

ORIGINAL RESEARCH ARTICLE

Forty-two normomenstruating adolescents with Müllerian obstructive anomalies: Presentation, pitfalls in the diagnosis and surgical management

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Abstract

Introduction: We analyzed the frequency, presentation and pitfalls in the diagnosis and surgical management of a large group of normomenstruating adolescents with obstructive reproductive tract anomalies.

Material and methods: Retrospective analysis of prospectively collected data. Of the 143 outpatients referred for severe dysmenorrhea and persistent pelvic pain, 42 (29.3%) young women with obstructive Müllerian anomalies and regular menstrual flow were identified. These patients were divided into four groups: (1) patients with duplicate uterine cavities, obstructed hemivagina and ipsilateral renal agenesis ($n = 34$); (2) patients with unicornuate uterus and noncommunicating cavitated rudimentary horn ($n = 5$); (3) patients with accessory cavitated uterine mass ($n = 2$); (4) patients with partially obstructed transverse vaginal septum ($n = 1$). All 42 patients were conservatively treated via laparoscopy and 35/42 patients had also vaginal surgery.

Results: Of the four groups, patients in groups 2 and 3 ($n = 7$) were conservatively managed by laparoscopy alone; for patients in groups 1 and 4 ($n = 35$), laparoscopy and the vaginal approach were used. Patients of group 1 were treated by resecting the obstructed vaginal septum with drainage of retained collections. In patients in group 2, surgery consisted of the removal of the rudimentary horn. Patients of group 3 were treated by the removal of myometrial neoformations. In the patient in group 4, treatment consisted of removal of the septum. All surgical procedures were successful and no major complications were recorded. Follow-up reports highlighted the disappearance of obstruction and clear improvement in pain symptoms.

Conclusions: Unilateral obstructive anomalies of the female genital tract are difficult to identify. Early diagnosis allows the preservation of reproductive activity and avoids potential complications.

Abbreviations: ACUM, accessory cavitated uterine mass; MRI, magnetic resonance imaging; ORTA, obstructive reproductive tract anomalies.

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KEYWORDS

dysmenorrhea, Herlyn–Werner–Wunderlich syndrome, Müllerian anomalies, OHVIRA syndrome, renal agenesis, unicornuate uterus

1 | INTRODUCTION

With the widespread improvement of non-invasive diagnostic techniques, obstructive reproductive tract anomalies (ORTA) associated with amenorrhea are now promptly diagnosed and treated.¹ Conversely, ORTA, which is associated with regular menstrual flow, albeit with some degree of dysmenorrhea, is challenging to diagnose, often leading to serious compromise of the patients' reproductive future.² Diagnostic delay is related to various factors. First, adolescents with obstructing genital malformations usually become symptomatic several years after menarche. Furthermore, dysmenorrhea is fairly common in teenagers, and pediatricians and pediatric surgeons are consulted before gynecologists. Severe dysmenorrhea in obstructive Müllerian anomaly may occur due to the following reasons: a uterus with duplicate cavities and imperforate hemivagina; a noncommunicating cavitated uterine horn in unicornuate uterus; an accessory cavitated uterine mass (ACUM); a microperforate transverse vaginal septum.³

The total and relative incidences of these anomalies are unknown, since most studies have reported the occurrence of ORTAs only in infertile patients. An incidence of ORTAs of between 0.1% and 3.8% in the general female population and 7% in all Müllerian anomalies is considered credible.^{4,5}

This report analyzed the symptoms, diagnostic difficulties and surgical solutions in a large group of adolescents with ORTA and normal periods who were referred to two tertiary centers for pediatric and adolescent gynecology.

2 | MATERIAL AND METHODS

2.1 | Study design and participants

This was an observational study performed at two tertiary centers with expertise in managing complex Müllerian anomalies with reconstructive surgery using minimal access techniques.

