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Original Research Article

A Rare Presentation of Uterus Didelphys in Labour

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Abstract:

Background: Mullerian Duct Anomalies (MDAs) are congenital abnormalities of the female reproductive system resulting from anomalous embryological development of the Mullerian Ducts. Among MDAs, a didelphys uterus, often referred to as a "double uterus," is one of the rarest. It occurs when two separate Mullerian Ducts develop independently, leading to the formation of two hemi-uteri, each with its own fallopian tube, ovary, and cervix. This anomaly can potentially lead to obstetric complications.

The Case: A 20-year-old primigravida presented at 30 weeks and 1 day for the first time in our OBGYN emergency department with preterm labor. History and records suggested that antenatally, she was suspected to have a uterine anomaly, possibly septate or bicornuate uterus and cervical incompetence based on findings from her first trimester ultrasound. The patient underwent assisted vaginal delivery with a right mediolateral episiotomy. A non-communicating thick longitudinal vaginal septum was unexpectedly discovered, with a wider vaginal segment noted on the right side.Vaginal speculum examination revealed the presence of two separate cervices. Neither the patient nor her husband had prior knowledge of this condition. 3 days post-delivery, pelvic MRI confirmed the presence of a didelphys uterus with a longitudinal complete vaginal septum. Subsequent abdominal CT scans showed no renal anomalies. The patient denied experiencing dyspareunia, dysmenorrhea, or chronic abdominal pain in the past.

Discussion: Achieving term pregnancies in cases of uterine didelphys is uncommon. Given the high incidence of obstetric complications and poor pregnancy outcomes associated with this condition, vigilant monitoring is essential both prenatally and throughout pregnancy.

Keywords: Bicornuate, Didelphys, Septum, Mullerian, Anomaly, Non-Communicating, MRI, Cervix.

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Introduction

Uterine didelphys is a congenital anomaly of the female reproductive system stemming from abnormal embryological development of the Mullerian Ducts. Also known as a double uterus, it represents one of the less common Mullerian Duct anomalies. These anomalies can arise due to failures in duct development, fusion, canalization, or reabsorption processes that typically occur between 6 to 12 weeks of gestation. [1]

According to the ESHRE (European Society of Human Reproduction and Embryology) nomenclature, uterine didelphys is classified as U2b, characterized by an internal indentation greater than 50% of the uterine wall and a straight external contour or one with an indentation less than 50%. This condition may also feature a double cervix or double vagina, with a longitudinal septum dividing them into two distinct structures. [2]

Many women with uterine didelphys are asymptomatic, although some may experience dyspareunia or dysmenorrhea due to variations in the longitudinal vaginal septum. Typically, these uterine anomalies are incidentally discovered. The estimated incidence of these abnormalities ranges from 0.5% to 5% in the general female population. [3]

In contrast, a bicornuate uterus (ESHRE class U3) is characterized by a fundus that is indented more than 1 cm or a fundal midline exceeding 50% of the uterine wall thickness. This anomaly results from partial rather than complete fusion of the Mullerian Ducts, and typically features a single cervix and vagina. Depending on the degree of fusion, there may be separation of the uterine horns. [4]

Didelphys uterus is characterized by complete failure of Mullerian duct fusion, resulting in separate uterine cavities and two cervices. A longitudinal

vaginal septum may be present, varying from thin and easily displaced to thick and rigid. Initial suspicion often arises during routine speculum examination, prompting further diagnostic investigation. Given the embryological association between Mullerian and Wolffian ducts, concurrent kidney abnormalities may also be observed in conjunction with uterine anomalies. [5]

It is widely accepted that uterine anomalies, including uterine didelphys and bicornuate uterus, are associated with adverse pregnancy outcomes. These may include increased risks of premature labor, spontaneous abortion, cesarean delivery (often due to fetal positioning challenges) and decreased live birth rates compared to individuals with normal uterine anatomy.

Unicornuate and didelphys uteri exhibit term delivery rates of approximately 45%, while untreated bicornuate and septate uteri have poorer outcomes with term delivery rates around 40%. Arcuate uterus shows a somewhat improved but still compromised pregnancy outcome with term delivery rates of about 65%.

