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# Herlyn-Werner-Wunderlich Syndrome in a young female presenting with dyspareunia - A Case Study

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## Introduction

Herlyn-Werner-Wunderlich (HWW) syndrome is a rare congenital Mullerian duct anomaly of the urogenital tract characterised by uterine didelphys with obstructed hemivagina and ipsilateral renal agenesis1. This case presents a unique occurrence of HWW syndrome diagnosed in a female presenting with dyspareunia.

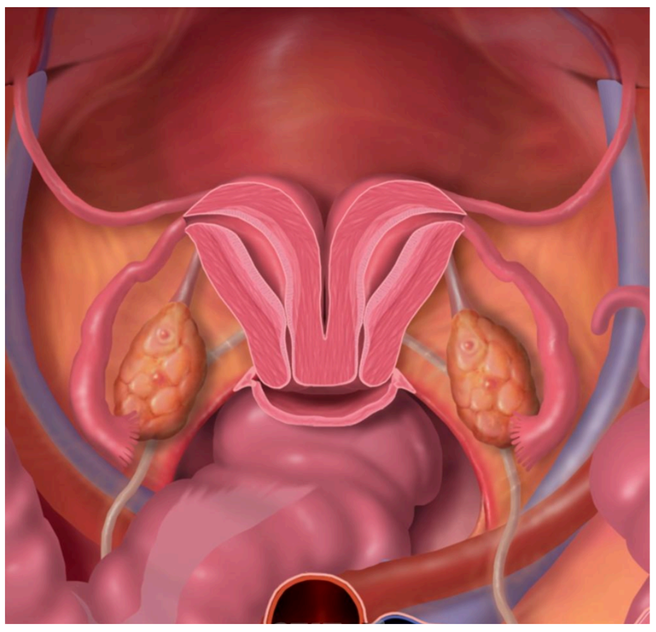

**Figure 1.** Graphic of a uterus didelphys shows complete duplication of uterine horns and cervices with no communication of the endometrial cavities. Image Credit: STATdx

## The Case

A nulliparous female presented to the Emergency Department with sudden onset, sharp vaginal pain following intercourse. She denied a history of dysmenorrhea, intermenstrual bleeding or post-coital bleeding. On speculum examination, a 5x3cm mass was noted at the right vaginal wall, with some brown discharge noted from the cervix.

Pelvic ultrasound demonstrated a suspected bicornuate uterus with a complex cystic foci in the upper vagina and cervix, no tubo-ovarian pathology was seen (see image 5-6). Magnetic resonance imaging (MRI) of the pelvis, demonstrated uterine didelphys and unilateral obstructed right hemivagina, with a possible right haematocolpos and right hydrosalpinx or haematosalpinx (See images 1-4). Based on the findings of the MRI, the right hemivaginal obstruction was thought to be due to a transverse vaginal septum.

A drainage of pyocolpos and excision of vaginal septum was performed. Surgical findings were consistent with transverse vaginal septum on the right half of the vagina. A right sided vagina with bulge around 6cmx5cm was noted with a single point draining pus noted. The left cervix was felt lateral to the bulge, and the right cervix was identified following the excision of the septum. An ultrasound renal tract performed post-operatively confirmed an absent right kidney.

On post-operative follow up at 8 weeks, the patient had resumed normal menstrual cycles, and reported no vaginal pain. On vaginal examination two cervices were felt, with no residual septum palpable.

