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Herlyn Werner Wunderlich Syndrome: A Very Rare Presentation in a Teenage Girl

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Abstract **Case Report**

Background: Uterus didelphys with blind hemivagina Ipsilateral renal agenesis (Herlyn Werner Wunderlich Syndrome) is a rare congenital anomaly. The true incidence of this anomaly is unknown, but it has been reported between 0.1% and 3.8%. Another name is"Double uterus-hemivaginal-renal agenesis syndrome." ***Case Summary:*** We present the case of Uterus didelphys with hemivagina with ovarian endometrioma ē Ipsilateral renal agenesis in a 14-year-old girl. Patient was symptomatically treated. She was then referred to BSMMU on 28-02-22 for further management. ***Conclusion:*** Neglected and inappropriately managed Uterus didelphys with hemivagina with ovarian endometrioma ē Ipsilateral renalagenesis can lead to endometriosis and infertility.

Keywords: Uterus didelphys, Herlyn Werner Wunderlich Syndrome, Double uterus-hemivaginal-renal agenesis syndrome.

INTRODUCTION

The Herlyn Werner Wunderlich Syndrome is a very rare combined Mullerian and Mesonephric duct abnormality, typically characterized by uterus didelphys and obstructed hemivagina with ipsilateral renal agenesis. There is failure of fusion of mullerian duct producing double uterus. Obstructed hemivagina results from transvaginal septum producing blind vagina. The renal agenesis of the same side seen. Usually it appears after menarche with pelvic pain which is progressive in nature and causes dysmenorrhea, it is secondary to hematocolpos. Diagnosis should be done before development of complications. So, awareness is necessary to diagnosis earlier and to treat this disorder in time. Ultrasonography of whole abdomen (KUB) is necessary but MRI is preferred. If any renal abnormity of encountered proper screening should be made to detect developmental congenital abnormalities of the genitourinary tract with both renal conditions. Patient with this syndrome is asymptomatic until menarche and can follow other complication which may be irreversible.

CASE REPORT

A 14 years old patient, weight 45 kg from Mothertak presents with a clinical feature of acute lower abdominal pain. She has cyclical dysmenorrhoea. Pain starts from the right lower back having radiator to the right thigh. Pain is progressive in nature increasing in the last few months. Her bladder & bowel habits are normal with no history of haematuria. Her menarche was at 11 years of age and irregular cycle with average flow. Dysmenorrhea started since 8 months.

Pain is occasionally associated with vomiting. She had occasional abdominal cramping. She is the first child of the non-consanguineous marriage, delivered by elective c- sections at term. Her birth weight was 3.2kg had no perinatal complications. The girl has well developed breasts according to her age. Duly immunized with EPI protocol. The general examination revealed a 45kg body weight. All parameters are within normal limits with normal hair distribution. The abdomen was soft, with no distention. No palpable mass or lesion was seen. Marphy's sign was negative. Mcburney's point was not tender.

