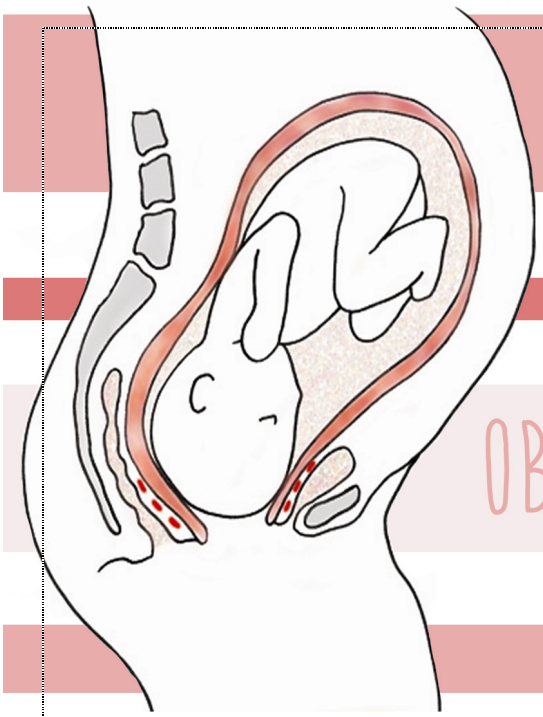

OBSTETRICS AND GYNECOLOGY

# (2) Embryology of female genital tract, congenital malformation and intersex


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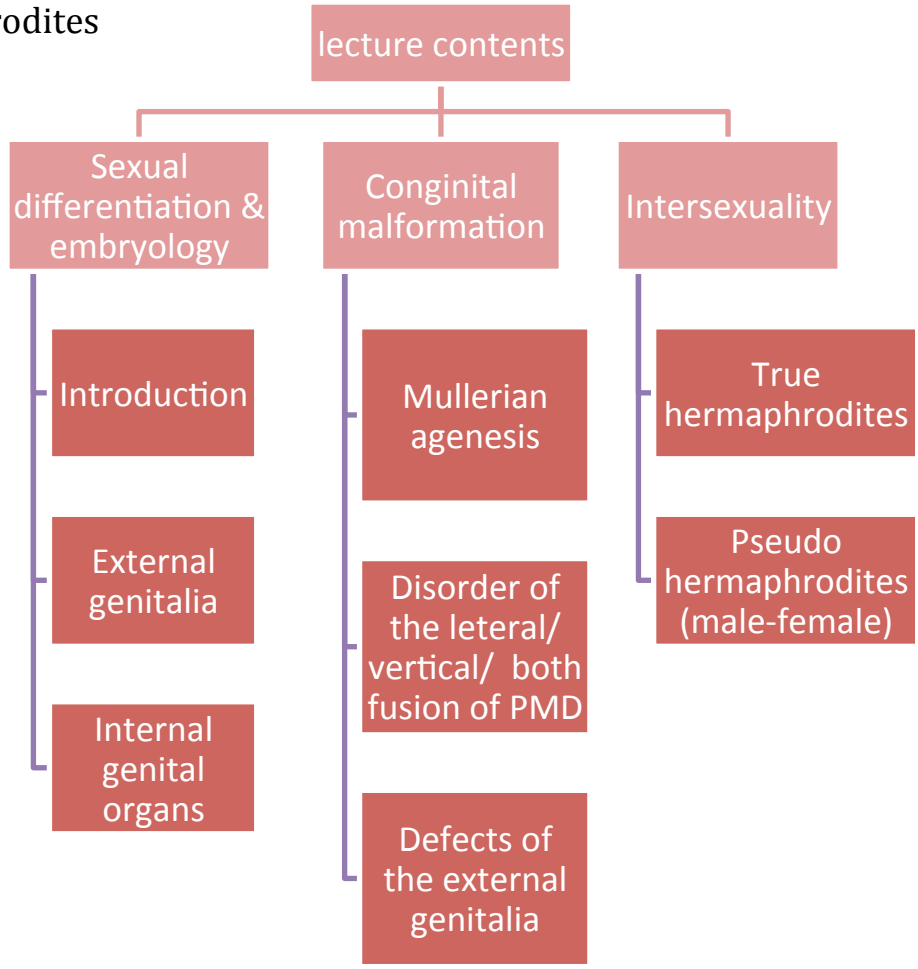
Doctor's note  Team's note  Not important  Important  431 teamwork

# Objectives:

1. Steps of the male and female sexual differentiation.
2. Development of the female external genitalia
3. Development of the female internal genital organs
4. Understand the clinical presentations, complications and management of the following malformations:
   a. Mullerian agenesis
   b. Disorders of lateral fusion of the mull ducts
   c. Disorders of vertical fusion of the mullerian duct
   d. Unusual configuration of vertical/lateral fusion defects
   e. Defects of the external genitalia

   f. A-What are the problems that can result in abnormal sexual development?

5. Know the types of intersex, how to differentiate between these types? What is the presentation? How to manage these cases?
   a. Masculinized female (female pseudohermophrodites)
   b. Undermasculinized male (male pseudohermophrodites)
   c. True hermaphrodites



# 1 - Sexual differentiation

## A - Introduction

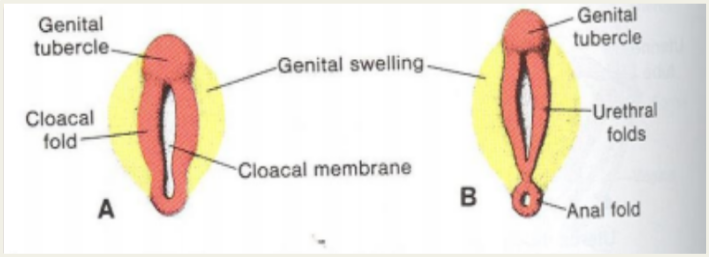
- The first step in sexual differentiation is the determination of genetic sex (XX or XY) at the time of fertilization.
- You should know that female reproductive system grow spontaneously without interference, but male reproductive system requires – primarily – the presence of Y chromosome.
- Gonads start differentiation to either ovaries or testes.
- In the absence of Y chromosome, they will develop into ovaries.
- In the presence of Y chromosome, testes determining factor (TDF-encoded by SRY gene on the Y chromosome) will influence the development of the testes.
- At this level, both males and females will have what's called paramesonephric duct (mullerian duct), which develops into uterus and fallopian tubes and the mesonephric duct (develop into the internal sexual parts of male reproductive system – this duct will regress spontaneously in the females)
- Development of internal and external sexual organs in female ♀ does **not** depend on the presence of ovaries (without hormones)
- In a male, testes produce:
  Testosterone (important for the development of the external genitalia) Mullerian inhibiting factor - MIF → important for paramesonephric duct regression (no MIF means that the male will have uterus and fallopian tubes despite the presence of testosterone) and continuation of the mesonephric duct.
- Male ♂ sexual development (internal and external) depends on the presence of functioning testes and responsive end organs.
- ♀ (46/XX) female exposed to androgens (androgenized) during development (in-utero), will be masculinized (she will have male external genitalia)

## B - External genitalia

### 1- **Undifferentiated stage** (4-8 weeks)

The neutral genitalia includes:

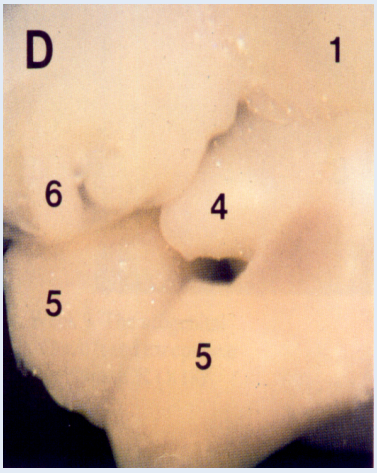
- genital tubercle (phalus)
- labioscrotal swellings
- urogenital folds
- urogenital sinus




### 2- **Male and female external genital development** (9-12 weeks)

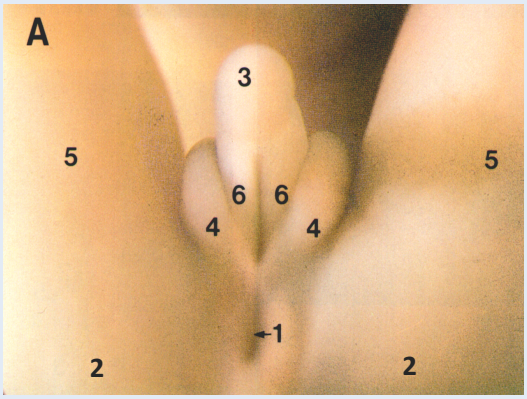
- By 12 weeks gestation, external genitalia can be differentiated
- The development of male genitalia requires the action of androgens, specifically DHT – dihydrotestosterone (testosterone should be converted into DHT by the action of 5 alpha reductase for the development of male external genitalia)

