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Complex Müllerian Duct Anomaly Presenting as a Rare OHVIRA Variant: Clinical and Surgical Perspectives

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Case Report

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Abstract

Background

Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA) syndrome, also referred to as Herlyn–Werner–Wunderlich syndrome, is an uncommon congenital anomaly of the genitourinary tract characterized by uterine malformations, an obstructed hemivagina, and ipsilateral renal agenesis. Atypical variants add further complexity, often posing significant diagnostic and therapeutic challenges.

Case Presentation:

We report the case of a 14-year-old girl presenting with primary amenorrhea and cyclical abdominal pain. Imaging revealed a bicornuate unicollis uterus with hematometrocolpos, absent left kidney, and a grossly dilated ectopic right ureter. Intraoperatively, the cervix was rudimentary and hypoplastic, precluding fertility preservation. A complete hysterectomy with right ureteric reimplantation was performed using the Politano–Leadbetter technique. Postoperative recovery was uneventful, and the patient experienced complete resolution of symptoms at follow-up.

Discussion

This case demonstrates an atypical variant of Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA) syndrome, characterized by bicornuate uterus, rudimentary cervix, segmental vaginal atresia, unilateral renal agenesis, and ureteric malpositioning. Such complex anomalies underscore the limitations of current classification systems and emphasize the importance of detailed anatomical description. Magnetic resonance imaging remains the diagnostic gold standard, while multidisciplinary planning is crucial for tailoring management to individual patients. When reconstructive options are not feasible, definitive surgical intervention is often required to achieve symptom relief and prevent long-term morbidity.

Conclusion

Atypical variants of OHVIRA syndrome underscore the importance of integrating embryological understanding, systematic renal screening, and collaborative surgical planning. Early recognition and tailored intervention are critical to achieving symptom relief and preventing long-term morbidity.

Introduction

Müllerian duct anomalies (MDAs) comprise a diverse spectrum of congenital malformations that arise from abnormalities in the development, fusion, or resorption of the paramesonephric ducts [1]. Given their shared embryological origin from the intermediate mesoderm, the genital and urinary systems

often exhibit concurrent anomalies. As a result, Müllerian duct malformations are frequently associated with renal abnormalities, including unilateral renal agenesis, ectopic kidneys, and ureteric defects [1].

Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA) syndrome, also known as Herlyn–Werner–Wunderlich syndrome (HWWS), is a complex congenital anomaly of the genitourinary tract characterized by a didelphic uterus, obstructed hemivagina, and ipsilateral renal agenesis, first described in 1922 [2]. Variants of OHVIRA syndrome such as bicornuate or didelphys uteri with a blind hemivagina, predispose patients to endometriosis and chronic pelvic pain due to retrograde menstruation [1].

Imaging is central to the diagnosis of Müllerian anomalies, with MRI serving as the gold standard for accurately delineating complex pelvic anatomy and associated renal malformations [3]. However, existing classification systems frequently fail to encompass atypical variants, highlighting the need for detailed descriptive reporting and tailored management approaches [4]. Although procedures such as uterovaginal canalization, vaginoplasty, and hemi-hysterectomy have been attempted in selected cases to preserve fertility, definitive management typically necessitates hysterectomy when the cervix is rudimentary or absent [5].

In the present case, a 14-year-old girl presented with primary amenorrhea and cyclical abdominal pain. Imaging revealed a bicornuate unicollis uterus with hematometrocolpos, absent left kidney, and a grossly dilated tortuous right ureter with ectopic implantation. Intraoperatively, the cervix was found to be rudimentary and hypoplastic, making fertility preservation unfeasible. A complete hysterectomy with right ureteric reimplantation was performed, and the patient experienced complete resolution of symptoms at follow-up.

