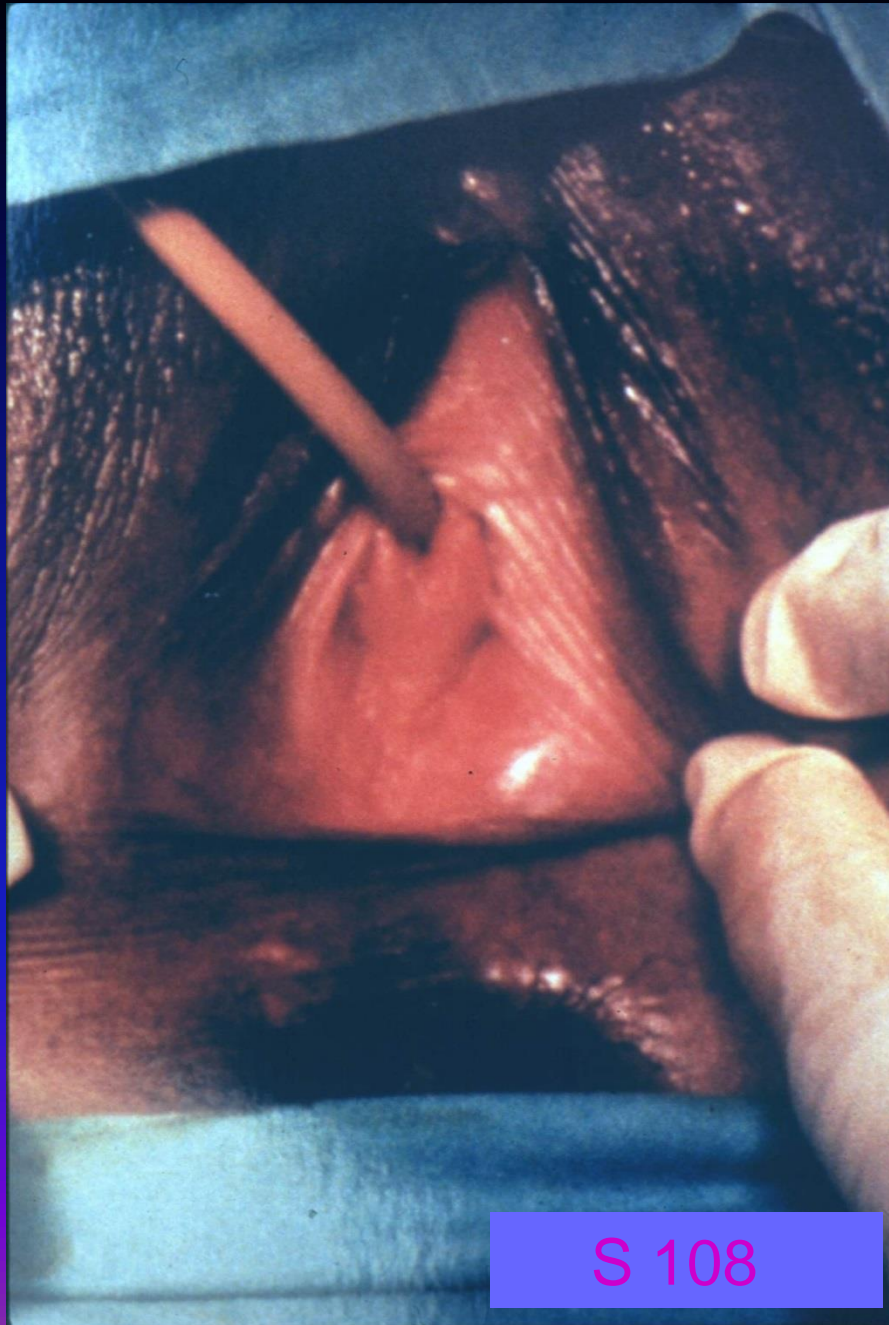


King Saud University Medical City
Department of Obstetrics & Gynecology
Course 482

CONGENITAL MALFORMATIONS
OF THE ♀ GENITAL TRACT

1-MULLERIAN AGENESIS

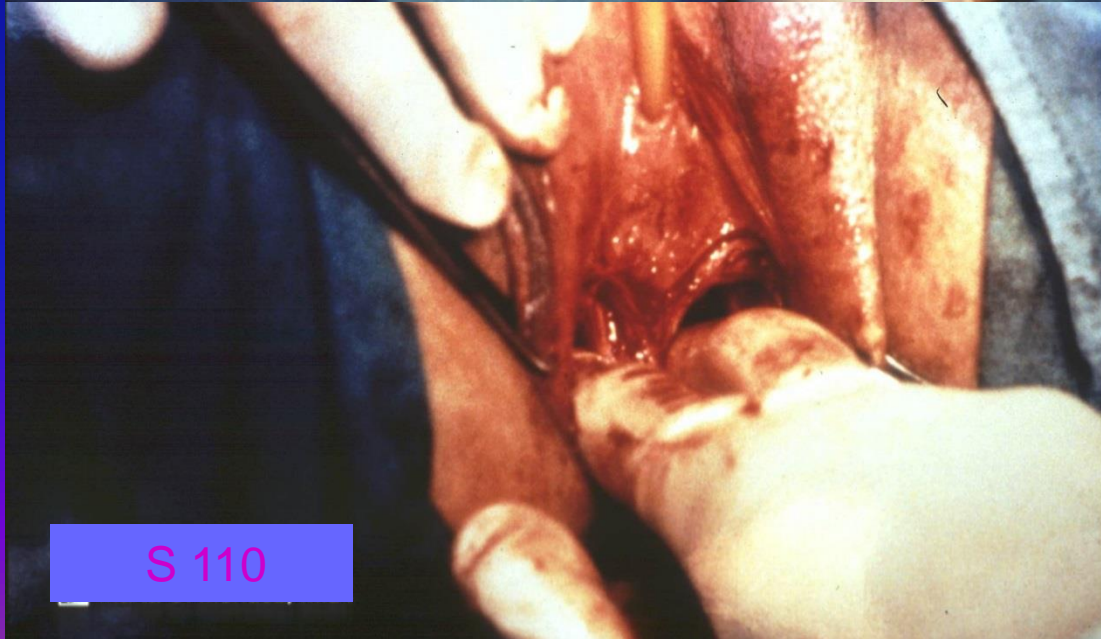
- Mayer –Rokitansky-Kuster-Huser syndrome
- Etiology ?
- Failure of mullerian duct development ⇨ absence of the upper vagina, cx & uterus (uterine remnants may be found)
- The ovaries & fallopian tubes are present
- Normal 46XX ♀ with normal external genitalia
- Pt present with 1ry amenorrhoea
- 47% have associated urinary tract anomalies
- 12% skeletal anomalies
- Rx ⇨
 - psychological counseling
 - surgical ⇨ - vaginoplasty
 - excision of uterine remnant (if it has functioning endometrium)
 - vaginal dilators



