

## Herlyn Werner Wunderlich Syndrome

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### ABSTRACT

Uterovaginal duplication with obstructed hemivagina and ipsilateral renal agenesis is referred to as the Herlyn Werner Wunderlich (HWW) syndrome. Here, we describe a 15 year old female patient with HWW syndrome, who was taken up for a diagnostic laparoscopy followed by vaginal septal resection. In addition to a definitive diagnosis, this approach helped in symptomatic relief to the patient. Prompt and accurate diagnosis and treatment of HWW syndrome can significantly prevent future complications.

**Keywords :** Herlyn werner wunderlich syndrome, laparoscopy, vaginal septal resection

### INTRODUCTION

Herlyn-Werner-Wunderlich (HWW) syndrome is a very rare congenital anomaly of the urogenital tract involving Müllerian ducts and Wolffian structures, and it is characterized by the triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis.<sup>[1]</sup> Combination of obstructed hemivagina and uterus didelphys was first reported in 1922.<sup>[5]</sup> The triad was reported in 1971 by Herlyn and Werner and again in 1976 by Wunderlich. The incidence of didelphys uterus, related to HWW, is approximately 1/2,000 to 1/28,000, and it is accompanied by unilateral renal agenesis in 43% of cases. The incidence of unilateral renal agenesis is 1/1,100 and 25-50% of affected women exhibits associated genital abnormalities.<sup>[2]</sup> A complete or partial vaginal septum is present in 75% of women with didelphys uterus.<sup>[3]</sup> The exact cause, pathogenesis and embryologic origin of HWW syndrome are unclear and remain a subject of discussion.<sup>[4]</sup>

### CASE REPORT

15 year old female patient presented with severe and increasing cyclical abdominal pain. She attained menarche 1 year back. The patient had irregular and scanty menstruation associated with dysmenorrhoea for the first 3 months, followed by 8 months of amenorrhoea. On physical examination, secondary sexual characters were well developed and on palpation lower abdomen tenderness was present. As the patient was unmarried and non cooperative, further examination was planned to be done under anaesthesia.

Routine investigations were normal. USG revealed absence of right kidney and a uterine didelphys with hematocolpos and also double endometrial echoes, and both ovaries were normal. MRI indicated a uterine-vaginal malformation consisting of uterus didelphys communicating with a double vagina, of which the right vagina was obstructed. There was a hyperintense

