# OHVIRA Syndrome with a Rare Presentation


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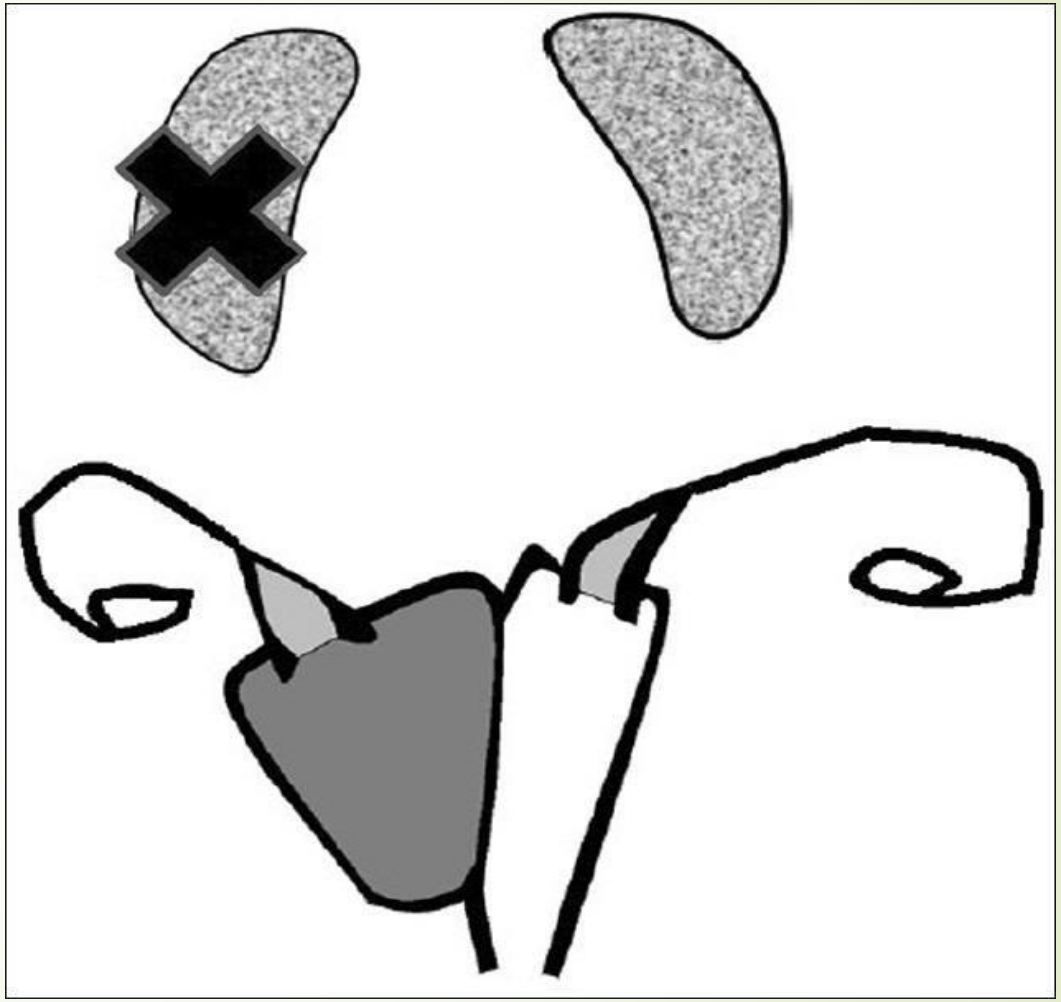
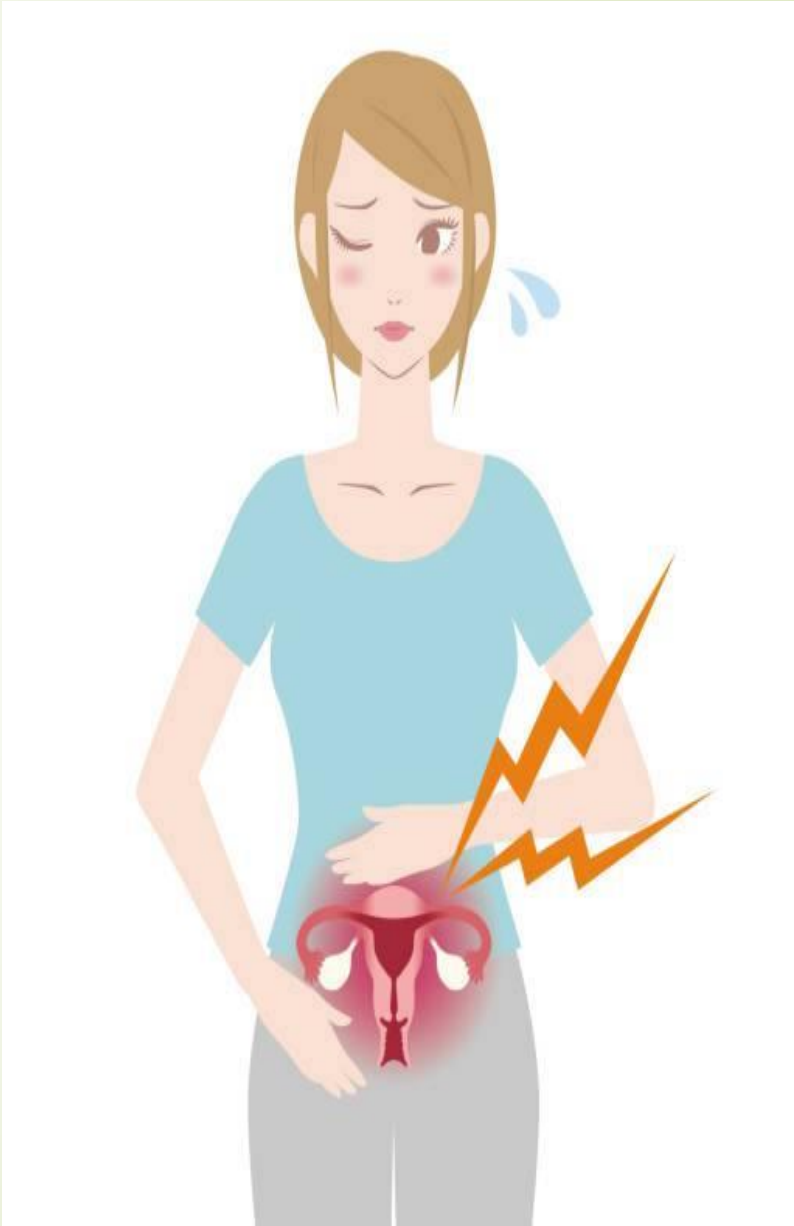


