International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Multimodality Imaging and Management of OHVIRA Syndrome: A Case from Diagnosis to Successful Pregnancy

Dr. Deep Kumar Roy[1], Dr. Hadiqa Shirin[2], Dr. Subhrasmita Bhoi[3], Dr. Rupak Bhuyan[4]

[1, 2, 3, 4]Jorhat Medical College Hospital, Jorhat, Assam, India

Abstract: ***A 24-year-old female presenting with primary infertility was diagnosed with OHVIRA syndrome, a rare congenital anomaly involving uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis. Multimodal imaging, including ultrasound and MRI, confirmed the diagnosis. Surgical correction led to remarkable outcomes, with the patient achieving spontaneous conception within six months. This case emphasizes the critical role of early diagnosis, targeted surgical intervention, and radiological follow-up in managing OHVIRA syndrome and preserving fertility.***

Keywords: OHVIRA syndrome, Herlyn-Werner-Wunderlich syndrome, uterus didelphys, Müllerian anomalies, infertility

1. Introduction

OHVIRA syndrome, also known as Herlyn-Werner-Wunderlich syndrome, is a rare congenital anomaly characterized by duplications of female genital tract with obstructed hemivagina and ipsilateral renal agenesis or rarely other renal abnormalities. There is also reported association of Gartner's duct cyst and pelvic endometriosis. The estimated incidence is around 0.1-3.8% of all Müllerian duct anomalies. Early diagnosis and appropriate management are crucial for preserving fertility and preventing complications.[1]

2. Case Presentation

A 24-year-old female presents with primary infertility, reporting 2 years of unsuccessful conception attempts. Her medical history is unremarkable, with regular menstrual cycles complicated by dysmenorrhea.

Diagnostic Assessment

Ultrasonographic evaluation reveals a uterine duplication anomaly, with two separate cavities and distinct endometrial echoes. The left hemivagina is dilated, containing echogenic fluid suggesting hematometrocolpos. Additionally, there is absence of the left kidney (renal agenesis) and a Bartholin's cyst on the right lateral side. Bilateral ovaries appear normal.

The MRI examination demonstrated a bicornuate bicollis uterus, featuring two distinct uterine bodies and cervices with an external indentation measuring 1.7 cm and an intercornual distance of 5.1 cm. Additionally, the study revealed a left-sided obstructed hemivagina with associated hematometrocolpos, a complete longitudinal vaginal septum, and renal agenesis involving the left kidney. The right kidney and collecting system were normal. A coincidental Bartholin's cyst was identified on the right lateral side, with no other associated anomalies detected.[2]

3. Treatment

The patient underwent vaginoplasty with excision of the vaginal septum and drainage of hematocolpos. Post-operative recovery was uneventful.

Follow-up and Outcomes

Post-surgical follow-up revealed remarkable outcomes, with complete resolution of symptoms observed at three months. Notably, the patient achieved spontaneous conception within six months. First-trimester ultrasonography at 12.4 weeks gestation demonstrated a single live intrauterine pregnancy in the right cornua of the bicornuate uterus, with an anterior placenta and normal cardiac activity, absent subchorionic hemorrhage. The left uterine cavity was noted to be empty. This successful pregnancy outcome underscores the efficacy of surgical intervention in correcting Müllerian duct anomalies, enabling fertile outcomes in patients with complex reproductive tract anatomy.

4. Discussion

OHVIRA (Obstructed Hemivagina and Ipsilateral Renal Anomaly) syndrome, presents a diagnostic challenge. Our case illustrates this complexity with a bicornuate uterus, differing from the more commonly reported didelphys uterus. Radiological imaging plays a crucial role in identifying this anomaly, with ultrasound (USG) and Magnetic Resonance Imaging (MRI) being essential diagnostic tools. While USG identified two uterine cavities, MRI provided detailed anatomical information, revealing a bicornuate uterus with two separate uterine bodies and cervices, obstructed left hemivagina with hematometrocolpos, and absent left kidney.[3]

The high spatial resolution and multiplanar capabilities of MRI enabled precise evaluation of reproductive tract anatomy. Post-treatment USG follow-up demonstrated successful conception, visualizing a single live intrauterine fetus in the right uterine cavity. This underscores the importance of radiological monitoring in assessing treatment outcomes and fetal well-being, highlighting the

complementary roles of USG and MRI in diagnosing and managing OHVIRA syndrome.[4]

5. Conclusion

Radiological imaging techniques, particularly MRI and USG, are critical for diagnosing OHVIRA syndrome and guiding effective surgical intervention. Our case highlights how early diagnosis and targeted management can significantly improve fertility outcomes, as demonstrated by the successful conception and pregnancy in this patient. The findings underscore the value of integrating advanced imaging modalities and surgical expertise in managing complex reproductive anomalies.

