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Case Report

# Herlyn-Werner-Wunderlich syndrome in a young female presenting with dysmenorrhea: A case report ☆

*Prajwal Dahal, MD*[a,*], *Ongden Yonjen Tamang, MD*[a], *Rudra Prasad Upadhyaya, MD*[a], *Kapil Dawadi, MD*[a], *Prajina Pradhan, MD*[a], *Sabina Parajuli, MBBS*[b]

[a] *Department of Radiology and Imaging, Grande International Hospital, Kathmandu, Nepal*
[b] *Department of Pathology, Bir Hospital, Kathmandu, Nepal*

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ABSTRACT

Herlyn-Werner-Wunderlich (HWW) syndrome, popularly known by acronym of obstructed hemivagina with ipsilateral renal agenesis (OHVIRA) is a rare Müllerian and Wolffian duct anomaly. The syndrome is classically described as triad of uterine didelphy, obstructed hemivagina, and ipsilateral renal agenesis. The symptoms are seen after menarche. Clinically, the patients present with dysmenorrhea, palpable paravaginal mass, increased frequency of micturition heaviness in pelvis and sometimes with infertility after marriage. Imaging is the cornerstone for diagnosis of the condition. Radiologists should screen for Müllerian duct anomaly if unilateral renal agenesis is detected and vice versa. Early diagnosis and surgical intervention help in prevention of complications like endometriosis, adhesions in lesser pelvis, infertility, pyosalpinx, abscess, and hematoma of cervix and vagina. We present a case of HWW syndrome in an adolescent female visiting the emergency department with complaint of dysmenorrhea. We diagnosed the case in ultrasonography (US) and confirmed in MRI.



## Introduction

The development of urogenital structures occurs at 6-12 weeks of gestation [1]. Mesonephric ducts, also known as Wolffian ducts and paramesonephric ducts also known as Müllerian ducts are important for the embryogenesis of the renal collecting system, uterus, cervix, and upper vagina [2]. The mesonephric ducts lie medially and stimulate the formation of paramesonephric ducts. The paramesonephric ducts cross the mesonephric ducts and fuse in the midline to form the uterus, cervix, and upper two third of the vagina [2,3]. The urogenital sinus forms the lower third of the vagina [2]. The vertical and horizontal septa usually disappear by the 12th

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* Corresponding author.
E-mail address: meprajwal7@gmail.com (P. Dahal).

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week of gestation to form an internally hollow uterus and a septa-free vagina [3]. The Wolffian duct also forms a collecting system of metanephros, the renal pelvis, and the ureter and facilitates further development of the kidneys [3]. Any abnormality in the Wolffian duct results absence of development of the collecting system, renal pelvis, and ureter and lateral displacement of the ipsilateral mesonephric duct, preventing fusion with the contralateral mesonephric duct [3]. This explains the co-existence of ipsilateral renal and Müllerian duct anomalies in HWW syndrome.

The patients with HWW present with symptoms of dysmenorrhea, para vaginal abdominal lump, increased frequency of micturition, and feeling of heaviness in the lower abdomen [4]. The symptoms start after menarche and the severity increases progressively. Rarely the patients present after marriage with complaints of primary infertility [2]. Diagnosis of HWW syndrome is done with imaging. US is cheap, readily available, radiation-free, and can provide real-time imaging in all planes [2]. The diagnosis can be made in US. However, MRI provides a better characterization of the anatomy and nature of hematocolpos/hematometra and aids in the planning of surgery [3]. Hence, MRI is recommended in all suspected cases of HWW syndrome [2]. Diagnostic laparoscopy is reserved for those cases in which anatomy is not clear in MRI [5].

