

# Successful pregnancy outcome in Herlyn-Werner-Wunderlich syndrome with pyocolpos: A case report and literature review

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## Abstract

The presence of pelvic pain, a pelvic/paravaginal mass, and purulent vaginal discharge in primigravida should raise the possibility of obstructed hemivagina and uterine didelphys. Though conservative management could result in successful pregnancy outcomes, early excision of vaginal septum and adequate drainage offer a shorter course of management and complication avoidance.

## KEYWORDS

OHVIRA syndrome, pyocolpos, urogenital anomalies, uterine didelphys

## 1 | INTRODUCTION

We report a new case with favorable outcome of Herlyn-Werner-Wunderlich syndrome coexisting with pyocolpos due to *Actinomyces* infection, diagnosed during pregnancy. Together with cases reported in the literature, it suggests that early excision of the vaginal septum and adequate drainage can shorten course of treatment and avoid unnecessary complications.

Herlyn-Werner-Wunderlich syndrome (HWWS) is a rare congenital malformation characterized by a triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis. It is also known as OHVIRA syndrome (obstructed hemivagina, ipsilateral renal anomaly) which has gained

popularity due to the report of cases with a nondidelphys uterus.<sup>1,2</sup> The incidence of HWWS/OHVIRA syndrome is the rarest type of Müllerian duct anomalies (MDAs), accounting for 0.1-3.8% of all MDAs of which its incidence is estimated to be 4.3-6.7%.<sup>1,3,4</sup> The HWWS was first described by Purslow (1922) and Embrey (1950) and became more popular in 1970s following the descriptions by Herlyn and Werner, and Wunderlich.<sup>1,2,5</sup> Didelphys uterus is a result of fusion arrest of the Müllerian ducts which should derive to uterus and cervix, whereas maldevelopment of the Wolffian ducts leads to abnormalities of kidneys and ureters.<sup>1</sup>

The onset of symptoms of HWWS usually occurs in the teenage years, shortly after menarche. Common presenting symptoms of HWWS are progressive cyclic pelvic pain

and a pelvic mass, leading to diagnosis of the condition in the majority of cases. The pelvic mass containing menstrual materials in the obstructed hemivagina, so-called hematocolpos, can be complicated with an ascending infection, leading to pyocolpos and mucopurulent vaginal discharge.<sup>1</sup>

There have been a handful of cases of HWWS with pyocolpos diagnosed during gestation which is considered a harmful condition.<sup>6–10</sup> Herein, we report a favorable pregnancy outcome of HWWS complicated with pyocolpos during pregnancy. Additionally, a short literature review of such cases focusing on presentations, diagnosis, management, and outcomes is provided.

## 2 | CASE PRESENTATION

A 33-year-old primigravida presented to our service at 17 weeks of gestation with a complaint of right pelvic pain and purulent vaginal discharge. She had been diagnosed 1 week earlier in another hospital, with septic abortion and had been treated with intravenous antibiotics including a suggestion of termination of the pregnancy.

The patient's past medical history was unremarkable. She had menarche at 13 years, with mild dysmenorrhea. Her menstruation cycles were regular and without intermenstrual bleeding.

Physical examination revealed a tense and tender cystic mass in her right pelvic region. The uterine height was deviated to the left and corresponding to the 17th gestational week. On pelvic examination, a single vagina with purulent discharge passing through a cervix deviated to the left was observed. The right lateral vaginal wall was bulging, with marked tenderness and without an opening identified. Bimanual pelvic palpation revealed an 8 × 8 cm tense and painful cystic mass in the right pelvic cavity.

