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The Herlyn-Werner-Wunderlich (HWW) syndrome– A rare case report

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Title: The Herlyn-Werner-Wunderlich (HWW) syndrome– A rare case report

Introduction: Herlyn-Werner-Wunderlich (HWW) syndrome is a rare female urogenital anomaly. It represents a diagnostic dilemma because of the regular menstruation and non-specific symptoms. It results from the combination of mesonephric and mullerian ducts anomaly. It is presented in post menarche young adults with irregular menses, lower abdominal pain and pelvic mass, which in most of the time is not ruled out due to its rarity. Early detection and septoplasty (surgical resection of obstructing vaginal septum) will relieve pain and prevent further complications such as endometriosis and infertility. The unique feature of our case is right sided renal agenesis; uterus didelphys with obstructed hemi vagina and to infected collection along the vaginal wall.

Epidemiology: Post menarche young females is the most common age of diagnosis. The exact incidence of this syndrome is unknown, but its estimated occurrence is 0.1%-3.8%[1]. Many of these are not detected during the women's lifetime unless they are symptomatic and evaluated properly.

Etiopathogenesis:

During fetal development, the Mullerian ducts represent the primordial components of the female reproductive system. They differentiate into the fallopian tubes, uterus, cervix, and the superior aspect of the vagina. The Wolffian duct gives rise to the ipsilateral ureteric bud and thus is responsible for the formation of the kidney. Embryologic arrest around 8 weeks of gestation that simultaneously affects the adjacent Mullerian and Wolffian ducts leads to didelphys uterus and renal agenesis. Accordingly, in the absence of the Wolffian duct on one side, the kidney and ureter (of the same side) will fail to fuse. On the side on which the Wolffian duct is missing, the Mullerian duct is displaced laterally and fails to adequately fuse with the urogenital sinus, leading to the formation of a blind sac, imperforate or obstructed hemivagina, right side in the present case. The distal part of vagina which arises from the urogenital sinus is not affected and develops normally[2].

Clinical presentation:

Usually present with pelvic/ lower abdominal pain secondary to hematocolpos and pelvic mass. May also present with fever, pain and abscess formation[1].

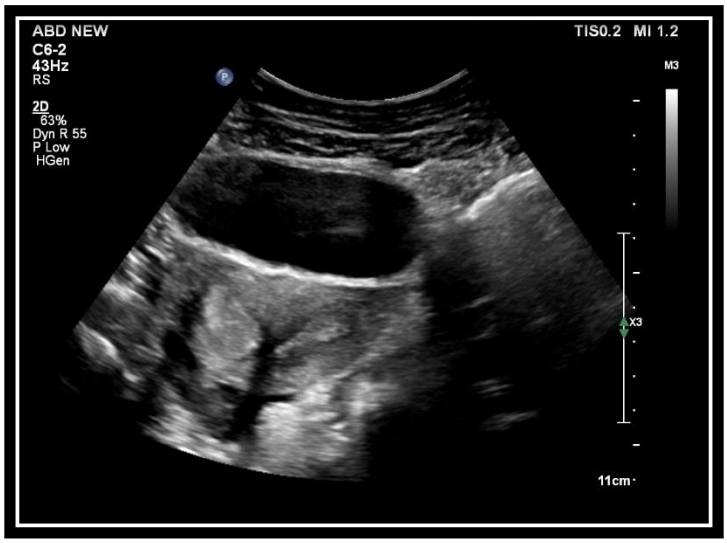
Case report: A 20-year-old unmarried female, student by occupation, well built and nourished presented with intermenstrual spotting from past 1 month. No h/o pelvic pain/ mass/ fever. No h/o of similar complaints in the past. Attained menarche at 14 years. Menstrual history otherwise normal.

Known case of right renal agenesis (diagnosed incidentally at the age of 8 years).

