Pediatric and Adolescent Gynecology
UNIVERSITY OF COLORADO **ANSCHUTZ MEDICAL CAMPUS**

Children's Hospital Colorado

Obstetric and Gynecologic Considerations in Patients with ARM

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Gynecological concerns arise during ALL stages of life

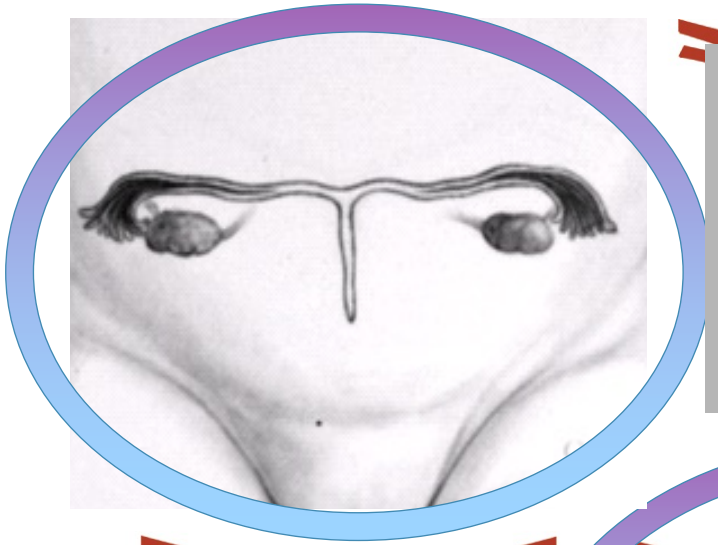
Infancy

Puberty & Menstruation

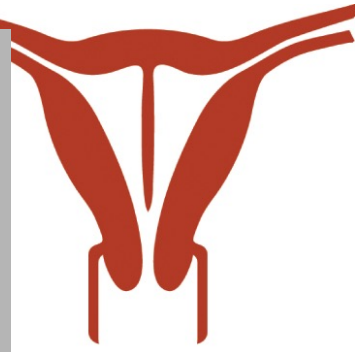
Sexual Intimacy

Fertility and Pregnancy

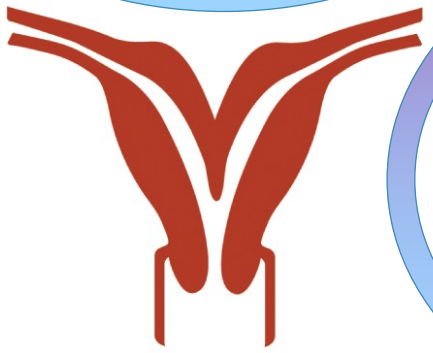
Mullerian Anomalies



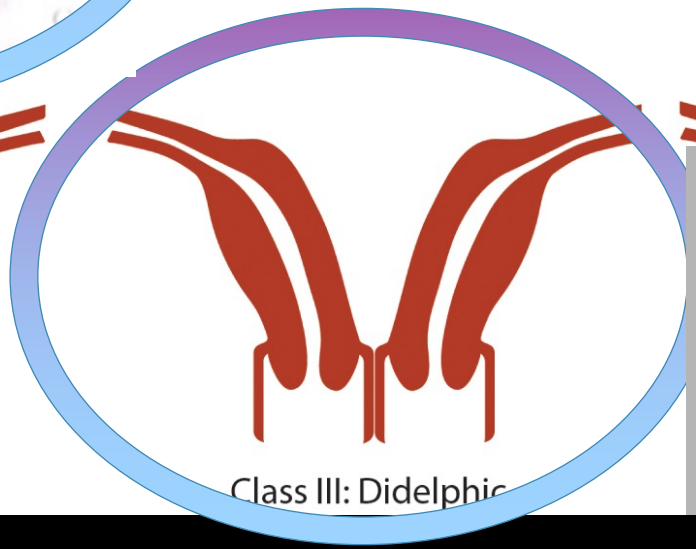
Vaginal Agenesis (5.6%)
- Cloaca 3.5 %
- Vestibular fistula 11 %
- Perineal fistula 1%



Class V: Septate



Class IV: Bicornuate



Class III: Didelphic

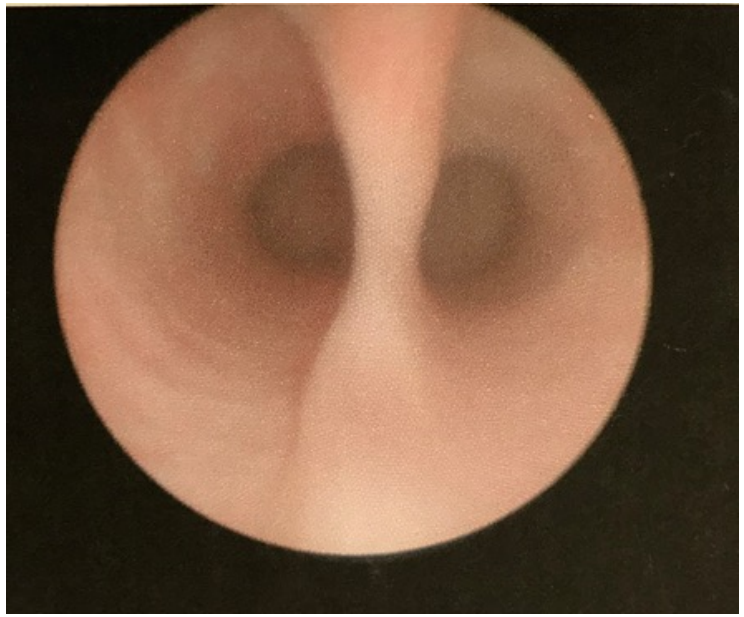
<u>Most Common</u>
Uterine Duplication (34%)
- Cloaca 60%
- Vestibular fistula 6%
- Perineal fistula 4%

What is the most common vaginal anomaly in patients with anorectal malformation?

