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Case Report

Herlyn-Werner-Wunderlich syndrome also known as obstructed hemivagina and ipsilateral renal anomaly: A case report and a comprehensive review of literature ☆

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ABSTRACT

Herlyn-Werner-Wunderlich syndrome, also known as obstructed hemivagina and ipsilateral renal anomaly (OHVIRA), is a Müllerian duct anomaly. It is a rare clinical condition consisting of a duplicated uterus with an oblique vaginal septum that causes partial genital tract outflow obstruction. A urinary tract anomaly, most commonly renal agenesis, is usually present on the obstructed side. The diagnosis of genital tract outflow obstruction is often delayed due to the normal functioning of the unaffected side. The most frequent complications are dysmenorrhea, chronic pelvic pain, infection, infertility and endometriosis. This report describes a 17-year-old G0P0 patient with a history of severe dysmenorrhea and left-sided renal agenesis, who was admitted for complaints of foul vaginal discharge over the past 3 months that was unsuccessfully treated with antibiotics. Transrectal ultrasound revealed the presence of 2 separate hemicavities on transverse and longitudinal views. A cystic lesion with ground-glass opacities was detected between the bladder and a normal-appearing cervix, which was determined to be hematocolpos. The diagnosis of OHVIRA was made. This case highlights the importance of excluding a Müllerian anomaly in the presence of renal system abnormalities. Being aware of the type of anomalies, combinations and variants is crucial to determine the diagnosis and the best surgical approach. Ultrasound was an in-

Abbreviations: AFS, American Fertility Society; ARSM, American Society for Reproductive Medicine; ESHRE–ESGE, European Society of Human Reproduction and Embryology–European Society for Gynecological Endoscopy; HWW, Herlyn-Werner-Wunderlich syndrome; OHVIRA, Obstructed hemivagina and ipsilateral renal anomaly.

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valuable imaging exam to determine the type of anomaly and its complexity. Awareness of this syndrome and its variants will prevent misdiagnosis and will help to define the appropriate treatment for these patients.

Introduction

Anomalies of the Müllerian system represent one of the most perplexing disorders in the field of gynecology. Herlyn-Werner-Wunderlich syndrome consists of a duplicated uterus with a vaginal oblique septum that causes partial genital tract outflow obstruction, frequently associated with renal agenesis [1–4]. Due to the Müllerian and renal system heterogeneity reported throughout the literature, the acronym OHVIRA (obstructed hemivagina and ipsilateral renal anomaly) was coined [5]. The following case report discusses the use of ultrasound in the diagnosis and surgical treatment of a case of Herlyn-Werner-Wunderlich syndrome.

Case report

A 17-year-old G0P0 woman, was admitted to our Gynecology Department in a clinical setting of foul discharge. Over the past 3 months, the patient had experienced a foul discharge and was treated with several cycles of antibiotics without success. The patient's medical history revealed left-sided renal agenesis. She was not sexually active yet, and menarche had occurred at 11 years of age. She had regular menstrual cycles with progressive and severe dysmenorrhea since menarche. Pelvic examination was limited and revealed normal external genitalia; since the patient was *virgo intacta*, the cervix and vagina were not appropriately assessed. However, a bulge on the left side of the vaginal wall and purulent secretions were observed. Rectal examination revealed the presence of a 5-6 cm, soft, painful mass on the left side of the vagina. Transrectal ultrasound was performed, and when scanning the uterine body, 2 separate hemicavities on transverse and longitudinal views were observed (Fig. 1A). A cystic lesion with ground-glass opacities measuring 59 mm × 39 mm × 57 mm was located between the bladder and a normal-appearing cervix (Figs. 1B–E). On probe pressure, the cystic lesion was painful and soft, giving the impression of an apparent pelvic abscess. However, the combination of the clinical setting and ultrasound imaging, pointed to a diagnosis of an obstructed hemivagina with ipsilateral renal anomaly. We proposed vaginoplasty and hysteroscopy, which were accepted by the patient and her mother. The procedure was carried out with a hysteroscope, virgin speculum, colposcope and bipolar electrocautery. Through vaginoscopy, bulging of the left vaginal wall was noted and located posteriorly, and the cervix was normal. This normal cervix gave access to a uterine cavity that revealed the presence of only 1 ostium and no communication with the other cavity. After completion of the hysteroscopy, ultrasound-guided surgery was performed. Through transrectal ultrasound, the most bulging point of the obstructed hemivagina was located midway between the bladder and rectum (Fig. 1F). A 5 mm incision was made, and purulent content was drained. Through the small incision, a Foley catheter was introduced and inflated into the hemivagina (Fig. 1G), and traction was applied. This allowed better access to the septum and created a good safety margin for the bladder and rectum. Under colposcopic visualization with bipolar electrocautery, a circular septectomy with ~3 cm in diameter was created around the Foley catheter (Fig. 1H). A left vaginoscopy was performed via the newly created connection. An inflamed cervix was observed and the cervix also gave access to a uterine cavity with only 1 ostium. No communication was found at the level of the cervices or uterine bodies. One month later, the septectomy was still patent, and a normal left cervix could be partly visualized. For the first time, the patient had completely painless menstruation.

