Abnormalities in female genital tract development

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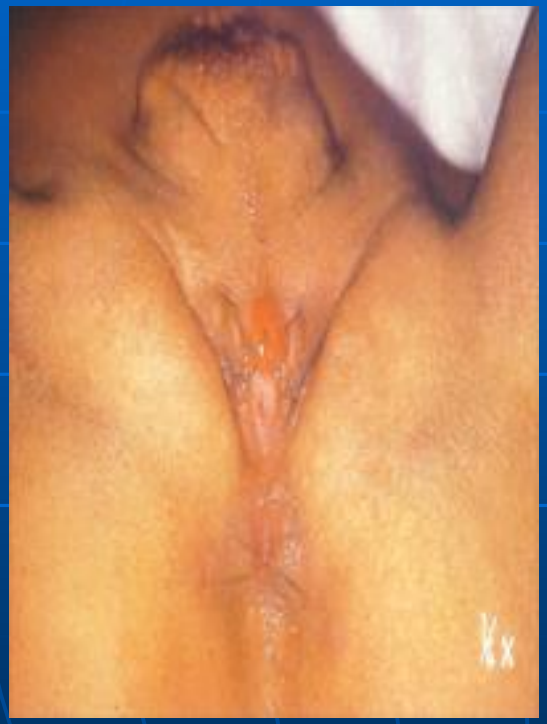
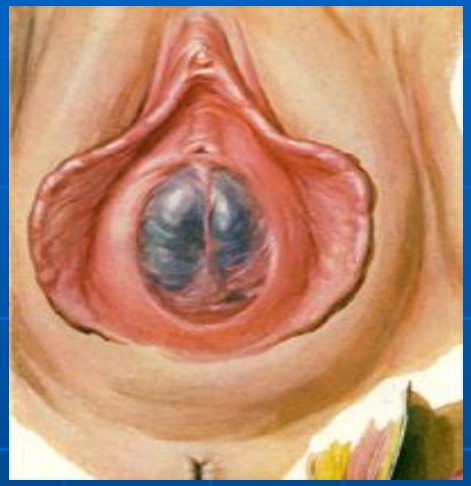
External genitalia

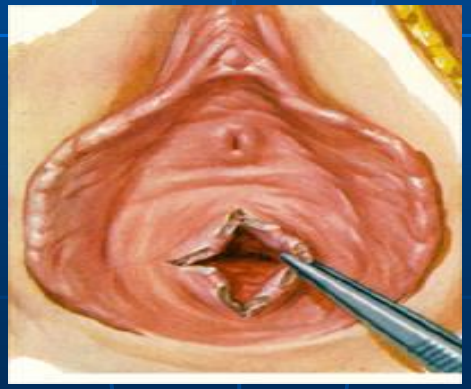
- Aplasia
- May present with or without developed hind limbs
- *Severe agenesis results in obliteration of perineum with absence of genital, urinary or anal orifices and fusion of lower limbs into one extremity (sirenomelia) incompatible with life.
- *With presence of lower limbs absent genitalia is noted by smooth perineum without orifices. Usually associated with extensive internal developmental defects



- Fusion
- Agglutination of the labia present from dense adhesions that hold the labia minora and majora together in the midline. This probably result from postnatal inflammation which is unnoticed
- Occasionally, fusion is complete as to stimulate the median raphe of male perineum and result in disposal of urinary stream. This is treated by separation of the adhesions and application of surface ointment to prevent recurrence

- **Imperforate hymen**
- The vaginal plate develops near the junction of the lower part of the vagina and the vestibule. Canalization of this plate is generally completed by the 6th. Month of fetal life. Failure of the final process of canalization occurs and results in imperforate hymen which obstruct the menstrual outflow
- Presentation:
- Usually present with primary amenorrhea with monthly pain
- Sometimes present with retention of urine due to collection of blood in vagina (hematocolpos), uterus (hematometra) and tubes (hematosalpinx)
- Treated by cruciate incision or excision of hymen





