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# A Rare Case of Herlyn-Werner-Wunderlich Syndrome

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## Abstract

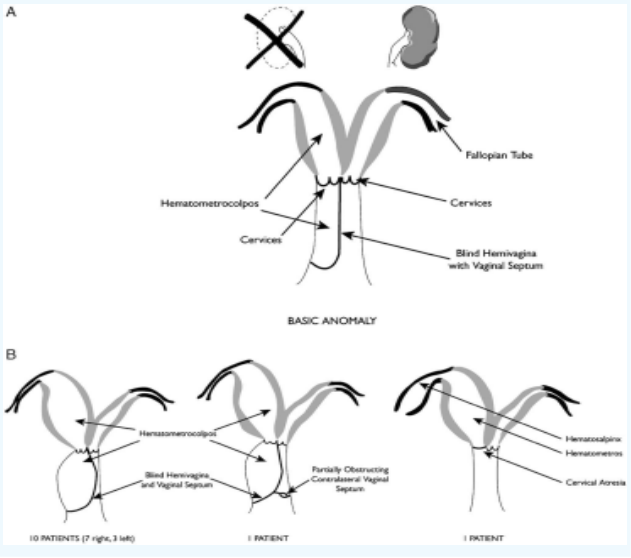
Herlyn-Werner-Wunderlich Syndrome (HWW syndrome) is a very rare Mullerian anomaly characterised by Uterine didelphys, Obstructed hemi cervix or hemi vagina and ipsilateral agenesis. Ovarian endometrioma may be present in 75% of the patients. Common presentation is progressive dysmenorrhea in the post menarche. There can be hemato-colpos and hemato-metra which can present as abdominal mass. Menstrual cycles may not be altered as there is no obstruction in the other vagina. The diagnosis is likely to be delayed unless the clinician has got a mind to think about it. It is invariably diagnosed by USG and MRI may be needed for accurate diagnosis and planning the surgical procedure. In this case, initial laparoscopy was done for endometrioma and noted uterine didelphys. The obstruction at the cervical level was not picked up and hence the patient had recurrence. Second laparoscopy we have done a hemi hysterectomy and ovarian cystectomy of right ovary. The patient became asymptomatic with preservation of fertility.

**Keywords:** Mullerian anomaly, Renal agenesis, Endometrioma.

## Introduction

Herlyn-Werner-Wunderlich Syndrome is a very rare congenital Mullerian anomaly characterised by Uterine didelphys, Obstructed hemi vagina and Ipsilateral renal agenesis (OVIHIRA Syndrome) (Fig: 1)[1,2].

**Fig: 1 – Schematic diagram depicting Herlyn-Werner-Wunderlich Syndrome**




It is commonly diagnosed during post menarche, but also it has been reported in neonates and adults with features of urinary obstruction and abdominal swelling[3,4]. Because of the rarity there is often a delay in diagnosis and affects quality of life. USG and MRI aid in the diagnosis of this syndrome. Treatment is basically surgical, like excision of transverse vaginal septum if the level of obstruction is at the hemi-vagina. If the obstruction is at the level of cervix leading to hematometra, patient may need laparoscopic hemi hysterectomy. Associated endometriomas can also be simultaneously treated. Very often renal anomalies will not have any impact on the quality of life. Fertility will not be hampered in 80% of the cases.

## Case Report

Miss. K. 20 years female was referred to us, with complaints of severe dysmenorrhoea since menarche. Her menstrual cycles were regular 3-4/30-35 days.

## Past History

Patient had undergone laparoscopy 2 years back for ovarian endometrioma. Intra-operative findings showed uterine didelphys with slightly enlarged right horn and right ovarian chocolate cyst. Cystectomy was done. Histopathology report was endometrioma.

