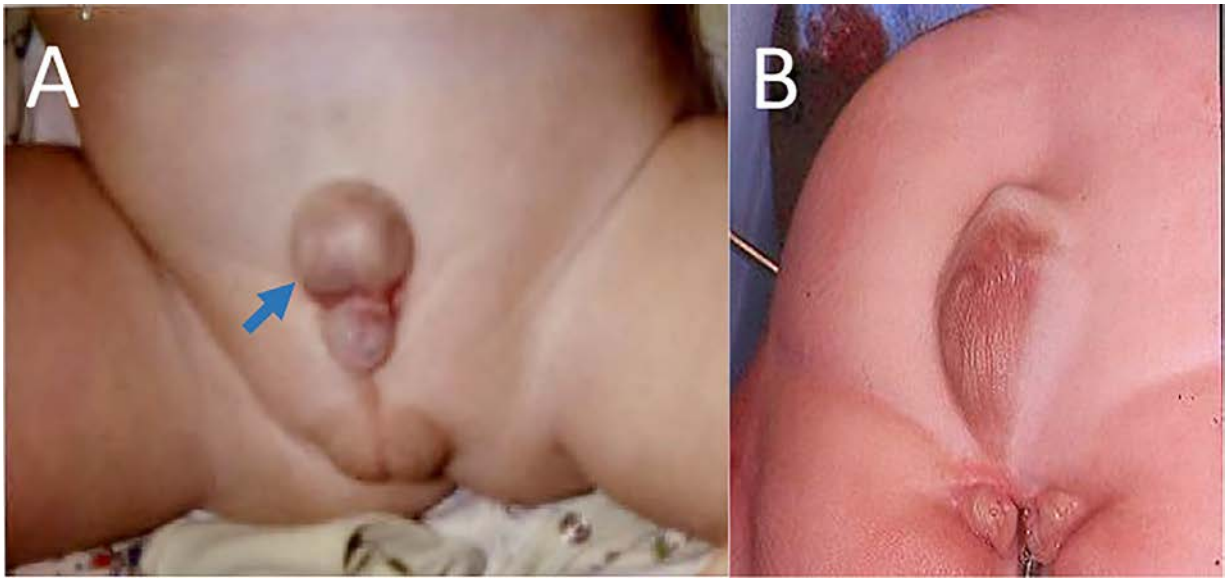

Figure 1. Skin covered variant in an infant male (A). The infraumbilical bulge with a thin membrane of skin is show by the blue arrow (A). In the female patient (B) the urethral sound can be seen against the infraumbilical bulge. (Color version available online.)

METHODS

The authors reviewed an Institutional Review Board approved database of 1336 EEC patients for those with variant presentations of EEC. Patients who presented to the authors' institution between 1975 and 2018 were included in the study. Cloacal exstrophy variants were not included in this study. The variant presentations were skin covered bladder exstrophy, duplicate bladder exstrophy, SVF, and epispadias with prolapsing bladder. Patients' records were reviewed for initial presentation, surgical management, and urinary continence outcomes. This report contains 19 patients the authors previously reported in 2005.[3]

All patients, whether for initial presentation or after referral/treatment, were evaluated by a senior pediatric urologist and a senior pediatric orthopedic surgeon. Initial evaluation involved a detailed prenatal and birth history when available, and physical examination. These patients may have been initially diagnosed and/or treated at a referring institution and then subsequently managed at the authors' institution. An examination under anesthesia with cystoscopy is typically performed by both the pediatric urologist. The pubic diastasis is measured radiographically, and the malleability of the pelvis is assessed by the pediatric orthopedic surgeon. Just as in CBE, patients with exstrophy variants who have a pubic diastasis of greater than 4 cm, have a nonmalleable pelvis, or undergo closure at an age greater than 3 days typically undergo pelvic osteotomy at the time of abdominal wall closure.

