Images — A series of congenital mesonephric/Wolffian duct abnormalities in the pediatric population

Meghan Cooper, DO[1]; Calvin Guiterrez, MS4[2]; Hubert Swana, MD[3]; Mark Rich, MD[3]; Lucas Wiegand, MD[1]

[1]Department of Urology, University of South Florida, Tampa, FL, United States; [2]University of Central Florida, Orange County, FL, United States; [3]Arnold Palmer Children's Hospital, Orlando, FL, United States

Cite as: Cooper M, Guiterrez C, Swana H, et al. Images — A series of congenital mesonephric/Wolffian duct abnormalities in the pediatric population. *Can Urol Assoc J* 2020;14(11):E611-2. http://dx.doi.org/10.5489/cuaj.6344

Published online June 5, 2020

Case presentation 1

A ten-month-old male developed a febrile urinary tract infection (UTI) and urinary retention after hypospadias repair. Urethral catheterization was performed without difficulty. Prenatal ultrasounds had demonstrated a solitary right kidney. A repeat ultrasound revealed a solitary right kidney and a tubular pelvic structure on the left side behind the bladder. Voiding cystourethrogram (VCUG) was obtained. This revealed an irregular bladder contour and reflux into what was believed to be dilated, duplicated left ureters that ended blindly in the retroperitoneum.

The patient was managed with antibiotic prophylaxis initially and, over the next several months, developed recurrent UTIs and urinary retention. At 18 months of age, the patient was taken to the operating room for cystoscopy and left retrograde urography that revealed a left-sided tubular structure that coursed cranially into the retroperitoneum then downward into the pelvis. Ureterovasal fusion was demonstrated with retrograde filling of the epididymis (Fig. 1).

Robotic-assisted laparoscopic excision of the left ureter was performed. Intraoperative examination of the renal fossa did not reveal a renal remnant. At two-year followup, he did well with resolution of his retention and infections.

Case presentation 2

A two-year-old boy was referred to the pediatric urology clinic for recurrent UTIs and episodes of right epididymo-orchitis. He was born with imperforate anus and underwent posterior sagittal anorectal repair at an outside institution.

Original ultrasound revealed a hydronephrotic right kidney with parenchymal thinning. Voiding cystourethrogram revealed grade 5 reflux of the right kidney, and diuretic renal scanning revealed <5% minimal function of the right kidney. Diagnostic cystoscopy and right retrograde were performed and demonstrated a dilated single ureter with grade 5 reflux and distal ureterovasal fusion. On the right side, the ureteral orifice was cannulated, and a sensor wire passed into the right ureterovesical junction and the ureterovasal fusion as it coursed retroperitoneally through the inguinal canal to the epididymis in the right scrotum. Injection of contrast filled the vas deferens and dilated proximal ureter (Fig. 2).

The patient underwent complete right nephroureterectomy and excision of distal ureterovasal fusion via a flank approach with a second lower inguinal incision. Postoperatively, the patient remained infection-free. He continued to do well other than requiring clean intermittent catherization due to neurogenic bladder from a tethered cord.

Discussion

The prevalence of genitourinary abnormalities in newborns is estimated to be 0.1–1%. Although these anomalies are relatively rare and often subclinical in nature, they can initially present incidentally during a workup for other pathologies.[1]

The establishment of the male internal reproductive system involves two key events: the formation of the testis and the maintenance and differentiation of the Wolffian (mesonephric) duct. The Wolffian duct is central in the development of the urogenital system and its role is well-established.[2] Embryologically, the mesonephric duct develops into the fetal kidney and bladder trigone, and the distal Wolffian ducts differentiate into the urogenital structures, including the epididymis, vas deferens, seminal vesicles, and ejaculatory ducts.[3] Although not essential for initial Wolffian duct formation, the later differentiation of this embryological structure relies on androgens derived from the Leydig cells.[2]

Genitourinary abnormalities due to Wolffian (mesonephric) duct malformations in pediatrics can present with symptoms of a UTI, epididymitis, during surgical exploration for undescended testes, or in association with other prenatal and postnatal malformations of the genitourinary mesonephric system.[1] In adults, cases can often present in a delayed fashion, typically