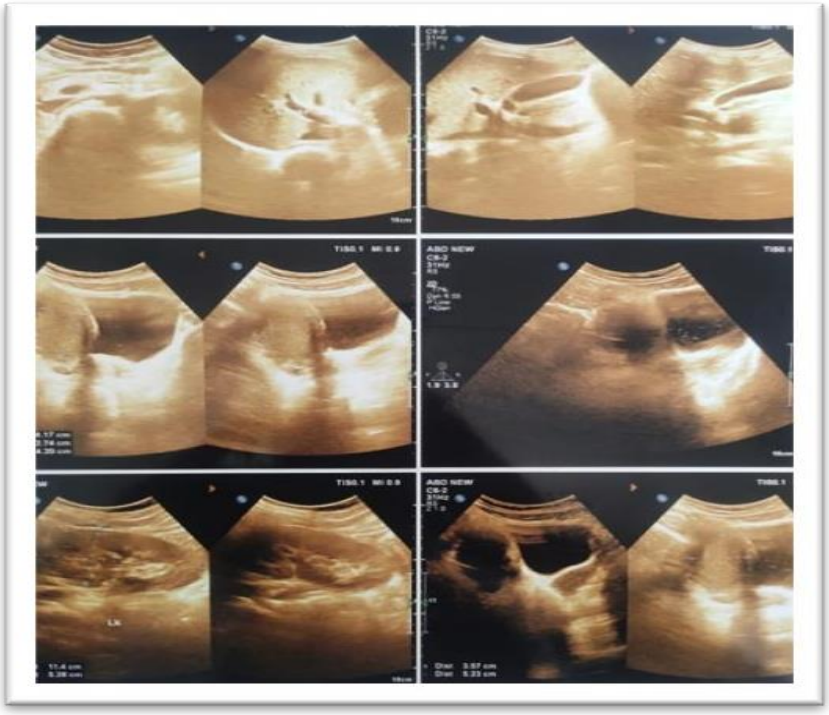
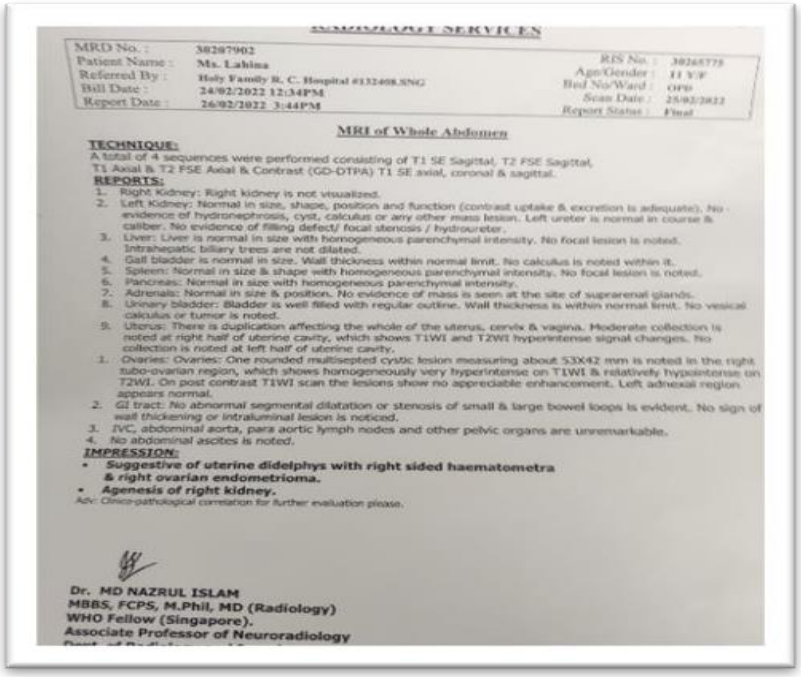


Figure I: Ultra sonogram Findings



MRD No. : 30287902
Patient Name : Ms. Lahina
Referred By : Holy Family R. C. Hospital #132408.SNG
Bill Date : 24/02/2022 12:34PM
Report Date : 26/02/2022 3:44PM

RIS No. : 30265778
Age/Gender : 11 YF
Bed No/Ward : OPD
Scan Date : 25/02/2022
Report Status : Final

MRI of Whole Abdomen

TECHNIQUE:
A total of 4 sequences were performed consisting of T1 SE Sagittal, T2 FSE Sagittal, T1 Axial & T2 FSE Axial & Contrast (GD-DTPA) T1 SE axial, coronal & sagittal.

