

## Herlyn-Werner-Wunderlich Syndrome on Imaging: Case Report

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### Abstract

Herlyn-Werner-Wunderlich Syndrome is a rare congenital anomaly affecting the urogenital tract, specifically involving the Mullerian ducts and mesonephric ducts. It is characterized by a triad of uterine didelphys with a blocked hemivagina and the absence of a kidney on the same side.

Symptoms typically manifest following menarche, presenting as pelvic pain, dysmenorrhea, and the development of a pelvic mass. In rare cases, it may lead to primary infertility in later years.

Diagnosis of this condition is typically accomplished through ultrasound and MRI imaging techniques.

In this case report, we present the clinical scenario of a 13-year-old patient who had experienced her third menstrual cycle and presented with pelvic pain lasting for a week.

**Keywords:** Didelphys uterus; Hemivagina; Hematocolpos; Imaging

### Introduction

Herlyn-Werner-Wunderlich Syndrome (HWWS), also known as OHVIRA, is a rare congenital malformation syndrome characterized by a triad of uterine didelphys with an obstructed hemivagina and missing kidney on the same side. This condition results from an uncommon combination of müllerian duct anomalies (MDAs) and mesonephric duct malformation within the female urogenital tract [1].

While the exact prevalence of this syndrome is unknown, it is estimated to affect between 0.1% to 3.8% of the population.

The syndrome was initially described by Herlyn and Werner in 1971, and in 1976, Wunderlich further characterized the association of right renal aplasia with a bicornuate uterus and a simple vagina with isolated accumulation of menstrual blood in the vagina [2].

Patients with HWWS typically remain asymptomatic until puberty and often develop symptoms shortly after their first menstruation. These symptoms commonly include pelvic pain, painful menstruation, or the presence of a palpable vaginal or pelvic mass.

In this case report, we present a case of uterus didelphys with an obstructed hemivagina diagnosed at the age of 14 years. Through this case, we will discuss the clinical, diagnostic, and therapeutic aspects of this uterine malformation [3].

### Case Report

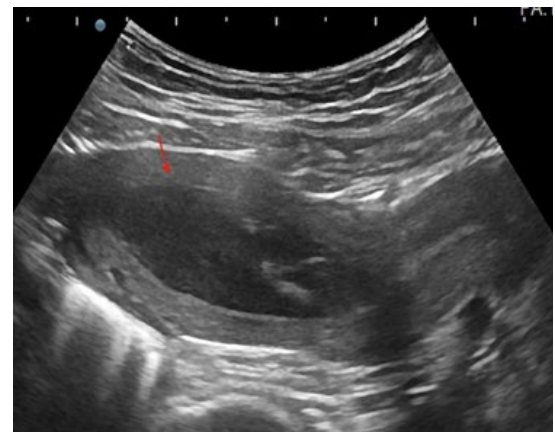
A 13-year-old unmarried girl with no significant medical or surgical history presented with acute pain in her lower right abdomen, which had been persistent for the past 7 days. She had her first period at the age of 13, and her menstrual cycles were irregular.

The patient did not report any fever, abnormal vaginal bleeding, nausea, vomiting, or diarrhea. During the physical examination, mild abdominal distension was observed along with tenderness in the lower right pelvic area. An abdominal ultrasound revealed the absence of the right kidney and a bicornuate uterus [4]. The right uterine horn appeared dilated and contained a cystic mass, with internal echoes indicating a hematoma. The left uterine horn and cervical canal appeared normal. Both ovaries were also found to be normal (Figure 1).

Following these findings, the medical staff recommended an MRI

for further evaluation. The MRI images revealed a didelphys uterus with the left uterine horn pushed backward by a distended right uterine horn. Additionally, the right hemi-vagina was obstructed and contained a cavity filled with menstrual blood (Figure 2). On axial T2 views, it was evident that the right kidney was absent, compensated by hypertrophy of the left kidney [5].

Based on these investigations, the patient received a diagnosis of right renal agenesis, uterus didelphys with an obstructed right hemivagina, and hematometocolpos.



