CONGENITAL MALFORMATIONS OF THE Q GENITAL TRACT

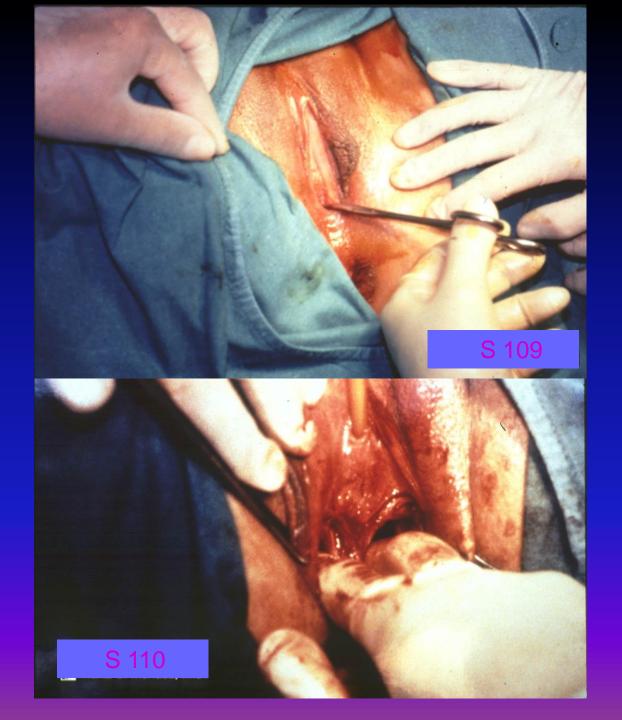
1-MULLERIAN AGENISIS

- Mayer Rokitansky-Kuster-Huser syndrome
- Etiology ?
- Failure of mullerian duct development
 ⇒ absence of the upper vagina, cx & uterus (uterine reminants may be found)
- The ovaries & fallopian tubes are present
- Pt present with 1ry amenorroea
- 47% have asociared urinary tract anomalies
- 12% skeletal anomalies
- Rx ⇒

psychological counseling surgical ⇒- vaginoplasty

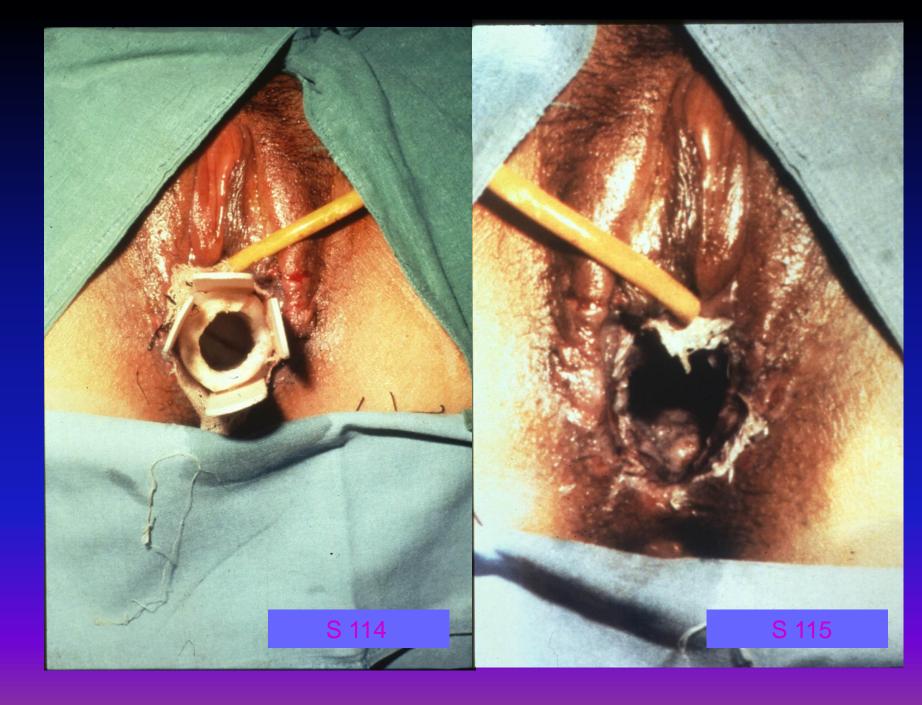
> excision of utrine reminant (if it has functioning endometrium)
> vaginal dilators

