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Diagnostic Dilemma and Management of Rare Mullerian Anomaly: OHVIRA Syndrome

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Abstract **Original Research Article**

Background: Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome is a rare female urogenital tract malformation characterized by the triad of uterine didelphys, obstructed hemivagina and ipsilateral renal anomaly. Obstructed anomalies, if not treated timely and adequately, may lead to chronic complications and thereby impairing a woman's reproductive life. ***Aims and Objectives:*** The aim of the study is to evaluate the reproductive outcome of patients with OHVIRA syndrome after treatment. ***Methods:*** The present prospective observational study was conducted in Reproductive Endocrinology and Infertility Department of BSMMU from September 2018 to September 2022. Among 84 patients of Mullerian Anomaly, 10 patients presented with obstructive features were enrolled in the study. Cases were analyzed according to detailed history, thorough examination, relevant investigations with TVS and MRI of whole abdomen and laparoscopy maintaining inclusion and exclusion criteria. ***Results:*** The mean age was 25.30 ± 5.93 years. In our study 90% patient presented with severe dysmenorrhoea. Right sided obstructed Hemivagina with right renal agenesis were more common 60%. Eighty percent patients had didelphys uterus, twenty percent had bicornuate uterus and twenty percent cases were associated with endometriosis. 60% patients were treated by excision of obstructed vaginal septum and 40% patients required excision of functioning non-communicating uterine horn. Improvement of symptom occurred in 90% patients. ***Conclusions:*** When diagnosis of OHVIRA Syndrome is not delayed and indication and timing of surgery are appropriate, complications can be avoided and fertility can be preserved.

Keywords: OHVIRA Syndrome, Infertility, Didelphys Uterus.

INTRODUCTION

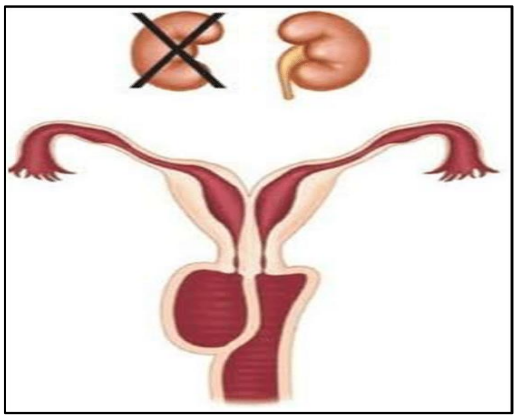
Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome is a rare female urogenital tract malformation [1]**.** It is a rare condition first reported in 1922 and subsequently described in detail by Herlyn, Werner, and Wunderlich after whom it was initially named HWW syndrome. It is characterized by the triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal anomaly [2]. Although the incidence of mullerian defects is about 1.1 - 3.5%, the incidence of OHVIRA syndrome is very low and is estimated to be between 0.1-3.5% of all mullerian anomalies [2, 3].

The exact etiology and pathogenesis of OHVIRA syndrome is still unknown. While the uterocervical development was believed to be from paired paramesonephric ducts; sinovaginal bulb from urogenital sinus was thought to develop into lower vagina; the kidneys and ureters were believed to arise from wolffian pro/meso/metanephros. Aberration in vertical or horizontal fusion or arrest of paramesonephric ducts during course of development

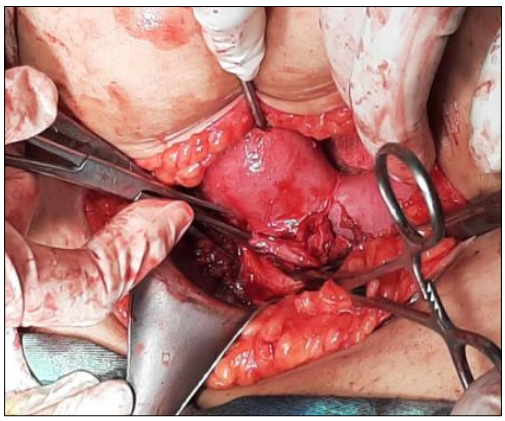
was understood to successfully explain various uterine anomalies. Some complex uterine anomalies such as OHVIRA syndrome (Obstructed Hemi Vagina with Ipsilateral Renal Agenesis) still defied the conventional theory of urogenital development until recently when theory of development of entire vagina from mesonephric ducts was postulated [4].

Due to failed positioning of paired paramesonephric duct, the two hemiuteri and hemicervices fail to unite, resulting in uterus didelphys. In OHVIRA syndrome, developmental arrest of ipsilateral mesonephric duct results in failure of distal hemivagina to develop, thereby resulting in obstructed hemivagina.