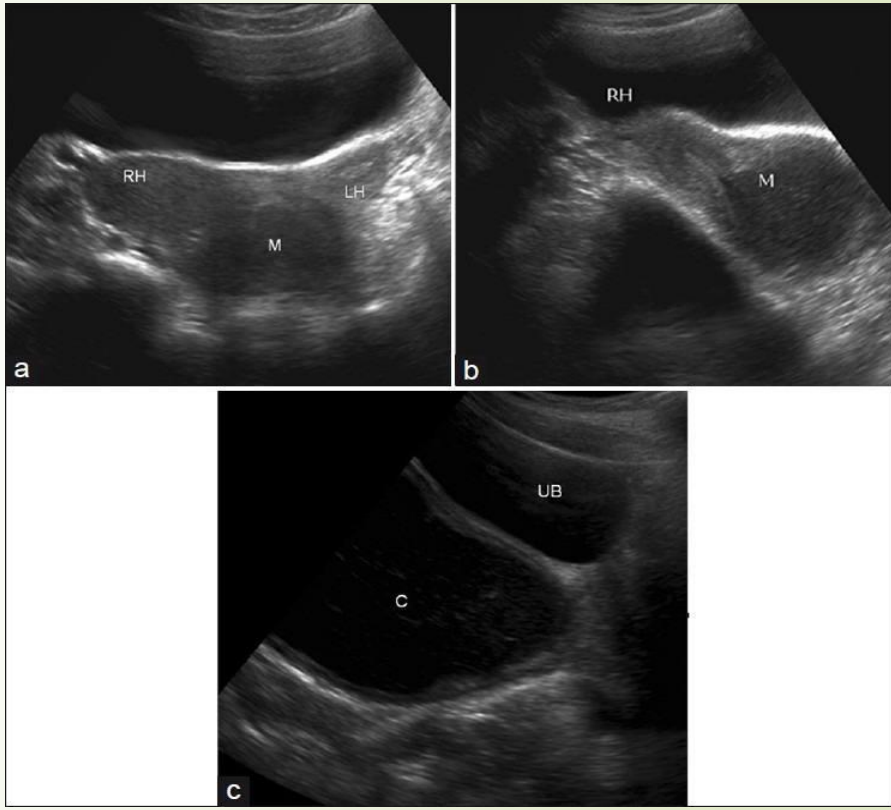
# case report presentation:

