

Original Research

Obstructed Hemivagina and Ipsilateral Renal Anomaly Syndrome: A Spectrum of Clinical Presentations, Diagnostic Challenges, and Surgical Management

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Academic Editor: Michael H. Dahan

Submitted: 26 June 2026 Revised: 10 August 2026 Accepted: 17 August 2026 Published: 29 September 2026

Abstract

Background: Müllerian anomalies are congenital abnormalities of the female reproductive tract that may present with a wide range of clinical manifestations, and some forms may remain undiagnosed until adolescence or until complications develop. This study aimed to evaluate the clinical presentations, radiological findings, diagnostic challenges, and surgical management of patients with obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome treated at a tertiary referral center. **Methods:** The study included patients diagnosed with OHVIRA syndrome from July 17, 2019, to October 1, 2024. Following ethics committee approval, cases were categorized into four different scenarios. Scenario 1: suspected OHVIRA syndrome during infancy; Scenario 2: obstructed hemivagina with hematocolpos at puberty; Scenario 3: spontaneous fistulization of hematocolpos; and Scenario 4: pregnancy following surgical management of OHVIRA syndrome. The diagnostic approach and follow-up processes for patients in each clinical scenario were summarized. **Results:** Six patients were categorized into four distinct clinical scenarios according to their clinical presentations. Five patients had confirmed OHVIRA syndrome, while one patient had suspected OHVIRA syndrome based on the presence of ipsilateral renal agenesis and a cystic vaginal lesion. Clinical presentations ranged from suspected OHVIRA syndrome presenting with mucocolpos during infancy to hematocolpos at puberty, spontaneous fistulization, and successful pregnancy following previous vaginal septum resection. Pelvic ultrasonography (USG) and magnetic resonance imaging (MRI) accurately identified uterus didelphys, obstructed hemivagina and hematocolpos. We present a case of mucocolpos and a case describing the pregnancy outcome following surgery for hematocolpos. Three patients underwent vaginoscopic hematocolpos drainage, one patient was diagnosed following spontaneous fistulization of hematocolpos, and five patients had favorable clinical outcomes without recurrent obstruction during follow-up. The remaining patient, who was followed during early childhood for mucocolpos, is awaiting menarche. **Conclusions:** OHVIRA syndrome encompasses a broad spectrum of age-dependent clinical presentations, which can pose significant diagnostic challenges. Early recognition and individualized management based on clinical and anatomical findings are essential, with USG and MRI playing key roles in establishing the diagnosis.

Keywords: fistulization; hematocolpos; mucocolpos; obstructed hemivagina and ipsilateral renal anomaly syndrome; placenta previa; vaginostomy

1. Introduction

Müllerian anomalies are developmental disorders that can present with a wide range of anatomical and clinical features. Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome, also known as Herlyn–Werner–Wunderlich syndrome, is a rare and complex congenital urogenital anomaly, with an estimated incidence of 0.1%–3.8% [1]. Characterized by uterus didelphys, obstructive hemivagina, and ipsilateral renal agenesis, this syndrome was first described in 1922 [2]. OHVIRA syndrome is one of the obstructive Müllerian anomalies increasingly diagnosed among adolescents in recent years [3]. Patients typically present shortly after menarche with abdominal pain or dysmenorrhea resulting from hematocolpos and the related anatomical disorders.

In gynecologic practice, ultrasonography (USG) is a primary imaging modality for evaluating Müllerian anomalies, and three-dimensional USG is currently recommended for the assessment of congenital uterine anomalies [4].

The diagnosis of unilateral renal agenesis, a significant congenital anomaly of the kidney and urinary tract (CAKUT), can be established during the antenatal period through ultrasonographic visualization of the fetal kidneys, which can be detected as early as 10 to 12 weeks of gestation [5]. Congenital unilateral renal agenesis occurs in approximately 1 in 2000 live births [6]. The incidence of urinary tract anomalies in all pregnancies has been reported to be approximately 2% [5]. Additionally, Müllerian duct anomalies are reported in 25% to 50% of women with unilateral renal agenesis. Although concomitant congenital re-



nal agenesis can be detected during antenatal ultrasound scans, Müllerian anomalies may remain unrecognized until later in life, when complications arise [7,8].