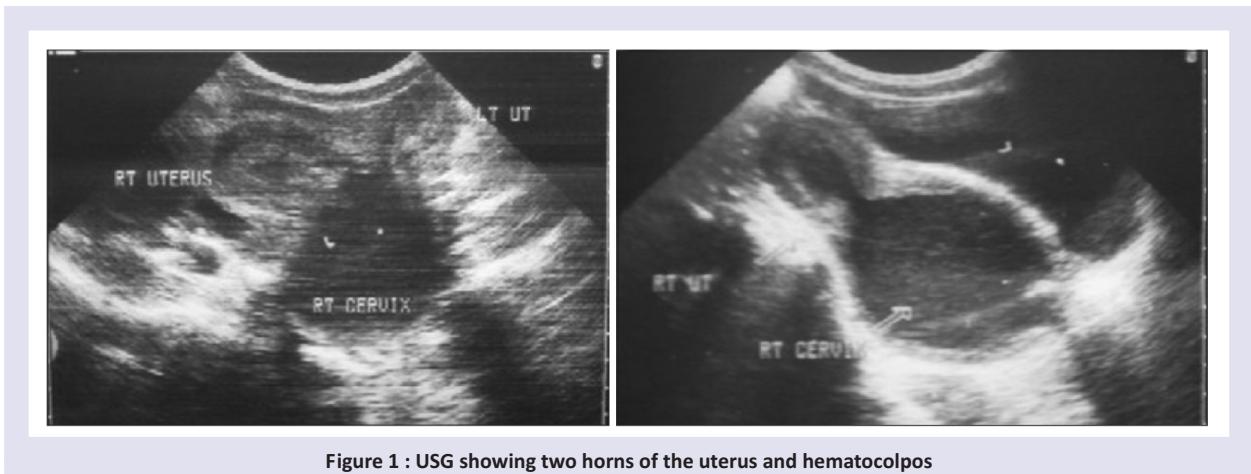


Figure 1 : USG showing two horns of the uterus and hematocolpos



Figure 2 : MRI showing absence of right kidney; and hematocolpos

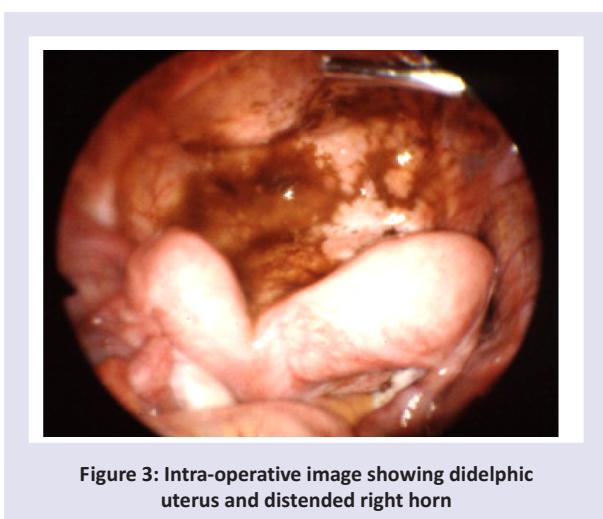


Figure 3: Intra-operative image showing didelphic uterus and distended right horn

collection of fluid both in the right uterus and right obstructed vagina suggestive of hematocolpos. Upper abdomen showed right renal agenesis. With this, the provisional diagnosis made as HWW Syndrome, the patient was posted for surgery. Under general anaesthesia per speculum examination revealed bluish bulging membrane at vagina and bimanual examination indicated a right-sided cystic pelvic mass, which was movable and on per rectal examination, suggested was a bulge s/o cystic mass.

**Diagnostic laproscopy findings :** Uterus didelphys seen. Right horn of the uterus had hematocolpos. Left horn appeared normal. Both tubes and ovaries were normal.

**Follow up :** Patient is asymptomatic and menstruating regularly after surgery.

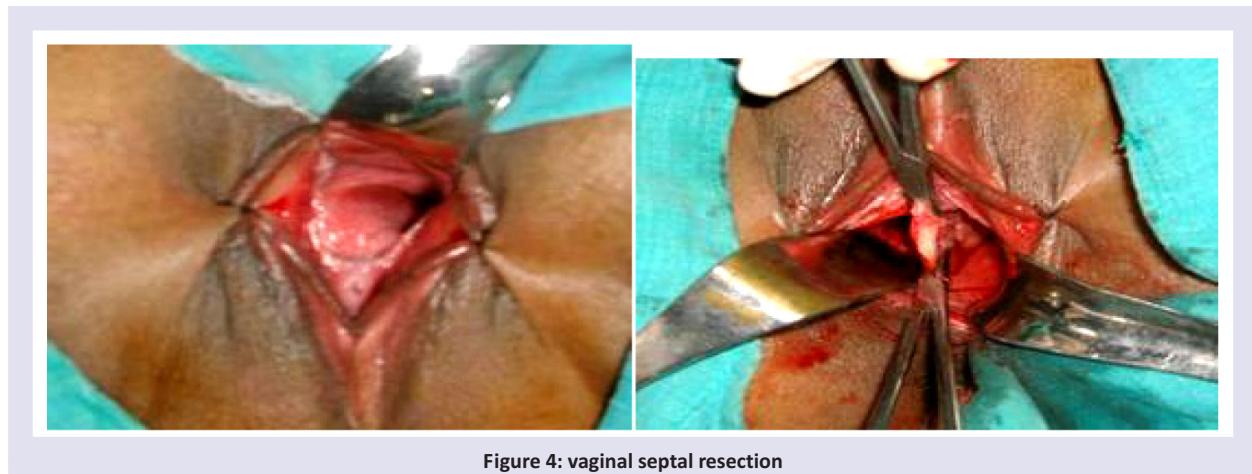


Figure 4: vaginal septal resection

## DISCUSSION

Mullerian (paramesonephric) duct anomalies are congenital anomalies of the female genital tract which result from non-development or non-fusion of the mullerian ducts or failed resorption of the uterine septum during the sixth to ninth weeks of fetal life causing a wide-ranging series of reproductive duct malformations.<sup>[6,7]</sup>

