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# A CASE REPORT OF OHVIRA SYNDROME – A RARE CONGENITAL ANOMALY

**Radio-Diagnosis**

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## ABSTRACT

Obstructed hemivagina with ipsilateral renal agenesis (OHVIRA) is a rare complex congenital structural abnormality of the female urogenital tract. The classical triad includes uterine didelphis, obstructed hemivagina and ipsilateral renal agenesis. It usually presents after menarche with non-specific and variable complaints of progressive pelvic pain, sometimes with regular menses causing a delay in the diagnosis. Mullerian duct anomalies (MDAs) are infrequently encountered in routine clinical settings, therefore making the diagnosis of OHVIRA even more challenging.[1] We report a case of a young adolescent female, referred to us for USG abdomen and pelvis, who presented with acute abdominal pain and acute urine retention to casualty. On USG, a provisional diagnosis of OHVIRA syndrome was given and MRI was suggested for confirmation and better delineation of the pelvis. The above diagnosis was confirmed on MRI, which is the gold standard imaging technique for assessing MDA.

## KEYWORDS

OHVIRA; Uterus didelphys; Renal agenesis, MDA

## INTRODUCTION

The female reproductive tract develops from a pair of Mullerian ducts which fuse between the 6th and 11th week of gestation to form the uterus, fallopian tube, cervix, and proximal two-thirds of the vagina. Any disruption in Mullerian duct development during embryogenesis can result in various Mullerian duct anomalies (MDAs).[2] However, the ovaries and the lower third of the vagina have different embryologic origins, which are primitive yolk sac and sinovaginal bulb. Therefore, MDAs are usually isolated anomalies and are not found in association with anomalies of the external genitalia or ovarian development.[3]

Normal development of the Müllerian ducts depends on completing three phases: Organogenesis, fusion and septal resorption. The formation of both Müllerian ducts characterizes organogenesis. Uterine agenesis/hypoplasia or a unicornuate uterus results from this failure. In the second phase, these Mullerian ducts fuse to form the uterus. Failure of which results in a bicornuate or didelphys uterus. The central septum, which was formed as a result of the second stage, is reabsorbed during the third phase and defects in this stage result in a septate or arcuate uterus.[2]

According to the classification system of the American Society of Reproductive Medicine, MDAs are classified into 5 classes, namely - I) Hypoplasia/agenesis, II) Unicornuate, III) Didelphys, IV) Bicornuate V) Septate.

In our study, the patient was diagnosed to have class III uterine anomaly, that is, didelphys uterus which results from complete non-fusion of both Müllerian ducts. The individual horns were fully developed and were normal in size. A deep fundal cleft and two cervices were present. A longitudinal or transverse vaginal septum may be present in didelphys uterus as seen in our case.

Association of MDA with renal anomalies is common and most frequently encountered with unicornuate uterus. Renal agenesis is the most commonly reported anomaly. Other anomalies include ectopic kidney, horseshoe kidney, renal dysplasia and duplicated collecting systems.

Diagnosis of MDAs is very important as it carries a high risk of infertility, endometriosis, and miscarriages. Therefore, early detection and classification are helpful in the appropriate course of action.

### Case Study

A 15-year-old female presented with complaints of acute urinary retention for one day and abdominal pain for 10 days. Gynaecological history revealed that she attained menarche 8 months back and has had regular, 28-day cycles with a 3-5-day flow since then, with a history of difficulty initiating voiding of urine and urgency during her previous menstrual cycle. On review, she had no complaints of fever, dysmenorrhea or abnormal vaginal discharge. She had a soft, non-tender abdomen and normal external genitalia on examination. Laboratory findings were unremarkable.

### USG Findings -

On Transabdominal USG, the right kidney was not seen in the right renal fossa. The left kidney was normal. A large, well-defined, thick-walled anechoic cystic lesion was noted in the right adnexa, suggestive of Hematometrocolpos. The uterine cavity was displaced towards the left adnexa; hence a possibility of uterus didelphys was given (Figure 1). Bilateral ovaries appeared normal. With the above findings, OHVIRA syndrome was given as a provisional diagnosis.

