Urethral duplication in an adult male: anatomic variants and operative approach to reconstruction

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Duplicated anterior urethras are a rare congenital anomaly that is exceedingly uncommon among adult patients, as surgical correction is often completed during childhood. We present the case of a 33-year-old man with uncorrected duplicated anterior urethras and urinary retention due to severe urethral stricture disease involving both of his uncorrected duplicated anterior urethras. This report highlights an operative management strategy for reconstruction to create a single, unobstructed urethra that terminates in an orthotopic meatus. Further, we provide an overview of the anatomic variants of urethral duplication.

Key Words: anterior urethral stricture, abnormalities, congenital, urethral obstruction

Introduction

Urethral duplication is a rare congenital anomaly that is most commonly reported in the pediatric literature. There are a variety of anatomic variants that range from blind-ending incomplete duplication to complete caudal duplication of the bladder and urethras.[1] Duplication most often occurs in the sagittal plane with the ventral urethra being the more functional conduit.[2] Adult cases of symptomatic duplication are quite rare, as boys that have symptoms often undergo urethral reconstruction during childhood.

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We present an operative management strategy for duplicated anterior urethras in an adult male that utilizes a combination of reconstructive techniques. Further, this report provides a description of the anatomic variants of urethral duplication.

Case report

The patient is a 33-year-old male with a history of VATER association (vertebral anomalies, anal atresia, cardiac malformations, trachea-esophageal fistula, and renal anomalies). His phenotype included high grade left ureteral reflux necessitating ureteral re-implantation as a child with subsequent moderate to severe left hydroureteronephrosis, left cryptorchidism requiring orchidopexy, and imperforate anus that was surgically repaired. He has neurogenic bladder and voided through an orthotopic and subcoronal

meatus since childhood with valsalva maneuvers that produced a weak urinary stream and sensation of incomplete emptying. His urinary urgency incontinence has been managed with oxybutynin. He has a long history of recurrent urinary tract infections for which he is on chronic suppressive antibiotics with two episodes of pyelonephritis in the past year. He has a history of infertility necessitating microTESE surgery.

He developed urinary retention and a suprapubic tube was placed within the past year. The dorsal urethra had an orthotopic meatus and 3 cm of normal caliber distal penile and fossa navicularis urethra. The ventral urethral meatus was subcoronal and approximately 5 Fr in size. Antegrade cystoscopy via the suprapubic tube tract identified bilateral patulous ureteral orifices, a single, intact bladder neck, and a 3 cm stone within the single prostatic urethra. Retrograde urethrogram demonstrated completely duplicated anterior urethras arising from the membranous urethra to an orthotopic and an ectopic urethral meatus, Figure 1a. The dorsal urethra had a long segment stricture from the distal penile urethra, extending to the membranous urethra and drained via the orthotopic meatus, while the ventral urethra opened via an ectopic, stenotic subcoronal meatus with panurethral stricture disease to the membranous urethra. Laser lithotripsy of the prostatic urethral stone was performed via the suprapubic tube tract prior to urethroplasty.

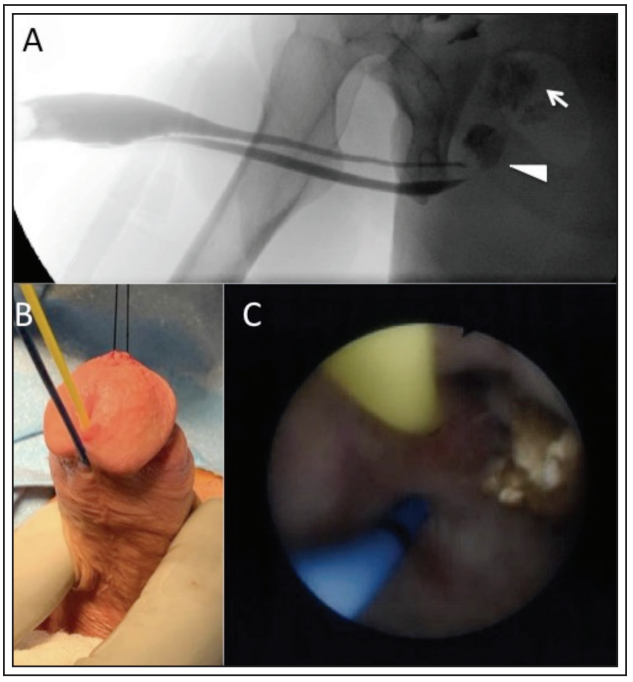

Figure 1. (A) Preoperative retrograde urethrogram illustrating duplicated urethras in a sagittal plane with strictured segments from the distal pendulous urethra to membranous urethra. The common membranous urethra contains a stone (triangle) and intraprostatic calcifications are demonstrated (arrow). **(B)** 5 Fr open ended catheters in the dorsal urethra with an orthotopic meatus (yellow) and the ventral urethra with an ectopic subcoronal meatus (blue). **(C)** Antegrade cystoscopy showing duplication arising at the bulbomembranous junction with a membranous urethral stone.

