COMPLEX MÜLLERIAN MALFORMATION: A CASE REPORT OF EARLY SURGICAL INTERVENTION

MALFORMAÇÃO MULLERIANA COMPLEXA: UM RELATO DE CASO DE INTERVENÇÃO CIRÚRGICA PRECOCE

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ABSTRACT

Müllerian malformations result from failures in the formation, fusion, or resorption of the Müllerian ducts, occurring between the 6th and 12th weeks of embryogenesis. The septate uterus is the most common anomaly, associated with dysmenorrhea, dyspareunia, and infertility. Early diagnosis is essential to prevent reproductive complications. This case report describes a 23-year-old patient with a complete uterine septum, true cervical duplication, and a longitudinal vaginal septum. The investigation included pelvic ultrasound and magnetic resonance imaging, confirming the complex malformation. Treatment involved hysteroscopic surgery with septoplasty and intrauterine device insertion to prevent synechiae. Postoperative follow-up revealed a residual septum, requiring a second septoplasty, which successfully restored the uterine cavity. The hysteroscopic approach proved effective in this case, suggesting potential improvement in reproductive prognosis and symptom relief. This case highlights the importance of clinical suspicion, detailed diagnosis, and early intervention to enhance patients' quality of life and pregnancy outcomes.

Keywords: Genital anomalies, septate uterus, hysteroscopy, infertility.

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RESUMO

As malformações mullerianas resultam de falhas na formação, fusão ou reabsorção dos ductos de Müller, ocorrendo entre a 6ª e 12ª semanas de embriogênese. O útero septado é a anomalia mais comum, associada a dismenorreia, dispareunia e infertilidade. O diagnóstico precoce é essencial para evitar complicações reprodutivas. Este relato descreve uma paciente de 23 anos com septo uterino completo, duplicação cervical verdadeira e septo vaginal longitudinal. A investigação incluiu ultrassonografia pélvica e ressonância magnética, confirmando a malformação complexa. O tratamento foi a histeroscopia cirúrgica com septoplastia e inserção de Dispositivo Intrauterino (DIU), para prevenção de sinéquias. Em revisão pós-operatória, constatou-se septo residual, necessitando nova septoplastia, com sucesso na reconstrução da cavidade uterina. A abordagem histeroscópica mostrou-se eficaz, sugerindo possível melhora no prognóstico reprodutivo e alívio dos sintomas. O caso destaca a importância de suspeitar clinicamente, diagnosticar de forma detalhada e intervir precocemente para melhorar a qualidade de vida e os desfechos gestacionais das pacientes.

Palavras-Chave: *Anomalias genitais, útero septado, histeroscopia, infertilidade.*

1 INTRODUCTION

Müllerian malformations typically arise between the 6th and 12th weeks of female embryogenesis due to developmental defects in the formation, fusion, or resorption of the Müllerian ducts (Laufer; Decherney, 2024).

The classification system proposed by the American Society for Reproductive Medicine (ASRM) is based on the extent and nature of anomalies affecting the Müllerian ducts. This classification encompasses nine distinct categories: Müllerian agenesis, cervical agenesis, unicornuate uterus, didelphys uterus, bicornuate uterus, septate uterus, longitudinal vaginal septum, transverse vaginal septum, and complex anomalies (Pfeifer *et al*., 2021). These conditions may be associated with menstrual disorders, infertility, and coexisting malformations of the urinary and musculoskeletal systems (Liao, 2020, p. 780).

Statistically, according to a cohort study by Reyes-Muñoz *et al*. (2019), the prevalence and distribution of various subtypes of Müllerian anomalies were evaluated. The septate uterus emerged as the most frequently observed anomaly, accounting for approximately 54.2% of cases, followed by arcuate uterus (15.8%), bicornuate uterus (10.7%), unicornuate uterus (8.5%), didelphys uterus (6.2%), hypoplasia/agenesis (3.4%), and unclassified anomalies (1.1%).