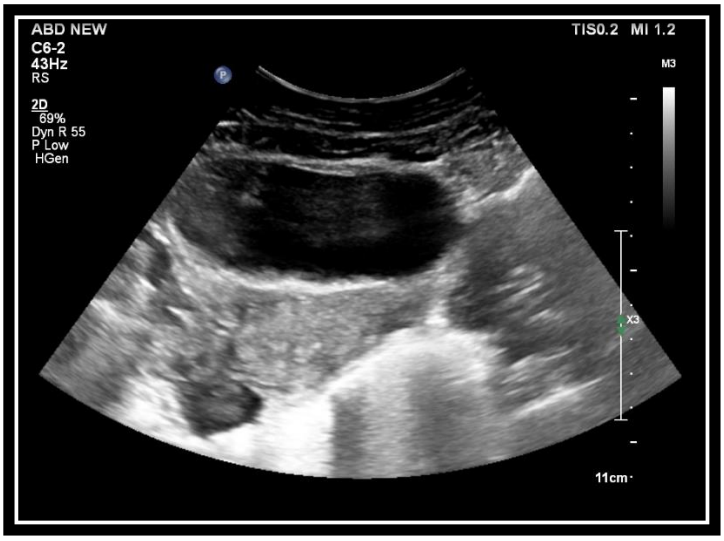
Pelvic examination by gynecologist revealed a cystic bulge to the anterior wall of vagina. PV examination was not performed for the sake of integrity of the hymen.

Imaging:

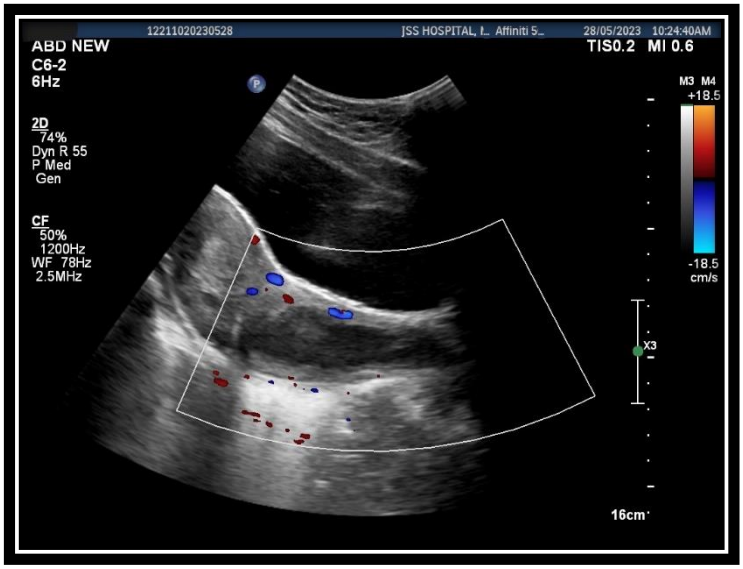
USG ABDOMEN AND PELVIS(TAS):



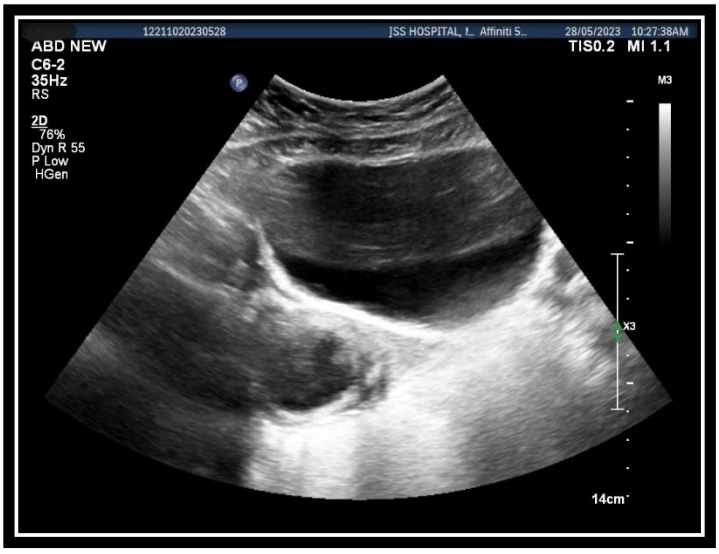
USG-Two separate uterine horns (cornua) (Endometrial thickness Right (15mm)>left (8mm).