REPORTS:
1. Right Kidney: Right kidney is not visualized.
2. Left Kidney: Normal in size, shape, position and function (contrast uptake & excretion is adequate). No evidence of hydronephrosis, cyst, calculus or any other mass lesion. Left ureter is normal in course & caliber. No evidence of filling defect/ focal stenosis / hydroureter.
3. Liver: Liver is normal in size with homogeneous parenchymal intensity. No focal lesion is noted. Intrahepatic biliary trees are not dilated.
4. Gall bladder is normal in size. Wall thickness within normal limit. No calculus is noted within it.
5. Spleen: Normal in size & shape with homogeneous parenchymal intensity. No focal lesion is noted.
6. Pancreas: Normal in size with homogeneous parenchymal intensity.
7. Adrenals: Normal in size & position. No evidence of mass is seen at the site of suprarenal glands.
8. Urinary bladder: Bladder is well filled with regular outline. Wall thickness is within normal limit. No vesical calculus or tumor is noted.
9. Uterus: There is duplication affecting the whole of the uterus, cervix & vagina. Moderate collection is noted at right half of uterine cavity, which shows T1WI and T2WI hyperintense signal changes. No collection is noted at left half of uterine cavity.
1. Ovaries: Ovaries: One rounded multiseipted cystic lesion measuring about 53X42 mm is noted in the right tubo-ovarian region, which shows homogeneously very hyperintense on T1WI & relatively hypointense on T2WI. On post contrast T1WI scan the lesions show no appreciable enhancement. Left adnexal region appears normal.
2. GI tract: No abnormal segmental dilatation or stenosis of small & large bowel loops is evident. No sign of wall thickening or intraluminal lesion is noticed.
3. IVC, abdominal aorta, para aortic lymph nodes and other pelvic organs are unremarkable.
4. No abdominal ascites is noted.

IMPRESSION:
- **Suggestive of uterine didelphys with right sided haematometra & right ovarian endometrioma.**
- **Agenesis of right kidney.**

Adv: Clinico-pathological correlation for further evaluation please.

Dr. MD NAZRUL ISLAM
MBBS, FCPS, M.Phil, MD (Radiology)
WHO Fellow (Singapore).
Associate Professor of Neuroradiology

Figure II: Ultra sonogram Findings

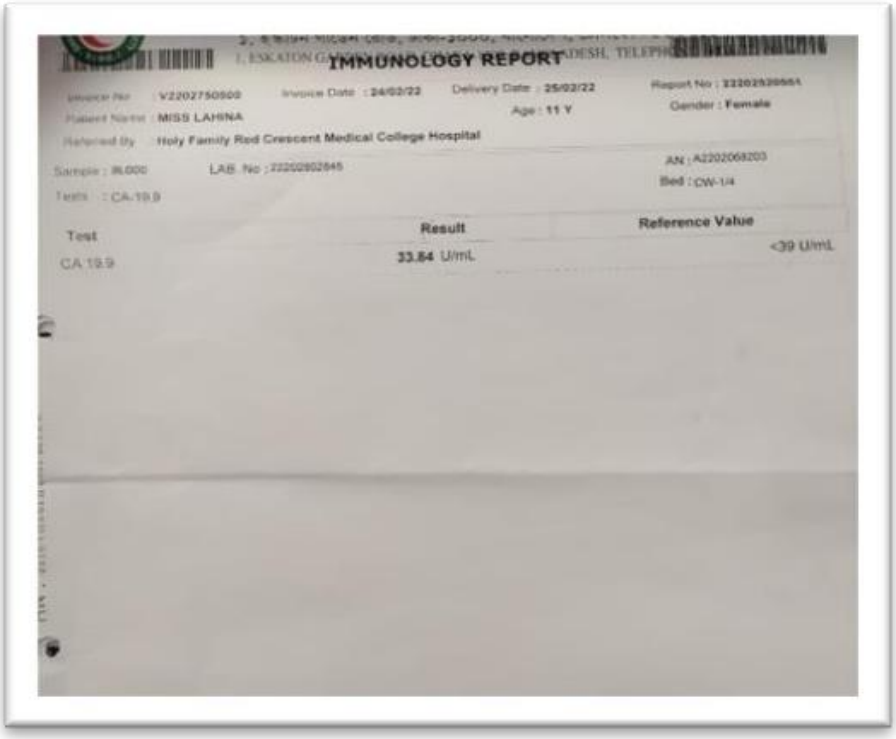
Ultrasonography of the abdomen and pelvis evaluated there was the absence of a right kidney in the night renal fossa which was occupied by bowel loops. The left kidney was normal. Uterus didelphys was noted, right uterine cavity and cervical canal showed a hypoechoic collection with multiple internal echos suggestive of haematocolpos. The left uterine horn with cervical canal appeared normal.

