

# Herlyn–Werner–Wunderlich Syndrome-Early Diagnosis with Ultrasonography in a 3-month-old Female Child

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## Abstract

Herlyn–Werner–Wunderlich syndrome is an uncommon, complex uterine anomaly - obstructed hemivagina with ipsilateral renal agenesis, usually presenting in adolescence. Here we report a rare presentation of this entity in a 3-month-old child, diagnosed on transabdominal ultrasonography alone. The patient underwent surgery wherein imaging findings were confirmed, and vaginoplasty was done. We present this case to highlight the role of ultrasonography in diagnosing this condition even though magnetic resonance imaging is the most accurate imaging modality in diagnosing this condition; and also the early age of presentation which is unusual. We describe the findings on ultrasound along with a review of the literature on Herlyn–Werner–Wunderlich syndrome.

**Key words:** Hemivagina, Ipsilateral renal agenesis, Mullerian anomalies, Obstructed hemivagina with ipsilateral renal agenesis syndrome, Uterus didelphys

## INTRODUCTION

Herlyn–Werner–Wunderlich syndrome is a rare Mullerian duct and Wolffian duct anomaly with uterus didelphys, unilaterally obstructed hemivagina, and ipsilateral renal agenesis (OHVIRA).<sup>1</sup> Due to functional patency of hemivagina and normal menstrual cycles, this type of obstructive anomaly initially remains unrecognized, usually presenting at puberty with recurrent pelvic pain and/or mass due to hematocolpos or hematometra. Moreover, the menstrual flow that comes from the patent hemivagina resembles normal menses leading to delay in diagnosis and surgery.<sup>2,3</sup> Developmental abnormalities of Mullerian duct have an estimated incidence of 1.1-3.5% among women.<sup>4,5</sup>

## CASE REPORT

The 3-month-old child was presented with a swelling at the introitus noticed by the mother; there were no other complaints such as pain, fever, or symptoms suggestive of urinary tract infection. She was referred for an ultrasound examination which revealed a bicornuate uterus with separate cervices (bicornis bicornis uterus), and an anechoic collection or cyst in the vagina. There was no evidence of hydrometra or hydrosalpinx. The right kidney was not visualized. Left kidney showed normal morphology (Figures 1-4).

On examination under anesthesia, the cyst revealed to be obstructed hemivagina filled with clear fluid. It was drained, and the septum excised. Further examination showed two cervices suggestive of uterus didelphys (Figures 5 and 6).

## DISCUSSION

Herlyn–Werner–Wunderlich syndrome comprises of obstructed hemivagina and ipsilateral renal agenesis and

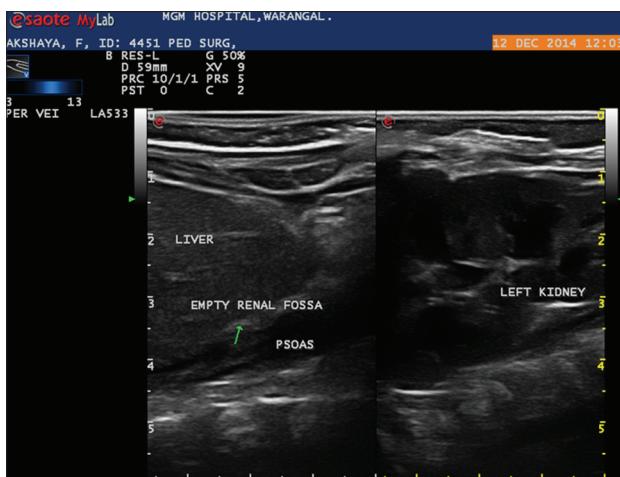
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**Figure 1:** Sagittal image showing empty right renal fossa.  
Compare with normal left kidney



**Figure 2:** Sagittal image showing fluid or cyst in vagina



**Figure 3:** Axial section at level of fundus showing bicornuate uterus

was first reported in 1922. It is more recently known by the acronym OHVIRA and commonly associated with uterine didelphys or rarely septate uterus.<sup>6</sup> The incidence



**Figure 4:** Axial section at level of cervix showing two cervices



**Figure 5:** Bluish cystic swelling at introitus



**Figure 6:** Obstructed hemivagina and patent hemivagina under anesthesia

of OHVIRA syndrome is very rare, and only isolated case reports have been published.<sup>4</sup>

Female reproductive tract develops both from paramesonephric and mesonephric ducts.

The OHVIRA syndrome occurs due to the developmental arrest of ipsilateral Wolffian duct resulting in developmental failure of distal hemivagina leading to obstructed hemivagina. The uterine didelphys commonly associated with OHVIRA syndrome occurs due to failure in the fusion of paired Mullerian ducts.<sup>4</sup> The female genital and urinary tracts have a similar embryological origin from the intermediate mesoderm, which is responsible for the increased association of their anomalies. Ipsilateral renal agenesis is a common urologic anomaly seen in OHVIRA syndrome.<sup>7,8</sup>

Typically, a patient with this rare condition presents after menarche with non-specific symptoms of recurrent pelvic pain with vaginal or pelvic mass. However in our case the patient presented at a very early age, which is very rare.<sup>9</sup> The pelvic examination may show a bulging vaginal mass, but sometimes the mass may be small and difficult to determine.<sup>10</sup> In general, ultrasonography is the initial screening tool in cases of suspected female genital tract anomalies. However, magnetic resonance imaging (MRI) is the most accurate diagnostic method as it better depicts uterine anatomy and has excellent tissue characterization, which play a crucial role in surgical planning.<sup>1,11</sup> In our case, however, ultrasonography alone was reliable in diagnosing this rare anomaly with proper delineation of uterine contour. The sonographic findings were clinically and peroperatively corroborated.

Mullerian agenesis and renal agenesis often coexist in Herlyn syndrome, so it is important to look for urinary tract anomalies in all cases of Mullerian duct anomalies.<sup>12</sup> Genitourinary anomalies can be associated with other abnormalities such as an atrial septal defect, coarctation of aorta, and spinal anomalies. Hence, a complete physical examination and relevant investigations are necessary in all patients.<sup>13</sup> Ipsilateral renal agenesis is found in 81% cases of OHVIRA syndrome and 25-50% show genital anomalies.<sup>14</sup>

The classic treatment is resection of the vaginal septum to relieve the obstructed hemivagina. Hemihysterectomy is no longer preferred, as the reported incidence of pregnancy is similar with hemihysterectomy and transvaginal repair. A successful pregnancy occurs in 87% of the patients with OHVIRA syndrome.<sup>10,12,13,15</sup>

## CONCLUSION

OHVIRA syndrome is an uncommon congenital anomaly with clinical significance and simple surgical management. Imaging, particularly MRI plays an important role in diagnosis. However, an initial screening ultrasound can sometimes diagnose it as in our case. An early correct diagnosis is a goal to relieve the symptoms and prevent complications caused by retrograde menstruation resulting in endometriosis and in preserving sexual and conceptional abilities.

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