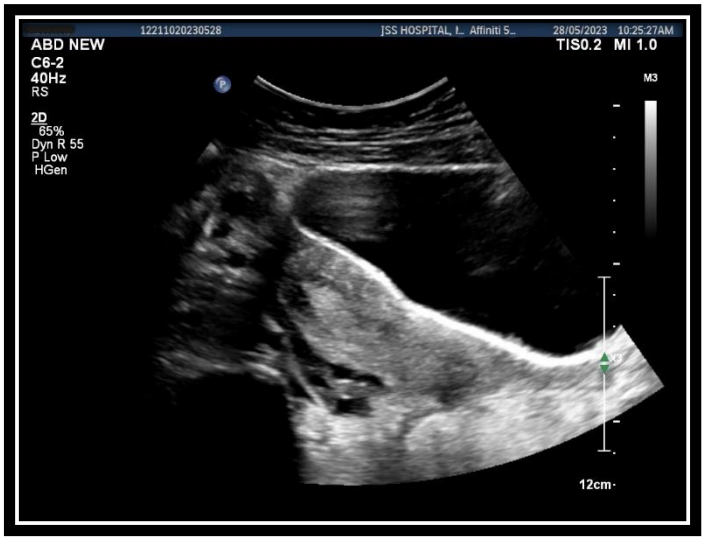
Endometrium extending till external OS of cervix – two separate cervices



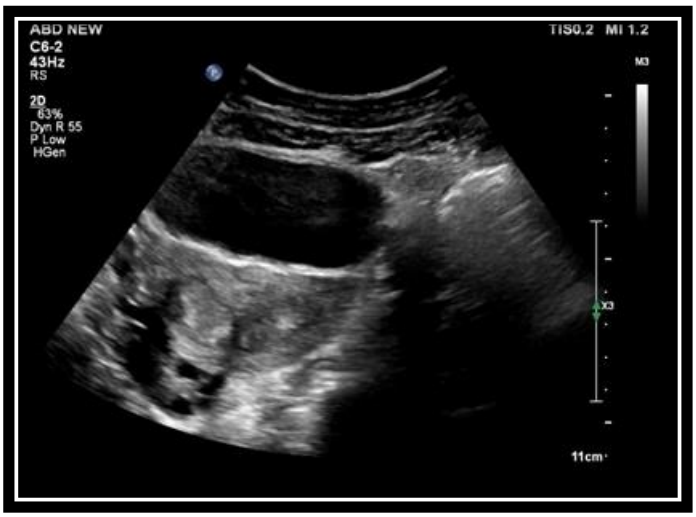
Right cornua of uterus, right cervix with a well-defined cystic in right vaginal cavity in sagittal plane



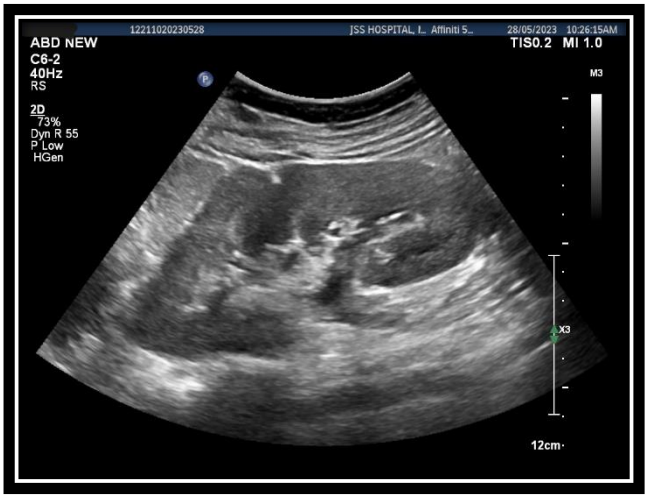
Urinary bladder with above mentioned collection in transverse plane

