

Mapping OHVIRA on MRI: Imaging Pearls for Uterus Didelphys with Obstructed Hemivagina and Renal Agenesis

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Clinical Image

OHVIRA (Obstructed hemivagina and ipsilateral renal anomaly), also called Herlyn-Werner-Wunderlich syndrome, is defined by the triad of a duplicated uterus, an obstructing longitudinal vaginal septum, and ipsilateral renal agenesis; it reflects the coupled embryologic development of the Müllerian and Wolffian systems in early gestation [1,2]. During weeks ~6-12, the mesonephric (Wolffian) duct both induces the ureteric bud (future kidney/ureter) and serves as a guide for lateral migration/fusion of the paramesonephric (Müllerian) ducts; ipsilateral mesonephric duct maldevelopment can therefore produce renal agenesis and simultaneously disrupt Müllerian fusion, yielding a complete bicorporeal uterus, while failure of canalization or resorption of the distal Müllerian/vaginal segment leaves a longitudinal septum that may be obstructing [1,2,9]. In contemporary reporting, the uterine, cervical, and vaginal compartments are coded separately using the ESHRE/ESGE system; the classic OHVIRA configuration corresponds to U3b C2 V2 (complete bicorporeal uterus with double cervix and an obstructing longitudinal vaginal septum) [3]. Obstruction on one side leads to retained menstrual blood (hematocolpos/±hematometra), cyclical pain, and-when diagnosis is delayed-retrograde spill with an increased risk of endometriosis and infection, phenomena repeatedly described in reviews and series [1,4,10].

Although uncommon and likely underrecognized because menses continue through the unobstructed side, reported incidence figures across heterogeneous cohorts range from approximately 0.1% to 3.8% [1,5]. In a systematic review of 734 patients, uterus didelphys accounted for ~83% of uterine configurations, ipsilateral renal agenesis for ~92%, the obstructed side was left in ~51%, and surgically confirmed endometriosis was present in ~14% [4].

MRI is preferred for mapping OHVIRA because it simultaneously depicts uterine duplication, the obstructing septum, and hemorrhagic complications with excellent soft-tissue contrast [2,6,7,9]. Typical appearances include two separate uteri with two cervices and preserved zonal myometrium, precise delineation of the obstructing longitudinal septum enabling standardized coding (U3b C2 V2), and a distended hemivagina containing blood products-hematocolpos that is hyperintense on T1-weighted images with variable T2 "shading"; hematometra and/or hematosalpinx may coexist depending on the level and completeness of obstruction [2,6,7,9]. A practical MR protocol uses high-resolution T2-weighted images in sagittal, axial, and coronal planes across the uterus and vagina to define septal length, insertion, and laterality; T1-weighted sequences (± fat suppression) confirm hemorrhagic content; contrast-enhanced and diffusion-weighted images are reserved for suspected complications such as infection or abscess [2,7,9]. Because Müllerian and Wolffian anomalies co-occur, the same examination should evaluate the urinary tract to confirm ipsilateral renal agenesis and identify ureteral variants (e.g. ectopic or blind-ending ureter), which MRI depicts well [6,8].

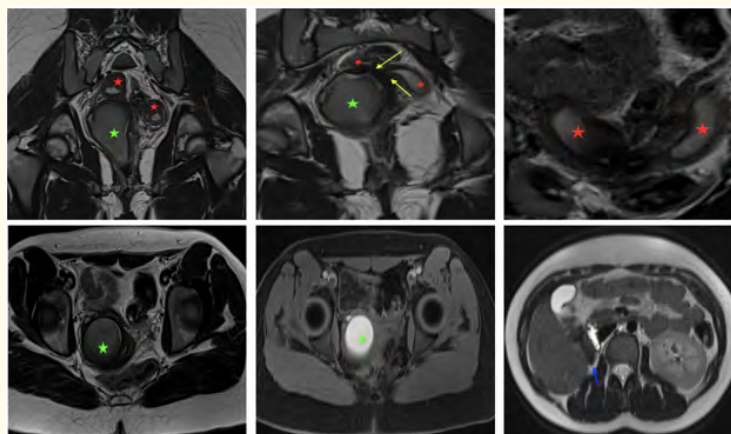


Figure 1: Pelvic MRI in a 14-year-old girl, menarche 8 months earlier, presenting with dysmenorrhea. The study shows two uterine cavities (red asterisks) with two cervixes (yellow arrows) and a vagina divided by a right-sided obstructing longitudinal septum, causing hematocolpos (green asterisk). Right renal agenesis is also present (blue arrow). Overall, the findings are consistent with a complete bicorporeal uterus (U3b), double cervix (C2), and an obstructing longitudinal vaginal septum (V2) - an OHVIRA configuration - complicated by right-sided hematocolpos.

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Conflict of Interest

The authors declare no conflict of interest.

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