# Surgical Approaches for Vaginal Septum Resection in OHVIRA Syndrome Cases

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## *Abstract*

Herlyn-Werner-Wunderlich syndrome (HWWS), also known as Obstructed Hemi-Vaginal Ipsilateral Renal Agenesis (OHVIRA) syndrome, is an uncommon congenital condition characterized by a didelphys uterus, obstructed hemivagina, and the absence of a kidney on the same side. Its rare occurrence makes diagnosis challenging and time-consuming, with surgical intervention representing the primary treatment option. In this study, we present three cases of OHVIRA that underwent surgical treatment with distinct resection approaches. Patients were within the reproductive age group and had been diagnosed with endometriosis for several years. They had been experiencing cyclic abdominal pain since menarche but received their OHVIRA diagnosis just one year before undergoing surgery. Surgical intervention for vaginal septum removal was performed, and the approach was determined based on physical findings observed during the preoperative assessment. In selected cases, a minimally invasive surgical approach using vaginoscopy with ultrasound guidance may be considered, contingent on conducting a thorough pre-surgical assessment. Key considerations in surgical planning encompass factors such as marital status, societal and cultural beliefs, vaginal length, the position of the vaginal septum, and its distance from the hymenal ring. Employing a minimally-invasive vaginoscopy approach offers advantages, including enhanced visualization, reduced risk of injury, and decreased postoperative discomfort. Diagnosing OHVIRA can be intricate, emphasizing the need for a comprehensive evaluation to determine the most suitable surgical strategies. In specific circumstances, the application of a minimally invasive approach involving vaginoscopy guided by ultrasound may be warranted.

**Keywords:** Herlyn-Werner-Wunderlich Syndrome (HWWS), Obstructed Hemi-Vaginal Ipsilateral Renal Agenesis (OHVIRA), Minimally-Invasive, Vaginoscopy.

## Introduction

Obstructed Hemi-Vaginal Ipsilateral Renal Agenesis (OHVIRA) syndrome represents a rare mullerian developmental anomaly frequently complicated by diagnostic delays and delayed treatment. Diagnostic delays often occur due to the normal onset of puberty and menstruation.[1] The precise incidence remains unknown, although various studies have reported an incidence range of 0.1% to 3.8%.[2] OHVIRA is characterized by the triad of uterine didelphys, a blocked hemivagina, and ipsilateral renal abnormalities.[3] Patients typically present with worsening dysmenorrhea and non-specific lower abdominal pain, although urine retention and pelvic masses are also possible manifestations.[4] Accurate diagnosis of OHVIRA is essential to ensure appropriate treatment. Delays in treatment can lead to fertility complications, including the development of endometriosis through retrograde menstruation, the formation of pelvic abscesses, and adhesions.[5] Surgery represents the only permanent solution for OHVIRA. In this report, we present three cases treated with different approaches, one utilizing direct vaginal access and the other employing a minimally-invasive technique with a vaginoscope.

## Case Report

### *Case one*

A woman in her 30s, married for 4 years, with chief complaint of dysmenorrhea since 13 years before admission. She had recurrent ovarian cysts and had ovarian cystectomy before. Her menstrual cycle was irregular and lasting 4-7 days with small amount of blood. Menstrual discomfort was manageable on a VAS of 2 and was eased with pain relieving drug. She has normal tanner stage (P4M4). She has no complaints about her bowel and urine habits. She was referred to a urogynecologist for a full evaluation and was diagnosed with hematocolpos and hematometra from an ultrasound examination. On physical examination, the external genitalia was normal. On speculum examination, seen bulging surface on the right-side vaginal wall located 4 cm from the hymenal ring. This finding corresponded to hematocolpos. The cervix located 6 cm from the hymen and pushed left side due to the hematocolpos. The cervix sondage was 7 cm. On vaginal examination, found enlarged uterus, with palpable mass size 3 cm on the right adnexa. Her blood test was within normal limit.