Indifferent stage



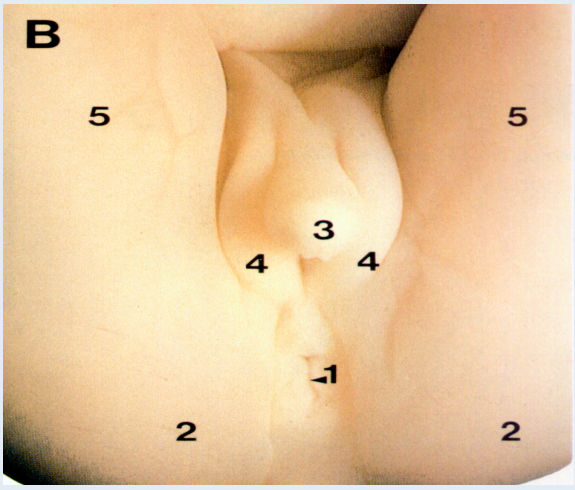

1- abdomen
4- genital tubercle
5- leg bud
6- midgut herniation to the umbilical cord

Week 9



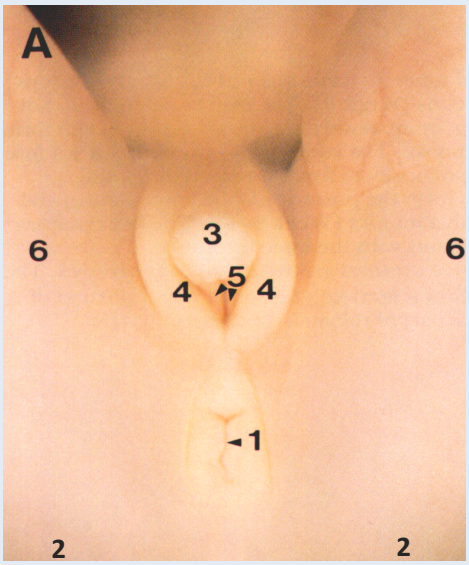

1- Anus
2- Buttocks
3- Clitoris
4- Labioscrotal swellings (labia majora)
5- Limbs
6- Urogenital folds (labia minora)

## Week 12



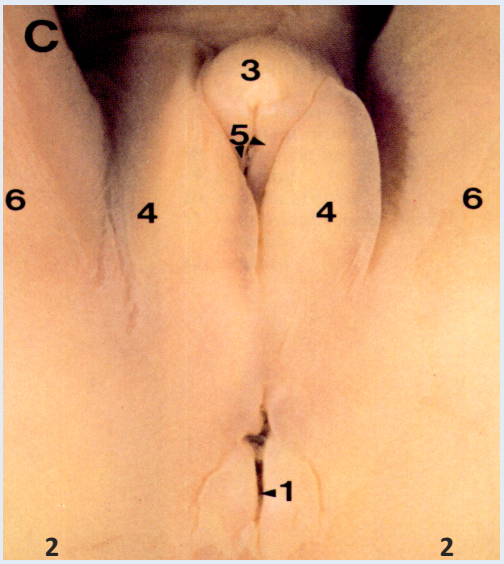

1- anus
2- buttocks
3- clitoris
4- labioscrotal swelling (labia majora)
5- leg
6- urogenital fold (labia minora)

## Week 13



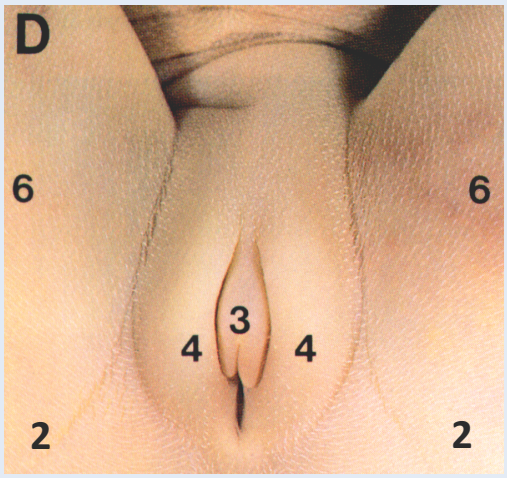

1- anus
2- buttocks
3- clitoris
4- labia majora
5- labia minora
6- leg

## Week 17



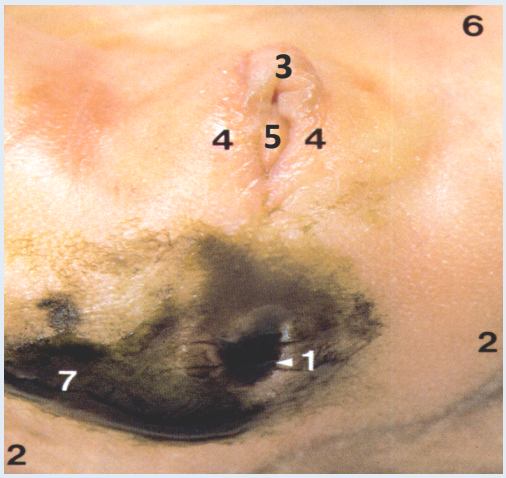

1- anus
2- buttocks
3- clitoris
4- labia majora
5- labia minora
6- leg

Week 20




- 1- anus
- 2- buttocks
- 3- clitoris
- 4- labia majora
- 5- labia minora
- 6- leg

Week 35




- 1- anus
- 2- buttocks
- 3- clitoris
- 4- labia majora
- 5- labia minora
- 6- leg
- 7- meconium

# C - Internal genital organs

## 1- Gonads

- Undifferentiated gonads begin to develop on the 5th week.
- Germ cells originate in the yolk sac and migrate to the genital ridge.
- In the absence of Y chromosome, the undifferentiated gonad develop into an ovary.
- 45/XO embryo (turner's syndrome) the ovaries develop but undergo atresia → streak ovaries (not normal ovaries).
- The gonads develop from the mesothelium on the genital ridge → 1ry sex cords grow into the mesenchyme → outer zone cortex and inner zone medulla.
- The **ovary develop from the cortex** and the medulla regress.
- The testes develop from the medulla and the cortex regress.
- The development of the testes requires the presence of SRY gene

(Sex determining region Y) found on Y chromosome.

- The ovary contains 2 million 1ry oocytes at birth (after birth no more oocytes formation)

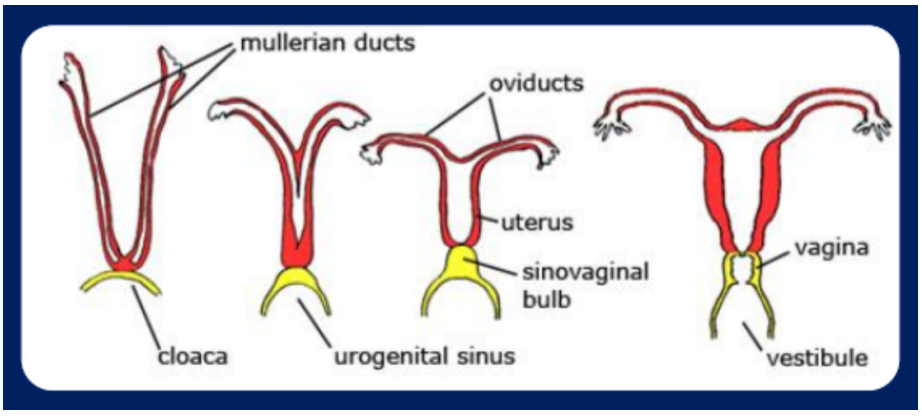
## 2- Uterus and fallopian tubes (from paramesonephric duct – PMN duct)

- Invagination of the coelomic epithelium on the cranio-lateral end of the mesonephric ridge → Paramesonephric ducts
- Fusion of the two PMN ducts (mullerian ducts) → uterus, cervix and fallopian tubes (at 8-11 week)
- 12-16 weeks → proliferation of the mesoderm around the fused lower part → muscular wall (muscles of the uterus)
- In the male fetus the testes secrete the mullerian inhibiting factor → regression of the mullerian ducts (explained before, deficiencies in MIF → abnormal male)

## 3- Vagina (2 parts)

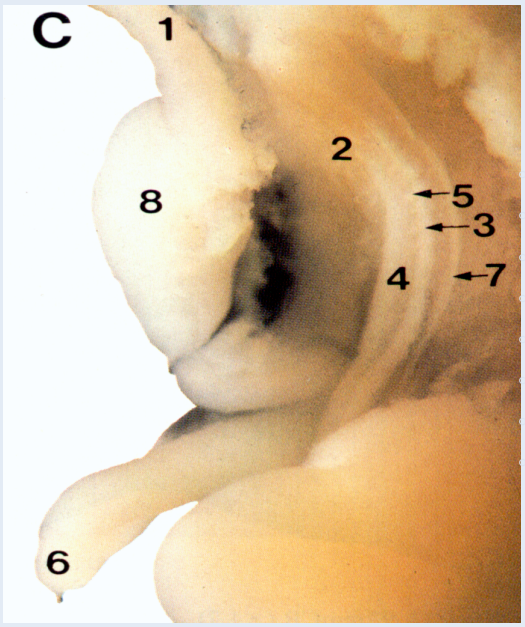
- The caudal ends of the mullerian ducts form the mullerian tubercle at the dorsal wall of the urogenital sinus.
- Mullarian tubercle is obliterated → vaginal plate → 16-18 week the central core breaks down → vaginal lumen.
- The upper 2/3 of the vagina → formed by mullerian tubercle.
- The lower 1/3 → urogenital sinus.