The study population consisted of adolescents with severe dysmenorrhea and persistent pelvic pain despite the use of non-steroidal anti-inflammatory drugs and oral contraceptives for at least 3–6 months. From 2010 to 2021, 157 patients were referred to the Center for Adolescent Gynecology of the State University of Milan (2010–2017), and University Vita Salute Ospedale San Raffaele, Milan (2018–2021) by primary care practitioners, pediatricians and gynecologists. Patients' original documents were reviewed and data were collected. Follow-up results were assessed using a questionnaire. Age at menarche, age at onset of dysmenorrhea, age at presentation to our clinic, pain control medications used prior to referral, characteristics of dysmenorrhea, vaginal discharge,

Key message

Unilateral obstructive Müllerian anomalies are not uncommon in adolescents and pose a diagnostic and surgical challenge. Early diagnosis allows the preservation of reproductive activity and avoids potential serious complications.

dyspareunia (for sexually active patients), pelvic pain, investigations and operative procedures prior to referral were analyzed. Fourteen patients had incomplete data or were lost to follow-up; these patients were excluded from the analysis. The remaining 143 patients were included in our cohort study.

2.2 | Intervention and outcome assessment

For the initial evaluation the patients' clinical histories were collected (particularly beginning, cyclicity, location and radiation of pelvic pain, vaginal bleeding or abnormal vaginal discharge) to assess for signs of normal or delayed puberty and to reveal other abdominal or systemic pathologies. When complete vaginal examination was not possible, rectal examination proved very useful. The following imaging techniques were used: abdominal and transvaginal/transrectal ultrasonography (all patients), magnetic resonance imaging (MRI) (if not performed before referral) and hysterosalpingography (in select cases).

Abdominal ultrasound was used to study the conformation of the uterus, presence of hematocolpos and/or hematometra and presence of menstrual or inflammatory fluid in the peritoneum, and to investigate the urinary system. On transvaginal/transrectal ultrasound the sonographer determined the characteristics of the uterine body and cervix. In the presence of paravaginal masses that might have been interpreted as imperforate hemivaginas, the sonographer attempted to clarify whether the hemivagina was related to one of the three types traced by Rock et al.⁶ MRI was used to obtain detailed information. In fact, because of its excellent soft tissue contrast, large field of view and absence of ionizing radiation, MRI remains the preferred imaging method in pediatric patients when transvaginal ultrasound is avoided. Hysterosalpingography was only used to document the communication between the obstructed and contralateral sides.

When anatomical anomalies of the genital system were excluded by instrumental investigations, a second- or third-line medical therapy with continuous oral contraceptives or progestogens⁷ was proposed. If the instrumental investigations were suggestive of an anatomical anomaly, particularly the presence of urinary system malformations, the patient underwent laparoscopy and surgical intervention on a case-by-case basis. Laparoscopy was used in all

surgical procedures, to perform operations with minimal access, to document the conformation of the genital system and to obtain information on associated pathologies.

2.3 | Ethics statement

This study was approved by the Ethics Committee of I.R.C.C.S. S.Raffaele Hospital, Milano on May 27, 2021 (protocol code GARA, register number CE 73/INT/2021).

3 | RESULTS

Between 2010 and 2021, 42 patients with obstructive Müllerian anomalies and regular menstrual flow were identified. These accounted for 29% of all referrals at our tertiary level clinic for severe dysmenorrhea and persistent pelvic pain. These patients were divided into four groups, in the order of frequency: (1) patients with duplicated uterine cavities, imperforate hemivagina and ipsilateral renal agenesis ($n = 34$); (2) patients with unicornuate uterus and noncommunicating cavitated rudimentary horn ($n = 5$); (3) patients with ACUM ($n = 2$); and (4) patients with partially obstructed transverse vaginal septum ($n = 1$).

3.1 | Group 1: obstructed hemivagina and ipsilateral renal agenesis

The patients in group 1 had Herlin–Werner–Wunderlich syndrome,⁸ also known as obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome.⁹ This rare congenital malformation simultaneously involves the Müllerian and Wolffian ducts. OHVIRA syndrome in the clinical-embryological classification corresponds to mesonephric anomalies 2.1.^{10,11} The patient characteristics and symptoms are presented in Table 1 and Figure 1A.

