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# Delayed diagnosis of a non-communicating right uterine horn: A case report and literature review

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ABSTRACT

*Background:* Accurate diagnosis of a müllerian anomaly is essential for appropriate management and prevention of complications. However, diagnosis is often missed or delayed.

*Case:* This is a case of a nulliparous woman with a müllerian anomaly diagnosed at the age of 36 despite prior evaluation with ultrasound and laparoscopy. Magnetic resonance imaging (MRI) suggested a unicornuate uterus with a right non-communicating rudimentary horn. Hysteroscopy and chromopertubation confirmed the diagnosis. The rudimentary horn was resected laparoscopically using bipolar energy.

*Conclusion:* An undiagnosed müllerian anomaly should be considered in the setting of persistent dysmenorrhea. Three-dimensional ultrasound or MRI should be used judiciously in patients with refractory dysmenorrhea or when ultrasound raises concern for a müllerian anomaly.

## 1. Introduction

Unicornuate uterus with a rudimentary horn is a rare type of müllerian anomaly found in approximately 0.4% of women [1]. The horn can be communicating or non-communicating, leading to variable clinical presentations. A non-communicating horn typically remains silent until adolescence, when the endometrium becomes functional. The most common presentation is dysmenorrhea; hematometra and endometriosis can also develop [1,2]. These clinical findings are thought to be explained by obstruction of the non-communicating horn, resulting in retrograde menstruation.

Diagnosis at the time of ectopic pregnancy in the rudimentary horn is also common. A uterine horn ectopic pregnancy can be life-threatening due to risk of uterine rupture, which can occur as early as the first trimester [3]. In addition, urinary tract anomalies are commonly associated with müllerian anomalies, and have been reported to be more frequent with a unicornuate uterus than with other müllerian anomalies [4].

The rarity and wide range of müllerian anomalies can make prompt diagnosis difficult. In 2021, the American Society of Reproductive Medicine (ASRM) developed a new classification system in attempt to expand and update the American Fertility Society (AFS) Classification from 1988. In that article, the authors comment that müllerian anomalies often remain undiagnosed for prolonged periods, and patients may undergo inadequate or inappropriate surgical interventions, which result in persistent pain or loss of reproductive function [5]. Accurate diagnosis is critical to appropriate management and prevention of complications. Our objectives are to report a late presentation of a unicornuate uterus with a non-communicating rudimentary horn and review management strategies as well as potential advantages of a combined diagnostic approach.

## 2. Case Presentation

A 36-year-old woman, G0, diagnosed with endometriosis by laparoscopy at age 15, presented for fertility evaluation. The patient had lifelong dysmenorrhea and had been on continuous hormonal suppression since adolescence. She was amenorrheic on this therapy. Transvaginal ultrasound performed at age 30 raised concern for a possible bicornuate versus septate uterus, but no additional work-up was performed at that time. On presentation at age 36, magnetic resonance imaging (MRI) was ordered, with findings consistent with a left unicornuate uterus with a right-sided non-communicating rudimentary horn containing active endometrium (Fig. 1). Renal anatomy appeared

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normal.

A combined surgical procedure in the operating room was recommended. Exam under anesthesia confirmed a single cervix without vaginal malformation. Diagnostic hysteroscopy revealed a small left uterine cavity with a single tubal ostium (Fig. 2). No communication to the right uterine horn was noted. Laparoscopy revealed a single uterus with a fundal indentation and findings suggestive of endometriosis (Fig. 3). During chromopertubation, spill was observed only from the left fallopian tube. The right uterine horn could be seen distinctly from the adjacent left unicornuate uterus, as it did not fill or distend. After injecting intrauterine vasopressin, the right uterine horn and fallopian tube were resected using bipolar energy. Chromopertubation was performed again, and no spill was noted at the site of uterine resection. The uterus was repaired with laparoscopic suture (Fig. 4). The patient tolerated the procedure well, was discharged the same day, and reported significant improvement in her pain at her post-operative appointment.

## 3. Discussion

This case highlights the potential for late diagnosis and misdiagnosis of müllerian anomalies. Our patient had a transvaginal ultrasound scan years prior to her presentation, which suggested a possible müllerian anomaly. Had she only undergone laparoscopy, it likely would have been concluded that she had an arcuate uterus, necessitating no further intervention. However, with MRI it became clear that her diagnosis was not an arcuate uterus but a unicornuate uterus with a functional non-communicating rudimentary horn, necessitating surgical intervention. Hysteroscopy and chromopertubation confirmed this diagnosis and were able to be performed at the same time as her therapeutic laparoscopic resection of the uterine horn.

Most patients with a rudimentary horn present in the third decade of life. Failure to make the diagnosis in a timely manner can result in adverse gynecologic and obstetric outcomes for patients. Persistent pain and loss of reproductive function can occur [5]. There is also increased risk of spontaneous abortion, ectopic pregnancy and associated rupture, preterm labor, cervical insufficiency, malpresentation, and fetal growth restriction [1,3].

