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Unresolving cyclic pelvic pain in young women due to rare structural uterine anomalies: a case series

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Abstract

Background There are many causes for cyclic pelvic pain unresponsive to anti-spasmodic medications in young women. An uncommon, although significant, association in patients presenting with such symptoms is structural uterine anomalies with rudimentary functional cavities. Ultrasound is the first-line modality for the investigation of dysmenorrhea, and the radiologist may be the first person to identify such an underlying condition in the patient. However, transabdominal ultrasound may provide only limited information. Magnetic resonance imaging (MRI) is the imaging modality of choice in such situations due to its high soft tissue resolution and multiplanar capability. This case series is unique in that it primarily highlights the MR imaging of rare structural uterine anomalies in three young females suffering from severe dysmenorrhea, along with an imaging approach.

Case presentation We present three cases of unresolving cyclic pain in young nulliparous females who were radiologically diagnosed with three rare uterine structural anomalies—accessory and cavitated uterine mass, Mayer–Rokitansky–Küster–Hauser syndrome, and functional non-communicating rudimentary horn in a hemiuterus or unicornuate uterus, which were treated with resection leading to symptom relief.

Conclusion In young women with recurrent cyclic pelvic pain, uterine structural abnormalities with functioning cavities should be considered as an etiology and evaluated with MRI. Owing to the non-specific nature of causes of dysmenorrhea, a schematic approach to imaging is essential for identifying the various underlying gynecological conditions, thus aiding in the prompt management of the patient.

Keywords Magnetic resonance imaging, Cyclic pelvic pain, Accessory and cavitated uterine mass, Mayer–Rokitansky–Küster–Hauser syndrome, Unicornuate uterus

Background

Müllerian duct anomalies may lead to non-resolving cyclic pelvic pain in young females if they involve some degree of outlet obstruction. The patients may present with primary amenorrhea or with regular menstrual cycles. Magnetic Resonance Imaging (MRI) plays a major role in the detection and characterization of these Müllerian duct anomalies and is the noninvasive imaging modality of choice. The currently followed classification system for congenital female genital anomalies has been proposed by the European Society of Human Reproduction and Embryology (ESHRE) and the European Society for Gynaecological Endoscopy (ESGE) [1], which is illustrated in Fig. 1.

Transabdominal ultrasound is the first-line radiological investigation for the evaluation of young women with cyclic pelvic pain. It effectively detects Müllerian anomalies, although the exact uterine zonal differentiation, structural anatomy of uterus, cervix, and vagina,

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UTERUS	MAIN CLASS	SUB CLASS
U0	Normal Uterus	
U1	Dysmorphic Uterus	a)T-Shaped b)Infantile c)Others
U2	Septate Uterus	a)Partial b)Complete
U3	Bicornuate Uterus	a)Partial b)Complete c)Bicorporeal septate
U4	Hemi Uterus	a)With rudimentary cavity (Communicating/not communicating) b)Without rudimentary cavity
U5	Aplastic Uterus	a)With rudimentary cavity (Communicating/not communicating) b)Without rudimentary cavity
U6	Unclassified malformations	

CO-EXISTENT CLASS (Cervical/Vaginal anomaly)	
C0	Normal cervix
C1	Septate cervix
C2	Double normal cervix
C3	Unilateral cervical aplasia
C4	Cervical aplasia
V0	Normal vagina
V1	Longitudinal non obstructing vaginal septum
V2	Longitudinal obstructing vaginal septum
V3	Transverse vaginal septum and imperforate hymen
V4	Vaginal aplasia

Fig. 1 European Society of Human Reproduction and Embryology and the European Society for Gynaecological Endoscopy (ESHRE/ESGE) consensus classification of Müllerian duct anomalies (Adapted from Reference 1)

the relationship of uterine buds with ovaries, and the status of fallopian tubes are demonstrated best on MRI. T2-weighted MR images depict the zonal anatomy of the uterus and shed details on whether the uterine cavity is functional. The MRI sequences need to be planned along the axis of the uterus. Alternatively, a 3-dimensional T2-weighted sequence of the pelvis can also be acquired. In addition, a coronal image with a large field of view is necessary to assess for the presence of any concomitant renal anomalies.