Magnetic resonance imaging (MRI) manifestation HWWS characterized by the double uterus, obstructed hemivagina and ipsilateral renal agenesis.

Table 1: Summary of Laboratory investigations of the case of HWWS, done in Holy Family Red Crescent Medical Hospital

Test	Result	Reference Value
Hemoglobin ESR Total count RBC	14.2 g/dl 22 mm/1st hr	F: 11-5- 165 g/dl F: 0-20
Platelets White blood cells	3,04,000 8,090	1,50,000 - 4,50,000 4,000 11,000
Differential count Neutrophil Lymphocyte Eosinophil PCV	66% 29% 2% 40.7%	40-75% 20-50% 1- 6% F-30-46%
S. Electrolytes Sodium Potassium C- Reactive Protein Creatinine	141.0 mmol/l 4.0 mmol/l 2 mg/dl 0.59 mg/dl	136-145 mml/l 3.5-5.2 mmol 0.0-5.0 mg/dl 0.6-1.3 mg/dl
Urine R/M/E Epithelial cell Pus cell Serum CA-125 CA 19.9	0-2 /HPF 2-4 /HPF 286.16 u/ml 33.84 u/ml	1-5 /HPF 0-5 /HPF Up to 35 u/ml <39 u/ml
DTPA Renal Study Left kidney Right kidney	Normal function Not visualized	
X-Ray of KUB Region	Normal X-Ray of KUB	
CXR, P/A view		
USG of W/A+KUB+PUR	- Features of UTI (Cystitis) - Features of fused ectopic Kidney at left renal fossa - Rd adnexal complex mass (dermoid cyed)	
MRI	- Suggestive of uterine didelphys ē right sided hematometra 4 right ovarian endometrioma. - Agenesis of right Kidney	

Others Reports



IMMUNOLOGY REPORT

Invoice No : V2202750500 Invoice Date : 24/02/22 Delivery Date : 25/02/22 Report No : 22202520661

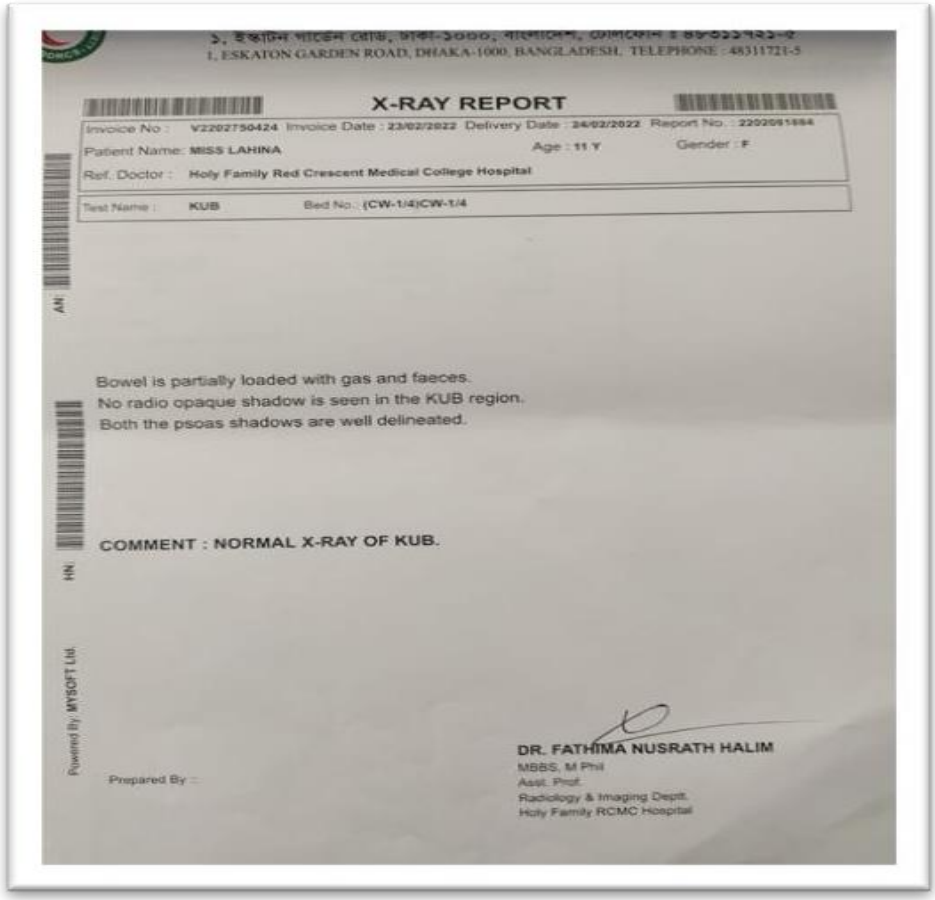
Patient Name : MISS LAHINA Age : 11 Y Gender : Female

