Challenging Surgical Cases

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Objectives

• Address the diagnosis, preoperative assessment and management of challenging surgical cases
• Provide innovative “tricks of the trade”
• Utilize group expertise to analyze difficult clinical situations
MT- A Challenging Case

• 15 year old referred for primary amenorrhea; possible vaginal anomaly
• No complaints of abdominal pain or genital bleeding.
• Pubertal milestones: Thelarche:2 years prior; Adrenarche ?2-3 years
  Growth velocity was normal

• Primary care physician: Noted on vulvar examination that she had an angle to her vaginal canal, ? Vaginal mass;
  otherwise physical exam/genital findings normal

History- MT continued:

• PMH: no prior illnesses
• PSH: none
• FH: Mother with menarche at age 14; No related family illness
• SH: High school student; normal development; lives with family unit; denies drugs alcohol or sexual activity.
  Swims in season and runs 1-2 miles most days.
• ROS: Denise headaches, weight gain or loss, increased exercise or activity; no history of constipation or urinary issues, relates good balanced diet without restrictions; dines with the family
Physical examination/Laboratory

- BP: 102/55, P 80
- WT: 56.6 Height: 172, BMI: 19.2
- Well developed well nourished female; no pain or distress
- Skin: no acanthosis, acne or hirsuitism
- Breasts: Tanner III
- Genitalia: Tanner III; normal external genitalia; introitus normal; Q-tip placed in vagina with normal vaginal length, some orientation to the left; cervix viewed on left; no vaginal mass palpated although vagina deviates to left

Ultrasound
Other:

- FSH: 6.74
- Prolactin: 3.6
- TSH: 0.432.
- Estradiol: 18
- Renal ultrasound: absent R kidney; otherwise normal
- Bone age: consistent with chronologic age

- **Diagnosis:**
  Primary amenorrhea consistent with constitutional delay;
  OHVIRA (Herlyn-Werner-Wunderlich)

- **Unique features of this case:** Patient going to South America in 2 months for extended summer mission trip. Wants to delay surgery until she returns but is worried about the consequences if menarche begins.
Unanswered questions:

• With no history of vaginal bleeding and apparent right vaginal obstruction – why does she have fluid in the obstructed right side?
• Patient desires menstrual suppression to allow timing of repair delayed. What is the best option for short term suppression?
• With no vaginal distention and need for suppression what are the anatomic considerations for repair? What technique should be used to identify anatomic landmarks?

Clinical Course /Plan

• Avoidance of long term suppression in this case
• Short term menstrual suppression with OCP while out of country. Patient has experienced spontaneous menstrual spotting but no symptoms to date
• Plan to stop OCP 2 months prior to repair
• Use of ultrasound guidance/ inflatable catheter to delineate obstructed side with vaginal resection
Interesting literature


KN- A challenging surgical case

- 46XY DSD patient first presented at age 12. History at birth was of ambiguous genitalia, had gonadectomy, hysterectomy, vaginoplasty, feminizing clitooplasty Sex of rearing: female

Surgical findings at EUA:
Epispadias, 9 cm torturous urethra, perineal pit 1-2 cm

Placed on hormonal regimen for pubertal induction
KN- A challenging case

- At age 18 underwent laparoscopic sigmoid vaginoplasty, vaginal flap, excision of mullerian remnant labiaplasty, cystoscopy, epispadius repair
- Started on vaginal dilations, did well initially with only complaints of mucous drainage
- 3 mo post op:
  - Complained of acute inability to pass dilator
  - Returned to OR with findings of normal vaginal length, stenosis 2 cm into vagina at area of perineal-graft junction
  - Had intraoperative dilation to #25 Hegar with inflatable stent placed

KN- A challenging case

- 6 month post-operative
  - Gradually had more pain/difficulty with dilation
  - Returned to OR for dilation under anesthesia
- 1 year post operative
  - Continued to have pain/difficulty with dilations with minimal compliance
  - Returned to OR with findings of now stenosis 3 cm from introitus. Lateral excision of stenotic area with free buccal flap placed
  - Inflatable dilator placed post operative
  - Intermittent dilations begun
KN- A challenging case

• 2 years post operative
  • Comained of progressive difficulty with dilation and pain
  • Returned to OR: Stenotic area excised laterally with mucosal mobilization over defect. Inflatable stent placed and patient resumed dilation

• 3 years post operative
  • Patient able to dilate to M/L dilator but continued to complain of pain and difficulty with dilation
  • Returned to OR: 28 Hegar dilator passed without difficulty; area with some erythema, friable
  • Started on Estrogen cream with improvement in tolerance to dilation

KN- A challenging case

• 5 years post operative
  • Lost to follow-up
  • In a relationship but unable to tolerate vaginal penetration
  • Can use large dilator but is painful
  • Using E2 cream and compliant with oral estrogens
Clinical considerations

• How can we increase vaginal compliance and function for this patient?
• The etiology of retraction of vaginal perineal suture line to an intravaginal place; short mesentery; early dilation
• Is dilation necessary after sigmoid vaginoplasty?
Interesting references


EP- A challenging case

• 7 year otherwise healthy girl with 6 month history prepubertal vaginal bleeding of uncertain etiology.
• On vaginoscopy, found to have an exophytic mass arising anterior to and involving the cervix. Final pathology: Clear Cell Carcinoma
• Workup revealed a T3B lesion with cancer extending to vagina with no metastatic disease
• Patient was referred to Sloan Kettering Cancer center for consideration for fertility sparing procedure
**EP- her clinical course**

- Patient was pretreated with 3 cycles of Cisplatinum/Taxol chemotherapy.
- Surgical procedure: Radical hysterectomy, repair of bladder wall, transposition of ovaries, partial vaginectomy, nephrectoureterectomy of atretic right kidney, lymph node dissection.
- Received 2 additional cycles of Taxol and Cisplatinum.
- Local brachytherapy administered after chemotherapy.

**Post operative course**

- Mother instructed to do post radiation dilations. Compliance with dilations was difficult, but continued for 1 year.
- Patient followed with 6 month EUA, vaginoscopy and pap smears which have all been normal.
- Patient with spontaneous signs of puberty age 10.
- 2 years post operative patient complained of incontinence – intermittent and primarily post voiding. She reported “being wet” sometimes after voiding. She denied dysuria, hematuria, but related some urgency. Placed on oxybutrin with some improvement.
3 years post operative:

Patient taken to the operating room for routine vaginal surveillance. At the time of vaginoscopy, lesion noted approximately 1 cm into introitus. No other lesions noted. When lesion explored closely appeared to be fistula with communication immediately inferior to bladder neck.

Clinical cases

• How to fix this now 13 year old with previously radiated pelvis now intrapubertal with urethral-vaginal fistula at the bladder neck.
• How long to continue surveillance?
• The role of vaginal dilation in children
Interesting references