### *Normal development*



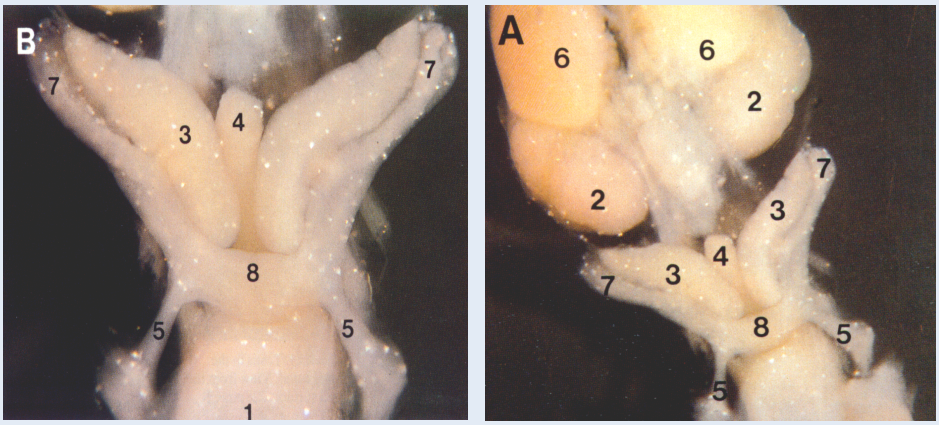

-The upper vagina, cervix & fallopian tubes are formed from the paramesonephric "PMN" (mullerian) ducts.
-the absence of Y chromosomal influence leads to the development of PMN system &total regression of the mesonephric system.

## Day 36-38



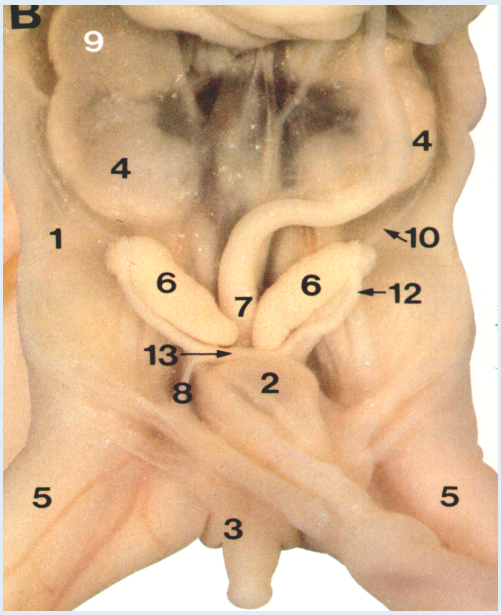

2- Gonadal ridge
3- Mesonephric duct
4- Mesonephric kidney
5- Mesonephric tubules
6- Midgut herniation
7- Paramesonephric duct
8- Stomach

## Week 8



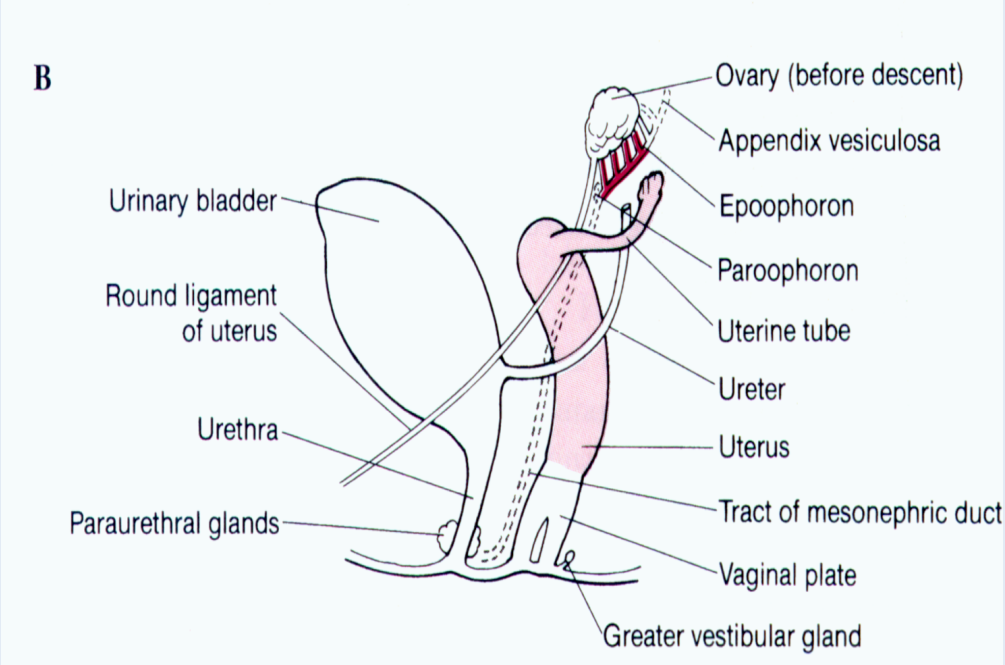

1- bladder
2- kidney
3- ovary
4- rectum
5- round ligament of the uterus
6- adrenal gland
7- fallopian tube
8- utero vaginal primordium

## Week 9

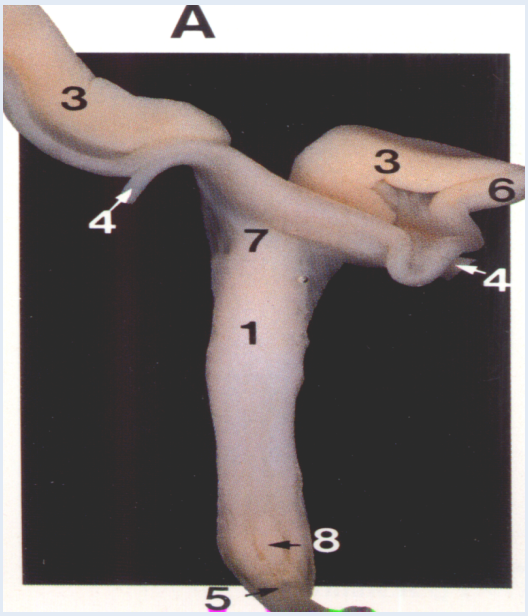



1- abdomen
2- bladder
3- external genitalia
4- kidney
5- leg
6- ovary
7- rectum
8- round ligaments
9- adrenal glands
10- suspensory ligament
12- uterine tube
13- uterovaginal primordium