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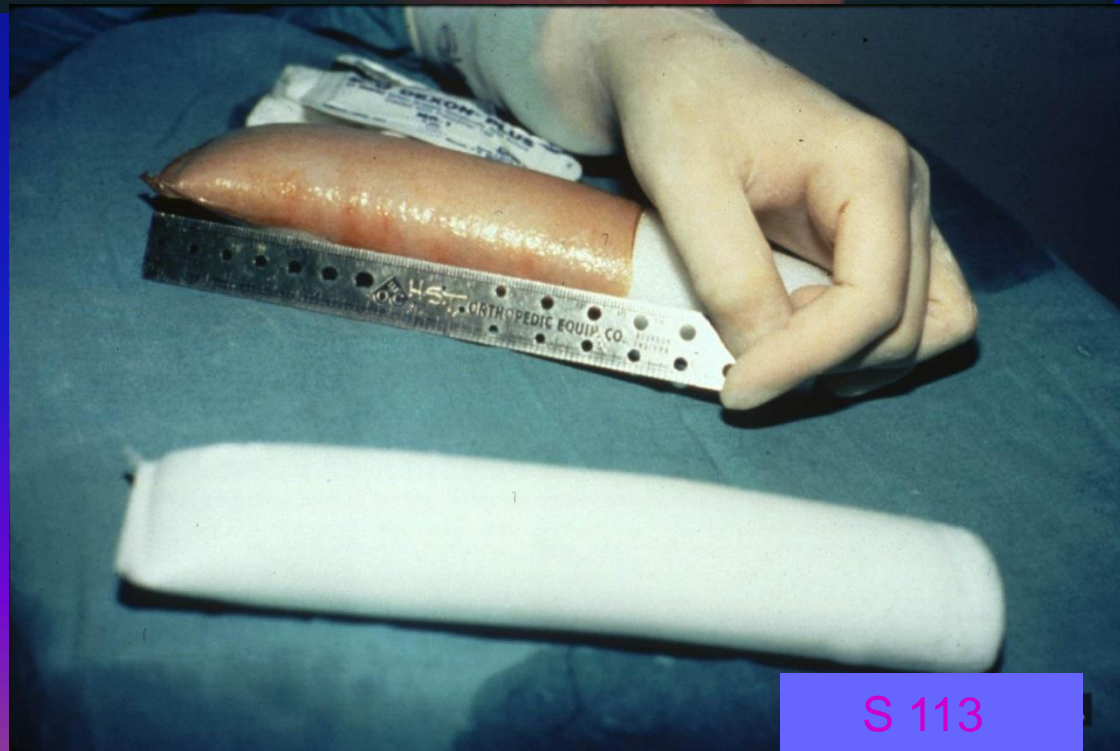
S 109