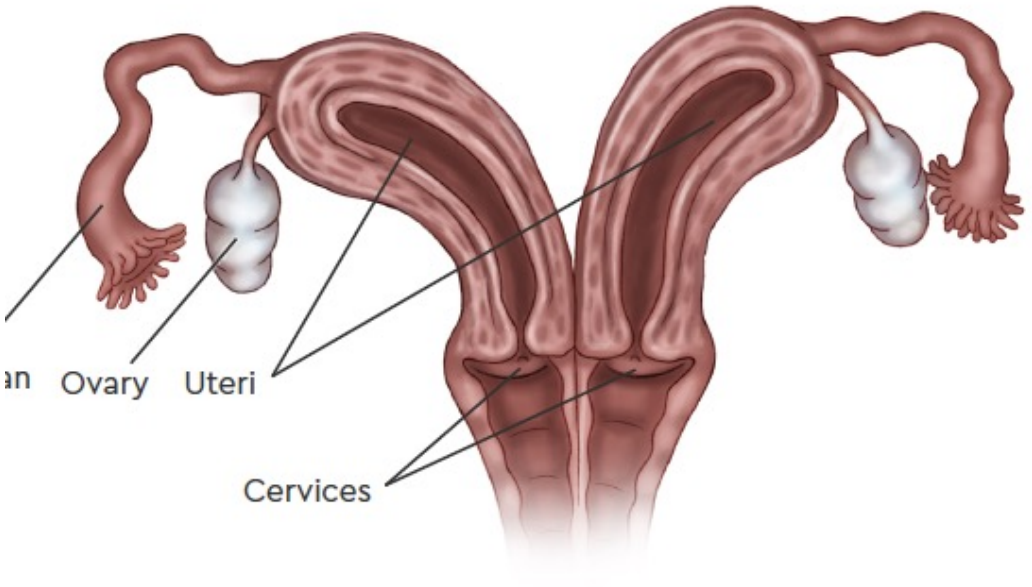
1. Vaginal atresia
2. Vaginal stenosis
3. Longitudinal vaginal septum
4. Transverse vaginal septum

Uterine and Vaginal Duplication

- Childhood – no implications
- Adolescence and Adulthood – dyspareunia and difficulty with menstrual hygiene
- Pregnancy - Didelphys uterus is associated with growth restriction, preterm labor/birth, fetal malpresentation and need for C-section

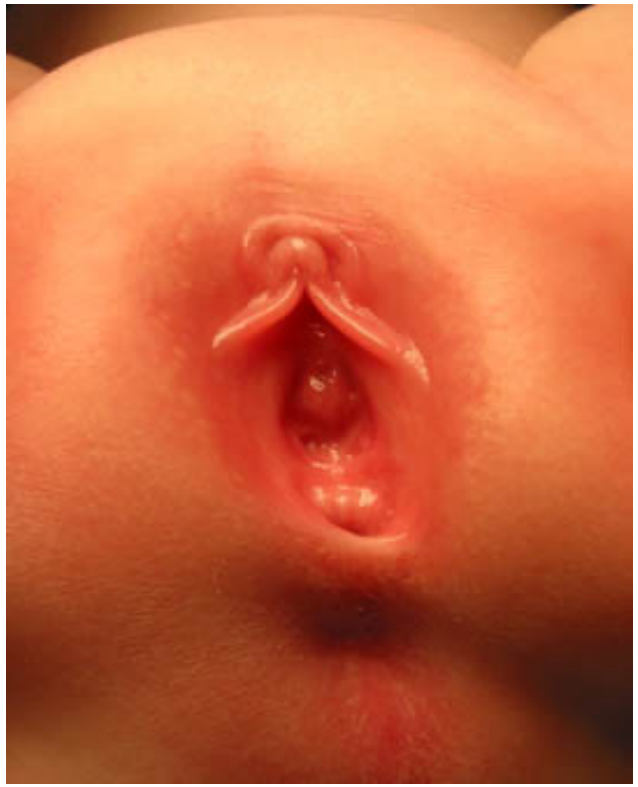


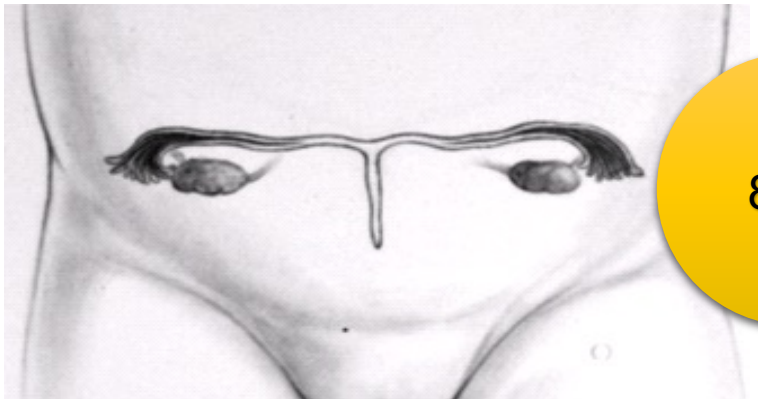
Pregnancy Surveillance

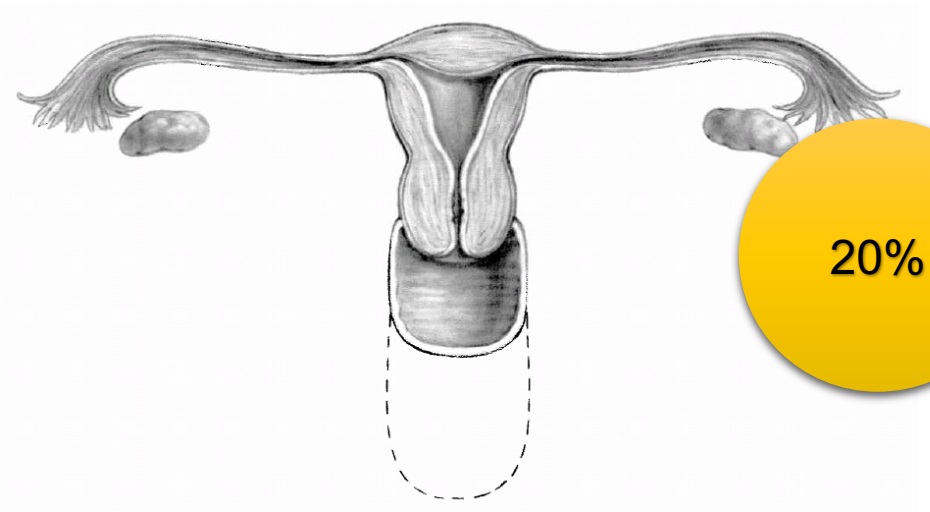


- Cervical length Q 2 weeks from 16-24 weeks gestation
- Fetal growth scans every 4 weeks

