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Role of magnetic resonance imaging in evaluation of ejaculatory duct in Zinner's syndrome: case series of five patients and review of the literature

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Abstract

Background: Zinner's syndrome is a mesonephric duct anomaly characterized by unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ipsilateral ejaculatory duct obstruction due to insult occurred at urogenital tract embryogenesis during the first trimester. In the third and fourth decades of life, it is frequently diagnosed when patients begin to be symptomatic, such as lower urinary tract symptoms, infertility and painful ejaculation.

Case presentation: Herein we illustrate case review including five patients diagnosed as Zinner's syndrome, three of them complaining from infertility; however, the remaining two cases were fertile and incidentally diagnosed.

Conclusions: Radiological investigations play significant role in the diagnostic and management processes including US, CT and MRI for detecting ipsilateral renal agenesis and unilateral seminal vesicles dilatation, but evaluation of ejaculatory duct can be done only by MRI and transrectal ultrasound; however, the latter may be sometimes nonconclusive or intolerable by patients, so MRI is considered the golden modality with its high capability to assess the lower male genital tract which played a significant role in our case series starting with detection of the presence or absence of the ejaculatory duct obstruction as well as the high delineation of the origin and nature the seminal vesicle cyst including also its size and content and finally by detection of the communication between ureteral bud and the seminal vesicles cystic dilatation.

Keywords: Zinner syndrome, Seminal vesicle cyst, Unilateral renal agenesis, Ejaculatory duct obstruction

Background

Ipsilateral renal agenesis, ipsilateral seminal vesicle (SV) cyst, and ipsilateral ejaculatory duct (ED) obstruction association is rare urogenital tract congenital malformation [1]. Mainly caused by arrest of development in early embryogenesis causing the caudal end of Müllerian duct affection. Initially, both male and female embryos have two pairs of genital ducts: mesonephric (wolffian) ducts and paramesonephric (müllerian) ducts. The normal development of the male genital tract is the result of the differentiation of wolffian derivatives and the involution of müllerian derivatives. However, wolffian and müllerian remnants sometimes persist in adult males [2, 3]. Zinner was the first one to report this syndrome in 1914 [4]. Till now few hundreds only of cases reported in the literature [5], mainly clinical presentation is nonspecific, resulting in delayed or missed diagnosis, most of patients remain asymptomatic until starting sexual activity. Although different radiological modalities play main role in the diagnosis, Magnetic resonance imaging (MRI) is the superior

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modality in the diagnostic arsenal in making conclusive diagnosis [6]. Although using an endorectal coil (ERC) in patients with suspected obstructive infertility is a valuable method to evaluate the lower male genital tract. ERC may be mandatory for 1.5 T imaging, but there are no clear recommendations at 3 T MRI which gives reproducible high diagnostic performance [7]. Accordingly, we are pleased to present five rare cases of this Müllerian ducts maldevelopment involving the encountered in our hospital.

Case presentation

We report an unusual five cases diagnosed as Zinner syndrome (ZS) during 4 years.

The first case was 23-year-old married male patient since three years presented to the urology outpatient clinic complaining from infertility with no detectable systemic or endocrinal diseases, primarily diagnosed as obstructive azoospermic patient, he underwent pelvi-abdominal ultrasound (US) which revealed non-visualization of the left kidney at its normal anatomical or ectopic sites associated with indeterminate pelvic retrovesical cystic structure, so pelvi-abdominal MRI was recommended for better assessment of the condition as MRI is considered the most conclusive diagnostic modality in cases of primary diagnosed male infertility with obstructive causes; due to its high-resolution properties in imaging of lower male genital tract, MRI showed absent left kidney, unilaterally cystically dilated SV accompanied by cystic tubular structure abnormally inserted in it (ureteric bud) as well as ED dilatation (Fig. 1).

The second case was 29-year-old infertile married male patient, initially diagnosed with obstructive azoospermia. Pelvi-abdominal ultrasound (US) revealed non-visualization of the right kidney through its expected sites as well as indeterminate pelvic cystic structure, so pelvi-abdominal MRI was performed for better evaluation which revealed right renal agenesis, with cystically dilated ipsilateral SV accompanied by tortuous elongated structure abnormally inserted inside (atretic ureter) (Fig. 2).

The third case was 34-year-old married male patient since three years complaining from infertility, referred to as obstructive azoospermic patient for radiological assessment. Pelvi-abdominal ultrasound revealed empty right renal fossa with hypertrophied left kidney as well as well-defined anechoic structure related to the posterior bladder wall to the right, so pelvi-abdominal MRI was performed which revealed absent right kidney, with right seminal vesicle cyst in addition to prominent ED (Fig. 3).

The fourth case was 25-year-old adult male who presented in the urology clinic with recurrent left loin pain. Patient underwent abdominal and pelvic US which showed empty right renal fossa with non-detectable right kidney; on the other hand, left mild hydronephrosis with non-dilated ureter was noted and diagnosed as pelviureteral junction (PUJ) narrowing; inferolateral to the urinary bladder, there is a multilocular cystic mass on the right side of the pelvis. Internal echoes were seen in the mass, most likely due to pus, bleeding or a high proteinaceous composition, and MRI was recommended. Two days later pelvi-abdominal MRI was done confirming the absence of right kidney with right SV dilatation with related tubular cystic structure with direct communication with right SV and was described as right rudimentary ureter with abnormal insertion into cystic dilated right SV with non-dilated ejaculatory ducts…. Final diagnosis was Zinner syndrome with left PUJ narrowing, by taking history from the patient about his marital status we noticed that he is married since three years and had one child, so in this case there was no obstructive effect (Fig. 4).

The fifth case was 27-year-old married male who is married since 5 years and had two children, he was presented with chronic renal impairment and on hemodialysis for 1 year, he came to start his preparation steps for kidney transplant, by routinely done pelvi-abdominal US, the left kidney was not visualized with incidentally discovered rounded anechoic retrovesical cystic structure was noted related to the posterior vesical wall to the left, MRI was done which confirm unilateral left kidney agenesis, with left sided seminal vesicle cyst measured (2.5×2.3 cm) and normal right one, and non-dilated ejaculatory ducts (Fig. 5).