This case is unique as it represents an atypical variant of OHVIRA syndrome, characterized by a bicornuate uterus with a rudimentary cervix, segmental vaginal atresia, unilateral renal agenesis, and ureteric malpositioning necessitating reimplantation. These complex presentations are infrequently documented in the literature and underscore the necessity of multidisciplinary evaluation with individualized surgical planning to optimize outcomes [6].

Case presentation

A 14-year-old girl presented to the outpatient clinic with a six-month history of cyclic lower abdominal pain. The pain was gradual in onset, progressive, and of moderate intensity. It was cyclical and occasionally associated with low-grade intermittent fever, relieved by over-the-counter analgesics. She had never attained menarche. On examination, her body mass index was 16, with a height of 4 feet 10 inches and a weight of 35 kg. Secondary sexual characteristics were well developed, corresponding to Tanner stage 5. The external genitalia appeared normal. Per vaginal examination revealed a vaginal canal measuring approximately 2 cm in length, with stenosis at the upper end of the vault, suggestive of a congenital anomaly. The abdomen was soft and non-tender, and systemic examination revealed no other significant findings.

Ultrasound of the abdomen and pelvis demonstrated absence of the left kidney in the left renal fossa. The right kidney showed moderate hydronephrosis with associated hydroureter, findings suggestive of ectopic ureteric implantation into the bladder. A bicornuate, anteverted uterus was identified, measuring 11 × 5.8 × 2.7 cm, with dilatation of the cervical canal. Approximately 50 ml of fluid was noted within the right endometrial cavity. Intravenous pyelogram demonstrated a solitary right kidney with moderate hydronephrosis. The right ureter was patent but appeared moderately dilated, elongated, and tortuous, with ectopic implantation into the posterior-inferior aspect of the distal bladder neck (as shown in Fig. 1). The patient underwent MRI for further evaluation; however, the imaging films were not available at the time of reporting. Findings were interpreted from the radiology report, which confirmed a bicornuate unicollis uterus with T1 hyperintense fluid in both endometrial cavities—the right horn measuring 6.7 × 3.4 cm and the left horn 4.2 × 2.0 cm. The right horn was continuous with a cystically ballooned cervix containing a well-defined collection (5.4 × 6.6 cm), extending to a blind-ending upper vagina, consistent with hematometrocolpos. A simple left adnexal cyst (2 × 3 cm) was noted, adjacent to a left ovary with small follicles. A distally dilated tubular structure in the left adnexa showed a blood-fluid level, suggestive of hematosalpinx. The right ovary was not visualized on MRI but appeared normal on ultrasound, located ectopically beneath the lower abdominal wall, lateral to the right uterine horn. The left kidney was absent, and there was moderate hydronephrosis of the right kidney with a dilated and tortuous right ureter opening posteroinferiorly into the distal bladder neck. Definitive confirmation of all the above-mentioned findings was obtained intraoperatively. Collectively, these findings were consistent with an atypical variant of OHVIRA syndrome (Obstructed Hemivagina and Ipsilateral Renal Anomaly).

Figure 1. IVP demonstrates a congenital solitary right kidney with secondary changes, including a dilated tortuous ureter and right-sided hydronephrosis. Findings suggest obstructive uropathy in the solitary functioning kidney.

Figure 1: IVP demonstrates congenital solitary right kidney with secondary changes: dilated tortuous ureter and right-sided hydronephrosis. Findings suggest obstructive uropathy in the solitary functioning kidney.

An initial hymenectomy was performed at a rural clinic; however, it failed to evacuate the hematocolpos, and the patient was subsequently referred to our hospital for definitive treatment.

At our gynecology clinic, she underwent detailed evaluation and preoperative investigations. A multidisciplinary team comprising a urologist, gynecologist, general surgeon, and anesthesiologist discussed various treatment options, including vaginoplasty, hemihysterectomy, and complete hysterectomy. Preoperatively, the team decided to proceed with either hemihysterectomy or complete hysterectomy with right ureteral reimplantation, depending on intraoperative findings and shared decision-making with the patient and her parents. The priority was placed on definitive symptom relief rather than fertility preservation. Intraoperatively, the cervix was found to be rudimentary and hypoplastic, making fertility preservation unfeasible, and a complete hysterectomy was performed along with right ureteric reimplantation.

