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Euro-Chinese consensus on accessory cavitated uterine malformation*,†

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ABSTRACT

Background: Accessory cavitated uterine malformations (ACUMs) are a rare obstructive uterine anomaly that remains poorly understood, posing challenges for clinical management. The aetiopathogenesis is hypothesised to involve the duplication and persistence of ductal Müllerian tissue usually near the round ligament attachment, potentially related to gubernaculum dysfunction. ACUM is specifically classified by Acién's system, though rare variants necessitate continued international research to refine classification frameworks.

Objectives: This consensus aims to develop good clinical practice recommendations for the pathophysiology, terminology, clinical presentation, diagnosis, and treatment of ACUM.

Methods: A working group consisted of Chinese and European experts, after approval from the European Society for Gynaecological Endoscopy, developed recommendations based on the best available evidence and experts' opinion.

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ABSTRACT

Results: Patients with ACUM present with typical symptoms such as dysmenorrhea and dyspareunia, and atypical symptoms, including gastrointestinal and generalised pelvic pain. Diagnostic criteria include isolated cavitated lesions in the anterolateral myometrium near the round ligament, lined by endometrial tissue and filled with haemorrhagic fluid, surrounded by a myometrial mantle with concentric orientation of myometrial fibres, and typically associated with a normal uterine cavity. Diagnosis is most accurately made through ultrasound and magnetic resonance imaging. Surgical excision of the ACUM is considered the definitive treatment offering near-complete symptom resolution, and minimally invasive approach should be preferred when possible. The timing of surgery and the interval before attempting pregnancy remain unclear. The mode of delivery post-surgery is individualised based on the degree of myometrial involvement.

Conclusions: The current consensus summarises the existing evidence on ACUM providing good clinical practice recommendations for their management. Existing gaps in the understanding and management of ACUMs, highlight the need for further research to guide clinical decision-making.

What is New? Good clinical practice recommendations for ACUM aiming to understand and optimise their management.

Keywords: Accessory cavitated uterine malformation, Müllerian anomalies, obstructive anomalies, adenomyotic cyst, cyclic pelvic pain, dysmenorrhea

Part I: Background

Embryology of the Internal Genitalia

The formation of the gonads begins as swellings located on either side of the dorsal mesentery, at the ventromedial surface of the mesonephros or Wolff's body. These protrusions form the gonadal or genital ridge as part of the primitive urogenital ridge.

During the sixth week, within the thickness of the urogenital ridge, the mesonephric excretory tubules converge in a mesonephric or Wolffian duct that descends to the cloacae, opening in the urogenital sinus. Meanwhile a longitudinal invagination of the coelomic epithelium is formed on the outer side of the urogenital ridge that originates the paramesonephric or Müllerian duct.[1]

This duct, at the top, opens into the coelomic cavity and descends in parallel and externally to the mesonephric duct. Then, both Müllerian ducts cross ventrally the mesonephric ducts, and grow in the caudomedial direction until fusing together and forming in the midline line a Y-shaped structure that is the uterine primordium, but without reaching the urogenital sinus (Figure 1).

Three portions can now be distinguished in the Müllerian ducts: a superior converging, a middle fused and an inferior diverging portion. The tubes come from the uppermost part of the Müllerian ducts, the converging portion, which remain separated and open into the coelomic cavity. The middle-fused parts of the paramesonephric ducts form the uterus, and the diverging portion forms the cervix up to the external cervical os.[2] And it's interesting to note that these different areas have also been related to different gene expressions of the HOXA family.[3]

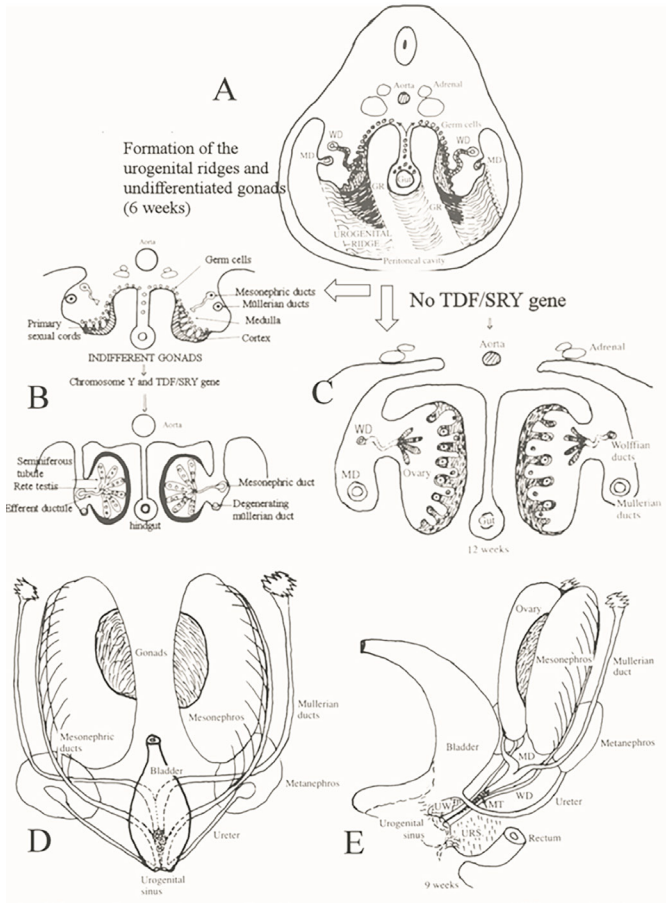

Figure 1. A) Urogenital ridge and undifferentiated gonads. B) Development of the gonads and Wolffian ducts in the male, and the Müllerian ducts in the female in (C). D) Development of the genital ducts in the female. The formation of the uterine primordia and opening of the mesonephric ducts to the urogenital sinus is shown. E) Lateral view showing the urorectal septum and the urogenital wedge (taken from Acién et al.[49] with permission).