Abdominal ultrasound was performed in all 34 patients; however, the double nature of the uterine body was successfully identified in only 32 patients, without determining whether it was a septate or didelphys uterus. In 16 cases, a paravaginal collection was suggested without specifying its characteristics, and in four cases the presence of haematosalpinx and inflammatory fluid in the pouch of Douglas was suggested. Ipsilateral renal agenesis was diagnosed in 33 of 34 patients (97.1%), whereas an ipsilateral dysplastic kidney was found in the remaining patient. Transvaginal (when possible) or transrectal ultrasound successfully identified uterine anomalies and hemivaginal obstruction; however, it failed to identify intervaginal or interisthmic communication between the two hemiuteri. MRI was equally precise in identifying the type of uterine anomaly and the collection in the imperforate hemivagina; however, it failed to identify the presence and site of communication between the two genital axes. MRI was also useful in providing helpful details for the surgery, such as the presence of a ureteral remnant in an imperforate hemivagina.

TABLE 1 Characteristics and symptoms of the patients in Group 1 ($n = 34$)

Mean age at presentation	13.8 years	range 11.5–19 years
Mean time between menarche and presentation	1.1 year	range 0.5–4.5 years
Dysmenorrhoea	34	100%
Chronic pelvic pain	34	100%
Acute abdominal pain	4	11.7%
Dyspareunia (in 14 sexually active patients)	6	42.8%
Intermenstrual bleeding	4	11.7%
Vaginal discharge	3	8.8%
Fever	4	11.8%
Paravaginal mass	34	100%
Obstruction side	Right side 58%	Left side 42%
Ipsilateral renal agenesis	33	97.1
Ipsilateral dysplastic kidney	1	2.9%

Hysterosalpingography was only used in select cases to document communication between the obstructed and contralateral sides.

All patients in this group were treated by resecting the obstructed vaginal septum and draining the retained collections (Figure 2). Diagnostic laparoscopy was performed at the time of surgery. Surgical findings clarified that in 33 of 34 cases, the uterine body was didelphys and in one it was completely septate. Of the 34 cases, there was no communication between the two Müllerian axes (Rock's type 1), in five the hemivagina was completely imperforate with inter-isthmic communication between the two hemiuteri (Rock's type 2) and in six the hemivagina was incompletely obstructed without communication between the two hemiuteri (Rock's type 3). In addition to a thorough examination of the abdomen and pelvis, laparoscopy revealed hemoperitoneum in four cases, inflammatory fluid in the pouch of Douglas in five cases and haematosalpinx ipsilateral to the area of fluid retention in five cases. Peritoneal endometriosis foci were observed in seven cases. The opening of the imperforate hemivagina was always sufficiently wide. In fact, the simple incision of the obstructing septum or its only partial removal very often leads to a reclosure of the septum with the recurrence of hematocolpos or pyocolpos.¹² However, too large an excision can lead to injuries to the urethra, bladder and rectum.² The surgical procedure was successful in all cases and no major complications were observed. The postoperative course was no pain on and the mean (\pm SD) hospital stay was 2.8 ± 1.6 days.

Follow-up reports were obtained for all 34 patients. Signs and symptoms associated with the obstruction were completely resolved. Dysmenorrhoea improved in over 90% of patients, and none of the patients complained of abdominal pain. Ultrasonography performed 1 and 6 months after surgery confirmed the disappearance of vaginal retention.