In our case series study we categorized the patients according to their clinical and radiological statuses (Table 1), the infertile category including the first three patients complaining with MRI signs of ejaculatory duct obstruction. On the other hand the second category includes the remaining two patients who were fertile but complaining from other urological symptoms with incidentally discovered signs of Zinner syndrome, with no MRI signs of ejaculatory duct obstruction.

Discussion

Zinner syndrome is one of the rarest congenital anomalies of the urogenital tract and is usually discovered and diagnosed in the 3rd-4th decade of life [8]. Zinner was the first to describe it, and there have been 200 incidences of seminal vesicle cysts linked to ipsilateral renal agenesis recorded in the literature [9]. Incidence is 1 in 3000 to 1 in 4000 newborns, [10] which reported 0.0046% according to Farooqui et al. [11].

Patients with this condition are usually normal but sometimes presents with lower urinary tract symptoms

as infertility, reduced urine output, frequency of micturation, and perineal pain [10].

Regarding infertility as a symptom which may be associated with Zinner syndrome, assuming that, in ZS, only one ejaculatory duct is affected, azoospermia should not be expected. However, azoospermia in ZS has been described in several literature reports [12]. The underlying pathogenesis of this aspect is not yet fully understood. One conceivable assumption is that unilateral testicular obstruction may cause antisperm antibody production, resulting in infertility despite the unobstructed contralateral testis [13]. Cito et al. proposed that due to

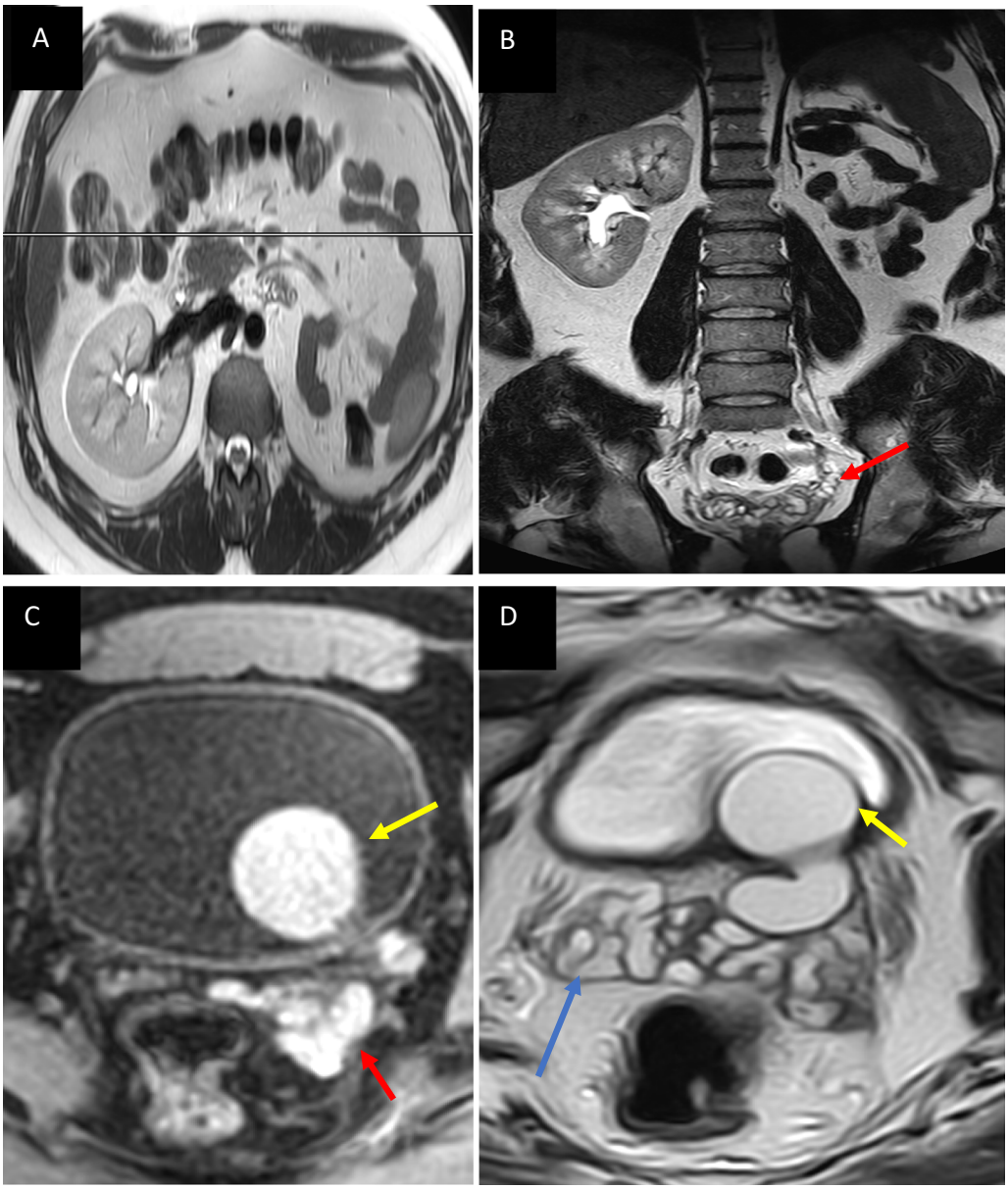


Fig. 1 **A** Axial T2WI of the abdomen and **B** coronal T2WI of the abdomen and pelvis show normal right renal unit and left renal agenesis and tubular cystic structure at the left side of the pelvis (red arrow), **C** and **D** high-resolution axial fat-suppressed T1WI and axial T2WI of the pelvis revealed normal right SV (blue arrow) and dilated left one with cyst (yellow arrow) measured (2.5 × 2.2 cm) compressing the left posterolateral vesical wall anteriorly, both of the cyst and the tubular structure display high SI at both T1WI and T2WI denoting hemorrhagic content, **E** and **F** high-resolution coronal T2WI of the pelvis show the left SV cyst with dilated ED (white arrow) as well as presence left sided pelvic cystic tubular structure (red arrow) is seen connected with the dilated left SV which resembles left ureteric bud with abnormal insertion with left SV, **G** and **H** high-resolution left parasagittal T2WI of the pelvis confirms the same findings

