

**LATEST 2024 MARROW
NEET-SS NOTES**



**UPDATED
OBSTYNE RESIDENCY
NOTES**

GYNECOLOGY

CONGENITAL MALFORMATIONS OF THE UTERUS

Relevant anatomy

00:00:43

- In females : genital tract is formed by the paramesonephric ducts (mullerian ducts).
- In males : From Wolffian duct (mesonephric duct).
- mullerian ducts (MD) are an invagination of coelomic epithelium (at 6 weeks) and grow downwards alongside mesonephric ducts enclosed in peritoneal folds that later give rise to broad ligament of the uterus.

In early intrauterine life, both mullerian duct and Wolffian duct are present in both the sexes, enclosed in broad ligament.

Remnants of the mesonephric duct (Wolffian Duct) :

- Epoophoron
 - Para oophoron
 - Gartner's duct
- } contents of broad ligament

In females : mullerian duct grows d/t lack of Anti-mullerian hormone (AMH) in intrauterine life.

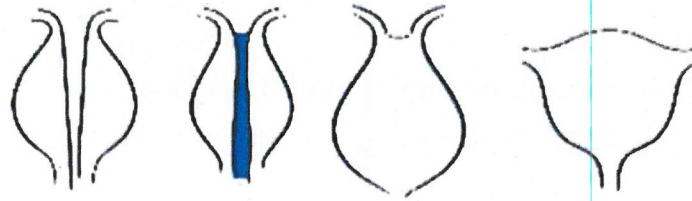
In males, AMH is formed by the sertoli cells of the testis.

During Paramesonephric duct elongation, certain Homebox genes (HOX genes) in group 9-13 play an important role in development.

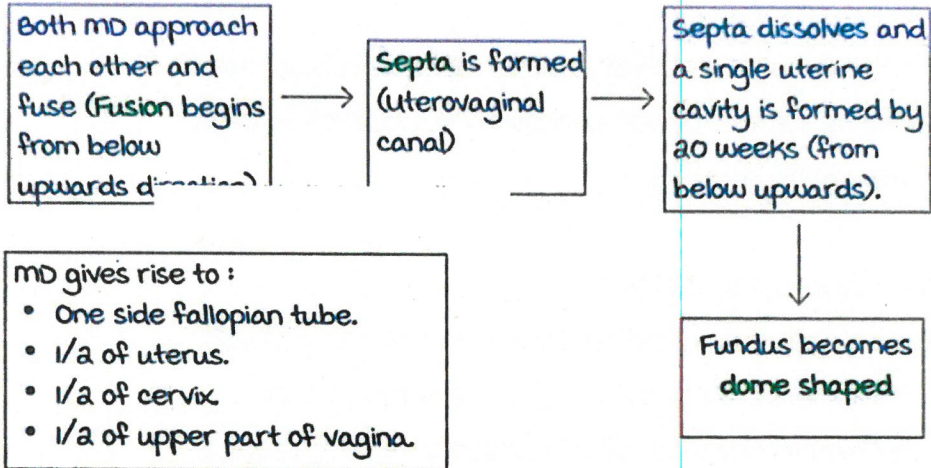
- HOX 9 : Fallopian tube.
- HOX 10, 11 : uterus.

At 10 weeks : The two distal parts of MD approach in the midline and fuse to form uterovaginal canal/septa.

At 12 weeks : mesonephric duct regresses.



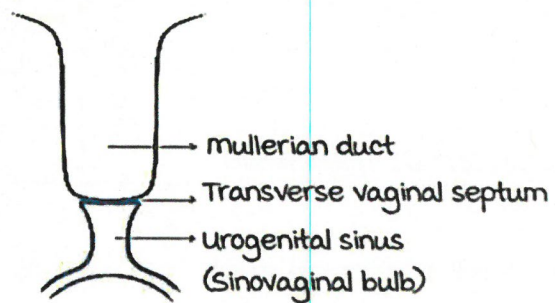
Stages of development



vaginal development :

- upper part (2/3rd or 1/3rd) : mullerian duct.
- Lower part (1/3rd or 2/3rd) : Sinovaginal bulb part of urogenital sinus.

