



NEET SS



GYNECOLOGY



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CONGENITAL MALFORMATIONS OF THE UTERUS

Relevant anatomy

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- 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.

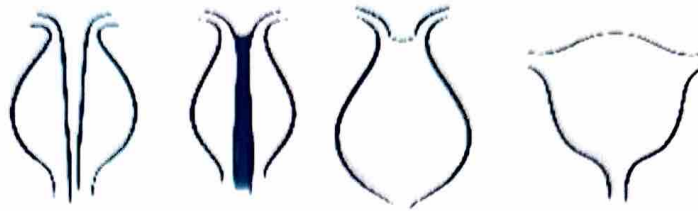
In males, AMH is formed by the sertoli cells of the testis.

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

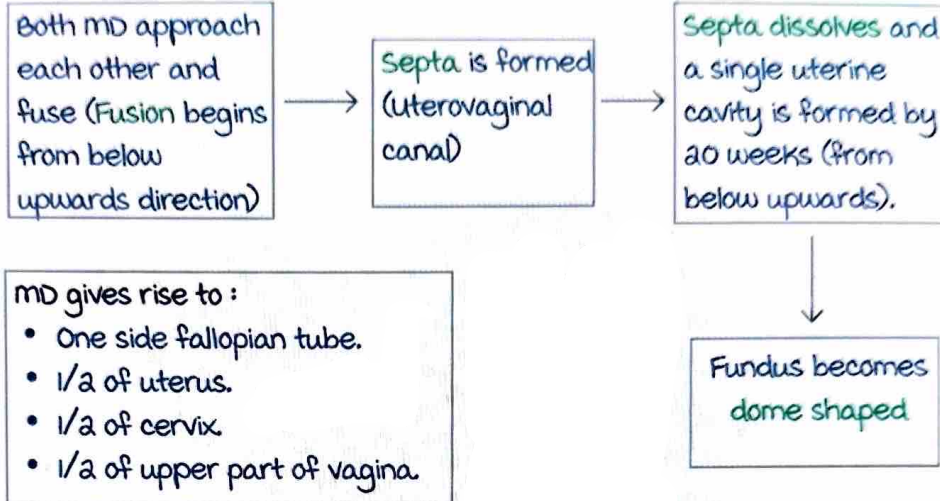
- HOX 9 : Fallopian tube.
- HOX 10, 11 : uterus.

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.



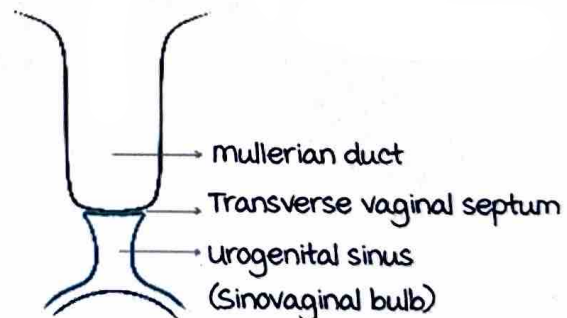
Stages of development



Vaginal development :

- upper part ($2/3^{\text{rd}}$ or $1/3^{\text{rd}}$) : mullerian duct.
- Lower part ($1/3^{\text{rd}}$ or $2/3^{\text{rd}}$) : 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.

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

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 tranverse serrations).
- Followed up with serial X-rays.

IOC of mullerian malformations : 3D USG.

Gold standard : MRI.

Last resort : Laproscopy + Hysteroscopy.

MC indication for doing surgery in mullerian malformations :
RPL.

Class I : Mullerian Agenesis

00:16:57

Description : Both mullerian ducts are absent

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

Associated problems :

- **Renal anomalies** (30-50%) : Renal agenesis, 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 :

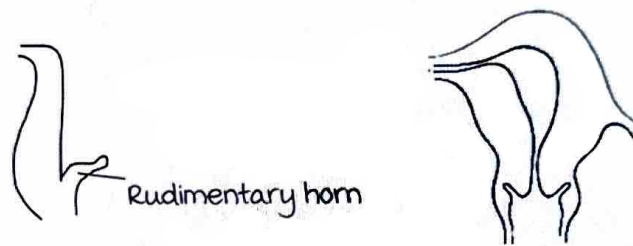
- Primary amenorrhea.
- Infertility

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



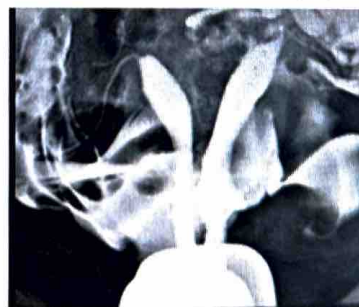
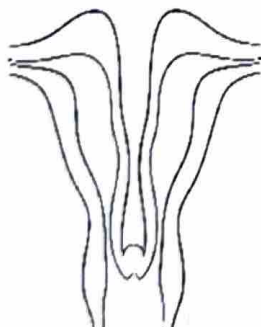
Unicornuate uterus

