MRI EVALUATION OF UTEROVAGINAL ABNORMALITY IN A YOUNG GIRL.

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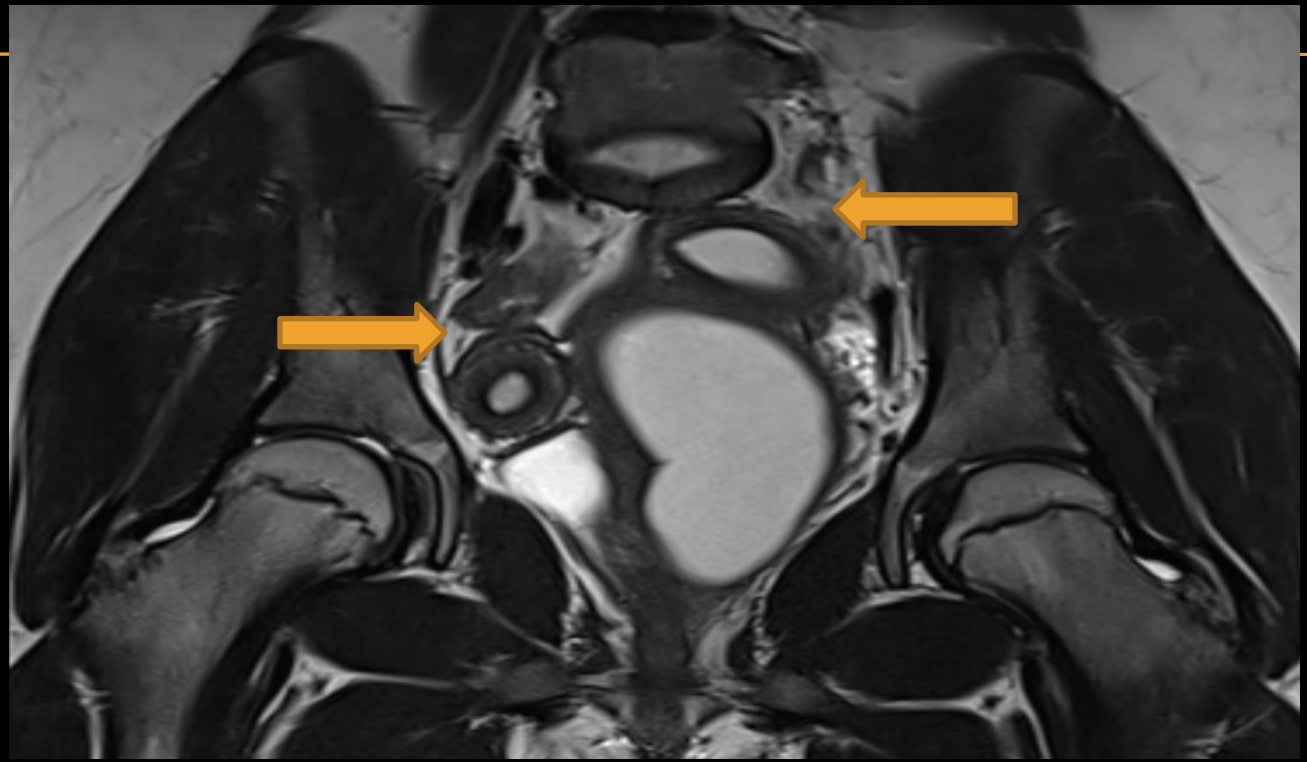
CLINICAL PROFILE:

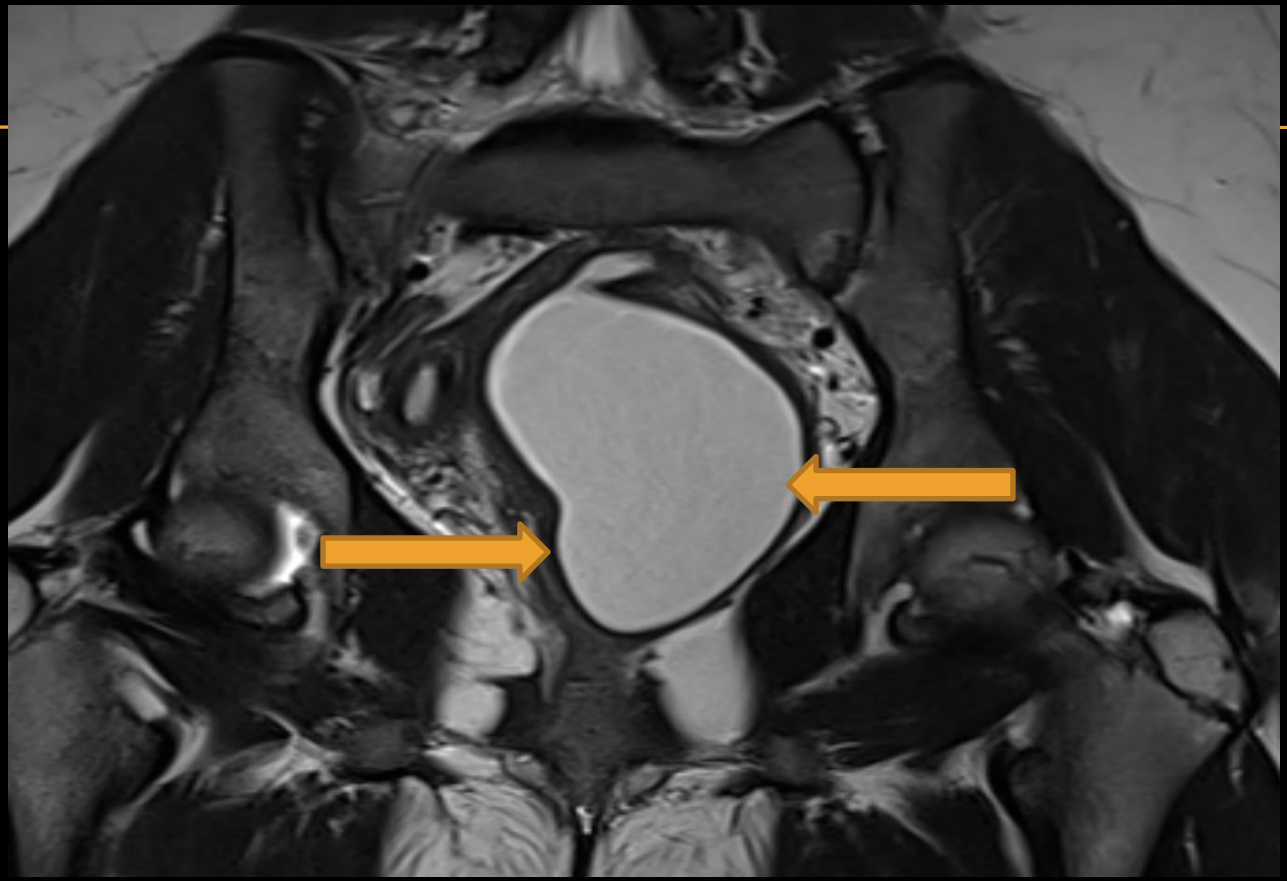
- A 11 year old female child came to our institution for MRI evaluation with history of irregular menstrual cycle.
- non specific pain in the abdomen and pelvic region.
- Menarche was attained 6 months back.
- No h/o trauma /nausea /vomiting/altered bowel and bladder habits

USG ABDOMEN AND PELVIS: (OUTSIDE REPORT)