Results and discussion

The first reported case of a duplicated uterus with obstructed hemivagina dates back to 1922, describing a sequential unilateral obstruction with hematocolpos, hematometra and hematosalpinx. However, no renal anomaly was mentioned [1]. In the 1970s, similar reports began to emerge. In 1971, Herlyn and Werner reported a case of a duplicated uterus and renal aplasia [2]. One year later, Amon et al. [3] described a case of a bicornuate uterus with a double cervix and a double vagina associated with right hematocolpos, hematometra, hematosalpinx, partial atresia of the vagina and right renal aplasia. In 1976, Wunderlich also described a bicornuate uterus with a simple vagina and isolated right hematocervix without connection of the right uterus to the vagina in association with aplasia of the right kidney and ureter [4]. In light of these findings, Herlyn-Werner-Wunderlich syndrome (HWW) was described. This syndrome classically consists of a triad that includes a congenitally duplicated uterus, an oblique septum that generates a blind hemivagina and obstructs the menses outflow of 1 uterine cavity and an associated ipsilateral renal anomaly (Fig. 2). However, the literature consistently reveals some heterogeneity in terms of anatomic Müllerian variants associated with this syndrome, which is likely the main reason why Smith and Laufer renamed the syndrome, only taking into consideration hemivagina obstruction and ipsilateral renal anomaly and suggesting the acronym OHVIRA [5].

The uterus, cervix, fallopian tubes and upper third of the vagina develop from the Müllerian ducts (also known as paramesonephric ducts) [6]. Müllerian congenital anomalies include developmental defects (agenesis/hypoplasia) and

