IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 25, Issue 4 Ser. 5 (April. 2026), PP 08-11
www.iosrjournals.org

Unmasking OHVIRA Syndrome – A Rare Cause Of Acute Urinary Retention In Adolescents

Dr. Arushi Pandya, Dr. Ankita Goyal
Resident, Department Of Obstetrics & Gynaecology, MGM Medical College, Navi Mumbai
Senior Resident, Department Of Obstetrics & Gynaecology, MGM Medical College, Navi Mumbai

Abstract
Background: *OHVIRA syndrome (Obstructed Hemivagina and Ipsilateral Renal Agenesis), also known as Herlyn–Werner–Wunderlich (HWW) syndrome, is a rare congenital Müllerian duct anomaly. It is characterised by the classic triad of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis.*
Case Presentation: *We present a 15-year-old adolescent girl who presented with acute urinary retention and lower abdominal pain in the post-menarcheal period. Pelvic ultrasonography and MRI revealed uterus didelphys with right hematocolpos, right hematometra, and absence of the right kidney — diagnostic of OHVIRA syndrome.*
Management & Outcome: *The patient was successfully treated with vaginal septal resection. Postoperatively, complete resolution of urinary symptoms and pelvic discomfort was achieved, with satisfactory healing on follow-up.*
Conclusion: *OHVIRA syndrome, though rare, is an important and potentially under-recognised cause of acute urinary retention in adolescent females. Early clinical suspicion, timely MRI evaluation, and prompt surgical management are pivotal for excellent outcomes and preservation of reproductive potential.*
Keywords: *OHVIRA syndrome; Herlyn–Werner–Wunderlich syndrome; uterus didelphys; hematocolpos; urinary retention; Müllerian duct anomaly; adolescent gynaecology; vaginal septal resection*

Date of Submission: 15-04-2026 Date of Acceptance: 25-04-2026

I. Introduction

OHVIRA syndrome — an acronym for Obstructed Hemivagina and Ipsilateral Renal Agenesis — is a rare congenital anomaly classified under Müllerian duct malformations. The syndrome is synonymously referred to as Herlyn–Werner–Wunderlich (HWW) syndrome in the European literature. It arises from aberrant embryological development of the paramesonephric (Müllerian) ducts, in conjunction with concurrent maldevelopment of the mesonephric (Wolffian) duct, which accounts for the characteristic association with ipsilateral renal anomalies.

The classical anatomical triad encompasses: (1) uterus didelphys — a complete duplication of the uterine corpus and cervix; (2) an obstructed hemivagina with accumulation of menstrual products; and (3) ipsilateral renal agenesis or dysgenesis. Due to the obstruction, retrograde accumulation of menstrual blood leads to hematocolpos and hematometra in the affected side, while the contralateral uterine cavity continues to drain normally.

The syndrome typically manifests in the post-menarcheal period, most commonly presenting with progressive, cyclical dysmenorrhea and pelvic pain resulting from menstrual retention. However, an uncommon and diagnostically challenging presentation is acute urinary retention, which occurs when a markedly distended hemivagina or hematocolpos exerts mass effect on the adjacent bladder neck or urethra.

Early and accurate diagnosis is critical. Delayed recognition predisposes to significant long-term morbidity including endometriosis, chronic pelvic pain, pelvic adhesions, and impaired fertility. With prompt surgical intervention, outcomes are excellent and reproductive potential is fully preserved. This report aims to highlight the urological presentation of OHVIRA syndrome and underscore the importance of maintaining a high index of clinical suspicion in adolescent females.

II. Aim

The objective of this case report is to:
(i) Highlight OHVIRA syndrome as an uncommon but clinically important cause of acute urinary retention in post-menarcheal adolescent females.
(ii) Emphasise the importance of maintaining a high index of suspicion when urinary complaints co-exist with cyclical pelvic pain in this demographic.
(iii) Demonstrate the diagnostic utility of multimodal imaging, particularly magnetic resonance imaging (MRI), in delineating the complex anatomy.

(iv) Illustrate that timely vaginal septal resection results in complete symptomatic resolution and preservation of reproductive function.

III. Case Presentation

History and Clinical Presentation

A 15-year-old adolescent girl was brought to the emergency department with a chief complaint of acute inability to void urine, accompanied by lower abdominal pain of approximately 24 hours duration. Her symptoms had developed gradually and were associated with significant suprapubic discomfort and a sense of pelvic fullness.

