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(CASE REPORT)

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# Ohvira síndrome: Case Report

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## Abstract

**Background**: OHVIRA (Obstructed Hemivagina and Ipsilateral Renal Anomaly) syndrome is a rare congenital condition characterized by uterine malformations, such as didelphic uterus with blind hemivagina and ipsilateral renal agenesis. It usually presents in adolescence with primary dysmenorrhea, cyclic pelvic pain and hematocolpos or hematometra. Its incidence is low, estimated between 1 in 2,000 to 1 in 28,000 women, and the available literature on this syndrome is limited.

**Case report**: 15-year-old female patient with a history of right renal agenesis diagnosed in childhood, who consulted for intense dysmenorrhea and recurrent vaginal bleeding, requiring hospitalization. The imaging studies, ultrasound and pelvic MRI, showed a didelphic uterus with obstructed hemivagina, compatible with OHVIRA syndrome. During laparoscopy, endometriosis foci were also identified. Combined laparoscopic and hysteroscopic surgical management was performed to resolve the obstruction and resect endometriotic lesions. The patient evolved with regular menstrual cycles and without symptoms.

**Discussion**: OHVIRA syndrome is an underdiagnosed pathology, associated with complications such as endometriosis, infertility and chronic pelvic pain. Its timely diagnosis by 3D ultrasound and magnetic resonance imaging is essential to plan the appropriate treatment and prevent long-term sequelae.

**Conclusions**: In patients with menstrual abnormalities and renal agenesis, the presence of Müllerian malformations, such as OHVIRA syndrome, should be suspected in order to make an early diagnosis and treatment, avoid future complications and preserve fertility.

**Keywords:** OHVIRA syndrome; Endometriosis; Uterine malformations; Dysmenorrhea; Hemivagina; Renal agenesis; Pelvic pain; Laparoscopic surgery; Herlyn-Werner-Wunderlich syndrome; Adolescents; Infertility; Quality of life

## 1. Introduction

OVHIRA syndrome refers to a series of congenital malformations including alterations of uterine formation such as didelphic uterus, total or partial blind hemivagina and ipsilateral renal agenesis. Didelphic uterus with obstructed hemivagina was first described in 1922, but the association with renal agenesis was reported in 1971 and at that time it was called Herlyn- Werner-Wünderlich syndrome. It is now known by the acronym OHVIRA (Obstructed Hemivagina and Ipsilateral Renal Anomaly)1. The incidence of Müllerian malformations is 2 to 3%, and the least common is OHVIRA syndrome, with only publications of isolated cases or case series, representing less than 10% of all Müllerian malformations; with a variable rate ranging from 1 / 2,000 to 1 / 28.It is a pathology that manifests itself in adolescents

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after menarche with primary dysmenorrhea and/or cyclic pelvic pain associated or not with vaginal bleeding and pelvic tumor by hematocolpos collection (hematocolpos), a 3D ultrasound and magnetic resonance imaging are useful to confirm the diagnosis.

The association of these three components is secondary to an anomaly in the embryological development caused by the proximity of the structures derived from the mesonephric primordium (Wolf's duct) and the paramesonephric primordium (Müller's duct)(4,5) ,These malformations can be the final product of a deficit in the fusion and canalization of Müller's ducts; or a defect in the distal union of these to the urogenital sinus or simply an alteration in the formation of the vaginal plate. In a high percentage (30- 40%) of the cases genitourinary anomalies are usually present; due to the coexistence and simultaneous development of both systems (Mullerian and Wollfian) for a considerable period from very early stages of embryogenesis 6, embryos initially have these 2 pairs of genital ducts; Wolff and Müllerian, The latter are located outside the ipsilateral mesonephric duct, but in its caudal portion they cross it ventrally to merge in the midline with each other. Shortly after the end of the paramesonephric ducts has reached the urogenital sinus, 2 massive evaginations, called synovaginal bulbs, detach from the pelvic portion of the sinus and give rise to the lower 2 thirds of the vagina. The upper third, however, derives from the uterine duct, of paramesonephric origin. On the other hand, the ureteral bud, an evagination of the mesonephric duct close to its outlet into the cloaca (origin of the collecting system), will subsequently enter the metanephric mesoderm to induce the development of the excretory units. The mesonephric or Wolffian ducts that give rise to the kidneys are also inducing elements for the proper fusion of the mullerian ducts. Thus, on the side where the Wolffian ducts are absent, the Mullerian duct is laterally displaced in such a way that it cannot fuse with the contralateral duct, resulting in a didelphic uterus; on the other hand the contralateral Mullerian duct gives rise to a vagina, while the displaced component forms a blind cul-de- sac as an imperforate or obstructed hemivagina. The vaginal introitus is not affected because of its origin in the urogenital sinus. Renal agenesis is presumed to be a consequence of premature degeneration of the ureteral bud, since if the ureteral bud does not reach the metanephric tissue cap, the latter does not proliferate7,8,9.

