Obstetric and Gynecologic Considerations in Patients with ARM

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Gynecological concerns arise during ALL stages of life

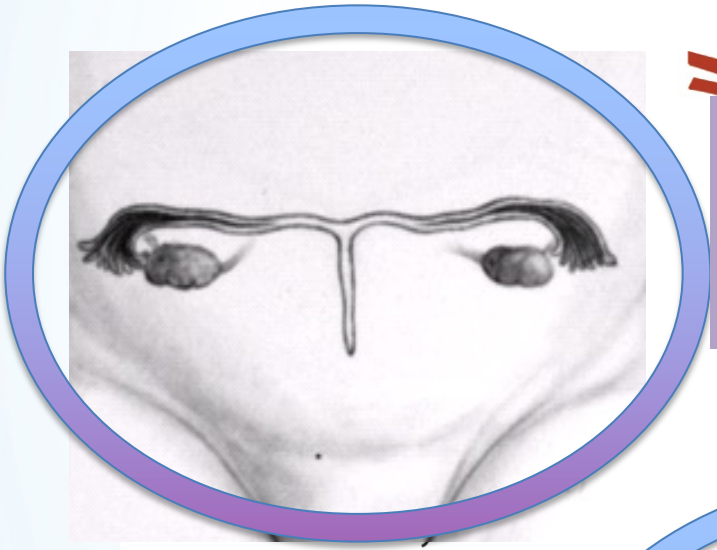
Infancy

Puberty

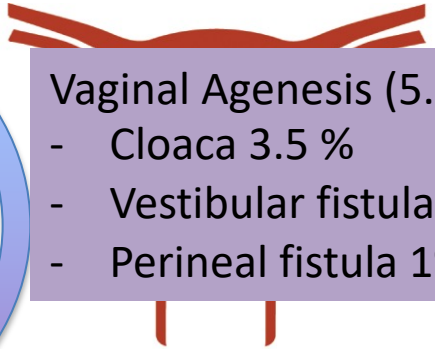
Sexual Intimacy

Obstetrics

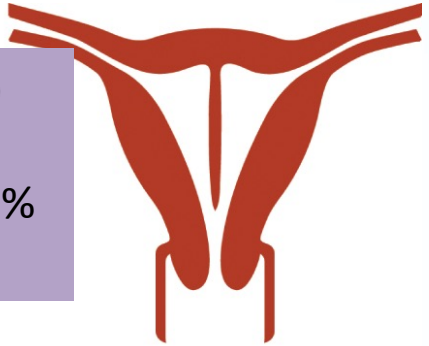
Uterine anomalies



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

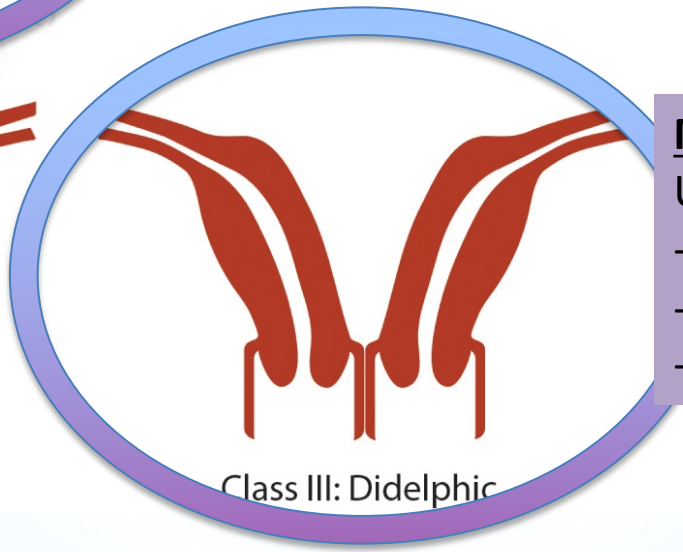


Class VI: Arcuate



Class V: Septate

Class IV: Bicornuate



Class III: Didelphic

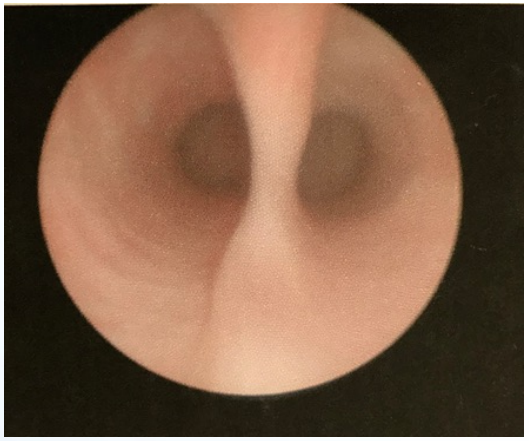
Class II: Unicornuate

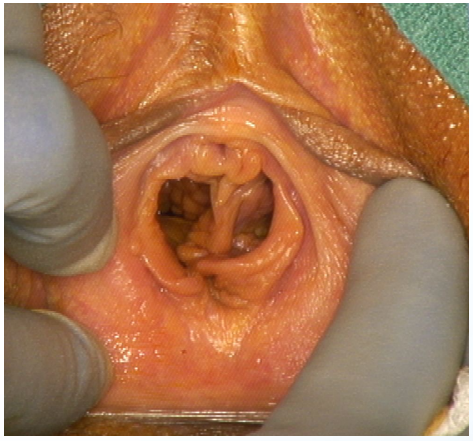
<u>Most Common</u>

Uterine Duplication (34%)
- Cloaca 60%
- Vestibular fistula 6%
- Perineal fistula 4%