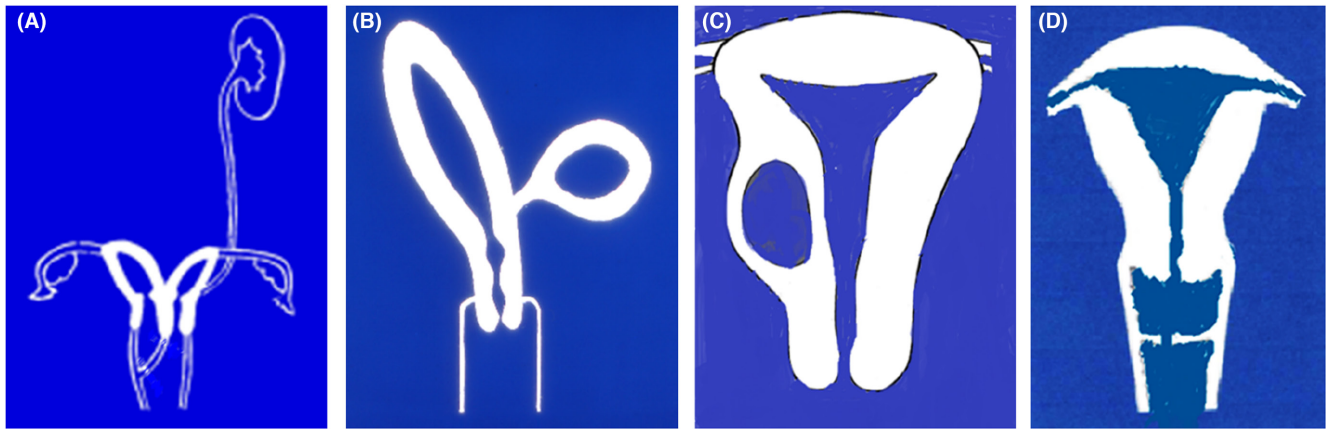


FIGURE 1 (A) Didelphic uterus with imperforate hemivagina and ipsilateral renal agenesis. (B) Right unicornuate uterus with cavitated noncommunicating rudimentary horn. (C) Accessory cavitated uterine mass (ACUM). (D) Transverse microperforate vaginal septum.

FIGURE 2 Didelphic uterus with left imperforate hemivagina. (A) In laparoscopy, enlargement of the left uterine horn is visible, ipsilateral haematosalpinx and left posterior isthmic swelling due to hematocolpos. (B) Vaginal time: opening of the obstructing vaginal septum with drainage of hematocolpos.

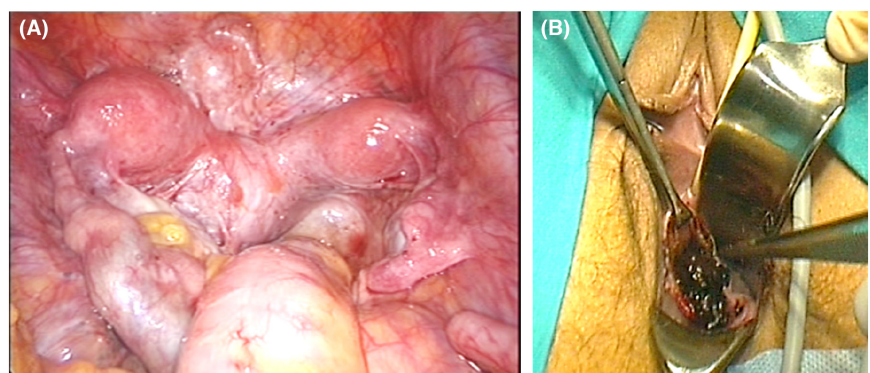


TABLE 2 Characteristics and symptoms of the patients in Group 2 (n = 5)

Mean age at presentation (year)	16.1	14.5–18
Mean time between menarche and presentation	3.5	1–6
Dysmenorrhea	5	100%
Recurrent pelvic pain	5	100%
Horn side (right/left)	3/2	60%/40%
Separated horn	3	60%
Non separated horn	2	40%
Urinary tract anomalies	1	Ipsilateral agenesis

3.2 | Group 2: non-communicating cavitated uterine horn in a unicornuate uterus

Patients in this group were diagnosed with a unicornuate uterus and a noncommunicating cavitated rudimentary horn (class IIB of The American Fertility Society classification).¹³ These cases, in the clinical-embryological classification, correspond to mesonephric anomalies 2.5, or may be isolated Müllerian anomalies 3.A.2.^{10,11} This was correctly diagnosed via ultrasound and confirmed via MRI in one case where the horn was widely separated from the

hemiuterus. The baseline characteristics and symptoms of the patients are presented in [Table 2](#) and [Figure 1B](#).

