Anorectal malformation in a female

Case # 2

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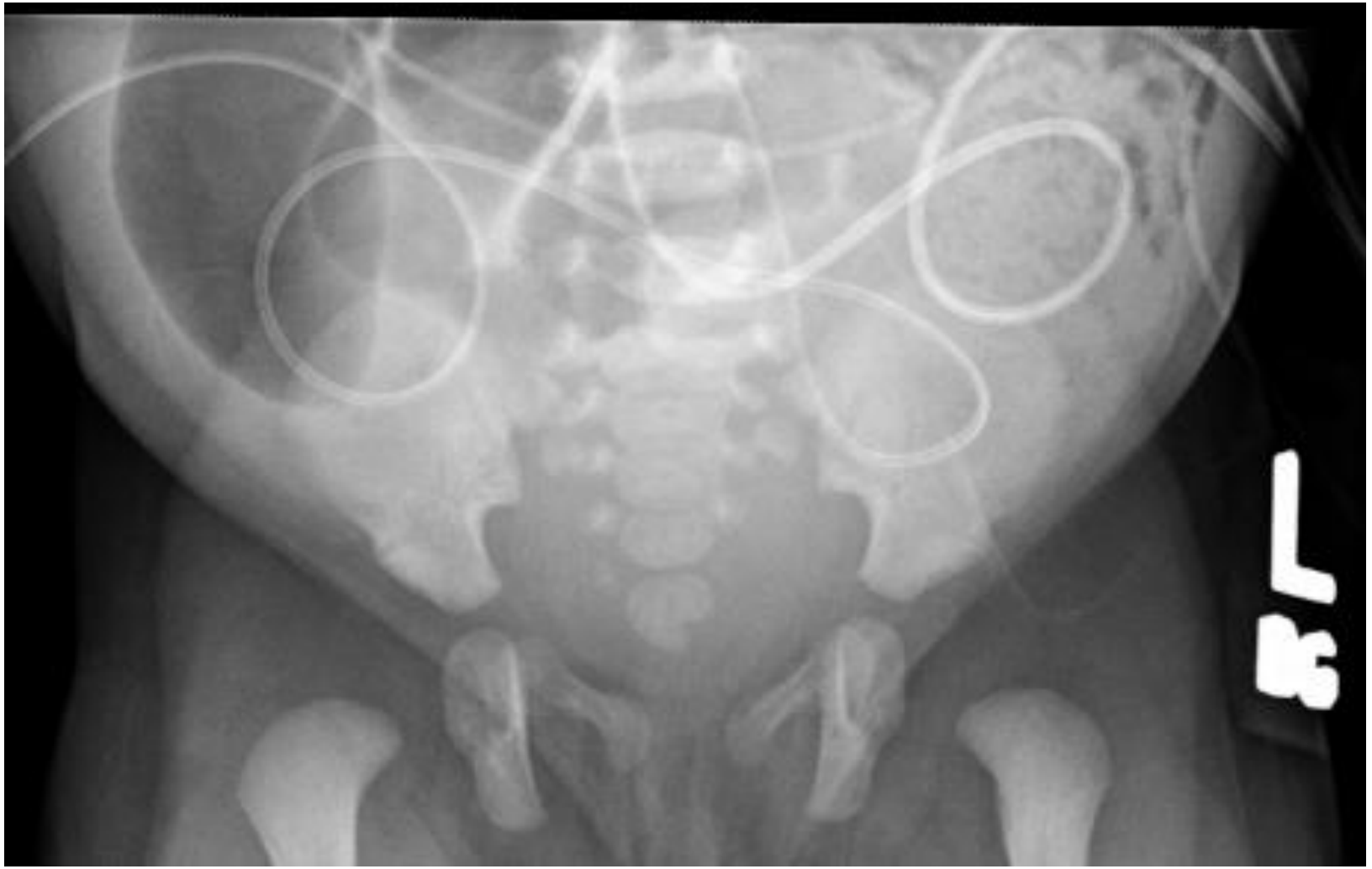
Presentation

- Full term, 4 kg, female patient

- On physical exam after birth: no anus
  - Orogastric tube decompression
  - NPO, TPN

What studies would you like to order?