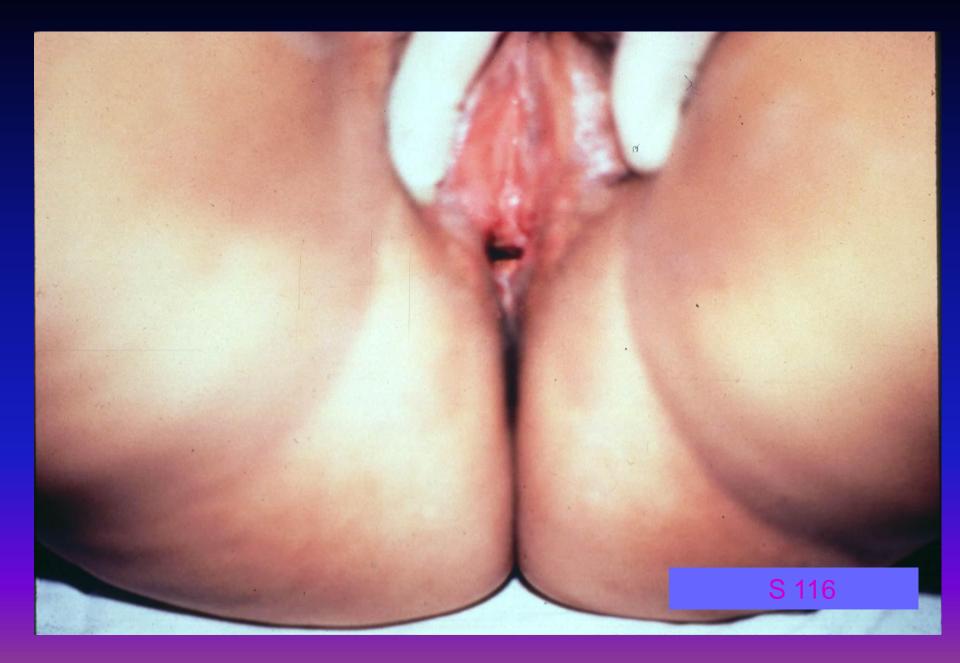


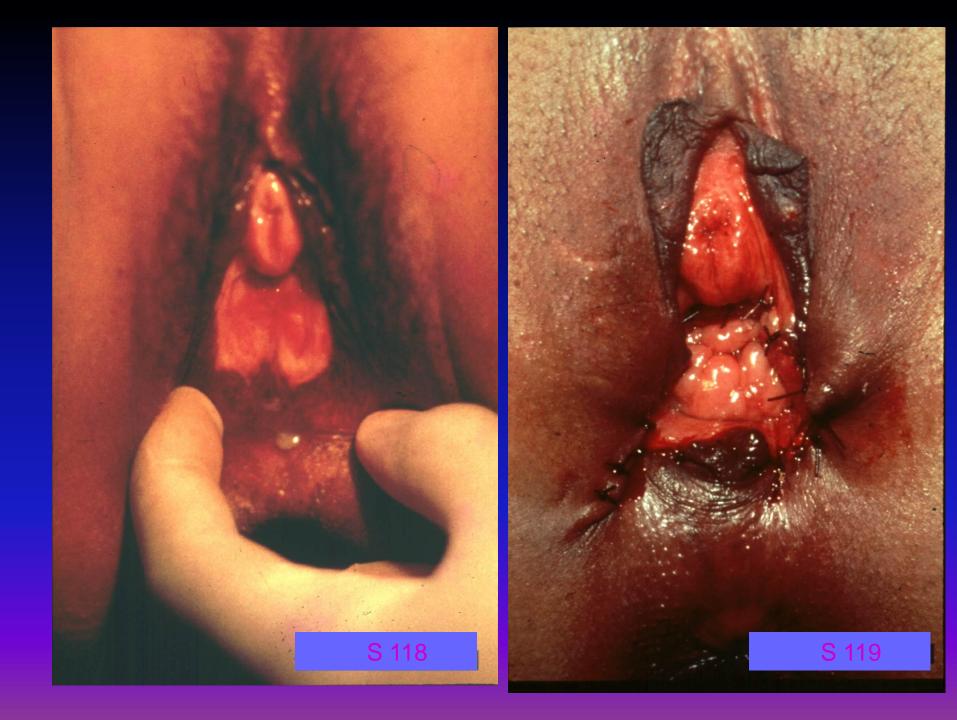














2-DISORDERS OF LATERAL FUSION OF THE MULL DUCTS

Incidence ? 0.1-2% 4% of infertile pt 6-10% recurrent abortion pt Most pt can conceive without difficulty 1 Incidence of :

