

# Normal and abnormal development of the female genital system



Components which form female and male reproductive systems are:

1. **Gonads** Ovaries and Testes

2. **Genital Duct Systems**

Mesonephric and Paramesonephric ducts

3. **External Genitalia**

λ Genotype of embryo 46XX or 46XY is established at fertilization

λ SRY (sex-determining region on Y) that encodes a protein called testis-determining factor (TDF) are responsible for male differentiation

# Indifferent Embryo

- λ Weeks 1-6 sexually indifferent or undifferentiated stage
- λ Week 7 begins phenotypic sexual differentiation
- λ Week 12 female or male characteristics of external genitalia can be recognized and completed at 20 weeks.

# At 4<sup>TH</sup> Week of gestation

λ **Mesonephric Duct**

λ extending from the

λ mesonephros

λ (Wolff's body)

λ to the cloaca

λ (urogenital sinus)

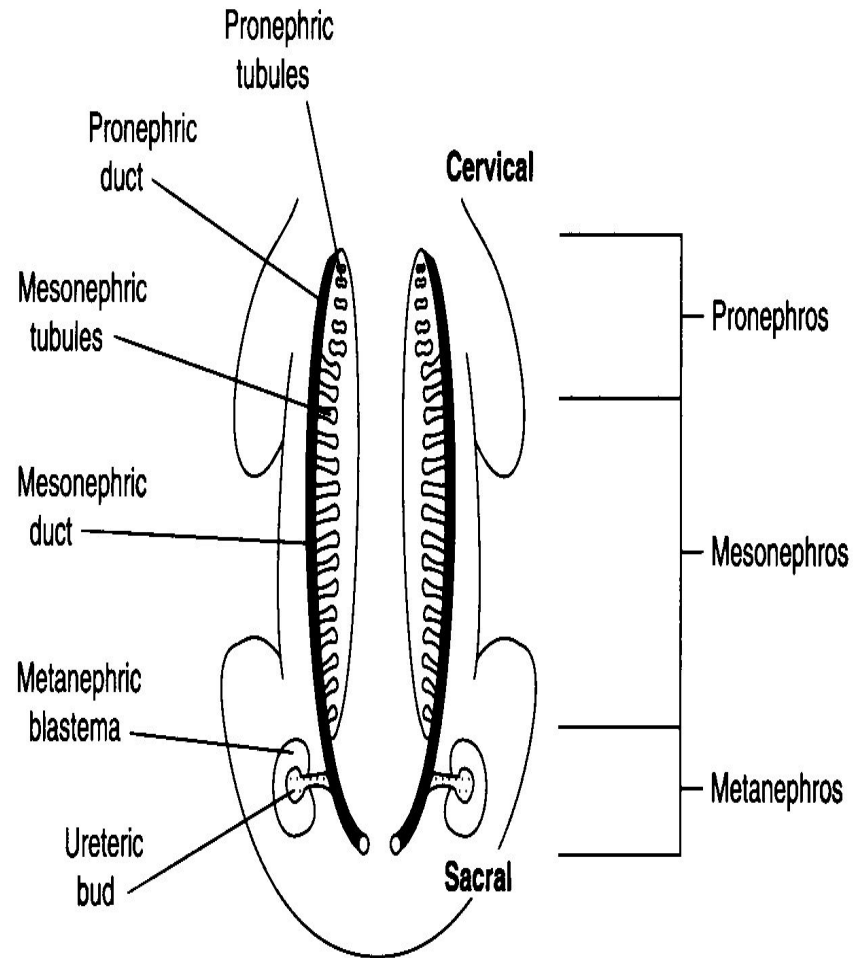
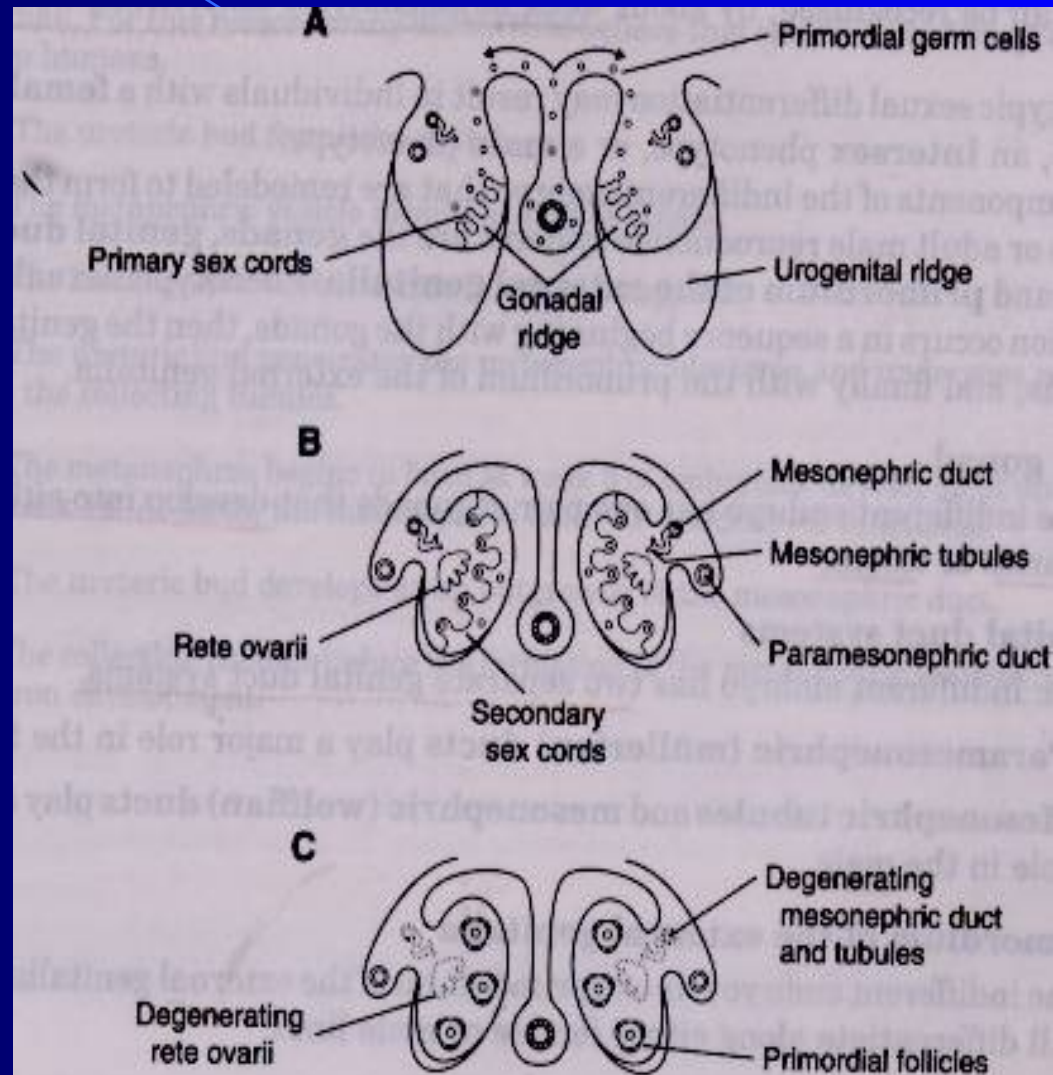


Figure 12.2 Frontal view of an embryo, depicting the pronephros, mesonephros, and metanephros. Note

# At 5<sup>TH</sup> Week of gestation

λ A Swelling on either side of dorsal mesentery on medial side of mesonephric duct forms the **urogenital ridge**

