A Rare Anomaly of the Müllerian System: OHVIRA Syndrome. Comprehensive Literature Review and Report of Seven Cases

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Purpose: Müllerian anomalies are rare, and the obstructed hemivagina and ipsilateral renal anomaly/agenesis (OHVIRA) syndrome is the rarest. Its subtle and nonspecific symptoms cause delays in diagnosis, especially in prepuberty. This study aims to remind this rare anomaly and review its symptoms, clinical and radiological findings, and treatment in pre- and post-pubertal girls.

Materials and methods: We conducted a retrospective chart review of cases of OHVIRA syndrome that were diagnosed and treated between May 2018 and June 2024. We evaluated the results and reviewed relevant literature.

Results: Seven patients were diagnosed with OHVIRA syndrome during the study period. Two patients were diagnosed in the pre-pubertal period, and all post-pubertal cases, except one, underwent surgery. Five (71%) of the seven patients had left-sided obstructed hemivagina with ipsilateral renal agenesis.

Conclusion: OHVIRA syndrome is a rare condition. Its diagnosis and management depend on the patient's age, symptoms, familiarity with the syndrome, and teamwork. Although a renal anomaly is part of the triad of this syndrome, OHVIRA syndrome often remains undiagnosed during infancy, even in cases of prenatally diagnosed renal agenesis. Screening for OHVIRA syndrome and associated urinary anomalies in cases of renal agenesis, particularly in prepubertal cases, is crucial.

Keywords: herlyn-werner-wunderlich syndrome; müllerian anomaly; obstructed hemivagina; ohvira syndrome; vaginal septum

INTRODUCTION

Obstructed hemivagina with uterine didelphys and ipsilateral renal anomaly is a rare congenital anomaly that was first reported in 1922. Laufer et al. proposed the acronym OHVIRA in 2007 to provide an everyday basis for describing the syndrome[1].

OHVIRA syndrome is associated with abnormal development of the Müllerian and Wolffian ducts and constitutes only 0.16%–10% of all Müllerian duct anomalies, with an incidence of 2%–3%[2-5]. It was also emphasized that the renal system should be evaluated simultaneously in Müllerian anomalies since OHVIRA cases accompanied unilateral renal agenesis[6,7].

OHVIRA syndrome is usually regarded as a post-pubertal disease and may present with acute abdomen, recurrent dysmenorrhea, mucoidal vaginal discharge, and hematometrocolpos[8,9]. In contrast, rare symptomatic presentation, vague clinical history, lack of awareness, and a wide variety of phenotypic types contribute to delayed diagnosis in the pre-pubertal age group. Delays in diagnosis may cause unnecessary interventions and failure to intervene promptly during the post-pubertal period. Consequently, the fertility of patients may be impaired because of late complications. Thus, when female patients present with dysplastic or absent kidneys, uterovaginal anomalies should be established to allow early recognition and management, especially in pre-pubertal girls[10,11].

The English literature reports mostly postpubertal cases, and prepubertal cases are rare. The rarest are those diagnosed during the prenatal or early infantile periods[11,12]. This study aimed to draw attention to this rare anomaly and discuss its presentation, diagnosis, and treatment or follow-up in pre- and postpubertal girls. Thus, we conducted a retrospective analysis of our cases and a comprehensive literature review.

MATERIALS AND METHODS

Study population

The clinical records of patients diagnosed and treated at an academic center for OHVIRA syndrome between May 2018 and June 2024 were retrospectively analyzed. The collected data included patient age, clinical history, physical examination results, ultrasonography (US), computed tomography (CT), magnetic resonance imaging (MRI) results, indications for surgery, details

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Table 1. Demographics of patients with OHVIRA syndrome

Variables	Prepubertal Patients (N=2)	Postpubertal Patients (N=5)
Age, months; mean ± SD (range)	23.5 ± 26.16 (5–42)	152.4 ± 10.03 (144–168)
Diagnostic method — Prenatal US	1	1
Diagnostic method — Abdominal US	2	4
Diagnostic method — Suprapubic pelvic US	2	4
Diagnostic method — MRI	—	2
Diagnostic method — Abdominopelvic CT	—	—
Presenting symptom — Prenatal diagnosed renal agenesis	1	2
Presenting symptom — Labia synechia	1	—
Presenting symptom — Chronic pelvic pain	—	3
Presenting symptom — Renal agenesis and contralateral VUR	—	1
Presenting symptom — Sudden pelvic pain	—	1
Treatment — Conservative	2	—
Treatment — Surgical intervention	—	4
Outcome — No complaints during follow-up	2	3 (in patients No. 3, 4, and 6)
Outcome — Immediate relief of pain after surgery	—	—
Associating urinary anomaly — Ipsilateral renal agenesis	1	4
Side — Right	—	1
Side — Left	1	2
Contralateral urinary duplication	—	1
Contralateral VUR	—	2
Follow-up, months; mean ± SD (range)	57.5 ± 54.44 (19–96)	36.5 ± 21.36 (13–60)