## Week 12

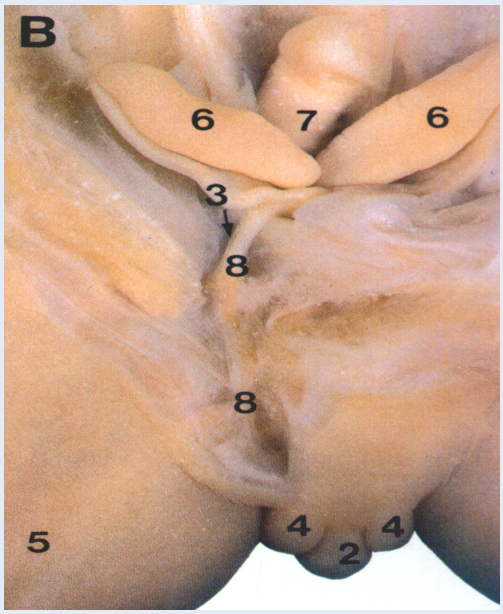


## Week 13 (dissected genital tract)



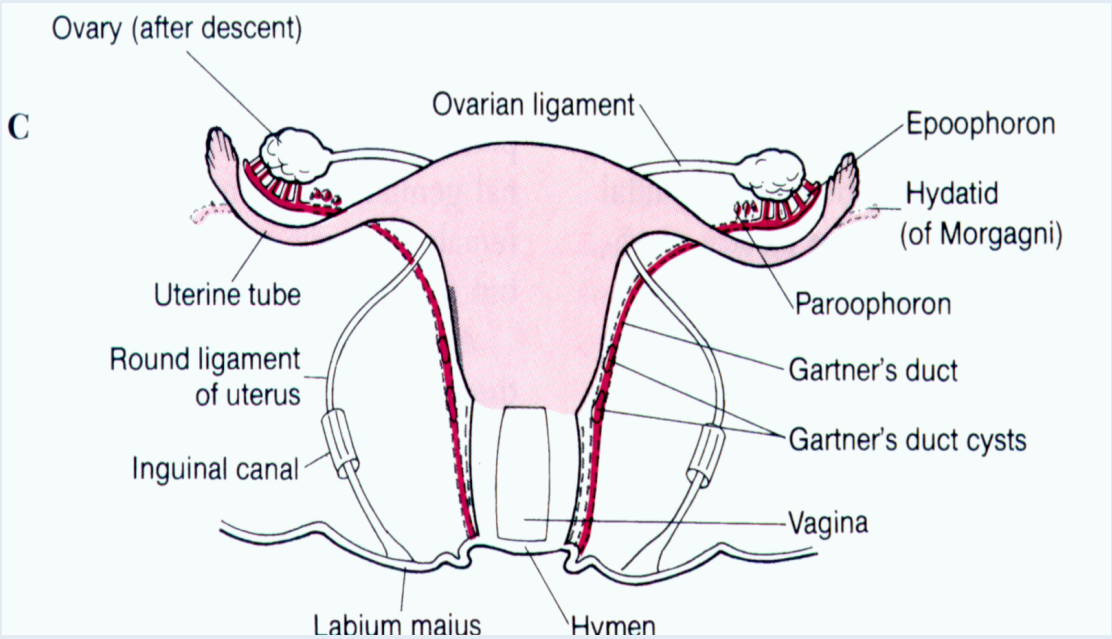

1- body of uterus
2- clitoris
3- ovary
4- round ligament
5- solid epithelium (vagina meets urogenital sinus)
6- fallopian tube
7- uterus
8- vagina

## Week 15




1- bladder
2- clitoris
3- vaginal process
4- labia majora
5- leg
6- ovary
7- rectum
8- uterine round ligament
9- uterovaginal primordium

## Newborn



Skipped by the doctor ^^

Male and Female Derivatives of Embryonic Urogenital Structures

| Embryonic Structure | Derivatives | |
|---|---|---|
| | Male | Female |
| Labioscrotal swellings | Scrotum | Labia majora |
| Urogenital folds | Ventral portion of penis | Labia minora |
| Phallus | Penis | Clitoris |
| | Glans, corpora cavernosa penis, and corpus spongiosum | Glans, corpora cavernosa, bulb of the vestibule |
| Urogenital sinus | Urinary bladder<br>Prostate gland<br>Prostatic utricle<br>Bulbourethral glands<br>Seminal colliculus | Urinary bladder<br>Urethral and paraurethral glands<br>Vagina<br>Greater vestibular glands<br>Hymen |
| Paramesonephric duct | Appendix of testes | Hydatid of Morgagni<br>Uterus and cervix<br>Fallopian tubes |
| Mesonephric duct | Appendix of epididymis<br>Ductus of epididymis<br>Ductus deferens<br>Ejaculatory duct and seminal vesicle | Appendix vesiculosis<br>Duct of epoophoron<br>Gartner's duct |
| Metanephric duct | Ureter, renal pelvis, calyces, and collecting system | — |
| Mesonephric tubules | Ductuli efferentes<br>Paradidymis | Ureter, renal pelvis, calyces, and collecting system<br>Epoophoron<br>Paroophoron |
| Undifferentiated gonad | Testis | Ovary |
| Cortex | Seminiferous tubules | Ovarian follicles |
| Medulla | — | Medulla |
| | Rete testis | Rete ovarii |
| Gubernaculum | Gubernaculum testis | Round ligament of uterus |

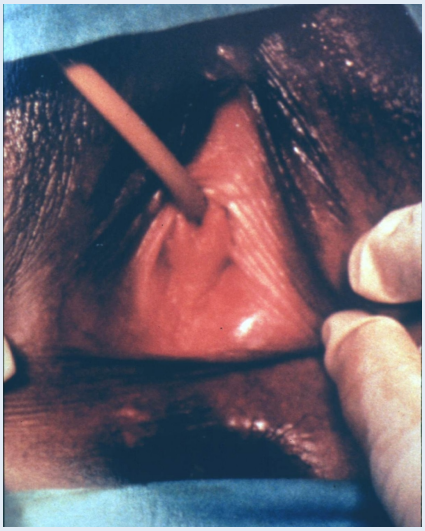
# 2 - Congenital malformation of the genital tract

## 1- Mullerian agenisis (mullerian duct is not formed either partially or completely)

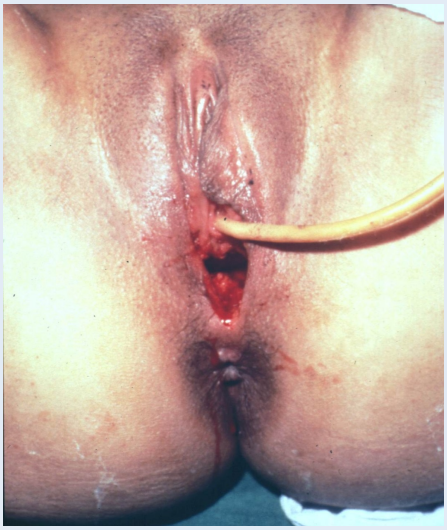
- Mayer –Rokitansky-Kuster-Huser syndrome (another name you don't have to memorize it ^^")
- Etiology is not known.
- Failure of mullerian duct development → absence of the upper vagina, cervix and uterus (uterine reminants may be found)
- The ovaries and fallopian tubes are present.
- Normal 46XX female ♀ with normal exrenal genitalia (the ovaries secret estrogen at the age of puberty → normal breast development)
- Patient presents with 1ry amenorroea.
- 47% have associated urinary tract anomalies (e.g. absent kidney)
- 12% skeletal anomalies (make sure the patient doesn't have it)

- Treatment:

a) Psychological counseling (supportive; you should tell her that she can't bear children, no period, she's infertile)
b) Surgical:
- vaginoplasty (only when she's ready to get married cause it needs regular dilatation, if you do it earlier it might get narrowed and closed again)
- excision of utrine reminant (if it has functioning endometrium)
- vaginal dilators
-



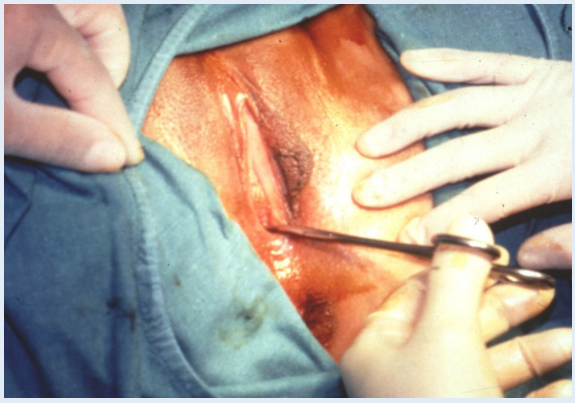
mullarian agenesis: no vagina, completely obliterated



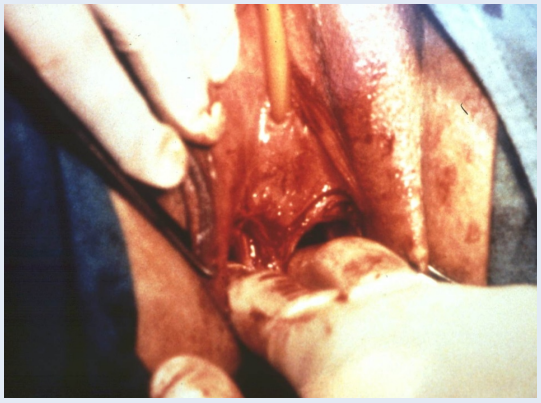
Sometimes they have what's called vaginal dimple, a very blind vagina that was developed from the urogenital tract

-Pt with 1ry amenorrhea, breast development & a 46XX karyotype have levels of testosterone appropriate of females.
-The clinical diagnosis may be caused by mullerrian defects that cause obstruction of the vaginal canal (e.g., imperforated hymen of a transverse vaginal septum) of by the absence of a normal cervix or uterus & normal fallopian tubes.