When the ovary is being formed, and therefore testosterone and anti-Müllerian hormone (AMH or Müllerian inhibitor factor) are absent, the Wolffian ducts become atretic and regress cranially, and the Müllerian ducts develop. However, the adequate development and fusion of the paramesonephric ducts, the reabsorption of the middle septum and the correct formation of the normal uterus are induced by the laterally located mesonephric ducts. These fusion and reabsorption processes begin at the uterine isthmus (which is the most proximate part between both Müller ducts, right above the internal cervical os) and progress simultaneously, but independently, in both cranial and caudal directions, acting the mesonephric ducts as guide elements.[4,5]

The gubernaculum forms from the caudal fold that provokes the mesonephros, elevating the covering peritoneum (Figure 2). It begins as a muscular cord-like structure that extends from the abdominal wall

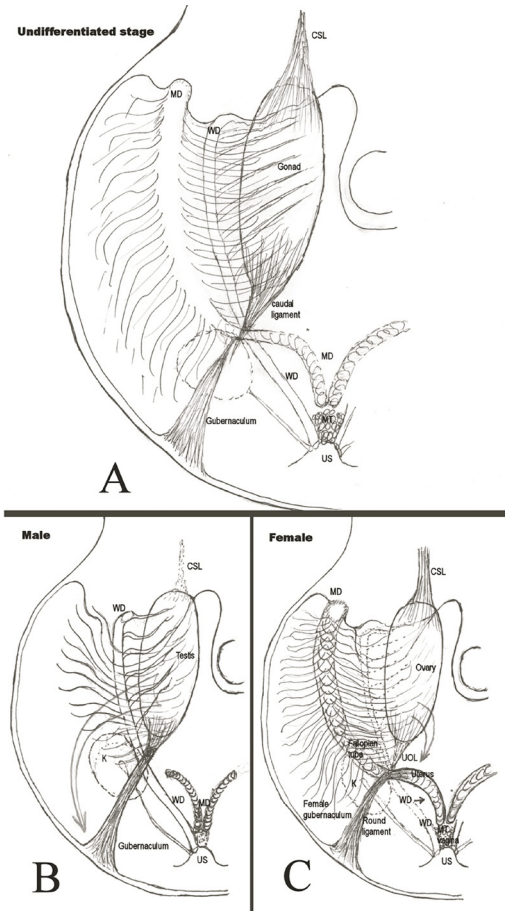

Figure 2. Schematic illustration of the possible development of the gubernaculum. A. At an Undifferentiated Stage. B. In Males. C. In Females. CSL, cranial suspensory ligament. WD, Wolffian duct. MD, Müllerian duct. MT, Müllerian tubercle. US, urogenital sinus. K, kidney. UOL, uteroovarian ligament (caudal ligament of the gonad) (taken from Acién et al.[7] with permission).

to the gonadal ridge. But the development of the paramesonephric or Müllerian duct interferes with the connection of this tissular column that has arisen between the inguinal cone and the caudal ligament of the gonad. Therefore, the gubernaculum then grows over the paramesonephric ducts, and its muscular fibres incorporate into the wall of the Müllerian ducts, becoming the round ligament. Behind and above, only atretic remnants of the mesonephric duct remain; and, the caudal ligament, uniting the gonad's inferior pole to the posterior wall of the Müllerian ducts, constitutes the utero-ovarian ligament.[6,7]

The female gubernaculum is likely formed by muscle fibres that are not of a mesonephric or paramesonephric origin, and their attachment to the Müllerian ducts allows the adequate development of the uterus. But the gubernaculum might also be responsible for many other specific human characteristics, including the uterus simplex, the anteflexion and low-intra-abdominal position of the uterus, and the disposition of uterine muscular fibres.[6,7]

Key question: What are the current theories on the aetiopathogenesis of accessory cavitated uterine malformation?

The pathogenesis of this entity is controversial. It is possible that the accessory cavitated uterine malformations (ACUMs) associated with an otherwise normal uterus should be considered Müllerian choristomas[8] as by definition the term refers to the growth of normal tissue at an ectopic location, thus suggesting developmentally misplaced Müllerian tissue. But where does this ectopic tissue come from and why?

During the eighth week of male embryo development, the production of testosterone and AMH begins. The consequence of this is that the mesonephric or Wolffian ducts develop while the paramesonephric or Müllerian ducts become atretic. Androgens, together with AMH and INSL-3 (insulin-like hormone), stimulate the growth of the tissular column which from the inguinal cone crosses the mesonephric or Wolffian duct to reach the caudal ligament at the inferior pole of the gonad. Thus, this third element in the crossing area, the gubernaculum, does not attach to the Müller duct and becomes the scrotal ligament, responsible for pulling down the gonad to the scrotum. Current thinking is that this process may be influenced by the cranial gonadal suspensory ligaments, hormones, genes as well as other factors[9] and its failure leads to cryptorchidism.