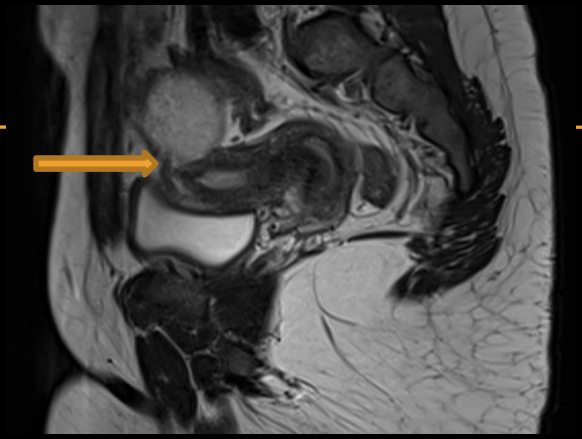
- Left kidney is not visualised in left renal fossa.
- Bicornuate uterus.
- A large collection (approx size 9.3x5.6x6.6cm) with dense internal echoes noted in cervical canal with extension into left uterine horn .
- Elongated cystic structure with incomplete septae noted in close proximity to the left uterine horn.
- Bilateral ovaries not be separately visualised.

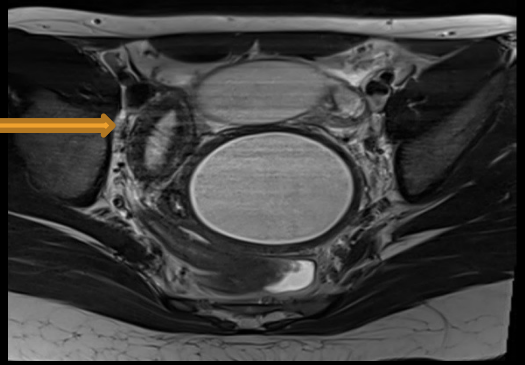


T2W CORONAL IMAGE SHOWING DUPLICATION OF UTERINE BODIES.

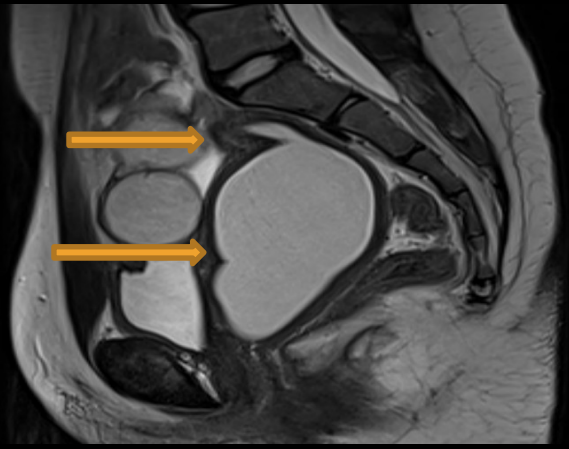


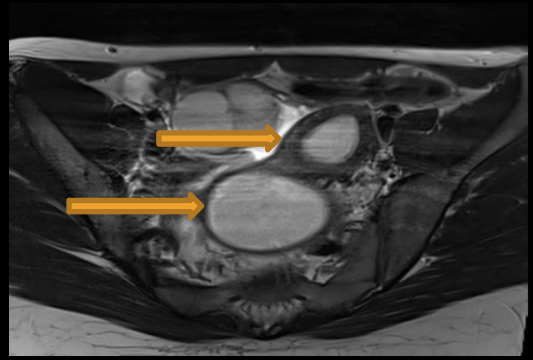
T2W CORONAL IMAGE SHOWING DUPLICATION OF UTERINE BODY AND CERVIX.





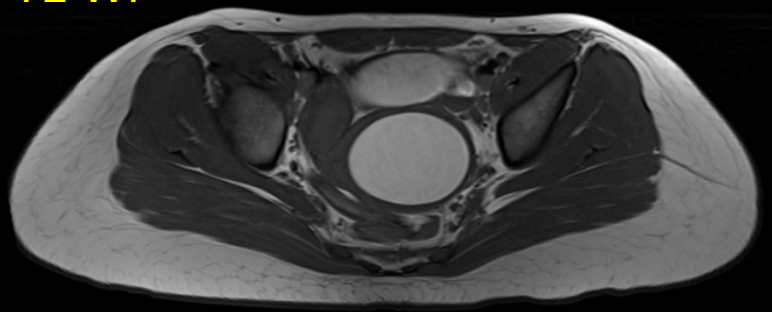
T2W SAGITTAL AND AXIAL IMAGE SHOWING THE RIGHT UTERINE BODY AND CERVIX.