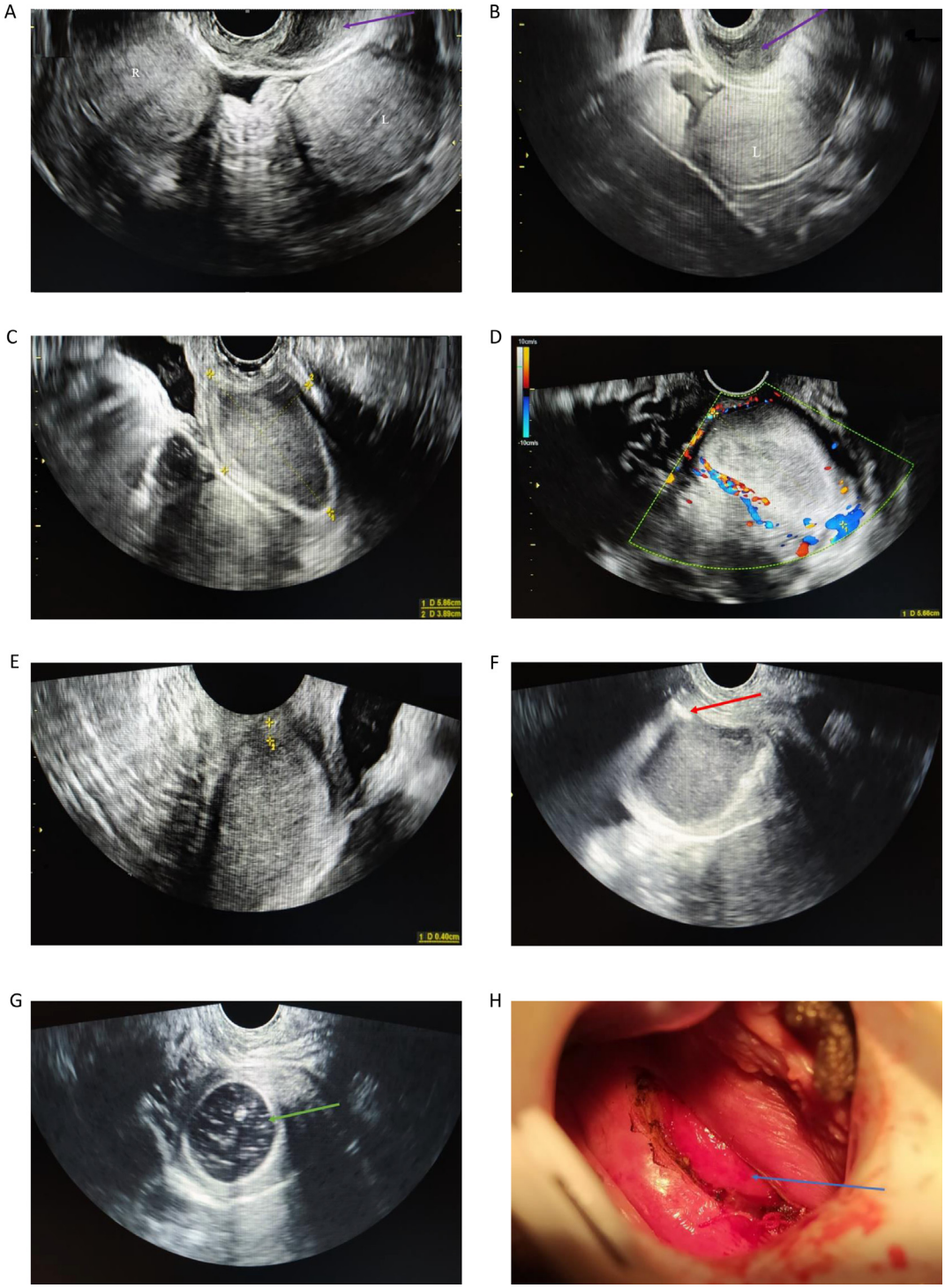

Fig. 1 – (A) Transverse view with 2 separate uterine bodies (R – right uterine body, L – left uterine body) and a cystic lesion suspected to be hematocolpos/pyocolpos on a left obstructed hemivagina (purple arrow); (B) Longitudinal view of the left hemiuterus (L – left uterine body) and cystic lesion suspected to be hematocolpos/pyocolpos of a left obstructed hemivagina (purple arrow); (C) Cystic lesion 59 x 39 x 57 mm with ground-glass content posterior to the bladder and anterior to the uterine body; (D) Cystic lesion with circumferential vascularization pattern and Doppler color score of 2; (E) Oblique septum thickness of 4 mm; (F) Transrectal ultrasound was used to locate the most bulging point of the obstructed hemivagina (red arrow), on a mid-distance between the bladder and rectum; (G) A 5 mm incision was made on the most bulging point of the obstructed hemivagina and a Foley catheter (green arrow) was introduced and inflated into the hemivagina; (H) Traction was applied on the Foley catheter and under colposcopy visualization, with a bipolar electrocautery, a circular septectomy with ~3 cm diameter was created around the Foley catheter. (Through the septectomy it is possible to partially visualize the left cervix [blue arrow]).

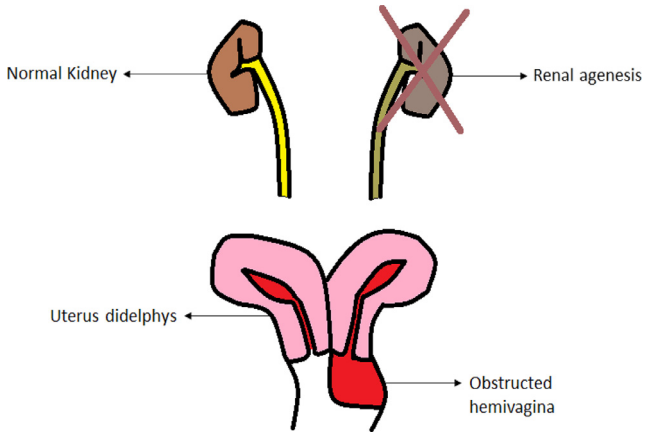

Fig. 2 – Classical Herlyn-Werner-Wunderlich syndrome.

Table 1 – Major symptom/sign[a] (Group A).

Abdominal/pelvic pain	142 (40,7%)
Cyclic pelvic pain	128 (36,7%)
Foul smelling vagina discharge	23 (66,0%)
Abnormal vaginal bleeding	8 (2,3%)
Infertility	8 (2,3%)
Urinary retention	7 (2,0%)
Other symptoms	33 (9,5%)

[a] 113 cases not specified

Table 2 – Major symptom/sign (Group B).

Dysplastic/Multicystic kidney	29 (49,2%)
Renal agenesis	23 (39,0%)
Pelvic/abdominal cystic mass	3 (5,1%)
Renal agenesis + Cystic mass	2 (3,4%)
Dysplastic/Multicystic kidney + Cystic mass	1 (1,7%)
Pelvic kidney suspicion	1 (1,7%)

Table 3 – Types of reported Mullerian anomalies according to the ARSM classification.

Uterus Didelphys	462 (88,7%)
Bicornuate Uterus	34 (6,5%)
Septate Uterus	25 (4,8%)

fusion defects (bicornuate uterus, didelphys uterus, septate uterus, transverse septum). Duplicated uterus is attributed to a failure in the fusion process of the Müllerian ducts at 9 weeks of gestation [7]. In a nonselected population, the prevalence of congenital uterine anomalies is approximately 5.5% [6]. Amongst the obstructed outflow forms that do not exhibit primary amenorrhea (ie, severe dysmenorrhea in the presence of menstrual cycles), HWW syndrome is the most common form [8].