HSG in normal uterus	HSG in unicornuate uterus
• 2 FT (thin and tortuous) • Single uterus. • Single cervix. • Single vagina. • Bilateral spillage of the dye (can be used to check patency of tubes).	• 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

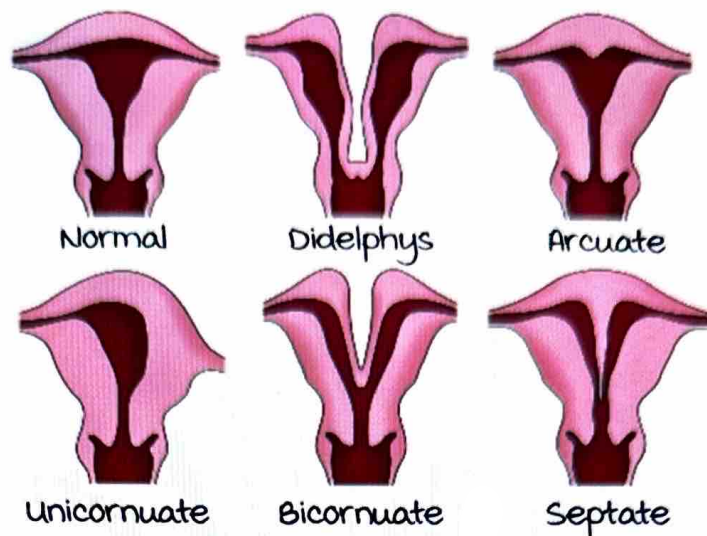
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Description : Failed fusion of the paired mullerian ducts.

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

Obstetric complications : RPL, fetal growth restriction.



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 :

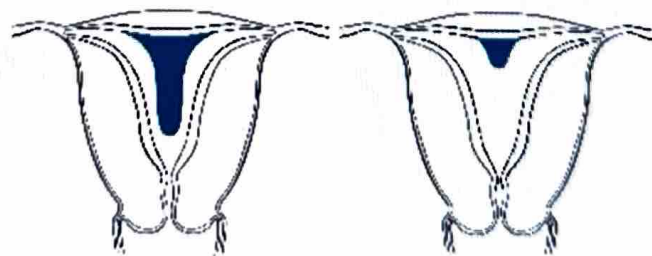
Normal fundus : Septate uterus

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

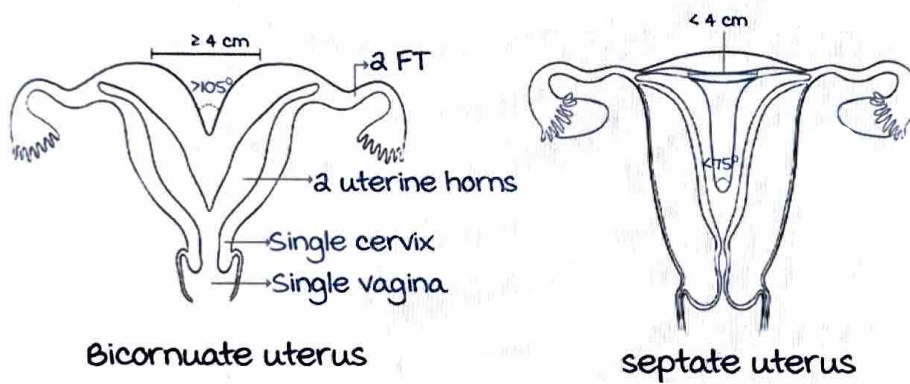
- Septate uterus results when both MD fuse, septa is formed but septa fails to resolve partially or completely.
- Outwardly, the uterus appears normal, but a septa is present inside the uterus

Bicornuate uterus vs septate uterus :