### *Case two*

A women in her 20s with chief complain of dysmenorrhea for 10 years. At first, the pain was endurable but worsened since 3 years ago. She has a normal menstrual cycle that lasts 7-10 with normal amount of blood. She has normal secondary development with tanner stage M4P4. She was diagnosed with endometriosis cyst and given progesterone therapy for 2 months to relieve the pain. However, the pain persisted and reoccurred every month. She was subsequently referred to a fetomaternal specialist for an ultrasound exam and diagnosed with vaginal septum. She was then referred to our hospital's urogynecology clinic. It took her 3 years from her first visit to the doctor to be diagnosed with OHVIRA syndrome and scheduled for surgery. She has been married for a year and complains with resisting sensation during sexual intercourse. She has no complaint about her urination or bowel habit. On physical evaluation, found normal external genitalia. On speculum examination, seen normal cervix located 6 cm from the hymenal ring and longitudinal vaginal septum located 5 cm from the hymenal ring. On vaginal examination, the uterus was normal in size with palpable mass size 4 cm on the left adnexa. Her Blood test was within normal limit.

### *Case three*

Young unmarried women on her 20s complained of dysmenorrhea for 2 months before admission. She mentioned her menstrual period to be regular, with duration of 4-7 days, and with normal amount. Due to her complained. She has normal secondary development tanner stage M4P4. She has no complaints about her bowel and urine habits, she was diagnosed with hematocolpos and hematometra from an ultrasound examination. On inspection, the hymen was intact. On rectal examination, cyst mass was palpable on the anterior rectum with size 3x2 cm, correspond to hematocolpos, the vagonal sondage 6 cm stucked from hymenal ring. The ultrasound result showed didelphys uterus with right endometriosis cyst and hematocolpos. On abdominal ultrasound, seen normal left kidney with unidentified right kidney. On vaginoscopy evaluation, seen right vaginalk bulging with identified portio on the left side.

### *Pre-surgical evaluation*

Surgical treatment should begin as soon as possible to alleviate symptoms and prevent complications once the diagnosis has been established. Comprehensive preoperative evaluation is needed to treat OHVIRA. We performed transvaginal ultrasound and Magnetic Resonance Imaging (MRI) for both patients. MRI should be recognized as the best modality to evaluate OHVIRA, particularly to identify any renal problems If surgical repair is not available, progesterone suppression is required. Reconstructive operation can be performed closing to menarche in adolescent at the age of 12-13 years old, while surgery in adult patient can be done during menstruation to allow septal border identification for incision. From the ultrasound, we found both patients presents with didelphys uterus, hematocolpos, and ovarian cyst, and abnormal ipsilateral kidney (Figure 1 and Figure 2).

