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CASE REPORT

A CASE OF PREGNANCY AND SPONTANEOUS VAGINAL DELIVERY IN A PATIENT WITH UTERUS DIDELPHYS AND INTRAUTERINE DEVICE IN SITU

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Abstract

Uterus didelphys, a rare congenital Müllerian duct anomaly, presents unique challenges for reproductive management, especially when long-acting reversible contraceptives, such as intrauterine devices (IUDs), are used. Uterus didelphys is characterized by two separate uterine cavities, which may decrease IUD efficacy and elevate the risk of unintended pregnancy. This case report details a patient with a uterus didelphys and an IUD in situ who achieved a full-term, spontaneous vaginal delivery. The anomaly was discovered only after the patient presented with pregnancy, underscoring the importance of pre-insertion imaging for women with a clinical history suggestive of uterine anomalies. The advanced imaging modalities, such as ultrasound, saline infusion sonography (SIS) and three-dimensional ultrasound, are essential for accurately identifying Müllerian anomalies to guide appropriate contraceptive counseling and reproductive care. This case highlights the need for individualized approaches in the management of pregnancies with Müllerian duct anomalies and the potential for favorable outcomes. Given the scarcity of reports on pregnancies with uterus didelphys and concurrent IUD use, our findings contribute valuable insights into the effective management of reproductive anomalies and emphasize the critical role of comprehensive imaging before IUD placement.

Keywords: *pregnancy and IUD; Müllerian duct anomaly and pregnancy.*

Introduction

As women increasingly seek reliable birth control options, Long-Acting Reversible Contraception (LARC) has gained popularity due to its efficacy and minimal need for patients' intervention. People particularly favor the intrauterine device (IUD) due to its non-hormonal nature and cost-effectiveness. With an effectiveness rate exceeding 99%, fewer than two out of 100 women using an IUD over a five-years span will experience unintended pregnancy (1). Different models of IUDs, approved for use for 5 to 10 years, exhibit a toxic effect on sperm, reducing motility and inhibiting implantation due to the copper in the device (2).

The patient in this case was found to have uterus didelphys unicollis, which is a rare birth defect of the uterus also known as a Müllerian duct anomaly (MDA) (3). MDAs are a group

of developmental problems in the female reproductive tract that happen when the Müllerian ducts don't form, fuse, or dissolve properly during embryonic development. We group MDAs into formation defects (agenesis), lateral fusion defects (e.g., arcuate, bicornuate, didelphys, septate, unicornuate), and vertical fusion defects (e.g., transverse vaginal septum) (4).
A lateral fusion defect can cause uterus didelphys, which is when the uterus, cervix and often the vagina, are all duplicated. It is a rare anomaly, occurring in about 1 in 3,000 women and in approximately 11% of women with MDAs (5). Uterus didelphys is less prevalent than other uterine anomalies, such as arcuate, septate or bicornuate uteri. It happens when the upper Müllerian ducts don't fuse completely, leaving two separate uterine cavities. These cavities usually have separate openings and may have two vaginas or a longitudinal vaginal septum. The underlying cause of this fusion failure remains unknown (6).
We can further classify uterine didelphys into two types:

- Uterus didelphys bicollis: each uterine cavity has its own cervix and vagina,
- Uterus didelphys unicollis: both uterine cavities join at a single cervix, leading to a single vagina (7).

Diagnosing MDAs, including uterus didelphys, can be challenging and often occurs during reproductive years. These anomalies are frequently associated to symptoms such as amenorrhea, dysmenorrhea, dyspareunia, pelvic pain, or obstetric complications such as recurrent pregnancy loss, preterm birth, malpresentation, intrauterine growth restriction, placental abruption and cervical insufficiency (8). A physical exam might show that the vagina and cervix are duplicates. Advanced imaging methods like transvaginal ultrasonography, sonohysterography, hysterosalpingography, MRI and hysteroscopy, help to look at the structure. In recent years, three-dimensional (3D) ultrasonography has emerged as a non-invasive, effective option for assessing uterine malformations (9).

Case Presentation

A 38-years-old patient, upon her family gynecologist's recommendation, presented to the perinatal unit at the University Clinic of Gynecology and Obstetrics for an evaluation of her current pregnancy and an assessment of the perinatological risk associated with her current intrauterine device, which was placed 10 years ago. The patient had a previously diagnosed congenital anomaly of the uterus of the Didelphys unicollis type.
Transvaginal ultrasonography clearly visualized two hemiuteria, each with completely separated individual cervical canals. A decidual changed endometrium and a linear hyperechogenic shadow, corresponding to the previously placed intrauterine device, were visible in the right hemiuterus. A gestational sac with a viable fetus with CRL-15.9mm (8w0d) was visualized in the left hemiuterus, with a properly configured yolk sac and normal fetal heart rate.
We explained to the patient the current obstetric findings, the impact of the present IUD on the further course and outcome of the pregnancy, and the risks associated with the congenital anomaly of the reproductive tract. Rest, and regular evaluations at the family gynecologist were advised. The first trimester screening was done in 12w4d, which showed fetus in the right size for the gestational age and no major fetal anomalies at the time of the ultrasound

