

DOI: <https://dx.doi.org/10.18203/2320-1770.ijrcog20264032>

Case Report

OHVIRA syndrome: a rare mullerian anomaly with diagnostic and management challenges: a case report and review of literature

Rao Preethi Venkatachala*, Shashikala Karanth

Department of Obstetrics and Gynaecology, St Johns Medical College and Hospital, Bengaluru, Karnataka, India

Received: 10 September 2026

Accepted: 08 October 2026

*Correspondence:

Dr Rao Preethi Venkatachala,
E-mail: preethibhat77@gmail.com

Copyright: © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ABSTRACT

Herlyn–Werner–Wunderlich syndrome, also known as OHVIRA syndrome is a rare complex congenital developmental anomaly characterized by the triad of uterine didelphys, obstructed hemivagina, causing hematometocolpos and ipsilateral renal anomaly. A 19-year-old nulligravida presented with foul-smelling vaginal discharge for 11 months and cyclical lower abdominal pain for 2 months. She had regular menstrual cycles since menarche. Examination revealed a single cervix with an anterolateral vaginal bulge draining mucopurulent discharge. Imaging with ultrasonography and MRI confirmed uterine didelphys, hematocolpos, right renal agenesis and hemivaginal septum. The patient had undergone prior aspiration elsewhere. Diagnostic hysteroscopy confirmed the findings. Excision of the vaginal septum revealed the obstructed cervix and a vaginal mould was placed postoperatively. The patient remains asymptomatic on follow-up and has conceived. High index of suspicion during evaluation of obstructive reproductive tract anomalies, early diagnosis through imaging modalities, multidisciplinary approach, timely surgical management and follow up yield good reproductive outcomes and quality of life.

Keywords: OHVIRA syndrome, Symptoms, Diagnosis, Management, Pregnancy

INTRODUCTION

Herlyn–Werner–Wunderlich syndrome, also known as OHVIRA syndrome, is a rare complex congenital developmental anomaly of the urogenital tract characterized by the triad of uterine didelphys, obstructed hemivagina causing hematometocolpos and ipsilateral renal anomaly. With a prevalence of 1:20000 in the general population, it accounts for 2-3% of all Mullerian anomalies.¹

Delayed or atypical presentations lead to diagnostic challenges and complications like endometriosis, infections or infertility. Here we present a case of OHVIRA syndrome with unusual presentation, delayed diagnosis and a good outcome

CASE REPORT

A 19-year-old nulligravida with a married life of 11 months and 3rd degree consanguinity, presented with foul smelling vaginal discharge in the past 11 months and cyclical lower abdominal pain since 2 months with no coital dysfunction. She had attained menarche at 13 years of age with regular menstrual cycles and spasmodic dysmenorrhoea.

Speculum examination revealed a single vagina and cervix on the left side and an anterolateral bulge draining mucopurulent discharge on the right side of vagina. On bimanual examination, uterus was anteverted, bulky, broad at the fundus with a depression at the middle – right horn connecting to a boggy non tender mass; the left horn connecting to the cervix.

Ultrasound abdomen-pelvis revealed uterine didelphys, hematocolpos, normal ovaries and right renal agenesis with compensatory hypertrophy of left kidney (Figure 1). MRI revealed uterine didelphys with hematocolpos of 6.3×3.2×4.1 cm communicating with right horn but not the left horn (Figure 2). Hemi vaginal septum and left hydrosalpinx were also noted along with the absence of right kidney (Figure 3).

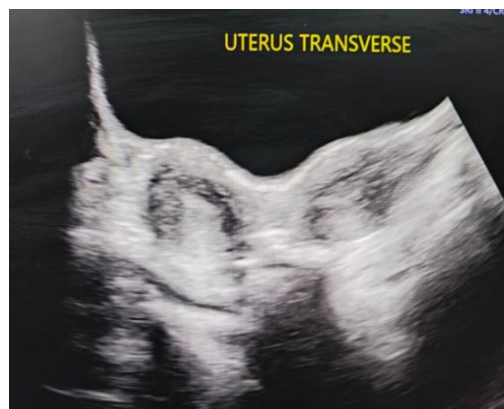


Figure 1: USG showing uterine didelphys.