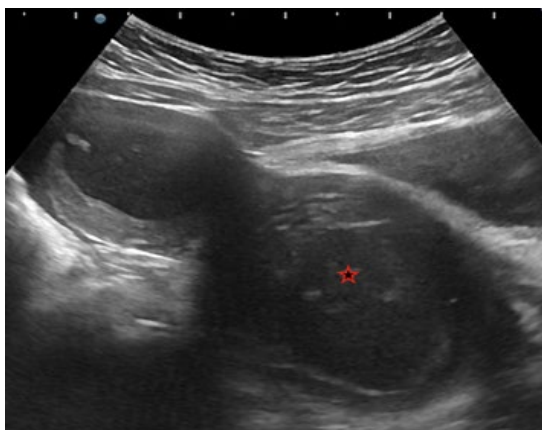
**Figure 1:** US of pelvis showing two uterine cavities, right side uterine cavity is grossly distended and fluid containing fine internal echoes within (red arrow).

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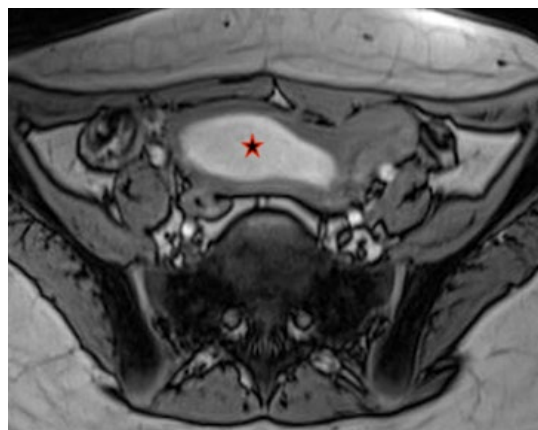
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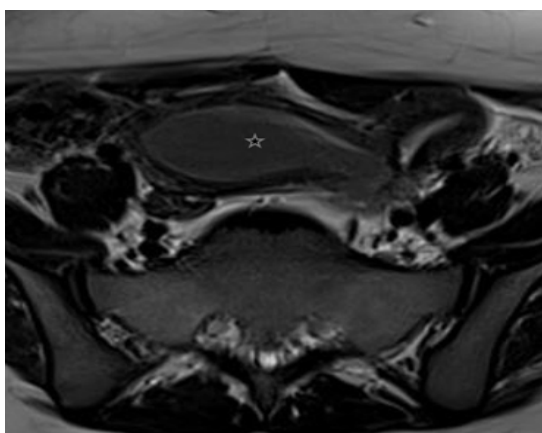
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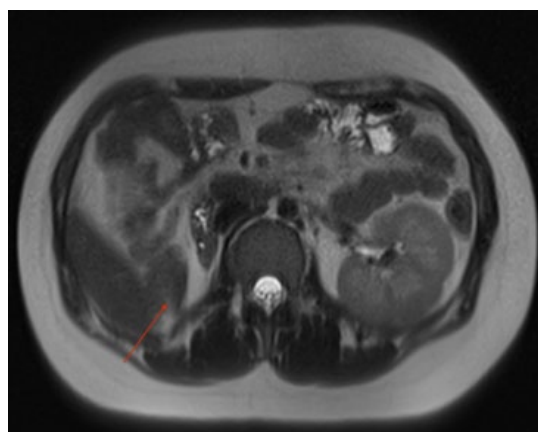
**Figure 2:** US of pelvis demonstrates cystic dilatation of cervix and vagina having internal echoes (red star).



**Figure 3 B:** T1W Axial MR images of pelvis showing a didelphys uterus with a right horn containing hematic retention cavity (red star).



**Figure 3 A:** T2W Axial MR images of pelvis showing a didelphys uterus with a right horn containing hematic retention cavity (white star).



**Figure 4:** T2W Coronal MR image of pelvis showing a blind right hemi-vagina containing a hematic retention cavity (red arrow).

Treatment commenced with hymenotomy, followed by the excision of the vaginal septum and drainage of a substantial amount of accumulated blood. Subsequently, reconstruction of the vaginal canal was performed, and a catheter was placed over the surgical resection site to prevent stenosis and facilitate drainage.

The primary goals of this treatment were to alleviate the patient's pain and prevent potential complications, such as endometriosis and infertility (Figure 3A).