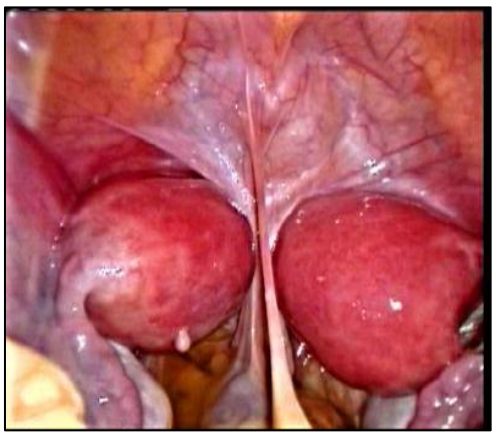
Though renal agenesis is the classic presentation, other anomalies include renal dysplasia and ectopic ureters [5]. In girls with unilateral renal anomaly it is particularly important to consider OHVIRA Syndrome. As well as renal anomalies ipsilateral to the obstructed hemivagina, contralateral renal anomalies have been reported in up to 50% [5, 6].



Graphical representation of the triad of OHVIRA syndrome



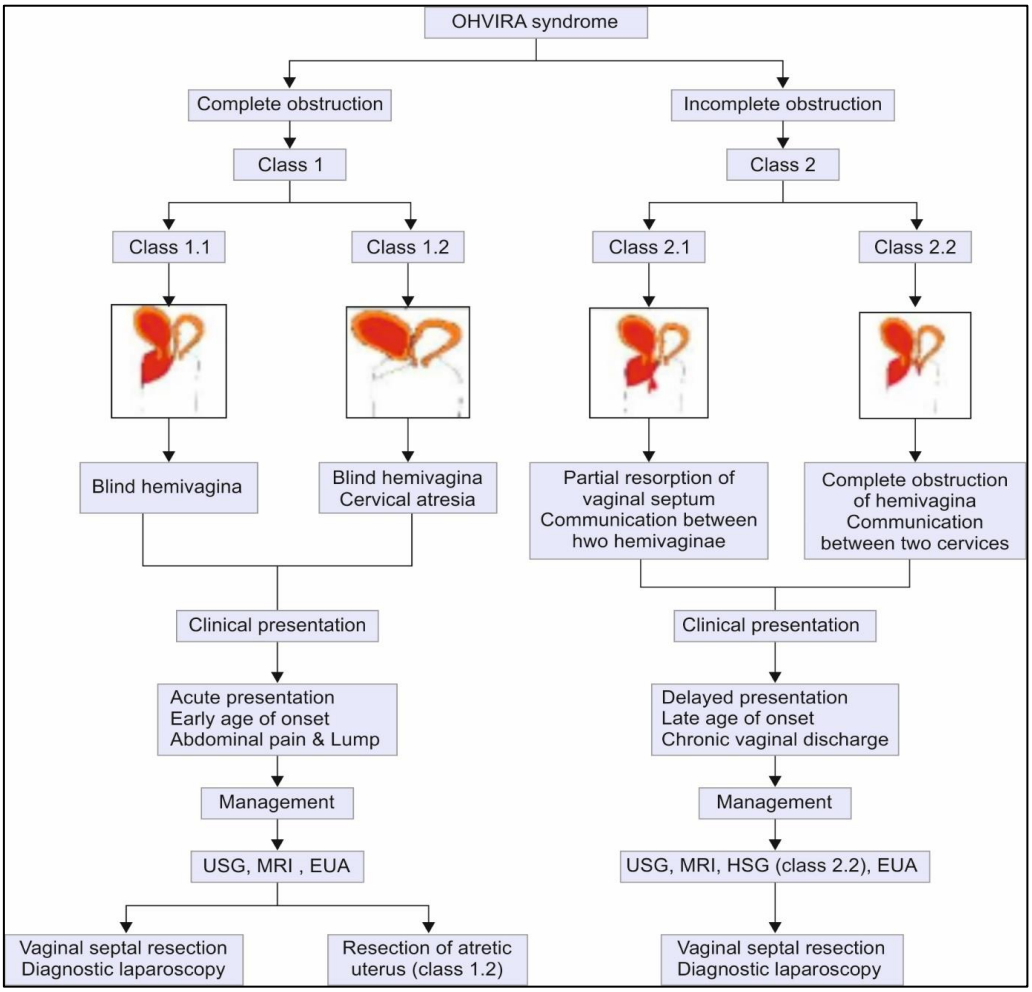
Drainage of Hematocolpos in OHVIRA Syndrome



Uterine didelphys (omental bands between two uterine horns)

As we all know female reproductive function depends on the intact genital tract along with a functional hypothalamic–pituitary–ovarian axis. Aberration in any of the systems in any form like OHVIRA Syndrome may significantly affect a woman's reproductive potential, thereby negatively affecting the quality of life.

