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Herlyn–Werner–Wunderlich syndrome: pre- and post-surgical MRI and US findings

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Abstract

Herlyn–Werner–Wunderlich syndrome (HWWS) is a rare congenital anomaly of the female urogenital tract that associates Müllerian duct anomalies with mesonephric duct anomalies. The triad of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis characterizes this syndrome. Patients generally present with non-specific symptoms after menarche. Pelvic pain, dysmenorrhea, and palpable mass due to hematocolpos or hematometra are the most common findings. Pyohematocolpos and pyosalpinx may appear as acute complications, while endometriosis and pelvic adhesions constitute potential long-term complications. When a prenatal diagnosis of unilateral renal agenesis in newborn girls is known, a gynecological imaging study should be performed to exclude uterine and vaginal abnormalities. These patients should be followed up to ensure that a timely surgical correction is performed. The diagnosis of HWWS is difficult due to the lack of specific symptoms or findings upon physical examination. An accurate imaging description of these congenital anomalies is crucial to guide patients toward surgical treatment, relieving acute complications, and preserving the normal fertility. The authors provide a pictorial review of the magnetic resonance imaging and ultrasonography findings of the HWWS with correlation to embryological, clinical, and surgical features.

Key words: Herlyn–Werner–Wunderlich syndrome—Müllerian duct anomalies—Mesonephric duct anomalies—Uterus didelphys—Hematocolpos—Renal agenesis

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Herlyn–Werner–Wunderlich syndrome (HWWS) is a rare congenital anomaly of the female urogenital tract that associates Müllerian (paramesonephric) and Wolffian (mesonephric) duct anomalies. It is characterized by the triad of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis [1, 2].

This syndrome was first reported in 1922 by Purslow [3], but its current denomination derives from two posterior reports. In 1971, Herlyn and Werner [4] reported the combination of an open Gartner duct cyst, homolateral renal aplasia, and double uterus (Herlyn–Werner syndrome). In 1976, Wunderlich [5] reported a rare form of bicornuate uterus with simple vagina and isolated hematocervix, in association with aplasia of the right kidney and ureter. The HWWS is also referred in the literature as the obstructed hemivagina and ipsilateral renal anomaly syndrome (OHVIRA) [6, 7].

The overall estimated occurrence of the HWWS is 0.1%–3.8% [8]. Its etiology, embryological basis, and pathogenesis are still under discussion and most peer-reviewed manuscripts regarding the HWWS are case reports and case series [2, 9]. This paper aims to provide a comprehensive review of the magnetic resonance imaging (MRI) and ultrasonography (US) findings of the HWWS with correlation to embryological, clinical and, surgical features.

Embryology

Müllerian duct anomalies (MDA) are congenital disorders that result from non-development (agenesis or hypoplasia), defective vertical or lateral fusion, or resorption failure of the Müllerian or paramesonephric ducts [2]. By the eighth week of gestation, the Müllerian ducts migrate to the midline and fuse to form the uterus, cervix, and upper vagina. While the cranial end of the fused ducts yields the future uterus and cervix, the caudal end contacts the posterior wall of the urogenital sinus to

form the Müllerian tubercle. This small bulge will form the upper two-thirds of the vagina. The cranial ends of the paramesonephric ducts remain unfused and open into the coelomic cavity as funnel-shaped structures to form the abdominal ostia of the fallopian tubes (Fig. 1) [10, 11].

The role of mesonephric ducts should also be highlighted. These are not only precursors and inducers of the female reproductive tract development but also act with the Müllerian tubercle to form part of the vagina [11]. Moreover, mesonephric ducts play an essential role in renal development (Fig. 1), explaining why renal tract anomalies are associated with MDA in approximately 30% of cases [2, 11–13].

Uterus didelphys accounts for near 5% of MDA and corresponds to a complete duplication of the uterus and the cervix. In 75% of the cases, it is associated with a longitudinal vaginal septum, either complete or incomplete (with fenestrations) [9]. A transverse vaginal septum resulting from defects in vertical fusion may be associated [9, 13]. According to the American Fertility Society (AFS) classification, uterus didelphys belongs to the Class III, which includes malformations arising from complete absence of fusion of the two Müllerian ducts [9, 12–14].

According to the most recent classification provided by the European Society of Human Reproduction and Embryology (ESHRE) and the European Society for Gynecological Endoscopy (ESGE) [15], the congenital anomalies of the HWWS are classified as U3c (bicorporal uterus, bicorporal septate), C2 (double "normal" cervix), and V2 (longitudinal obstructing vaginal septum).

In a recent paper, Wang et al. [16] reviewed a Chinese classification of the HWWS based on morphological features of the vaginal septum. This classification firstly reported by Bian et al. in a Chinese-written manuscript, classifying the vaginal septum into three types: type I, with an imperforate vaginal septum; type II, with a perforate vaginal septum; and type III, with an imperforate vaginal septum and a cervical fistula. This classification is not widely used in western countries.

Some variants of the HWWS have been already reported. Dorais et al. [17] described the case of a 14-year-old girl with a non-communicating uterine horn, ipsilateral renal agenesis, and Gartner duct cyst (which comes from the Gartner duct, a remnant of the mesonephric ducts) (Fig. 1) [10]. Nabeshima et al. [18] reported another variant in a 12-year-old girl with an obstructed non-communicating uterus didelphys without vaginal septum or Gartner duct cyst.

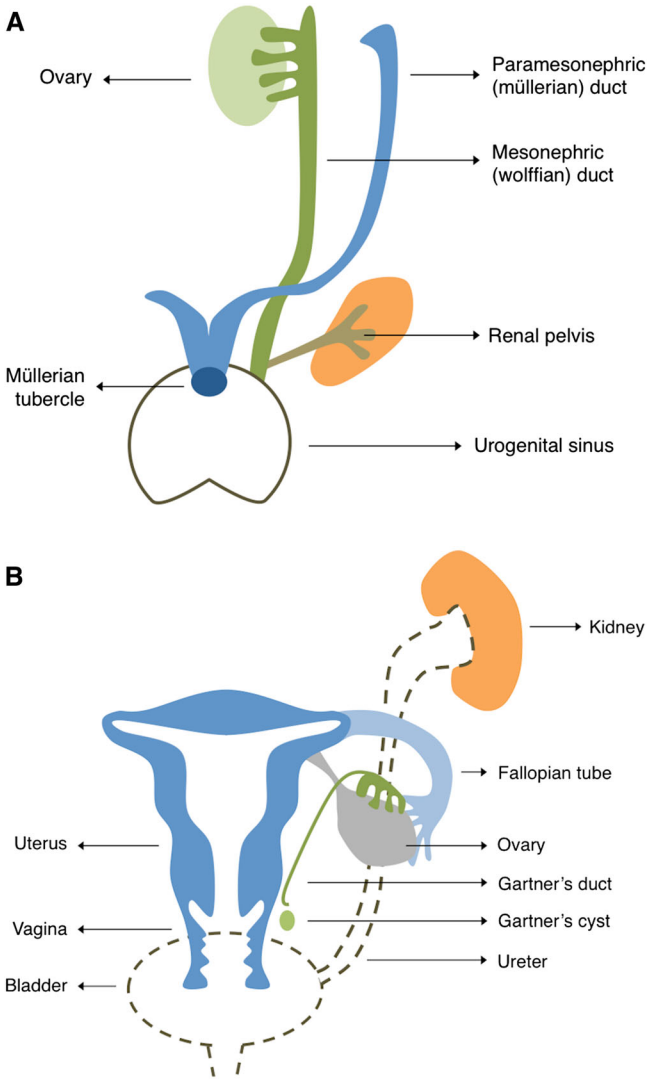


Fig. 1. Embryological development of the female genital tract. Genital ducts at approximately 6 weeks (**A**) and mature female genital duct system (**B**).

Clinical presentation

Most patients present with non-specific postpubertal symptoms, generally from 2 to 12 months after menarche [19]. In a study including 70 patients with HWWS, Tong et al. [1] reported a mean age at onset of symptoms of 17 years, ranging from 10 to 44. In the same manuscript, the authors concluded that the mean age of diagnosis is significantly different between patients with complete (13 years) and incomplete obstruction (25 years).