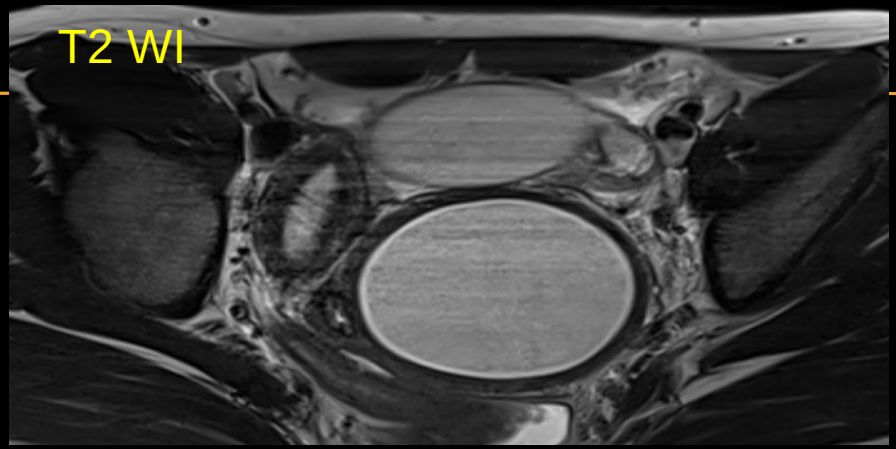


T2W SAGITTAL AND AXIAL IMAGE SHOWING LEFT UTERINE BODY AND CERVIX WITH T2 HYPERINTENSE COLLECTION.