Transverse vaginal septa is formed by fusion which dissolves by 20 weeks forming a single vaginal canal.



Complications in a female with mullerian malformations :

- Obstetric complications :**
1. Recurrent pregnancy loss (RPL).
 2. Abortion.
 3. Preterm labor.
 4. malpresentations.
 5. Ectopic pregnancy ; unicornuate pregnancy.

- Gynaecologic complications :**
1. Infertility.
 2. Endometriosis.
 3. Dysmenorrhea : Generalised (w/L dysmenorrhea : Unicornuate uterus)
 4. Outflow tract obstruction : Hematometra.

Active space

NOTE : In young pubertal females with C/O endometriosis :
Always rule out mullerian malformations.

uterus

m/c complaints in female with mullerian malformations :
Obstetrics complications (RPL) > infertility.

1st Investigations : Incidental finding on USG (RPL) / HSG
(infertility).

HSG is not IOC : It cannot differentiate between bicornuate
& septate uterus as the outer contour & fundus of uterus
cannot be visualised on HSG.

Hysterosalpingography (HSG) :

- A water soluble iodinated radioopaque dye is passed inside the uterus with the help of Leech-Wilkinson cannula (funnel shaped with transverse serrations).
- Followed up with serial X-rays.

IOC of mullerian malformations : 3D USG.

Gold standard : MRI.

Last resort : Laparoscopy + Hysteroscopy.

MC indication for doing surgery in mullerian malformations :
RPL.

Class I : Mullerian Aggenesis

00:16:57

Description : Both mullerian ducts are absent

- No Fallopian tube (distal part present).
- No upper vagina (Generally complete vaginal aggenesis)
- No uterus.
- Ovaries are normal (develop from genital ridge)
- No cervix

Associated problems :

- **Renal anomalies (30-50%)** : Renal aggenesis, Horse-shaped kidney.
- **Skeletal anomalies (10-15%)**.

NOTE : Hence in all cases of MRKH syndrome, intravenous pyelography (IVP) and skeletal X-rays must be done.

MRKH syndrome D/D : Androgen Insensitivity syndrome (AIS)

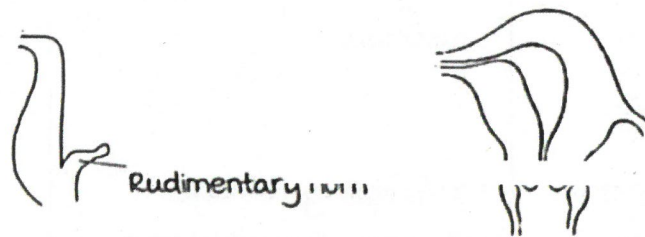
- Obstetrical complications :
- Primary amenorrhea
 - Infertility

MRKH : Mayer-Rokitansky Kuster Hauser syndrome.

MURCS : Mullerian agenesis, Renal anomalies, Cervical somites.

Class II : Unicornuate Uterus

00:20:52



Description : Only one side Mullerian duct develops to form

- Fallopian tube : 1
- uterus, cervix, upper vagina : 1/2

Other side either **complete agenesis** or **rudimentary horn** (communicating or non-communicating).

Associated problems :

Overall increased chances of :

- Endometriosis.
- Infertility.
- **Ipsilateral renal anomalies** (2nd m/c Mullerian malformation associated with renal anomalies).

Non-communicating horn with active endometrium present :

- Cyclical u/L dysmenorrhea.
- u/L hematometra.

Obstetric complications :

- Increased spontaneous abortion.
- Increased preterm deliveries.
- If pregnancy occurs in rudimentary horn : uterine rupture (prior to 20 weeks).
- Ectopic pregnancy in the rudimentary horn.

NOTE : Mullerian malformation associated with increased risk of ectopic pregnancy : Unicornuate uterus.