Accessory and cavitated uterine mass (ACUM) is an under-recognized uterine structural abnormality [2], with an accessory cavity containing functional endometrial and myometrial elements separate from the native endometrial cavity [3]. Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome occurs due to defective embryonic development of the paramesonephric ducts which leads to varying degrees of hypoplasia or aplasia of the Müllerian duct structures [4, 5]. Both conditions result in rudimentary uterus-like structures with variable stages of differentiation and pose a diagnostic dilemma to the radiologists as they may closely mimic a hemiuterus with a non-communicating rudimentary horn. However, this case series is unique in that it focuses on the MR imaging and a few tell-tale signs of this modality which help in the differentiation of these pathologies.

Case presentation

Case 1

A 17-year-old female presented in August 2021 with cyclic lower abdominal pain for the past 7 months. She had attained menarche at the age of 16 years. Hence, the pain was clinically diagnosed as primary dysmenorrhea and the patient was given anti-spasmodic medications, but they were not beneficial.

Subsequently, the patient underwent USG followed by MRI of the pelvis. On USG, there was a well-defined round thick-walled cystic lesion along the right lateral wall of the uterus with fine internal echoes. No communication with the uterine cavity could be demonstrated, and a possibility of a non-communicating rudimentary uterine horn was considered. MRI was done for further evaluation, which also demonstrated a well-defined, round, thick-walled peripherally enhancing cystic lesion along the right lateral wall of the uterus, at the site of insertion of the right round ligament. The wall of the cystic lesion appeared isointense to myometrium on T1W and hypointense on T2W sequences, while its contents were hemorrhagic (Fig. 2). It showed no communication with

