

Wolffian system agenesis with contralateral ejaculatory duct stenosis: First case in the literature

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ABSTRACT

Congenital agenesis of seminal vesicles and vas deferens is rare and its co-existence with structural abnormality on the contralateral side is even rarer. We report a case of an adult male presenting with infertility due to obstructive azoospermia, in which transrectal ultrasonography (TRUS) and magnetic resonance imaging (MRI) revealed the agenesis of Wolffian derivatives on the left side with stenosis of the ejaculatory duct on the contralateral side. This dual combination of uncommon anomalies is the rarest phenomenon and poses as a diagnostic dilemma for the radiologist. Emphasis is given to MRI as a one-stop shop modality in the evaluation of complete genitourinary system; as well as on the importance of TRUS as a screening tool in patients with infertility due to obstructive azoospermia.

Keywords: Azoospermia; ejaculatory duct; anomaly; seminal vesicles; Wolffian duct.

Introduction

Genitourinary abnormalities due to embryological malformations in the Wolffian (mesonephric) duct derivatives are usually encountered in an adolescent with a primary complaint of infertility.^[1] Timely detection of the anomaly can be helpful for the treatment planning. Transrectal ultrasonography (TRUS), due to its wider availability, is the imaging modality of choice for screening in this group of patients; however, it is underutilized mainly because of the patient discomfort and due to the lack of skilled imaging specialists. Role of pelvic magnetic resonance imaging (MRI) is indispensable, not only in delineating the genitourinary anatomy but also acting as a visual delectation for the concerned urologist and in treatment planning. MRI also provides a complete screen-shot from the level of kidneys up to the scrotum where all the findings can be consummated to reach an accurate diagnosis.

In this manuscript, we report an exclusive scenario where TRUS revealed one-sided ejaculatory duct stenosis and MRI provided information regarding agenesis of the Wolffian duct derivatives on the contralateral side. To our knowledge, none of the available literature documents the combination of above two rare events in the same patient.

Case presentation

A 36-year-old male presented to the Department of Urology of our hospital with complaints of infertility and low volume ejaculate. His history revealed six years of marriage and the couple had been evaluated under the basic infertility protocol. The patient was aware of the presence of male factor infertility and he had been referred by a local gynecologist for consultation with the urologist. Clinical examination was unremarkable with the normal size of the external genitalia and normal history of penile tumescence. Semen analysis revealed

