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CASE REPORT

# Herlyn-Werner-Wunderlich syndrome: a case report

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## Abstract

Herlyn-Werner-Wunderlich syndrome is a rare congenital disease of the Müllerian ducts, which consists of didelphys uterus, obstructed hemivagin and unilateral renal agenesis; in which nondiagnosis or early treatment can lead to secondary complications, such as pelvic inflammatory disease, tube-ovarian abscess and future infertility. This work consists of a case report of an 11-year-old child, G0P0, with abdominal pain in cramps of severe intensity and voiding difficulty. We describe the evaluation, confirmation of the diagnosis with imaging tests and the clinical and surgical management of the patient, as well as a literature review.

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## INTRODUCTION

Herlyn-Werner-Wunderlich Syndrome (SHWW) is characterized by the presence of hemivaginal septum, didelphic uterus and ipsilateral renal agenesis. Herlyn-Werner syndrome was described in 1971 as the presence of renal agenesis and ipsilateral blind hemivagina. In 1976, Wunderlich described the association of right renal aplasia with a bicornuate uterus and simple vagina in the presence of a hematocervix, resulting from the lack of communication between the right hemiuterus and the vagina. The combination of these two syndromes is called SHWW, which results from a defect in genito-gonadal embryogenesis, with failure of the mullerian structures[1-3].

SHWW is a rare variant of Müllerian anomalies and patients are usually asymptomatic until menarche. Unlike the most common congenital anomalies of the genital tract that lead to amenorrhea, such as imperforate hymen, vaginal atresia and/or transverse vaginal septum, SHWW usually has normal menstruation, associated with cyclic abdominal pain[4,5]. This is because only one uterine horn is obstructed. Thus, the patient regularly menstruates through the other horn[3,4].

SHWW is also known in the literature as obstructed hemivagina and ipsilateral renal anomaly syndrome (OHVIRA)[5], variants of Mullerian anomalies. These congenital anomalies occur in 0.1% to 6% of women[6-8].

The diagnosis can be delayed by the use of non-steroidal anti-inflammatory drugs (NSAIDs) and oral contraceptives for patients with dysmenorrhea, which can mask pain and reduce or eliminate menstruation[6,9,10]. When the diagnosis is delayed it can lead to infertility or obstetric complications. On physical examination, a painful suprapubic mass and a paravaginal cystic mass can be palpated. Imaging investigation can include Ultrasonography (USG), Computed Tomography (CT) and Nuclear Magnetic Resonance (MRI) of the abdomen and pelvis, with MRI being the gold standard[9,11,12].

The objective of this case report is to present a rare syndrome, with non-specific signs and symptoms, commonly confused with physiological conditions or other specific comorbidities of the woman's reproductive period. The existence of SHWW should be more widespread among pediatricians to facilitate the early diagnosis of the disease, thus avoiding future complications.

## CASE REPORT

GFSC, 11 years old, G0P0, referred to Mário Palmério Hospital Universitário (MPHU), Uberaba, MG, in her second menstrual cycle 18 days after menarche, without the use of medications and previous surgeries, due to severe abdominal pain with cramping, voiding difficulty and lower abdominal US suggestive of Herlyn-Werner-Wunderlich syndrome. On physical examination, on admission, she had an intact and patent hymen.

Imaging exams (USG and MRI) of the abdomen and pelvis at the beginning of hospitalization showed a vagina with mid-distal duplication with dilatation on the left, secondary to heterogeneous content, as shown in Figure 1A. In addition to a uterus with two independent horns, the one on the right being laterally flexed, with an approximate volume of 17 ml and a homogeneous, centered endometrial sign, measuring 4.0 mm in thickness, and the one on the left, with distended endocervical and endometrial cavities filled with blood content. (approximate volume of 33ml), as shown in Figure 1B. And a small amount of free liquid in the posterior cul-de-sac. In addition to a vicarious kidney on the right associated with probable renal agenesis on the left.