Pelvic pain, dysmenorrhea, and palpable mass due to hematocolpos or hematometra are the most common findings [2, 20]. Primary amenorrhea, dyspareunia, urinary retention, spontaneous rupture of the hematocolpos, infertility, and obstetric complications are less common manifestations [13, 21, 22]. Patients may also complain of mucopurulent discharge in the presence of vaginal communications, as well as intermenstrual bleeding when uterine cavities communicate [23]. In rare cases, a large hematocolpos results in a huge paravaginal

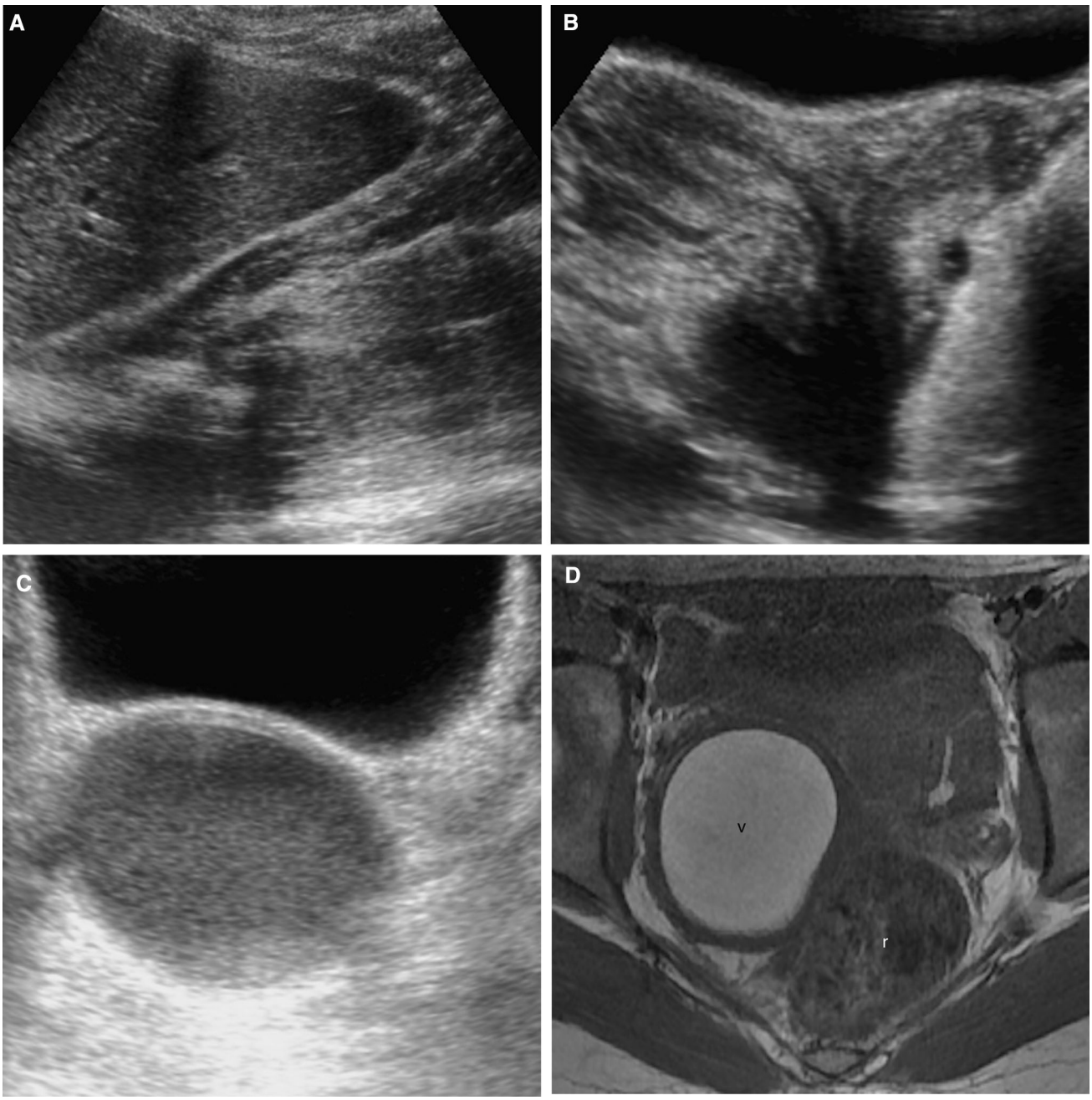
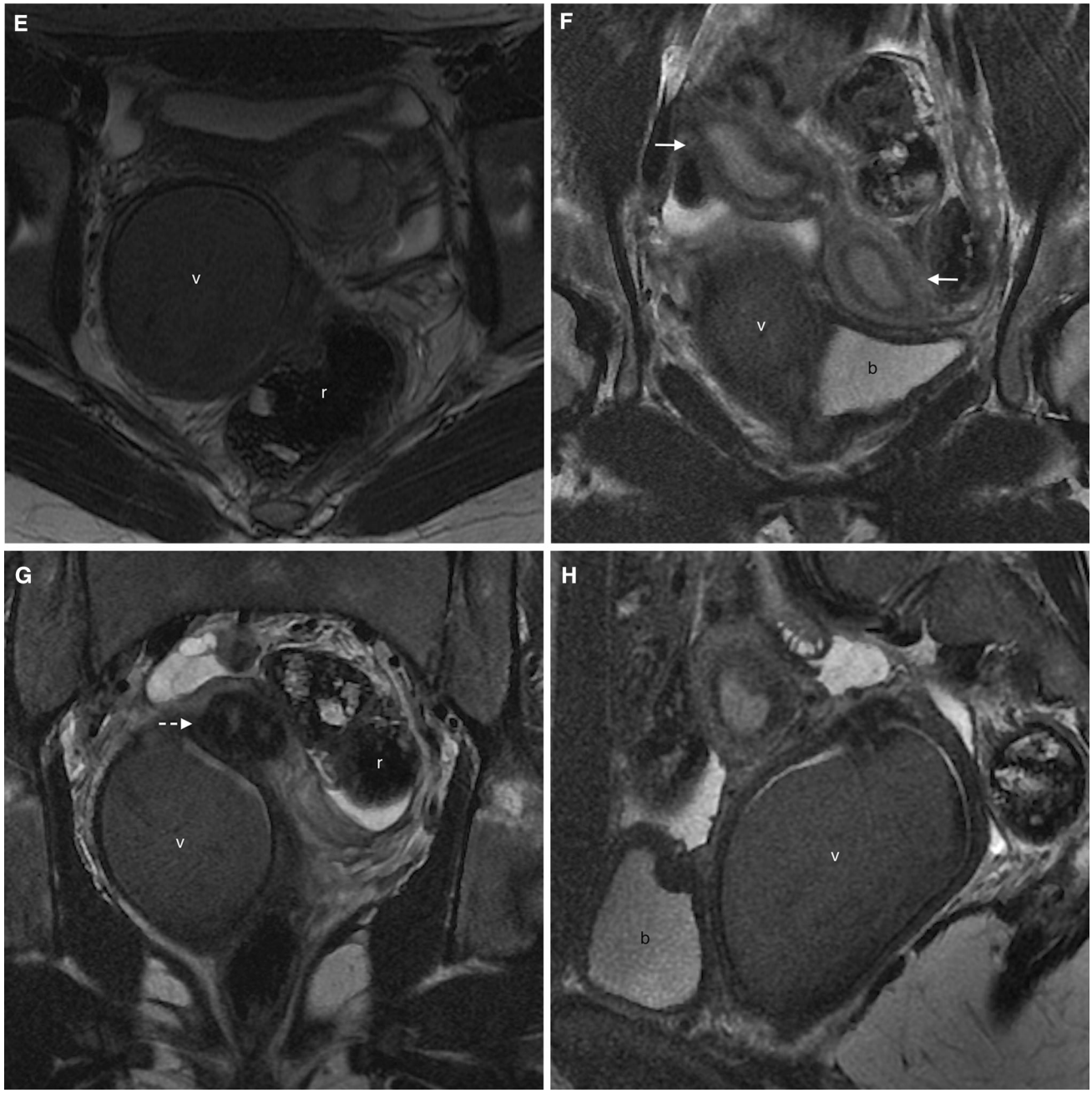


Fig. 2. Images in a 13-year-old girl who presented to the emergency department with a 3-month history of dysmenorrhea (**A**–**C** US; **D** axial T1WI; **E** axial T2WI; **F**, **G** coronal T2WI; **H** sagittal T2WI). US revealed the absence of the right kidney (**A**), two uterine horns (**B**), and a dilated round cavity with homogenous, hypoechogenic content, and posterior enhancement (**C**). MRI was performed, showing a dilated right hemivagina with hematic content (*v*), hyperintense on T1WI (**D**) and hypointense on T2WI (**E**–**H**). Two uterine horns (**F** *arrows*) and two cervices (**G** *dashed arrow*) are clearly depicted. These findings are compatible with uterus didelphys and, in combination with right hematocolpos and ipsilateral renal agenesis, correspond to the HWWS. A schematic representation (**I**) shows the septum position and the obstructed right hemivagina (*pink*). Patient underwent a corrective septectomy 3 months later. On the post-surgery MRI (1 year later), uterus didelphys is easily identified, with two non-dilated uterine cavities (**J**) and two independent cervices (**K** *dashed arrow*). No vaginal obstruction is found. A schematic representation (**L**) reveals a post-surgical non-septate single vagina. *b* bladder; *r* rectum; *v* obstructed hemivagina.