Referred By : Holy Family Red Crescent Medical College Hospital

Sample : BLOOD LAB No : 22202802645 AN : A2202068203

Tests : CA-19.9 Bed : CW-1/4

Test	Result	Reference Value
CA 19.9	33.84 U/mL	<39 U/mL



১, ইস্কাটন গার্ডেন রোড, ঢাকা-১০০০, বাংলাদেশ, টেলিফোন : ৪৮৩১১৭২১-৫
1, ESKATON GARDEN ROAD, DHAKA-1000, BANGLADESH, TELEPHONE : 48311721-5

X-RAY REPORT

Invoice No : V2202750424 Invoice Date : 23/02/2022 Delivery Date : 24/02/2022 Report No. : 2202/01884
Patient Name: MISS LAHINA Age : 11 Y Gender : F
Ref. Doctor : Holy Family Red Crescent Medical College Hospital
Test Name : KUB Bed No. : (CW-1/4)CW-1/4

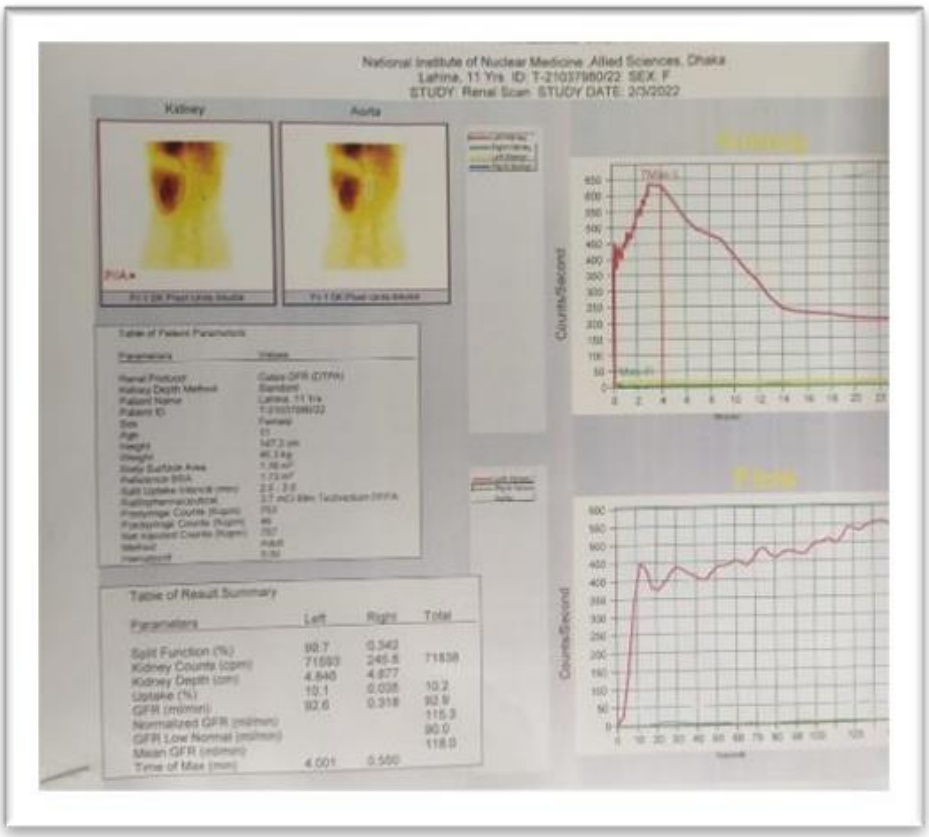
AN:
HN:
Powered By: MYSOFT Ltd.