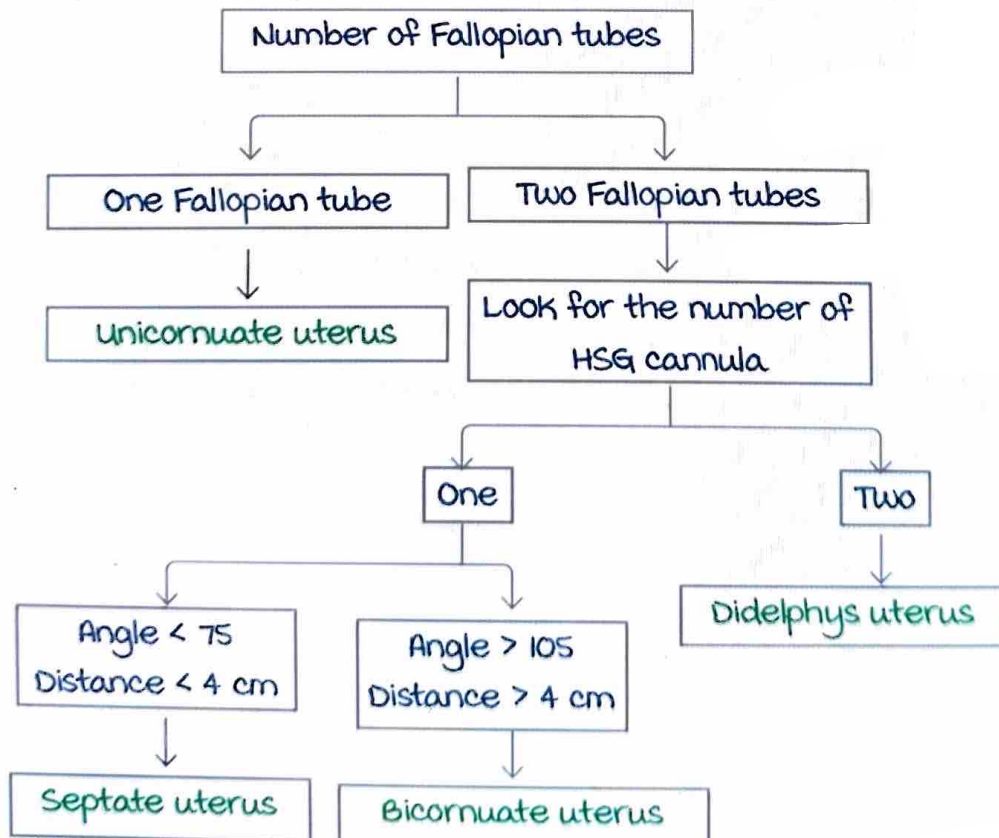
Should be distinguished by looking at fundus of the uterus (on USG).

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 :





Bicornuate uterus



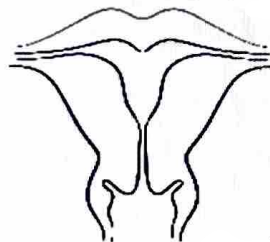
Septate uterus

	Bicornuate uterus	Septate uterus
Obstetric complications	• RPL (repeated pregnancy loss) • PTL (preterm labor)	m/c uterine anomaly leading to: • 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.

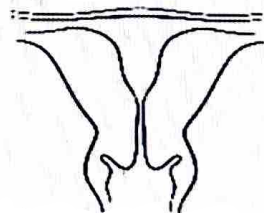
Class VI : Arcuate Uterus

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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 :

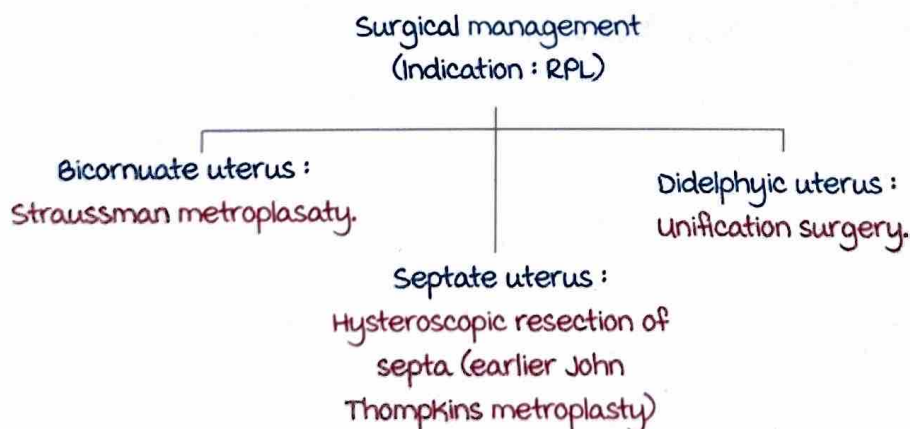
- Increased incidence of cryptorchidism.
- Testicular hypoplasia.
- Hypospadias.
- micropallus.
- Renal anomalies.

Note :

DES exposure does not lead to renal abnormalities in females.

Mx of Mullerian Anomalies

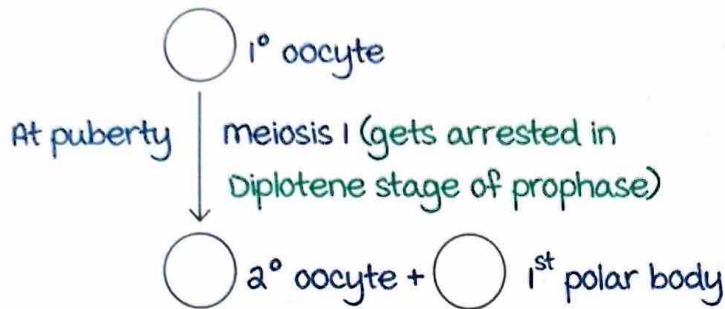
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MENSTRUAL CYCLE