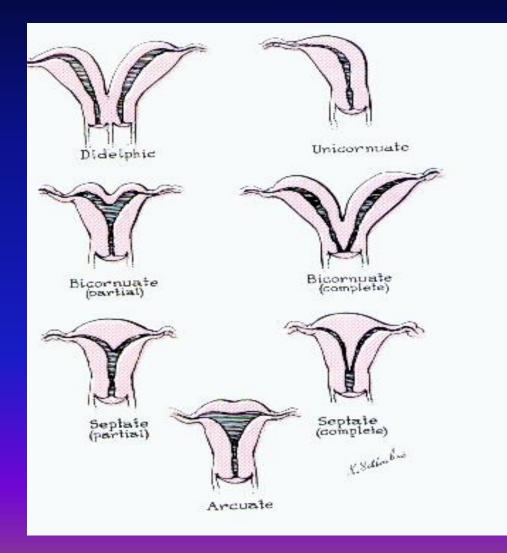
- ◊ recurrent abortions
- ◊ premature birth
- ◊ fetal loss
- ◊ fetal malpresentation
- $\diamond C S$
- ◊ cx incompetence

2-DISORDERS OF LATERAL FUSION OF THE M D

CLINICAL PRESENTATION

 Shortly after menarche ⇒if there is obstruction to uterine blood flow ♣Difficulty in intercorse ⇒ longitudinal vaginal septum Dysmenorrhea or menorrhagia Abnormality detected on D&C ♣U/S, laparoscopy or laparotomy ♣Palpable mass Complications of pregnancy ♣HSG ⇒ during infertility or RFL investigations

NON OBSTRUCTIVE MALFORMATIONS OF THE MULLERIANE DUCTS



2-DISORDERS OF LATERAL FUSION OF THE MULL DUCTS

A-Uterus didelphus

- Complete duplication of uterus, cx & vagina (due to failure of fusion of the two Mull ducts)
- ? 1 pregnancy wastage
- Dx ⇒ HSG or at laparoscopy / laparotomy
- Rx ⇒ If affecting pregnancy outcome ⇒ surgical correction (metroplasty)

B-Bicornuate uterus

- Incomplete fusion of the two Mull ducts
- ↑ pregnancy wastage
- Dx ⇒ HSG or at laparoscopy / laparotomy
- Rx ⇒ If affecting pregnancy outcome ⇒ surgical correction (metroplasty)

2-DISORDERS OF LATERAL FUSION OF THE MULL DUCTS

C-Septate uterus

External contour of the uterus is normal but there is intrauterine septum of varying length & thickness Worst pregnancy outcome Dx ⇒ both HSG & laparoscopy Rx ⇒ Hystroscopic excision of the septum

D-Unicornuate uterus

Due to development of only one Mull duct Almost all pt have associated single kidney Pregnancy outcome ⇔ similar to pt with didelphic uteri Dx ⇔ HSG or surgery Rx ⇔ NO corrective surgery ⇒ if pt has associated cx incompetence ⇔ cx cerclage 2-DISORDERS OF LATERAL FUSION OF THE MUL DUCTS

E-Unicurnuate with rudimentary horn

Noncommunicating horn 90% Present with cyclic pelvic pain, mass, ectopic pregnancy in the rud horn or endometriosis Rx is surgical excision **Communicating horn** Present with ectopic pregnancy in the rud horn or 1 pregnancy wastage

3-DISORDERS OF VERTICALE FUSION OF THE MULLERIAN DUCTS

A- VAGINAL SEPTUM

- Faults in the junction between the Mull. Tubercle & the urogenital sinus ⇒ could be very thick or thin
- 85% in upper two third of the vagina
- Pt present 1ry amenorrhea, hematocolpus, mass or cyclic abdominal pain
- 1 incidence of endometriosis
- Rx ⇒ surgical exision

