

MRI for the diagnosis of uterine anomalies

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Congenital uterine anomalies
and reproductive outcome

Purpose

To present a pictorial review of typical Müllerian duct anomalies diagnosed on MRI in order to highlight its role and to encourage its widespread use.

Background

Mullerian duct anomaly:

- Developmental cause of infertility and amenorrhea
- Classified into various structural abnormalities according to the time of developmental interruption
- Correct classification crucial for determining indications for surgery and for planning surgical approach
- Accurate classification might not be possible in up to 25% of the cases

Background

Müllerian duct anomalies (MDAs) result from non-development or partial or complete nonfusion of the Müllerian ducts. They occur in 1-15% of women.¹

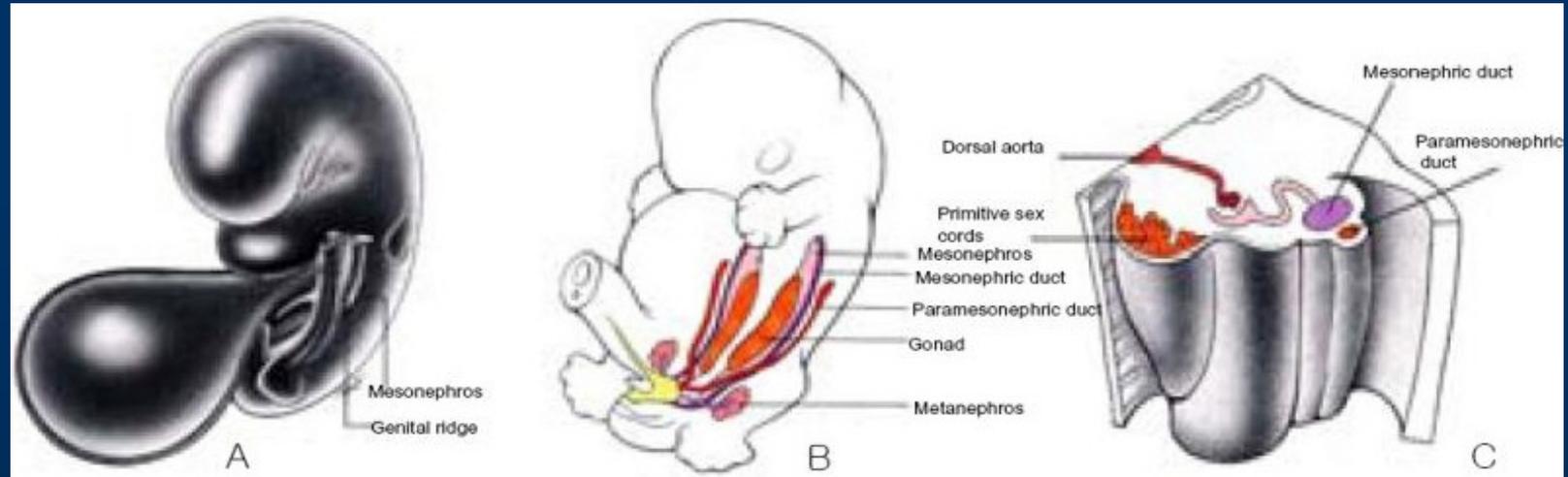
MDAs are clinically relevant because they are associated with an increased incidence of impaired fertility and menstrual disorders.² In particular, women with MDAs have a significant risk of obstetric complications, such as spontaneous abortion, stillbirth, and preterm delivery.^{3,4}

MDAs may be associated with renal anomalies, particularly renal agenesis or ectopia which occurs in 50% of patients with vaginal agenesis, and may be seen in obstructed duplications.^{5,6}

Background

Ultrasound, hysterosalpingography and laparoscopy or surgery have until now been the mainstays for the diagnosis of MDAs.⁷ All of these modalities have inherent limitations, however, particularly in the differentiation between septate and bicornuate uteri.⁸

MRI has been shown to be an accurate and non-invasive method for the evaluation of MDAs.^{9,10} MRI is also helpful in elucidating the etiology of obstructed MDAs and is particularly useful in patients in whom surgical unification is anticipated.^{11,12}

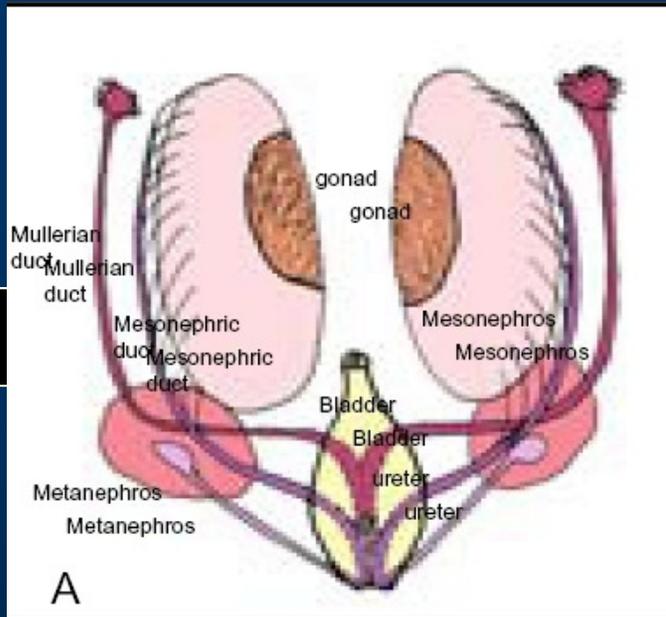


The embryonic development of the uterus, fallopian tube and vagina begins in the 3th or 4th week of gestation and progressing into the second trimester of pregnancy.

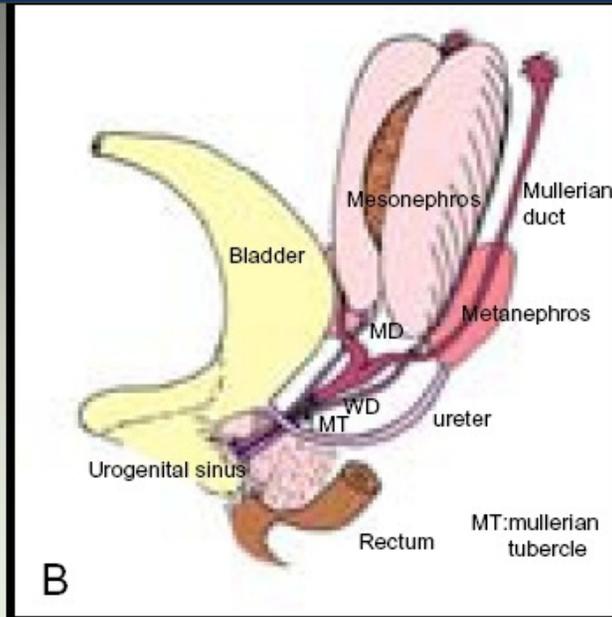
Gonadal development results from a migration of primordial germ cells into the genital ridge. The development of uterus, fallopian tubes, cervix and upper two thirds of vagina results from formation and reshaping of the Mullerian duct (paramesonephric duct). These ducts are invaginations of coelomic cavity, which rapidly form into tubular structures, the paramesonephric ducts.

Paramesonephric ducts open into the coelomic cavity and parallel the mesonephric ducts in caudal development.

7-8 weeks

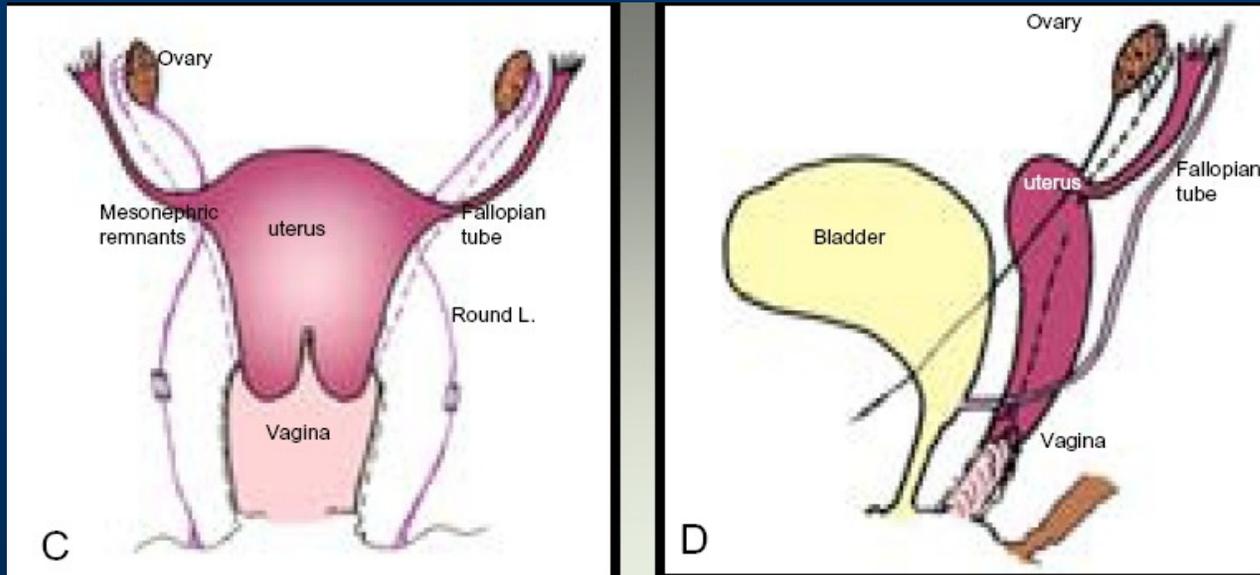


9 weeks



images from Langman's Medical Embryology

The paramesonephric ducts fuse in the midline in their caudal region forming the uterus cervix and upper two thirds of vagina. The fusion is followed by a slow resorption of the intervening wall septum, forming a single cavity. Initially the uterus is bicornuate and by the third month, the fundus begins, creating the morphology of adult uterus. A mesenchymal coat covers the fused Mullerian ducts giving rise to the myometrium and perimetrium. The cranial portions of the unfused ducts develop into the fallopian tubes.