ACUM could be caused by the duplication and persistence of ductal Müllerian tissue in the critical area at the attachment level of the round ligament, possibly related to a gubernaculum dysfunction or abnormal traction and, as such, of congenital origin.[10] Alternatively, increased tension or traction of the gubernaculum could prevent fusion of the Müllerian ducts or traction of a hemi-uterus or rudimentary horn towards the inguinal duct and its herniation.[6,7] The observation of a tubal rudiment adjacent to the ACUM would speak in favour of detached Müllerian choristoma arising from abnormal gubernaculum traction in a female embryo.[11]

Key point

- *ACUMs could be caused by the duplication and persistence of ductal Müllerian tissue in the critical area at the attachment level of the round ligament, possibly related to a gubernaculum dysfunction or abnormal traction.*

Key question: What is the most appropriate terminology for accessory cavitated uterine malformation?

Various terms have been used for ACUM, including adenomyotic cyst, juvenile cystic adenomyosis, myometrial cyst, and uterine-like mass.

It has often been published under the term "Juvenile cystic adenomyoma", but it actually refers to the same pathology as the ACUM.

The Pros and Cons of using the words "malformation" and "mass" are shown in the Table 1.

We recommend using the word "malformation" rather than "mass". Not only is it a more accurate reflection of what ACUMs are, but the word "mass" implies uncertainty of the nature of a lesion which can lead to unnecessary concern over possible malignancy.

Key point

- *ACUM is the preferred terminology.*

Key question: How is accessory cavitated uterine malformation classified according to the existing classification systems?

Over the last two centuries, we have gained better knowledge on the embryology and pathogenesis of congenital malformations of the female genital tract. There have been different attempts to classify female reproductive tract anomalies.[6,7,12-20]

Despite many classification systems of female genital tract anomalies being available, Acién's proposal was the first and only one to include ACUM as a gubernaculum anomaly.

The classification system is based on the embryological development and the clinical presentation of the anomaly.

Key point

- *Acién's classification is the only system which specifically refers to the ACUM.*
- *Clinicians who care for patients with Müllerian anomalies should be mindful of the existence of other rare, unique and potentially very complex variants.*
- *Continued international efforts are needed to conduct high-quality studies that offer evidence-based data to improve the classification systems and their applicability in clinical practice.*

Part II: Clinical Presentation, Diagnosis and Differential Diagnosis

Key question: What are the typical and atypical symptoms in patients with accessory cavitated uterine malformation?

ACUMs were associated with substantial pelvic pain symptoms in all published cases. The most frequently reported symptoms are severe menstrual pain that can be central or ipsilateral to the side of the ACUM, and chronic pelvic pain. Other symptoms reported in the

Table 1. Pros and Cons of using the words "malformation" and "mass".

	Accessory cavitated uterine mass	Accessory cavitated uterine malformation
Pros	It highlights the fact that the anomaly looks like a tumour, plus it is often present in a uterus which is otherwise completely normal, making it different from other uterine malformations like rudimentary horns or the Robert's uterus	It highlights the fact that the anomaly is a malformation
Cons	A mass, can be benign or malignant	The term anomaly is preferred nowadays to refer to malformations plus, fewer papers are retrieved in PubMed using the term "mass"

literature include dyspareunia and hypogastric pain. The pelvic pain is thought to be caused by the accumulation of an increasing volume of menstrual fluid from the functioning endometrium lining the ACUM, within a cavity that has no outflow. The presumed mechanism for this causing pain is that it leads to increased pressure within the ACUM and subsequent stretching of the cavity.

Like other obstructive uterine anomalies, it tends to present in young women and girls. Nevertheless, while there are case reports describing diagnosis at as young an age as 13 years old,[21] the mean age at diagnosis in the larger case series' varies from 21 years old to 29 years old.[10,22-29] This most likely reflects the commonly experienced delays in reaching a diagnosis of ACUM, rather than being an accurate description of the onset of symptoms, which is classically described as starting with menarche or soon afterwards.

Key points

- *Typical symptoms: dysmenorrhea, dyspareunia and recurrent pelvic pain.*
- *Atypical symptoms: gastrointestinal pain and generalised pelvic pain, as can be seen in any chronic/ recurrent pain problem.*
- *ACUMs should be considered in all young women presenting with severe menstrual pain symptoms after menarche.*

Key question: What are the diagnostic criteria for an accessory cavitated uterine malformation?

ACUMs are almost certainly underdiagnosed, due to a lack of awareness by patients and clinicians, as well as the absence of widely agreed-upon diagnostic criteria. Failure to diagnose ACUMs will often condemn women to years of debilitating pain while trialling empirical, often ineffective, treatments. Many will undergo unnecessary investigations, procedures and operations in an attempt to diagnose and treat their pain. There may also be additional psychological consequences from experiencing ongoing, debilitating symptoms without a clear explanation. Failure to diagnose ACUMs denies women the opportunity for surgical excision, which in most cases substantially reduces or even completely cures the pain symptoms.

Several criteria have been proposed for the diagnosis of ACUMs, as detailed below.