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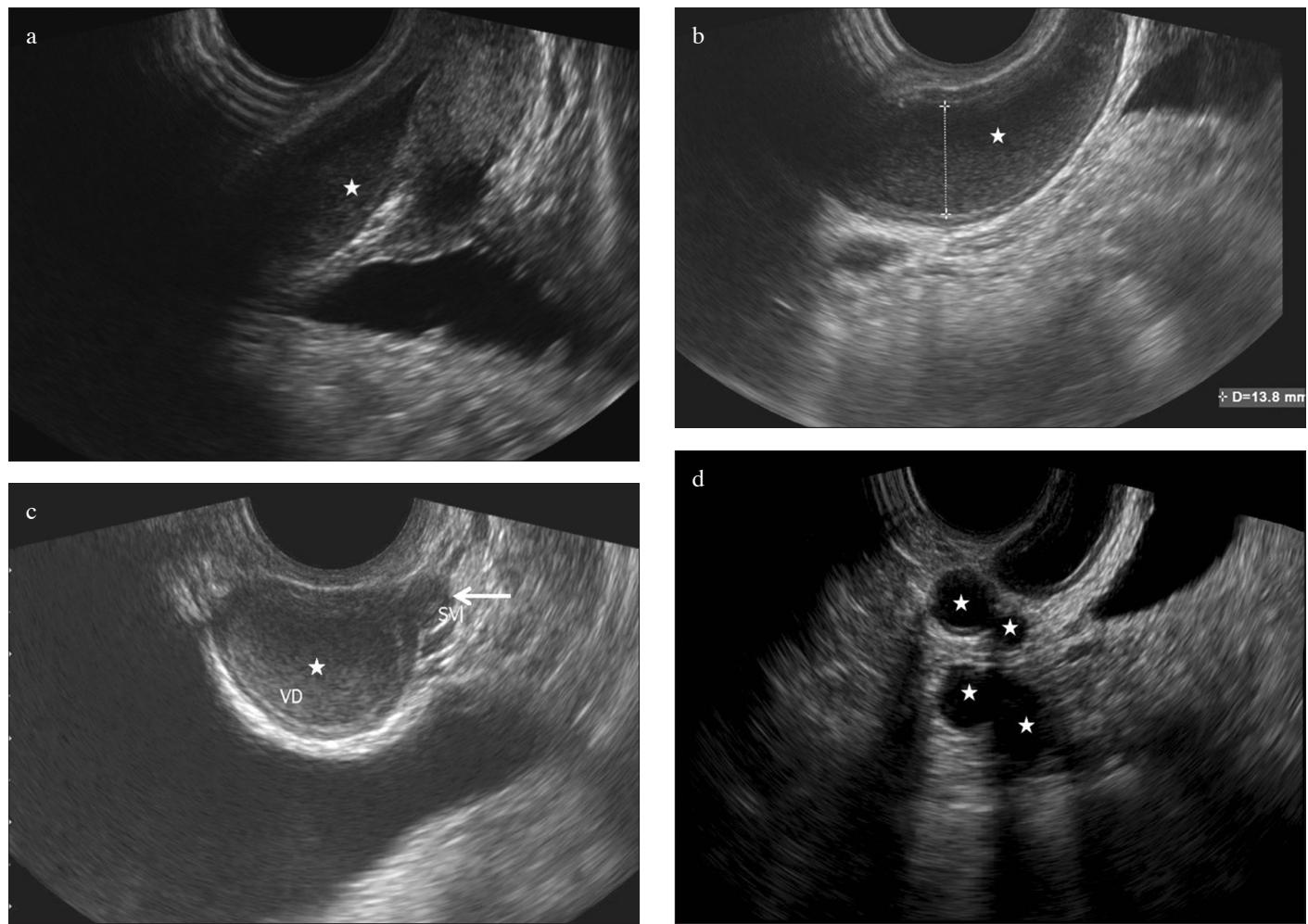


Figure 1. a-d. (a) TRUS image along midsagittal-sagittal plane, at the level of bladder neck and prostate showing smooth tapering of the dilated tubular structure (white star). (b) Right parasagittal image showing vas deferens distended with low-level echoes (white star). (c) Transverse section of distended right vas deferens (white star) and prominent seminal vesicle (arrow). (d) Multiple segments of tortuous vas deferens seen on further lateral angulations of the transducer (white stars)

hypovolemic ejaculate (0.4 mL) with an alkaline reaction, azoospermia (even after centrifugation) and absent seminal fructose indicative of obstructive azoospermia. The hormonal profile including testosterone (4.83 ng/mL); Luteinizing hormone (LH-2.44 mU/mL) and Follicle Stimulating hormone (FSH-1.86 mU/mL) was unremarkable. In consideration of obstructive azoospermia TRUS was recommended for the patient.

Transrectal ultrasonography revealed the normal size and echotexture of the prostate with the normal anatomical orientation of the collapsed urinary bladder and continuation of the urethra into the prostate. Midsagittal and the right parasagittal sweep of the transducer (Figure 1a and b) showed a dilated tubular structure of calibre 13.8 mm tapering towards the midline and filled with dense low-level internal echoes. Prominent right seminal vesicle and tortuous vas deferens were also visualized in transverse section (Figure 1c and d). Left seminal vesicle and

vas deferens were not identified and the diagnosis of right-sided ejaculatory duct stenosis was made. After TRUS, the patient was advised to get pelvic MRI done to delineate complete genitourinary anatomy and to confirm the sonographic findings.