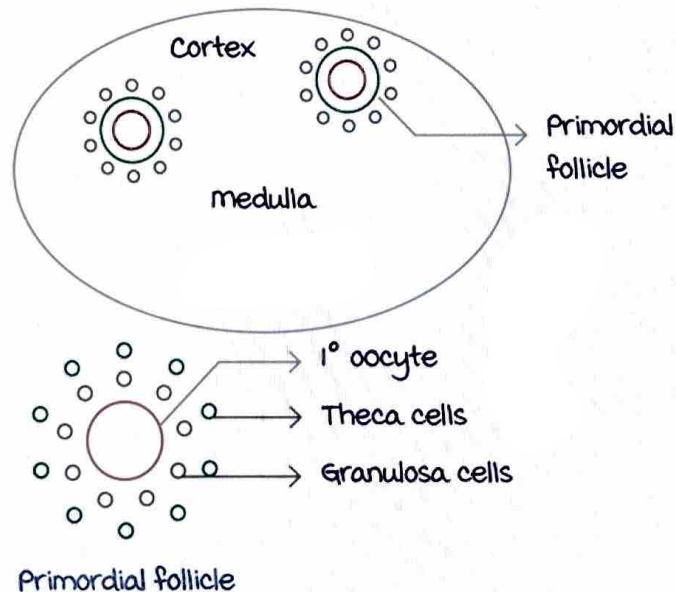
Oogenesis

00:01:24



Structure of Ovary :

Before puberty :



Number of follicles :

- maximum : 5th month of intrauterine life (6-7 million).
- At birth : 1-2 million follicles.
- At puberty : 4-5 lakh follicles.

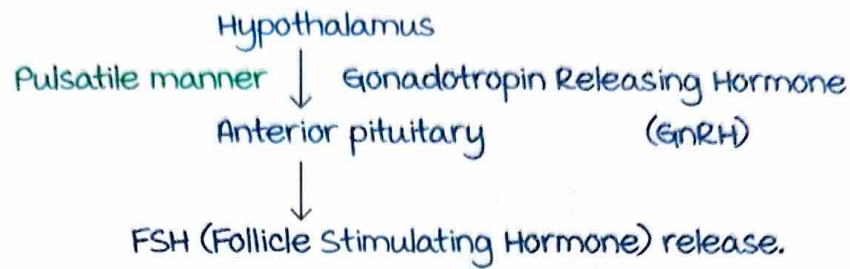
Note :

Initial recruitment of follicles is hormone independent.

Role of FSH

00:08:55

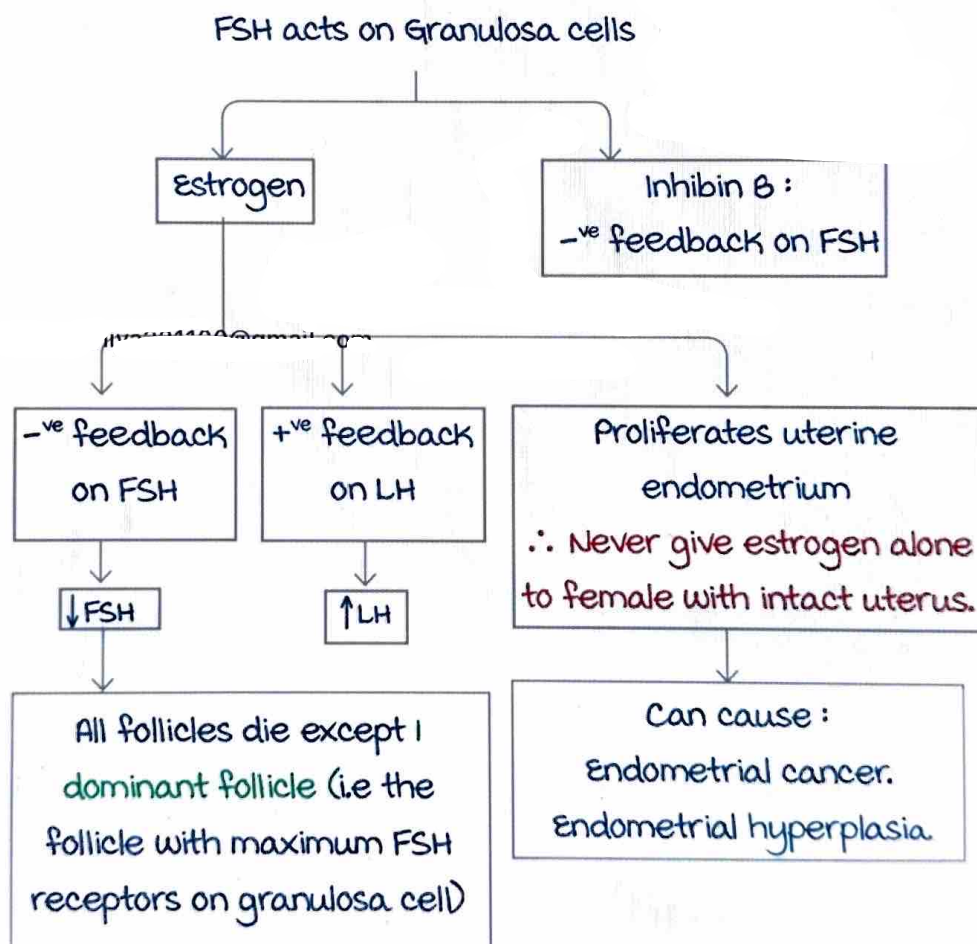
At puberty, Hypothalamic Pituitary Ovarian Axis (HPOA) becomes functional.