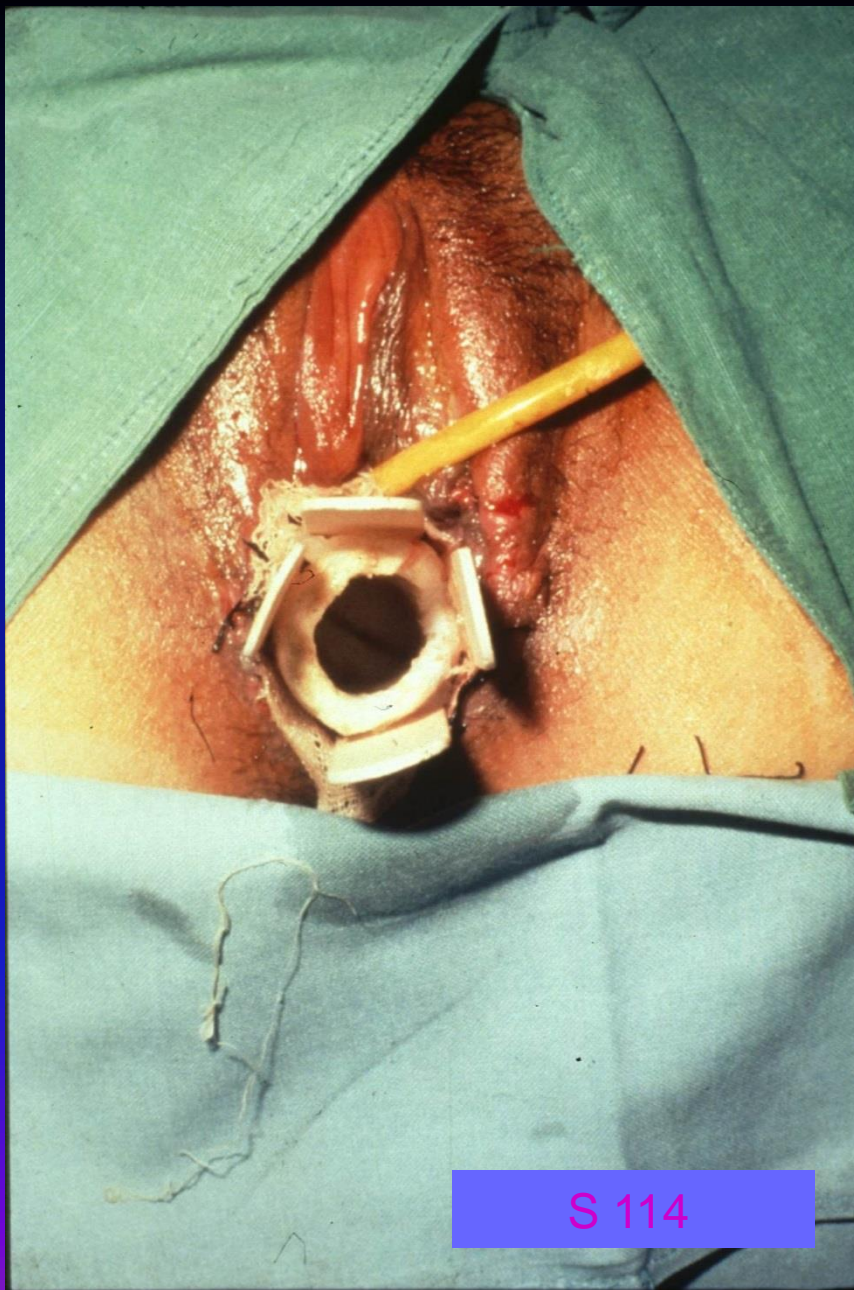


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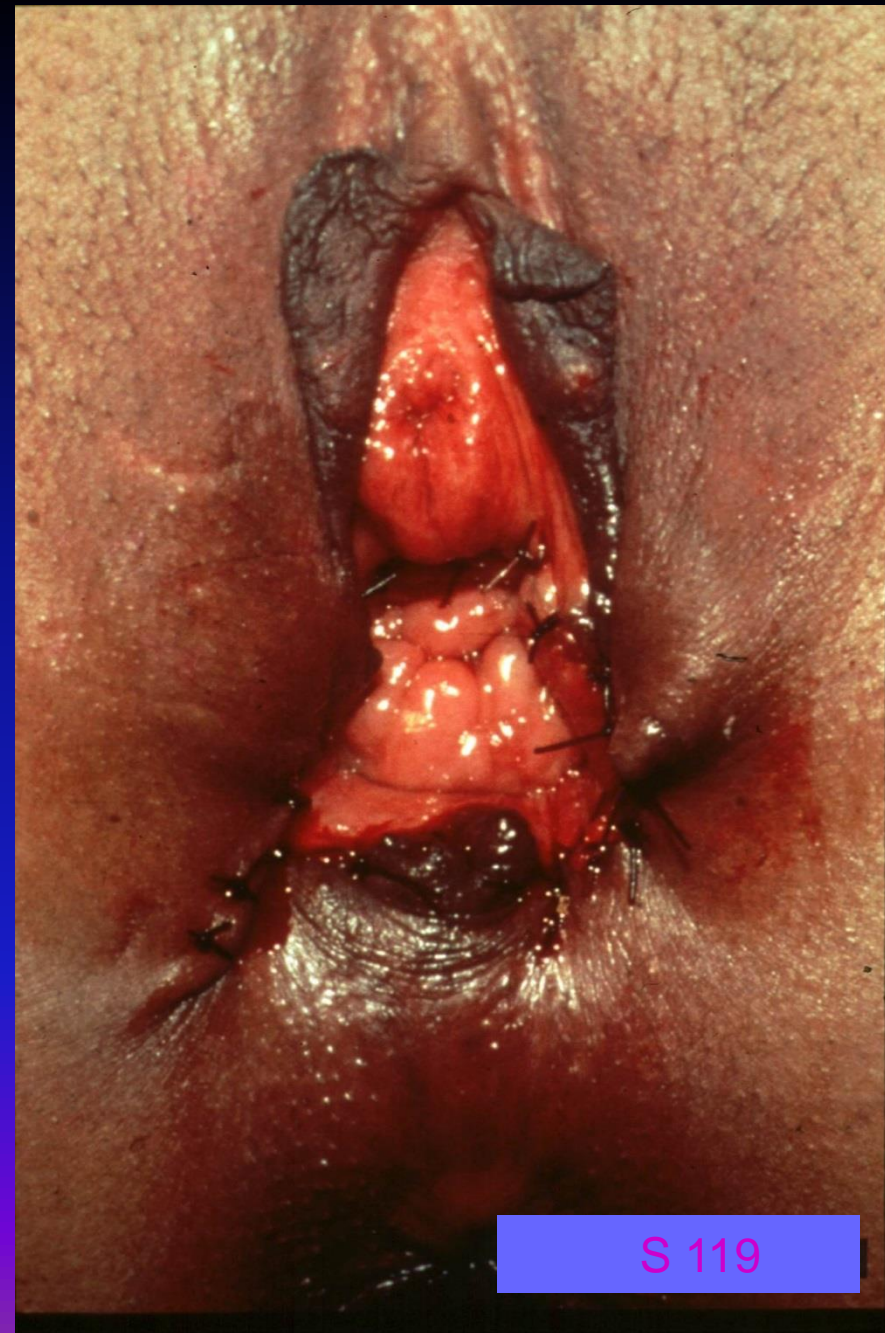
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s120

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

↑ Incidence of :

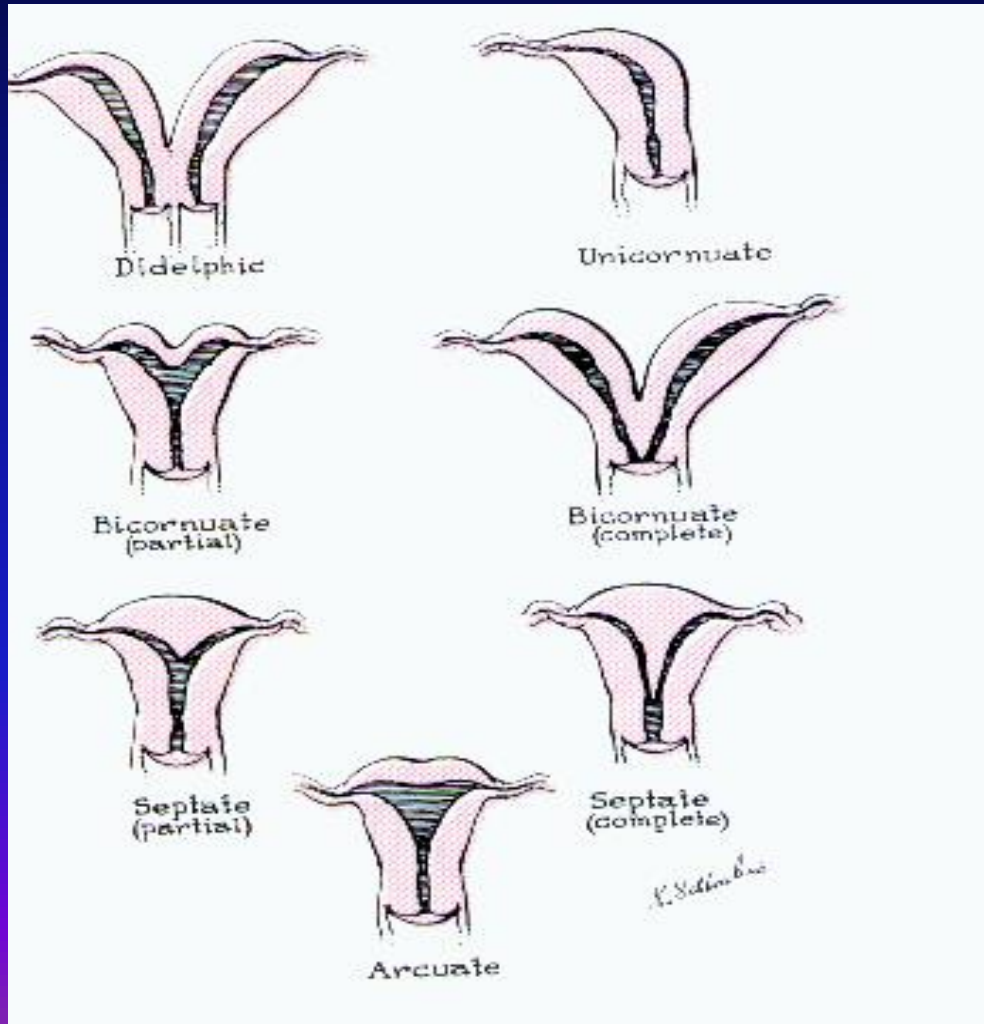
- ◇ recurrent abortions
- ◇ premature birth
- ◇ fetal loss
- ◇ fetal malpresentation
- ◇ C S
- ◇ cx incompetence