This condition has important implications for adolescent gynecology, future fertility, and obstetric outcomes. Accurate diagnosis of this rare condition requires a high index of clinical suspicion. Recognizing the diverse clinical presentations is essential for timely diagnosis and appropriate management. Herein, we aimed to present four different clinical scenarios associated with OHVIRA syndrome based on our institutional experience.

2. Materials and Methods

We retrospectively analyzed data from six patients evaluated between July 17, 2019, and October 1, 2024, who presented with clinical and imaging findings suggestive of OHVIRA syndrome. Among these patients, five fulfilled the diagnostic criteria for confirmed OHVIRA syndrome, defined by the presence of uterine duplication, an obstructed hemivagina, and an ipsilateral renal anomaly. One patient was classified as having suspected OHVIRA syndrome because ipsilateral renal agenesis had been identified during the prenatal period, whereas the subsequent vaginal cystic lesion could not be definitively confirmed as an obstructed hemivagina. Clinical presentation, physical examination findings, imaging findings, diagnostic evaluations, treatment procedures, and follow-up data were retrospectively reviewed and obtained from the patients' medical records. The study was approved by the Bursa City Hospital Ethics Committee (Approval No.: 2024-19/6). The study was conducted at Bursa City Hospital.

The patients were categorized into four clinical scenarios (Table 1). Scenario 1: suspected OHVIRA syndrome in infancy; Scenario 2: obstructed hemivagina-hematocolpos at puberty; Scenario 3: spontaneous fistulization of hematocolpos; and Scenario 4: pregnancy following surgical management of OHVIRA syndrome. The diagnostic evaluation, treatment, and follow-up of patients at our institution were conducted as follows. All patients were scheduled for their first post-operative follow-up visit at 3 months. Follow-up consisted of gynecological examination and pelvic USG to evaluate vaginal patency, menstrual irregularities, reoperation, and resolution or recurrence of hematocolpos. Clinical and laboratory findings suggestive of infection were also assessed during follow-up.

3. Results

3.1 Suspected OHVIRA Syndrome in Infancy

A 13-mm cystic structure was identified on the right side of the pelvis during suprapubic USG in a 2-year, 2-month-old child (Fig. 1). Mucocolpos was diagnosed through USG (GE Voluson S10 BT 18, SPC330) examination in the patient with renal agenesis, diagnosed in the antenatal period. Physical examination excluded an imper-

forate hymen. Considering the limitations of imaging in evaluating prepubertal Müllerian structures, a Gartner duct cyst was also considered in the differential diagnosis of this patient. Given the suspicion of OHVIRA syndrome, the family was counselled regarding the need for gynecological examination following the first menstruation.

3.2 Hematocolpos

3.2.1 Case 1

A 12-year-old girl who was scheduled to undergo surgery for an endometrioma at another hospital, presented to our tertiary care center. Laboratory findings, including tumor markers such as cancer antigen 125, lactate dehydrogenase, human chorionic gonadotropin, and carcinoembryonic antigen, as well as infection parameters such as C-reactive protein, were within normal limits. USG revealed bilateral normal ovarian tissue. However, the presence of uterus didelphys and right-sided hematocolpos prompted evaluation of the urinary tract, which revealed right renal agenesis. The magnetic resonance imaging (MRI) report revealed a didelphys uterus or bicornuate uterus with hematometra in the right uterine cavity. However, the MRI images demonstrated two separate uterine bodies and two distinct cervixes, consistent with uterus didelphys. The left uterine cavity appeared normal. Transhymenal vaginotomy was performed with a resectoscope. The left cervix was visualized in the left apical portion of the vagina, while a swollen area was observed at the right vaginal wall. With the monopolar hook electrode tip, an incision was made in the lower part of the area where the swelling was most prominent, and the organized hemorrhagic content was drained. To prevent restenosis at the incision site, a 22-Fr Foley catheter was placed through the incision, and the catheter balloon was inflated with 30 mL saline. The patient was monitored for 72 hours with a Foley catheter in the vagina and received metronidazole and ceftriaxone antibiotic prophylaxis. During the 5-year follow-up period, the patient remained asymptomatic with no need for recurrent surgery. MRI and intraoperative images of the patient are presented in Fig. 2.