## Case presentation

A 14-year-old female from a remote district of the country presented to the emergency department of a tertiary care center with complaints of severe right lower abdominal pain and vomiting for 2 days, increasing over a few hours. The pain was radiating to the back. The patient was menstruating for the last 2 days. The patient provided a history of similar pain of progressively increasing intensity during menses for the last 2 years. The patient had menarche at 12 years of age. According to the patient, her menstrual cycle was regular with blood flow for 4 days, soaking 3-4 home made pads per day. On deep palpation, a firm lump was palpable in the right lower abdomen. The lump was mildly tender. Blood examination reports were normal except for mildly reduced hemoglobin level (11 mg/dL). The serum creatinine and serum urea levels were 0.7 mg/dL and 17.8 mg/dL respectively. An US examination was performed which revealed non-visualization of the right kidney in the right renal fossa and possible ectopic sites and uterine didelphy. An isoechoic collection measuring ~7.2 × 5.6 cm was present in the cervix of the uterus on the right side of the abdomen. The left kidney and ureter were normal. MRI pelvis revealed uterine didelphy with hematometra and hematocolpos in the uterus in right side of the abdomen. There was no communication between the endometrial cavities of the 2 uteruses. The uterus on the right side of the pelvis had a blind-ending vagina with vertical septa. A small dysplastic kidney was seen in the right renal fossa. The renal parenchyma could not be appreciated in the dysplastic kidney. A few subcentimetric size simple cysts were seen replacing the renal parenchyma. Diagnosis of HWW syndrome was made and the patient was referred to the gynecology department for further management.

## Imaging

US was done on Toshiba Canon Aplio 500 platinum ultrasound machine using a 6-19 MHz convex transducer. In US, the right kidney could not be visualized in the expected anatomical location and possible ectopic sites. Two separate uteruses were visualized in the pelvis (Fig. 1A). Each uterus had a separate cervix (Fig. 1B). There was an isoechoic collection measuring ~7.2 × 5.6 cm in the cervix and upper vagina of the uterus on the right side of the pelvis (Figs. 1B and C). External os of the cervix was patulous. The uterus in the left half of the pelvis was normal (Fig. 1D). The 2 ovaries were visualized, one attached to each uterus. Other abdominal organs were normal. Although diagnosis of HWW syndrome was made on US, MRI pelvis was advised for proper anatomical definition. MRI can properly evaluate uterine contour, shape of endometrial cavity, location and orientation of vaginal septum, type of collection in obstructed hemivagina and associated urinary tract malformation. MRI pelvis was done in a 1.5 Tesla Philips MRI machine which revealed similar findings. A small ~ 3.5 cm size multicystic dysplastic kidney was present in the right renal fossa (Fig. 2). The right ureter could not be visualized. Two uteruses were present in the pelvis. Each uterus had a separate cervix (Fig. 3A). A collection measuring ~ 10.1 × 5.7 cm was present in the cervix and upper vagina of the uterus located on the right side of the pelvis (Figs. 3B and 4). The collection demonstrated mixed signals in T2-weighted images (Figs. 3A and B) and high signal in T1-weighted images. The high signal in T1-weighted images was not suppressed in T1 fat-saturated images (Fig. 4). A vertical vaginal septum was present in vagina obstructing the flow of collection in the cervix and upper vagina (Fig. 5).

Comparison of US and MRI findings for evaluation of our case of HWW syndrome can be found in Table 1.

## Discussion

HWW syndrome was initially reported in 1922 by Purslow [3,5]. Since 2007, the term OVHIRA syndrome has been used instead of HWW syndrome [6]. The reported incidence of HWW syndrome is 0.16%-10% of all Müllerian anomalies [7]. HWW syndrome is characterized by the coexistence of renal and Müllerian anomalies on the same side. The most common renal anomaly associated with the syndrome is renal agenesis and the most common Müllerian anomaly associated with the syndrome is uterine didelphys with vertical vaginal septum [1]. Other renal anomalies like megaureter, ectopic ureter, dysplastic kidney, and renal duplication may be associated with the syndrome [3]. Other Müllerian anomalies like the septate uterus and bicornuate uterus may be present [8]. The syndrome occurs due to abnormality of the Wolffian and Müllerian ducts. The diagnosis is usually made after menarche during puberty. The signs and symptoms are because of ob-