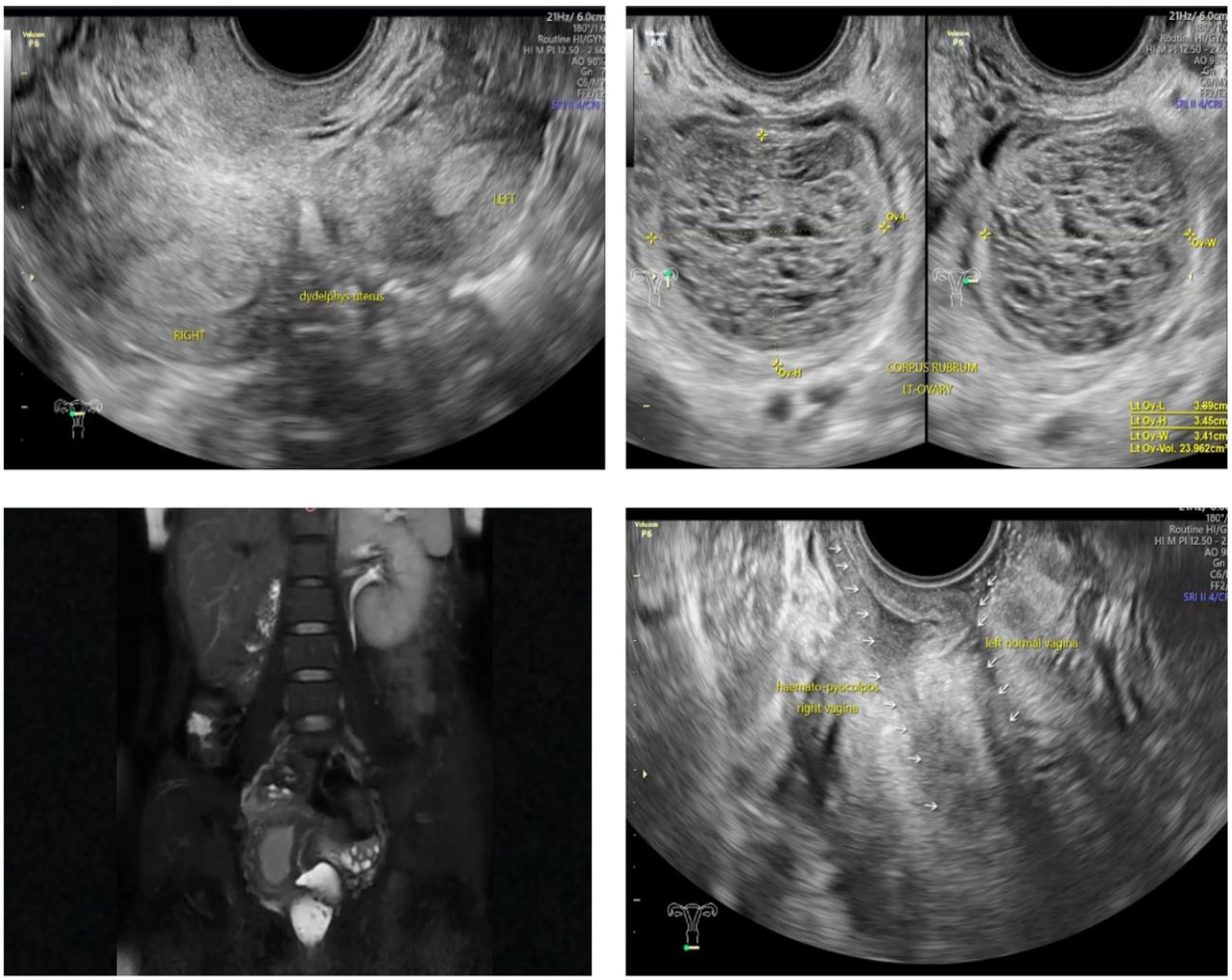


**Figure 1:** Ultrasound finding in Case 1 A) didelphys bicolis uterus, size: right 84x33x29 mm, vol 42.75 $cm^3$, left 81x32x25 mm, vol 33.4 $cm^3$. Homogenic myometrium. Endocervix and cervix within normal limit. B) Left ovary forming corpus rubrum, size 39x35x34 mm, vol. 24 $cm^3$. no sign of solid part, papil growth, vascularization or free fluid. C) No sign of widened pelvio-calyces system of left kidney. Unidentified right kidney (MRI). D) Longitudinal vaginal septum divided left and right side, seen hemato-pyocolpos at right vagina. Positive sliding sign. US guided tenderness not found.

