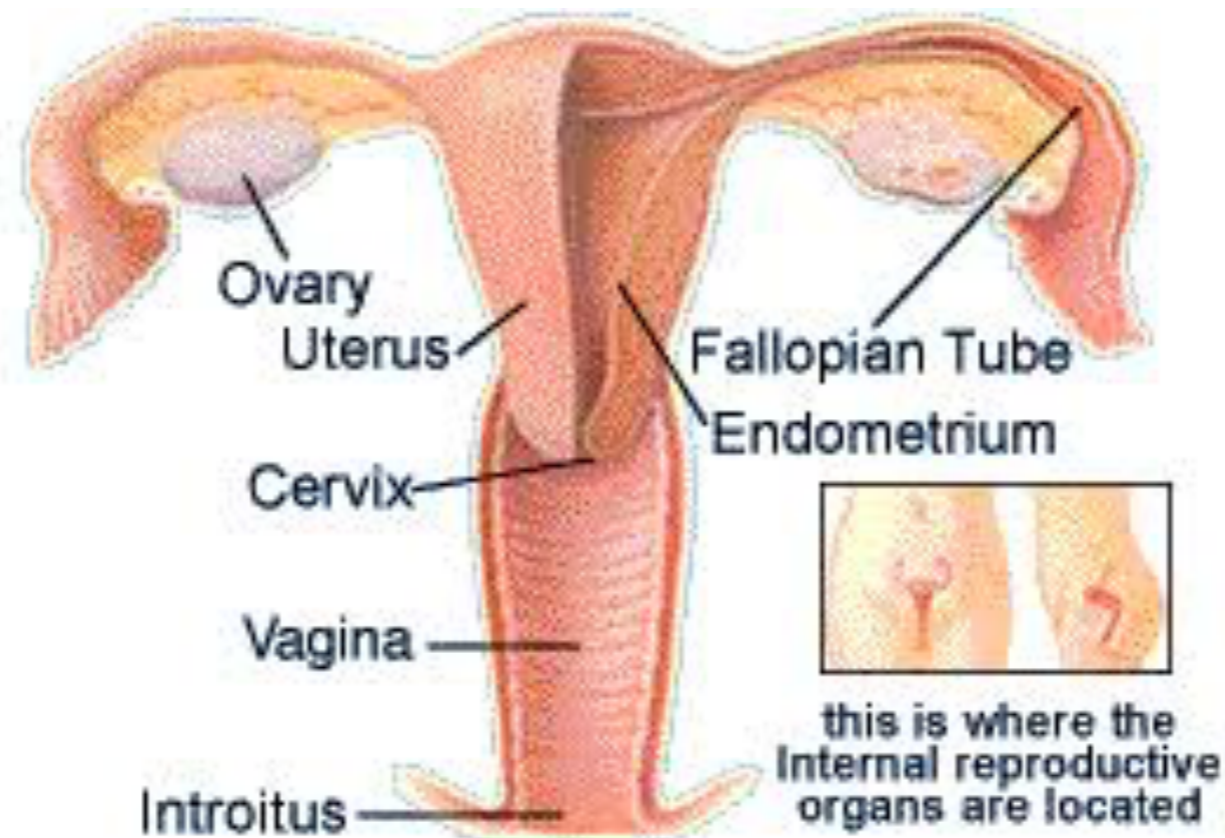


CONGENITAL ANOMALIES OF FRS

*PREPARED BY
DR.JUSTIN JEYA AMUTHA*

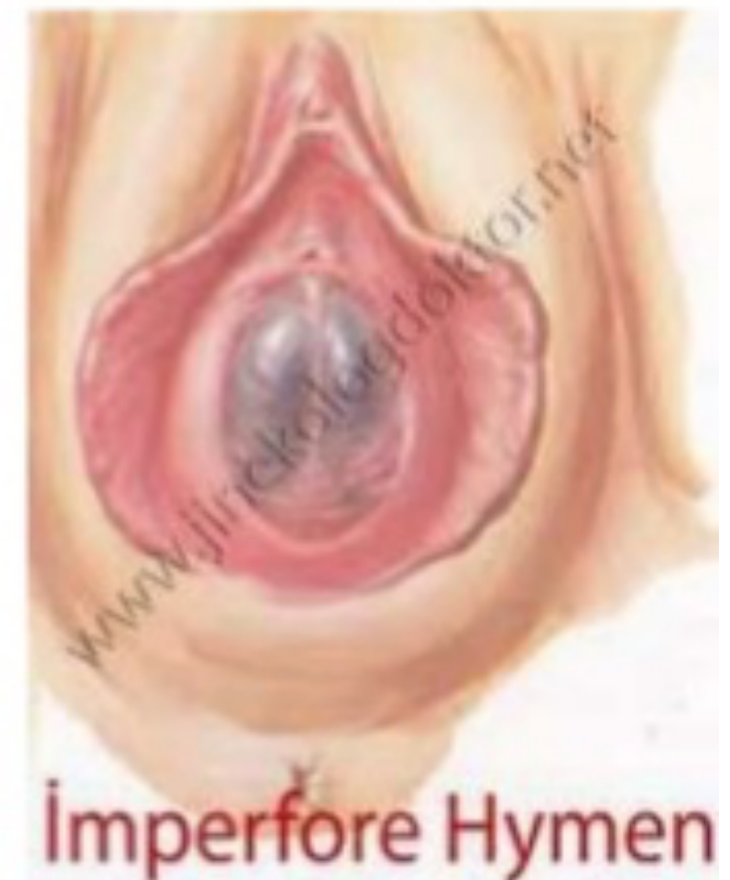
NARROW INTROITUS

- Revealed after marriage..
- **DYSPAREUNIA MAY BE THE FIRST COMPLAINT** or it may be detected during investigation of infertility
- Treatment is effective by manual stretching under general anaesthesia or by surgical enlargement.



HYMEN ABNORMALITY

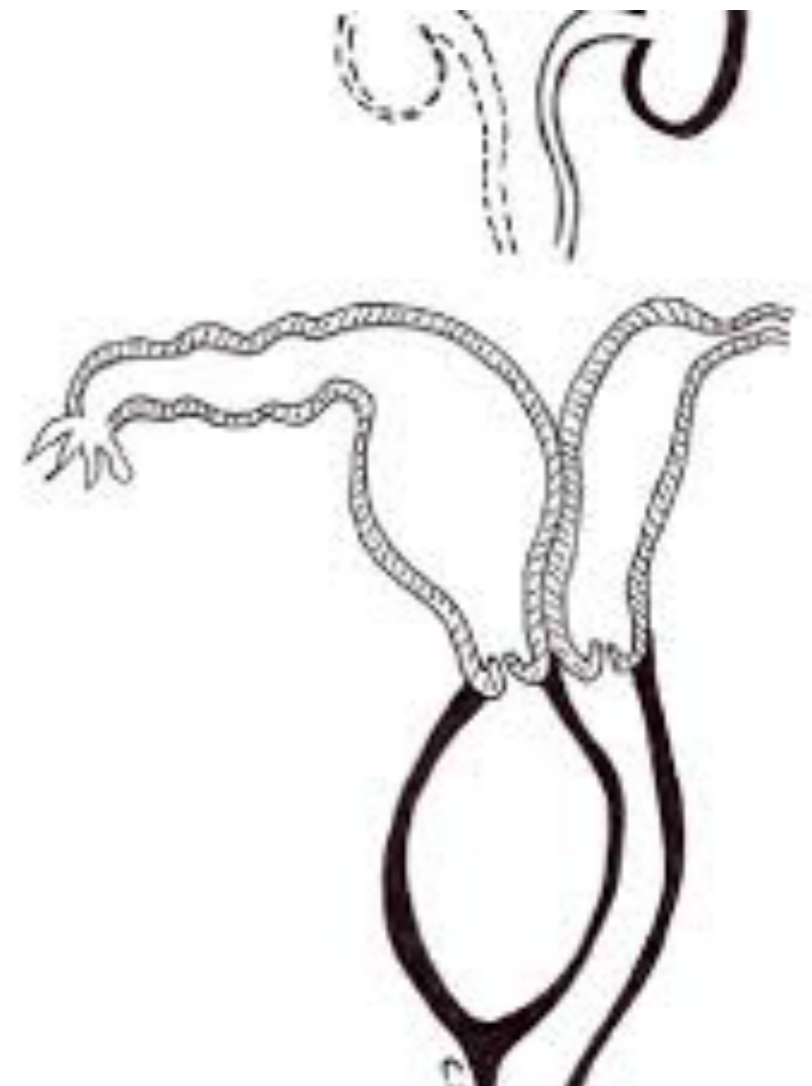
- Imperforate hymen → of significance abnormality.
 - Always unnoticed until 14 – 16 yrs.
 - Uterine functioning is normal → menstrual blood is pent up inside vagina behind the hymen (**CRYPTOMENORRHOEA**)
 - Depending upon the amount of blood so accumulated, it first distends the vagina (**HAEMATOCOLPOS**)
 - The uterus is next involved and cavity dilated (**HAEMATOMETRA**)
 - If late and neglected, tubes may also be distended after the fimbrial ends becoming closed by adhesions (**HAEMATOSALPINX**)