Vaginal Agenesis

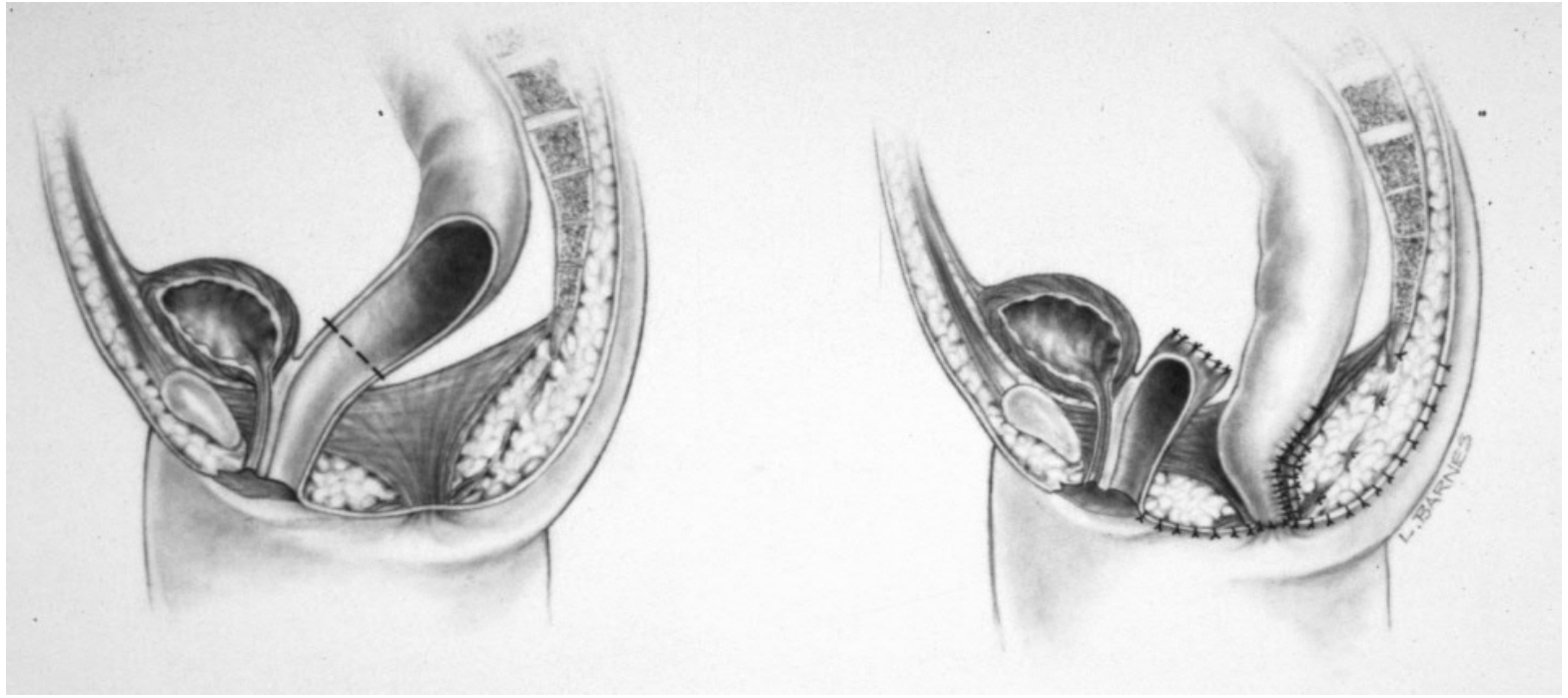






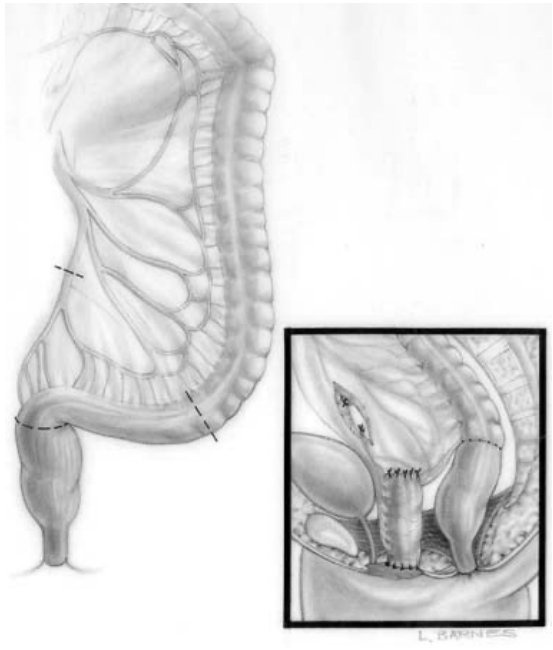
Vaginal Replacement for Patients with Vaginal Agenesis

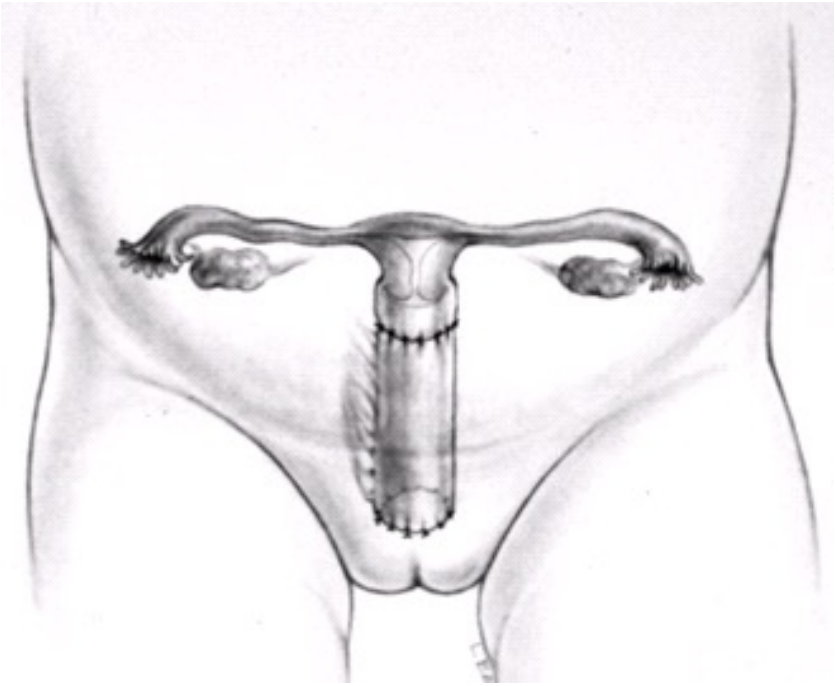
POOR Bowel Control Expected



Vaginal Replacement for Patients with Vaginal Agenesis

GOOD Bowel Control Expected





What is the frequency of vaginal agenesis in patients with anorectal malformation?

