IMAGING IN UTERINE ANOMALIES

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DEFINITION

- Uterine anomalies –

Type of female genital malformation resulting from an abnormal development of the mullerian duct.

MULLERIAN DUCT ANOMALIES

- The müllerian ducts form most of the female genital tract.
- The caudal fused portion of müllerian ducts form the uterovaginal pri mordium, which gives rise to the uterus and the superior part of the vagina. The vagina, divide into upper and lower portions, arising from the müllerian ducts and urogenital sinus,respectively
- The endometrial stroma and myometrium are derived from splanchnic mesenchyme.
- Uterine development is regulated by the HOXA10 gene.

- MDA Commonly categorized as developmental anomalies (müllerian hypoplasia, unicornuate uterus), fusion anomalies (didelphys and bicornuate uteri), resorption anomalies (septate uterus).

- MDAs result from arrests of development of the uterovaginal primordium during the **eighth week** by:

(1) Incomplete development of a paramesonephric duct

(2) Failure of parts of one or both paramesonephric ducts to develop

(3) Incomplete fusion of the paramesonephric ducts

(4) Incomplete canalization of the vaginal plate to form the vagina.

Hysterosalpingography (HSG), 3D US or MRI remains the standard modality for evaluation of MDA.

CLASSIFICATION

	Main class Uterine anomaly	Main sub-class	Co-existent sub-class Cervical/vaginal anomaly
Class 0	Normal uterus		*Cervix* C0: Normal C1: Septate C2: Double 'normal' C3: Unilateral aplasia/dysplasia C4: Aplasia/dysplasia *Vagina* V0: Normal vagina V1: Longitudinal non-obstructing vaginal septum V2: Longitudinal obstructing vaginal septum V3: Transverse vaginal septum/imperforate hymen V4: Vaginal aplasia
Class I	Dysmorphic uterus	a. T-shaped b. Infantilis	
Class II	Septate uterus	a. Partial b. Complete	
Class III	Dysfused uterus (including dysfused 'septate')	a. Partial b. Complete	
Class IV	Unilaterally formed uterus	a. Rudimentary horn with cavity (communicating or not) b. Rudimentary horn without cavity/aplasia (no horn)	
Class V	Aplastic/dysplastic	a. Rudimentary horn with cavity (bi- or unilateral) b. Rudimentary horn without cavity (bi- or unilateral)/aplasia	
Class VI	Unclassified malformations		

Classification of MDAs according to ASRM Guidelines AND ESHRE/ESG

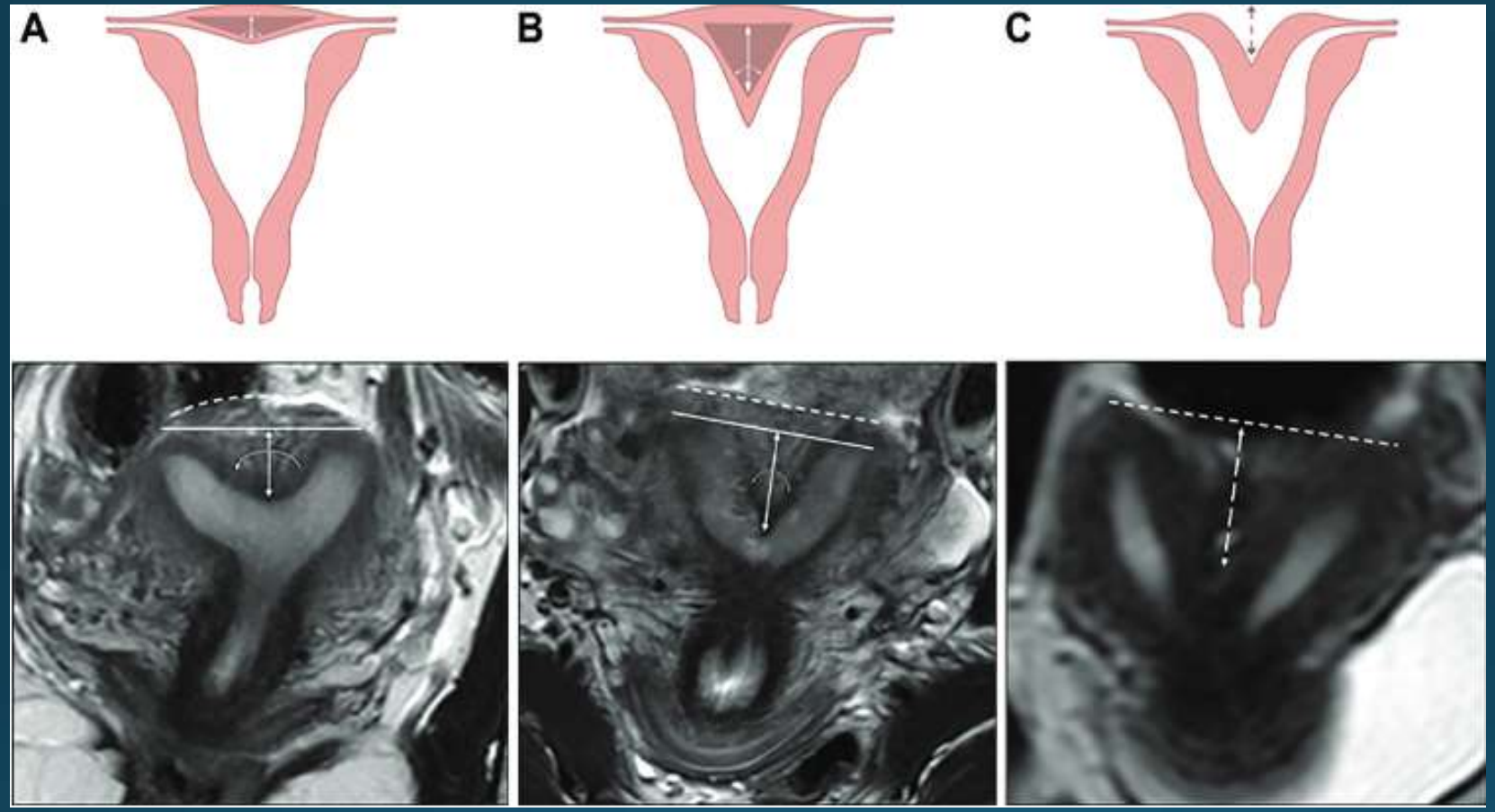
ASRM GUIDELINES		ESHRE/ESG CLASSIFICATION	
Normal or Arcuate Uterus		**Normal Uterus (Class Uo)**	
• Fundus	Convex	• Fundus	Normal outline or external indentation <50% of uterine wall thickness.
• Internal indentation	Indentation depth < 1 cm, angle of divergence more than 90°	• Internal indentation	Straight or curved interostial line with internal indentation <50% of myometrial thickness.
• Cervix	Single	• Cervix	Normal
• Vagina	Single	• Vagina	Normal

ASRM GUIDELINES		ESHRE/ESG CLASSIFICATION	
Resorption anomaly class V: Septate		**Resorption Anomaly: Septate uterus (class U2)**	
• Fundus	Convex or flat; minimal external indentation < 1 cm	• Fundus	Normal outline or external indentation < 50% of uterine wall thickness.
• Internal indentation	Muscular or fibromuscular septum • Complete division to the internal cervical os • Partial division above the internal cervical os	• Internal indentation	More than 50% of uterine wall thickness. • Partial – Septum dividing the cavity above the internal cervical os • Complete – septum extending to the internal cervical os
➢ According to ASRM classification	➢ 1.5 cm, < 90°		
• Cervix	Single, septate or double	• Cervix	Single or septate
• Vagina	Single, longitudinal or transverse septum	• Vagina	Single or septate