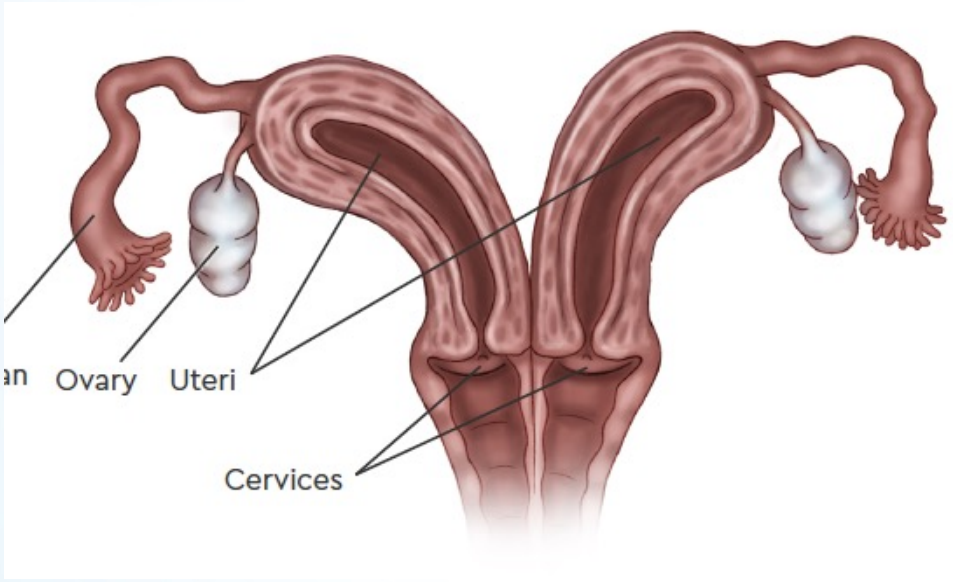
Uterine and Vaginal Duplication

- 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





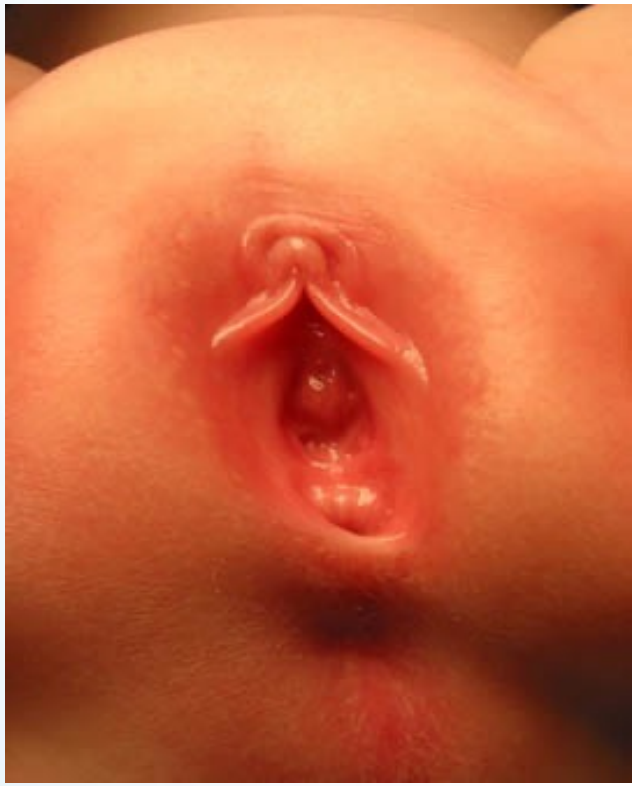
Pregnancy Surveillance

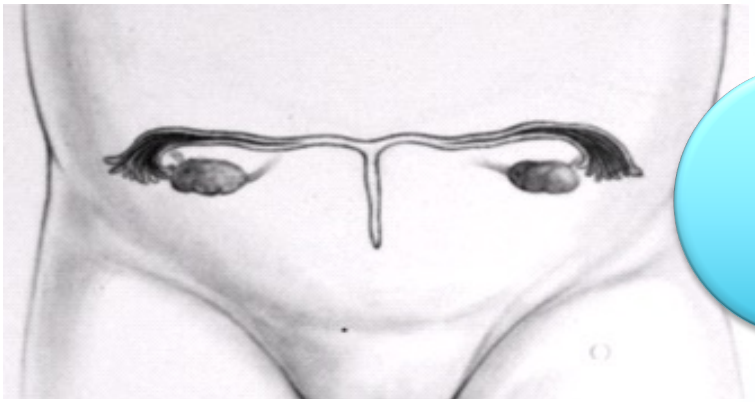


Cervical length Q 2 weeks from 16-24 weeks gestation

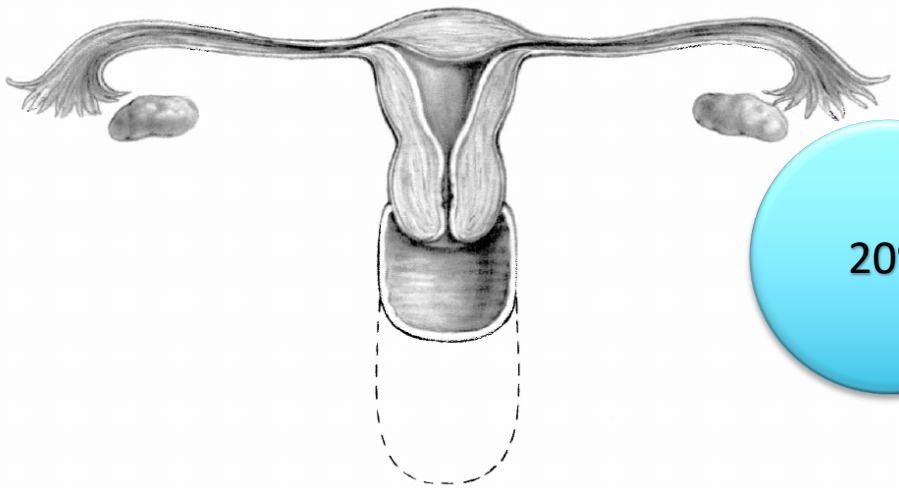
Fetal growth scans every 4 weeks

Vaginal Agenesis



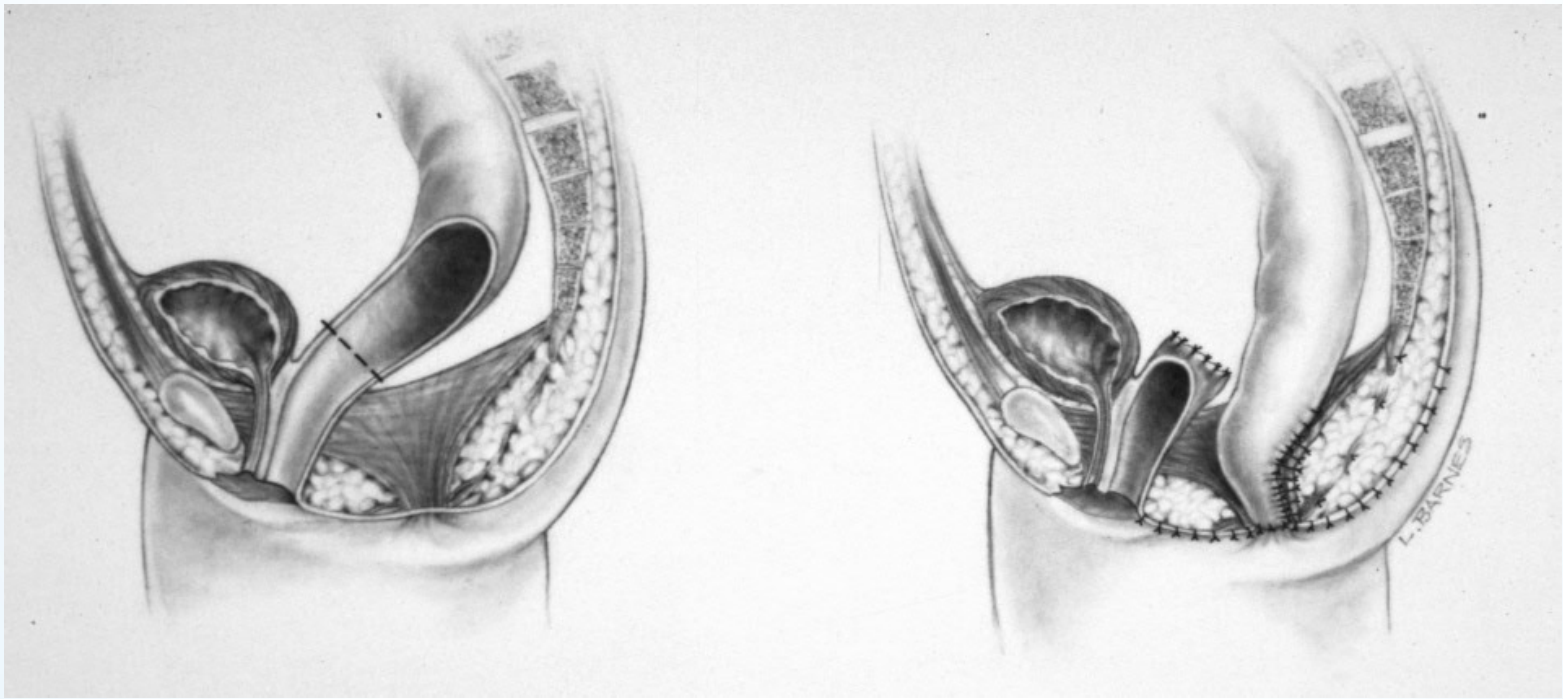


80%

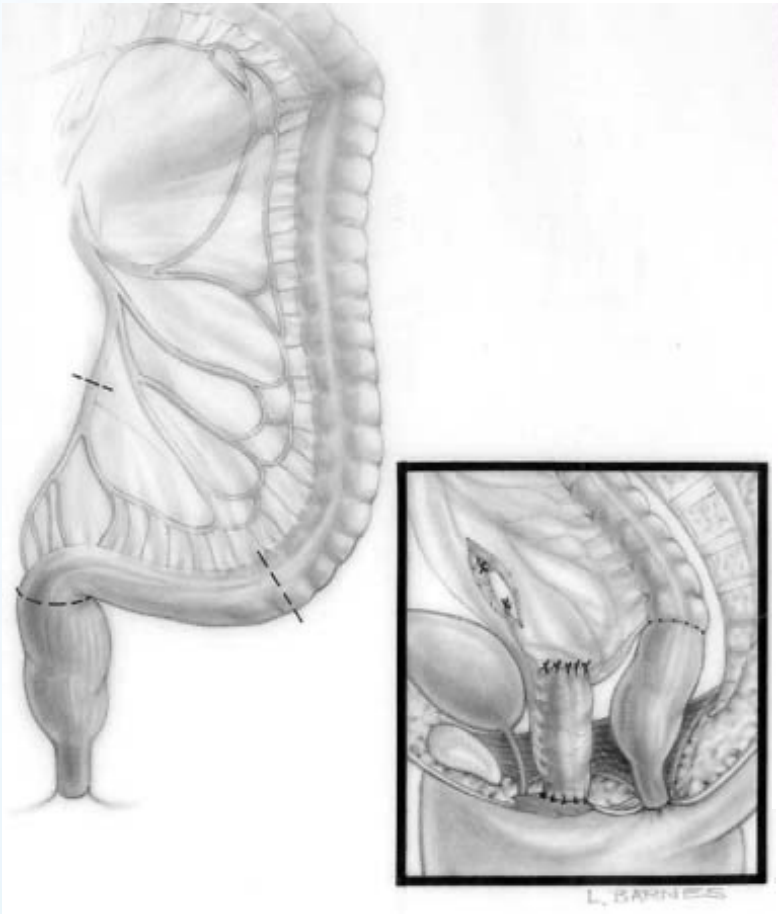


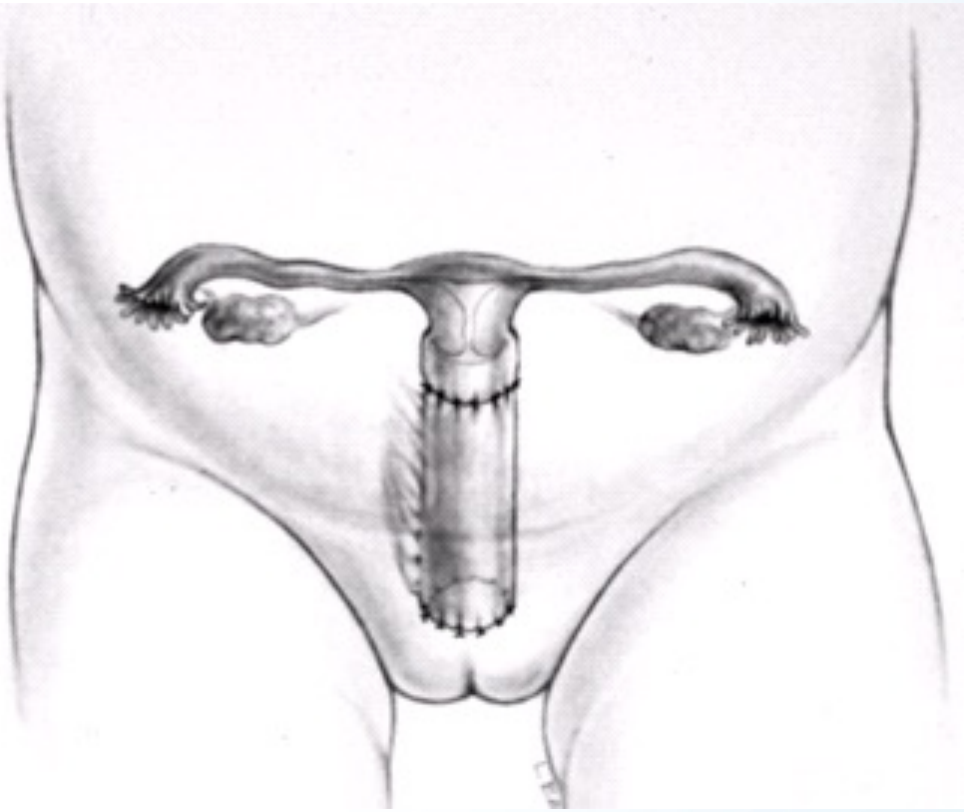
20%

Vaginal Replacement for Patients with Vaginal Agenesis

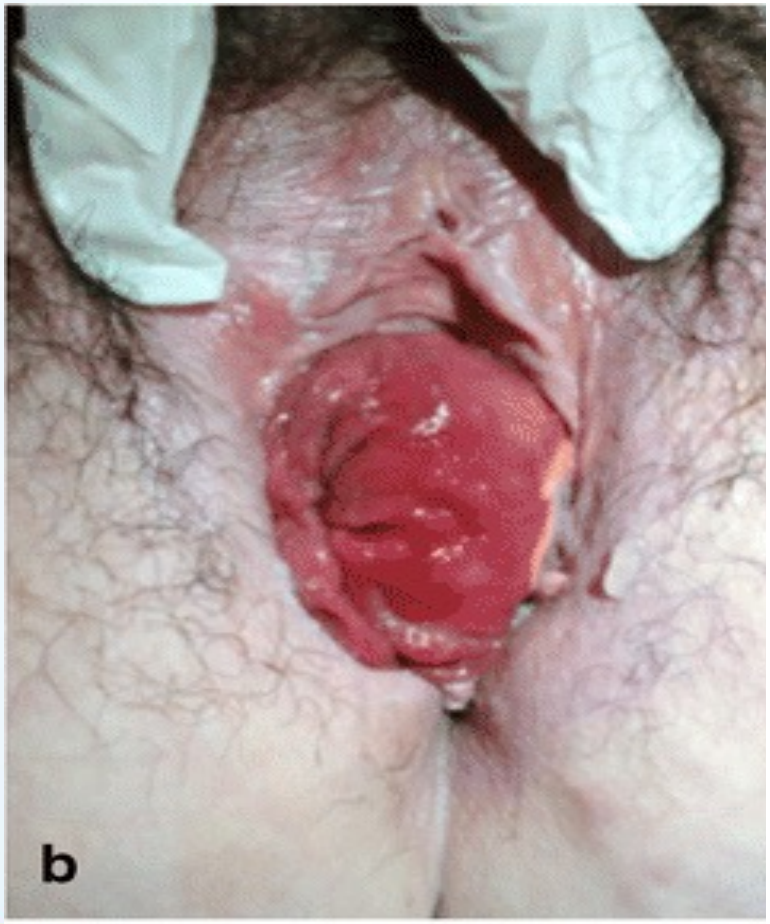


Vaginal Replacement for Patients with Vaginal Agenesis





