

Uterine anomalies in pregnancy

Clinical pearls for assessment and management of the pregnant patient with a uterovaginal anomaly

by ANNE-MARIE AMIES OELSCHLAGER, MD

Uterine anomalies are associated with higher rates of pregnancy complications, including spontaneous abortion, ectopic pregnancy, preterm labor, malpresentation, and abruption. Obstetricians should understand the embryology, incidence, classifications, and management of patients with uterine anomalies during pregnancy.

Embryology

The process of uterine and vaginal development begins in the first trimester. The bilateral müllerian (paramesonephric) ducts develop into the uterus, fallopian tubes, cervix, and proximal vagina. These ducts fuse and internal canalization of each duct will produce 2 channels divided by longitudinal septum. The septum will typically resorb throughout the second trimester. The urogenital sinus will also separate

into the distal vagina and urethra. Resorption anomalies represent persistence of the longitudinal uterine or vaginal septum. Transverse anomalies include obstructing vaginal septa or cervical atresia.

Classification and diagnosis of uterovaginal anomalies

The American Society for Reproductive Medicine (ASRM) Müllerian Anomalies Classification (MAC) system, updated in 2021,¹ categorizes the range of anomalies by descriptive terms and includes müllerian agenesis, cervical agenesis, unicornuate uterus, uterus didelphys, bicornuate uterus, septate uterus, longitudinal vaginal septum, transverse vaginal septum, and complex anomalies. An online interactive classification is available online. To view the ASRM MAC 2021 online, please scan the QR code.



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Diagnosis

To differentiate uterine anomalies, imaging or direct visual examination of the contour of external uterus and the endometrial cavity is required. To differentiate the bicornuate from septate from arcuate uterus, ideally, a line can be drawn from cornua to cornua. If the external contour indents more than 10 mm, the uterus is bicornuate. If there is less than 10-mm external contour indentation and less than a 10-mm fundal cavity indentation with the leading angle of the indentation greater than 90°, the diagnosis is arcuate uterus. If the indentation extends more than 10 mm into the cavity and the leading



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angle is less than 90°, the diagnosis is septate uterus.

Optimal tests include 3D transvaginal ultrasound, MRI, saline sonohysterography, or laparoscopy or laparotomy combined with hysteroscopy or hysterosalpingogram (HSG; **Figures 1 and 2**).^{2,3} Less sensitive diagnostic tests include 2D transvaginal ultrasound, clinical assessment at cesarean section, as well as hysteroscopy or HSG without external uterine contour evaluation.⁴ A pregnancy will alter the contour of the uterine fundus and repeat imaging after resolution of the pregnancy is more likely to result in an accurate diagnosis.

Incidence and associated anomalies

Studies using optimal imaging have identified uterine anomalies in 5.5% to 9.8% of the general population, in 16.7% of those with recurrent miscarriage, and in 24.5% of those with a history of miscarriage and infertility.⁵⁻⁸ A study of uterine evaluation during cesarean sections noted that 5% were found to have a uterine anomaly: 71.0% were septate uteri, 19.4% were bicornuate uteri, 6.4% were unicornuate uteri, and 3.2% were didelphic uteri.⁴

Anorectal, cardiac, limb, ear, skeletal, and especially renal anomalies have all been commonly associated with müllerian anomalies. Obstetricians should have a low threshold to consider renal imaging, as they may be present in 30% of those with fusion anomalies.^{9,10}

Obstetric complications

Arcuate uterus is the most common anomaly identified, but it does not appear to be associated with a higher rate of infertility or miscarriage by prospective studies.^{5,11} In general, the arcuate uterus is considered a benign



FIGURE 1. MRI images allow visualization of the patent and nonpatent uterine anatomy. This image is diagnostic for a left unicornuate uterus with patent vagina (outlined arrow) and a functional right noncontiguous uterine horn (solid arrow).^{2,3} (Image courtesy of Anne-Marie Amies Oelschlager, MD.)



FIGURE 2. A complete septate uterus. The arrow points to the septum at the level of the internal os of the cervix. The septum extends into the cervix. Note that the leading angle of the septum in the endometrial cavity is less than 90°, and there is a flat external fundal contour.^{2,3} (Image courtesy of Anne-Marie Amies Oelschlager, MD.)

variant; however, the remaining uterine anomalies confer a significantly higher risk of obstetric complications.