ASRM GUIDELINES		ESHRE/ESG CLASSIFICATION	
Developmental anomaly, class I – II 1. **Unicornuate** • Uterine body • Cervix • Vagina • Association	Elongated (banana shaped) and deviated to the right or left pelvis Isolated (35%) or with rudimentary horn Single Single Renal agenesis on the same side as absent or rudimentary horn	**Developmental anomaly** **Dysmorphic uterus (class U I)** T shaped Infantalis **Hemiuterus (Class U4)**	Small uterus with normal outline but abnormal shape of uterine cavity Narrow uterine cavity with thickened lateral wall and normal corpus to cervix ratio Narrow uterine cavity with normal lateral wall and abnormal corpus to cervix ratio Unilateral formed uterus, sub divided by a functional rudimentary cavity

ASRM GUIDELINES		ESHRE/ESG CLASSIFICATION	
2. **Hypoplasia or agenesis** • Uterine body • Cervix • Vagina	MRKHS or vaginal agenesis Unilateral or bilateral rudimentary horns or absent horns. Absent or replaced by triangular soft tissue. Hypoplastic or absent	**Aplastic Uterus (class U5)** **Unclassified (class U6)**	absence of any fully formed uterine cavity Rudimentary horns knee manifest with or without a functional cavity Ectopic mullerian tissue anomalies

ASRM GUIDELINES		ESHRE/ESG CLASSIFICATION	
Fusion anomaly class III-IV:		**Fusion Anomaly:**	External indentation >50% of uterine wall thickness
1. Bicornuate		**Bicorporeal uterus (class U3)**	
• Fundus	External indentation > 1 cm	• Partial Bicorporeal	Indentation dividing the body above the cervix.
• Internal indentation	Muscular or fibromuscular septum- Complete or Partial	• Complete Bicorporeal	Indentation dividing the body to the cervix.
• Cervix	Single (unicollis) , septate (bicollis)	• Bicorporeal septate uterus	Both fusion and resorption defects with midline external indentation more than 150% of uterine wall thickness
• Vagina	Single, longitudinal or transverse septum		
2. Didelphys			
• Uterine body	2 separate uterine cavities with double cervix		
• Cervix	Double	• Cervix	Single, septate, double or unilateral aplasia (eg. OHVIRA)
• Vagina	Longitudinal or oblique septum (eg. OHVIRA)	• Vagina	Single or septate



Arcuate uterus	Septate uterus	Bicornuate uterus
(Fundal contour normal,	(Fundal contour flat,	External uterine fundal indentation > 1cm
< 1 cm, more than 90°)	> 1.5 cm, < 90°)	

ESHRE/ESGE classification system

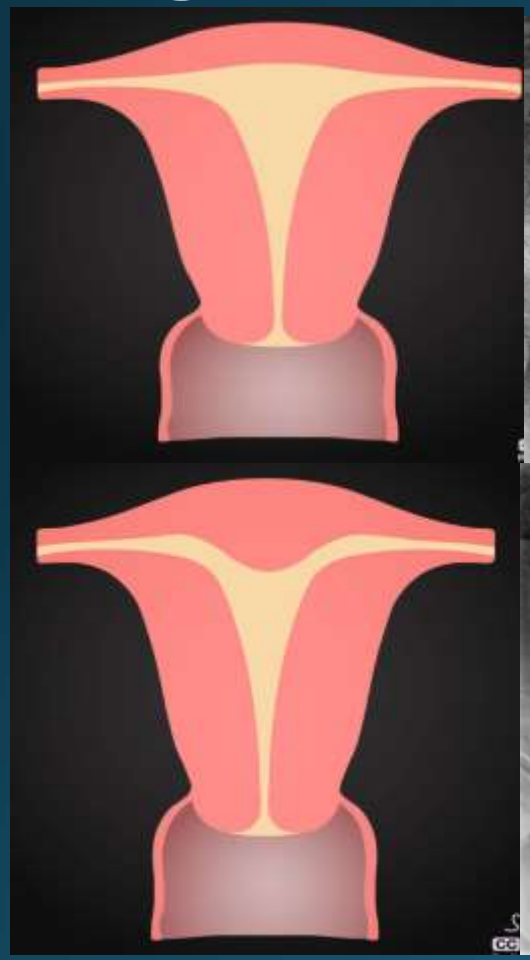
This classification system divides uterine anomalies into six categories on the basis of uterine wall thickness.

- (i) Anatomy is the basis for the systematic categorization of anomalies.
- (ii) Deviations of uterine anatomy deriving from the same embryological origin are the basis for the design of the main classes.
- (iii) Anatomical variations of the main classes expressing different degrees of uterine deformity and being clinically significant are the basis for the design of the main sub-classes.
- (iv)Cervical and vaginal anomalies are classified in independent supplementary sub-classes

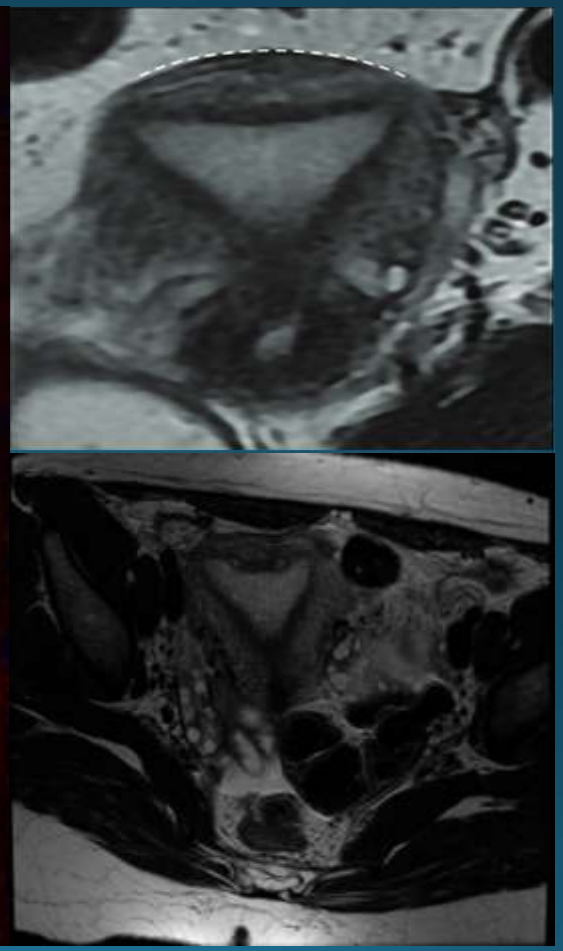
Class Uo

- Incorporates all cases with normal uterus.
- Anormal uterus is any uterus having either straight or curved interostial line but with an internal indentation at the fundal midline not exceeding 50% of the uterine wall thickness.

Normal or Arcuate Uterus – 3D Ultrasound and MRI imaging



HSG: Indentation of uterine endometrial canal 1 cm in its maximum depth



Normal uterine fundal contour with a smooth indentation of fundal endometrial canal.