All patients in this group underwent laparoscopic removal of the rudimentary horn¹⁴ ([Figure 3](#)). In all cases the salpinx ipsilateral to the rudimentary horn was also removed, and in two of these cases a haematosalpinx had developed. In two cases, hemoperitoneum was observed, and in three cases, peritoneal foci of endometriosis were identified. All patients were discharged 48 hours after the procedure. Clinical evaluation 12 months after the procedure revealed a marked improvement of painful symptoms in all patients.

3.3 | Group 3: ACUM

This malformation in the embryological-clinical classification corresponds to 4. gubernaculum dysfunction 4.^{10,11} Two patients were evaluated for severe dysmenorrhea and cyclic pelvic pain associated with the presence of a cavitated myometrial mass ([Figure 1C](#)).

The first patient, aged 15 years, had recently been hospitalized for severe dysmenorrhea and fever. Transrectal ultrasound revealed, within a normal profile uterus, a round area (2.6 × 3.2 cm) located to the left of the endometrial cavity, slightly imprinting the uterine contour that was initially interpreted as a myoma with cystic degeneration. Subsequent MRI revealed a homogeneous hyperintense cavity content in T1-weighted sequences and led to a diagnosis of cystic adenomyoma.

The second patient, aged 16 years, was referred to our clinic for persistent dysmenorrhea and disabling postmenstrual pelvic pain, despite the use of non-steroidal anti-inflammatory drugs and oral contraceptives for approximately 8 months. She underwent abdominal/transrectal ultrasound and MRI. Imaging revealed a normal uterine contour and endometrial lining, with a hypoechoic nodule in the right myometrium measuring 3.5×2.8 cm.

Considering the severity of the symptoms, poor responsiveness to medical therapy, and imaging results, both patients underwent laparoscopic removal of myometrial neoforations. In both cases, the uterus was of normal conformation, with normal tube implantation and the presence of an anterior intramural subserosal mass to the left. The ovaries appeared normal and no endometriotic lesions were observed. Transcervical methylene blue dye test confirmed the patency of both tubes. Both the nodules were completely resected, and histological examination revealed the presence of a small endometrial cavity in both cases (Figure 4).

The patients were discharged on postoperative day 2. No intraoperative or postoperative complications occurred, nor did the patients complain of pelvic pain recurrence at the 18-month follow-up appointment.

3.4 | Group 4: transverse microperforate vaginal septum

Patients with partially obstructed vaginal septum in the clinical-embryological classification correspond to isolated Müllerian anomalies affecting the Müllerian tubercle 3.B.2 or segmentary atresias.^{10,11} A 17-year-old female patient was referred to our clinic for chronic dysmenorrhea; she was experiencing pain and difficulty in penetration during vaginal intercourse. Gynecological exploration performed during menstruation showed a transverse septum of the middle third of the vagina with a peripheral opening that appeared insufficient to allow a valid menstrual flow. By introducing a