The patient had attained menarche recently and reported a history of cyclical lower abdominal pain coinciding with her menstrual cycles. There was no history of prior gynaecological evaluation. Her medical history was otherwise unremarkable, and she had not undergone any prior surgical procedures. Family history was non-contributory.

Examination Findings

General examination revealed a well-nourished adolescent in mild-to-moderate distress due to pain and urinary discomfort. Vital signs were within normal limits. Abdominal examination demonstrated suprapubic fullness consistent with bladder distension. There was no peritoneal guarding or rigidity. Given the acute urinary retention, urethral catheterisation was promptly performed, which drained a significant volume of urine and provided immediate relief from urinary discomfort.

Following bladder decompression, the patient continued to report pelvic pain, and clinical suspicion for an underlying pelvic structural abnormality was raised. A further systematic clinical evaluation was undertaken, with the patient referred for radiological investigation.

Investigations

Pelvic ultrasonography was the initial imaging modality employed. It revealed findings suggestive of a septate or duplicated uterine configuration and raised the possibility of ipsilateral renal absence. Notably, the right kidney was not visualised in its normal renal fossa, raising concern for right renal agenesis.

Given the complex findings and high clinical suspicion, Magnetic Resonance Imaging (MRI) of the pelvis and abdomen was subsequently performed. MRI is the gold standard for delineating Müllerian anomalies due to its superior soft-tissue contrast and multiplanar capability. MRI findings included:

• Uterus didelphys — complete duplication of the uterus with two separate uterine corpora and cervices
• Right-sided hematocolpos — markedly distended right hemivagina filled with retained menstrual blood, causing significant mass effect on the bladder base and urethral neck, accounting for the presenting urinary retention
• Right hematometra — blood accumulation within the right uterine cavity
• Non-communicating left uterine cavity — the left uterus drained normally with no obstruction
• Right renal agenesis — confirmed absence of the right kidney, with compensatory hypertrophy of the left kidney

These findings were diagnostic of OHVIRA (Herlyn–Werner–Wunderlich) syndrome, confirming all three components of the classical triad.

IV. Management

Multidisciplinary Approach

A multidisciplinary team (MDT) was convened, involving consultants from the Departments of Obstetrics and Gynaecology, Radiology, and Urology. Radiological findings were reviewed in detail to delineate the exact anatomical configuration and plan a definitive surgical approach. The urology team provided assessment regarding the single functioning left kidney and peri-operative renal monitoring.

Surgical Intervention

The patient underwent surgical management in the form of vaginal septal resection with simultaneous drainage of the retained menstrual blood (hematocolpos). The procedure was performed under general anaesthesia with the patient in the lithotomy position. Following thorough preoperative counselling and consent, the obstructing vaginal septum was excised, and the collected haematocolpos was fully drained.

The procedure was completed without intraoperative complications. Adequate decompression of the obstructed right hemivagina was achieved. Intraoperative findings corroborated the preoperative MRI assessment. Haemostasis was secured, and the surgical site was inspected for integrity.

Postoperative Course

The patient had an uneventful postoperative recovery. Urinary catheterisation was maintained for 48 hours postoperatively and subsequently removed once normal voiding was confirmed. The patient experienced complete resolution of urinary symptoms and marked relief of pelvic discomfort from the first postoperative day.

Follow-up evaluation at six weeks demonstrated satisfactory healing at the surgical site with no evidence of re-accumulation, recurrence of haematocolpos, or return of urinary symptoms. The patient reported normal, pain-free menstruation following surgery. Renal function remained stable, with the single left kidney confirmed to be functioning adequately on follow-up ultrasonography.

V. Discussion

OHVIRA syndrome, first described by Herlyn and Werner in 1971 and subsequently by Wunderlich in 1976, remains an uncommon condition with an estimated prevalence of approximately 0.1–3.8% among women with Müllerian anomalies. The embryological basis lies in aberrant lateralised Müllerian duct fusion during organogenesis between the 6th and 20th weeks of gestation. As the Müllerian and Wolffian ducts share a common developmental pathway in early embryogenesis, unilateral defects in Müllerian fusion are often accompanied by ipsilateral Wolffian duct regression failure — accounting for the consistent ipsilateral association of the renal anomaly.