B-Cx AGENISIS / DYSGENISIS

- V rare
- Difficult, unsuccessful surgical correction
- Rx ⇒ hysterectomy

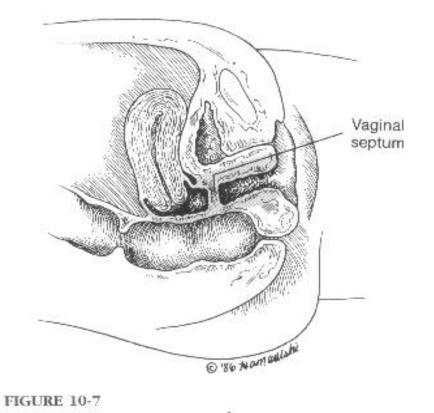
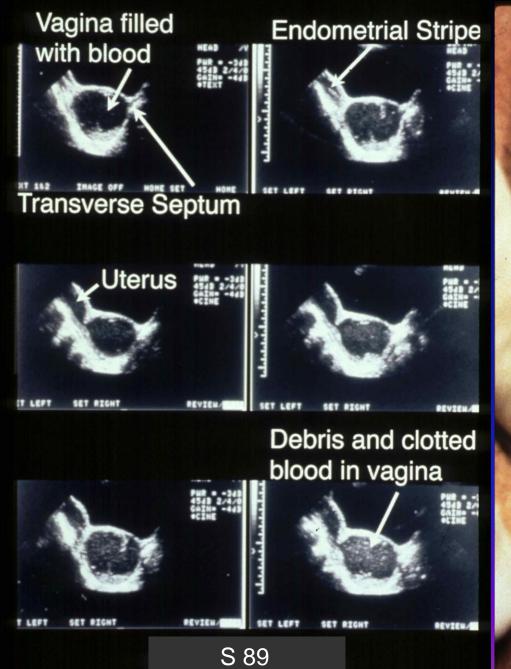
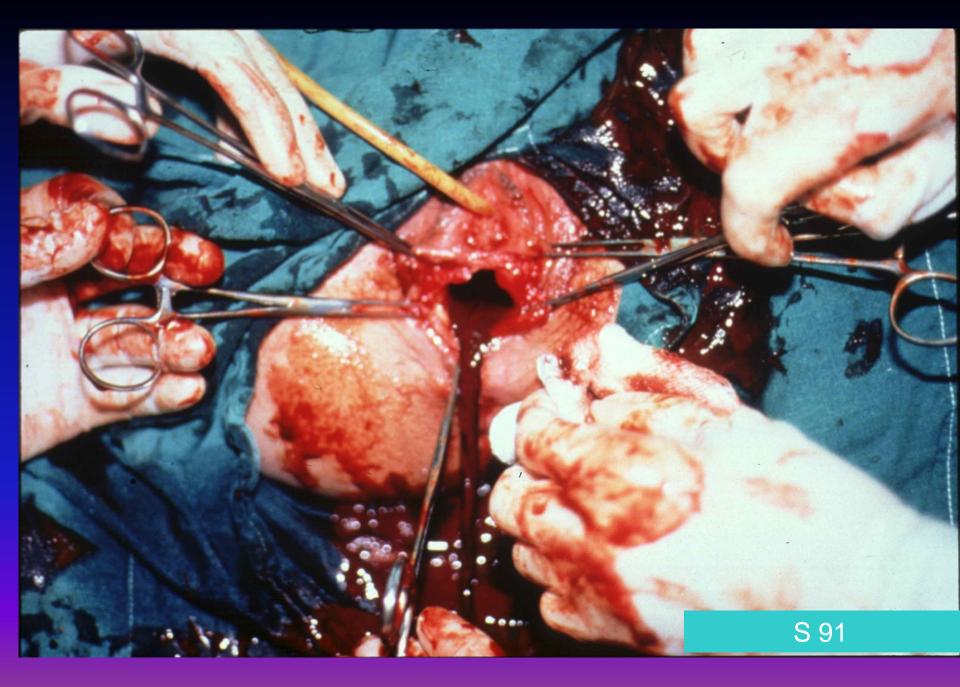


Diagram of transverse vaginal septum.







Long vag septum

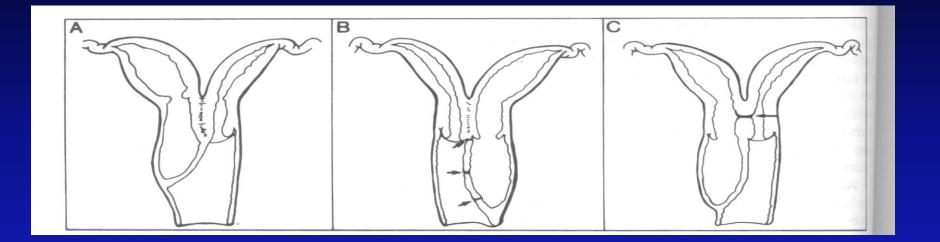




4-UNUSUAL CONFIGRATION OF VERTICAL/LATERAL FUSION DEFECTS

- Combined lateral & verticle defects
- Do not fit in other categories
- EXAMPLE, double uterus with obstructed hemivagina

Double uterus with vaginal obstruction



A-Complete vaginal obstruction B-Incomp vag obst

C-Comp obst with comm double uterus

5-DEFECTS OF THE EXTERNAL GENITALIA

- Ambigious genitalia
 congenital adrenal hyperplasia hermaphrodites
- Defects of the clitoris
 ⇒ uncommon
 ⇒ bifid clitoris
 hypertrophied
 ⇒ androgen effect

• IMPERFORATE HYMEN

Hymen is formed at the junction of the urogenital sinus & sinovaginal bulbs

Pt presents with 1ry amenorrhea with cyclic abdominal pain or hematocolpus /hematometra

Rx ⇔cruciate incision