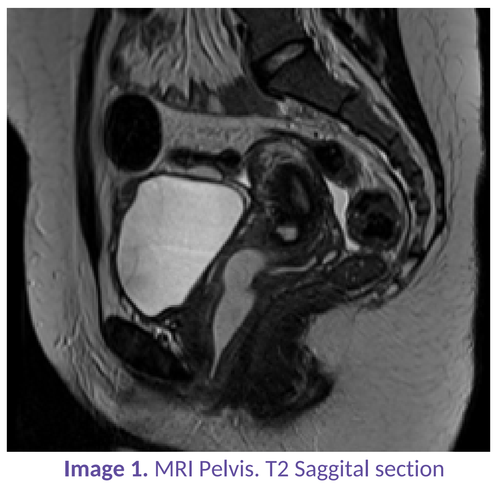

**Image 1.** MRI Pelvis. T2 Saggital section demonstrating pyocolpos and one side of the duplicated uterine cavities.

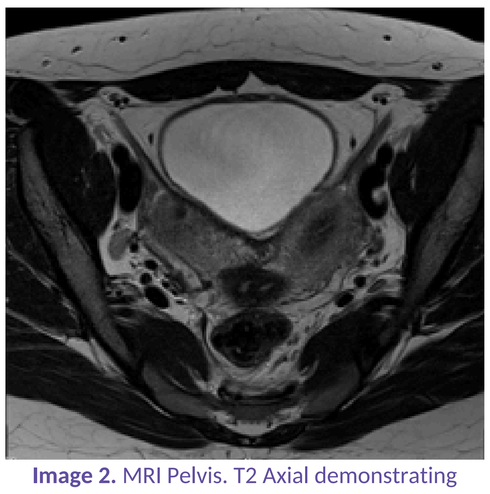

**Image 2.** MRI Pelvis. T2 Axial demonstrating fundus and body of each component of uterus.

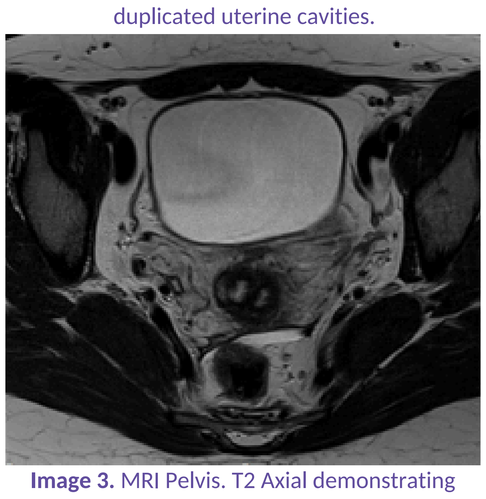

**Image 3.** MRI Pelvis. T2 Axial demonstrating duplication of cervix.

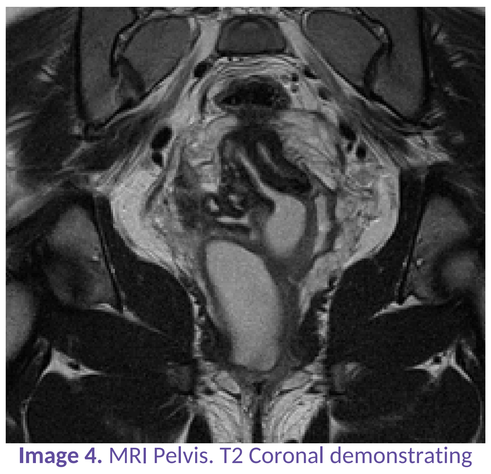

**Image 4.** MRI Pelvis. T2 Coronal demonstrating duplication of endocervical cavity.

## Discussion

Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA), also known as Herlyn-Werner-Wunderlich (HWW) syndrome, is a rare congenital anomaly of the female reproductive and urinary systems1. The condition is characterized by the presence of uterus didelphys with an obstructed hemivagina and the absence of the ipsilateral kidney2 (See Figure 1).

**OHVIRA is the most common obstructive uterine anomaly2 with an estimated global incidence of 0.1-3.8%3** and is thought to arise secondary to anomalous development of the both the paramesonephric and mesonephric ducts during embryological development4.The mesonephric ducts play a significant role in inducing the normal development of the paramesonephric ducts as well as giving origin to the formation of the kidneys. Subsequently, the abnormal development of the mesonephric ducts gives rise to the unilateral renal agenesis and imperforated hemivagina characteristic to OHVIRA syndrome4.