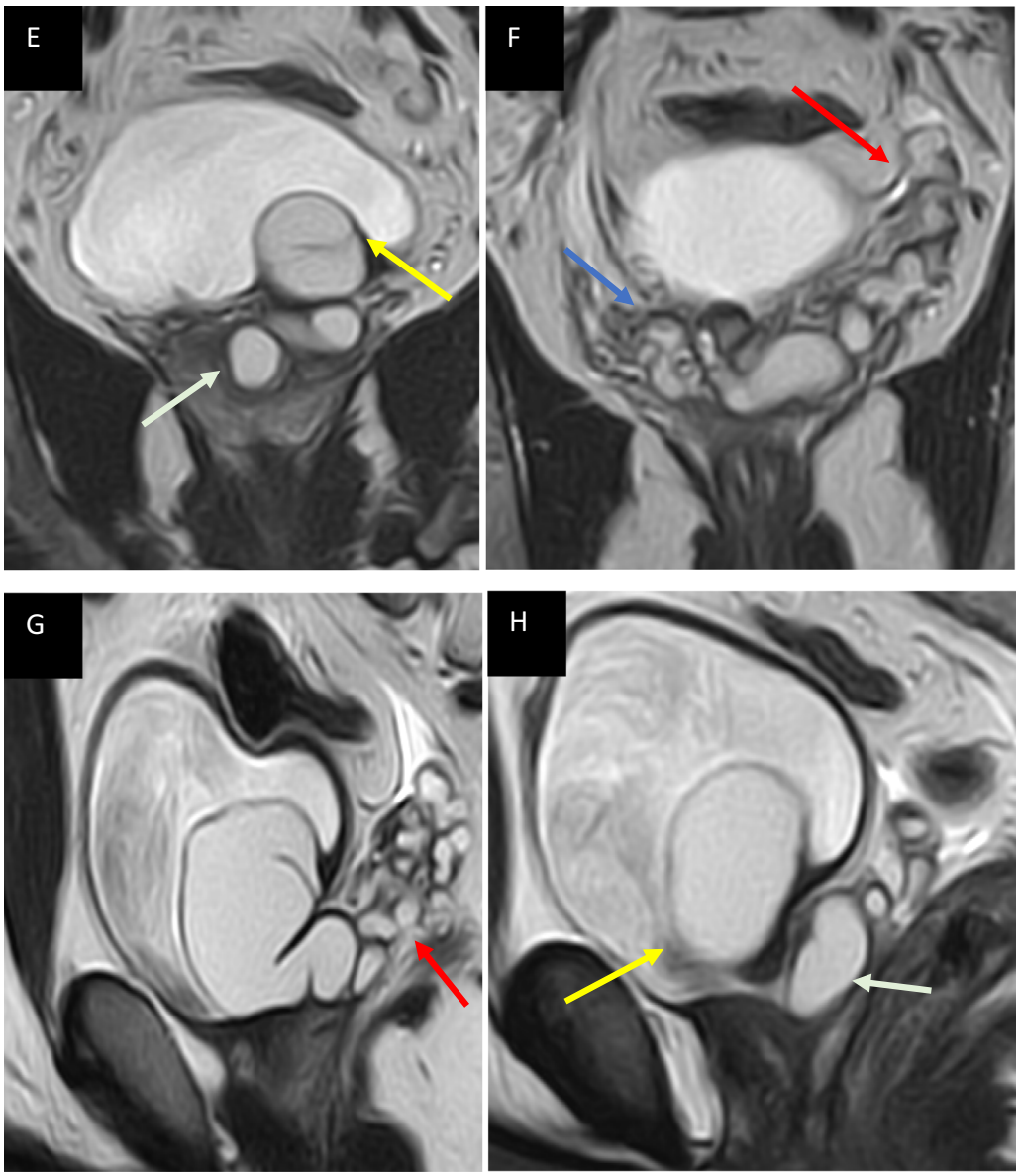


Fig. 1 continued

long-lasting obstruction reactive oxygen species may mediate reproductive toxicity in patients with ZS, thus reducing the sperm count by germ cell apoptosis [14].

Another hypothesis is that free semen passage in the normal contralateral duct is blocked because of a congenital defect in the ejaculatory duct area. Nevertheless, further studies are needed to identify the pathomechanism of infertility in patients with ZS. Because of the high rate of infertility associated with ZS, affected patients should be examined regarding their fertility status [13].

The seminal vesicles and kidneys are embryologically of the same origin; The metanephric blastema, which is produced by the ureteral bud that arises from the dorsal portion of the distal mesonephric duct, forms the kidney. Most of the genital tract is formed by the mesonephric duct, which includes the epididymis, vas deferens, ejaculatory duct, and seminal vesicle. Zinner syndrome can be caused by any ureteral bud or mesonephric duct abnormality [15].

US, CT, and MRI are the radiological modalities used to diagnose and evaluate Zinner syndrome. US and CT are commonly employed in this situation to detect the absence of the ipsilateral kidney or to indicate various

anechoic structures in the pelvis resembling the seminal vesicle cyst or dilatation [16, 17].

