

Original Research

Received 25 May 2026

Revised 26 June 2026

Accepted 21 July 2026

Natural Resources for Human
Health 2026, Vol. 6 (10s): 1062–1066

KEYWORDS:

Herlyn-Werner-Wunderlich
syndrome; OHVIRA syndrome;
uterine leiomyoma; renal agenesis;
hysterectomy; Mullerian duct
anomalies; magnetic resonance
imaging.

Recurrent Uterine Leiomyomas in Herlyn-Werner-Wunderlich Syndrome (OHVIRA): A Saudi Arabian Case Report and Review of the Literature

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ABSTRACT: Herlyn-Werner-Wunderlich (HWW) syndrome, also known as obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome, is a rare congenital Mullerian anomaly characterized by uterine didelphys, an obstructed hemivagina, and an ipsilateral renal anomaly. It is a rare condition, with some published series estimating an incidence below 1% (approximately 0.1-0.8%), although broader estimates of 0.1-3.8% have also been reported [11]. Surgical treatment during adolescence is directed at relieving vaginal obstruction while preserving reproductive function; however, long-term gynecologic follow-up remains important. We report a 45-year-old nulliparous woman with known HWW/OHVIRA syndrome and right renal agenesis who had previously undergone surgery for the obstructed hemivagina and associated right-sided Mullerian abnormalities. She later developed progressive abnormal uterine bleeding, pelvic pressure, and urinary frequency secondary to recurrent uterine leiomyomas. Pelvic MRI demonstrated multiple intramural and subserosal fibroids, with a dominant lesion measuring 7.2 x 7.5 x 6.8 cm and displacing the endometrial cavity. Her history included two prior myomectomies and seven unsuccessful intrauterine insemination (IUI) attempts. After counseling regarding conservative and definitive treatment options, she elected definitive surgical management. This case highlights the potential for significant adult gynecologic pathology in patients with previously treated HWW/OHVIRA syndrome and emphasizes the role of MRI in preoperative assessment of complex Mullerian anatomy.

INTRODUCTION

Herlyn-Werner-Wunderlich (HWW) syndrome, also termed OHVIRA syndrome, is characterized by uterine didelphys, an obstructed hemivagina, and an ipsilateral renal anomaly [1-3]. **The syndrome is rare; some studies have estimated its incidence at approximately 0.1-0.8%, i.e., less than 1% [11], while other reviews report a wider range of 0.1-3.8%.** Initial surgical management commonly consists of resection of the obstructing vaginal septum with drainage of hematocolpos, thereby relieving symptoms and helping preserve reproductive function [1,4]. Despite successful correction of the obstructive anomaly, patients remain susceptible to gynecologic disorders later in life. Uterine leiomyomas occurring in the setting of Mullerian anomalies are uncommon in the published literature [5-9]. Management of recurrent symptomatic leiomyomas in patients with altered Mullerian anatomy requires careful consideration of symptoms, previous operations, reproductive goals, and the technical challenges associated with distorted pelvic anatomy. We report recurrent uterine leiomyomas in an adult patient with previously treated HWW/OHVIRA syndrome and review the relevant literature.

Case Presentation

A 45-year-old married nulliparous woman with a known history of HWW/OHVIRA syndrome and right renal agenesis presented to the gynecology outpatient clinic with one year of progressive abnormal uterine bleeding, pelvic pressure, and increased urinary frequency. Her bleeding had been sufficiently severe to require blood transfusion on two occasions. She had undergone previous surgical treatment for the obstructed hemivagina and associated Mullerian abnormalities, including posterior vaginal wall surgery and subsequent resection of right-sided rudimentary Mullerian structures and a right vaginal wall cystic lesion. She also had a history of two myomectomies and seven unsuccessful IUI attempts.

Her menstrual cycles were regular, although menstrual flow had progressively increased, with frequent pad changes. She reported no current medication use, hormonal contraception, or intrauterine contraceptive device use. Her last menstrual period was 15 August 2026. Abdominal examination demonstrated a palpable pelvic mass below the umbilicus. On gynecologic examination, the cervical os was closed and there was no active bleeding. Cervical stenosis made initial cervical and endometrial sampling technically difficult.