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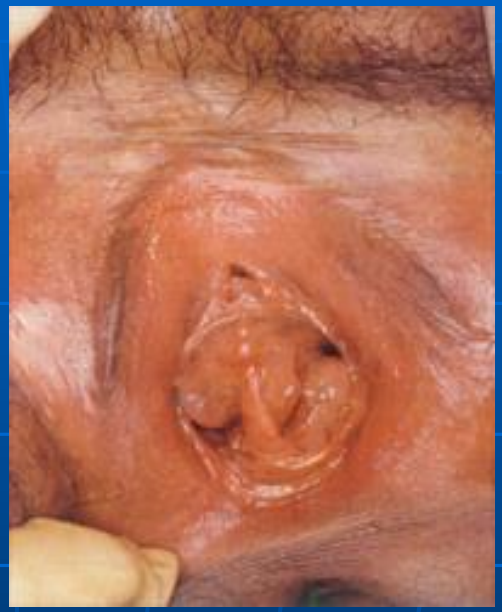
Vaginal anomalies

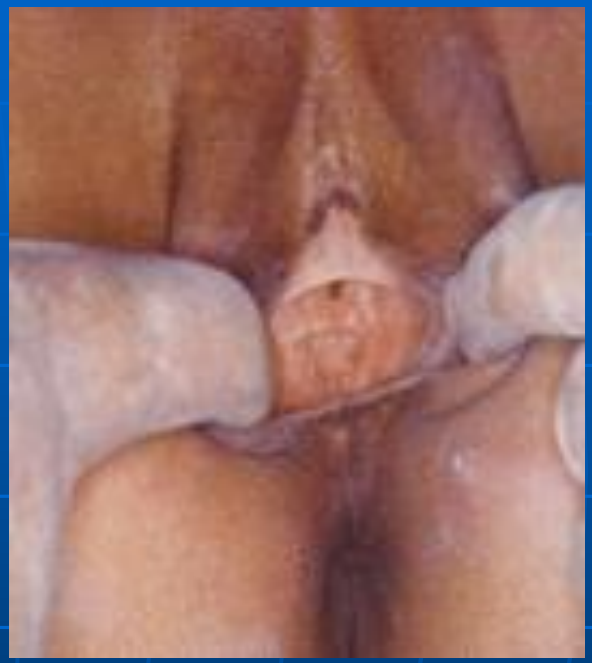
- **Vaginal agenesis**
- This could be partial or complete
- *complete absence of vagina is generally a Mullerian agenesis with absent uterus
- Patient usually have normal external genitalia and lower vagina derived from the urogenital sinus
- Ovaries are normally developed
- Present usually with primary amenorrhea
- In 50% urinary anomalies present e.g. absent or pelvic kidney
- Anomalies of spinal cord is seen in some cases
- Treatment: 1-construction of new vagina
- (McIndoe'operation)
- 2- Gradual dilatation (Frank's method)

- Frank's non-operative procedure used progressive larger dilators inserted by the patient to gradually deepen and widen the canal
- Vaginal construction (McIndoe's operation)
- It involves the development of a space between the bladder and rectum by blunt dissection and inserting a skin-graft-covered mold (from thigh) into the cavity allowing epithelialization. This undergoes metaplasia and loss its keratinization.
- -timing is important since narrowing due to scarring occurs unless regular coital activity is undertaken
- N.B Testicular feminization
- This is XY person with no external genitalia, instead a blind vaginal pouch present, but here, testes are present .treated by creation of new vagina and removal of testes after growth to avoid the possibility of future malignancy (30%)

- **Longitudinal septa**
- Partial or complete septa extending antroposteriorly in the vagina results from failure of disappearance of the fused Mullerian structure, this may be associated with normal uterus or duplicate uterus
- Occasionally the septum may not be in the midline and may not communicate externally leading to lateral pouch containing menstrual blood