Magnetic resonance imaging was done on a 1.5 T superconducting system (Magnetom Avanto, Siemens Medical System, Erlangen, Germany) using body array coil with respiratory gating. MRI images confirmed the TRUS findings and showed dilated and tapering distal part of right vas deferens and mildly dilated seminal vesicle duct with absence of their left-sided counterparts (Figures 2a and 2b). Dilated and tortuous right vas deferens was seen along the right lateral wall (Figure 2c). Coronal screen-shot of the lower abdomen and inguinoscrotal region showed empty left renal fossa (Figure 3a) and ipsilateral tubular ectasia of the rete testes (Figure 3b). No other ectopic location of the left kidney was visualized and the agenesis of the left

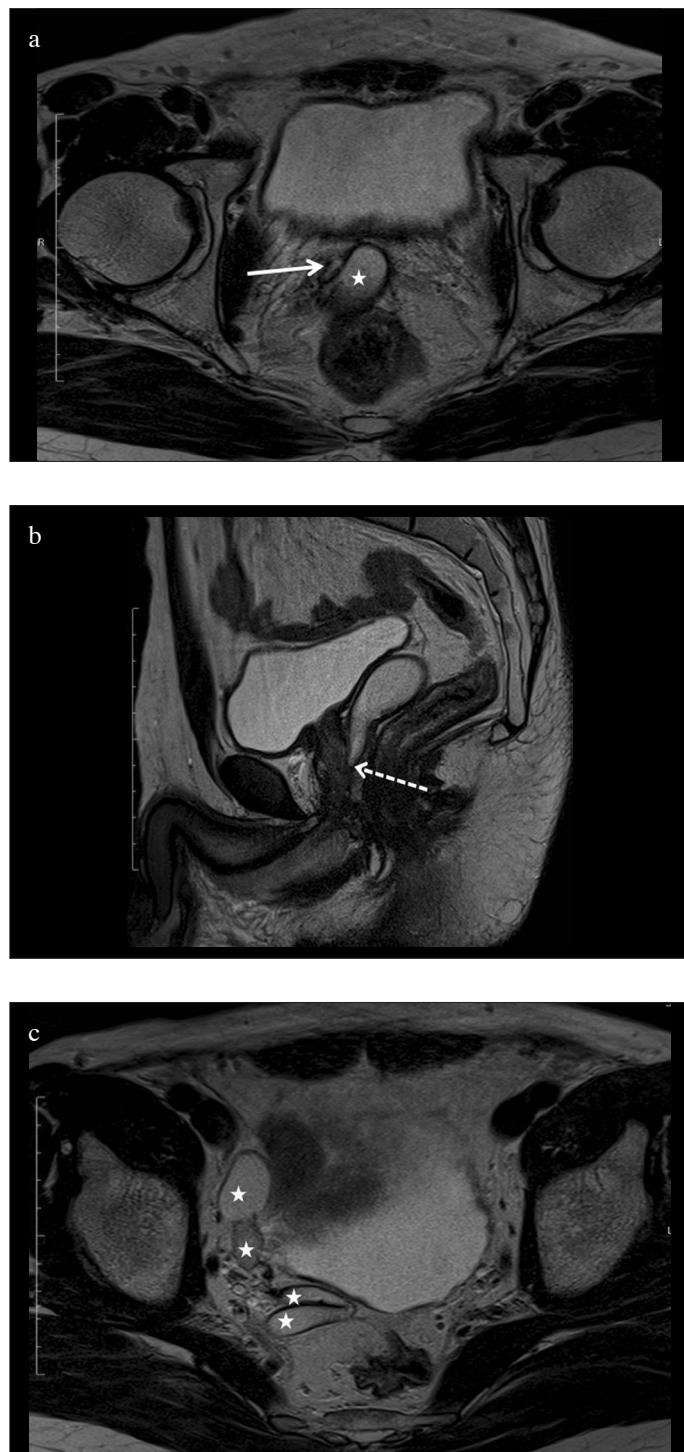


Figure 2. a-c. Pelvic MRI images (a) Axial T2-W sequence showing right corrugated seminal vesicle and its duct (arrow) and dilated right vas deferens (white star). Non-visualized left-sided seminal vesicle and vas deferens. (b) mid-sagittal T2-W image showing distal tapering and stenosis of the ejaculatory duct (dashed white arrow). (c) Axial T2-W image showing dilated and tortuous right vas deferens along right lateral pelvic wall (white stars)