1. 80%
2. 60%
3. 40%
4. 20%
5. 5%

Considerations with NeoVagina

Benefits

- Usually good length and width
- Naturally lubricating
- Does not require routine dilation

Risks

- Excessive mucus production
- Stenosis
  - Introital (mild)
  - Anastomosis
- Prolapse
- Diversion colitis & IBD

Additional Considerations

- No cases of successful pregnancy

- Cases of dysplasia and adenocarcinoma are reported
  - Any bleeding, pain, nodules or stenosis should be evaluated
    - Exam
    - Vaginoscopy
    - Biopsy

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ORIGINAL ARTICLE

Neovagina stricture complicated by high-grade dysplasia in a patient with history of cloaca and ulcerative colitis: a case report and review of the literature

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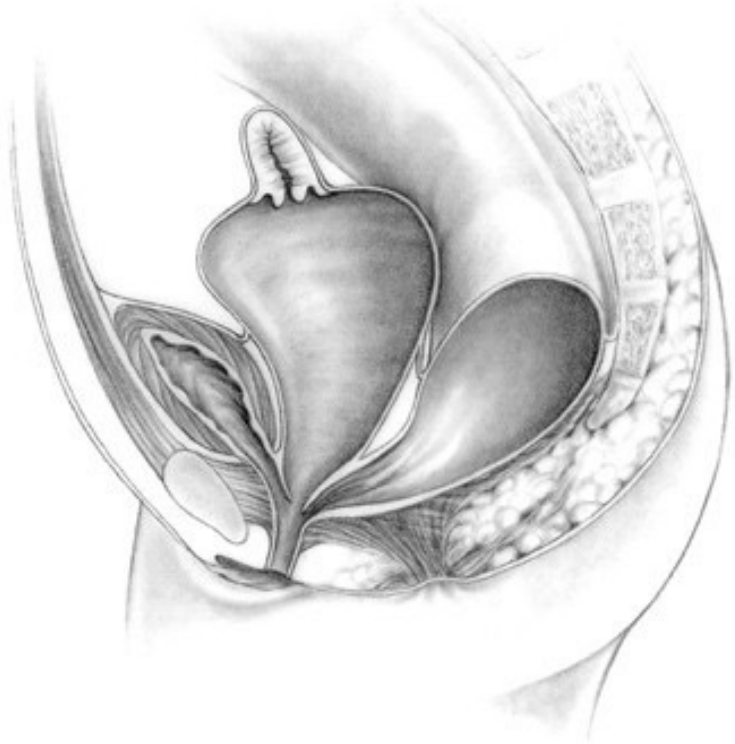
Abstract
Vaginoplasty with colon is a common technique for vaginal replacement in patients with cloaca. Malignancy in the neovagina is a rare outcome and typically presents decades after reconstruction. We present a case of an adolescent female with history of cloaca, ulcerative colitis, and high-grade dysplasia of the sigmoid neovagina.

Keywords Neovagina · Dysplasia · Ulcerative colitis · Vaginal stenosis · Vaginal stricture

Infancy

- Hydrocolpos
  - Distension of vagina(s) with fluid, urine and/or mucus
  - More common with uterine duplication and longer common channel
  - Can be identified antenatally



What is the incidence of hydrocolpos in patients with cloaca?

1. 90%
2. 70%
3. 50%
4. 30%
5. 10%

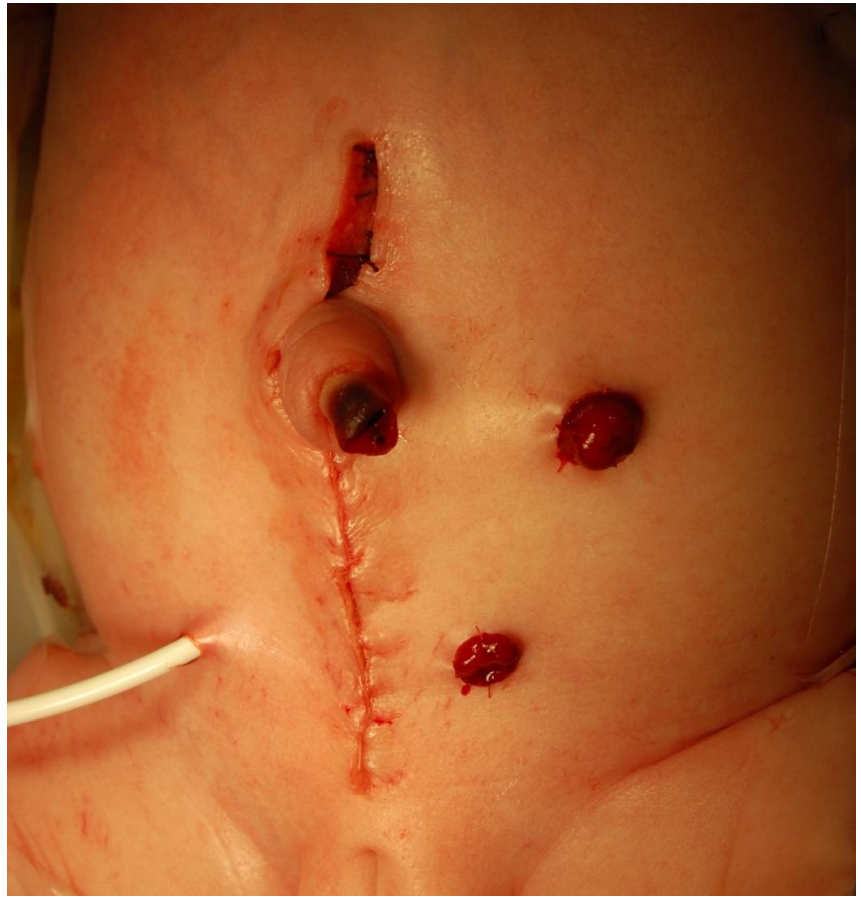
What are the indications to drain a hydrocolpos?

1. Compression of the ureters and hydronephrosis
2. Risk of infection
3. Drainage uncessary in asymptomatic patients
4. Answers 1 and 2

What is the preferred method of draining a hydrocolpos?