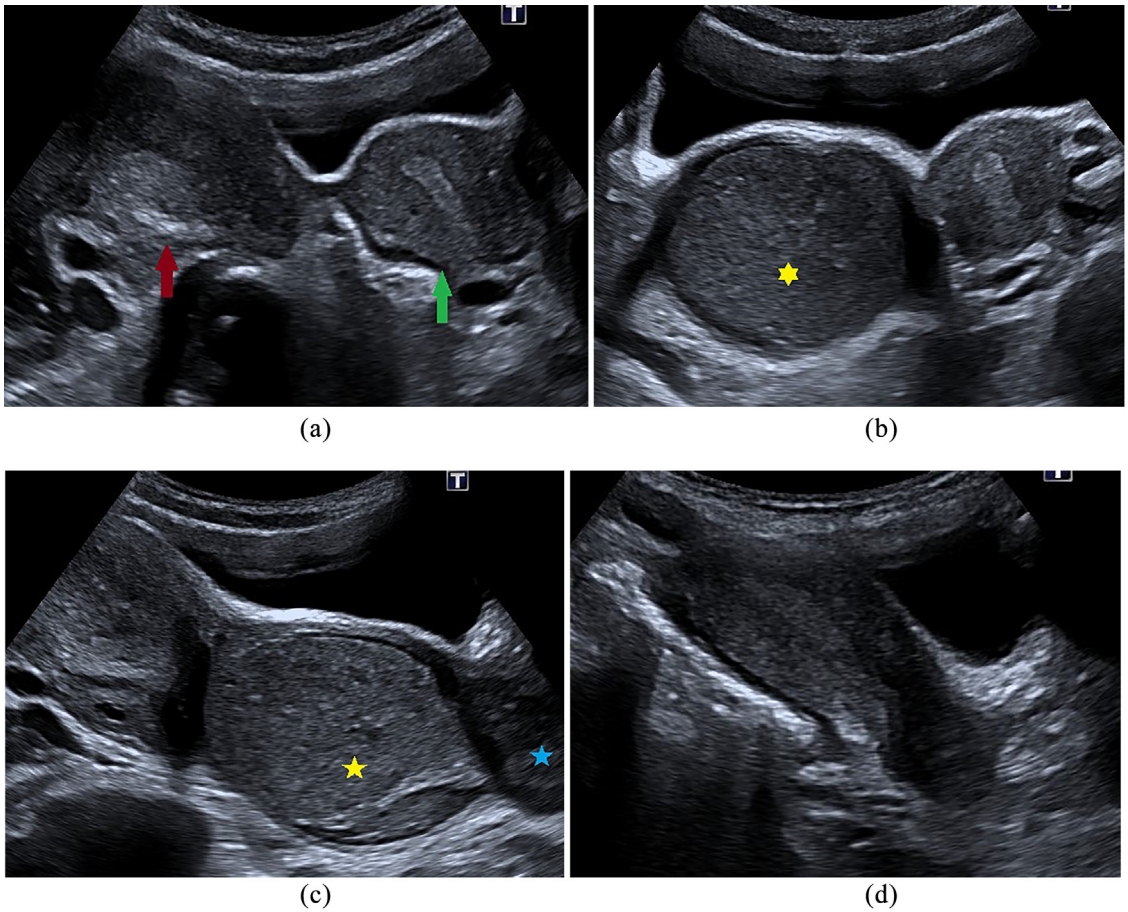


**Fig. 1 – (A) High-resolution ultrasound image of pelvis. Uterine didelphy is seen with 2 separate uteruses on the right side (marked with a red arrow) and the left side (marked with a green arrow) of the pelvis. (B) High-resolution ultrasound image of the pelvis in the axial plane. The cervix of the uterus on the right side of the pelvis is filled with isoechoic contents and is grossly distended (marked with a yellow star). (C) High-resolution ultrasound image of the uterus on the right side of the pelvis in the sagittal plane. A blind-ending vagina is seen in the uterus on the right side of the pelvis. The cervix (marked with a yellow star) and blind-ending vagina (marked with a blue star) are filled with isoechoic contents and are grossly distended. (D) High-resolution ultrasound image of the uterus on the left side of the pelvis in the sagittal plane. The uterus including the cervix appears normal.**