The tip of the fused paramesonephric ducts project into the genital ridge caudally forming the paramesonephric tubercle. The tubercle reaches the urogenital sinus, dense column of cells is formed, creating the vaginal plate. The bulbs proliferate in a cranial direction, separating the uterus from urogenital sinus. This movement of tissue results in canalization and formation of vagina. Remnants of the paramesonephric systems at the tip of cervix, anastomose with sinus fat and create the vaginal fornices. Full canalization of the Fallopian tubes, uterus, and vagina is complete by the 18th week of fetal life.

Classification of Müllerian duct anomalies¹³

- **Class I: Segmental Agenesis or Hypoplasia**

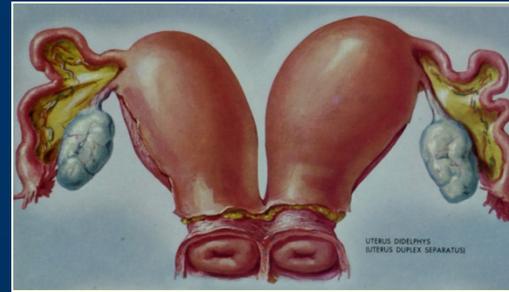
- A. Vaginal
- B. Cervical
- C. Fundal
- D. Tubal
- E. Combined

- **Class II: Unicornuate**



- A.1. Rudimentary horn contains endometrium.
Horn may or may not communicate with main uterine cavity.
- A.2. Rudimentary horn without endometrium
- B. No rudimentary horn

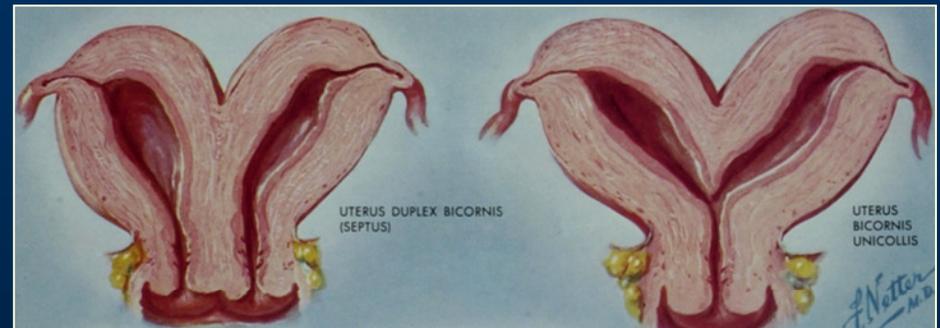
- **Class III: Uterus Didelphys**



- **Class IV: Bicornuate**

A. Complete - division down to internal os

B. Partial



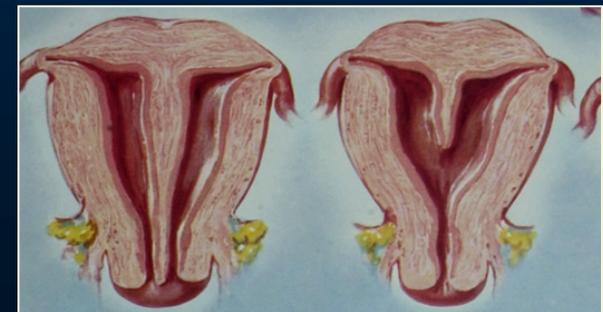
- **Class V: Septate**

A. Complete down to internal/external os

B. Incomplete

- **Class VI: Arcuate**

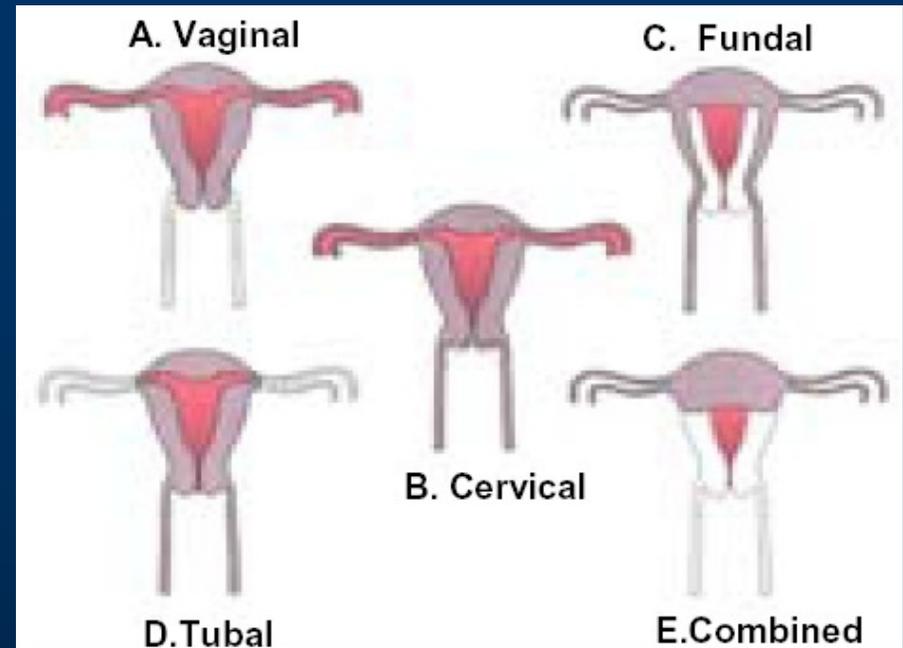
normal contour, small fundal cleft



- **Class VII: T-shaped uterus** < use of DES

images from F.H. Netter

**Class I:
Segmental Agenesis or
Hypoplasia**



Complete or partial agenesis of the upper genital tract with associated skeletal anomalies is the Mayer-Rokitansky-Kuster-Hauser syndrome, thought to result from excess Müllerian inhibiting hormone.

The incidence of vaginal agenesis is about 1:4000 live births. The ovaries can be normal because they are derived separately from the mesodermal epithelium.

MRI shows the absent vaginal stripe and permits evaluation of the status of the uterine size and endometrial stripe.

Class I: Segmental Agenesis or Hypoplasia

Agenesis of the distal one third of the vagina is due to the failure of the urogenital sinus to develop.

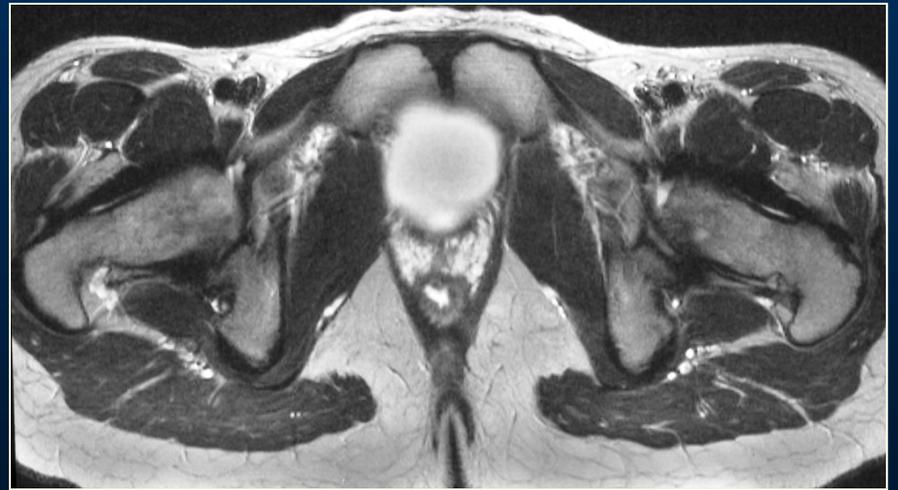
In adolescence, patients present with amenorrhea and an enlarged uterus.

The differential diagnosis is an imperforate hymen.

Axial images are best for revealing the absence of the vagina.

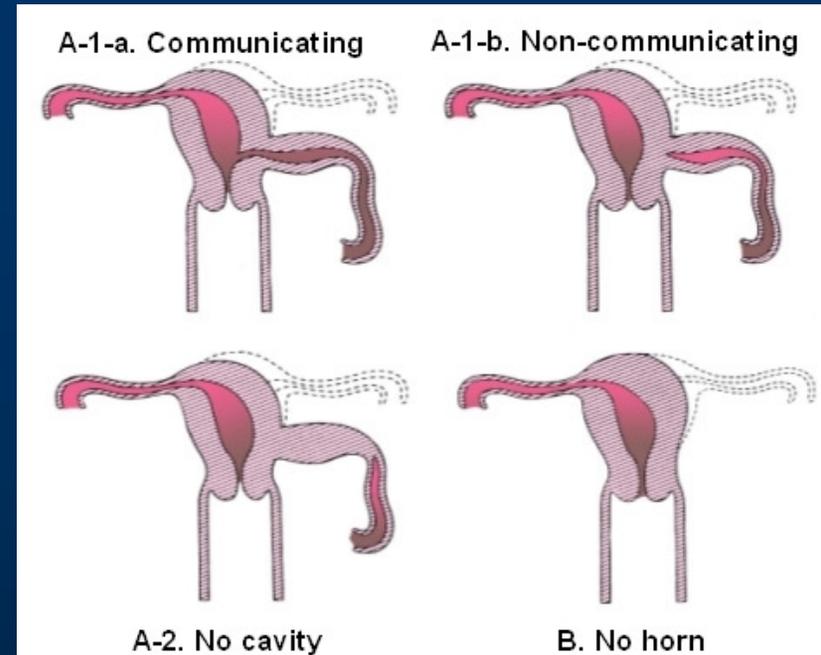
The uterus is usually markedly dilated and filled with blood that shows high signal intensity on T1-weighted and T2-weighted images.