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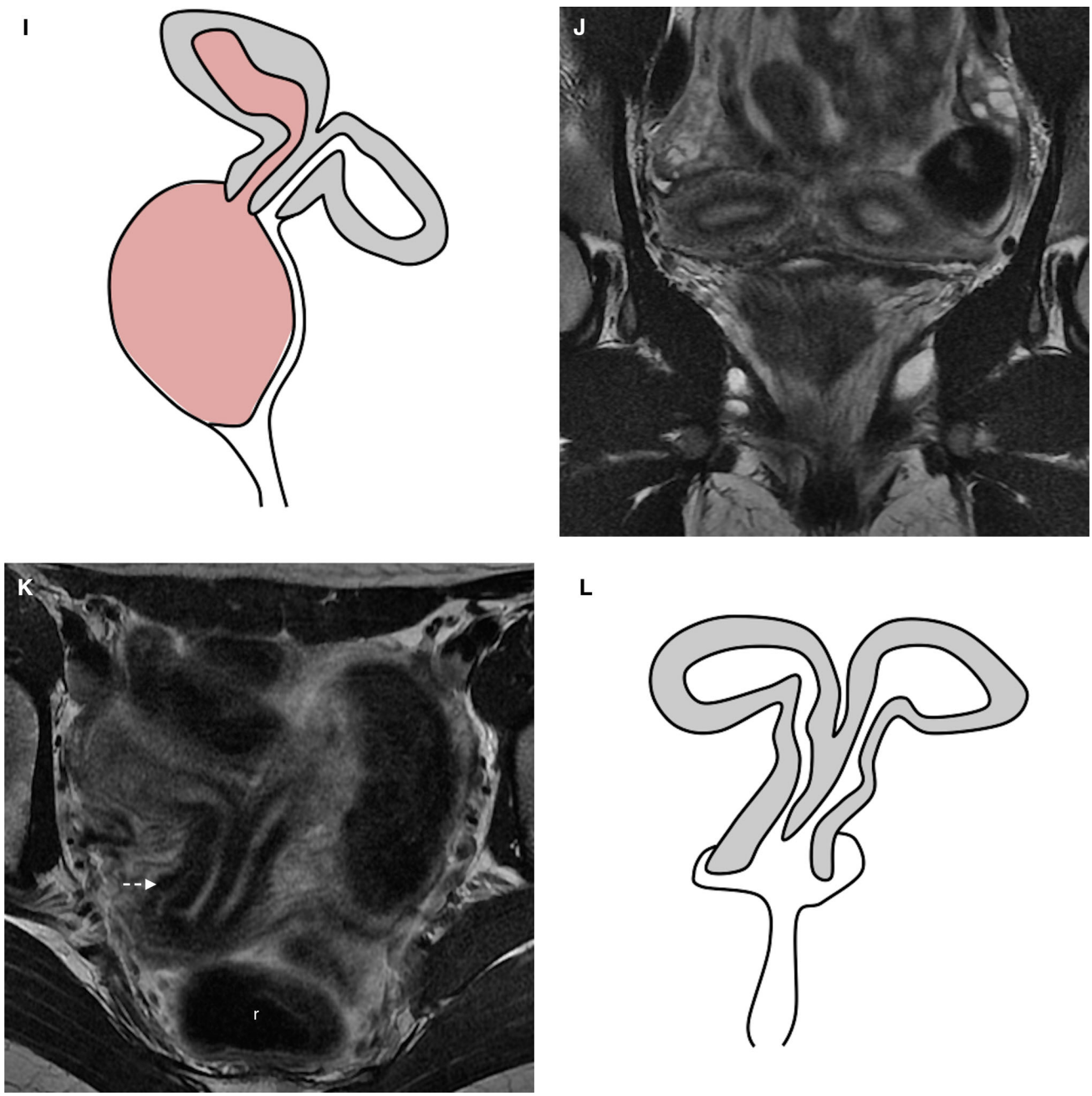
mass that may mimic ischiorectal swelling on initial examination, as reported by Asha and Manila [24].

Pelvic pain is generally intermittent and worsens during menses. Hematocolpos and hematometra are more prevalent at the right side and result from retained menstrual flow. These manifestations are more painful when the vaginal septum fuses with the vaginal wall, leading to a complete obstruction [1, 2, 20, 25]. If there is not a complete obstruction, symptoms may delay or be only mild [12].

When no fusion occurred between the septum and the vaginal wall, the former extends either completely or partially from the cervices to the vaginal introitus. In these cases, two vaginal openings are present and patients are frequently asymptomatic and only incidentally detected [25]. These vaginal duplications are commonly mistaken for HWWS, but actually do not belong to the typical presentation [13].

HWWS is often misdiagnosed even after menarche. Several reasons may justify the delayed symptoms: first,



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due to its distention proprieties, the vagina can accommodate a large volume of blood; second, some of the blood is absorbed between menses [23]; third, anti-inflammatory drugs and oral contraceptives are usually prescribed to relieve dysmenorrhea [2, 26]; and finally, some patients presenting with isolated vaginal discharge are erroneously treated with long-term antibiotics. At the emergency department, the diagnosis is also challenging and frequently simulates inflammatory and infectious disorders like tubo-ovarian abscess [25].

Only a few cases of prepubertal presentation are reported. Pansini et al. [27] reported the case of a 5-month-old infant with an acute urinary retention due to didelphys uterus associated with an obstructed hemivagina. In these early cases, the initial manifestations result from collected secretions within the obstructed hemivagina by the effects of maternal hormones [9]. In another report, Hansen and DeWitt [28] described the case of a 5-year-old female presenting with an 8-month history of recurrent, infectious vaginal discharge secondary to uterus