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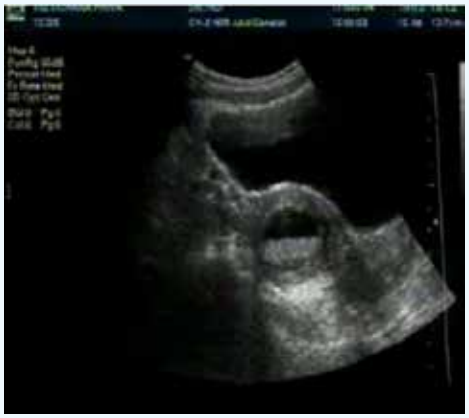
She was free of symptoms for one year. After that she developed dysmenorrhoea again and came to our hospital for further management.

## Clinical Examination

Her vitals were stable. Abdomen was soft, no mass was palpable.

## USG Findings

**Fig 2 : USG showing right hematometra with normal left horn**



USG showed absent right kidney with uterine didelphys. These was haemato-metra of right horn of uterus and recurrent right ovarian endometrioma. Left kidney, left horn of uterus and ovary were normal. MRI was done and showed similar findings (Fig: 3 & 4).

## Treatment

As she was symptomatic, repeat laparoscopy was done, which showed similar findings (Fig: 5). Hysteroscopy was also done which showed normal left uterine cavity and right horn which was non communicating. Hence excision of right horn was done with removal of endometrioma of right ovary. Post-operative period was uneventful. Patient was followed up and was free of dysmenorrhoea. There was no recurrence of endometrioma also. Patient got married at the age of 22 yrs. and had spontaneous conception within a year. She delivered vaginally at 37 weeks of gestational age.

This rare case of Herlyn Werner Wunderlich Syndrome, when diagnosed earlier and treated both quality of life and fertility can be preserved.

**Fig: 5 – Laparoscopic view**

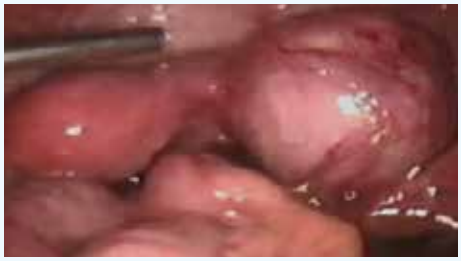
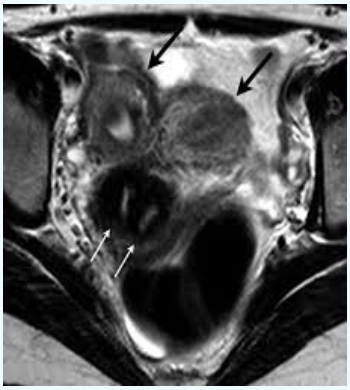
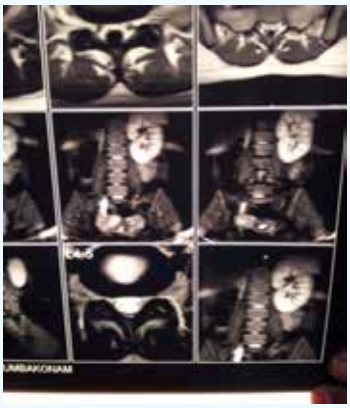