NORMAL UTERUS

ARCUATE UTERUS

Class U1 or Dysmorphic uterus

- Incorporates all cases with normal uterine outline but with an abnormal shape of the uterine cavity excluding septa.
- Further subdivided into three categories:-

➢ ClassU1a or T-shaped uterus characterized by an arrow uterine cavity due to thickened lateral walls with a correlation 2/3 uterine corpus and 1/3 cervix.

➢ Class U1b or uterus infantilis characterized also by a narrow uterine cavity without lateral wall thickening and an inverse correlation of 1/3 uterine body and 2/3cervix.

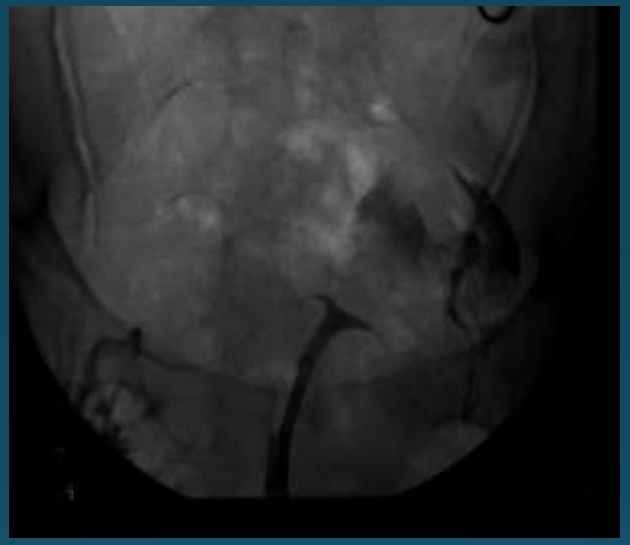
ClassU1c or others

- Include all minor deformities of the uterine cavity including those with an inner indentation at the fundal midline level of 50% of the uterine wall thickness.

T-shaped uterus

- Most commonly associated abnormality from in utero diethylstilbestrol (DES) exposure

Radiographic features



Hysterosalpingogram (HSG)
Typically shows a narrowed irregular endocervical canal. The opacified endometrial cavity appears small, with a shortened upper uterine segment, resulting in the characteristic T-configuration.

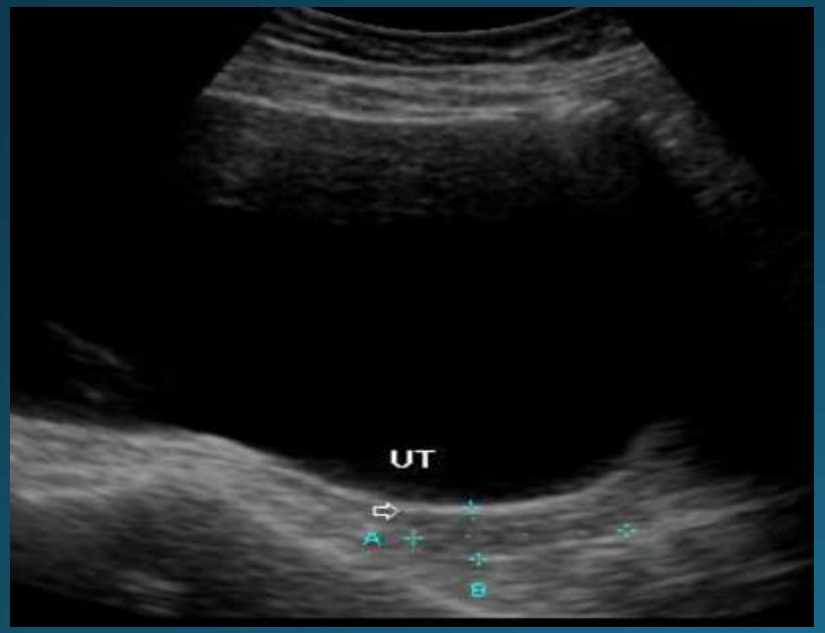
Ultrasound:
A narrow uterine cavity, with thickened lateral walls that resemble a "T" shape

Uterine hypoplasia

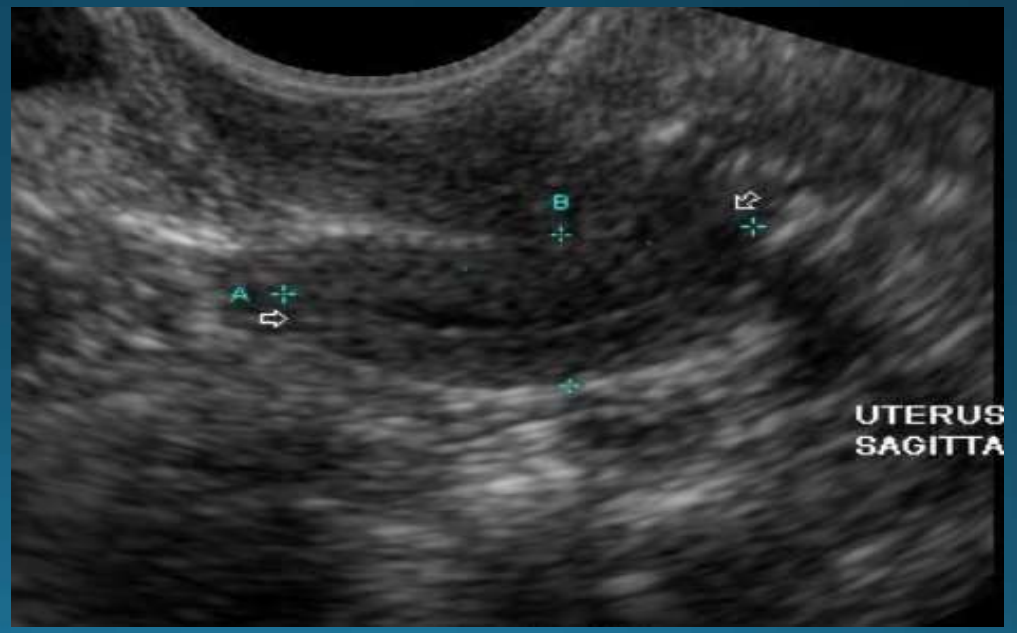
- Congenitally very small uterus
- Types
  - Simple hypoplasia: form of the uterus is normal, but is small in size.
  - Elongated hypoplasia: fundus is normal, but the length is normal or more than normal.
  - Malformative hypoplasia: uterus is arcuate or T- or Y-shaped.

IMAGING

- Sonography and MRI are best imaging tools for the diagnosis of hypoplasia of the uterus.
- In an ultrasound, hypoplasia of the uterus is usually indicated if the distance between the cornu is less than 2 cm or if the distance from the internal os to the fundus is less than 3 to 5 cm.