Neo Vagina Risks



Risks

- Excessive mucus production
- Introital stenosis (usually mild)
- Prolapse
- Diversion colitis
- IBD

Neo Vagina Risks

Pediatric Surgery International
https://doi.org/10.1007/s00383-020-04838-2

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

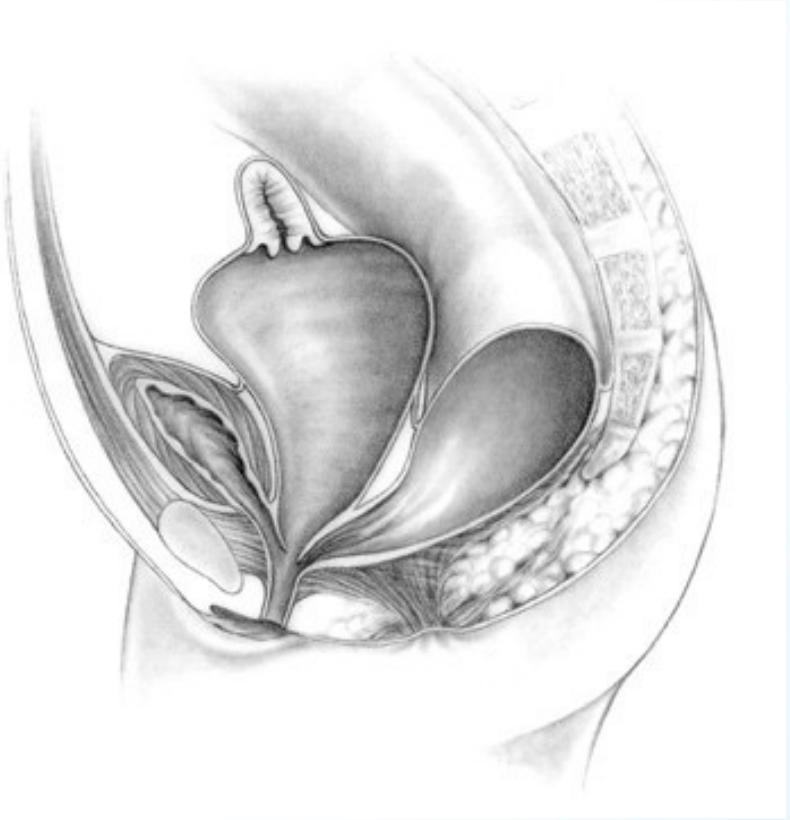
Cases of dysplasia and adenocarcinoma are reported

Any bleeding, pain, nodules or stenosis should be evaluated

- Exam
- Vaginoscopy
- Biopsy

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?

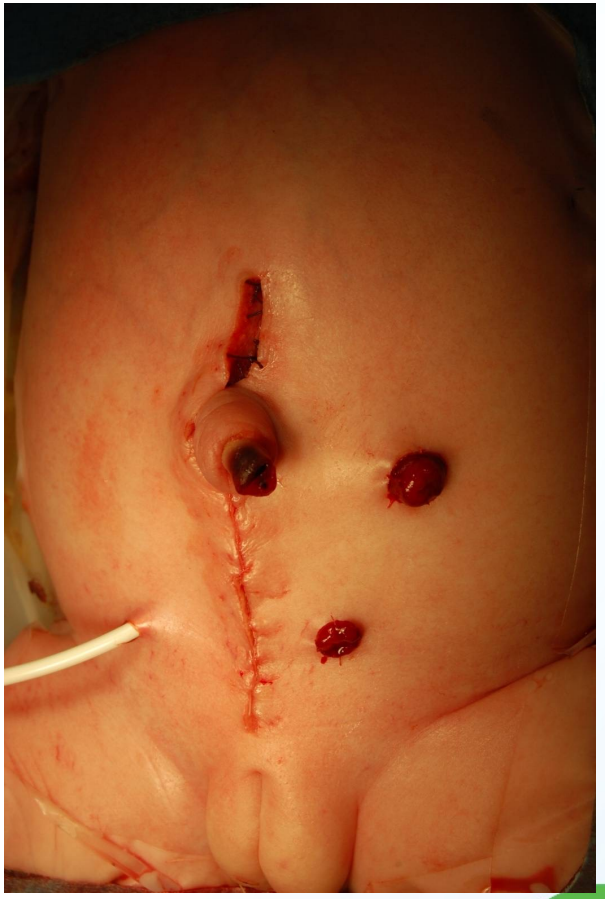
1. Compression of the ureters and hydronephrosis
2. Risk of infection
3. Drainage uncessary in asymptomatic patients
4. Answers 1 and 2