Functions of FSH :

- Prevents the follicles from undergoing atresia.
- Stimulates follicles.

Role of FSH :

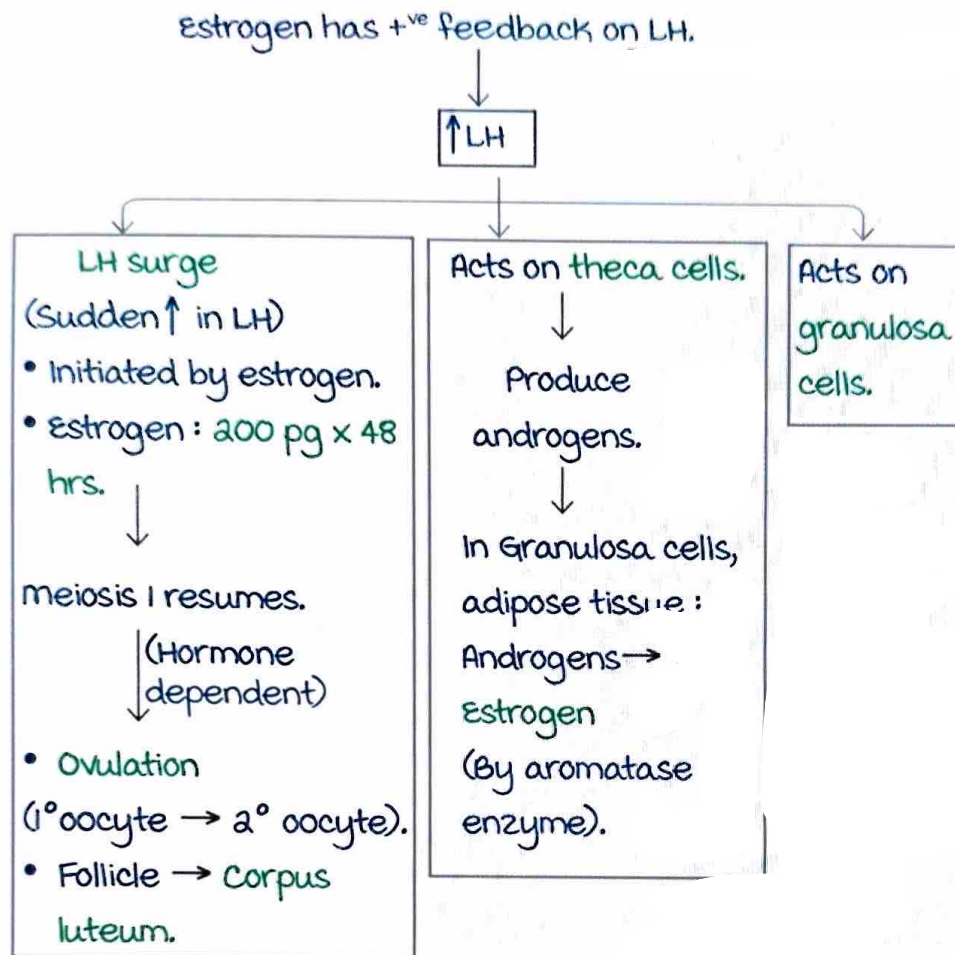


Note :

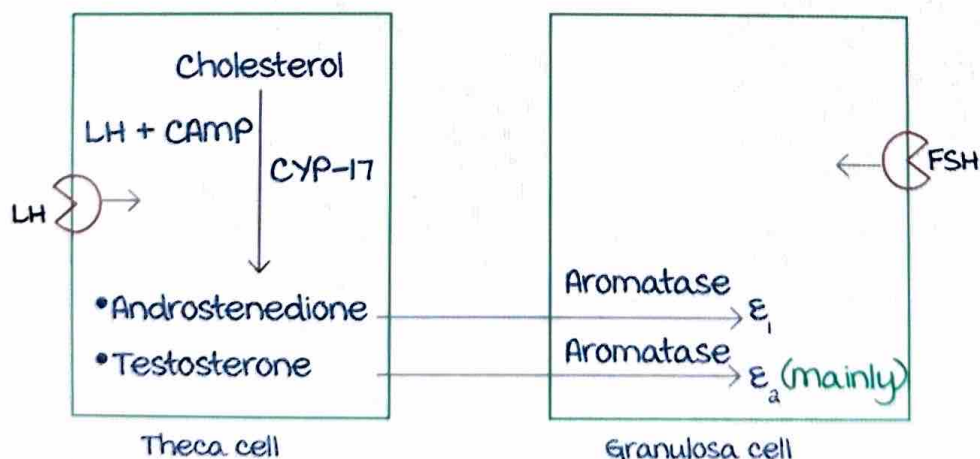
FSH receptors are present on granulosa cells.