Transabdominal and transvaginal ultrasound scans revealed a double uterus and a normally developed fetus consistent with 17 week of gestation residing in the left uterus. The right uterus appeared normal in size, with smooth endometrial lining through its cervix. A 6 × 6 cm mass of mixed echogenicity was confirmed at the right pelvic cavity. Both

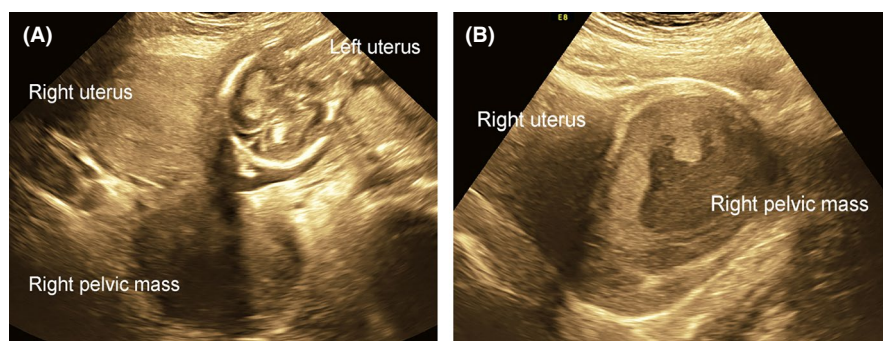
ovaries appeared normal. Uterine didelphys and an obstructed right hemivagina with pyocolpos were diagnosed (Figure 1).

Magnetic resonance imaging (MRI) of lower abdomen demonstrated the presence of two separate uteri, cervixes, and vaginas; obstructed right hemivagina with fluid-filled; and agenesis of right kidney, with compensatory hypertrophic left kidney (Figure 2).

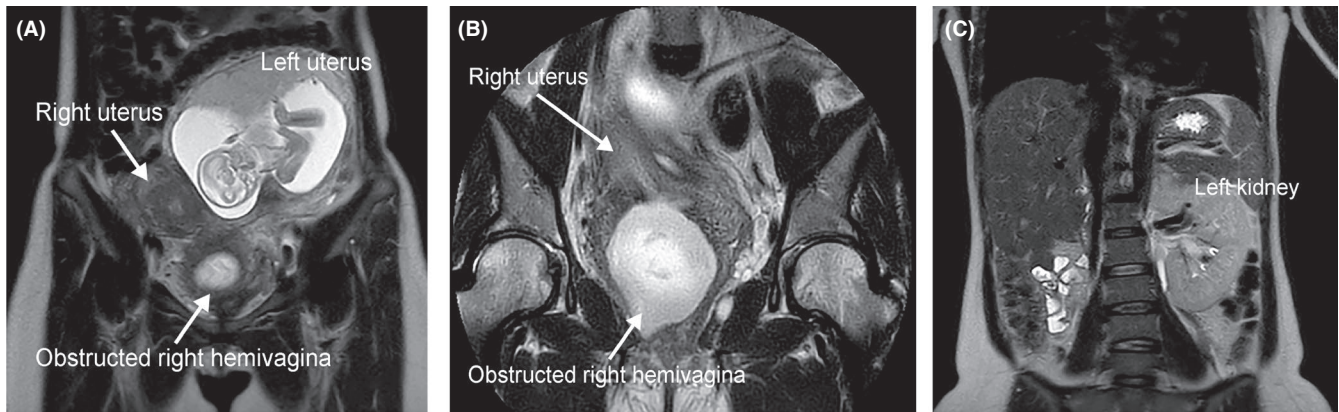
Results of blood tests were as follows: hemoglobin 11.5 g/dL, white blood cell count 14 100 cells/cu.mm with 85% segmented neutrophils, ESR 48 mm/h, and CRP 3.7 mg/L. Renal and liver function tests were unremarkable.

A needle aspiration for drainage of the pyocolpos was performed by a transvaginal ultrasound-guided puncture technique using a high-frequency vaginal sector transducer equipped with a needle guide (16 gauge). A 100 mL of foul-smelling pus-like discharge was aspirated from the most protruding part of the vaginal wall. The patient was relieved of her symptoms after aspiration. At 48 hours after the first drainage, a transabdominal ultrasound revealed a re-accumulation of fluid in the right hemivagina with pocket measuring 2 × 2 cm. On day 5 after the aspiration, the size of the pelvic mass was increased to 3.5 × 3 cm, without clinical deterioration. Gram stain revealed mixed organisms including gram-positive cocci and bacilli and few gram-negative bacilli. Acid-fast and modified acid-fast stained samples were negative. Bacterial cultures showed *Fusobacterium nucleatum* and *Actinomyces radingae*. The plans at that time were to provide conservative treatment and continue metronidazole for 10 days and amoxicillin for 6 months, according to bacterial susceptibility tests. The patient was discharged home on day 6th after the drainage.