3.2.2 Case 2

The patient was 11 years and 10 months old at the time of presentation to our clinic. During nephrology follow-up for left renal agenesis, USG revealed a thick-walled, dense cystic lesion, measuring 102 × 55 mm, extending from the posterior bladder towards the left adnexal space. Based on these findings, a pediatric surgery consultation was requested.

Based on the appearance of hematocolpos, MRI suggested an imperforate hymen as the preliminary diagnosis (Fig. 3). Gynecological examination revealed normal menstruation and hymenal patency. The patient underwent transhymenal vaginoscopic drainage of the left hematocolpos with a preliminary diagnosis of left-sided hematocolpos. As

Table 1. Patient-level summary of OHVIRA syndrome cases across four clinical scenarios.

Scenario / Case	Age	Presentation / chief complaint	Menarcheal status	Renal anatomy	Genital anatomy	Imaging findings	Treatment	Follow-up
Scenario 1: suspected OHVIRA syndrome in infancy	2 years 2 months	Incidental finding (suprapubic USG)	Pre-menarchal	Right renal agenesis (antenatal diagnosis)	13 mm cystic structure, right pelvis; imperforate hymen excluded; Gartner duct cyst in differential diagnosis; mucocolpos	Suprapubic USG: 13 mm cystic structure, right pelvis	Conservative management; gynecological examination planned after first menstruation	Awaiting menarche
Scenario 2 – Case 1: hematocolpos	12 years	Referred for endometrioma surgery (outside hospital)	Post-menarchal	Right renal agenesis	Uterus didelphys; right hematocolpos; bilateral normal ovarian tissue	USG: uterus didelphys, right hematocolpos MRI: didelphys/bicornuate uterus, hematometra in right cavity; left cavity normal	Transhymenal vaginotomy; monopolar hook incision; drainage of organized hemorrhagic content; 22-Fr Foley catheter (balloon 30 mL saline); 72 h antibiotic prophylaxis (metronidazole + ceftriaxone)	5 years; no complaints; no re-operation
Scenario 2 – Case 2: hematocolpos	11 years 10 months	Incidental on nephrology follow-up	Post-menarchal (normal menses)	Left renal agenesis	Left hematocolpos; normal hymenal patency on examination	USG: 102 × 55 mm thick-walled cystic lesion MRI: hematocolpos (initial dx: imperforate hymen)	Transhymenal vaginoscopic drainage; 22-Fr Foley catheter; 72 h monitoring	18 months; no restenosis
Scenario 2 – Case 3: hematocolpos	12 years 2 months	Routine gynecology check-up; no pain or dysmenorrhoea	Post-menarchal (menarche 3–4 months prior)	Right renal agenesis (neonatal diagnosis)	Uterus didelphys; ~5 × 3.5 cm hypoechoic mass, right upper vagina (hematocolpos)	USG: uterus didelphys, 5 × 3.5 cm smooth-contoured hypoechoic mass MRI: confirmed USG findings	Vaginotomy; plasmakinetic hook incision; drainage of brown organized hematoma; 22-Fr Foley catheter; hymen intact throughout	14 months; uneventful

Table 1. Continued.

Scenario / Case	Age	Presentation / chief complaint	Menarcheal status	Renal anatomy	Genital anatomy	Imaging findings	Treatment	Follow-up
Scenario 3: fistulised hematocolpos	12 years	Abdominal pain	Post-menarchal	Right renal agenesis	Uterus didelphys; right hematocolpos (51 × 49 mm); spontaneous fistula to vagina; OHVIRA syndrome	USG: 51 × 49 mm cystic mass MRI: uterus didelphys and hematocolpos confirmed (OHVIRA syndrome)	Transhymenal vaginoscopy; fistula identified (spontaneous drainage)	3 years 7 months; no re-loculation
Scenario 4: pregnancy following surgical management of OHVIRA syndrome	32 years (G1P0)	Antenatal care; placenta previa	Post-menarchal (adult, pregnant)	Unilateral renal agenesis	Uterus didelphys; pregnancy in left uterine cavity; placenta previa; vaginal septum resection 15 years earlier	USG: 14-week fetus in left uterine cavity; uterus didelphys; placenta previa	Elective cesarean section at 37 weeks (placenta previa)	3335 g infant; Apgar 9/10; no post-operative complications

Abbreviations: USG, ultrasonography; MRI, magnetic resonance imaging; OHVIRA, obstructed hemivagina and ipsilateral renal anomaly; G1P0, gravida 1, para 0.