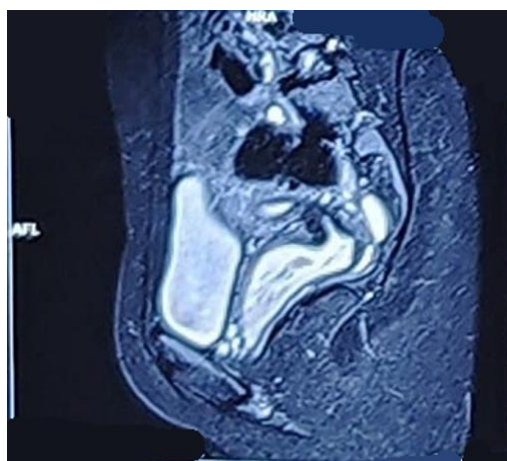


Figure 2: MRI showing hematocolpos.



Figure 3: MRI showing right renal agenesis.

Aspiration of hematocolpos under ultrasound guidance had been attempted at a local clinic and then referred to us for further management. A diagnostic hysteroscopy confirmed the diagnosis of OHVIRA syndrome – single cervix on the left side and a transverse vaginal septum on the right side (hemivagina) on vaginoscopy; regular endometrial cavity through the left cervix on hysteroscopy; uterus didelphys on laparoscopy (Figure 4). Excision of the vaginal septum revealed the right cervix (Figure 5). Surgicel was placed and vaginal mould inserted to maintain patency and to prevent adhesion formation (Figure 6). Patient is under follow up and doing well. She has conceived with a dating scan confirming the pregnancy.

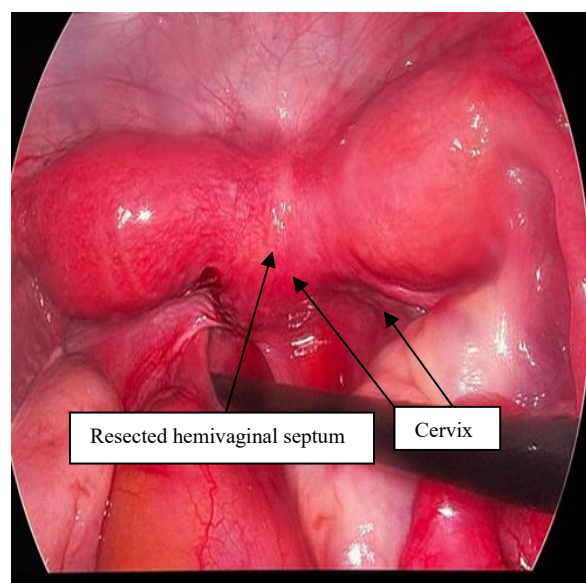


Figure 4: Laparoscopic view.



Figure 5: Postoperative picture.



Figure 6: Per Speculum - postoperative 2 weeks.

DISCUSSION

OHVIRA syndromes are rare and often under reported due to challenges in diagnosis as they present with diverse and non-specific symptoms. Delay in diagnosis is due to its varied presentation from acute abdomen due to hematocolpos at puberty to regular menstruation with slow progression of hematocolpos at a later age.² The most common clinical presentation is that of pelvic pain shortly after menarche in association with a vaginal or pelvic mass and normal menstrual periods. Rarely, it presents as abnormal vaginal discharge, infertility, acute abdominal pain, vomiting, fever and endometriosis.

A review of previously reported cases (Table 1) shows that most patients present during adolescence with cyclical abdominal pain and pelvic mass with ipsilateral renal agenesis.¹⁻⁵ In contrast, our patient presented atypically with chronic vaginal discharge and delayed diagnosis following prior intervention.

Table 1: Summary of reported cases of OHVIRA syndrome.

Author (Year)	Age (years)	Presentation	Uterine Anomaly	Vaginal Obstruction	Renal Anomaly	Management	Outcome
Joel GD et al, 2024	15	Cyclical pain, abdominal mass	Uterine didelphys	Obstructed hemivagina	Ipsilateral renal agenesis	Septal excision	Symptom relief
Elgohary MA et al, 2023	13	Acute abdominal pain, hematocolpos	Uterine didelphys	Obstructed hemivagina	Renal agenesis	Surgical drainage + septal resection	Good recovery
Rudra S et al, 2017	17	Dysmenorrhea, pelvic mass	Uterine didelphys	Hemivaginal septum	Renal agenesis	Septal excision	Improved symptoms
Akay E et al, 2025	19	Delayed diagnosis, pelvic pain	Uterine didelphys	Obstructed hemivagina	Renal agenesis	Multidisciplinary surgery	Good outcome
Agarwal N et al, 2021	16	Dysmenorrhea, mass	Uterine didelphys	Hemivaginal obstruction	Renal agenesis	Septal excision	Symptom resolution

OHVIRA syndrome is classified under class III as per the ASRM classification and under class U3b C2 V2 as per the ESHRE classification. At times, obstructed hemivagina and ipsilateral renal agenesis may be associated with a bicornuate or septate uterus (class U2b as per ESHRE/class IVa or Va as per ASRM); constituting OHVIRA variants. Other malformations, such as multicystic dysplastic kidney and renal duplication, have also been reported.²

The management of a single kidney should not be overlooked. About 15% of patients have accompanying contralateral urologic anomalies, with vesicoureteral reflux being the most common, raising concern for the possible damage to the solitary kidney.