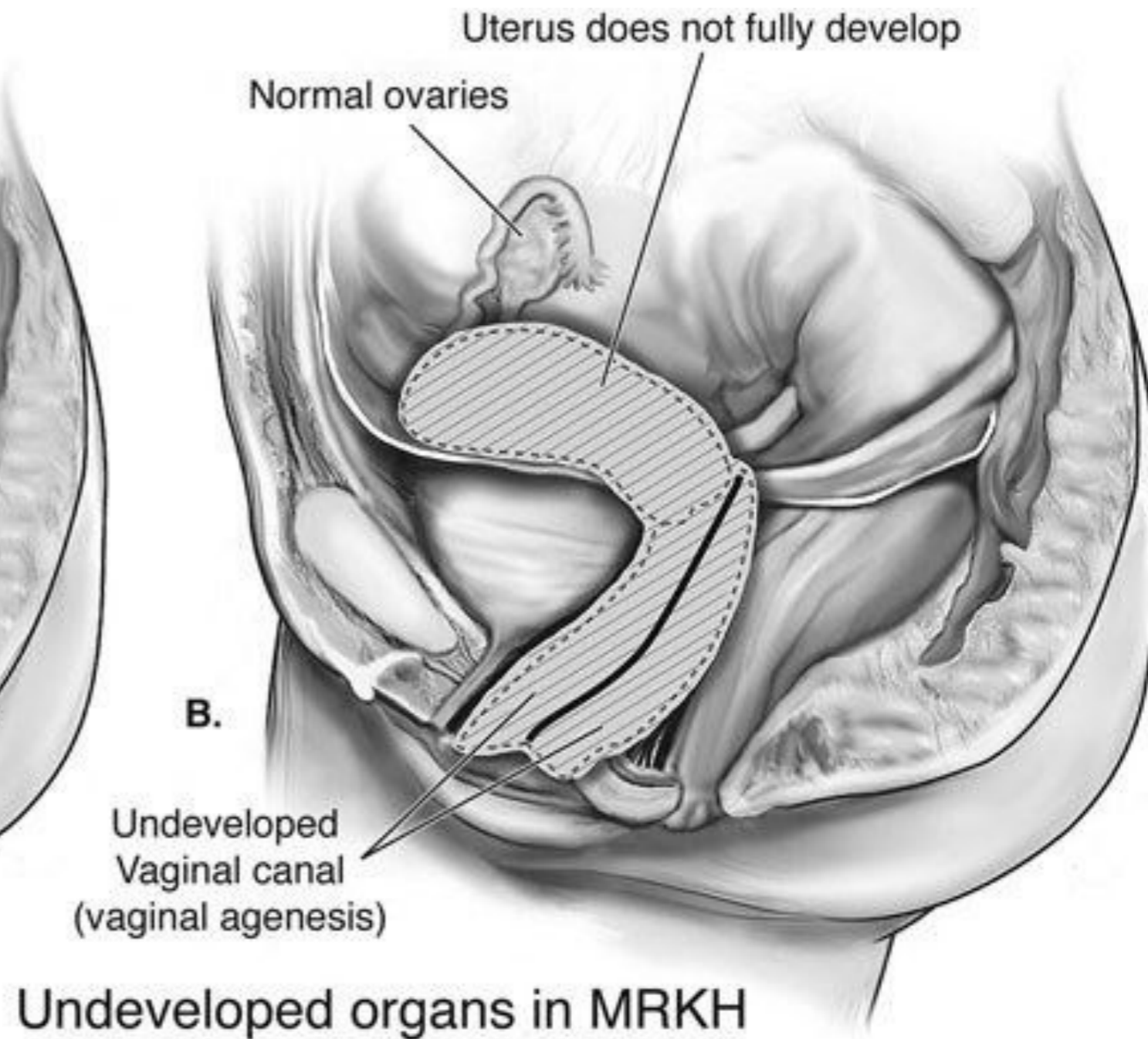
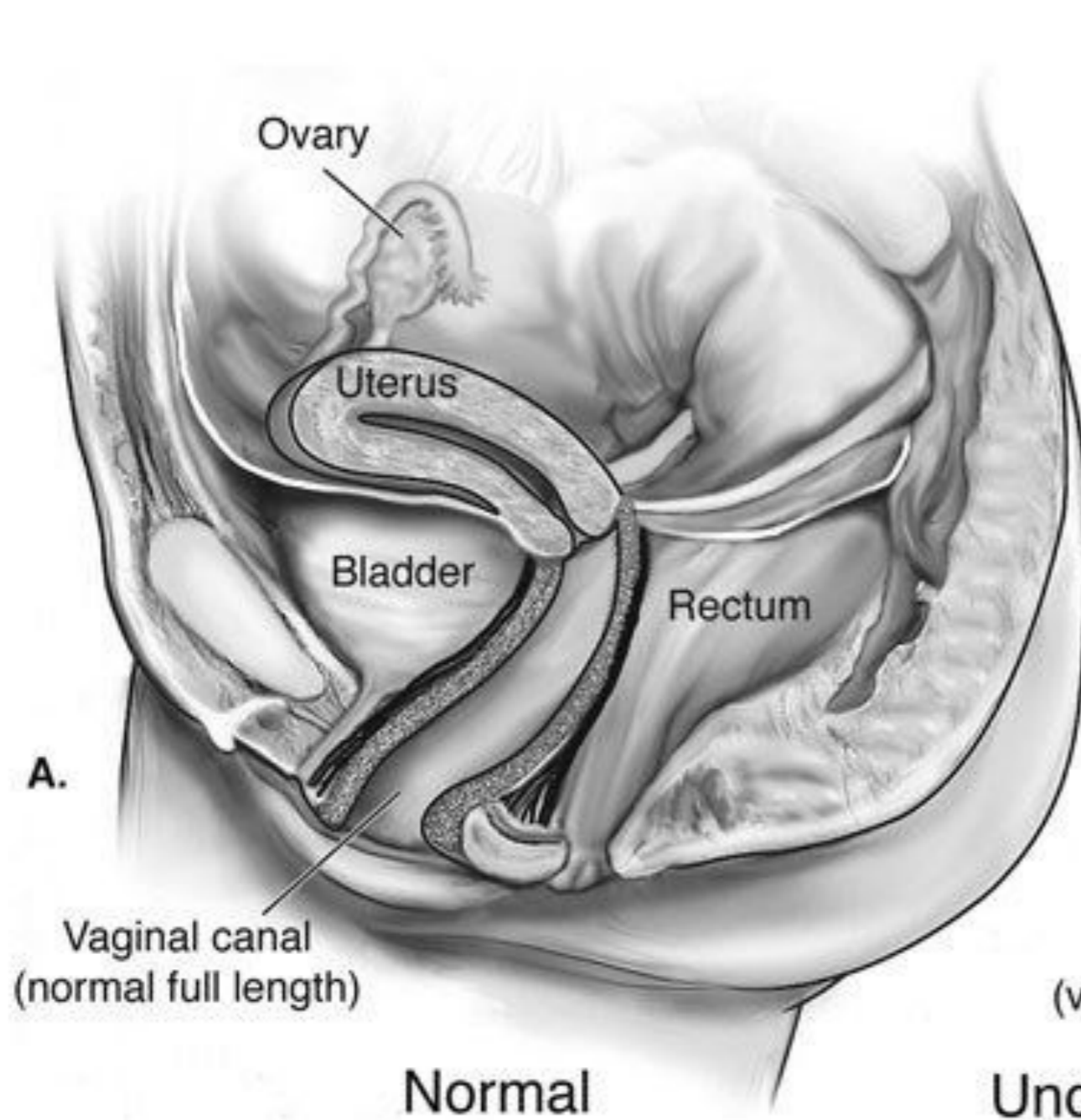
VAGINAL ABNORMALITIES

Developmental abnormalities of the normal single vagina include:

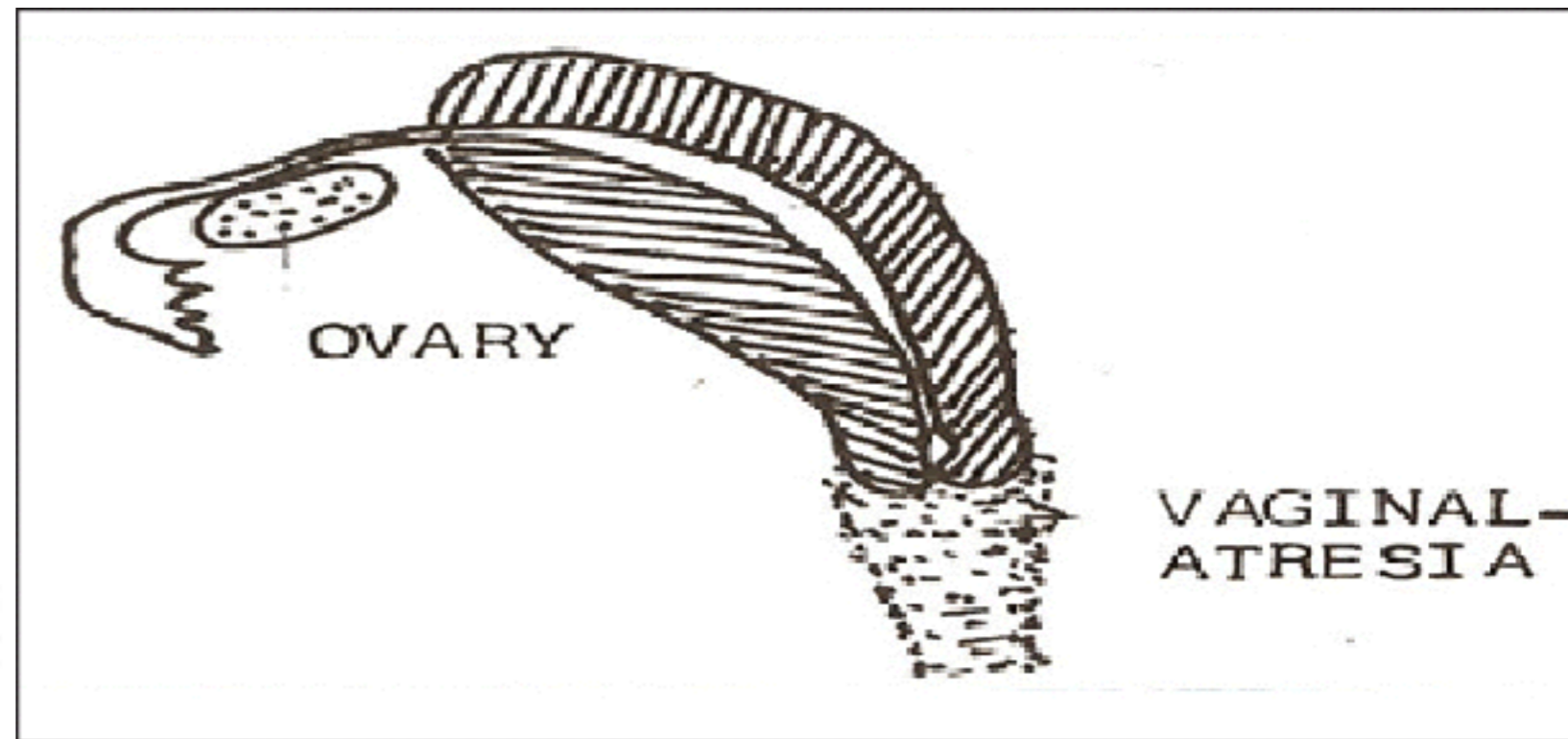
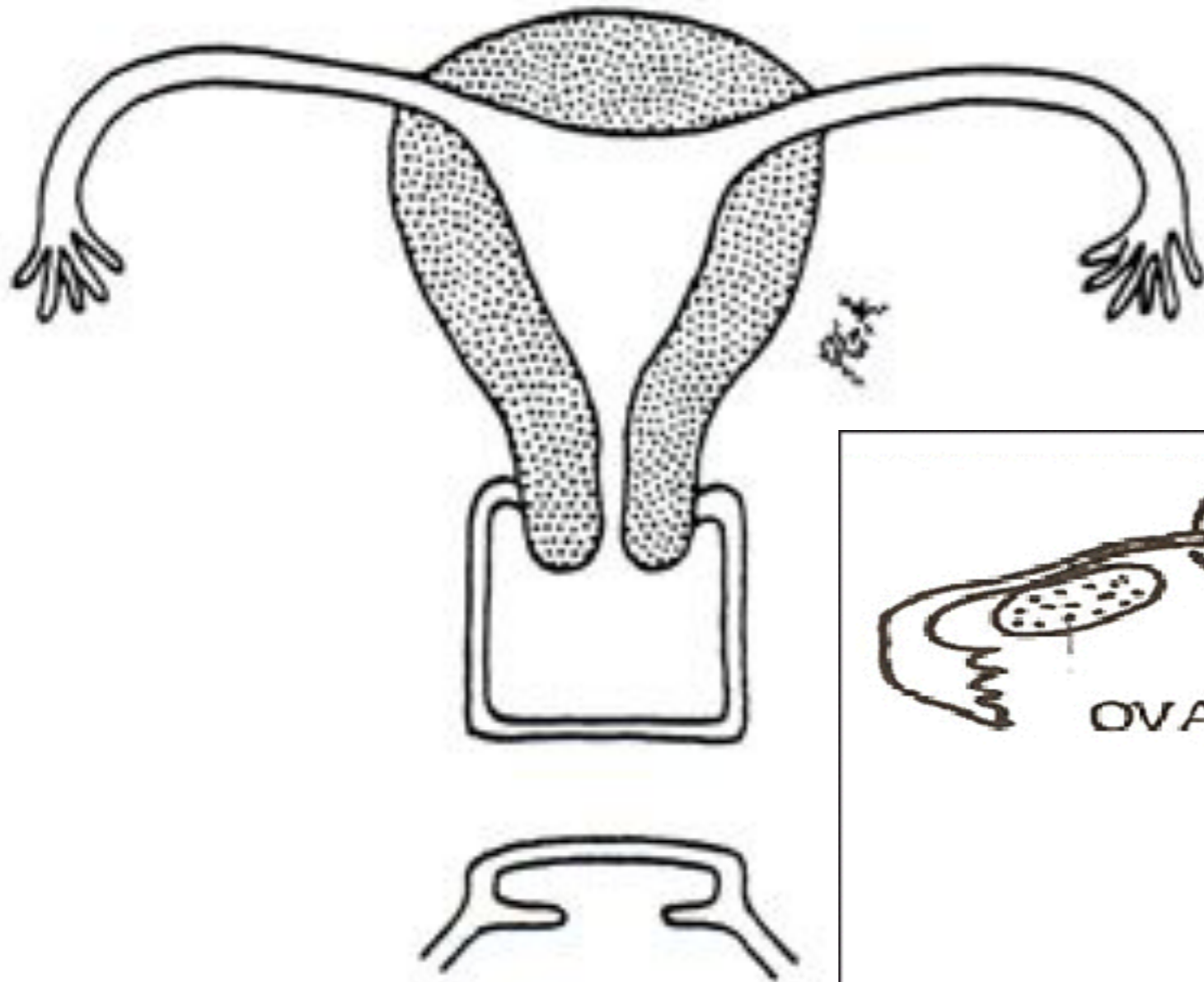
- Vaginal agenesis
- Vaginal atresia
- Double vagina
- Longitudinal vaginal septum
- Transverse vaginal septum



VAGINAL AGENESIS

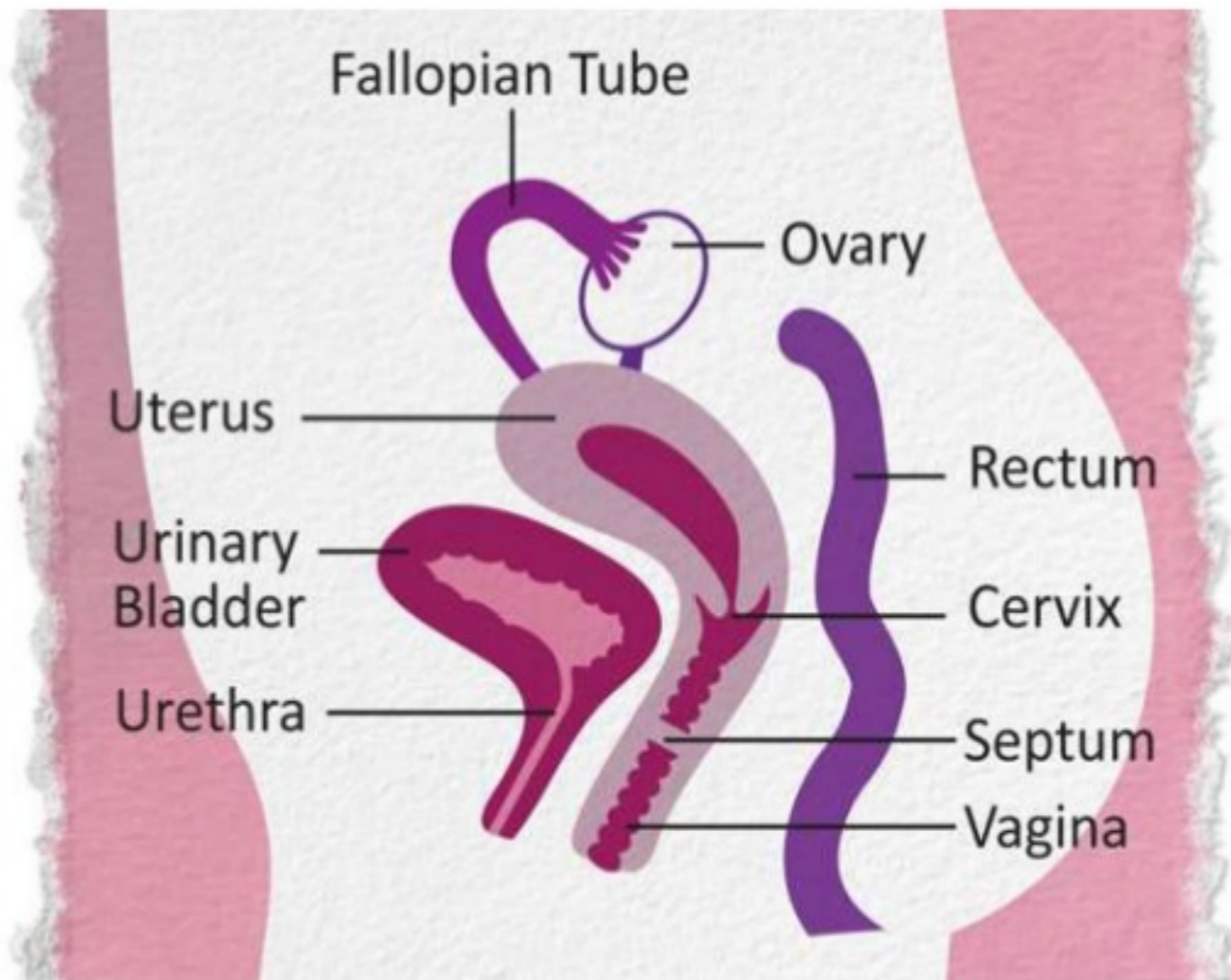


VAGINAL ATRESIA



TRANSVERSE SEPTUM ABOVE THE LOWER ONE - THIRD...