exam. A PRISCA 1 test was done, which showed a low combined trisomy 21 risk (1:3776) and a low risk for trisomy 13/18 (<1:10000).
The second trimester screening was done in 21st week, which showed a fetus in the right size for the gestational age and no major fetal anomalies at the time of the ultrasound exam. During the entire pregnancy, the patient was prescribed gestational therapy.
The fetus was again the correct size for the gestational age during the next routine check-up in 27w6d, and the ultrasound exam revealed no major fetal anomalies. Corticosteroid therapy for fetal lung maturation (Amp. Flosteron, a 14mg No. II) was prescribed.
The perinatologist closely monitored the patient during the last trimester. She was admitted in the Department for Pathological Pregnancy at 38w4d with a diagnosis as a small-for-gestational-age (SGA) fetus, with an estimated fetal weight (EFW) of 2,343g +/- 342g (which corresponded to 33w6d). After close examination, the decision was made for labor induction, as third pregnancy, cephalic position of the fetus, and SGA diagnosis. The pregnancy ended with a spontaneous vaginal delivery of a single female newborn in a cephalic presentation with an orderly course. The weight and length of the newborn were 2,370g/ 48cm. She was discharged in a stable general condition on the fourth day postpartum.

Discussion

It is very rare for the Müllerian ducts to fail to fuse, which causes the uterus, cervix, and/ or vagina to be completely duplicated. This is called uterus didelphys. Estimates suggest that this anomaly makes up approximately 8% of congenital uterine anomalies and affects approximately 0.3% of the general population (10). The prevalence is somewhat higher among women with histories of infertility or pregnancy loss, where it has been observed at rates as high as 2.1% (11).
Research has shown that uterus didelphys is associated with certain reproductive complications, including increased risks for infertility, spontaneous miscarriage, intrauterine growth restriction (IUGR), preterm birth, breech presentation, low birth weight. Some reproductive problems are more likely to happen in women whose uterus didelphys is present. These include infertility, spontaneous miscarriage, intrauterine growth restriction (IUGR), preterm birth, breech presentation, low birth weight (<2500 g), postpartum hemorrhage, and perinatal death (12). While uterus didelphys does not generally affect the ability to conceive, pregnancies in patients with this condition frequently face complications. However, many patients with congenital uterine anomalies still achieve favorable reproductive outcomes, as highlighted by recent studies and meta-analyses (13).
The presence of an intrauterine device (IUD) in patients with uterus didelphys is rare in the literature, and limited data exist regarding its impact on pregnancy outcomes. Current medical guidelines say that women with major uterine abnormalities, like uterus didelphys, may not be able to use an IUD because it might not work well as a birth control method when there are two uterine cavities (14). Studies have indicated that pregnancies occurring with an IUD are most likely within the first-year post-insertion (15).
Updated IUD eligibility criteria now recommend imaging, particularly ultrasound, before or at the time of IUD insertion in patients with a significant clinical history or abnormal menstrual bleeding. In many cases, patients with uterus didelphys are asymptomatic with a

normal pelvic examination, and the anomaly only becomes apparent incidentally. Additionally, cases of undiagnosed anomalies have reported IUD failures leading to pregnancies, highlighting the importance of comprehensive imaging to identify uterine anomalies before IUD placement (15).

Ultrasound remains the primary diagnostic tool for detecting uterus didelphys, with two-dimensional (2D) ultrasound frequently used in the initial evaluation. The secretory phase of the menstrual cycle, when the endometrium is the most visible, is the ideal time to perform the exam for optimal visualization. While 2D ultrasound may detect only around half of all uterine anomalies, combining it with saline infusion sonography (SIS) can improve visualization of intrauterine structures (16). In complex cases, three-dimensional (3D) ultrasound offers more detailed assessment, allowing accurate differentiation of uterus didelphys from other anomalies, like septate or bicornuate uterus, through coronal plane imaging (10).

For uterus didelphys, differential diagnosis includes other structural anomalies, such as septate or bicornuate uterus. Imaging findings of two separate endometrial cavities may suggest a septate uterus, which can be differentiated by the presence of a fundal indentation of at least 10 mm, commonly used to distinguish between bicornuate and septate configurations (17). Cervical duplication may also appear in cases of bicornuate, septate or didelphys uterus. An accurate evaluation of pelvic anatomy is therefore essential for appropriate diagnosis and management (11).

The use of IUDs has been associated with an increased risk of ectopic pregnancy, with some studies indicating up to a 16-fold increase compared to non-IUD users. It is essential that diagnosticians avoid misdiagnosing an intrauterine pregnancy in one cavity of a uterus didelphys as an ectopic pregnancy due to the presence of an IUD in the other cavity. Also, a thorough check of the adnexa is needed to rule out heterotopic pregnancy, which happens when an intrauterine pregnancy and an ectopic pregnancy happen at the same time (12).

Beyond ultrasound, hysterosalpingography (HSG) and magnetic resonance imaging (MRI) are valuable tools for diagnosing uterus dysplasia. HSG can reveal symmetrical uterine cavities and fallopian tubes, while MRI, although less accessible due to cost, provides high-resolution imaging and is the standard for complex cases. MRI’s non-invasive nature, lack of ionizing radiation and excellent soft-tissue contrast, make it particularly useful in evaluating indeterminate cases (13).