**Fig 3 & 4 : MRI of the patient**

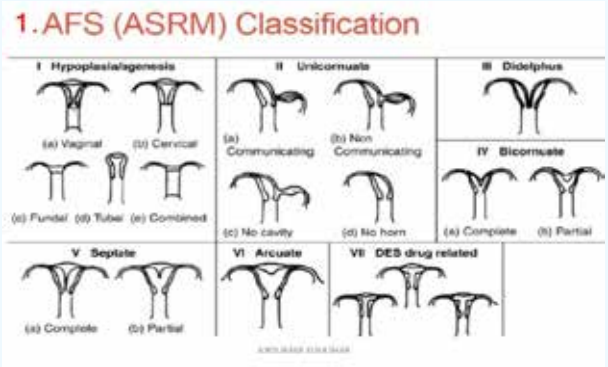




## Discussion

Mullerian duct anomalies result due to non-development, defective fusion or defective regression of septum during foetal development. Herlyn Werner Wunderlich Syndrome is a rare Mullerian ductal anomaly, characterised by a triad of obstructed hemi-vagina, uterine didelphys and ipsilateral renal agenesis. It is classified as Type III Mullerian anomaly (Fig: 6) associated with non-development of meso nephric duct and accounts to 5% of Mullerian duct anomalies[1,2,5]. Though the first case was reported in 1922 in a patient with unilateral hemato-colpos, hemato-metra and haemato-salpinx, the triad was first described only in 1971.

**Fig 6 : Depicts AFS (ASRM) Classification. Our patient fell under Class III**




This is most commonly observed in adolescent girls and young women. They commonly present with symptoms of severe progressive dysmenorrhoea, abdominal pain, and

pelvic mass. The diagnosis may be difficult due to rare occurrence and hence high index of suspicion is required. We need to search for renal anomaly whenever there is Mullerian anomaly and vice versa.

## Incidence

The estimated of incidence of MDA is 2-3% and in women with reproductive disorders, there is higher incidence of MDA[2,5,6]. Uterine didelphys accounts to 11% of MDAs. Renal anomalies are associated in 43% of the patients[2,3]. 75% of the patients with uterine didelphys have complete or partial vaginal septum[4]. The incidence of uterine didelphys is 1/2000 – 1/28,000. Incidence of HWW syndrome varies between 0.1 – 3.8% in the literature [3,7].

HWW Syndrome is a disorder of lateral fusion between the two Mullerian Ducts[8]. Studies have shown uterine organogenesis is controlled by HOX and WNT genes. Usually this defect starts occurring at 8 weeks of gestation, in which there is non-fusion of Mullerian duct or failed resorption of utero vaginal septum[1]. Renal defects include agenesis of ipsilateral kidney, dysplasia and ureteric abnormality. Right side has more predilection than left side[2,3]. Our patient had anomaly in the right side only. If complete obstruction occurs, patients present with symptoms in the younger age group. If there is incomplete obstruction, they present with features of dysmenorrhea seen in the later age.

## Presenting Features

Common presenting features are abdominal mass due to haemato-colpos and haemato-metra, lower abdominal pain, dysmenorrhea, retention of urine and constipation. The diagnosis may be delayed if there is patency of one hemi vagina through which the patient will be menstruating. Rarely, this menstrual blood can get infected leading to pyocolpos, pyometra and tubo-ovarian mass. Such patients present with features of severe fever, chills, nausea and vomiting.

## For diagnosis

Ultrasonography usually gives a correct picture, by showing uterovaginal duplication, haemato-colpos or haemato-metrocolpos, absence of ipsilateral kidney and ovarian endometrioma[7,9,10]. However, MRI is considered as imaging modality of choice by various authors before planning for surgical intervention[8,10,11].

## Treatment

Modality of treatment in the presence of vaginal septum is septostomy. If there is obstruction at the level of cervix, like cervical agenesis, laparoscopic excision of the non-communicating horn of uterus is the treatment of choice [7,9,10]. Removal of endometriotic cyst and conserving the affected ovary should be done. Mullerian anomalies with obstruction are associated with endometriosis in 75% of cases. Usually the endometriosis resolves once the obstruction is relieved.

Prognosis is usually very good with preservation of fertility and relief of symptoms[3,7,9,10]. 80% of patients conceive spontaneously and there is always risk of abortion (74%), premature delivery (22%) and LSCS (80%) of the patients[12,13,14].

To summarise, a gynaecologist should always have a high index of suspicion regarding MDA, when they see a patient with renal abnormality and vice versa. Clinical examination supported by USG and MRI will help to clinch the diagnosis and plan the treatment. Proper diagnosis and treatment will reduce the risk of endometriosis and infertility. When these patients conceive, they should be counselled regarding the high risk of miscarriage and premature labour.

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