A unicornuate uterus may be difficult to diagnose using two-dimensional ultrasound (2D-US) [6]. Studies report the sensitivity of standard ultrasound for diagnosis is only 26% [3]. On 2D-US, identifying the rudimentary uterine horn can be challenging. Furthermore, the smaller size and lateral deviation of a unicornuate uterus may not be appreciated [7]. The ASRM recommends MRI or three-dimensional ultrasound (3D-US) to screen for a unicornuate uterus [5]. Both studies are superior to 2D-US and can provide information about both the external contour of the uterus and the endometrial cavity [8].

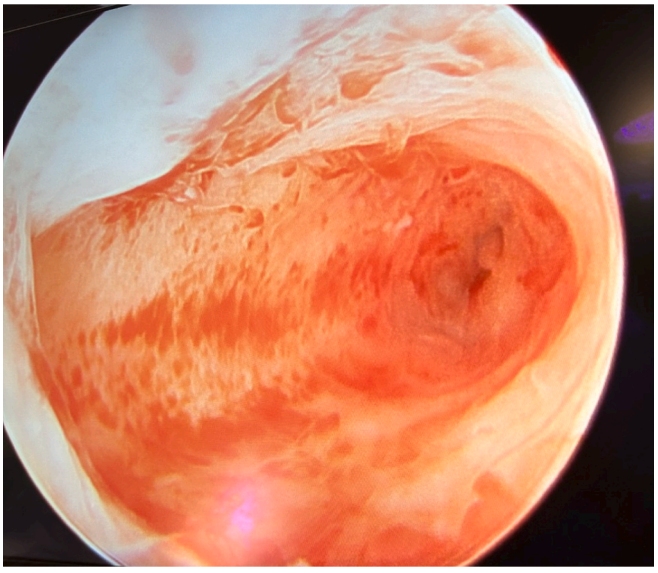

**Fig. 2.** Hysteroscopic findings demonstrating left unicornuate uterus with a single tubal ostium and no communication to right uterine horn.

For many years, MRI has been considered the gold standard for diagnosing a unicornuate uterus [5]. Multiplanar capabilities and excellent soft-tissue resolution allow diagnosis of all subtypes of unicornuate uterus and differentiation from other müllerian anomalies [7]. Furthermore, diagnostic accuracy is maintained at any point in the menstrual cycle and is not dependent on technical skill like sonography. MRI also has the added advantage of being able to diagnose an associated renal anomaly.

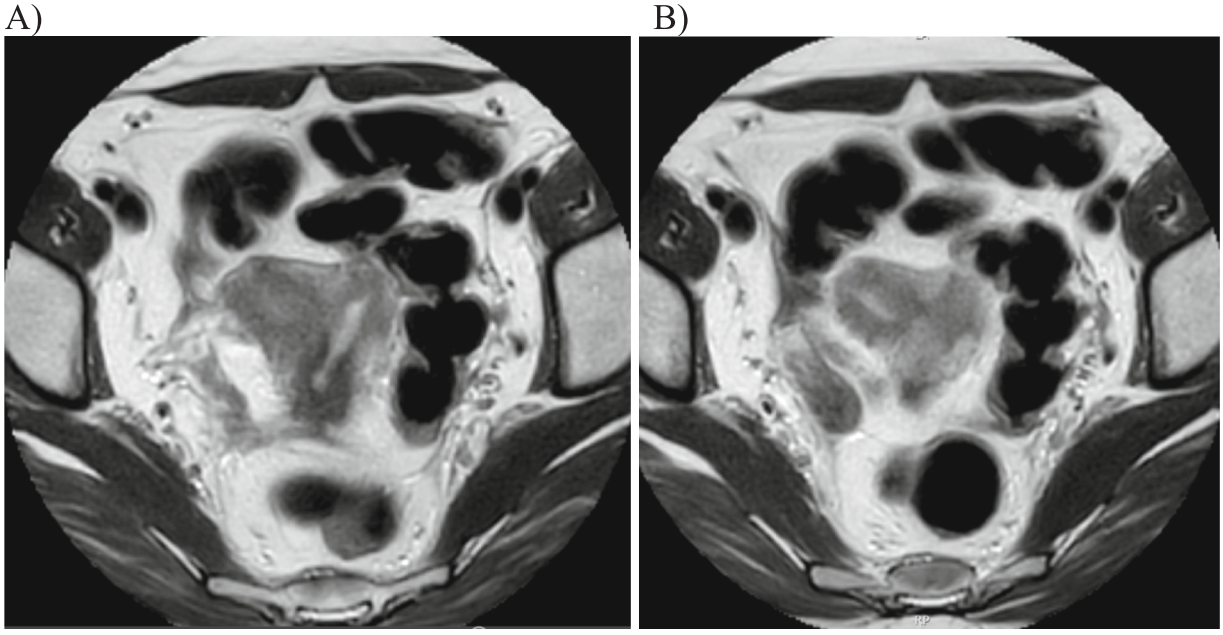


**Fig. 1.** Axial T2-weighted magnetic resonance imaging (MRI) images demonstrating left unicornuate uterus (A) and right functional non-communicating rudimentary horn (B).