ESHRE/ESGE classification
Female genital tract anomalies

	Uterine anomaly		Cervical/vaginal anomaly	
	Main class	*Sub-class*	*Co-existent class*	
U0	Normal uterus		C0	*Normal cervix*
U1	Dysmorphic uterus	a. T-shaped b. Infantilis c. Others	C1	*Septate cervix*
U2	Septate uterus	a. Partial b. Complete	C2	*Double 'normal' cervix*
U3	Bicorporeal uterus	a. Partial b. Complete c. Bicorporeal septate	C3	*Unilateral cervical aplasia*
U4	Hemi-uterus	a. With rudimentary cavity (communicating or not horn) b. Without rudimentary cavity (horn without cavity/no horn)	C4	*Cervical aplasia*
U5	Aplastic	a. With rudimentary cavity (bi- or unilateral horn) b. Without rudimentary cavity (bi- or unilateral uterine remnants/aplasia)	V0	*Normal vagina*
U6	Unclassified malformations		V1	*Longitudinal non-obstructing vaginal septum*
			V2	*Longitudinal obstructing vaginal septum*
			V3	*Transverse vaginal septum and/or imperforate hymen*
			V4	*Vaginal aplasia*
U			*C*	*V*

These anomalies can be classified into four main types based on abnormal Mullerian duct development:

1. Complete or partial failure of Mullerian duct development (agenesis; unicornuate uterus without a rudimentary horn).
2. Failure of ducts to canalize (unicornuate uterus with a rudimentary horn lacking proper cavities).
3. Incomplete fusion of Mullerian ducts (bicornuate or didelphys uterus).
4. Incomplete reabsorption of the uterine septum (septate or arcuate uterus).

Diagnosing these conditions traditionally involves invasive methods such as hysteroscopy, hysterosalpingography, and laparoscopy/laparotomy, which rely on subjective interpretation by clinicians. Initial imaging with 2D ultrasound is often insufficient for precise classification of Mullerian duct anomalies (MDAs). However, 3D ultrasound is gaining popularity as it offers a noninvasive alternative, providing detailed coronal views that facilitate morphological classification by visualizing both the endometrial cavity and uterine fundus.

Magnetic resonance imaging (MRI) is equally valuable in diagnosing MDAs, offering noninvasive imaging that can also detect associated urinary tract abnormalities concurrently.

In this case report, we discuss a rare instance of didelphys uterus in a woman with incompetent cervix and preterm onset of labor, who ultimately delivered vaginally without complications.

Presentation of the case

A 20-year-old primigravida with a suspected uterine anomaly at 30 weeks and 1 day presented to emergency labor and delivery room for preterm onset of

labor with a living fetus. During her prenatal course, records of maternal-fetal medicine (MFM), first trimester ultrasound detected two definite uterine horns, with cervical incompetence, but was unable to confirm the presence of two cervices. At a subsequent visit, the ultrasound was unable to determine whether the uterus was bicornuate or didelphys, as well as the number of cervixes. No further imaging was ordered to confirm the type of uterine anomaly present. Following clinic visits, throughout the pregnancy course were unremarkable. The patient did not report having dyspareunia, dysmenorrhea, or chronic abdominal pain in the past.

Patient had uncomplicated prenatal care and did not have any signs of bleeding or threatened preterm labor, her lab reports (B positive) and vital signs remained within normal limits, and fetal ultrasoundtracings also remained within normal limits. Fetus showed appropriate growth and the pregnancy was carried in the right half of uterine cavity.

At 30 weeks and 1 day, the patient presented with sudden vaginal leakage of clear fluids with some bloody mucoid discharge along with pain in the abdomen. On physical exam she was not in any distress, normotensive, and afebrile. On speculum exam the patient had a non-communicating, thick longitudinal vaginal septum, however, the patient and her husband were not aware of the patient's condition until that day and the right cervix was fully dilated and effaced with fetal head at station 0, absent membranes and clear liquor with the left cervix closed. The fetal heart tracing was category one, showing a fetal heart rate of 150 at baseline, moderate variability, with accelerations and no decelerations. The bedside sonogram at that time showed the fetus to bein vertex position, placenta positioned anteriorly. At the end of this work-up, the patient was admitted for Premature Rupture of Membranes (PROM) and Preterm Labor and underwent assisted vaginal delivery under antibiotic coverage.