Acién et al.[10]

1. Isolated accessory cavitated mass,

2. Normal uterus (endometrial cavity), tubes, and ovaries,

3. Surgical case with excised mass and with pathological examination,

4. Accessory cavity lined by endometrial epithelium with glands and stroma,

5. Chocolate-brown–coloured fluid content,

6. No adenomyosis (if uterus removed), but there could be small foci of adenomyosis in the myometrium adjacent to the accessory cavity.

Takeuchi et al.[22]

1. Solitary myometrial cyst measuring >1 cm surrounded by hypertrophic endometrium, independent of the uterine lumen,

2. Found in women ≤30 years of age,

3. Associated with severe dysmenorrhea.

Chun et al.[30]

1. Age of onset of severe dysmenorrhea within 5 years after menarche or ≤18 years of age,

2. No history of suspected endometrial or uterine injuries (delivery, myomectomy or dilatation and curettage),

3. Presence of a cystic lesion ≥0.5 mm indicated by imaging studies or observed during surgery.

Naftalin et al.[25]

1. Solitary cavitated lesion with a,

2. Myometrial mantle and,

3. Echogenic contents in the anterolateral wall of the myometrium beneath the insertion of the round ligament,

4. Ruling out obstructive congenital anomalies, such as communicating and non-communicating horns is crucial to diagnosis.

Timmerman et al.[31]

1. A uterine abnormality, presenting as a cavitated lesion surrounded by a myometrial mantle, in continuity with the anterolateral uterine wall, and located beneath the insertion of the round ligament and the interstitial portion of the fallopian tubes.

2. The appearance on imaging reflects the surrounding rim of functional endometrium and the haemorrhagic content of the cyst.

3. To distinguish ACUMs from other uterine abnormalities, a normal uterine cavity should be visualised.

There is, unsurprisingly, substantial overlap between these diagnostic criteria but there are specific criteria that apply to most cases but are not ubiquitous. Acién et al.'s.[23] criteria describe a normal uterus (endometrial cavity), tubes and ovaries as a criterion, although not indispensable. More specifically, they state that the patient must not have adenomyosis apart from small foci of adenomyosis surrounding the ACUM. Women may often however have coincidental uterine or ovarian pathology such as fibroids, a dermoid cyst or even adenomyosis elsewhere in the uterus that would not need to influence the diagnosis of an ACUM. Further, Acien's criteria include surgical excision of the ACUM for a definitive diagnosis, but there are increasing numbers of case descriptions of women with ACUMs not undergoing surgical excision. Increasing imaging quality means that the diagnosis can be confidently reached without a requirement for surgical excision. Nevertheless, as the original description of ACUMs, Acién et al.'s.[23] criteria have formed the basis of all the subsequent descriptions.

Both Takeuchi et al.[22] and Chun et al.'s[30] diagnostic criteria include stipulations about age. While these criteria help focus on the younger age group in which women with ACUMs frequently present, there are many case reports that describe women with ACUMs presenting outside of these age-related criteria. Chun et al.[30] go on to state that no history of suspected endometrial or uterine trauma should have occurred, including delivery. However, there are multiple case reports of women with ACUMs being diagnosed despite having had children or having previously undergone uterine surgery. More recent diagnostic criteria by Naftalin et al.[25] and Timmerman et al.[31] have focused more on the imaging appearance of ACUMs while still maintaining Acién et al.'s[23] original focus on the importance of ensuring that other uterine anomalies with similar appearances to ACUMs are excluded. These criteria have evolved over time as more has been learnt about ACUMs. Mindful that there remains a great deal about ACUMs that we do not yet know, it is important that diagnostic criteria account for this uncertainty and do not become overly prescriptive.

Key points

- *In order to diagnose an ACUM, the following criteria should be fulfilled:*
- *An isolated cavitated lesion located in the anterolateral myometrium, in the proximity of the round ligament.*
- *The cavity is lined by endometrial tissue and typically filled with haemorrhagic/menstrual fluid.*
- *The cavity is surrounded by a myometrial mantle with concentric orientation of myometrial fibres.*
- *They are typically associated with a normal uterine cavity.*

Additional notes

- While large ACUMs may enlarge to *involve* the posterolateral myometrium, because they are thought to originate from the gubernaculum, they should predominantly be within the anterolateral myometrium.
- ACUMs are found within the myometrium but the extent of their involvement with the myometrium can vary. They can be completely embedded within the myometrium or substantially outside the myometrium with minimal myometrial involvement. A grading system could be used to describe this based on the FIGO classification of type 4, type 5 and type 6 fibroids, as this will inform the extent of surgical dissection necessary, as well as the risk of uterine cavity breach.[32]

Key question: What are the diagnostic tools for diagnosing accessory cavitated uterine malformation?

Transvaginal ultrasound is the primary diagnostic tool in gynaecology and, in expert hands, is sufficient to diagnose ACUMs with confidence. Nevertheless, not all gynaecologists or sonographers will have the experience or confidence to diagnose ACUMs on ultrasound alone. Magnetic resonance imaging (MRI) is important in these circumstances, and with expert radiologists, the diagnosis can be made with confidence. Furthermore, given that the population in which ACUMs will be suspected often includes young women and girls in whom transvaginal ultrasound might not be appropriate, MRI should be considered in preference to transvaginal ultrasound. Consideration can also be given to transrectal ultrasound, which gives equivalent views to transvaginal ultrasound, although it may also not be appropriate or considered acceptable by the patient.