The incidence of preterm birth before 37 weeks is approximately 40% for patients with known uterine anomalies. The highest risk of preterm labor is associated with uterine didelphys (56%), unicornuate uterus (43%), bicornuate uterus (39%), and septate uterus (31%).¹² Congenital uterine anomalies are also associated with increased risk of first and second trimester loss, cervical insufficiency, premature rupture of membranes, fetal malpresentation, fetal growth restriction, placental abruption, placenta previa, retained placenta, and cesarean birth (**Table**).¹³⁻¹⁵ For this reason, consultation with a maternal-fetal medicine specialist during pregnancy is recommended.

Management of obstetric risk

Limited data suggest that cervical length screening for short cervix (< 25 mm) and fetal fibronectin screening may be useful for predicting preterm birth in those with septate uteri. The data have not shown a consistent predictive benefit for those with unicornuate, didelphic, or bicornuate uteri.^{16,17}

Most studies of 17-hydroxyprogesterone caproate (17-OHPC) for prevention of premature birth did not include those with a history of congenital uterine anomalies. In a small retrospective cohort study of 48 women with congenital uterine anomalies, there was an equally high rate of recurrent preterm birth in those who received 17-OHPC and those who did not. 17-OHPC has not been demonstrated to prolong pregnancy in women with uterine malformations or prenatal diethylstilbestrol exposure.¹⁸

Patients with a history of cervical atresia may have had reconstructive surgery to create a patent cervix. In those who can become pregnant, case reports

have suggested that abdominal cerclage with planned cesarean delivery is warranted.¹⁹ There are limited data to suggest that cerclage may decrease the rate of preterm delivery in those with bicornuate uteri with cervical incompetence.²⁰

Historically, hysteroscopic uterine septum resection has been recommended for those with a history of recurrent pregnancy loss, preterm labor, and infertility.²¹ In case control studies, hysteroscopic metroplasty has been associated with a higher live birth rate and lower spontaneous abortion rate but has also been associated with higher rates of preterm delivery, cesarean section, and fetal malpresentation compared with those with uterine septum who did not have surgery.^{22,23} A multinational multicenter trial randomly assigned 80 women with a septate uterus to septum resection or expectant management. Live births occurred in 31% who had septum resection compared with 35% who did not have surgery.²⁴ It appears that uterine septum resection may benefit select patients with recurrent fetal loss, but their risk of obstetric complications is still significantly elevated. If the septum extends into the cervix, resection of the cervical component has not been typically recommended due to concern for an increased risk of cervical incompetence.

Delivery considerations

The incidence of cesarean delivery in those with uterine anomalies is

TABLE. Risk of Obstetric Complication for Those With Identified Uterine Anomalies vs Those Without Identified Uterine Anomalies¹³⁻¹⁵

Obstetric outcome	Odds ratio
First trimester loss	1.8
Second trimester loss	2.9
Placenta previa	4.0-5.8
IUGR	1.5-3.8
IUFD	1.6
Abruption	1.9-5.2
PTD < 34 weeks	4.9
PTD < 37 weeks	3.0-3.9
PPROM	2.5-3.0
Cervical insufficiency	15.1
Fetal malpresentation	7.9-11.1
Cesarean delivery	2.1-13.5
Retained placenta	1.7

IUFD, intrauterine fetal demise; IUGR, intrauterine growth restriction; PPRM, preterm premature rupture of membrane; PTD, preterm delivery.

significantly higher, likely because of a combination of higher rates of abruption, preterm labor, and malpresentation. The risks of abruption and failure of external cephalic version (ECV) may be higher in patients with uterine anomalies; however, there have been case reports of successful ECV.²⁵

If a longitudinal vaginal septum is identified during a pregnancy, the septum can be resected prior to labor in the operating room or during the first stage or early second stage of labor prior to pushing. Clamping and resecting the intervening septal tissue should be done with extreme care to avoid the bowel, urethra, and the cervix. Use of the LigaSure device (Medtronic) has also been described.²⁶ After vaginal delivery, the vagina should be carefully inspected, hemostasis assured, and gaps in the mucosa should be reapproximated using absorbable suture.²⁷

Many patients with obstructive uterovaginal anomalies will present in adolescence or prior to puberty. Some patients may have had vaginal septum resection for a transverse vaginal septum or may have had a vaginal pull-through procedure for distal vaginal atresia. Interposition vaginal grafts, including buccal mucosa or bowel grafts, may have been used to bridge the gap between the upper and lower vagina. In these situations where interposition grafts or residual septal tissue is present, and there is a concern for vaginal obstruction, planned cesarean delivery is recommended. Additionally, for those who have had anorectal malformations with a history of anoplasty or reconstructive urological procedures where continence may be negatively impacted by vaginal delivery, planned cesarean delivery should be offered. For any patient with



CLINICAL PEARLS

- Uterine anomalies are found in approximately 5% to 10% of the general population. One in 8 with recurrent miscarriage and 1 in 4 of those with a history of miscarriage and infertility have uterine anomalies.