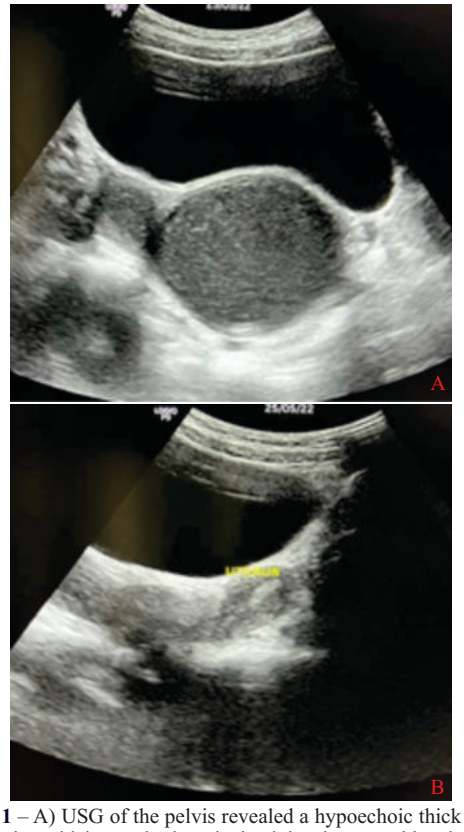


**Figure 1** – A) USG of the pelvis revealed a hypoechoic thick walled cystic lesion with internal echoes in the right adnexa pushing the uterus to the left side. B) Normal looking uterus in the left adnexa.

**MRI Findings -**

Contrast enhanced pelvic MRI was performed for further evaluation, which demonstrated two separate uterine horns (Figure 2). The two separate uterine horns were seen separated by an isointense septum which appeared to be terminating at the level of introitus (Figure 3). The endometrium of the right horn was mildly dilated (~12.1mm).

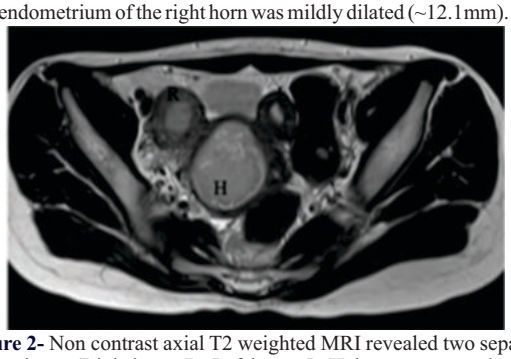


**Figure 2-** Non contrast axial T2 weighted MRI revealed two separate uterine horns (Right horn– R ; Left horn – L; H- hematometrocolpos)

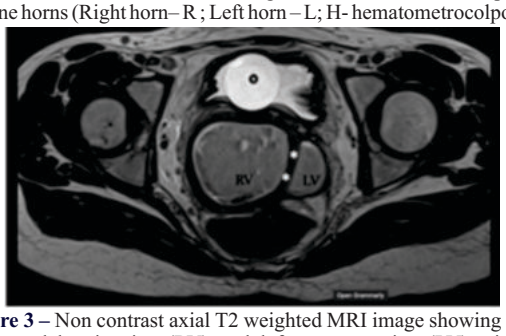


**Figure 3 –** Non contrast axial T2 weighted MRI image showing right obstructed hemivagina (RV) and left patent vagina (LV) with an isointense vaginal septum (*)

Dilated fluid-filled tubular structure containing contents that are hyperintense on both T1W and T2W sequences was noted with an open communication to the right uterine horn, suggestive of dilated vagina containing old blood products- Hematometrocolpos (Figure 4 and Figure 5)

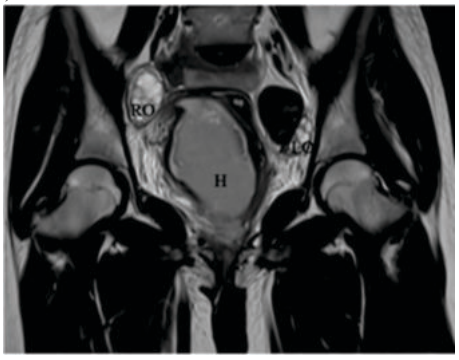


**Figure 4** - Non contrast axial T2 weighted image showing a large right hematometrocolpos (H) and normal right (RO) and left ovaries (LO)

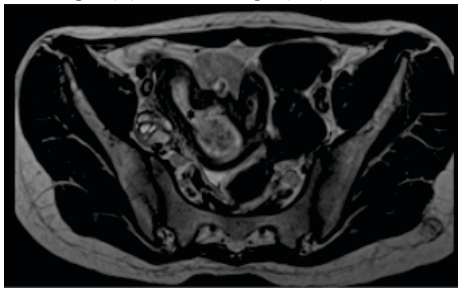

**Figure 5 –** Non contrast axial T2 weighted MRI image showing communication of the pelvic mass with the right horn (*)

Left hemivagina appeared normal with minimal blood products within and had a patent external opening.

Bilateral ovaries appeared normal.

The final diagnosis on MRI was uterine didelphys with an obstructed right hemivagina. In association with ipsilateral right renal agenesis, the diagnosis of OHVIRA was confirmed.

**Operative Findings –**

The patient was taken for Vaginal septum resection and diagnostic laparoscopy, which confirmed two uterine horns and a patent left cervix (Figure 6). Septal resection was done and around 250ml of chocolate-coloured fluid was drained in keeping with old collected blood. The resected septal wall was sutured to the anterior and posterior vaginal walls.