After a successful closure, patients are followed annually with cystoscopy and cystography. For the patients who were not continent after the closure, the bladder neck and bladder capacity were assessed for bladder neck reconstruction (BNR) candidacy. Urinary continence was assessed at least 6 months after any continence procedure(s). Patients who displayed volitional voiding per urethra and dry intervals greater than 3 hours, even without a continence procedure, were considered continent of urine.

RESULTS

Cohort Characteristics

Of the 1336 patients with EEC managed at the authors' institution, a total of 44 (3.2%) variant bladder exstrophy patients (27 males and 17 females) were included in this study. Table 1 displays the number of patients within each identified variant type and their respective surgical repair outcomes. The presentation and management of each variant category was also examined.

Skin-Covered Exstrophy

Of the 44 variant EEC patients, 19 (43%), 12 male and 7 female, were identified as skin-covered variants. The median pubic diastasis was 5 cm (IQR 4-6.8). Eight skin-covered patients (42%) had associated gastrointestinal anomalies (Supplementary Table 2). Seventeen patients (90%) underwent abdominal wall closure at a median age of 112 days (range 1-921). Eleven of these 17 closures (65%) were performed with a pelvic osteotomy. Of the 17 primary closures, 15 (88%) were successful. Two patients were seen in consultation only and lost to follow-up prior to their primary closure.

In total, 7 skin-covered variants underwent a procedure for urinary continence with a median follow-up of 10.2 years (range 0.3-35.7). Two patients, who previously had a successful closure with pelvic osteotomy underwent BNR at the age of 5.5 and 8.3 years, and remained incontinent at their most recent follow-up. Five patients underwent bladder neck transections (BNT) with continent urinary diversions and were continent. Of these 5 patients, 3 underwent augmentation cystoplasty (AC). There were 3 patients without a continence procedure who had dry intervals greater than 3 hours, and voided per urethra. Urinary continence status is summarized in Supplementary Table 1.

Anterior-Posterior Duplicate Exstrophy

Three patients, 2 male and 1 female, with duplicate exstrophy were identified. All 3 underwent successful closure, 2 with pelvic

Table 1. Displays the number of bladder exstrophy variants managed at the authors' institution and the outcomes of the respective surgical repairs

Variant Type	N, (%)	Closure, n (% of Group)	Age at Closure, Days (Range)	Pelvic Osteotomy, n (% of Closure)	Successful Primary Closure, n (% of Closure)
Skin-covered	19 (43%)	17 (89%)	112 (1-921)	11 (65%)	15 (88%)
Duplicate exstrophy (anterior-posterior)	3 (7%)	3 (100%)	23 (1-246)	2 (67%)	3 (100%)
Duplicate bladder (side-side)	2 (5%)	0	0	1 (50%)	0
Superior vesical fissure	6 (14%)	6 (100%)	1 (1-7)	1 (17%)	6 (100%)
Epispadias with bladder prolapse	14 (32%)	8 (87%)	287 (169-2010)	7 (88%)	8 (100%)

osteotomy (67%). One patient was previously reported in a case report.[5] In all cases, the anterior, rudimentary accessory bladder was excised. In 2 patients (male and female), both ureters inserted into the subjacent posterior bladder. In 1 male patient, the excised tissue was used for an epispadias repair and neourethra. One patient presented with the left ureter inserting into the anterior bladder. During the abdominal wall closure at 8 months, the left ureter was reimplanted into the posterior bladder, and the anterior bladder excised. A schematic of duplicate exstrophy presentations is shown in Figure 2. Follow-up time for duplicate exstrophy patients ranged from 6.4 to 25.9 years. One duplicate exstrophy patient is continent per urethra without a continence procedure. Two other patients underwent BNR, 1 with AC and stoma. All 3 patients are continent for greater than 3 hours; 2 void per urethra and 1 catheterizes per continent stoma.