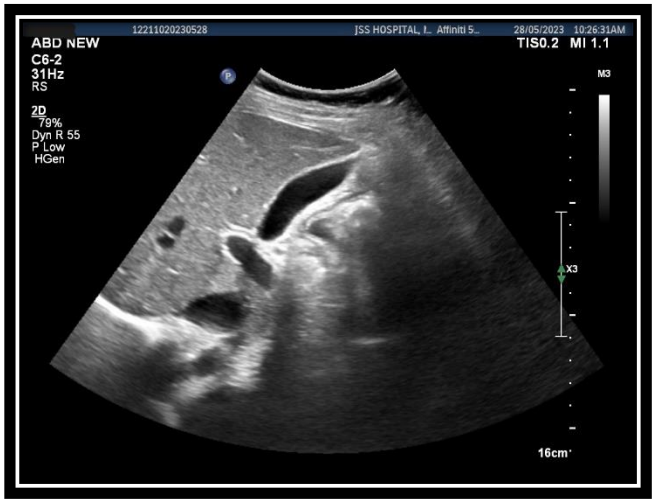


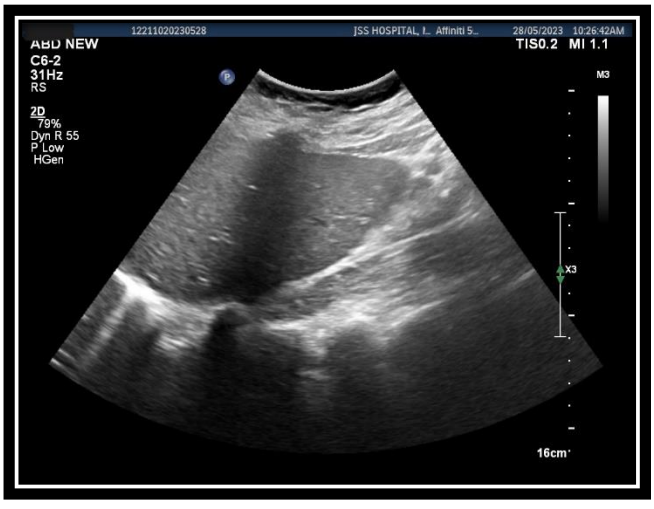
Left cornua of uterus and left cervix with collapsed left vagina.



