

SURGICAL APPROACHES TO CYCLIC PELVIC PAIN IN MAYER–ROKITANSKY–KÜSTER–HAUSER SYNDROME

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Abstract

Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome is a rare congenital disorder characterized by the absence or underdevelopment of the uterus and the upper two-thirds of the vagina in individuals with a normal female karyotype (46,XX) and fully functioning ovaries. Although primary amenorrhea is often the initial clinical manifestation leading to diagnosis, a subset of patients with MRKH syndrome present with cyclic chronic pelvic pain. This symptom is typically associated with the presence of rudimentary uterine structures containing functional endometrial tissue, which responds to hormonal fluctuations, resulting in hematometra, inflammation, and fibrosis within the pelvic cavity.

The management of cyclic chronic pelvic pain in MRKH syndrome presents significant clinical challenges, particularly due to the anatomical variability of the rudimentary structures and the risk of misdiagnosis or delayed treatment. Imaging modalities such as MRI and transabdominal or transrectal ultrasound are critical for the identification of endometrial cavities in non-communicating uterine remnants. Surgical intervention remains the primary therapeutic approach in symptomatic cases. Among the surgical options, endometrial ablation (cauterization) of the rudimentary uterus has emerged as an effective method for eliminating functional endometrium, thereby alleviating pain and reducing the risk of endometriosis and future recurrence.

This article provides a comprehensive overview of the etiopathogenesis, clinical presentation, diagnostic workup, and therapeutic strategies for cyclic pelvic pain in MRKH patients, with a special focus on the role of endometrial cauterization. Furthermore, the article underscores the importance of a multidisciplinary approach involving gynecologists, radiologists, surgeons, psychologists, and pain management specialists to ensure timely diagnosis, individualized treatment planning, and holistic patient care. Recognizing and addressing chronic pain in this population not only improves quality of life but also mitigates the psychological burden associated with this complex congenital condition.

Keywords: Mayer–Rokitansky–Küster–Hauser syndrome, MRKH, chronic pelvic pain, rudimentary uterus, endometriosis, endometrial ablation, multidisciplinary approach, uterine agenesis, pelvic pain syndrome, reconstructive gynecology.





Introduction

Mayer–Rokitansky–Küster–Hauser syndrome (MRKH) is a rare congenital anomaly of the Müllerian ducts, characterized by the absence or severe underdevelopment of the uterus and the upper two-thirds of the vagina in women with a normal 46,XX karyotype. Despite the absence of a uterus, patients typically exhibit normal development of secondary sexual characteristics and have functioning ovaries. The prevalence of MRKH is estimated at approximately 1 in 4,500 to 5,000 female births.

One of the complications seen in a subset of patients with MRKH is cyclic chronic pelvic pain (CPP), which is often associated with functioning endometrial tissue located in rudimentary uterine structures. These patients may present with pelvic pain that occurs in sync with the menstrual cycle, despite the absence of menstruation (primary amenorrhea). The pain is usually due to the accumulation of menstrual-like blood in non-communicating uterine remnants, resulting in hematometra or functional obstruction, and may be further exacerbated by secondary endometriosis due to retrograde bleeding.

Pathophysiology of Pelvic Pain in MRKH Syndrome

The pathogenesis of cyclic CPP in MRKH syndrome is most commonly linked to the presence of functional endometrial tissue in rudimentary uterine horns. These structures, although not connected to the vaginal canal, may still contain endometrium that responds to hormonal stimulation. The accumulation of menstrual blood in these non-draining cavities leads to hematometra, inflammation, and pressure on adjacent tissues, causing significant cyclic or even persistent pain.

Furthermore, retrograde menstruation from these rudimentary horns can result in secondary pelvic endometriosis, further intensifying the pain and contributing to infertility in cases of associated anomalies. In rare instances, obstructed rudimentary horns can also cause infections or abscesses.

The clinical presentation can be misleading, as these patients lack outward menstrual bleeding. They may instead report monthly lower abdominal or pelvic pain, frequently misdiagnosed as gastrointestinal or musculoskeletal in origin. Careful gynecological evaluation is critical for early diagnosis and management.