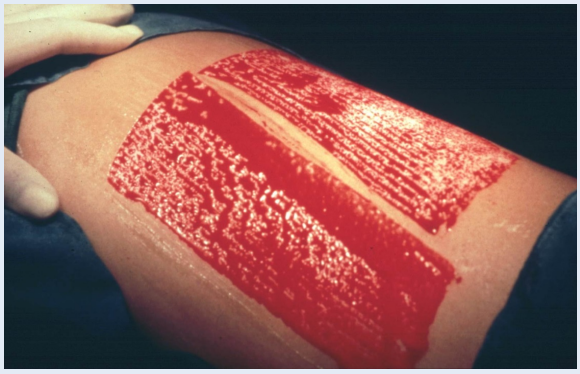
## Vaginoplasty:



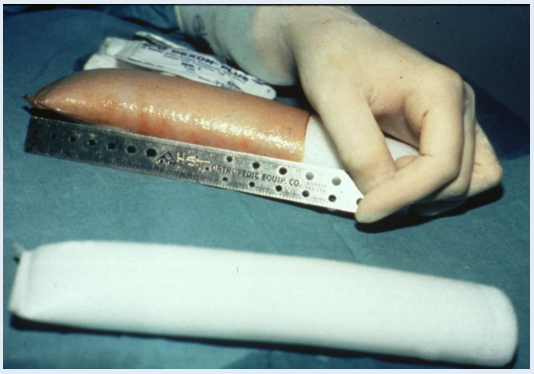
1- Make an opening



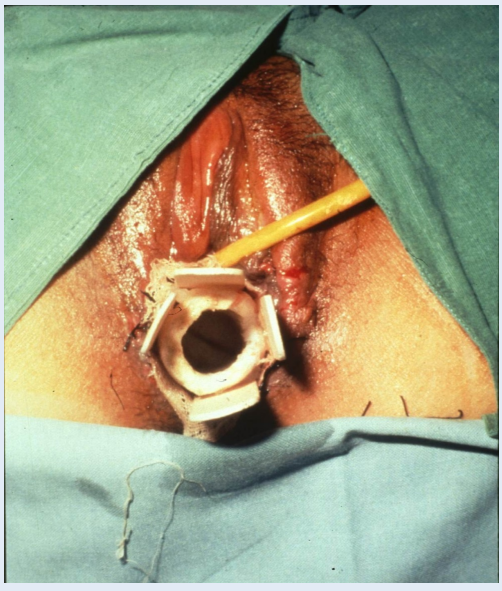
2- Dilate the area between the bladder and rectum to form vaginal space



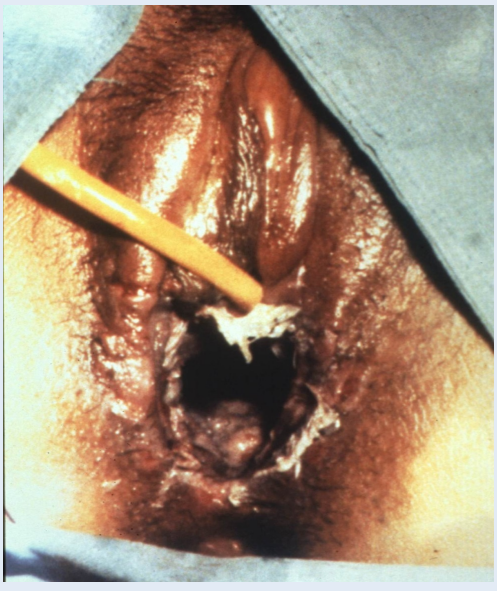
3- Skin grafting from the thigh



4- Placing the graft on the vaginal mold



5- Insert it; suture it with the surrounding skin



6- After suturing and removal of the mold

*(In the first 6 months she needs to wear this mold all the time so the skin does not shrink and collapse. After that, she can remove it but she needs a regular intercourse)*

* All the congenital malformations of the uterus can be associated with cervical +incompetence (which by itself can cause 2nd trimester abortion. treatment → cervical circulage)

## 2- Disorders of **lateral** fusion of the mullerian duct

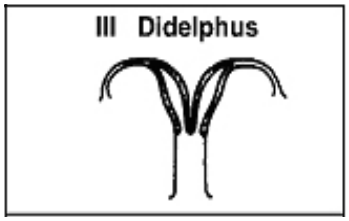
(either completely or partially)

- Incidence
  - Generally 0.1-2%
  - 4% of infertile patients
  - 6-10% recurrent abortion patients
- Most patients can conceive without difficulty.

- High Incidence of:
  - Recurrent abortions
  - Premature birth
  - Fetal loss
  - Fetal malpresentation
  - Cesarean section incidentally
  - Cervical incompetence (loose cervix, they will have mid trimester abortion – cervical cerclage to prevent)
- Clinical presentation:
  - Shortly after menarche → if there is obstruction to uterine blood flow (one tube is opened the other is obstructed → cyclic pain + mass)
  - Difficulty in intercourse → longitudinal vaginal septum (lower part didn't open completely)
  - Dysmenorrhea or menorrhagia
  - Abnormality detected on D&C (sometimes, incidentally discovered)
  - Ultra sound, laparoscopy or laparotomy
  - Complications of pregnancy (abortion, pre-term labor)
  - HSG (hysterosalpingogram) → when investigating for infertility or recurrent fetal loss

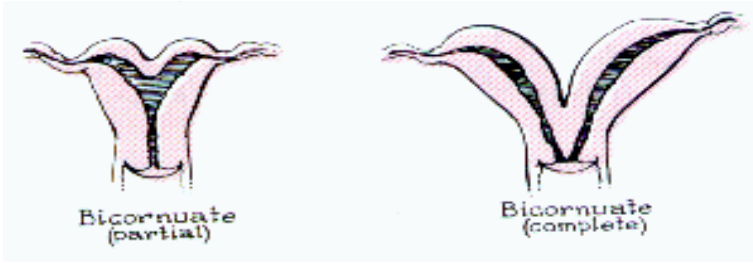
## A- Uterus didelphus (No fusion at all. not a serious condition, can be left without treatment)

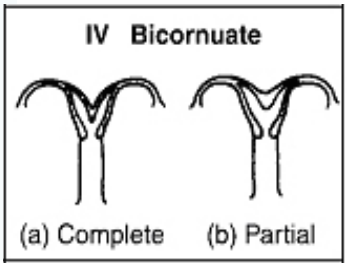

- Complete duplication of uterus, cervix and vagina (due to failure of fusion of the two mullerian ducts)
- Might increase pregnancy wastage.
- Diagnosis → HSG or at laparoscopy or laparotomy.
- Treatment → If affecting pregnancy outcome → surgical correction (also called uteroplasty or hysteroplasty)




## B- Bicornuate uterus

- Incomplete fusion of the two mullerian ducts
- Increase pregnancy wastage (pre term labor 25-26 weeks, late or mid-trimester abortion 17-18 weeks.
- Surgical interventions are required according to the degree of the pre maturity, late preterm → no medical interfere.
- Diagnosis → HSG or at laparoscopy or laparotomy
- Treatment → If affecting pregnancy outcome (or causing recurrent pregnancy loss) → surgical correction (metroplasty): cut the septum.

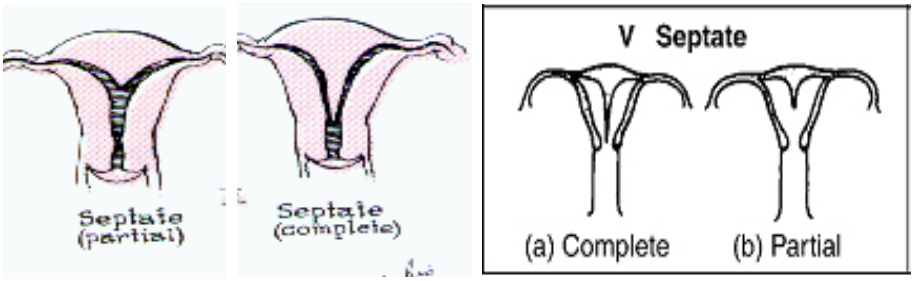


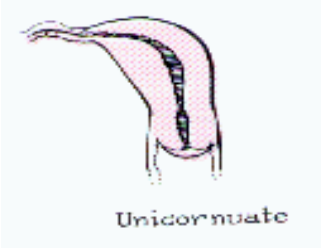

## C- Septate uterus

- External contour of the uterus is normal but there is intrauterine septum of varying length thickness.
- Worst pregnancy outcome (they will have early -1st trimester- abortion especially if the embryo implants in the septum)
- Diagnosis → both HSG & laparoscopy (or at the time of D&C)

- Treatment → Hystroscopic excision of the septum (easier) they present earlier in comparison to the bicurnate.