Taking into consideration the different classification systems of mullerian anomalies, OHVIRA syndrome is classified under class III as per the ASRM classification and under class U3b C2 V2 as per the ESHRE classification [7, 8]. At times, obstructed hemivagina and ipsilateral renal agenesis may be associated with a bicornuate or septate uterus (class U2b as per ESHRE/class IVa or Va as per ASRM); constituting OHVIRA variants. Based on a retrospective study including 70 patients by Zhu *et al*., OHVIRA syndrome is classified into two classes depending on whether the obstruction of the hemivagina is complete or incomplete (Flowchart 1) [9].



In most cases, OHVIRA isn't diagnosed until a girl reaches puberty and experiences menstrual problems or other symptoms. Clinical symptoms of this syndrome vary widely. OHVIRA should be considered as a differential diagnosis in young women and adolescents presenting with pelvic pain, tenderness, dysmenorrhea, pelvic mass, persistent vaginal discharge, urinary symptoms and primary infertility. The diagnosis is likely to be missed or delayed due to the occurrence of normal menstrual flow from the patent hemivagina. Diagnosis of OHVIRA syndrome requires a multimodal approach, which includes a detailed history, meticulous examination, and appropriate imaging studies. Ultrasound and MRI are the established imaging modalities for diagnosing this condition with the ultrasound being the first-line option. However, expertise is required to diagnose the condition as the problem arises due to the small size of the uterus, nonreactive endometrium, and distended vagina in prepubertal and pubertal girls [10]. MRI is the gold standard investigation available to confirm the diagnosis, providing detailed information of both internal and external uterine anatomy, and to diagnose associated extragenital anomalies [11].

Laparoscopy can help in confirming the diagnosis, detection, and treatment of the associated complications and was considered to be the gold standard in the management of OHVIRA syndrome but due to the availability of better imaging facilities – 3D

ultrasound and MRI, it is no longer necessary in every case.

Though OHVIRA syndrome is a complex and rare anomaly, it's treatment, however, is relatively simple, comprising either resection or excision of the vaginal septum [12].

Obstructed anomalies, if not treated timely and adequately, may lead to chronic complications related to retrograde menstrual flow like hematocolpos, pyocolpos, pelvic adhesions and endometriosis and negative psychological impact, thereby impairing a woman's quality of life [13]. So, the purpose of this original study is to inform all clinicians, particularly those in reproductive medicine, that OHVIRA Syndrome is a rare congenital anomaly that requires a high index of suspicion and early diagnosis and treatment can avoid long term complications and can improve reproductive outcome.

MATERIALS AND METHODS

The present prospective observational study was conducted in Reproductive Endocrinology and Infertility Department of BSMMU from September 2018 to September 2022.

Among 84 patients of Mullerian Anomaly, 10 patients presented with obstructive features were enrolled in the study. Consent from every patient was taken. Cases were analyzed according to detailed history, thorough examination, relevant investigations with TVS and MRI of whole abdomen and laparoscopy maintaining inclusion and exclusion criteria.

Inclusion Criteria: Didelphys and bicornuate uterus with obstructive features were included in the study.

Exclusion Criteria

Mullerian agenesis, septate uterus, unicornuate uterus, arcuate uterus, transverse vaginal Septum patients were excluded from the study. After Operation patients were followed up for at least 1 year.

RESULTS

Table-1 describes the demographic characteristics of the patient. The mean age was 25.30 ± 5.93 years. All cases of OHVIRA Syndrome were from middle socio-economic condition 6(60%). Maximum patients were from urban area 6(60%). In our study 9(90%) patient presented with severe dysmenorrhoea and 1(10%) with secondary infertility. Right sided obstructed Hemivagina with right renal agenesis were more common 6(60%) than the left side 4(40%).

Six patients (60%) were undergone for Laparoscopic evaluation followed by drainage of the haematocolpos of that side by resection of vaginal septum. Four (40%) patients required excision of functioning, non-communicating uterine horn due to previous treatment failure with vaginal septal resection outside BSMMU. Uterine didelphys was found in 8(80%) patients, bicornuate uterus in 2(20%) and endometriosis was associated in 2(20%) patients.

After Operation patients were followed up for 1 year. Regarding improvement of symptom all the patients 9(90%) previously suffering from dysmenorrhoea is now enjoying normal menstruation without having any pain. But no patient conceived yet.