On the other hand, MRI is considered the best modality in diagnosing this case due to its high-resolution criteria in evaluating the abdomen and pelvis [10]. It is considered the modality of choice for the accurate anatomical demonstration of the male genital tract and hence confirming the periprostatic cystic lesion whether originating from seminal vesicle or not [15]. On MRI the seminal vesicle cyst appears hypointense on T1 and hyperintense on T2 weighted images; high protein cyst being exception which would have high signal on T1 weighted and intermediate to low signal on T2 weighted images [18], Intracystic vegetation's can be a sign of malignancy, although malignant degeneration remains exceptional with only 3 reported cases in the literature [6]. Seminal vesicle cyst needs to be differentiated from ejaculatory duct cyst, ureterocele and urinary bladder

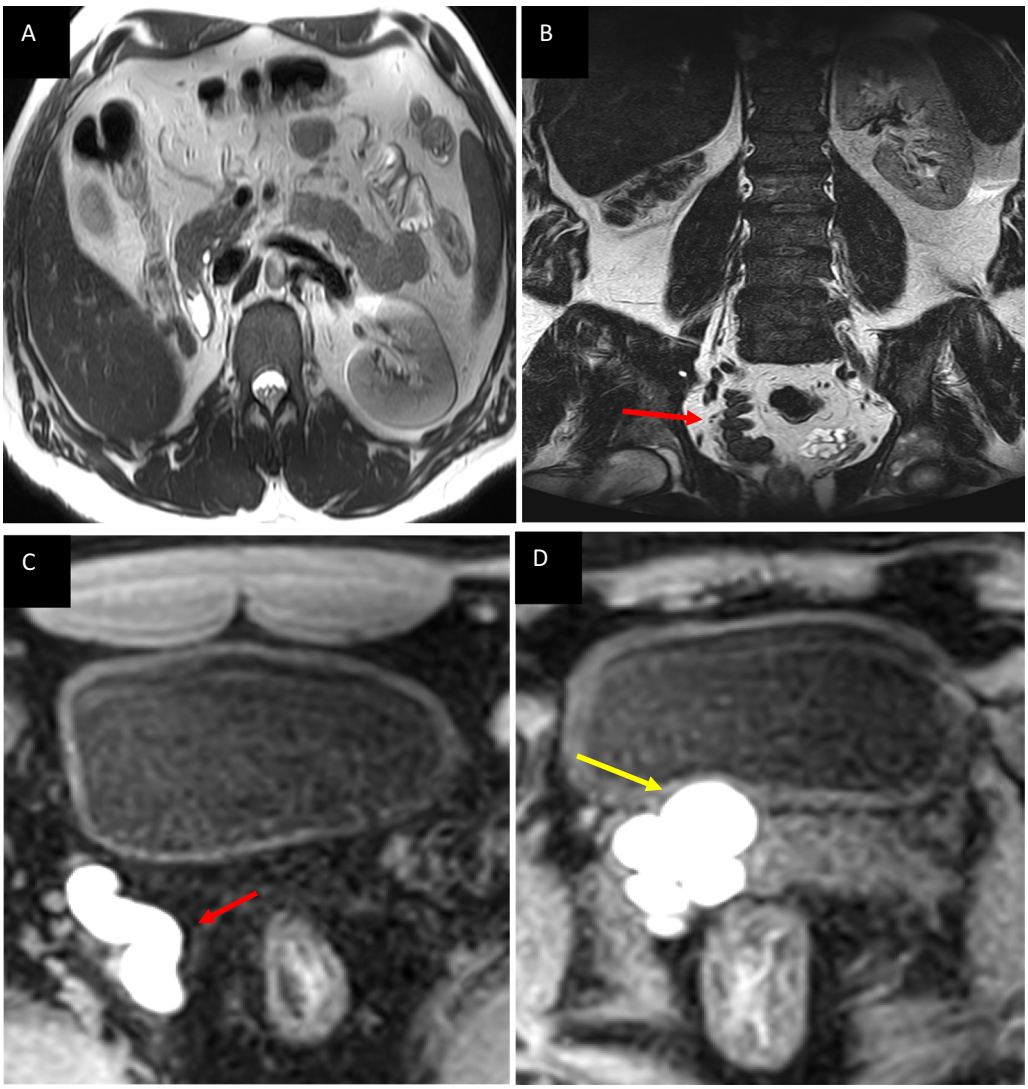
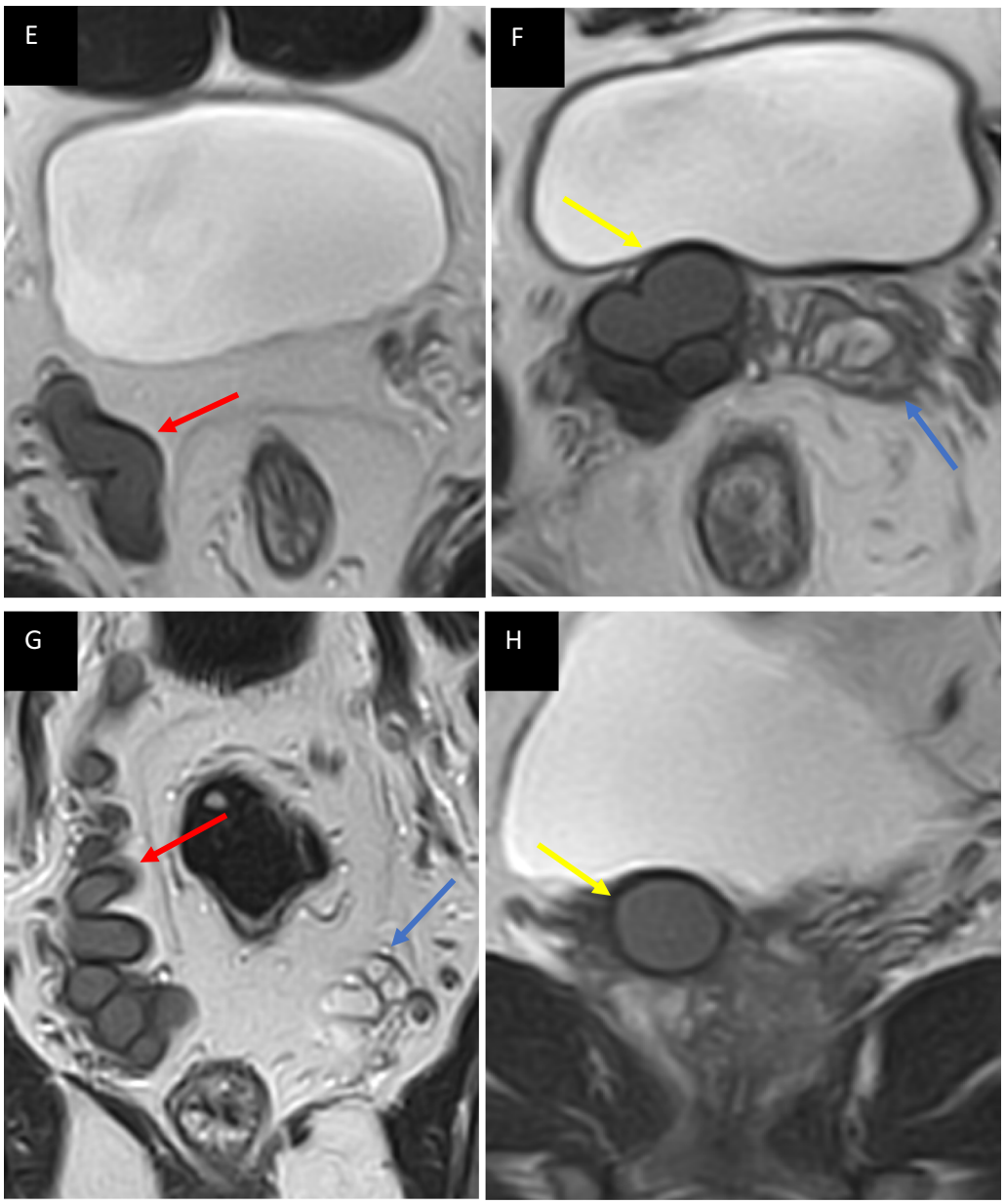