After discussing potential management options including cystectomy with ileal conduit, creation of a catheterizable channel, or continued suprapubic cystostomy drainage, the patient ultimately expressed his desire to void via his orthotopic meatus. He elected to proceed with reconstruction of his duplicated anterior urethras.

The patient was brought to the operating room, administered general anesthesia and culture directed antibiotics, and was placed in the dorsal lithotomy position. Open-ended catheters (5 Fr) were placed across both urethras into the bladder. Antegrade cystoscopy via the suprapubic tract identified both catheters joining in a common membranous urethra 1 cm distal to the verumontanum as well as a recurrent membranous urethral stone, Figure 1b and 1c.

A ventral midline incision was made from the distal penile shaft to the perineum. The urethras were in a sagittal (dorsal and ventral) orientation. The urethra with a subcoronal meatus was located ventrally and had a normal corpus spongiosum, which was dissected free to separate the two urethras from the distal penile urethra to the bulbomembranous junction. In contrast, the dorsal urethra with the orthotopic meatus lacked a normal corpus spongiosum and was surrounded by fibrotic tissue that was densely adherent to the tunical albuginea of the corpora cavernosa. This precluded it from being freed circumferentially. A longitudinal urethrotomy was made on the dorsal aspect of the ventral urethra from the membranous urethra to a point 1 cm proximal to the coronal sulcus. A similar urethrotomy was made on the ventral aspect of the dorsal urethra, Figure 2a. The membranous urethral stone was extracted.

A 3 cm segment of the proximal bulb of the dorsal urethra was obliterated and therefore excised. However, the remaining dorsal and the entirety of the ventral urethral plate appeared healthy (0.5 cm or greater in width) allowing for them to be sutured together longitudinally with 5-0 monocryl to form a single urethral plate from the distal penile to the mid bulbar urethra, Figure 2b. Distally, the urethra of the orthotopic meatus was opened ventrally and combined with the ventral urethra, while the mucosa of the

ventral urethra was excised to form a single urethra with an orthotopic meatus. The combined urethral plate was tubularized with 5-0 monocryl down to the mid bulbar urethra and accommodated a 24 Fr bougie á boule sound.

There was a 4 cm area of proximal bulbar urethra that was excised in its entirety due to severe fibrosis and narrow plate (1-3 mm wide). This was replaced with a 4.0 cm long by 2.0 cm wide buccal mucosa graft, Figure 2c. The dorsal graft bed was largely avascular given the dense fibrosis surrounding the excised portion of the dorsal urethra. Therefore, a healthy, vascularized graft bed was created by rotating the bulbospongiosus muscle over top of the corporal bodies, while keeping the blood supply intact to provide a healthy bed for the buccal mucosa graft. The ventral urethra was closed over the dorsal buccal mucosa graft with 5-0 monocryl suture and the urethral anastomosis was completed, Figure 2d. The penile urethral mucosa was quite thin necessitating coverage with two dartos interposition flaps. A series of interrupted and running 4-0 monocryl sutures were used to close the penile and scrotal skin. Proximally, the remaining bulbospongiosus muscle was used to cover the bulbar urethra followed by closure of Colles fascia and skin with 4-0 vicryl. A 14 Fr urethral Foley catheter was placed and a 16 Fr suprapubic tube was replaced.