2-DISORDERS OF LATERAL FUSION OF THE M D

CLINICAL PRESENTATION

- ♣ Shortly after menarche ⇒ if there is obstruction to uterine blood flow
- ♣ Difficulty in intercourse ⇒ 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)
- ? ↑ 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 MULLERIAN DUCTS

E-Unicornuate with rudimentary horn

Noncommunicating horn 90%

Present with cyclic pelvic pain, mass, ectopic pregnancy in the rud horn or endometriosis

Rx \Rightarrow surgical excision

Communicating horn

Present with ectopic pregnancy in the rud horn or \uparrow 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
- ↑ incidence of endometriosis
- Rx ⇒ surgical exision

B-Cx AGENISIS / DYSGENISIS

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

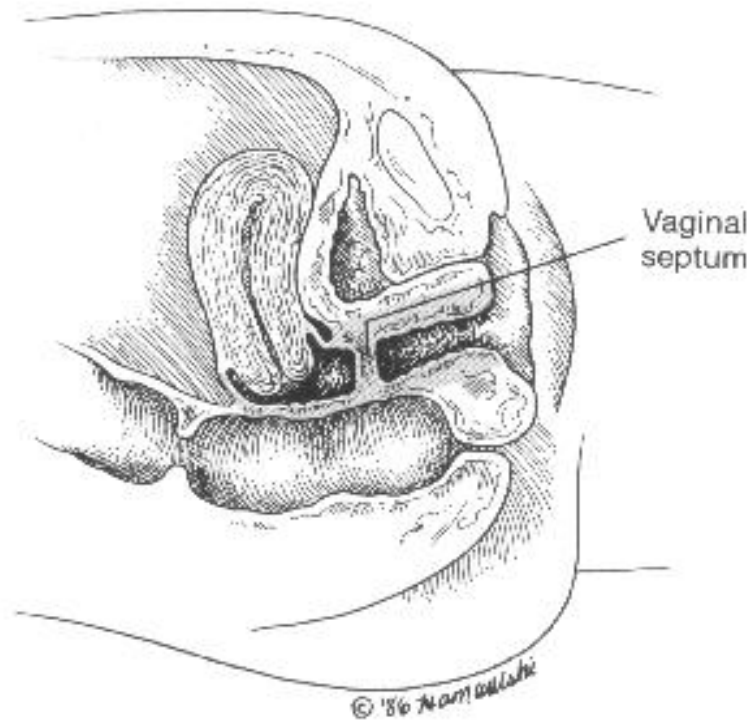
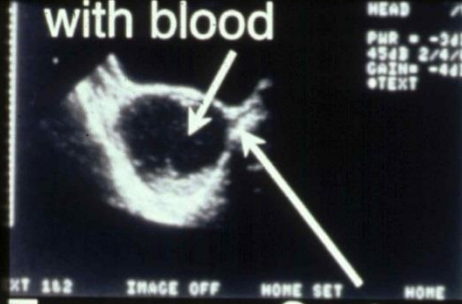


FIGURE 10-7
Diagram of transverse vaginal septum.