Bowel is partially loaded with gas and faeces.
No radio opaque shadow is seen in the KUB region.
Both the psoas shadows are well delineated.

COMMENT : NORMAL X-RAY OF KUB.

Prepared By :

DR. FATHIMA NUSRATH HALIM
MBBS, M Phil
Asst. Prof.
Radiology & Imaging Deptt.
Holy Family RCMC Hospital



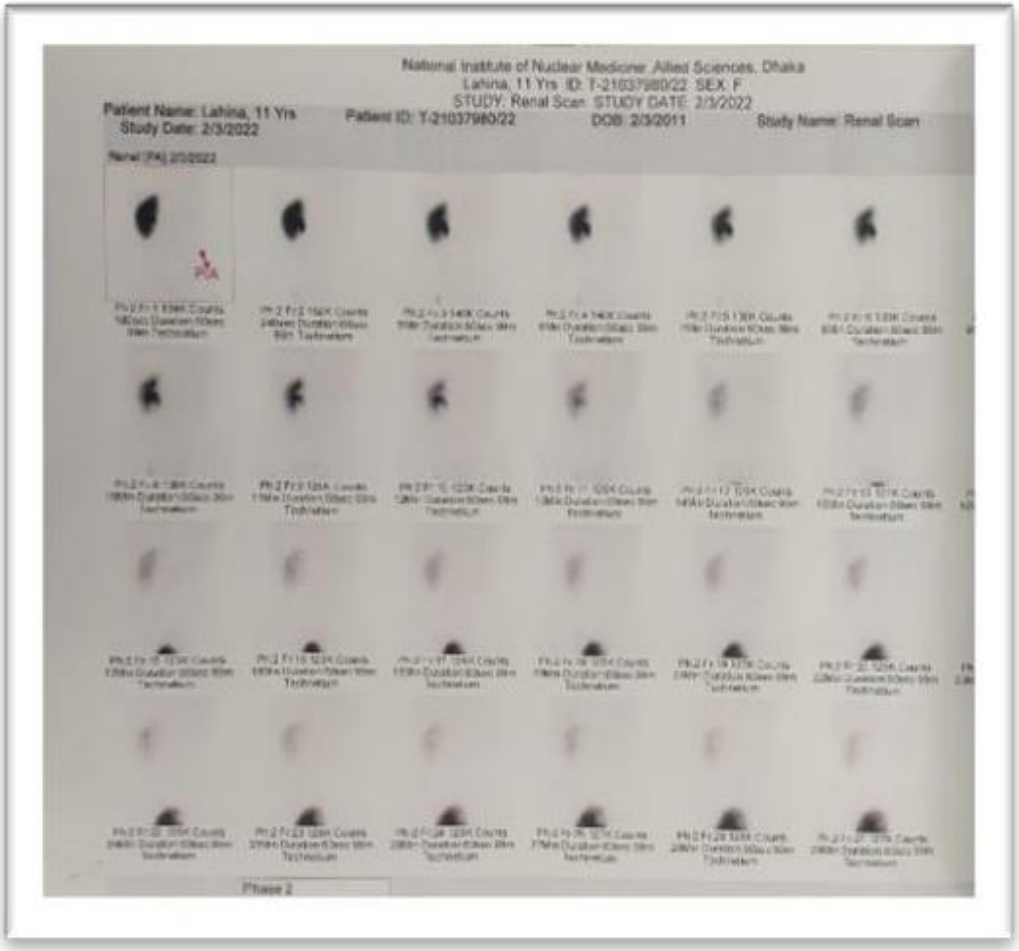
National Institute of Nuclear Medicine, Allied Sciences, Dhaka
Lahina, 11 Yrs ID: T-21037980/22 SEX: F
STUDY: Renal Scan STUDY DATE: 2/3/2022