Patient had a vaginal delivery of a live baby boy with weight of 1120 grams with Apgar Score7 at 1 min with right mediolateral episiotomy and without tear of vaginal septum. Placenta was removed by controlled cord traction and episiotomy wound repaired via chromic catgut in layers under aseptic precautions.

Pelvic MRI was performed 3 days after successful vaginal delivery and confirmed this Mullerian Duct anomaly to be didelphys uterus which had a longitudinal, complete vaginal septum. Subsequent abdominal CT scan didn’t reveal any renal anomaly.

Referred By: OBS & GYNAE	OPD/IPD No- 2368661
Part Examined: PELVIS	Date:- 30/03/2024

MRI - PELVIS (WITHOUT IV CONTRAST):

Imaging Technique: Coronal – STIR, T1 SE. Axial – T2 FS, T1 SE. Sag – T1 FS.

OBSERVATIONS:

Two distinct well defined uterine corpus are noted with an indentation in the external fundal contour. They show well defined zonal differentiation with well formed endometrial cavities. These corpi are extending caudally to form two distinct cervices and further seen distally opening into vaginal canal separated into two halves by a intervening vertical septum along the entire length.

The right uterine corpus is comparatively bulky (right measures ~ 3.5 x 3.7 x 8 cm (AP x TR x CC) & left measures ~ 2.6 x 2.9 x 3.6 cm (AP x TR x CC).

- **The two ovaries are seen in relation to the two distinct uterine bodies.**
- **Exophytic cyst noted in surface of right ovary measuring ~ 14 x 13 mm.**

URINARY BLADDER: Normal.

FLUID: Nil.

LYMPH NODES: Nil.

RECTUM, ANAL CANAL & VISUALIZED BOWEL: Normal.

VISUALIZED SPINE: Normal.

IMPRESSION:

- **Complete bicorporeal uterus (U3bC2V1 ESHRE classification) with double cervix and longitudinal non obstructing vaginal septum.**

Note: This is only a Radiological Impression & not a Diagnosis and has its limitations. Therefore it should be interpreted in correlation with clinical &/or pathological findings.

Discussion

A didelphys uterus is a rare Mullerian Duct anomaly compared to other classifications outlined by Buttram and Gibbons. Existing data on its clinical significance and outcomes primarily stem from small retrospective, observational, or case studies. These studies yield mixed results due to the anomaly's low incidence relative to more common malformations like arcuate, septate, and bicornuate uteri. [6]

Most women with a didelphys uterus are asymptomatic, though some may experience dyspareunia or dysmenorrhea due to a thick, occasionally obstructive vaginal septum. This septum can lead to complications such as hematocolpos/ hematometrocolpos, resulting in chronic abdominal pain. Rarely, cases have been

associated with endometriosis or genital neoplasms. [7]

Untreated didelphys uteri generally show better fertility outcomes compared to other Mullerian Duct abnormalities but still exhibit poorer reproductive performance than women with normal uterine anatomy. Pregnancy risks include increased rates of spontaneous abortion, fetal growth retardation, and premature delivery, with term delivery rates estimated at approximately 45% or lower, akin to those observed in unicornuate uterus cases. However, these rates are not as compromised as those seen in septate or bicornuate uteri, which are more prevalent among Mullerian Duct anomalies. [8]

Literature on didelphys uterus suggests varying pregnancy outcomes, with some studies indicating more favorable results compared to other anomalies, while others demonstrate poorer outcomes. Surgical correction (metroplasty) is generally not indicated for didelphys uterus, unlike for septate or bicornuate uteri, where it has shown improvement in reproductive and gestational outcomes in select cases. [9]

Didelphys uterus can also occur as part of Herlyn-Werner-Wunderlich syndrome (HWWS), also known as OHVIRA syndrome, characterized by didelphys uterus, obstructed hemi-vagina, and ipsilateral renal agenesis. This rare syndrome can present with symptoms like lower abdominal pain or a protruding vaginal mass post-menarche. Diagnosis typically involves pelvic ultrasound followed by MRI to confirm the condition, especially in cases presenting with obstructed hemi-vagina and renal anomalies. [10

In conclusion, while didelphys uterus is uncommon and presents challenges in diagnosis and management, understanding its clinical implications and associated syndromes like HWWS is crucial for appropriate patient care and management.