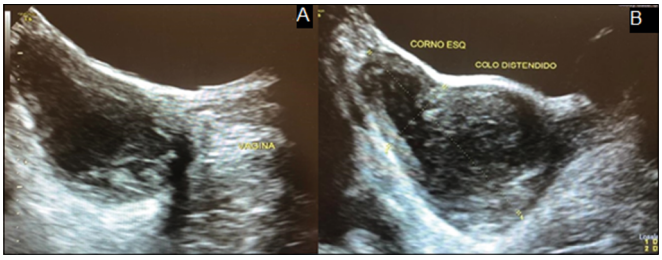



**Figure 1. A.** Ultrasonographic image of the vagina with mid-distal duplication with dilatation on the left; **B.** Ultrasonographic image of the left horn with increased volume and distended neck.

Due to the didelphic uterus with a moderate volume hematocolpo-hematometrium on the left, associated with a small amount of free fluid in the posterior cul-de-sac, a surgical approach was chosen for drainage, initially via the vaginal route. As the access route to the hematocolpos via the vaginal route was not identified, it was decided to convert the surgery to laparotomy. Moderate amount of chocolatey blood was identified in the abdominal cavity, right and left ovaries and right tube were identified without alterations, dilated left tube with presence of brownish bloody material inside, identified uterus with two horns, the left horn being dilated with increased size in relation to the right horn (about 2 times the volume of the right). Approximately 30 ml of thick, darkened/chocolate blood was drained, manual milking and aspiration were performed with a probe introduced through the horn incision, extending to the left cul-de-sac and left parametrial region.

After the procedure, the patient was discharged with daily desogestrel for follow-up at the gynecological endocrinology outpatient clinic of the MPHU. On an outpatient basis, it was decided to discontinue use for monitoring hematocolpos formation until the actual formation of a bulging of the left hemivagina and definition of the entry point for septoplasty and hematocolpos drainage.

After 147 days of the procedure, the patient returned to our service with a complaint of intense, colicky pain in the lower abdomen. The new MRI of the Pelvis showed a didelphys uterus with a large hematometrocolpus on the left, exerting an expansive effect on the vaginal vault and pelvic structures. Elongated image in blind background to the left

of the midline containing hemorrhagic material, which may correspond to hemivagina. Uterine component on the right and anterolaterally deviated ovaries , but without significant morphological changes.

After the 153rd day, the patient underwent hematocolpos drainage again. During the procedure, a softened area on the left side of the vaginal wall was visualized, a puncture of the area was performed with aspiration with exit of a large amount of clot, on average 500 ml. A marsupialization of the orifice is carried out.

The patient remains under follow-up at our service at the Gynecological Endocrinology outpatient clinic, regularly, for follow-up. The last USG performed showed an endometrial cavity with preserved thickness, as shown in Figure 2. The patient has remained asymptomatic since then, using Desogestrel 75 mcg daily.

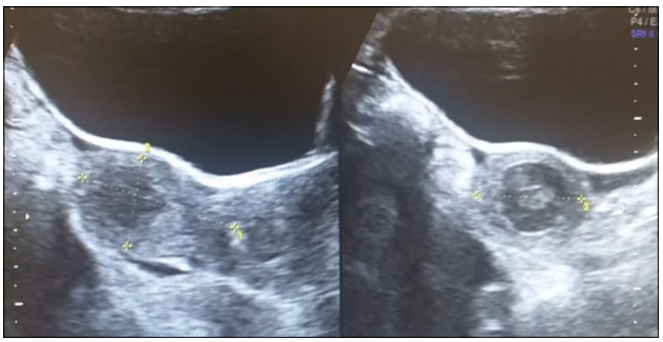


**Figure 2. A.** Ultrasonographic image of the vagina with mid-distal duplication with dilatation on the left; **B.** Ultrasonographic image of the left horn with increased volume and distended neck.

