



OBSTRUCTED HEMIVAGINA IPSILATERAL RENAL AGENESIS (OHVIRA) IN MALAYSIA: A CASE REPORT

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INTRODUCTION

- The incidence of Herlyn-Werner-Wunderlich Syndrome or Obstructed Hemivagina Ipsilateral Renal Agenesis (OHVIRA) syndrome among the female population was documented as ranging from 0.1 to 3.8%.¹
- The median age of presentation is between 14-16 years median of age².
- The commonest presentation is abdominal mass or cyclical pelvic pain, often diagnosed during puberty².
- Complications of OHVIRA include subfertility, chronic pelvic pain, and endometriosis³.
- MRI is considered the gold standard diagnostic tools, especially in aiding further surgery planning⁴.

CASE REPORT

A 14-year-old Malay girl with no known medical illness, presented to the hospital with suprapubic pain for 1 week. However, she denies any other symptoms. Upon further history, patient attains menarche at 12 years old with regular cycle of about 28 to 30 days. Each cycle consists of 5 to 6 days of menses, no dysmenorrhea or menorrhagia. Abdominal examination revealed mass palpable at 14 weeks. External genitalia examination noted bulging observed at left vaginal wall. Transabdominal ultrasound shows normal uterus seen, but noted hematocolpos with the size of 8.9cm x 7cm and hematometra seen.

The right kidney was visualized under ultrasound but left kidney was absent. Her renal function test was otherwise normal. A magnetic resonance imaging (MRI) of the pelvis was then performed, which revealed complete duplication of the uterus and cervix in keeping with didelphys. There was also a well-defined cystic structure posterior to the bladder, suggestive of hematocolpos and the left uterine cavity was mildly distended suggestive of hematometra. The right kidney was normal in size and in its normal location. but the left kidney was not visualized. MRI findings were suggestive of Herlyn-Werner-Wunderlich syndrome. Subsequently, an examination under anaesthesia (EUA) and vagina septoplasty and drainage of hematocolpos were done. Intraop finding noted bulge from the left vagina wall.

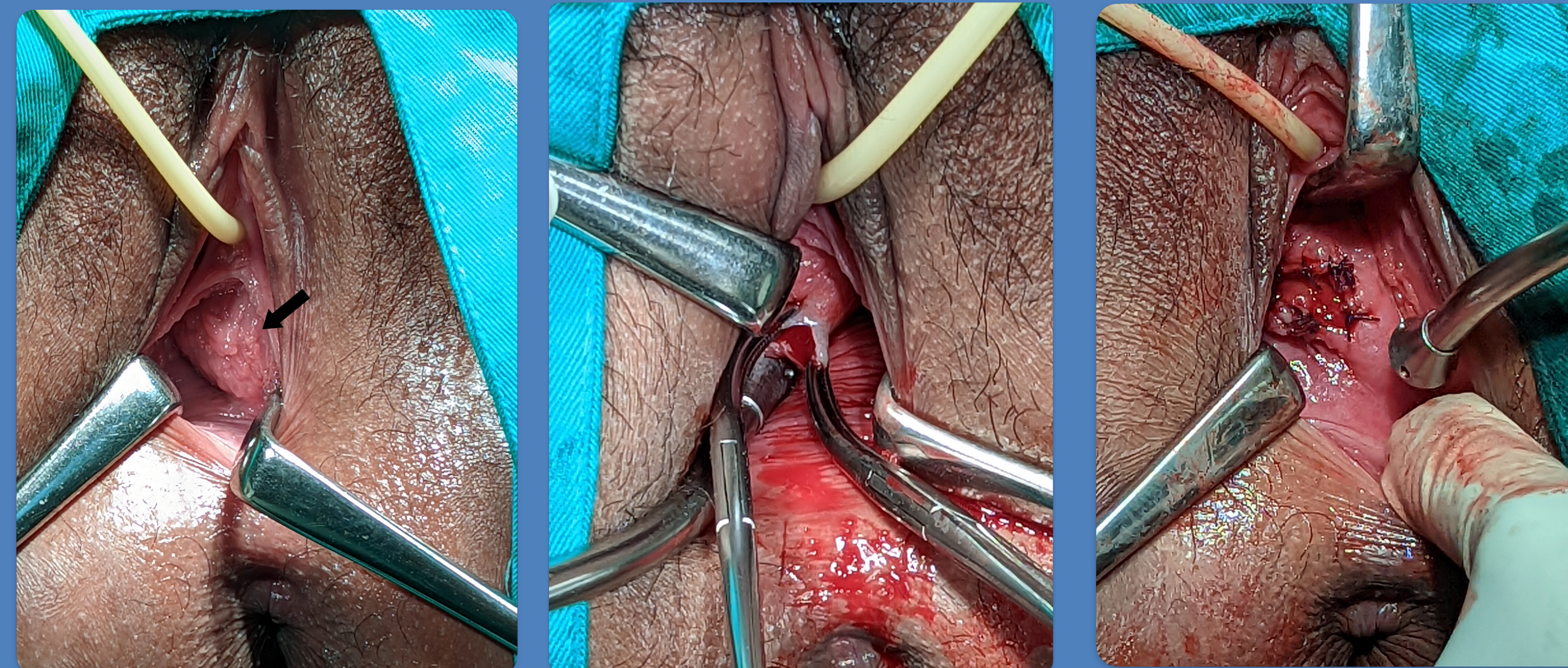


Figure 1. A: Intraoperative finding shows a bulge at the left vagina, resulted from hematocolpos (black arrow). B: The resection of the vaginal septum with drainage of the hematocolpos containing chocolate material content. C: The incised septum was sutured to the vaginal wall and the cavity was left open.

