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### Abdominal Imaging / Imagerie abdominale

# Pictorial Review of Complications of Uterine Anomalies

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Uterine anomalies are a potential cause for amenorrhea, infertility, and abnormal obstetrical outcomes in young women [1]. Uterine anomalies occur due to failed formation or fusion of the paired Müllerian ducts early in embryonic life and are also referred to as Müllerian duct anomalies. Uterine anomalies may also be associated with symptoms of pain or a mass due to hematometros, hematocolpos, endometriosis, or ectopic pregnancy [2]. The overall incidence of uterine anomalies is often quoted to be 2%-3%. However, the incidence is up to 5%-10% in women with recurrent first trimester miscarriage and up to 25% in women with recurrent second trimester pregnancy loss [1].

#### **Embryology of Müllerian Duct Anomalies**

The Müllerian ducts are present in both the male and female embryo before 6 weeks' embryonic life. Between 6 and 12 weeks embryonic life, the Müllerian ducts fuse to form the uterus, cervix, upper two-thirds of the vagina and fallopian tubes in the female embryo. In the male embryo, Müllerian inhibiting substance produced by sertoli cells in the testis inhibits Müllerian duct development beyond 6 weeks [1,2]. The lower vagina is formed from sinovaginal buds after fusion with the caudal most portion of the Müllerian ducts forming the vaginal plate [1]. The ovaries arise from the primitive yolk sac [2].

#### Müllerian Duct Classification

Müllerian duct anomalies are classified by the American Society of Reproductive Medicine 1988 classification scheme

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(Figure 1) [3-5]. Type 1 anomalies are due to hypoplasia or agenesis of the vagina, cervix, fundus, tubes, or a combination of these structures. Type 2, 3, and 4 anomalies are due to failed fusion of the Müllerian ducts. Type 2 anomalies are variants of a unicornuate uterus including a unicornuate uterus without a rudimentary horn and a unicornuate uterus with a rudimentary horn. The rudimentary horn may or may not contain functioning endometrium. A rudimentary horn that contains functioning endometrium may or may not communicate with the cervix. Type 3 anomalies are complete failure of fusion of the Müllerian ducts resulting in uterine didelphys with 2 separate vaginas (the upper two-thirds), 2 separate cervices, and 2 separate uteri. Type 4 anomalies are incomplete fusion of the Müllerian ducts resulting in a bicornuate uterus with 2 separate uterine cavities and 1-2 cervices. There is only 1 vagina. Type 5 and 6 anomalies are due to failed resorption of the septum after the Müllerian ducts have fused. Type 5 anomalies are the septate uterus with a septum that may be composed of muscle and/or fibrous tissue. Type 6 anomalies are the arcuate uterus where there is thickening of the myometrium at the fundus but without a septum. Type 7 anomalies are teratogenic effects from diethylstilbesterol that results in a small uterus with a narrow irregular endometrial canal that has a T-shape [6].

#### **Imaging Evaluation of Uterine Anomalies**

Women with uterine anomalies may present with amenorrhea, infertility or abnormal obstetrical outcomes including recurrent miscarriages, second trimester pregnancy loss, preterm delivery, malpresentation, and intrauterine fetal demise [1]. They may also present with symptoms of pain or a mass due to endometriosis, hematocolpos, or hematometros [2]. Some women are asymptomatic and the uterine anomaly is

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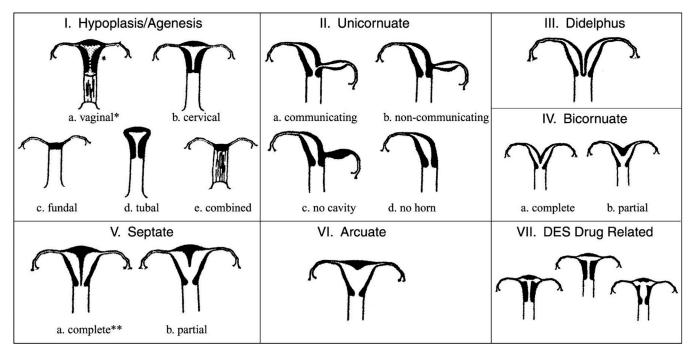


Figure 1. Classification of Müllerian duct anomalies by the American Society of Reproductive Medicine 1988. \*Uterus may be normal or take a variety of abnormal forms. \*\*May have 2 distinct cervices. DES = diethylstilbestrol. Reproduced from *Fertility and Sterility* [3] with permission from Elsevier. Image provided by the Radiological Society of North America [5].

diagnosed on imaging performed for an unrelated problem. Women with primary amenorrhea are often first evaluated with ultrasound (US). Infertility is often due to blocked fallopian tubes and is evaluated early with hysterosalpinography. Women with abnormal obstetrical outcomes may be evaluated with magnetic resonance imaging (MRI) after their obstetrical US. The single best imaging test to diagnose uterine anomalies is MRI [7]. MRI is performed with T2weighted imaging through the pelvis in 3 planes with oblique coronal imaging through the uterus to evaluate the external fundal contour. T2-weighted imaging is best performed without fat suppression to take advantage of the natural contrast between the signal intensity of the uterus and the surrounding fat. Axial T1-weighted imaging is performed to evaluate for blood products that may be seen with endometriosis and hematometrocolpos. Fat suppressed axial T1-weighted imaging is used to differentiate blood products from an incidental dermoid cyst and may be performed as a spoiled gradient echo sequence to save time. Gadolinium-enhanced imaging is not necessary to evaluate uterine anomalies. 3D US has also been shown to be reliable in evaluating the internal morphology and external contour of the uterus to diagnose uterine anomalies in recent studies [8,9]. The imaging evaluation of uterine anomalies should include limited imaging of the kidneys since renal anomalies occur in 30-50% of cases, most commonly unilateral renal agenesis [1,10,11]. Limited MRI of the kidneys may be performed as a large field of view coronal T2 breath-hold sequence. The results of imaging are combined with the findings on pelvic exam and in some cases hysteroscopy and laparoscopy to arrive at a precise diagnosis [7].

#### **Examples of Uterine Anomalies**

Primary amenorrhea is often first evaluated with US followed by MRI. Complete Müllerian duct agenesis is Mayer-Rokitansky-Kuster-Hauser syndrome characterized by an absent uterus, cervix and upper two-thirds of vagina. Patients may still have ovaries because the ovaries arise from the primitive yolk sac [2]. There may be a lower 1/3 to the vagina because it forms from the sinovaginal bud (Figure 2) [1,2]. Other cases of primary amenorrhea may be due to partial agenesis or hypoplasia of the Müllerian ducts as shown in Figures 3 and 4. Endometriosis is commonly encountered in uterine anomalies due to obstructed flow and retrograde menstruation (Figure 3) [2]. Hematometros and hematosalpinx may also occur due to obstructed menstruation (Figure 4). Women with agenesis or rudimentary unilateral uteri have an increased incidence of absent or ectopic ovaries that may be located anywhere between their position of origin near the kidneys to the inguinal canal (Figure 3) [1,10,12,13].

Unicornuate uterus has been implicated in miscarriage, cervical incompetence, malpresentation, preterm labor, and intrauterine growth restriction [1]. The small size of the unicornuate uterus is not a favorable environment for development of a pregnancy, and the live birth rate is only 50% [1]. Other patients may present with symptoms due to the rudimentary horn. A total of 74% of patients with a unicornuate uterus have a rudimentary horn (Figure 5) [4]. A nonfunctioning rudimentary horn without an endometrium is of no clinical significance. However, a functioning rudimentary horn with an endometrium requires surgical removal

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