A comprehensive literature search of PubMed utilized the terms Herlyn-Werner-Wunderlich syndrome" or "obstructed hemivagina and ipsilateral renal anomaly" to review case reports and case series published prior to December 2020. The original articles were selected based on features such as age, major symptom, obstructed side, ipsilateral/contralateral renal anomaly, Müllerian anomaly, types of diagnostic imaging, obstruction mechanism and treatment. Data extraction compiled 521 into a descriptive analysis [12–14,19–153].

Data analysis was conducted on 521 cases reported in case reports and case series, which were organized into 2 groups. Group A (n=462) included cases diagnosed after birth, and Group B (n=59) included cases whose diagnostic investigation started prenatally.

Regarding age, Group A had a mean age at diagnosis of 16.64 years (range 0.5-65 years) when excluding case series that reported age with mean or median values.

In this group, the major symptoms/signs that prompted the diagnostic work-up where available, were abdominal/pelvic pain and cyclic pelvic pain (Table 1).

In Group B, as the syndrome was suspected prenatally, the main ultrasound features that prompted investigation were dysplastic/multicystic kidney (49.2%) and renal agenesis (39.0%). On rare occasions, the features reported were pelvic/abdominal cystic mass combined with renal agenesis or isolated pelvic/abdominal cystic mass (Table 2).

There are multiple classification systems for Müllerian anomalies but there is no consensus on which system is more appropriate. The American Society for Reproductive Medicine (ARSM; formerly known as the American Fertility Society [AFS]) published a classification system in 1988, which has endured over time and is the most used system in the included articles. For that reason, this review uses the ARSM terms and definitions. A more recent classification system was published by the European Society of Human Reproduction and Embryology–European Society for Gynecological Endoscopy (ESHRE–ESGE 2013) for female genital tract anomalies, which classifies anomalies regarding the characteristics of the uterine body, cervix and vagina. When comparing both classification systems, uterus didelphys from the ARSM corresponds to bicorporeal uterus subtype b (U3b). In fact, when using the ESHRE-ESGE system, classic OHVIRA is classified as U3bC2V2. It is important to note that since 2021, ARSM adopted a more holistic classification system, taking into account the uterine body, cervix and vagina, similar to the ESHRE-ESGE system.

Regarding the type of uterine anomaly, uterus didelphys was the most commonly reported (88.7%), followed by bicornuate uterus and septate uterus (Table 3). A diagram depicts all variants reported by type of Müllerian anomaly (Figs. 3–5). However, the authors would like to highlight some limitations of the Müllerian anomaly classification. Historically, MRI was considered the gold-standard diagnostic imaging technique for assessing a Müllerian anomaly. Three-dimensional (3D) ultrasound imaging has improved and allows a very accurate characterization of Müllerian anomalies and often dismisses the need for further imaging techniques. The combination of an imaging technique with hysteroscopic and/or surgical findings, further increases the chance of correct classification of the anomaly. When reviewing all reported cases, we found that the uterine anomalies were classified by imaging techniques, such as ultrasound, CT scan or MRI, and