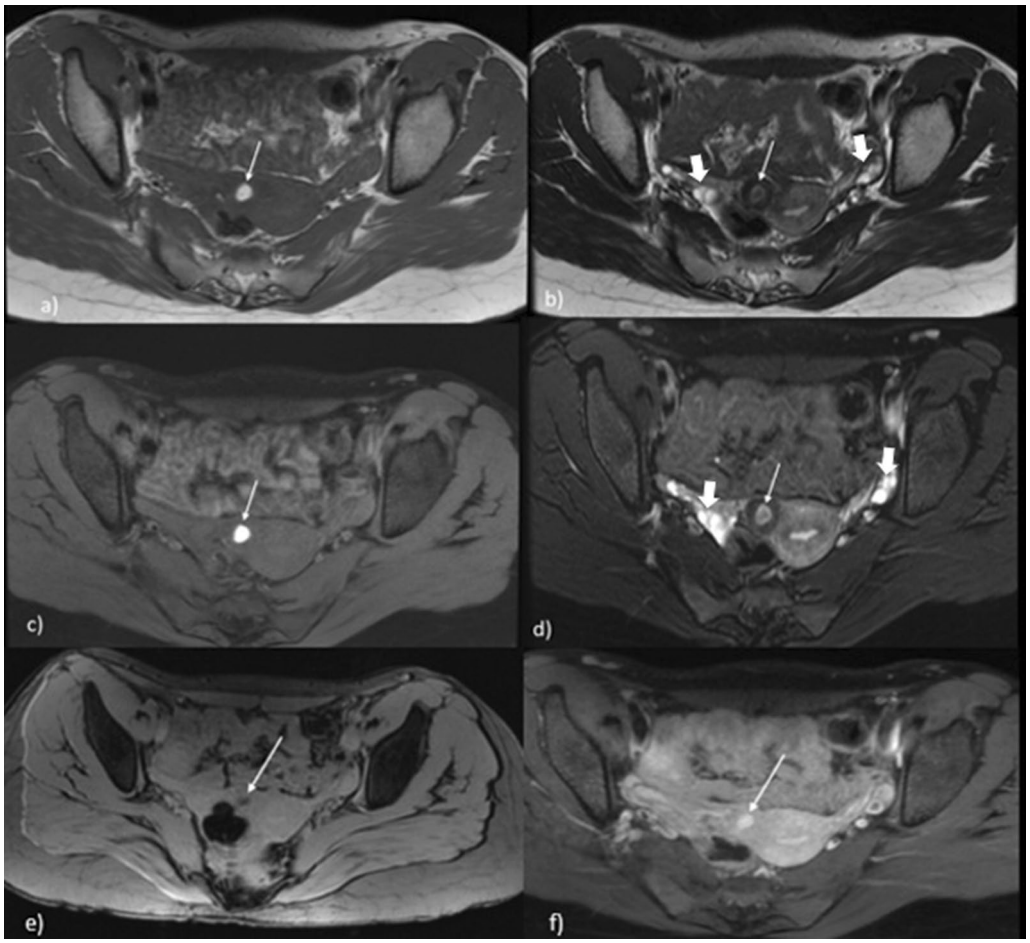

Fig. 2 MRI pelvis of a 17-year-old female with cyclic pelvic pain. Axial T1W SE image (**a**) shows a round hyperintense lesion (white arrow) in the right anterolateral myometrium, at the site of attachment of round ligament. Axial T2W SE image (**b**) reveals a thick-walled, round lesion with hypointense wall and shading of internal contents. Uterine endometrium is seen separately from the lesion. Both ovaries are normal (short white arrows). Corresponding axial T1W FS (**c**) and axial T2W FS (**d**) show no suppression of signal on fat saturated images, suggesting blood products. Axial T2* gradient-recalled echo (GRE) image (**e**) shows blooming of contents, confirming bleed. Axial T1W FS post-contrast image (**f**) shows isoenhancement of the wall relative to the myometrium. No enhancement of the intraluminal contents seen. Imaging features are suggestive of accessory and cavitated uterine mass (ACUM)

the endometrial cavity or any mass effect on the endometrium. The uterus was otherwise normal, and both horns were well developed. Both ovaries were also normal.

Given the typical radiological appearance and characteristic location of the cavity in an otherwise normal uterus, the diagnosis of ACUM (ESHRE/ESGE U6C0V0) was made. The patient underwent a laparoscopic tumorectomy of ACUM in December 2021 which led to complete symptomatic relief.

ACUM is myometrial-lined cavities located in the anterolateral aspect of the uterus, at the site of insertion of the round ligament, predominantly on the right side [2]. They are usually diagnosed in nulliparous females who are < 30 years of age [2]. According to a recent hypothesis, ACUM is thought to represent a new type of unclassified Müllerian duct, which is congenital in origin due to defective function of the female gubernaculum or ectopic Müllerian remnants [2]. In the appropriate clinical setting, ACUM can be diagnosed if it fulfills the following criteria: (i) an isolated accessory cavitated mass; (ii) a normal uterus (endometrial lumen), fallopian tubes, and ovaries; (iii) an accessory cavity lined by functional endometrium; (iv) blood degradation products in cavity; and (v) absence of adenomyosis in the rest of the uterus, although there could be small foci of adenomyosis in the myometrium adjacent to the accessory cavity [2].

The differential diagnoses of ACUM include:

Rudimentary and cavitated uterine horns seen in other uterine structural anomalies

Visualizing a normal uterus in ACUM as compared to the banana-shaped developed horn in a hemiuterus helps in differentiating between these two entities [6]. Similarly, rudimentary functioning horns in MRKH can be identified easily due to associated aplasia or hypoplasia of the uterus and other Müllerian duct derivatives.

Uterine leiomyoma with cystic or red degeneration

The larger size, multiplicity of lesions, and mass effect on the endometrial cavity usually help in the diagnosis [7]. However, a definite diagnosis may require laparoscopy and histopathology to identify the presence of endometrial lining in ACUM.

Focal cystic adenomyosis

This occurs in older multiparous women between 35 and 40 years, is usually ill-defined, and located away from the junctional zone, with no significant mass effect on the endometrial cavity. The rest of the uterus may show features of focal or diffuse adenomyosis [8]. In *cornual ectopic pregnancy,* although the location of the lesion may be similar to ACUM, MRI helps in accurately localizing the eccentric position of the gestational sac, surrounded by a thin rim (< 5 mm) of the myometrium and situated more than 1 cm from the lateral wall of the endometrial cavity [9]. Besides, the clinical presentation of secondary amenorrhea and urine pregnancy test positivity easily differentiates between the two conditions.

Case 2

A 14-year-old female presented to our department in February 2022 with primary amenorrhea and severe unresolved cyclic lower abdominal pain for six months. She had normal secondary sexual characteristics and hormonal assays.