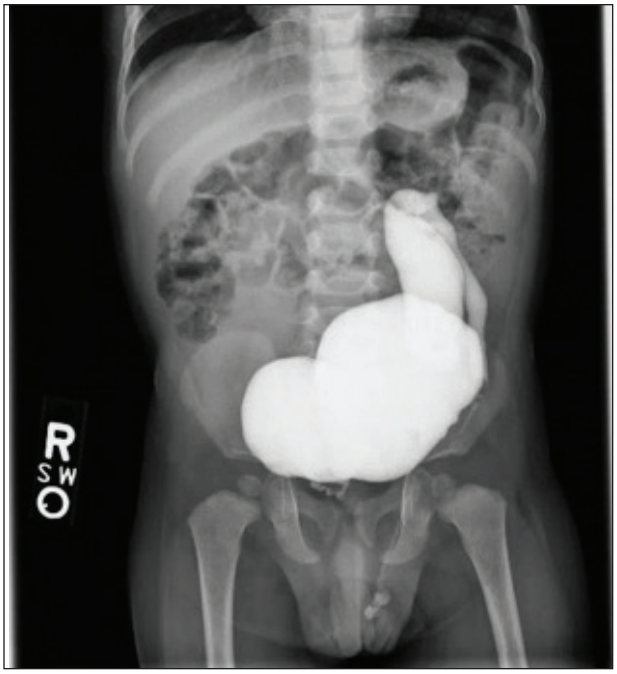

Fig. 1. Case 1: Ureterovasal fusion was demonstrated with retrograde filling of the epididymis.

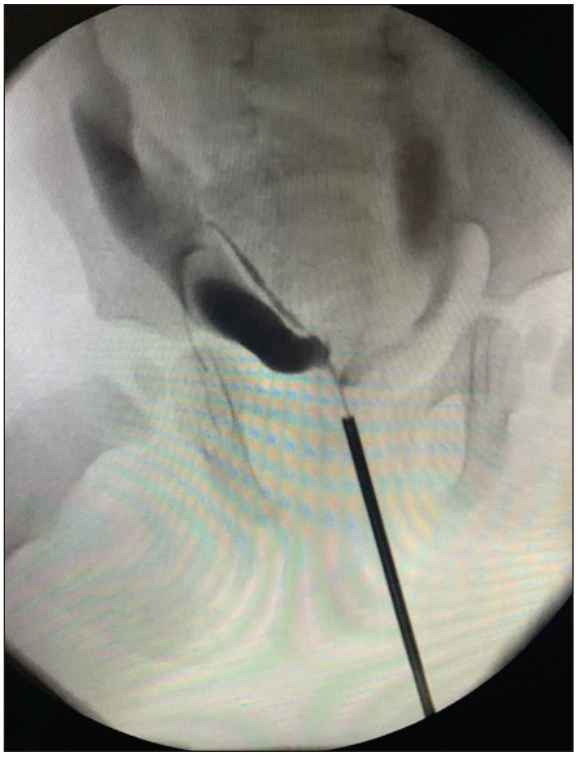

Fig. 2. Case 2: Injection of contrast filled the vas deferens and dilated proximal ureter.

detected with the onset of sexual activity. Adults with Wolffian duct abnormalities often present with symptoms of bladder dysfunction, UTIs, pelvic and ejaculatory pain, or infertility.[4]

Ectopic ureters with ureterovasal fusion are uncommon and often associated with ipsilateral renal agenesis, as well as anorectal abnormalities.[5] The embryological etiology of this malformation results from the ureteral bud developing more cephalad on the mesonephric duct preventing it from achieving an independent orifice within the urinary triad.[6] Therefore, the ureter remains attached to the distal Wolffian duct, which ultimately becomes the seminal vesicle and ejaculatory duct. A rudimentary renal unit is often detected on the involved side and its ureter fuses with the seminal vesicle or ejaculatory duct.[3]

In these cases, there was noted to be ectopic ureteral insertion into the Wolffian duct structures. Each case offers a unique presentation of this genitourinary anomaly, as the first patient had a solitary kidney with no renal remnant found, while the second case demonstrated a hydronephrotic, non-functional renal unit. However, both patients were diagnosed during a workup for recurrent UTIs. This supports the concept that these anomalies are often diagnosed via ultrasound during investigation for UTI, epididymitis, or flank pain in adolescent males; in older males, the symptoms often present as prostatitis, pain on ejaculation, or infertility.

Conclusions

Our two case reports demonstrate the rare clinical presentation of ureterovasal fusion in the adolescent male and offer interesting pathological anatomy. As exhibited in our cases, surgical intervention is often warranted in these patients rather than conservative therapy due to the persistence of symptoms and/or recurrence of infection.

Competing interests: The authors report no competing personal or financial interests related to this work.

This paper has been peer-reviewed

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Correspondence: Dr. Meghan Cooper, Department of Urology, University of South Florida Morsani College of Medicine, Tampa, FL, United States; Meghan.a.cooper@gmail.com