International Journal of Medical Science in Clinical Research and Review
Online ISSN: 2581-8945
Available Online at http://www.ijmscrr.in Volume 7|Issue 02 (March-April) |2024 Page: 184-186

Case Report

Vaginoscopic Septotomy of a Blind Right Vagina in Virgin Adolescent with d'Herlyn-Werner-Wunderlich Syndrome: A case report

Authors:
AMAL MOUZALI[1,2], Faycal Makhlouf [3], Nabila Mechri[1], Faiza Nemmour[1], Youcef Tayebi[1]
[1,2]Gynecology and Obstetrics Department. Douera University Hospital, Algiers, Algeria
3: Epidemiology Department. Tipaza Hospital. Algeria
Corresponding Author:
AMAL MOUZALI
Gynecology and Obstetrics, Department. Douera University Hospital, Algiers, Algeria, ORCID: https://orcid.org/0000-0002-5985-7143

Article Received: 05-February-2024, Revised: 25-February-2024, Accepted: 16-March-2024

ABSTRACT:
Herlyn-Werner-Wunderlich syndrome is a rare congenital anomaly of the urogenital system. It is defined by the triad of a didelph uterus, a blind hemivagina and ipsilateral renal agenesis. Diagnosis is usually made shortly after menarche. We present a case report and surgical video of a 14-year-old virgin girl treated by vaginoscopically septotomy of her complet blind right hemivagina. Therefore, we preserved the integrity of the hymen, concluding that endoscopy is useful and effective in this pathology.

Keywords: Hysteroscopy, Herlyn-Werner-Wunderlich syndrome

INTRODUCTION:

Herlyn-Werner-Wunderlich syndrome is a rare congenital anomaly of the urogenital tract and it's also called OHVIRA syndrome (Obstructed hemivagina and ipsilateral renal anomaly). It is defined by the triad of a didelphys uterus, a blind hemivagina and ipsilateral renal agenesis. The diagnosis is usually made shortly after menarche [1,2].

METHODS:

A case report, surgical video and technique for a vaginoscpic section and resection of a hemiblind vagina at Douera university hospital in Algeria, for management of hematocolpos and right hematometria causing intense dysmenorrhea. Informed Consent was obtained from the patient and her parents for presentation and publication.

CASE REPORT:

We present a case report of a 14-year-old virgin girl which was referred to our hospital's gynecology clinic for intense dysmenorrhea that she has had since her menarche (04 months before). She had normal menstruation from the non-obstructed side, but her hemi-vaginal obstruction caused cyclical intense pain of 10/10 from visual analogue scale VAS. Ultrasound and MRI explorations as showed in Figure 01, revealed the presence of a uterovaginal malformation represented by blind right hemivagina, which caused right hematometria as well as a unilateral renal agenesis, classified U3b, C2, V2 according to ESHRE /ESGE classification of congenital anomalies. Early intervention was needed to reduce risk of endometriosis and infertility.

DESCRIPTION OF SURGICAL TECHNIQUE:

The treatment was a vaginal septotomy with hysteroscopic edges resection in this virgin adolescent. Under rachianethesia we performed vaginoscopy through the intact hymen and the patent left vagina, we found a left cervix and a right vaginal wall bulging caused by the hemaocolpos behind this blind oblique vaginal septum. We introduced the Gubbini mini resectoscope 18,5 French through the intact hymen and we made about 06 cm of lengh section on the right vagina using a bipolar current mode after vaginal distension with saline solution. Figures 2,3. The straight vertical vaginal section allowed drainage of dark bloody secretions that had collected behind the blind vagina. The opening of the right vagina made it possible to identify the right cervix which was widened. After that we resected the anterior and posterior edges of the septum. Figure4. In the same time, we performed left and right hysteroscopy and we found an endometritis on the right uterus caused probably by the right hematometria. This resection was done without any complications per or post sugery. To be sure that the vagina remains permeable, we carried out a vaginoscopic control in the office 05 months

after the first surgery and the result was good with patency of the right vagina and therefore her problem was definitely resolved. Figure 5

DISCUSSION:

Blind vagina in the OHVIRA syndrome, was treated in the past by vaginal root which caused injuries to the hymen. However, keeping the vagina intact is really important in some culture as ours, indeed endoscopy allows us to perform cutting with precision. The vagina remains patent without any other procedures like a balloon inside and restenosis of the vagina was not observed at follow-up. Previously, laparoscopy was used for the diagnosis of OHVIRA or Herlyn-Werner-Wunderlich syndrome, but recently thanks to advanced imaging technologies, laparoscopy should be reserved only for cases where the diagnosis is not clear after imaging.

Kim et al [3], reported hysteroscopic resection of the septum under transabdominal guidance in order to preserve the hymenal integrity. But in our case, we didn't use any ultrasound exam within the septum resection and we could keep the hymen intact. Moreover, the vagina kept patent after the office vaginoscopy control 05 months after the first surgery. In our case the right vagina side was blinded also in the study of Uribe et al in 2019(4), this side anomaly was observed in about half of the patients. Gungor et al, stated that the main treatment is single-stage vaginoplasty and most OHVIRA cases do not develop recurrent occlusion during follow-up and may deliver [5]. It's clear that this septotomy by resectoscope through an intact hymen is easy and safe and allows to keep the hymen integrity of the adolescent, as several authors mentioned on their publications [6,7,8,9,10] Indeed, office vaginoscopy is worth doing in the virgin adolescent.

CONCLUSION:

Vaginiscopic septotomy is a virginity sparing surgery that is described as the treatment of choice for the blind vaginal anomaly in the OHVIRA syndrome. It's a safe, feasible and effective procedure with a low rate of complications.