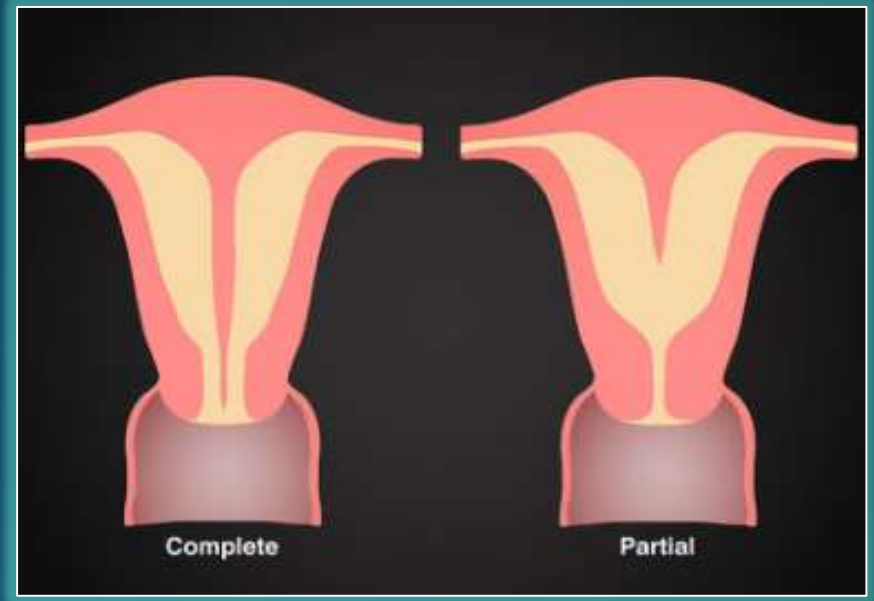
Small uterus



Thin endometrium

Class U2 or septate uterus

- incorporates all cases with normal fusion and abnormal absorption of the midline septum.
- Septate is defined as the uterus with normal outline and an internal indentation at the fundal midline exceeding 50% of the uterine wall thickness.
- This indentation is characterized as septum and it could divide partly or completely the uterine cavity.



ClassU2 a or partial septate uterus

- Septum dividing partly the uterine cavity above the level of the internal cervical os.

ClassU2 b or complete septate uterus

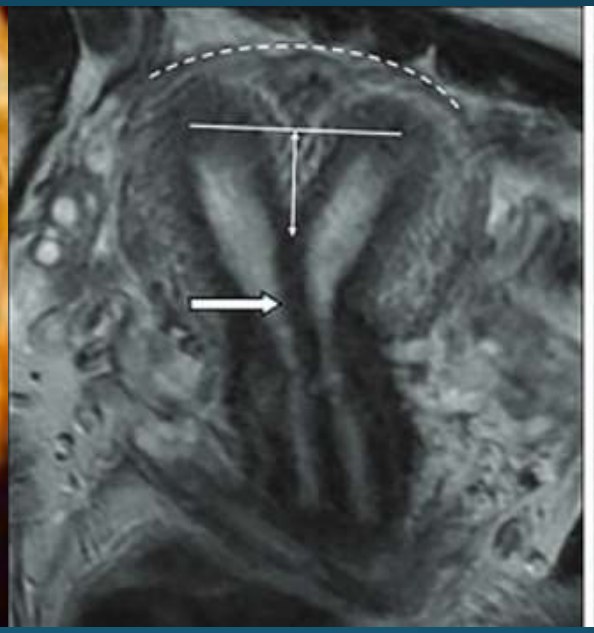
- Septum fully dividing the uterine cavity upto the level of the internal cervical os.
- **Associations**
  - Associated longitudinal vaginal septum may be present in ~25% of cases
  - As with other Mullerian duct anomalies, abnormalities of the renal tract may also be present

Septate Uterus – Imaging

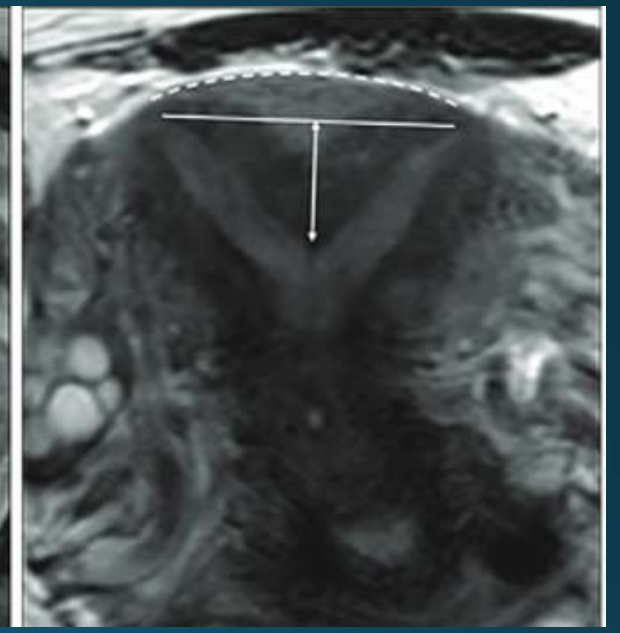
HSG: Indentation of uterine endometrial canal which measures >1.5cm in its maximum depth. (< 90°)

Complete septate

Partial septate



Complete septate uterus



Partial septate uterus

Fundal contour convex, > 1.5 cm, < 90°)

- Ultrasound:
  - Fundal myometrial depth measures ~ 1.2 cm which is more than that of arcuate uterus and less than that of septate uterus suggestive of partial septate/subseptate uterus.

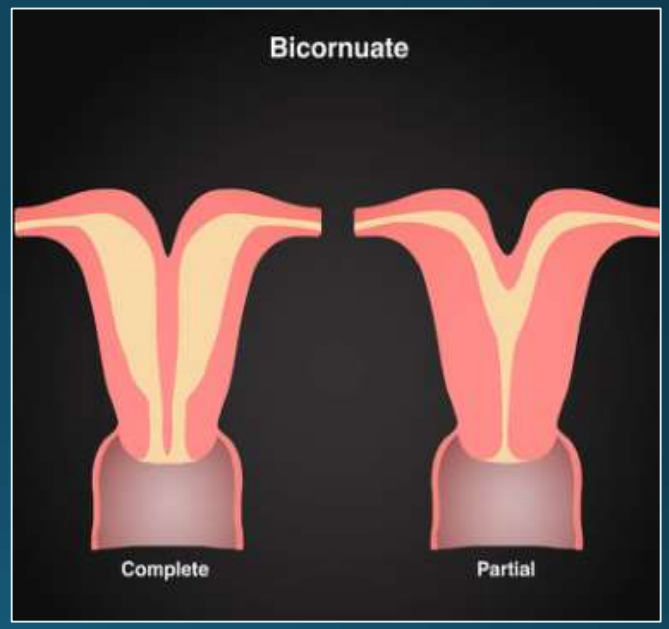
ClassU3 or bicorporeal uterus

- Incorporates all cases of fusion defects.

defined the uterus with an abnormal fundal outline

Characterized by the presence of an external indentation at the fundal midline exceeding 50% of the uterine wall thickness.

This indentation could divide partly or completely the uterine corpus also associated with an inner indentation at the midline level that divides the cavity as in septate uterus.



- **ClassU3a or partial bicorporeal uterus**

An external fundal indentation partly dividing the uterine corpus above the level of the cervix.

- **Class U3b or complete bicorporeal uterus**

An external fundal indentation completely dividing the uterine corpus upto the level of the cervix.