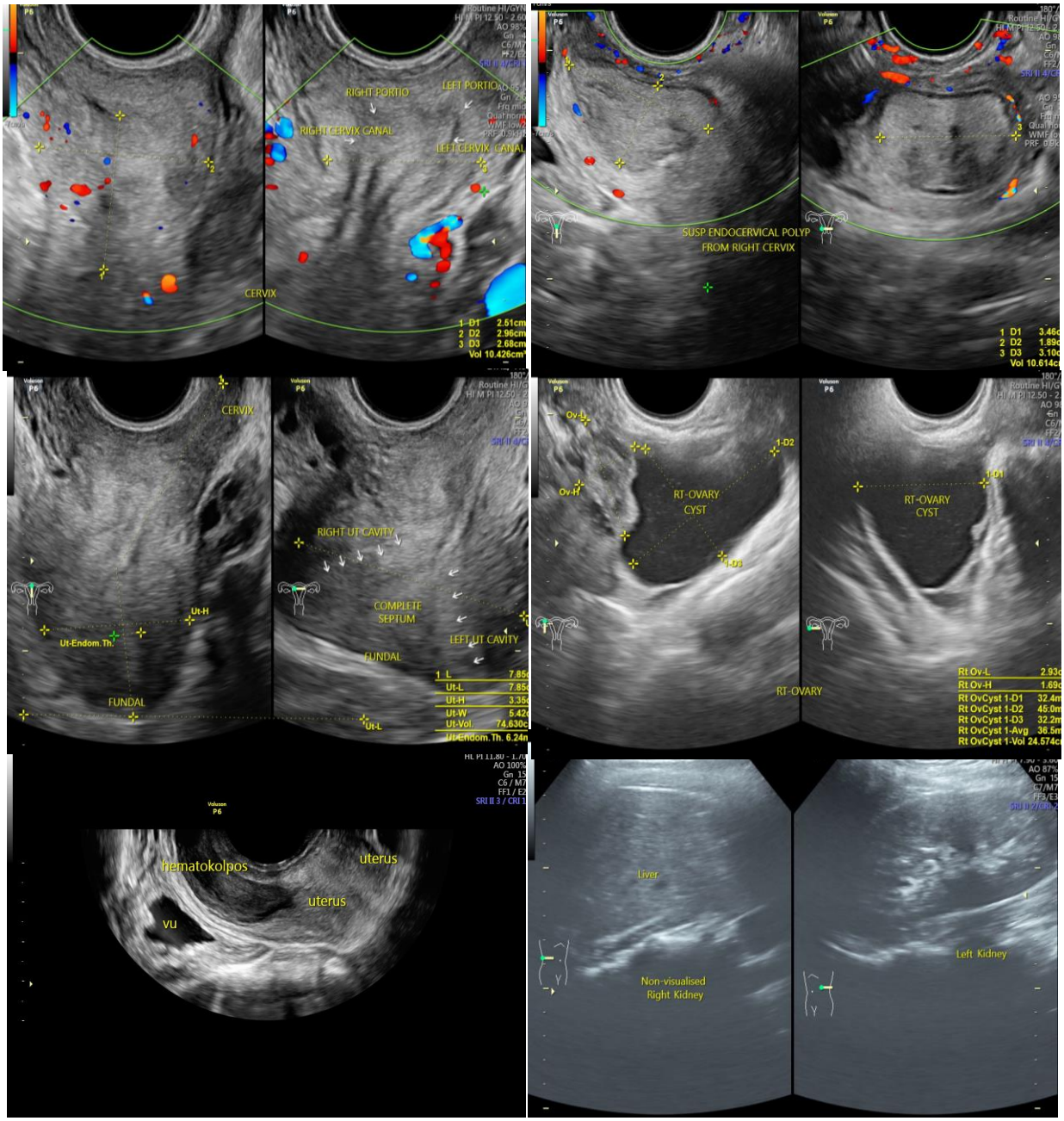


**Figure 2:** Ultrasound finding in case 2 A) Didelphys bicolis anteflexed uterus, size: Right 57x25x34mm, Left 54x26x35mm. Homogenic myometrium. Right Endometrium thickness 7.5 mm, left 6.6 mm, no sign of triple line, homogenous, no sign of intra-cavity mass, regular stratum basale. Endocervix and cervix within normal limit B) Right ovary normal size 29x24x21 mm, vol. 7.9 cm3, no sign of solid part, papil growth, vascularization or free fluid. Left ovary enlarged forming unilocular cystic mass, with picture of ground-glass, size 38x35x38 mm, vol. 26.4 xm3. no sign of solid part, papil growth, vascularization or free fluid. Accordance to small left ovary endometriosis cyst. Positive sliding sign. US guided tenderness not found. C) No sign of widened pelvio-calyces system of right kidney. Unidentified left kidney. D) Hematocolpos filled left vagina closed by septum with 6 mm thickness.