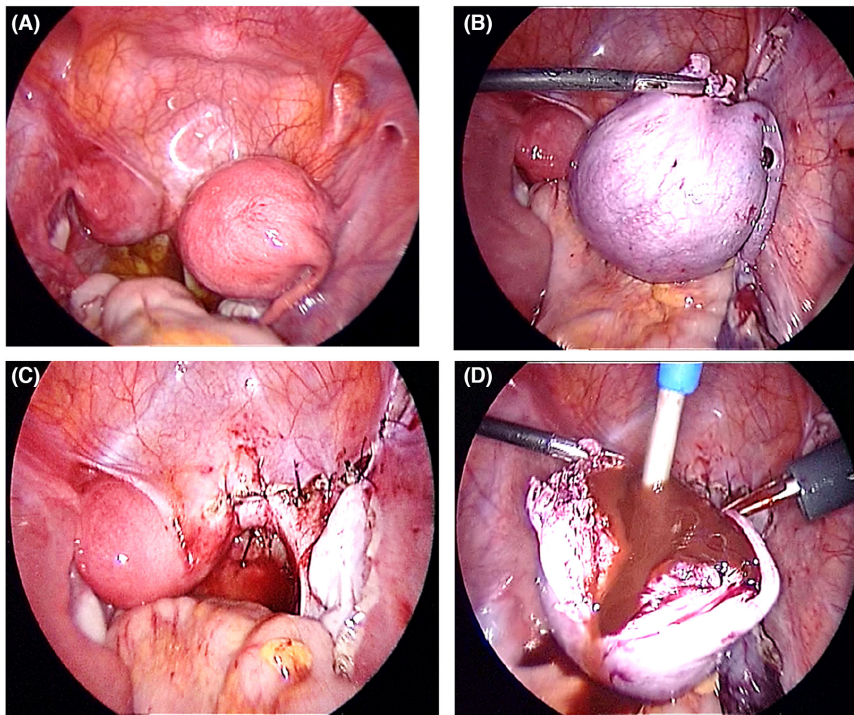


FIGURE 3 Left unicornuate uterus with right rudimentary horn. (A) Preoperative picture. (B) The rudimentary horn was freed from its vascular connections en bloc with the ipsilateral salpinx. (C) End of intervention. (D) The opening of the rudimentary horn highlights the presence of hematometra.

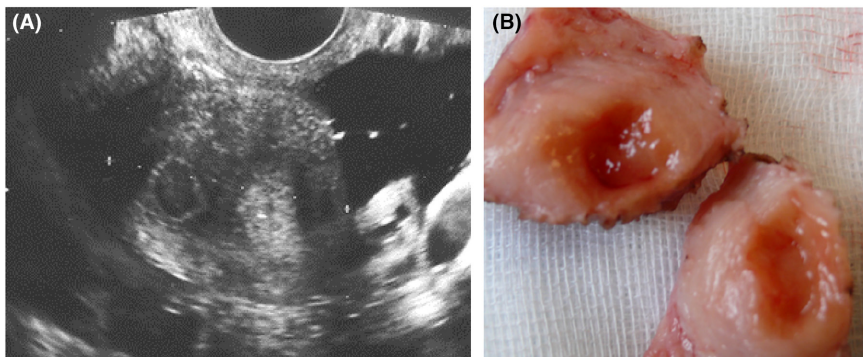


FIGURE 4 (A) Transvaginal ultrasound: the right-sided nodule with hypoechoic content. (B) Examination of nodule revealed the presence of a small endometrial cavity.

small catheter through this communication, vaginography was performed, which allowed measurement of the thickness of the septum and the presence of an undamaged upper vaginal third, with normal fornices. Vaginal surgery was recommended for septum removal, and the procedure was no pain on. Along with the removal of the septum, laparoscopy with dye test was performed, which revealed a normal uterus and adnexae and no visible endometriotic lesions. Transcervical dye testing confirmed the tube patency. No vaginal adhesions were observed 1 year after the operation, and the patient reported experiencing no pain on vaginal intercourse (Figure 1D).