The patient initially underwent USG of the pelvis. This showed a well-defined round to oval cystic lesion with thick walls in the right adnexa, along with a tubular cystic lesion with incomplete septation and internal echoes within it. No separate uterine structures could be demonstrated apart from the above-mentioned lesions. Subsequently, the patient underwent MRI, which showed a rudimentary horn in the right hemipelvis, with T1W and T2W hyperintense contents within the lumen, without signal suppression on T1FS images, suggestive of hematometra. A large tubular cystic lesion with incomplete septa was seen in the right adnexa abutting the right ovary, which had similar signal intensity contents, suggestive of hematosalpinx. There was complete aplasia of the cervix and vagina; however, both ovaries were normal (Fig. 3). No renal or lumbosacral vertebral anomalies were evident.

The case was diagnosed as atypical MRKH syndrome with an aplastic cervix, vagina, and a functional rudimentary right horn with hematometra and right hematosalpinx (ESHRE/ESGE U5aC4V4). The patient is currently awaiting laparoscopic removal of the functioning horn.

MRKH syndrome is suspected when a patient presents with primary amenorrhea with normal secondary sexual characteristics due to preserved ovarian function. MRI can demonstrate a spectrum of anomalies ranging from the complete absence of a uterus and upper 2/3rd of the vagina with ectopic ovaries, and rudimentary horns connected to each other by a fibrous cord, to the presence of a rudimentary, hypoplastic uterus with one to three layers of uterine differentiation [5]. The rudimentary uterus is defined as one which does not have conventional differentiation of corpus uteri and cervix uteri [5]. It can be located in the pelvis, mostly being attached to the ovary with a constant caudal relationship [5] and is well depicted on MRI unlike with transabdominal ultrasound.

Case 3

A 16-year-old female presented to us in October 2021 with complaints of non-resolving dysmenorrhea for the past 8 months with normal hormonal assay and secondary sexual characteristics.

Transabdominal USG demonstrated two separate uterine cavities, of which the right uterine cavity was filled with anechoic contents showing fine internal echoes. There was a well-defined tubular anechoic structure with incomplete septation seen in the right adnexa. A provisional diagnosis of hemiuterus with right hematometra and hematosalpinx was made. Subsequently, MRI of the pelvis of the patient revealed a banana-shaped uterus (hemiuterus) with normal zonal differentiation in the left hemipelvis, contiguous with a normal cervix and vagina. A large myometrial-lined cavity was seen in the right hemipelvis, which was connected to the left-sided hemiuterus by a fibrous band. This cavity contained heterogeneously hyperintense contents on T1WSE, T2WSE, and T1WFS (fat saturated) sequences, suggesting hematometra in a non-communicating, functional, rudimentary right horn. Right-sided hematosalpinx was also seen (Fig. 4). Bilateral ovaries were visualized separately and appeared normal.

Hence, the diagnosis of a hemiuterus with a non-communicating, functional, right rudimentary horn with hematometra and right hematosalpinx (ESHRE/ESGE U4aC0V0) was made. Surgical excision of the obstructed horn was done laparoscopically for the patient in