Diagnostics

The diagnostic approach for CPP in MRKH syndrome begins with a detailed clinical history, emphasizing the timing and nature of pain, and any association with cyclic hormonal changes. A gynecological examination may reveal vaginal shortening or agenesis, leading to suspicion of a Müllerian anomaly.

Ultrasound imaging (transabdominal or transrectal) can detect the presence of pelvic masses or rudimentary uterine horns. However, the gold standard for anatomical assessment is magnetic resonance imaging (MRI). MRI provides detailed visualization of uterine remnants, endometrial tissue, and any associated renal or skeletal anomalies (which are common in MRKH type II).

In some cases, diagnostic laparoscopy is performed to confirm the presence of endometrial tissue and rule out other causes of pelvic pain such as adhesions or endometriosis. Laparoscopy also allows for simultaneous therapeutic intervention.





Treatment: Endometrial Ablation of Rudimentary Uterine Horns

The cornerstone of treatment for CPP in MRKH patients with functioning endometrial remnants is surgical intervention. The primary goal is to eliminate the source of pain while preserving surrounding structures and avoiding unnecessary radical surgery.

Endometrial ablation (coagulation or electrofulguration) is a minimally invasive, fertility-sparing approach, especially valuable in:

- Patients not suitable for full horn resection.
- Patients with inaccessible or deeply located rudiments.
- Those undergoing concurrent vaginoplasty.

During laparoscopic surgery, bipolar or monopolar coagulation is used to ablate the functional endometrium lining the rudimentary horns. This results in fibrotic transformation and cessation of bleeding activity from the endometrial tissue.

In our study, we applied bipolar electrocoagulation to endometrial zones under laparoscopic visualization. Special care was taken to avoid thermal injury to adjacent organs such as bladder, ureters, and bowel. In some cases, the procedure was combined with neovaginoplasty using techniques such as Vecchietti or McIndoe procedures.

Materials and Methods

We observed a cohort of patients with confirmed MRKH syndrome and cyclic pelvic pain. All had imaging evidence of non-communicating rudimentary uterine horns with functional endometrium.

- Sample size: 14 patients aged 14–23.
- Diagnostic tools: MRI, ultrasound, and laparoscopy.
- Intervention: Laparoscopic bipolar endometrial ablation.
- Evaluation tools: Visual Analogue Scale (VAS) pre- and post-operatively.
- Follow-up: 6 to 12 months post-procedure.

Results

Following surgical ablation:

- 92% of patients reported significant pain reduction, with average VAS scores decreasing from 7.8 to 2.1.
- 6% (1 patient) achieved complete pain relief.
- 1 patient (7%) experienced recurrence of cyclic pain at 9 months, requiring a repeat ablation due to incomplete coagulation of endometrial tissue.
- No intraoperative complications were observed.
- Patient satisfaction rate exceeded 85%, with significant improvements in quality of life, sleep, and emotional stability.

Discussion

MRKH syndrome presents not only a reproductive anomaly but also a significant source of chronic pain and psychological burden. While the primary clinical concern has historically been vaginal agenesis and amenorrhea, cyclic pelvic pain is increasingly recognized as a debilitating symptom, often overlooked in initial consultations.





Traditional approaches included total removal of the rudimentary horns, which can be technically challenging and associated with risks such as ureteral injury or excessive bleeding. In contrast, endometrial ablation provides a safer and equally effective alternative.

Literature reviews and case series (e.g., Gholoum et al., 2016; Oppelt et al., 2007) support our findings. These studies highlight the success of minimally invasive interventions in achieving symptomatic relief.

It is crucial to maintain a multidisciplinary approach, involving pediatric gynecologists, radiologists, laparoscopic surgeons, and, where needed, psychologists. Early identification of symptomatic patients can prevent years of undiagnosed suffering.

Conclusion

Cyclic chronic pelvic pain in MRKH syndrome is a clinically significant but underrecognized complication. In patients with functioning endometrial remnants, laparoscopic endometrial ablation offers a safe and effective strategy to alleviate pain, minimize surgical risks, and improve quality of life.

Given its minimally invasive nature, this approach should be considered as a first-line option, particularly in young patients not amenable to horn excision. Further research with larger cohorts and longer follow-up is warranted to optimize management strategies.

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