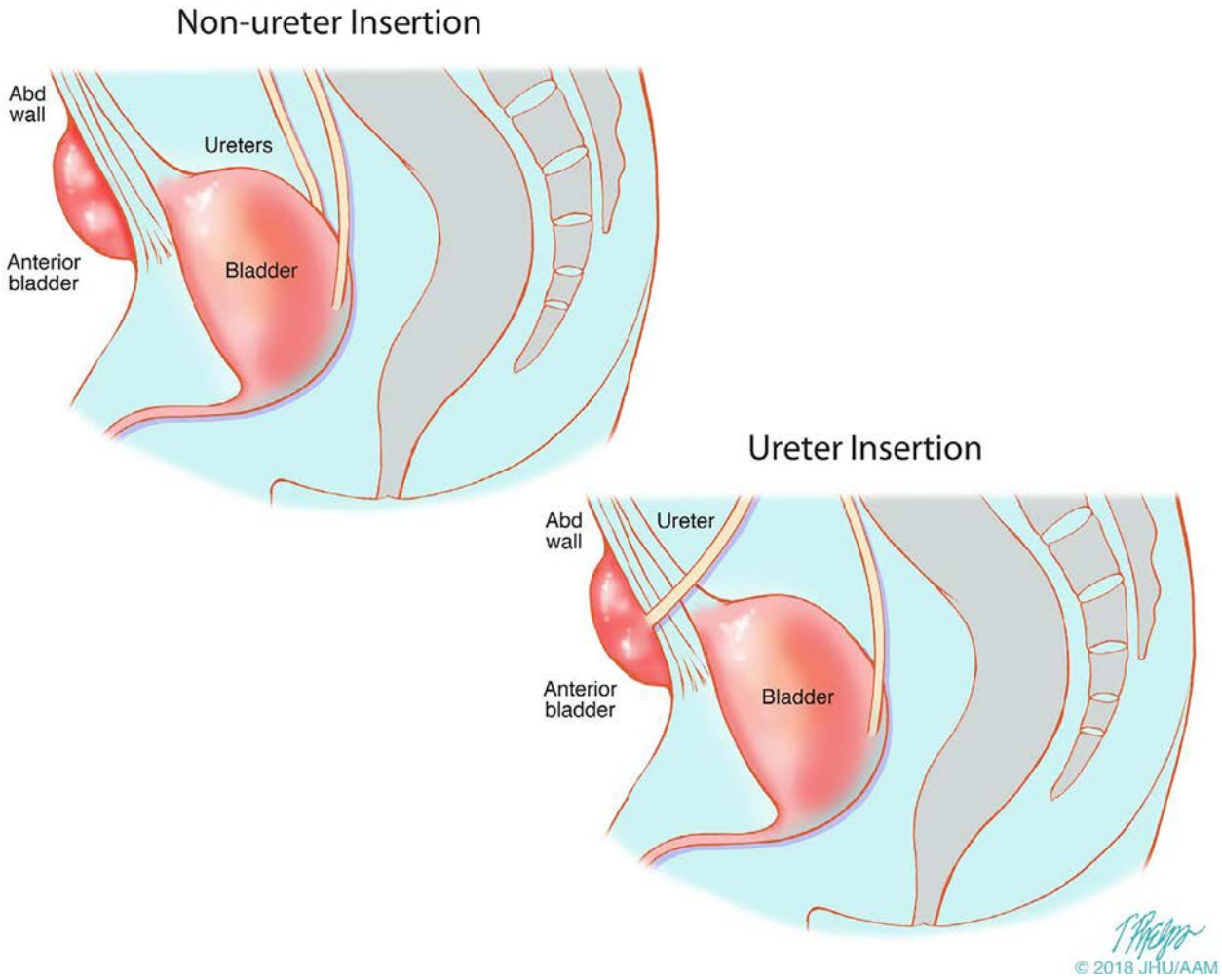

Figure 2. The presentation of duplicate exstrophy may be categorized by the insertion of the ureter. The anterior bladder without ureter insertion (left) is more common than anterior bladders with ureter insertion (right). (Color version available online.)