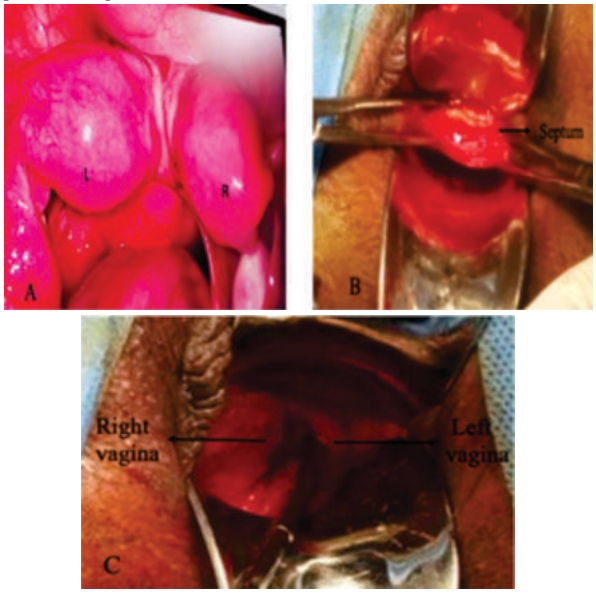


**Figure 6:** Intraoperative images showing – A) Two uterine horns. B) Transverse vaginal septum which is held with Allis forceps. C) Patent left vagina and blind ending right vagina.

**DISCUSSION**

OHVIRA is a rare congenital Mullerian tract anomaly. The worldwide prevalence of MDAs is only 2-3%. OHVIRA was first mentioned in 1922. The association of Renal agenesis with ipsilateral blind hemivagina was first reported in 1971 as Herlyn-Werner syndrome and the association of renal aplasia, bicornuate uterus with isolated hematocervix was first reported in 1976 by Wunderlich, so it was previously known as Herlyn-Werner-Wunderlich syndrome.[4]

The exact etiology and pathogenesis of OHVIRA are unknown but are thought to be multifactorial. It is assumed to be caused by a developmental anomaly in both the Mullerian (paramesonephric, forms female organs) and Wolffian (mesonephric, forms male organs) ducts.[5] In females, Mullerian ducts are present lateral to the Wolffian ducts and grow downwards and towards the midline and finally cross Wolffian ducts and fuse with each other to form the uterovaginal canal, which later on develops into the fallopian tube, uterus and upper two thirds vagina. After fusing, the Mullerian ducts join the urogenital sinus which forms the lower third of the vagina. Wolffian ducts, in addition to giving rise to the ureter and kidneys, also induce the development and fusion of Mullerian ducts. Each Mullerian duct forms its own hemiuterus and cervix. On the side of the absent Wolffian duct, the Mullerian duct is pushed laterally and will not fuse with the contralateral duct, resulting in a didelphic uterus. The contralateral ducts follow the original pathway fusing centrally with the urogenital sinus forming its own vagina, while the displaced duct forms a blind sac, resulting in an imperforate or obstructed hemivagina. Sometimes may be accompanied by a transverse or longitudinal septum.[6]

USG and MRI are incredibly useful in diagnosing and categorizing MDAs.[7] USG is usually the first line of investigation for a suspected MDA or in any pelvic symptoms as it is widely available and non-invasive. The presence of hematocolpos, which appears as a fluid collection with low-level echoes, can aid in the identification of uterine anomalies (bicornuate/didelphic). USG also allows the identification of Renal anomalies (absent kidney; multicystic kidney; blind megaureter) easier, which is a component of OHVIRA. However, MRI remains the gold standard imaging for the diagnosis of Mullerian

anomaly with higher sensitivity. The number and disposition of the uterine horns, cervices, continuity with the vaginal channels (obstructed and unobstructed) and the nature of the echogenic contents in the vaginal channels are all more precisely identified on MRI.

Vaginoplasty is the treatment of choice for OHVIRA primarily to relieve the obstruction and to restore normal vaginal function. Hemihysterectomy should be avoided because the incidence of pregnancy in both horns is said to be equal. It is preferred only in cases where vaginoplasty does not relieve the hematometrocolpos or if the vaginal stenosis reoccurs.

### Outcome And Follow Up

The patient came for a follow up to obstetrics and gynecology department after 2 months and had no complaints and the examination was unremarkable.

## CONCLUSIONS

As MDA are a rare entity, knowledge of the syndrome is essential for early diagnosis and treatment in order to avoid complications. When encountering adolescent patients with non-specific abdominal or pelvic symptoms or mass, clinicians and radiologists should raise high suspicion of OHVIRA syndrome. It would be wise to check for genital anomalies whenever a multicystic dysplastic kidney or absent kidney is encountered in a fetus or newborn and should be followed up till puberty.

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