1. Serial dilation of the common channel
2. Placement of a catheter by endoscopy
3. Needle aspiration
4. Transabdominal catheter placement

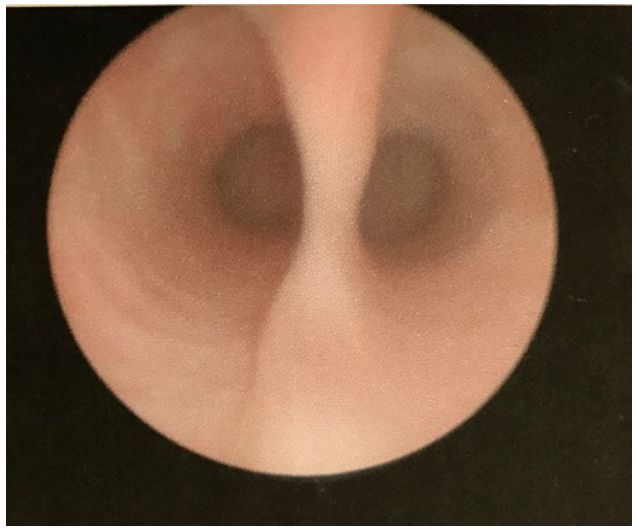
Hydrocolpos Treatment

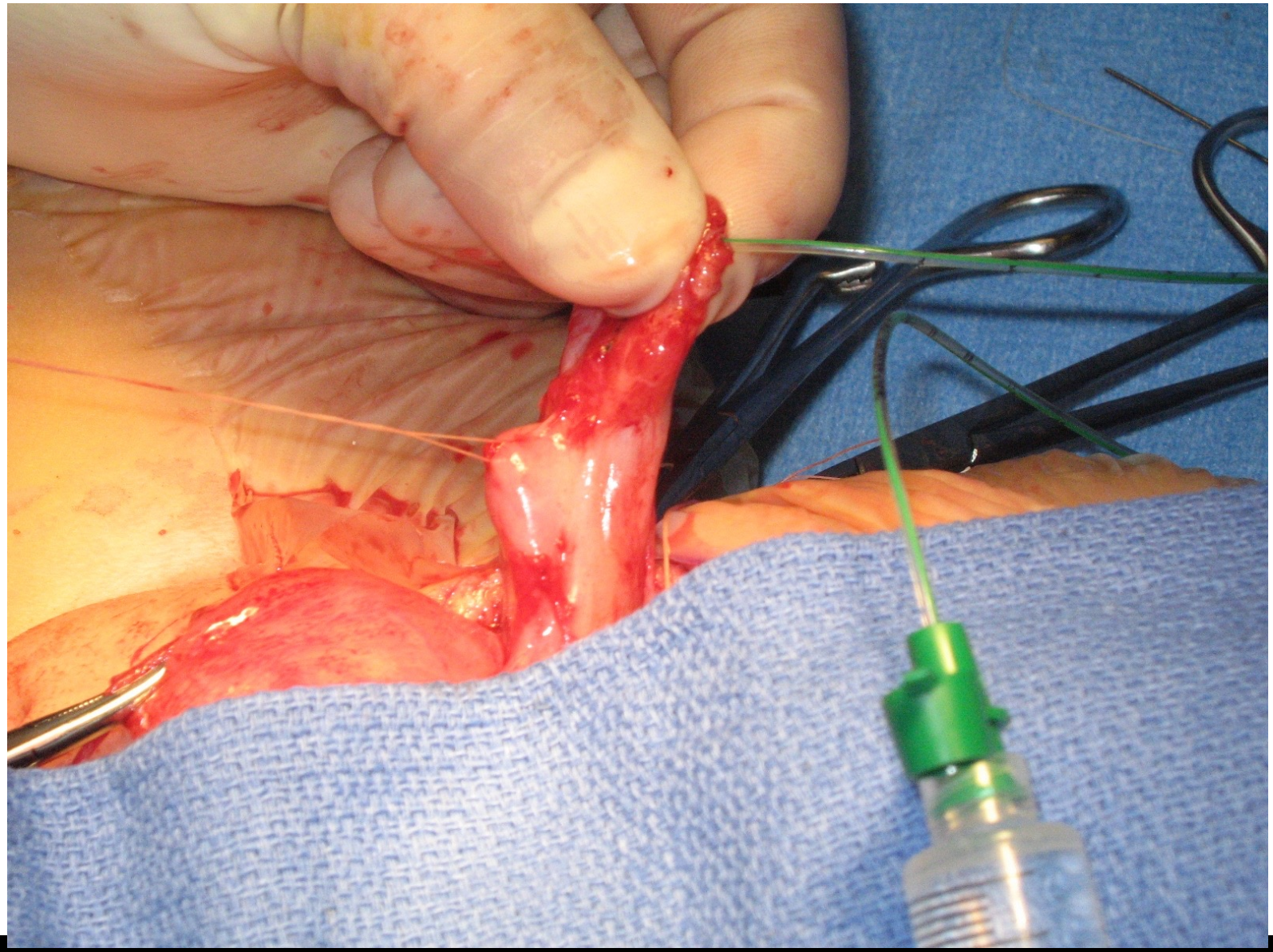


- Vaginostomy
- If a vaginal septum is present, a window is created to drain both vaginas

Evaluation of Gynecologic Anatomy

- At time of primary repair
  - EUA and Vaginoscopy
- With any abdominal surgery
  - Inspection
  - Checking patency



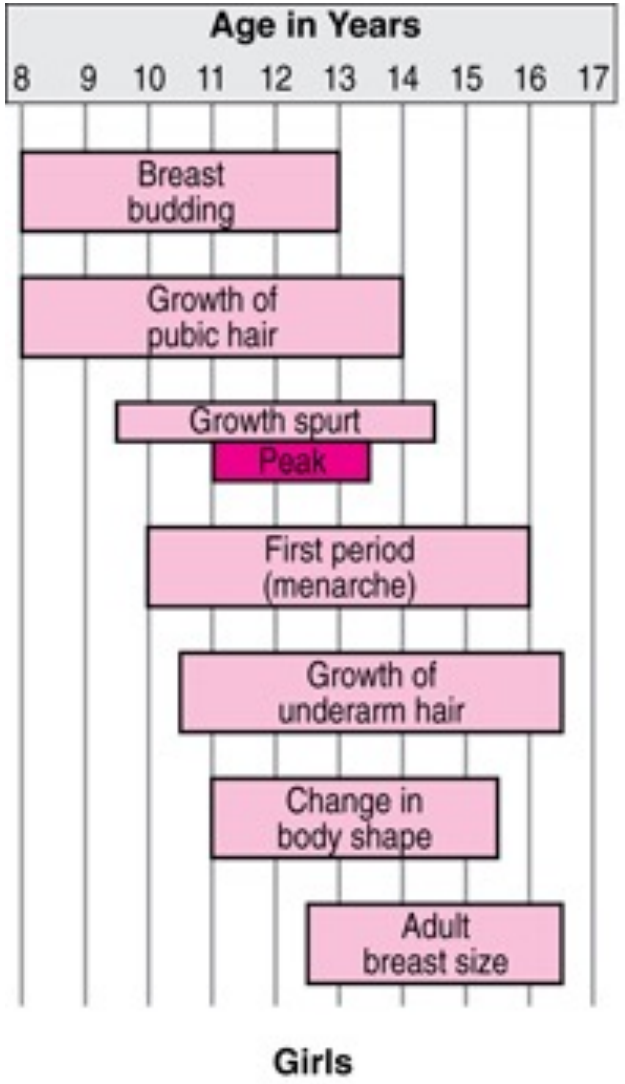


Infancy

- Determining gynecologic anatomy can be challenging
- With uncertainty, caution should be taken when considering removing uterine tissue