Surgery to create a neovagina is often performed.

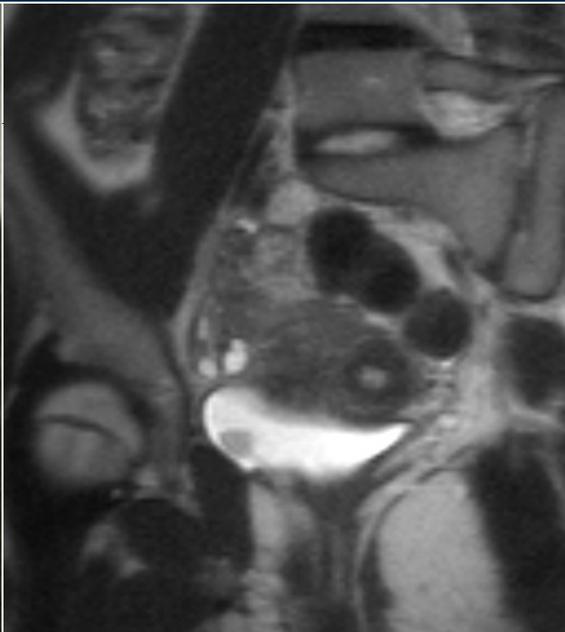
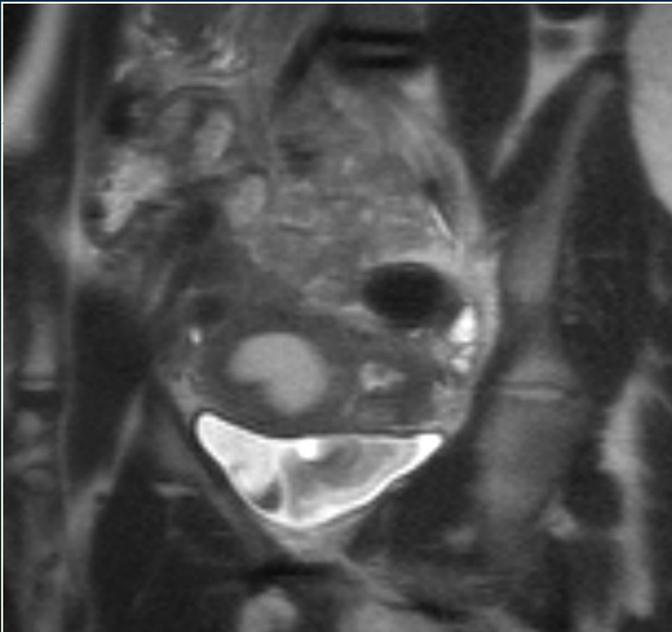
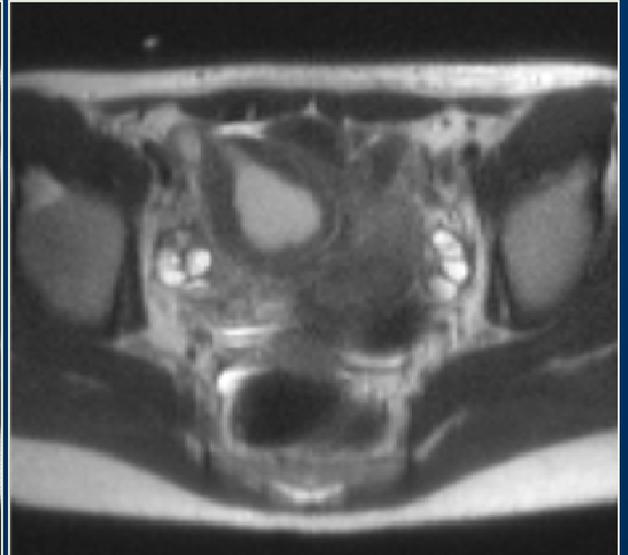
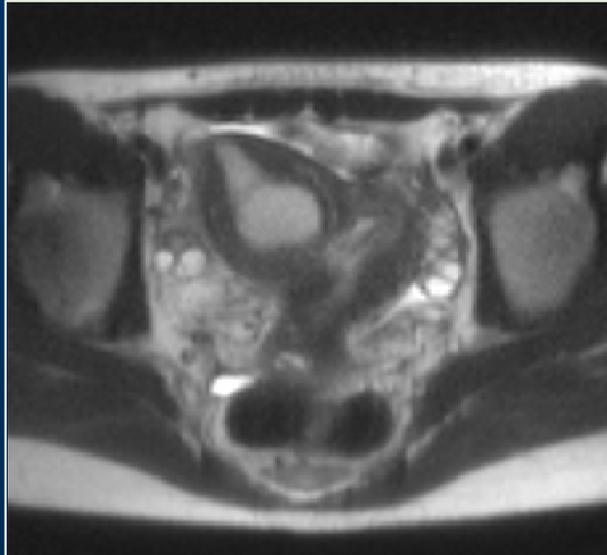
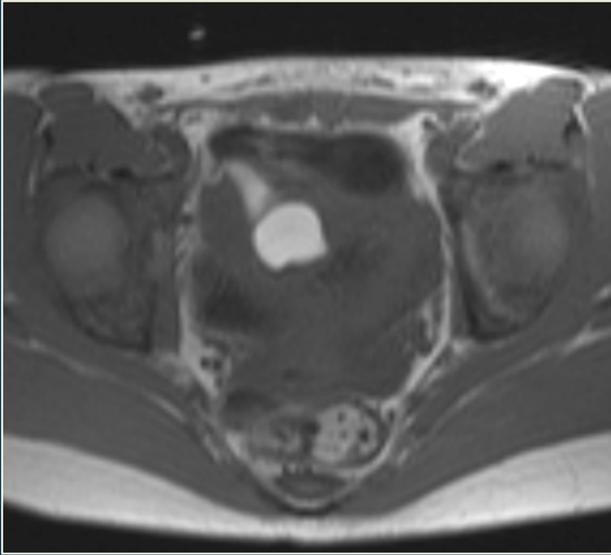