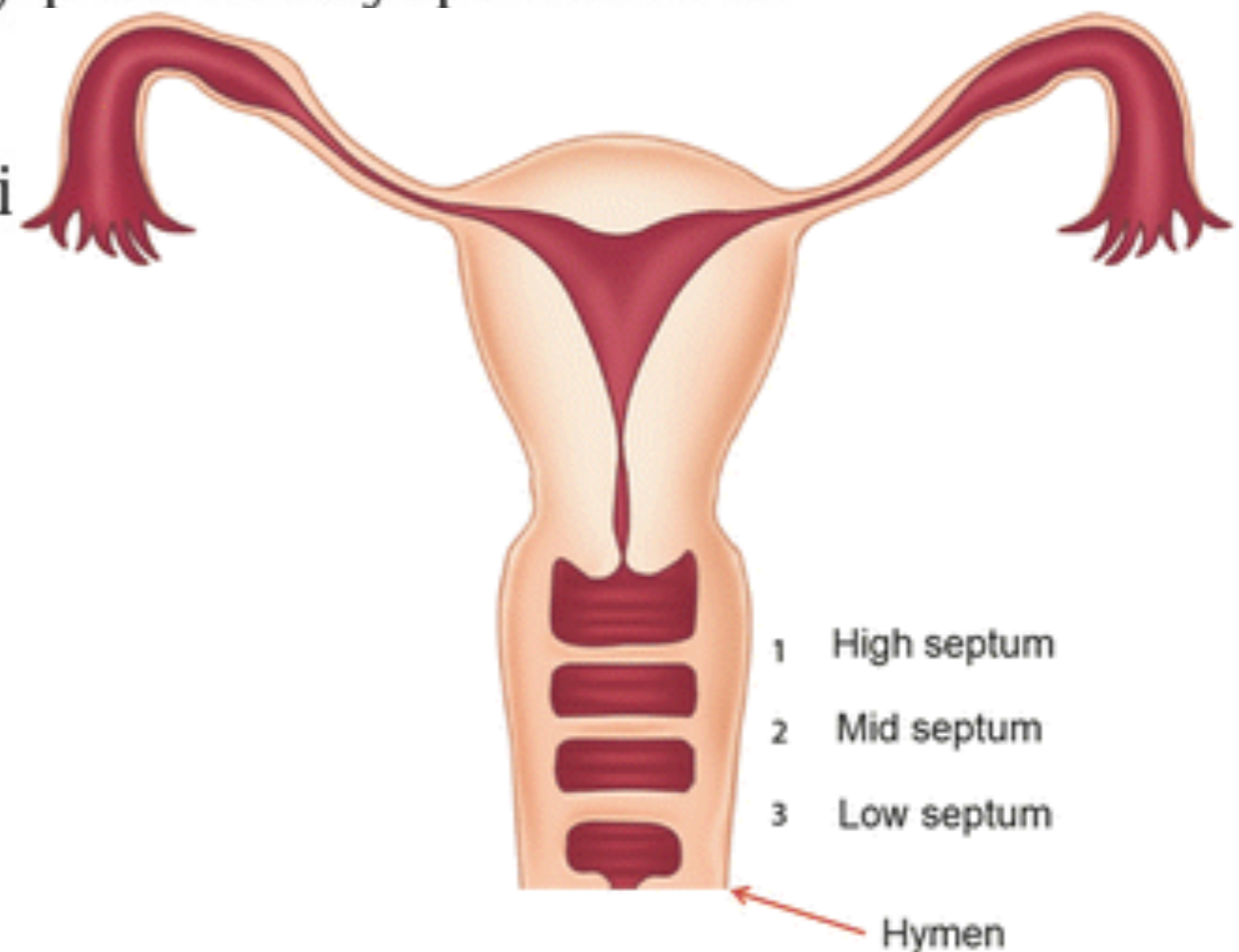
- Usually comes earlier than the previous one with the septum placed higher and higher



m

SEPTATE VAGINA

- Complete or incomplete longitudinal septum
- May be assoc with double uterus and double cervix
- May be asymptomatic or may produce dyspareunia or may obstruct delivery
- Treatment: septum to be exci



CERVICAL ABNORMALITIES

Atresia.

- The entire cervix may fail to develop.
- This may be combined with incomplete development of the upper vagina or lower uterus

Double cervix.

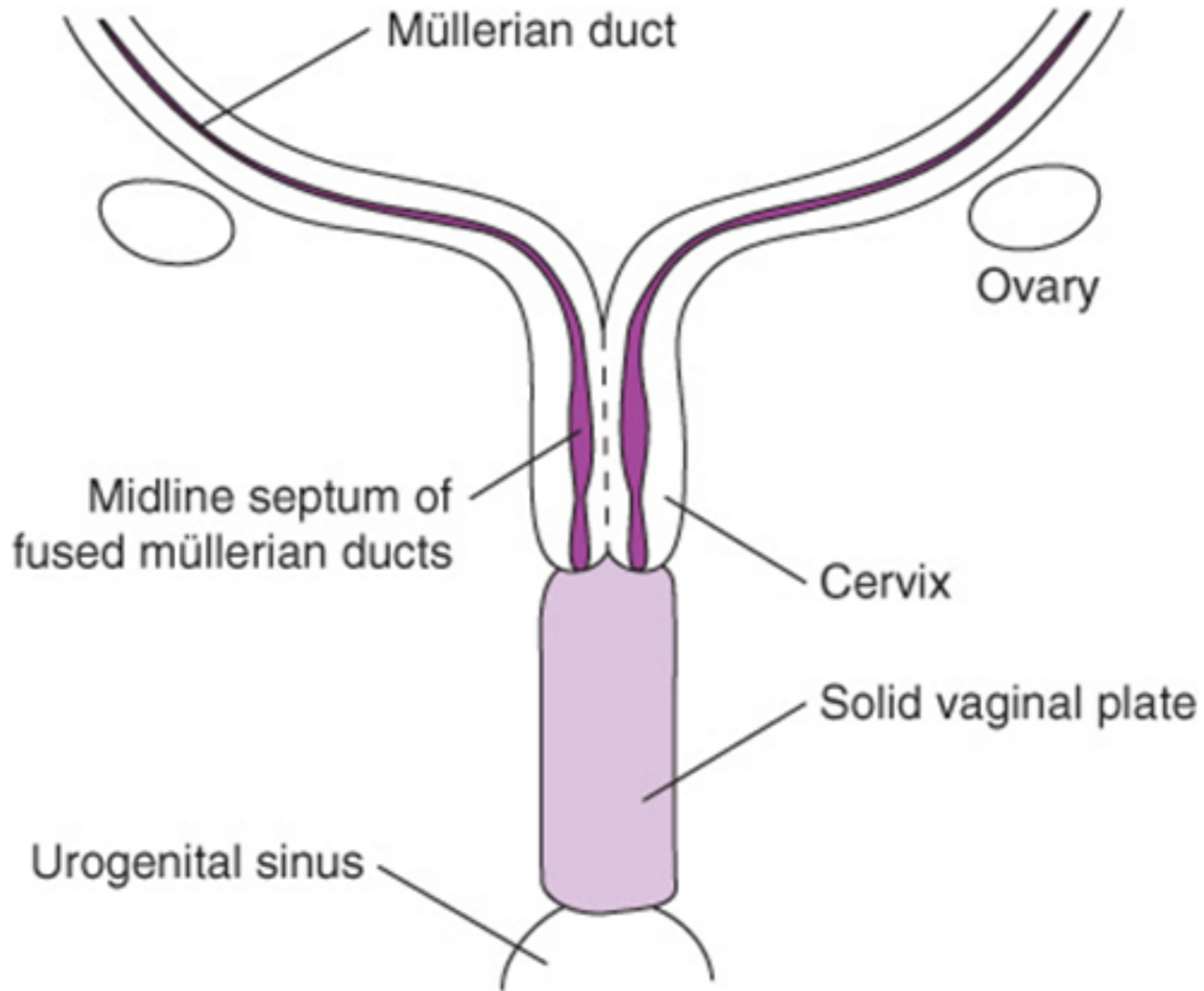
- Each distinct cervix results from separate müllerian duct maturation.
- Both septate and true double cervixes are frequently associated with a longitudinal vaginal septum.
- Many septate cervixes are erroneously classified as double.

Single hemicervix.

- This arises from unilateral müllerian maturation.

Septate cervix.

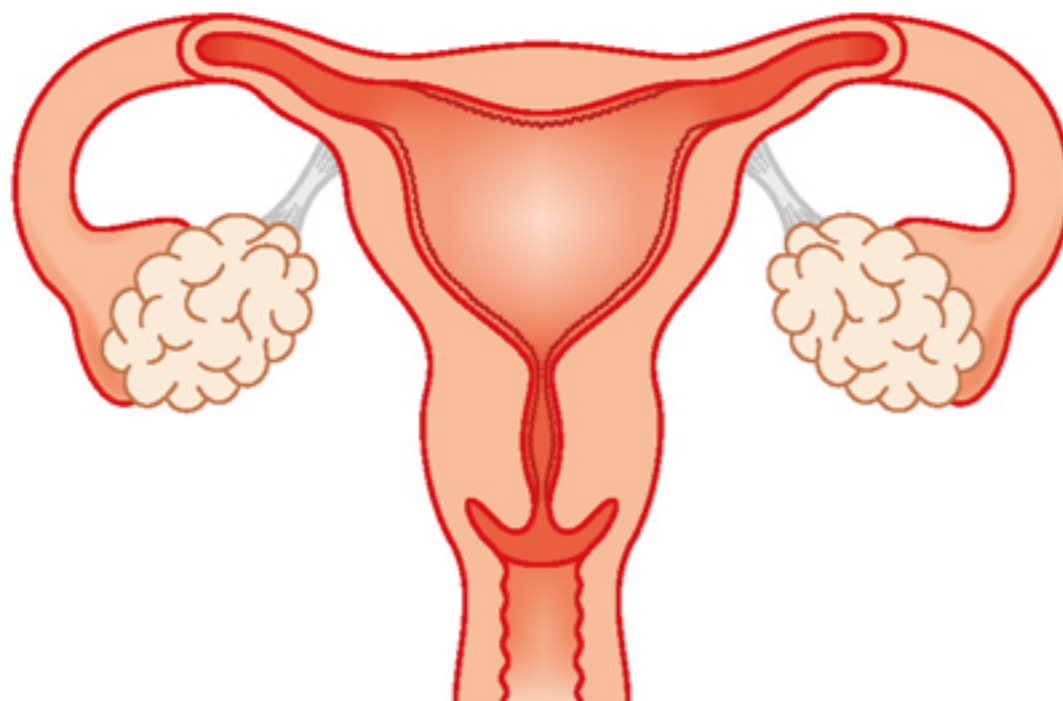
- This consists of a single muscular ring partitioned by a septum.
- The septum may be confined to the cervix, or more often, it may be the downward continuation of a uterine septum or the upward extension of a vaginal septum.