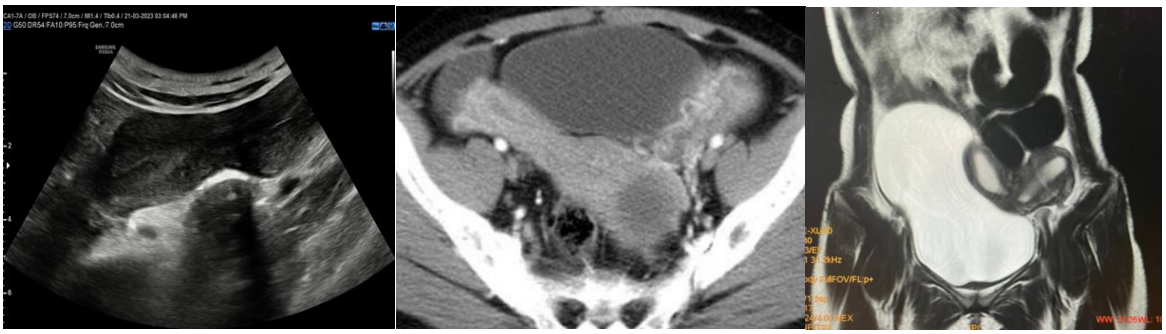
References

[1] KIMBLE RMN, KHOO SK, BAARTZ D, KIMBLE RM. The obstructed hemivagina, ipsilateral renal anomaly, uterus didelphys triad. Australian and New Zealand Journal of Obstetrics and Gynaecology. 2009 Oct;49(5):554–7.

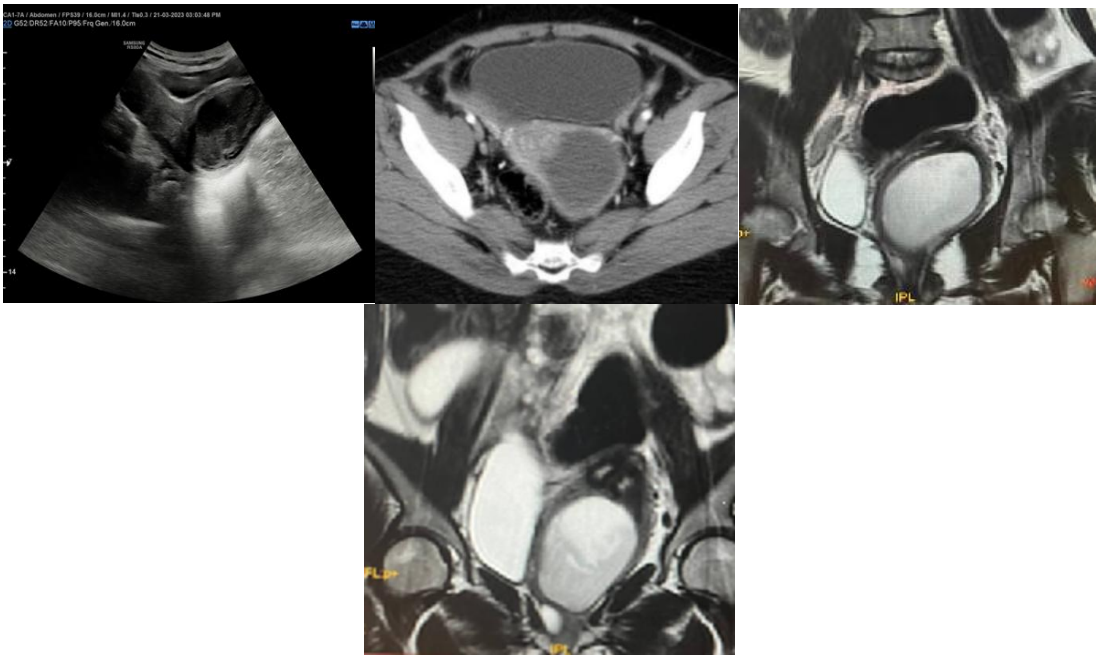
[2] Bajaj, SunilK, Ritu Misra, BrijB Thukral, and Rohini Gupta. 2012. "OHVIRA: Uterus Didelphys, Blind Hemivagina and Ipsilateral Renal Agenesis: Advantage MRI." *Journal of Human Reproductive Sciences* 5 (1): 67. https://doi.org/10.4103/0974-1208.97811.