Chocolate material was aspirated from the left vaginal wall and septoplasty was then performed by doing incision over the vaginal septum, draining up to 200ml of chocolate material content. Post septoplasty, two cervixes were felt and they were both normal. The hymenal ring and posterior fourchette were then intersected. Patient was well post operatively and was discharged day 1 post operation. Transabdominal scan during clinic follow up noted didelphys of uterus, with no more haematometra or hematocolpos seen.

DISCUSSION

The definitive management of OHVIRA syndrome is vaginoplasty, which is resection of the vaginal septum to drain the hematocolpos and hematometra. Hemihysterectomy is rarely needed and often was only carried out due to complications from OHVIRA such as infection^{3,4}. Laparoscopy may be used if there is presence of endometriosis or hydrosalpinx at the same setting of valvuloplasty. In our case, we performed a single stage vaginoplasty under open method. A two stage vaginoplasty may also be done whereby in the first stage, the hematocolpos was drained first and then followed up by complete resection of the vaginal septum in a different setting⁵. However, in certain case reports, vaginoplasty can also be done via vaginoscope and resectoscope⁵. This may be the preferable method in the future especially in certain culture where virginity-sparing is one of the main concerns. Yamada et al (2018) in their case study reported their first successful case of vaginoscopic resection of vaginal septum for an OHVIRA patient in Japan. Meanwhile, Ludwin et al (2018) in their case reports of 4 cases are able to introduce a new method of vaginoscopic resection of the vaginal septum under trans-rectal ultrasound guidance (TRUS) with pre- and post-op use of a novel 3-dimensional ultrasound technique with virtual speculoscopy⁸. In all cases, hymenal integrity of the patients were intact and all patients achieved optimum post operative outcome. Although minimally invasive surgery with good outcome have been reported in multiple case reports in other countries^{7,8,9}, the open method is still the surgery of choice in Malaysia.

CONCLUSION

In conclusion, a high index of suspicion among clinicians is paramount in early detection of OHVIRA syndrome to prevent complications. While single stage vaginoplasty remains the surgical treatment of choice in Malaysia, minimally invasive procedure such as vaginoscope and resectoscope may be attempted in the future, especially in young women.

REFERENCES

1. Kudela G, Wiernik A, Drosdzol-Cop A, Machnikowska-Sokolowska M, Gawlik A, Hyla-Klekot L, et al. Multiple variants of obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome – one clinical center case series and the systematic review of 734 cases. *Journal of Pediatric Urology*. 2021;17(5). doi:10.1016/j.jpurol.2021.06.023
2. Smith NA, Laufer MR. Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome: management and follow-up. *Fertility and sterility*. 2007 Apr 1;87(4):918-22.
3. Tan YG, Laksmi NK, Yap TL, Sadhana N, Ong CC. Preventing the O in OHVIRA (obstructed hemivagina ipsilateral renal agenesis): early diagnosis and management of asymptomatic Herlyn–Werner–Wunderlich syndrome. *Journal of Pediatric Surgery*. 2020 Jul 1;55(7):1377-80.
4. Bajaj SK, Misra R, Thakral BB, Gupta R. OHVIRA: Uterus didelphys, blind hemivagina and ipsilateral renal agenesis: Advantage MRI. *Journal of human reproductive sciences*. 2012 Jan;5(1):67.
5. Kriplani A, Dalal V, Kachhawa G, Mahey R, Yadav V, Kriplani I. Minimally invasive endoscopic approach for management of OHVIRA syndrome. *The Journal of Obstetrics and Gynecology of India*. 2019 Aug 1;69:350-5.
6. Yamada Y, Kawaguchi R, Iwai K, Waki K, Kawahara N, Kimura F. Successful vaginoscopic excision of the vaginal septum in a virgin girl of obstructed hemivagina and ipsilateral renal anomaly: Case report and review of literature. *Journal of Obstetrics and Gynaecology Research*. 2023 Jan;49(1):350-5.
7. Ludwin A, Ludwin I, Bhagavath B, Martins WP, Lindheim SR. Virginity-sparing management of blind hemivagina in obstructed hemivagina and ipsilateral renal anomaly syndrome. *Fertility and Sterility*. 2018 Oct 1;110(5):976-8.