Puberty

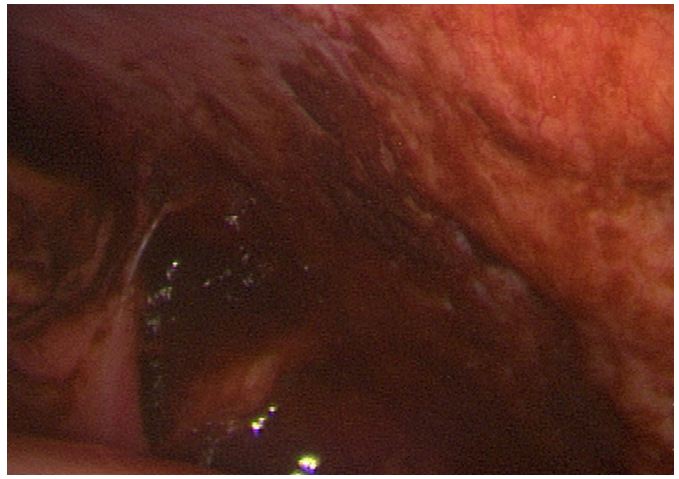
- Patients with ARM have normal ovaries
- Pubertal events occur as expected and follow a typical sequence
  - Menarche at age 12.4 (avg)