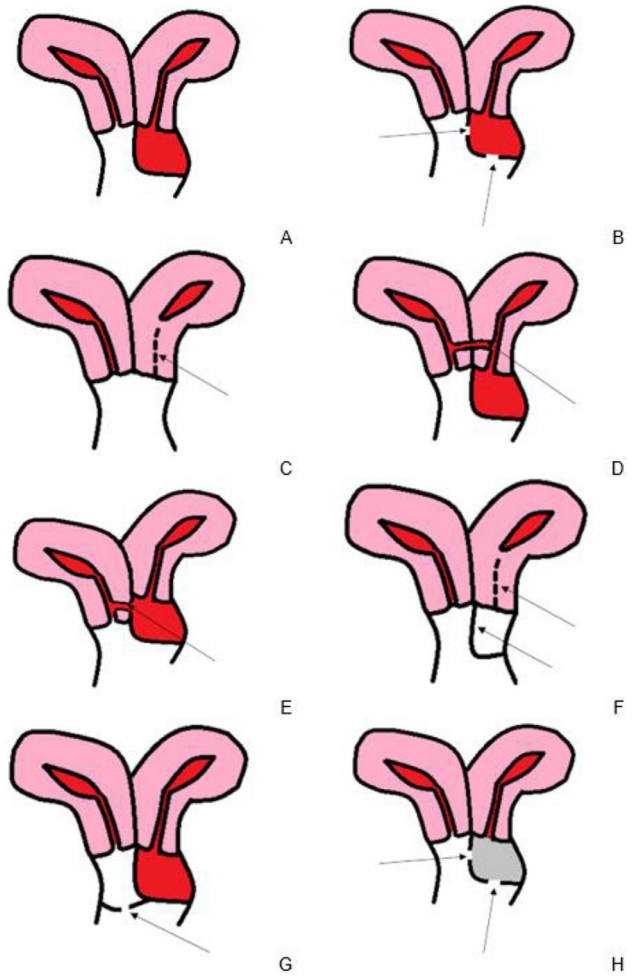

Fig. 3 – Reported variants in uterus didelphys. (A) Classical type with longitudinal obstructing vaginal septum (U3bC2V2); (B) Longitudinal non-obstructing vaginal septum (U3bC2V1); (C) Cervical aplasia (U3bC3V0); (D) Cervical communication (U3bC2V2 + Cervical communication) Fistula between obstructed hemivagina and nonobstructed cervix (U3bC2V2 + cervico-vaginal communication); (F) Cervical aplasia associated to a septum (U3bC3V2); (G) Classical type combined with fenestrated transverse septum (U3bC2V3 (nonobstructing septum)); (H) Septum microperfuration due to infection/distension (originally, U3bC2V2).

by endoscopic/surgical findings. Sometimes, authors changed the imagiological classification because of surgical findings. This indicates that Müllerian anomalies classified by imaging techniques may not be precise, which suggest that using congenital anomaly classification as an outcome measure is questionable due to the lack of standardization of definitions and classifications.

Due to the absence of ultrasound imaging features, it is not possible to attribute the septate/bicornuate uterus of ARSM (1988) a correct ESHRE-ESGE classification. All U3b (uterus didelphys) variants are reported in Table 4 and a schematic illustration is depicted in Fig. 3.

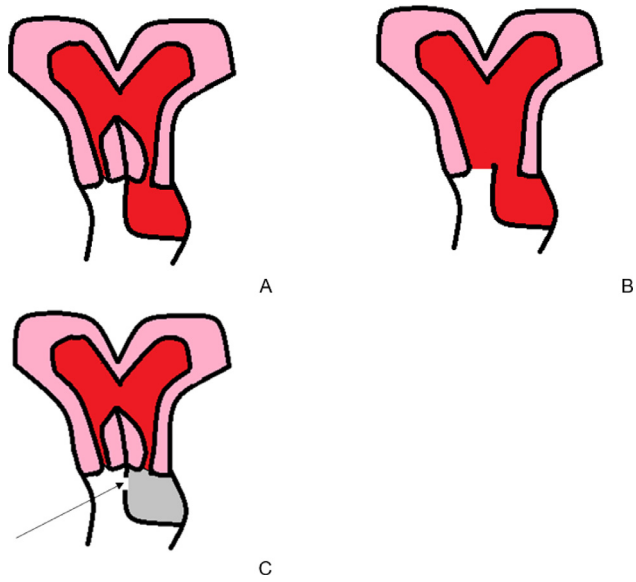

Fig. 4 – Reported variants in bicornuate uterus. (A) Bicornuate uterus bicollis with longitudinal obstructing vaginal septum (B) Bicornuate uterus unicollis with longitudinal obstructing vaginal septum; (C) Bicornuate uterus bicollis with septum microperfuration due to infection/distension.

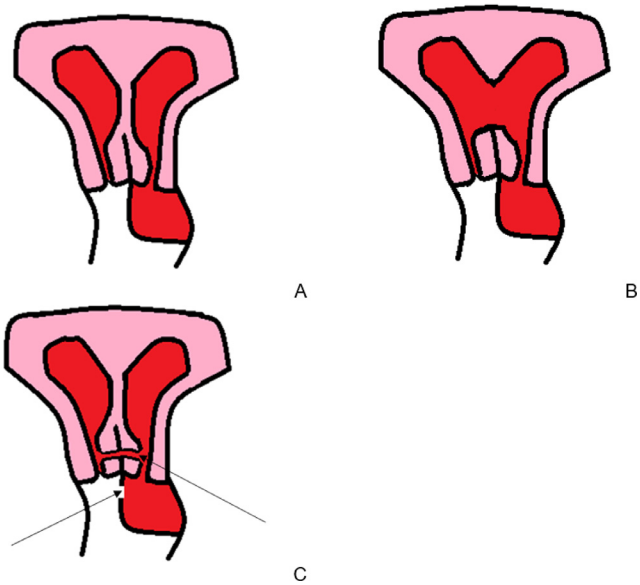

Fig. 5 – Reported variants in septate uterus. (A) Complete septate uterus with longitudinal obstructing vaginal septum; (B) Partial septate uterus with longitudinal obstructing vaginal septum; (C) Complete uterus septate with cervical communication and longitudinal nonobstructing vaginal septum.

Table 4 – Types of reported Mullerian anomalies according to the ESHRE-ESGE classification.