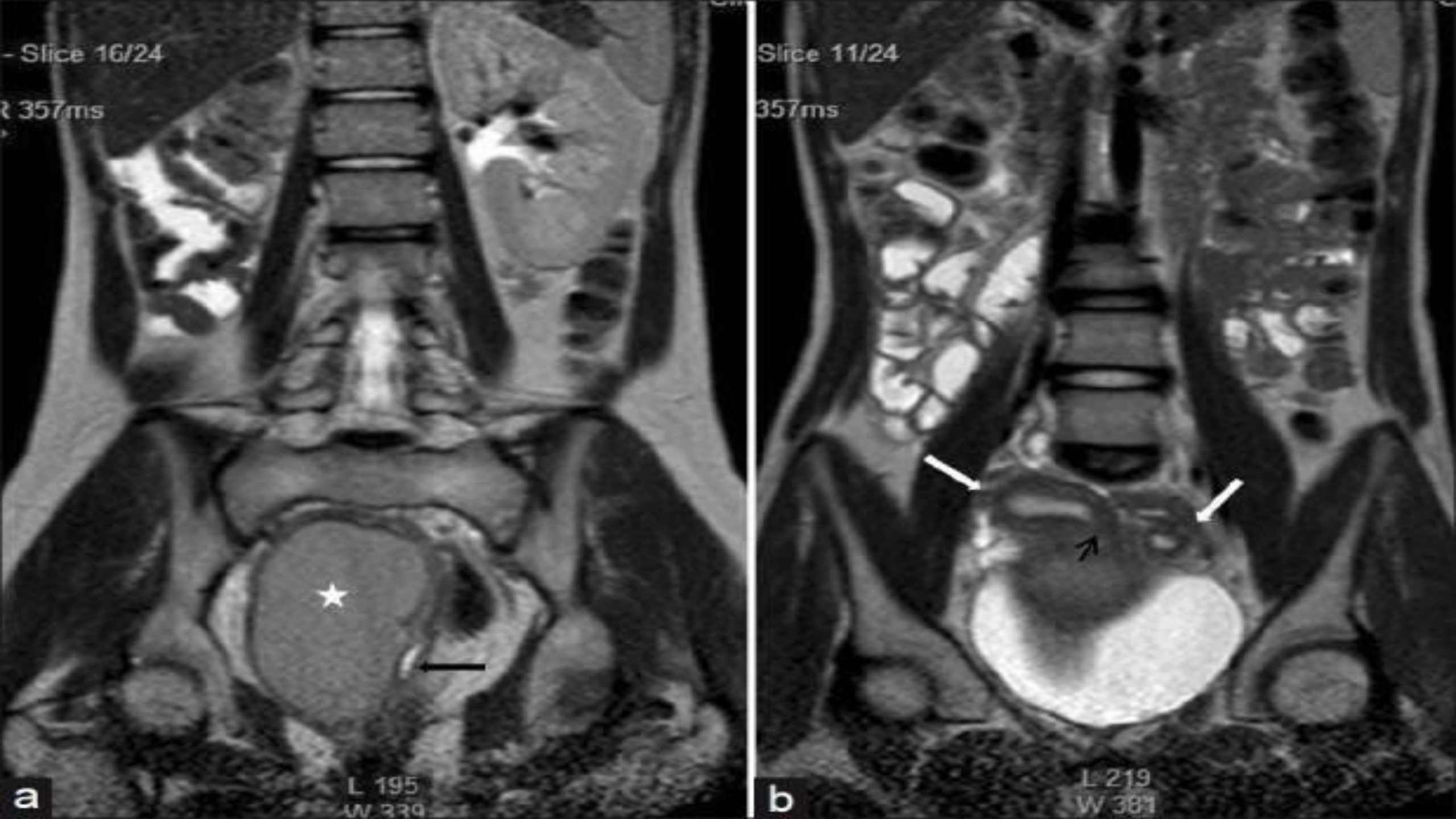
- 15-year-old girl came to our casualty with acute urinary retention for 1 day. She was not able to void urine though she had the urge to void. She also had complaints of colicky pain in the abdomen, which started with her present menstrual cycle, with a gradual increase in severity. Her last menstrual cycle was 15 days back. She went to a local hospital and was referred to our institution with Foley's catheter in situ.
- She attained menarche 8 months back. Her prior cycles were on time and she bled for 3–4 days not associated with dysmenorrhea. General physical examination was unremarkable with stable vitals.
- She had normal secondary sexual characteristics (Tanner staging of breast, axilla and pubic hair were IV).
- On abdominal examination there was no palpable mass or tenderness. Local examination revealed normal-looking external genitalia.