It is definitely an anomaly with complications that can compromise the quality of life of the adolescents affected, with risk to their sexual and reproductive future; the presence of cases such as these reflects the importance of suspecting this pathology in cases of dysmenorrhea and menstrual cycle alterations in adolescent patients who have recently undergone menarche; procedures such as hysteroscopy in conjunction with ultrasonography and magnetic resonance imaging can provide accurate diagnoses and detect complications associated with persistent obstruction of the genital tract, in addition to serious associations such as endometriosis.10,11.

## 2. Clinical case description

We describe the case of a 15-year-old female patient with a history of menarche at age 12, right renal agenesis, recurrent urinary tract infection since age 4, with multiple antibiotic treatment, who consulted for 3 years of evolution characterized by disabling dysmenorrhea, vaginal bleeding of moderate amount leading to school absenteeism and multiple visits to the emergency department for pain management, which led to request complementary diagnostic imaging studies, In the transabdominal ultrasound of the pelvis, two uterine bodies were visualized, compatible with a didelphic uterus, and a contrasted abdomino-pelvic MRI was requested, which confirmed the presence of uterine malformation described in the ultrasound, The right hemivagina was partially distended with a large hematocolpos of 6 x 10 cm, and the left hemivagina was normal (Figure 1). Additionally, there was evidence of a complete thin longitudinal septum of 1-3 mm, with a transverse septum of similar thickness that gives it a bilobed appearance at the junction of the middle third , finally, it was confirmed the presence of a single left kidney of normal appearance, without other lesions (Figure 2). Physical examination showed mild pain on deep palpation in the hypogastrium, with no signs of peritoneal irritation at the time of examination; external genitalia of normal appearance. Given the persistence of the clinical picture and the imaging findings, hysteroscopy and diagnostic laparoscopy with therapeutic intent were ordered. The surgical protocol was performed in three stages; first diagnostic hysteroscopy was performed using Glycine 1.5% Glycine was used as distension medium, the right cervix was identified with normal exocervix and endocervix, uterine cavity displaced to the right, decreased in size, tubular, without being able to observe the ostium due to the presence of tortuosity, Secondly, surgical hysteroscopy was performed where the resection of the vaginal septum was started longitudinally along its entire length using monopolar energy by resectoscope with cutting loop tip, after draining the hematic content, the identification of a second left cervix with uterine cavity and normal ostium was achieved. The tissue of the vaginal septum was sent for anatomopathological study which was reported as fibro-leiomyomatous tissue. As a last surgical step, exploratory laparoscopy was performed, both uterine bodies were identified with normal tubes, right uterus of normal size and enlarged left uterus. Red vesicular lines suggestive of endometrial tissue were observed at the level of the serosa in the anterior face of the uterus and another one in the right lateral pelvic wall of approximately 2 cm, from which a sample was taken for histological study which reported chronic inflammation with atypical cells and fibrosis (Figure 4 and 5).