U3bC2V2[a]	399
U3bC2V1	40
U3bC3V0	12
U3bC2V2 + Cervical communication	7
U3bC2V2 + Cervical-vaginal communication	1
U3bC3V2	2
U3bC2V3 (nonobstructing septum)	1
Total	462[b]

[a] 22 cases in which microperfuration has occurred due to infection/distension in a previous obstructing septum were regarded as longitudinal obstructing vaginal septum (V2)

[b] Septate uterus [25] and Bicornuate uterus [34] were not included due to the lack of information regarding ultrasound features, which does not allow a feasible ESHRE-ESGE classification.

Table 5 – Types of anatomical obstruction.

Longitudinal obstructing/nonobstructing vaginal septum	506 (97.1%)
Cervical aplasia	12 (2.3%)
Cervical aplasia + Longitudinal obstructing vaginal septum	2 (0.3%)
Longitudinal obstructing vaginal septum (on the affected side) + Transverse nonobstructing vaginal septum (on the usually normal side)	1 (0.1%)
Total	521

Table 6 – Anatomical location of incomplete obstructions.

Longitudinal nonobstructing vaginal septum	44 (75,8%)
Cervical communication	9 (15,5%)
Cervical-vaginal communication	1 (1,7%)
Septum communication + Cervical communication	1 (1,7%)
Uterine cavities communication	3 (5,2%)
Total	58

Table 7 – Associated ipsilateral renal anomalies.

Agenesis		461 (88.5%)
	No specifications	454/461
	Agenesis with ectopic ureter insertion to the vagina	7/461
Dysplasia		51 (9.8%)
	No specifications	9/51
	Not specified ectopic/orthotopic kidney position with ectopic ureter insertion to the vagina	30/51
	Ectopic kidney position	4/51
	Ectopic kidney position with ectopic ureter insertion to the vagina	7/51
	Orthotopic kidney position with ectopic ureter insertion to the vagina	1/51
Hypoplasia		3 (0.6%)
	Ectopic	1/3
	Ectopic kidney position with ectopic ureter insertion to the vagina	2/3
Dysplasia and Hypoplasia[a]		3 (0.6%)
	No specifications	2/3
	Ectopic	1/3
Dupplex collecting system		1 (0.2%)
No anomaly, but contralateral renal anomaly (Fig. 7)		1 (0.2%)

[a] These 6 cases described as “hypoplastic” probably belong to the “dysplastic” group. Literature often mixes both terms due to similar/overlapping definitions.

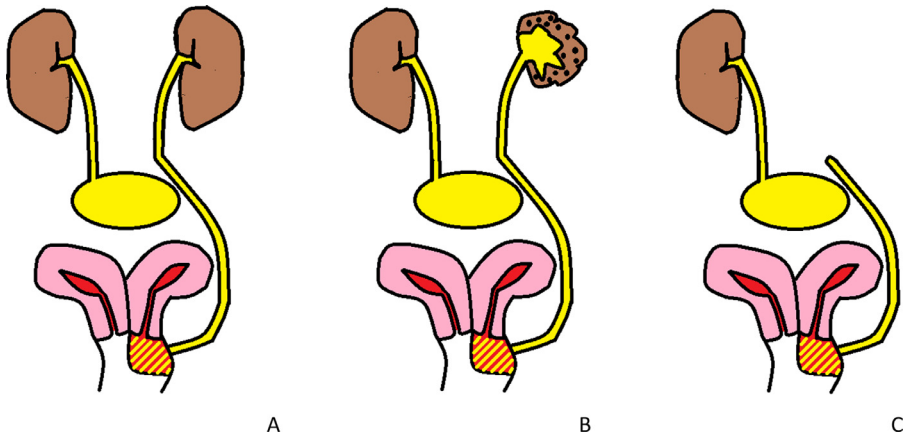

Fig. 6 – Herlyn-Werner-Wunderlich Syndrome spectrum (A) Ectopic ureter insertion in the obstructed hemivagina; (B) Dysplastic process secondary to the ectopic ureter insertion in the obstructed hemivagina; (C) Renal involution generating agenesis with ectopic ureter insertion in the obstructed hemivagina.

One classification system suggests that the syndrome should be classified as type 1 or type 2 according to the obstruction mechanism (complete or incomplete) of the hemivagina. Type 1 represents complete obstruction, including type 1.1 (classic HWWS) and type 1.2 (cervical atresia). Type 2 represents incomplete obstruction, including type 2.1 with incomplete septum and type 2.2 with outflow through a connection to the contralateral cervix [156]. However, this classification does not include bicornuate and septate uteri. For obvious anatomic reasons, it is impossible to extrapolate this classification to these 2 Müllerian anomalies (Figs. 4 and 5).

Regarding the type of anatomical obstruction, vaginal septum comprised 97.1% of all cases (Table 5), and a schematic illustration is depicted in Fig. 2–4.