Transabdominal pelvic ultrasonography performed on 30 October 2025 demonstrated a bulky uterus with a well-defined heterogeneous posterior myometrial lesion measuring approximately 7 x 6 cm and showing a swirling echotexture. The endometrium could not be clearly distinguished from the lesion. A left adnexal cyst measuring approximately 3.7 x 3 cm was also described. Renal ultrasonography confirmed absence of the right kidney.

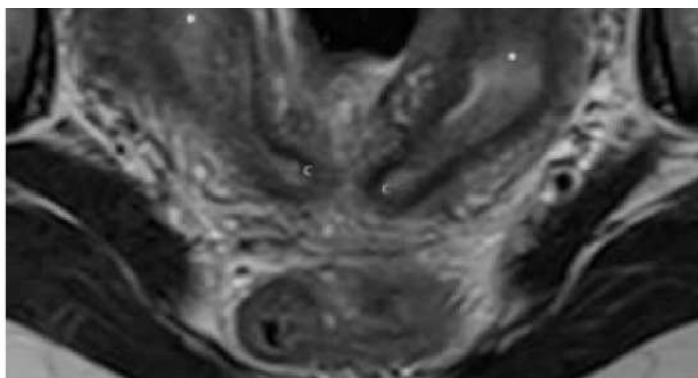


Figure 1. Axial T2-weighted pelvic MRI demonstrating uterine didelphys, with two separate uterine horns and endometrial cavities, consistent with the Mullerian anomaly associated with OHVIRA syndrome.

Contrast-enhanced pelvic MRI performed on 27 July 2026 demonstrated multiple intramural and subserosal uterine leiomyomas. The dominant intramural mass measured 7.2 x 7.5 x 6.8 cm and displaced the endometrial cavity. The imaging findings were consistent with abnormal uterine bleeding secondary to multiple fibroids and were important for defining the altered pelvic anatomy before definitive surgery.

Figure 1. Preoperative axial T2-weighted pelvic MRI demonstrating uterine didelphys, with two separate uterine horns and endometrial cavities, consistent with the Mullerian anomaly associated with OHVIRA syndrome.

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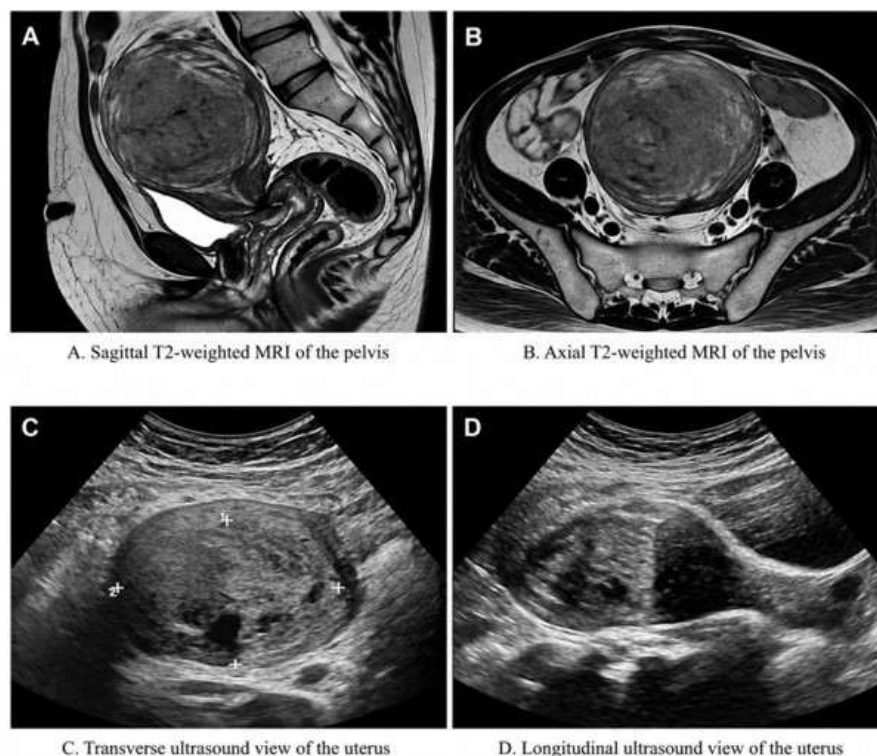


Figure 2. Preoperative pelvic imaging before hysterectomy. (A) Sagittal T2-weighted MRI of the pelvis. (B) Axial T2-weighted MRI of the pelvis. (C) Transverse ultrasound view of the uterus. (D) Longitudinal ultrasound view of the uterus.

