

# Unexplained Abdominal Pain and Sepsis in Herlyn-Werner-Wunderlich Syndrome: A Diagnostic Challenge

## Objectives

1. To describe a rare and severe presentation of **Herlyn-Werner-Wunderlich (HWW) syndrome** complicated by pelvic abscess, tubo-ovarian abscess, and sepsis
2. To highlight diagnostic challenges and delays in care associated with obstructive Müllerian anomalies
3. To emphasize the importance of early surgical correction and multidisciplinary management in preventing life-threatening complications
4. To review potential infectious and anatomic sequelae of untreated vaginal septum in patients with HWW syndrome

## Introduction

- Herlyn-Werner-Wunderlich (HWW) syndrome is a rare congenital Müllerian duct abnormality (MDA) characterized by uterine didelphys, obstructed hemivagina, and ipsilateral renal anomalies.

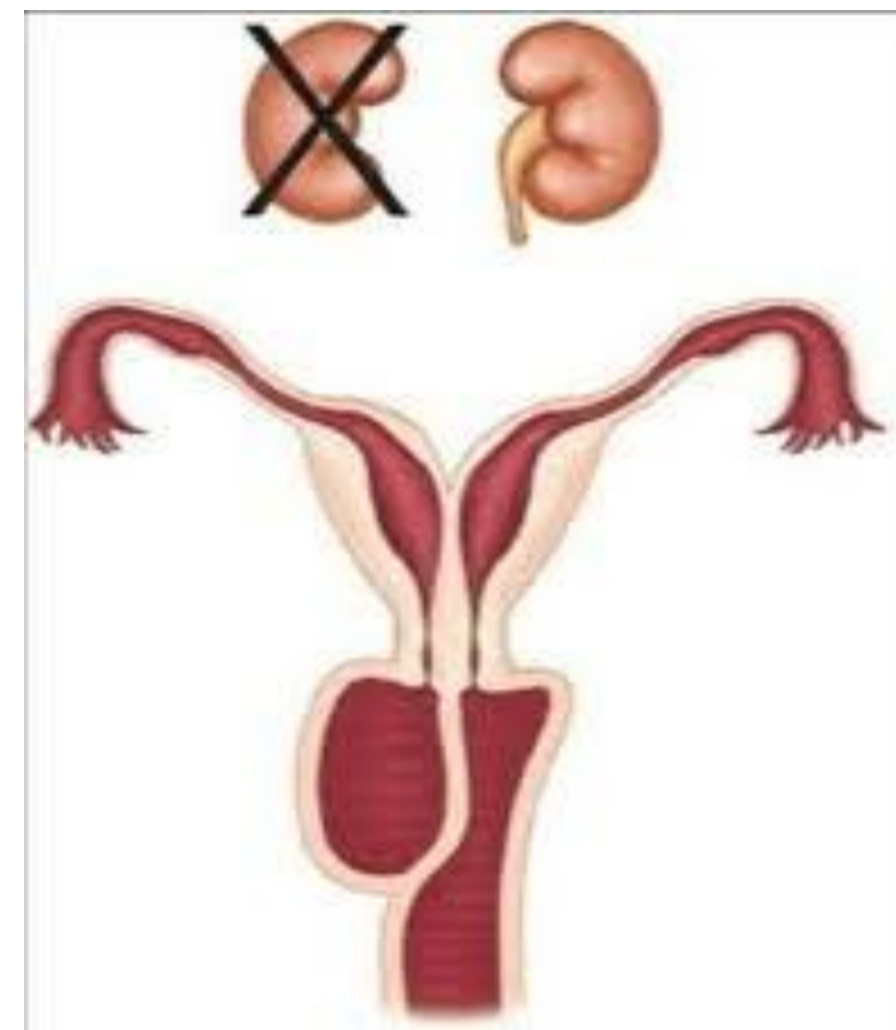


Figure 1: Schematic of HWW syndrome<sup>2</sup>

- The syndrome is classified based on the level of vaginal obstruction:
  - Class 1: completely obstructed hemivagina
  - Class 2: Incompletely obstructed hemivagina
- Typical presentation can include:
  - Cyclic pelvic or lower abdominal pain (worsens with menses)
  - Progressive dysmenorrhea
  - Tubo-ovarian abscess/pelvic abscess
  - Endometriosis
  - Infertility or recurrent pregnancy loss
  - Acute abdomen or sepsis (rare but severe)
- Diagnosis of HWW is often made through ultrasonography and confirmed via MRI.
- Early diagnosis is crucial to reduce complications. Menstrual suppression with OCPs can help prevent accumulation and obstruction.
- Long-term complications can include, but are not limited to, the following:
  - chronic pelvic pain
  - endometriosis
  - pelvic adhesions from retrograde menstruation

## Case Description

A 21-year-old female G1P0010 with a known history of HWW syndrome presented with severe left lower quadrant abdominal pain for three days. During her first year of life, she underwent a left nephrectomy for a poorly functioning polycystic kidney. Also, she was treated one year prior for a chlamydia infection.

She had previously been evaluated for similar complaints of left lower quadrant abdominal pain twice in the past four months. Unfortunately, due to insurance issues, she had not been able to obtain vaginal septum resection.

On physical exam, the rupture site revealed copious purulent, foul-smelling discharge and scant vaginal bleeding. MRI revealed HWW anatomy, left pelvic abscess 12cm in size, and likely reactive lymph nodes. She was admitted for a tubo-ovarian abscess (TOA) and started on ceftriaxone, doxycycline, and metronidazole.