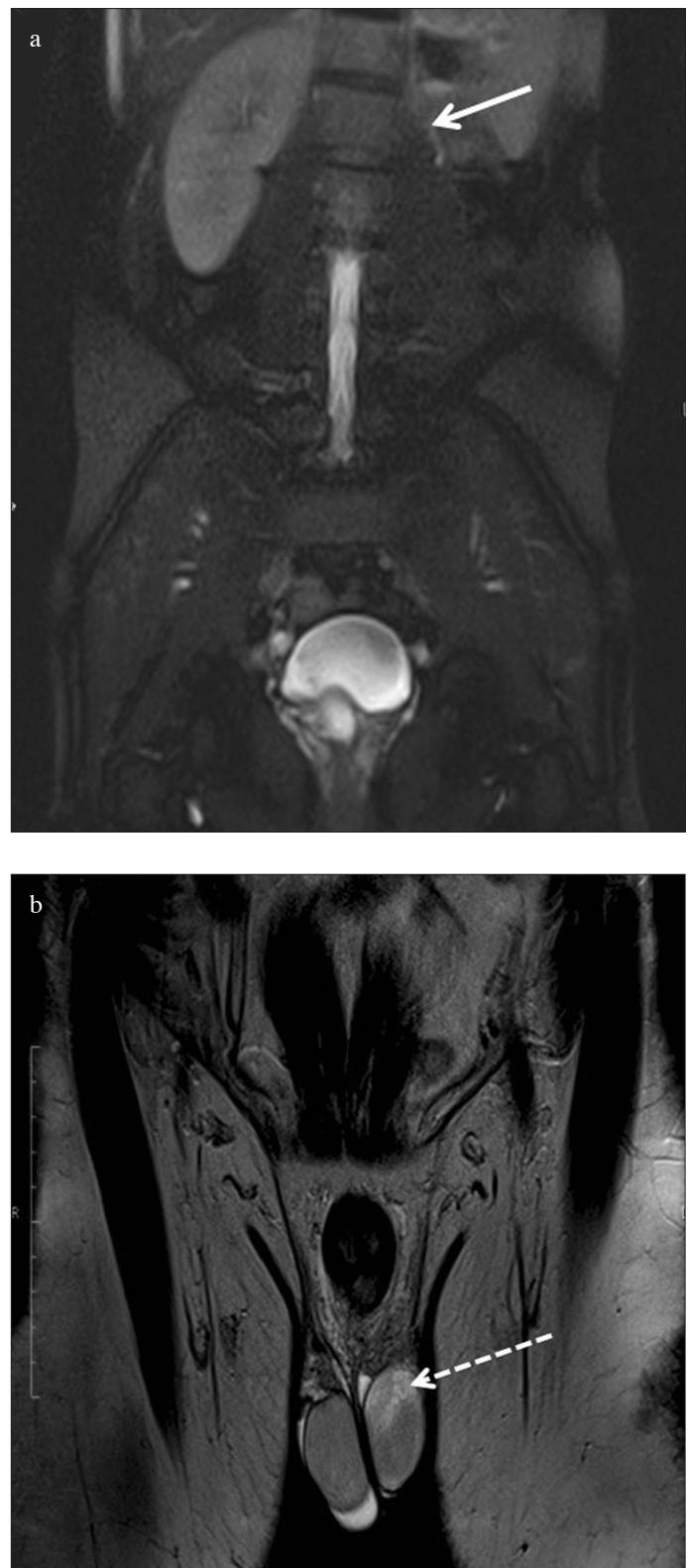


Figure 3. a, b. T2-W coronal images (a) showing absent left kidney (white arrow) (b) serpiginous hyperintense signal along mediastinum of the left testes suggesting tubular ectasia of rete testis (dashed arrow)