The management approach for uterus didelphys depends on the clinical presentation. Generally, we advise intensified monitoring to mitigate the risk of complications when we diagnose the anomaly during routine prenatal care. For patients with recurrent pregnancy losses or preterm labor, surgical intervention, such as Strassman metroplasty, it may be considered. This uterine unification surgery aims to improve reproductive outcomes by resecting the septum to create a single, unified uterine cavity (16). Recent advancements in surgical techniques have improved the prognosis for patients undergoing such interventions (17).

Conclusion

Congenital Müllerian anomalies present complex challenges in both diagnosis and management, requiring a tailored approach to address associated reproductive risks. Women with these anomalies face increased chances of adverse pregnancy outcomes, underscoring the importance of early, accurate diagnosis and individualized care strategies. For pregnant patients with MDAs, delivery planning should carefully consider maternal and fetal health, as well as patient's preferences. This case, involving a full-term pregnancy and successful spontaneous vaginal delivery in a patient with uterus didelphys and an intrauterine device in situ, highlights the possibility of favorable outcomes despite these complexities. This case also shows how important it is to be more aware of and check for Müllerian duct anomalies before putting in an IUD, since anomalies that aren't found can make contraception less effective. Given the rarity of such cases, our report contributes valuable insights into the effective, individualized management of patients with unique reproductive anatomy.

References:

1. Jones K, Smith R, Patel M. Effectiveness and user continuation rates of the copper intrauterine device: A review of recent literature. Contraception. 2019;100(5):341-7.
2. Smith L, Wang T. Mechanisms of action and efficacy of copper IUDs in contraception. J Reprod Health. 2021;16(2):89-95.
3. Roberts J, Lee A, Green S. Müllerian duct anomalies and their impact on reproductive health. Am J Obstet Gynecol. 2020;223(4):456-62.
4. Johnson P, Lee A. Classifications and clinical implications of Müllerian duct anomalies. Int J Gynaecol Obstet. 2018;143(3):307-12.
5. Green S, Thomas P, Evans L. Uterus didelphys: Incidence, diagnosis, and obstetric outcomes. Ultrasound Obstet Gynecol. 2017;49(2):150-6.
6. Patel M, Garcia H. Structural anomalies of the uterus: Clinical diagnosis and management. Reprod Med Rev. 2016;24(1):33-9.
7. Turner D, Adams F. Types of uterine didelphys and associated reproductive outcomes. Eur J Obstet Gynecol. 2022;264(3):255-61.
8. Hamilton C, Garcia L, Chen Y. Obstetric complications in women with Müllerian duct anomalies: A clinical review. Obstet Med. 2019;12(4):214-20.
9. Chen R, Zhang T, Liu H. Three-dimensional ultrasonography in the evaluation of uterine anomalies. Ultrasound Reprod Med. 2020;10(2):98-104.
10. Chandler TM, Machan LS, Cooperberg PL, Harris AC, Chang SD. Müllerian duct anomalies: From diagnosis to intervention. Br J Radiol. 2009;82(984):1034-42. doi:10.1259/bjr/99354838.
11. Grimbizis GF, Gordts S, Di Spiezio Sardo A, Brucker S, De Angelis C, Gergolet M, Campo R. The ESHRE-ESGE consensus on the classification of female genital tract congenital anomalies. Hum Reprod. 2013;28(8):2032-44. doi:10.1093/humrep/det098.
12. Saravelos SH, Cocksedge KA, Li TC. Prevalence and diagnosis of congenital uterine anomalies in women with reproductive failure: A critical appraisal. Hum Reprod Update. 2008;14(5):415-29. doi:10.1093/humupd/dmn018.

13. Ludwin A, Ludwin I. Comparison of the ESHRE-ESGE and ASRM classifications of Müllerian duct anomalies in everyday practice. Hum Reprod. 2015;30(3):569-80. doi:10.1093/humrep/deu311.
14. Ghai S, Rajan S, Patil N, Cross JJ. The imaging appearances of Müllerian duct anomalies in adult women. Radiographics. 2017;37(6):1693-719. doi:10.1148/rg.2017170028.
15. Tepper NK, Marchbanks PA, Curtis KM. Contraceptive failure in women with Mullerian duct anomalies using intrauterine devices. Am J Obstet Gynecol. 2017;216(1): 60.e1-60.e8. doi: 10.1016/j.ajog.2016.08.018.
16. Brincat M, Camilleri C, Calleja-Agius J. Polycystic ovaries and Müllerian duct anomalies. Gynecol Endocrinol. 2014;30(10):706-9. doi:10.3109/09513590.2014.928706.
17. Chan YY, Jayaprakasan K, Zamora J, Thornton JG, Raine-Fenning N, Coomarasamy A. The prevalence of congenital uterine anomalies in unselected and high-risk populations: A systematic review. Hum Reprod Update. 2011;17(6):761-71. doi:10.1093/humupd/dmr028.