Abbreviations: OHVIRA, Obstructed Hemivagina Ipsilateral Renal Anomaly/Agenesis; US, Ultrasonography; CT, Computerized Tomography; MRI, Magnetic Resonance Imaging; VUR, Vesicoureteral Reflux.

of operations, and follow-up status.

All the patients' families provided written informed consent regarding the operation type, possible complications, and possible publication of the results. The Institutional Review Board of Prof. Dr. Cemil Taşcıoğlu City Hospital, Training and Research Center, approved the study design (22.07.2024/14).

Inclusion and exclusion criteria

Patients with a more than one-year follow-up period were included in the study.

Operative technique

The patients underwent a detailed physical examination in the proper lithotomy position under general anesthesia before any intervention and were evaluated endoscopically. For endoscopic examination of the vesical and vaginal cavities and visualization of the uterine cervixes before and after surgical excision of the vaginal septum, we used a 9 Fr pediatric cystoscope (Storz®, Germany). While septum excision was performed on all adolescent patients, only vaginoscopy and cystoscopy were performed following physical examination in the pre-adolescent patients because neither a visible vaginal mass protruding outward was detected on physical examination nor associated complications were found. Thus, asymptomatic prepubertal patients were followed up conservatively.

Vaginal septum excision and hydrometrocolpos evacuation were performed in adolescent patients. To perform vaginal septum excision, a no. 16 needle was inserted into the bulging area, which is visible and accessible through the introitus vagina without hymen defloration in most cases. The location of hematocolpos was confirmed by blood-stained turbid fluid aspiration. An incision was made using monopolar cautery from where the needle was inserted. Through this incision, the septum was incised longitudinally, and a septal strip that was one–two centimeters thick and three–four centimeters long was removed using a vascular sealing device. Excision was stopped approximately one centimeter close to the intercervical junction to prevent excessive bleeding or injury to the uterine cavity. However, bleeding occurred in two cases at the proximal end of the excision line, and continuous suturing with 3/0 absorbable sutures along the excision line was sufficient to stop the bleeding. All surgical procedures were performed under general anesthesia.

Postoperative evaluation

In the first two operated adolescent cases, we left a no. 16 Foley catheter in the previously obstructed hemivagina after septum excision for three to five days to prevent post-excisional obstruction and to help maintain blood flow. However, we noticed that the Foley catheter dislodged shortly after the operation; therefore, we decided not to place a Foley catheter in the other cases. During subsequent visits, the dimensions of both the cavity and opening of the vagina were evaluated endoscopically one month and three months after surgery. All patients were followed up regularly by a pediatric nephrologist twice a year and once a year by our pediatric surgery team, except for one, to control renal function and the Müllerian system.

Statistical analysis

Descriptive statistics were used, and the results are expressed as percentages.

Methodology of literature review

Specifically, we conducted a systematic search of PubMed, Scopus, and Web of Science databases using the keywords: "Herlyn-Werner-Wunderlich syndrome," "Müllerian anomaly," "obstructed hemivagina," "OHVIRA syndrome," "Vaginal septum," and "case report" (with appropriate MeSH terms where applicable). The search included articles published between January 1979 and December 2024. We included English-language publications that reported individual cases or small case series with sufficient clinical and surgical data. Articles without original patient data or duplicate cases were excluded.