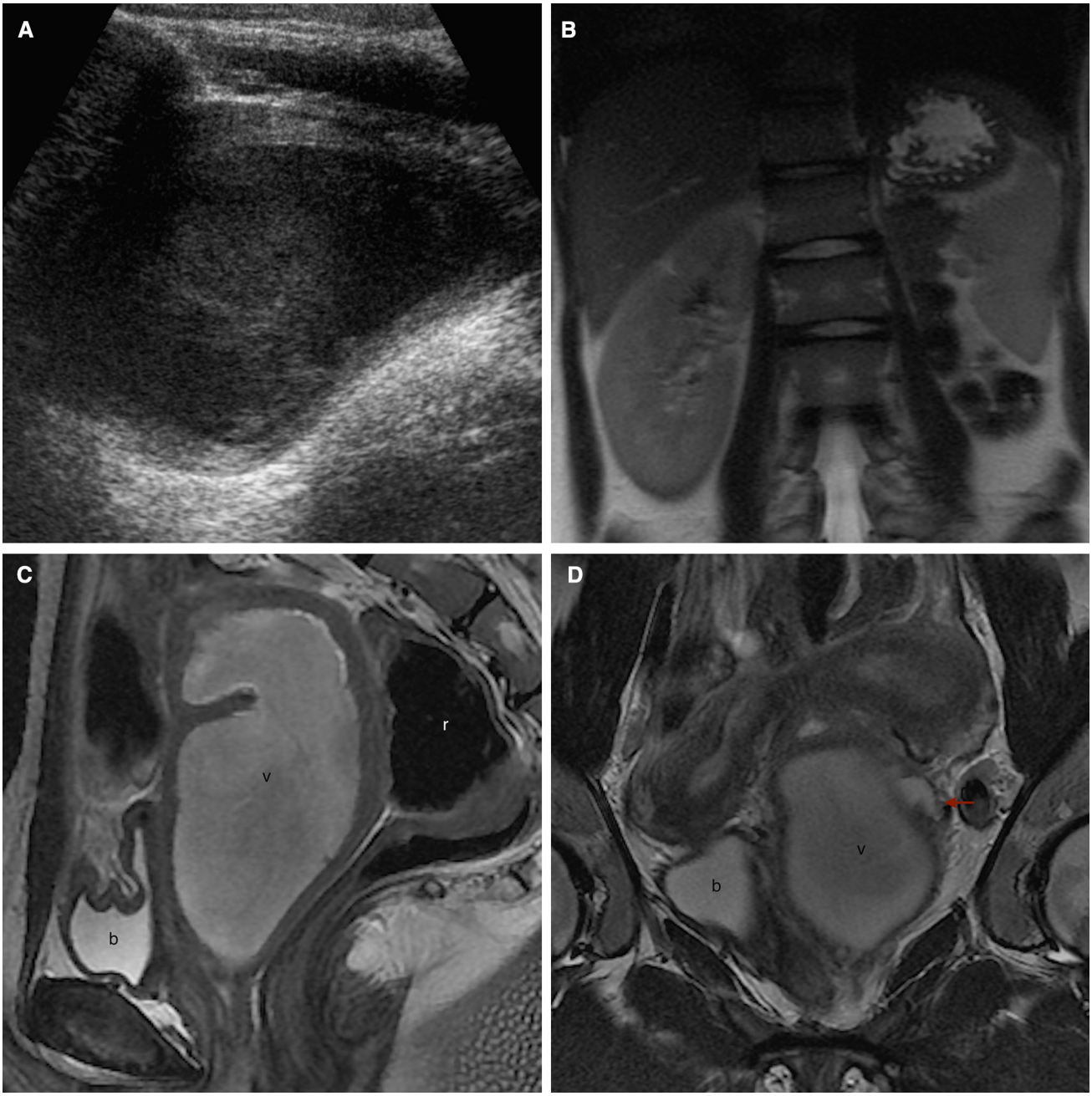
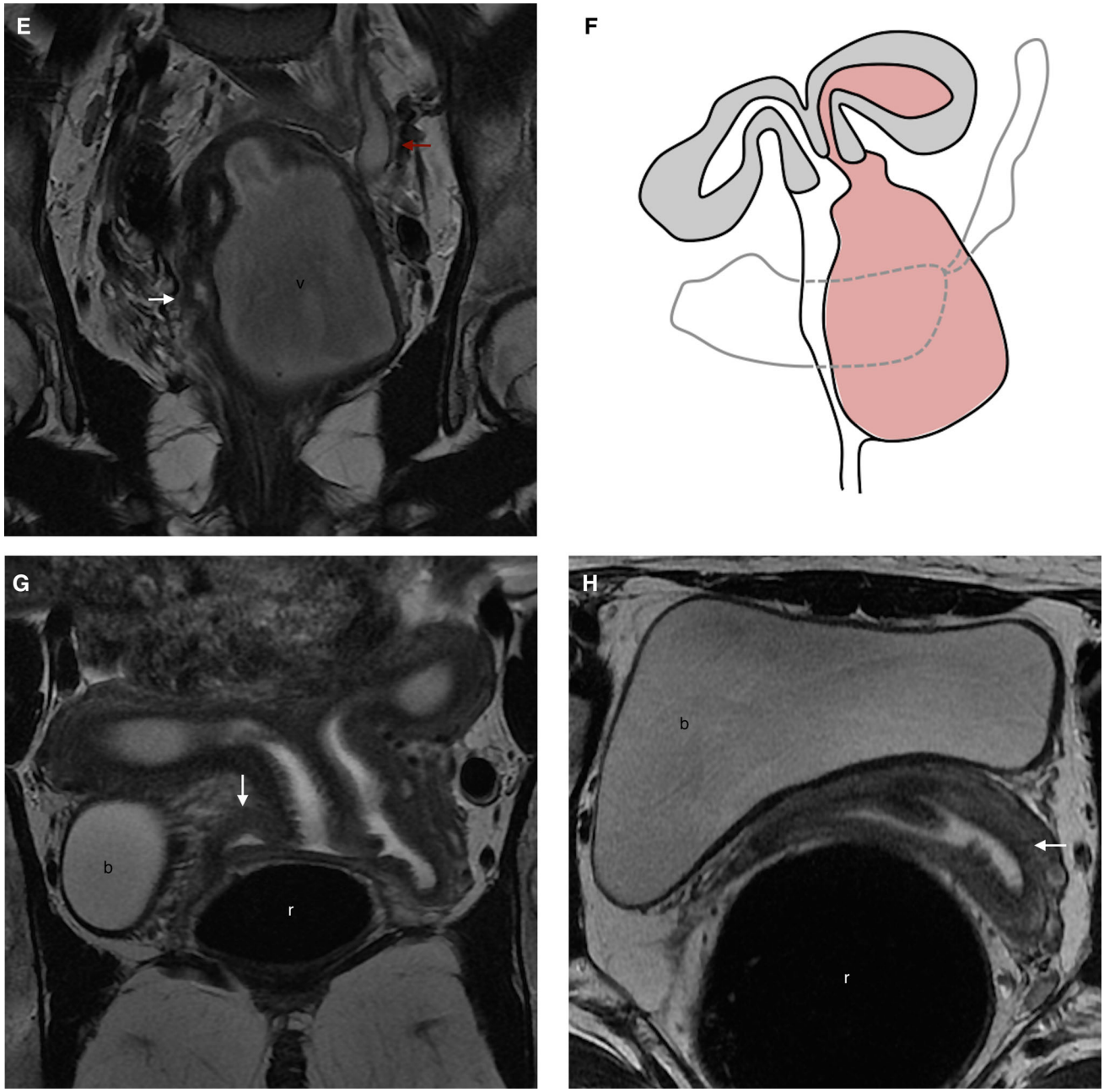


Fig. 3. Images in a 16-year-old girl who presented to the emergency department with a 2-month history of severe pelvic pain and anomalous uterine hemorrhage (**A** US; **B** coronal T2WI; **C** sagittal T2WI; **D**, **E** coronal T2WI; **G** coronal T2WI; **H** axial T2WI). US revealed the absence of the left kidney (not shown) and a pelvic, thick-walled cystic structure with homogeneous, low-level internal echoes, and posterior enhancement (**A**). MRI showed the left renal agenesis (**B**), a huge left hematocolpos (**C**–**E** *v*), and a uterus didelphys (**D**). The non-obstructed right hemivagina (**E** *white arrow*) may be also seen. A dilated left ureteral stump (**D**, **E** *red arrows*) is also identified, coursing lateral, and posterior to the hematocolpos. This triad—uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis—characterizes the HWWS. A schematic representation (**F**) shows the septum position and the obstructed right hemivagina (*pink*), as well as the dilated left ureteral stump (*gray line*). Patient underwent a corrective septectomy 4 months later. On the post-surgery MRI (1 year later), uterus didelphys is clearly depicted (**G**), as well as a single non-obstructed vagina (**G**, **H** *arrow*). A schematic representation (**I**) better illustrates the post-surgical anatomy. *b* bladder; *r* rectum; *v* obstructed hemivagina.



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didelphys with partial hemivagina obstruction. Nevertheless, prepubertal cases are not always symptom-rich. Actually, the normal external genitalia appearance and age-appropriate development may mask asymptomatic abnormalities of the internal reproductive organs [13]. So, the prepubertal diagnosis of the HWWS generally implies a high index of suspicion, which may come from prenatal or neonatal renal US. If renal agenesis is found

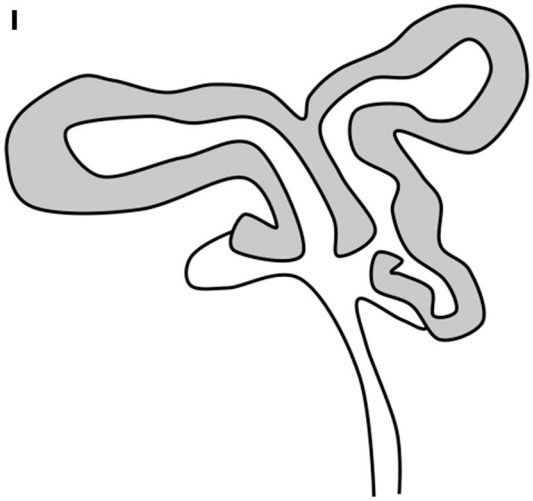


Fig. 3. continued

in prenatal or neonatal US, particular attention should be provided to the external genitalia and, even in the absence of a pelvic or vulvar mass, the risk of further genital anomaly should be highlighted [19].

Complications

Pyohematocolpos, pyosalpinx, and pelviperitonitis may appear as acute complications, while endometriosis, pelvic adhesions, infertility, and obstetric complications constitute potential long-term problems [2, 13].

In some cases, the hemivaginas communicate through a partially fenestrated septum or the two cervices communicate through a fistula, which may lead to pyocolpos due to infection of the retained menstrual blood. These patients tend to present with purulent vaginal discharge [1, 25]. Peritonitis is a rare acute complication and occurs when hematocolpos progress to hematosalpinx and rupture [12].