4 | DISCUSSION

Dysmenorrhea and pelvic pain are common complaints of adolescent women.¹¹ In most cases it is a primary dysmenorrhea linked to the physiological production of prostaglandins and leukotrienes during menstruation. These patients usually respond well to over-the-counter analgesic regimens such as non-steroidal anti-inflammatory drugs or continuous oral contraceptive therapy.¹⁵ In cases where the symptomatology persists or does not significantly regress after at least three cycles of this therapy, instrumental investigations and laparoscopy are indicated to exclude the presence of Müllerian anomalies, endometriosis, or other organic pathologies. The incidence of “secondary” dysmenorrhea is low but varies depending on the diagnostic tools used and on the profession of the authors.^{16,17}

An analysis of over 10 years' data from our clinic indicates that almost 30% of adolescents referred for severe dysmenorrhea and pelvic pain resistant to medical therapy had an obstructive Müllerian anomaly. This percentage is surprisingly high; however, this is mainly associated with the fact that our center is a tertiary level unit for referring patients with suspected malformative pathologies of the female genital tract. However, it should be noted that we only considered teenagers, although many ORTAs become symptomatic even after the age of 20. In our study, endometriosis was only reported in association with Müllerian anomalies because laparoscopy, which is the only investigation capable of highlighting the minimal and mild stages of the disease that are typical of women below the age of 20, is not used as a primary diagnostic tool for endometriosis.

The subanalysis of our data, ie the percentage incidence of the various types of ORTA, has a more general value and compares well with other studies in the literature.^{18,19} Even in our study group, Herlin-Werner-Wunderlich syndrome was confirmed as the most frequent Müllerian obstructive anomaly in adolescents with dysmenorrhea not responding to medical therapy. The low incidence of this syndrome sometimes reported in the literature, is attributable to an often difficult diagnosis and to the lack of awareness of this condition.⁴ As a result, these young patients frequently undergo unnecessary interventions for pelvic mass leading to other gynecologic complications such as abscess formation and retrograde menstrual flow endometriosis. It should also be noted that some of the ORTAs, when the obstruction of the Müllerian half-axis is incomplete (communicating forms), may present even after 20 years of age. The same can be said of rudimentary

horns cavitated in unicornuate uterus. Indeed, many noncommunicating horns with endometrial cavity become clinically symptomatic during or after the third decade of life with dysmenorrhea.²⁰ However, the early onset of clinical symptoms is typical of ACUM. ACUM is a rare pathology, observed in young women, which causes nonresponsive severe dysmenorrhea and recurrent pelvic pain, and always begins early after menarche and persists during the postmenstrual period.²¹ ACUMs are problematic because of their broad differential diagnosis, which includes rudimentary cavitated uterine horns, adenomyosis, and degenerated leiomyomas. The criteria used for the diagnosis are as follows:²² (i) presence of an accessory intramyometrial cavitated mass, (ii) normal appearance of the uterus, fallopian tubes and ovaries, and (iii) the cavity must not communicate with the normal endometrial cavity. Modalities used to evaluate the uterine cavity include hysterosalpingography, saline infusion ultrasound and hysteroscopy. Currently, ACUM is considered a type variety of Müllerian anomaly, possibly related to dysfunction of the female gubernaculum, which was previously mistaken for juvenile cystic adenomyosis.²³

The use of laparoscopy has proved very useful in all cases. Surgery was almost always performed shortly after the diagnosis and investigation of the case. However, in some cases, delayed surgical management was preferred, given the patient's young age. In these cases, menstrual suppression is achieved with continuous oral contraceptives, progestogens or GnRH agonist injections.

5 | CONCLUSION

Unilateral obstructive anomalies of the female genital tract are difficult to identify. Therefore, it is important to maintain an elevated index of suspicion in high-risk groups. These conditions include severe unresponsive dysmenorrhea in postmenarchal girls and, above all, the presence of urinary tract malformations. Early diagnosis allows the preservation of reproductive activity and avoids potential serious complications.

AUTHOR CONTRIBUTIONS

FF developed the concept and wrote the article. EF and MP performed the outpatient follow-up examinations. SG participated in writing the article. FP analyzed the data. MC coordinated efforts. All authors contributed to editorial changes and approved the final article.

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CONFLICT OF INTEREST

The authors have stated explicitly that there are no conflicts of interest in connection with this article.

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