Ultrasonography

On ultrasound, ACUMs are visualised as cavitated lesion with a myometrial mantle and echogenic contents seen in the antero-lateral wall of the myometrium or within the broad ligament (Figure 3). While the myometrial mantle will likely be of similar echotexture to the surrounding myometrium, the concentric orientation of its muscle

fibres means that it can be clearly distinguished from it. The endometrial lining will often be visible. The fluid within the cavity should be echogenic and is most often seen as being of "ground glass" echogenicity, equivalent to the altered blood content seen in endometriomas. The contents have also been reported as hyperechogenic.

3D ultrasound can also be useful in confirming the diagnosis, although it can be difficult to get a single clear image of both the uterine cavity and the ACUM within it, because they are rarely in the same plane. Coronal 3D ultrasound image should reveal a circular cavity adjacent to the otherwise normal uterine cavity with no communication between the two cavities (Figure 4). 3D ultrasound is also crucial to excluding other uterine anomalies, and so in women with ACUMs, the main uterine cavity will be visible with both uterine horns.

Key point

- *In expert hands, the diagnosis of ACUM can confidently be made on transvaginal or transrectal ultrasound.*

Magnetic resonance imaging

On MRI, ACUMs will be seen to have a central cavity, surrounded by a well-defined ring with low T1 and T2 signal enhancements, which is similar to that of the junctional zone (Figures 5, 6). The surrounding myometrial mantle has been described as thickened and hypointense on T2-weighted images, which demonstrates myometrial hypertrophy. In addition, the cavities had a thin inner lining

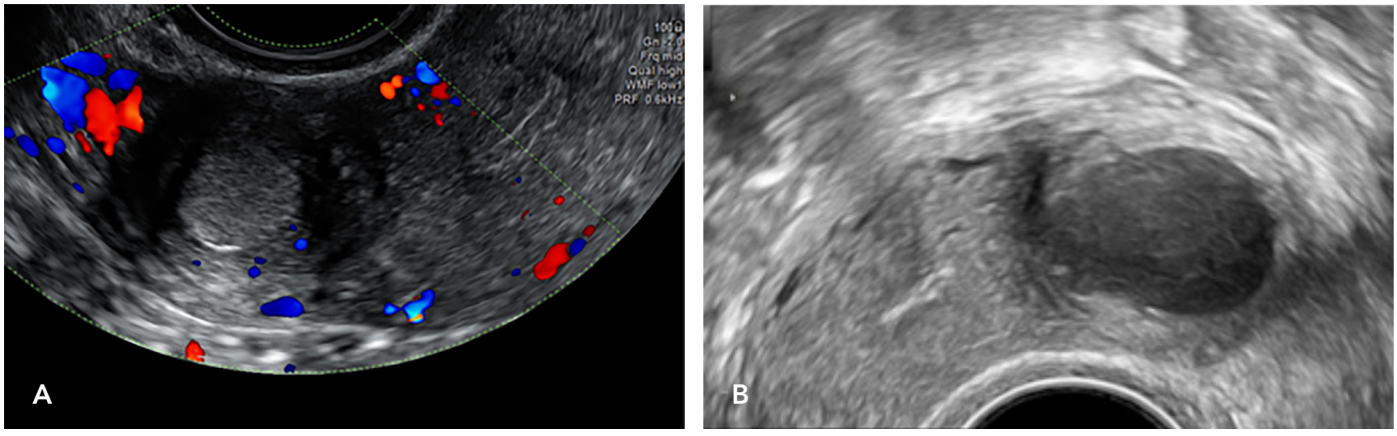

Figure 3A, B. 2D Ultrasound images of ACUMs in the left lateral myometrium showing the myometrial mantle and haemorrhagic content that can be hyperechogenic **(A)** or "ground glass" in appearance **(B)**.

ACUM: Accessory cavitated uterine malformation.

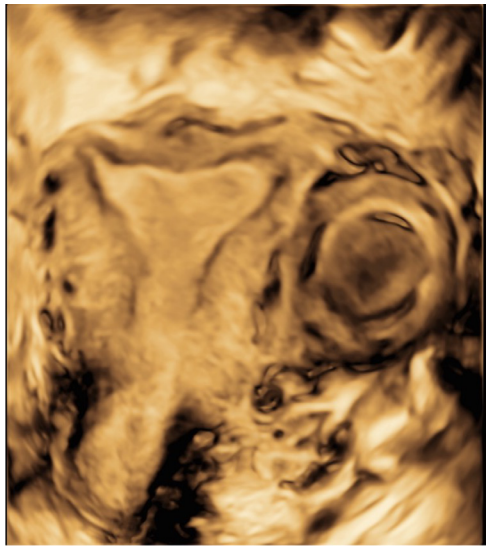

Figure 4. 3D ultrasound image of ACUM in the left lateral myometrium.

ACUM: Accessory cavitated uterine malformation.