Pelvic endometriosis is a well-known long-term complication of the HWWS. Tong et al. [1] reported a prevalence of 17.1% to endometriosis among 70 patients with HWWS. Actually, endometriosis is not more common in Müllerian anomalies as a whole, but its incidence increases when outflow obstruction, hematosalpinx, hematometra, or hematocolpos are present. The onset of pelvic endometriosis in patients with HWWS often occurs in adolescence, and some authors believe that long-term continual reverse menstrual flow due to hemivagina obstruction is the main pathological contributor [8, 29]. In another study of Tong et al. [8], all of the 14 ovarian endometriotic cysts found among 94 women with HWWS were ipsilateral to the vaginal septum.

Besides relieving symptoms, early diagnosis and treatment of endometriosis may avoid major anatomical distortion due to pelvic adhesions therefore preventing infertility. Ovarian endometriomas should be removed, especially if large, in order to preserve healthy ovarian tissue and reduce recurrence of endometriosis. On the other hand, surgical correction of the obstructive anomaly usually leads to significant regression of endometriosis [8]. However, Silveira and Laufer [29] reported the cases of five patients in whom endometriosis developed or persisted after surgical correction. The authors advocate that persistence of prior peritoneal implants, coelomic metaplasia, metastases, immunologic deficiency, and genetic predisposition may explain these non-successful cases.

The risk of infertility has constituted matter of concern in patients with HWWS. Nonetheless, some studies like that of Tong et al. [1] showed that the incidence of primary infertility is not increased among women with HWWS. Actually, infertility is a potential complication of HWWS when the diagnosis is delayed and treatment is inappropriate, and appears to be more related to endometriosis and infectious complications than to uterus didelphys [20, 23, 29].

The risk of abortion, preterm delivery, and neonatal morbidity is higher in patients who conceived before surgical treatment [12, 21, 30]. However, women who carry a pregnancy to term often have no obstetric difficulties [13].

Watanabe et al. [31] reported two cases of lower genital tract adenocarcinomas in patients with HWWS, one endometrioid adenocarcinoma within the obstructed cervix and one primary clear cell carcinoma within the obstructed upper vagina. Although malignancy is not a well-established complication yet, the authors recommended periodic imaging evaluation of the blind side of the vagina and uterus.

Diagnostic procedures

US and MRI are the most commonly used imaging modalities for pre-surgical diagnosis of the HWWS [1]. US is a readily available and non-invasive technique that does not use contrast material or radiation. Transabdominal US is unreliable and particularly limited in the presence of insufficient bladder distension, bowel interposition, uterine retroflexion, and leiomyomas [32]. On the other hand, transvaginal US improves anatomical depiction and better assesses contour anomalies like fundal clefts. According to Pellerito et al. [32], transvaginal US may suffice to make a correct diagnosis. However, this is not a valid option in the pediatric set before the beginning of sexual life.

Three-dimensional (3D) US has been developed over recent years. It may be used with transabdominal or transvaginal probes and allows the radiologist to view

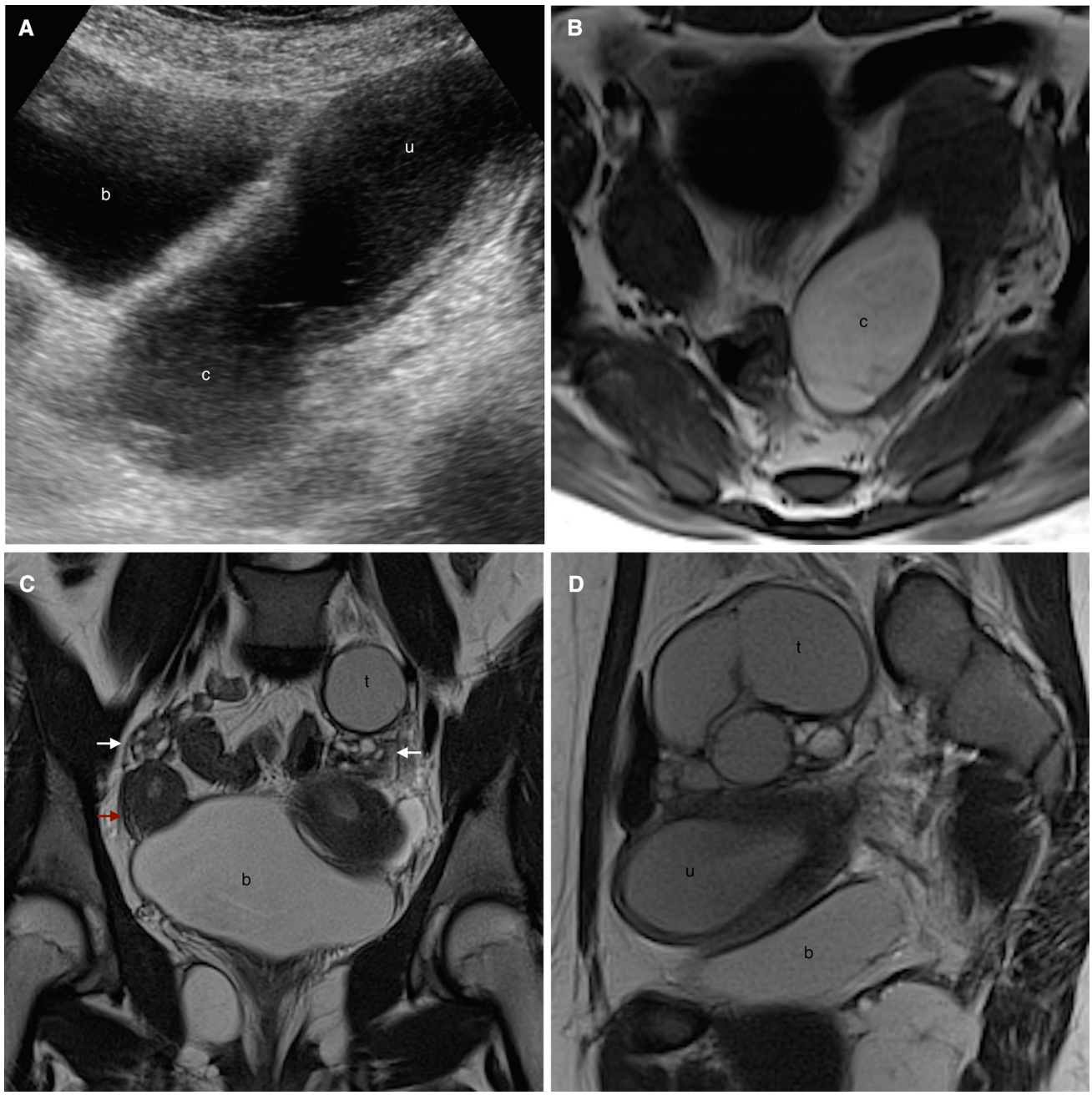
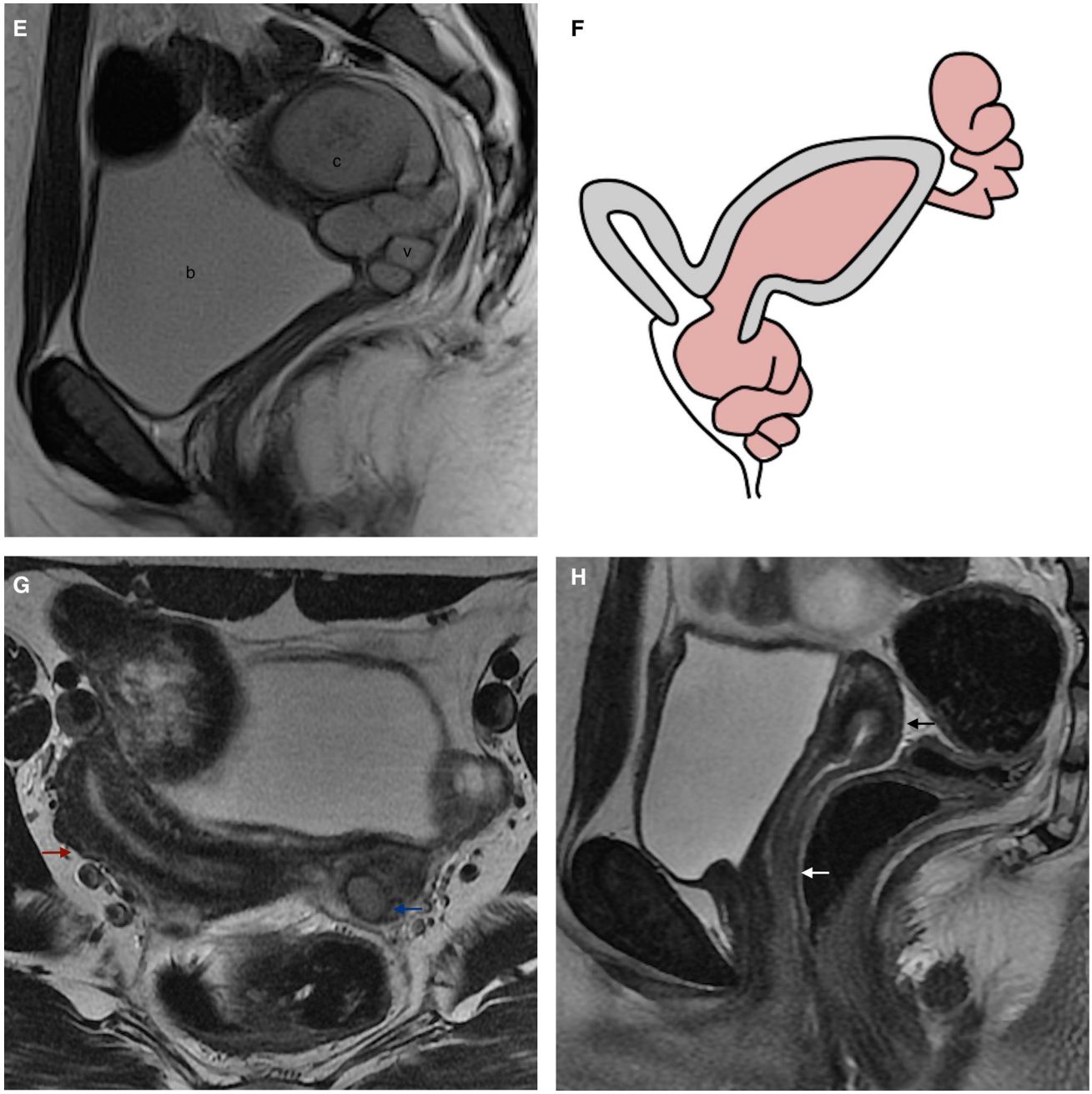