Nevertheless, these anomalies often go undetected at birth, as asymptomatic uterine malformations hinder early diagnosis. This contributes to underreporting and potential methodological errors, with many cases being identified only when reproductive issues arise later in life (Liao, 2020). Therefore, it is essential to conduct a thorough clinical investigation that includes a comprehensive physical and gynecological examination, along with the support of appropriate imaging and diagnostic tools, to ensure accurate diagnosis and optimal management (Liao, 2020).

We report a rare case of a patient presenting with a complete uterine septum, true cervical duplication, and a longitudinal vaginal septum in the upper third of the vagina, culminating in a diagnosis of a complex Müllerian anomaly. The patient underwent early hysteroscopic intervention. This condition does not conform to classical theories of embryological development or to the ASRM classification system, and only a limited number of similar cases have been documented in the literature (Saygili-Yilmaz *et al*., 2004). Nonetheless, this case report adheres to the current recommendations established by the American Society for Reproductive Medicine (ASRM).

2 CASE NARRATIVE

A 23-year-old nulligravid, mixed-race woman presented with dysmenorrhea and deep dyspareunia. During routine gynecological follow-up, transvaginal pelvic ultrasonography revealed a congenital uterine malformation, specifically a complete septate uterus. Given the clinical findings and in accordance with medical guidelines, a renal tract ultrasound was requested to investigate associated anomalies, which returned normal bilateral findings.

As the diagnostic process progressed, a pelvic magnetic resonance imaging (MRI) scan was performed, as it is considered a highly accurate modality for the evaluation of Müllerian anomalies. The MRI identified a complex malformation characterized by a complete septate uterus with a single fundus, two uterine cervices, and a longitudinal vaginal septum in the upper third of the vagina, as illustrated in Figure 1 A and 1 B.

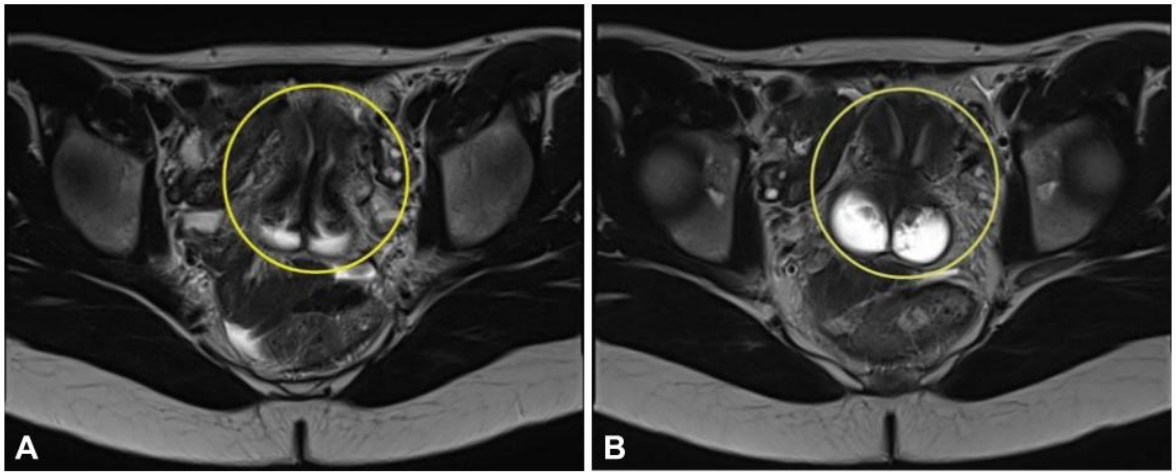

Figure 1. Axial pelvic magnetic resonance imaging: (A) longitudinal section demonstrates an elongated endometrial cavity divided by a T2 hypointense septum, consistent with a fibromuscular tissue, resulting in complete partitioning of the uterine cavity; (B) longitudinal section demonstrates cervical duplication, with two distinctly separate uterine cervixes, each maintaining anatomical continuity with its respective endocervical canal. This finding is compatible with a Müllerian duct anomaly.
Source: The authors (2025).