CONFLICT OF INTEREST STATEMENT:

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

FUNDING:

There were no sources of funding in this work.

Ethical Approval:

Our institution does not require ethical approval for reporting individual cases or case series.

Data available on Request:

through the corresponding author.

CONSENT:

A consent form has been completed by the Douera Hospital.

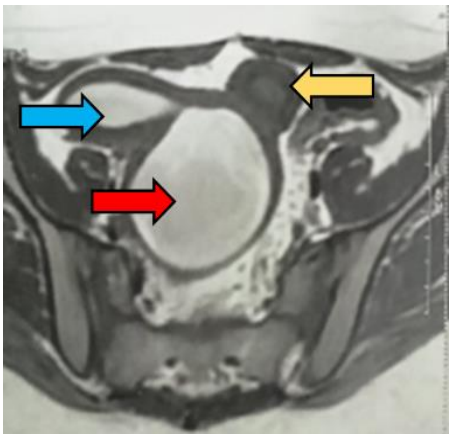

Figure 1: MRI showed Uterus didelphys with a blind right hemivagina which caused hematocolpos (red arrow)and hematometria(blue arrow). The normal left uterine horn is showed by the (yellow arrow)

Figure2: Opening of the right bulging vaginal wall

Figue3: Resection of vaginal septum

Figure 4: Resection of edges of vaginal septum

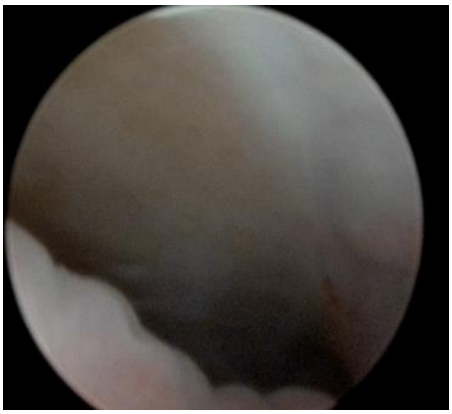

Figure 5: Office vaginoscopy 05 months after surgery

REFERENCES:

1. Herlyn U, Werner H. Simultaneous occurrence of an open Gartner-duct cyst, a homolateral aplasia of the kidney and a double uterus as a typical syndrome of abnormalities. Geburtshilfe Frauenheilkd 1971;31:340–7.
2. Wunderlich M. Unusual form of genital malformation with aplasia of the right kidney. Zentralbl Gynakol 1976;98:559–62.
3. Kim TE, Lee GH, Choi YM, Jee BC, Ku SY, Suh CS, et al. Hysteroscopic resection of the vaginal septum in uterus didelphys with obstructed hemivagina: a case report. J Korean Med Sci 2007;22(4):766–9.
4. Siu Uribe A, Vargas Cruz V, Murcia Pascual FJ, Escassi Gil A, Garrido Pérez JI, Antón Gamero M, Paredes Esteban RM. Síndrome de OHVIRA: características clínicas y complicaciones, nuestra experiencia [Clinical characteristics and complications in patients with OHVIRA (obstructed hemivagina and ipsilateral renal anomaly) syndrome. Our experience]. Cir Pediatr. 2019 Jan 21;32(1):11-16. Spanish. PMID: 30714695.
5. Gungor Ugurlucan F, Dural O, Yasa C, Kirpinar G, Akhan SE. Diagnosis, management, and outcome of obstructed hemivagina and ipsilateral renal agenesis (OHVIRA syndrome): Is there a correlation between MRI findings and outcome? Clin Imaging. 2020 Feb;59(2):172-178. doi: 10.1016/j.clinimag.2019.11.013. Epub 2019 Nov 30. PMID: 31821975.
6. Moneir WM, Almodhen F, Almaary J, Almatar Z, Alaqeel A. Vaginoscopic Incision of Vaginal Septum With Preservation of the Hymen in a Child With Obstructed Hemi-Vagina Ipsilateral Renal Agenesis (OHVIRA) Syndrome. Cureus. 2022 Oct 19;14(10):e30450. doi: 10.7759/cureus.30450. PMID: 36407203; PMCID: PMC9672824.
7. Cheng C, Subedi J, Zhang A, Johnson G, Zhao X, Xu D, Guan X. Vaginoscopic incision of oblique vaginal septum in adolescents with OHVIRA syndrome. Sci Rep. 2019;9(20042) doi: 10.1038/s41598-019-56471-2. [PMC free article] [PubMed] [CrossRef] [Google Scholar]
8. Johary J, Xue M, Xu B, Xu D, Aili A. Use of hysteroscope for vaginoscopy or hysteroscopy in adolescents for the diagnosis and therapeutic management of gynecologic disorders: A systematic review. J Pediatr Adolesc Gynecol. 2015;28:29–37. doi: 10.1016/j.jpag.2014.02.014. [PubMed] [CrossRef] [Google Scholar]
9. Shih CL, Hung YC, Chen CP, Chien SC, Lin WC. Resectoscopic excision of the vaginal septum in a virgin with uterus didelphys and obstructed unilateral vagina. Taiwan J Obstet Gynecol. 2010;49:109–111. doi: 10.1016/S1028-4559(10)60022-8.
10. Kriplani A, Dalal V, Kachhawa G, Mahey R, Yadav V, Kriplani I. Minimally Invasive Endoscopic Approach for Management of OHVIRA Syndrome. J Obstet Gynaecol India. 2019 Aug;69(4):350-355. doi: 10.1007/s13224-019-01240-4. Epub 2019 Jun 10. PMID: 31391743; PMCID: PMC6661036.