Although the majority of OHVIRA diagnoses occur at an early reproductive age, usually a few months after menarche due to severe dysmenorrhea and pelvic pain, some patients have a later diagnosis. This subgroup of patients can be partially explained by incomplete outflow obstruction, even in the presence of a longitudinal obstructing vaginal septum. In this review, incomplete outflow tract obstruction was reported in 10.9% of all cases (58/521). The most common mechanisms were a longitudinal nonobstructing vaginal septum (75.8%) and cervical communication (15.5%), which allows menstrual blood flow between the 2 hemiuteruses. Rarer mechanisms include a combination of fenestrated septum associated with cervical communication, fistula formation and uterine cavity communication (as in partial septate uterus or partial bicornuate uterus) (Table 6).

From a nephro-urological perspective, case series report a 63% rate of unilateral renal agenesis associated with uterus didelphys, which increase to 90% when associated with obstructed hemivagina [9]. As described, renal agenesis has been the classical form of urological pathology associated with the syndrome and has always been defined by radiological methods [10]. However, over time, some other forms have been reported, such as dysplastic, hypoplastic, multicystic, polycystic, atrophic and ectopic kidney [5,11]. The data in this review allowed the classification of 521 cases. Renal agenesis was by far the most common anomaly, with a rate of 88.5% (Table 7), but the authors believe that this rate is overestimated. There are multiple reported cases of dysplastic kidneys with ectopic ureter insertion in the blind hemivagina, mainly in case series that are focused on the prenatal, neonatal and prepubertal periods [10,12,13]. In fact, there are some reported cases of dysplastic kidneys with ectopic ureters inserted into the hemivagina (with some residual function) that were not detected on imaging exams, including magnetic resonance [31], in which a continuous urine leak was triggered after vaginal septum resection [17,18]. This has led to questions the true physiopathology of Herlyn-Werner-Wunderlich syndrome. Several authors have suggested that agenesis is the final stage of a spectrum, which results from a process of dysplasia secondary to the insertion of the ureter into a blind hemivagina, generating a major obstruction that compromises the normal development of the kidney. Invariably, this leads to the involution of the nephro-urological system (Fig. 6) [10,13,14]. Right-sided vaginal obstruction and renal anomalies have been reported with higher frequency since the earliest reported cases [15] and a systematic review reported a 65% rate of right-sided anomalies [16]. In fact, of 512 reported anomalies (311 right-sided, 201 left-sided, 9 cases not included [8 not specified and 1 with contralateral renal anomaly]), we have verified a slightly higher rate for right-sided anomalies, supporting the literature (60.7% vs 39.3%). Of all analyzed cases, a very unusual case reported a renal anomaly associated with contralateral obstructed hemivagina (Fig. 7) [16].

Symptoms usually start soon after menarche with severe dysmenorrhea due to partial outflow obstruction and, subsequently, a pelvic mass is found on clinical and/or imaging assessment. However, this syndrome may remain unrecognized for several years. One of the most likely causes is the existence of communication between the 2 hemivaginas [97]. In fact, in this review, when considering the presence of a communication, the median age at diagnosis was 19 years of age vs. a median of 14 years of age when completely obstructed. In addition, patient tolerance to pain and the use of painkillers

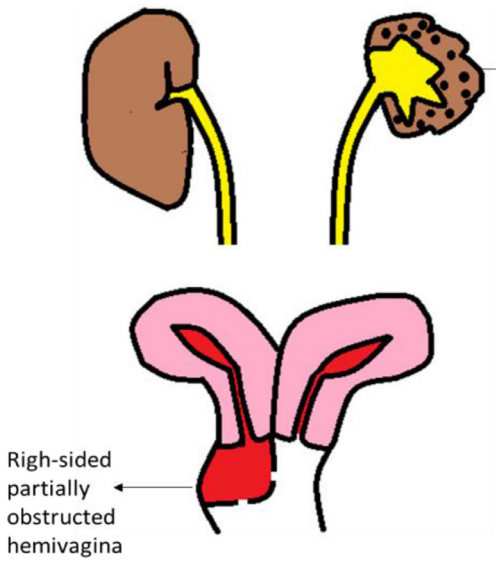

Fig. 7 – Obstructed hemivagina with incomplete septum and contralateral renal anomaly.

may also may delay the diagnosis. The range of volumes from small volumes, such as 300-500 ml [75,85,86,88,96,110] to high volumes as 1000-2000 mL [54,111], mean that the distensibility degree of the vagina/uterus and the threshold to trigger pain are different for each person. On the other hand, with the advent of better ultrasound quality and imaging, prenatal diagnosis has been progressively added as a new presentation of renal agenesis or multicystic kidney, as an indirect sign of a Müllerian anomaly.