Normal uterus



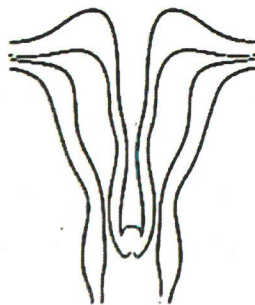
Unicornuate uterus

HSG in normal uterus	HSG in unicornuate uterus
<ul style="list-style-type: none"> • 2 FT (thin and tortuous) • Single uterus. • Single cervix • Single vagina. • Bilateral spillage of the dye (can be used to check patency of tubes). 	<ul style="list-style-type: none"> • Single FT (unicornuate uterus). • U/L spillage of dye. • Banana shaped uterus.

Communicating rudimentary horn should be obliterated to prevent ectopic pregnancy and uterine rupture in the horn.

Class III : Uterine Diadelphys

00:27:02

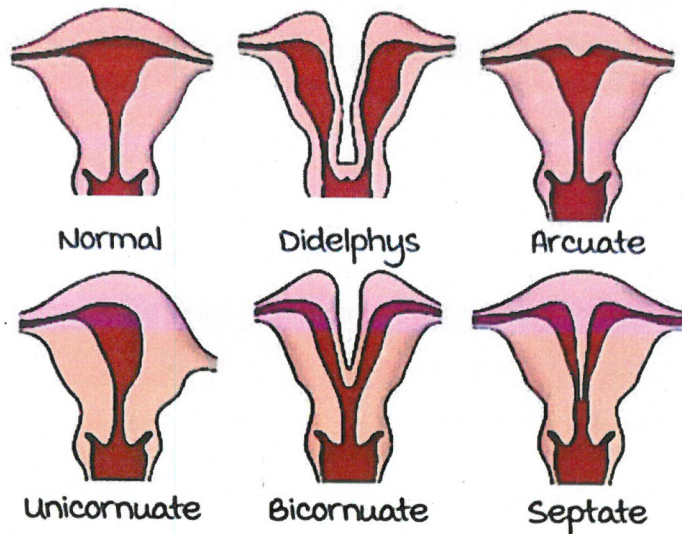


Description : Failed fusion of the paired mullerian ducts.

- Fallopian tubes, uterus, cervix, upper part of vagina : 2 in number.
- a Leech wilkinson canula needed to visualise the uterus.
- Good reproductive outcome when compared to other malformations.

Obstetric complications : RPL, fetal growth restriction.

Active space



Class IV : Uterus Bicornuate

00:28:58

Description : Incomplete fusion of mullerian duct.

- Fallopian tube, uterus : 2
- Cervix : 1 or 2
- Always single vagina (because fusion occurs from below upwards.)

Bicornis unicollis : 2 uterus 1 cervix

Bicornis Bicollis : 2 uterus 2 cervix

Distinguishing feature :

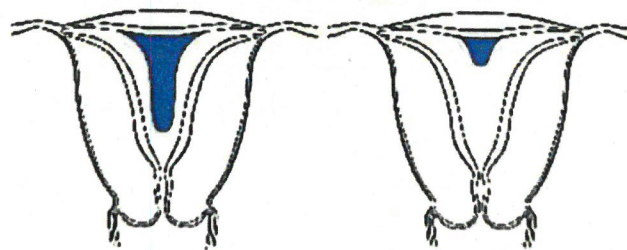
Normal fundus : Septate uterus

Depression on the fundus (divided) : Bicornuate uterus.

The two cannot be distinguished on HSG (Hence not IOC).