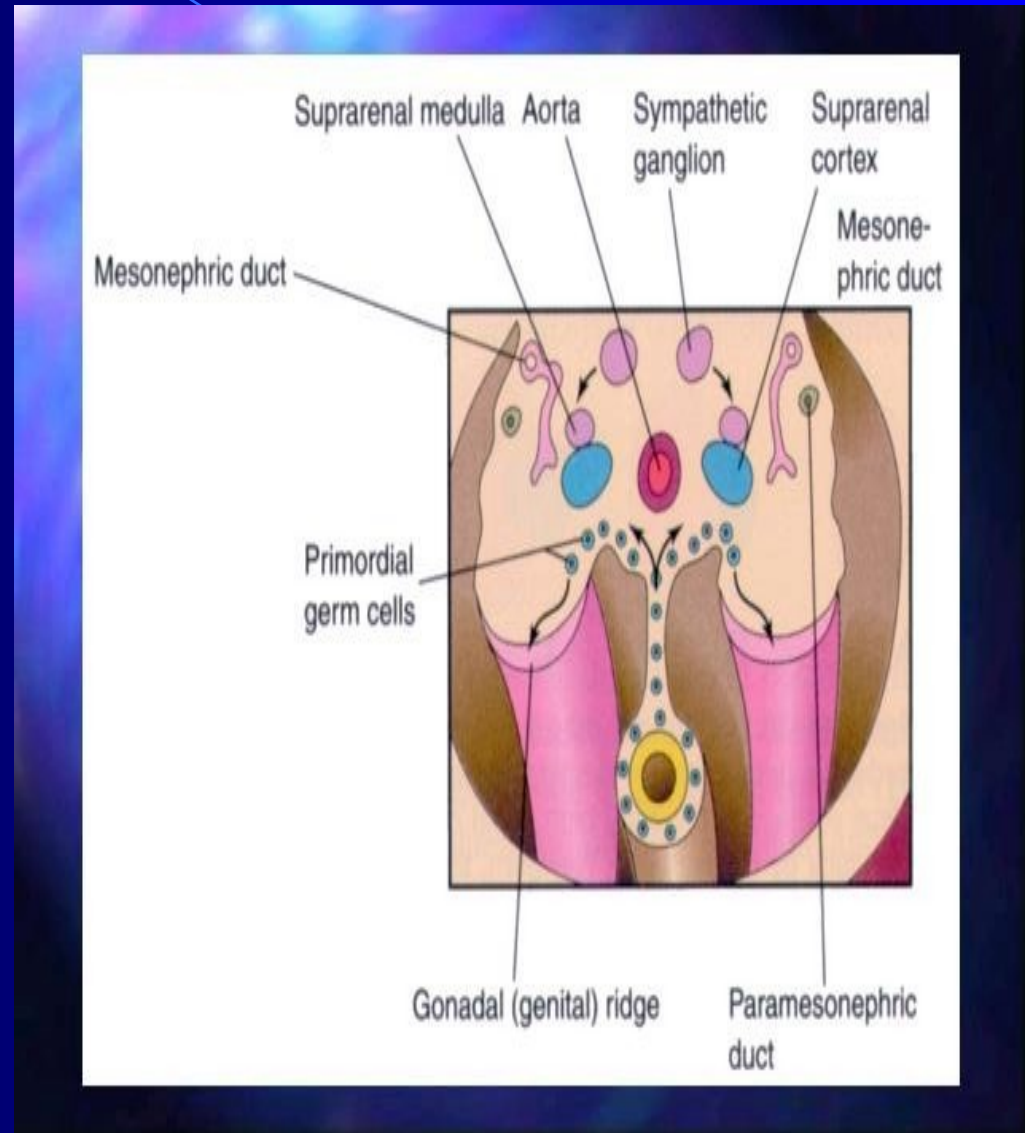


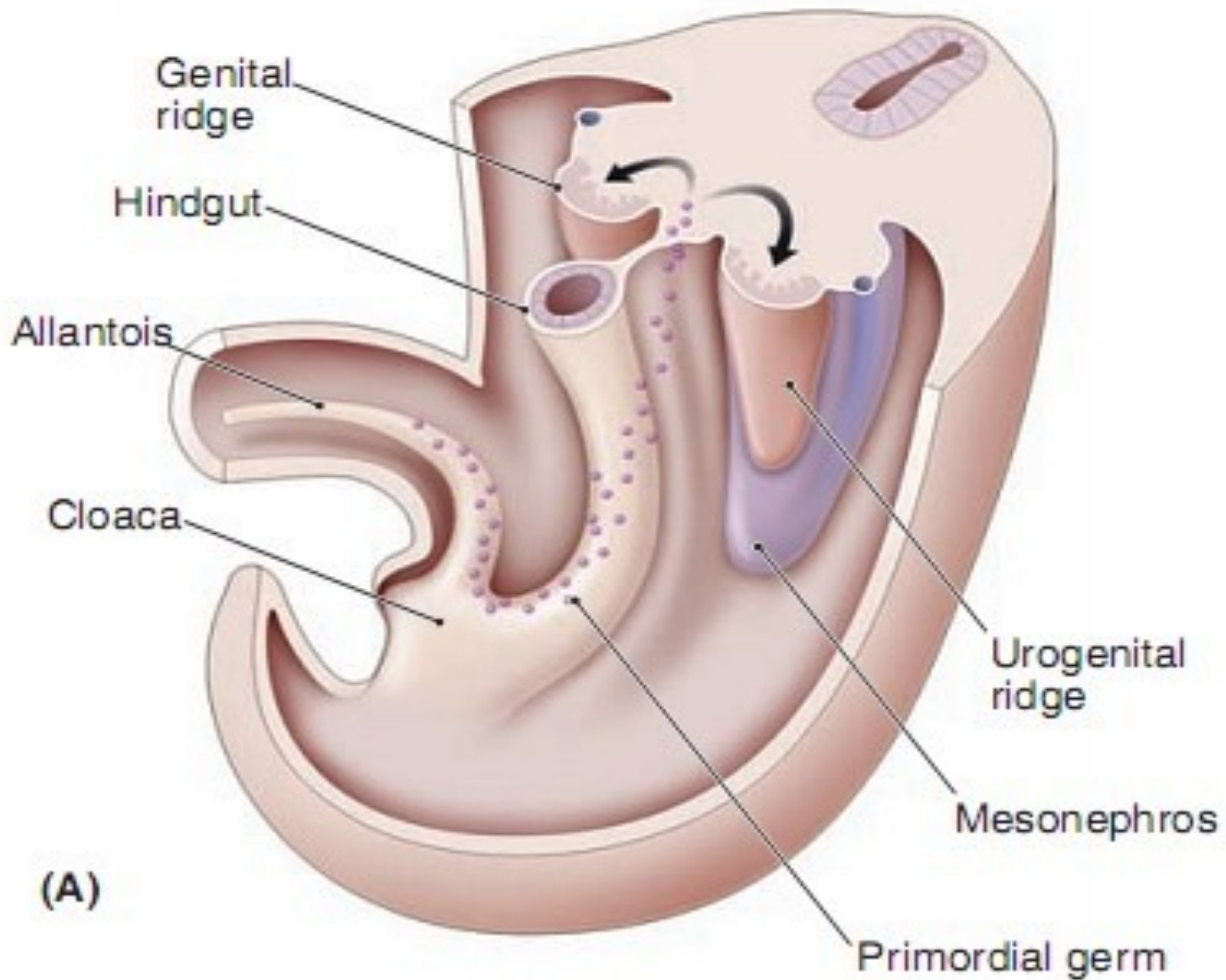
-1. Diagram indicating the differentiation of the gonad in the female. (A) Gonad in the

# Formation and Differentiation of Gonads

Primitive germ cells migrate from yolk sac through dorsal mesentery to reach genital ridge

These germ cells stimulate coelomic epithelium and underlying mesoderm to proliferate and form primitive sex cords