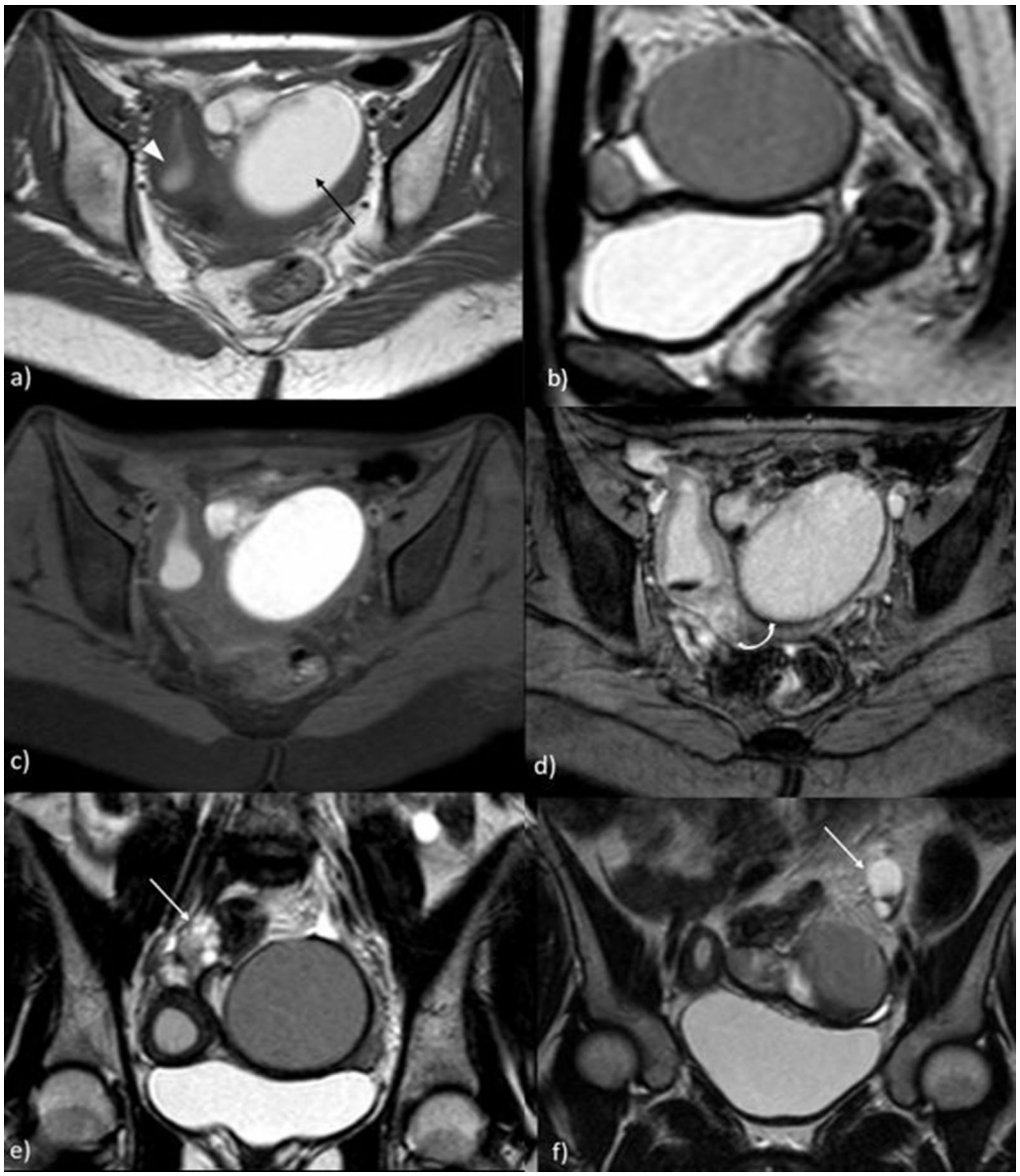

Fig. 3 MRI pelvis of a 14-year-old female with primary amenorrhea and cyclic lower abdominal pain. Axial T1W SE image (**a**) reveals a small rudimentary uterus in right hemipelvis, with hyperintense contents (white arrowhead) within the endometrial cavity and a large dilated right fallopian tube with hyperintense contents (black arrow). Mid-sagittal T2W SE sequence (**b**) depicts shading within the rudimentary horn and tube, with non-visualization of cervix and vagina. Axial T1 FS (**c**) shows no suppression of T1 hyperintense contents, confirming hematometra and hematosalpinx. Axial T2*GRE (**d**) shows hemosiderin lining of the hematosalpinx (curved arrow) and dependent blooming foci in the rudimentary horn. Cervix and vagina were not visualized on MRI, suggesting their aplasia. Right ovary (**e**) and left ovary (**f**) (white arrows) are normal. Features are consistent with Mayer-Rokitansky-Küster-Hauser syndrome with functional rudimentary right horn with hematometra, right hematosalpinx, and aplastic cervix and vagina

January 2022, followed by total relief from the debilitating symptoms.

Hemiuterus results from unilateral uterine development, with a rudimentary (functional) horn (Class U4a) or without rudimentary horn (Class U4b—non-functional or aplastic cavity) [1]. Various other conditions may result in similar clinical presentation, such as endometriosis, adenomyosis, and entities causing vaginal obstruction like obstructed hemivagina with ipsilateral renal anomaly (OHVIRA) syndrome [10]. A high index of clinical suspicion and a simple approach to imaging patients with recurrent cyclic pain enable the radiologist to clinch the diagnosis (Fig. 5).

