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SUCCESS OF PREGNANCY AND CHILDBIRTH IN SYNDROME *HERLYN-WERNER-WUNDERLICH (HWW)* / SYNDROME *OHVIRA*: *LAPORAN KASUS LANGKA*

Patricia Riwiris Sry Rezeki Samosir[1], Dhaniel Theolia Persadanta Ginting[2], Kevin Nicholas Rumahorbo[3], Edwin Martin Asroel[4]
[1 3]*Faculty of Medicine, University of North Sumatera, Medan, Indonesia*
[2] *General Practitioner, Stella Maris Women's & Children's Hospital, Medan, Indonesia*
[4]*Obstetrician and Gynecologist Subspecialist in Fetomaternal, Stella Maris Women's and Children's Hospital, Medan, Indonesia*
Email : patricia.r.sam@gmail.com

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ABSTRACT

Herlyn-Werner-Wunderlich (HWW) syndrome also known as OHVIRA (Obstructed HemiVagina and Ipsilateral Renal Agenesis) syndrome is a rare form of Müllerian Duct Anomalies (MDA). Uterine malformations may be associated with adverse obstetric outcomes, including increased rates of miscarriage, malpresentation, and preterm birth. Case Illustration: A 29-year-old woman at 36-37 weeks' gestation was diagnosed with Premature Rupture of Membranes with OHVIRA syndrome. The patient underwent a cesarean section under spinal anesthesia. A baby girl was born with a birth weight of 2980 grams and an Apgar score of 8/9. The uterus appeared didelphysic. This case study examines a nulliparous woman with premature rupture of membranes (PROM) and OHVIRA syndrome. Delivery was assisted by cesarean section. Conclusion : OHVIRA syndrome is a rare condition that carries many risks in pregnancy. However, there have been several cases of successful pregnancies and deliveries. Suggestion: A multidisciplinary approach and patient education are needed to increase the success of pregnancy and childbirth.

Keywords: *OHVIRA, Müllerian duct, Didelphys.*

INTRODUCTION

The Mullerian duct is the embryological origin of the fallopian tubes, uterus, cervix, and the upper two-thirds of the vagina. Various malformations can occur due to impaired Mullerian duct development resulting in agenesis, hypoplasia, or fusion defects in one or both ducts (1). Major structural malformations such as unicornurate, bicornnuata, and didelfic uterus affect the external form, while minor malformations such as septate or arcutic uterus are confined to the internal cavity (2)

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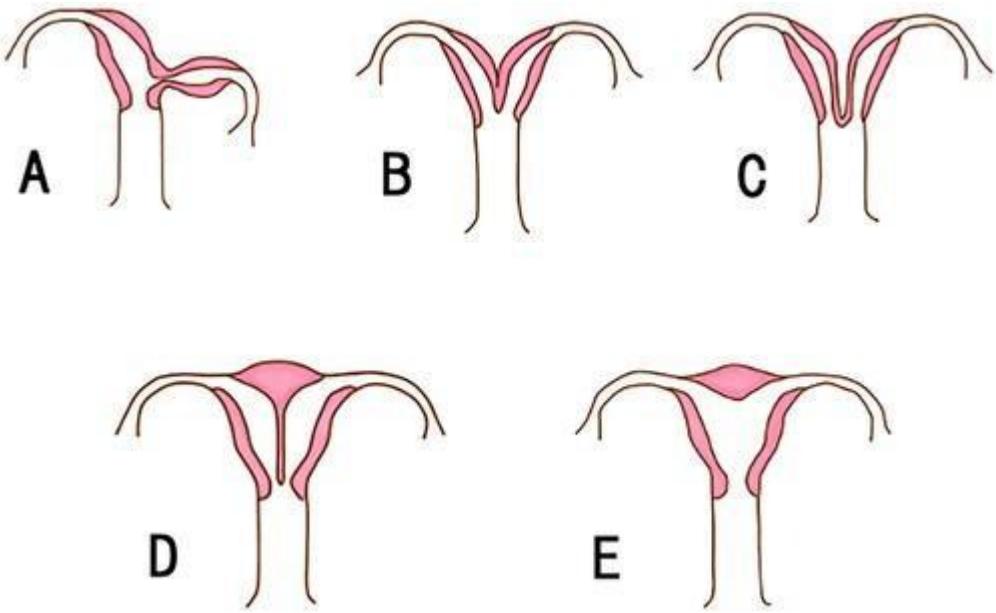


Figure 1. Types of uterine congenital abnormalities. (A) unicornual uterus, (B) bicornual uterus, (C) uterus double, (D) blocked uterus, and (E) arquatic uterus (2).

Congenital uterine malformations are estimated to occur in about 0.4–5% of the general female population (2). A systematic review estimated the overall prevalence of uterine malformations to be 5.5% in the general population, 8% in infertile women, 13.3% in women with a history of miscarriage, and 24.5% in women who had miscarriage and infertility, suggesting a link between congenital uterine malformations and infertility and poor pregnancy outcomes (3). Uterine malformations may be associated with adverse obstetric outcomes, including increased miscarriage rates, malpresentation, and preterm birth (3).