Fig. 1. Ultrasonographic view of the mucocolpos on the right lateral vaginal wall.

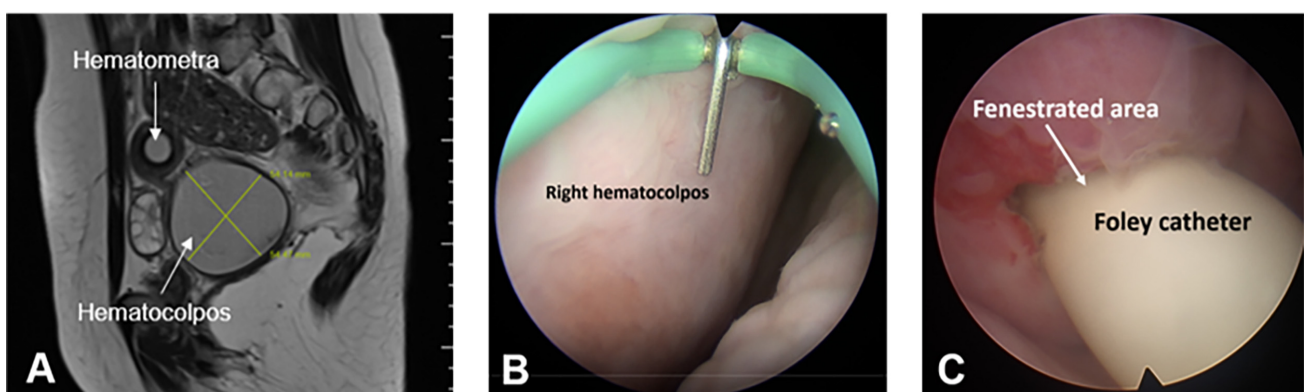


Fig. 2. MRI and intraoperative images of the Case 1 in Scenario 2. (A) Magnetic resonance imaging of hematometra and hematocolpos. (B) Vaginoscopic view of the right hematocolpos. (C) Appearance of catheterization after hematocolpos drainage.

in the first case, the patient was monitored for 72 hours with a Foley catheter placed at the incision site. No evidence of restenosis was found during the 1-year, 6-month follow-up period.

3.2.3 Case 3

The patient, who had been diagnosed with right renal agenesis in the neonatal period, presented to the gynecol-