The proposed management was elective surgical hysteroscopy. The patient underwent hysteroscopic septoplasty with simultaneous insertion of a TCu 380A copper intrauterine device (IUD) to prevent the formation of intrauterine synechiae, which are known to develop within approximately six weeks following the procedure (Dason; Mathur; Murji, 2021).

During the scheduled postoperative follow-up at three months, a residual septum was noted. A second hysteroscopic septoplasty was performed and proved successful, with a subsequent surgical review confirming the presence of a single uterine cavity. Despite advances in reproductive medicine and in the development of diagnostic imaging techniques, studies focusing on Müllerian anomalies, particularly those involving a septate uterus associated with other complex malformations, are still limited in the current scientific literature (Chang *et al*., 2004).

An advanced literature search conducted on PubMed using the descriptors “Infertility” [MeSH Terms] OR “Intrauterine Devices” [Text Word], “Hysteroscopy” [MeSH Terms] OR “Uterine Septoplasty” [Text Word], and “Septate Uterus” [MeSH Terms] OR “Müllerian Malformations” [Text Word] yielded only 11 articles. Notably scarce were detailed case reports involving multiple anatomical anomalies and specific interventions such as hysteroscopy combined with intrauterine device (IUD) placement for synechiae prevention.

This paucity of data highlights the importance of case reports such as the present one, which contribute significantly to the clinical knowledge surrounding early and individualized management of rare uterine malformations. Moreover, such publications enhance the evidence base regarding reproductive outcomes and the efficacy of minimally invasive procedures such as hysteroscopic septoplasty, ultimately informing improved therapeutic approaches and clinical decision-making.

This paper presents case reports encompassed by an advanced literature review, discussing the clinical data and therapeutic approaches employed, as well as the outcomes pertaining to the patients' conditions (Table 1).

Table 1 - Comparative table of case reports of complex Müllerian malformations and their specificities.

Study	Clinical presentations	Diagnostic methods	Therapeutic approaches	Outcomes
Chang *et al*., 2004	Dysmenorrhea, dyspareunia, infertility	Pelvic magnetic resonance imaging and hysteroscopy	Hysteroscopic septoplasty	Improvement of symptoms; some term pregnancies
Saygili-Yilmaz *et al*., 2004	Dysmenorrhea, dyspareunia	Pelvic magnetic resonance imaging	Septoplasty	Anatomical correction; uncertain reproductive prognosis
Reyes-Muñoz *et al*., 2019	Infertility, some patients with menstrual complaints.	Hysteroscopy and laparoscopy	Variable depending on the anomaly (including septoplasty)	Anatomical correction in most cases; partial improvement in reproductive outcomes.
Present case	Dysmenorrhea, dyspareunia	Transvaginal ultrasound and magnetic resonance imaging	Septoplasty with DIU insertion	Anatomical correction after two procedures; symptomatic improvement.

Caption: Comparative table of case reports on complex Müllerian malformations, including descriptions of clinical presentations, diagnostic methods, therapeutic approaches, and outcomes.
Source: The authors (2025).

3 PATIENT PERSPECTIVE

This study was conducted with the patient's informed consent, in accordance with the principles of the Free and Informed Consent Form (TCLE). The patient expressed that her primary motivation in contributing to this report was to encourage other women, once diagnosed, to undergo surgical intervention early in the clinical course. She emphasized that timely surgical management may lead to better clinical outcomes and greater confidence regarding obstetric prognosis.

4 DISCUSSION

A septate uterus is associated with adverse reproductive outcomes, including recurrent pregnancy loss, unexplained primary infertility, malpresentation, and preterm delivery. It may also affect menstrual health, presenting with symptoms such as dysmenorrhea, amenorrhea, dyspareunia, and chronic pelvic pain (Laufer; Decherney, 2024).