Herlyn-Werner-Wunderlich syndrome (HWW) is a rare form of Mullerian Duct Anomalies (MDA). The characteristic trias of this syndrome include uterine didelphys, blocked hemivagina, and ipsilateral renal agenesis hence, also known as OHVIRA syndrome (Obstructed HemiVagina and Ipsilateral Renal Agenesis) (4). The incidence of uterine didelphys is about 1 in 2000 to 1 in 28,000 and is accompanied by unilateral renal agenesis in 43% of cases. The incidence of unilateral renal agenesis is 1 in 1100 and between 25% to 50% of affected women have associated genital anomalies (4). The most common symptoms are dysmenorrhea, abdominal pain, and abdominal mass due to hematococpus. The exact cause, pathogenesis, and embryological origin of OHVIRA syndrome remain unclear (1).

The writing of this case report aims to report the success of pregnancy and childbirth in patients with OHVIRA Syndrome. This case report also aims to improve understanding of the characteristics, diagnosis, and management of pregnancy in patients with these conditions so that it can help in making appropriate clinical decisions and improve patient prognosis.

CASE ILLUSTRATION

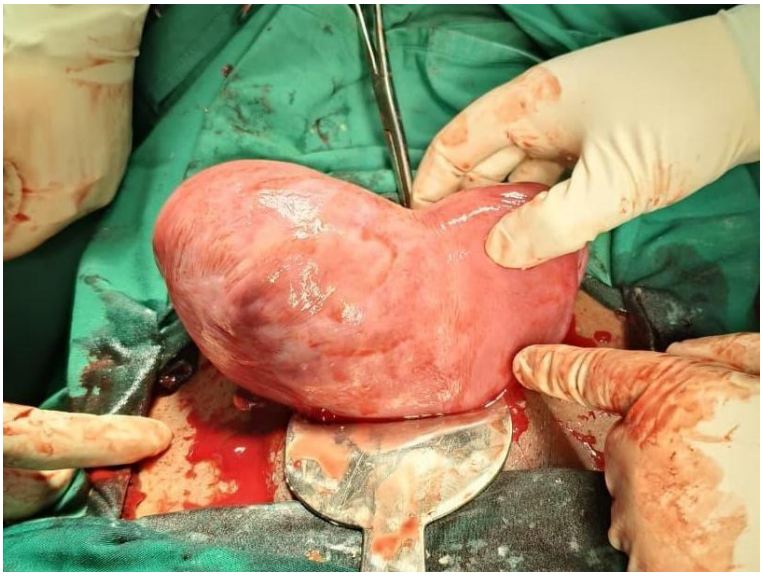
A old woman 29 years old came in a wheelchair to the emergency room of the hospital on December 4, 2025 at 18.49 WIB. The patient is in the first pregnancy (G1P0A01) with a gestational age of 36-37 weeks. The patient came in with complaints of seeping out amniotic fluid this morning and a slight mule stomach. The last period is on March 21, 2025. Estimated delivery date December 28, 2025. Patients with a history of single kidney based on ultrasound examination. During the antenatal period, through ultrasound examination, the fetus appears to develop according to the gestational age and no congenital abnormalities are detected. The patient has an allergy to Cyclopentasiloxane cream.

The patient's general state is good and fully conscious (GCS 15). Vital signs of patients with blood pressure 110/80 mmHg, Pulse 82 times/minute, Breathing 20 times/minute, Body temperature 36.8 C, Oxygen saturation 99% of the water room, pain scale 4/10. The patient's weight is 59 kg, the patient's height is 150 cm. On physical examination, anemic conjunctiva was found. His inadequacy, fetal movement is present.

Routine blood test for anemia (Hb 8.8 g/dL). There were no signs of infection (Leu 10.21 103/µL). Blood sugar levels are checked within normal limits (GDS 112 mg/dL). Bilirubin test within normal limits (Bil Tot 0.51 mg/dL). Examination of liver function within normal limits (SGPT 14 U/I). Examination of kidney function within normal limits (Ur 8 mg/dL; Cr 0.5 mg/dL). Serological examination of HbsAg and non-reactive HIV blood.

On December 8, 2025, at 36-37 weeks gestation, 5 and 5 caesarean sections were performed to deliver the patient's fetus under spinal anesthesia because his mother was inadequate. The operation proceeds without complications. Baby girl was born with a BBL of 2980 gr, PB 48 cm, APGAR Score 8/9, Abnormalities: Not found. The uterus appears to be didelphys. Approximate amount of bleeding 400 ml, diuresis 300 ml. The condition of the mother and child after the surgery is in good condition and healthy without complications.

The working diagnosis is Premature Ruptured Amniotic Rupture in G1P0A0 with Gestational Age of 36-37 weeks with a Live Single Fetal and Buttock Presentation with OHVIRA Syndrome. The post-surgical diagnosis on December 8, 2025 is Post Sectio Caesarea G1P1A0 with OHVIRA Syndrome.