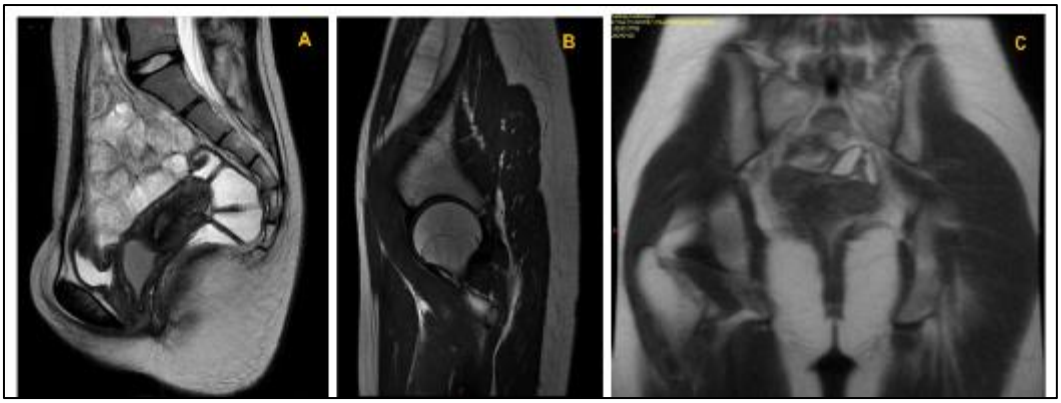


**Figure 1** A; Contrasted MRI mid-sagittal view, right uterine cavity is visualized distended and B; contrast allows visualization of right hematocolpos, right side uterine cavity and right hemivagina are distended with a fluid of mixed intensity that is probably a blood component suggestive of right hematocolpometra. C; Normal left hemivagina

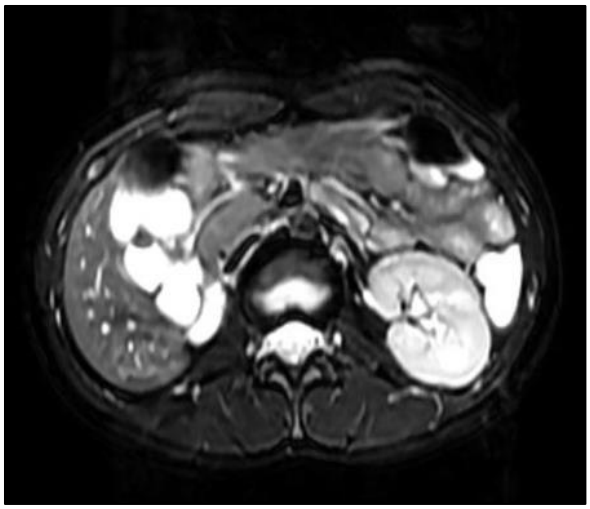

**Figure 2** Axial section of the abdomen, showing a single kidney on the left side

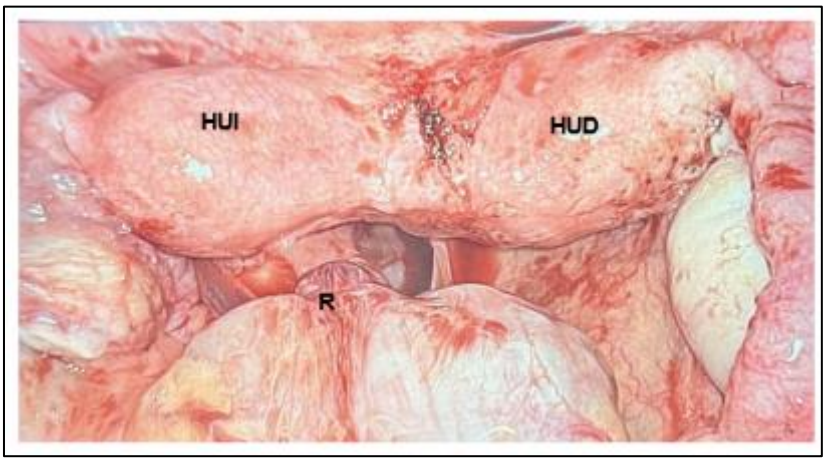


**Figure 3** Intraoperative image shows didelphic uterus and multiple foci of endometriosis; *right uterine hemibody (RUB), left uterine hemibody (LUB), rectum (R)*