USG and MRI can clinch the diagnosis.³ Ultrasound is a noninvasive, widely available imaging modality that helps in the accurate diagnosis of this condition. However, the vaginal septum is difficult to visualize on ultrasound and is best shown on MRI. MRI is more sensitive in detecting uterine contour, shape of the intrauterine cavity and character of septum compared to other imaging modalities, but it is less adequate in diagnosing endometriosis, pelvic inflammation and adhesions. Hence laparoscopy has been suggested as the gold standard of diagnosis.

Management varies from simple excision of vaginal septum to hysterectomy based on whether the vaginal obstruction is incomplete or complete with cervical atresia.¹⁻⁴ Surgical resection of the vaginal septum is done

conventionally under direct vision or using cautery or laser. This conventional approach may cause hymenal rupture and sometimes vaginal tears in adolescents, which leads to postoperative pain and adhesion formation. To overcome these issues, minimally invasive “vaginoscopic no-touch technique” with excellent visualization has been introduced for a longitudinal vaginal septum resection.⁶ Using a hysteroscopic L-hook electrode, the vaginal septum is incised followed by placement of a 14/16 Fr Foley's catheter, inflated with 50–80 ml saline in the previously obstructed hemivagina.⁵ In young girls, hysteroscopic excision of the septum under transabdominal ultrasound guidance may be performed to preserve hymenal integrity. Because of the associated risk of ascending infection, simple incision and drainage of the obstructed hemivagina is not recommended.

Medical management with continuous oral contraceptive pills or medroxyprogesterone can be advised to suppress menstruation until definitive surgery is planned. Postoperatively, antibiotics, analgesics and progesterone tablets for menstrual suppression are prescribed. Those who are at high risk for stricture or adhesion formation need bed rest, vaginal moulds, indwelling catheter and deep vein thrombosis prophylaxis. Psychological counselling with respect to the diagnosis, classification of anomaly, consequences, management options and timing of the surgery is important and should not be underestimated because of the association of this syndrome with the sexual, reproductive and social stigma. Multidisciplinary approach involving a gynaecologist, radiologist and urologist as well as preoperative counselling of patient are essential tenets of care.

CONCLUSION

High index of suspicion during evaluation of obstructive reproductive tract anomalies, early diagnosis through imaging modalities, multidisciplinary approach, timely

surgical management and follow up yield good reproductive outcomes and quality of life.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

REFERENCES

1. Joel GD, Basetti B, Mallula BV. A rare case report of urogenital anomaly in a teenage girl: Herlyn-Werner-Wunderlich syndrome/OHVIRA syndrome (obstructed hemivagina and ipsilateral renal anomaly). *Egypt J Radiol Nucl Med*. 2024;55:177.
2. Elgohary MA, Naik R, Elkafafi M, Hamed H, Ali Y. Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome: a case report. *J Pediatr Surg Case Rep*. 2023;95:102662.
3. Rudra S, Dahiya N. OHVIRA syndrome: a rare variant of Mullerian duct anomaly. *Int J Reprod Contracept Obstet Gynecol*. 2017;6(1):326-8.
4. Akay E, Uzun Gül AD. OHVIRA syndrome: a case report treated with delayed diagnosis and multidisciplinary approaches. *Taiwan J Obstet Gynecol*. 2025;64(4):693-9.
5. Sharma R, Mishra P, Seth S, Agarwal N. OHVIRA syndrome—diagnostic dilemmas and review of literature. *J South Asian Feder Obst Gynae*. 2020;12(6):421-6.
6. Cheng C, Subedi J, Zhang A, Johnson G, Zhao X, Xu D, et al. Vaginoscopic incision of oblique vaginal septum in adolescents with OHVIRA syndrome. *Sci Rep*. 2019;9:20042.

Cite this article as: Venkatachala RP, Karanth S. OHVIRA syndrome: a rare mullerian anomaly with diagnostic and management challenges: a case report and review of literature. *Int J Reprod Contracept Obstet Gynecol* 2026;15:4663-6.