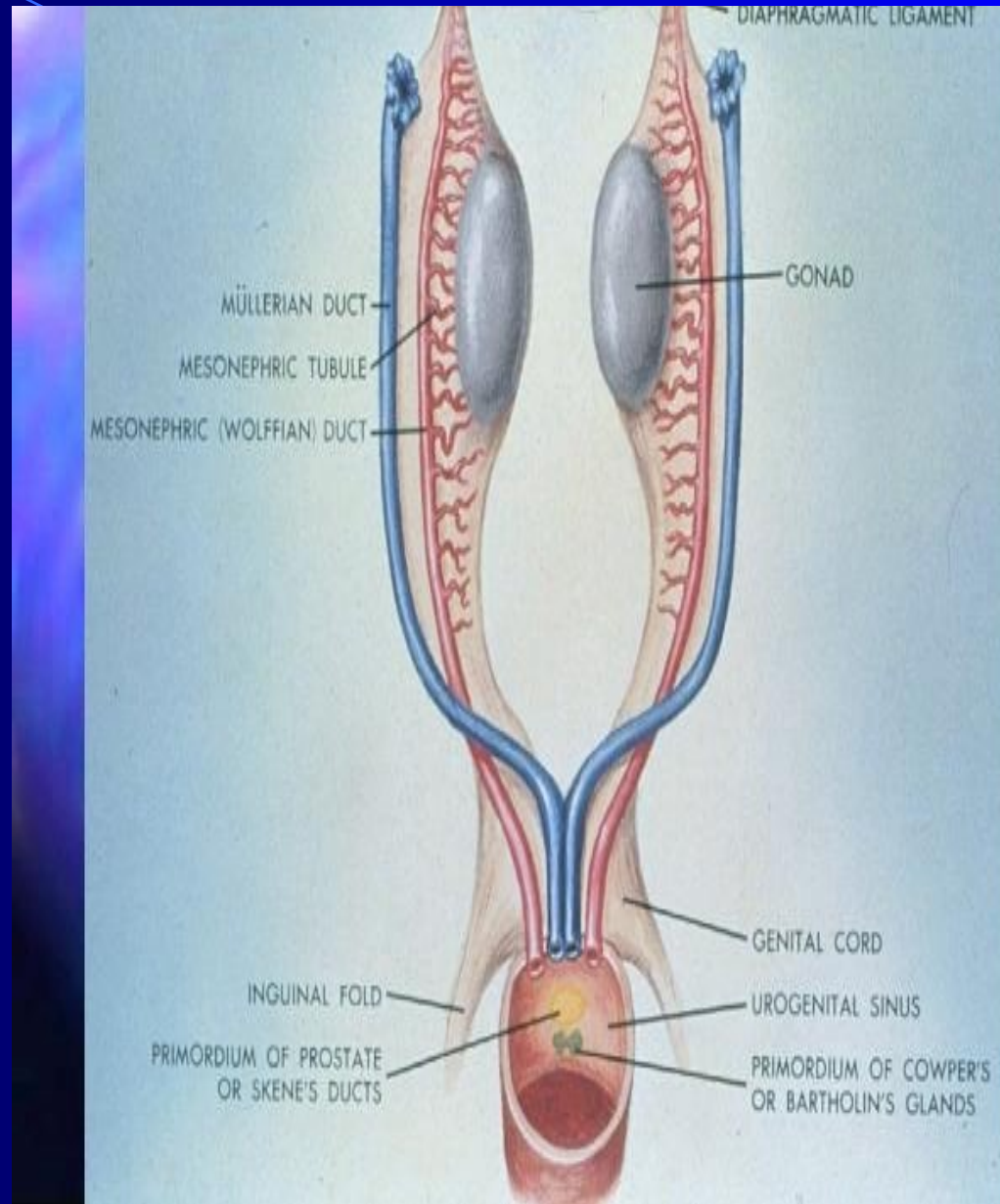




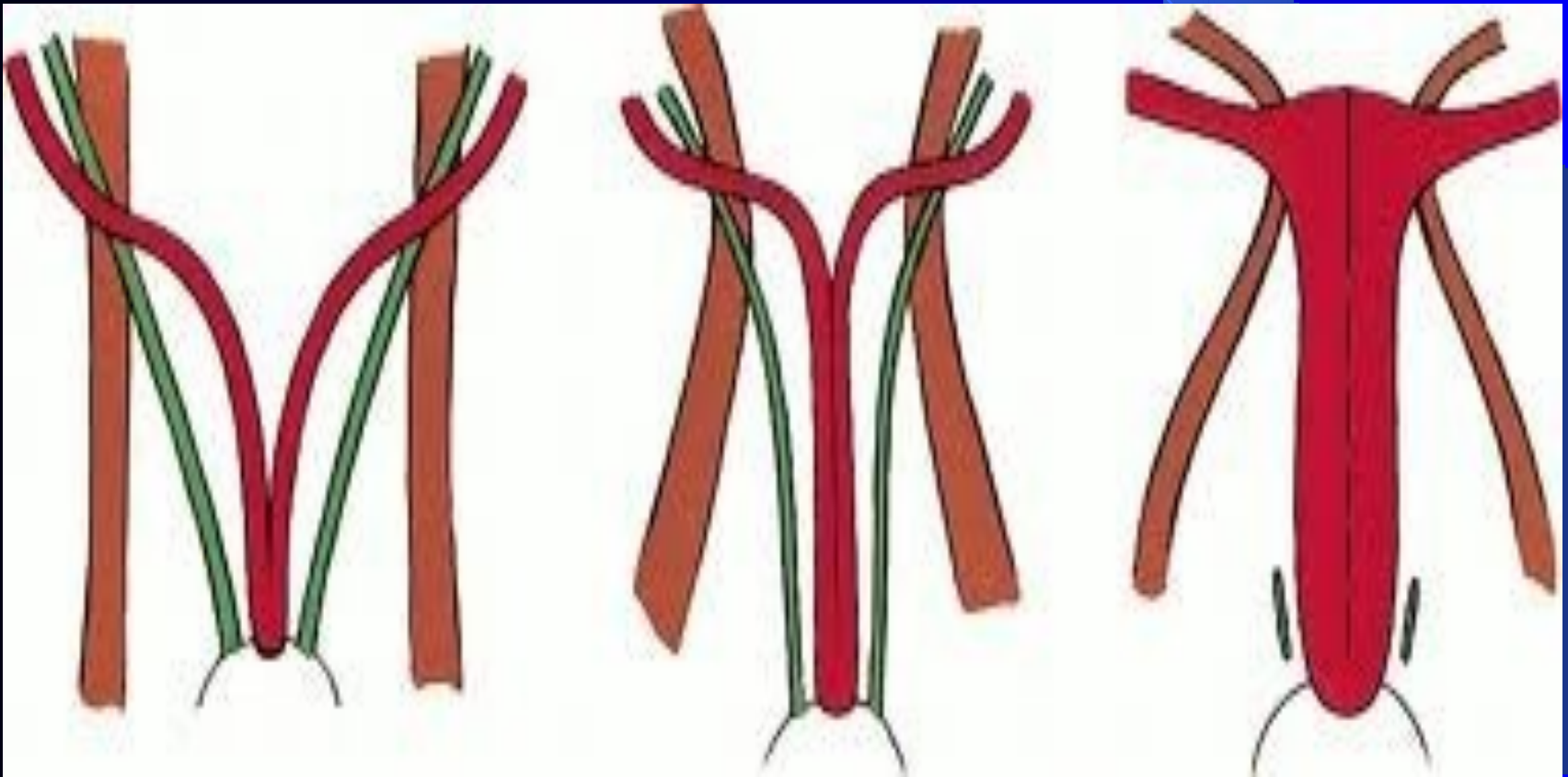


At 6<sup>TH</sup> week  
gestation

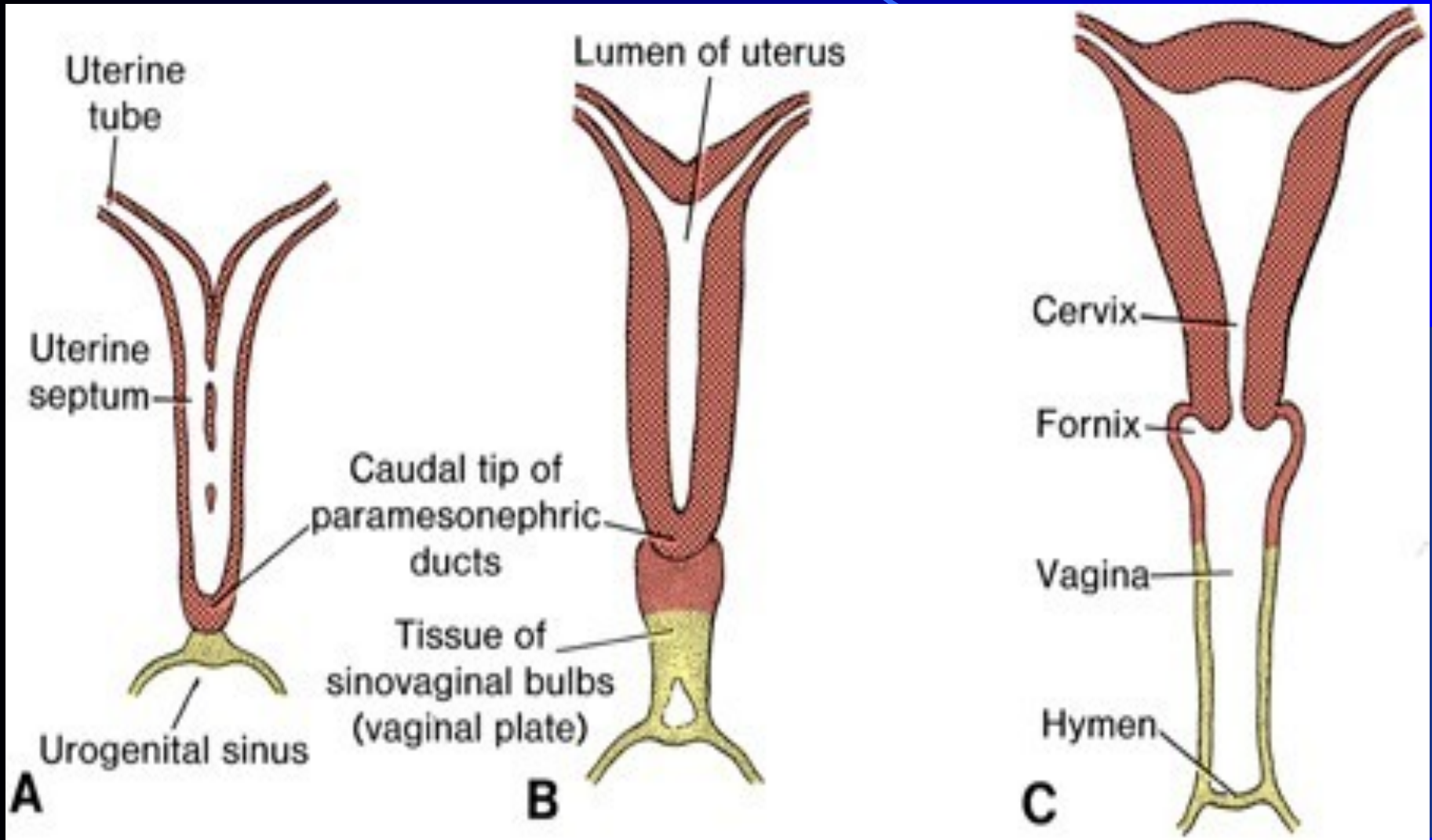
Paramesonephric  
or Mullerian Duct  
develops lateral to  
the Mesonephric  
"wolffian" Duct



<sup>λ</sup>The middle and caudal parts of the Mullerian ducts undergoes medial migration and fusion.



# Development of the vagina



## 2 main Principle

- λ Internal genital organs develop in close association with urinary tract **So** gross malformation of uterus and tube are commensally associated with anomalies of kidney and ureter.
- λ 2-Development of gonads is separate from that of the ducts **So** functional ovary are usually present when uterus, vagina are absent

# Female Genital Duct Formation

- λ In ovary the absence of testosterone inhibits the development of the mesonephric ducts.
- λ
- λ The atretic remains form the epoophoron, paraoophoron and Gartner's ducts.
- λ In absence of AMH, paramesonephric ducts form the female internal genital tract.

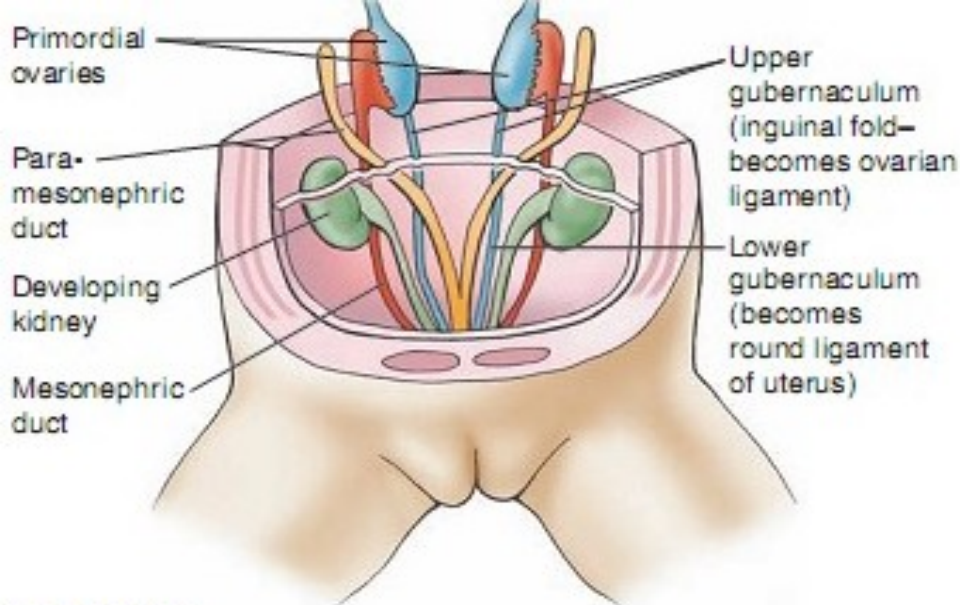
# Descend of the Ovary

The ovary descends from its position high up the posterior abdominal wall to the edge of pelvic brim by the mesonephric ligament, which becomes differentiated into:

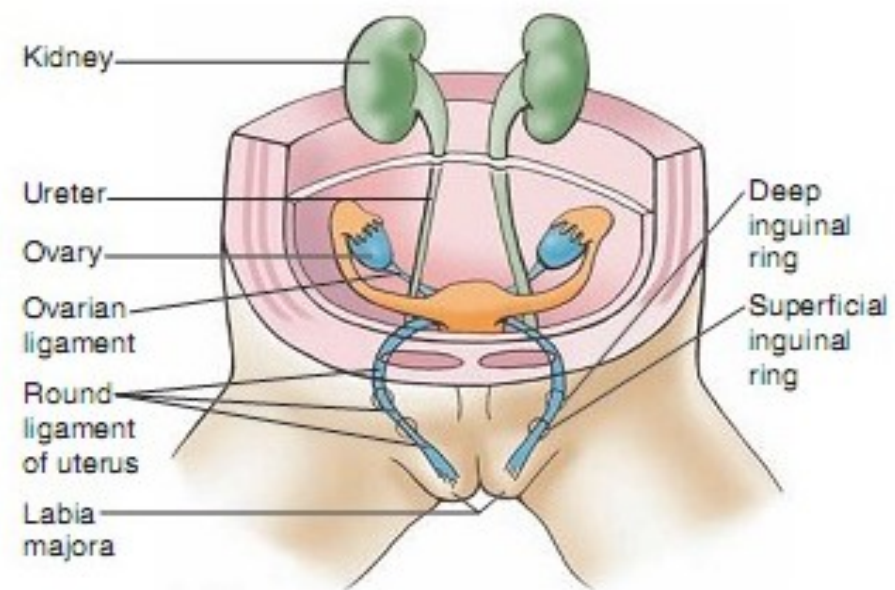
**Cranial Part** → Suspensory ligament of the Ovary

**Caudal Part:**

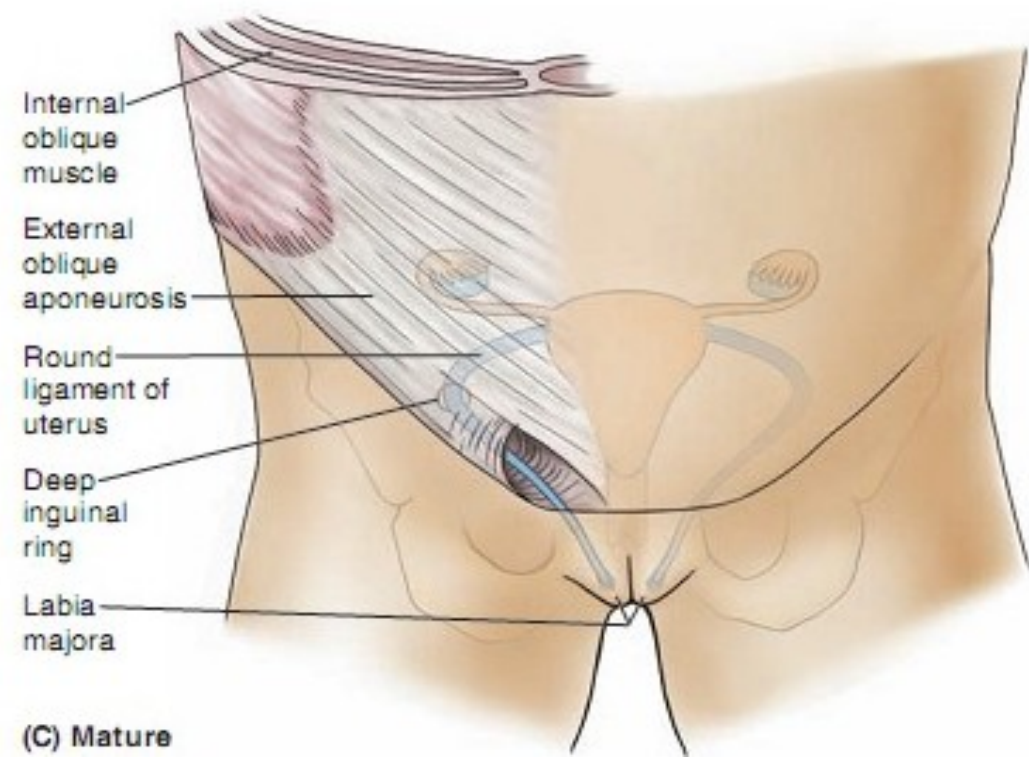
1. **Upper part** → Ovarian ligament
2. **Lower part** → Round ligament



