# A Curious Radiological Presentation of a Rare Case: Herlyn-Werner-Wunderlich Syndrome

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## ABSTRACT

An uncommon and intriguing radiological representation of Herlyn-Werner-Wunderlich syndrome (HWW), a rare Mullerian duct anomaly, is presented in this case report. Uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis are the three characteristics that define HWW. This case study illustrates the difficulty in diagnosing HWW by describing an unusual radiological manifestation in a young female patient. The patient's imaging studies revealed some unusual results, such as asymmetrical pelvic structures and peculiar anatomy of the urinary and reproductive systems. Because of this unusual presentation, this anomaly presented difficulties for both diagnosis and subsequent management. The radiological representation was different from standard HWW patterns, highlighting the significance of thorough imaging analysis and clinical suspicion to make an accurate diagnosis. Comprehending these non-traditional manifestations of HWW is essential for prompt detection and suitable treatment measures to avert possible consequences linked to this uncommon illness.

**Key Words:** *Mullerian duct anomalies, Female urogenital disease, Diagnostic imaging.*

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## INTRODUCTION

The Herlyn-Werner-Wunderlich (HWW) syndrome was first described in 1922 and is characterised by ipsilateral renal anomalies, uterine didelphys, and blind hemivagina.[1] Later reports described deviations from the classical form: Herlyn and Werner described kidney aplasia, an open Gartner's duct cyst, and a double uterus without obstruction, and Wunderlich identified cervical obstruction.[2] Clinical presentations include dysmenorrhea, dyspareunia, and possible side effects such as endometriosis or pelvic inflammatory disease. Since surgical intervention is frequently required, early and accurate diagnosis is crucial.[3] Considering the complexity of the syndrome, which includes a variety of uterine and renal symptoms, a thorough assessment is essential. Understanding intricate anatomical variations is aided by MRI.

## CASE REPORT

A 17-year girl complained of intermittent lower abdominal pain that had gotten significantly worse over the previous week. The pain had been present for three years. The menarche and a regular menstrual cycle began at 10 years of age.

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Unremarkable external genitalia were found upon physical examination. Despite a history of regular menses and normal external genitalia, the patient's primary complaint of intensified lower abdominal pain prompted an investigation into possible underlying causes for the recent exacerbation of symptoms.

Using a convex probe operating at 3.5 MHz, pelvic ultrasonography revealed the presence of two separate uterine cavities (Figure 1).

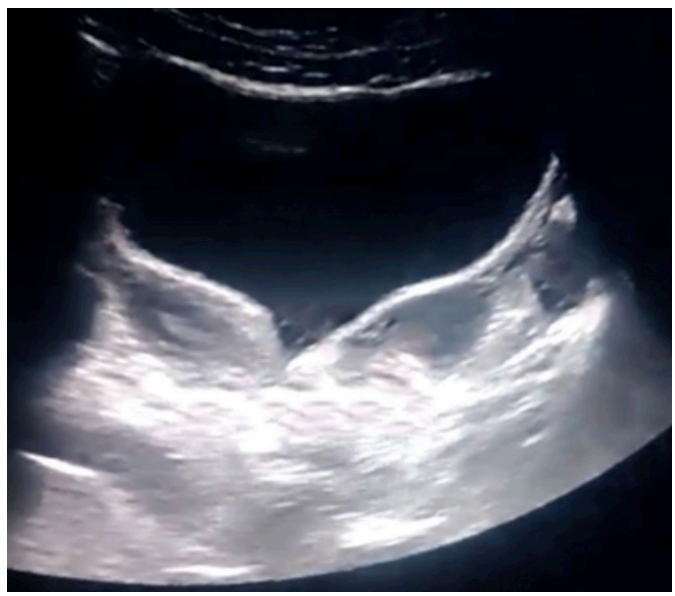

**Figure 1: Gray-scale ultrasound of the pelvis, showing a duplicated uterus with two separate endometrial cavities.**

A pelvic MRI revealed unilateral haematocolpos on the right side along with total duplication of the uterus, cervix, and vagina. Both ovaries showed many follicles, mostly smaller than 10 mm, and measured 3.1 × 1.9 × 2.1 cm (6 ml) on the right and 3 × 1.4 × 2.2 cm (4.6 ml) on the left. There was no evidence of pelvic ascites or lymphadenopathy, and the remaining pelvic organs were normal (Figure 2).

CT scan abdomen showed hypertrophied left kidney and right renal agenesis. The remaining viscera in the abdomen looked normal (Figure 3). The duplicated uterus was also appreciated on CT pelvic images (Figure 4).

