

Obstructed hemivagina with ipsilateral renal agenesis (OHVIRA) syndrome. A case report and review of the updated literature

Síndrome de hemivagina obstruida con agenesia renal ipsilateral (OHVIRA). Presentación de caso y revisión de la literatura actualizada

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Summary
The OHVIRA syndrome, previously known as Herlyn-Werner-Wunderlich syndrome, is a rare female urogenital malformation characterized by the coexistence of müllerian and renal anomalies, prominently featuring a didelphys uterus, obstructed hemivagina, and ipsilateral renal agenesis. Diagnosis is typically established after menarche, with the most common clinical symptoms including nonspecific pelvic pain associated with amenorrhea and palpable abdominopelvic mass secondary to hematocolpos. Frequent complications include chronic pelvic pain, infections, infertility, and endometriosis. In this context, the contribution of diagnostic imaging (ultrasound and magnetic resonance imaging) is essential for precise characterization of these findings. Furthermore, a comprehensive understanding of this syndrome, its multiple manifestations, and classification is crucial for implementing appropriate and timely treatment. We present the case of an 11-year-old female patient, accompanied by an updated review of the literature.

Resumen
El síndrome de OHVIRA, anteriormente conocido como síndrome de Herlyn-Werner-Wunderlich, es una malformación urogenital femenina rara, que se caracteriza por la coexistencia de anomalías müllerianas y renales, entre las que se destacan un útero didelfo, hemivagina obstruida y agenesia renal ipsilateral. El diagnóstico suele establecerse después de la menarquia. La sintomatología clínica más común consiste en dolor pélvico inespecífico asociado a amenorrea y masa abdominopélvica palpable secundaria a hematocolpos. Entre las complicaciones más frecuentes se encuentran dolor pélvico crónico, infecciones, infertilidad y endometriosis. En este escenario, la contribución de las imágenes diagnósticas (ecografía y resonancia magnética) son esenciales para una caracterización precisa de estos hallazgos. Asimismo, la comprensión integral de este síndrome, sus múltiples manifestaciones y clasificación son cruciales para implementar un tratamiento adecuado y oportuno. Se describe el caso de una paciente femenina de 11 años de edad acompañado de una revisión actualizada de la literatura.

Introduction
OHVIRA syndrome, an acronym for Obstructed Hemivagina with Ipsilateral Renal Anomaly, formerly known as Herlyn-Werner-Wunderlich syndrome—named after its first discoverers, Herlyn and Werner in 1971 and Wunderlich in 1976—is a female urogenital anomaly originating in the Müllerian ducts. It is estimated that this condition, although rare and poorly documented, affects between 0.1% and 3.8% of the general population (1, 2). It is characterized by uterine malformations, an obstructed hemivagina, and renal and/or ipsilateral urinary tract anomalies. Its main manifestations include a didelphic uterus, as the most common Müllerian malformation, associated with a hemivagina separated by a septum that can lead to hematocolpos in cases of complete obstruction, along with an ipsilateral renal anomaly; agenesis is prevalent. However, there are various presentation variants of this condition (2). We report a new case of this rare syndrome, supplemented by an updated review of the literature, emphasizing the importance of diagnostic imaging in the identification and timely management of this condition.

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Clinical case report

An 11-year-old female patient, with a history of right renal agenesis diagnosed in March 2021 via UroCT (Figure 1), who presents to the emergency department with progressive abdominal pain of two-week duration localized in the right iliac fossa and radiating to the ipsilateral lower limb, associated with a two-month delay in menstruation following menarche six months prior. She denies a history of sexual activity. Physical examination reveals stable vital signs, tenderness on palpation of the lower right quadrant, and signs of peritoneal irritation. The pelvic examination is limited; no vaginal or cervical evaluation is performed; external genitalia appear normal on inspection. A transabdominal pelvic ultrasound reveals a double uterus, or didelphic uterus, with hematometra in the right horn (Figure 2a), hematocolpos of approximately 122 cm^3, and a hemorrhagic cyst on the right ovary (Figure 2b). The workup is supplemented with magnetic resonance imaging (MRI), which confirms the Müllerian malformation, showing two hemuteruses with separate endometrial cavities, hematometra in the right hemuterus, and vaginal distension due to hematocolpos caused by an obstructed hemivagina, in addition to a hemorrhagic cyst on the right ovary (Figures 3–6). The pediatric surgery department performed drainage of the hematocolpos and hematometra, as well as a laparoscopic vaginoplasty in a single procedure. During genitoscopy and colposcopy, a double cervix was confirmed, with a septum dividing a right atresic hemivagina from a left permeable hemivagina; the septum was removed, leaving a single vaginal canal. The postoperative period proceeded without complications, and the patient's clinical course was favorable.