Fig. 4. Images in a 12-year-old girl who presented to the emergency department with a 1-month history of severe dysmenorrhea (**A** US; **B** axial T1WI; **C** coronal T2WI; **D**, **E** sagittal T2WI; **G** axial T2WI; **H** sagittal T2WI). US revealed the absence of the left kidney (not shown) and a pelvic, thick-walled tubular structure with heterogeneous, mixed echogenicity, and posterior enhancement (**A**). MRI showed a uterus didelphys with a non-dilated right horn (**C** *red arrow*) and left hematocolpos (*v*), hematocervix (*c*), hematometra (*u*), and hematosalpinx (*t*). Blood appeared hyperintense on T1WI (**B**) and slightly less intense on T2WI (**C**–**E**). The hematocolpos essentially involved the upper vagina and was heterogeneous and lobulated, suggesting co-infection. Both ovaries were identified (**C** *white arrows*), the left one encased in a dilated Fallopian tube (*t*). A schematic representation (**F**) shows the septum position, the obstructed right hemivagina and blood regurgitation to the ipsilateral Fallopian tube (*pink*). This case exemplifies an acute complication with hematosalpinx and probable pyocolpos. Ten days later, the patient underwent a left hemihysterectomy with salpingectomy and preservation of the left ovary. On the post-surgery MRI (1 year later), single uterus (**G** *red arrow*) and vagina (**H** *white arrow*) are seen, as well as a remnant hemorrhagic cystic structure (**G** *blue arrow*). A schematic representation (**I**) better illustrates the post-surgical anatomy. *b* bladder, *c* cervix, *r* rectum, *t* Fallopian tube, *u* uterine body, *v* obstructed hemivagina.



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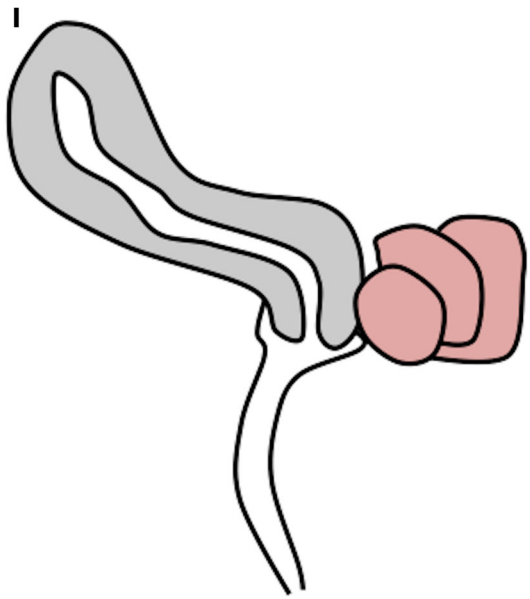
images of the three orthogonal planes simultaneously. 3D US appears to provide high-quality images similar to those yielded by MRI and achieve high-accuracy levels in the diagnosis of congenital uterine anomalies. Moreover, its lower cost and superior tolerance by patients constitute relevant advantages [11].

According to our experience, transabdominal US should be performed prior to the transvaginal approach even in sexually active patients. The former enables a global evaluation of the pelvis, which is particularly useful in patients with uterus didelphys since the uterine horns may diverge and be laterally placed within the pelvis. In these cases, an isolated transvaginal approach may not suffice if the field of view is too limited. At our institution, we perform transabdominal US with a convex probe with a frequency bandwidth ranging from 6 to 2 MHz (Siemens Acuson Antares). When indicated, transvaginal US is performed using an endocavitary probe with a frequency bandwidth ranging from 4 to 9 MHz (GE Voluson E8).

US usually identifies two hemiuteri compatible with uterus didelphys and dilatation of the proximal vagina



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(Figs. 2, 3, 4). The content of the dilated reservoir is hemorrhagic, ranging from anechoic fluid to heterogeneous collections with mixed echogenicity. In the set of infection, some echogenic foci representing gas bubbles may be found. In some cases, hydrometra or hematometra may be found as a result of regurgitation of the vaginal content to the endometrial cavity. The distinction between an obstructed vagina and an adnexal cystic lesion like endometriomas or other complex ovarian cysts may be difficult [9].

MRI is safe, non-invasive, and free of ionizing radiation. It better depicts pelvic anatomy and documents uterine anomalies, thus being useful in the management of surgical treatment options [9, 23]. MRI is more accurate than hysterosalpingography (HSG) or US not only in the evaluation of the uterine contours, the shape of the endometrial cavity and the course of the vaginal septum, but also in the detection of acute and chronic complications [9]. High-resolution T2WI should be obtained in three different planes, one of them parallel to the long axis of the uterus, in order to better understand the uterine morphology and the features of the septum [9]. Sometimes, the non-obstructed hemivagina is flattened by the hematocolpos and difficult to recognize (Fig. 3). T1-weighted images (T1WI) are generally obtained in the axial plane, showing the high signal intensity of the hemorrhagic fluid within the obstructed hemivagina (Figs. 2, 4). In some cases, this fluid looses signal intensity from T1WI to T2-weighted images (T2W1), due to high concentration of protein and iron from recurrent hemorrhage (Fig. 2). This T2 shortening is known as shading sign and was initially described as an MRI sign of ovarian endometriomas. In the set of the HWWS, it is not uncommon to find endometriomas and post-obstructive hematosalpinx, both potentially hyperintense on T1WI. If any doubt persists, a complementary fat-saturated T1WI sequence should be performed in order to exclude a fat-containing mass. Intravenous contrast material should not be routinely used but may be helpful in the setting of infectious complications or incidental findings. At our institution, MRI is performed on a 3.0 Tesla (T) magnet (Signa HDxt, GE Healthcare). Specific parameters are provided in Table 1.

HSG provides valuable information regarding the interior cavity of the uterus. However, this technique fails in the characterization of some uterine subtypes since only patent cavities are demonstrated and the external contour and rudimentary non-communicating horns are not visualized. Moreover, it implies contrast material and radiation exposure. Thus, HSG is not currently considered a first-line modality when Müllerian duct anomalies are suspected [11, 32].

Computerized tomography (CT) does not easily depict pelvic anatomy and is not recommended in the differential diagnosis of congenital uterine anomalies [9]. The exposure to ionizing radiation and the eventual intravenous administration of iodinated contrast also constitute important disadvantages in the pediatric context. However, CT may be required in the set of acute abdomen when US is not conclusive. A spontaneously hyperdense cystic lesion may be found, corresponding to an obstructed, hemorrhagic, or infected hemivagina [26].