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

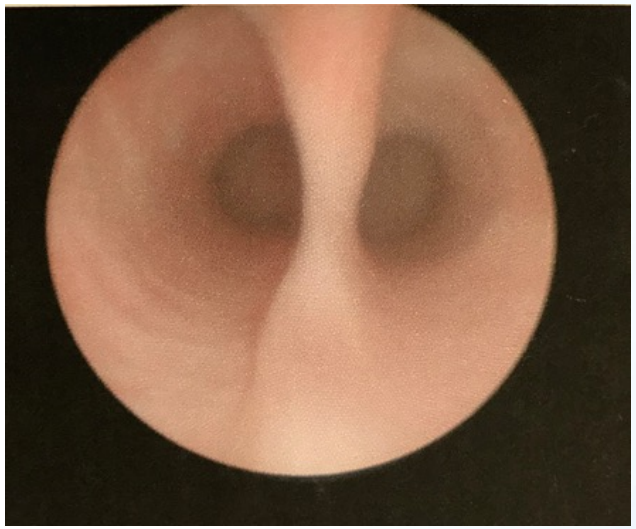
Hydrocolpos Treatment

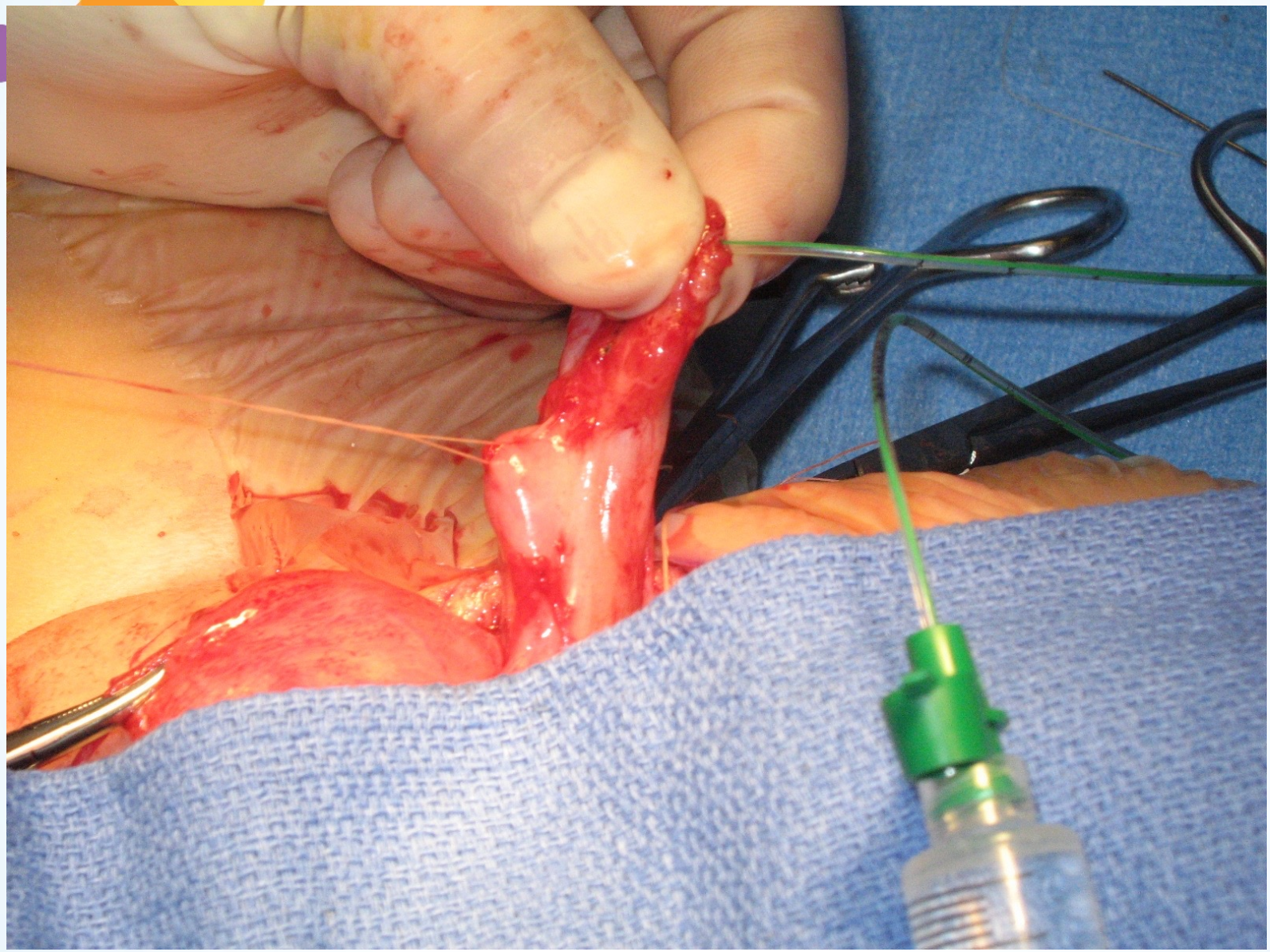
- Vaginostomy
- If a vaginal septum is present, a window is created to drain both vaginas

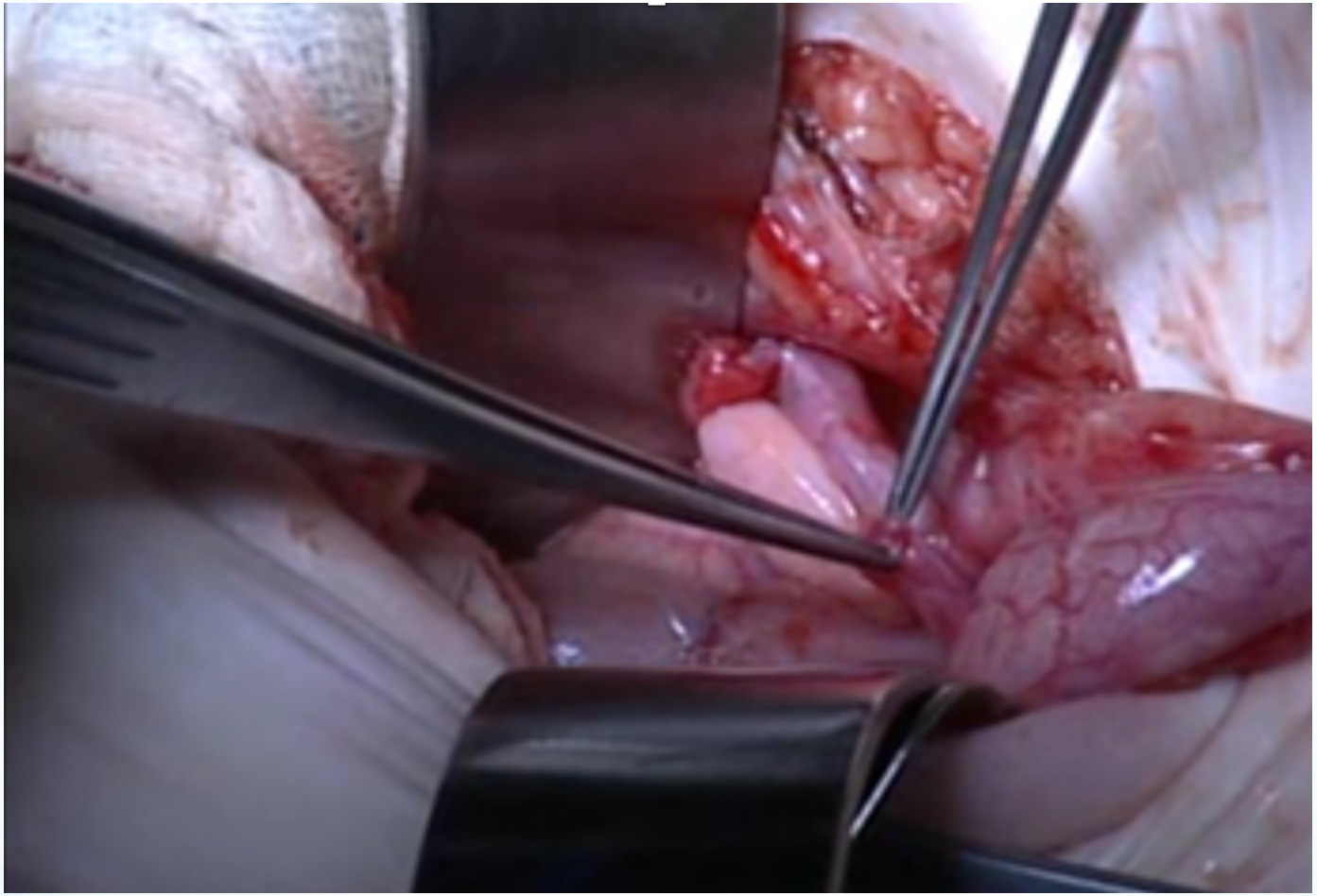


Evaluation of Gynecologic Anatomy

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







Infancy

- Determining gynecologic anatomy can be challenging
  - Example: In a review by Warne et al, out of 10 patients diagnosed with uterine hypoplasia, 6 went on to have normal menstruation
- With uncertainty, caution should be taken when considering removing uterine tissue