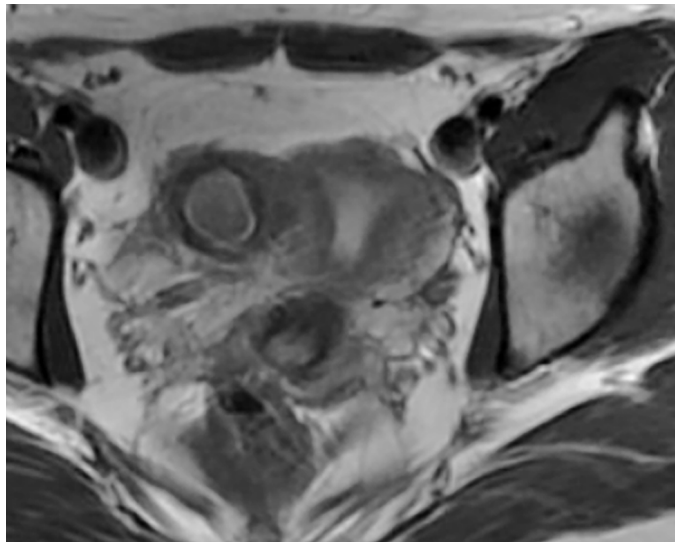

Figure 5. MRI of ACUM in the right lateral myometrium.

MRI: Magnetic resonance imaging, ACUM: Accessory cavitated uterine malformation.

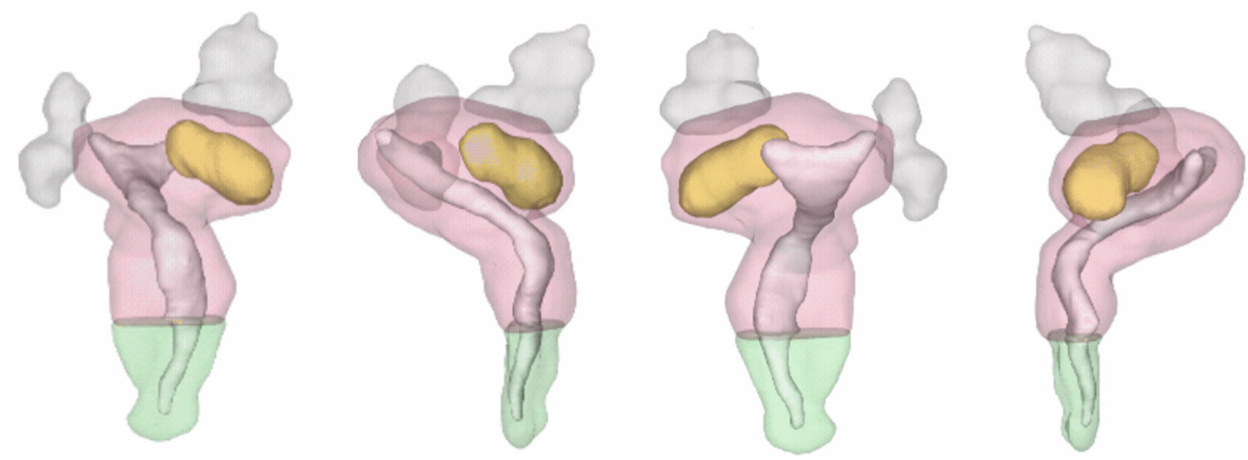

Figure 6. Three-dimensional reconstruction of ACUM based on MRI.
ACUM: Accessory cavitated uterine malformation, MRI: Magnetic resonance imaging.

that moderately enhanced after gadolinium contrast and appeared hyperintense on T2-weighted images, indistinct from endometrium. The internal content of the cavities displays high T1 signal intensity, which persists after fat saturation and is indicative of haemorrhagic content. Some lesions will demonstrate T2 shading, which is seen in ovarian endometriomas.

Key point

- *If there is diagnostic uncertainty after ultrasound examination, consideration should be given to using MRI.*

Histology (microscopy)

Microscopically, the cavity of the lesion is lined with functional endometrium consisting of glands and stroma and blood is seen within the cavitation (Figure 7). Studies report that the endometrial tissue within ACUMs positively stains for CD10, oestrogen receptors (ER) and progesterone receptors (PR), which are markers of normal endometrium. They also reported that the myometrial mantle of the ACUMs contained smooth muscle cells that stained positive for desmin, ER, and PR. The myometrium surrounding the cavitated lesion may be hypertrophic and will often contain foci of adenomyosis.

Key point

- *Not all histopathologists are familiar with ACUMs so it is important to let them know what that you suspect an ACUM and ensure they are aware of their histopathological features.*

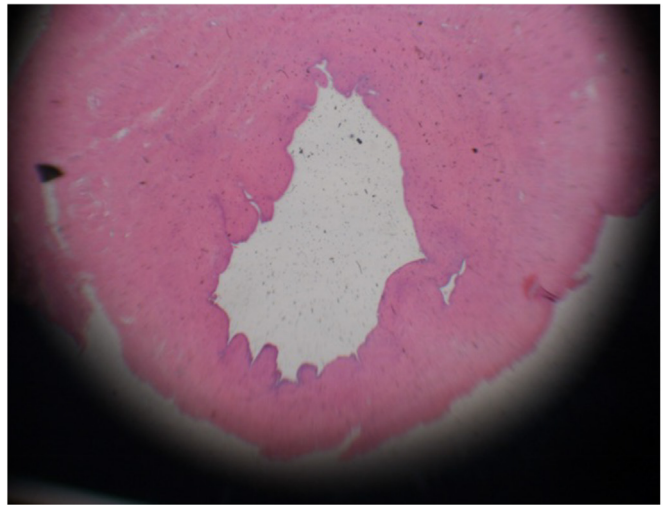

Figure 7. Microscopic pathologic image of ACUM.
ACUM: Accessory cavitated uterine malformation.