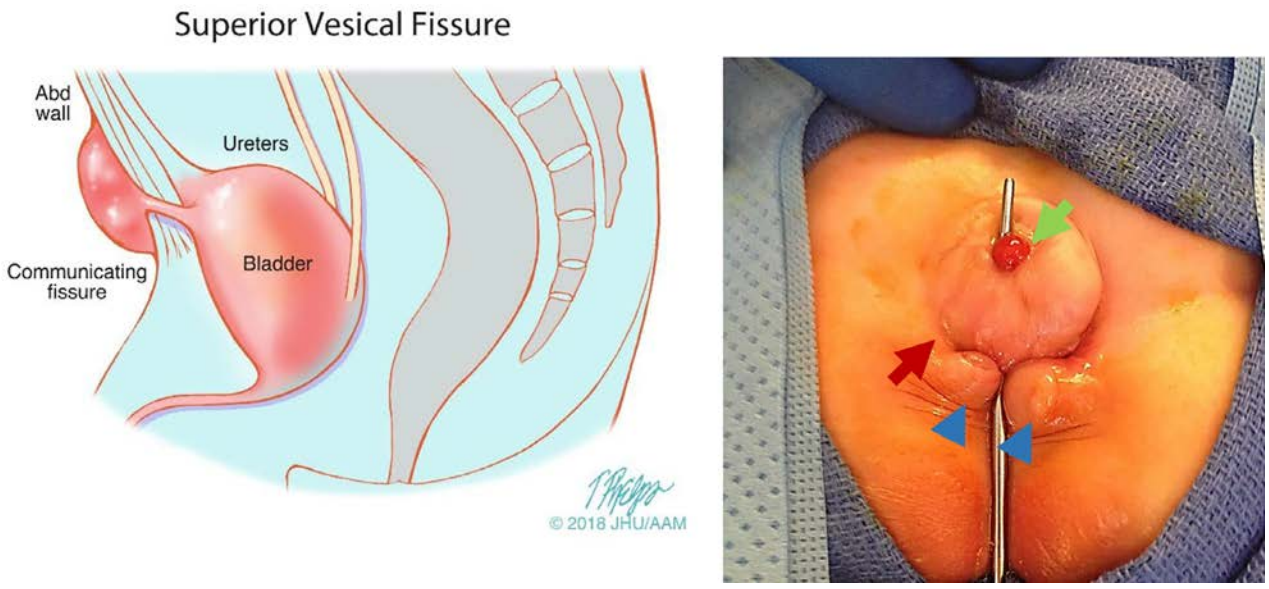

Figure 3. A schematic diagram of the superior vesical fissure is show (right). This depicts a posterior bladder with a fistulous communication to the skin. On the right, an infant male with superior vesical fissure and bifid phalli (blue arrows) is shown. The bladder (red arrow) is directly beneath the skin. The rectus abdominal muscles are diverged and a pubic diastasis of 4.6 cm is present. A bladder polyp (green arrow) is present where the bladder communicates with the skin. (Color version available online.)

Side-to-side Duplicate Exstrophy

Two patients, male and female, presented with a side-to-side duplicate bladder and duplicate genitalia. At the age of 1 years old, the male patient, with a 4 cm pubic diastasis underwent removal of the right bladder, ureter, and atrophic right kidney (Supplementary Figure 1). A right penectomy was performed and the hemi-scroti was approximated. The abdominal wall was adequately closed without osteotomy. The female patient, previously reported in a case report, initially underwent resection of duplicated bowel within the first month of birth without osteotomy.[6] No intervention for her duplicate bladder or Müllerian structures was performed. In adulthood, she was found to have a 10 cm pubic diastasis, which was corrected with bilateral pelvic osteotomies. Without any continence procedures, both patients are dry for greater than 3 hours and void per urethra.

Superior Vesical Fissure

A SVF was found in 6 patients, 5 male and 1 female (Fig. 3). Five patients had a repair of this defect between 1 and 7 days old by closure of the bladder defect and abdominal wall without osteotomy. One patient underwent closure at 8 months old with osteotomy. The female patient also presented with a small omphalocele, which was repaired at the same time. The median follow-up time was 16.6 years (range 3.6-36.3). Five patients are dry for greater than 3 hours. Four void spontaneously per urethra while 1 patient, who also underwent AC and continent urinary diversion voids via stomal catheterization. Only 2 patients underwent a BNR. The remaining patient is awaiting his BNR.