segmental vaginal hypoplasia
with hematometra,
and left sided hematosalpinx

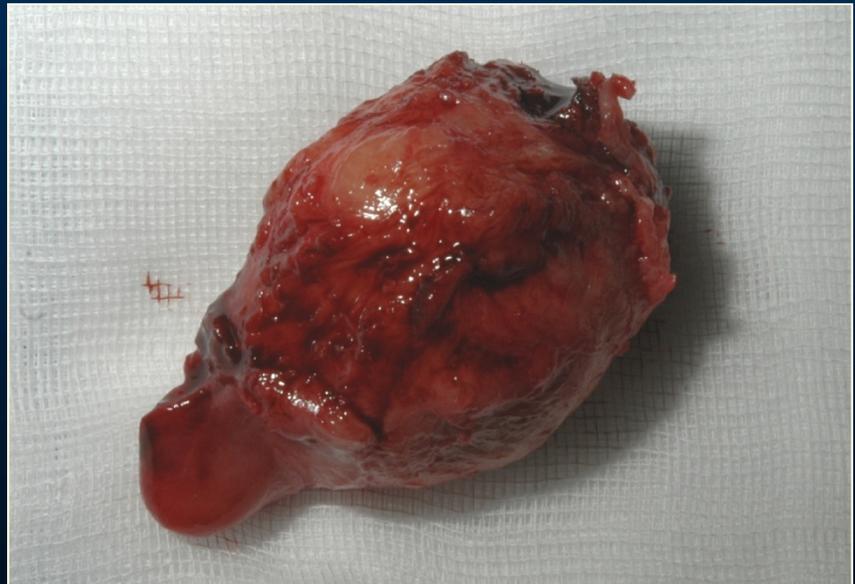
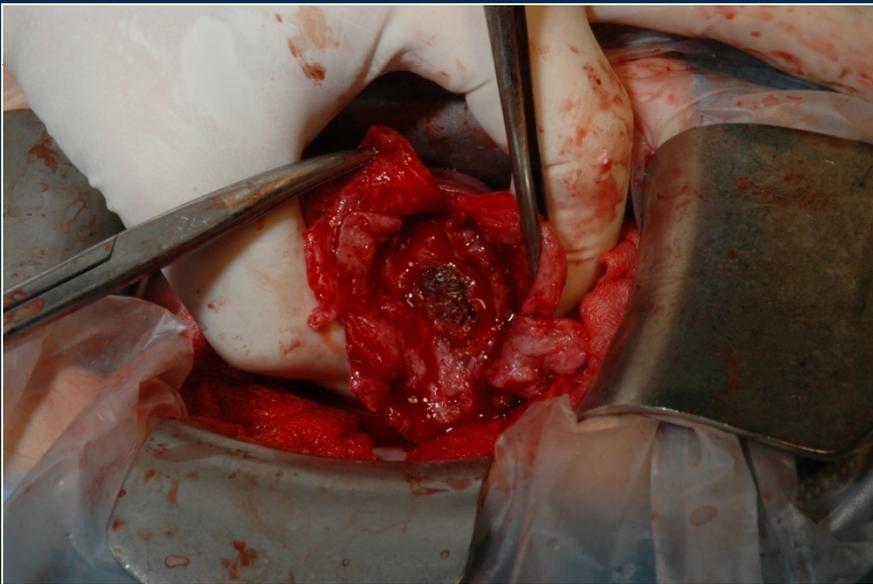
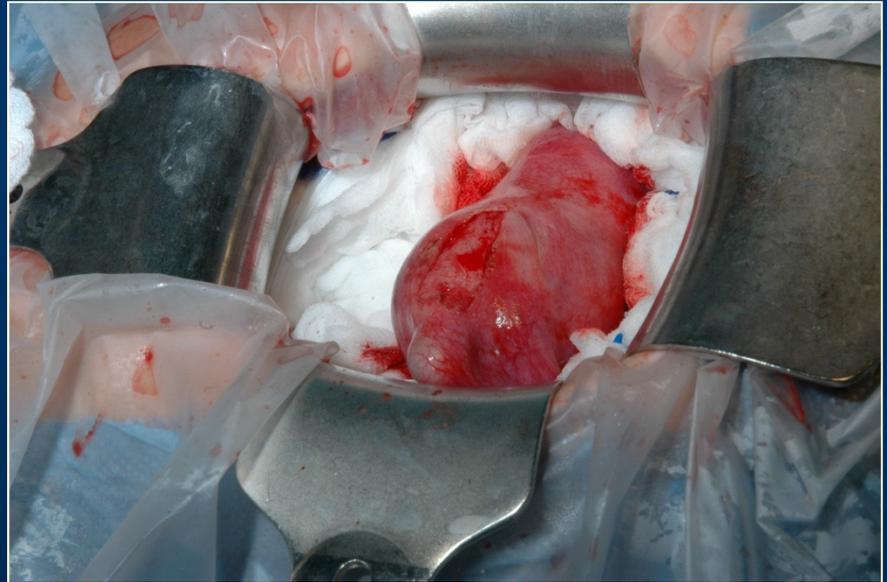
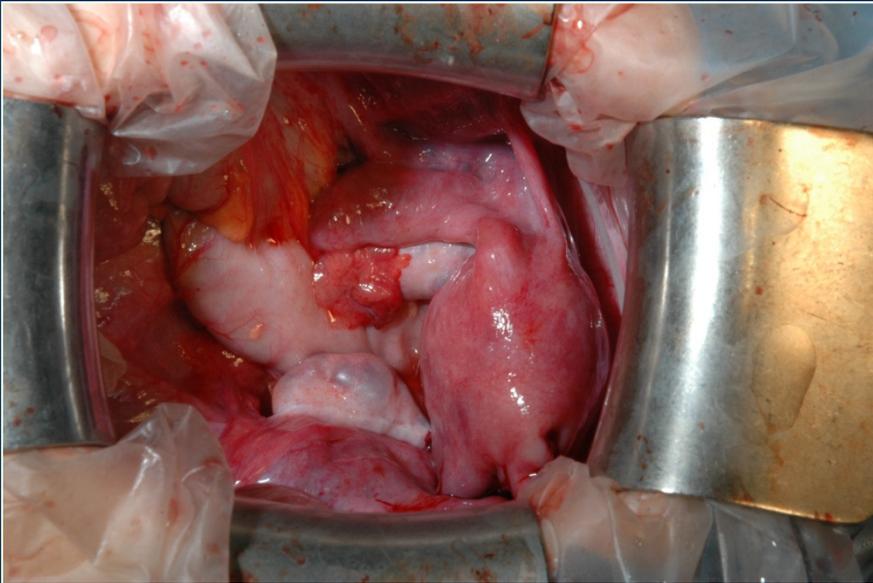
Class II: Unicornuate uterus



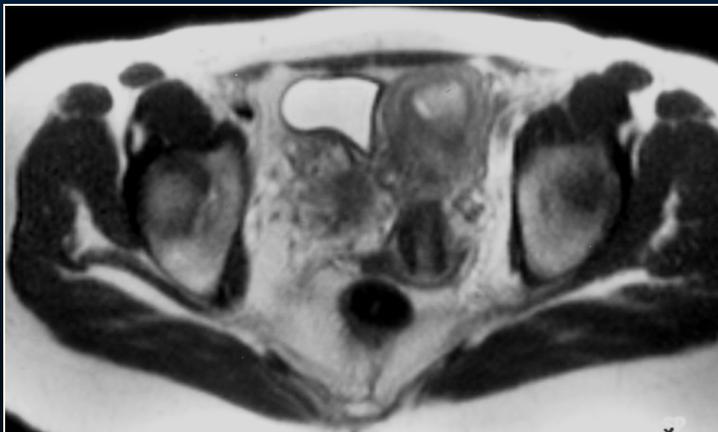
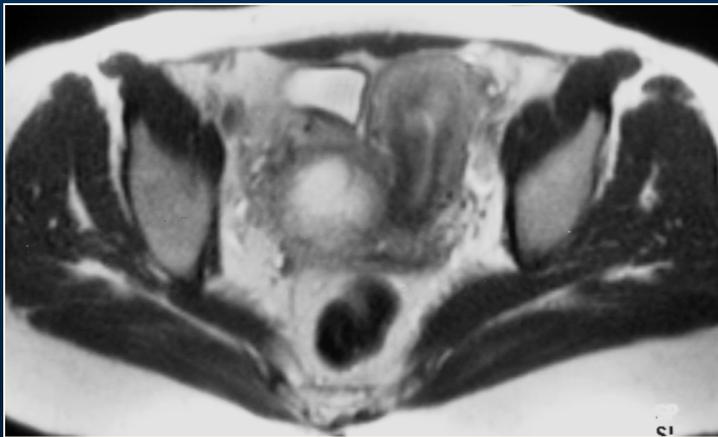
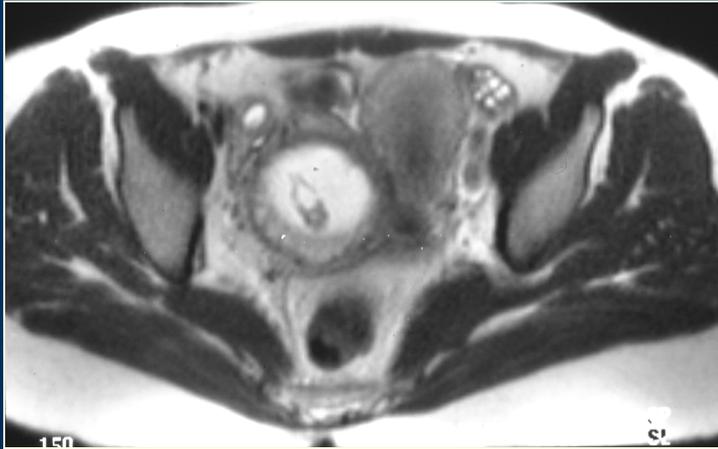
Failure of one Müllerian duct to develop causes the formation of a single, small lateroflexed uterine cavity (20%). A rudimentary horn is often present (65%), which may be communicating or noncommunicating. On HSG, contrast filled endometrial cavity has a fusiform shape and tapers at its apex, where it joins with a single fallopian tube. The MR imaging criteria for a unicornuate uterus was an elongated, curved uterus, whose external configuration appeared banana shaped. The endometrial/myometrial width and ratio were normal.



unicornuate uterus with rudimentary horn at the right side with endometrial tissue resulting in hematometra and hematosalpinx

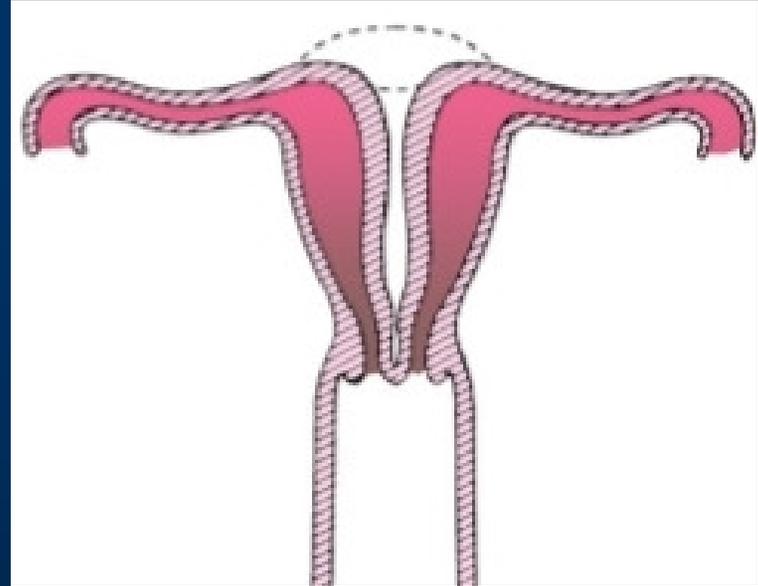


resection of the rudimentary horn.



unicornuate uterus with pregnancy in the rudimentary horn containing endometrium without communication with main uterine cavity