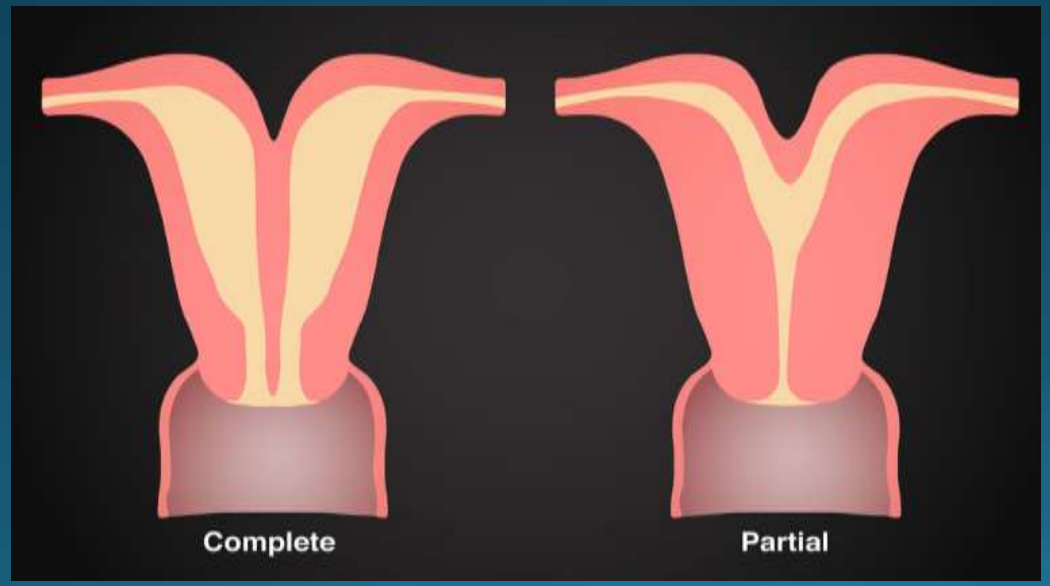
- **ClassU3c or bicorporeal septate uterus**

Presence of an absorption defect in addition to the main fusion defect. The midline fundal indentation exceeds by 150% the uterine wall thickness

- **Associations**
  - Associated longitudinal vaginal septum may be present in ~25% of cases
  - As with other Mullerian duct anomalies, abnormalities of the renal tract may also be present

Subtypes

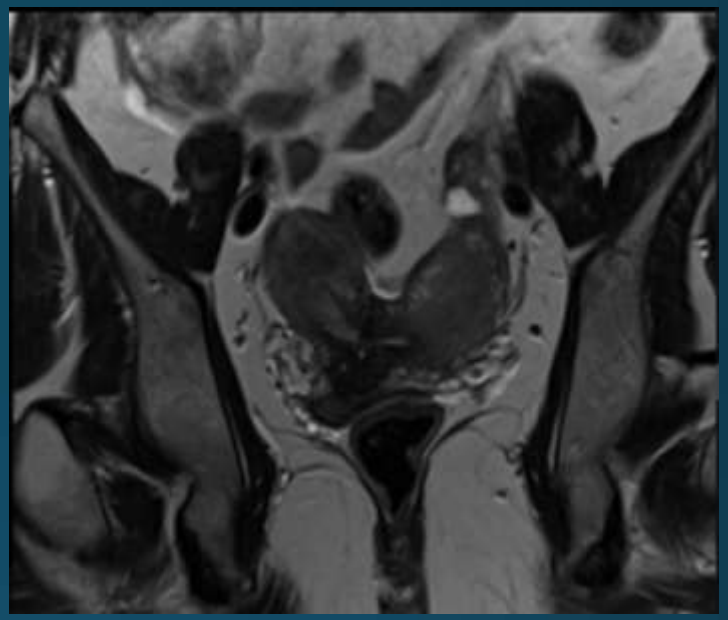
- A bicornuate uterus is divided according to the involvement of the cervical canal:
  - Bicornuate bicollis: two cervical canals; central myometrium extends to external cervical os
  - Bicornuate unicollis: one cervical canal; central myometrium extends to internal cervical os



Bicornuate Uterus – Imaging

HSG:
Two diverging endometrial cavities are seen with increased intercornual distance.
However single cervical canal is noted.

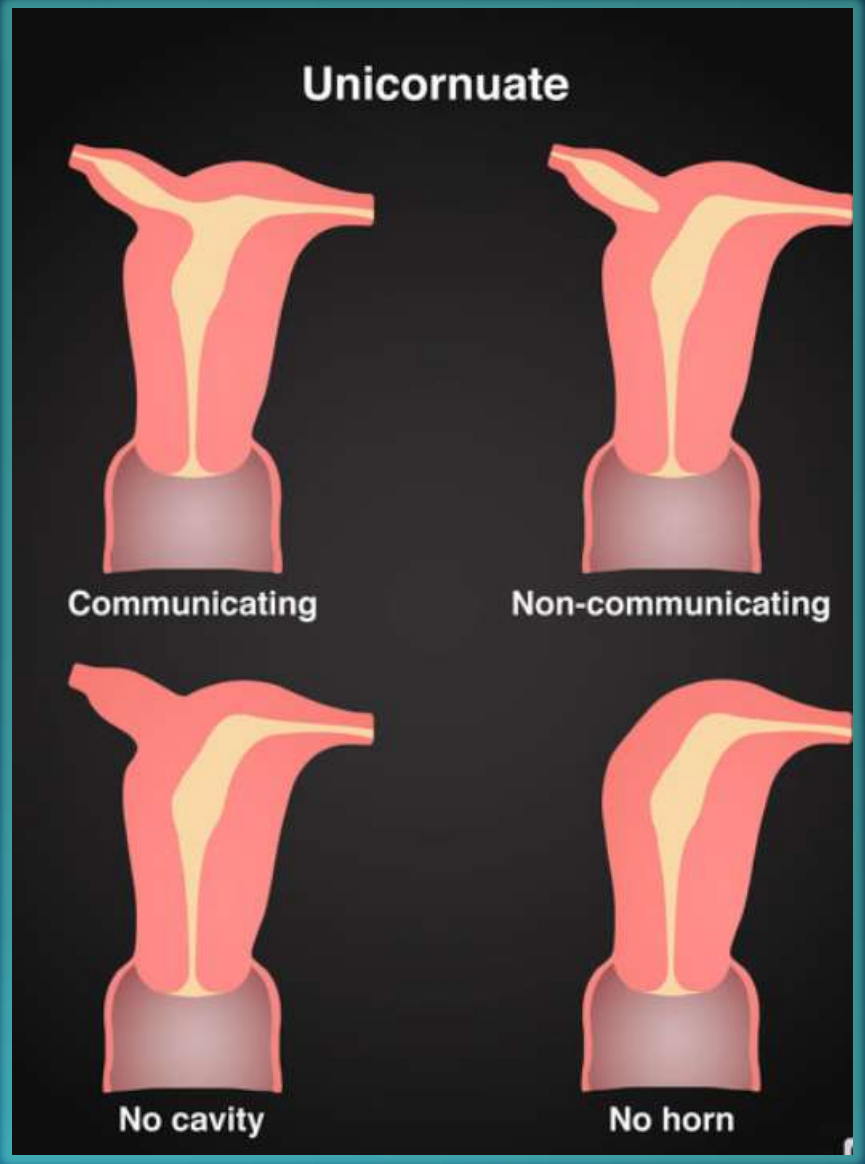
Ultrasonography:
External indentation > 1 cm
Muscular or fibromuscular septum causing inner indentation at the midline level that divides the uterine cavity



Coronal T2WI:
External indentation > 1 cm
Muscular or fibromuscular septum dividing the body above the lumen of cervix

ClassU4 or hemi-uterus

- Incorporates all cases of unilateral formed uterus.
- Defined as the unilateral uterine development; the contralateral part could be either incompletely formed or absent.