Gambar 2. Uterus Ibu durante operasi.

DISCUSSION

This case reported a 29-year-old nullipara woman (G1P0A0) with Premature Rupture of Amniotic Membrane with OHVIRA Syndrome. Childbirth assistance is carried out by sectio caesarean section.

OHVIRA syndrome is described as a malformation of the Müllerian duct. In the 8th week of pregnancy, the two Müllerian or paramesonephric ducts fuse to form the uterus, then the proximal two-thirds of the vagina and cervix with the central septum begin to be absorbed in the 9th week until one cavity is finally left. The incidence of unilateral renal agenesis is described at 1:1000 live births. The relationship between renal malformations and Mullerian duct malformations is described between 30 and 70% and is apparent from the interaction between the paramesonephic and mesonephric ducts during renal development. This failure of interaction between the paramesonephric and mesonephric ducts is believed to be the cause of kidney anomalies in OHVIRA Syndrome (5).

The fertility of patients with uterine didelphys has been shown to be better than that of patients with other Mullerian duct abnormalities but still lower than that of women with normal uterine anatomy. For uterine didelphys, vaginal delivery can be considered but cesarean delivery is preferable. Compared to a normal uterus, patients with didelphys uterus have a full-term delivery rate of about 45%. In these patients it reaches the gestational age of 36-37 weeks. The patients had higher rates of poor obstetric outcomes such as spontaneous miscarriage 32%, premature birth 28%, intrauterine growth retardation, malpresentation, preterm labor, and preterm birth. This complication is suspected to occur due to impaired uterine stretching ability, reduced uterine cavity size and abnormalities in the function of the endometrium, myometrium, and cervix. Malpresentation and dystocia due to obstruction of the pelvic entrance by the uterine cavity that are not pregnant are the two main reasons for the high rate of cesarean sections in patients with uterine didelphys. In this patient, the fetus in the buttocks presentation position is accompanied by a

condition of premature rupture of amniotic. In uterine didelphys with obstructive hemi-vagina, ipsilateral urinary tract malformations such as renal agenesis are common in about 15–30%, the right side is usually affected twice as often as the left side (1). OHVIRA syndrome is usually characterized by a double uterus, a unilaterally blocked vagina, and ipsilateral renal agenesis (6).

Some of the main complaints in OHVIRA syndrome are lower abdominal pain, dysmenorrhea, recurrent UTIs, urinary retention, chronic pelvic pain, and recurrent vaginal discharge. Most of these complaints are caused by fluid accumulation in the blocked hemivagina. In further circumstances it can lead to chronic infections (7).

Ultrasound can be useful in detecting obstructive presentations, but it is not the most reliable diagnostic method. MRI shows up to 95% correlation with laparoscopic findings making it an efficient examination method for diagnosing Mullerian duct malformations (8). In this patient, the shape of the uterus is seen in the form of didelphys when the sectio caesarean section is performed. The findings on the MRI included complete duplication of the uterine and endocervical cavities (uterus didelphys). Typically, cystic structures look distal and are associated with one of the endocervical canals that represents a blocked hemi-vagina. A blocked vagina usually contains blood or protein fluid that appears bright on a T1-weighted image. Levels of fluids may be visible. Other findings including hematometrosis and or hematosalpinx can be seen as the intensity of bright T1 filling the uterine cavity or fallopian tubes. Simultaneously assess Mullerian duct malformations and associated urinary tract malformations. Surgery to diagnose uterine malformations is rarely performed. Laparoscopy is the gold standard in diagnosing female genital tract malformations, but it is only performed if MRI is not available or fails to establish a diagnosis with MRI (1).

Mullerian duct malformations do not require surgical correction if the patient is asymptomatic and does not have obstruction. In this patient, no surgical procedure was performed on uterine malformations, only sectio caesarea surgery was performed to deliver the patient's fetus with good maternal and child conditions. The surgical goals of Mullerian duct malformations are normal menstrual flow, and pain reduction (1).

CONCLUSION

OHVIRA syndrome is a rare case that has many risks in pregnancy, but there are a number of cases that have been successful in pregnancy and childbirth with the condition of the mother and fetus in good condition despite a number of complications. This success is greatly influenced by optimal antenatal monitoring. Proper management during pregnancy and the selection of appropriate delivery methods also play an important role in minimizing maternal and neonatal complications.

SUGGESTIONS

It is necessary to take a multidisciplinary approach involving obstetrics and gynecology, radiology, and urology specialists in treating patients with OHVIRA syndrome, especially during pregnancy. A number of studies and case

reports are also needed to evaluate the success of pregnancy and childbirth, prognostic factors, and the best management strategies in patients with this condition. Patients with OHVIRA syndrome also need to be educated about pregnancy risks, the importance of regular control during pregnancy, and careful pregnancy planning to improve pregnancy and delivery success.

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