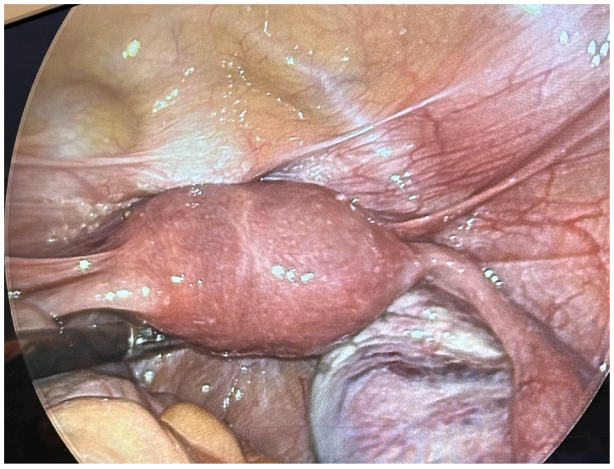

**Fig. 3.** Laparoscopic findings demonstrating an arcuate-appearing uterus.

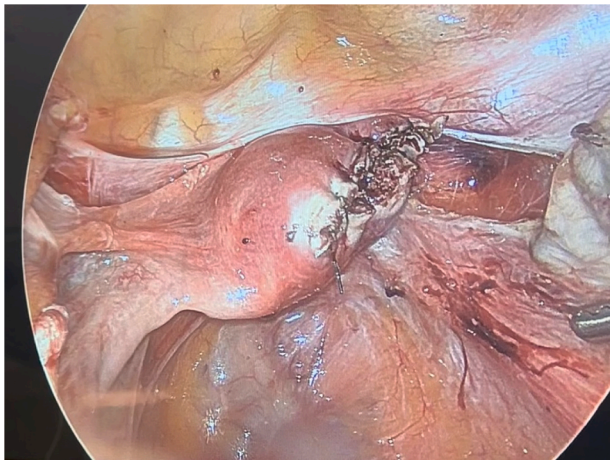

**Fig. 4.** Laparoscopic findings following resection of non-communicating uterine horn and closure of the uterus.

Although not currently considered the gold standard, studies have found a good level of agreement between 3D-US and MRI for diagnosing various müllerian anomalies [8]. As opposed to 2D-US, 3D-US allows for reconstructed images to be obtained in the coronal plane and clearly depicts the deviation of the unicornuate uterus and characteristic appearance of the endometrium [7]. While MRI can be performed at any point in a patient's menstrual cycle with high diagnostic accuracy, 3D-US has higher sensitivity and specificity for diagnosing müllerian anomalies in the luteal phase of the cycle [9].

The clinical utility of MRI and 3D-US is indisputable. The challenge for the clinician is determining which patients should undergo one of these higher-end imaging studies. On average, MRI is hundreds to thousands of dollars more expensive than traditional ultrasound. 3D-US, though less expensive, is less readily available and requires more technical skill than traditional ultrasound. These barriers explain why these studies should not be ordered routinely for all patients. However, there are clinical instances in which they clearly offer substantial advantage.

When 2D-US raises concern for a possible müllerian anomaly, we recommend further evaluation with either 3D-US or MRI. We also recommend that müllerian anomaly be considered in the setting of persistent or refractory dysmenorrhea. Additionally, when diagnostic laparoscopy is planned to evaluate dysmenorrhea, concomitant hysteroscopy and/or tubal evaluation should be considered given the potential to improve diagnostic capability with limited additional risk and minimal added operating time. Laparoscopy alone is not sufficient to exclude the diagnosis of a müllerian anomaly, because it does not provide evaluation of the internal anatomy of the uterus. Nor does normal renal anatomy exclude the diagnosis of a müllerian anomaly. Müllerian anomalies are frequently associated with renal anomalies due to the close association of genital and urinary embryological development. The kidney is of wolffian origin, whereas the uterus, cervix, and upper vagina are of müllerian origin. Unicornuate uterus is frequently associated with ipsilateral renal agenesis due to failure of both wolffian and müllerian ducts to develop on that side. However, these can be isolated defects as well. While renal agenesis is predictive of an obstructive ipsilateral müllerian anomaly over 50% of the time, normal renal anatomy does not exclude müllerian obstruction [3]. It is important to have a broad differential diagnosis and retain clinical suspicion for müllerian anomaly in patients who fail to respond to medical management.

*Contributors*

Lindsay A. Hartup contributed to patient care, conception of this case report, completion of the literature review, acquiring and interpreting the data, drafting the manuscript, and revising the manuscript.

Katherine E. Kostroun contributed to patient care, conception of this case report, and revising the article critically for important intellectual content.

Belinda J. Yauger contributed to patient care, conception of this case report, acquiring and interpreting the data, and revising the article critically for important intellectual content.

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*Patient consent*

We received our patient's written consent to submit her case for publication.

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*Conflict of interest statement*

The authors declare that they have no conflict of interest regarding the publication of this case report.

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