(A) 2 months



(B) 15 weeks



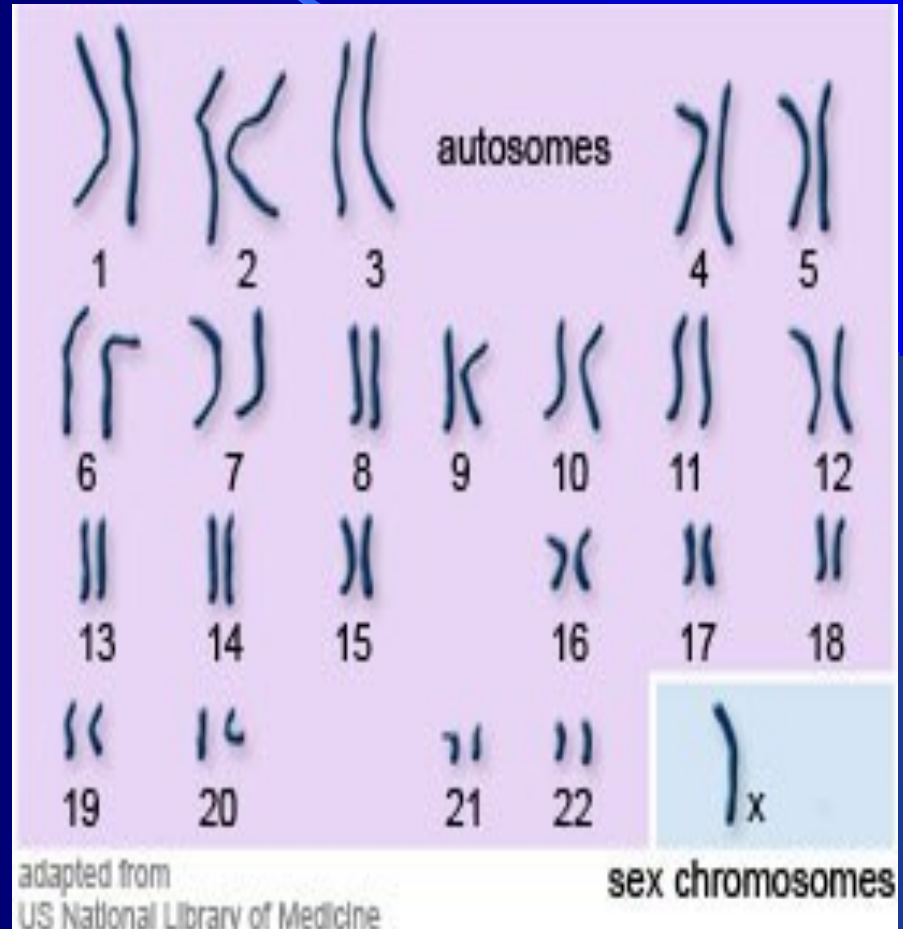
(C) Mature

# Abnormalities of the ovaries:





- λ 1) **agenesis** or complete absence.
- λ 2) **Gonadal dysgenesis** "streak gonads" as in Turner syndrome.
- λ 3) **Failure of descent** into the pelvis.
- λ 4) **Ovotestis** "true hermaphrodite"  
In which combined ovarian and testicular tissues seen.



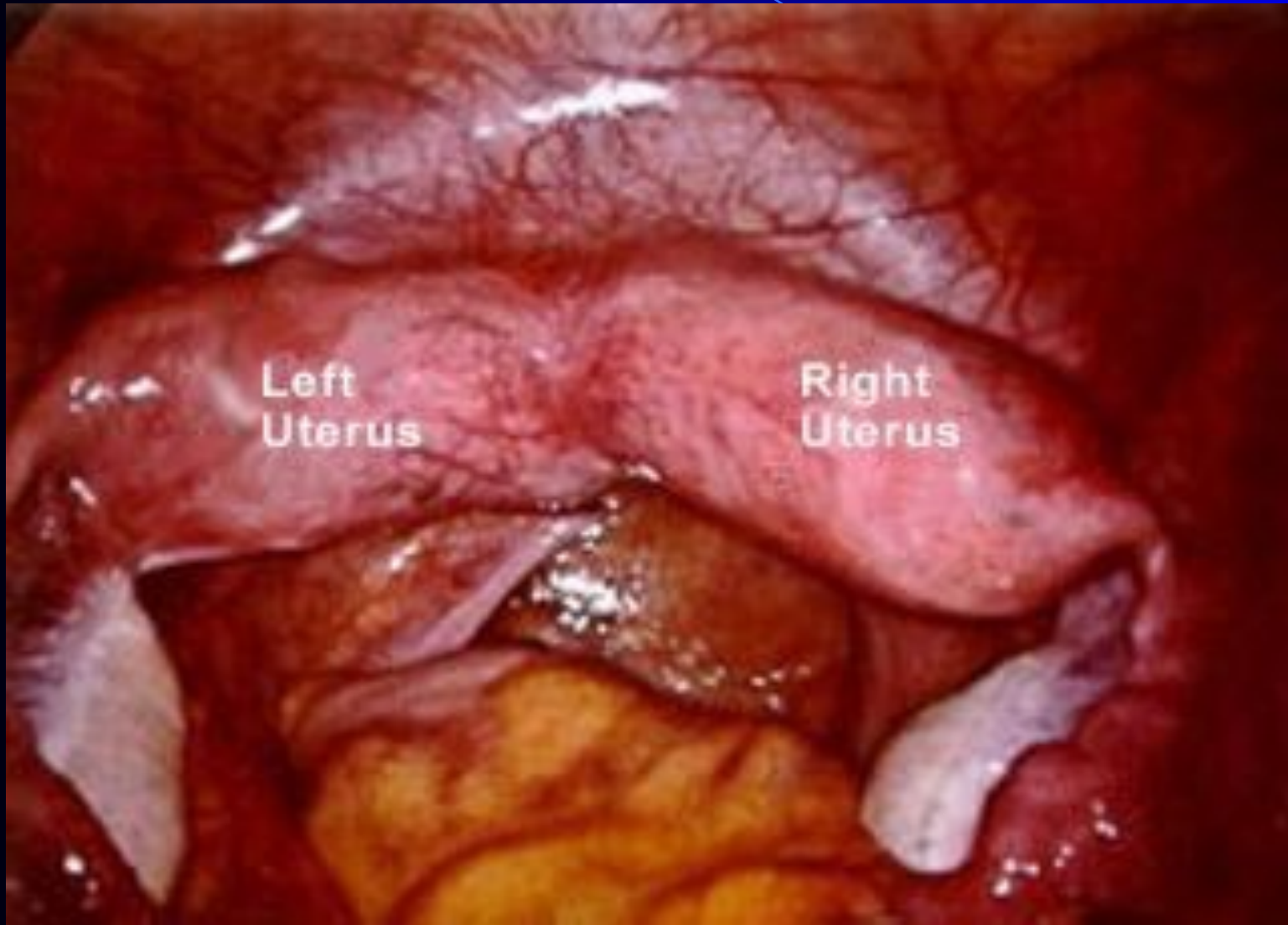
# Turner syndrome" ovarian "dysgenesis



# Development external genitalia

- λ Early, similar in both sexes
- λ 6<sup>th</sup> wk, three external protuberance surround cloacal membrane, the left and right genital swellings meet anteriorly to form the genital tubercle.
- λ 12<sup>th</sup> wk identify difference.  
- λ Genital swelling labioscrotal folds  
scrotum or labia major
- λ Genital tubercle phallus  penis or clitoris 

# Mullerian Anomalies



# Why is this important?

λ Majority have no problem conceiving, but have higher rates of:

- 1. Spontaneous Abortion
- 2. Premature Delivery
- 3. Infertility
- 4. Abnormal Fetal Lie
- 5. Dystocia at delivery
- 6. Dysmenorrhea, endometriosis
- 7. Cervical incompetence

# Uterine Anomalies

```
graph TD; A[Uterine Anomalies] --> B[Absence of Uterus]; A --> C[Fusion anomalies];
```

Absence of Uterus

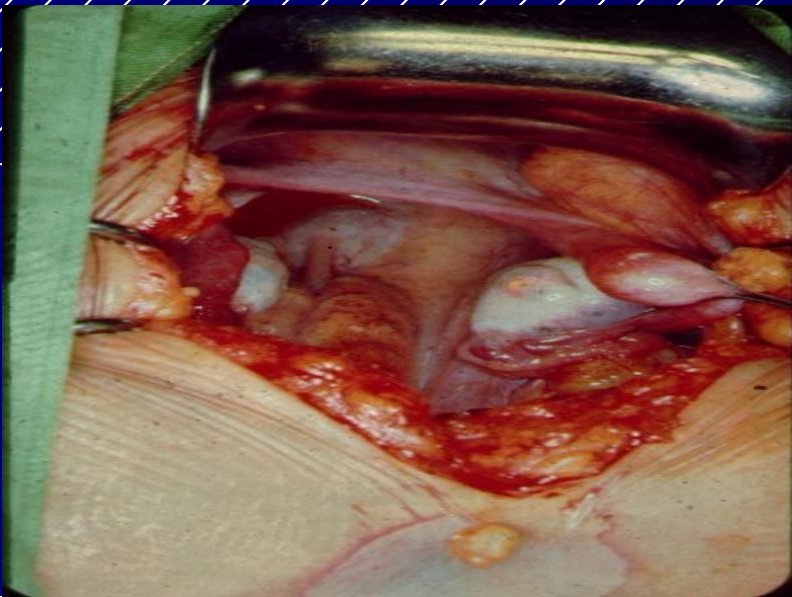
Fusion anomalies

# Classification into 4

## λ groups:

1. Aggenesis of uterus/vagina: Rokitansky-Kuster-Hauser Syndrome.
- λ 2-Unilateral development : Unicornate uterus
- λ 2. Defects in Vertical Fusion (obstructive or non-obstructive)
- λ 3. Lateral Fusion defects (obstructive or non-obstructive).

# Mayer-Rokitansky-Kuster-Hauser Syndrome (utero-vaginal agenesis)



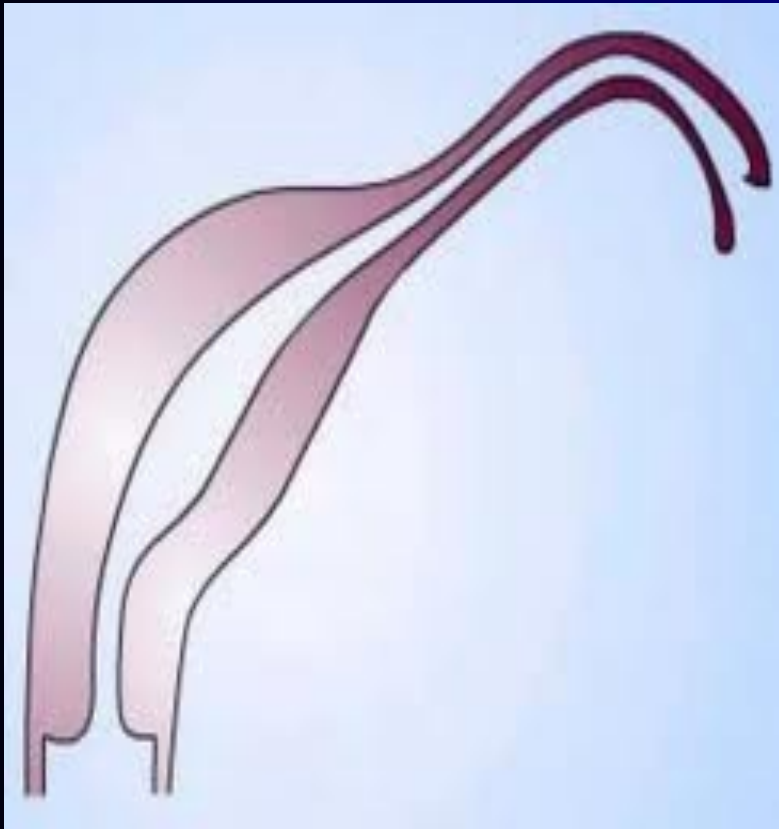
- 15% of primary amenorrhoea
- Normal secondary development & external female genitalia
- Normal female range testosterone level
- Absent uterus and upper vagina & normal ovaries
- Karyotype 46-XX
- 15-30% renal, skeletal and middle ear anomalies