- **Transverse septum**
- Rare occurs at the junction of upper and middle 1/3 of vagina
- When there is no perforation in the septum retention of mucoid material may occur before puberty leading to mucocolpos
- After puberty menstrual flow becomes hidden and accumulate in the vagina, uterus and tubes giving a picture like imperforate hymen
- If the septum is perforated patient may complain of dyspareunia and it may cause dystocia in labor



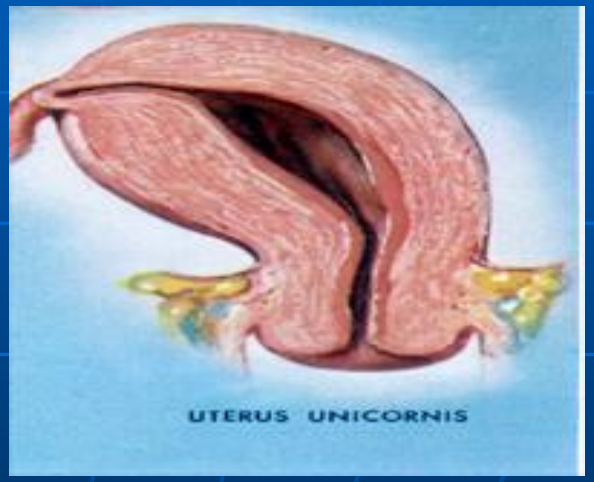
Internal organ anomalies

- Abnormalities of the upper Mullerian ductal system are enormous variety, but can generally be grouped into five categories:
- 1-Agenesis
- 2-Unicornuate uterus
- 3-rudimentray uterine horn
- 4-Symmetric duplication

- **Agenesis**
- Complete failure in the development of the Mullerian duct results in the absence of uterus, vagina and tubes
- Usually vulva is normal with normal hymen
- It is essentially a case of vaginal agenesis

- **Unicornuate uterus**
- Absence of one Mullerian duct results in a unicornuate uterus with only one fallopian tube
- Although vagina and cervix may be normal in both appearance and function they are in fact represent one-half the fully developed organs
- It is relatively rare and usually associated with absence or gross malformation of the urinary tract on the site of the missing Mullerian duct

- Complications:
- 1-infertility
- 2-recurrent abortion
- 3-prematurity
- 4-Abnormal fetal presentation
- 5-abnormal placental implantation