Hysteroscopy allows direct visualization of the intrauterine cavity and ostia but does not evaluate the external contour of the uterus. Thus, it is frequently inconclusive and implies further investigation. Moreover, this procedure is invasive, usually needs anesthesia, and is less appropriate in pediatrics [11].

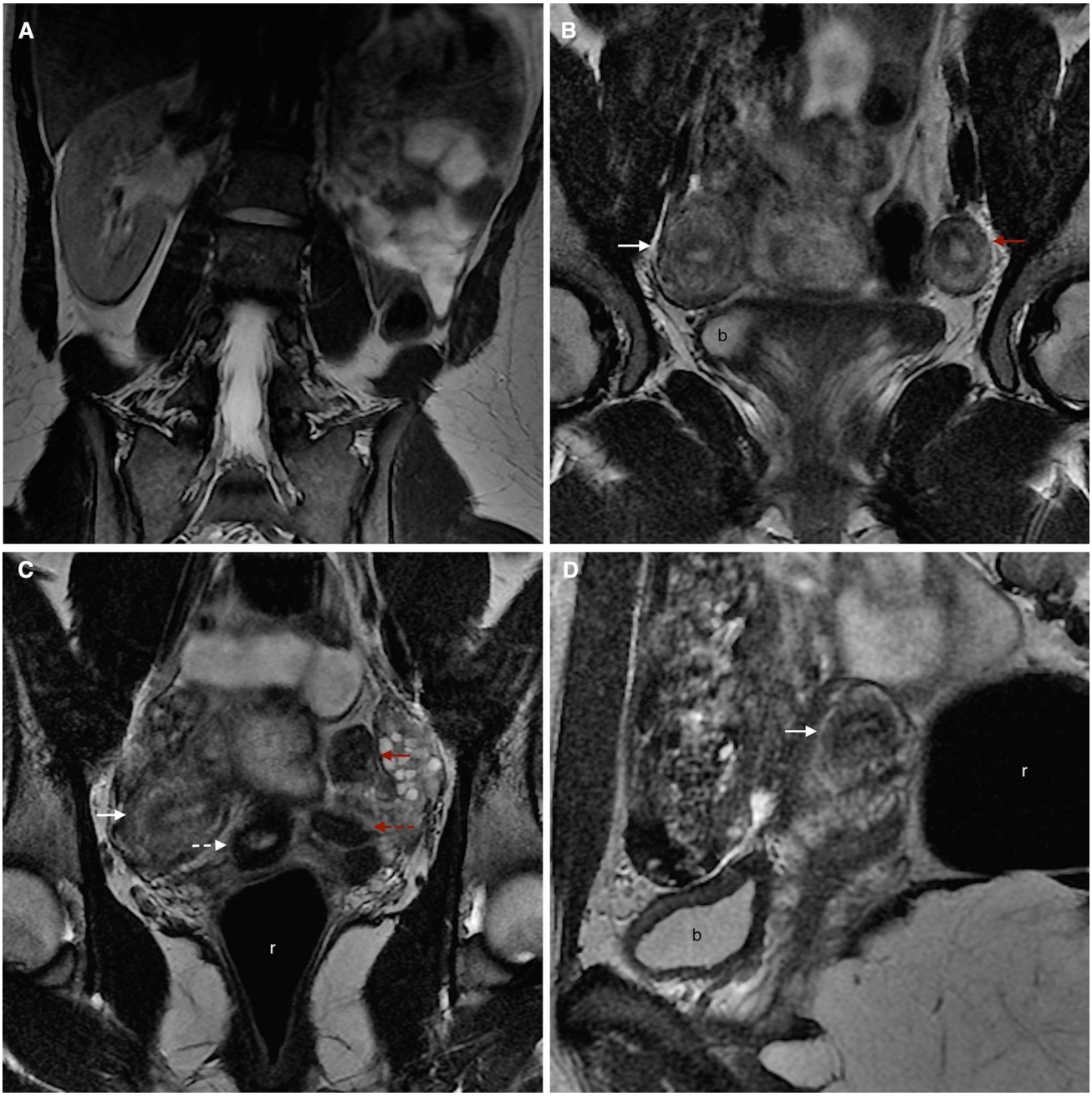
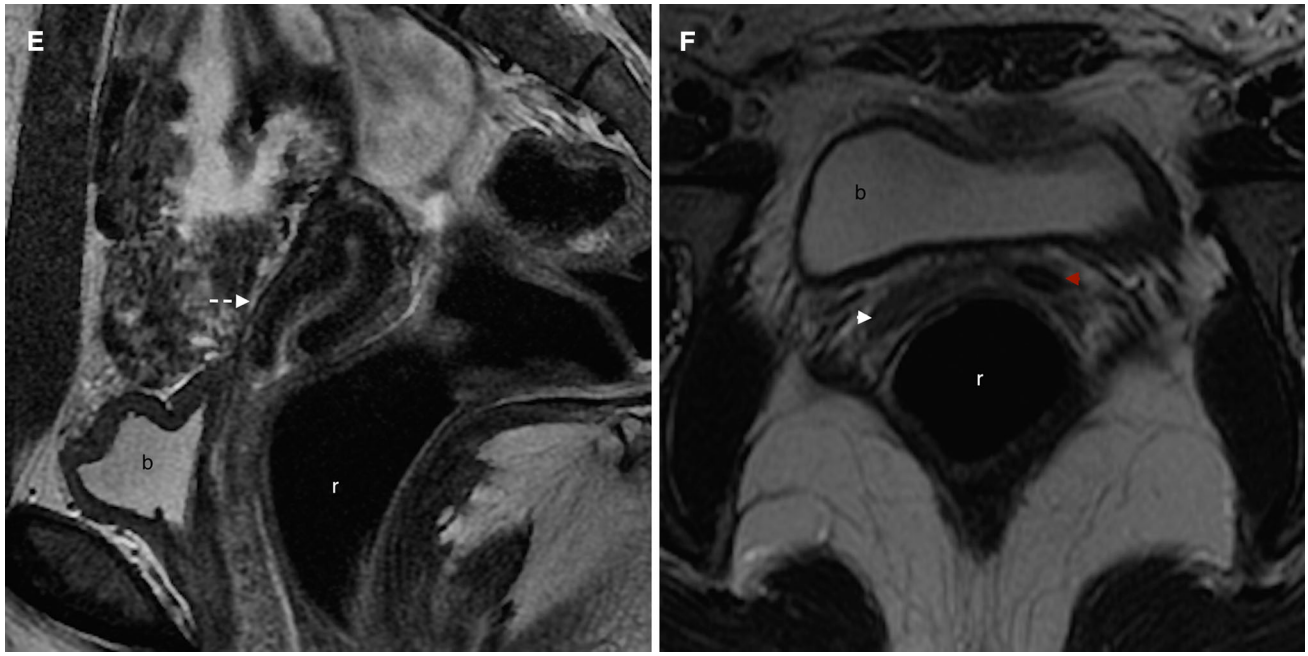


Fig. 5. **A**–**F** Images in a 17-year-old girl who complained with a 6-year history of severe dysmenorrhea (**A**–C coronal T2WI; **D**, **E** sagittal T2WI; **F** axial T2WI). MRI showed a left renal agenesis (**A**) and a uterus didelphys. Two independent uterine horns and two non-communicating cervices are recognized. The right uterine horn (**B**–**D** *white arrow*) and cervix (**C**–**E** *white dashed arrow*) have a normal appearance. The left uterine horn is smaller (**B**, **C** *red arrow*), and the ipsilateral cervix appears thinner and tortuous (**C** *red dashed arrow*). Two independent hemivaginas seem to be found at a lower level (**F** *white and red arrow-heads*). Despite the absence of obstruction, these findings appear to be related to the HWWS. Long-term oral contraceptives may explain the lack of hematocolpos. The existence of incomplete vaginal septa or longitudinal non-fused septa may also be the cause. *b* bladder, *r* rectum.

Laparoscopy and laparotomy may also be used for diagnostic purposes but are expensive and invasive procedures with anesthesia- and surgery-related risks [32]. However, some authors like Zurawin et al. [26] advocate that laparoscopy should be the gold standard for the complete evaluation of congenital anomalies of the female reproductive tract. Despite referring that MRI may correctly anticipate the diagnosis, the authors consider that laparoscopic evaluation better identifies endometriosis, pelvic infection, and adhesions.



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Treatment

The main purposes of the surgical treatment in HWWS are to relieve symptoms and guarantee successful reproductive outcomes [13].