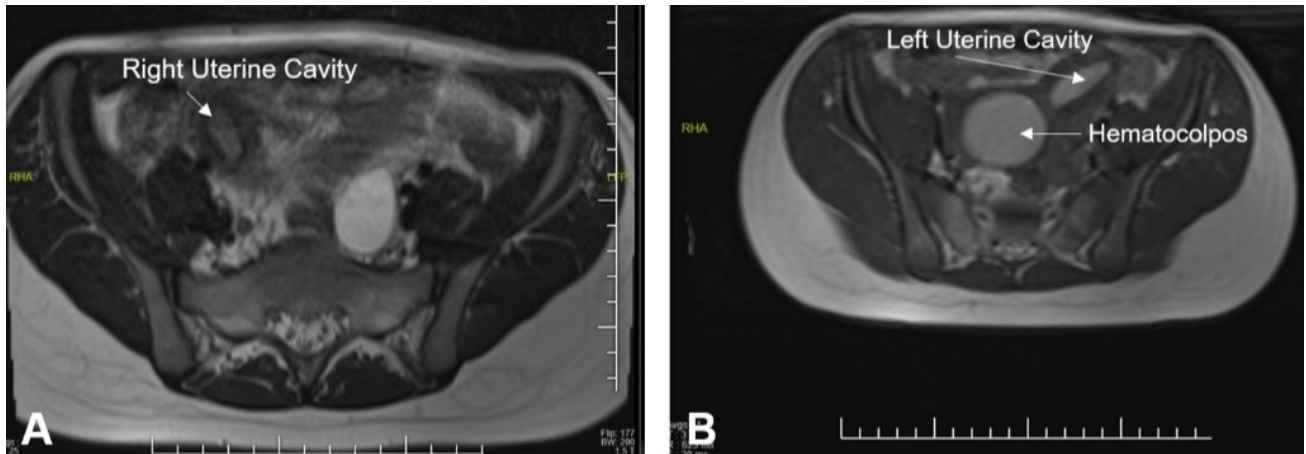


Fig. 3. MRI images of Case 2 in Scenario 2. (A) Normal right uterine cavity. (B) View of the left uterine cavity and left hematocolpos.

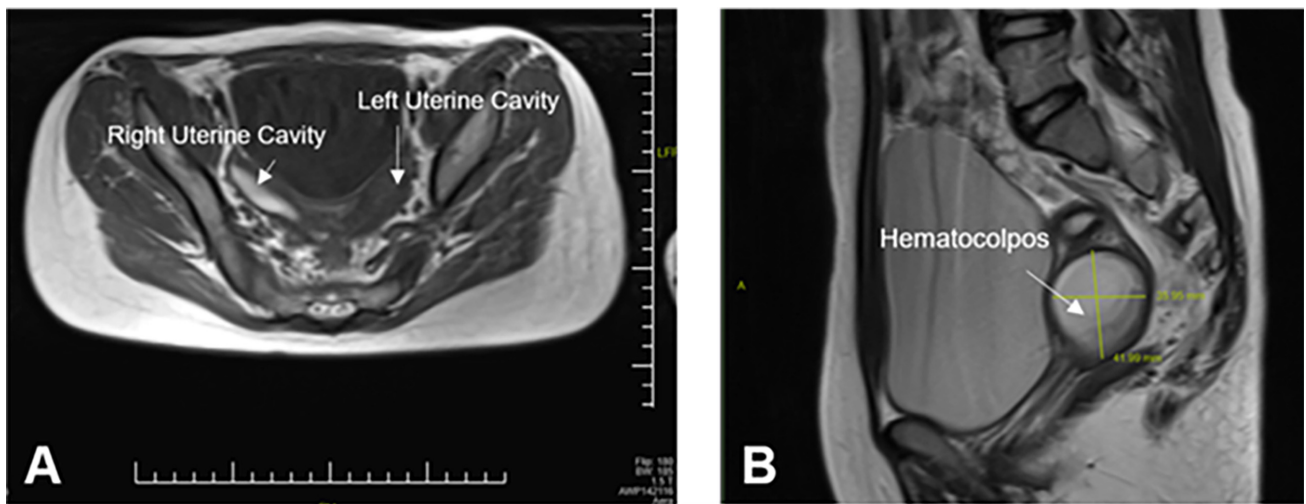


Fig. 4. MRIs of Case 3 in Scenario 2. (A) Image of both uterine cavities. (B) Image of hematocolpos.

ogy outpatient clinic for routine evaluation at 12 years and 2 months of age. Menarche had occurred 3–4 months prior to presentation. The patient reported no abdominal pain or dysmenorrhea. USG revealed uterine didelphys. At the upper vaginal level on the right side, distal to the cervical os, a hypoechoic mass consistent with a thick-walled hematoma with smooth contours, approximately 5×3.5 cm, was observed, consistent with hematocolpos. MRI also supported the ultrasonographic findings (Fig. 4).

Vaginotomy revealed the left cervical ostium, with a bulge of the vaginal wall observed to the right of the cervix. The bulging area was incised using a plasmakinetic hook. A brown, locally organized hematoma was drained. Entry into the hemivagina through the incised area revealed a cervical opening. The hematoma within the hemivagina was completely drained by irrigation. Subsequently, a 22-Fr Foley catheter was inserted into the incised area. The hymen remained intact during and after the procedure. The 14-month follow-up period was unremarkable.