1. Sacral x-ray AP and lateral
2. Kidney ultrasound
3. Spinal ultrasound
4. Babygram
5. Echocardiogram
6. All of the above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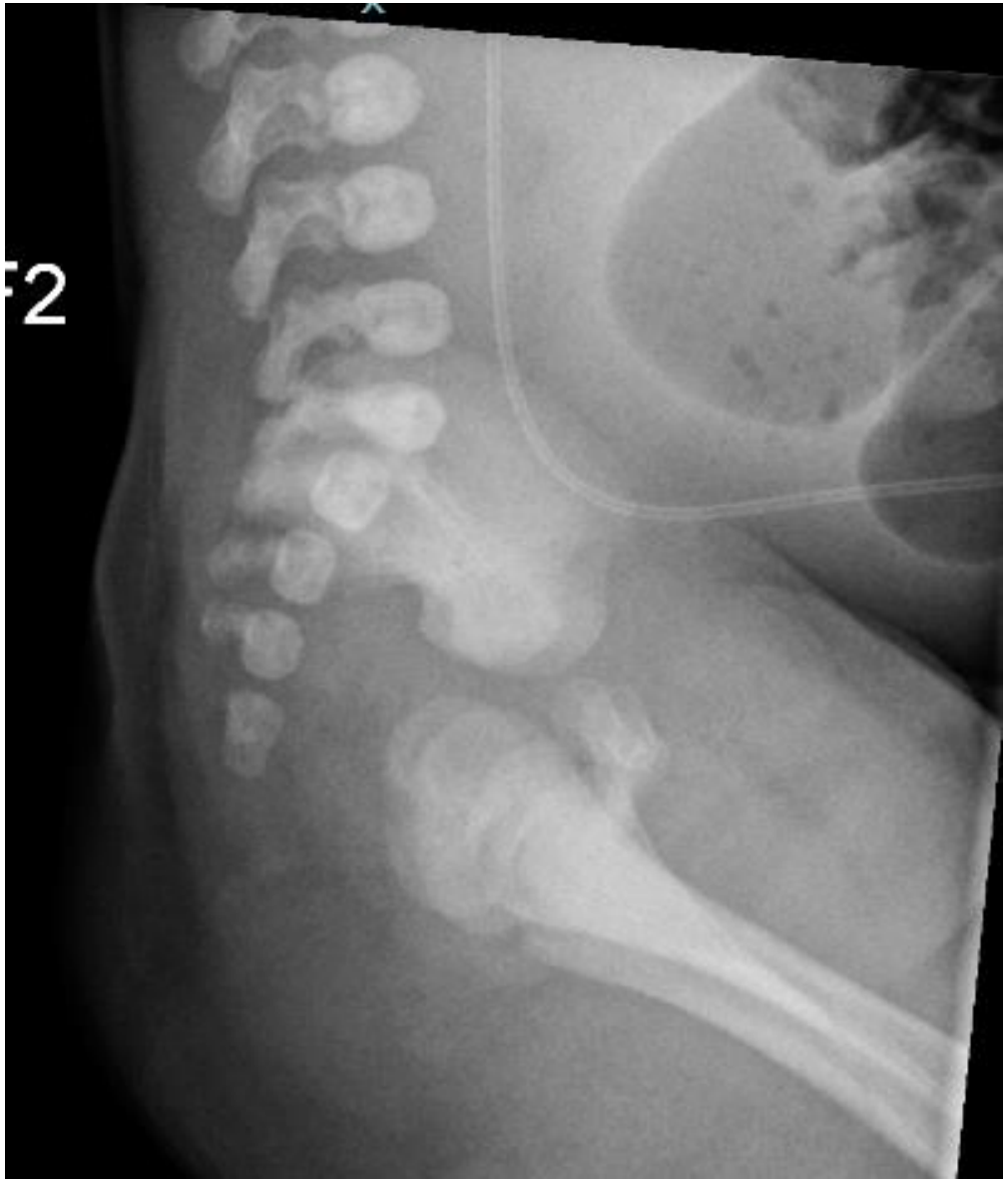
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