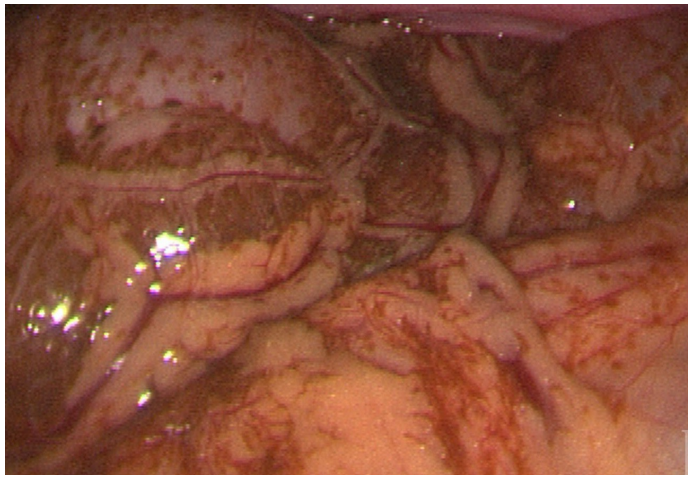


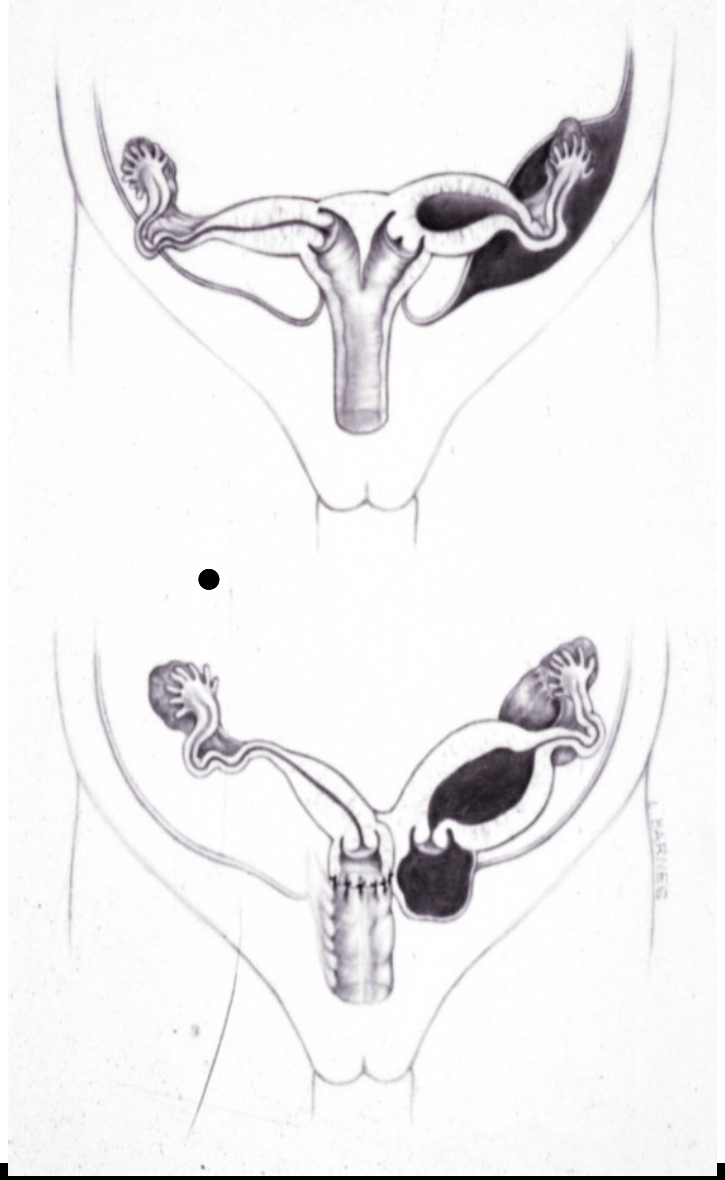
Girls

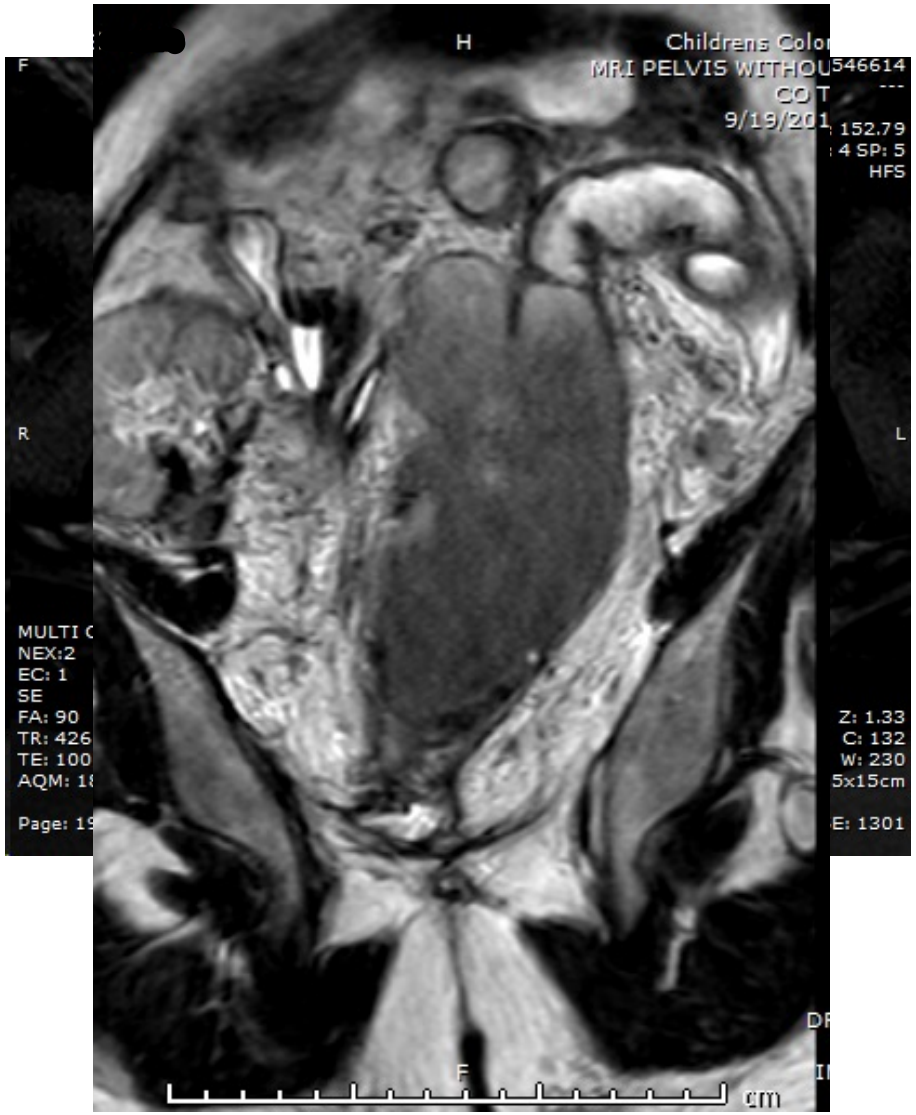
Puberty

- During puberty, focus is on menstruation
  - Amenorrhea: 20-30%
  - Obstructed menstruation: 36-41%
    - Congenital and acquired causes
    - Retrograde flow > cyclic abdominal pain > endometriosis





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MRI PELVIS WITHOU
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546614
152.79
4 SP: 5
HFS
R
L
MULTI
NEX:2
EC: 1
SE
FA: 90
TR: 426
TE: 100
Z: 1.33
C: 132
W: 230
5x15cm
E: 1301
cm

Assessment of Reproductive Structures

HOW?

- Ultrasound/MRI
- Vaginoscopy
- Saline pertubation of fallopian tubes
- Examination of introitus and perineal body

WHEN?

- At initial repair
- With any surgery
- **During puberty**
  - **6-12 months after thelarche**
- Pelvic pain

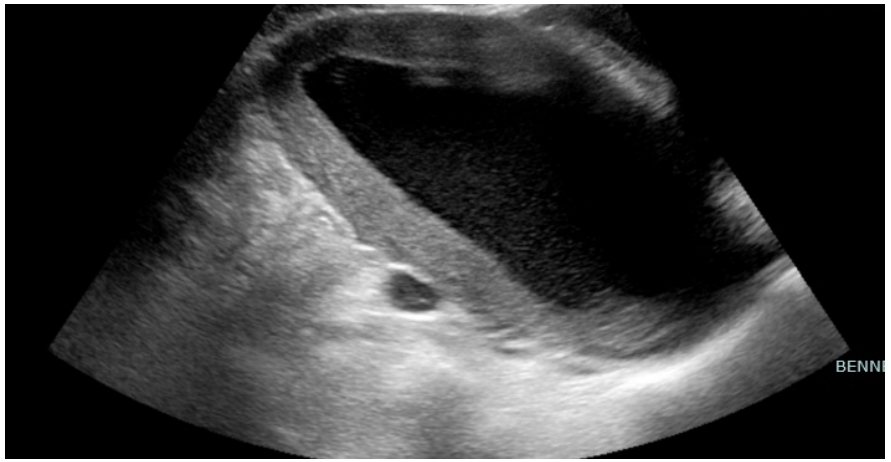
Treatment for Menstrual Obstruction

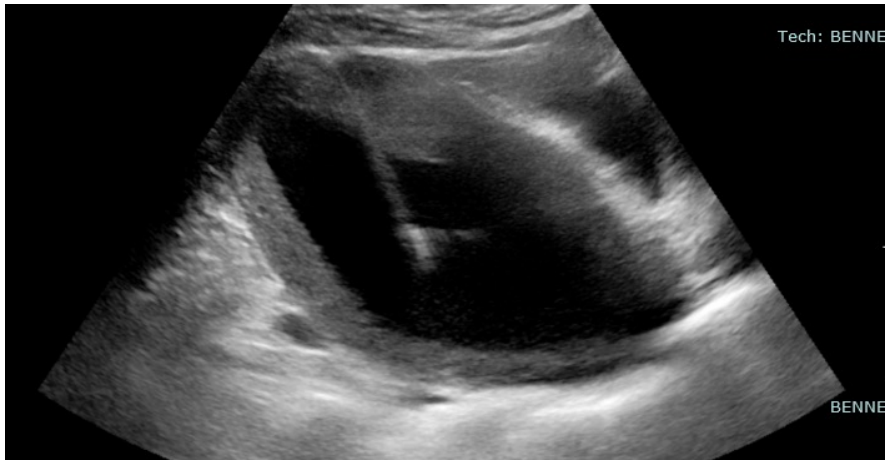
Pre Operative Strategies

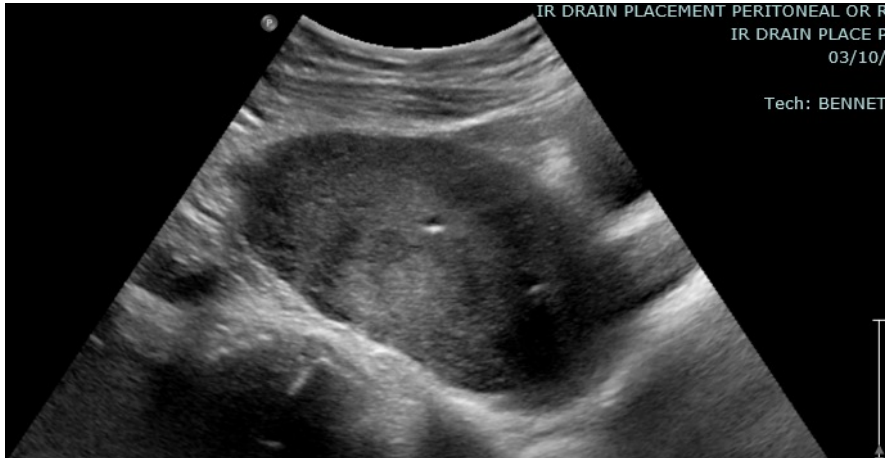
- Menstrual Suppression
- US Guided Drainage
- Vaginostomy