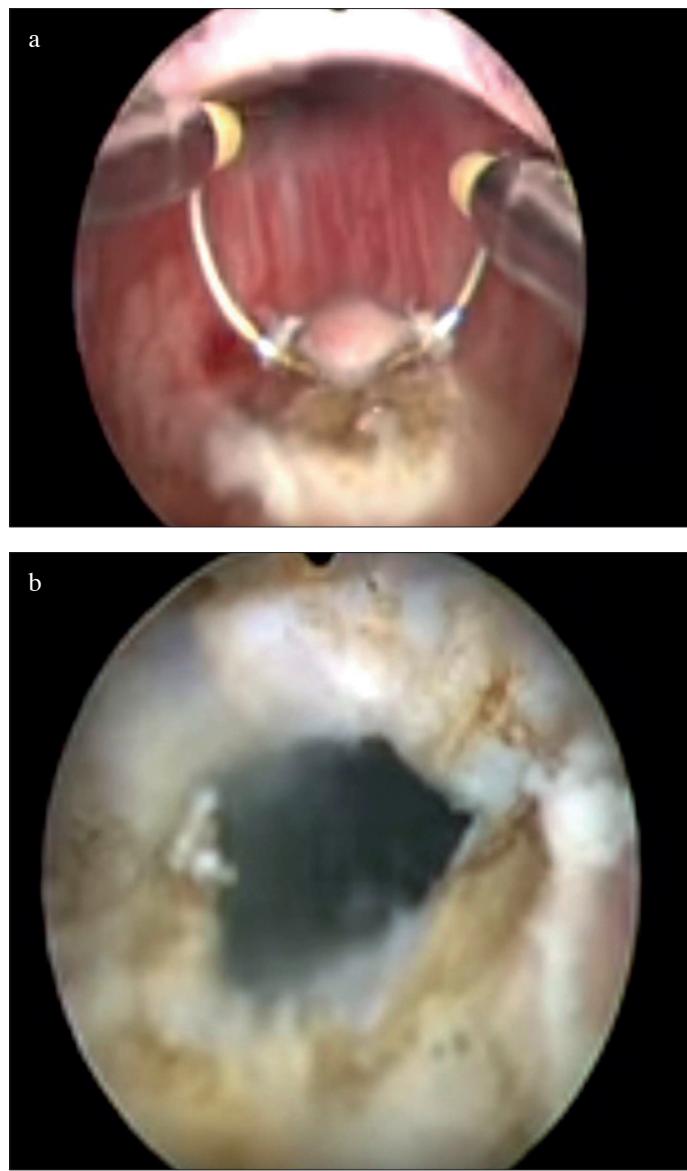


Figure 4. a, b. (a). Transurethral resection of the verumontanum and ejaculatory duct stenosis, TURED. (b) Expression of cloudy fluid through the opening

side Wolffian system derivatives (kidney, vas deferens, seminal vesicle and efferent ducts) and right-sided ejaculatory duct stenosis were diagnosed.

On cystourethroscopy, the verumontanum and urethra were essentially normal. The patient underwent transurethral resection of the ejaculatory duct (TURED) which was immediately followed by a gush of the chronic inspissated material through the urethra (Figure 4a and b). The patient was advised to refrain from sexual activity for 7-10 days and received human chorionic gonadotropin (β -HCG) injections and oral tamoxifen. Semen analysis was done after 4 weeks of TURED which revealed

normal volume ejaculate with positive fructose, however; sperm quality was poor. The sperm count was low (10 million/mL) with 70% dead/immotile sperms and 5% with normal morphology. This can be attributed to the fact that chronic ejaculatory duct stenosis can lead to variable fibrosis/stenosis of the vas deferens and consequent low sperm quality. The patient didn't return and was lost to extended follow-up.

Discussion

Congenital anomalies of the seminal vesicle and vas deferens include unilateral or bilateral agenesis of seminal vesicles or seminal vesicle cysts and most of them are associated with other genitourinary abnormalities. Unilateral agenesis of seminal vesicle is associated with ipsilateral renal agenesis in 79% of the cases. Similarly, unilateral agenesis of the vas deferens is associated with ipsilateral seminal vesicle, and ipsilateral renal agenesis in 86%, and 26% of the cases, respectively.^[2] Bilateral seminal vesicle agenesis is usually a manifestation of cystic fibrosis and it is almost always associated with vas agenesis and normally developed kidneys.^[3,4]