# RKH Syndrome: Diagnosis

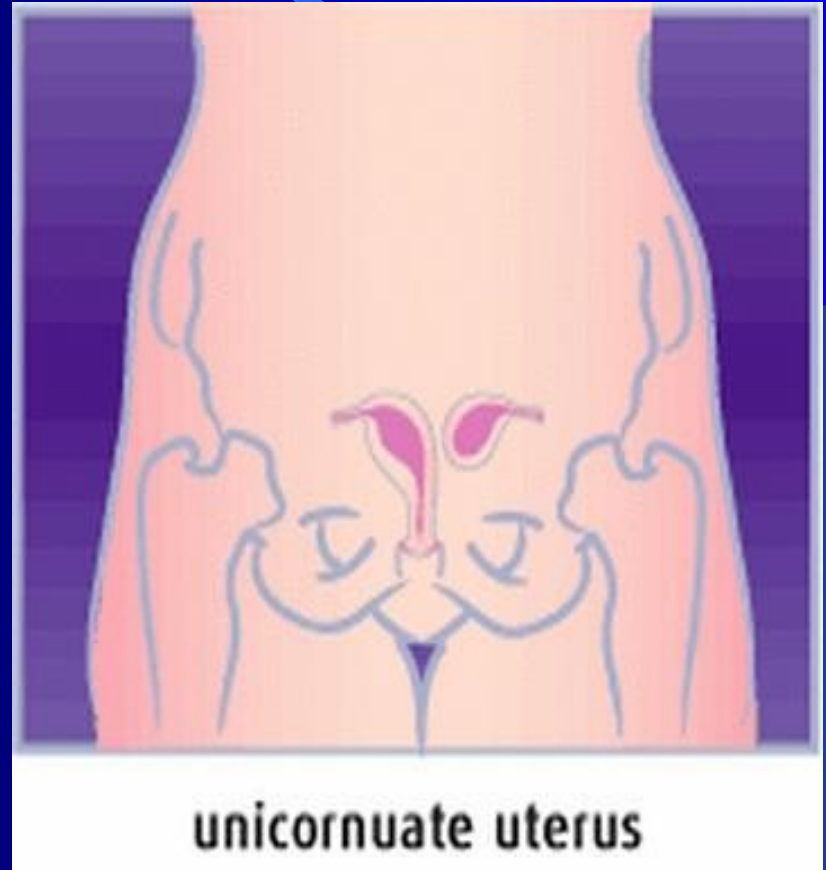
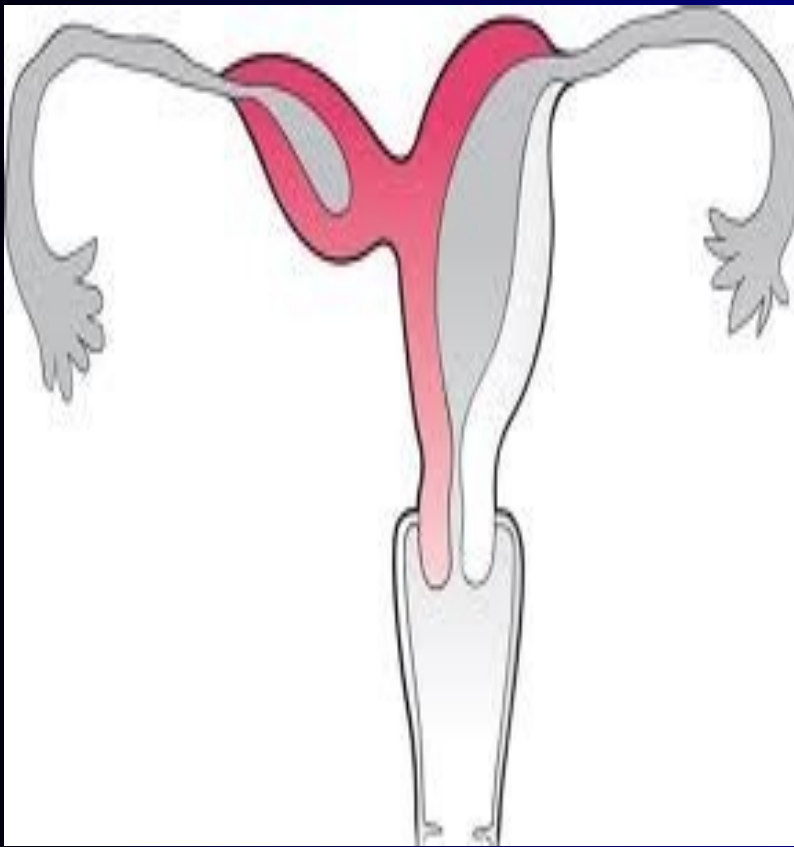
- λ Expected Menarche
- λ Difficult to differentiate from imperforate hymen
- λ No uterus on exam, U/S, MRI, Laparoscopy, IVP
- λ Confused with Androgen Resistance Syndrome with shallow pouch and no uterus.
- λ Determine karyotype.



# Unicornuate Uterus

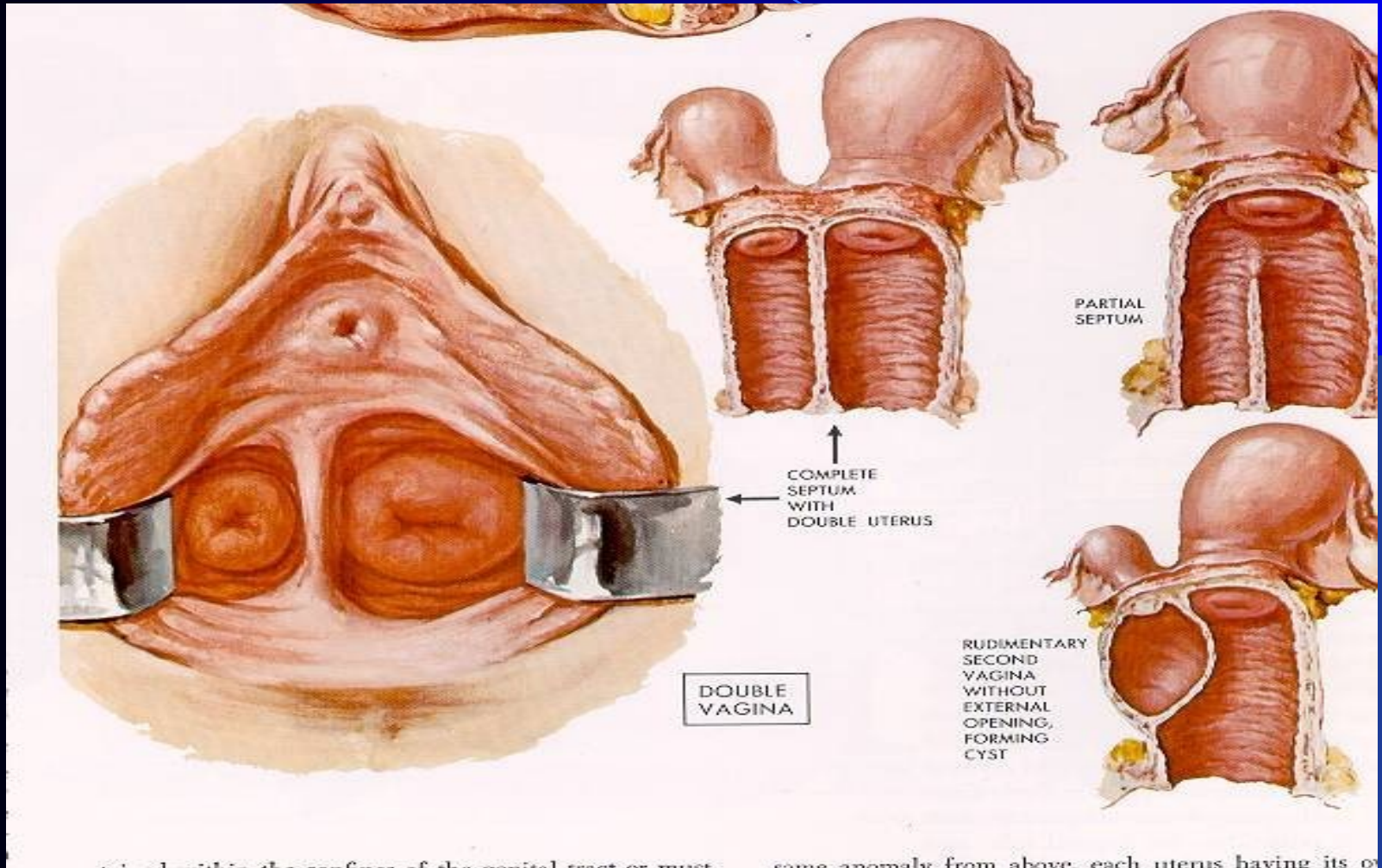


# Unicornuate Uterus



# Lateral Fusion Defects:

Most common type of mullrian defects



... and ... to the surface of the genital tract or must ... some anomaly from above, each uterus having its o...

# Lateral Fusion Defects

λ

•

λ

Result from failure of fusion of the  
mullerian ducts

λ

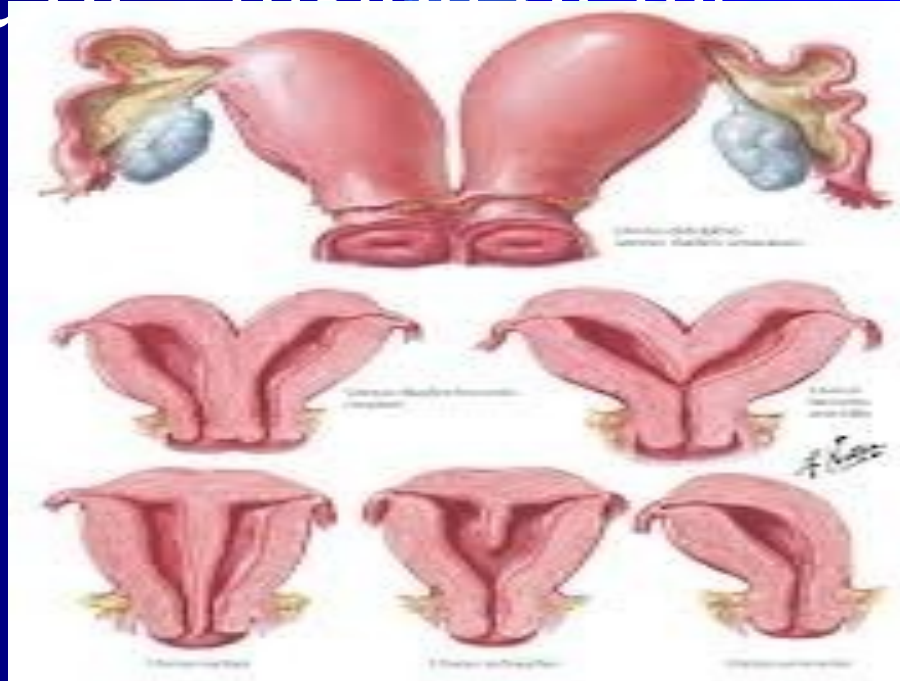
, or failure of

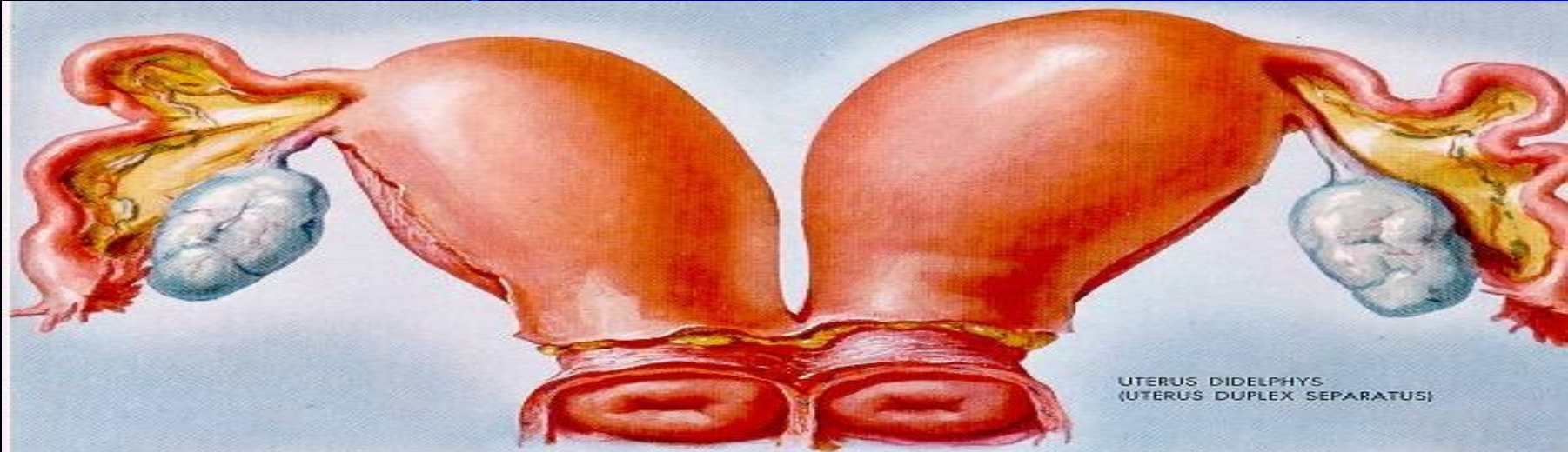
λ

absorption

λ

of the septum.