**Table 1 – Comparison of US and MRI findings for evaluation of our case of HWW syndrome.**

| Discrepancies in findings of ultrasonography and MRI | Similarities in findings of ultrasonography and MRI |
|---|---|
| 1) Dysplastic kidney was not visualized in US but clearly depicted in MRI | 1) Uterine didelphy was seen in both US and MRI |
| 2) Vaginal septum was not visualized in US but clearly seen in MRI | 2) The ovaries were visualized in both US and MRI |
| 3) Nature of collection in cervix and obstructed hemivagina could not be assessed in US. MRI demonstrated the collection to be blood products. | 3) Collection in cervix and obstructed hemivagina was visualized in both US and MRI |

structed flow in one of the uteruses. Imaging is the diagnostic modality of choice. US is the initial modality of imaging. MRI helps in better delineation of anatomy and pre-operative planning. Diagnostic laparoscopy is done if anatomical delineation with MRI is not satisfactory. American Society of Reproductive Medicine has classified Müllerian duct anomalies as uterine agenesis or hypoplasia [type 1], unicornuate uterus [type 2], uterine didelphy [type 3], bicornuate uterus [type 4], septate uterus [type 5], arcuate uterus [type 6], "T" shaped uterus related to diethylstilbestrol use [type 7] [9]. Uterine didelphy associated with our case is classified as a type 3 anomaly.

In 2015, Zhu et al. [8] proposed a detailed classification of HWW syndrome. According to them, HWW can be classified

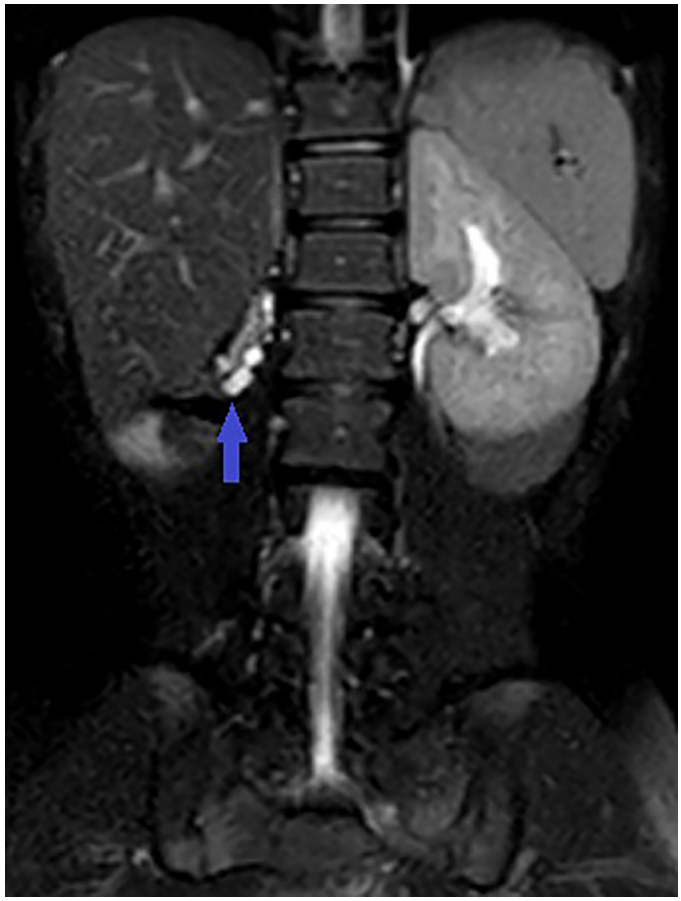

**Fig. 2 – Coronal STIR image of the abdomen at the level of bilateral renal fossae. A small multicystic dysplastic kidney is seen on the right side (marked with a purple arrow). The right ureter is not visualized. Visualized left kidney and left upper ureter are normal.**