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

Mullerian Duplication (34%)

Cloacas	59%
Vestibular Fistula	6.2%
Perineal Fistula	3.8%

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

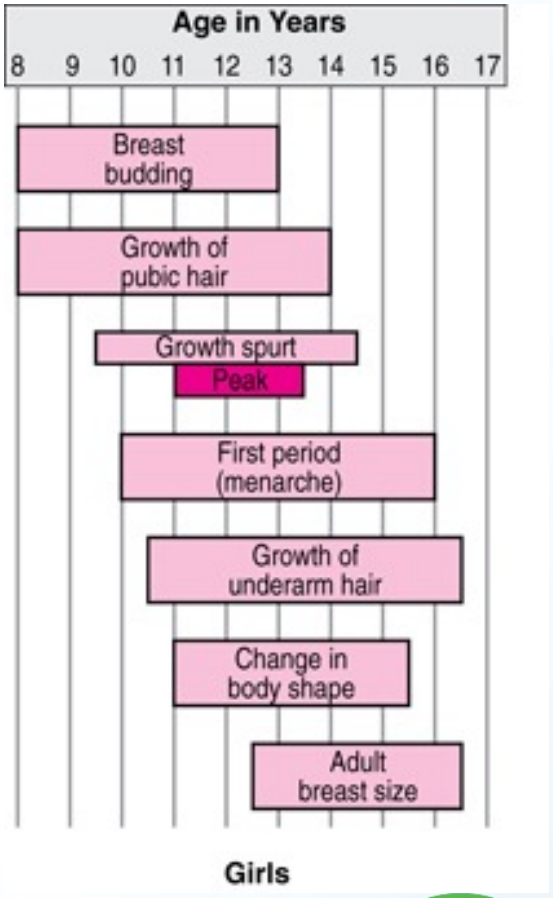
1. 80%
2. 60%
3. 40%
4. 20%
5. 5%

Vaginal Agenesis (5.6%)

Cloaca	17 (3.5%)
Rectovestibular Fistula	34 (10.7%)
Rectoperineal Fistula	1 (1%)

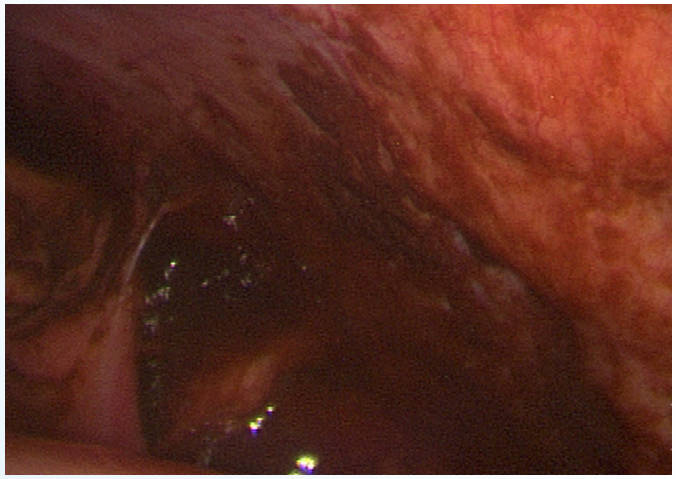
Puberty

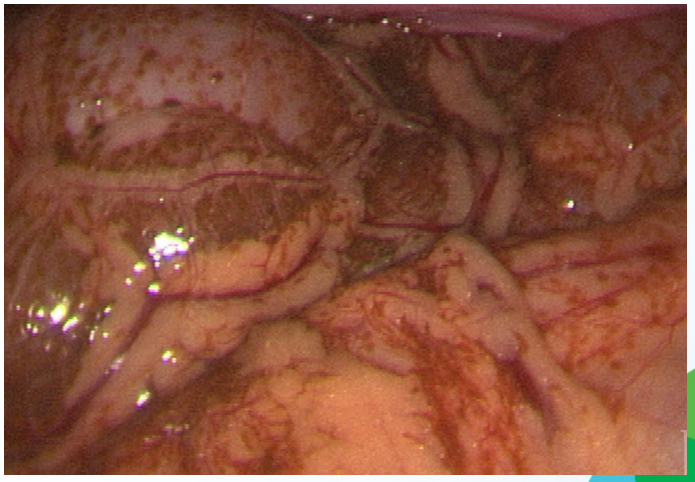
- Patients with ARM have normal ovaries
- Pubertal events occur as expected and follow a typical sequence
  - Menarche at age 12.4 (avg)