Kidney Aorta

Table of Result Summary

Parameters	Left	Right	Total
Split Function (%)	99.7	0.342	
Kidney Counts (cpm)	71593	246.6	71839
Kidney Depth (cm)	4.846	4.877	
Uptake (%)	10.1	0.035	10.2
GFR (ml/min)	92.6	0.318	92.9
Normalized GFR (ml/min)			115.3
GFR Low Normal (ml/min)			90.0
Mean GFR (ml/min)			118.0
Time of Max (min)	4.001	0.560	

Counts/Second



DISCUSSION

The Herdlyn-Werner-Wunderlich syndrome (Renal agenesis with ipsilateral blind hemivagina) shortly known as HWWS was initially described by Herlyn and Werner in 1971. But HWWS was not first reported by Herlyn and Werner. However, HWWS is such a rare congenital anomaly of the genital area which was first reported by Purslow in 1922 [1]. Almost after fifty years of Purslow's reports, the association of renal agenesis with ipsilateral blind hemivagina was reported as Herlyn-Werner Syndrome. Following a few years later in the presence of an isolated hematocervix" in 1976, Wonderlich labeled it as" an association of right renal aplasia with a bicornuate uterus and simple vagina [2]. The true incidence of Hwws is unknown, but it has been reported between 0.1 to 3.8%. The estimated overall prevalence of Mullerian duct anomaly is believed to be 2-3% of women. Uterus didelphys constitutes approximately 11% of MDA's. Associated renal anomalies are present in 43%. Among the patient with uterus didelphys, 75% have a partial or complete vaginal septum which is usually longitudinal in HWWS. After the proper treatment, about 80% of patients with uterus didelphys are able to get pregnant with elevated rates of premature delivery (22%) of Abortion (74%). C/S may be needed in over 80% of patients [3]. HWWS is a combination of Mullerian of 4 mesonephric duct anomaly which is characterized by the classical triad of uterus didelphys, obstructed hemivagina of ipsilateral renal agenesis. Mullerian duct develops from the coelomic epithelium ultimately forming the two uterovaginal canals, On the other hand, the sino vaginal bulbs rise from the urogenital sinus to meet the caudal end of the fused Mullerian ducts. To form the vaginal plate. Then the lower part of the vagina is formed by the reabsorption to canalization of the vaginal plate. Uterus didelphys results from the failure of fusion of the paired Mullerian ducts, Obstructed hemivagina represents the failure of vertical fusion resulting in non-resorption of tissue between the vaginal plate that rises from the urogenital sinus & the caudal aspect of paired Mullerian ducts. The renal agenesis on the ipsilateral side of the obstructed hemivagina indicates the developmental anomaly of one walfian duct. HWWS is classified into two, one is complete where there is completely obstructed hemivagina and another is incompletely obstructed hemivagina. There are several clinical presentations including abdominal pain, dysmenorrhea, urinary retention, hematosalpinx, endometriosis, and rupture of a tube- ovarian abscess. If HWWS is not treated properly or if there is a delay in diagnosis of this symptom many complications may arise such as

