

# Herlyn-Werner-Wunderlich Syndrome: Uterus Didelphys and Obstructed Hemivagina with Unilateral Renal Agenesis

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**H**erlyn-Werner-Wunderlich syndrome is a rare congenital Mullerian ductal anomaly characterized by the triad of obstructed hemivagina, uterus didelphys, and unilateral renal agenesis [1]. This anomaly is generally observed in post-menarche adolescents and young women with irregular menses, dysmenorrhea, abdominal pain, and pelvic mass [2,3]. The diagnosis may be difficult due to the infrequency of this syndrome, and a high index of suspicion is required. Early detection is important as immediate surgical resection of the obstructing

vaginal septum can provide pain relief and prevent further complications [4,5]. In young adolescent girls where postponing surgery is warranted, a continuous pill regimen of GnRH analogues may be used to maintain amenorrhea. We describe the case of Herlyn-Werner-Wunderlich syndrome in a young woman with computed tomography imaging findings.

A 20 year old woman with no medical or surgical history was admitted to hospital because of lower abdominal pain and urinary retention. The patient reached menarche at age 12 and had a medical history of hypomenorrhea and dyspareunia. Physical examination revealed a large bulge on the right lateral wall of the vagina and a palpable pelvic mass. Routine laboratory tests were unremarkable. Ultrasound scan and CT scan revealed a large hematocolpos and a double uterus.

The right hemiuterus was enlarged due to hematometra, while the left hemiuterus was rudimentary [Figure 1]. In addition, renal agenesis of the right kidney with compensatory hypertrophy of the left kidney was identified [Figure 2].

At examination under general anesthesia, an obstructed right hemivagina was identified with a longitudinal vaginal septum. A vaginal septectomy was performed and 1000 ml of chocolate-colored fluid was drained. Right and left cervixes were palpable. The diagnosis of Herlyn-Werner-Wunderlich syndrome was made based on imaging and the clinical presentation.

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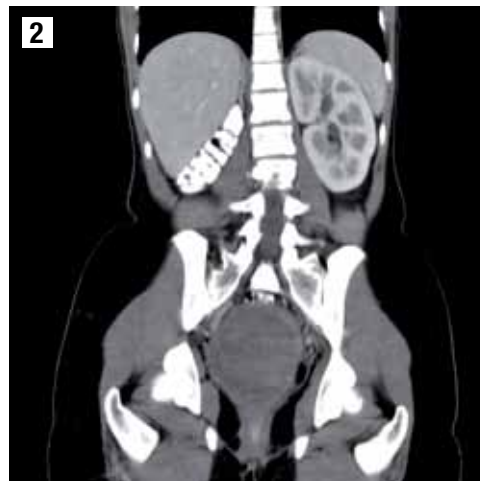
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**Figure 1.** Coronal CT image demonstrates a complete view of the pelvic anatomic defects: enormously dilated hemivagina due to the presence of vaginal septum, mild dilatation of the right hemiuterus (hematometra) and a rudimentary left uterine cavity



**Figure 2.** Coronal CT image demonstrates the absence of right kidney and a compensatory hypertrophy of the left kidney. A large hematocolpos due to the obstructed right hemivagina is observed

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