Granulosa cell tumour of ovary : Feminizing tumor.

Tumor marker for granulosa cell tumor of ovary : Inhibin B.



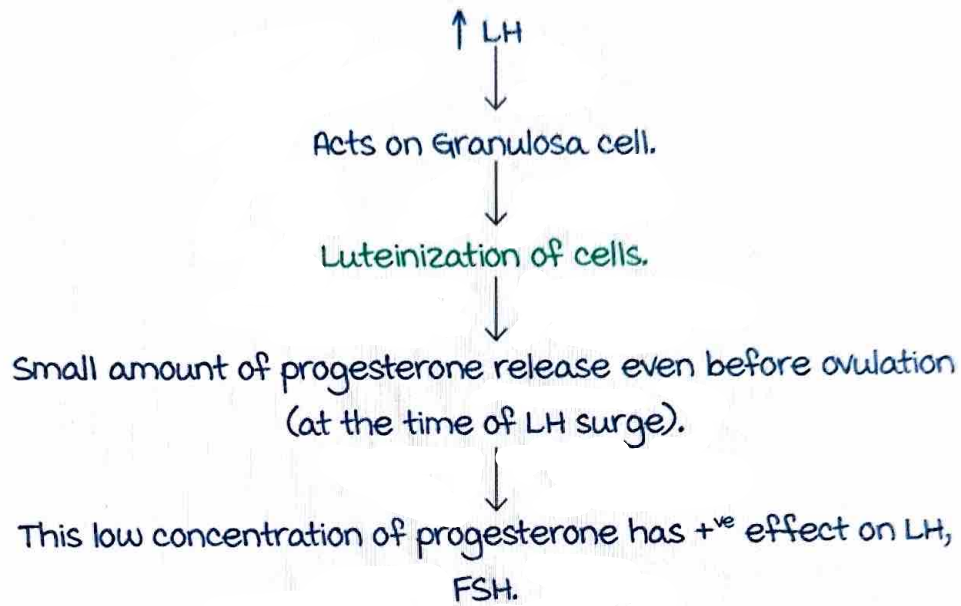
Two cell two gonadotropin theory



Aromatase enzyme absent.
CYP 17 present.

Aromatase present.
CYP 17 absent.

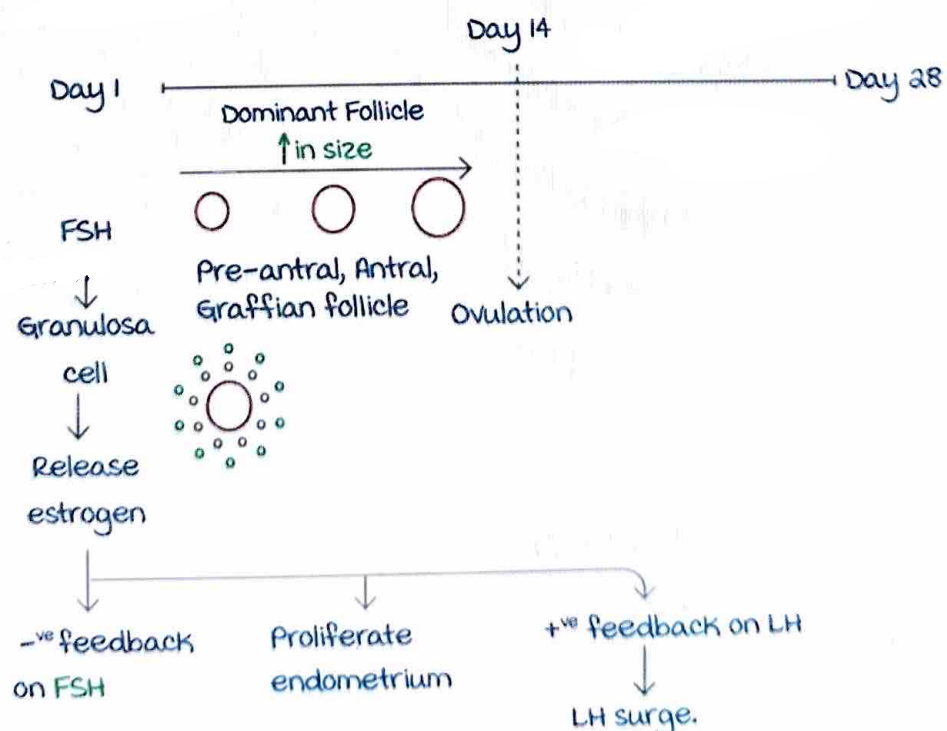
Adipose tissue :
Androstenedione — aromatase → E_i.



- In females, LH receptors are present on theca cell, granulosa cell.
- Progesterone appears earliest in menstrual cycle at LH surge (32 to 36 hrs before ovulation).
- There is LH and FSH surge before ovulation.