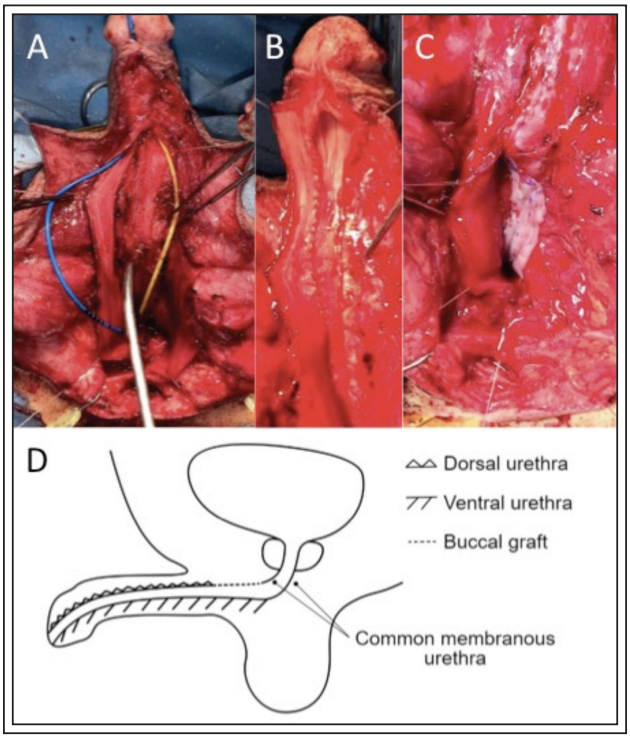

Figure 2. Intraoperative photos demonstrating **(A)** the urethral plates of the ventral urethra with normal corpus spongiosum (blue catheter) and the dorsal urethra with fibrotic attachments to the corpus cavernosum (yellow catheter). **(B)** Longitudinal anastomosis of the dorsal and ventral urethral plates. **(C)** Proximal ventral bulbar urethra and dorsal buccal mucosa graft (buccal graft supported by bulbospongiosus muscle). **(D)** Sagittal illustration showing the components that contributed to the reconstructed urethra including the plates from the dorsal and ventral urethras along with the dorsal buccal mucosa graft.

The patient was discharged home on postoperative day zero with chlorhexadine gluconate 0.12% oral rinse, "magic" mouthwash (diphenhydramine, lidocaine, Maalox oral rinse), and acetaminophen.

His urethral catheter was removed for a voiding cystourethrogram 3 weeks postoperatively, Figure 3. This demonstrated a widely patent urethra with no contrast extravasation. The suprapubic catheter was removed at that time. He did not require narcotic pain medication postoperatively.

Cystoscopy at 5 months postoperatively demonstrated a patent reconstructed urethra. Given his neurogenic bladder, he utilizes valsalva maneuvers to void via his orthotopic meatus. He is able to empty his bladder well, is continent and is very satisfied with his voiding status. His erectile function is normal and he is sexually active.

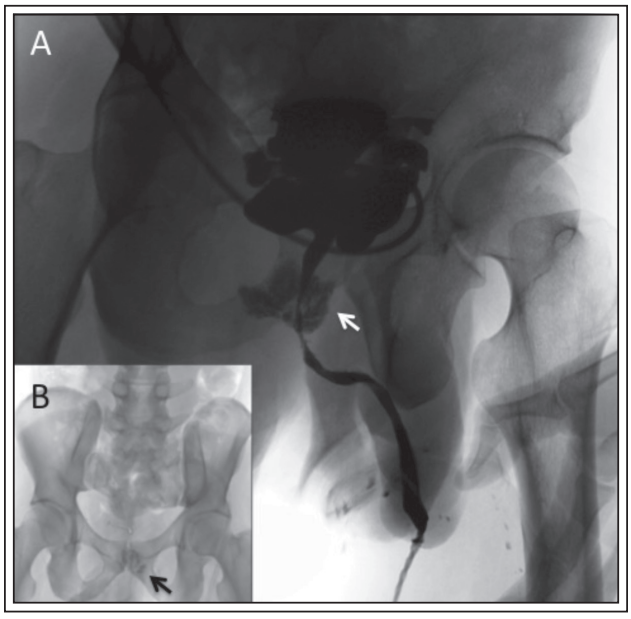

Figure 3. (A) Postoperative voiding cystourethrogram demonstrating a trabeculated bladder and a widely patent, single reconstructed urethra. Intraprostatic calcifications (arrow) were identified on **(B)** scout film.

Discussion

Urethral duplication is a rare entity in the pediatric literature, with just over 170 cases reported.[2] Symptomatic duplicated urethras are often surgically repaired during childhood. Therefore, an adult with symptomatic duplicated urethras is extremely rare. We highlight important considerations involving the complicated anatomy and operative technique to reconstruct these difficult strictures.

The embryonic development of duplicated urethras is not fully established and may vary according to the type of duplication. In general, it is thought to involve misalignment of the terminal portion of the cloacal membrane with the urogenital sinus and genital tubercle.[3] The majority of cases of urethral duplication occur in the sagittal plain with a dorsal and ventral urethra. Effman et al developed a classification system for duplicated urethras ranging from type 1 involving incomplete duplication with a single blind ended urethra, to type 3 involving complete caudal duplication of the urethras and bladder. Type 2 duplication is subdivided into type 2A-I with complete duplication of both urethras beginning at the bladder, type 2A-II with incomplete duplication arising from the proximal urethra, type 2A-IIY involving termination of the ventral urethra in the perineum, and type 2B with duplicated proximal urethras that converge into a single distal channel, Figure 4.[1] The current case is most consistent with a type 2A-II, as duplication arose at the level of the membranous urethra. Small series in the pediatric literature suggest that type 2 duplication is most common and occurs in approximately 63% of cases, while type 1 is present in 25%.[2]