# Radilolgy:

- Abdomen and pelvic ultrasonography (USG) revealed right kidney could not be visualized (renal agenesis), a large well-defined thick-walled anechoic cystic lesion (12 x 6 x 7.2 cm) in the right adnexa? hematometrocolpos, and a possibility of uterus didelphys.
- Further evaluation by magnetic resonance imaging (MRI) was advised. The MRI of abdomen and pelvis showed two separate uterine horns (5.1 x 2.5 x 3.1 cm on the right side and 4 x 2.6 x 1.9 cm on the left side), two endometrial cavities, cervix, and vagina – suggestive of the uterus didelphys (type III), large dilated fluid-filled right hemivagina with communication with the right uterine horn suggestive of the obstructed vagina with hematometrocolpos, ipsilateral renal agenesis.
- Based on the imaging findings of unilateral renal agenesis, uterus didelphys, and unilateral obstructed hemivagina with resultant hematometrocolpos, the case was diagnosed as OHVIRA syndrome.

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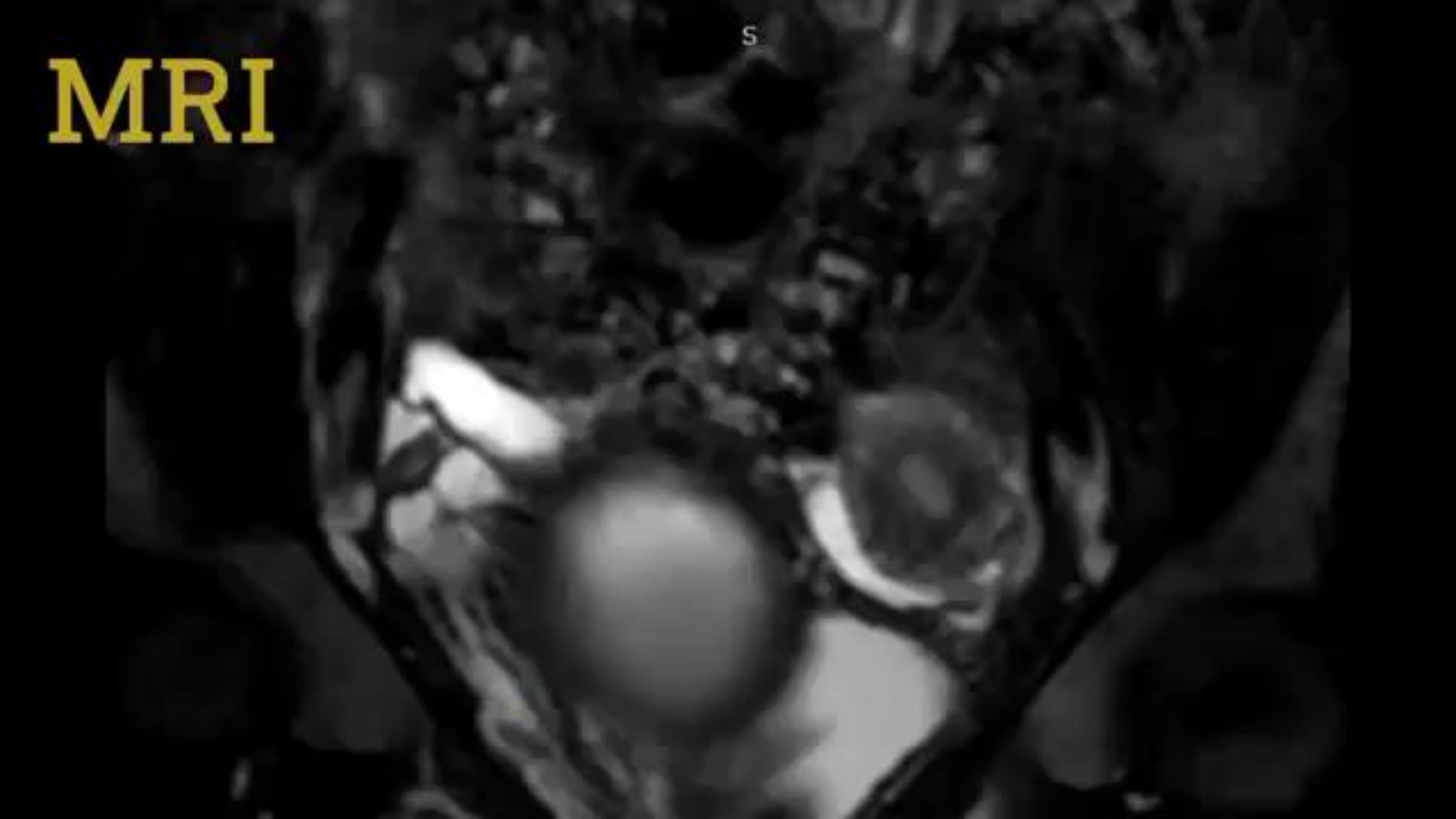
MRI
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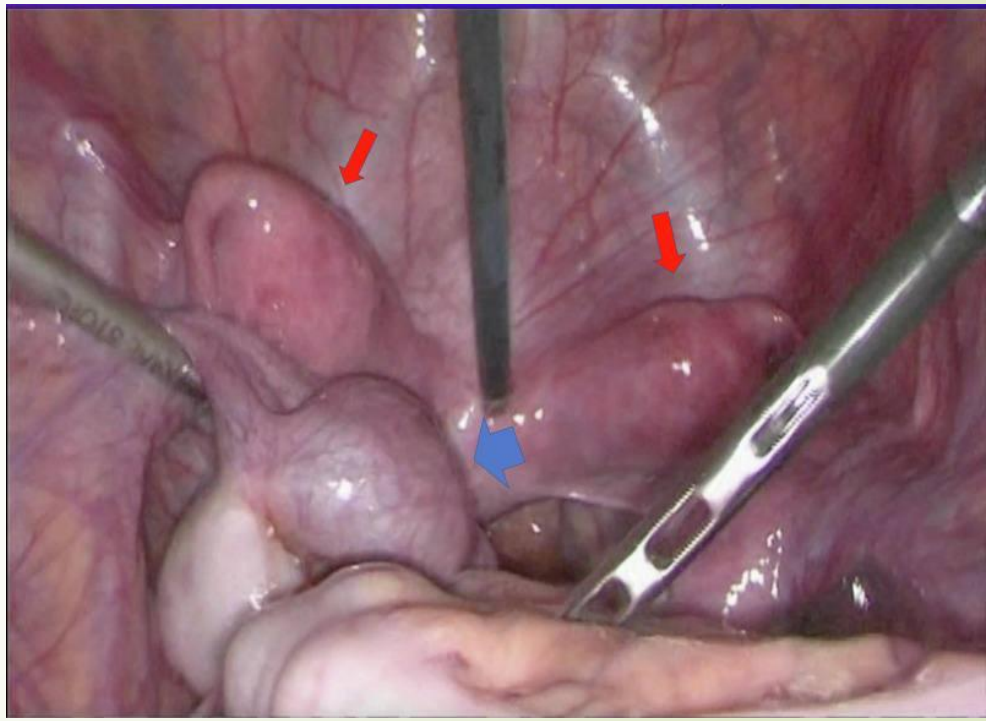
# What we do :