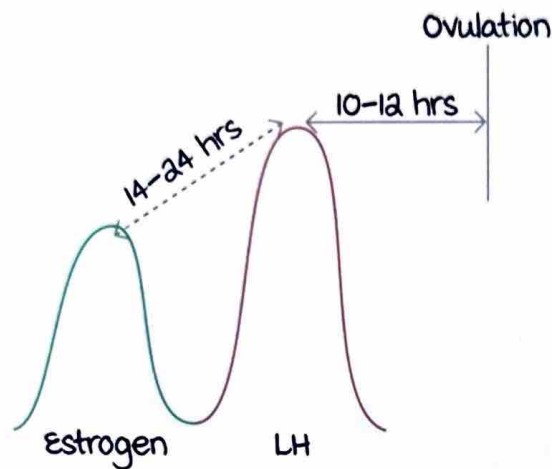
Follicular/Proliferative phase of menstrual cycle

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- Ovarian cycle is initiated by FSH.
- Size of follicle, just before ovulation : 18 to 20 mm.
- For LH surge to occur, Estrogen levels : 200 pg x 48 hours.

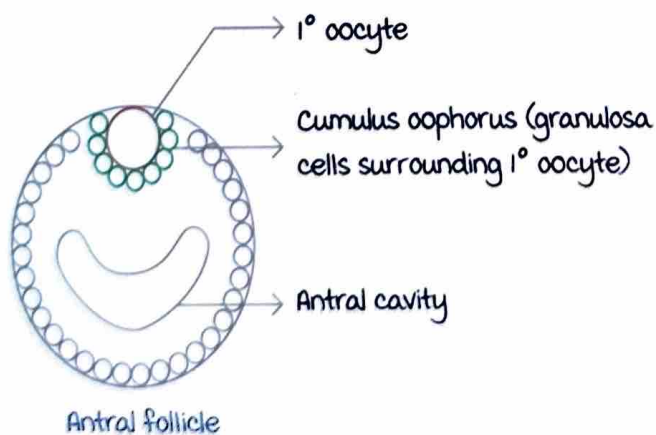
- LH surge $\xrightarrow[24-36 \text{ hours}]{32-36 \text{ hours/}}$ Ovulation.
- LH peak $\xrightarrow{10-12 \text{ hours}}$ Ovulation.



- Time interval b/w estrogen peak to LH peak : 14-24 hours
- Time interval b/w estrogen peak to ovulation : 24-36 hours.
- LH surge :
 - Initiated by estrogen.
 - maintained by both estrogen & progesterone.
- Ovulation is due to LH surge only.
- meiosis I is resumed due to LH surge (32-36 hours before ovulation).

Ovulation

00:48:42



Normally :

- Antral cavity fluid : Estrogen + growth factor + LH.
- LH appears in the antral cavity fluid only towards mid cycle.

Anovulation :

If LH appears in antral cavity fluid early in the cycle :

- Leads to atresia of follicle.
- Decreases the mitotic activity of granulosa cells which leads to Anovulation.

Secretory phase of menstrual cycle

00:53:02

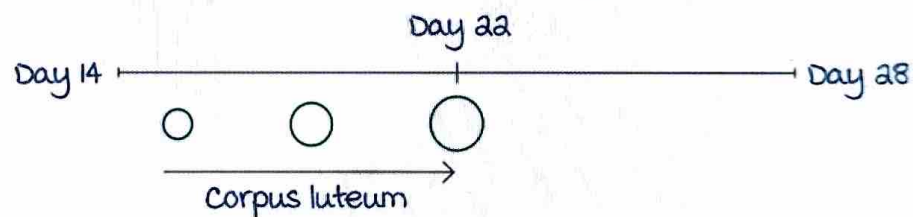
1° oocyte → 2° oocyte.

Follicle → Corpus luteum.

LH : maintains corpus luteum in a non pregnant female.

Corpus luteum :

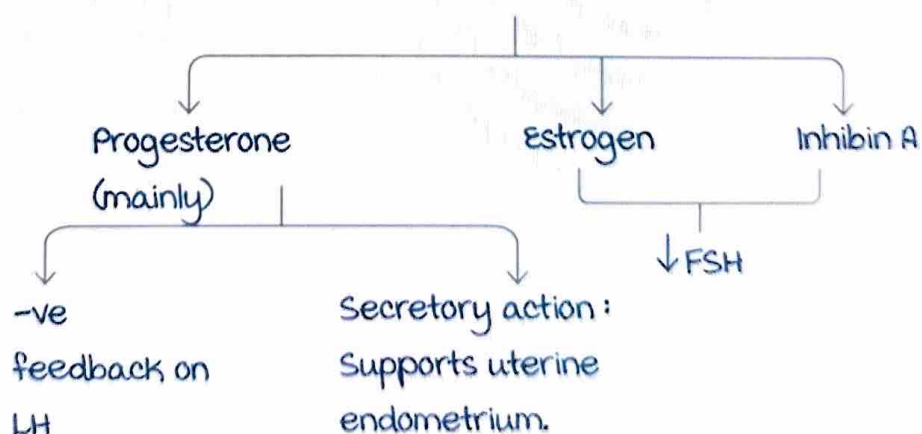
- Corpus luteum starts growing under the effect of LH.
- Day 22 of cycle/8 days after ovulation :
Attains maximum size and activity.



