

# Case Report: A Rare Cause of Complicated Urinary Tract Infection in a Woman with Herlyn-Werner-Wunderlich Syndrome

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## Abstract

**Introduction:** Urinary tract infection is a common disease in the general population. However, in patients with frequent urinary tract infection, it is important to determine any treatable cause to avoid recurrence.

**Case Presentation:** Herlyn-Werner-Wunderlich syndrome or OHVIRA syndrome is a very rare congenital anomaly with uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis. The earliest presentation of this syndrome is hematocolpos that develops during menstruation and results in dysmenorrhea and a pelvic mass shortly after menarche. Herein, we report a patient with Herlyn-Werner-Wunderlich syndrome manifested with unusual symptoms, delayed onset and without surgery. The unique point of this patient is the partial obstruction of cervico-vaginal junction.

**Conclusions:** Early diagnosis and timely treatment of OHVIRA syndrome can prevent long-term complications, such as recurrent urinary tract infection and infertility. A high index of suspicion is required, even though OHVIRA syndrome is extremely rare and may have an atypical presentation.

**Keywords:** Urinary Tract Infection, HWWS, OHVIRA Syndrome

## 1. Introduction

Urinary tract infection (UTI) is very common in women. However, in patients with repeated UTI, the possibility of underlying causes or risk factors of complicated UTI should be investigated. They can include biologic or genetic factors, behaviors, pelvic anatomy, postmenopause, special virulence of pathogens, or incomplete treatment. OHVIRA syndrome is the triad of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis. It was first reported in 1950 (1) but the incidence is still low (between 0.5 and 5.0%) (2, 3). Almost all patients are diagnosed at their menarche due to obstruction of menstrual blood. Besides, nearly all patients should receive operation for the structural abnormalities. Herein, we report a woman with recurrent UTI diagnosed at the age of 43 years old and without any surgery. We also discussed the unique points of this patient and conducted an extensive literature review.

## 2. Case Presentation

This 43-year-old woman was previously in good health. She visited the emergent department of Taichung Veterans general hospital in Taichung city of Taiwan due to fever with chills and left flank pain on 14th September 2015. Post-void dribbling with difficulty in emptying bladder and de-

creased urinary amount for each micturition were also noted during the two-day period prior to admission. Physical examination revealed left flank knocking pain and abdominal dullness over suprapubic area. Laboratory data revealed 22000/cumm of white blood cell, 10 mg/dL of C-reactive protein, 2.1 meq/L of serum potassium, 2.35 mg/dL of serum creatinine, and pyuria (10-20/high power field). Abdominal computed tomography (Philips diamond select brilliance CT 64-slice) showed solitary left kidney with mild hydronephrosis. The patient also complained about severe nausea and vomiting for one week. She was then admitted to the department of nephrology. All data are summarized in Table 1. Soon after administration of 1st generation of cephalosporin and Foley insertion, she became afebrile. The urinary culture yielded Proteus. The hypokalemia was deemed to be most likely vomiting-related. Pyelonephritis and hypokalemia are commonly diagnosed and treated. However, to our surprise, only the left kidney was observed (Figure 1A), and two uteruses were found (uterus didelphys) (Figure 1B). The urinary bladder was compressed by the two uteruses (Figure 1B), which caused left mild hydronephrosis and compensated hypertrophy. In addition, right cervico-vaginal partial obstruction (obstructed hemivagina-communicant) was identified (Figure 1C). After meticulous tracing of her medical history, she claimed she had seven spontaneous abortions (G9P2SA7),

and she never experienced any surgical interventions except two caesarean sections. She had chronic pelvic pain, recurrent severe dysmenorrhea, spotting, and intermenstrual bleeding since her menarche. All manifestations of OHVIRA syndrome (uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis) were found in this patient. Family history was checked but she did not have any family history regarding this congenital abnormality. We treated her by antibiotics and the insertion of Foley. There was not any surgery for her because of well response for medical treatments and stable vital signs. Soon, she was discharged after one-week intravenous antibiotics and received one-week oral antibiotics at outpatient department, too. After this episode, she was under regular follow-up and there was no more UTI within one year. This study had been approved by patient herself and she signed the informed consent.

**Table 1.** Summary of Data

Variables	Value	Unit
Age	43	years
Body weight	91	kg
Body height	153	cm
Body mass index	38.87	kg/m <sup>2</sup>
<b>Vital signs</b>		
Blood pressure	130/80	mmHg
Heart rate	80	/min
Respiratory rate	16	/min
Body temperature	39.8	°C
White blood cell	22000	/cumm
C-reactive protein	10	mg/dL
Serum potassium	2.1	meq/L
Serum creatinine	2.35	mg/dL
Urinary white blood cell	10 - 20	/high power field
<b>Tumor markers</b>		
CEA	2.8	ng/mL
CA 125	20	u/mL
CA 153	15	u/mL
CA 199	25	u/mL

Abbreviations: CBC, Coulter® LH750 Analyzer from Beckman Coulter®; Urinalysis, iChem® VELOCITY™ from Iris®; Urinary culture, BACTECTM FX from BD; Serum potassium creatinine and CRP, COBAS® c501 from ROCHE; Tumor markers, Specific IMx tumor marker assays from Abbott.