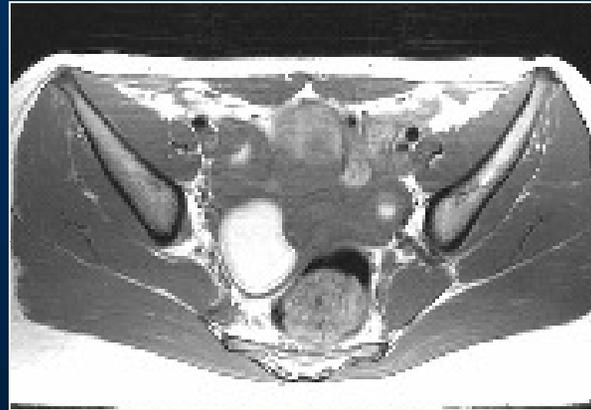
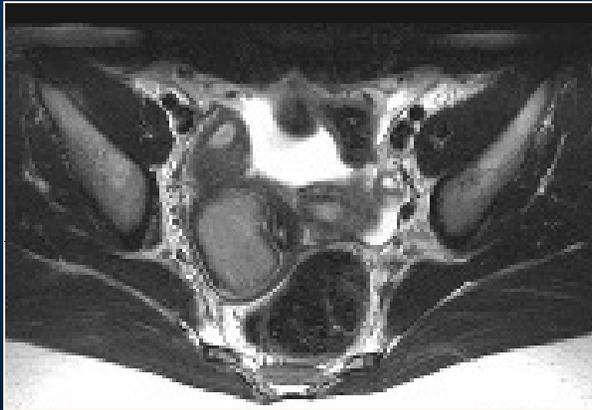
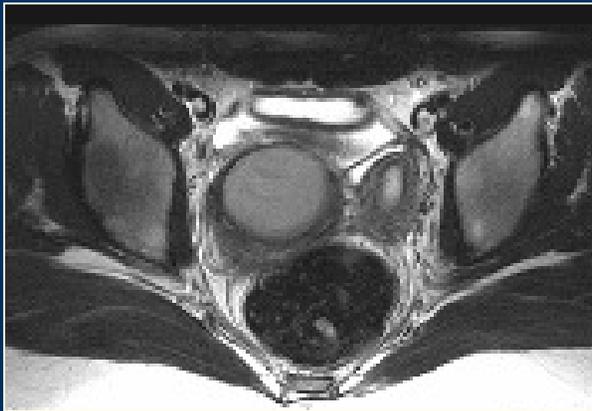
**Class III:
Uterus didelphys**



This class III anomaly is characterized by complete duplication of the uterine body and cervix which is due to a nonfusion of the Mullerian ducts (5%). A longitudinal vaginal septum is associated in 75%. These patients are usually asymptomatic unless there is an associated transverse vaginal septum causing hematometrocolpos. Complications of the uterine didelphys include preterm abortions (32-52%), premature labor (20-45%), fetal growth retardation, and malpresentation similar to the unicornuate uterus.

Class III: Uterus didelphys

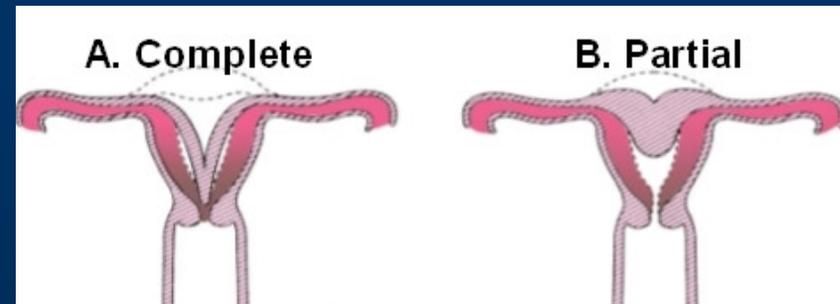
The MRI appearance is that of two separate uterine bodies, two cervixes, and often a vaginal septum. Unlike the longitudinal vaginal septum, if there is a transverse vaginal septum, there are usually symptoms of obstruction of the involved side. Vaginal septectomy may be performed. If a patient has a history of recurrent spontaneous abortion or premature labor attributable to the uterine anomaly, there can be an attempt at surgical union of the uterine bodies, but not the cervixes because an incompetent cervix will usually result. There is otherwise no surgical repair offered for the uterus didelphys.



uterus didelphys

with obstructing
transverse septum upper-third right vagina
resulting in hematocolpos and hematometra,
and ipsilateral renal agenesis (67%)

Class IV:
Bicornuate uterus

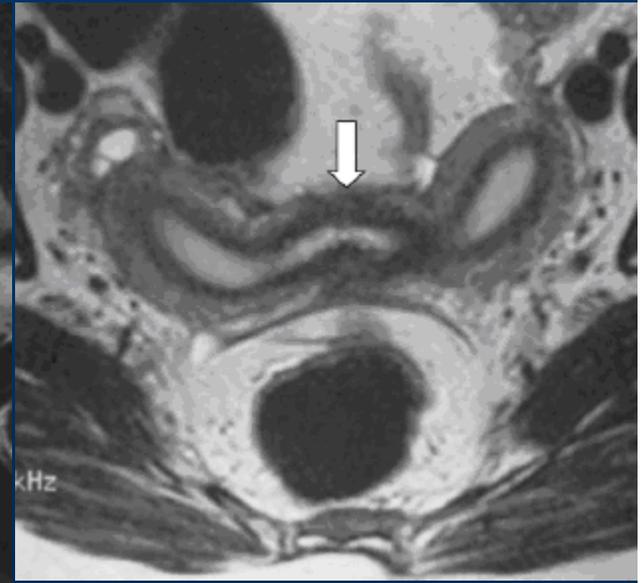
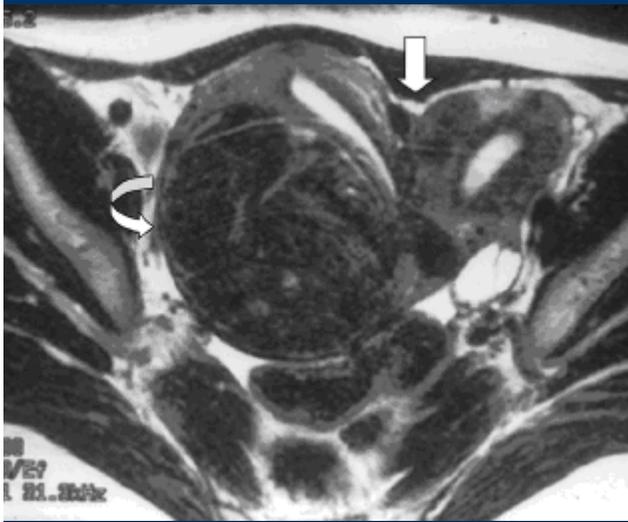


Incomplete fusion of the Müllerian ducts causes a bicornuate uterus (10%). Patients have two widely divergent ($>75\%$) uterine horns and a thick septum with MR imaging signal intensity usually equal to that of myometrium. The long axis view of the uterus (usually the off axis coronal) T2-weighted FSE scans are crucial to making the distinction. The fundal contour must be carefully inspected. There is deep indentation in the bicornuate (and also didelphys) uteri, but the septate uterus is usually flat or with a tiny indentation (less than 1cm). The bicornuate uterus has a wide intercornual distance.

Bicornuate uterus

Partial failure of Müllerian duct fusion. The resulting septum is composed of myometrium and surgery is required for correction. The septum may extend to the external os (**bicornuate bicollis uterus**) or to the internal os (**bicornuate unicollis uterus**). Spontaneous abortion rates: 28-35% , premature birth rates: 14-23%, and fetal survival rates: 57-63%. Highest associated prevalence (38%) of cervical incompetence.

MRI: increased intercornual distance, outward fundal concavity with a cleft of at least 1.0 cm, the horns demonstrate normal uterine zonal anatomy, normal endometrial and myometrial width and ratio