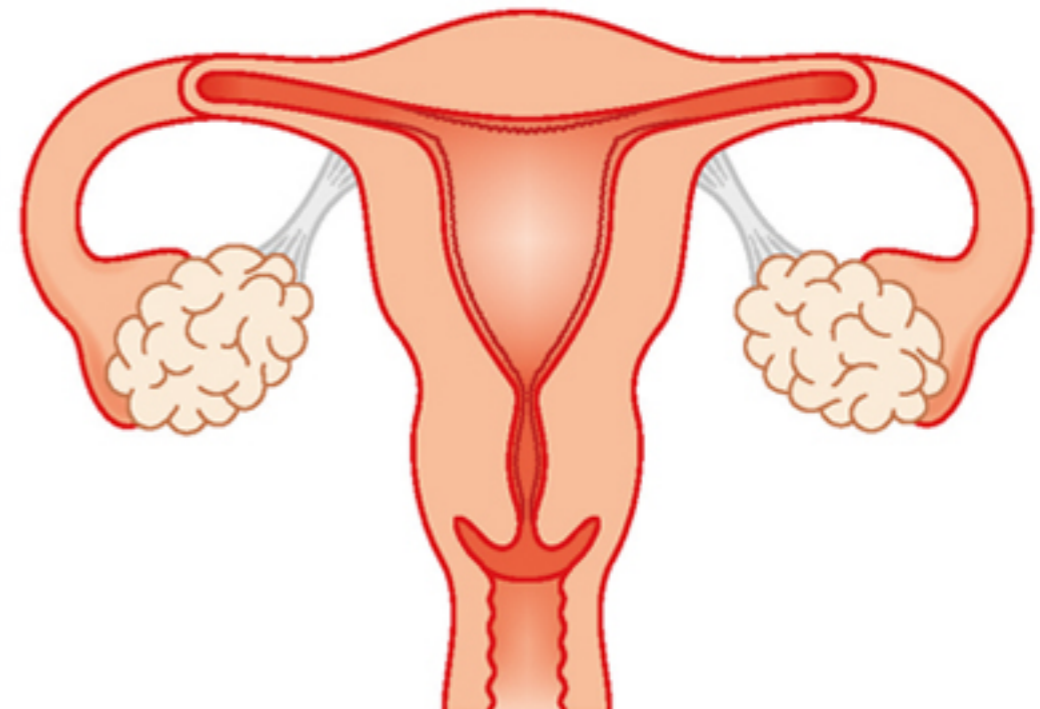
TYPES:

- **Arcuate**

- Fundus looks concave with heart shaped cavity outline
- This looks more like a normal uterus, except it has a dip, or slight indentation at the top.



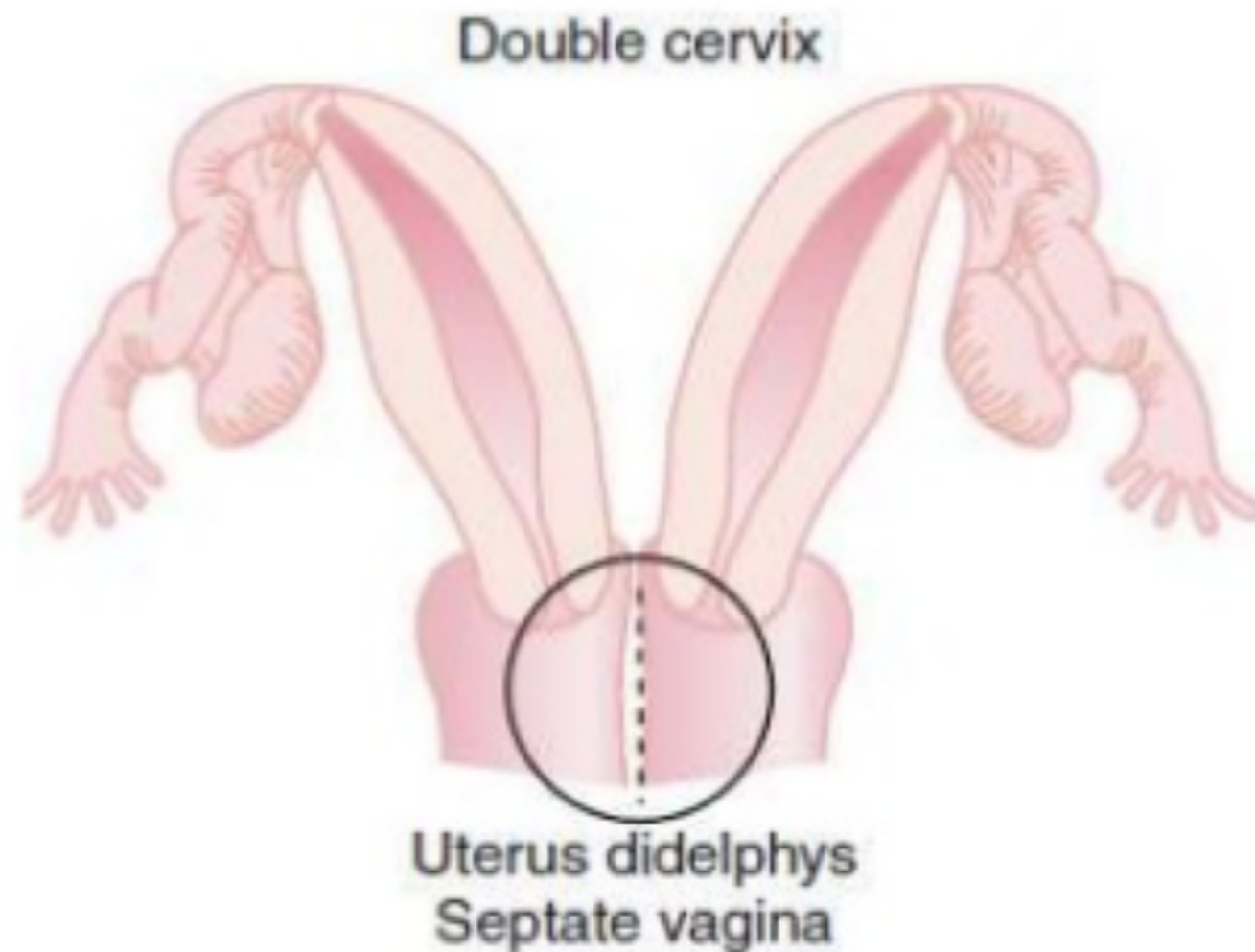
6. Arcuate uterus

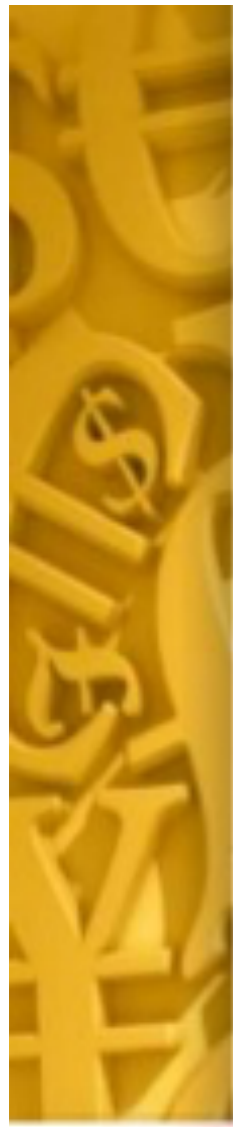


1. Normal uterus

Uterus didelphys

Two separate uterine bodies, each with its own cervix and attached fallopian tube and vagina





- Uterus bicornis
 - There are varying degree of fusion of the muscle walls of the two ducts.
 - Uterus bicornis bicollis : 2 uterine cavity with double cervix with or without vaginal septum.