Conclusion

The didelphys uterus is an exceedingly rare Mullerian Duct anomaly with diverse reproductive and gestational outcomes compared to more prevalent abnormalities. The issue of fertility remains debated due to limited data on surgical interventions such as metroplasty, which are generally not recommended. However, excision of the vaginal septum may be necessary if symptomatic. Cesarean delivery is not typically warranted unless there is a thick, inflexible vaginal septum posing a risk of vaginal dystocia. Importantly, cervical incompetence is not usually associated with didelphys uterus. Diagnosis of didelphys uterus should prompt investigation for renal anomalies to exclude Herlyn-Werner-Wunderlich (OHVIRA) syndrome.

Currently, the available literature on didelphys uterus is sparse. Further research is essential to better understand its reproductive and gestational outcomes, enabling clinicians to provide informed guidance and care to affected individuals.

Uterine anomalies pose significant challenges during labor and delivery, often remaining undetected until pregnancy confirmation. To prevent emergency situations with inadequate preparation, women identified with possible uterine anomalies during first trimester ultrasounds should undergo comprehensive evaluation, including imaging such as magnetic resonance imaging (MRI), especially in cases where the anomaly is unclear. Planning for delivery should be meticulous to ensure appropriate resources are available for a safe outcome for both mother and neonate.

Patients with uterus didelphys are categorized as high-risk and warrant careful prenatal care. A thorough diagnosis and evaluation of the anomaly are crucial for proper delivery planning. Based on our review of the literature, cesarean section is considered a reasonable, though not mandatory, approach for uterus didelphys. While there have been reports of successful vaginal deliveries in these cases, surgical delivery is generally deemed safer for managing this type of Mullerian anomaly.

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Informed Consent: Written informed consent was obtained from the patient for their anonymized information to be published in this article.

Patient's Consent: Informed written consent has been obtained from the patient for all case details and images to be reported in the journal.

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References

1. Wong JWH. A case of vaginal birth after cesarean delivery in a patient with uterine didelphys. Case Rep Obstet Gynecol. 2019; 20 19: 3979581.
2. Carrington BM, Hricak H, Nuruddin RN, et al. Mullerian ducts anomalies: MR imaging evaluation. Radiology. 1990; 176:715-20.

3. Cansole D, Tamburrini S, Barresi D, et al. The value of MR imaging in the evaluation of Mullerian ducts anomalies. Radiol Med. 2001; 102: 226-32.
4. Brown MA. MR imaging of benign uterine disease. Magn Reson Imaging Clin N Am. 2006; 14:439-53.
5. Heinonen PK. "uterus didelphys: a report of 26 cases" European journal of obstetrics & gynecology & reproductive biology. 1984;17(5): 345-50.
6. Pui M. "Imaging diagnosis of congenital uterine malformation" Computerized medical imaging and graphics. 2004;28(7):425-33.
7. Hua M, Odibo AO, Longman RE, et al. Congenital uterine anomalies and adverse pregnancy outcomes. Am J Obstet Gynecol 2011; 205 (6): 558.e1–558.e1.
8. Console D, Tamburrini S, Barresi D, et al. The value of the MR imaging in the evaluation of Müllerian duct anomalies. Radiol ed. 2001; 102 (4): 226–232.
9. Heinonen PK. Clinical implications of the didelphic uterus: long-term follow-up of 49 cases. Eur J Obstet Gynecol Reprodiol. 2000;91 (2):183-90.
10. Grimbizis GF, Camus M, Tarlatzis BC, Bontis JN, Devroey P. "Clinical implications of uterine malformations and hysteroscopic treatment results". Human Reproduction Update. 2001; 7(2): 161-74