Fig. 2 **A** Axial T2WI of the abdomen and **B** coronal T2WI of the abdomen and pelvis show normal left renal unit and right renal agenesis and tubular cystic structure at the right side of the pelvis displays low T2WSI (red arrow), **C**–**F** high-resolution axial fat-suppressed T1WI and axial T2WI of the pelvis revealed normal left SV (blue arrow) and dilated right one with cyst (yellow arrow) measured (2 × 1.8 cm) slightly indenting the right posterolateral vesical wall anteriorly; both of the cyst and the tubular structure display high T1WSI and low T2WSI denoting hemorrhagic or high proteinaceous content (red arrow), **G** and **H** high-resolution coronal T2WI of the pelvis show the right SV cyst as well as presence of right sided pelvic cystic tubular structure (red arrow) seen connected with the dilated right SV which resembles right rudimentary ureteric bud with abnormal insertion into right SV, **I** and **J** high-resolution right parasagittal T2WI of the pelvis confirm the same findings



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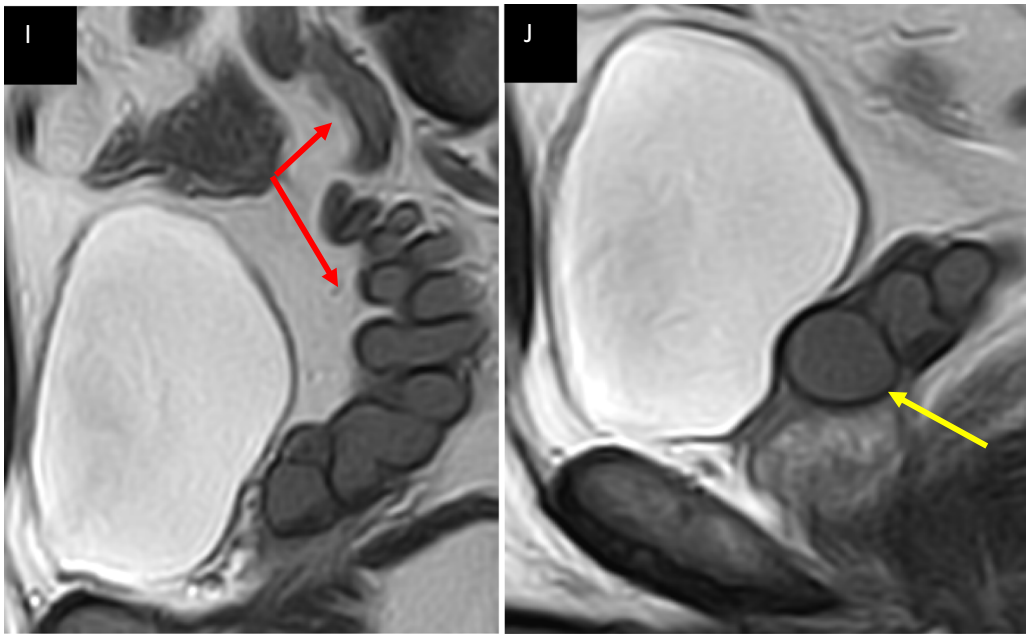