## DISCUSSION

The American Society for Reproductive Medicine classifies SHWW as a class III uterine and class IIa vaginal anomaly, due to failure of lateral and vertical fusion of the mullerian structures[13]. The proximity of the mesonephric and paramesonephric ducts explains the association between genital tract anomalies and renal anomalies, which is why renal agenesis is found in the syndrome[10,14,15].

Unlike other genital tract anomalies that occur with amenorrhea, SHWW has a normal menstrual pattern or small-volume bleeding[10,12,15,16]. In addition to the uncharacteristic symptomatology in the first months, the use of contraceptives in patients with dysmenorrhea can suppress the menstrual cycle and the symptoms of colic-like pain and increased abdominal volume[16].

On physical examination, a suprapubic mass is identified that is painful on palpation and may have urinary incontinence on mobilization. The bimanual touch shows the hemivaginal septum bulged by hematocolpos or not. As there is a diagnosis in young people without sexarche, making bimanual examination difficult, anal examination can be performed, which shows vaginal bulging. The investigation includes Pelvic, Kidney and Urinary tract USG, and the gold standard, which is pelvic MRI[9,11,12]. The images show the presence of didelphys uterus, distention of the horn by hematometra and hematocolpos, hydronephrosis and ipsilateral renal agenesis[9,12,14,15].

Treatment is aimed at relieving the symptoms caused by this distention of the uterine horn and preserving the patient's fertility; in addition to avoiding immediate complications, such as urinary infection and symptoms of pelvic inflammatory disease[10,14,16,17], and late complications, such as pyosalpinx, endometriosis, pelvic adhesions and spontaneous abortions[6,17-19]. It should also provide the patient with a normal postoperative life, with the ability to maintain normal sexual relations and reproductive life[15,17,19].

In SHWW, surgical treatment is necessary and consists of complete excision or marsupialization of the vaginal septum, with drainage of the hematocolpos, which can be performed with preservation of the hymen. Hemihysterectomy, with or without salpingo-oophorectomy, is rarely indicated and should be avoided in order to achieve the greatest chances of success in relation to reproduction. Even though the prognosis is unknown, the use of laparoscopy to access the uterine anatomy is advocated and used[12]. In cases of pyometra with complicated pyosalpinx, hemihysterectomy with adnexectomy is necessary, which may compromise the patient's future[17,18].

In patients without sexarche, hysteroscopy with resection of the vaginal septum can be performed because it does not alter the integrity of the hymen. On the other hand, ipsilateral hysterectomy can be performed in cases of repetitive atresia and severe infection, but it is not totally recommended[19]. When it is necessary to postpone the surgery, gonadotropin-releasing hormone analogues can be used, with the aim of maintaining amenorrhea[18,19].

Patients with the syndrome must be regularly monitored by a specialized team in order to avoid possible complications. An appropriate multidisciplinary approach is also necessary, since the disease affects the renal system and can psychologically affect the patient.

## CONCLUSION

The characteristics of SHWW can delay the diagnosis and, consequently, the treatment, generating a greater risk of immediate and late complications. Diagnosis before menarche is rare, and is common in cases of renal complications[3,4]. The prescription of non-steroidal anti-inflammatory drugs and oral contraceptives in patients with dysmenorrhea, in addition to masking the pain, increases the risk of complications[9,11,12]. Therefore, SHWW should be suspected in adolescents with pelvic mass, renal agenesis, menstrual alterations and cyclical pelvic pain[1-3].

However, it should be remembered that SHWW is underdiagnosed and, in many cases, diagnosed late, because, among other factors, it is not well known in the pediatric

environment. Delay in diagnosis worsens the prognosis, with the development of acute (hematocolpos or pyohematocolpos) or chronic complications, such as pelvic adhesions, endometriosis and difficulties in maintaining pregnancy until delivery[10,16,17].

Thus, the existence of SHWW should be more widespread among pediatricians to facilitate the early diagnosis of the disease, avoiding future complications. Diagnosis before menarche is challenging and can be performed if there is a high degree of suspicion.

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