Discussion

Herlyn-Werner-Wunderlich syndrome was originally characterized by a didelphic uterus, an obstructed hemivagina with hematocolpos, and ipsilateral renal agenesis (1, 2). However, subsequent cases have revealed a variety of associated anomalies, leading to the introduction of the term OHVIRA, proposed by Smith and Laufer in 2007, to specifically refer to the obstruction of a hemivagina with some form of ipsilateral renal anomaly, thereby broadening the spectrum of manifestations (1, 3). The etiology of the syndrome is not fully understood, but it is hypothesized that it may arise from an anomaly in the fusion of the Müllerian ducts during fetal development, likely between the 8th and 12th weeks of gestation, when it also interferes with the development of the caudal portion of one of the Wolffian ducts (the primitive precursor of the ureteric bud), causing renal malformation or agenesis (3).

The prevalence of Müllerian duct anomalies is reported to be as high as 7% in the general population, and at least 30% of these cases are associated with some form of renal anomaly, with a higher incidence on the right side (4). The most common Müllerian anomaly in more than 80% of cases is a didelphic uterus, while a bicornuate or septate uterus is observed in a small percentage of cases. Renal agenesis is the most common abnormality, present in 85% of cases, followed by anomalies such as ectopic kidney, horseshoe kidney, and duplex kidney. Cases with ectopic ureteral insertion into the hemivagina have been documented (1). It is important to note that nearly 50% of the renal anomalies detected during the prenatal period are multicystic renal dysplasia, which in most cases regress to subsequently become renal agenesis (5).

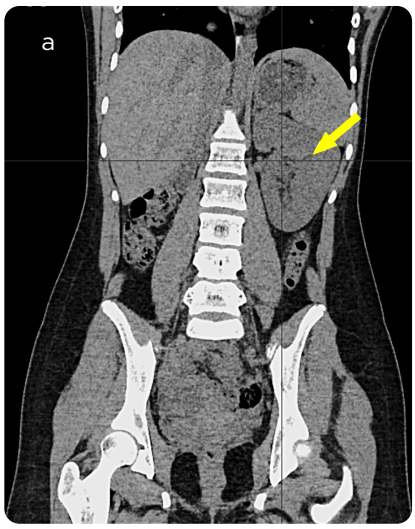
In 2015, Zhu et al. proposed a classification of OHVIRA/Herlyn-Werner-Wunderlich syndrome based on partial or complete obstruction of the hemivagina (Figure 7). Class I is characterized by a completely obstructed hemivagina and is subdivided into two subtypes: subtype 1.1, which presents a blind hemivagina without connection to the duplicated uterus, and subtype 1.2, which shows cervicovaginal atresia without uterine union, with obstruction or underdevelopment of the cervix and vagina. This group of patients, who typically begin experiencing symptoms around the age of 12, are more likely to develop complications such as hematometra, hematocolpos, hemoperitoneum, and endometriosis (6).

Class II is characterized by incomplete obstruction of the hemivagina and is further divided into two subtypes: subtype 2.1, which involves partial resorption of the vaginal septum and a small opening between the hemivaginas; and subtype 2.2, which involves complete obstruction of one hemivagina with a connection between the two cervixes. Symptoms in this group tend to appear several years after menarche, with purulent or bloody discharge and an increased risk of ascending genitourinary infections, such as pelvic inflammatory disease (PID) (6). This classification does not apply to cases of bicornuate or septate uterus and does not consider the type of renal anomaly. However, it is essential for planning surgical or medical treatment, as well as for determining the prognosis and potential complications.

The main imaging modalities used for diagnosis are ultrasound and magnetic resonance imaging (MRI). Ultrasound is a convenient and noninvasive option and is the first-line method for detecting Müllerian and renal malformations. However, soft-tissue resolution is limited, and diagnostic accuracy may depend on the operator. A pelvic ultrasound examination can reveal a double uterus (didelfic or bicornuate, among others) and detect its main complications, such as hematocolpos or hematometrocolpos, depending on the degree of distension; however, it has limitations in adequately characterizing the obstructed vaginal septum and the level of obstruction; Similarly, with an abdominal approach, the presence or absence of renal agenesis (1, 7).