### 3. Discussion

OHVIRA syndrome or Herlyn-Werner-Wunderlich syndrome (HWWS) remains a very rare congenital anomaly of the urogenital tract involving Mullerian and Wolffian (mesonephric) ducts atonalties (4-6), presenting the triad of didelphys uterus, obstructed hemivagina, and ipsilateral renal agenesis. It was first reported in 1950 (1) but the incidence remains still low (between 0.5 and 5.0%) (2, 3). Most data on the clinical significance and outcomes are based on small retrospective, observational, or case studies. In terms of embryology, nonfusion of the Müllerian ducts or failed resorption of the uterine septum cause duplication of the uterus (7). The lack of a caudal opening on one side of the duplicated vagina may result from the failure in formation of uterovaginal canal, mesodermal proliferation, or the vaginal canalization (8).

Typically, most women with didelphys uterus only are asymptomatic (9). However, in OHVIRA syndrome, obstructed hemivagina usually causes menstrual blood to accumulate in the obstructed side, which will distend the vagina, uterus and fallopian tubes, and it may cause pelvic pain or mass, hypermenorrhoea, intermittent vaginal spotting, menometrorrhagia, malodorous vaginal discharge, and urinary symptoms. This condition can cause renal agenesis on “the same” side of hematometrocolpos or hematocolpos because of embryologic arrest at the 8th week of gestation, which simultaneously affects the Mullerian and metanephric ducts. The incidence of renal anomalies is 20% in OHVIRA syndrome (10). In addition to symptoms of the reproductive system, there are still some rare presentations, such as acute retention of urine, pyelonephritis, and vomiting (11, 12). In this case, the patient also had urinary retention because of compression of the urinary bladder and pyelonephritis-related fever. The poor compliance of urinary bladder led to the incomplete emptying of urinary bladder and susceptible UTI. The nonspecific manifestations and extreme rarity represents a diagnostic dilemma. Because the onset of presentations usually occurs around menarche, young adolescent girls with the above conditions should be examined carefully. If she cannot receive surgery immediately, continuous GnRH analogues may be used to maintain amenorrhea to avoid obstructed hemivagina-related infections. In our case, the delayed diagnosis let her suffered from chronic pelvic pain, dysmenorrheal, and UTI. She used to take non-steroidal anti-inflammatory drug to relieve her dysmenorrhea, which caused renal dysfunction. The renal dysfunction was also superimposed with recurrent UTI.

The prognosis of this rare disease depends on the surgery, including excision of the vaginal septum, which helps in relieving obstruction. After successful surgery,



**Figure 1.** A, Solitary left kidney and agenesis of right kidney; B, Uterus didelphys and compression of urinary bladder; C, Non-communicating, thick vaginal septum over right side.

pelvic endometriosis due to retrograde menstrual seeding will decline and 87% of patients will still have successful pregnancy (13). For this patient, even without surgery for OHVIRA syndrome or HWWS, seven spontaneous abortions (G9P2SA7) were noted but she had two sons without complications.

There are some unique considerations for the affected patient. First, she had never received surgical intervention for HWWS, and this was the most delayed diagnosis in literature review. The main treatment for OHVIRA syndrome or HWWS is surgery as soon as possible. In our case, however, because cervico-vaginal obstruction was still with partially communicant, she did not have significant hematocolpos, pelvic pain, palpable abdominal, pelvic or vaginal mass (mucocolpos or pyocolpos), which made the delayed diagnosis of HWSS. According to this patient, we believe surgery may not be always necessary if partial obstruction of cervico-vaginal junction exists. Medical therapy at first and close monitoring of any complication may be alternative treatment in some patients like our case. Secondly, most symptoms of OHVIRA syndrome or HWWS did not progress further to cause urinary disturbance. Nevertheless, in this case, bilateral uterus compressed the urinary bladder, causing poor compliance of the bladder and recurrent UTI. Thus, even in the case of very common diseases such as UTI, clinicians should pay more attention to the point that if they are combined with other genital organ dysfunctions.

The strong point of this study is unusual presentations, clinical course and management of OHVIRA syndrome or HWWS, including late onset, presented by UTI, and no surgery. We still should keep in mind that very common complications (like UTI) may be due to very rare diseases.

On the contrary, there is a limitation for this study that we did not perform genetic and chromosomal tests for this patient. However, these tests may not be necessary because of typical manifestations of OHVIRA syndrome or HWWS in such cases.

### 3.1. Conclusion

Early diagnosis and timely treatment of OHVIRA syndrome can prevent long-term complications, such as recurrent UTI and infertility. A high index of suspicion is required, even though OHVIRA syndrome is extremely rare and may have an atypical presentation.

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### Footnotes

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