Class V : Septate Uterus

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Complete septa

Partial septa

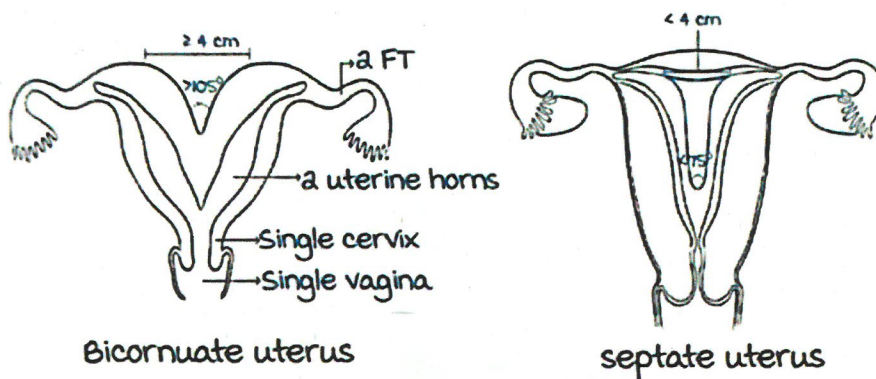
- Septate uterus results when both MD fuse, septa is formed but septa fails to resolve partially or completely.
- Outwardly, the uterus appears normal, but a septa is present inside the uterus

Bicornuate uterus vs septate uterus :

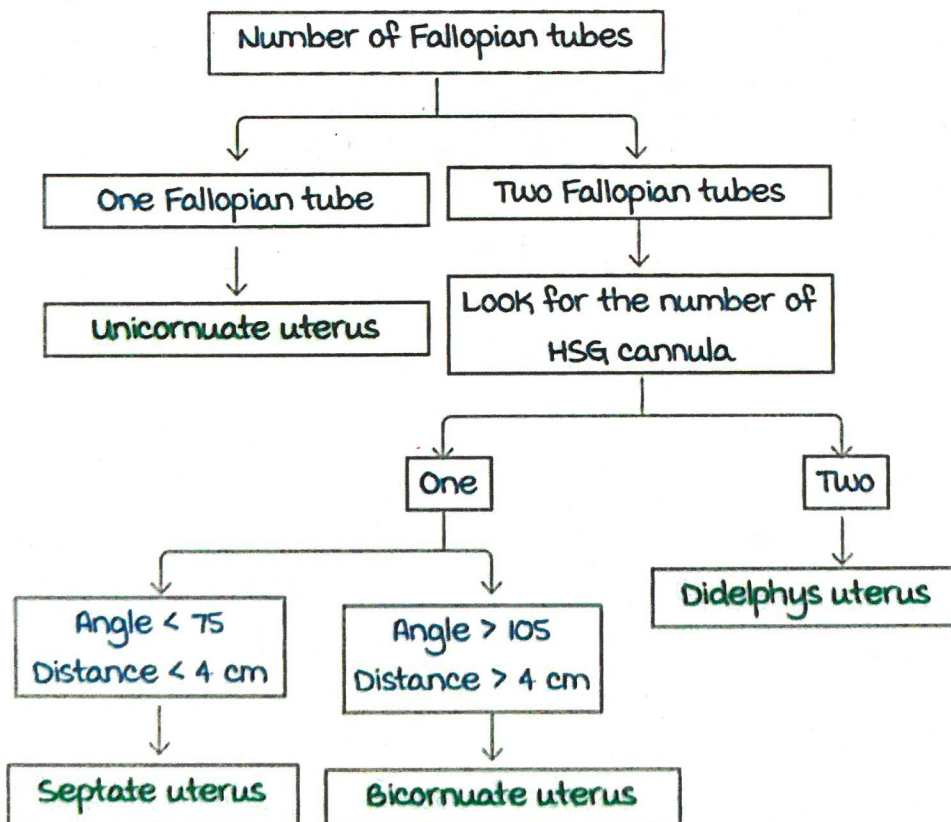
Should be distinguished by looking at fundus of the uterus (on USG).