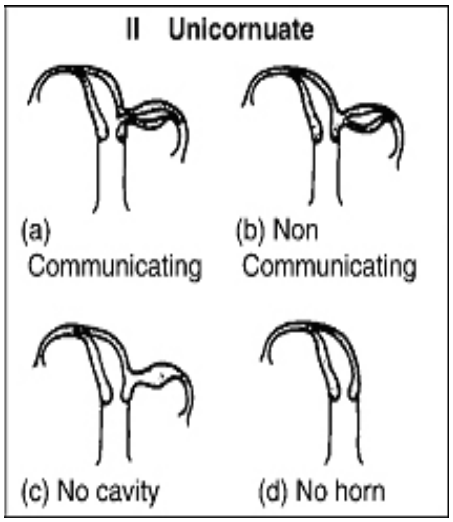



**D- Unicornuate uterus** (patients are normal usually unless the other duct is partially developed it will form what it called rudimentary horn – explained next)




- Due to development of only one mullerian duct.
- Almost all patients have associated single kidney.
- Pregnancy outcome → similar to patient with didelphic uteri.
- Diagnosis → HSG or surgery.
- Treatment → NO corrective surgery, unless patient has associated cervix incompetence → cervical cerclage.

**E- Unicurnuate with rudimentary horn** (the problem begins when the rudimentary horn is obstructed and the blood is collected inside it, sometimes ectopic pregnancy can occur)




1. Non-communicating horn 90%
   - Present with cyclic pelvic pain, mass
   - Complication: ectopic pregnancy in the rudimentary horn, endometriosis
   - Treatment → surgical excision

2. Communicating horn
   - Present with ectopic pregnancy in the rudimentary horn or increased pregnancy wastage.

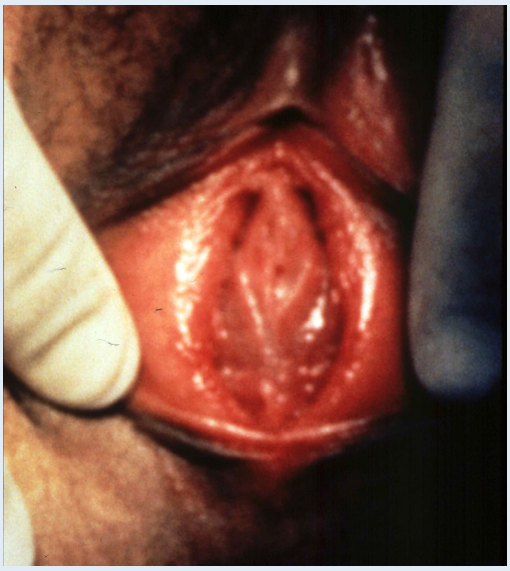
# 3- Disorders of **vertical** fusion of the mullerian duct

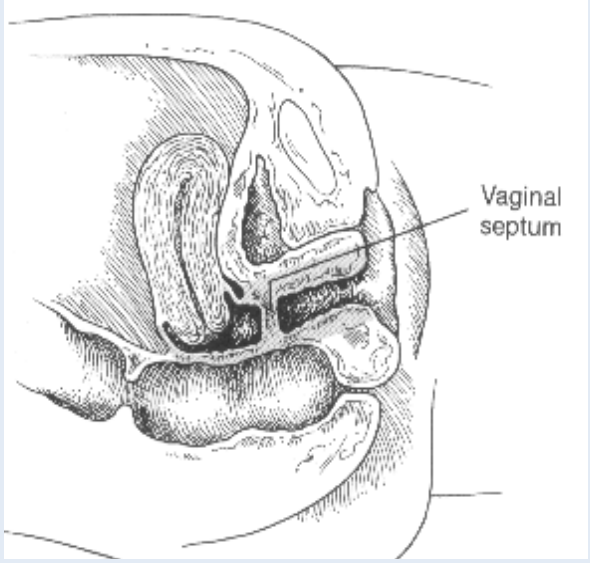
(fusion between the caudal (lower) part of the mullarian duct and the urogenital sinus to form the vagina)

## A- Vaginal septum

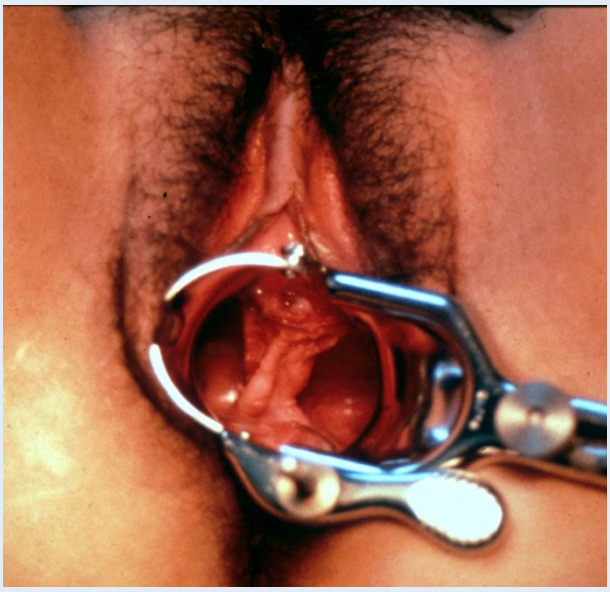
- Faults in the junction between the mullerian tubercle & the urogenital sinus → could be very thick or thin.
- 85% in upper two third of the vagina (sometimes it's lower and you can see it in the vaginal examination)
- Patient presents 1ry amenorrhea, hematocolpos*, mass (at the age of menarche, because blood is collected inside) or cyclic abdominal pain.
- High incidence of endometriosis
- Treatment → surgical excision of the septum (The higher the septum the more difficult to treat)

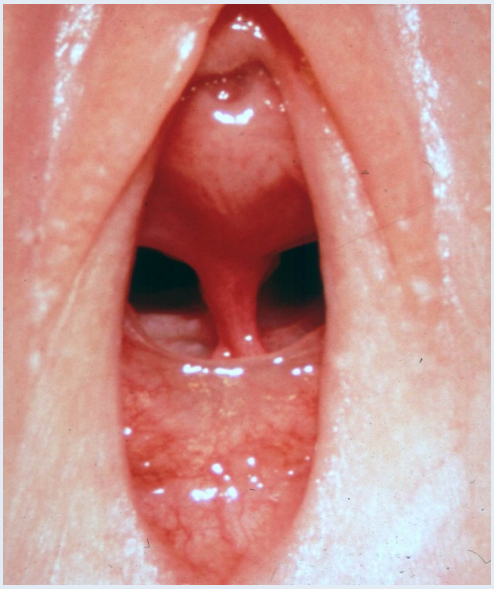
Blood collected in the vagina: hematocolpos*
Blood collected in the uterus: hematometra





Longitudinal vaginal septum



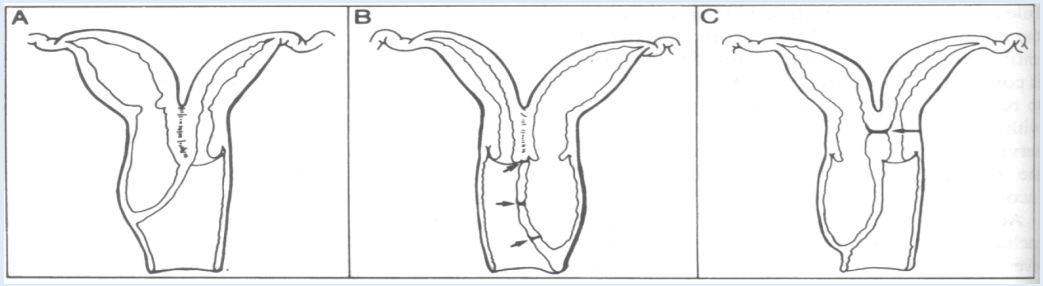


**B- Cervical agenesis / dysgenesis** (cervix is not form from the lower part of the mullarian duct)

- Very rare.
- Difficult to form a cervix, unsuccessful surgical correction (it won't work for a long time, it will get obliterated rapidly)
- Treatment → blood will keep collecting inside so the only treatment is hysterectomy or suppressing menstruation medically (depo provera)
- She can get pregnant through IVF (in vitro fertilization), delivery by CS – this is theoretical, most likely it won't be successful.

## 4- Unusual configuration of vertical/lateral fusion defects

- Combined lateral and vertical defects.
- Do not fit in other categories.
- E.g., double uterus with obstructed hemivagina.




A- Complete vaginal obstruction – vaginal septum (Will cause hematocolpos and hematometra)

B- Incomplete vaginal obstruction – fenestrated vaginal septum (Blood might move from the vagina and get infected – worst scenario)

C- Complete vaginal obstruction with double uterus.