DISCUSSION

OHVIRA syndrome typically presents after menarche with progressive dysmenorrhea, pelvic pain, and a pelvic mass related to hematocolpos [2,3,6]. Resection of the obstructing vaginal septum is the treatment of choice and can prevent long-term complications such as pelvic adhesions and endometriosis [1,4,7]. Delayed or missed diagnosis may occur even in adulthood, and the association between a Mullerian anomaly and a solitary kidney should raise clinical suspicion for OHVIRA syndrome [7,8].

Development of secondary gynecologic pathology, including uterine leiomyomas, in patients with previously treated OHVIRA syndrome is less frequently reported. Leiomyomas are estrogen-responsive benign smooth-muscle tumors and may substantially worsen heavy menstrual bleeding and pressure symptoms when they occur in anatomically abnormal uteri [9,10].

Imaging is essential both for diagnosis of Mullerian anomalies and for assessment of subsequent pelvic pathology. Pelvic ultrasonography is an appropriate initial modality, whereas MRI provides superior delineation of complex Mullerian anatomy, the relationship of leiomyomas to the endometrial cavity, and surgical planes in patients with distorted anatomy [9,10]. In this patient, MRI demonstrated extensive recurrent leiomyomatous disease and provided important information for operative planning.

Management should be individualized according to age, symptom burden, prior operations, reproductive goals, and the anatomy of the remaining uterus. Uterine-preserving surgery may be appropriate in patients who desire future fertility, whereas definitive surgery may be considered in women with recurrent symptomatic fibroids after multiple previous procedures and no further reproductive plans. In patients with renal anomalies and previous pelvic surgery, careful identification and protection of the solitary ureter are particularly important.

CONCLUSION

This case demonstrates recurrent symptomatic uterine leiomyomas in a 45-year-old woman with previously treated HWW/OHVIRA syndrome and right renal agenesis. Adult patients with congenital Mullerian anomalies may develop common gynecologic disorders that become clinically and surgically complex because of altered anatomy and previous operations. MRI is valuable for accurate anatomic characterization and preoperative planning, while definitive management should be individualized according to symptoms, reproductive goals, and surgical history.

Authors' Contributions

Both authors participated in the clinical management of the patient, manuscript preparation, critical revision, and approval of the final manuscript.

Patient Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying radiological images. Documentation of consent should be retained and submitted according to the target journal's policy.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have influenced the work reported in this manuscript.

REFERENCES

- [1] Arakaki R, Yoshida K, Imaizumi J, Kaji T, Kato T, Iwasa T. Obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome: a case report. *Int J Surg Case Rep.* 2023;107:108368.
- [2] Bezabih NA, Mehammed AH, Gebresilassie MY, Regasa ND. A rare case of OHVIRA (obstructed hemivagina and ipsilateral renal anomaly) syndrome: case report. *Radiol Case Rep.* 2024;19(9):3667-3671.
- [3] Saifi I, Kashikar SV, Parihar P, Saifi AI, Ansari KK. MRI findings in a rare case of left-sided obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome in a 21-year-old female. *Cureus.* 2023;15(10):e46704.
- [4] Kerimkulova MK, Oliveira Souza Lima SR, Elamin AE, Hamid YH, Faran N, Khan I. Multidisciplinary management of obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome in an adolescent with complex congenital anomalies: a case report. *Cureus.* 2024;16(3):e56961.
- [5] Haghgoo A, Faegh A, Nasiri S, Akhbari F. Vaginoscopic resection of hemivagina in a 20-year-old virgin female with prior misdiagnosis of OHVIRA syndrome as a bicornuate uterus: a case report. *Clin Case Rep.* 2024;12(3):e8661.
- [6] Akay E, et al. OHVIRA syndrome: a case report treated with delayed diagnosis and multidisciplinary approaches. *Taiwan J Obstet Gynecol.* 2025;64(4):693-699.
- [7] Lim LM, Sivapatham L, Chong ASL, Wahab AVA. Late-onset OHVIRA syndrome in a 49-year-old woman with severe dysmenorrhea: a case report on vaginoscopic management.
- [8] Lecka-Ambroziak A, Skobejko-Wlodarska L, Ruta H. The need for earlier diagnosis of obstructed hemivagina and ipsilateral renal agenesis/anomaly (OHVIRA) syndrome in case of renal agenesis in girls: case report and review of the literature. *J Clin Med.* 2023;12(23):7284.
- [9] Tu W, Yano M, Schieda N, Krishna S, Chen L, Gottumukkala RV, Alencar R. Smooth muscle tumors of the uterus at MRI: focus on leiomyomas and FIGO classification. *RadioGraphics.* 2023;43(6):e220161.
- [10] Paek, J. J., Kim, J., Bae, S., Paek, K., & Lee, Y. (2026). Chive-fortified fermentation enhances gamma-aminobutyric acid production by *Levilactobacillus brevis* PL9014. *International Journal of Probiotics & Prebiotics*, 21(1), 1–6. <https://doi.org/10.37290/ijpp.v21i1.12>
- [11] Thakur, S., Anvesh, G., Soman, R. J., Soman, D., & Pokuri, V. K. (2026). Evaluation of *Bacillus clausii* probiotic formulations in the prevention of antibiotic-associated diarrhea in infants and children: A randomized, double-blind, placebo-controlled study. *International Journal of Probiotics & Prebiotics*, 21(1), 1–14. <https://doi.org/10.37290/ijpp.v21i1.23>