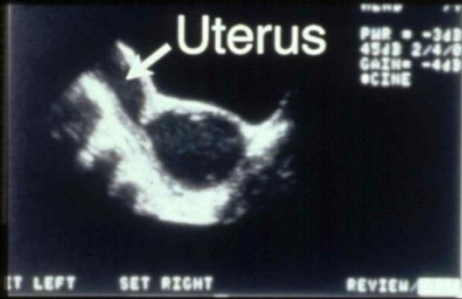
Vagina filled with blood



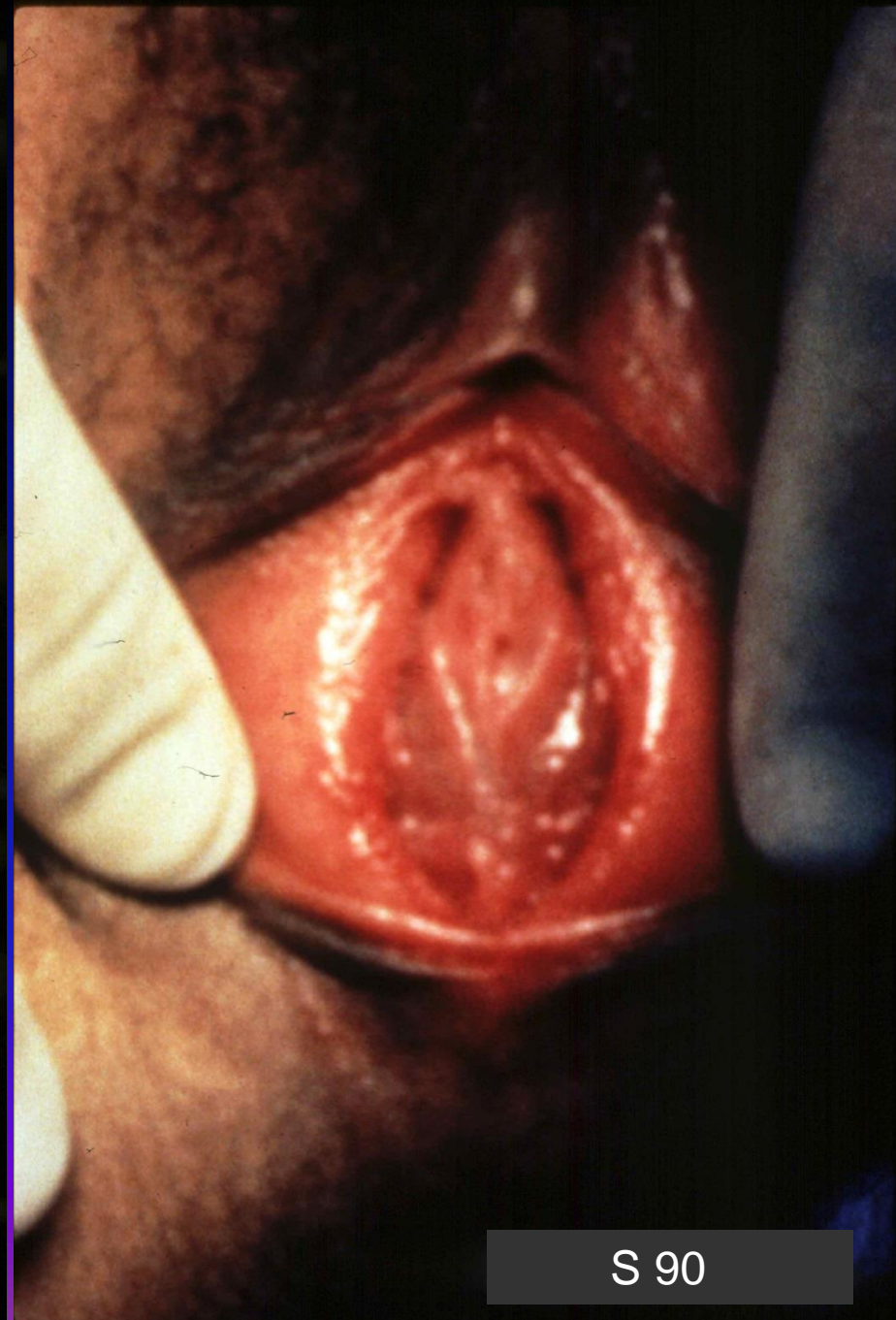
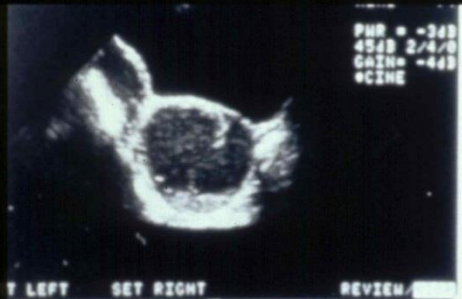
Endometrial Stripe