Table 1: Demographic Profile

Parameters	No. of patients (n=10)	Percentage
Age in years		
<25	05	50%
>25	05	50%
Residence		
Urban	06	60%
Rural	04	40%
Socio-economic Status		
Lower	02	20%
Middle	06	60%
High	02	20%

Table 2: Presenting Symptoms

Parameters	No. of patients (n=10)	Percentage
Dysmenorrhoea	09	90%
Infertility	01	10%

Table 3: Associated Renal Malformation

	No. of patients (n=10)	Percentage
Right Renal Agenesis	06	60%
Left Renal Agenesis	04	40%

Table 4: Operative Findings

	No. of patients (n=10)	Percentage
Didelphys Uterus	**08**	**80%**
Bicornuate Uterus	**02**	**20%**
Obstructed Hemivagina (Right)	**06**	**60%**
Obstructed Hemivagina (Left)	**04**	**40%**
Associated Endometriosis	**02**	**20%**

Table 5: Operative Procedure

	No. of patients (n=10)	Percentage
Excision of the obstructed vaginal septum	**06**	**60%**
Excision of functioning, non communicating uterine horn	**04**	**40%**

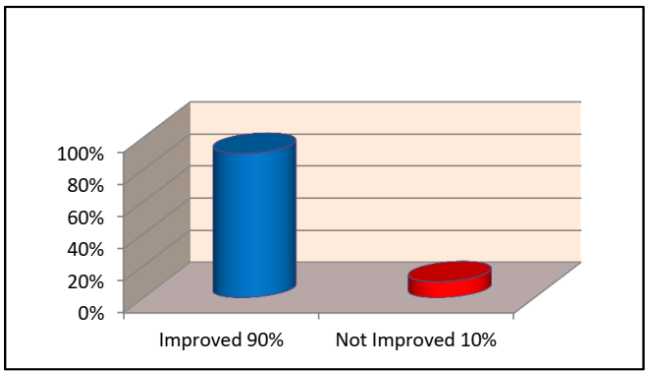


Fig. 1: Improvement of Symptom

DISCUSSION

In the literatures OHVIRA syndrome has shown varied predilection related to the sides involved—some claiming right sided abnormality being more common, some left sided, while others claimed equal prevalence on both sides [14, 15]. In our study it was more common on right side (60%). Although OHVIRA syndrome is common with didelphic uterus, but also found with bicornuate uterus [16]. In this study, most of the patients (80%) had didelphys uterus whereas 20% was with bicornuate uterus.

In a completely obstructed hemivagina (class 1), with the onset of menarche, there is a progressive collection of blood in the vagina, uterus, and fallopian tubes. If the obstruction is still not relieved, reflux of menstrual blood in the peritoneal cavity causes endometriosis, pelvic adhesions, and pelvic infections, leading to infertility. In patients with class 1, HWWS presents early after menarche with severe dysmenorrhoea, nonspecific lower abdominal pain, abdominal lump, peritonitis, and urinary tract obstruction. In patients with incomplete obstruction (class 2), it usually presents late with recurrent and persistent purulent vaginal discharge or PID. In our study 90% patients presented with severe dysmenorrhoea and 10% with secondary infertility.

Though OHVIRA syndrome is a complex and rare anomaly, its treatment, however, is relatively simple, comprising either resection or excision of the vaginal septum. Only in class 1.2 (cervico-vaginal atresia with complete obstruction) ipsilateral hysterectomy, either abdominal or laparoscopic, would be required as in these cases apart from an obstructing vaginal septum, there is obstruction at another higher level, i.e., cervix [9]. For those who refuse partial hysterectomy, the option of uterovaginal canalization but adequate counselling regarding restenosis of the neocervix and future complication is must [17]. In our study, 60% patients were undergone for Laparoscopic evaluation followed by drainage of the haematocolpos of that side by excision of obstructed vaginal septum. Forty percent patients required excision of functioning, non-communicating uterine horn due to previous treatment failure.

In timely and adequately treated patients with OHVIRA syndrome, reproductive performance is consistent with that of a didelphic uterus. Pregnancy rate in obstructed mullerian anomalies varies from 37–40% [18]. Though no patients conceived yet in our study but 90% patients improved from dysmenorrhoea and are enjoying normal menstrual cycle without having any pain. Hence, early diagnosis and treatment entail a good reproductive outcome of this rare syndrome.

CONCLUSION

When diagnosis of OHVIRA Syndrome is not delayed and indication and timing of surgery are appropriate, complications can be avoided and fertility can be preserved.

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CONFLICT OF INTEREST: The authors have no conflicts of interest.

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