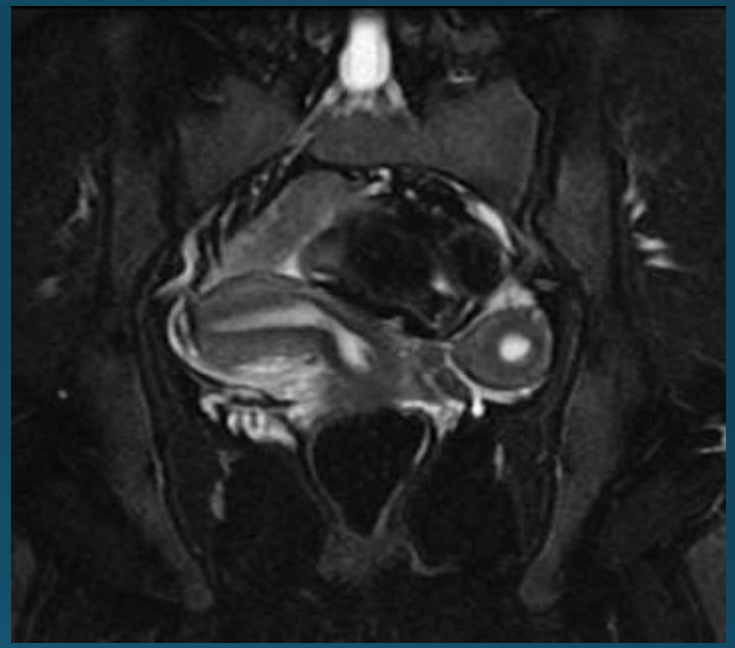
- Class U4a or hemi-uterus with a rudimentary (functional) cavity characterized by the presence of a communicating or non communicating functional contralateral horn.

- ClassU4b or hemi-uterus without rudimentary (functional) cavity characterized either by the presence of non-functional contralateral uterine horn or by aplasia of the contralateral part.

Hemi Uterus – Imaging

HSG : The endometrial cavity assumes a fusiform (banana type) shape, tapering at the apex and draining into a single Fallopian tube. The uterus is generally shifted off the midline.

3D ultrasound :
Endometrial lining deciated to right side.



MRI
Curved and elongated uterus: banana-shaped external uterine contour
Reduced uterine volume
Normal myometrial zonal anatomy
Rudimentary horn seen on left side

ClassU5 or aplastic uterus

Formation defect characterized by the absence of any fully or unilaterally developed uterine cavity.

- ClassU5a or aplastic uterus with rudimentary (functional) cavity characterized by the presence of bi-or unilateral functional horn
- ClassU5b or aplastic uterus without rudimentary(functional)cavity characterized either by the presence of uterine remnants or by full uterine aplasia.

Mayer-Rokitansky-Kuster-Hauser (MRKH)

- Complete absence of the Mullerian ducts is termed Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome
- Includes the absence of the upper 2/3 of the vagina as well.
- Arise during embryogenesis, with arrested development of the paramesonephric ducts at ~7 weeks after fertilisation.
- Characterized by normal external genitalia and absence or reduced development of the uterus and upper two-thirds of the vagina.
- The development of kidneys, ureter, and bladder occurs concomitantly at around the 6^{th}-12^{th} weeks of gestation.

- Two different forms are described:
  - The **typical form** (**type A**) :
    - characterized by the congenital absence of the uterus and upper 2/3 vagina with normal ovaries and fallopian tubes
  - The **atypical form** (**type B**) :
    - includes associated abnormalities of the ovaries and fallopian tubes and renal anomalies
- Associations:
  - Renal tract anomalies including renal agenesis, ectopic kidney, fused kidney, renal hypoplasia, and horseshoe kidney.
  - Skeletal: vertebral anomalies.

- MRI needed to fully define the anatomy as small uterine remnants can be present, difficult to identify sonographically.
- It is the imaging modality of choice after an initial assessment with ultrasound, allowing the characterization of the uterine buds and presence of functioning endometrium within them.

Sagittal/Axial T2: Absence of uterus and vagina

ClassU6 - Unclassified

- Infrequent anomalies, subtle changes or combined pathologies could not be allocated correctly to one of the six groups.
- Furthermore, the system is designed to include, all cases resulting from formation, fusion or absorption defects of normal embryological development.

Uterus didelphys (double uterus)

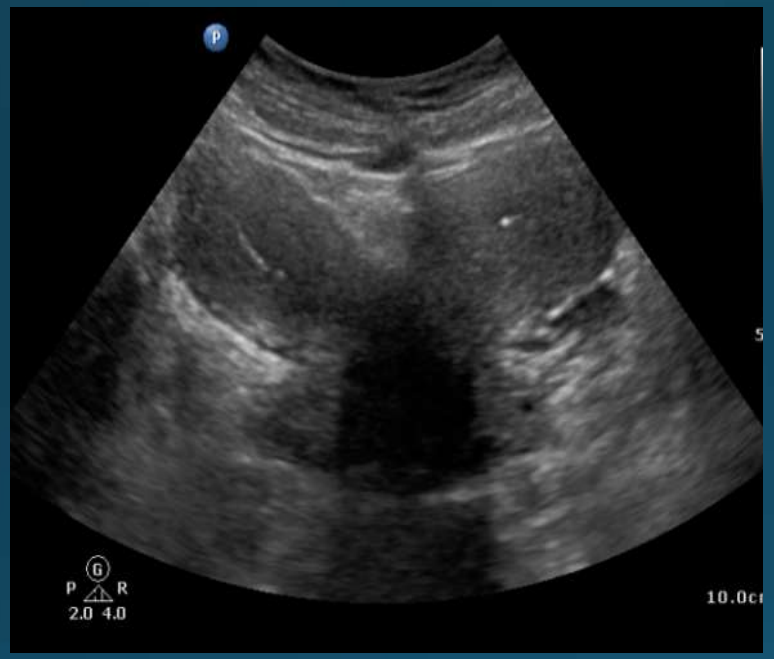
- Results from failure of fusion of the inferior parts of the paramesonephric ducts.