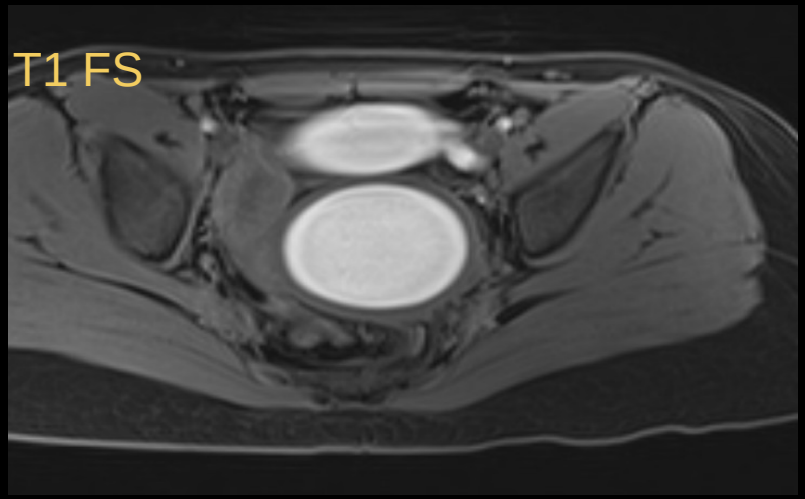
T1 WI



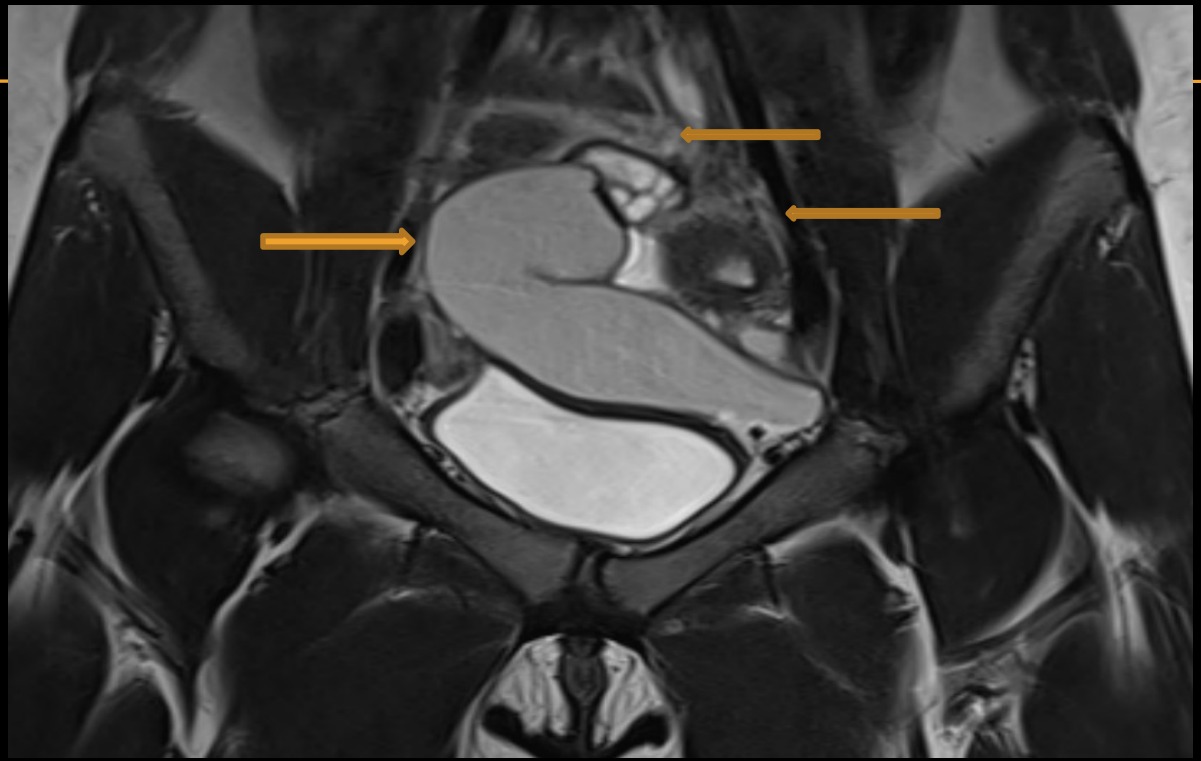
T2 WI



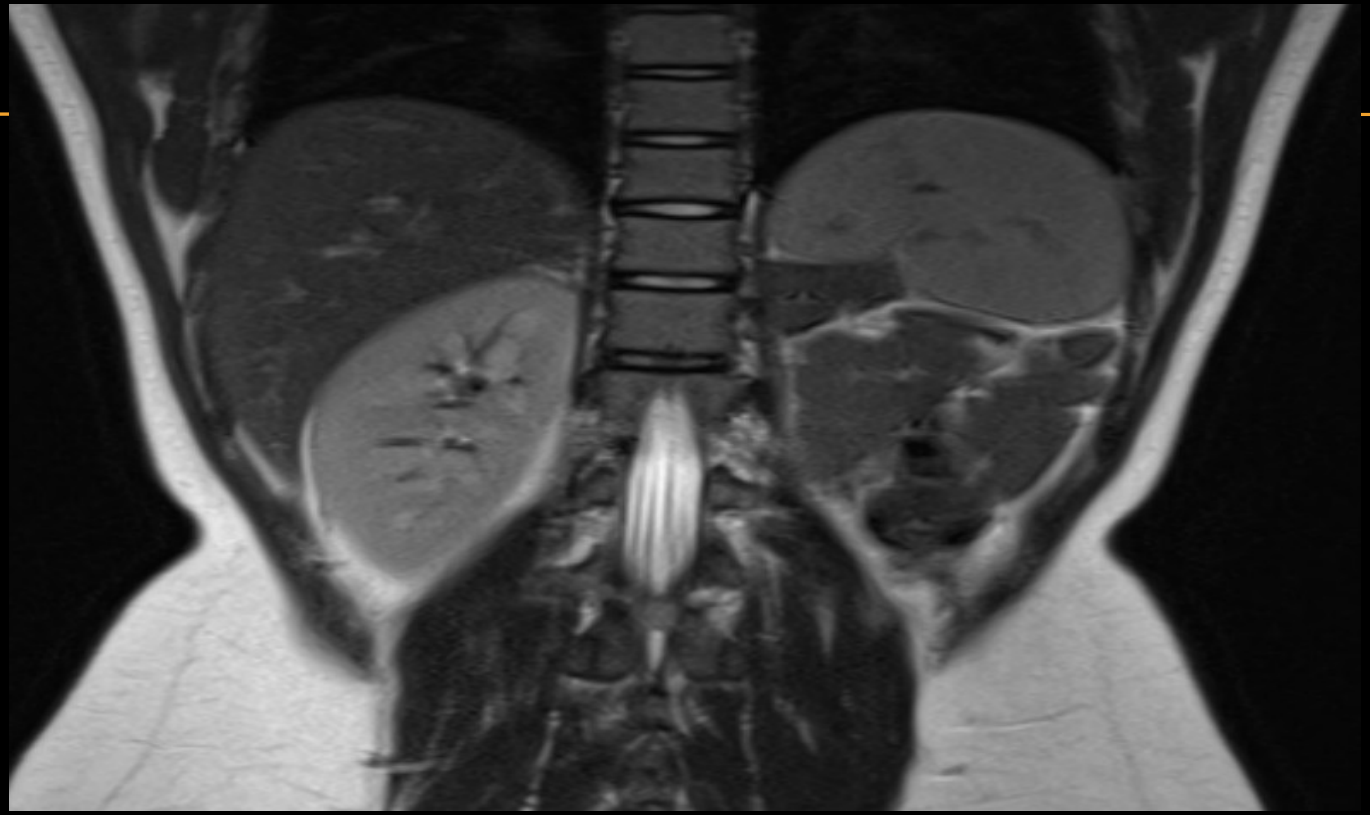
T1 FS



LEFT LOWER UTERINE BODY AND CERVIX APPEARED GROSSLY DISTENDED WITH COLLECTION APPEARING HYPERINTENSE ON T1WI AND T2WI AND SHOWING NO SUPPRESION ON T1FS SEQUENCE.



T2W CORONAL IMAGE REVEALING A DILATED TORTOUS FALLOPIAN TUBE CONTAINING COLLECTION CORRESPONDING TO BLOOD PRODUCTS.