## 5- Defects of the external genitalia

**a. Ambiguous genitalia**

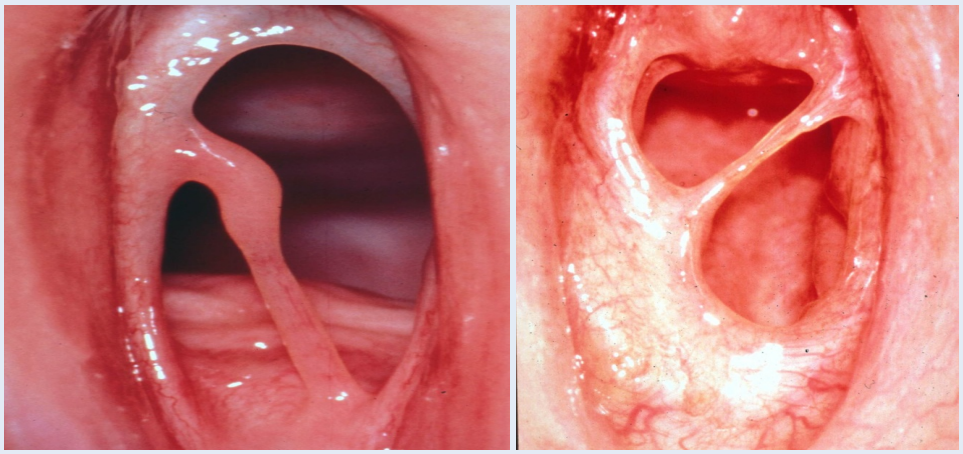
- Most common cause: congenital adrenal hyperplasia (will be discussed)
- Hermaphrodites: (will be discussed)
- (external resource, only if you're interested, it'll organize the whole idea in your brains 🤯: http://www.mayoclinic.org/diseases-conditions/ambiguous-genitalia/basics/causes/con-20026345)

**b. Defects of the clitoris**

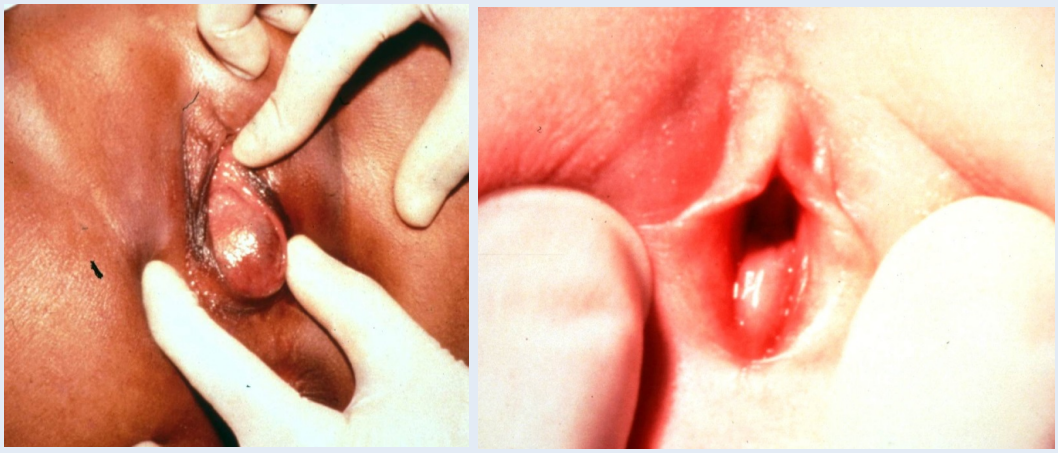
- Bifid clitoris (rare) (bifid means: divided by a deep cleft or notch into two parts)
- Hypertrophied clitoris (enlarged - androgen effect)

**c. Imperforate hymen**

- Hymen is formed at the junction of the urogenital sinus & sinovaginal bulbs (it's different from vaginal septum, grossly hymen is very thin, septum is much more thicker and it's usually diagnosed by ultrasound)
- Patient presents with 1ry amenorrhea with cyclic abdominal pain or hematocolpus / hematometra (mimics cervical agenesis and vaginal septum) can be seen with a naked eye in vaginal examination.
- Treatment → cruciate incision



Incomplete vaginal septum, very simple procedure, removed in the clinic.



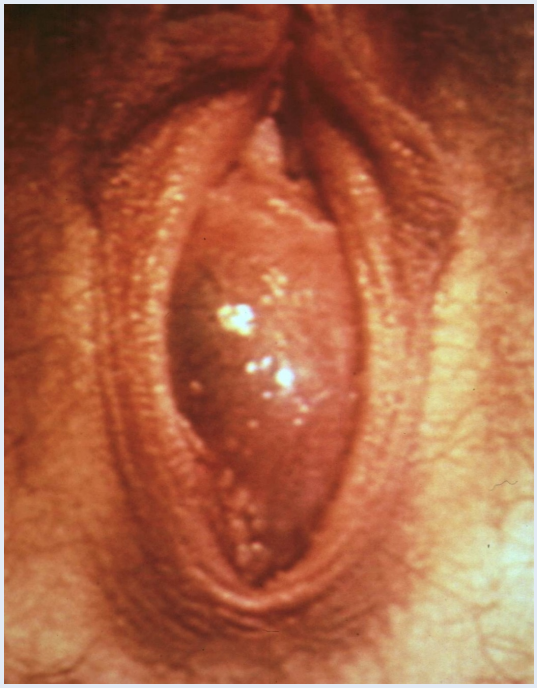
Mucocele: babies with imperforate hymen will have mucus collected in the vagina where it looks like this.

When there is bluish fluid collected behind the hymen then it is usually blood (at the age of menarche)

How to differentiate between vaginal septum and imperforate hymen?

Imperforate hymen: very thin bulging membrane with collection of blood behind it.

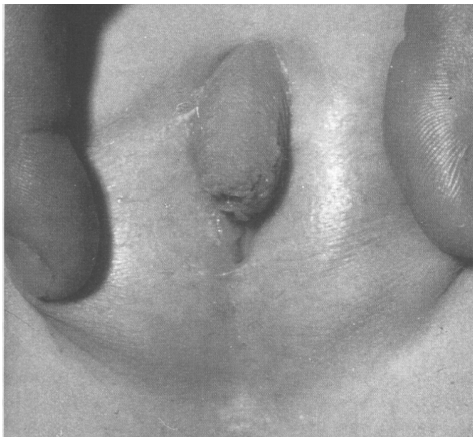
Septum will look like a blocked vagina (it might be septum or mullarian agenisi → US to diagnose)



## 3 - Intersexuality

| **True hermaphrodites:** The presence of testicular and ovarian tissue in the same individual. (extremely rare) | **Pseudo hermaphrodites:** | |
|---|---|---|
| | male | female |
| Most common: 46XX male due to the presence of the SRY gene (Sex determining Region Y) *very rare.* | 1- Chromosomal abnormality: Mosaicism associated with gonadal dysgenesis→ 45XO/46XY (means some cells have 46 chromosomes, others have 45)<br>* Mosaicism (*noun)* the property or state of being composed of cells of two genetically different types.<br>2-Testes are incapable of producing testosterone.<br>3-End organs are incapable of utilizing testosterone e.g. 5α reductase deficiency, failure of testosterone binding to receptors (androgen insensitivity)<br>4-Defficient production of MIF → female internal genital organs in a normal male. | 6- Masculinization of the external genitalia in a normal genetic female (46XX) due to high androgen e.g. congenital adrenal hyperplasia, prenatal exposure to male hormones (rare condition) |

1. Masculinized female - female **Pseudo**hermaphrodites (completely normal female, only external genitalia is abnormal) - 46XX

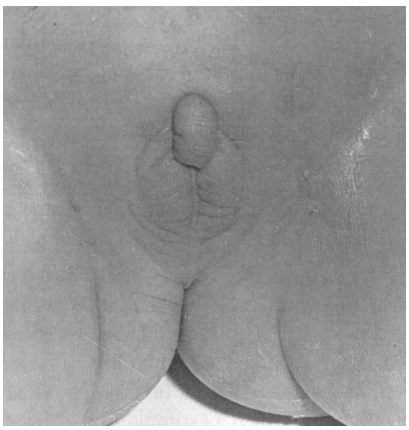


   a. **Congenital adrenal hyperplasia (CAH):** (enzymatic defect in the adrenal gland where the pathway of producing cortisone and aldosterone is blocked, androgen will be collected → high androgen in blood results in masculinizing effect)

   - The most common cause of female intersex.
   - Deficiencies of the various enzymes required for cortisol and aldosterone biosynthesis (21-hydroxylase, 11β-hyroxilase , 3βhydroxisteroid dehydrogenase) – *memorize*
   - 21-hydroxylase deficiency is the commonest defect 90%
   - Affected female may present at birth with ambiguous genitalia:
     - Enlargement of the clitoris
     - Excessive fusion of the genital folds obscuring the vagina & urethra
     - Thickening and rugosity of the labia majora resembling the scrotum
   - A dangerous salt losing syndrome due to deficiency of aldosterone may occur in some patient (considered as emergency)
   - Delayed menarche and menstrual irregularities.
   - Investigations:
     - Karyotyping:
       - 17-α-hydroxiprogestrone – *will be elevated*
       - 17-ketosteroids (androgens) in urine
     - Electrolytes
     - U/S (for internal genitalia → comfort the parents, tell them it's a normal female, and the situation is correctable surgically)
   - Treatment:

1. Cortisol or its synthetic derivatives (hormone replacement) → suppress the adrenals → lower androgen production.
2. Corrective surgery.
   - Clitroplasty (at the neonatal period to reduce the size of it)
   - Division of the fused labiocsrotal folds to create the vagina (should be delayed until puberty)

**b. Mother exposure to androgens:**



- Rare (because most women know they should avoid drugs during pregnancy)
- Androgen secreting tumours e.g. luteoma, arrhenoblastoma, or drugs.
- Female newborn will have androgenized external genitalia: not very well developed scrotum where the labia majora look darker and fused in the middle – mimics males'.
- Notice the enlarged clitoris in the picture and the obliterated Labioscrotal swellings.