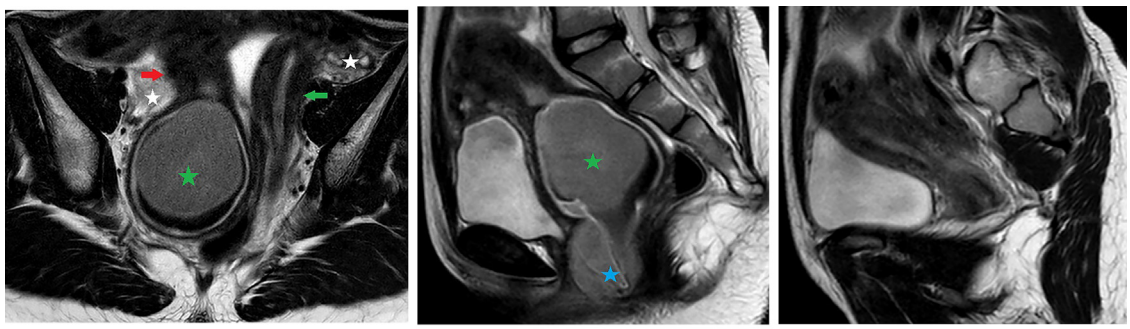


**Fig. 3 – (A) High-resolution oblique coronal T2-weighted image of the pelvis. Uterine didelphy is seen (the uterus on the right side of the pelvis is marked wired arrow and the uterus on the left side of the pelvis is marked with a green arrow). The cervix of the uterus on the right side of the pelvis is grossly distended and filled with contents showing T2 mixed signal (marked with a green star). The ovaries are separately visualized (marked with white stars). (B) High-resolution T2-weighted sagittal image of the uterus on the right side of the pelvis. A blind-ending vagina is seen on the right side of the pelvis. The cervix (marked with a yellow star) and blind-ending vagina (marked with a blue star) are filled with contents showing mixed signals and are grossly distended. (C) High-resolution T2-weighted sagittal image of the uterus on the left side of the pelvis. The uterus including the cervix appears normal.**

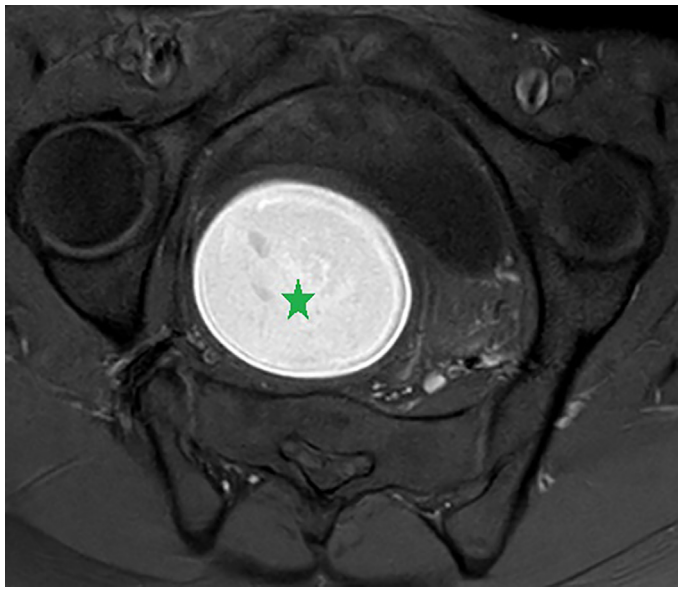

**Fig. 4 – High-resolution T1 fat-saturated axial image of pelvis. The contents in the cervix of the right side of the uterus show a high signal and are not suppressed in fat-saturated images. This is suggestive of hematometra and hematocolpos.**

