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OHVIRA Syndrome with Spontaneous Pregnancy After Prolonged Primary Infertility

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Abstract

OHVIRA syndrome (Obstructed Hemivagina and Ipsilateral Renal Anomaly), also known as Herlyn–Werner–Wunderlich syndrome, is a rare congenital Müllerian anomaly characterized by a uterine malformation, an obstructed hemivagina, and ipsilateral renal agenesis. Diagnosis is usually made during adolescence due to cyclical pelvic pain caused by hematocolpos, but delayed diagnosis may lead to endometriosis, adhesions, and infertility.

We report the case of a 36-year-old woman with a history of OHVIRA syndrome diagnosed at the age of 26 after several years of cyclical pelvic pain. Imaging revealed a bicornuate uterus with an obstructed uterine horn and right renal agenesis. Surgical management consisted of resection of the obstructed uterine component with drainage of retained contents, resulting in significant symptomatic improvement. Despite appropriate surgical correction, the patient experienced ten years of primary infertility, likely related to tubal obstruction, adhesions, and possible residual endometriosis. She ultimately achieved a spontaneous pregnancy without assisted reproductive techniques and is currently 25 weeks pregnant with a favorable evolution.

This case highlights the importance of early diagnosis and timely surgical treatment of OHVIRA syndrome to optimize reproductive outcomes. Although prolonged infertility may occur due to secondary pelvic damage, spontaneous conception remains possible. Pregnancies in these patients require close multidisciplinary follow-up due to increased obstetric and renal risks.

Abbreviations: HSG: Hysterosalpingography

Introduction

OHVIRA syndrome (Obstructed Hemivagina and Ipsilateral Renal Anomaly), also known as Herlyn-Werner-Wunderlich syndrome, is a rare congenital anomaly of the Mullerian structures characterised by a didelphic or bicornuate uterus, an obstructed hemivagina and an ipsilateral renal anomaly **[1]**. This malformation accounts for 2-3% of all Müllerian anomalies, with an estimated incidence of between 0.1-3.8% **[2]**.

The diagnosis is typically made during adolescence following

cyclical pelvic pain and a pelvic mass resulting from haematocolpos **[1]**. However, in some patients, the discovery occurs later, revealed by primary infertility secondary to peritoneal adhesions or endometriosis **[3]**. Early surgical treatment, performed by resection of the vaginal septum and drainage of the retained contents, significantly improves subsequent fertility and reduces obstetric complications such as premature delivery or abnormal foetal presentation **[4]**. This case report describes a 36-year-old patient who was 25 weeks pregnant and underwent surgery in 2015 for OHVIRA syndrome, illustrating the diagnostic and therapeutic challenges as well as the favourable reproductive outcomes possible after conservative treatment.

Clinical Case

Initial Clinical Presentation A 26-year-old female patient with no significant surgical history consulted for chronic cyclical pelvic pain that had been developing over several years.

The interview revealed progressive dysmenorrhoea and debilitating pelvic discomfort during menstruation.Pelvic imaging (ultrasound and MRI) revealed: Bicornuate uterus with small (hypoplastic) left horn, no bilateral ovarian abnormalities, Single left kidney (right renal agenesis), Enlarged right horn, site of fluid retention suggestive of haemometrium.

An exploratory laparotomy was performed, confirming the imaging findings:

- Bicornuate uterus with blind right horn (imperforate)
- Retained uterine contents (haematometra)
- Normal ovarian appendages bilaterally
- Single left kidney
- No additional renal abnormalities

Surgical Treatment

The patient underwent resection of the obstructed uterine half, allowing for:

- Complete drainage of retained fluid
- Restoration of functional vaginal anatomy
- Prevention of long-term complications (endometriosis, infertility)

The postoperative period was uneventful, with no complications and a marked improvement in pain symptoms within the first month following surgery.

Post-operative progress and infertility assessment

Despite regular gynaecological check-ups and regular unprotected sexual intercourse, the patient presented with primary infertility over a period of ten years. A thorough infertility assessment was undertaken in the context of a lack of spontaneous conception.

Additional imaging results:

Follow-up pelvic MRI revealed a 7 cm cystic formation in the right lumbopelvic region, coming into contact with the cervico-isthmic region of the uterus, suggestive of a hydrosalpinx or adnexal collection.

Hysterosalpingography (HSG) revealed:

- Endometrial pseudopolypoid hyperplasia
- Moderately suspended left fallopian tube with localised peritoneal mixing
- Radiological appearance suggestive of a post-infectious origin of this obstruction
- Obstruction of the right fallopian tube with a club-like appearance at its distal end

Hormonal and male factor investigation:

Sperm analysis of spouse: no significant abnormalities

Complete hormonal assessment (FSH, LH, prolactin, testosterone): results within normal limits Spontaneous Pregnancy and Current Follow-up

After ten years of primary infertility, the patient successfully conceived spontaneously, without resorting to assisted reproductive technology. The pregnancy was diagnosed early and is being closely monitored by an obstetrician, given the underlying uterine malformation and potential obstetric risks. At the time of this report, the patient is 25 weeks pregnant, with a satisfactory gestational outcome to date.