UTERUS DIDELPHYS  
(UTERUS DUPLEX SEPARATUS)



UTERUS DUPLEX BICORNIS  
(SEPTUS)



UTERUS  
BICORNIS  
UNICOLLIS

*F. Netter*  
© C



UTERUS SEPTUS



UTERUS SUBSEPTUS



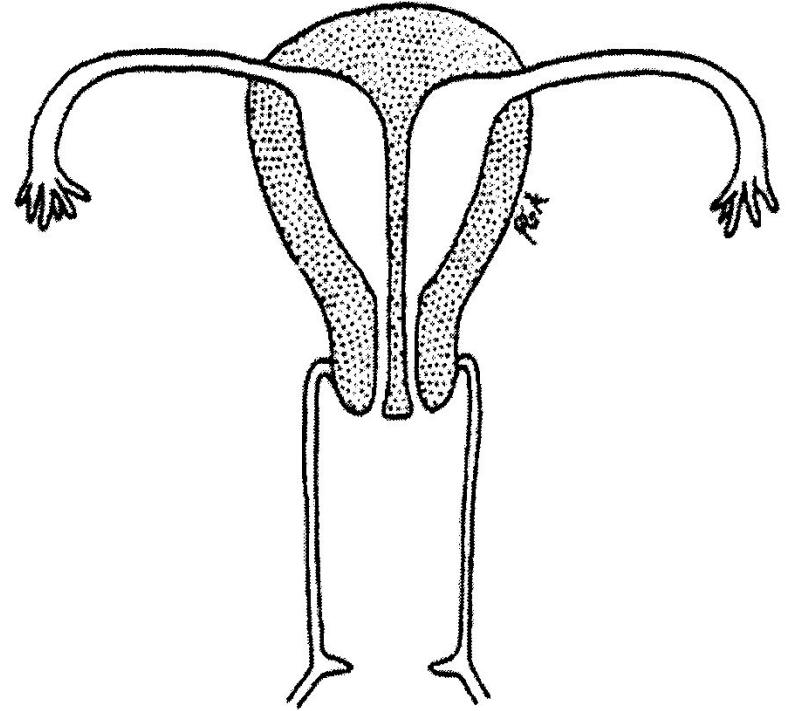
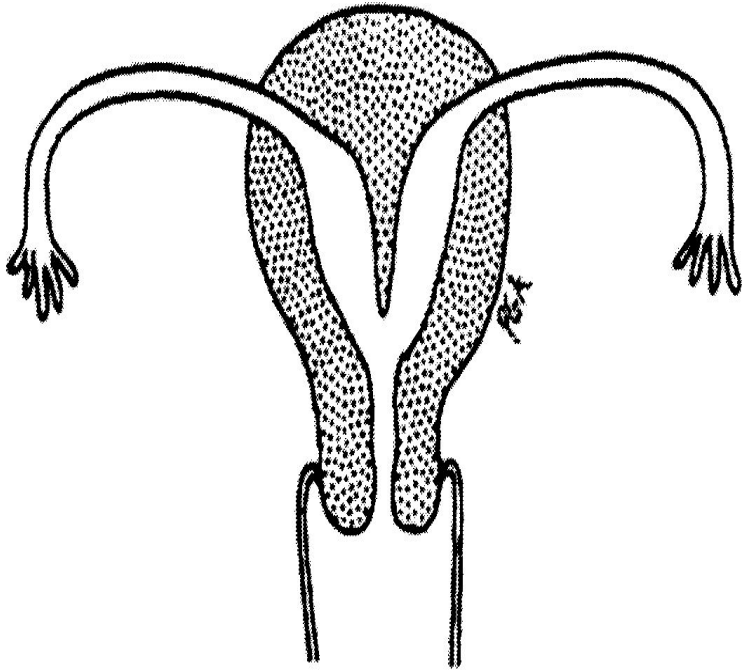
UTERUS UNICORNIS



# Septate Uterus

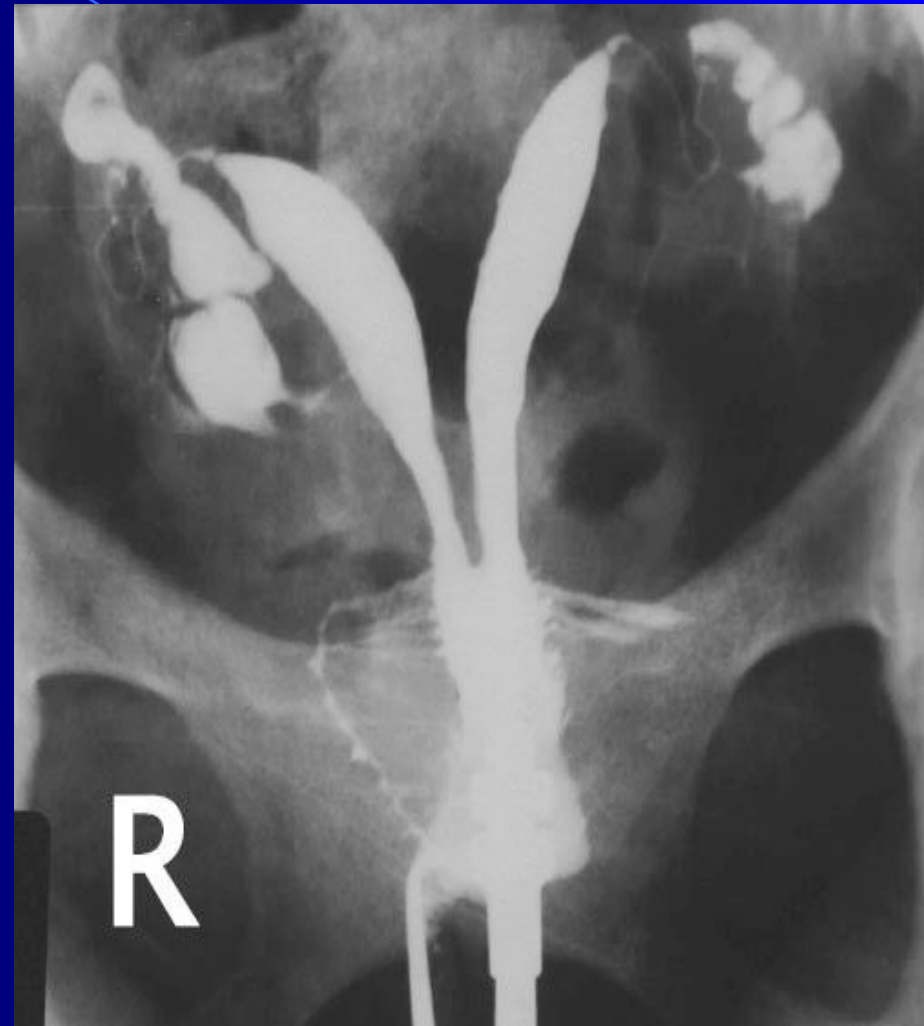
λ Defective resorption of the septum between the fused müllerian ducts results in a uterine septum, which may extend either partially down the uterus or the full length to the cervix. Normal external surface

# Septate Uterus: Partial and Complete

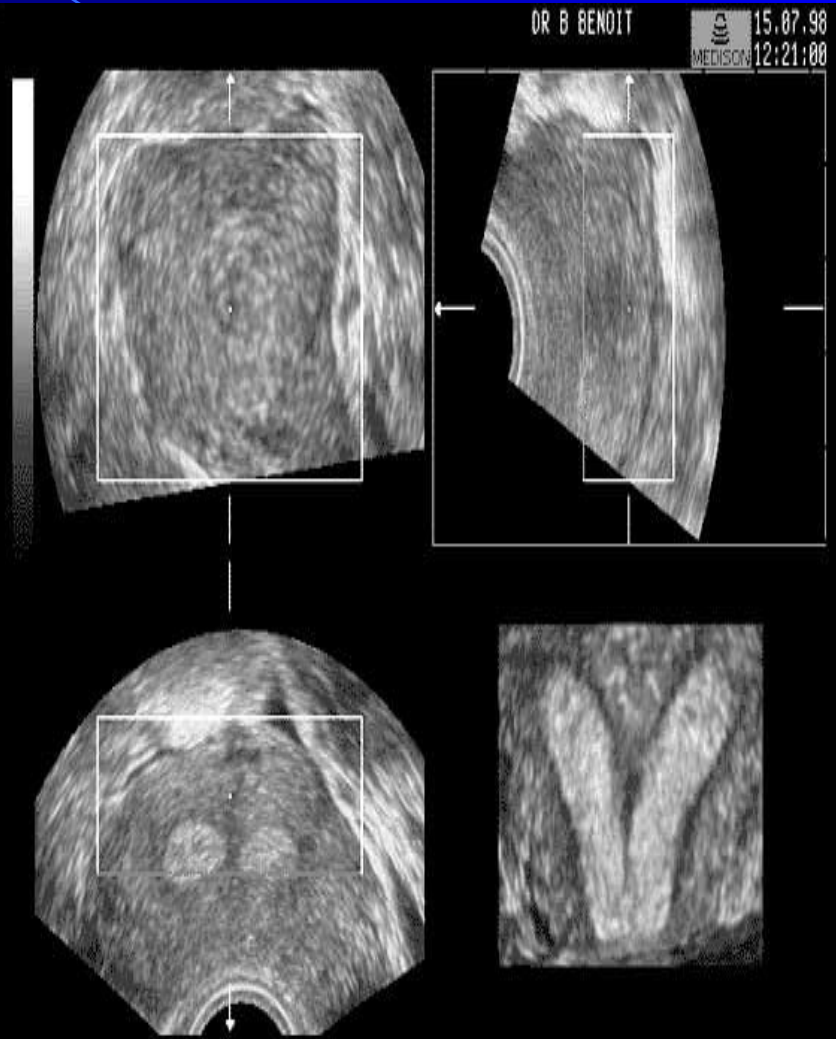
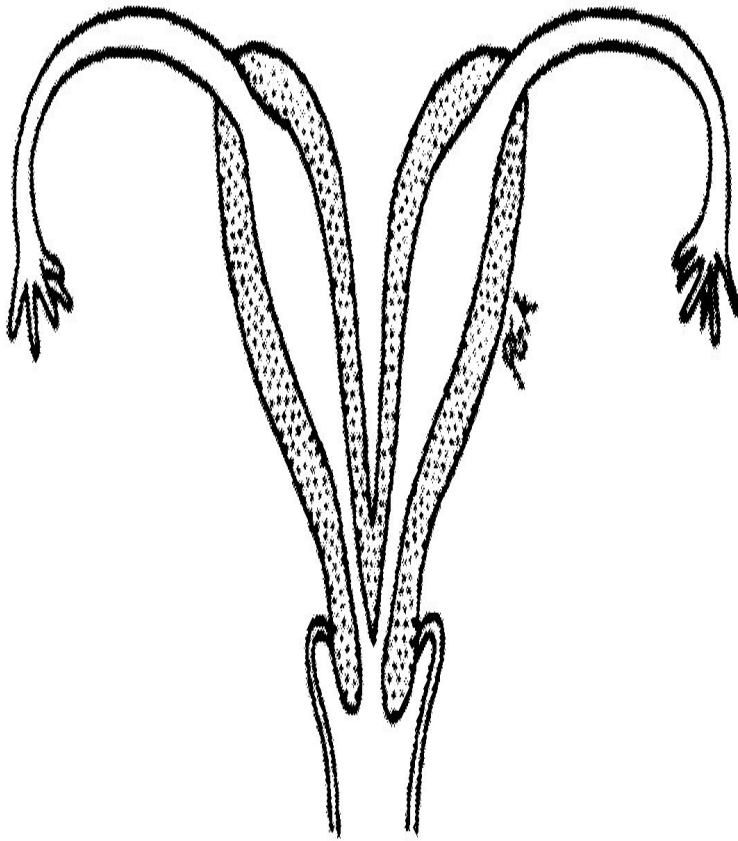