MRI provides high-resolution, multiplanar, and multisecession soft-tissue imaging, which facilitates detailed visualization of the morphology of the uterus, cervix, and vagina. It also allows for accurate assessment of anomalies such as cervical agenesis or aplasia, the level of obstruction of the vaginal septum, as well as communication between two cervixes or hemivaginas. Images in T1WI and T2WI sequences allow for the determination of the location and volume, as well as the composition and stage of a contained hematoma (hematometra, hematocolpos, hematometrocolpos). DWI sequences and diffusion coefficient maps are highly sensitive and specific methods for diagnosing complications related to retrograde flow and resulting endometriosis, as well as infectious processes such as pyosalpinx, pyohyematocolpos, pelvic peritonitis, abscesses, and pelvic adhesions, providing a crucial reference for their classification and the development of appropriate surgical strategies (8).

Vaginoplasty, which involves resection of the septum and drainage of the hematocolpos, is the primary treatment for relieving the obstruction. In more complex cases, a two-stage vaginoplasty may be performed (1,3). Additionally, other procedures may be considered, such as ureteronephrectomy, salpingectomy, or cervicoplasty, which should be tailored according to the classification of each case and any additional complications (1, 4).



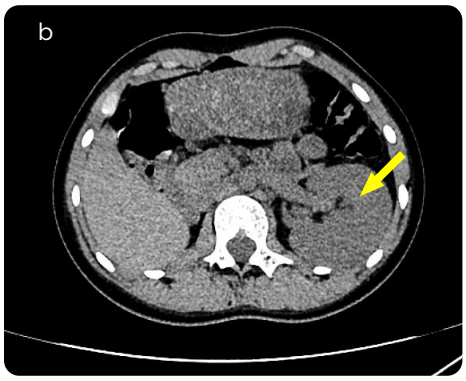
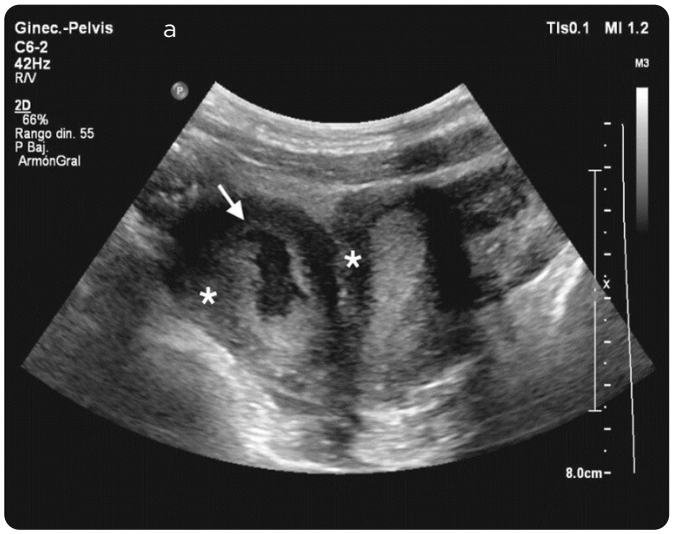


Figure 1. Uro-CT. (a) Coronal and (b) axial sections in soft-tissue window. Empty right renal fossa due to congenital agenesis. Left kidney slightly enlarged due to compensatory hypertrophy (yellow arrows).



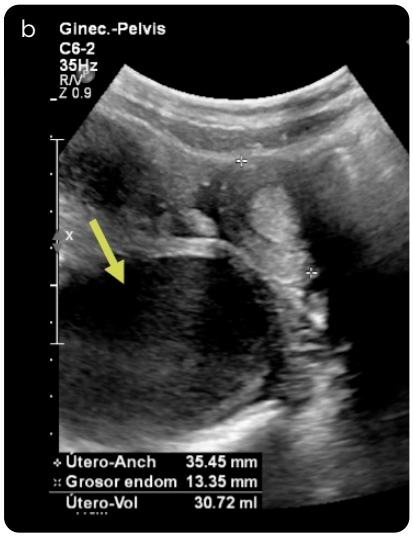


Figure 2. Longitudinal (a) and transverse (b) sections of transabdominal pelvic ultrasound in B-mode.
a) Two uterine bodies (asterisks) are observed, both with endometrial thickening. The right endometrial cavity is distended with heterogeneous, bloody fluid (white arrow). b) A unilocular cyst with internal echoes is observed in the right adnexa, without a solid nodular component or Doppler flow, suggestive of a hemorrhagic cyst (yellow arrow).