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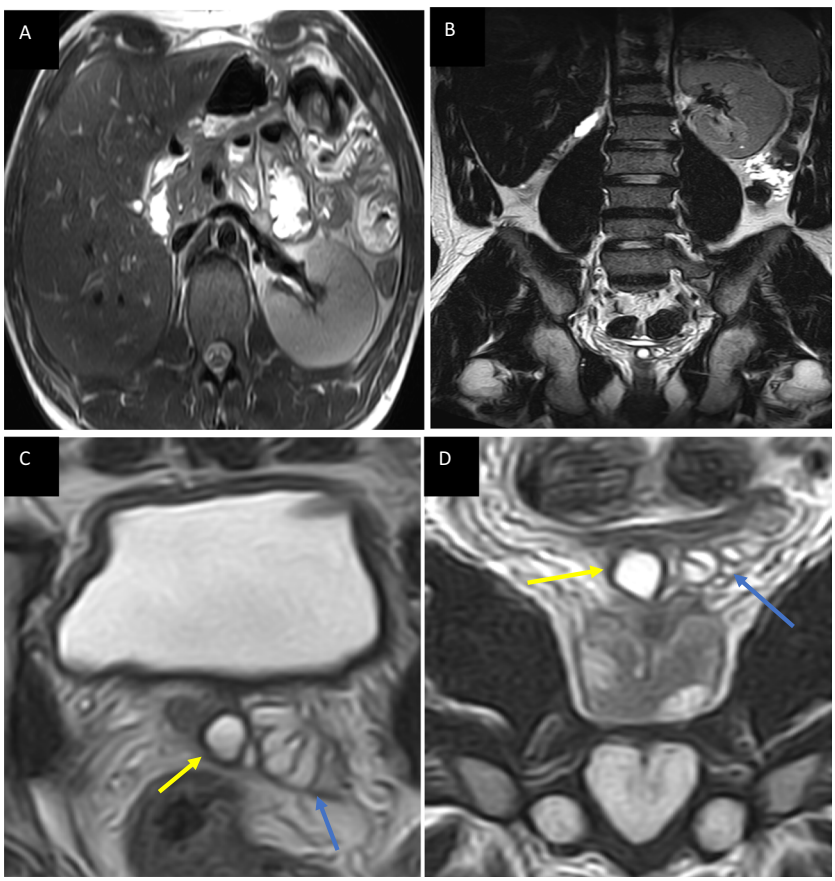
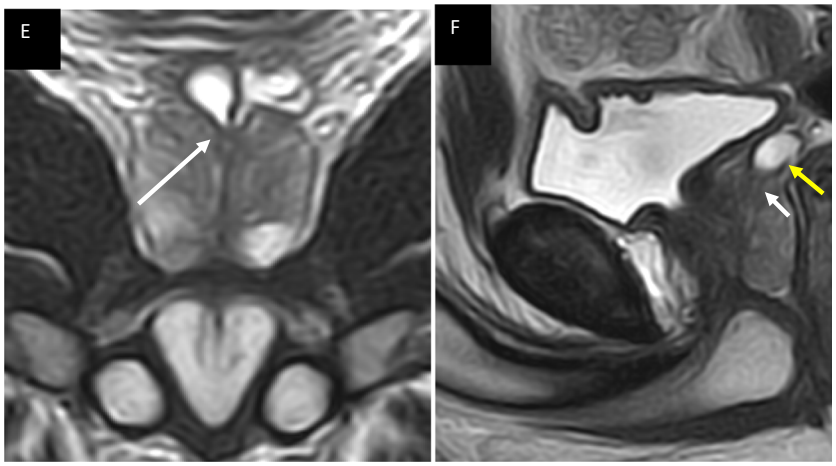


Fig. 3 **A** Axial T2WI of the abdomen and **B** coronal T2WI of the abdomen and pelvis show normal left renal unit and right renal agenesis, **C**–**F** high-resolution axial, coronal and right parasagittal T2WI of the pelvis revealed normal left SV (blue arrow) and right SV cyst (yellow arrow) measured (1.7 × 1.3 cm). It displays high T2WSI and low T1WSI (not shown) denoting clear fluid content, as well as ED obstruction (White arrow)



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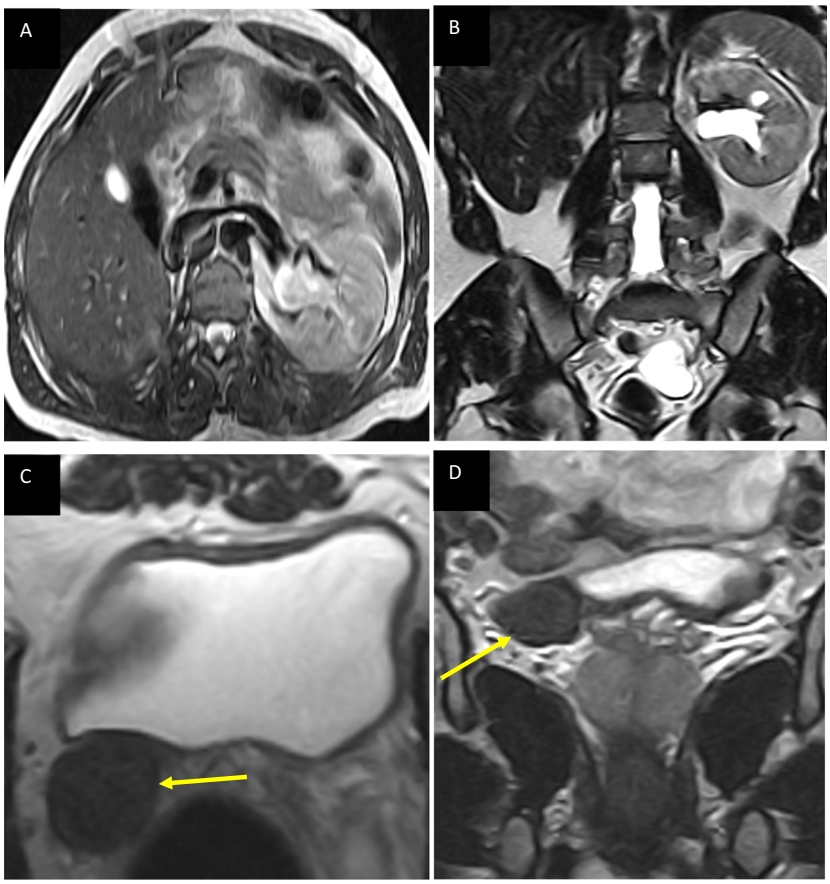

Fig. 4 **A** Axial T2WI of the abdomen and **B** coronal T2WI of the abdomen and pelvis show mild left hydronephrosis with dilated renal pelvis and non-dilated ureter suggesting PUJ narrowing as well as right renal agenesis, **C**–**G** high-resolution axial, coronal and right parasagittal T2WI of the pelvis revealed normal left SV and right SV cyst (yellow arrow) measured (1.7 × 1.3 cm) as well as presence of right sided pelvic cystic tubular structure (red arrow) seen connected with the dilated right SV which resembles right rudimentary ureteric bud with abnormal insertion into right SV, both the cyst and the tubular structure display low T2WSI and high T1WSI (not shown) denoting hemorrhagic or high proteinaceous content