RESULTS

Seven patients aged five months–14 years were diagnosed with OHVIRA syndrome during the study pe-

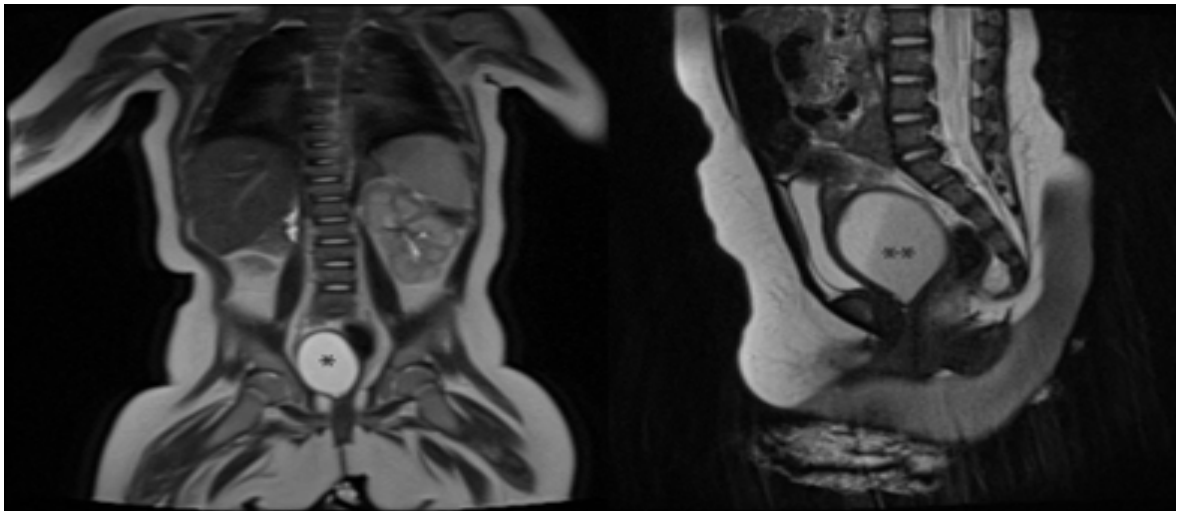

Figure 1. Magnetic resonance images of right-sided hydrometrocolpos (3 × 2 cm in size) in a five-month-old patient prenatally diagnosed with right renal agenesis. (Black asterisk: right hydrometrocolpos in the coronal plane; double black asterisk: hydrometrocolpos in the sagittal plane).

riod. One post-pubertal patient presented with acute abdominal pain. One of the post-pubertal patients was diagnosed with a right duplicated urinary system and reflux to it and left renal agenesis during early infancy. However, the patient was not diagnosed with OHVIRA syndrome until menarche onset. A pediatric nephrologist referred a post-pubertal patient because she was diagnosed with right vesicoureteral reflux (VUR) with left renal agenesis. Two of the seven patients were diagnosed while being investigated for genitourinary anomalies or complaints during the prepubertal period. One prepubertal patient was prenatally diagnosed with right renal agenesis. She was referred to our clinic by the same pediatric nephrologist when she was five months old for further investigation to detect other possible associated anomalies. The second prepubertal patient was referred to our department for the investigation of recurrent labial synechiae. Urinary US incidentally revealed left renal agenesis while investigating whether the upper urinary system was affected by a chronic obstruction. None of the prepubertal patients presented with acute or chronic abdominal symptoms.

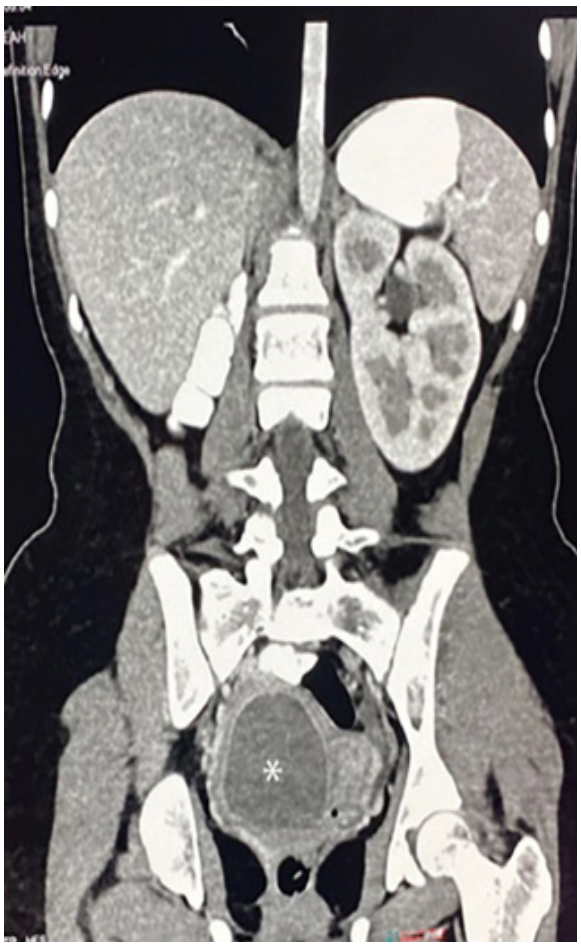

Figure 2. Magnetic resonance image of right hematometrocolpos (6 × 7 cm) in a 12-year-old girl (white asterisk: right hematometrocolpos).