Definitive Surgical Management

- Vaginoplasty
  - Consider age of patient and need for post op dilation
- Septum resection
- In rare cases, hysterectomy





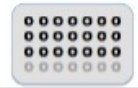


Ultrasound Guided Drainage

- Typically performed by Interventional Radiology
- Percutaneous approach preferred
- Tissue plasminogen activator (TPA) to break up clot

Puberty

- Hormonal suppression of menses
- Obstructed menstruation
- Dysmenorrhea
- Heavy menstrual bleeding
- Hygiene concerns

Treatment Choice
Norethindrone Acetate 
Pill 
Vaginal Ring 
Skin Patch 
Depo-Provera 
Progestin IUD 
Implant 

Sexual Function

- Factors affecting sexual function
- Anatomy
- Relationship status
- Medical comorbidities
  - Fecal or urinary incontinence
  - Spinal cord anomalies
  - Adhesive disease
- Psychosocial factors
- Body image

Sexual Function

- Introital or vaginal stenosis
  - Vaginal dilation
  - Introitoplasty
  - Distal vaginal replacement

- 20-50% of patients need a second (or third) vaginal surgery during adolescence or adulthood

Sexual Function

- Outcome data is VERY limited
  - Schmidt et al. 23 Females with ARM (18-56 yo)
    - 65% had sexual intercourse
      - 63% of those reported dyspareunia
  - Kyrklund et al. 10 Females with vestibular/perineal fistula
    - No difference in experience with intercourse, # stable relationships, and orgasm. Coital debut delayed
  - Studies are in progress!

Fertility

- Outcome data is limited
- Spontaneous conception is possible
- Females with complex ARM appear to have lower child birth rates
  - Mullerian anomalies
  - Iatrogenic damage
  - Adhesive disease related to prior surgeries or endometriosis
  - Psychosocial factors

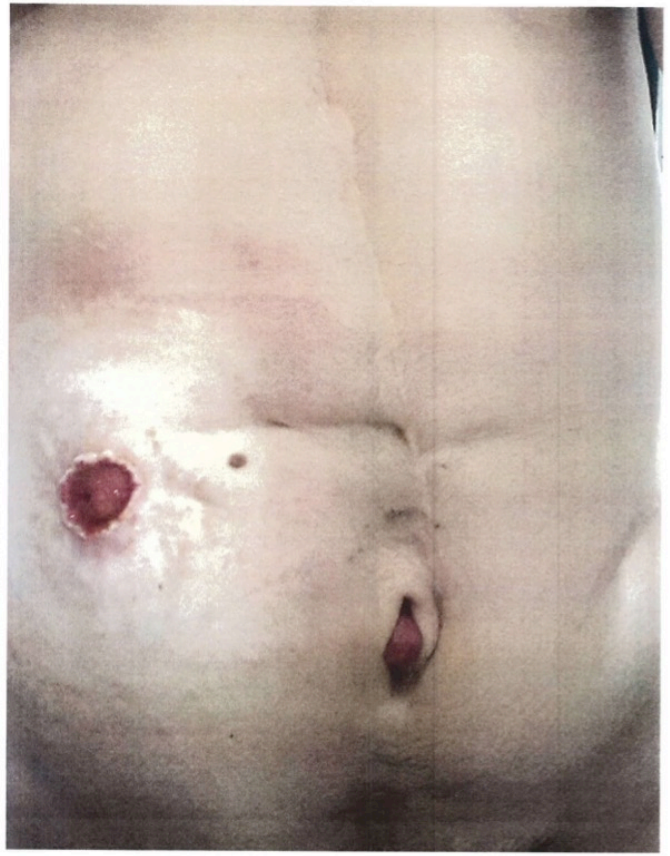
Fertility

- Evaluate anatomy early and often
- Prevent menstrual obstruction
- Work up for infertility and refer to a specialist

Should we consider fertility preservation in cases of complex ARM?

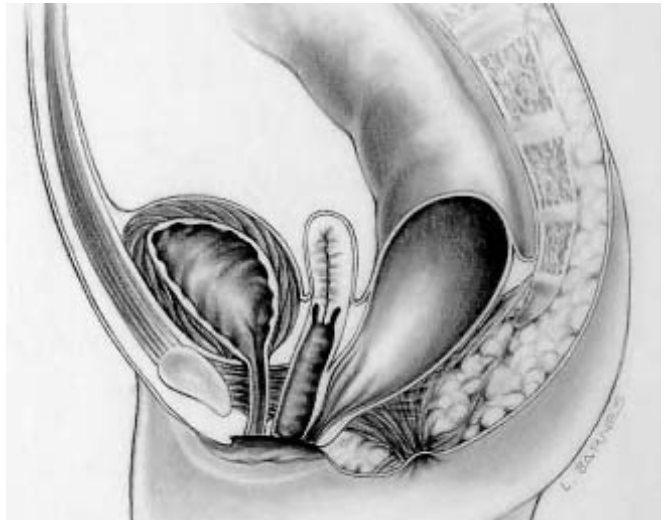
Pregnancy

- Patients need preconception counseling
  - Multiple co morbidities
    - Cardiac
    - Gastrointestinal
    - Spinal and vertebral
    - Renal anomalies and kidney disease
  - Mullerian anomalies are common
  - Complicated surgical history



A 32 year old female with history of rectovestibular fistula is pregnant at 35 wk GA. She has good bowel control and an adequate perineal body What mode of delivery would you recommend?

1. Vaginal delivery, avoid assisted vaginal delivery
2. Vaginal delivery or assisted vaginal delivery
3. C-section
4. I have no idea



Mode of Delivery

- Dependent on type of ARM and surgical history
  - Rectovestibular or rectoperineal fistula – probably okay for vaginal birth
    - Evaluate perineum
    - Minimize risk for sphincter injuries in patients with good bowel control
  - Cloaca or vaginal replacement – consider C-section

Unrepaired ARM in Adulthood

- Chronic and severe constipation
- Dyspareunia
- Injury with sexual activity

*The **mission** of **Pediatric and Adolescent Gynecology** is to promote and advance reproductive health through excellence in patient care, education, research and advocacy.*

*Our **vision** is to elevate reproductive health across the lifespan.*

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