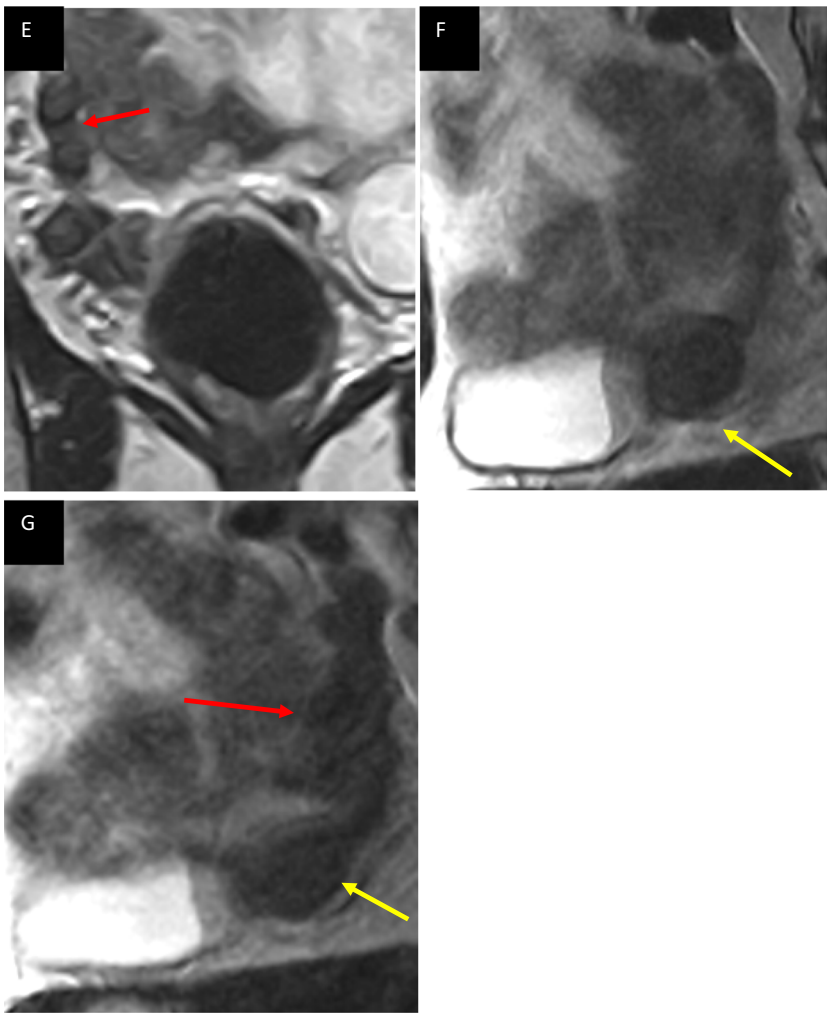

Fig. 4 continued

diverticulum. MRI plays an important role for distinguishing these possibilities in addition it can show the connection between ectopic ureter and seminal vesicle as A convoluted tail communicating with the cystic lesion and seminal vesical, which quite often is lost at other radiological modalities [15].

The treatment plan depends on patient symptoms and clinical status as patients who are asymptomatic, usually have regular routine follow-up until they begin to complain symptoms, and it includes transrectal ultrasound (TRUS) guided aspiration of the seminal vesicles cysts which has short-term effect on relief of patient symptoms and signs. The other option is surgical intervention which may be optimum in some cases using different techniques including open, laparoscopic and robotic assisted surgeries [8].

Conclusions

Urogenital system developmental anomalies are often not taken in consideration by clinicians when patients present with indeterminate symptoms referred to the urinary tract. Radiological investigations act as the cornerstone in the diagnostic and management processes including US, CT and MRI for detecting ipsilateral renal agenesis and unilateral SV dilatation, but evaluation of ejaculatory duct can be done only by MRI and transrectal ultrasound, however the latter may be sometimes non-conclusive or intolerable by patients. In addition, seminal vesicle cyst needs to be differentiated from ejaculatory duct cyst, ureterocele and urinary bladder diverticulum. MRI plays an important role for distinguishing these possibilities as well as it can show the connection between