In six cases, cystoscopy revealed no additional anomalies, and we observed no ureteral orifice on the ipsilateral agenetic side. In one case with a duplex system, the right ureteral orifice appeared deformed because of a previous cystoscopic antireflux procedure. Post-excisional vaginoscopy revealed an inflamed hemivagina and a flat uterine cervix with a dilated ostium.

In the control vaginoscopy, no fibrosis/stricture was observed in all operated patients at one and three months after the operation.

The details and descriptive characteristics of the cases are as follows.

Patient one

A five-month-old girl was referred to our department with prenatally diagnosed right-sided renal agenesis. Postnatal ultrasonography (US) revealed a 10 × 20 mm cystic mass in the right adnexal region. MRI confirmed uterus didelphys and possible obstructed hemivagina (**Figure 1**). A detailed physical examination under anesthesia revealed a slightly bulging septum visible through the introitus vagina. Surgical intervention was postponed and watchful observation was planned. During the 19-month follow-up period, renal function was normal and there was no increase in the size of the cystic mass.

Patient two

A three-and-a-half-year-old girl presented with recur-

rent labial synechia. Abdominal US showed left renal agenesis. Suprapubic pelvic Doppler US and MRI revealed uterus didelphys and an 8 × 5 mm anechoic fluid at the vaginal level. Vaginoscopy revealed a bulging vaginal septum on the left side. Surgical intervention was postponed, and watchful observation was planned. During the 96-month follow-up period, renal function was normal and there was no increase in the size of the cystic mass.

Patient three

A 12-year-old girl presented with a two-month history of pelvic pain. Abdominal US revealed left renal agenesis. Suprapubic pelvic Doppler US revealed a normal uterus and a 64 × 31 mm cyst in the left adnexal region. MRI confirmed left renal agenesis and revealed uterus didelphys with hematocolpos. Physical examination under general anesthesia revealed a bulging septum that was visible through the introitus vagina. Vaginal septum excision, as described in the methods section above, resulted in outflow of old menstrual blood and immediate pain relief after surgery. During the 60-month follow-up period, renal function was normal, and hematocolpos did not recur.

Patient four

A 14-year-old girl presented with a six-month history of pelvic pain. Suprapubic pelvic Doppler US showed a normal uterus and ovaries with a hemorrhagic cyst in the left adnexal region, which was 60 × 35 mm. MRI revealed left renal agenesis, uterus didelphys, and hematometrocolpos (**Figure 2**). A detailed physical examination was performed under general anesthesia; however, no septa were observed. In the endoscopic evaluation, the vaginal septum was observed to be protruding on the left side, and surgical intervention was performed. Pain relief was immediate after surgery. During the 48-month follow-up period, renal function was normal, and hematocolpos did not recur.

Patient five

A 13-year-old girl with a history of left renal agenesis and right-sided VUR was referred to our department. Abdominopelvic CT demonstrated uterus didelphys and hematocolpos within the left uterine cavity, measuring 55 × 60 × 100 mm. A detailed physical examination under general anesthesia revealed a bulging septum visible through the introitus vagina. Vaginoscopy and surgical excision were performed as described in the methods section. During the 25-month follow-up period, renal function was normal and hematocolpos did not recur.

Patient six

A 12-and-a-half-year-old girl with sudden pelvic pain was admitted to the emergency department. Clinical history revealed constipation lasting for the last year and menometrorrhagia in the previous six months. Abdominal US showed an 84 × 54 mm cystic lesion located in the cervix uteri. Abdominopelvic CT revealed right renal agenesis and a 69 × 75 mm right cystic mass lesion located deep in the pelvis extending through the right adnexa, suggesting hematocolpos. MRI confirmed uterus didelphys and hematocolpos on the right side. Vaginoscopy and surgical excision were performed as described in the methods section. Postoperatively, pelvic pain was relieved. During the 13-month follow-up period, renal function was normal and hematocolpos did not recur.