T2W CORONAL IMAGE SHOWING LEFT RENAL AGENESIS

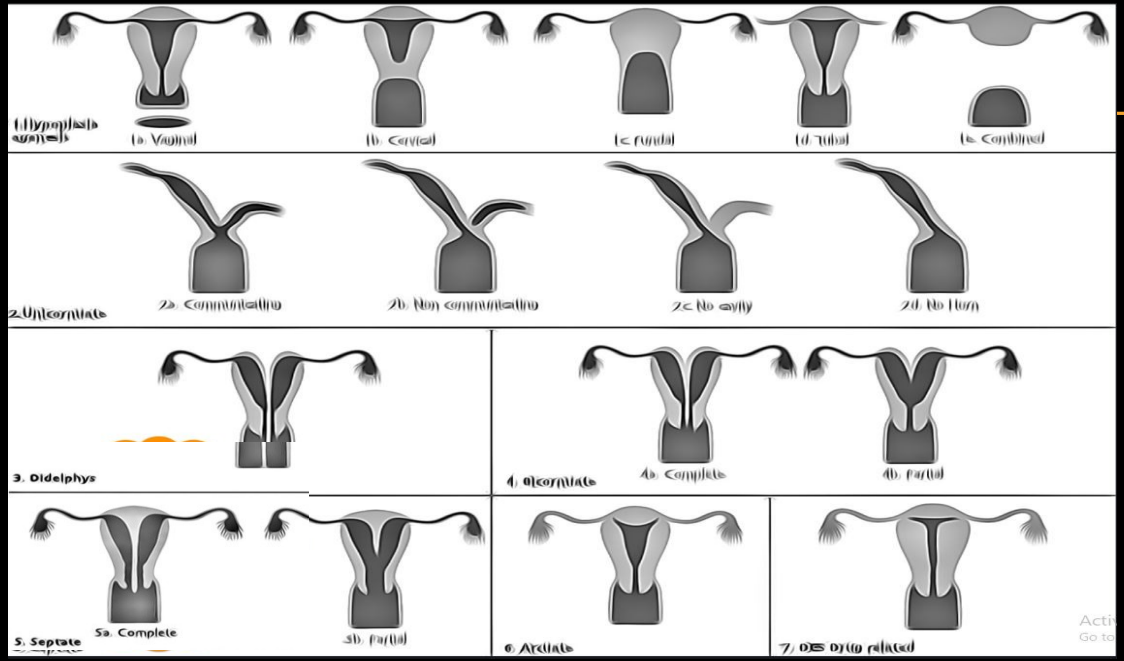
IMPRESSION:

- Left renal agenesis.
- Uterine didelphys.
- Left hemivaginal obstruction causing left hematometrocolpos and hematosalphinx.

- -This is most likely suggestive of OHVIRA (Obstructed hemivagina and ipsilateral renal anomaly) syndrome/ Herlyn-Werner-Wunderlich (HWW) syndrome.

DISCUSSION:

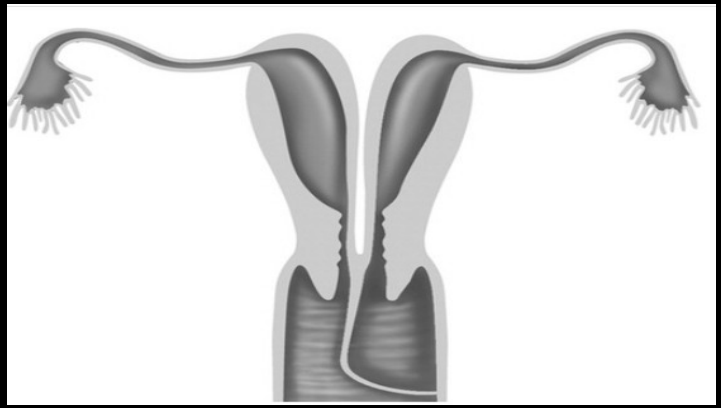
- The müllerian ducts are paired embryologic structures that undergo fusion and resorption in utero between the 6th and 11th week of gestation to give rise to the uterus, fallopian tubes, cervix, and upper two-thirds of the vagina. Interruption of normal development of the müllerian ducts can result in formation of müllerian duct anomalies (MDAs).
- MDAs are not associated with anomalies of the external genitalia or ovarian development as they have different origin.
- MDAs are commonly associated with renal anomalies, including renal agenesis, ectopia , hypoplasia, fusion, malrotation, and duplication.