Polycystic morphology of ovary

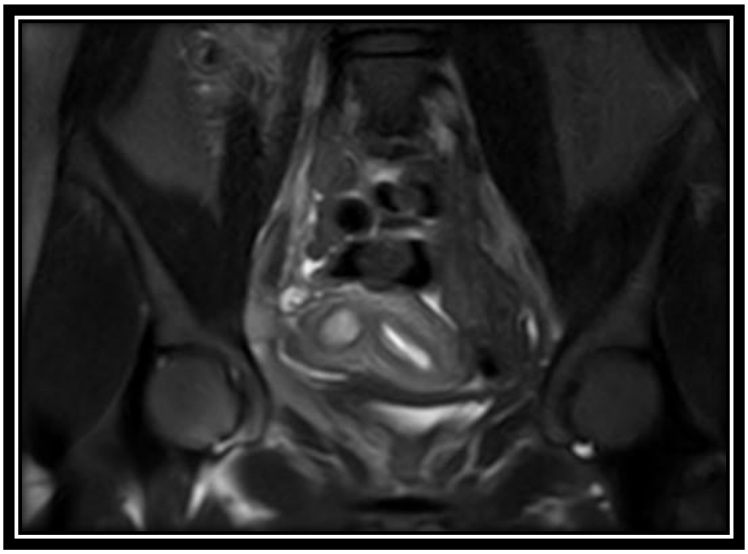


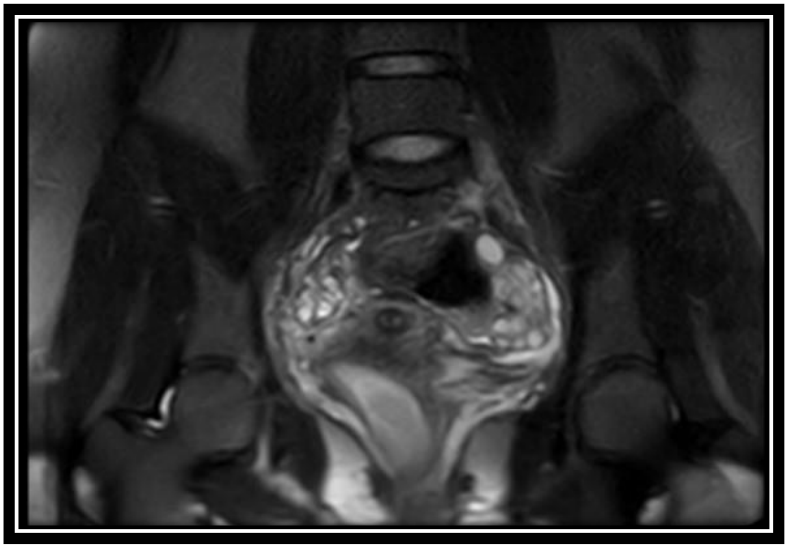




Absent right kidney with normal left kidney

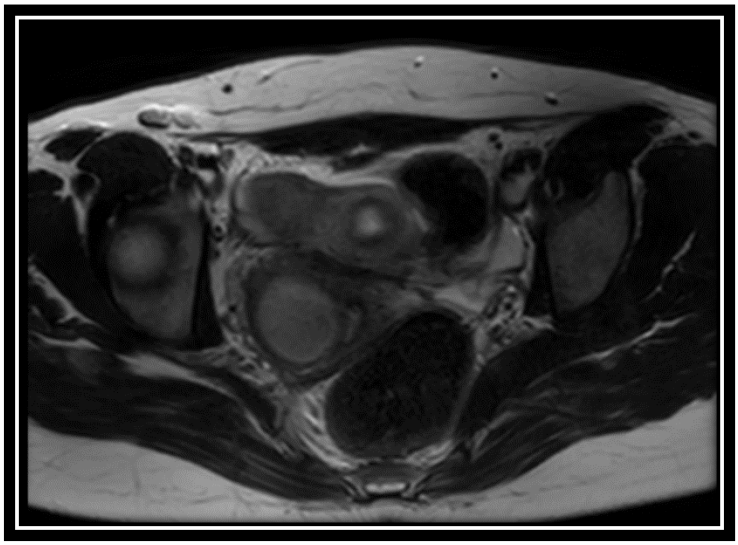
CE MRI ABDOMEN AND PELVIS :