- Optimal diagnostic tests include 3D transvaginal ultrasound, MRI, saline sonohysterography, laparoscopy or

laparotomy combined with hysteroscopy or hysterosalpingogram.

- Congenital uterine anomalies are associated with increased risk of first and second trimester loss, preterm labor, cervical insufficiency, premature rupture of membranes, fetal malpresentation, fetal growth restriction, placental abruption, placenta previa,

retained placenta, and cesarean birth. Consultation with a maternal-fetal medicine specialist is recommended during pregnancy.

- Ultrasound guidance is recommended for management of first and second trimester therapeutic and incomplete abortion, as well as for removal of retained placenta.

a uterovaginal anomaly, aberrant urologic anatomy, including pelvic kidneys and ectopic and duplicate ureters, are common. Obstetricians should be vigilant during operative deliveries to avoid injury to the urinary system.

Obstetricians should be prepared for an increased rate of retained placenta after vaginal delivery in patients with uterine anomalies. Ultrasound-guided management of instrumentation during removal of retained placental tissue is recommended.

Management of therapeutic and spontaneous abortion

For those with 2 cervices, a history of vaginal septum resection, or long interposition vaginal grafts, uterine evacuation can be challenging, with increased difficulty visualizing and dilating 1 or both cervices. For those with a uterine septum, bicornuate uterus, or uterine didelphys, care must be taken to avoid inadvertent instrumentation of the wrong cervix and uterine horn. For those with congenital reproductive tract anomalies, procedures should be performed with ultrasound guidance.²⁸ Operative hysteroscopy may facilitate guided entry into the cavity if dilation with ultrasound guidance alone is not successful.²⁹ Although rare, there are case reports of ectopic pregnancies occurring in asymmetric or noncommunicating uterine horns. MRI may be useful if a pregnancy is visualized by ultrasound but is not accessible through uterine instrumentation and cannot be visualized through hysteroscopy.³⁰

Conclusion

Uterine anomalies are common in the general population and are associated with a higher risk of obstetric complications. Optimal imaging is recommended for accurate diagnoses. Consultation with a maternal-fetal medicine specialist is recommended during pregnancy. ■

FOR REFERENCES VISIT

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Pregnancy-related homicides are leading cause of maternal death

by LINDSEY CARR, EDITOR

Women in the United States are more likely to be murdered during pregnancy or soon after childbirth than to die from the country's top 3 leading causes of maternal death—high blood pressure disorders, hemorrhage, or sepsis—according to research in the *BMJ*. Most of these pregnancy-related homicides result from a combination of intimate partner violence and firearms, according to Rebecca Lawn, PhD, of Harvard TH Chan School of Public Health, and colleagues.

They argue that society must address “male violence,” or the violence committed by men, to save the lives of hundreds of women in the United States every year. Although intimate partner violence is common worldwide, with 1 in 3 women reporting experiences of physical, sexual, or psychological abuse/violence by a partner in their lifetime, rates in the United States are significantly higher than rates in other high-income countries.

The homicides committed by an intimate partner in the United States are overwhelmingly tied to firearms, and homicides related to pregnancy are no different. Recent reports found that firearms were used in 68% of pregnancy-related homicides between 2008 and 2019, with Black women at a substantially higher risk of murder than White or Hispanic women. These domestic homicides are also associated with

state-level firearms legislation and gun ownership, and the plethora of loopholes allowing access to remain.

The recent decisions regarding women's reproductive rights in the United States place further emphasis on these issues, Lawn and her colleagues said. Specifically, reproductive coercion is a common aspect of intimate partner violence and increases the risk of unwanted pregnancy. Restricting access to abortion care further endangers women, as unwanted pregnancies can often amplify risks in abusive relationships.

Lawn and her colleagues pointed out that pregnancy typically means more contact with health care providers, which can provide opportunities for screening and other approaches to help women at risk of intimate partner violence. These interventions from doctors and health care teams may help stop a pattern of abuse that could eventually lead to homicide or other negative health outcomes, the authors said.

All causes of maternal mortality are important, the authors said, but the tragedy here is that pregnancy-associated homicide is preventable. They concluded with an emphasis on the need to end male violence, including gun violence, for the health and safety of women everywhere. ■

REFERENCE

1. Lawn RB, Koenen KC. Homicide is a leading cause of death for pregnant women in US. *BMJ*. 2022;379:o2499. doi:10.1136/bmj.o2499