Even with severe dysmenorrhea, the presence of menstrual outflow delays the suspicion of an outflow obstruction. Menarche and physiological changes are easily evoked as justification for the symptoms. Introduction of a birth control pill relieves the menstrual symptoms, which further delays the true diagnosis. Due to the young age of the patients, most of the time, a full gynecological exam is not performed, which also interferes with a diagnosis. Therefore, a high suspicion index is needed to obtain the diagnosis.

Ultrasound imaging is an optimal screening exam for this condition and should be considered in cases of severe dysmenorrhea. Of the primary imaging diagnostic tools, ultrasound and magnetic resonance imaging (MRI) are the mainstays [53,154]. In this review, in 44.4% of cases, a combination of ultrasound and MRI was used as the imaging work-up. On the other hand, work-up was performed with ultrasound alone in 27.7% of cases. Other imaging exams, such as computed tomography scans [102,135,153], 3D-MRI [134], 3D-CT scans [78] and colpography [105] have been reported.

A single-stage vaginoplasty with drainage of the obstructed side and resection of the septum is the gold standard treatment [5,81,139]. There are several reported surgical approaches, including, laparotomic, laparoscopic [125] and transvaginal approaches, which allow relief of the obstruction and the pain associated with the distension generated by retained menses. The transvaginal approach is the most commonly used and different techniques are reported, such as sharp dissection [22], hysteroscopy with monopolar electrocautery [146], hysteroscopy with bipolar electrocautery [40], resectoscopy with bipolar electrocautery [46], bipolar electrocautery [102] and CO_2 laser [136]. However, septum resection may not be possible when the vaginal septum is in a proximal position, and a hemihysterectomy may be necessary [139]. In fact, the greater the distance between the septum and perineum, the more likely a hemihysterectomy will be necessary [139]. However, due to the known variants, vaginoplasty may not be the best approach. The associated nephroureteric issues and complications related to long-term retrograde menstruation may need a different approach. A 3O (obstruction, ureteric orifice, and outcome) subclassification system associated with OHVIRA has been proposed to guide surgical management. This system considers the mechanism of obstruction, the presence/absence of ectopic ureter insertion and the possible long-term complications of the syndrome. In addition to vaginoplasty, other strategies, such as hemihysterectomy, cervicoplasty, ureteronephrectomy, salpingectomy and ovarian cystectomy, may be needed depending on the clinical scenario [155].

Conclusion

Herlyn-Werner-Wunderlich syndrome is a rare clinical condition that is traditionally defined by a triad of uterus didelphys, OHVIRA. Due to the complex Müllerian variants that have been reported, more recent literature has used the term OHVIRA, only taking into consideration the outflow obstruction and ipsilateral renal anomaly.

Classically, the main symptom of the syndrome is represented by pelvic cyclic pain that starts shortly after menarche. The normal outflow of the unobstructed side and the assumption that dysmenorrhea is the most likely cause at this age group delays the suspicion of an outflow tract obstruction and the true diagnosis. A new diagnostic trend has emerged in the literature. With the development of prenatal diagnosis, renal anomalies have suggested this diagnosis before birth, particularly the presence of a dysplastic kidney. In fact, prenatally diagnosed cases have supported the theory that there is no true agenesis. It is thought that renal agenesis is generated by ectopic ureter insertion into the obstructed hemivagina leading to dysplasia, atrophy, involution and agenesis.

Performing vaginoplasty with excision of the obstructing septum is the treatment mainstay. Timely surgical intervention prevents complications such as endometriosis, hemato/pyosalpynx, hemato/pyocolpos, hemato/pyometra and pelvic adhesions. A Müllerian anomaly alone can greatly impact the chance of conception. The clinical scenario of a Müllerian anomaly with complications associated with outflow obstruction has an even greater impact on future obstetric outcomes. This is enough reason to be aware of this syndrome and to determine the appropriate treatment. Depending on the complexity of the Müllerian anomaly and associated complications, a simple septum excision may not be sufficient.

In conclusion, in the presence of a urologic anomaly in a female patient, Müllerian anomalies should be excluded. Aware-

ness of the type of anomalies, combinations and variants is crucial to determine the diagnosis and the best surgical approach. Ultrasound is an invaluable imaging exam to determine the type of anomaly and its complexity. Awareness of this syndrome and its variants will prevent misdiagnosis and will help to define the appropriate treatment for these patients.

Author contributions

All authors participated in the medical care offered to the patient; Costa, C., Pereira H. and Borges, AL. performed the surgery; Borges AL., Costa, C. and Martins, A. conceptualized the case report; Borges AL. and Costa, C. collected data and wrote the manuscript draft; Martins, A., Sanha, N. and Pereira H. reviewed and edited the manuscript; all authors have approved the final manuscript.

Patient consent

Informed written consent was obtained by the patient's mother.

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