A Pfannenstiel incision was made to access the abdomen and pelvis. Intraoperative findings included a bicornuate uterus of approximately 10-week size with no distinct cervix and a dilated blind-ending vaginal canal containing hematocolpos (as shown in Fig. 2A). The right ovary was rudimentary, while the left ovary appeared normal but was associated with an adnexal endometriotic cyst that drained 15 ml of chocolate-colored fluid. The right ureter was grossly dilated and tortuous, with multiple kinks at different levels (as shown in Fig. 2B). Approximately 150 ml of hematocolpos was evacuated. Following hemostasis, hysterectomy was performed. The right ureter was carefully mobilized by dissecting adhesions to facilitate reimplantation (as shown in the Fig. 2C). It was clamped and ligated 1 cm proximal to the ectopic vesicoureteric junction, then reimplanted at the anatomical site using the Politano-Leadbetter technique. A 6 Fr JJ stent was placed, and the anastomosis was secured with 2 – 0 vicryl sutures. A pelvic drain was inserted, and after ensuring hemostasis, the abdominal wall was closed in layers. The patient was catheterized with a silicone Foley catheter. Estimated intraoperative blood loss was approximately 800 ml.

A: Imaging reveals left ovarian endometriotic cyst with contralateral rudimentary ovary and associated right ureteric dilatation.

B: Imaging reveals Bicornuate uterus: dilated cervix and canal with hematocolpos drainage.

C: Image showing a dilated right ureter with a feeding tube passed for identification, dissected from the bladder and prepared for ureteric reimplantation.

Postoperatively, the patient was commenced on intravenous piperacillin–tazobactam and metronidazole. She remained hemodynamically stable with adequate urine output and normal laboratory parameters. The pelvic drain collected 150 ml over the first 36 hours and was removed on the 3rd postoperative day. An X-ray KUB performed on day 2 confirmed correct positioning of the JJ stent (as shown in Fig. 3). Her recovery was uneventful, and she was discharged on the 4th postoperative day with outpatient follow-up. The Foley catheter and JJ stent were removed at 6 weeks, and at the 2-month follow-up, she reported complete resolution of symptoms.

Figure 3. X-ray KUB demonstrating the right-sided JJ ureteric stent in situ.

Figure 3: X-ray KUB demonstrating right-sided JJ ureteric stent in situ.

Discussion

Müllerian anomalies encompass a diverse group of congenital malformations resulting from defects in ductal development, fusion, or resorption, and are often linked to endometriosis and renal abnormalities owing to the common embryological origin of the genital and urinary systems from the intermediate mesoderm [1]. The relationship between Müllerian anomalies and endometriosis is variable, with obstructive forms including cervicovaginal agenesis, transverse vaginal septum, and non-communicating rudimentary horns demonstrating the strongest association, thereby supporting

retrograde menstruation as a principal pathogenic mechanism [1]. However, endometriosis has also been documented in non-obstructive Müllerian anomalies, implicating alternative mechanisms such as coelomic metaplasia or persistence of embryonic remnants [1]. Importantly, the ESHRE/ESGE consensus emphasizes that anatomy should serve as the basic characteristic for grouping anomalies, with uterine morphology used as the key organ in classification, thereby ensuring clinical relevance and management correlation [7].

Phenotypic variability significantly influences clinical outcomes, with functional rudimentary structures containing active endometrium showing a stronger association with endometriosis and pain than non-functional remnants [1]. The coexistence of bicornuate uterus with cervicovaginal agenesis is exceptionally rare, necessitating individualized management that balances symptom relief with fertility preservation; although uterovaginal anastomosis and metroplasty have been attempted, outcomes remain variable, and hysterectomy is often required when reconstruction is not feasible. [8].