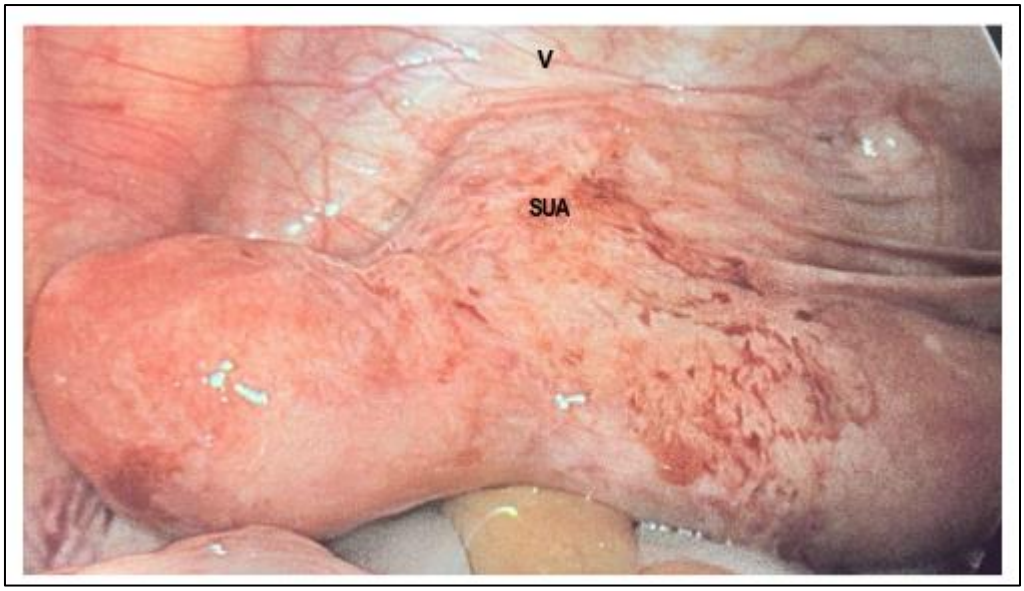


**Figure 4** Intraoperative image shows didelphic uterus and multiple foci of endometriosis; anterior uterine segment (AUS), bladder (V)

A follow-up after two months was performed with MRI, which reported a descended uterus, both hemiuterus in anteversoflexed position with symmetrical hemicavities. Clinical follow- up continued for 12 months, the patient presented menstrual cycles without dysmenorrhea and decreased bleeding, remaining currently asymptomatic.

## 3. Discussion

OHVIRA syndrome is an underdiagnosed entity, although congenital, it is usually identified during adolescence, generally between 12 and 16 years of age, after multiple admissions to hospital services, deterioration of the quality of life due to marked symptoms and even impacting fertility. Rarely cases have been reported in newborns or infants, manifested by mucocolpos secondary to a septated hemivagina12,13, identification of the uterine anomaly at early ages is difficult, due to the small size and tubular morphology of the uterus in infancy, This presents a clinical and diagnostic challenge14, as in the case of our patient the symptoms of the syndrome are not specific, which limited an initial diagnostic suspicion, dysmenorrhea represented a disabling symptom that considerably deteriorated her quality of life.

Transabdominal pelvic ultrasound, although it does not make a definitive diagnosis in most cases, is a complementary method of initial approach that should not be missing, this study presents a diagnostic accuracy for uterine malformations of between 90% and 92%, being also an accessible, fast and non-invasive method[1]5,16. Contrasted MRI, as a gold standard, presents a sensitivity and specificity close to 100%1[7](,) [18]is a tool that allows a more detailed characterization of the anatomical alterations that make up this syndrome, by means of which we were able to confirm the presence of a uterine septum dividing two complete cavities (didelphic uterus).

The surgical approach of our patient was performed in three stages, seeking a better characterization of the anatomical alterations and greater success in their surgical correction, where the main objective was to resect the vaginal septum and marsupialization of the blind vagina, to facilitate drainage of the hematic content and prevent recurrence through subsequent dilatations. This procedure can traditionally be performed in one or two surgical stages, as required by the case19,20,21. Recently, complementary techniques have been used with laparoscopy, cystoscopy and vaginoscopy, and even hysteroscopic resection of the septum, seeking to alleviate symptoms, preserve future fertility and prevent complications. The placement of temporary prosthetic devices - syringes, endotracheal tubes, tracheotomies or tracheobronchial stents - in the created vaginal canal has also been described, to avoid stenosis in the immediate postoperative period21,22.

Late diagnosis and treatment can trigger complications, since persistent obstruction favors retrograde bleeding into the abdominal cavity, leading to endometriosis in varying degrees of severity, formation of pelvic adhesions and ascending infections. This can translate into chronic pelvic pain, considerably affecting the quality of life of these patients.

## 4. Conclusion

This case leads us to think about the association of classic gynecological symptoms such as chronic pelvic pain and menstrual alterations, with anomalies such as unilateral renal agenesis, to the possible diagnosis of Müllerian malformations and with this we can later specify an OHVIRA syndrome. Magnetic Resonance Imaging not only gives us an accurate diagnosis but also provides us with a useful preoperative window to reduce complications. Finally, minimally invasive surgical management by hysteroscopy and laparoscopy should be provided in a timely manner and should be aimed at restoring functionality, so that the surgeon obtains precision and favors a faster recovery of the patient, in addition to preserving future fertility in our patients.

## Compliance with ethical standards

### *Disclosure of conflict of interest*

No conflict of interest to be disclosed.

### *Statement of informed consent*

Informed consent was obtained from all individual participants included in the study.

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