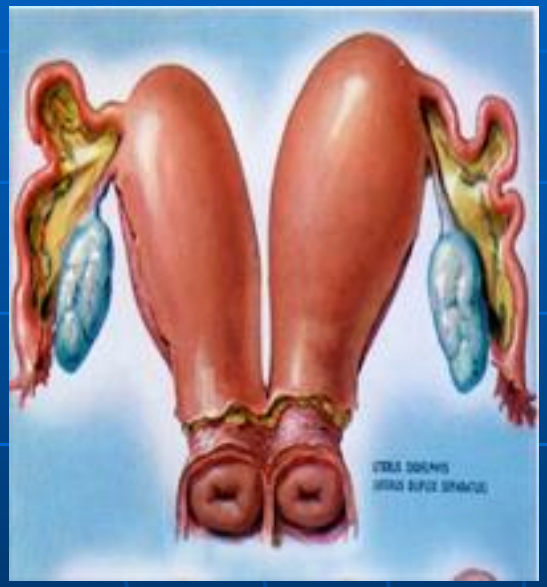


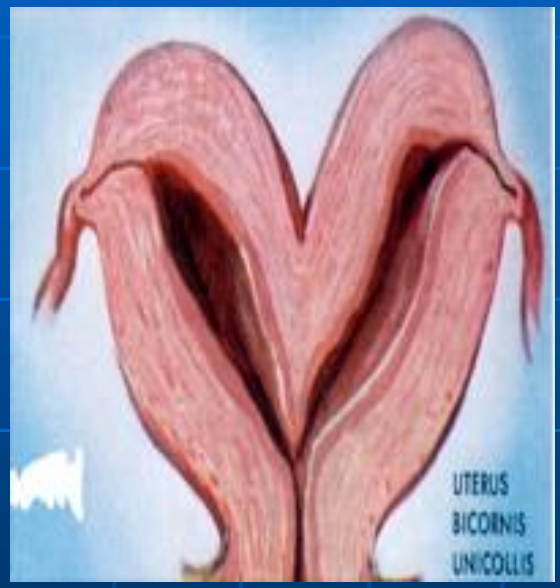
- **Rudimentary uterine horn**
- Incomplete development of one Mullerian duct ,is more common
- Most are non-communicating and are connected to the opposite unicornuate uterus by fibrous plica
- Often discovered in asymptomatic patient during laparoscopy

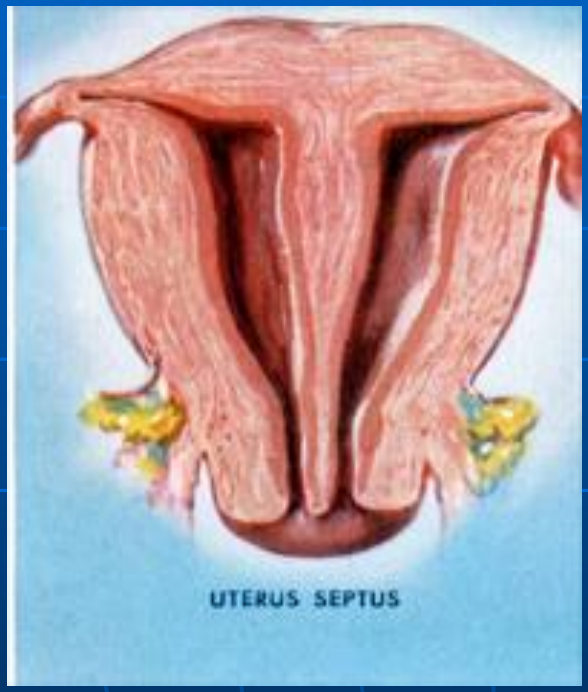
- Communicating horn in some cases may present and if pregnancy occurs it leads to rupture in the second trimester

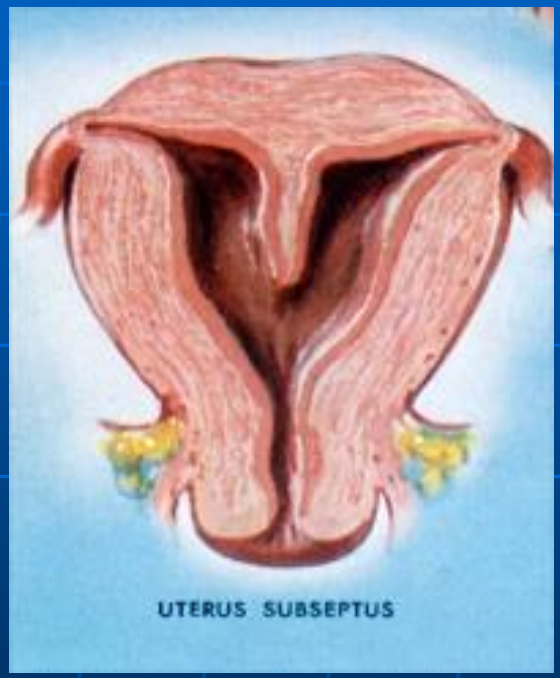
- Treated by removal of the rudimentary horn

- Symmetric duplication may result in
- Uterus didelphia
- Bicornuate
- Septate uterus




UTERUS
BICORNIS
UNICOLLIS


UTERUS SEPTUS


UTERUS SUBSEPTUS