[3] M. Alexandra Friedman, Aguilar L, Heyward Q, Wheeler C, Caldamone AA. Screening for Mullerian anomalies in patients with unilateral renal agenesis: Leveraging early detection to prevent complications. Journal of Pediatric Urology. 2018 Apr 1;14(2):144–9.

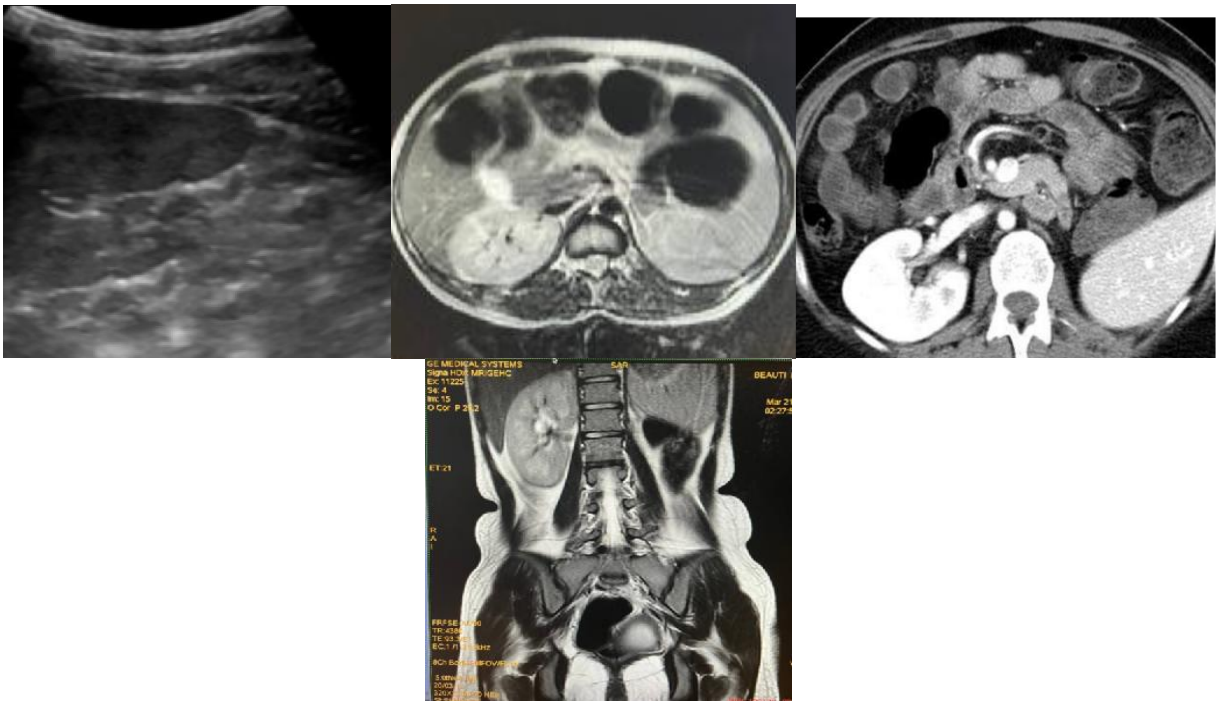
[4] Bermejo C, Martínez Ten P, Cantarero R, Diaz D, Pérez Pedregosa J, Barrón E, et al. Three-dimensional ultrasound in the diagnosis of Müllerian duct anomalies and concordance with magnetic resonance imaging. Ultrasound in Obstetrics and Gynecology. 2010 Jan 5;35(5):593–601.



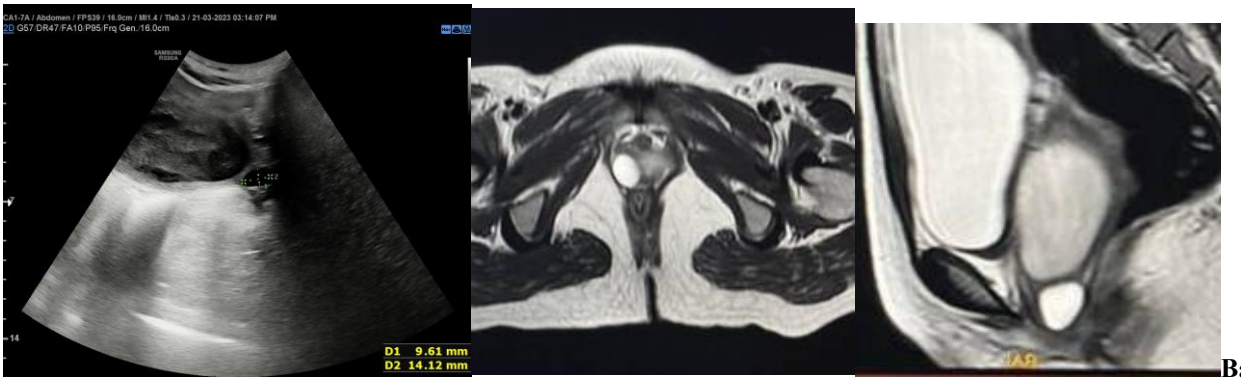
Two uterine cavities (Bicornuate Uterus- MRI)



Two separate vaginal cavities with hematocolpos in the left side. Compression and displacement of right vaginal cavity.