The classical presentation, as described in the literature, is cyclical pelvic pain and dysmenorrhea commencing shortly after menarche. The obstructed hemivagina retains menstrual blood with each cycle, leading to progressively enlarging hematocolpos. The contralateral, unobstructed uterine cavity allows menstruation to proceed normally, often delaying clinical recognition.

Acute urinary retention as the presenting complaint — as observed in our case — represents a rare and atypical but well-documented complication of OHVIRA syndrome. The mechanism is mechanical: progressive distension of the right hematocolpos exerts direct pressure on the bladder base and urethra, causing functional urinary outflow obstruction. This presentation may be mistakenly attributed to primary urological pathology, deferring the gynaecological diagnosis.

Ultrasonography is typically the first-line imaging modality owing to its wide availability and non-invasive nature. However, MRI is considered the gold standard for characterising Müllerian anomalies due to its superior soft-tissue contrast, multiplanar capacity, and ability to delineate the precise uterovaginal and renal anatomy. In our patient, MRI confirmed uterus didelphys, right hematocolpos with mass effect on the bladder, and right renal agenesis — all three elements of the diagnostic triad.

Vaginal septal resection remains the definitive surgical treatment and has consistently demonstrated excellent outcomes across reported series. Early intervention prevents progression to endometriosis, retrograde implantation, pelvic adhesion formation, and tubal compromise — all of which have adverse implications for future fertility. In our patient, a single-stage vaginal septal resection resulted in complete decompression, prompt resolution of urinary symptoms, and restoration of normal menstrual function.

The broader significance of this case lies in the diagnostic challenge it presents. Urinary retention in a post-menarcheal girl, particularly when accompanied by cyclical pelvic pain, should prompt the clinician to consider OHVIRA syndrome in the differential diagnosis. The absence of the ipsilateral kidney — frequently identified incidentally — should heighten suspicion. Increased awareness among emergency physicians, paediatricians, and gynaecologists is essential for timely diagnosis and intervention.

VI. Conclusion

This case underscores the importance of OHVIRA (Herlyn–Werner–Wunderlich) syndrome as a rare but clinically significant cause of acute urinary retention in adolescent females. Although the syndrome is uncommon, its presentation with urinary symptoms can mimic primary urological disease, leading to diagnostic delays with potential long-term reproductive consequences.

Key clinical take-aways from this case include: (i) maintaining a high index of suspicion for Müllerian anomalies in post-menarcheal adolescents presenting with urinary retention, particularly when accompanied by a history of cyclical pelvic pain; (ii) the indispensable role of MRI in confirming diagnosis and surgical planning; and (iii) the efficacy and safety of vaginal septal resection as a definitive, fertility-preserving surgical intervention.

Heightened clinician awareness across emergency medicine, paediatrics, radiology, and gynaecology is critical for early recognition, timely referral, and optimisation of outcomes in young patients with this complex congenital anomaly.

Conflict of Interest Statement

The authors declare no conflict of interest.

Patient Consent

Informed consent was obtained from the patient's parent/guardian for publication of this case report and associated clinical details. Patient identifiers have been withheld to preserve confidentiality.

References

[1]. Smith NA, Laufer MR. Obstructed Hemivagina And Ipsilateral Renal Anomaly (OHVIRA Syndrome): Management And Follow-Up. Fertility And Sterility. 2007;88(5):1261–1265.
[2]. Orazi C, Inserra A, Lucchetti MC, Schingo PM, Ferro F. Herlyn-Werner-Wunderlich Syndrome: Uterus Didelphys, Blind Hemivagina And Ipsilateral Renal Agenesis. MRI Findings And Clinical Importance. Radiology. 2007;245(2):559–563.
[3]. Rock JA, Jones HW. Te Linde's Operative Gynecology. 11th Ed. Philadelphia: Wolters Kluwer; 2015.
[4]. Acién P. Incidence Of Müllerian Defects In Fertile And Infertile Women. Human Reproduction. 1997;12(7):1372–1376.
[5]. 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(4):340–347.
[6]. Wunderlich M. Unusual Form Of One-Sided Aplasia Of The Kidney Associated With Ipsilateral Blind Uterine Horn And Vaginal Aplasia. Zentralbl Gynakol. 1976;98(9):559–562.