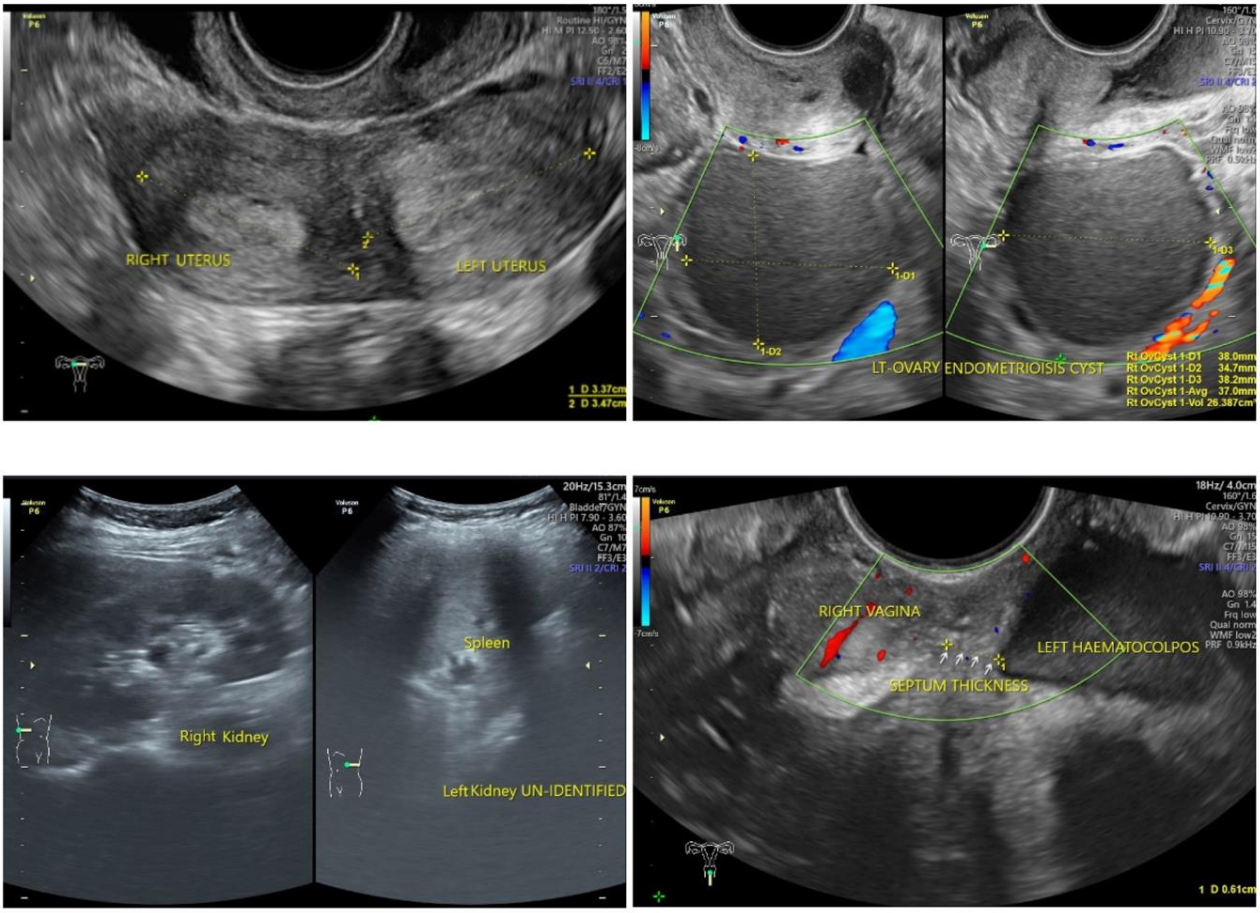


**Figure 3:** Ultrasound finding in case 3 A) There are two endocervix and portio, with polyp on the right endocervix (B). C) Two uterine cavities were detected, separated with complete endometrial septum D) Right ovarian cyst size 32x20x18 mm, vol. 6.21 cm3, with hypoechoic appearance suspected endometriosis cyst E) Hematocolpos appearance with unilocular and smooth echogenic material, size 33 x 35 x38 mm, vol. 23.9 cm3 F) Unidentified right kidney with normal left kidney.

## *Surgical approaches*

## *Identification of septum location, border, and direction*

This step is done with direct visualization and to identify the uretra, cervix, bulging area, border of the septum, and vaginal mucose. We perform clamping of the longitudinal septum on the lateral side, at the border of the septum and the lateral vaginal mucosa. Using uterine sondage, the distance between the cervix and septum to the hymen were evaluated. This step is crucial to determine the difficulty of the surgery and whether assisted approach with vaginoscopy is preferred.