Associated with

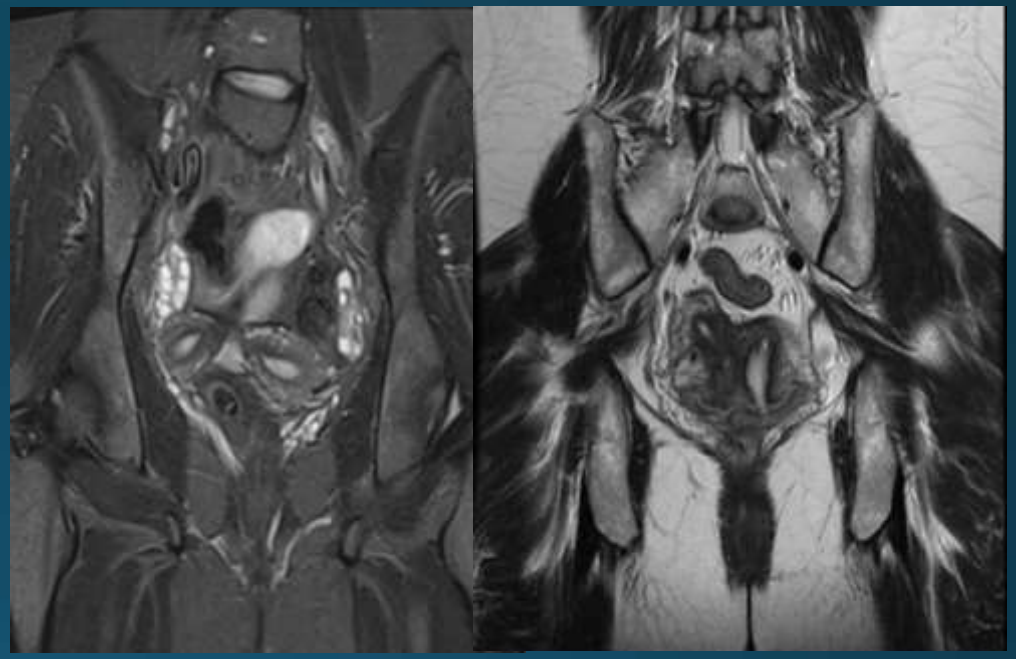
- Renal agenesis
- Vaginal septum which can include a transverse vaginal septum
  - There is a vaginal septum in 75% of cases, and obstruction to one horn is possible from occasional transverse septae

Uterus didelphys – Imaging

HSG:
Two separate endocervical canals that open into separate fusiform endometrial cavities, with no communication between the two horns. Each endometrial cavity ends in a solitary fallopian tube.



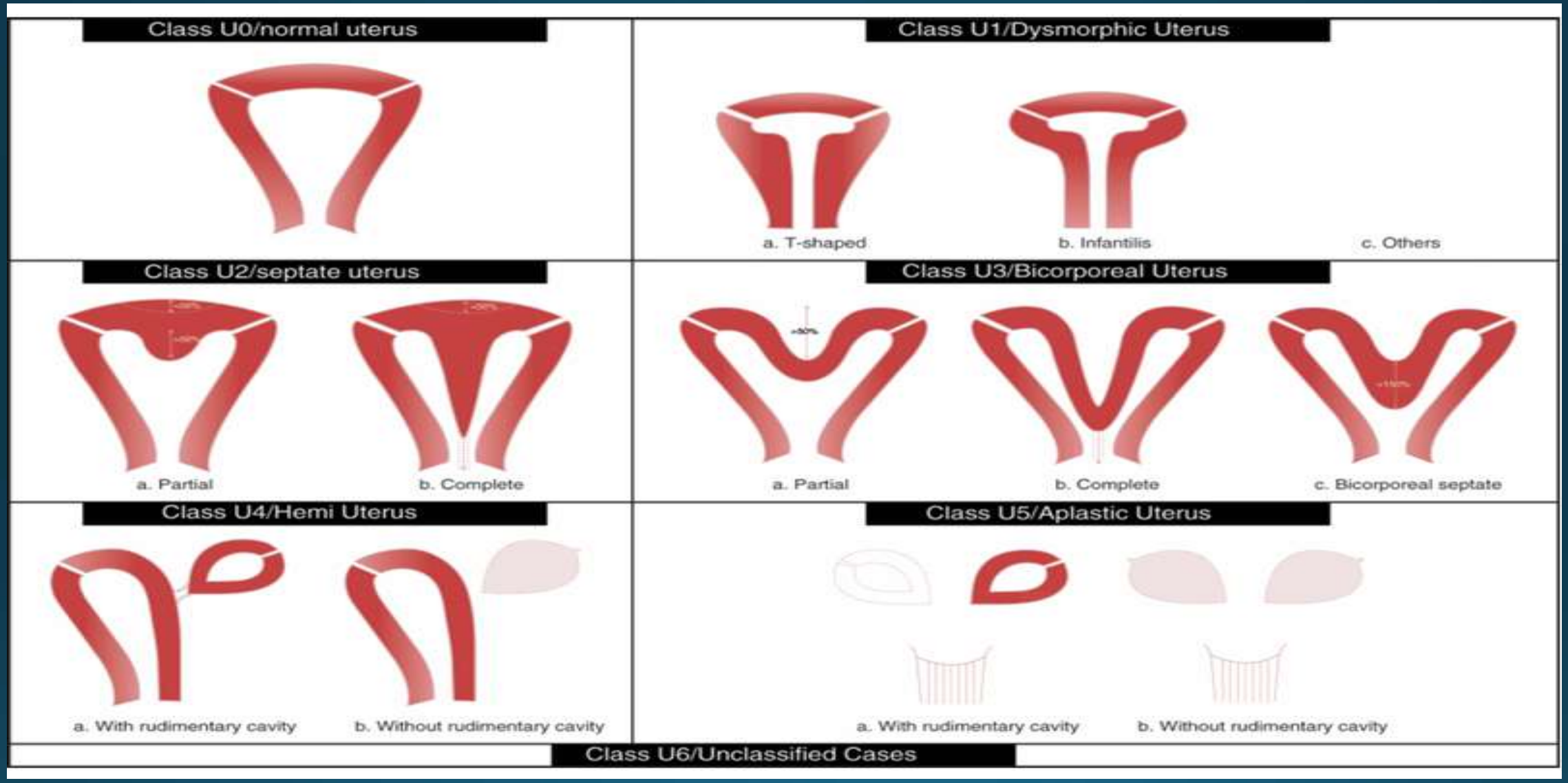
Ultrasound:
Separate divergent uterine horns with a large fundal cleft.
No evidence of communication between endometrial cavities.



Coronal T2:
Separate uterine horns

Differential diagnosis

Feature	Septate Uterus	Bicornuate Uterus	Uterine Didelphys
Embryological Origin	failure of resorption of longitudinal septum that partially divides the uterine cavity	Complete or partial failure of Müllerian duct fusion	Complete failure of Müllerian duct fusion
External Uterine Contour	Normal convex fundus	Deep indentation at the fundus (>1cm)	Two separate uterine bodies with distinct fundus
Fundal Indentation Depth	<1cm	>1cm	Complete separation
Intercornual Angle	<75°	<105°	Each uterine horn is separate
Cavitary Anatomy	Single endometrial cavity	Two separate endometrial cavities	Two separate endometrial cavities
MRI Characteristics	Hypointense (T2 WI) fibrous septum extending into the cervix	Deep fundal cleft with two horns	Complete separation into two distinct uterine bodies



ESHRE/ESGE classification of uterine anomalies : schematic representation

ClassU2: internal indentation > 50% of the uterine wall thickness and external contour straight or with indentation < 50%
ClassU3: external indentation > 50% of the uterine wall thickness, ClassU3b: width of the fundal indentation at the midline > 150% of the uterine wall thickness)

ASRM MÜLLERIAN ANOMALIES CLASSIFICATION 2021

Scan QR code to view the ASRM MAC 2021 tool (page 1 of 2)