Patient seven

A 12-year-old girl presented with abdominal pain after onset of menarche. Clinical history revealed that she was diagnosed with a right duplicated urinary system with grade three reflux to it and left renal agenesis during early infancy. She was treated and operated on for urinary reflux during childhood. Pelvic US revealed a cystic lesion located in the uterine cervix. MRI confirmed left renal agenesis, uterus didelphys, and an 80 × 75 × 60 mm cystic mass compatible with hydrometrocolpos on the left side. The patient's creatinine level was 1.2 mg/dL at admission, and she was under nephrology follow-up. However, the patient was lost to clinical follow-up before the surgical intervention. During the nephrology follow-up, it was noticed that the patient had been operated on, and there were no clinical complaints related to the Müllerian anomaly.

Demographics of the patients were given in Table 1.

DISCUSSION

It is generally accepted that the uterus, fallopian tubes, cervix, and upper two-thirds of the vagina develop from paired Müllerian ducts. By contrast, the lower third of the vagina develops separately from the urogenital sinus. Recent studies have shown that the vagina develops from the sinovaginal bulbs, which originate from the Wolffian ducts. During embryogenesis, Wolffian ducts (mesonephric channels) give rise to the kidneys. OHVIRA syndrome is considered a part of the spectrum of Müllerian anomalies. Therefore, its pathogenesis can be explained by partial embryologic arrest at the 8th week of gestation, which simultaneously affects the Müllerian paramesonephric and Wolffian mesonephric ducts[8,13–16].

OHVIRA syndrome is usually diagnosed during puberty, shortly after menarche. The typical symptom that leads to the diagnosis is cyclic, increasing lower abdominal pain. The main reason for cyclical pelvic pain and menometrorrhagia is menstrual outflow obstruction caused by obstructed hemivagina. Obstruction usually causes hematometrocolpos and haematosalpinx after the onset of hormonal activity[17]. There are only a few reports of OHVIRA cases presenting with acute abdominal pain, abnormal vaginal discharge, or even acute retention of urine. Lim et al. reported that the percentage of patients presenting with lower abdominal pain was less common in adults than in adolescents[18]. However, it was also stated that this syndrome may go unrecognized during puberty unless suspected due to normal menstrual flow through the unobstructed hemivagina and dysmenorrhea-like symptoms, which are expected at this age[5,8]. On the other hand, OHVIRA syndrome can present as early as in the neonatal period, depending on anatomy and cervical development[12,19,20].

Despite every effort, patients with OHVIRA syndrome may be misdiagnosed as having an acute abdomen and undergoing emergency surgery. Lim et al. reported that two of 18 patients (22.2%) in their series underwent emergency surgery because of suspicion of acute abdomen and ovarian torsion[18]. In the care of adolescents presenting with acute abdomen and associated menstrual irregularities, a strong index of suspicion is necessary to avoid misdiagnosis or unnecessary delays in diagnosis and treatment. In our study, three (60%) of the five

post-pubertal patients presented with lower abdominal or pelvic pain lasting for at least two months (six months in one case). Only one (20%) of the five post-pubertal patients was admitted with acute abdominal pain. One (20%) post-pubertal patient did not complain of pain. These findings emphasize that the clinical presentation of this syndrome is diverse and can be misdiagnosed as an acute abdomen in some cases.

In previous literature, it has been reported that there is a 94.5% to 100% incidence of ipsilateral renal anomaly or agenesis among patients with OHVIRA syndrome[11,14]. In our study, six patients with obstructed hemivagina had ipsilateral renal agenesis as a urinary anomaly with a normal contralateral kidney. One (14%) post-pubertal patient also had a contralateral duplex system and vesicoureteral reflux to this duplicated system. Furthermore, in contrast to some reports stating that right-sided OHVIRA is more prevalent than left-sided[5], 71% of the cases were left-sided in our study. This finding was consistent with that of a previous systematic review that stated that left-sided anomalies were slightly more prevalent in OHVIRA cases[11]. Although we detected an additional renal anomaly other than ipsilateral renal agenesis in only one case in our OHVIRA series, this finding should be alarming to evaluate the contralateral kidney's functions and confirm that this single kidney is intact in OHVIRA syndrome.