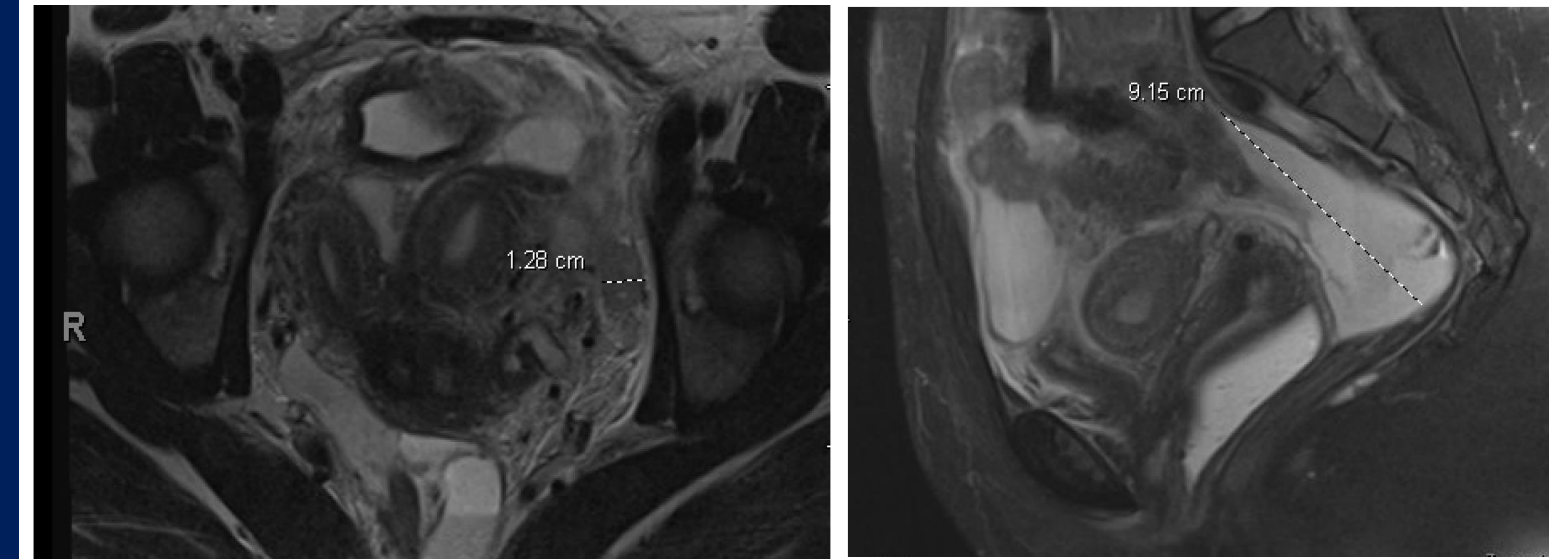
The following day, she underwent incision and drainage of the left hemivagina and pyometrocolpos, with partial vaginal septum resection. The surgery led to some immediate pain improvement. On post-operative day 1, she acutely decompensated with persistent tachycardia to 130 beats per minute, tachypnea, fever (38.5 °C), new oxygen requirement with pleuritic chest pain. Her CTA and echocardiogram were negative. Internal medicine recommended a transition to broader antibiotics (vancomycin and piperacillin-tazobactam). CT abdomen and pelvis showed a persistent 12cm pelvic abscess, which was subsequently drained by interventional radiology, yielding 110mL of purulent fluid. The pelvic abscess fluid grew *Staphylococcus epidermidis*.

After some improvement over the next three days, her condition worsened, accompanied by a new fever, leukocytosis (WBC 24,000 cells/ $\mu$ L), and a platelet count of 953,000/ $\mu$ L. The patient's abdominal pain worsened, with concerns for peritonitis. An abdominal x-ray was non-diagnostic.

Due to suspected peritonitis, persistent fluid collections on imaging, and failure to improve on antibiotic therapy, a second surgery was recommended. This was two weeks following the first surgery with Gynecologic Oncology staffing. The patient underwent a diagnostic laparoscopy that was converted to an exploratory laparotomy, ileocecectomy (10 cm removed by general surgery), left salpingo-oophorectomy, and lysis of adhesions. Intra-operative findings included a perforated appendicitis, pelvic abscess, TOA, uterine didelphys, and severe adhesions throughout and extending to the liver capsule—infected disease consult post-operatively suspected Fitz-Hugh-Curtis syndrome.

The patient did well and was discharged five days after surgery. Two months later, the patient noted resolved abdominal pain, regular cycles on oral contraceptive pills, and denied vaginal discharge nor other symptoms.

## MRI Imaging



MRI Images of pelvic abscess.

## Discussion

- Pelvic inflammatory disease, TOA, and sepsis are uncommon but serious sequelae of untreated vaginal obstruction in HWW syndrome.
- Diagnostic complexity is further increased by overlapping gastrointestinal and gynecologic symptoms, prior pelvic infections, and limited access to surgical care.
- Early definitive surgical management of the obstructing vaginal septum is critical in preventing recurrent infections, abscess formation, and long-term morbidity.
- This case reinforces the need for prompt diagnosis of Müllerian anomalies, removal of barriers to surgical care, and coordinated multidisciplinary management when patients present with unexplained abdominal pain and sepsis.
- Increased awareness of these rare but serious complications may facilitate earlier intervention and improved outcomes in similar patients.

## References

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