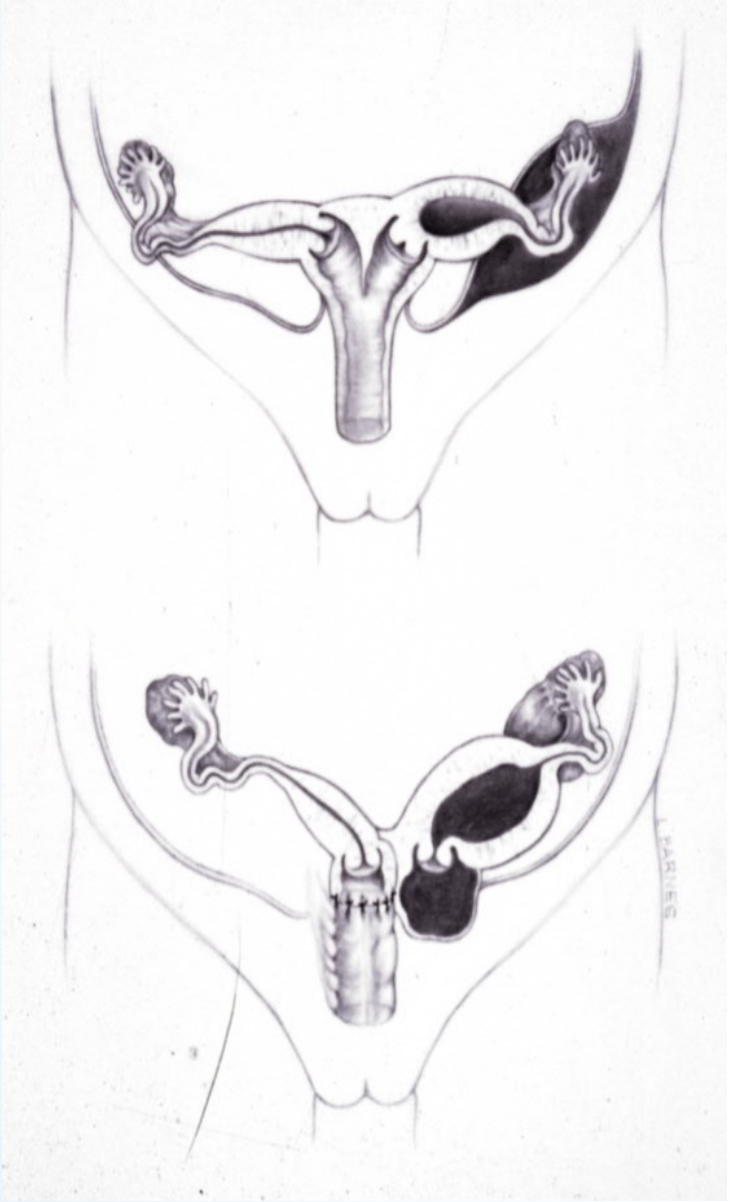


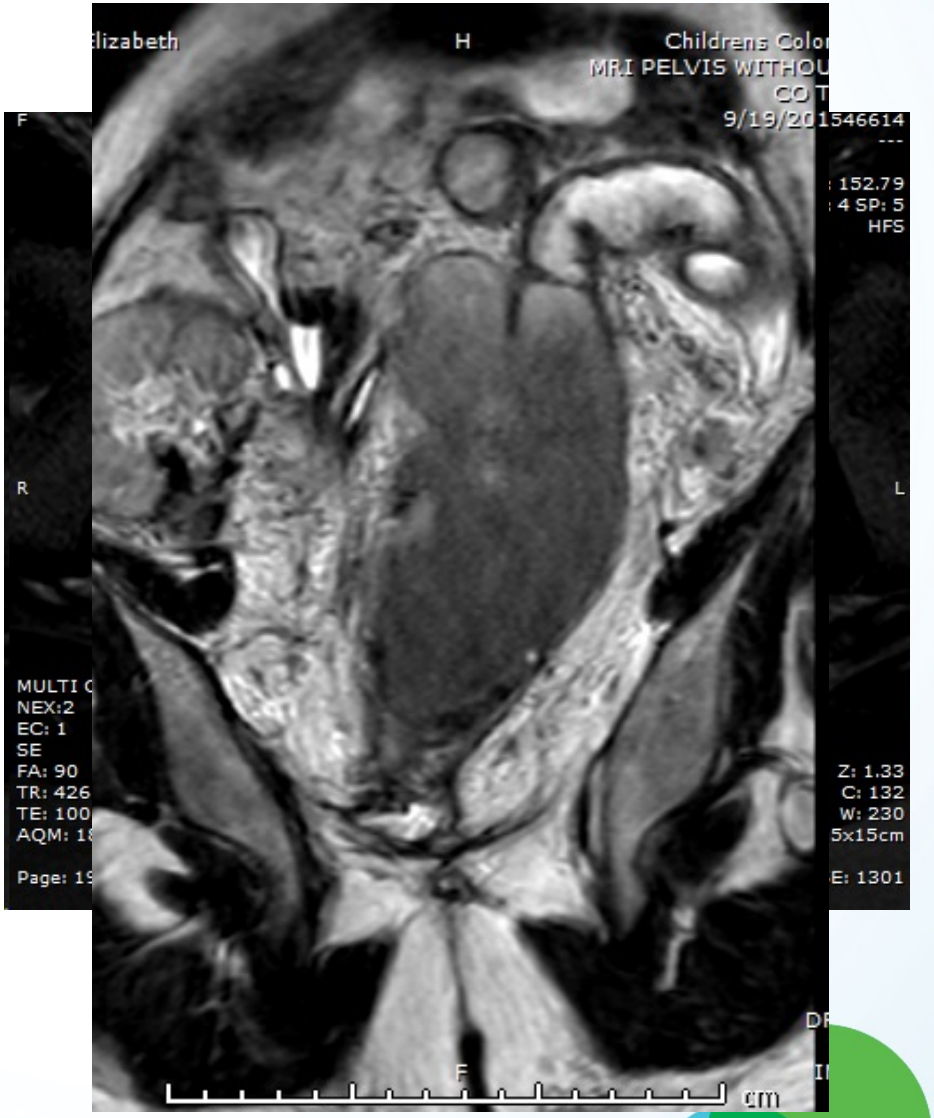
Puberty

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








Childrens Color
MRI PELVIS WITHOU
9/19/201546614
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 Options

Pre Operative

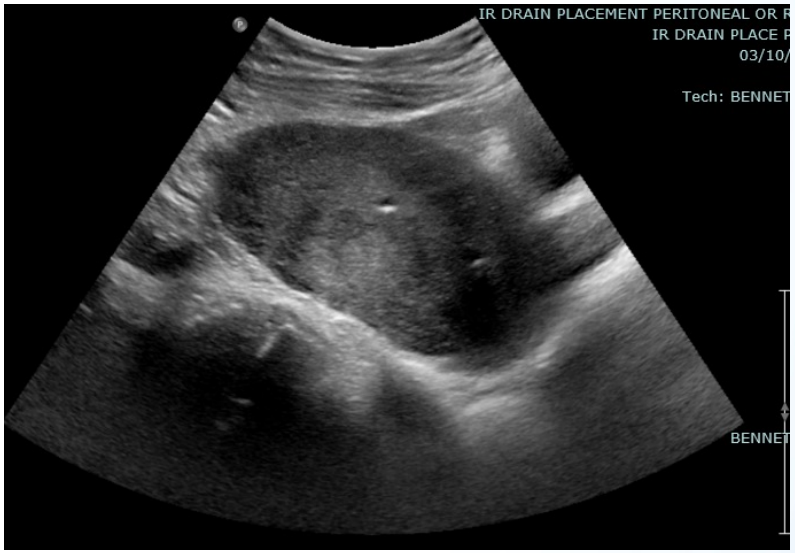
- Menstrual Suppression
- US Guided Drainage
- Vaginostomy

Definitive Surgical Management

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

Ultrasound Guided Drainage

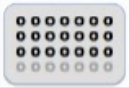
- Typically performed by Interventional Radiology
- Percutaneous approach preferred
- Tissue plasminogen activator (TPA) to break up clot



Puberty

Hormonal suppression of menses

- Obstructed menstruation
- Dysmenorrhea
- Heavy menstrual bleeding
- Hygiene concerns

Treatment Choice	How to Use	What to expect after the first 3-6 months
Norethindrone Acetate 	Take 1 pill every day	• Usually no periods when taken every day • Less cramping and less pain.
Pill 	Take 1 pill every day	• Lighter and regular periods. • Less cramping and less pain. • Clearer skin. • No weight gain. • Can be used in a certain way to have a period only every 4 months, or to have no periods at all.
Vaginal Ring 	Change every month	
Skin Patch 	Change every week	
Depo-Provera 	Shot every 3 months	• Lighter or no periods after 6-9 months of use. • Less cramping and less pain. • May cause increased appetite.
Progestin IUD 	Doctor places inside the uterus. IUD works for 5 years.	• Lighter or no periods. • Less cramping and less pain. • No weight gain.
Implant 	Doctor places under skin of arm. Implant works for 3 years.	• May have no periods, or irregular bleeding. • Less cramping and less pain. • No weight gain.

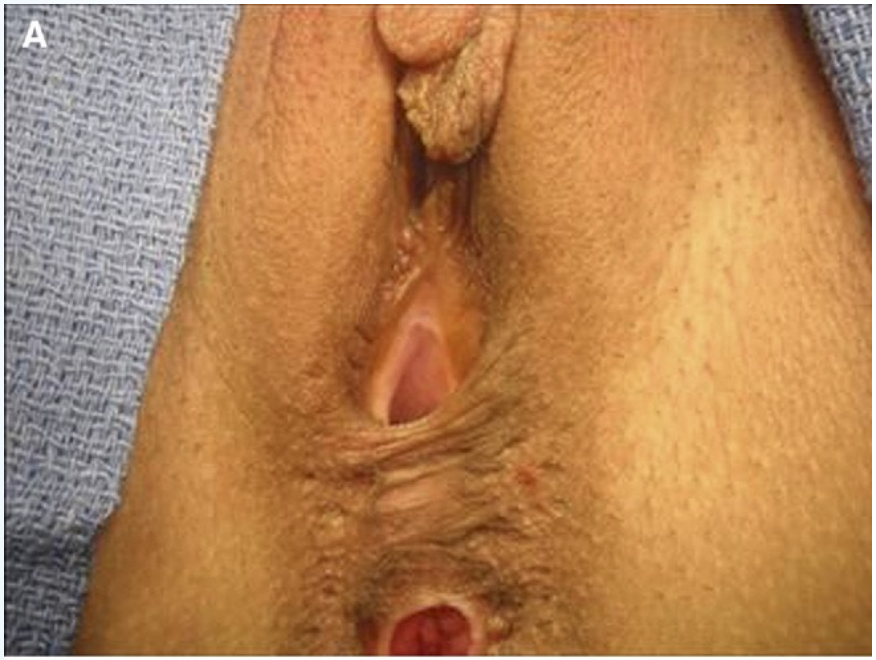
Sexual Function

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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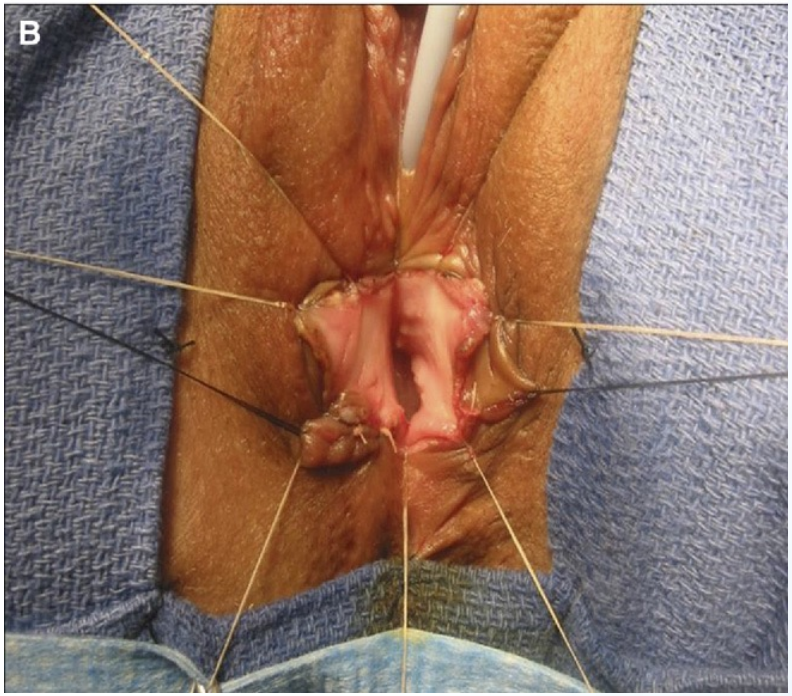
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Sexual Function

- 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

Sexual activity and fertility in adolescents and adults with cloaca.