Uterus duplex bicornis



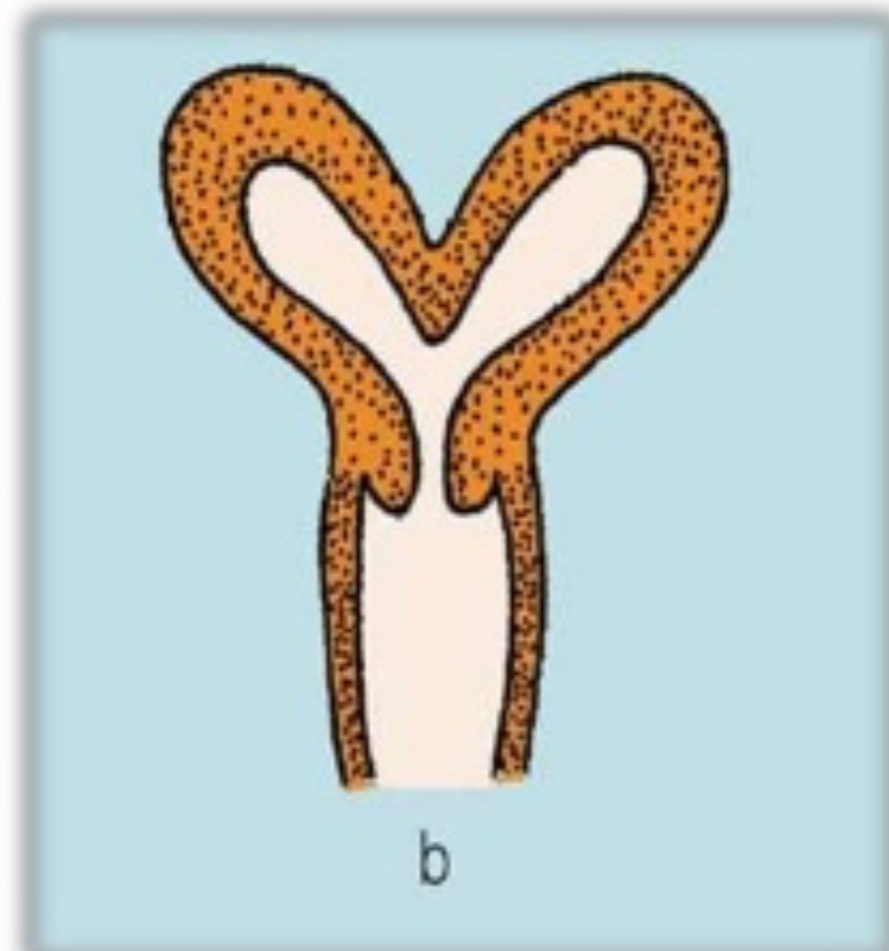
Bicornuate bicollis

F. Sullivan
2008



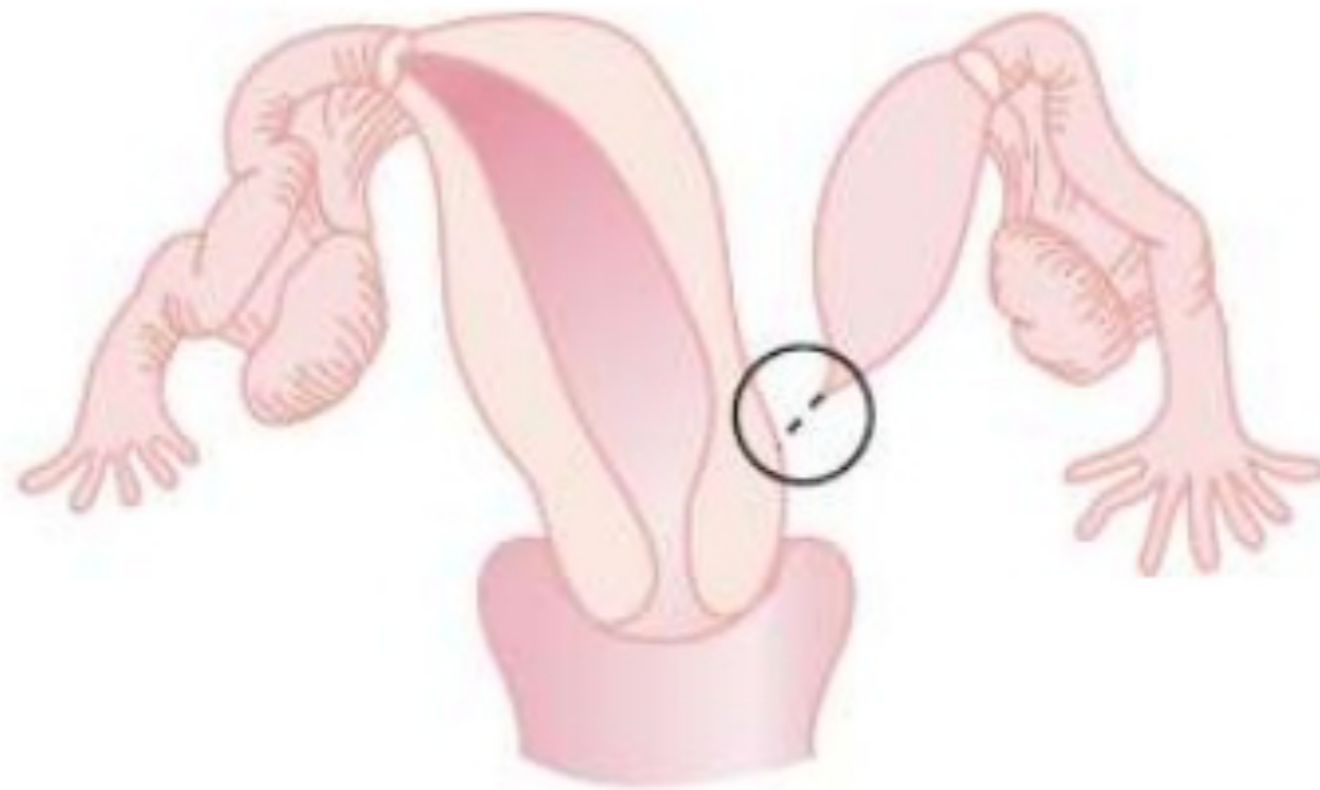
- Uterus bicornis unicollis

- 2 uterine cavity with one cervix
- Horns may be equal or one horn may be rudimentary and have no communication with the developed horn.



A bicornuate uterus with a rudimentary horn

Represents a fusion failure



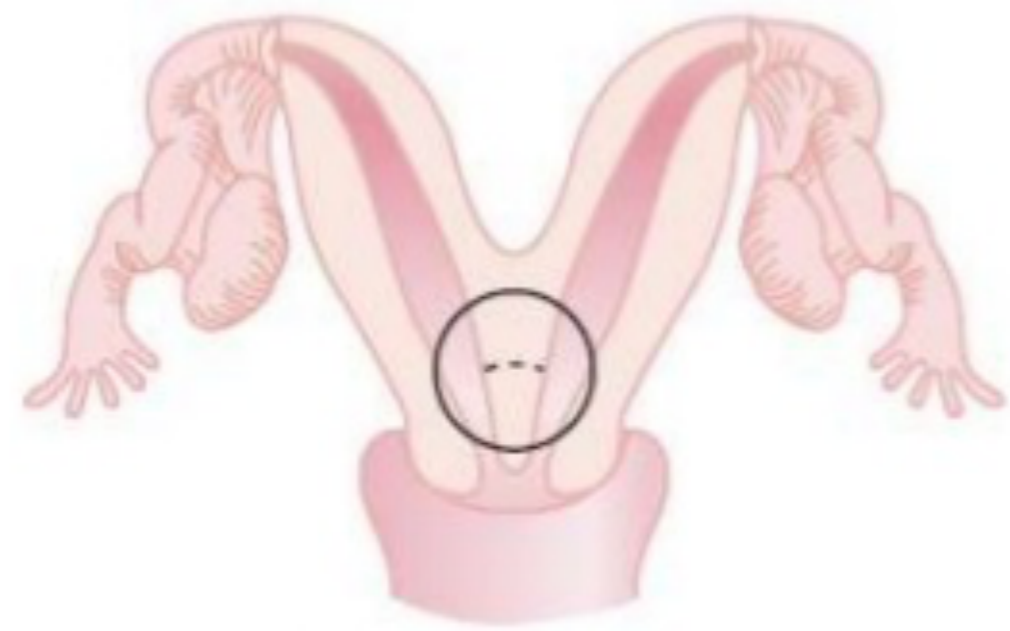
Bicornuate uterus
Rudimentary horn

bicornuate uterus with or without double cervixes

- Bicornuate and unicornuate uteri are associated with second-trimester pregnancy loss, malpresentation, and preterm labor and delivery