Abdominal ultrasound can be used as guidance to visualized and reconstruct the position of the uterus, septum, hematometra, and bladder, as we performed in our second patient. In the second case, we found that the septum was positioned lower than we expected. We can see from the ultrasound that the the bladder was position very closely to septum and cervix. This might be due to the weight of the hematocolpos that pull other structures downward. To avoid injury, we proceed with ultrasound guided.

## *Hydro-dissection and aspiration to identify hematocolpos*

If the needle is correctly positioned, menstrual blood can be observed during aspiration. In cases where the accumulated blood volume is insufficient to clearly identify the septal border, the addition of a 20 cc injection of normal saline into the septal space can facilitate the process.

## *Identification of medial and lateral border of the vaginal septum*

Precise identification of the septal border is of utmost importance to minimize the risk of vaginal and cervical injury during the resection. To assist in this process, we place a simple suture, using 2.0 PGA suture, at both the medial and lateral aspects of the septum (as seen in case 2). This suture placement serves as a valuable guide during the resection, particularly when the septum is situated at a considerable distance from the hymen.

## *Septum resection*

a) Excision with direct visualization

In the first case, we initiate the procedure by making an incision at the predetermined location, guided by the needle marking the hydrodissection spot. The incision is a 0.5 cm longitudinal one, and we proceed to widen it using a dilator until reaching the base of the septum. Subsequently, we conduct a circular excision on the anterior, posterior, and lateral borders of the septum. The final step involves performing interrupted sutures using PGA 3.0 suture along the edges of the excision site, securing it to the vaginal mucosa.

b) Septum resection with resectoscope

In the second case, the resection was executed from both the lateral and medial sections of the septum, using the border sutures as guides. The septum was determined to have a thickness of 1.5 cm, and the resection was performed until we could successfully identify both cervixes.