# Septate Uteri



# Bicornuate Uterus



# Bicornuate Uterus

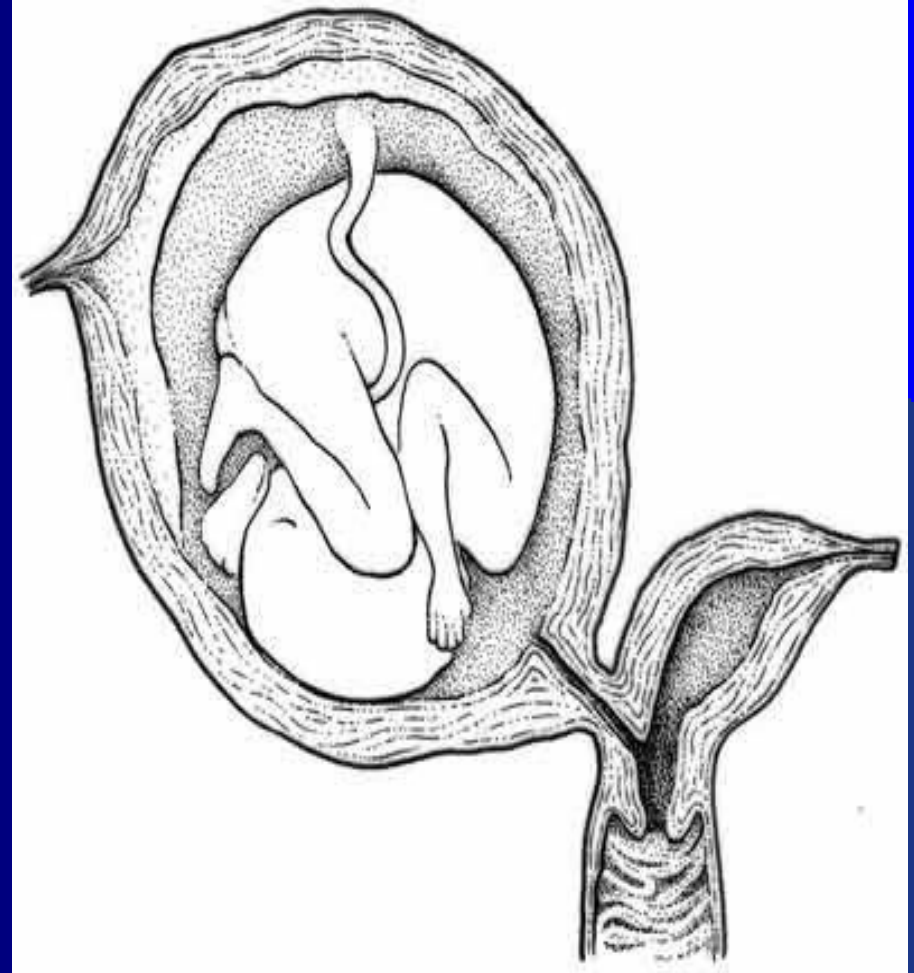
– Fundus

– Inverted fusion of müllerian ducts

– Variable degree of separation of horns that can be complete, partial or minimal

– HSG not diagnostic , need laparoscopy

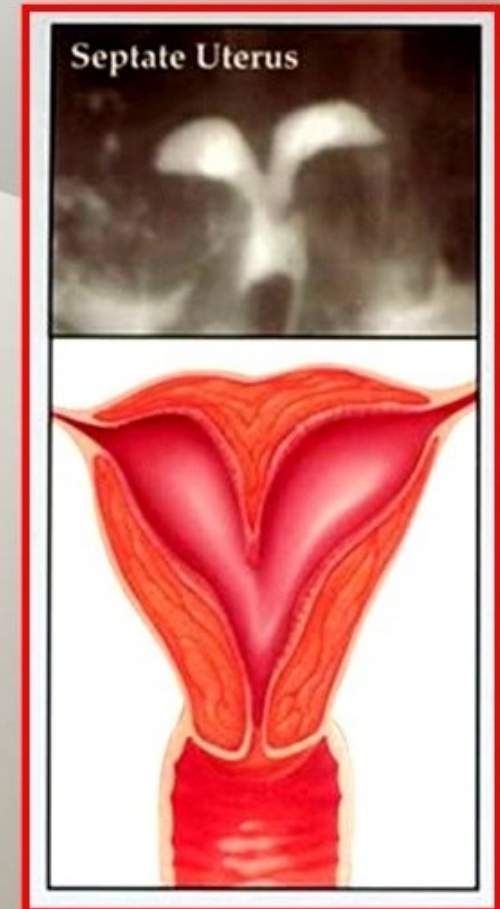
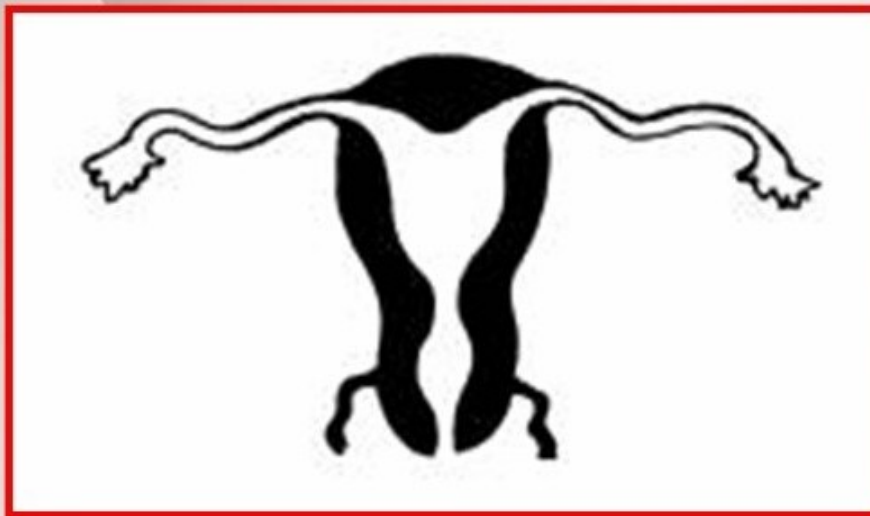
# Bicornuate uterus with unilateral pregnancy



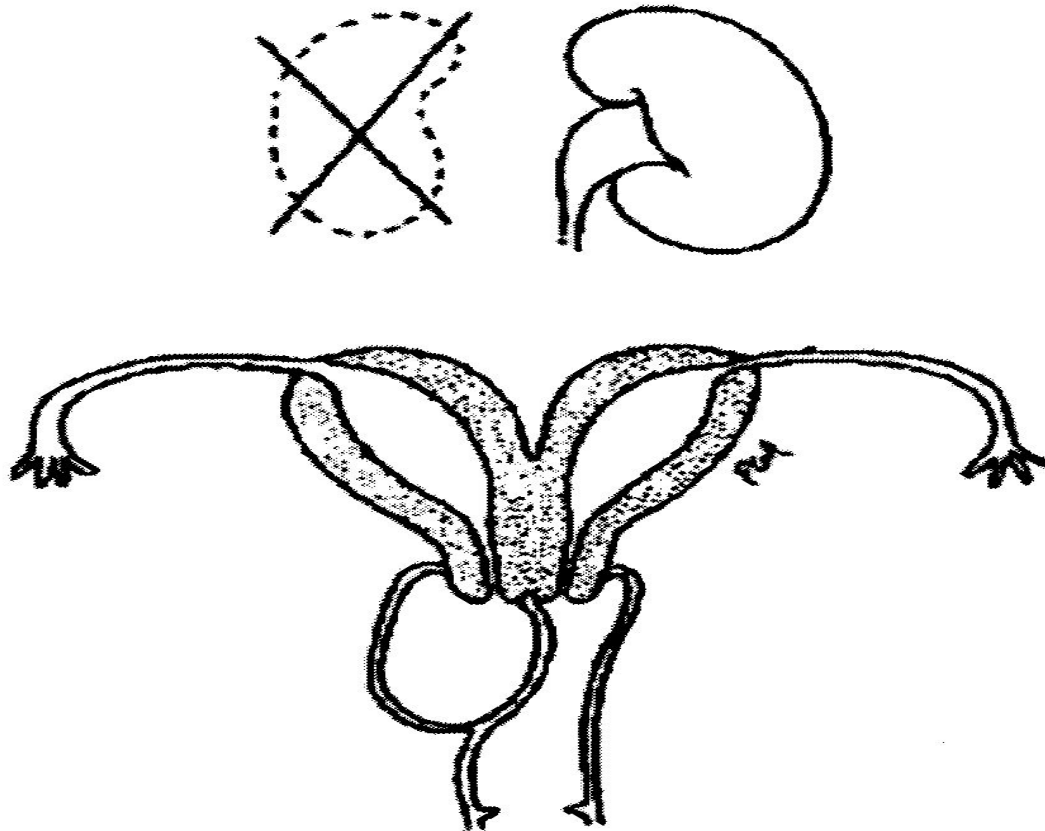
- **Arcuate uterus (Codiformis):**

A depression at the fundus (planeform uterus).

- **Septate or subseptate uterus.**



# Obstructive Defect of Lateral Fusion:



# Vertical Fusion obstructive and non-obstructive Defects

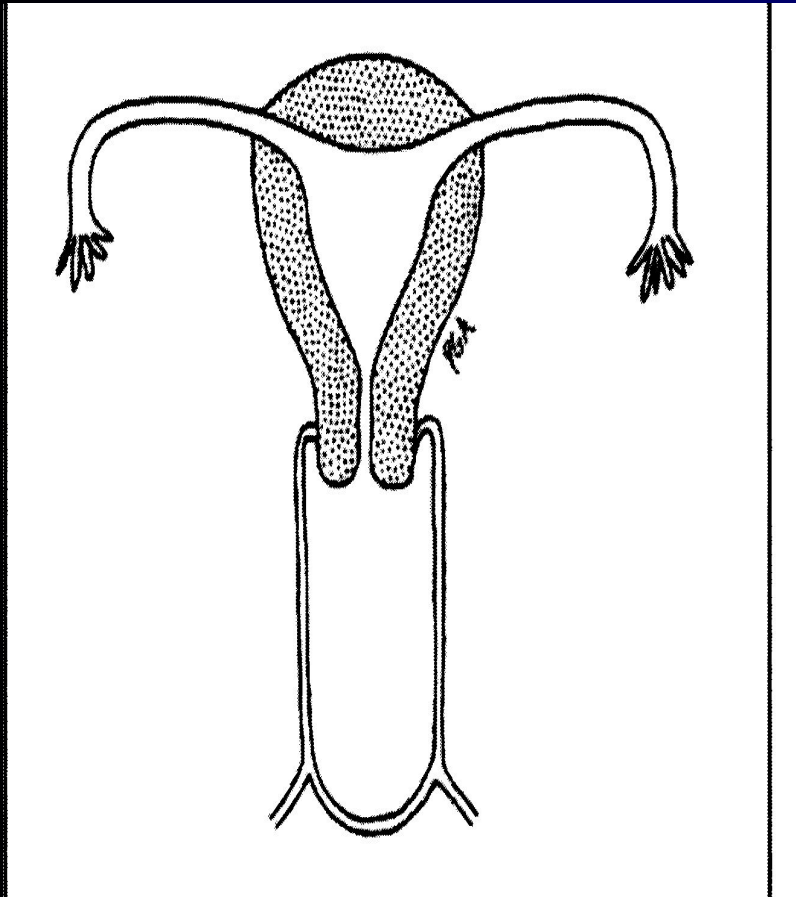
- λ . Can be considered in two categories:
  - λ 1. Imperforate Hymen
  - λ 2. Transverse Vaginal Septum