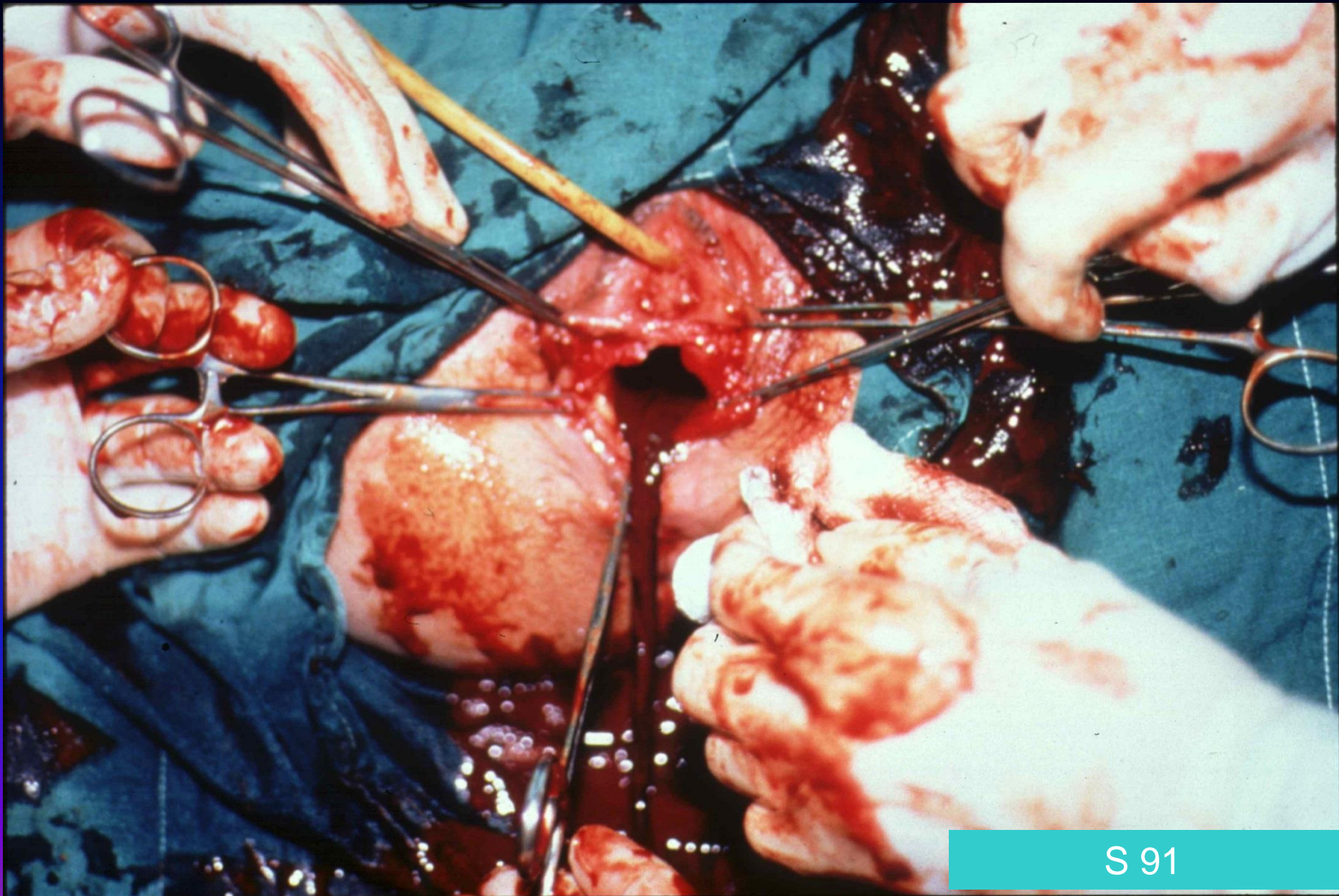


Transverse Septum



Debris and clotted blood in vagina



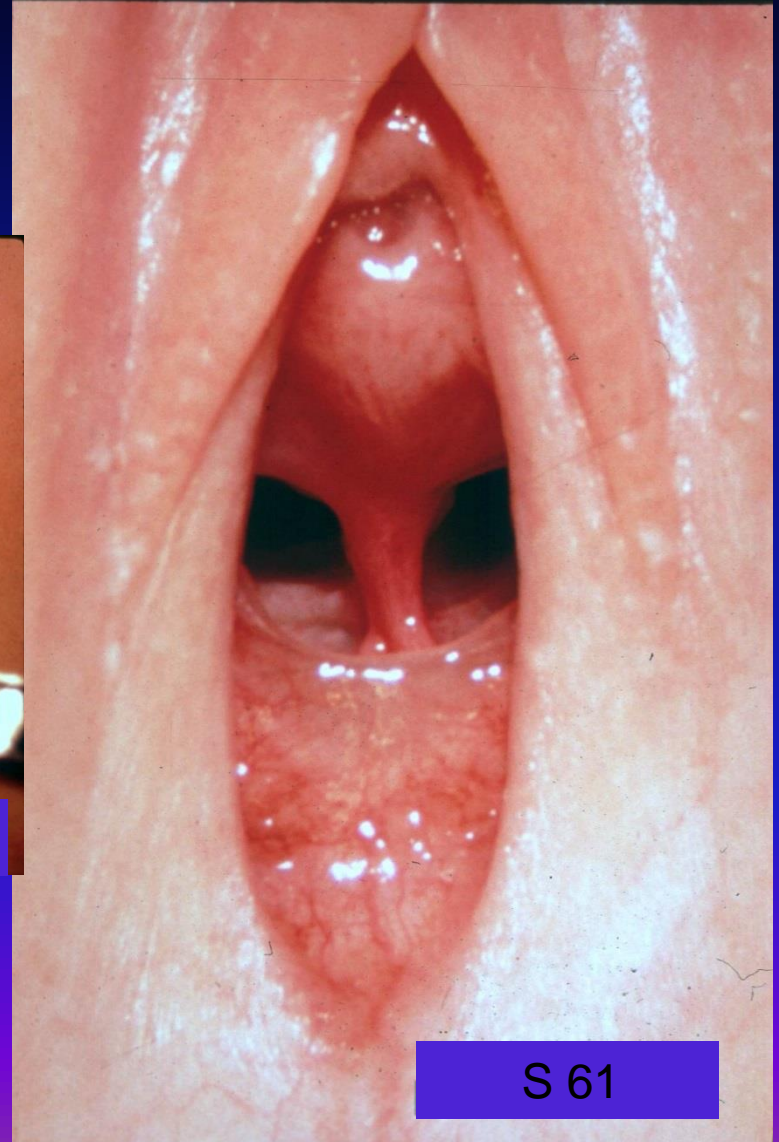


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Long vag septum



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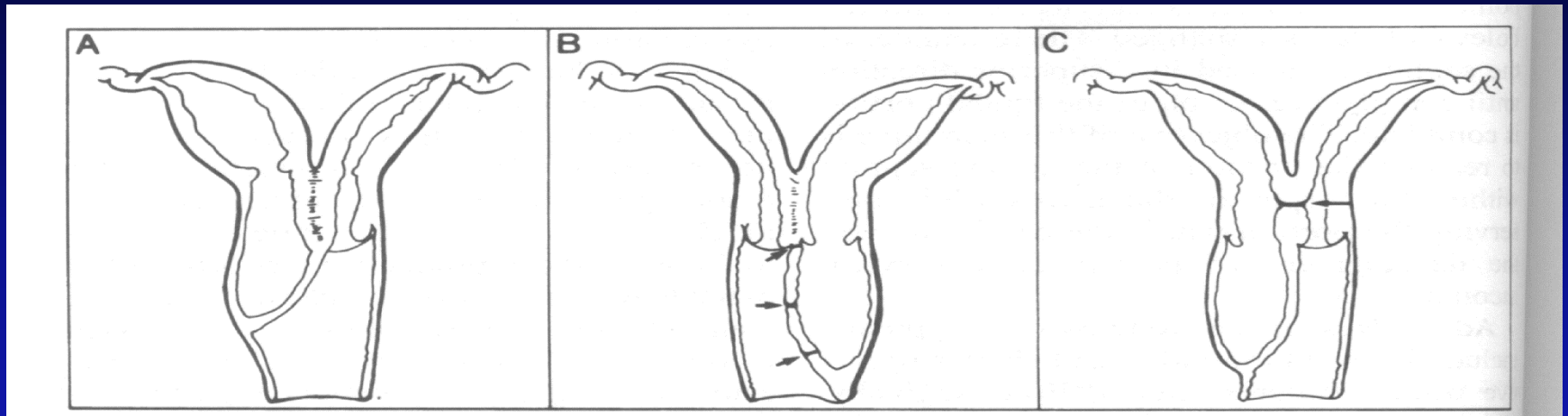


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4-UNUSUAL CONFIGURATION OF VERTICAL/LATERAL FUSION DEFECTS

- 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

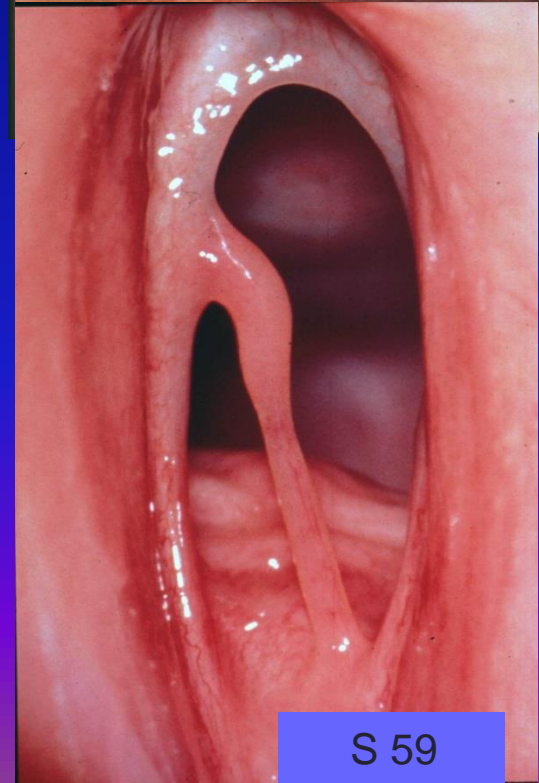
- Ambiguous genitalia ⇨ congenital adrenal hyperplasia
hermaphrodites
- Defects of the clitoris ⇨ uncommon ⇨ bifid clitoris
hypertrophied ⇨ androgen effect
- IMPERFORATE HYMEN
Hymen is formed at the junction of the urogenital sinus
& sinovaginal bulbs
Pt presents with 1ry amenorrhea with cyclic abdominal
pain or hematocolpus /hematometra
Rx ⇨ cruciate incision