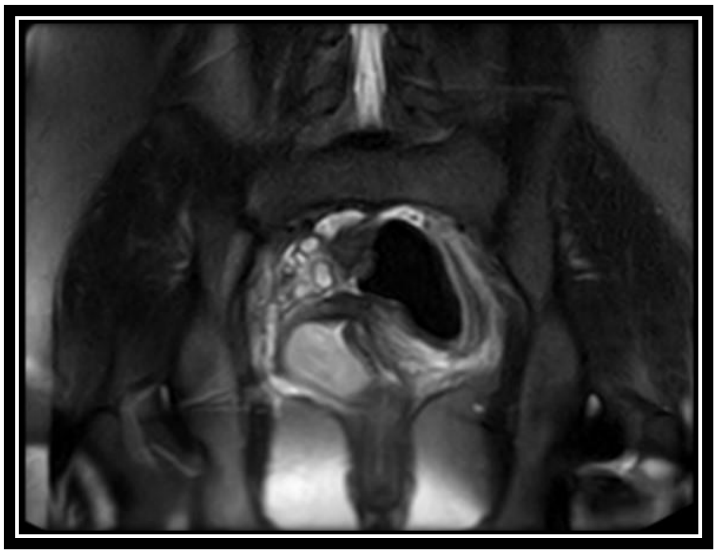




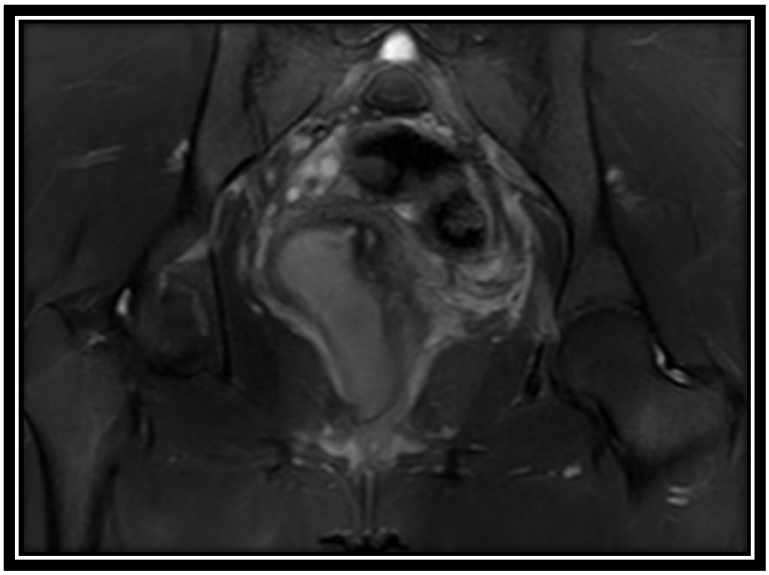
T2 Wt images Pelvis coronal sections showing two separate uterine cornua with normal myometrium traversing between two extending up till the external cervical OS inferiorly resulting in two separate cervices.

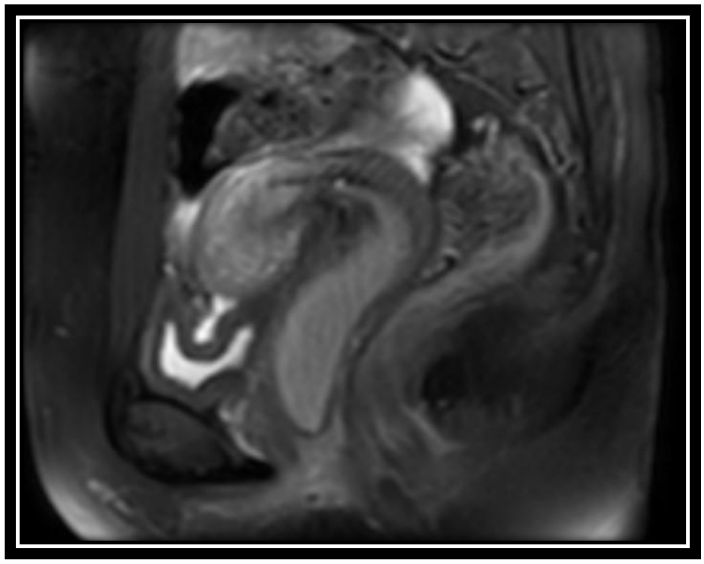
Bilateral ovaries show polycystic morphology