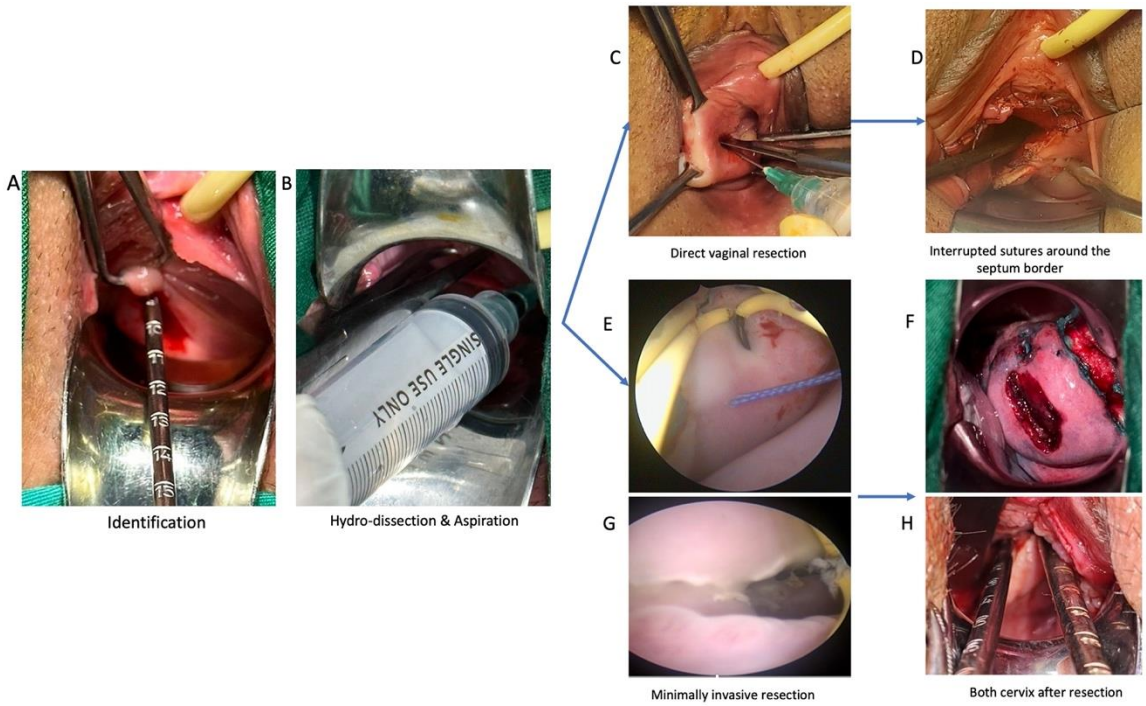


**Figure 4:** Surgical steps for septum resection: A) Identification of septum location, border, and direction, perform clamping of the septum. Intraoperative US guided to determine the location of other organs B) hydro-dssection and aspiration. Case 1. C) Using guiding sutures on the medial and lateral border of the septum. Septum resection with direct approach and using needle to determine the location. The incision is widen using dilator D) Final view from the direct approach, with interrupted suture circuling the septum. E) septum resection with resectoscope, the incision is done between the lateral and medial border with suture as guidance (Blue line). F) Final view for minimally invasive resection approach (case 2) where no suture is needed. In experienced surgeon, the procedure can also be done without suture guided as in case 3 with the same precise result (G & H)

## Discussion

Obstructed Hemi-Vagina And Ipsilateral Renal Agenesis Syndrome (OHVIRA) is a rare Mullerian duct anomaly characterized by an obstructive vaginal septum, associated ipsilateral renal anomalies, and varying forms of uterine deformities. OHVIRA cases are typically classified into three types based on the extent of septal obstruction: Type I: A complete hemi-vaginal septum without any opening, Type II: A complete hemi-vaginal septum with a small pinpoint-sized hole allowing limited menstrual blood drainage, Type III: A complete hemi-vaginal septum with a cervical fistula connecting both cervix.

The specific type of OHVIRA has implications for the patient's symptoms and potential complications.[6,7] Diagnosis of OHVIRA syndrome necessitates a comprehensive evaluation involving a review of medical records, physical examinations, and multimodal imaging techniques, such as ultrasonography (USG), magnetic resonance imaging (MRI), or direct visualization.[8,9] In many cases, OHVIRA is diagnosed late, as was the case with both of our patients, leading to the exacerbation of complications. Anticipated long-term complications include infertility and the risk of infection.For instance, the first patient experienced primary infertility for four years, underwent surgery for cyst removal, and may require additional surgical interventions in the future to address the cyst and achieve fertility. OHVIRA not only diminishes fertility but also increases the patient's overall morbidity.[10,11]

OHVIRA can be managed through a single-step septum resection.[12] Traditionally, septum resection is performed under direct visualization, with the septum being palpated for resection. However, the conventional approach carries the risk of hymenal damage, vaginal tears, increased postoperative pain, and adhesion formation. Hymenal reconstruction may be considered for adolescents and patients who wish to preserve the hymenal ring structure.[13,14] Although there are limited reported cases, a minimally invasive approach employing vaginoscopy and a "no-touch"

technique may be appropriate for adolescent and virgin patients. This method offers enhanced safety, improved visualization, reduced post-operative pain, and a decreased risk of injury.[14]

In our study, both married patients were managed differently. The choice of surgical technique should take into account various factors, including marital status, the position and distance of the septum in relation to the hymen, cyst thickness, types of organ malformations, and any concurrent conditions such as cysts or infections. Ultrasound guidance may be necessary to prevent organ injury, and in specific cases, an abdominal approach may be required to manage OHVIRA. Postoperative follow-up is crucial to assess the risk of adhesions, stenosis, and potential complications. Patients may experience vaginal discharge after the operation, necessitating comprehensive counseling before the procedure.

## Conclusion

In conclusion, OHVIRA presents a complex and challenging clinical scenario. Delayed diagnosis, as commonly observed in our cases, amplifies the risk of long-term complications, particularly infertility and infection. Effective management of OHVIRA typically involves single-step septum resection, and our discussion highlighted the advantages of minimally invasive approaches, particularly vaginoscopy with a "no-touch" technique for certain patient populations. The choice of surgical technique should be carefully tailored to individual patient characteristics, including marital status, septum positioning, concurrent conditions, and the need for ultrasound guidance.

## Conflict of interest

none

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