3.3 Fistulized Hematocolpos

A 12-year-old girl presented with abdominal pain, and USG revealed a 51×49 mm cystic mass. MRI confirmed the diagnosis of uterus didelphys and hematocolpos (Fig. 5). The patient had been diagnosed with right renal agenesis, and the findings supported OHVIRA syndrome. Following the scheduled preoperative evaluation, transvaginal vaginoscopy revealed a fistula to the vagina from the right-sided hematocolpos. The fistulized cavity was irrigated to ensure adequate drainage of the hematocolpos. As satisfactory drainage had been achieved, no vaginal septum resection was performed. No reaccumulation was detected in the patient during the 3-year, 7-month follow-up period.

3.4 Pregnancy Following Surgical Management of OHVIRA Syndrome

A 32-year-old gravida 1, para 0 (G1P0) patient presented to the obstetric clinic for routine pregnancy follow-up. The patient had a known history of unilateral renal agenesis and reported undergoing vaginal septum resection at a university hospital 15 years earlier for hematocolpos.

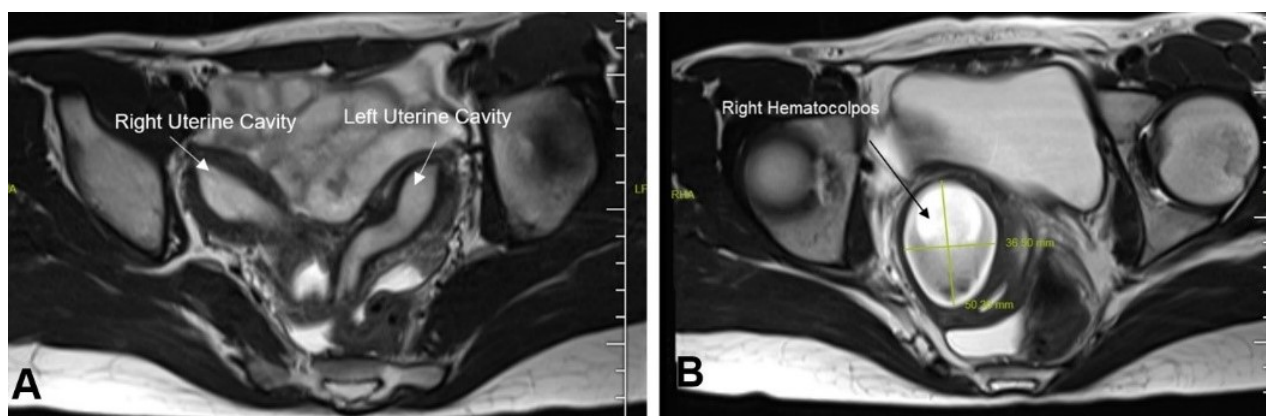


Fig. 5. MRI of uterus didelphys and hematocolpos prior to fistulization. (A) The view of two separated uterine cavities. (B) The view of the right hematocolpos.

USG at 14 gestational weeks revealed a fetus located in the left uterine cavity and an appearance consistent with uterus didelphys. Due to subsequent ultrasonographic findings consistent with placenta previa, the patient was followed up in the perinatology department. Cesarean section was planned for the case of placenta previa at 37 weeks of gestation, with fetal biometry consistent with gestational age. A 3335 g infant was delivered, with Apgar scores of 9 and 10 at 1 and 5 minutes, respectively. The post-operative follow-up was uneventful.

4. Discussion

Müllerian anomalies, with a prevalence of 5.5%, are embryologic abnormalities associated with infertility and pregnancy complications [9]. According to the American Society for Reproductive Medicine Müllerian Anomalies Classification 2021 (ASRM MAC2021), Müllerian anomalies are classified into nine categories. Uterus didelphys with an obstructed hemivagina is classified as a specific variant within the uterus didelphys category [10].