- Examination under anesthesia and diagnostic laparoscopy was planned.
- shows the on per speculum examination of the left side cervix, which was visualized and confirmed by placing Hegar's cervical dilator, whereas the right-side cervix could not be visualized. A thick septum was noted. Diagnostic laparoscopy was done, which showed two uterine horns. shows that on the lower aspect of right uterine horn, a bulge was visualized (hematocolpos), which was probably pressing on the neck of the bladder and that was the reason for the retention of urine
- Vaginally septal resection was done and around 150 mL of chocolate color fluid (old collected blood) was drained. This was confirmed laparoscopically by a gradual reduction in the size of the bulge. Then right-side cervix was visualized, confirmed by placing Hegar's cervical dilator which could be passed till the fundus of the right uterine horn. The resected septal wall was sutured to anterior and posterior vaginal walls (like marsupialization). She was treated with hemivaginal septal resection and drainage of hematometrocolpos.
- Following surgery, the catheter was removed and she voided urine normally, post-op period was uneventful. She came for follow-up after her next menses and she had no complaints.

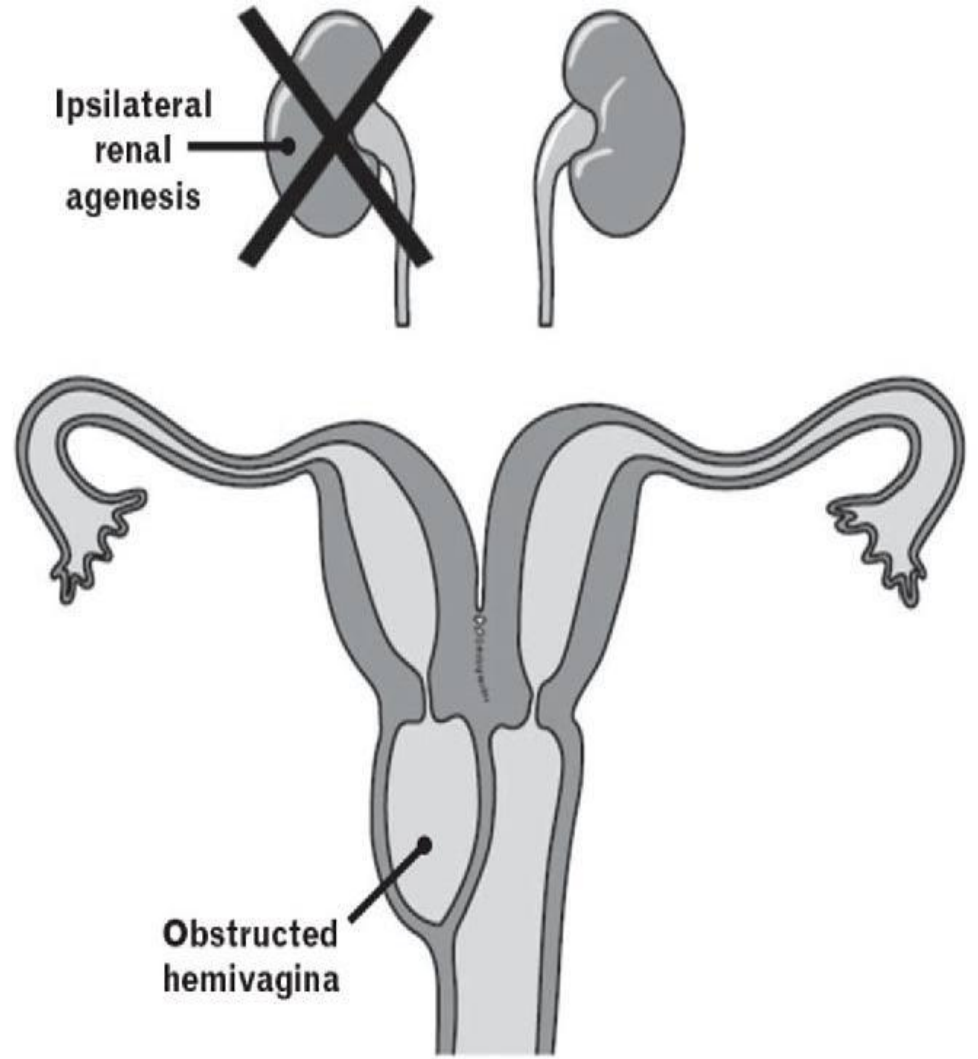


# discussion:

- The diagnosis of OHVIRA syndrome is mostly done after menarche because patients usually present with cyclic, increasing lower abdominal pain, and progressive dysmenorrhea due to the formation of hematocolpos resulting from long-standing retained menstrual blood in the obstructed hemivagina. But many a times, it may go undiagnosed because the menstrual flow from the normal side could be misleading. Dysmenorrhea, of course, is a common symptom in this age group and commonly gets treated by analgesics without thorough evaluation.

- Rarely, it can present as acute abdominal pain, abnormal vaginal discharge, infertility. Initial clinical diagnosis is incorrect in the majority of cases, because of its rare incidence and misleading presenting signs and symptoms. But there are no case reports of acute retention of urine in OHVIRA syndrome cases. Funda et al. showed that no patients presented with acute urine retention in a set of 32 patients with OHVIRA syndrome. All of them either presented with pain in the abdomen and dysmenorrhea; one with primary infertility and one with acute pain in the abdomen

- Right-sided abnormalities are more common (60–70%) compared with the left sided abnormalities for reasons unknown. Even in our case, it is on the right side. In girls with unilateral renal anomaly and a Mullerian anomaly, kindly look to rule out OHVIRA syndrome.
- Other than renal agenesis, renal dysplasia, renal cysts, double or ectopic ureter can be associated. Some other associations reported include high bifurcation of aorta, duplication of inferior venacava (IVC), intestinal malrotation, and ovarian malposition.
- Ultrasonography and MRI are effectively used in the diagnosis of genitourinary anomalies. A 100% accuracy has been reported for MRI. I



- توصيات خاصّــة :
- ١. دوماً نضع تشوهات السبيل التناسلي في ذهننا خصوصاً عند الفتيات مع بدء الطمث وعسراته
- ٢. ينصح البحث عن تشوّهات مرافقة بحال وجود تشوهات سبيل تناسلي ( تشوهات السبيل البولي خصوصاً )
- ٣. نضع الاعراض النادرة في ذهننا مع ضرورة الربط مع المعطيات الأخرى