Epispadias With Major Bladder Prolapse

There were 14 patients (7 male and 7 female) with epispadias and bladder prolapse, of which 9 (64%) underwent a bladder repair at a median age of 287 days (range 169-2010) (Supplementary Figure 2). The median pubic diastasis was 4 cm. There was a significant delay in median age of bladder repair in patients with epispadias and bladder prolapse when compared to the other variant subtypes (287 vs 23 days; $P = 0.014$). Seven of these patients underwent pelvic osteotomy at the time of bladder repair as the median pubic diastasis was 4 cm. All of the female and 3 male patients had their epispadias repaired at the time of bladder repair. Two male patients underwent epispadias repair 7.5 and 37.6 months after bladder repair. Four patients (28.6%) with epispadias and bladder prolapse, 2 male and 2 female, underwent epispadias repair without bladder repair at a median age of 18.9 months (range 12.1-25.7). Finally, 1 patient was lost to follow-up immediately after presentation, and thus no surgical information is available. Three patients underwent a BNR, of which 2 are continent for greater than 3 hours and void per urethra. Four patients have undergone a BNT, AC, and CUD with appendicovesicostomy and are continent per stomal catheterization.

DISCUSSION

A classification scheme for exstrophy variants has been devised to categorize such presentations since proper recognition is important for subsequent management.[2] Importantly, these categories are not mutually exclusive, as a patient may exhibit characteristics of 2 or more variant presentations. Thus far, variants of bladder exstrophy have been classified as skin-covered, duplicate BE, or SVF. A variant of epispadias, presenting with prolapsing bladder, is related to bladder exstrophy since there is more bladder involvement than classic penopubic or complete epispadias along with a widened pubic diastasis. This study shows that despite the range in presentation, the surgical and urinary continence outcomes of variant

presentations are quite optimistic. Still, the approach to management of bladder exstrophy variants is quite similar to that of CBE. This is primarily because the wide pubic diastasis is a major driving force in the management of each variant. In fact, most patients within each variant underwent pelvic osteotomy for proper closure. So despite the range in presentations, the pelvic defect is a unifying consideration. Once the diastasis is addressed, the abdominal wall reconstruction is often successful, despite the fact that patients present at a wide range of ages.

The skin-covered exstrophy is the most common variant, in our series spanning 40 years. In addition to the urinary and musculoskeletal defects, a number of other anomalies may be found concomitantly. Sahoo et al described 5 patterns observed with skin-covered exstrophy, including a female preponderance, a split symphysis and low-set umbilicus, gastrointestinal anomalies, genitourinary tract anomalies, and a catch-all "other" anomalies.[7]

In the present study, the authors observed more males with skin-covered exstrophy than females (63% vs 37%), revealing a sex distribution that is similar to CBE and contrary to previous reports of skin-covered variants.[8] However, like observations in the literature, gastrointestinal anomalies without spinal defects were frequent, with anorectal malformations being the most common.[8] The incidence of imperforate anus was 16% in this group, nearly nine times the frequency of the same anomaly in CBE.[9] A number of embryologic hypotheses have been proposed for isolated cases of imperforate anus. In the setting of skin-covered bladder exstrophy, the theory postulated by Pauli[10] of lower mesodermal deficiency may align with embryologic mechanisms for exstrophy proposed by Marshall and Meucke.[2,11] A segment of sequestered bowel was also relatively common in these patients, and up to 20% of skin-covered variants in the literature present with this phenomenon. Secondary closure of the abdominal parieties during embryogenesis is thought to cause sequestration of visceral organs.[12] Renal anomalies are markedly more frequent in skin-covered variants compared to CBE. Renal defects are present in 2.8% of patients with CBE, while over 40% of skin-covered variants had some renal anomaly—13 times that found in CBE.[13]