Although the co-occurrence of urinary and Müllerian anomalies is well recognized, the appropriate emphasis on this association in clinical practice remains uncertain [11,12,13]. In a systematic review by Bonetti et al. [13], which included 35 studies published between 1988 and 2022 and a total of 526 patients, the mean age at symptom onset was 14.45 years, whereas the mean age at diagnosis was 16.36 years, indicating a potential diagnostic delay. In our patients, vaginoscopy enabled diagnosis in the early post-menarchal period. The American College of Obstetricians and Gynecologists recommends that adolescents have their first reproductive health visit between 13 and 15 years of age, although earlier evaluation may be appropriate when clinically indicated [14,15]. This assessment provides an opportunity to identify OHVIRA syndrome early and prevent potential long-term complications.

In a study conducted in our country, the median age at diagnosis of unilateral renal agenesis was 2 years, whereas in our clinical practice, the age at first diagnosis ranges from

the antenatal period to 50 years [16]. Renal agenesis has been reported in 20% of patients with uterus didelphys [9]. However, a recently published review reported that ipsilateral renal agenesis, predominantly on the right side, was detected in 89% of OHVIRA cases [13].

La Scola et al. [6] recommend screening for genital anomalies using abdominopelvic USG between thelarche and menarche in all girls diagnosed with congenital solitary kidney. In contrast, our asymptomatic patient with antenatally diagnosed renal agenesis was scheduled for gynecological evaluation after menarche, in accordance with our institutional follow-up practice. In our clinical experience, evaluating anomalies in the internal genitalia can be challenging due to their immature appearance on pelvic USG performed at an early age. Although rare, antenatal diagnosis is possible; similar to our case, in which renal agenesis was diagnosed during the antenatal period and mucocolpos was subsequently identified during infancy, Kim et al. [17] also diagnosed OHVIRA syndrome in a female fetus at 32 weeks of gestation. Advances in imaging techniques and increased clinical awareness have facilitated the early diagnosis of Müllerian anomalies. OHVIRA syndrome can be diagnosed even during infancy based on the presence of mucocolpos resulting from the obstructed hemivagina [18]. Although mucocolpos cases associated with imperforate hymen have been reported in the literature, an imperforate hymen was not present on genital examination in our patient. The presence of accompanying renal agenesis therefore led us to make a preliminary diagnosis of OHVIRA syndrome [19]. As illustrated by our patient in Scenario 1, when unilateral renal agenesis is detected in a female fetus during second-trimester screening, the possibility of an associated Müllerian duct anomaly should also be considered.

The use of USG as the primary imaging method in gynecological practice allows separate visualization of both ovarian tissues, facilitating the diagnosis of hematocolpos. This approach may help prevent misdiagnosis of endometrioma, as occurred in the first case of the Scenario 2. The

ability to visualize the ovarian tissues separately represents another advancement in the diagnosis of obstructive Müllerian anomalies.

A recent study reported that, as in our case, hysteroscopy and intraoperative USG can be used more effectively for surgery [20]. As in our cases, vaginoscopic drainage of the obstructed hemivagina has been reported as an effective treatment modality [21]. Vaginoscopic incision of the vaginal septum has even been reported in the premenarchal period in patients with mucocolpos [22]. In patients with obstructive symptoms, prompt drainage is mandatory. In accordance with the patients' religious beliefs and social background, transhymenal vaginoscopic drainage was performed, as previously reported, and a Foley catheter was placed inside the fenestrated area to prevent reobstruction [23]. However, urgent treatment of obstructive symptoms should always be guided by clinical indications. A comprehensive review and meta-analysis showed that vaginal septum resection was performed in 83% of cases, making it the most commonly performed surgical procedure [13]. As Bonetti et al. [13] also noted, more complex surgical procedures should be reserved for recurrent or complicated cases. In our cohort, the first post-operative follow-up examination was performed at 3 months, and the longest follow-up period was 5 years. None of the patients required reoperation. Patients were followed for menstrual irregularities, ultrasonographic evidence of relocation or recurrent hematocolpos, and clinical and laboratory signs of infection. The favorable outcomes observed in our patients may be related to the lower incision site near the bulging area, although this observation should be interpreted with caution given the limited number of cases. None of the adolescent patients received post-operative oral contraceptive therapy, as complete drainage had been achieved.