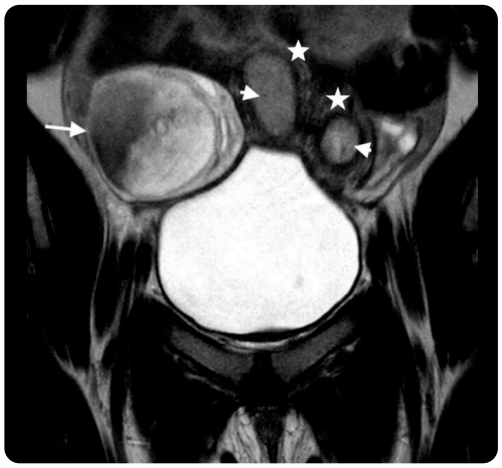


Figure 3. Pelvic MRI. T2-weighted FAT SAT sequence, coronal section. In the right adnexal region, an oval, heterogeneous lesion is observed, predominantly high signal intensity with a poorly defined low-signal component adjacent to the right uterine body (white arrow), consistent with a hemorrhagic cyst. Both uterine bodies (stars) show endometrial thickening (white arrowheads).



Figure 4. Pelvic MRI. T1-weighted + C FS sequence, coronal section. Both uterine bodies (white arrows) show homogeneous enhancement with contrast. The right uterine cavity is distended by high-signal fluid, suggestive of hematometra. Right adnexal hemorrhagic cyst without pathological enhancement (curved arrow). The bladder, marked with a black star.

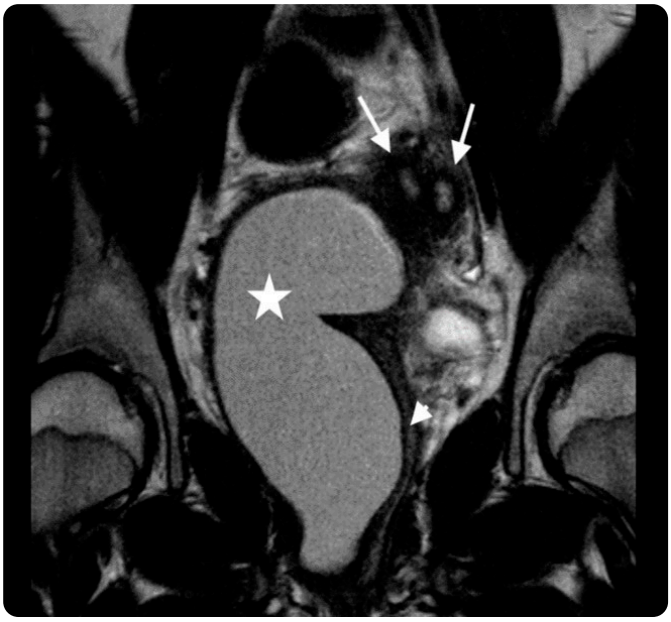


Figure 5. Pelvic MRI. T2-weighted sequence, coronal section. Two cervical canals (white arrows); at the caudal level (star), the right hemivagina is occupied by high-signal material that exhibits the same behavior on the T1-weighted sequence, confirming a hematoma. The collapsed left hemivagina, separated by a septum, is identified, indicating a fusion defect (arrowhead).



Figure 6. Pelvic MRI. Plain T1-weighted sequence, axial slice. Right ovarian cyst (white arrow). Occupation of the right hemivagina by high-signal material corresponding to hematocolpos (black star).