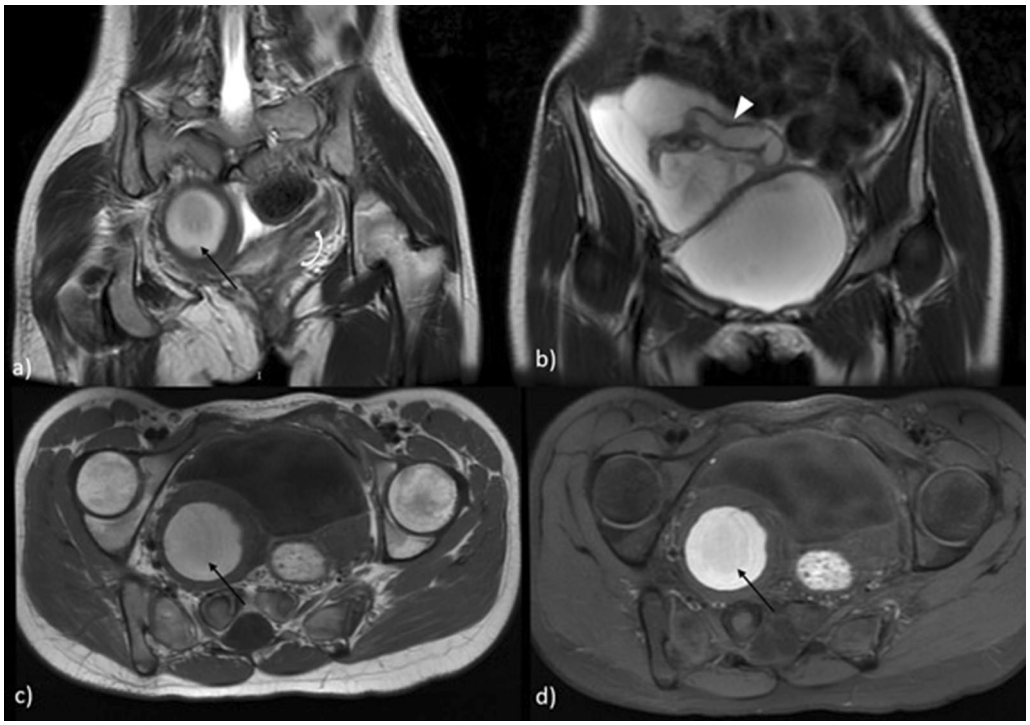

Fig. 4 MRI pelvis in a 16-year-old female presented with unresolving dysmenorrhea. Coronal T2W SE images (**a**, **b**) shows a small hemiuterus in the left hemipelvis (curved white arrow) with another separate myometrium-lined structure in the right hemipelvis with hyperintense contents (black arrow) within the lumen. A dilated tortuous right fallopian tube (white arrowhead) with hematosalpinx (confirmed on T1W images—not shown in image) is seen. Axial T1 SE image (**c**) and axial T1 FS (**d**) shows hyperintense contents within the structure in right hemipelvis suggesting a rudimentary horn with hematometra (black arrow). Features are consistent with a hemiuterus with a non-communicating, functional, rudimentary right horn