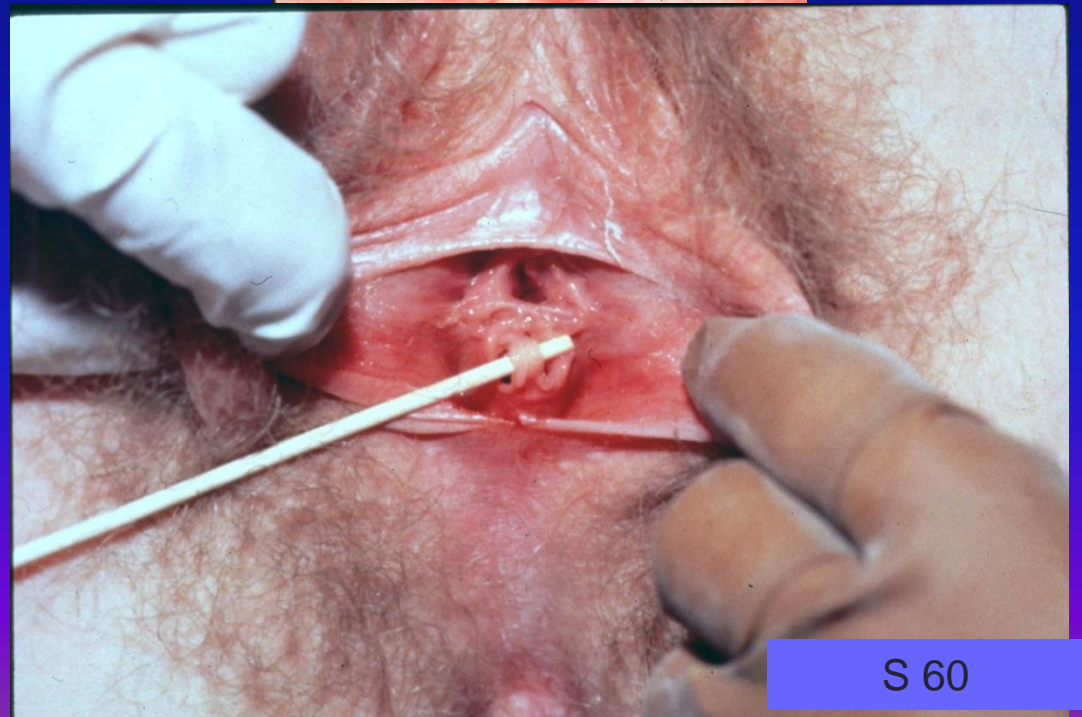
S 57



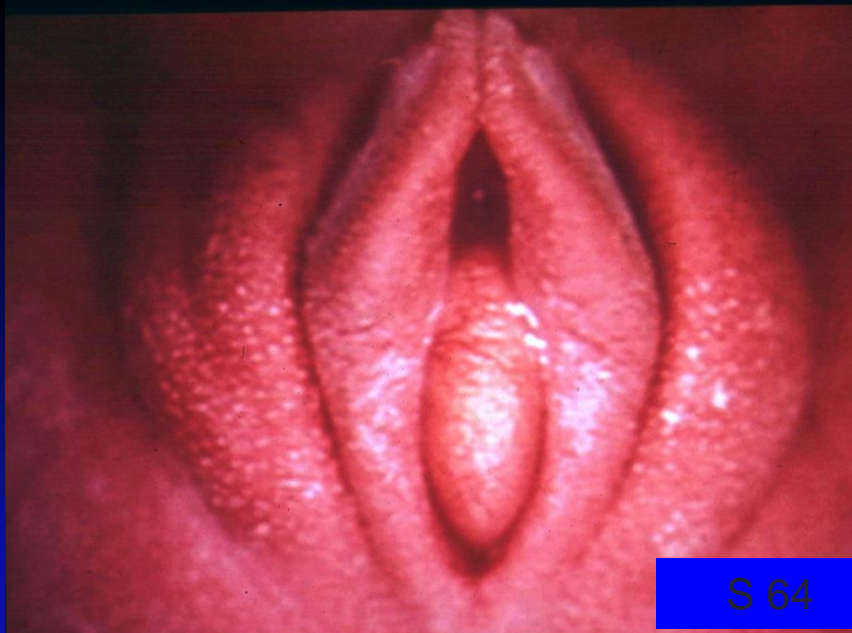
S 58



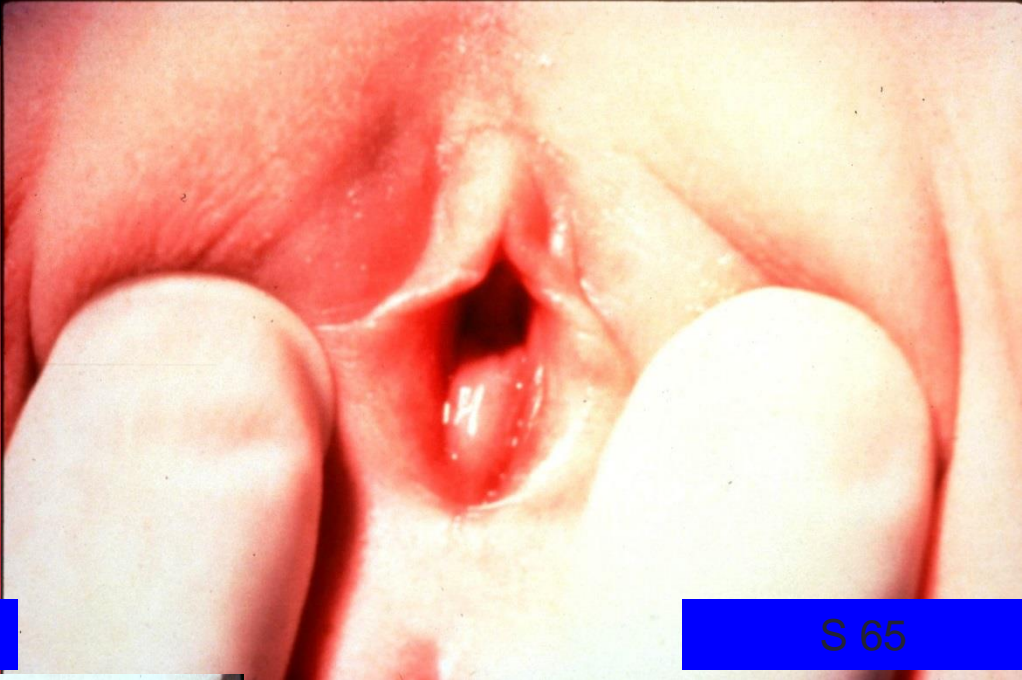
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