	Sexual activity (%)	Adequate vagina for intercourse (%)	Problems in intercourse (%)	Number of patients with offspring
Hendren[4] ($n = 24$)	71	n.r	n.r	6
Warne et al.[19] ($n = 21$)	57	86	n.r	0
Couchman et al.[13] ($n = 19$)	42	n.r	13	1
Rintala (2015) ($n = 27$)	52	81	21	3

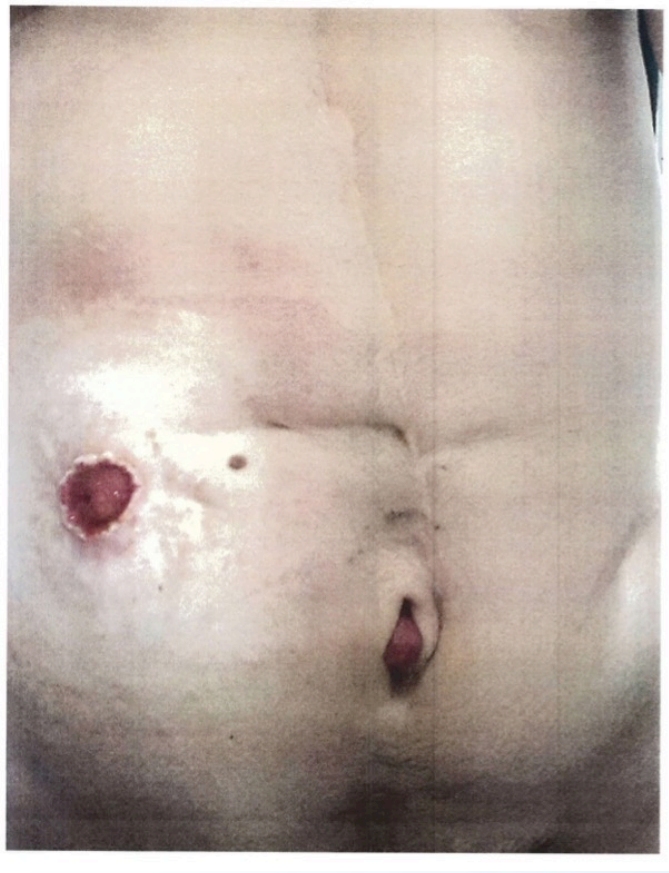
n.r, not reported.

Reproduction

- 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
  - Psychosocial factors

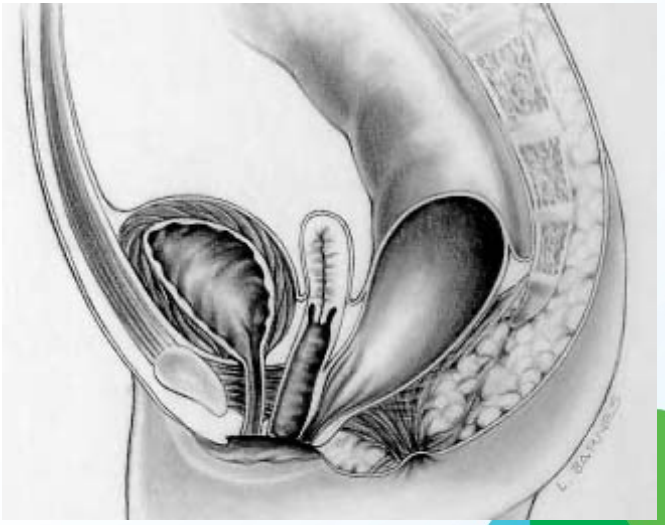
Reproduction

- 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

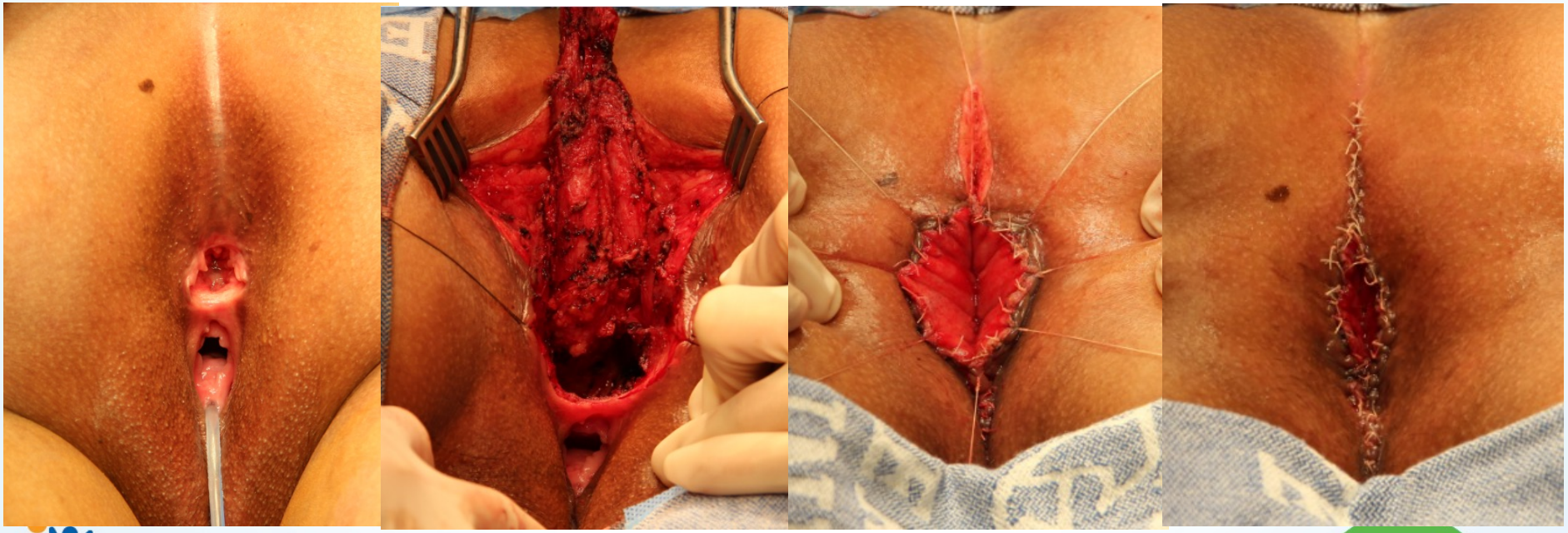
QUESTIONS

Case Presentation

26 year old female with chronic and severe constipation, diagnosed with rectovestibular fistula during adulthood

Case Presentation

Patient underwent diverting colostomy followed by posterior sagittal anorectoplasty



Case Presentation

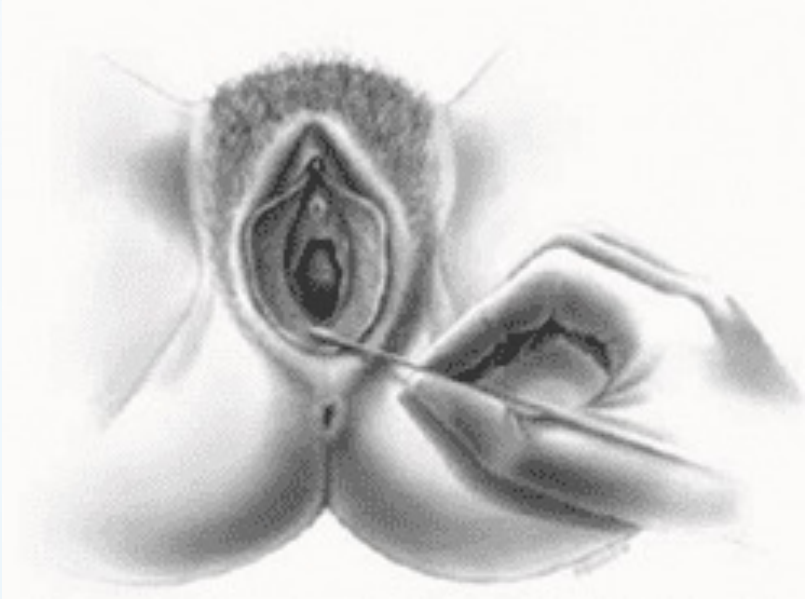
At a follow up visit, she reports long history of pain with intercourse. She describes a burning sensation with insertion of the penis and significant discomfort with deep penetration. She is unable to engage in vaginal-penile intercourse because of this pain