Differential diagnoses

Obstructive congenital uterine anomalies are key differentials of ACUMs and, therefore, excluding them is crucial to making the diagnosis. Regardless of the imaging modality used, it is important to demonstrate that there is no connection to either the uterine cavity or to the Fallopian tubes and that there are two normal interstitial portions of the Fallopian tubes. Those without sufficient expertise in ultrasound and MRI may consider other more invasive investigations, such as saline infusion sonography (SIS), hysterosalpingography (HSG), hysterosalpingo sonography using saline or foam (HyCoSy/HyFoSy) or hysteroscopy, to exclude other congenital anomalies, but these modalities should only be required in rare circumstances.

Other important differentials of ACUMs include cystic adenomyomas, unicornuate uteri with functioning rudimentary horns, complete septate uterus with unilateral cervical aplasia (Robert's uterus) and degenerating fibroids. It is particularly important to differentiate these entities because the management options and strategies vary greatly. Knowledge of their different features can help distinguish them (Table 2). Other potential differentials that could be confused with ACUMs are endometriomas that are adherent to the lateral aspect of the uterus and, more rarely, ectopic pregnancies, including interstitial, intramural and rudimentary horn pregnancies.

Key point

- *As exclusion of other uterine anomalies is crucial to the diagnosis, in the rare circumstances where ultrasound and MRI have failed to clarify the morphology of the uterine cavity, consideration could be given to more invasive tests such as saline infusion sonohysterography, HyCoSy/HyFoSy or hysteroscopy.*

Part III: Treatment and Counselling

Key question: What are the clinical indications and available treatment options for accessory cavitated uterine malformation?

Treatment aims to alleviate pain and to restore normal anatomy. Reported ACUM management options range from medical treatment to surgery. Factors that influence decision-making include the severity of symptoms, age, and patient preferences.[25,33,34]

Surgical treatment

Surgery is considered the definitive treatment for ACUM and has shown excellent results in symptom relief, pregnancy prognosis and long-term management.[10,22-25,28,34-36] There is no direct evidence to guide the timing of ACUM surgery or on the role of preoperative gonadotropin-releasing hormone (GnRH) agonist therapy.

Table 2. Features of accessory congenital uterine malformations and relevant differential diagnoses.

	Accessory cavitated uterine malformations	**Cystic adenomyomas**	**Rudimentary horn with functioning endometrial cavity**	**Complete septate uterus with unilateral cervical aplasia (Robert uterus)**	**Degenerated fibroids**
Location	Located in the anterolateral myometrium, in proximity to the round ligament	Located in the myometrium	Located at the lateral cornual aspect or lateral and distinct from the myometrium	Located in the lateral aspect of the uterus with a thin septum between cavities that can be bulging	Can be located anywhere in the myometrium
Pathophysiology	Have a myometrial mantle and endometrial lining	Absence of myometrial mantle	No myometrial mantle	Will not have a myometrial mantle that is distinct from the surrounding myometrium	Fibroid pseudo capsule and heterogeneous aspect
Relation to the uterine corpus	Typically bulges outside the uterine corpus	Typically, entirely within the myometrium	It can be separate from the main uterine body or not, but the uterine cavity will be unicornuate.	Typically, within the uterine corpus	Could be within the body of the uterus or pedunculated
Content	Typically, echogenic cavity content	Often anechoic cavity content	There is usually not hematometra found because the content refluxes into the abdomen	Typically, echogenic content	Can be echogenic or anechoic content
Age	Commonly found in young women or teenagers	Commonly found in older parous women	Can be found in any age group	Commonly found in young women or teenagers	Can be found in any age group

Surgical approaches include laparoscopy, robot-assisted laparoscopy, and laparotomy[10,23,37] and they involve the excision of the ACUM.

Irrespective of the surgical approach, a systematic approach is required. Firstly, incision and circumferential enucleation of the ACUM is performed with or without preceding injection of dilute vasopressin along the uterine-ACUM interface for haemostasis. It should be noted that finding the cleavage plane can be challenging, as the typical pseudocapsule present in fibroids will not be found in ACUM. Assisted by ancillary instruments, such as a tenaculum or suction device, the procedure is completed by transecting the ACUM from its attachment and closing the uterine defect with sutures.

Considering that ACUM is a benign condition, its contents are not thought to pose any threat if they leak. Thus, various techniques, such as morcellation and specimen retrieval in endo bags, have been described for removing specimens of ACUM.[31]

Adhesion barrier agents can be used during surgery to prevent postoperative adhesions.[35]

The boundaries of an ACUM can sometimes be imprecise and so some authors have described using intraoperative ultrasound to help with lesion localisation and excision,[31] also using intraoperative 3-dimensional ultrasonography, which not only can clearly locate the nodule but also show the thickness of the myometrium overlying the cystic cavity.[36]

For older patients who do not desire future pregnancies, hysterectomy may be recommended as it offers permanent relief from dysmenorrhea. Total laparoscopic hysterectomy is a common, safe, and minimally invasive option for women with benign gynaecological conditions like ACUM.[33]