Diagnostic challenges persist, as MRI remains the preferred non-invasive modality for identifying Müllerian anomalies, yet existing classification systems often fail to encompass atypical variants; in such circumstances, detailed descriptive reporting together with interdisciplinary review is essential to guide appropriate management [3]. Moreover, Müllerian anomalies are often accompanied by renal and skeletal malformations, reflecting the shared embryological origins of these systems [3].

From an embryological standpoint, distal mesonephric anomalies—such as uterine duplicity with ipsilateral blind hemivagina—are frequently misdiagnosed; these conditions typically manifest with pelvic pain, dysmenorrhea, hematocolpos, and occasionally urinary symptoms from ectopic ureters. Systematic cataloguing of anomalies according to embryological processes facilitates anticipation of associated renal malformations and supports individualized management strategies [4]. Congenital absence of a kidney is the most frequent renal malformation in women with genital anomalies, particularly when obstructive cervical or vaginal anomalies coexist, underscoring the need for systematic renal screening [9]

Unilateral renal agenesis shows a strong association with bicornuate and didelphys uteri, and these combinations predispose affected women to gynecological pathologies, particularly endometriosis [6]. Laterality analyses indicate that right-sided agenesis is more frequently associated with distal mesonephric anomalies, such as blind hemivagina, whereas left-sided agenesis more commonly corresponds to agenesis or hypoplasia of the urogenital ridge [6]. Cervical agenesis, although extremely rare, presents significant diagnostic and management challenges; while reconstructive procedures such as uterovaginal anastomosis may be attempted, outcomes remain inconsistent, and hysterectomy is often necessary when reconstruction proves unfeasible [5].

Bicornuate uterus is a rare anomaly, with an estimated prevalence of 0.4% in the general population; when accompanied by cervical atresia, patients typically present with pelvic pain resulting from hematometra and retrograde menstruation. Management remains controversial, spanning from definitive hysterectomy to conservative canalization approaches [10]. Other reports highlight that complex

anomalies often elude traditional classification systems, necessitating descriptive reporting and individualized assessment to ensure appropriate clinical management [3], [11].

The clinical implications are substantial: women with Müllerian duct anomalies not only exhibit renal agenesis but also a broader spectrum of urological abnormalities, including multicystic dysplastic kidneys, duplicated systems, and pelvic kidneys. Such congenital anomalies, particularly when they result in a solitary kidney, confer increased risk of renal injury, hypertension, and progression to end-stage renal disease, underscoring the importance of early identification, counseling, and long-term surveillance [12]. Even with the use of advanced modern imaging techniques, classifying Müllerian duct anomalies often remains challenging. When a definitive category cannot be established, it is more appropriate to provide a thorough anatomical description rather than attempting to fit the anomaly into an existing classification system. In these complex situations, detailed morphological assessment together with interdisciplinary review of imaging findings offers the best strategy to optimize patient management [3].

Our patient's case exemplifies the complexity of Müllerian duct anomalies and their frequent association with urological malformations. She presented with an atypical OHVIRA variant, combining a bicornuate uterus, rudimentary cervix, segmental vaginal atresia, unilateral renal agenesis, and ureteric malpositioning requiring reimplantation. In line with prior reports emphasizing individualized assessment and multidisciplinary planning [3], [4], a team of specialists prioritized definitive symptom relief over fertility preservation. Intraoperative findings confirmed a hypoplastic cervix and complex pelvic anatomy, leading to complete hysterectomy with right ureteric reimplantation. This case underscores that when classification proves difficult, descriptive reporting and collaborative management remain central to ensuring accurate documentation and optimal outcomes

Conclusion

Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA) syndrome, also known as Herlyn–Werner–Wunderlich syndrome, represents a rare but clinically significant subset of Müllerian duct anomalies. This case highlights an atypical variant characterized by bicornuate uterus, rudimentary cervix, segmental vaginal atresia, unilateral renal agenesis, and ureteric malpositioning requiring reimplantation. Such complex presentations underscore the limitations of existing classification systems and the importance of detailed anatomical description, systematic renal screening, and multidisciplinary management. Definitive surgical intervention, guided by individualized assessment and intraoperative findings, remains central to achieving symptom relief and preventing long-term morbidity. Ultimately, this case reinforces the need for integrating embryological understanding with clinical practice to optimize outcomes in patients with rare Müllerian anomalies.