uterus bicornuate

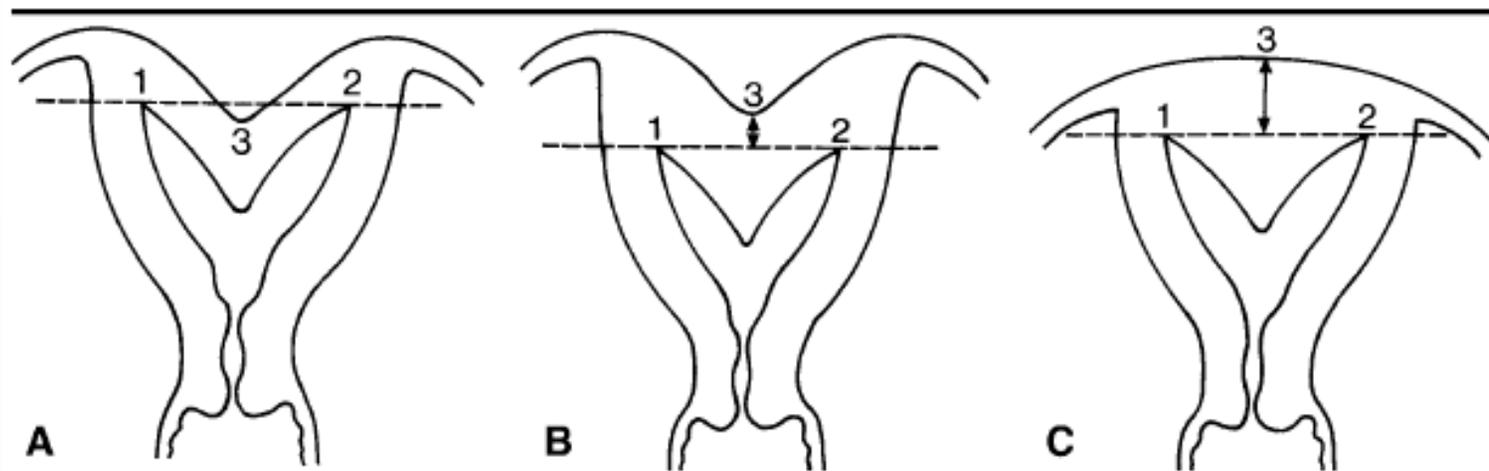
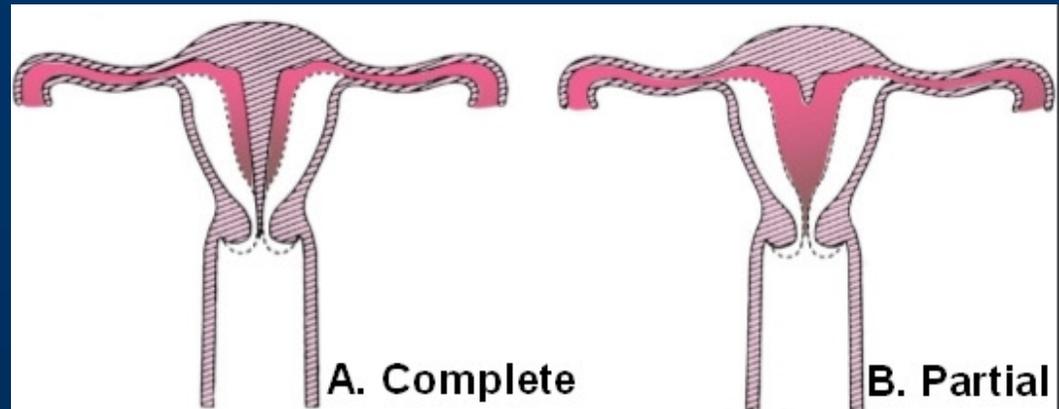


Figure 7. Classification criteria for US differentiation of septate from bicornuate uteri. A, When apex (3) of the fundal external contour occurs below a straight line between the tubal ostia (1, 2) or, B, 5 mm (arrow) above it, the uterus is bicornuate. C, When apex is more than 5 mm (arrow) above the line, uterus is septate.

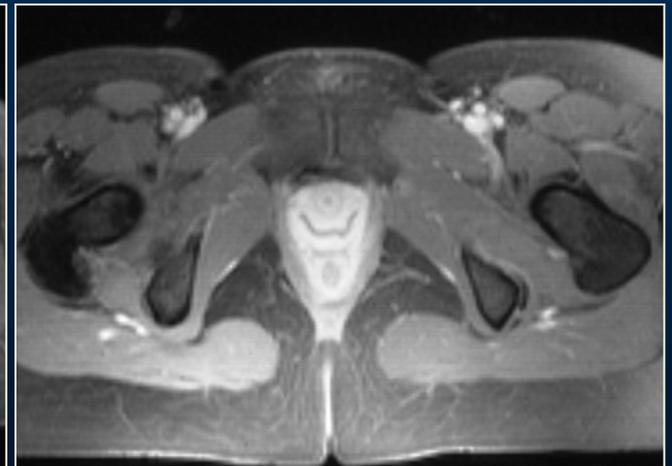
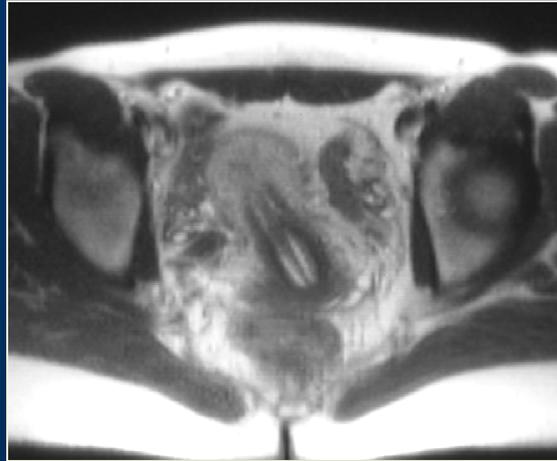
From Troian RN et al. Radiology, 2004;233:19-34

**Class V:
Uterus septate**

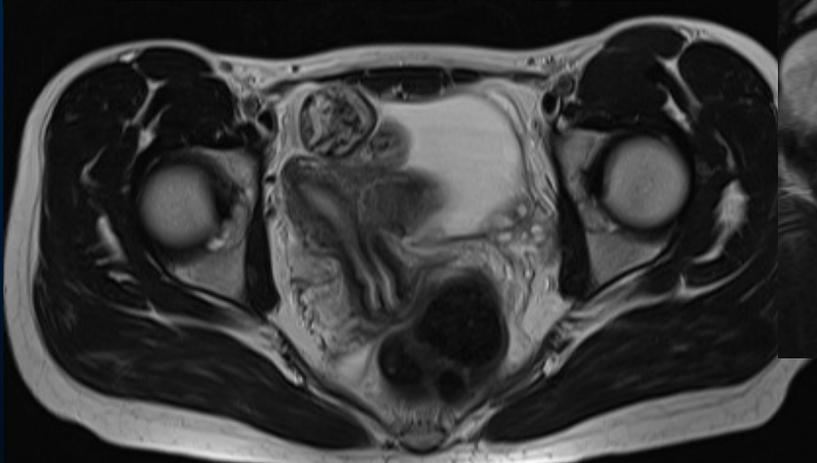
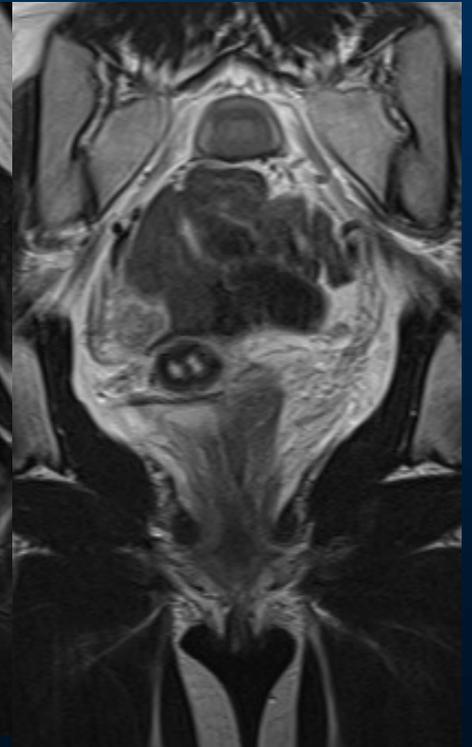
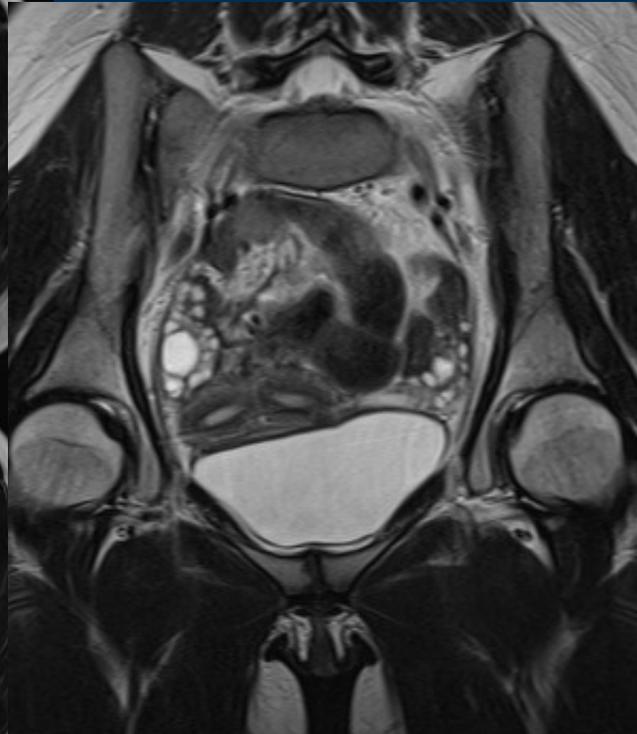
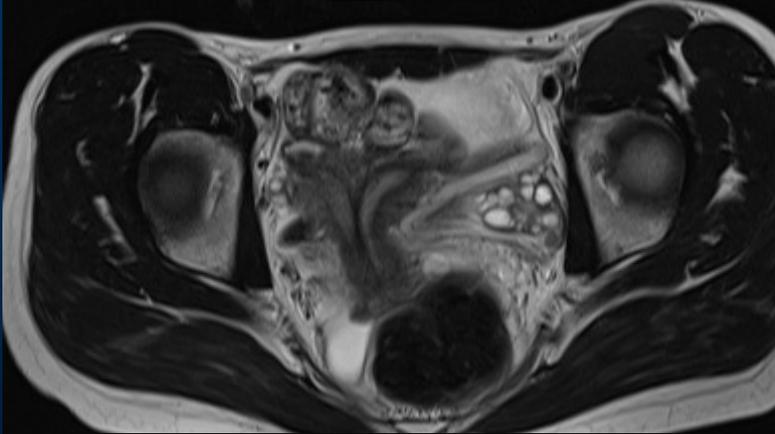
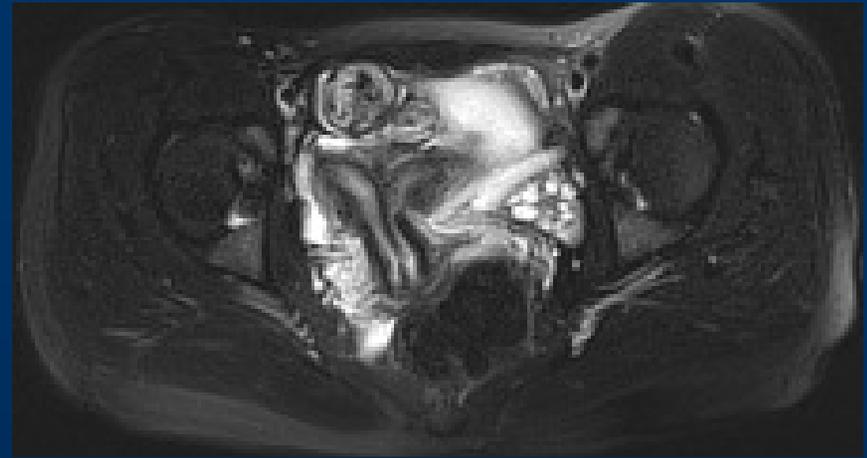


The class V septate uterus is the most common Müllerian anomaly (55%), and is due to partial or complete failure of resorption of the mid-line uterovaginal septum. It is associated with a very high incidence of preterm labor and spontaneous abortion (26-94%) usually in the mid-trimester.