Bicornuate uterus



Bicornuate uterus
Double cervix

Unicornuate uterus

Failure of formation

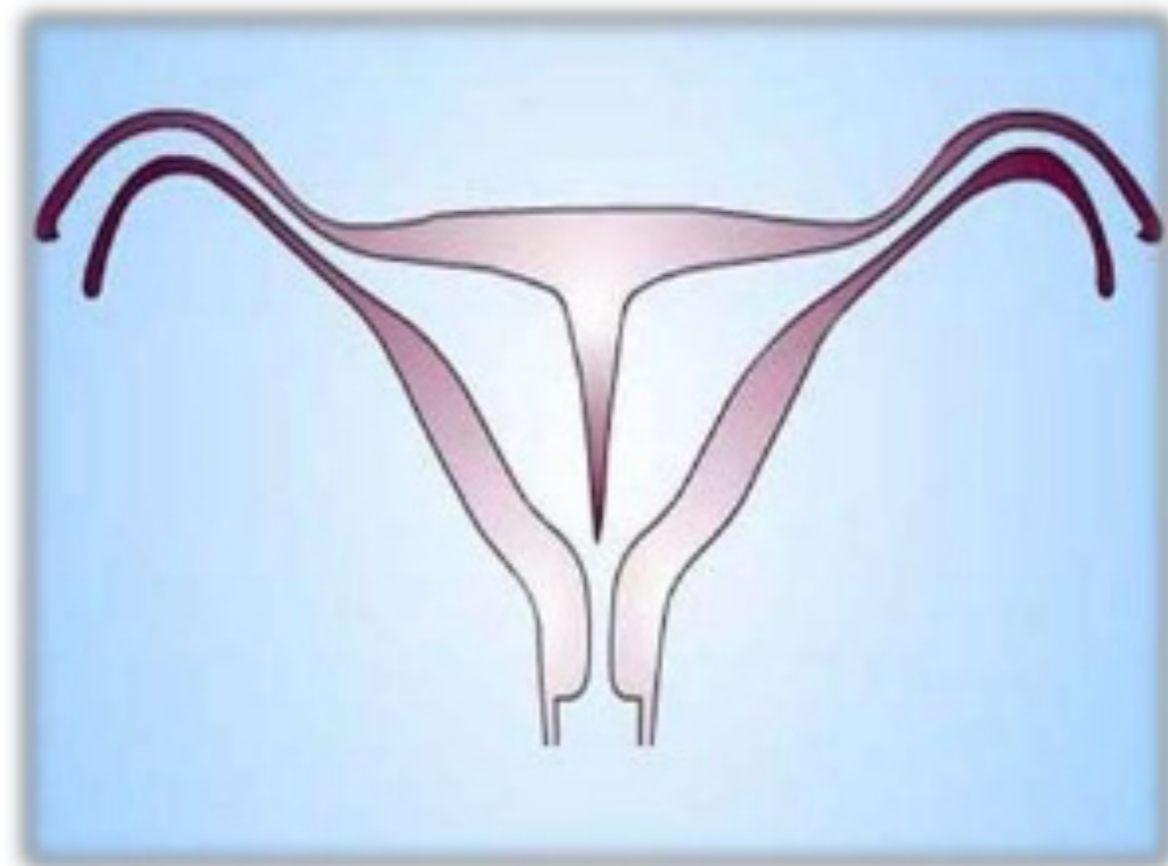


Unicornuate uterus



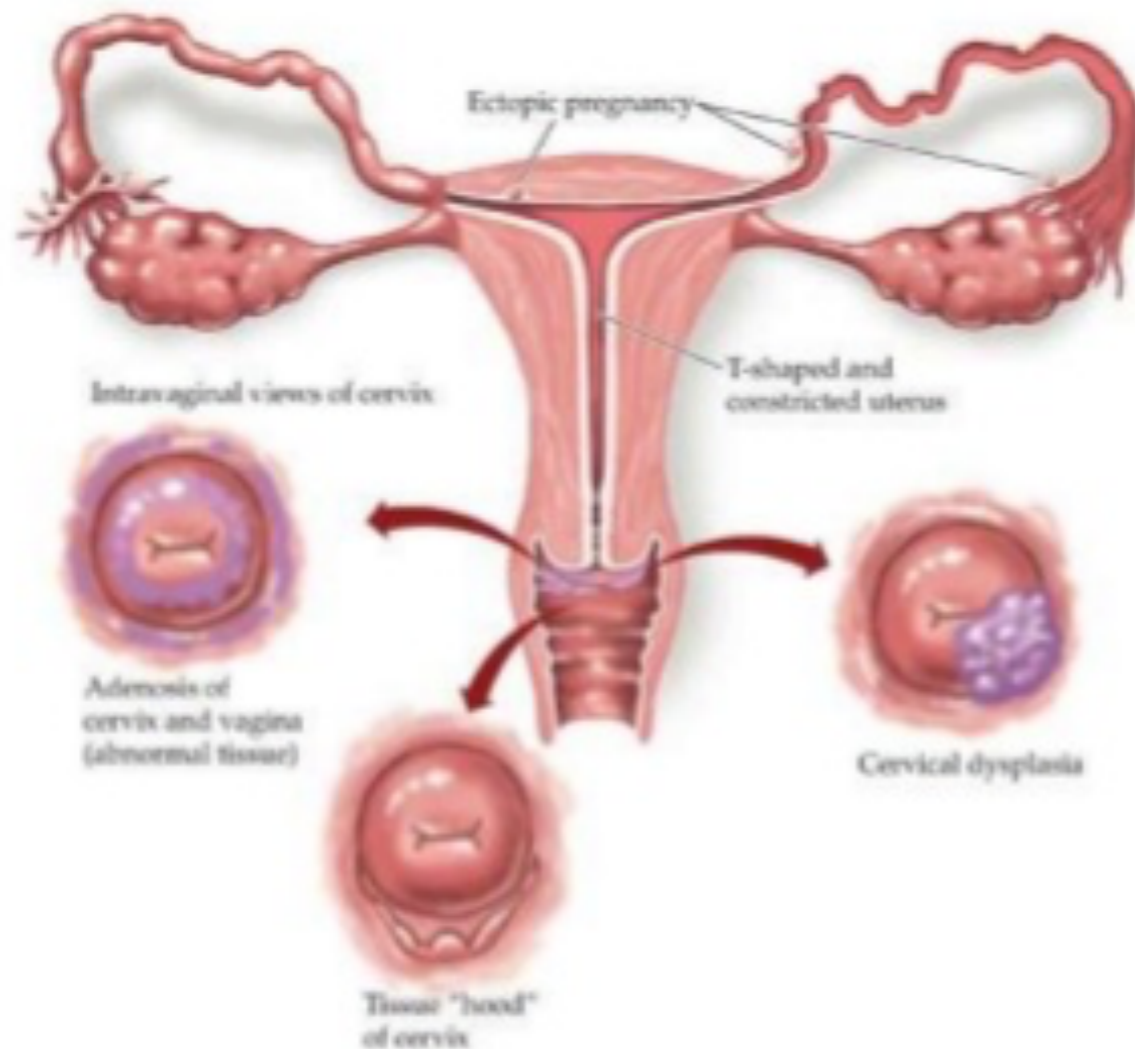
- Septate uterus

- 2 mullerian ducts fused together but there is persistence of septum in between the two either partially or completely



DES-exposed female's infant

- T-shaped endometrial cavity
- cervical collar deformity



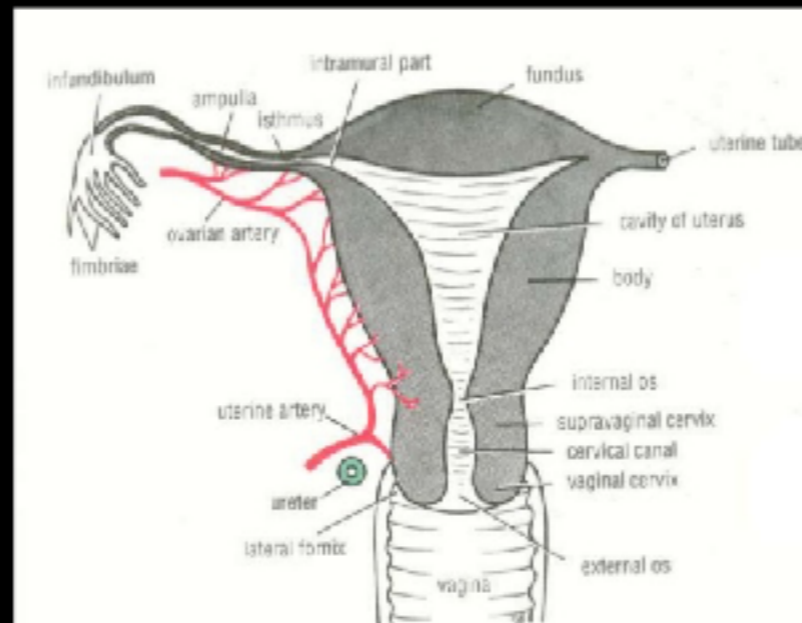
- **METROPLASTY**

- Women with septate or bicornuate anomalies and poor reproductive outcomes may benefit from uterine surgery
- Repair of a bicornuate uterus is by transabdominal metroplasty involving septal resection and fundal recombination
- Repair of a septate uterus is usually by hysteroscopic septal resection

FT ABNORMALITY

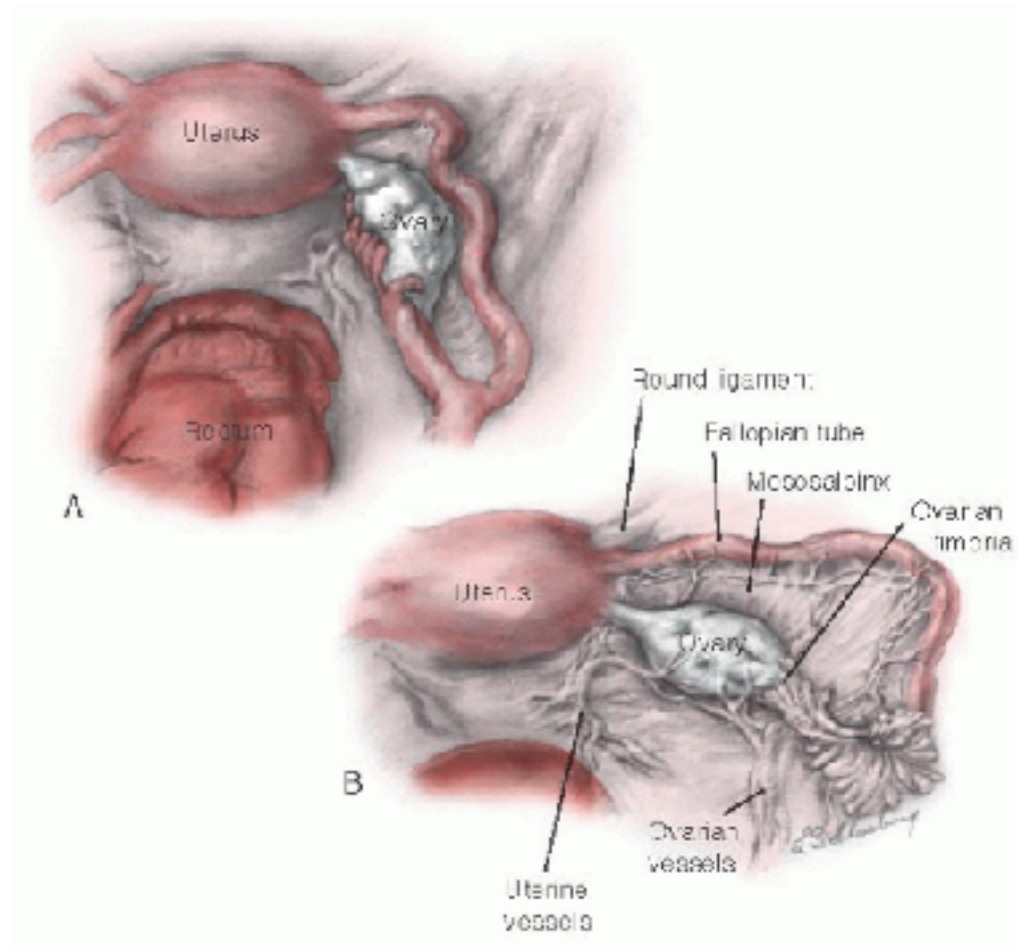


- Tubes may be unduly elongated
- Rarely tube may be absent on one side
- These may lower the fertility or favour ectopic pregnancy



OVARY ABNORMALITY

- There may be streak of gonads or gonadal dysgenesis which are usually associated with errors of sex chromosomal pattern.
- No treatment will help this condition
- Accessory ovary may be present
- Supranumerary ovaries may be present in the broad ligament or elsewhere (rare)