Declarations

Consent to Publish

Written informed consent for publication of the patient's clinical details and accompanying images was obtained from the patient's parent/legal guardian.

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Author Contribution

Adnan Rehman performed the surgical procedure, contributed to data collection, and reviewed the manuscript. Gulmadin Hikmat drafted and wrote the manuscript. Muhammad Umar Haneef and Ayesha Tasneem participated in the surgical procedure. All authors reviewed and approved the final manuscript.

References

1. Bhamidipaty-Pelosi S et al. Müllerian anomalies and endometriosis: associations and phenotypic variations. Dec 01 2024 BioMed Cent Ltd. 10.1186/s12958-024-01336-1
2. Elgohary MA, Naik R, Elkafafi M, Hamed H, Ali Y. Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome: A case report. J Pediatr Surg Case Rep. Aug. 2023;95. 10.1016/j.epsc.2023.102662.
3. Kagan Arleo E, Troiano RN. Complex Mullerian Duct Anomalies Defying Traditional Classifica tion: Lessons Learned, *Journal of Fertilization: In Vitro - IVF-Worldwide, Reproductive Medicine, Genetics & Stem Cell Biology*, vol. 01, no. 03, 2013, 10.4172/2375-4508.1000115
4. Acién P, Acién M. Diagnostic imaging and cataloguing of female genital malformations. Oct 01 2016 Springer Verlag. 10.1007/s13244-016-0515-4
5. Dwivedi D, Jain M, Jain S, Jain S. A rare case report on complete cervical agenesis with vaginal atresia and suspended didelphys uterus with hematometra and left haematosalpinx. Int J Reprod Contracept Obstet Gynecol. Feb. 2020;9(3):1274. 10.18203/2320-1770.ijrcog20200914.
6. Acién P, Acién M. Unilateral renal agenesis and female genital tract pathologies, *Acta Obstet Gynecol Scand*, vol. 89, no. 11, pp. 1424–1431, Nov. 2010, 10.3109/00016349.2010.512067
7. Grimbizis GF et al. Aug., The ESHRE-ESGE consensus on the classification of female genital tract congenital anomalies, *Gynecol Surg*, vol. 10, no. 3, pp. 199–212, 2013, 10.1007/s10397-013-0800-x
8. Meena J, Bharti J, Roy KK, Kumar S, Singhal S, Shekhar B. Bicornuate Uterus with Complete Cervico-Vaginal Agenesis and Skeletal Deformity: A Case Report, *Journal of Obstetrics and Gynecology of India*, vol. 69, pp. 67–70, Apr. 2019, 10.1007/s13224-018-1107-7
9. Acién P, Navarro V, Acién M. Embryological–clinical classification of female genital tract malformations – a review and update, Jul. 01, 2025, *Elsevier Ltd*. 10.1016/j.rbmo.2024.104751

10. Dohbit JS, Meka E, Tochie JN, Kamla I, Mwadjie D, Foumane P. A case report of bicornis bicollis uterus with unilateral cervical atresia: An unusual aetiology of chronic debilitating pelvic pain in a Cameroonian teenager. BMC Womens Health. Jun. 2017;17(1). 10.1186/s12905-017-0396-9.
11. Dixit R, Duggireddy CS, Pradhan GS. Mullerian anomalies: revisiting imaging and classification. Dec 01 2025 Springer Sci Bus Media Deutschland GmbH. 10.1186/s13244-024-01879-2
12. Mooren ERM, Cleypool CGJ, de Kort LMO, Goverde AJ, Dik P. A Retrospective Analysis of Female Müllerian Duct Anomalies in Association With Congenital Renal Abnormalities, *J Pediatr Adolesc Gynecol*, vol. 34, no. 5, pp. 681–685, Oct. 2021, 10.1016/j.jpag.2021.04.013