Most cases of OHVIRA are diagnosed in the first few years after menstruation, with common presenting symptoms including amenorrhea, dysmenorrhea, or recurrent abdominal pain5. The case described involves a diagnosis of a woman in her mid-twenties, which is significantly later than the median age of diagnosis5. It is possible that diagnosis was delayed due to the single point of drainage in the pyocolpos noted during surgical intervention preventing significant mass or collection. It also appears that the patient was menstruating normally through the left cervix, which was not obstructed by the vaginal septum. **MRI imaging is the gold standard in diagnosis of HWW1**. In this case, the patient was misdiagnosed with bicornuate uterus in earlier life, however the MRI examination performed prior to surgical intervention clearly demonstrated two separate uteruses with two cervices (see image 2 and 7).

Complications associated with delayed diagnosis of HWW include haematocolpos, dysmenorrhea, recurrent abdominal pain or pelvic pain, vaginal or pelvic mass, abnormal vaginal discharge, and urinary dysfunction3. For this reason, most patient with HWW are diagnosed within the first few months of menarche4. Fertility is typically not impacted in patients with HWW who undergo surgical excision of the obstructed vaginal septum, with a fertility rate of up to 87% noted following surgical correction6.

## Conclusion

HWW is one of the most common obstructive uterine anomalies noted in the literature, with an incidence of 0.1-3.8% globally3. While the majority of cases are diagnosed in early adolescence, the case presented highlights variances in typical presentation of HWW that can lead to diagnosis in later life.

HWW is an easily treated congenital anomaly, with complete resolution of symptoms and likely normal fertility outcomes following appropriate surgical intervention.

A greater understanding of HWW and the variety of ways in which it can present allows for improved recognition and subsequent improvement in timely intervention for women presenting with HWW.

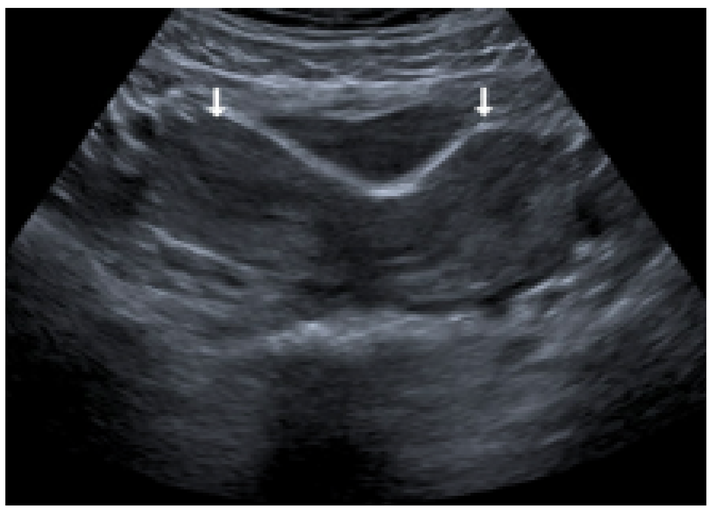

**Image 5.** USS Pelvis. Transverse view demonstrating duplication of uterine cavity.

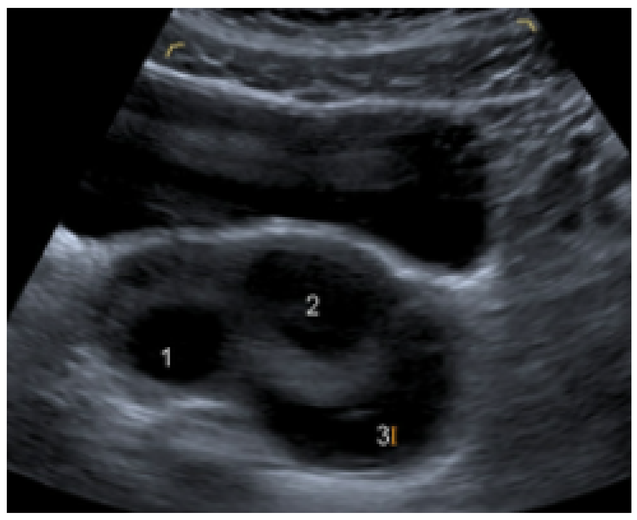


**Image 6.** USS Pelvis. Lower-level section demonstrating left cervix (1), right cervix (2), and pyocolpos (3).

## References

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