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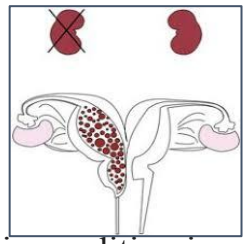
Title: OHVIRA Syndrome :A rare cause of abdominal pain in an adolescent girl

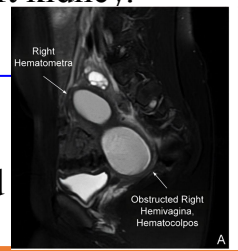
INTRODUCTION : Obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome is a rare congenital defect of the Müllerian ducts characterized by **uterus didelphys**, **unilateral obstructed hemivagina**, and **ipsilateral renal agenesis**. It was previously known as **Herlyn-Werner-Wunderlich syndrome.** Typically diagnosed after menarche, this condition is often associated with severe dysmenorrhoea and abdominal pain. Diagnosis can be delayed due to normal onset of menses from non-obstructive side. In this report, we present a case of OHVIRA syndrome in a 16-year old girl who presented with lower abdominal pain.

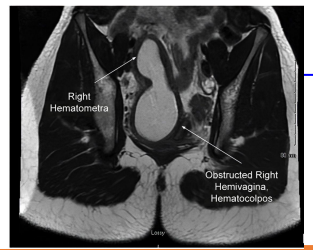


CASE REPORT : A 16 year old girl presented to the OPD of Motherhood Hospital, Indore with complain of right side abdominal pain since 5 days along with 1 episode of vomiting. She had menarche at 13 years of age with history of dysmenorrhoea during menstrual cycle. On examination, she was hemodynamically stable with normal external genitalia. Lower right abdominal tender mass with guarding was noted.

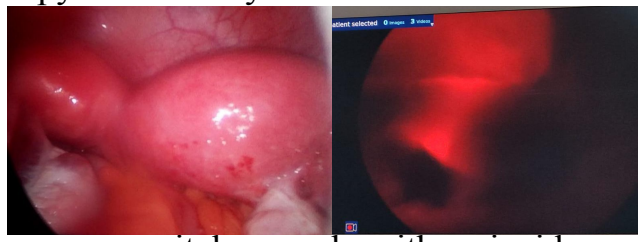
INVESTIGATION	FBC, U/E, Inflammatory markers-normal, B-HCG -neg.
USG and MRI Abdomen and Pelvis	Uterus with 2 cavities with distension of right horn and cervix s/o Hematometra (11*7*6). Right kidney absent with hypertrophied left kidney.

Thus a **diagnosis of uterine didelphys with Obstructed right hemivagina with right renal agenesis** made.Parents were counseled and the patient was planned for examination under anesthesia (EUA) and lap- hysteroscopy followed by definitive treatment in the same sitting.





PROCEDURE: Resection of vaginal septum was done with drainage of 500ml of old blood from obstructed hemivagina. Two cervices were visualised



DISCUSSION: OHVIRA syndrome is a rare congenital anomaly with an incidence rate ranging from **0.1% to 3.8%** in the general population. In 1992, **Pedro Acien** proposed that the vagina is embryologically derived from the **mesonephric ducts**. The caudal portion of the mesonephric ducts enlarges to form the sinovaginal bulbs which later fuse to form the vaginal plate. So, in case of distal agenesis of a mesonephric duct, there will be **agenesis of the ureteral bud** on that side, hence ipsilateral renal agenesis, along with associated uterine malformations. OHVIRA syndrome is classified under **class III (ASRM classification)** and under **class U3bC2V2(ESHRE classification).** Diagnosis of OHVIRA syndrome requires a multimodal approach, which includes a detailed history, meticulous examination, and appropriate imaging studies (USG and MRI).Its treatment comprises of excision of the vaginal septum. Complicated cases may require ipsilateral hysterectomy.

CONCLUSION: Obtaining a detailed menstrual history and identifying the relationship between renal and uterovaginal abnormalities are critical steps in the diagnostic process. Maintaining a **high index of suspicion** in any adolescent female with vague abdominal pain is essential to achieve timely diagnosis and avoid complications.

REFERENCES:1)Acien P. Embryological observations on the female genital tract.2)The ESHRE/ESGE consensus on the classification of female genital tract congenital anomalies.3)Heinonen PK. Renal tract malformations associated with Müllerian duct anomalies.