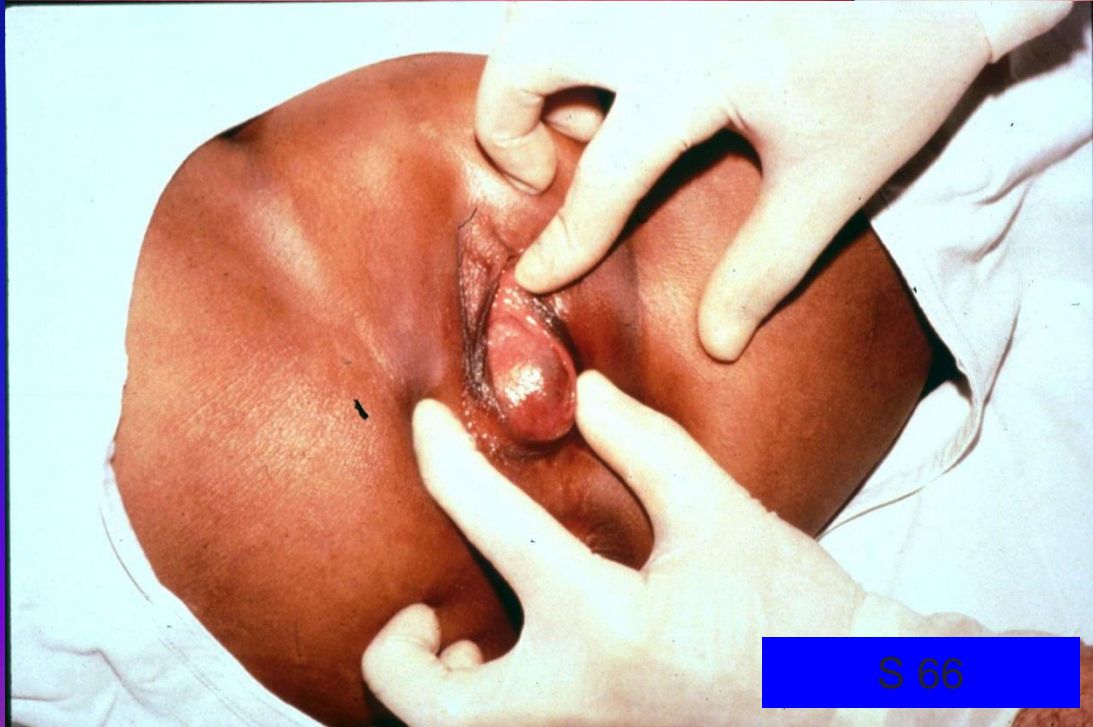
S 60



S 64



S 65



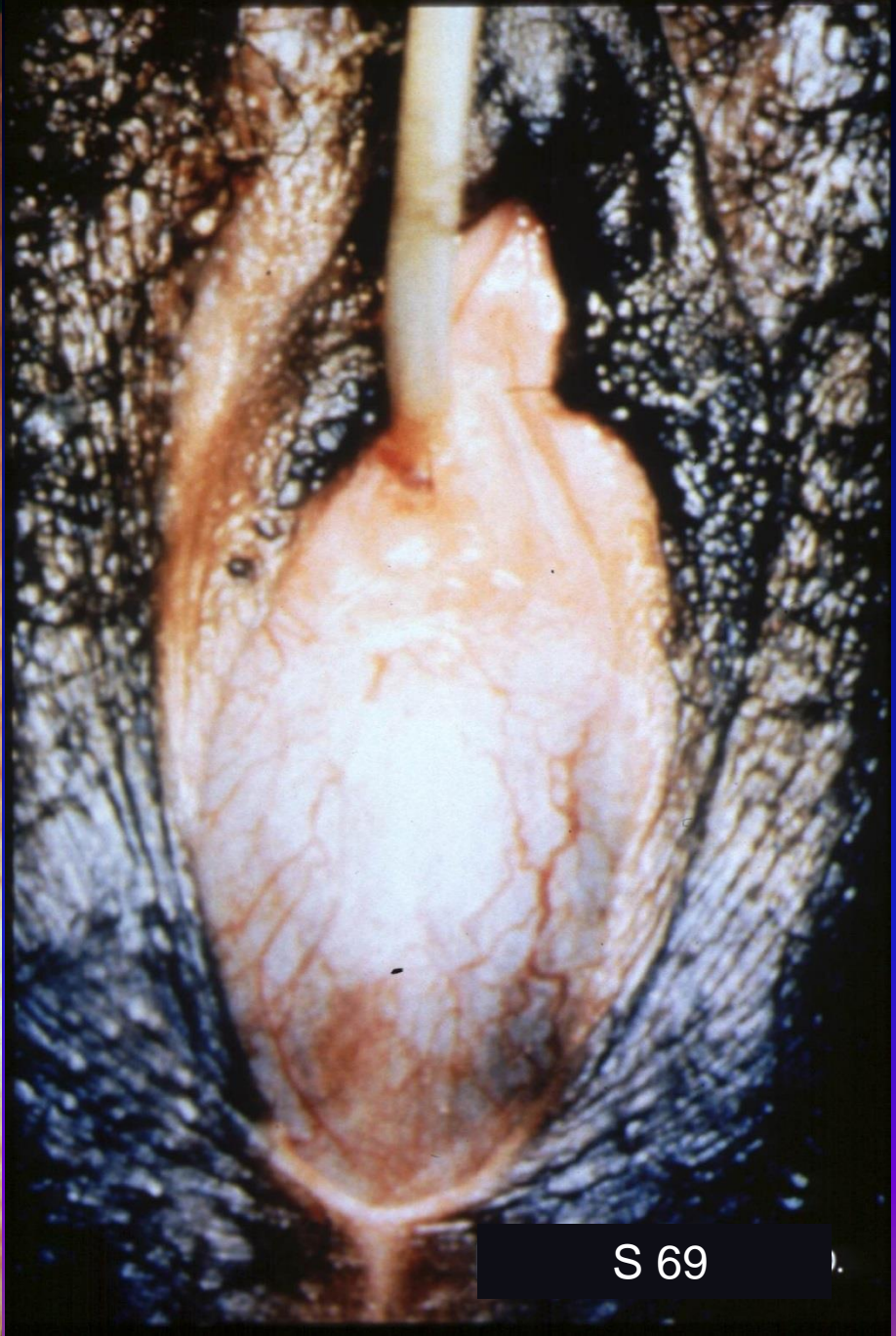
S 66



S 67



S 70



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