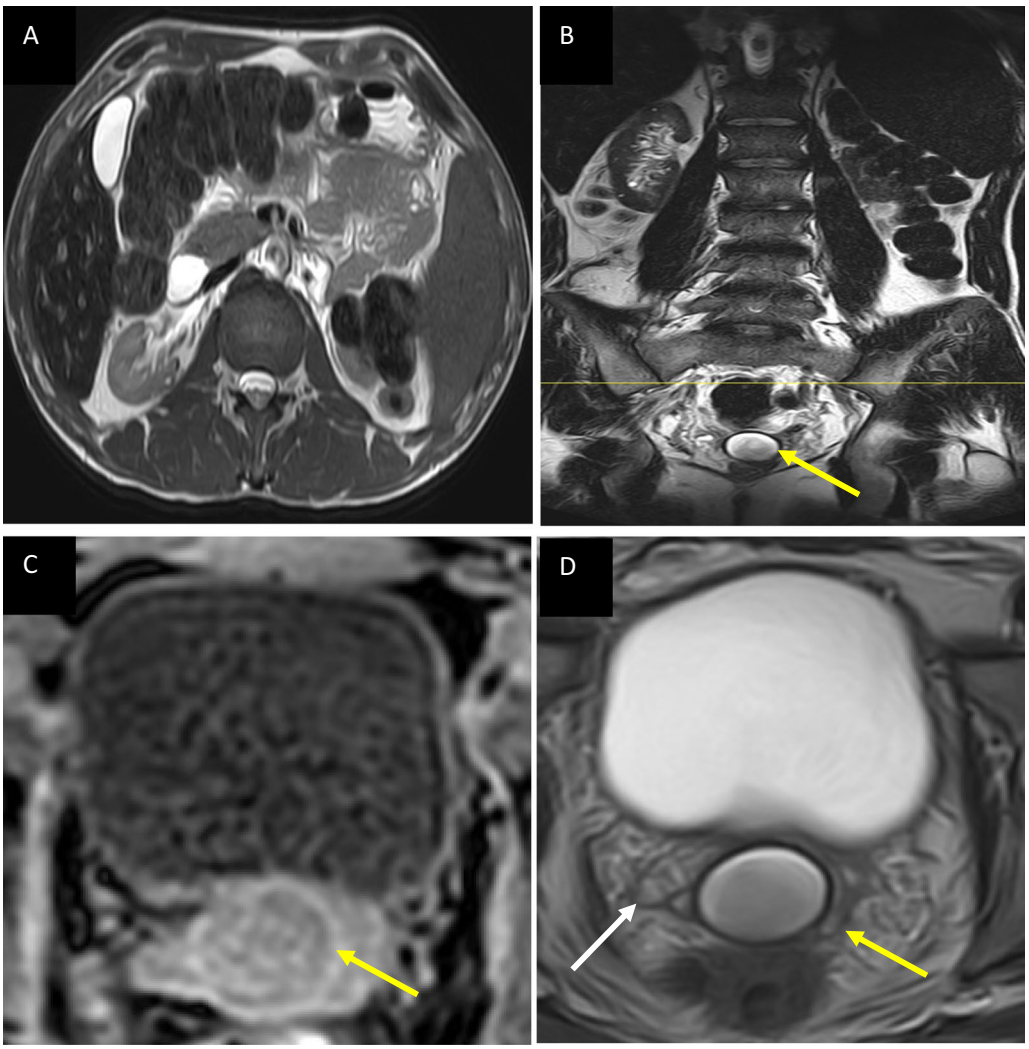

Fig. 5 **A** Axial T2WI of the abdomen and **B** coronal T2WI of the abdomen and pelvis show Small sized right kidney with pyelonephritic changes (in the form of reduced parenchymal thickness, increased thickness of sinus fat, irregular contour) and left renal agenesis, (**C**–**F**) high-resolution axial fat-suppressed T1WI and axial, coronal and right parasagittal T2WI of the pelvis revealed normal right SV (white arrow) and left SV cyst (yellow arrow) measured (2.5 × 2.3 cm). It displays high T2WSI and intermediate to low T1WSI denoting clear fluid content

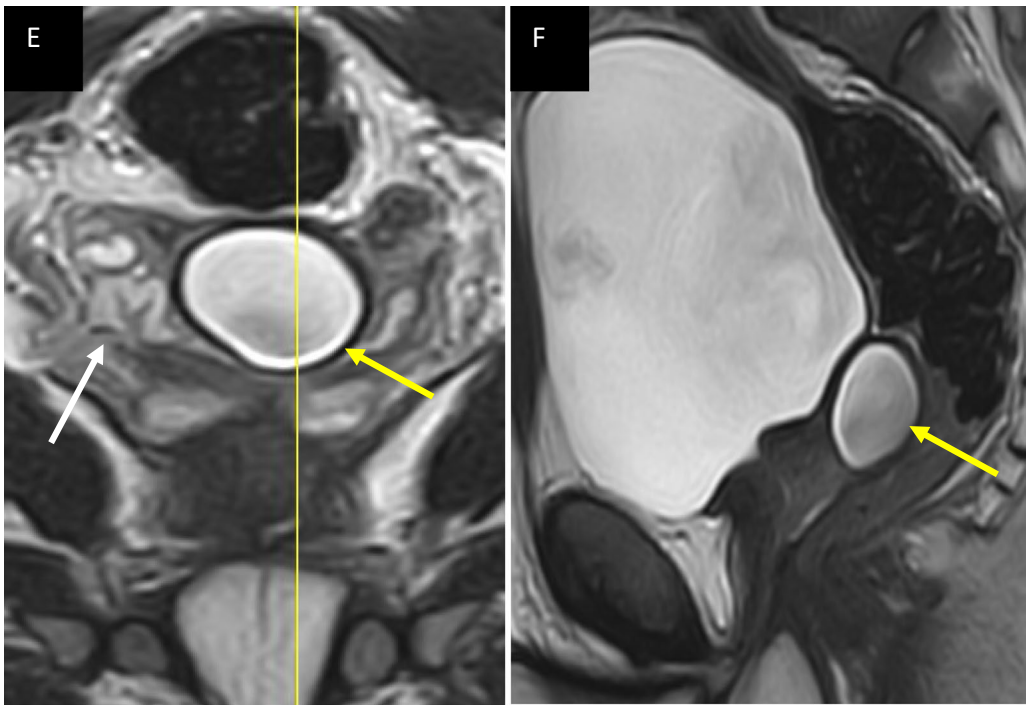

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Table 1 Semen analysis of the infertile cases

Parameter value	Case 1	Case 2	Case 3	Reference values
Color Milky white	Milky white	Milky white	Milky white	
Consistency Fluidly	Fluidly	Fluidly	Fluidly	
Volume (ml)	2.5	2.7	2.1	> 1.5
Concentration (millions/ml)	1.0	2.3	0.8	> 15
pH	8.1	8.3	8.7	> 7.2
Total spermatozoa (millions)	2.5	3	2.1	> 39
Progressive mobility (%)	0	0	0	> 32
Spermatozoa vitality (%)	0	0	0	> 58
Total mobility	0	0	0	> 40

ectopic ureter and seminal vesicle as a convoluted tail communicating with the cystic lesion and seminal vesical, which quite often is lost at other radiological modalities.

Learning points

1. Knowledge of the anatomy and embryology of genitourinary tract is important to make an accurate diagnosis of developmental anomalies.
2. According to our study, we concluded that not all patients diagnosed as Zinner syndrome should complain from ejaculatory duct obstruction and infertility.
3. Multiplanner MRI is the best imaging modality for diagnosing developmental anomalies of male genital tract due to its high-resolution anatomical and pathological soft tissue delineation which confirm the diagnosis especially the diagnosis of ejaculatory duct obstruction.

Abbreviations
US: Ultrasonography; ERC: Endorectal coil; CT: Computed tomography; MRI: Magnetic resonance imaging; SV: Seminal vesicle; ED: Ejaculatory duct; ZS: Zinner syndrome.

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Author contributions
HF and AA contributed to the data collection. AE and MB performed data analysis and writing. ME and HS performed supervision. They all approved the final version of the manuscript.

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Availability of data and materials
The datasets used and/or analyzed during this case report are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate
This study was approved by the Research Ethics Committee of the Faculty of Medicine at Mansoura University in Egypt on 20/02/2022; reference number of approval: MS/17.09.92.

Consent for publication
Patient included gave written informed consent to publish the data contained within this case report.

Competing interests
The authors declare that they have no competing interests.

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