# Imperforate Hymen



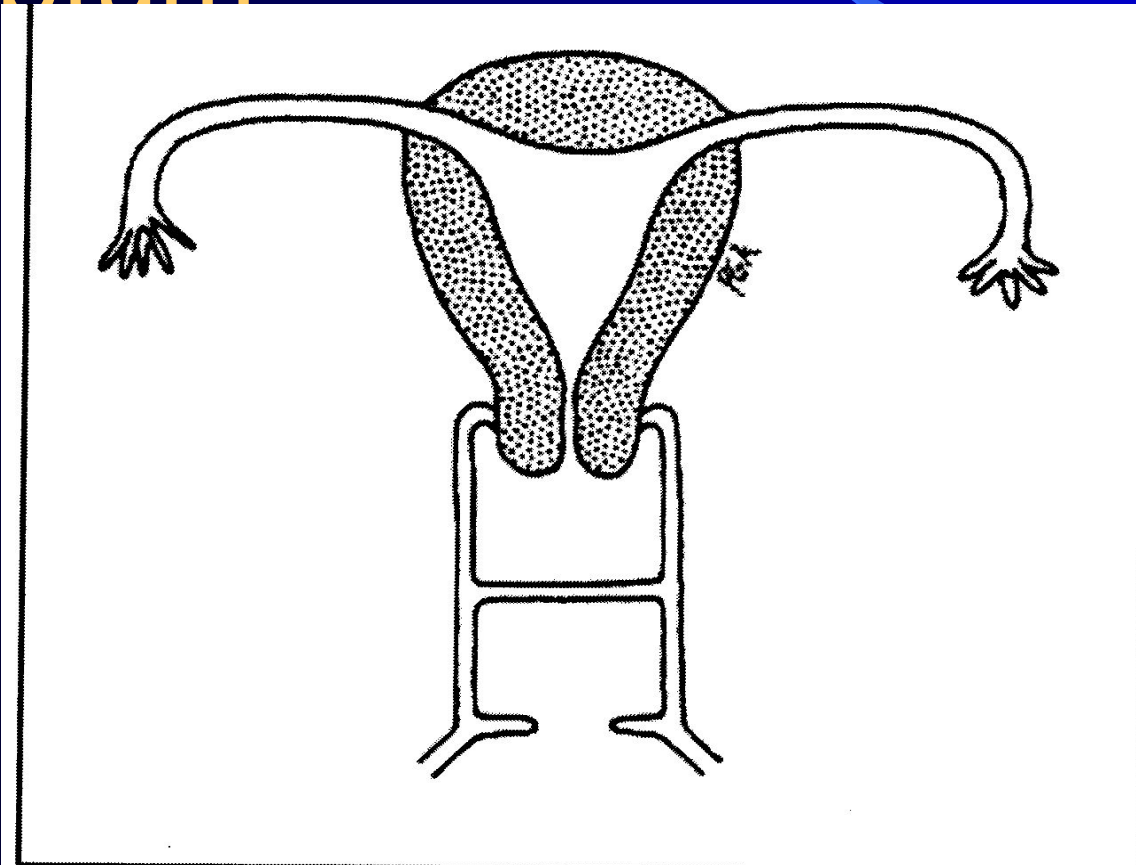


# Imperforate Hymen: Diagnosis/ Treatment

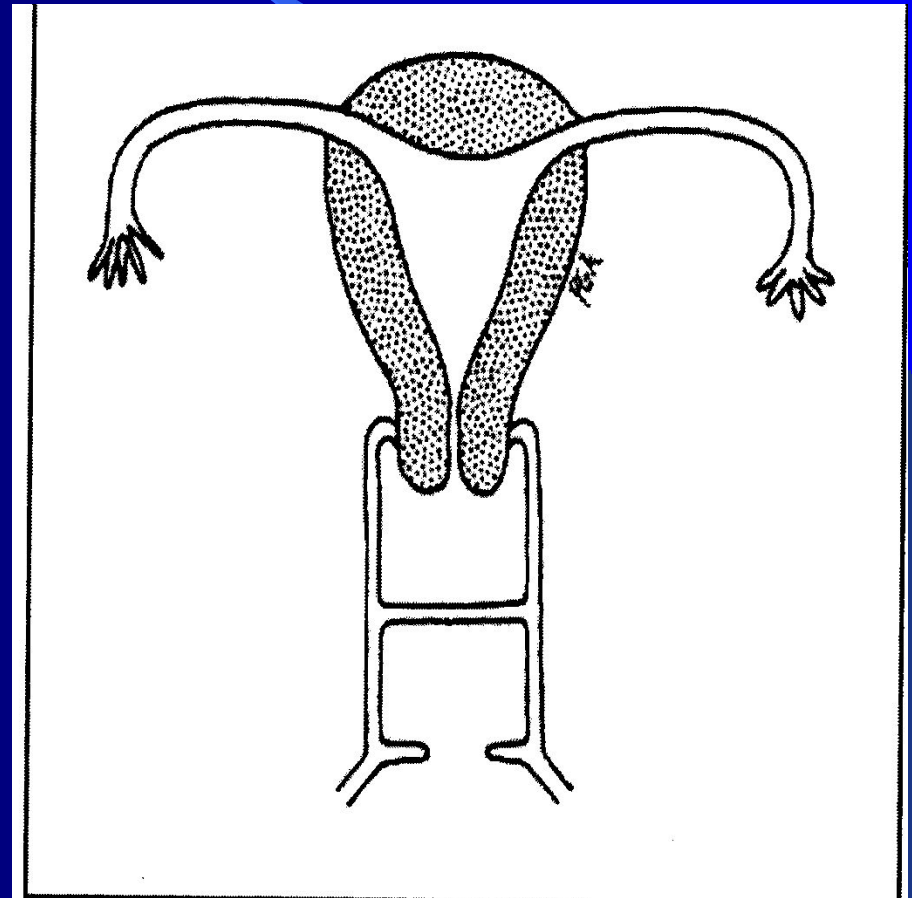
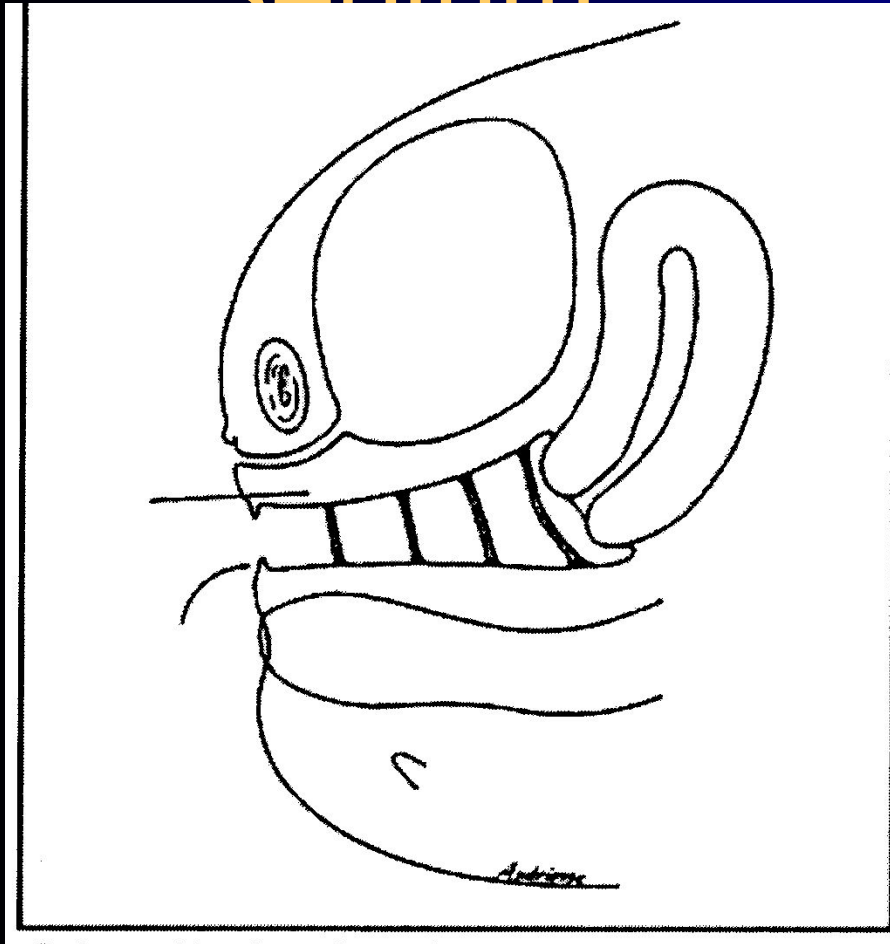


- λ Cyclic pelvic pain due to hematocolpos, hematometria, or hematosalpinx
- λ Bulging hymeneal membrane or a blind-ending pouch on exam.
- λ Pelvic/Rectal exam, U/S, MRI
- λ Rarely urologic anomalies.
- λ Tx: Cruciate incision

# Transverse Vaginal Septum



# Transverse Vaginal Septum



# Transverse Vaginal Septum: Presentation/Diagnosis

- λ Cyclical pain due to hematocolpos or hematometria.
- λ Blind-ending pouch.
- λ No bulging at outlet, hydromucocolpos or hematocolpos, rectal exam or U/S, MRI.
- λ Thickness varies and site varies in vaginal canal.

# Summary

- λ Understanding the embryologic origin of the defect of mullerian anomalies is key to its correct diagnosis
- λ Presentation: Obstetrical problems, dysmenorrhea, amenorrhea
- λ Diagnosis : Pelvic/Rectal exam, U/S, HSG, Laparoscopy, Hysteroscopy, MRI

# Questions & Answers

- **Where are Gartner ducts located?**
- **Gartner duct cysts are persistent portions of what embryonic structure?**
- **The portion of the gubernaculum between the ovary and uterus becomes what structure?**
- **The portion of the gubernaculum between the uterus and the labium majus becomes what structure?**

○ **Where are Gartner ducts located?**

*In the lateral walls of the vagina.*

○ **Gartner duct cysts are persistent portions of what embryonic structure?**

*Mesonephric duct.*

○ **The portion of the gubernaculum between the ovary and uterus becomes what structure?**

*The ligament of the ovary (utero-ovarian ligament).*

○ **The portion of the gubernaculum between the uterus and the labium majus becomes what**

**structure?**



**○ Failure of the development of adhesions between the uterus and what structure can result in the ovary migrating through the inguinal canal to the labium majus?**

**○ What is the name of a pouch of peritoneum analogous to the saccus vaginalis in the male, which accompanies the gubernaculum in the inguinal canal?**

- **Name the three coats of the ureter.**
- **The epithelium lining the ureter is of what type?**

**○ Failure of the development of adhesions between the uterus and what structure can result in the ovary migrating through the inguinal canal to the labium majus?**

**The gubernaculum.**

**○ What is the name of a pouch of peritoneum analogous to the saccus vaginalis in the male, which accompanies the gubernaculum in the inguinal canal?**

○ **Name the three coats of the ureter.**

**Fibrous, muscular, mucosal.**

○ **The epithelium lining the ureter is of what type?**

**Transitional.**



THANK YOU