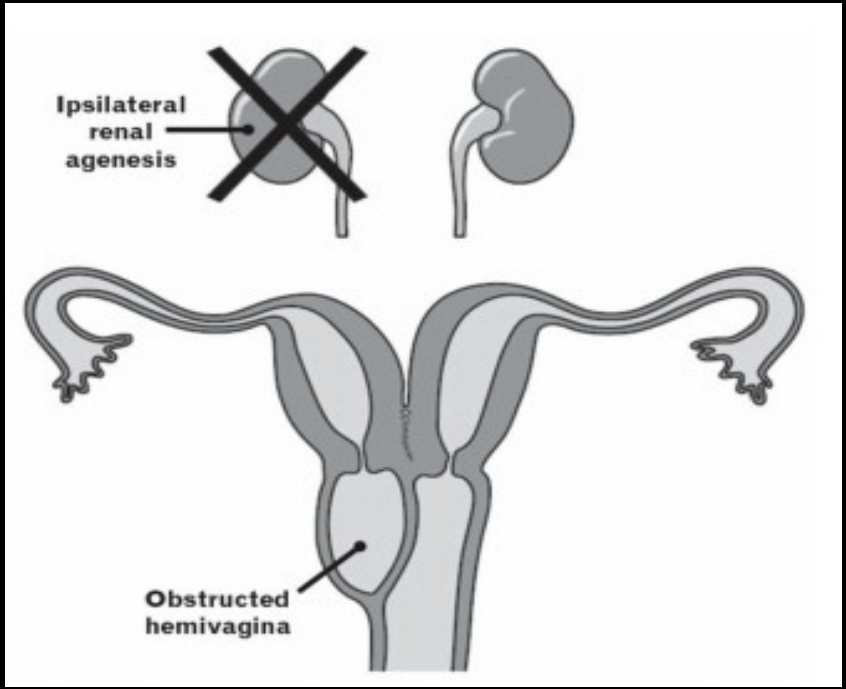
Complete failure of the fusion of the mullerian duct results in the two separate right and left hemiuteri and cervix.

TYPE III MDA

MDA CLASSIFICATION BY AMERICAN FERTILITY SOCIETY (AFS).



OBSTRUCTED HEMIVAGINA WITH IPSILATERAL RENAL AGENESIS (OHVIRA)



- Syndrome consists of
1. uterine didelphys ,
2. unilateral low vaginal obstruction,
3. ipsilateral renal agenesis
- All 3 components being secondary to mesonephric duct-induced müllerian anomalies.
- Developmental arrest of ipsilateral mesonephric duct results in failure of distal hemivagina to develop, thereby resulting in obstructed hemivagina.

- OHIVARA syndrome present either early in infancy because of collection of secretions in the obstructed vagina under influence of maternal hormones or within 1 - 2 years of onset of menarche due to development of hematocolpos , hematometra, or even hematosalpinx.
- The patients present with cyclical dysmenorrhea, which later evolves into persistent pelvic pain.
- Nearly all of them are initially misdiagnosed clinically as endometriosis or tubercular collection on clinical grounds.
- Treatment invariably requires surgical intervention in the form of excision or complete division of vaginal septum to relieve obstruction.

WHY IS MRI ESSENTIAL?

- Limitation of TVS and HSG evaluation in female child and adolescent girls.
- TAS is limited because of field of view and distortion of normal anatomy due to abnormality.
- MRI gives a detailed delineation of uterovaginal anatomy hence is the investigation of choice for evaluation of Mullerian duct anomaly with reported accuracy of 100% in the evaluation.
- Noninvasive and non-ionizing investigation add to the advantage.

REFERNCES:

Magnetic Resonance Imaging in Diagnosis and classification of Uterovaginal Congenital Anomalies

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ABSTRACT

Uterine congenital anomalies are clinically relevant because these are associated with increased incidences of infertility and menstrual disorder. These are frequently not diagnosed at birth. These anomalies usually diagnosed at child-bearing age when reproductive malfunction arise. When normal mullerian duct development interrupted at any stage, it results in mullerian abnormality. Magnetic resonance imaging (MRI) has an excellent role in evaluation of mullerian duct anomalies. Ultrasonography is the primary investigation; however, MRI is an excellent noninvasive investigation for accurate evaluation of uterine congenital anomalies. MRI is a very good modality to evaluate the vaginal malformation, which usually difficult to evaluate by ultrasound.

KEYWORDS: *Congenital, infertility, mullerian, uterus, vagina*

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- ***THANK YOU…***