- [12] Reddy, C. P., Keziah, M. S., & Madempudi, R. S. (2026). Efficacy and safety of *Bacillus clausii* UBBC-07 in chronic kidney disease: A double-blind randomized placebo-controlled study. *International Journal of Probiotics & Prebiotics*, 21(1), 15–23. <https://doi.org/10.37290/ijpp.v21i1.27>
- [13] Raj, M., Kumar, V., Bansal, C., Sharma, A., Rawat, V., Madan, Z., Upadhyay, S. K., Singh, R., Yadav, D., & Singh, M. (2026). Sustainable green synthesis of zinc oxide nanoparticles from *Citrus sinensis* peel extract for enhanced antibacterial activity. *Annals of Biology*, 42(2).
- [14] Souza, K. F., Gonçalves, L. C. O., & Espindola, F. S. (2026). A broad overview of the multifaceted royal jelly and its applicability in health and performance. *Current Topics in Nutraceutical Research*, 24(1), 4–14. <https://doi.org/10.37290/ctnr.v24i1.8>
- [15] Ghosh, S., Sagar, L., Reddy, M. D., & Bochalaya, R. S. (2026). Influence of methods of establishment and nitrogen levels on yield, yield attributes and nitrogen uptake of dry seeded rainfed lowland rice. *Annals of Biology*, 42(2).
- [16] Ostojic, S. M., Prémusz, V., & Ács, P. (2026). Co-administration of creatine and vitamin D and telomere length in older men: An exploratory pilot trial. *Current Topics in Nutraceutical Research*, 24(1), 1–3. <https://doi.org/10.37290/ctnr.v24i1.28>
- [17] Singh, S., Bochalaya, R. S., & Singh, S. (2026). Strategies for enhancing carbon sequestration in plants and soil through agricultural activities. *Annals of Biology*, 42(2).
- [18] Husen, T. K., Faris, M. N., & Taher, M. A. (2025). Multi-pollutants water cascade analysis for industrial water minimization. *Current Topics in Nutraceutical Research*, 23(2), 9–17. <https://doi.org/10.37290/ctnr.v23i2.25>
- [19] Gupta, V., Gill, K., & Bhatia, S. (2026). A multi-level analysis of barriers in the Pradhan Mantri Bhartiya Janaushadhi Pariyojana. *Annals of Biology*, 42(1), 142–151.
- [20] Kassem, O., Tremblay, A., Ghaly, M. S. W., Oula, M.-L., Bronner, S., & Binda, S. (2025). A subjective evaluation of the effects of a probiotic formulation on skin quality in young adult females: Open-label proof-of-concept study. *International Journal of Probiotics & Prebiotics*, 20(1), 1–9
- [21] Valletta R, Corato V, Lombardo F, Avesani G, Negri G, Steinkasserer M, et al. Leiomyoma or sarcoma? MRI performance in the differential diagnosis of sonographically suspicious uterine masses. *Eur J Radiol*. 2024.
- [22] Santos XM, Dietrich JE. Diagnosis and treatment in a tertiary hospital of a series of complex genital malformations corresponding to double uterus with obstructed hemivagina and ipsilateral renal agenesis. [Published literature reports an estimated incidence of approximately 0.1-0.8%].