On HSG :

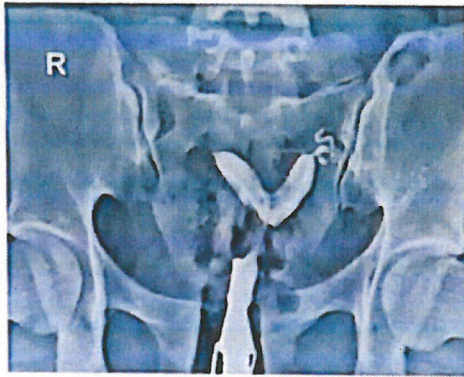
	Bicornuate uterus	Septate uterus
External contour	Divided	Normal
Intercornual angle	$> 105^\circ$	$< 75^\circ$
Distance between 2 horns	≥ 4 cm	< 4 cm



Algorithm to diagnose mullerian malformations :



Active space



Bicornuate uterus



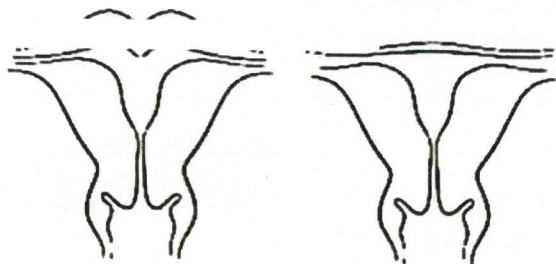
Septate uterus

	Bicornuate uterus	Septate uterus
Obstetric complications	<ul style="list-style-type: none"> • RPL (repeated pregnancy loss) • PTL (preterm labor) 	m/c uterine anomaly leading to: <ul style="list-style-type: none"> • 1st trimester abortion • Infertility • possibly congenital malformations.
Surgery (in case of RPL)	Straussman metroplasty. Pregnancy planned after repair; deliver by C-section.	Hysteroscopic resection of the septum.

Class VI : Arcuate Uterus

00:42:06

Associated problem : Entire uterus is formed except there is slight indentation of fundus/flat topped uterus



Indentation of fundus

Flat topped fundus

Overall best reproductive outcome in all mullerian malformations.

Class VII - Diethylstilbesterol (DES) induced reproductive tract abnormalities

00:42:45

DES is a synthetic non-steroidal estrogen which was prescribed to millions of pregnant females earlier.

Led to abnormal development of the reproductive tracts of daughters of the pregnant females :

- T-shaped uterus.
- Clear cell adenocarcinoma of the vagina and cervix (due to suppression of WNT-4 gene; HOX gene).
- Vaginal adenosis.
- Cervical collar.
- Genitourinary malformations.
- Fallopian tube abnormalities like absent fimbriae.

These females in adulthood showed :

- Earlier menopause.
- Increased risk of Breast cancer.

males exposed to DES :

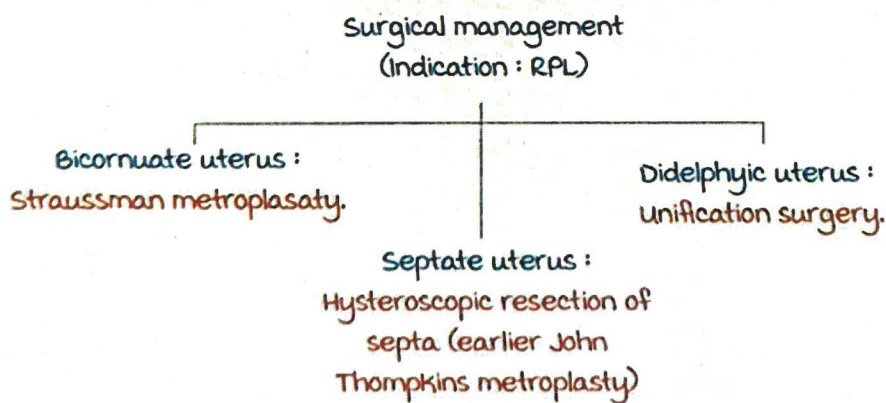
- Increased incidence of cryptorchidism.
- Testicular hypoplasia.
- Hypospadias.
- Microphallus.
- Renal anomalies.

Note :

DES exposure does not lead to renal abnormalities in females.

Mx of Mullerian Anomalies

00:45:30



Active space