

## Surgical approach of the OHVIRA-like syndrome: a didactic video

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### Abstract

Introduction.

Herlyn-Werner-Wunderlich (HWW) syndrome, also known as OHVIRA (obstructed hemivagina and ipsilateral renal anomaly) syndrome, is a rare congenital urogynecological malformation, typically characterized by the triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis. The multiple anatomical variations of this syndrome challenge the prompt diagnosis and the therapeutic approach. Surgical treatment may not always be clear until completing diagnostic with endoscopic exploration. Case presentation. An adolescent female, virgin, with a history of left renal agenesis was addressed to the Mother and Children Department with complaints of chronic abdominal pain, worsened in the last weeks, mainly irradiated to the left iliac fossa. Physical examination showed a painful abdomen and normal external genital organs. Abdominopelvic ultrasound and MRI revealed a uterus didelphys and suspected a blind left hemivagina with 8 cm presumed hematocolpos. The association with a

left renal agenesis was suggestive of an OHVIRA syndrome. A vaginoscopy followed by a laparoscopy were planned to complete the diagnostic. Unexpectedly, vaginoscopy didn't show either the hematocolpos, second hemivagina, or cervix, with no possibility then to drain it transvaginally. Laparoscopy revealed a probably obstructed left hemi-uterus associated with a large hematocervix and hematosalpinx; on the right side, a normal hemiuterus and adnexa with a functional right tube.

Peritoneal endometriosis was identified at multiple sites of the abdominal cavity. A left hemi-hysterectomy was decided intraoperatively, which is illustrated in this video as the surgical approach to a variant of HWW syndrome classified by Zhu L. 2015 as cervicovaginal atresia without communicating uteri (OHVIRA type 1.2.). An extensive adhesiolysis to progressively "enucleate" the dilated cervix from the surrounding peritoneum was performed until the complete resection of the rudimentary left uterus and tube without

complications. Desogestrel-only pill was then started to treat endometriosis and regular follow-up was established. Conclusion. HWW syndrome may be suspected in young patients with abdominal pain, a pelvic mass (suggestive of hematocolpos) and renal agenesis. Early diagnosis and timely proper management of this

**Keywords:**

syndrome are crucial as they would prevent urogynecological complications such as endometriosis, infection, and infertility. Even if a conservative approach is recommended, some anatomical variants may require a hemihysterectomy.

Benign Gynecology, Gynecologic Surgical Procedures, Herlyn-Werner-Wunderlich (HWW) syndrome, Laparoscopy, Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome

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