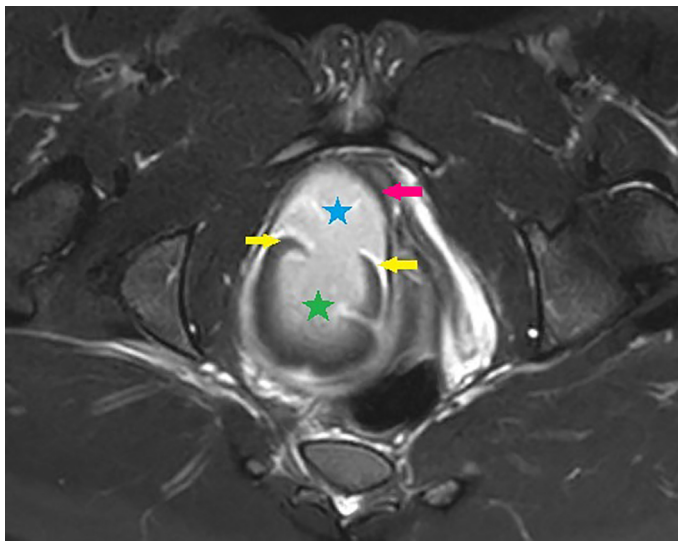

**Fig. 5 – High-resolution T2 STIR axial image of the pelvis at the level of the external os of the cervix of the uterus on the right side of the pelvis. The external os is marked with a yellow arrow. The cervix is marked with a green star and the upper vagina is marked with a blue star. A vertical vaginal septum is seen (marked with a pink arrow).**

into 4 different variants depending on the presence or absence of vaginal septum and communication between endometrial cavities of 2 uteruses.

Zhu et al. classification of HWW can be found in Table 2.

Clinical features of the syndrome vary depending upon the type. Types 1.1 and 1.2 present with pain starting after menarche and paravaginal lump. Types 2.1 and 2.2 present with prolonged and heavy menstruation rather than pain. Pain and paravaginal lump are seen after a few years. Presentation with genital tract infections is also common in these types.

A systematic review was done by Vercellini et al. [10] of Milan University which revealed the presence of anomalies on the right side in 65% of HWW syndrome. In our case, too, a right-sided anomaly is seen. Determining the sidedness of anomaly in OHVIRA syndrome is important for surgical planning. Some studies have shown an association of HWW with skeletal anomalies but Cappello et al. [6] have concluded that

**Table 2 – Zhu et al classification of HWW.**

| Type of HWW | Features |
|---|---|
| Type 1.1 | No communication between the uterine cavities and the vagina |
| Type 1.2 | The closed cervix is embedded in the blindly ending vagina |
| Type 2.1 | There is communication between both vaginas |
| Type 2.2 | The vaginal septum is complete but there is communication between 2 cervices or the blind-ending vagina and second cervix |

skeletal anomalies are not typically associated with HWW syndrome. They also concluded that right-sided anomalies are more common.

Excision of the vaginal septum and drainage of hematometra/ hematocolpos is the definitive treatment [3]. Early intervention helps in the prevention of complications like endometriosis, pelvic adhesion, primary infertility, pelvic abscess, etc.

If early intervention is done, fertility is preserved in most cases. Few studies are done which show an 87% pregnancy rate in patients with HWW syndrome and positive obstetric outcome in 62% of cases without complications [6]. Chances of miscarriage and preterm delivery are high if conception occurs in the uterus with a blind-ending vagina.

## Treatment and follow-up

The objectives of treatment are to relieve the obstruction, which cures pain and increases the chances of conception [11]. The definitive treatment for OHVIRA syndrome is vaginoplasty. Usually, a single-stage vaginoplasty is performed, but sometimes a 2-stage or multistage vaginoplasty is required [11]. The advantages of vaginoplasty include the alleviation of symptoms associated with OHVIRA syndrome, improved quality of life, and the preservation of fertility. Immediate postoperative complications of vaginoplasty include infection, bleeding, and postoperative pain. Late postoperative complications of vaginoplasty may include vaginal stenosis, fistula formation, and dyspareunia [11]

The patient was referred to the gynecology department. The patient was counseled for vaginoplasty. The benefits and risks of the surgery were explained to the patient. The patient denied any surgical intervention and was lost to follow-up. Attempt was made to contact the patient for further counseling, but all the efforts went in vain.

## Learning points

- HWW syndrome is classically characterized by a triad of uterine didelphy, hemivagina obstruction, and ipsilateral renal agenesis. But other forms of renal anomalies and uterine anomalies may be present.
- Radiologists, pediatricians, and gynecologists should be aware of the syndrome. The presence of uterine anomaly should be looked for if renal agenesis is detected in females and vice versa.
- Early diagnosis and surgery prevent complications.
- Fertility is preserved in most of the cases and obstetric outcome is good [5].

## Patient consent

Written informed consent was obtained from the patient's father for publication of the case report including accompanying images.

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