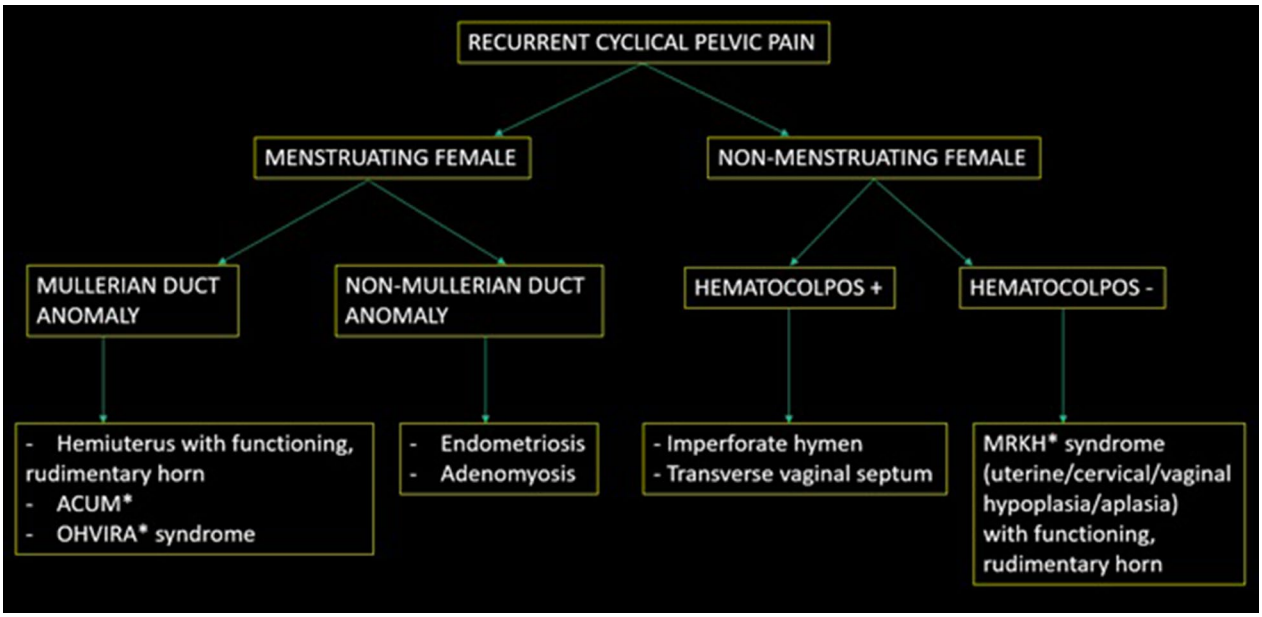

Fig. 5 Schematic approach to young women with recurrent cyclic pelvic pain. MRKH*-Mayer-Rokitansky-Küster-Hauser syndrome, ACUM*-Accessory and cavitated uterine mass (ACUM), OHVIRA*-Obstructed hemivagina with ipsilateral renal anomaly

Conclusions

In young women with recurrent cyclic pelvic pain, uterine structural abnormalities with functioning, endometrium-lined, obstructed cavities should be considered as an etiology and evaluated with MRI. Owing to the nonspecific nature of causes of dysmenorrhea, a systematic radiological approach in assessing the uterine, cervical, and vaginal morphology as well as the relation of the functional cavity with the uterus and ovaries is essential for identifying the various underlying gynecological conditions, thus aiding in the prompt management of the patient. Ultrasound may help in initial evaluation by detecting a non-communicating cavity; however, MRI plays a crucial role in the definitive identification of functioning rudimentary uterine horns or cavities, thereby guiding their surgical removal, which leads to complete resolution of symptoms [5].

Abbreviations

MRI	Magnetic resonance imaging
ACUM	Accessory and cavitated uterine mass
MRKH	Mayer–Rokitansky–Küster–Hauser syndrome
ESHRE	European Society of Human Reproduction and Embryology
ESGE	European Society for Gynaecological Endoscopy
OHVIRA	Obstructed hemivagina with ipsilateral renal anomaly
T1WSE	T1-weighted spin echo
T2WSE	T2-weighted spin echo
T1WFS	T1-weighted fat saturated

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Author contributions

VK contributed to concepts, design, definition of intellectual content, literature search, data acquisition, data analysis, manuscript preparation, and manuscript editing. AR was involved in concepts, design, definition of intellectual content, data acquisition, data analysis, manuscript preparation, manuscript editing, and manuscript review. SG contributed to data acquisition, data analysis, manuscript preparation, and manuscript review. AS was involved in data acquisition, data analysis, manuscript preparation, manuscript editing, and manuscript review. RGG contributed to data acquisition, data analysis, manuscript preparation, and manuscript review. All authors have read and approved the manuscript.

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Availability of data and materials

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

Declarations

Ethics approval and consent to participate

The consent for this report was obtained from the Institutional Ethics Committee, and informed consent was taken from the patients.

Consent for publication

Appropriate written consent to publish information presented in the manuscript was obtained from the participants' legal guardians as patients were minors.

Competing interests

The authors declare that they have no competing interests.

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