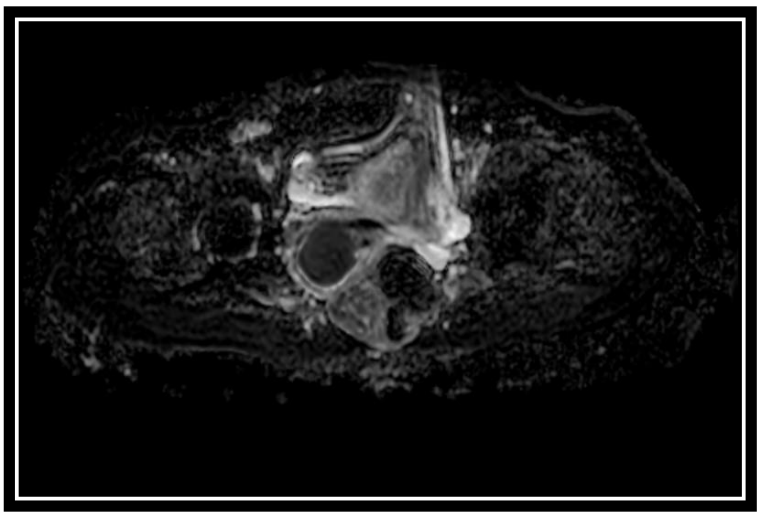


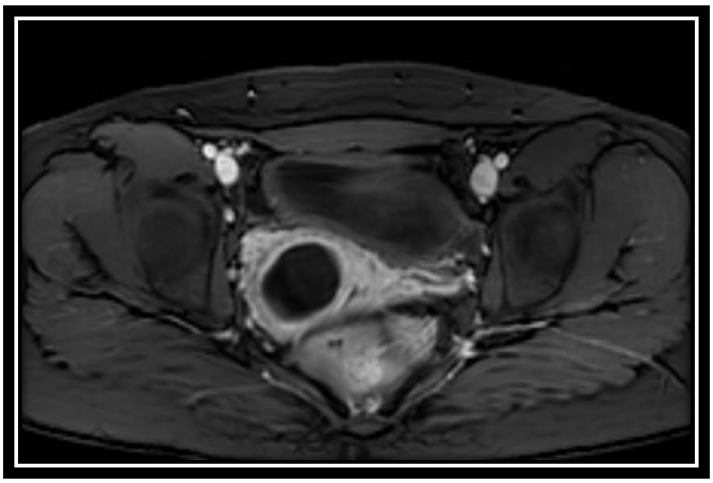
T2 Wt images Pelvis axial and coronal sections showing dilated right vaginal cavity with water intensity collection measuring ~ 7.4 x 3.5 x 3.7 cm, volume 52cc appearing hyperintense on T1wt images causing mass effect over collapsed left vaginal cavity.





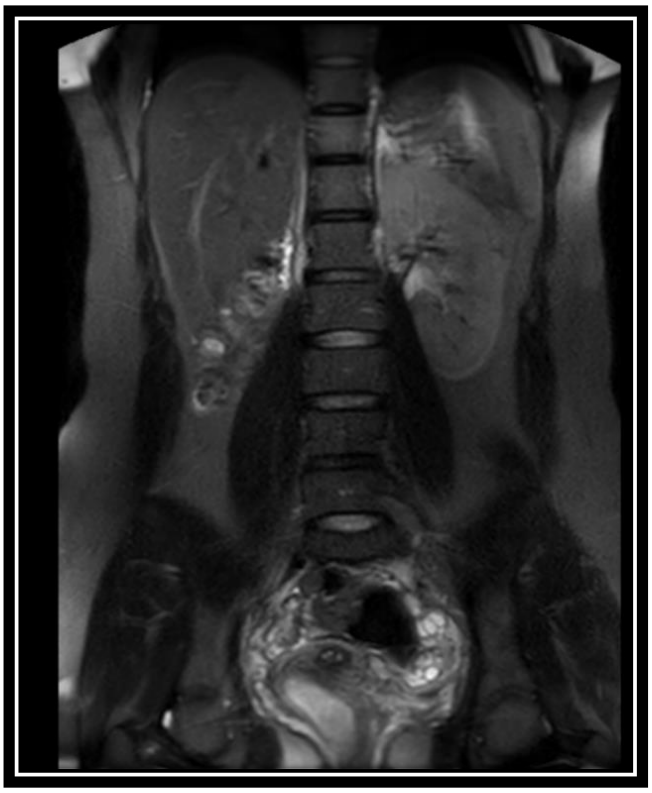
T1 Wt images Pelvis axial and sagittal sections showing dilated right vaginal cavity with above mentioned collection appearing hypointense on T1wt images.