Multiple surgical interventions have been reported in a patient who developed recurrent pelvic abscesses following delayed diagnosis [24]. Nejkovic and Stanojevic [25] also reported a radical surgical procedure, including hemihysterectomy with vaginectomy, in a 28-year-old patient with a delayed diagnosis. Although laparoscopy has been advocated for the diagnosis of accompanying adhesions and endometriosis, routine laparoscopic surgery is not currently recommended [2]. Minimally invasive surgical management is currently favored. As demonstrated in our cases, incision of the vaginal septum while preserving the hymen is one available surgical option. Due to the incorrect cervical anatomy caused by hematocolpos and hematometra, we believe that making a wide proximal incision may pose a risk of subsequent cervical insufficiency.

Wozniakowska et al. [26] reported a patient with a Müllerian anomaly consistent with uterus didelphys detected during appendectomy and presenting with purulent vaginal discharge after menarche. In that patient with right renal agenesis, a pelvic mass was palpated during rectal examination, and vaginoscopy revealed a fistula tract from the right hematomyometra to the left vaginal opening. Unlike

the case reported by Wozniakowska et al. [26], our patient in Scenario 3 had no preoperative findings to support infection, such as leukocytosis or elevated C-reactive protein levels.

Previous studies have reported that OHVIRA syndrome, like other Müllerian anomalies, is associated with adverse obstetric outcomes [27]. However, this association does not imply that these patients will invariably experience preterm birth. The reported live birth rate of 73% is encouraging, and one of our patients with OHVIRA syndrome achieved a term live birth [13]. Unlike the patient with OHVIRA syndrome reported by Cappello et al. [28], who underwent cesarean section for breech presentation at 33 weeks and 5 days of gestation, our patients with OHVIRA syndrome delivered at term. In a case series evaluating obstetric outcomes in patients with OHVIRA syndrome who underwent surgical drainage of hematocolpos, Bunnell et al. [27] also reported term and near-term deliveries. In our case, the occurrence of placenta previa was considered an independent obstetric finding rather than a consequence of OHVIRA syndrome.

Limitations

This study has several limitations. The small sample size and retrospective design limit the ability to draw definitive conclusions regarding long-term outcomes. However, due to the rarity of OHVIRA syndrome, this case series provides valuable insights into its diverse clinical presentations, diagnostic challenges, and surgical management. Additionally, the reproductive outcome observed in a single patient should be interpreted as an individual clinical observation rather than as evidence of long-term fertility outcomes. Further studies involving larger patient cohorts and longer follow-up periods are needed to better define optimal management strategies for OHVIRA syndrome.

5. Conclusions

Raising awareness of OHVIRA syndrome may facilitate earlier diagnosis. All female patients diagnosed with renal agenesis should undergo pelvic USG for possible Müllerian anomalies after thelarche; conversely, patients diagnosed with Müllerian anomalies should also be evaluated for associated renal anomalies. In addition to acute complications, potential long-term complications such as infertility, endometriosis, miscarriage, and preterm birth, should be considered. For patients diagnosed with OHVIRA syndrome during childhood or adolescence, management should be individualized according to clinical symptoms, the degree of obstruction, anatomical findings, and the risk of recurrence.

Availability of Data and Materials

The datasets used and analyzed during the current study are available from the corresponding author upon reasonable request.

Author Contributions

ZA: Conceptualization, Investigation, Writing-Original Draft, Writing-Review & Editing. OOU: Investigation, Data Curation, Writing-Review & Editing. SRO: Methodology, Supervision, Writing-Review & Editing. SS: Investigation (surgical management), Writing-Review & Editing. BU: Investigation (renal anomaly assessment), Writing-Review & Editing. All authors read and approved the final manuscript. All authors have participated sufficiently in the work and agreed to be accountable for all aspects of the work.

Ethics Approval and Consent to Participate

The study was carried out in accordance with the guidelines of the Declaration of Helsinki. The study was approved by the Bursa City Hospital Ethics Committee (Approval number: 2024-19/6). The requirement for informed consent was waived by the Bursa City Hospital Ethics Committee due to the retrospective nature of the study. Our institution is a research center, so all patients are informed of this point prior to examinations.

Acknowledgment

Not applicable.

Funding

This research received no external funding.

Conflicts of Interest

The authors declare no conflicts of interest.

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