Medical treatment

Medical treatments for ACUM generally focus on pain relief and symptom management and are based on non-steroidal anti-inflammatory drugs (NSAIDs) and analgesics or on hormonal treatments [include continuous oral contraceptive pills (OCPs)],[34-43] the levonorgestrel-releasing intrauterine system (LNG-IUS, e.g., Mirena), and gonadotropin-releasing hormone agonists (GnRHa).[22,38]

These therapies may temporarily reduce symptoms, allowing patients to defer or avoid surgery or to help them manage their symptoms while awaiting surgery.[44] It is unclear whether factors such as age at the start of treatment, the size of the ACUM, or other morphological characteristics influence the success of medical treatment.[31] However, if medical treatment is not effective, conservative minimally invasive surgery should be considered, always considering fertility preservation in young patients.[40]

Sclerotherapy and radiofrequency ablation

An alternative treatment is sclerotherapy with ethanol, as described by Merviel et al.[45]

The procedure typically involves general anaesthesia and ultrasound guidance to insert a needle through the vaginal wall and into the ACUM. After aspirating the cyst's contents, 96% ethanol is injected to fill the cavity for about 15 minutes, then drained.

This method can offer temporary relief from symptoms but is rarely a permanent solution. Risks include leakage of the sclerosing agent into the peritoneal cavity.

More recently, lauromacrogol has also been introduced as a sclerosing agent for ACUM.[46] This compound offers the dual benefits of sclerotherapy and local anaesthesia, although its long-term efficacy and safety remain under investigation.

Radiofrequency ablation has also been used with similar results as ethanol sclerotherapy in terms of symptom relief.[47,48]

Key points

- *Treatment aims to restore uterine anatomy through excision of the ACUM and to alleviate symptoms.*
- *ACUM typically requires treatment in case of severe dysmenorrhea or chronic/recurrent pelvic pain.*
- *Surgical management, consisting of ACUM removal from the myometrium and suturing of the uterine defect, is the definitive treatment, and it has shown nearly complete remission of symptoms. Options include laparotomy, laparoscopy, and robot-assisted laparoscopy. A minimally invasive approach should be preferred when possible.*
- *Medical management including administration of NSAIDs, OCP, LNG-IUS, and GnRH agonists. Medical management provides temporary relief but is often not a definitive solution.*
- *Sclerotherapy is an alternative for those who wish to avoid surgery, though it may lead to recurrence.*

Key question: What are the optimal skills and facilities to remove the accessory cavitated uterine malformation while protecting the uterine myometrium wall?

The surgical management of ACUM requires a meticulous approach to achieve complete lesion excision while preserving myometrial integrity. Optimal outcomes depend on precise surgical techniques, surgeon expertise in minimally invasive gynaecologic surgery, and selective use of intraoperative imaging guidance when necessary.

Key points

- *Ability to accurately estimate the penetration depth in the myometrium to remove the lesion while minimising risks.*
- *Use of intraoperative ultrasound, including 3D ultrasonography, for precise localisation and excision of the ACUM.*
- *Surgical skills and experience to apply proper surgical techniques including careful enucleation of the ACUM using mechanical, monopolar or bipolar energy, with various tools assisting in the dissection and suturing (especially for laparoscopy/robotics).*

Key question: What is the best timing of surgery for accessory cavitated uterine malformation?

The optimal timing for surgical intervention in ACUM remains poorly defined due to limited evidence, but clinical decisions should prioritise symptom severity, reproductive goals, and patient priorities.

Key point

- *There is no direct evidence to guide the timing of ACUM surgery.*

Key question: What is the recommended interval before attempting pregnancy after surgery?

There is no direct evidence to guide decision making on the interval before embarking on pregnancy.

Key point

- *After surgery for ACUM, a recommended waiting period of 4-6 months is advised before attempting pregnancy; this allows the proper healing of the myometrium.*

Key question: What is the recommended mode of delivery for future pregnancy?

The mode of delivery following ACUM excision lacks standardised guidelines due to insufficient outcome data, necessitating individualised decision-making based on surgical characteristics and obstetric context. Delivery planning should account for the depth of myometrial resection during ACUM excision, analogous to the FIGO classification for fibroids (e.g., FIGO type 4–5 lesions involving >50% myometrial thickness may warrant heightened surveillance for uterine rupture).

Key point

- *There are no data to determine the optimal mode of delivery after ACUM excision. Caesarean sections and vaginal deliveries are described in literature. In determining the mode of delivery after ACUM excision, consideration should be given to the depth of myometrial involvement/FIGO type of ACUM.*

Conclusion

There remains a great deal that is unknown about ACUMs, which provides challenges for clinicians managing patients with this malformation. The embryological origin remains unclear, although it´s possibly related to a gubernaculum dysfunction or abnormal traction. Existing classification systems, except for Acién´s, while widely utilised, do not adequately incorporate ACUM. To diagnose an ACUM, the following criteria should be fulfilled: to be an isolated cavitated lesion located in the anterolateral myometrium, in the proximity of the round ligament, with a cavity lined by endometrial tissue and typically filled with haemorrhagic/menstrual fluid. They should be surrounded by a myometrial mantle with typical concentric orientation of the myometrial fibres, with a normal uterine cavity. Current management strategies prioritise complete surgical excision of the lesion, preferably via minimally invasive techniques, to achieve symptom resolution. There is a paucity of high-quality evidence to guide clinical decision-making regarding aspects of optimal surgical intervention and, more specifically regarding the management of future pregnancies.

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