Discussion

Pathophysiology and Complications of Untreated OHVIRA Syndrome

OHVIRA syndrome results from an abnormality in the embryonic development of the Mullerian structures, characterised by asymmetric fusion or the presence of an obstructive vaginal septum. This obstruction causes menstrual blood to accumulate in one of the uterine horns,

creating a state of chronic haemometria. This pathological situation promotes the development of endometriosis through retrograde menstrual regurgitation and the return of endometrial cells to the peritoneal cavity, contributing directly to infertility **[2,4]**. Secondary peritoneal adhesions, resulting from chronic inflammation and endometriosis, compromise tubal function and significantly reduce the likelihood of natural conception **[3]**.

Impact of Surgical Treatment on Fertility

Early resection of the obstructed hemivagina effectively prevents the development of endometriosis and infertility due to menstrual flow **[5]**. Published series report a post-surgical pregnancy rate of up to 85% in patients treated within years of diagnosis **[2,6]**. In the patient presented, the initial surgery at the age of 26 restored the vaginal anatomy and eliminated the source of haematometra, thereby reducing the long-term risks of progressive endometriosis **[7]**.

Analysis of Prolonged Infertility

Despite appropriate surgical correction and anatomical restoration, this patient experienced ten years of primary infertility before achieving a spontaneous pregnancy. This prolonged duration can be explained mainly by:

1. Chronic peritoneal adhesions: The prolonged inflammation preceding the surgical intervention likely generated dense adhesions, reducing tubal mobility and compromising gamete transport.

2. Distal tubal obstruction: The HSG documented obstruction of the right tube with morphology suggestive of post-infectious sequelae, limiting tubal patency.

3. Associated hydrosalpinx: The lumbopelvic cystic formation detected on MRI probably represents a hydrosalpinx, a condition known to reduce fertility rates.

4. Residual endometriosis: Pseudo-polypoid endometrial hyperplasia may represent sequelae of previous endometriosis.

These multiple factors, combined with the residual anatomical abnormality (bicornuate uterus), likely contributed to prolonged infertility, despite partial restoration of normal anatomy **[1,4,8]**.

Expected Obstetric Complications and Prognosis Subsequent pregnancies in patients with a history of OHVIRA syndrome carry increased risks that are well documented in the literature:

- Prematurity: reported incidence of 36% in published series
- Abnormal presentation (breech or transverse): estimated risk of 38%
- Need for caesarean delivery: frequency of 67%
- Intrauterine growth restriction: secondary to inadequate placental perfusion
- Pre-eclampsia: increased risk associated with unilateral renal agenesis and reduction in total nephron mass

Rigorous multidisciplinary follow-up involving an obstetrician, urologist and nephrologist is essential to monitor:

- The appearance of signs of pre-eclampsia
- The progress of the pregnancy through regular serial ultrasound scans (foetal growth, amniotic fluid volume, foetal well-being)
- Maternal renal function (creatininaemia, albuminuria, blood pressure monitoring)
- The development of the foetal position to anticipate vaginal delivery or planned extraction

Despite these documented risks, the favourable outcome of this pregnancy at 25 weeks of amenorrhoea suggests a potentially favourable obstetric prognosis if appropriate monitoring is maintained until term **[1,6]**.

Clinical Implications and Recommendations

This case highlights the importance of early diagnosis of OHVIRA syndrome in adolescent girls and young women presenting with chronic cyclical pelvic pain.

Early surgical diagnosis and treatment significantly optimis long-term reproductive outcomes and reduce the risk of complications. Even in cases of prolonged infertility, the possibility of spontaneous conception persists, as illustrated by this case after a decade of infertility **[9]**.

Conclusion

This clinical case illustrates the reproductive success that can be achieved following conservative surgical treatment of OHVIRA syndrome, performed as soon as the diagnosis is made. Although the patient had experienced prolonged

primary infertility for ten years as a result of the sequelae of the Müllerian anomaly, she ultimately achieved a viable spontaneous pregnancy, currently at 25 weeks of amenorrhea. This case demonstrates the crucial importance of accurate diagnosis and timely surgical treatment of OHVIRA syndrome to optimise reproductive outcomes. Special obstetric vigilance and multidisciplinary follow-up are essential for subsequent pregnancies in these patients, given the increased risks of maternal and foetal complications. Systematic screening for Müllerian anomalies in young infertile patients is justified to prevent long-term complications and improve the chances of future reproduction **[9,10]**.

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