Several pathophysiological mechanisms have been proposed to explain the correlation between a septate uterus and poor pregnancy outcomes. These include altered vascularization of the uterine septum and the opposing endometrial surface, increased intrauterine pressure, cervical insufficiency, abnormalities in myometrial contractility, and reduced expression of hormone receptors in the myometrium and/or endometrium (Pace; Júnior; Pereira, 2021).

The primary treatment modality is hysteroscopic septal resection. However, this procedure carries risks such as uterine perforation, infection, and postoperative intrauterine adhesions. Therefore, a follow-up surgical evaluation after septoplasty is essential to ensure optimal outcomes. Although such complications are generally rare, they can be serious if not identified promptly. When performed by experienced professionals, the incidence of severe adverse outcomes remains low (Pace; Júnior; Pereira, 2021).

The Randomized Uterine Septum Trial (TRUST), a large multicenter international randomized controlled trial, examined various postoperative adjunctive therapies for adhesion prevention. These included intrauterine devices, estrogen therapy, Foley catheter placement, aspirin, and hyaluronic acid instillation, among others. While these interventions are supported by some evidence, few have been

specifically evaluated in the context of postoperative care following septal resection. Consequently, there remains a lack of robust evidence to support the routine use of specific adjuncts, underscoring the need for clinicians to share their surgical experiences to advance treatment strategies (Dason; Mathur; Murji, 2021).

The reported outcomes were diverse: some patients achieved complete anatomical correction and successful pregnancies, while others required reintervention due to residual septa, similar to the course observed in the present case. A successful septoplasty is believed to restore a unified uterine cavity, with the potential to support a physiological term pregnancy. Additionally, it may increase the likelihood of alleviating menstrual symptoms and promote more favorable reproductive outcomes (Laufer; Decherney, 2024).

Thus, this report is primarily limited by the absence of long-term follow-up, particularly regarding reproductive outcomes, as the patient did not achieve pregnancy during the observation period. Although anatomical correction was achieved, it is not feasible to estimate future fertility, risk of miscarriage, or potential obstetric outcomes based on this single case alone. Consequently, as an isolated report, its conclusions should not be generalized, underscoring the necessity for additional studies with extended follow-up.

Given the patient's reproductive potential and the complexity of the malformation, long-term follow-up is recommended. This should include periodic pelvic exams to ensure uterine cavity integrity, evaluation of menstrual symptoms, and pre-conception counseling should future pregnancy attempts be made. In the event of a future pregnancy, first-trimester ultrasound, cervical length surveillance, and placentation assessment are recommended, as these measures can aid in identifying complications more commonly associated with uterine anomalies (Laufer; Decherney, 2024).

5 CONCLUSION

Müllerian malformations represent a rare condition in the general population but can significantly impair a woman's reproductive health. Therefore, there is a clear need for increased awareness and understanding of these anomalies, with emphasis on early diagnostic assessment, guided medical counseling, and timely specialized

treatment. Such an approach is crucial to improving menstrual, sexual, and gestational outcomes and, ultimately, enhancing the quality of life for affected individuals.

In the present case, early surgical hysteroscopy enabled the reconstruction of the uterine cavity by restoring normal uterine anatomy, thereby increasing the intrauterine space. This anatomical correction may support appropriate fetal development and potentially increase the chances of carrying a pregnancy to term, reflecting the therapeutic value of early intervention in complex uterine malformations.

ACKNOWLEDGMENTS

We extend our sincere gratitude to all our undergraduate colleagues who contributed to the development of this work. Special thanks to our advisor for their technical expertise and continuous guidance throughout the process. We also acknowledge the professors for their valuable feedback, support, and patience. Finally, we thank the University Center of Espírito Santo (UNESC), whose academic environment was fundamental to our educational journey.

ETHICAL APPROVAL

This study was conducted in full compliance with ethical standards and was duly approved by the Research Ethics Committee for Human Studies of the University Center of Espírito Santo (CEP/UNESC), under opinion number 7.696.791 (CAEE number 90174225.0.0000.5062).

DECLARATION OF CONFLICT OF INTEREST

The authors declare that they have no conflicts of interest related to this publication.

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