Although renal anomaly is part of the triad of this syndrome, OHVIRA syndrome often remains undiagnosed in infancy, even in cases of prenatally diagnosed renal agenesis. Therefore, screening for OHVIRA syndrome in female infants with renal agenesis, detected during the prenatal period, has been advocated[19–22]. Early diagnosis and pre-symptomatic elective surgery may reduce long-term morbidity and prevent possible complications such as endometriosis, infertility, and miscarriage in OHVIRA[10,23,24]. Prenatal or postnatal detection of renal abnormalities, whether accompanied by a pelvic mass, indicates the probability of a genital anomaly in a female infant[10,11,15,25]. As a result, in suspected cases, especially in girls with renal agenesis, performing diagnostic pelvic imaging at the time of renal agenesis diagnosis or planning to perform it at the beginning of puberty is crucial[21,25]. In such cases, postnatal screening enables the diagnosis of obstructive uterovaginal duplication before puberty[25,26]. In our study, one of the post-pubertal patients was diagnosed with left renal agenesis during early infancy, but her OHVIRA diagnosis was delayed until menarche. A pediatric nephrologist referred to one of the pre-pubertal cases and one post-pubertal case, which was diagnosed as renal agenesis, for further investigation of associated Müllerian anomalies. However, the remaining four cases in our study were diagnosed incidentally during the investigation of acute or chronic abdominal/pelvic pain. These findings suggest that a) some female renal agenesis patients are still not investigated for OHVIRA syndrome, and b) teamwork, especially with the help of a pediatric nephrologist, is a crucial part of diagnosis and treatment.

Routine US and MRI are helpful in detecting this anomaly. MRI is the primary modality used for the diagnosis and preoperative planning of OHVIRA syndrome. It evaluates uterine morphology, detects communication between the uterine and vaginal lumen, characterizes fluid content, and diagnoses complications[13]. Although US and MRI findings are sufficient in most cases, laparoscopy is rarely required to clarify diagnosis and treatment, especially in post-pubertal and multiparous patients[1]. In our study group, suprapubic pelvic Doppler US detected a pelvic mass in both patients. A CT scan and MRI provided a definitive diagnosis, and two (33%) of the seven patients were diagnosed with OHVIRA syndrome before puberty.

Several authors have reported the management of OHVIRA syndrome in detail. Vaginal septum excision is offered to all patients in the adolescent or adult age group with OHVIRA to prevent any potential long-term complications and to preserve fertility and sexual functions. In prepubertal OHVIRA cases, patients without symptoms should undergo regular follow-up until the onset of menarche[1,23,24,27]. The treatment of girls younger than ten years is rarely mentioned in the literature[7,18,28]. Maternal estrogen in the serum of newborns causes the accumulation of vaginal secretions or vaginal bleeding for a maximum of six months after delivery. Therefore, problems may arise and require early onset interventions. Conversely, rapid recovery may occur because of a decrease in the effect of maternal estrogen. Consequently, OHVIRA patients with prenatal findings or those diagnosed in the early postnatal period do not always require early surgery[29]. However, surgical intervention may become necessary in younger patients because of complications such as hydronephrosis, hydropyocolpos, abdominal pain, and recurrent urinary tract infection due to the long-term accumulation of vaginal fluid[15]. In our study group, surgical intervention and excision of the vaginal septum was the standard approach for all post-pubertal patients, and asymptomatic pre-pubertal patients were followed up conservatively. The main limitation of this study is the small number of OVHIRA cases. Consequently, the clinical presentations and associated anomalies may differ across large series.

CONCLUSIONS

OHVIRA syndrome is uncommon and can present at any age or in any manner. The management of this rare condition depends on the patient's age and symptoms. It should be suspected in female patients with pre- or postnatally detected unilateral renal agenesis, and in adolescent girls with cyclical pelvic pain and renal agenesis. Teamwork and early diagnosis of this rare disorder and other associated urinary tract anomalies are crucial for planning a proper management program, choosing the correct treatment, and preventing possible long-term complications.

SUMMARY

OHVIRA is a rare Müllerian anomaly often missed in girls with renal agenesis. In our seven cases, most were left-sided. Early imaging and team-based care enable timely septum excision in symptomatic adolescents and conservative follow-up prepubertally.

CONFLICT OF INTEREST

The authors declare no conflicts of interest.

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