DWI sequence showing diffusion restriction in the above-mentioned sequence

Post contrast T1 Fat sat axial images showing peripheral enhancement in above mentioned collection



T2 Wt coronal sequence of abdomen and pelvis showing normal left kidney with absent right kidney, above mentioned T2 hyperintense collection

Final diagnosis: Uterus didelphys with obstructed hemi-vagina with a collection in right vagina causing mass effect over collapsed left vagina and ipsilateral renal agenesis (OHVIRA) syndrome/ The Herlyn-Werner-Wunderlich (HWW) syndrome.

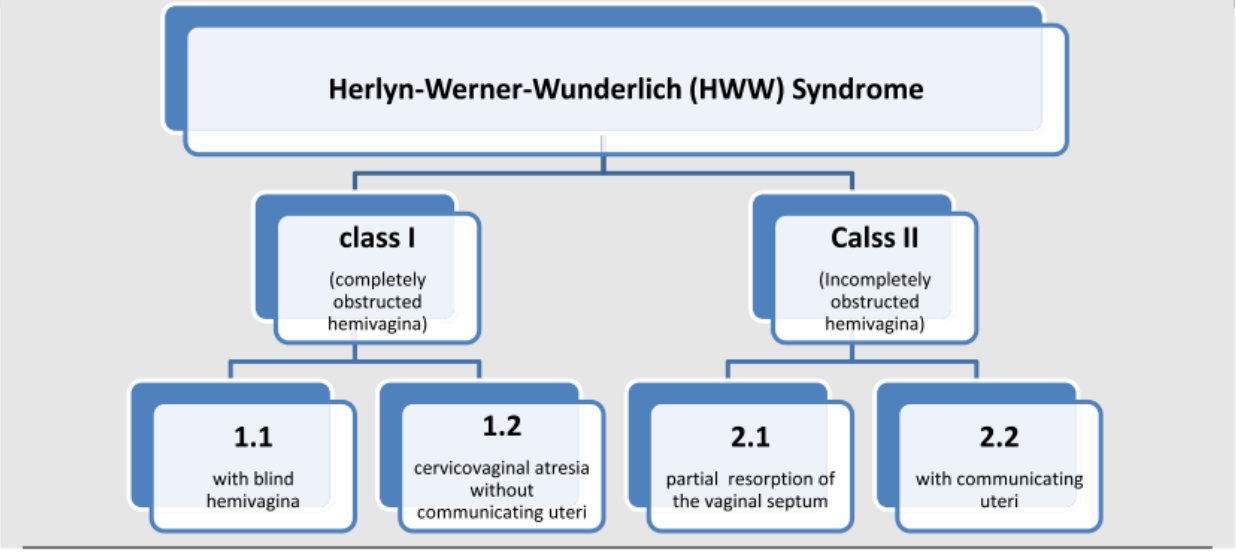
Treatment:

Surgical correction is the treatment of choice

Teaching points:

- Young female patients presenting with lower abdominal/pelvic symptoms especially when associated with renal anomaly/agenesis MDA should be considered as differential diagnosis.
- Prompt diagnosis and excision of the obstructed vaginal septum can relieve these symptoms completely and prevent further sequelae like infertility.
- USG and MRI play major role in diagnosing this condition since most patients will have normal clinical examination and laboratory findings although mass can be palpated in few cases.

Table 1 – Lan Zhu et al. new classification of HWW syndrome.



References

1. Zahran KM, Abd El Aal DE, Othman MH, Ahmed ER. Uterus didelphys with imperforate hemivagina and ipsilateral renal agenesis complicated by hematocolpos, hematometra and hematosalpinex. The challenge of intact hymen. Middle East Fertility Society Journal. 2011 Dec 1;16(4):291-4.
2. Hayat AM, Yousaf KR, Chaudhary S, Amjad S. The Herlyn-Werner-Wunderlich (HWW) syndrome–A case report with radiological review. Radiology Case Reports. 2022 May 1;17(5):1435-9.
3. Bhoil R, Ahluwalia A, Chauhan N. Herlyn Werner Wunderlich syndrome with hematocolpos: an unusual case report of full diagnostic approach and treatment. International Journal of Fertility & Sterility. 2016 Apr;10(1):136.