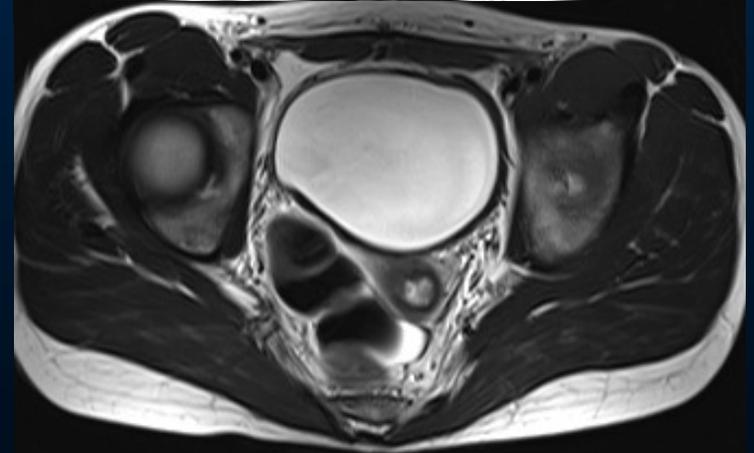
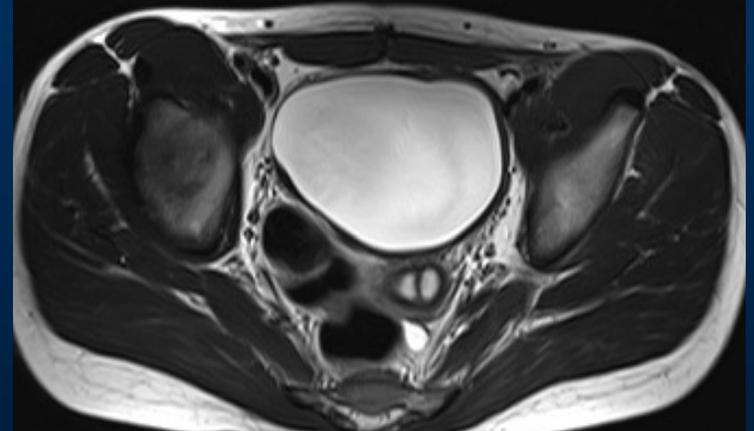
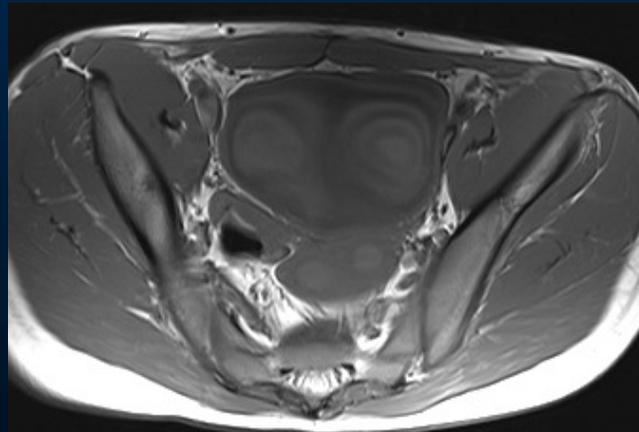
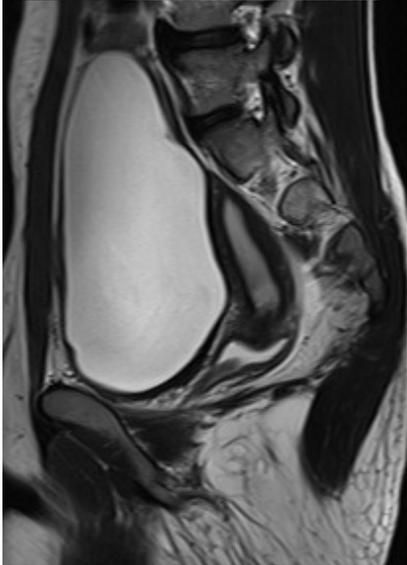
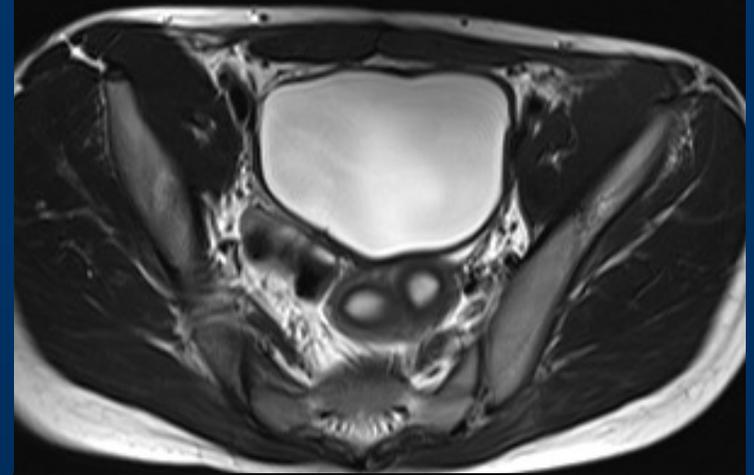
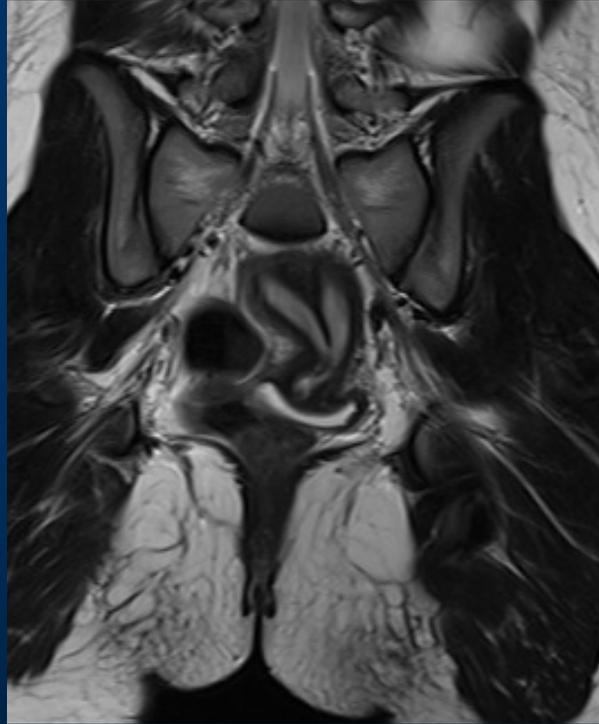
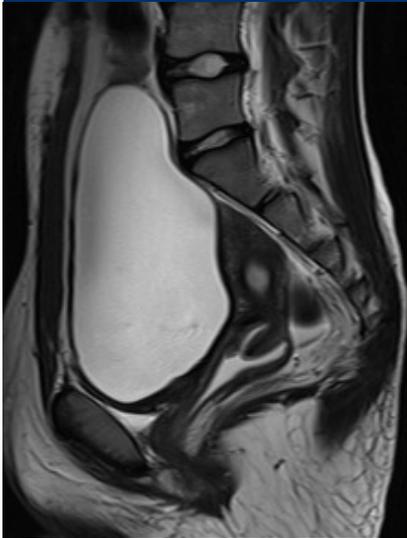
On MRI, generally normal sized uterus, two high signal endometrii are seen and only low signal intensity zone separates the relatively small cavities, characteristic of fibrous tissue in MRI. The septum may be composed of low signal intensity fibrous tissue, intermediate SI myometrium or both. *Intercornual distance shows normal (2-4cm) (HSG criterium).* The fundal contour is normal, with an outward fundal convexity.



uterus septate

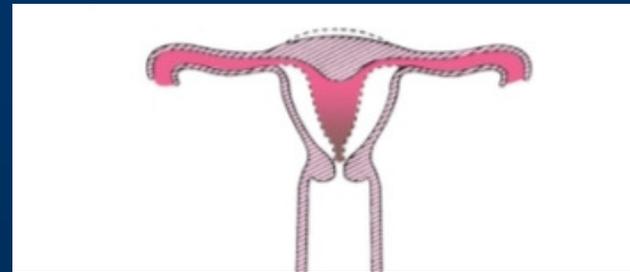


uterus septate (complete)