## Discussion

Herlyn-Werner-Wunderlich Syndrome (HWWS) is a rare congenital malformation affecting the urogenital tract, specifically involving the Müllerian and mesonephric ducts [6]. Overall, congenital anomalies of the Müllerian tract are estimated to affect approximately 2 to 3% of women [7].

The precise pathogenesis of the HWWS pattern is not yet completely understood, but it is believed to have multiple contributing factors [8]. It may result from factors such as non-development (agenesia or hypoplasia), defective vertical or lateral fusion, or resorption failure of the Müllerian ducts [6]. HWWS is characterized by the presence of uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis (Figure 3B) [6].

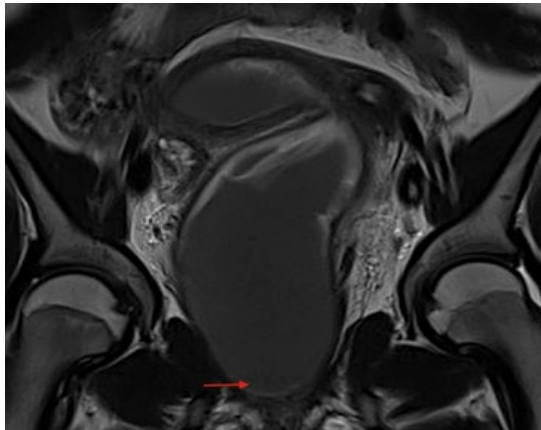
However, HWWS syndrome has been associated with various other malformations, including bicornuate and septate uterus, unilateral

cervical obstruction, and incomplete vaginal septum [14]. Additionally, urinary tract anomalies may also occur, such as dysplastic kidney, ureteral anomalies like blind ectopic ureter with vaginal or cervical insertion, and paravaginal cervical cysts [14].

Uterus didelphys is classified as Class 3 according to the American Fertility Society classification [10]. According to the European Society of Human Reproduction and Embryology (ESHRE) classification, our patient can be categorized as the main class U3 (bicornuate uterus), sub-class U3b (complete), with concurrent classes C2 (double 'normal' cervix) and V2 (longitudinal obstructing vaginal septum) (Figure 4) [11].

Patients with HWWS syndrome are typically asymptomatic until they reach puberty, at which point they may experience symptoms such as pelvic pain, dysmenorrhea, or the presence of a palpable mass in the vagina or pelvis [6]. The palpable mass often results from hematometocolpos, which occurs due to the obstructed hemivagina.

Ultrasound is the initial imaging modality used to evaluate suspected Müllerian duct anomalies. It can aid in establishing the diagnosis by revealing a didelphys uterus and the presence of blood retention in the hemivagina and/or the uterine cavity [8, 12]. MRI is an excellent imaging modality for assessing complex Müllerian duct anomalies and associated pelvic findings [6]. It typically reveals complete duplication of the uterine and endocervical cavities, along with the presence of a hematoma collection connected to one of the endocervical canals due



**Figure 5:** T2W axial MR image showing right renal agenesis with compensatory left renal hypertrophy (red arrow).

to the obstructed hemivagina [13]. Laparoscopy is considered the gold standard for diagnosing female genital tract abnormalities, but it is usually reserved for cases where MRI is unavailable or fails to provide a diagnosis [14].

Late diagnosis of this syndrome can lead to various complications, including endometriosis, pelvic adhesions, and infertility, all of which are related to chronic obstructed hematometocolpos (Figure 5). Therefore, early diagnosis and treatment are essential to prevent these complications [9]. The treatment of choice is vaginal septum excision, typically performed using laparoscopic or endoscopic techniques, as it ensures adequate drainage for hematometocolpos [9].

## Conclusion

Herlyn-Werner-Wunderlich Syndrome (HWW), a rare congenital urogenital anomaly, presents with pelvic pain and dysmenorrhea from the first menarche.

Due to its rarity, diagnosing HWW syndrome can be challenging. It should be considered as one of the potential differential diagnoses in young patients experiencing pelvic symptoms, particularly when there is an associated renal abnormality or agenesis.

The diagnosis is typically confirmed through a combination of pelvic ultrasound and magnetic resonance imaging (MRI), which remains the preferred diagnostic approach.

The primary treatment for HWW syndrome is surgical, involving

the complete removal of the vaginal septum to enable continuous drainage of menstrual retention.

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