## 2. Undermasculinized male - male pseudohermophrodites

**a. Anatomical testicular failure:** (testes are not developed at all or developed abnormally)

I. Normal chromosomes 46XY

- Patients display variable features: mild or severe (e.g. pure gonadal dysgenesis)
- (If sever, no gonadal secretions: patient looks like a normal female and you don't discover it till age of puberty when they present with amenorrhea, if mild with some gonadal secretions: looks like a female with mild masculinization)
- Uterus present (if testes are not secreting MIF)

II. Mosaicism 45XO/46XY

- Variable features – various degrees of ambiguous genitalia: normal female, ambiguous genitalia, nearly normal male

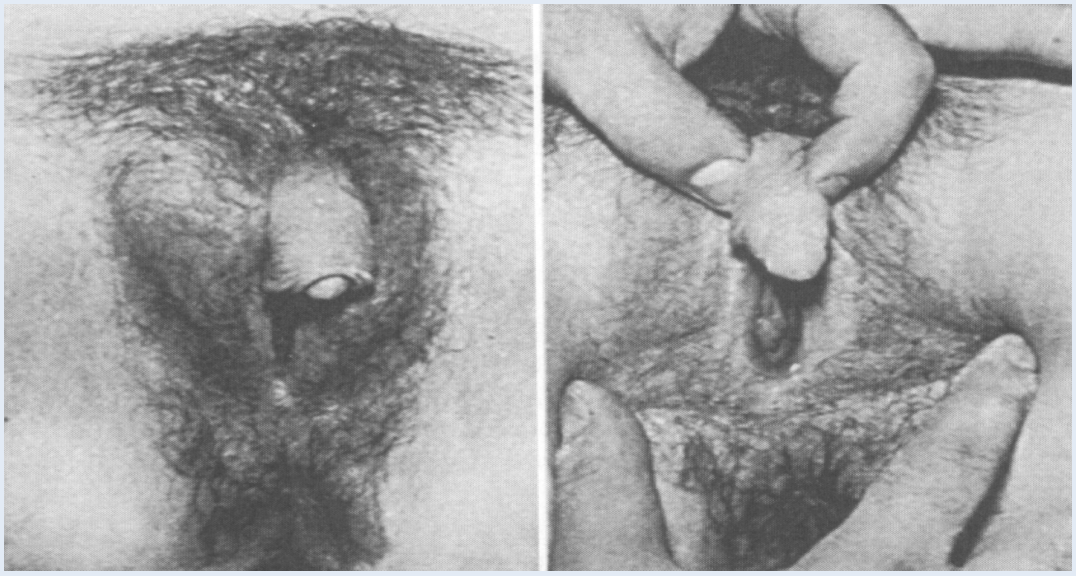
**b. Enzymatic testicular failure :**

- Enzymatic defects in the biosynthesis of testosterone.
- These defects are usually incomplete → Varying degrees of masculinization of the external genitalia, uterus and tubes → absent if the testes produce MIF.

**c. End-organ insensitivity:** (usually partial enzymatic defect, so they have some of the DHT still working. If it is complete enzymatic block, they will have female external genitalia)

I. 5α reductase deficiency (Autosomal recessive)

- Formation of the male external genitalia requires 5α reductase for the conversion of testosterone to dihydrotestosterone - DHT
- Formation of the internal wolffian structures (males' ductal part) respond directly to testosterone.
- Clinical features:
  - Female external genitalia with mild masculinization
  - Absent uterus (proper MIF secretions)
  - At puberty → high testosterone secretion → virilization (male 2ndry characteristic)



Male patient with a vaginal opening

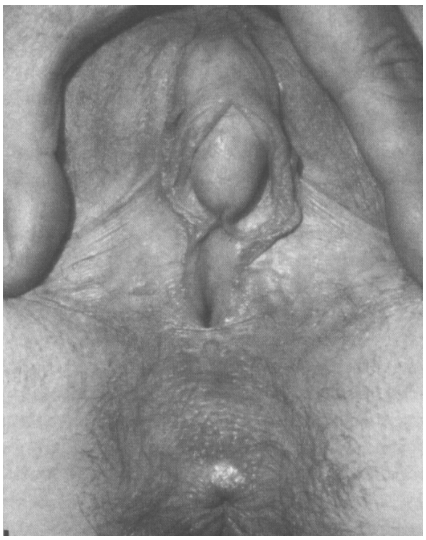
II. Androgen insensitivity (or testicular feminization TF – this term is not used anymore)

- Etiology
  1) Lack of androgen receptors → complete androgen insensitivity (classical TF) but they have MIF → so no uterus.
  2) Receptors are present but either few or inactive → incomplete androgen insensitivity.

| ***Complete Androgen Insensitivity*** | ***Incomplete Androgen Insensitivity*** |
|---|---|
| • Normal female external genitalia with blind vagina<br>• Absent uterus<br>• Breast development<br>• Present with 1ry amenorrhea<br>• Testes found in abdomen or inguinal canal<br>• Normal male Testosterone level<br>• Treatment:<br>◦ Gonadectomy after puberty due to high incidence of Malignant change (5%)<br>◦ Estrogen replacement | • Ambiguous genitalia with varying degrees.<br>• Breast development.<br>• Masculinization at puberty. |

## 3. True hermaphrodites

- The gonads of hermaphrodites possess testicular and ovarian tissue simultaneously as ***ovotestis***. (ovotestis: an organ producing both ova and spermatozoa)
  Patient might have:
  - Ovarian and testicular tissue at the same time (ovotestis)
  - Ovotestis on one side and ovary or testis on the other
  - Ovary on one side and testis on the other
  - Bilateral ovotestis.
- Varying degrees of sexual ambiguity
- Possible karyotypes:
  - 46XX most common
  - 46XX/XY
  - 46XY
  - 46XY/47XXY



✠ **Klinefelter Syndrome** (47XXY)

- Normal male external genitalia
- Tall stature
- Gynecomastia
- Azospermia (infertility)

# Summary

1. Sexual differentiation:
   - At the time of fertilization, sex of embryo is determined → according to the presence or absence of Y chromosome, testes or ovaries develop → with testes, male external and internal genital organs develop. No testes, female external and internal genital organs develop.
   - External genitalia can be differentiated grossly at the 12th week.

2. Congenital malformations:

| Mullerian agenesis | Disorders of lateral fusion of the mull ducts | Disorders of vertical fusion of the mullerian duct | Unusual configuration of vertical/lateral fusion defects | Defects of the external genitalia |
|---|---|---|---|---|
| — | 1. didelphus<br>2. bicornuate<br>3. septate<br>4. unicornuate<br>5. unicurnuate with rudimentary horn | 1. vaginal septum<br>2. cervical agenesis/dysgenesis | — | 1. Ambiguous genitalia<br>2. Defects of the clitoris<br>3. Imperforate hymen |

3. Intersex

| True Hermaphrodites | Pseudo Hermaphrodites | |
|---|---|---|
| 46XX most common | Undermasculinized male:<br>Anatomical testicular failure<br>Enzymetic testicular failure<br>End-organ insensitivity | Masculinized female:<br>Congenitall adrenal hyperplasia (CAH) – most common<br>Exposure of the mother to androgens |

# MCQ's :

**1- Fusion of the PMN happens at?**

a- 12-16 W
b- 16-18 W
c- 5th W
d- 8-11 W

**2- The upper 2\3 of the vagina develop from ..... while the lower 1\3 from .....?**

Mullerian Tubercle, Urogenital Sinus

**3- In mullerian agenesis, which one will be absent?**

a- Upper vagina and Cervix
b- Lower vagina and Cervix
c- Upper vagina, Uterus and Cervix
d- Lower vagina, Uterus and Cervix

**4- Abnormal shape of the uterus is diagnosed by?**

a- US
b- Speculum
c- HSG
d- only Laparotomy can diagnose it

**5- Masculinized female may have ....?**

a- Internal organs
b- 45XO
c- Low Androgen
d- 21, Hydroxylase deficiency

1=d

3=c

4=c

5=d

For mistakes or feedback

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