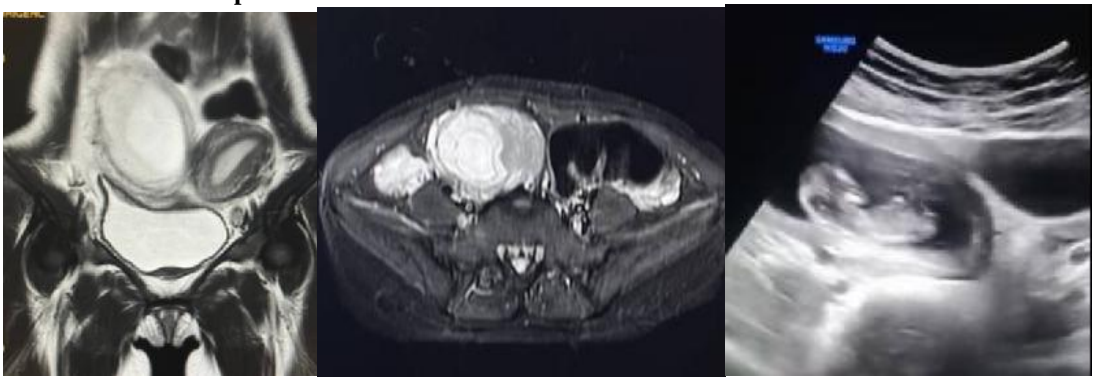


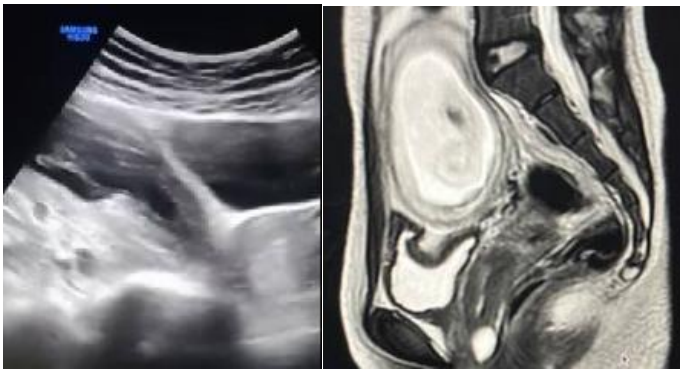
Absent left kidney



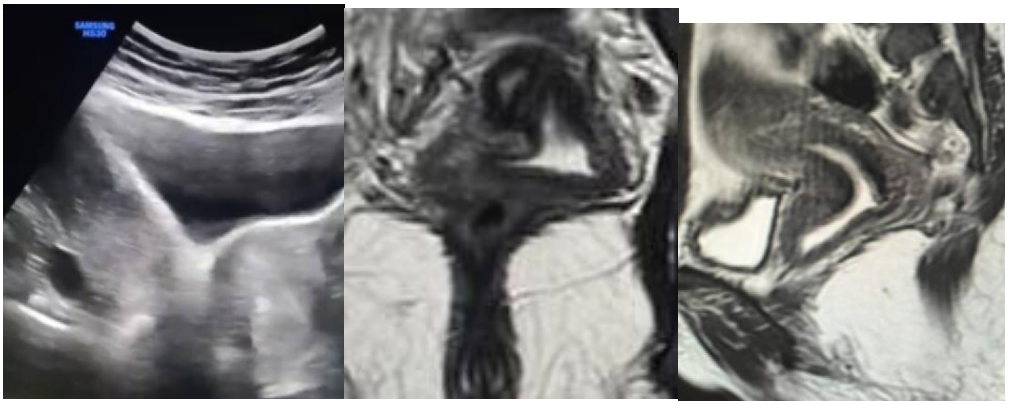
Bartholin's Cyst

Post surgical treatment follow up after four months





Bicornuate uterus with intrauterine gestation of 12.4 weeks in the right cornua, anterior placenta.



No evidence of hematocoplos noted in the left vaginal cavity