An ultrasound follow-up at 2 weeks later demonstrated the pelvic mass which was not tender but increasing in size, measuring 6 × 6 cm. At 23 weeks of gestation, a transabdominal ultrasound was performed and showed the tense pelvic mass measured 9.2 × 9.3 × 8.6 cm. Partial excision of the vaginal septum was performed, draining of ≈1000 mL of foul-smelling blood-tinged discharge. A 9-day course of intravenous ceftriaxone and metronidazole were administered in addition to the prolonged treatment with oral amoxicillin. Follow-up ultrasounds in a week later and at every 2–3 weeks afterward revealed no fluid recollection in the pelvic cavity.



**FIGURE 1** Axial sonogram at 17 wk of gestation. A, a single viable fetus in the left uterus. B, a mixed echogenic pelvic mass next to the right uterus



**FIGURE 2** T2-MRI of the abdomen at 17 wk of gestation. A, the gravid left uterus with a residing fetus. B, right uterus connected to the fluid-filled pelvic mass at the level of upper part of the right vagina, consistent with an obstructed right hemivagina with a pyocolpos. C, presence of one hypertrophic kidney on the left only, confirming right renal agenesis

**TABLE 1** Characteristics of six cases of uterine didelphys with coexisting pyocolpos diagnosed during pregnancy

Case	1	2	3	4	5	6
Age (y)	18	23	33	27	16	33
GA (wk)	9	10	11	27	18	17
Classical presentations <sup>a</sup>	Yes	Yes; vaginal bleeding	Yes	Yes; vomiting	Yes; lumbar pain	Yes
Previous misdiagnosis	Septic abortion, ectopic pregnancy	PID	None	Unclear	None	Septic abortion
Connection between uteri	Isthmus	Vaginal septum	NA	NA	NA	Isthmus
Hemivagina	Rt.	Rt.	Lt.	Rt.	Rt.	Rt.
Single kidney	No	Yes	No	No	Yes	Yes
Pathogens	<i>Streptococcus</i> and <i>Klebsiella species</i>	NA	<i>Pediococcus species</i>	NA	<i>Staphylococcus hemolyticus</i>	<i>Fusobacterium</i> and <i>Actinomyces species</i> <sup>b</sup>
Treatment	Conservative treatment at 9-wk GA; incision and drainage at 21-wk GA	Excision of vaginal wall and drainage	Resection of vaginal wall and continuous drainage by Foley catheter insertion	Incision and drainage	Repeated needle aspirations	Needle aspiration at 17-wk GA; partial excision of vaginal wall and drainage at 23-wk GA
Time to complete resolution (days) <sup>c</sup>	140	5	7	Few	Unclear	49
Pregnancy outcome	Term	Term	Term	Term	PPROM at 36-wk GA	Term
Mode of delivery	C/S, elective	Vaginal delivery	C/S, elective	Vaginal delivery	C/S, obstructed labor	C/S, malpresentation
Reference	Karpathios et al 1992 <sup>6</sup>	Rana et al 2008 <sup>7</sup>	Park et al 2013 <sup>8</sup>	Sahu et al 2015 <sup>9</sup>	Albulescu et al 2018 <sup>10</sup>	Present case

Abbreviations: C/S, cesarean section; GA, gestational age; Lt., left; NA, not available; PID, pelvic inflammatory disease; Rt., right.

<sup>a</sup>Classical presentations include purulent vaginal discharge, pelvic pain, and mass.

<sup>b</sup>*F nucleatum* and *A radingae*.

<sup>c</sup>Resolution of the pyocolpos.