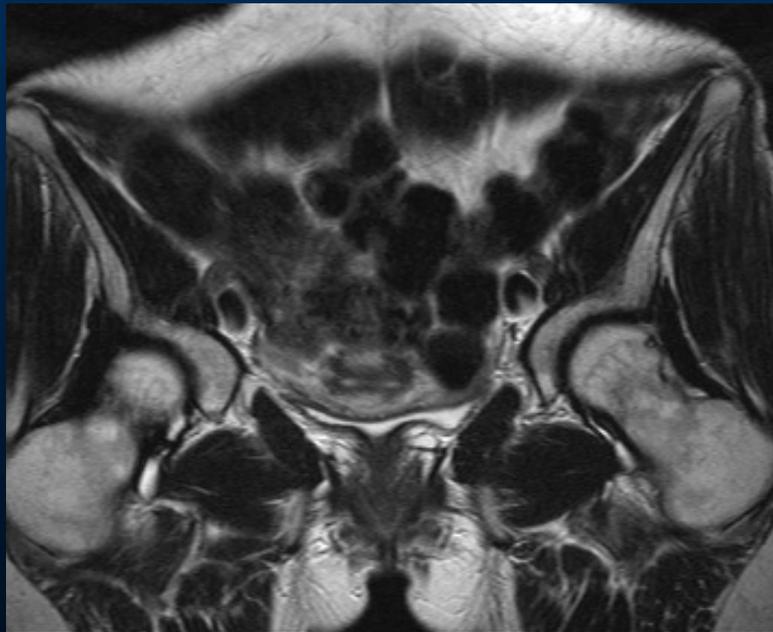
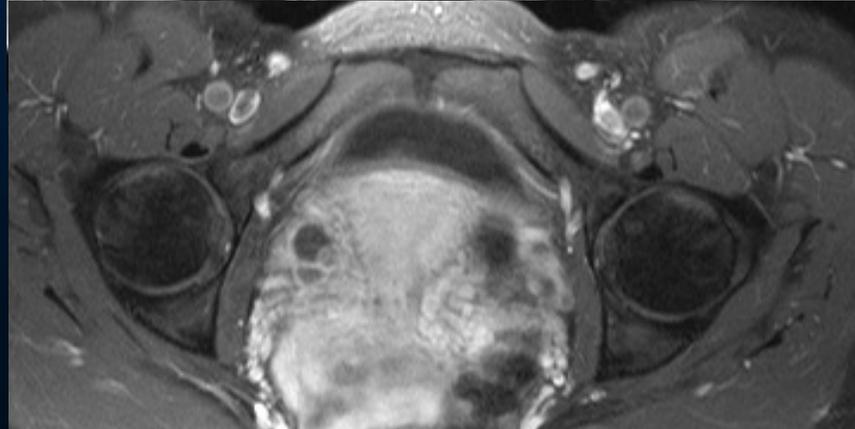
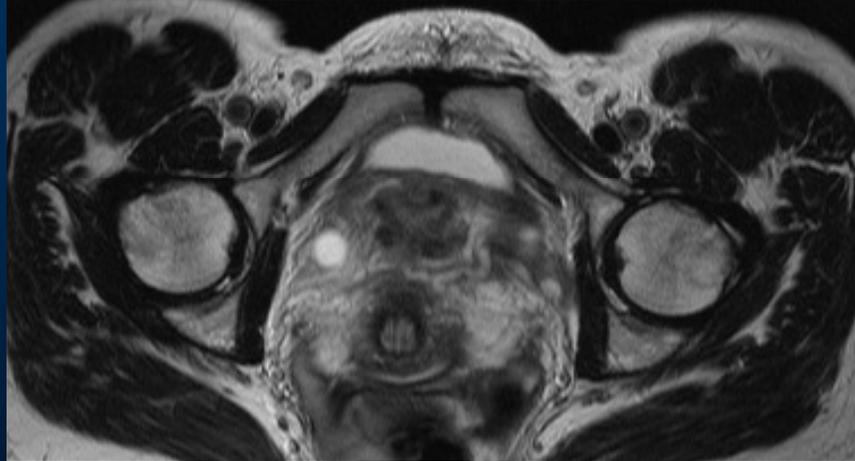
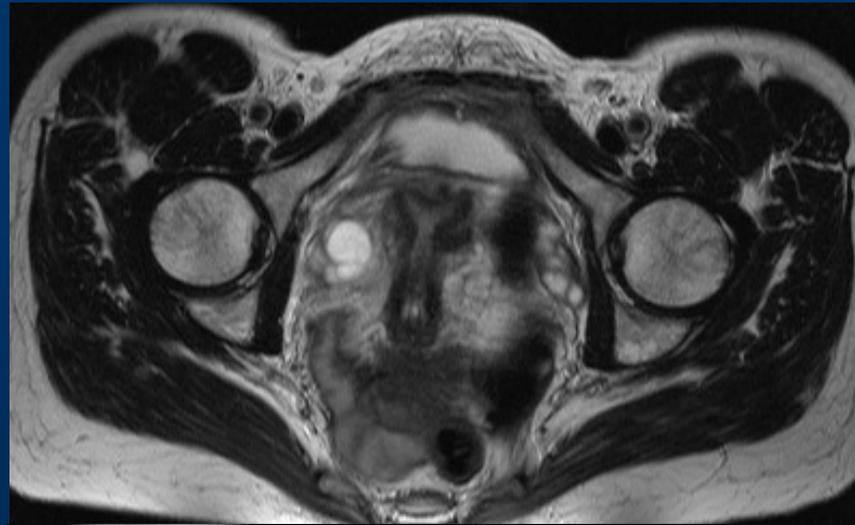
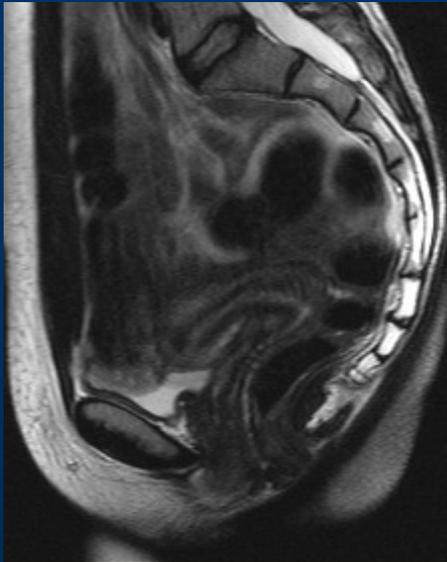


uterus septate (partial)

Class VI: Arcuate uterus



Arcuate uterus is the result of a nearly complete resorption of the uterovaginal septum. This is a rather insignificant anomaly of the uterine cavity in which, generally, no abnormalities in the contour of the uterus are visible. The long axis view of the uterus (usually the off axis coronal) T2-weighted FSE scans are crucial to making the distinction. The fundal contour must be carefully inspected. There is deep indentation in the bicornuate (and also didelphys) uteri, but the septate uterus is usually flat or with a tiny indentation (less than 1cm). The bicornuate uterus has a wide intercornual distance.



uterus arcuate

Class VII: DES uterus (diethylstilbestrol exposure)

Between 1945 and 1970, DES a synthetic estrogen, was prescribed to many pregnant women for treatment of threatened abortion and severe hyperemesis gravidarum. The drug was ineffectual in the prevention of miscarriages and was banned in 1971; however, DES was later found to have a profound effect on the development of the Müllerian system in female fetuses. DES-related Müllerian defects were once very common, accounting for approximately 20% of Müllerian anomalies. However, with the decrease in the size of the DES-exposed population after the drug was removed from the market, the incidence of these anomalies has since declined. For female offspring exposed to DES in utero, an increased incidence of vaginal and cervical clear-cell adenocarcinoma have been documented. Structural abnormalities have been observed in up to two thirds of women exposed to DES in utero making it the single most common cause of Müllerian anomalies.

Class VII: DES uterus (diethylstilbestrol exposure)

These include hypoplastic uterine cavity, shortened upper uterine segment, T-shaped uterus, hypoplastic cervix, transverse septa, circumferential vaginal or cervical ridges, and cervical hood or collar. Vaginal adenosis, cervical ectropion, and hypoplastic cervix have been noted. In addition, abnormal fallopian tubes with shortening, narrowing and absent fimbria have been described. There is an increased incidence of infertility, spontaneous abortion, ectopic pregnancies, and preterm labor in women with these structural abnormalities. The differential diagnosis of DES exposure by imaging criteria includes infantile uterus and multiple uterine adhesions (Asherman's syndrome).

Complications associated with MDAs.:

Impaired fertility and obstetric complications

Increased incidence of endometriosis

Obstructed uterine drainage in 1. segmental vaginal agenesis or hypoplasia; 2. unicornuate uterus with rudimentary horn which contains endometrium that does not communicate with the main uterine cavity, and 3. uterus didelphys, when one of the duplicated vaginas has an obstructing transverse septum in its upper-third. All types present with hematometra, but only the patients with distal vaginal agenesis or hypoplasia and the obstructed didelphys also have hematocolpos. The hematocolpos associated with uterus didelphys can be differentiated from hematocolpos due to an imperforate hymen, by demonstration of the level of the obstruction. In uterus didelphys the obstruction is in the upper-third of the vagina while the imperforate hymen causes obstruction at the lower-third of the vagina.

Conclusion:

- **Optimal imaging technique is the key to high quality examinations.**
- **Basic knowledge of congenital anomalies and physiological changes is mandatory for good clinical practice.**

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