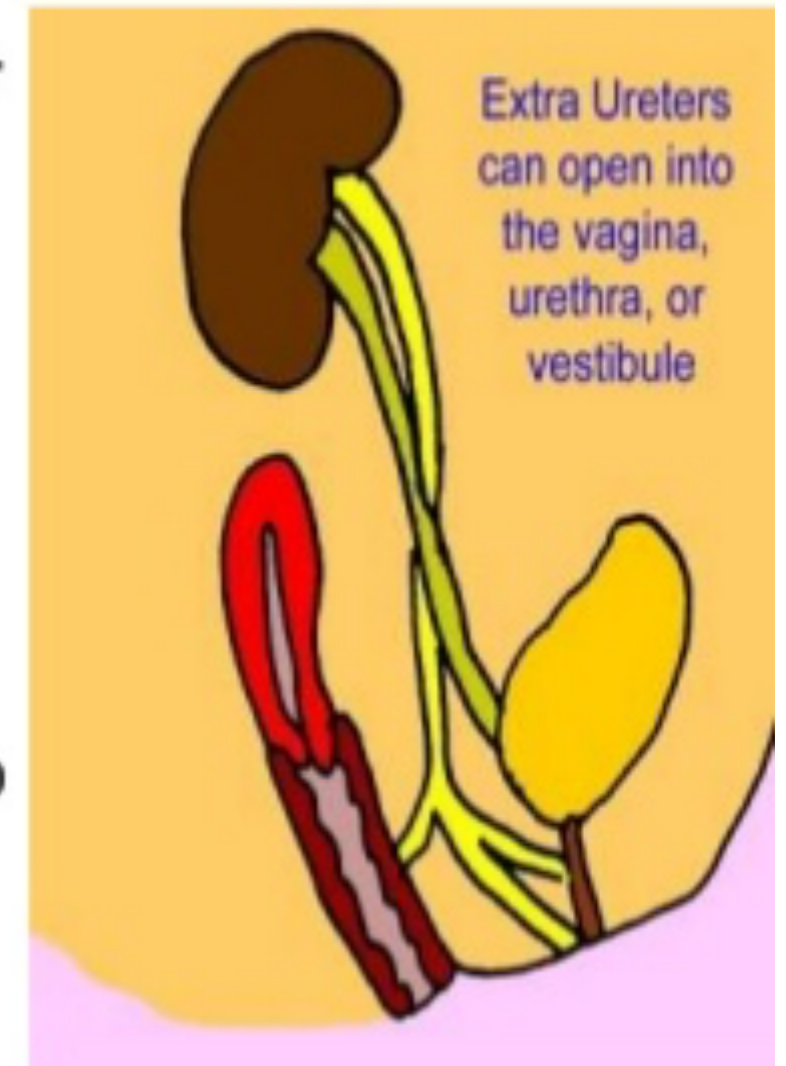


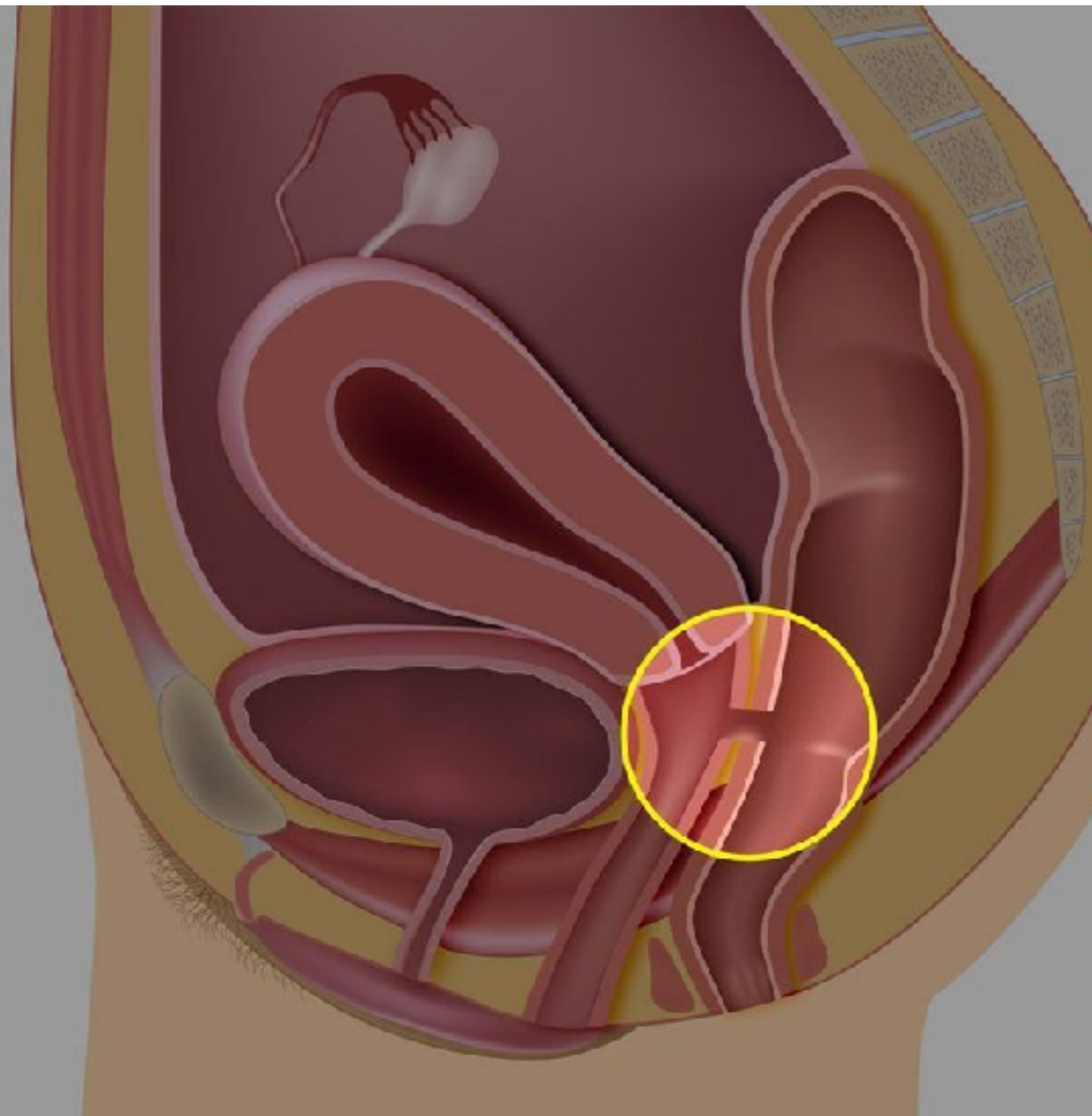
PERINEAL OR VESTIBULAR ANUS

- Detected at the time of birth
- Usual anal opening site is evidenced by anal pit
- Either anus will be situated close to the posterior end of the vestibule or in the vestibule.
- In vagina (rarely) → (congenital rectovaginal fistula)
- Opening is usually sufficiently big
- Future reproduction → not a problem
- Ceaserian section adviced in the future

ECTOPIC URETER

- Additional ureteric opening is usually in the vestibule close to the urethra or in the vagina.
- Symptom:-
 - Uncontrollable wetness
- Partial nephrectomy and ureterectomy may be indicated or implantation of the ectopic ureter into the bladder may be done.





Normal Anatomy

Intestines

Bladder

Uterus

Rectum

Vagina



Rectocele

Rectocele

VULVAR ABNORMALITIES

Atresia

- Complete *atresia of the vulva* includes *atresia of the* introitus and lower third of the vagina.
- In most cases, however, atresia is incomplete and results from adhesions or scars following injury or infection
- The defect may present a considerable obstacle to vaginal delivery, deep perineal tears may result.

Labial Fusion

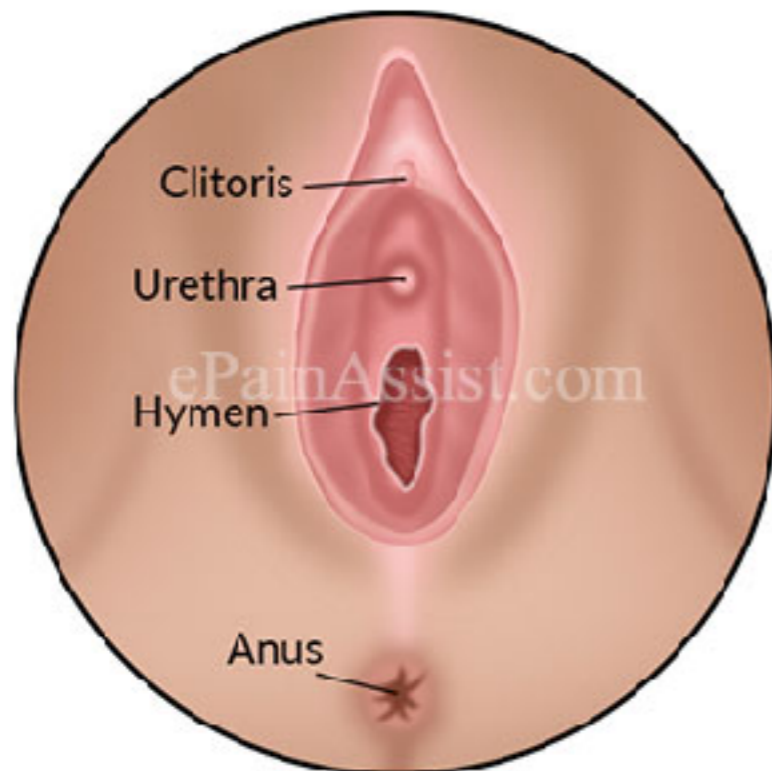
- Most commonly due to congenital adrenal hyperplasia.

Imperforate hymen is persistence of the fusion between the sinovaginal bulbs at the vestibule

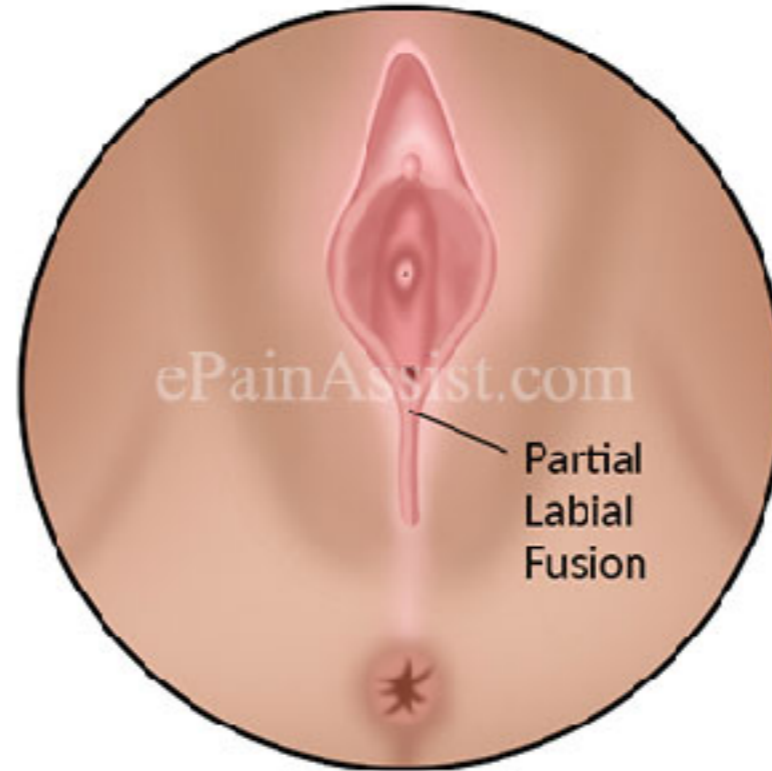
- Associated with primary amenorrhea and hematocolpos and is not encountered often during pregnancy.



Labial Fusion



Normal



Labial Fusion