Due to a breech presentation, an elective cesarean section was done at 38 weeks of gestation, resulting in a birth of a male infant, with birth weight 3370 grams and no neonatal complication. Intraoperatively, the uterine didelphys with minimal intrapelvic adhesion was observed. There was no maternal complication and the patient was discharged on day 4.

Literature review of the cases with uterine didelphys diagnosed with coexisting pyocolpos during gestation revealed only five patients, as detailed in Table 1.<sup>6-10</sup> Of them, only two cases had ipsilateral renal agenesis, or confirmed to be HWWS.<sup>7,10</sup> Including our case, all were primigravida and conceived naturally without any medical or surgical history. The mean age at presentation was  $25 \pm 7.3$  (range 16-33) years and the mean age at menarche was  $14 \pm 0.7$  years. The median duration between menarche and presentation was 13.5 (range 2-20) years. The mean gestational age at presentation was  $15.3 \pm 6.8$  (range 9-27) weeks. Half of the cases presented in the first trimester and the remainder in the second trimester. The most common clinical presentations were pelvic pain, pelvic mass, and purulent vaginal discharge. Among patients with available data, a communication of two uteri was demonstrated, including a fistula at the isthmus or a small opening on the vaginal septum. The obstructed hemivagina predominantly occurred on the right side, at 83.3% of the cases (5/6). Five out of six patients (83.3%) gave deliveries at term. Cesarean section was the most frequent mode of delivery, accounting for 66.7% (Table 1).

### 3 | DISCUSSION

The clinical presentations of uterine didelphys and pyocolpos in the study pregnancies are similar to those observed in nonpregnant women. The mean age at diagnosis in affected pregnant women is late, at 25 years, which is similar to the nonpregnant women with incomplete obstructed hemivagina.<sup>1</sup>

According to a recent classification based on the degree of obstruction, HWWS is categorized into two types, including class 1: completely obstructed hemivagina (30% of cases) and class 2: incompletely obstructed hemivagina (70%) which is subdivided according to the level of connection between the two uteri, that of via vaginal septum (class 2.1) and the other at isthmus (class 2.2).<sup>11</sup> The onset of clinical manifestations is much later in women with incompletely obstructed hemivagina and purulent/bloody vaginal discharge can be the chief complaints.<sup>11</sup> The obstructed hemivagina and ipsilateral renal agenesis are preferential on the right side in 60% of cases.<sup>1,11</sup>

The present case represented a late diagnosis, 20 years after menarche. We hypothesize that during the monthly cycle, the patient's menstrual materials could overflow from the obstructed hemivagina to the contralateral cervical passage, not resulting in a hematocolpos and that the retained

menstrual substances were adequately resorbed between the periods. However, throughout the pregnancy, the material excreted from the decidua was multiplied beyond its usual resorption capacity and an increasing size of the gravid uterus led to an anatomical distortion and narrowing of the passage connecting the two uteri, leading to formation of the hematocolpos and eventually pyocolpos when it became complicated with ascending infection. As the gravid uterus grew out of the pelvic cavity, the tightened passage reopened, allowing overflows of the retained content in the obstructed right hemivagina to the contralateral cervix and resulting in purulent vaginal discharge.

Without a diagnosis of HWWS prior to a gestation, establishing a diagnosis of HWWS coexisting with pyocolpos during pregnancy can be difficult and complicated by misdiagnosis, such as septic abortion, ectopic pregnancy, and pelvic inflammatory disease, as noted in 4 out of five cases (Table 1).<sup>6,7,9</sup> Usually, ultrasonography provides sufficient information for establishing diagnosis of Müllerian duct anomalies (MDAs) including HWWS. However, MRI, with multiplanar image acquisition, can provide more details of the complex abnormalities and echogenicity of tissues which can be useful for subclassification of the MDAs and differentiating blood from pus collected in the obstructed hemivagina.<sup>1,11</sup> The information obtained from MRI would be useful for planning of medical management. Based on the data of six case detailed here, HWWS coexisting with pyocolpos was successfully diagnosed by using transabdominal ultrasonography alone, though transvaginal ultrasound and/or MRI were shown to provide more accurate information and associated renal anomalies.<sup>6-10</sup>