Salle et al have outlined three basic principles when evaluating a patient with duplicated urethras – establish an anatomical understanding of the abnormality, identify the functional urethra, and investigate other anomalies.[2] Retrograde urethrogram is useful to delineate the anatomy and should be performed via the orthotopic and the ectopic meatus, which can be identified on detailed physical exam. Voiding cystourethrogram can be useful to distinguish between blind ending urethras versus narrow caliber urethral strictures that inhibit retrograde contrast flow. Cystourethroscopy allows for further evaluation of external sphincter function and bladder neck competence.

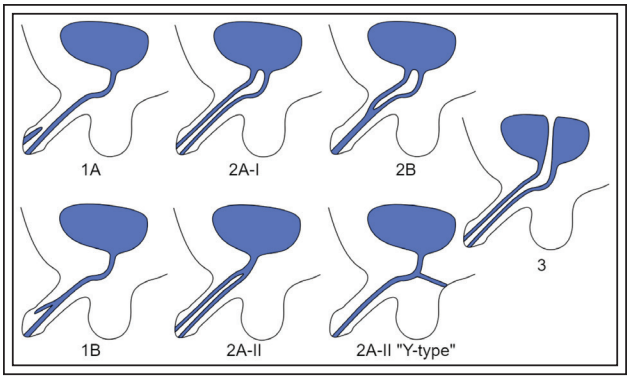

Figure 4. Effman classification system for duplicated urethras.[1]

Abdominal imaging should be performed to identify any upper urinary tract abnormalities. At minimum, an ultrasound should be obtained with consideration of computed tomography (CT) if there is suspicion for additional abnormalities. In this case, the patient had a history of high grade left vesicoureteral reflux and had undergone a left ureteral re-implant with subsequent development of moderate to severe ipsilateral hydroureteronephrosis identified on CT scan and limited split renal function (19%) on diuretic renal scan.

The ventral urethra is most often the functional urethra and is associated with the external sphincter and the verumontanum.[2] Further, the functional urethra is often ectopic with the more orthotopic urethra being hypoplastic. The dorsal urethra can also be surrounded by tunica albuginea between the corpora cavernosa.[4] This was observed intraoperatively in the current case, as the dorsal urethra with the orthotopic meatus lacked a normal corpus spongiosum and was surrounded by fibrosis that was fused to the corpora cavernosa. In contrast, the ventral urethra had a healthy appearing corpus spongiosum but terminated in a subcoronal hypospadic opening.

Urethral duplication has been associated with other congenital anomalies. Bladder duplication is a rare occurrence in the setting of completely duplicated urethras that is most often observed in the coronal orientation with each bladder associated with a single ureter.[5] Less common is a sagittal orientation of duplicated bladders with a non-functional anterior bladder.[6] Several reports and small series have described pediatric patients with VATER association and duplicated urethras.[7,8] The current case had several VATER anomalies including imperforate anus and vesicoureteral reflux.

Surgical repair in the adult patient is complex and can require a combination of reconstructive techniques. There have been multiple proposed techniques for reconstruction in the pediatric literature including the use of preputial island flaps, scrotal flaps, and buccal mucosa grafts as well as anastomosis of the dorsal and

ventral urethras in an end-to-side fashion or complete excision of the non-functional dorsal urethra.[4,9,10]

In this case, the ventral and dorsal urethral plates were > 5 mm and therefore adequate for a longitudinal anastomosis to combine the two urethras into one as far proximally as the mid bulbar urethra. The dorsal urethra had an obliterated proximal bulbar urethra and was excised, which allowed for the ventral urethra to be augmented with a dorsal onlay buccal mucosa graft. Blood supply was an important consideration at several points. First, the dorsal urethra lacked a healthy corpus spongiosum and was encased in fibrosis. To avoid further compromise of an already anomalous blood supply, the dorsal urethra was left in situ and not mobilized. Second, the graft bed in the proximal bulbar urethra was densely fibrotic. A bulbospongiosus muscle flap interposition was therefore utilized to provide a healthy, vascularized graft bed for optimized graft take.

In conclusion, we describe the operative management of uncorrected urethral duplication in an adult. There are multiple anatomic variations in urethral duplication, which can influence the surgical approach in symptomatic patients. Ultimately, surgical repair in the adult can be a successful endeavor that may require a combination of reconstructive techniques in order to create a single channel to void. □

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