

# UTERINE DOPPELGANGER :A CASE REPORT ON HERLYN WERNER WUNDERLICH SYNDROME

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## INTRODUCTION

Herlyn-Werner-Wunderlich (HWW) syndrome is a rare congenital anomaly characterized by uterus didelphys with blind hemivagina and ipsilateral renal agenesis. It is one of the rare anomalies of the genitourinary system and is usually identified in the first few years after menarche (1). OHVIRA syndrome belongs to the group of ORTAs (Obstructive reproductive tract abnormalities) with incidence varying between 0.1% and 3.8% in the general female population and 7% in all mullerian anomalies.(2)

## CASE REPORT

A 13-year-old female came to Queen Mary Hospital with complaints of intermenstrual spotting and dysmenorrhea for the past three months. Patient attained menarche four months back. Her cycles were regular but associated with heavy flow and pain. Patient first went to a private hospital where she got a USG done which was suggestive of a bicornuate bicollis uterus with right hematometra. On examination, her breast and pubic hair development corresponded to Tanner stage 4. Her abdomen was soft non-tender, no mass or associated organomegaly was seen. On local examination vulva and labia appeared normal. After consent, a gentle per vaginal examination was done which was suggestive of a canalized vagina and a bulge in the right upper vagina wall. On per rectal examination, the uterus appeared normal in size. MRI pelvis was suggestive of uterus didelphys with right sided obstructed hemivagina and ipsilateral renal agenesis.

On examination under anesthesia, a bulge was seen on the right anterolateral wall of vagina with a single cervical opening and a pulled up cervix. On diagnostic laparoscopy, two horns of uterus were seen resembling a horse-shoe in appearance with the right horn being larger than left horn. Both ovaries and fallopian tubes appeared normal. A decision was made to proceed through abdomino perineal route. Per vaginally a uterine sound was inserted into the left horn of uterus and 20 mL tarry fluid was drained. Per abdominal, both horns were palpated and a vertical incision was made on the right horn to expose the internal os. A Hegar dilator was introduced through the incision, passing through the internal os with its lower end felt through the bulge on the right anterolateral vaginal wall. The lower edge of the vaginal septum was identified and resected, thus canalising the vagina leading to two cervix and two vaginas.



INTRAOPERATIVE  
IMAGES



## DISCUSSION

Due to overlapping symptoms with other conditions, OHVIRA syndrome is often misdiagnosed. It is characterized by a combination of Mullerian anomaly, mesonephric duct anomaly, vaginal septum and renal anomalies. Diagnostic approach involves a thorough history, physical examination, and imaging. The primary management for obstructive symptoms is vaginoplasty, which can be performed by a single-stage approach. Surgical correction leads to normal sexual function and successful pregnancies.(3)

## CONCLUSION

Management of OHVIRA syndrome requires a comprehensive careful diagnostic evaluation, and surgical approach. Early diagnosis and intervention significantly improves patient outcomes. It represents a diagnostic dilemma because of the regular menstruation and nonspecific lower abdominal pain. Patients with this anomaly usually present after menarche with pelvic pain and/or an abdominal mass.(4)

## REFERENCES

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