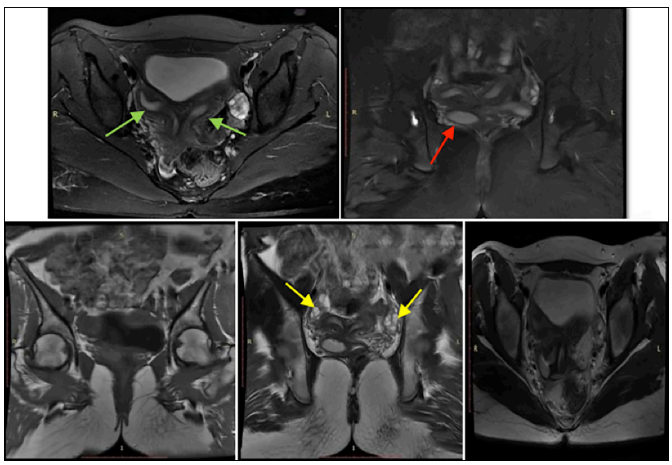

**Figure 2: An MRI of the pelvis reveals complete duplication of a morphologically normal uterus (green arrow), cervix, and vagina, as well as unilateral haematocolpos (right side, red arrow). Both ovaries had more than 10 follicles (yellow arrow), none of which were larger than 10 mm in diameter.**

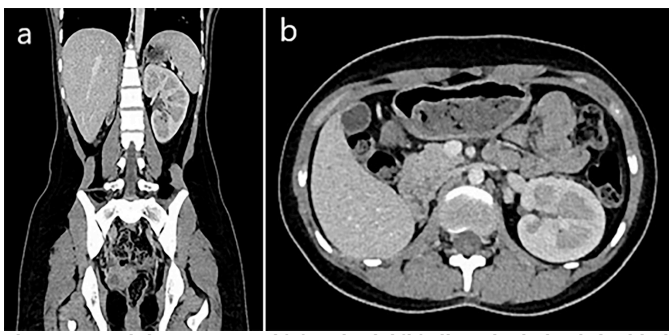


**Figure 3: CT abdomen coronal (a) and axial (b) slices depicting left-sided hypertrophied kidney with agenesis of the right kidney.**

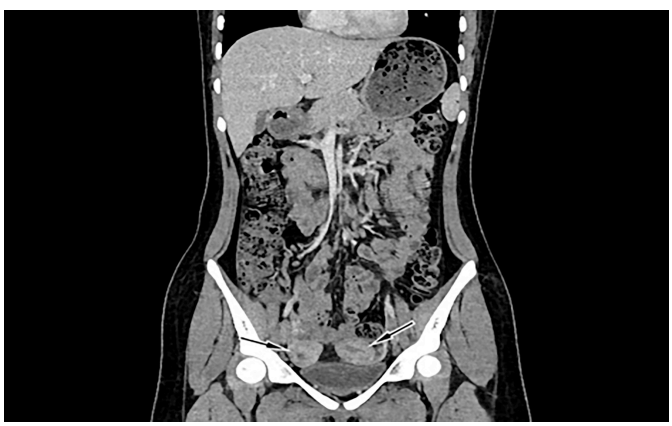

**Figure 4: CT abdomen and pelvis coronal slice showing two uterine cavities with separate endometrium (black arrows).**

## DISCUSSION

A rare class of uterine developmental defects, known as Mullerian duct anomalies (MDAs), or uterus didelphys, is frequently associated with renal anomalies. Of MDAs, uterine didelphys accounts for 11%. A subgroup of MDAs, known as HWW syndrome, is characterised by renal abnormalities, uterine didelphys, and obstructed hemivagina.[4-7] Anatomical characteristics of obstetric malformations resulting from MDAs are varied.[4] With an estimated incidence ranging from 0.1 to 3.8%, HWW syndrome is a rare urogenital anomaly typified by ipsilateral kidney absence, hemivagina, and uterine didelphys.[5] Clinical manifestations include pelvic pain, dyspareunia, and dysmenorrhea, which are frequently linked to endometriosis.[3]

The main preoperative diagnostic method is ultrasound, and different surgical procedures are intended to reduce symptoms and maintain fertility. Although ultrasound is initially useful in raising suspicion of the pathology, MRI outperforms it in detailing complex malformations, guiding preoperative planning, and aiding in maintaining fertility.[7]

To prevent complications, early detection and intervention are essential. One such intervention is hemivaginal septum excision. Hysteroscopic therapy performed in-office provides secure, anesthetic-free relief while maintaining fertility. Treatment involves draining haematocolpos and cutting the blocked vagina. Symptoms such as haematocolpos and lower abdominal pain guide the course of treatment.[4] For efficient symptom relief and fertility preservation, laparoscopic techniques and particular surgical procedures are taken into consideration in complicated cases.[5]

Considering the psychological and social effects of congenital genital malformations, specialised and comprehensive care is essential, particularly for adolescent patients. To ensure efficient management and follow-up, patients and specialised centres must work together. Patients find that incorporating psychological support into clinical care greatly helps them deal with the difficulties associated with these congenital defects. Innovations in treatment approaches, like one-step procedures, improve patient convenience by encouraging effective symptom relief while maintaining fertility.[6]

This case study highlights the difficulties in diagnosing HWW syndrome, a rare but serious MDA, and the value of sophisticated imaging. The rare presentation highlighted in this article adds to the diagnostic narrative of HWW syndrome, supporting early and correct detection to avert serious sequelae. To address the wider effects on patients' lives, it emphasises the need for a multidisciplinary approach to treatment that includes psychological support.

### PATIENT'S CONSENT:

Informed consent was obtained from the patient's parents to publish this case.

### COMPETING INTEREST:

The authors declared no conflict of interest.

**AUTHORS' CONTRIBUTION:**

MTK, RR: Literature review, drafted an initial document, created images, and amended the final draft.
MS: Oversaw the research and revision.
GJ: Revised the manuscript and edited images.
All authors approved the final version of the manuscript to be published.

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