Figures



Figure 1

Figure 1: IVP demonstrates congenital solitary right kidney with secondary changes: dilated tortuous ureter and right-sided hydronephrosis. Findings suggest obstructive uropathy in the solitary functioning kidney.

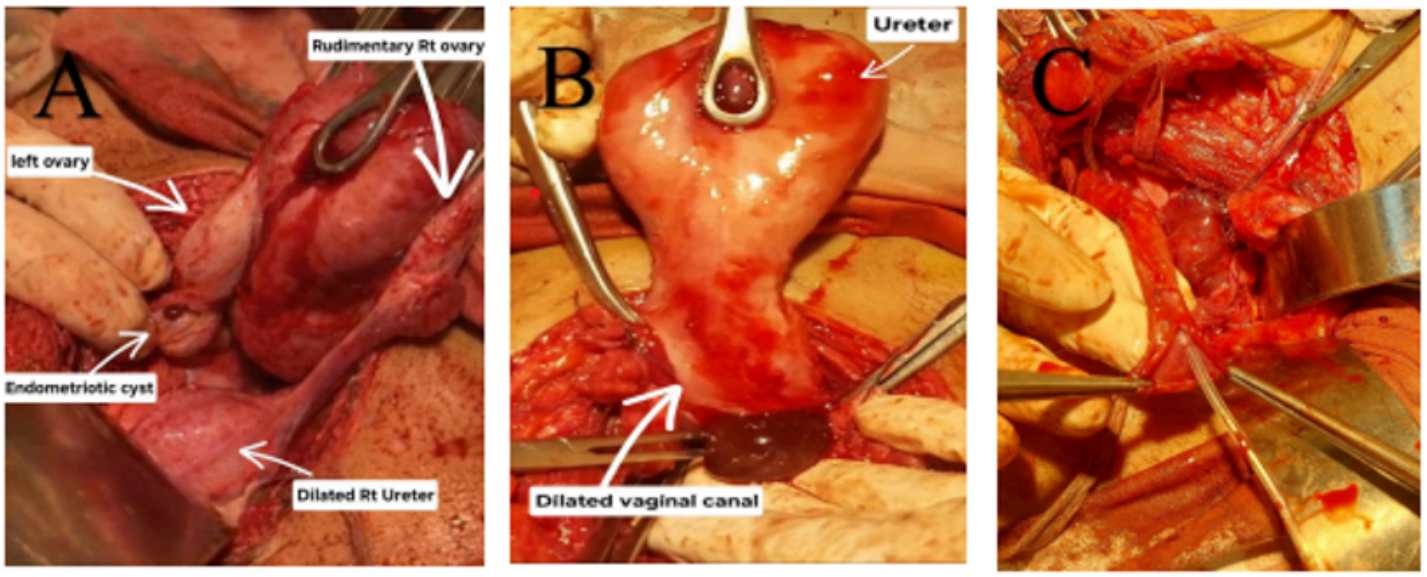


Figure 2

Figure 2: intraoperative images

A: Imaging reveals left ovarian endometriotic cyst with contralateral rudimentary ovary and associated right ureteric dilatation.

B: Imaging reveals Bicornuate uterus: dilated cervix and canal with hematocolpos drainage.

C: Image showing a dilated right ureter with a feeding tube passed for identification, dissected from the bladder and prepared for ureteric reimplantation.



Figure 3

Figure 3: X-ray KUB demonstrating right-sided JJ ureteric stent in situ.