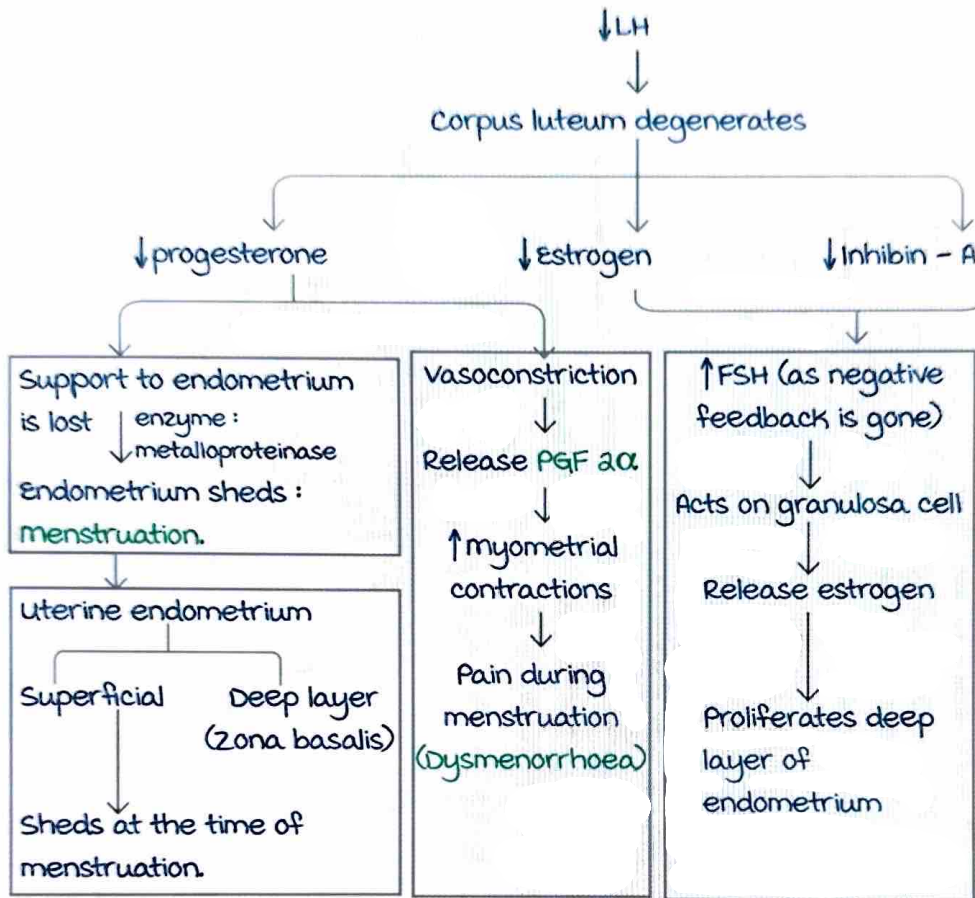
Hormones produced by corpus luteum

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Hormones released by corpus luteum

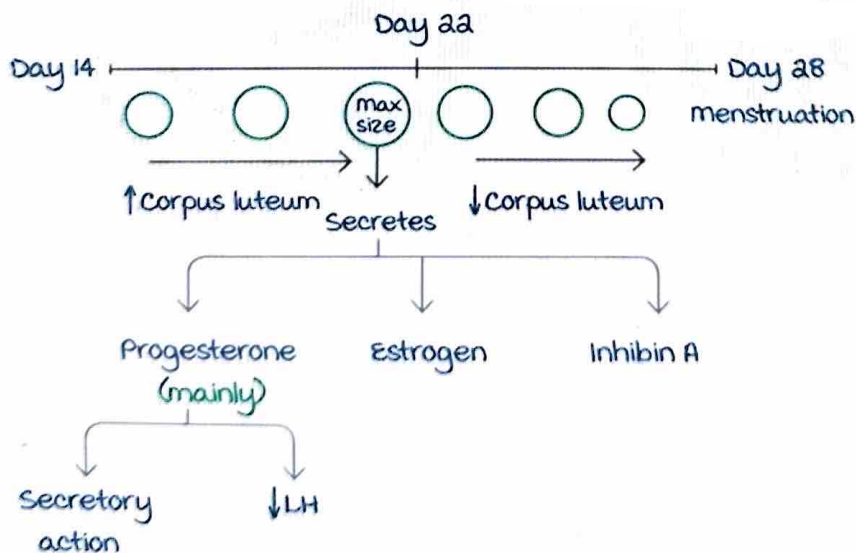


Progesterone	Low concentration	↑ LH, ↑ FSH.
	High concentration	↓ LH, ↓ FSH.



Note :

Deep layer is responsible for regeneration of entire endometrium in next cycle.



2nd half of menstrual cycle : **Luteal/Secretory phase**