The imaging findings can be dated back to the embryological origin of the genitourinary system which develops from a common mesodermal ridge-intermediate mesoderm, differentiating into pronephros, mesonephros and metanephros. The nephrotomes develop from the regressing pronephros and forms the nephric ducts. These tubules later fuse with mesonephros to form mesonephric (Wolffian) duct. A paramesonephric duct (Müller) is also formed from the coelomic epithelium. Under the influence of Mullerian inhibiting substance and testosterone, the paramesonephric duct regresses and the mesonephric duct develops into bladder neck, seminal vesicle, vas deferens, efferent ducts, epididymis and paradidymis.^[5] Ureteric bud arises from the mesonephric duct, near the end of the 5th week of gestation and combines with the metanephric blastema to form the primitive renal tissue in the 6th week.^[6] This development occurs in the region of the pelvis and metanephric blastema migrates cranially to occupy the renal fossa. Seminal vesicle and vas deferens develop later near the 12th week of gestation. Hence, embryological insult prior to the 7th week of gestation can result in ipsilateral renal agenesis or other renal anomalies.^[7]

Hypoplasia of the seminal vesicle and congenital seminal vesicle cyst are other anomalies described in the literature. There are no definite criteria for the hypoplasia; however, MRI may show few septae. Zinner et al.^[8] was the first to describe the triad of unilateral renal agenesis, seminal vesicle cyst and ejaculatory duct obstruction; better known as the Zinner syndrome. This syndrome is considered as the male counterpart of the Mayer-Rokitansky-Kustner-Hauser (MRKH) syndrome in females.^[9] Casey et al.^[10] had reported a unique pentade of mesonephric

duct abnormalities on imaging, including cystic dysplasia of rete testis, seminal vesicle cyst, ipsilateral renal agenesis, partial hemi-trigonal development, and epididymal dilatation.

Ejaculatory duct obstruction is a rare but surgically correctable cause of male infertility. Use of TRUS has resulted in an increase in the incidence of diagnosis of this disorder. The ejaculatory ducts develop from the most distal vas, which is also a part of the Wolffian duct system. It enters the prostate obliquely at its base, traverse antero-medially through the prostatic glandular tissue, and opens at the verumontanum. Ejaculatory duct obstruction can be either congenital or acquired.^[11] Congenital causes include atresia or stenosis of the ducts and cysts along the utricle, obstructive Müllerian, and Wolffian ducts. Anomalous development of the distal part of mesonephric duct leads to ejaculatory duct atresia leading to chronic accumulation of secretions in the vas deferens and the seminal vesicle.^[12] Acquired causes may include infection, inflammation or trauma, iatrogenic and non-iatrogenic etiologies.^[11,13,14] Calculus formation as the sequel to infection may also cause obstruction. Several authors have found that patients with congenital or non-infectious causes of ejaculatory duct obstruction show a better response to treatment than those with infectious etiology.^[11,14] The combination of findings in our case can be presumed and correlated with embryological insult prior to the development of ureteral bud on the left side leading to the complete left-sided agenesis as well as maldevelopment of the distal part of the right Wolffian duct causing ejaculatory duct stenosis.

Congenital agenesis of the seminal vesicle is not surgically correctable and requires no treatment in cases with a patent contralateral ductal system. In symptomatic patients, surgical options include vaso-epididymostomy, TURED, and microscopic epididymal, testicular, or vasal sperm aspiration.^[15] TURED has become a standard procedure in cases of ejaculatory duct stenosis^[14,16] and has shown improvement in semen profile and pregnancy outcomes.^[17] Operative success and termination of the procedure for TURED are defined as fluid expression from both ejaculatory ducts. Close and extended follow-up of the patient is required as there are rare postoperative complications like damage to the bladder neck, urinary incontinence and sometimes extensive fibrosis causing scarring and azoospermia.

In conclusion, combination of wolffian system agenesis and contralateral ejaculatory duct stenosis is an extreme and rare phenomenon, but it should always be kept in a list of differential diagnosis while evaluating the cases of male infertility. TRUS is an important screening modality in cases of male infertility, primarily cases with obstructive azoospermia, because of the ease of availability and high-resolution assessment of the basic urogenital anatomy. MRI, in addition, proves to be a confirmatory cross-sectional modality, helpful in supporting/rejecting the

provisional diagnosis as well as enhancing the diagnostic yield as illustrated in the case. A better understanding of the anatomy and pathology of the ejaculatory ducts will also help to improve the diagnostic and therapeutic procedures for this disorder.

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