Dyspareunia

- Prevalence is 8-22% in the general population
- Outcome data on sexual dysfunction in patients with ARM is limited
  - In one study, dyspareunia was reported in 63 % of women who were sexually active

Schmidt et al. “Sexual function in adults with anorectal malformation: psychosocial adaptation. German Network for Congenital Uro-REctal Malformations (CURE-Net)

Examination: the vestibule



Using a cotton swab, test for pain on

- Inner thigh
- Labia majora
- Interlabial sulcus
- Clitoris
- Vestibule

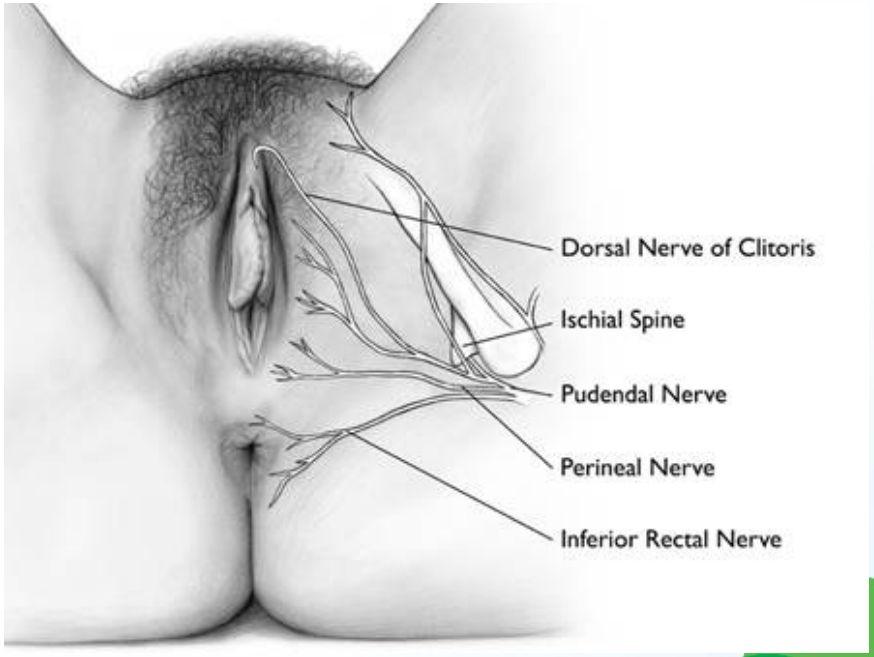
Vulvar Pain

Specific causes:

- Infectious
- Inflammatory
- Neoplastic
- Neurologic
  - Pudendal nerve injury

Vulvodynia: vulvar pain without identifiable cause

- Generalized
- Localized

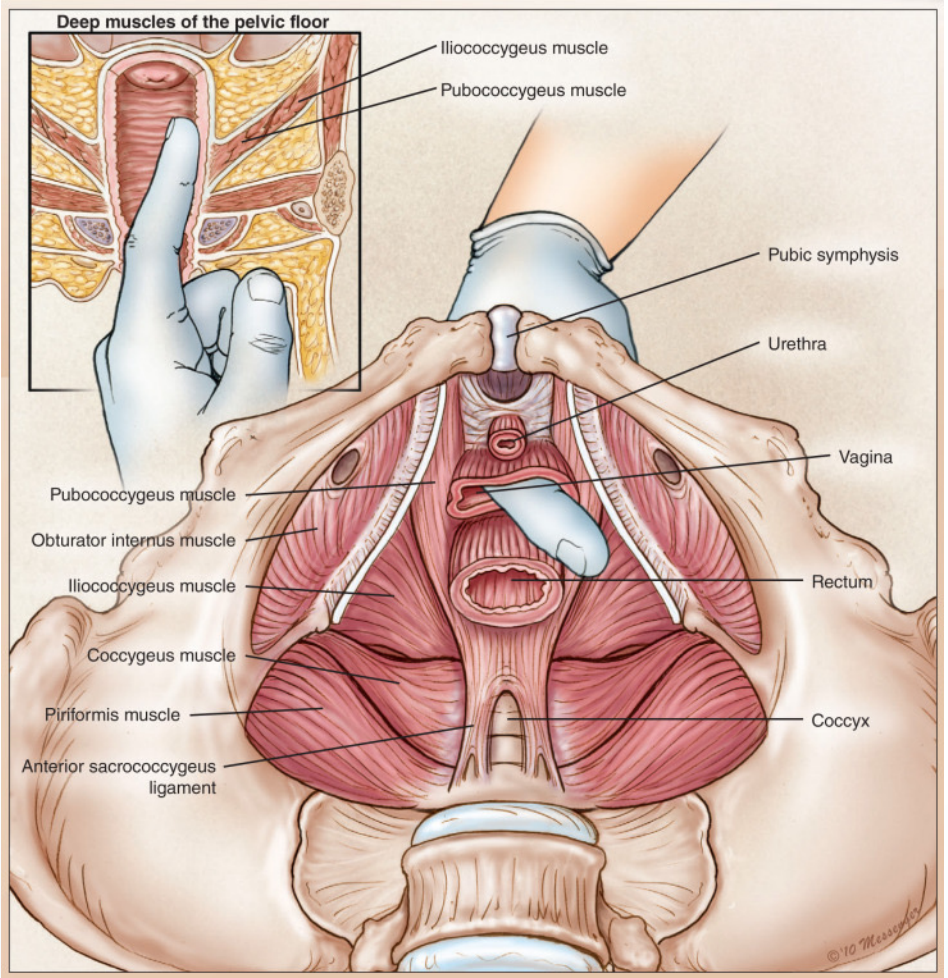


Treatment of Vulvar Pain

- Good vulvar hygiene
- Symptoms relief
- Stress reduction & exercise
- Lubrication

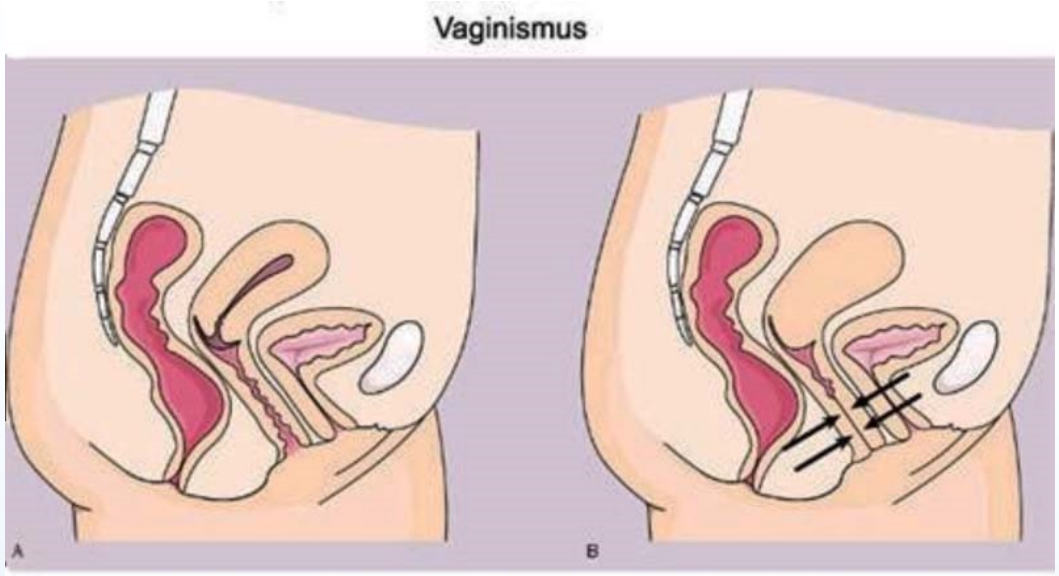
- Medications
  - Topical Lidocaine ointment
  - Estrogen cream
  - Compounded preparations (gabapentin, amitriptyline, baclofen, ketamine)
  - Oral anti depressants and anticonvulsants

Examination: the Pelvic Floor



Vaginismus

- Involuntary contraction of the muscles of the pelvic floor surrounding the vaginal orifice
- Aversion to vaginal penetration (sexual and nonsexual) due to actual or anticipated pain



Vaginismus is an involuntary constriction of the outer vaginal muscles, prohibiting intercourse. The drawing on the left (A) illustrates the relaxed vagina, while the drawing on the right (B) illustrates vaginismus.

Treatment of Vaginismus

- Pelvic floor physical therapy
  - Kegal
  - Pelvic drop
  - Myofascial release of muscle tension in pelvic floor, thighs, and abdomen, with or without biofeedback
- Desensitization with dilator therapy
  - Gradual increase in size
  - Topical anesthetic
- Sex therapy
- Benzodiazapines
- Biofeedback

Case Presentation

Our patient was treated with lidocaine ointment, dilators, vaginal valium, and pelvic floor physical therapy. Discomfort improved at follow up visits. She is now married and can comfortably engage in penetrative intercourse.


questions