The paramesonephric ducts of the genetically female embryo fuse together in the midline and form the uterus, cervix and the upper four-fifths of the vagina. The lower 20% of the vagina are formed from sinovaginal bulbs which are protrusions of the urogenital sinus.<sup>[8]</sup> The urinary and genital systems arise from a common ridge of mesoderm arising along the dorsal body wall and rely on normal development of the mesonephric system. Hence, abnormal differentiation of the mesonephric and paramesonephric ducts may also be associated with anomalies of the kidneys.<sup>[8]</sup> Renal agenesis is the most common anomaly although horseshoe or pelvic kidney, cystic renal dysplasia, duplication of the collecting system and ectopic ureters have all been described.<sup>[8,9]</sup>

Renal agenesis is predictive of an ipsilateral obstructive Mullerian anomaly greater than 50% of the time.<sup>[9]</sup> These anomalies have a right-sided dominance, twice as often as on the left side.<sup>[10,11]</sup> Such relationship between female genital and urogenital anomalies should lead us to examine the urogenital system when a genital anomaly is identified and vice-versa.

HWW syndrome is usually discovered at puberty with non-specific symptoms, like increasing pelvic pain, dysmenorrhea and palpable mass due to the associated haematocolpos or hematometra, which result from retained, longstanding menstrual flow in the obstructed vagina. A right sided prevalence has been described. It is postulated that the right side is more susceptible to

hypoxic damage than the left side due to a precocious mitochondrial maturity on the left side, resulting in less tissue damage following hypoxia.<sup>[12]</sup>

If treatment is delayed, complications may develop, such as endometriosis caused by retrograde menstruation, infections and pelvic adhesions, which in turn might cause obstruction of the genital organs. Clinical suspicion and awareness of the syndrome are therefore imperative to making a timely diagnosis and preventing these complications.<sup>[13]</sup>

CT and ultrasound are the most widely used diagnostic tools.<sup>[13]</sup> However, MRI is considered to be more sensitive for imaging soft-tissue anatomy and delineating subtle findings seen in congenital anomalies. Hence, it should be obtained before any surgical intervention.<sup>[14]</sup> Laparoscopy is not mandatory but could be helpful in confirming the diagnosis when radiologic imaging is inconclusive, especially in those cases with endometriomas warranting resection.<sup>[15]</sup>

As obstructive genital lesions may be associated with other anomalies such as coarctation of the aorta, atrial septal defects and abnormalities of the lumbar spine, a complete physical examination and abdominal tests may be indicated.<sup>[14]</sup> Resection of the vaginal septum is the treatment of choice of obstructed hemivagina.<sup>[6]</sup>

Women with uterus didelphys have a reasonable chance of getting pregnant, but the abortion rate is high (74%) and premature delivery is common (22%).<sup>[16]</sup> A caesarean section is required in 82%.<sup>[11]</sup> Evaluation of the genital tract by means of MRI scanning is recommended in all girls with known renal abnormalities detected antenatally or thereafter, before the onset of menstruation. This enables us to diagnose some patients before menarche and carry out a surgical correction of the obstruction before any damage has occurred because of haematocolpos,

haematometra and retrograde menstruation.<sup>[11]</sup>

## CONCLUSION

The prompt and accurate diagnosis of female reproductive tract disorders including HWW syndrome, is necessary to prevent complications and preserve future fertility. Early recognition of this relatively rare syndrome would lead to the immediate, proper surgical intervention.

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## CONFLICT OF INTEREST:

None

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