infertility, endometriosis and spontaneous abortion. As an initial investigation, we perform an abdominal USG which will reveal any anatomic anomaly. MRI is the leading choice for diagnosing the HWWS. However, it should be noted that MRI may not define cervical anatomy which may change the operative management [4]. In most cases, the first choice of management is surgical resection of the vaginal septum and preserving the hemiuterous which relieves the obstruction, improves the symptoms, and clogs the complications of retrograde flow. Routine follow-up and USG is important for this syndrome as it is associated with ipsilateral renal agenesis. Laparoscopy also can be done if the patient has concurred endometriosis or any other adhesions or rare occasion's laparoscopy may be done for confirming the diagnosis. One group used indigocermine die injected into the cervix during laparoscopy to aid in anatomic classification [5]. Most patients with OHVIRA syndrome can be treated solely with single-stage vaginoplasty. Routine laparoscopy is not essential to management. Vaginal stenosis is a postoperative possibility, and may be associated with vaginal adenosis [6]. The complexity of these cases, especially the evolving manifestation of cervical aplasia postoperatively, illustrates the need to recognize limitations in imaging and divergence in definitive management [7].

CONCLUSION

In the conclusion part, I would write that several issues make it difficult to make an accurate diagnosis of HWW syndrome. An unusual presentation of regular menstruation and accurate and nonspecific abdominal pain are the two common problems, and these require special clinical suspicion. But at the same time, there are some pre-cautionary issues that might be helpful to minimize the severeness of this syndrome. For example, early identification of these problems (unusual presentation of regular menstruation, and non-specific abdominal pain) warrants medical consciousness of such anomaly. In addition, there are some medical practices such as Ultrasonography and MRI findings that can collectively play a role to diagnose this abnormal syndrome. Last but not the least, a collective approach from multidisciplinary perspectives including expertise from gynecologists, Radiologists, pediatricians, and pediatric Surgeons is unequivocally important to avoid any complications and achieve better treatment.

Abbreviations

HWWS: Herlyn Werner Wunderlich Syndrome USG: Ultrasonography.
MRI: Magnetic Resonance Imaging.
BSSMU: Bangabandhu Sheikh Mujib Medical University Hospital.
OHVIRA: Obstructed hemivagina and ipsilateral W/A: Whole renal anomaly.
PV- Per vaginal UTI- Urinary tract infection.

AUTHOR CONTRIBUTION

All authors made a significant contribution to the work reported, whether that is in the conception, study design, data analysis, and interpretation.

FUNDING

No funding source.

DECLARATIONS

Consent for Publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

REFERENCES

1. Purslow, C. E. (1922). A case of unilateral haematocolpos, hematometra and haematosalpinx. *J Obstet Gynaecol Br Emp*, *29*, 643. https://doi.org/10.1111/j.1471-0528.1922.tb16100.x
2. Jia, G., Chai, W., Cui, M., Wen, Y., Cui, L., & Gong, F. (2018). A case report on Herlyn–Werner–Wunderlich syndrome with spontaneous abortion. *Medicine*, *97*(36).
3. Piccinini, P. S., & Doski, J. (2015). Herlyn-Werner-Wunderlich syndrome: a case report. *Revista Brasileira de Ginecologia e Obstetrícia*, *37*, 192-196.
4. *J Pediatr Adolesce Gynecol*, 27 (2014), PP, 375-378, 10,1016/ j. jpag 2014. 02. 001
5. Ottman, N., Ruokolainen, L., Suomalainen, A., Sinkko, H., Karisola, P., Lehtimäki, J., ... & Fyhrquist, N. (2019). Soil exposure modifies the gut microbiota and supports immune tolerance in a mouse model. *Journal of allergy and clinical immunology*, *143*(3), 1198-1206. Doi: 10.1016/J. fertnstert, 2006.11.015
6. Smith, N. A., & Laufer, M. R. (2007). Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome: management and follow-up. *Fertility and sterility*, *87*(4), 918-922. doi: 10.1016/j.fertnstert.2006.11.015. Epub 2007 Feb 22. PMID: 17320871.
7. Sabdia, S., Sutton, B., & Kimble, R. M. N. (2014). The obstructed hemivagina, ipsilateral renal anomaly, and uterine didelphys triad and the subsequent manifestation of cervical aplasia. *Journal of pediatric and adolescent gynecology*, *27*(6), 375-378. 1.doi: 10.1016/j.jpag.2014.02.001. Epub 2014 Sep 23. PMID: 25256882.