The skin covered variant of exstrophy requires formal abdominal wall closure to approximate the rectus muscles and fascia, and position the bladder deep within the pelvis. Pelvic osteotomy is frequently performed because: (1) the primary closure was often performed more than 72 hours after birth and (2) the pubic diastasis may be more than 4 cm. Meeting these criteria, or having a pelvis that is not easily approximated during an examination under anesthesia, will necessitate a pelvic osteotomy for closure.[14] Likewise, 2 patients were referred to the authors' institution following a failure of their primary abdominal wall closure, neither of which underwent pelvic osteotomy. The authors speculate that a pelvic osteotomy results in a more secure closure.

Skin-covered exstrophy variants achieve a varying degree of urinary continence. A number of reports show that continence and urethral voiding can be attained without a continence.[8,15] In this series, 4 skin-covered exstrophy patients achieved dry intervals greater than 3 hours and voided per urethra. Notably, these patients had a successful primary abdominal wall closure. Minimal bladder neck involvement may also contribute to optimistic continence outcomes. Other factors are still necessary to consider for urinary continence. For example, 5 patients in this series required a BNT and CUD due to after poor bladder growth and leaking of urine. As such, close follow-up and assessment of continence is necessary, even in patients with a skin-covered variant.

This series presents 3 patients with AP duplicate bladder exstrophy, but with 2 different types. The embryologic origin of the AP duplicate bladder variant remains uncertain. Marshall and Meucke[2] report that this is an extension of the SVF variant, while Sheldon et al[16] speculate that this is a formal duplication of the bladder in a coronal plane from a frontal septation. Still, the AP duplicate bladder variant may be defined by the insertion of the ureters. The anterior exstrophic bladder may be either ureteral-inserting or nonureteral-inserting. These 2 presentations can be viewed in Figure 2. The nonureteral inserting is the most common with a patch of dry mucosa on the abdomen.[17] With such cases, the anterior accessory bladder is typically excised and the abdominal wall defect is repaired. This may also require pelvic osteotomy to address the musculoskeletal defects. The other type of AP duplicate exstrophy variant occurs when a ureter inserts into the anterior exstrophic bladder. The contralateral ureter inserts into the posterior, otherwise normal bladder. This form is excessively rare. Repair of this presentation involves reimplantation of the anterior ureter into the subjacent bladder.

All 3 AP duplicate exstrophy patients attained urinary continence. Two required a BNR (1 with AC and continent mitrofanoff diversion) for continence management. Bladder exstrophy and variants may exhibit pathologies beyond gross anatomic presentations. As such, bladder growth and continence should be assessed even in exstrophy variants that appear to have relatively benign manifestations.

We also present here the collateral or side-to-side variant of bladder exstrophy, in which 2 closed bladders are present within the pelvis. Ninety percent of these variants may also present with other congenital anomalies, including duplication of the external genitalia.[18] Indeed, both patients in this series had duplicated genitalia. In the few reported cases of females with this condition, no intervention is necessary for the duplicate genitourinary system[19]; however, a recent prior report describes joining the bladders, resecting the duplicated urethra, and creating a continent appendicovesicostomy.[20] Management is quite individualized depending on the presentation. The male patient in the present study underwent cystoprostatectomy and penectomy of the duplicate system as well as

removal of the duplicated anorectum. However, in a similarly presenting patient, Azmy and colleagues reported joining the 2 bladders and retaining the duplicate penis.[21] Over 40% of patients with duplicate bladders may also have anorectal malformations or duplications of the hindgut. It has been hypothesized that during development, incomplete "twinning" of the primitive hindgut, from which the bladder and urethra derive, prevents normal fusion of the caudally advancing urorectal septum. In this manner, duplicated hindgut derivatives and lower urinary tract structures may occur. Urorectal abnormalities may also lead to Müllerian duct separation, thus permitting duplicated Müllerian derivatives in females.[22,23] Regardless of embryologic theories, duplicate bladders are exceptionally rare, and careful assessment of the presentation with an individualized surgical approach will result in satisfactory outcomes.