As for pathogens involved in the pyocolpos reviewed here, they were mixed microorganisms of commensal flora of human skin, mouth, digestive, and female urogenital tracts which were facultative or obligate anaerobic bacteria.<sup>6-10</sup> Interestingly, *Actinomyces radingae* was found to be an organism involved in causing pyocolpos of the present patient. *Actinomyces* species form fungus-like branched hyphae and infection with this organism demands a prolonged course of beta-lactam antibiotics for 6-12 months.<sup>12</sup> These data addressed the necessity of aerobic and anaerobic bacterial culture to identify causative organisms which can lead to appropriate treatment. Generally, *Actinomyces* species are known to cause opportunistic infection in immunocompromised hosts and women with the use of intrauterine devices.<sup>12</sup> Inadequate drainage of *Actinomyces* infection can lead to a chronic pelvic abscess and severe tissue fibrosis causing bowel adhesion and obstruction. We could not identify a predisposing factor related to *Actinomyces* infection of the present patient. To our knowledge, this is the first case of *Actinomyces* infection associated with obstructed hemivagina.

The most effective treatment for HWWS with clinical complications due to the obstructed hemivagina in nonpregnant

women is resection of vaginal septum as much as possible and drainage of the retained fluids.<sup>11</sup> This procedure is also considered safe for pregnant women.<sup>6</sup> Nonetheless, conservative treatment with antibiotics combined with a less invasive procedure of needle aspiration was attempted in three (cases 1, 5, 6) out of six cases due to various reasons and demonstrated that it was not efficient for adequate drainage of pus. These measures could unnecessarily increase risk of complications because of multiple general anesthesia and procedure-related problems such as preterm labor. It also resulted in prolonged course of procedural intervention and hospital stay (Table 1) and complicated pelvic fibrosis due to delayed drainage of the pus collected. These data suggest the benefit outweighing the risk of excision of the vaginal septum and adequate drainage of pus in pregnant women with HWWS-associated pyocolpos.

Of note, we first demonstrate the utility of transvaginal ultrasound-guided approach, a technique commonly used for oocyte retrieval in an in vitro fertilization unit, that it can be used safely for needle aspiration and drainage of fluid collected in the obstructed vagina. This procedure is considered safe and less invasive with an advantage of real-time visualization under ultrasound guidance and it can be done under local anesthesia. However, a successful drainage would depend on nature of the retained fluid.

Concerning fertility, pregnancy rate in women with uterine didelphys is reduced, at 85% and most of the pregnancies (64%) occur in the uterus contralateral to the obstructed hemivagina.<sup>3</sup> Successful pregnancy outcomes in HWWS have been reported at 57%-68%, while abortion rates was noted at 30%-33%, perinatal mortality rate of 3.6%, preterm labor at 21%-29%, and increased incidence of fetal malpresentation.<sup>4,13,14</sup> Fifty-four percent (15/28) of women with HWWS conceived and delivered their first babies at term prior to surgical treatment for obstructed hemivagina.<sup>1</sup> The present patient and the cases reviewed demonstrated successful pregnancy outcomes.

## 4 | CONCLUSION

The presence of pelvic pain, a pelvic/paravaginal mass, and purulent vaginal discharge in primigravida should raise the possibility of obstructed hemivagina and uterine didelphys. Though conservative management could result in successful pregnancy outcomes, early excision of vaginal septum and adequate drainage offer a shorter course of management and complication avoidance.

## CONFLICT OF INTEREST

The authors of this study have no conflicts of interest to declare.

## AUTHOR CONTRIBUTIONS


CT: involved in study design, patient care, data collection and literature review, and manuscript preparation. WD, SD, YT and CS: provided patient care and/or surgical management and clinical data. DW: performed critical revision of the manuscript. All the authors reviewed the manuscript.


## ETHICAL APPROVAL

The study was approved by the Ramathibodi Hospital Institutional Review Board (COA. MURA2019/1076 and COA. MURA2020/536) and complied with the Declaration of Helsinki. Written informed consent was obtained from the patient for publication of this case report and the images shown.

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