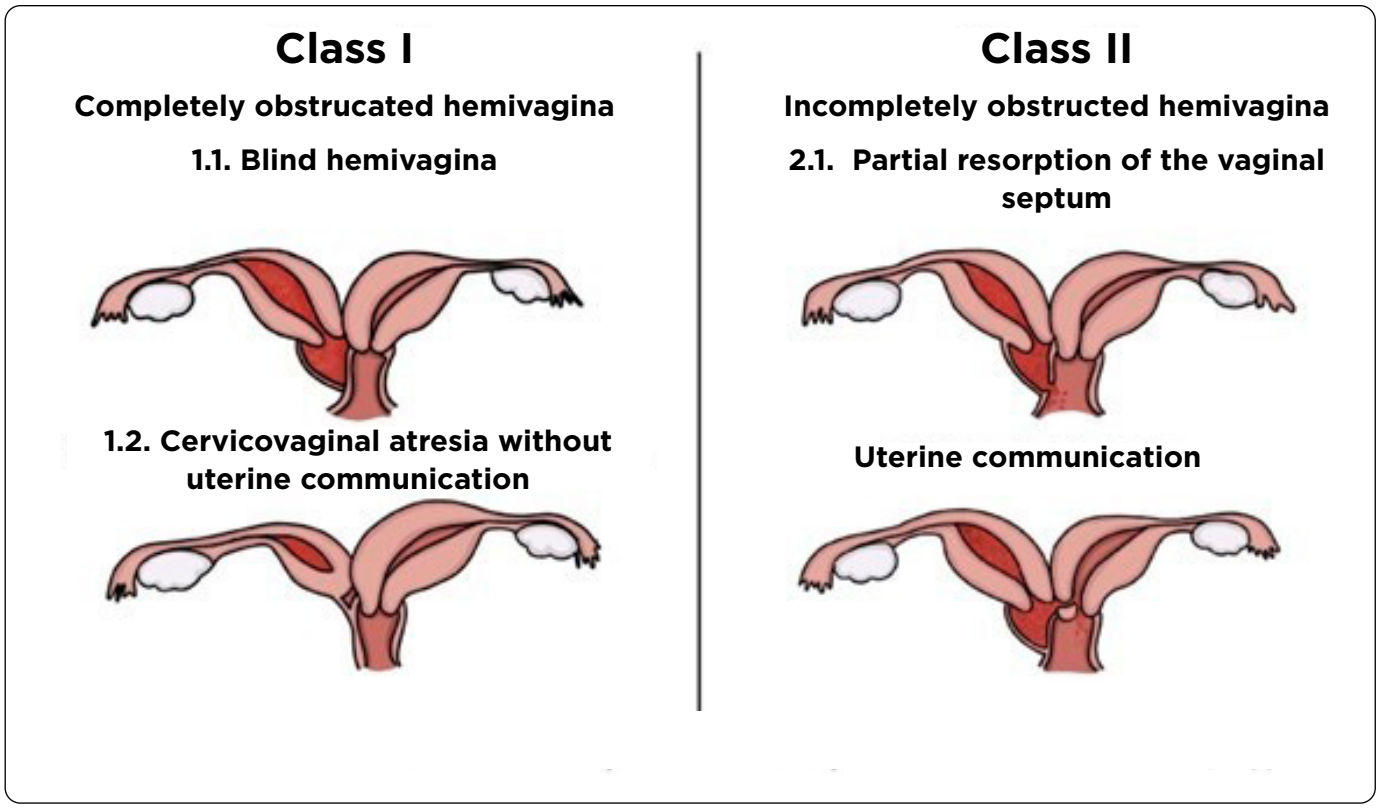


Figure 7. Classification of OHVIRA/HWW syndrome proposed by Zhu in 2015 (6).

In general, Müllerian anomalies alone do not prevent pregnancy, nor is there sufficient evidence of a link to infertility; however, they are associated with a higher incidence of pregnancy complications such as preterm birth, preterm rupture of membranes, or perinatal mortality (4, 5).

Recent studies have proposed new recommendations for implementing ultrasound screening for Müllerian anomalies in young women (prior to menarche) with a prenatal diagnosis of renal malformations, such as unilateral renal agenesis, multicystic renal dysplasia, or renal ectopia. Pelvic ultrasound, being a low-cost, highly accessible technique with good diagnostic performance, allows for the early detection of these anomalies, facilitating timely intervention and controlled clinical management, which could significantly reduce the risk of long-term complications (9, 10).

Conclusion

OHVIRA syndrome is a complex clinical condition with heterogeneous manifestations and diverse imaging findings, underscoring the need for a multidisciplinary approach to ensure optimal management. Imaging modalities, particularly pelvic ultrasound and MRI, are essential for accurate diagnosis, anatomical classification, and characterization of associated anomalies. MRI, given its superior tissue resolution and ability to delineate pelvic anatomy, constitutes the gold standard for preoperative evaluation and surgical planning.

The close embryological correlation between the renal and Müllerian systems requires a systematic bidirectional evaluation: when renal anomalies are diagnosed in the prenatal or neonatal period (such as renal agenesis or ectopia), concomitant Müllerian malformations must be ruled out; conversely, every patient with Müllerian anomalies should be evaluated by abdominal ultrasound to identify possible associated renal defects. Surgical treatment, with vaginoplasty as the primary procedure, must be tailored to each patient's specific anatomy and symptoms, with the goal of resolving the obstruction and preserving reproductive function. Early diagnosis, supported by appropriate imaging studies, combined with comprehensive and personalized care, is crucial for optimizing functional outcomes, preventing long-term complications, and improving the quality of life for those affected.

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