The SVF variant of exstrophy presents with a bladder defect communicating anteriorly with the skin.[24] This defect also has a wide range of presentations, from a small fistulous communication to a large eversion of bladder mucosa.[25] Because the bladder neck is not characteristically involved, continence and volitional voiding may be attained without a continence procedure. In very rare instances when the bladder neck is involved, it is known as an inferior vesical fissure.

Multiple variant presentations within the epispadias spectrum have been described.[26-28] An epispadias with bladder prolapse has been classified as a variant of penopubic epispadias in males or complete epispadias in females; however, there are close structural parallels to bladder exstrophy.[3] Frequently, the repair involves surgical correction of the bladder prolapse, and pelvic osteotomy for apposition of the pubic symphysis. Nine patients in this series underwent surgical repair of the prolapsed bladder, 7 with pelvic osteotomy, highlighting the need to address bladder involvement and associated musculoskeletal defects. The challenge in this variant is timely diagnosis, and the bladder defect, unfortunately, may not be noticed until later. In fact, prolapse may only present with crying, straining, or other Valsalva maneuvers. As the classic form of complete epispadias in females may not be apparent until the time of toilet training in females, the diagnosis of bladder prolapse may be further delayed. Like CBE, the continence outcomes may range and are dependent on the growth and quality of the bladder.

Though BE variants may have many different presentations, their surgical managements share the same principles as the management of CBE. Particularly, (1) the pelvic defect must be addressed, with bilateral osteotomy when indicated; (2) radical dissection of the bladder and urogenital fibers from the pubis to the levator hiatus; (3) appropriate draining of the ureters; and (4) a secure abdominal wall closure. While there are nuances in the closure, these shared principles are paramount for a successful outcome in BE variants.

CONCLUSION

Variants of bladder exstrophy are rare. Proper and early recognition of these infrequent presentations if crucial for appropriate management. Surgical repair is often successful and urinary continence can be achieved in many without a continence procedure. Still, assessments of bladder quality, growth and dry intervals are necessary as some bladder exstrophy variants will require an outlet procedure and/or augmentation to be dry.

SUPPLEMENTARY MATERIALS

Supplementary material associated with this article can be found, in the online version, at https://doi.org/10.1016/j.urology.2018.10.049.

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EDITORIAL COMMENT

This manuscript is extremely valuable as it brings this unique cohort to the rest of the pediatric urology community. It is unlikely this type of experience will be replicated.

One of the takeaways from this paper is that pubic separation remains an important consideration in these variants and many patients still require osteotomies similar to the classic exstrophy patients.

I would encourage the authors to publish an atlas and/or videos of the steps of the procedures they have used in correcting these variants.

Lastly, it is undoubtedly disheartening for parents to hear their child has a condition that is even rarer than the rare classic bladder exstrophy. This data can at least reassure families that these anatomic abnormalities can be corrected surgically and that ultimate continence can be achieved even if the repair is not done in the neonate.

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AUTHOR REPLY

Indeed, in both bladder exstrophy and epispadias variants, the pubic separation is an important consideration in the reconstructive process. Some centers are performing pelvic osteotomies in the epispadias population.[1] As addressing the bony pelvic abnormalities is a critical step in exstrophy management, this principle certainly applies in even rare exstrophy variants.

We appreciate the thought of offering an atlas or video of the surgical procedure, and it may be a future endeavor. The wide range of variant presentations may seem daunting, however, we do maintain that the unifying characteristic of the variants, as well as classic bladder exstrophy, is the correction of the widened pubic diastasis.

Finally, we completely agree that this data do show optimistic outcomes for these patients. Though rarer than the classic form of bladder exstrophy, the infrequent bladder neck involvement in the variant form appears to be a "silver lining" with regards to urinary continence. Surely this will be useful in parent counseling. Still, assessment of the patient's bladder and bladder neck function are paramount for eventually establishing urinary continence.

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