The treatment of choice for patients with HWWS is the surgical excision and marsupialization of the longitudinal vaginal septum (Figs. 2, 3) [25]. Some surgeons prefer to perform the vaginal septum resection only after a large hematocolpos develops. It distends and thins the septal tissue consequently becomes easier to excise [26, 33]. Care must be taken during the dissection in order to preserve the hymen and avoid damage to the bladder or the rectum [26].

Some asymptomatic patients with HWWS are suspected after the identification of a renal agenesis at prenatal or neonatal US (Figs. 5, 6). Some surgeons decide to closely follow these patients and ensure that surgical correction is performed before hormonal impregnation at puberty. In young adolescents, postponement of surgery may be required; in these cases, gonadotropin-releasing hormone analogs may constitute a valid option for maintaining amenorrhea, particularly if cervical atresia coexists [1].

Some morphological features increase the complexity of the surgery and the post-surgical risk of contracture, stenosis, infection, and reoperation need. These include thick septum, narrowed area of resection, collapsing sidewalls, and high septum location. Vaginal packing, vaginal dilators, and vascular stents are reported in the literature as useful to maintain the vaginal patency in these complicated cases [33].

The use of laparoscopy at the time of vaginal septectomy has been used to assess the uterine anatomy. However, the impact of this approach on definitive treatment remains unclear, and some authors believe that a timely diagnosis avoids unnecessary laparoscopies [13]. Endoscopic ablation of the vaginal septum has been reported as an alternative to the conventional open surgical excision. This is a minimally invasive technique that lacks major morbidity and appears to achieve equivalent results to those of open surgical excision, as reported by Roth et al. [34]. Hemihysterectomy with or without salpingo-oophorectomy should be avoided in order to preserve fertility. It is rarely indicated when resecting the vaginal septum is not enough to relieve the hematometra (Fig. 4) [13, 23]. Metroplasty, a reconstructive uterine surgery, may also be indicated in some variants. Nabeshima et al. [18] reported the case of a 12-year-old girl with an obstructed non-communicating uterus didelphys without vaginal septum in whom a laparoscopic metro plastic surgery was successfully performed. Women with a uterine septum and recurrent pregnancy loss may also benefit from metroplasty [21].

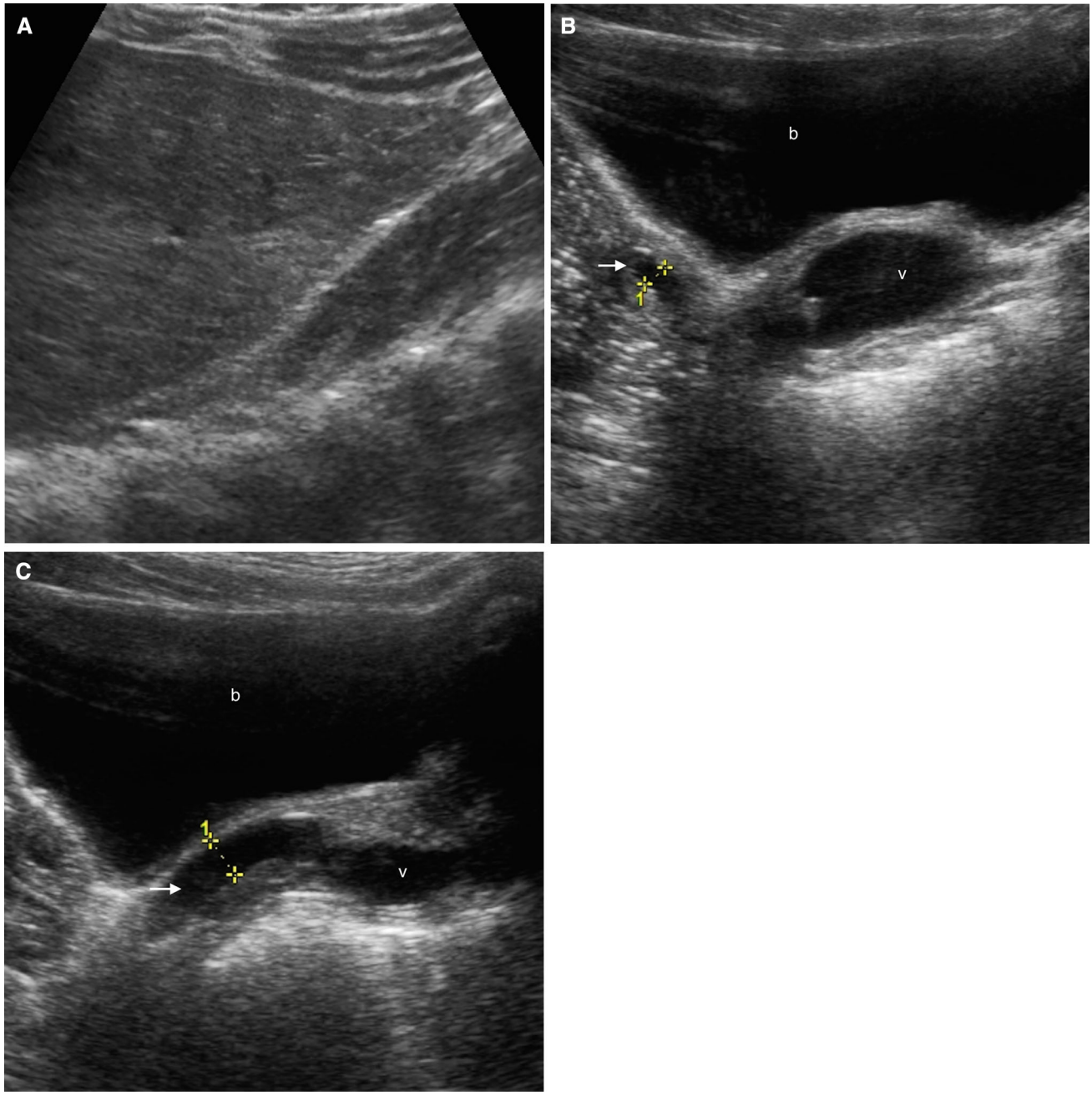

Fig. 6. US images in a 3-year-old patient with prenatal diagnosis of right multicystic dysplastic kidney. Postnatal right kidney resorption occurred (**A**). The right ureteral stump is dilated (**B**, **C** *white arrows*) and ends in a tubular structure coursing posterior to the bladder (*b*). These findings suggest the presence of an obstructed hemivagina in association to an ectopic ureter. The uterus is not easily identified due to its immature morphology. This case probably corresponds to a variant of the HWWS. A rigorous follow-up should be performed in order to characterize the uterine anatomy and guide the patient toward timely and appropriate surgical correction. *b* bladder, *v* hemivagina.

Conclusion

The triad of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis characterizes the HWWS. This syndrome usually presents with non-specific symptoms after menarche and is frequently mistaken for inflammatory and infectious pelvic disorders. Anti-inflammatory drugs, oral contraceptives and/or antibiotics are therefore prescribed, relieving symptoms and delaying the diagnosis. Before puberty, the diagnosis is even more challenging, unless early acute complications occur, leading to emergent imaging evaluation. If renal agenesis is found at prenatal or

Table 1. MRI protocol

	Axial T2WI	Coronal T2WI	Sagittal T2WI	Axial T1WI	Axial FS T1WI	Dynamic CE FS T1WI[a]	Coronal T2WI[b]
Sequence type	frFSE	frFSE	frFSE	FSE	FSFSE	GRE	SSFSE
FOV (cm)	18	20	22	32	32	34	48
Matrix	352 × 256	384 × 256	384 × 256	512 × 320	384 × 256	320 × 256	512 × 320
Slice thickness (mm)/spacing (mm)	3.5/0.3	3/0.3	4/0.4	5/0.5	5/0.5	3/0	6/0.6
TR	4860	5160	3440	760	680	3.5	1800
TE	120	120	100	Minimum full	Minimum full	1.7	100
Flip angle (°)	90	90	90	90	90	12	90

[a]Phases at 50, 100, 150 and 200 s after contrast administration
[b]Upper abdomen
FS, fat saturation; CE, contrast-enhanced; frFSE, fast relaxation fast spin-echo; FSE, fast spin-echo; GRE, gradient echo; SSFSE, single-shot fast spin-echo; FOV, field of view; TR, time to repetition; TE, echo time

neonatal US, the risk of further genital anomaly should be considered.

Pediatricians, radiologists, gynecologists, and pediatric surgeons should become familiar with this syndrome and other congenital anomalies of the female genital tract. Despite its rarity, an accurate preoperative diagnosis can be achieved and depends on a high clinical suspicion. Prognosis may be excellent if a timely corrective surgery is performed, thus preventing long-term complications and preserving future fertility.

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