MÜLLERIAN AGENESIS

MÜLLERIAN AGENESIS

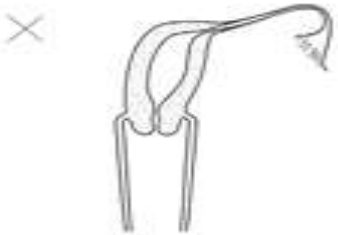
MÜLLERIAN AGENESIS WITH R/L ATROPHIC UTERINE REMNANT WITH FUNCTIONAL ENDOMETRIUM

CERVICAL AGENESIS

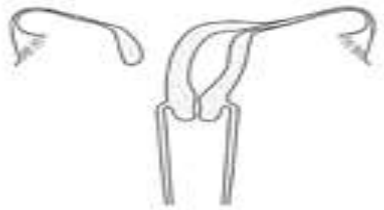
CERVICAL AGENESIS

DISTAL CERVICAL AGENESIS

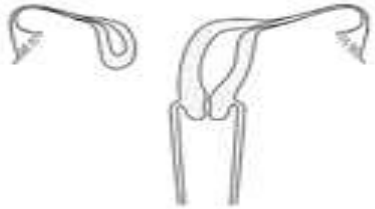
UNICORNUATE UTERUS



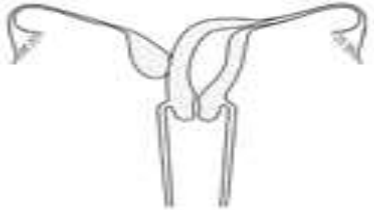
R/L UNICORNUATE UTERUS



R/L UNICORNUATE WITH R/L DISTAL ATROPHIC UTERINE REMNANT



R/L UNICORNUATE WITH R/L DISTAL UTERINE REMNANT WITH FUNCTIONAL ENDOMETRIUM



R/L UNICORNUATE WITH R/L ASSOCIATED ATROPHIC UTERINE REMNANT

R/L UNICORNUATE WITH R/L UTERINE HORN COMMUNICATING AT LEVEL OF CERVIX

UTERUS DIDELPHYS

UTERUS DIDEPHYS AND LONGITUDINAL SEPTUM

UTERUS DIDELPHYS AND +/- LONGITUDINAL VAGINAL SEPTUM OF VARIABLE LENGTH

UTERUS DIDELPHYS AND OBSTRUCTED R/L HEMIVAGINA

BICORNUATE UTERUS

BICORNUATE UTERUS

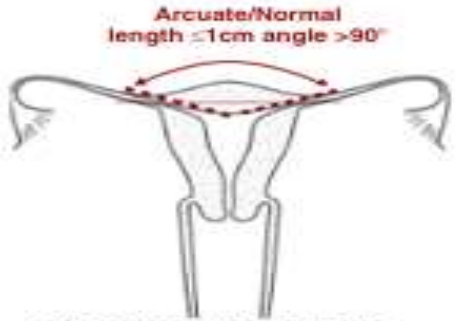
BICORNUATE UTERUS WITH R/L COMMUNICATING TRACT

UTERUS BICORNUATE BICOLLIS

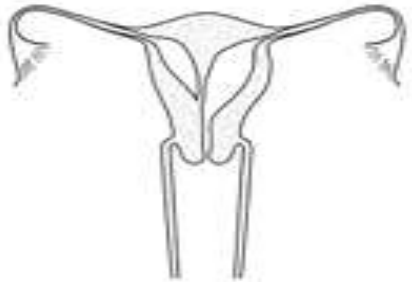
COMBINED BICORNUATE SEPTATE UTERUS

SEPTATE UTERUS

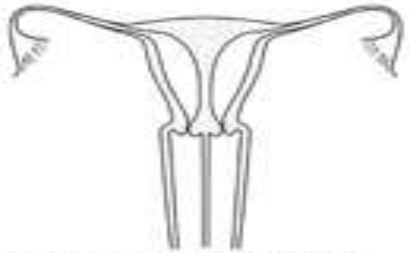
PARTIAL SEPTATE UTERUS



NORMAL/ARCUATE UTERUS



ROBERT'S UTERUS



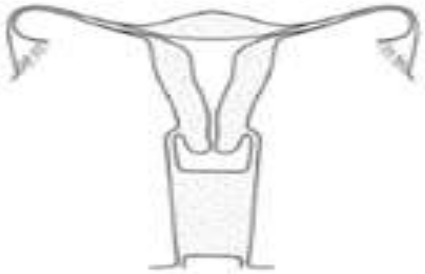
COMPLETE SEPTATE UTERUS WITH DUPLICATED CERVICES AND LONGITUDINAL VAGINAL SEPTUM

COMPLETE SEPTATE UTERUS WITH SEPTATE CERVIX AND LONGITUDINAL VAGINAL SEPTUM

COMPLETE SEPTATE UTERUS, DUPLICATED CERVICES, AND OBSTRUCTED R/L HEMIVAGINA

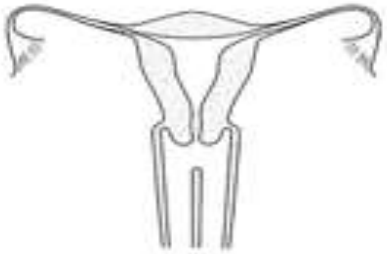
TRANSVERSE VAGINAL SEPTUM

MID VAGINAL SEPTUM



DISTAL VAGINAL AGENESIS

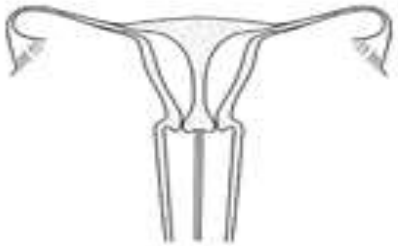
LONGITUDINAL VAGINAL SEPTUM



LONGITUDINAL VAGINAL SEPTUM OF VARIABLE LENGTH

LONGITUDINAL VAGINAL SEPTUM OF VARIABLE LENGTH AND UTERUS DIDELPHYS

OBSTRUCTED R/L HEMIVAGINA AND UTERUS DIDELPHYS



LONGITUDINAL VAGINAL SEPTUM OF VARIABLE LENGTH AND COMPLETE SEPTATE UTERUS WITH DUPLICATED CERVIX

OBSTRUCTED R/L HEMIVAGINA AND COMPLETE SEPTATE UTERUS WITH DUPLICATED CERVICES

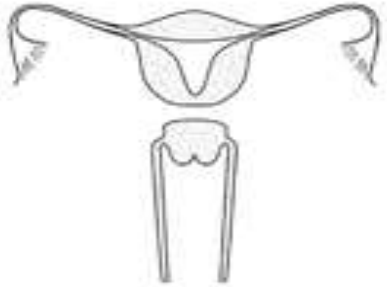
COMPLEX ANOMALIES

BICORNUATE UTERUS WITH BILATERAL OBSTRUCTED ENDOMETRIAL CAVITIES

UTERUS DIDELPHYS WITH COMMUNICATING HEMIUTERI AND UNILATERAL R/L CERVICO-VAGINAL ATRESIA

OBSTRUCTED R/L HEMIVAGINA, HEMIUTERUS AND SINGLE CERVIX WITH SEPARATE CONTRALATERAL R/L PATENT HEMIUTERUS, CERVIX AND VAGINA

BICORNUATE UTERUS WITH R/L COMMUNICATING TRACT AND TRANSVERSE VAGINAL SEPTUM



UTERUS ISTHMUS AGENESIS

THANK YOU