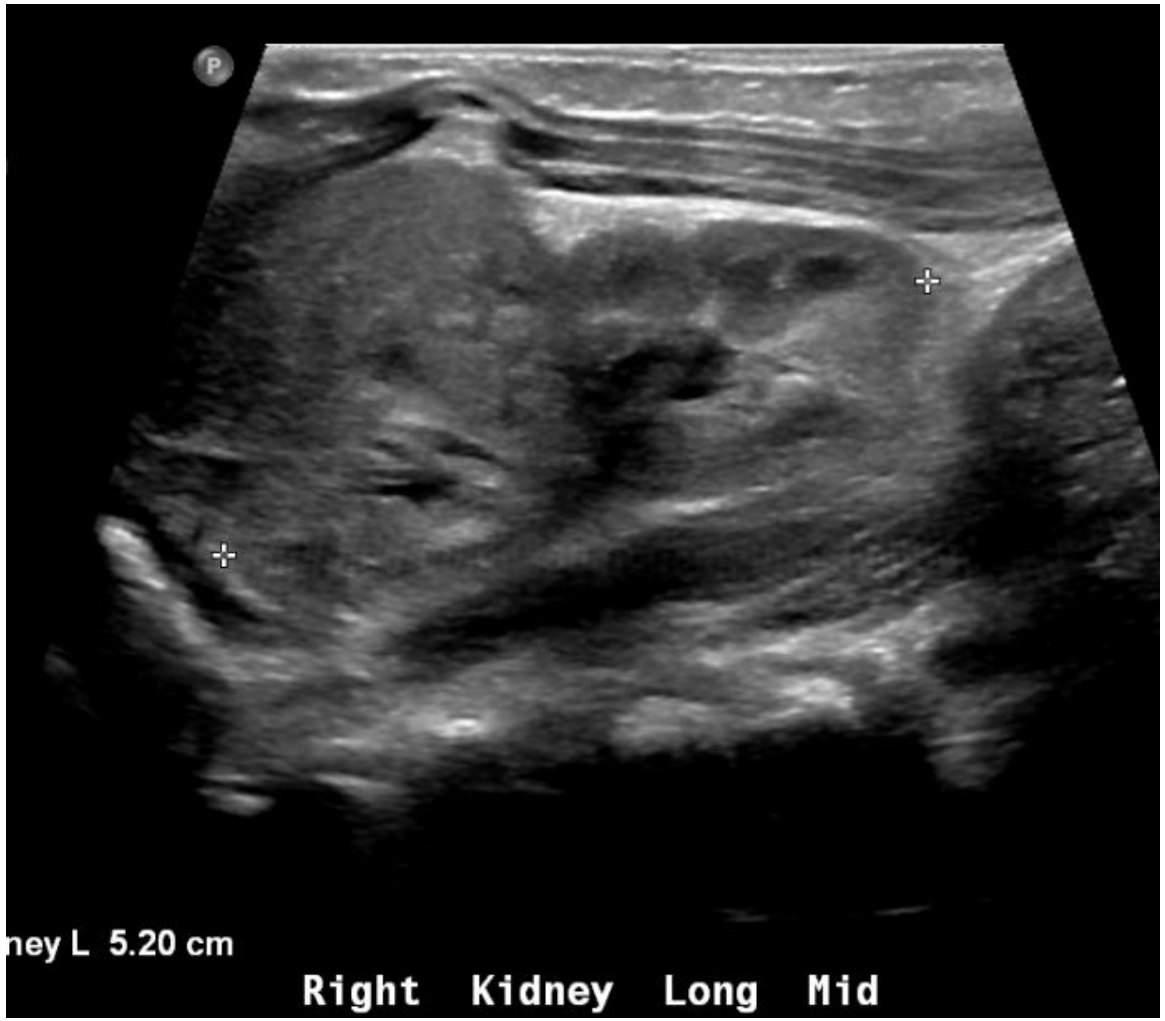
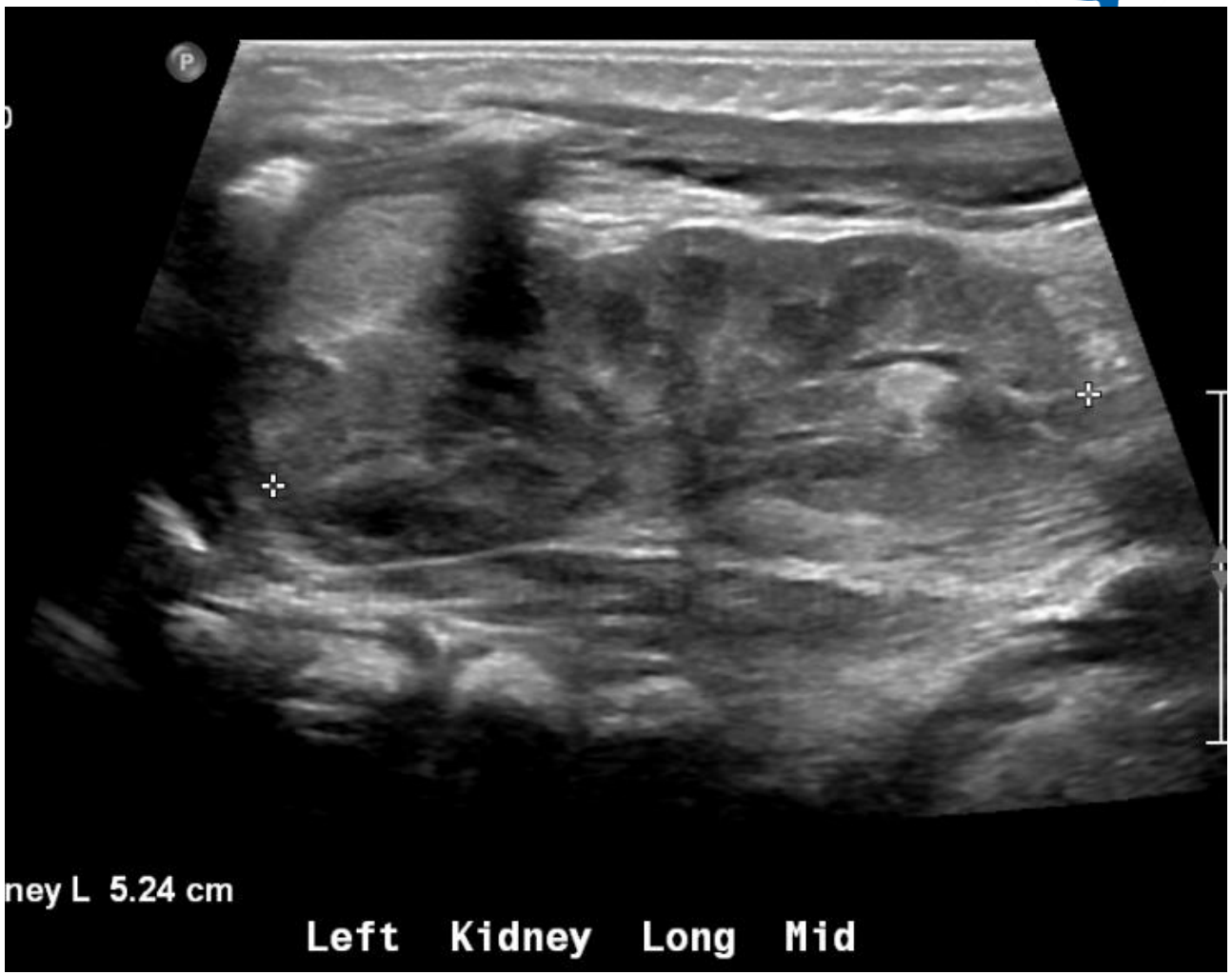
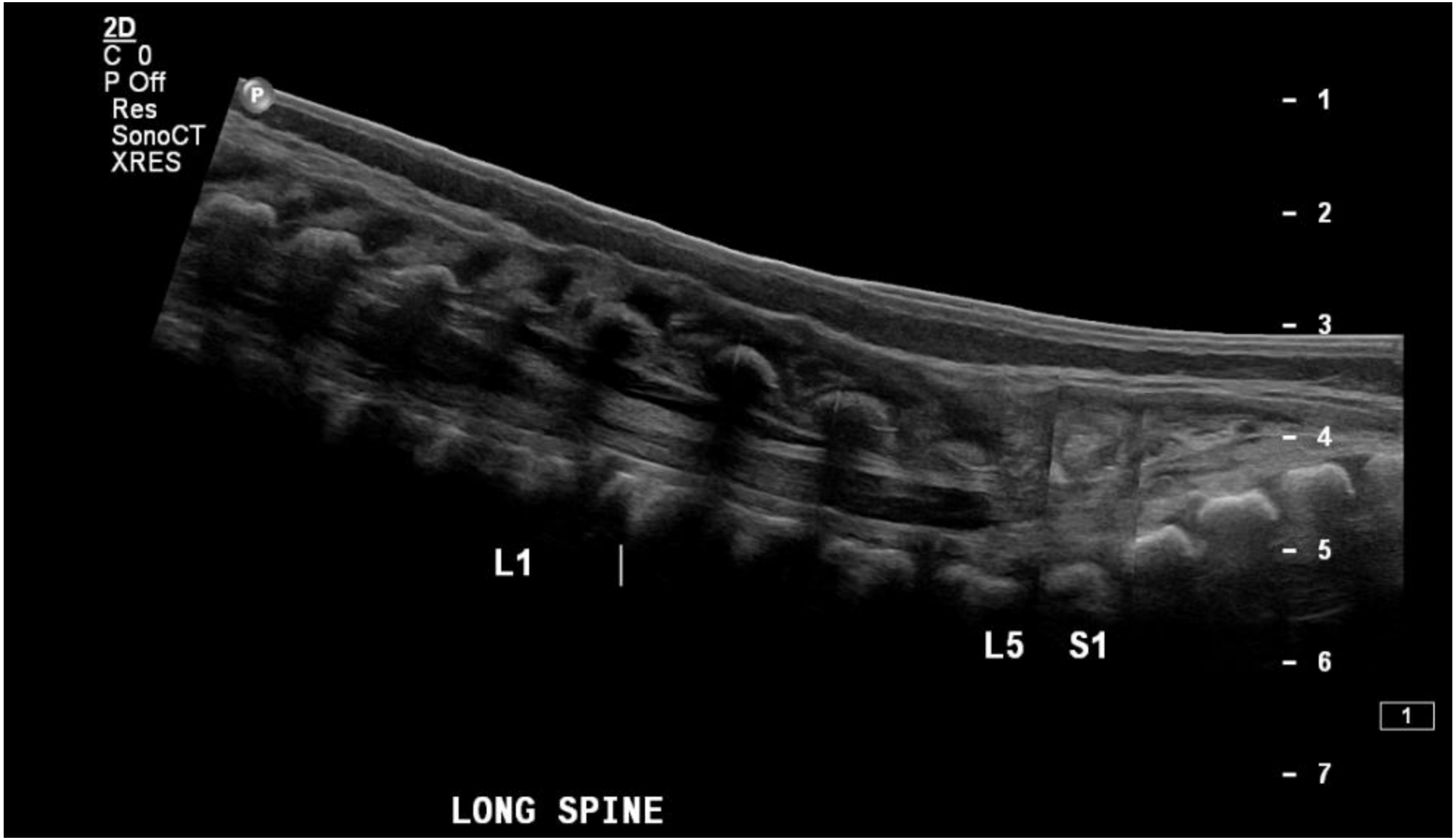

ney L 5.20 cm
Right Kidney Long Mid


ney L 5.24 cm
Left Kidney Long Mid


2D
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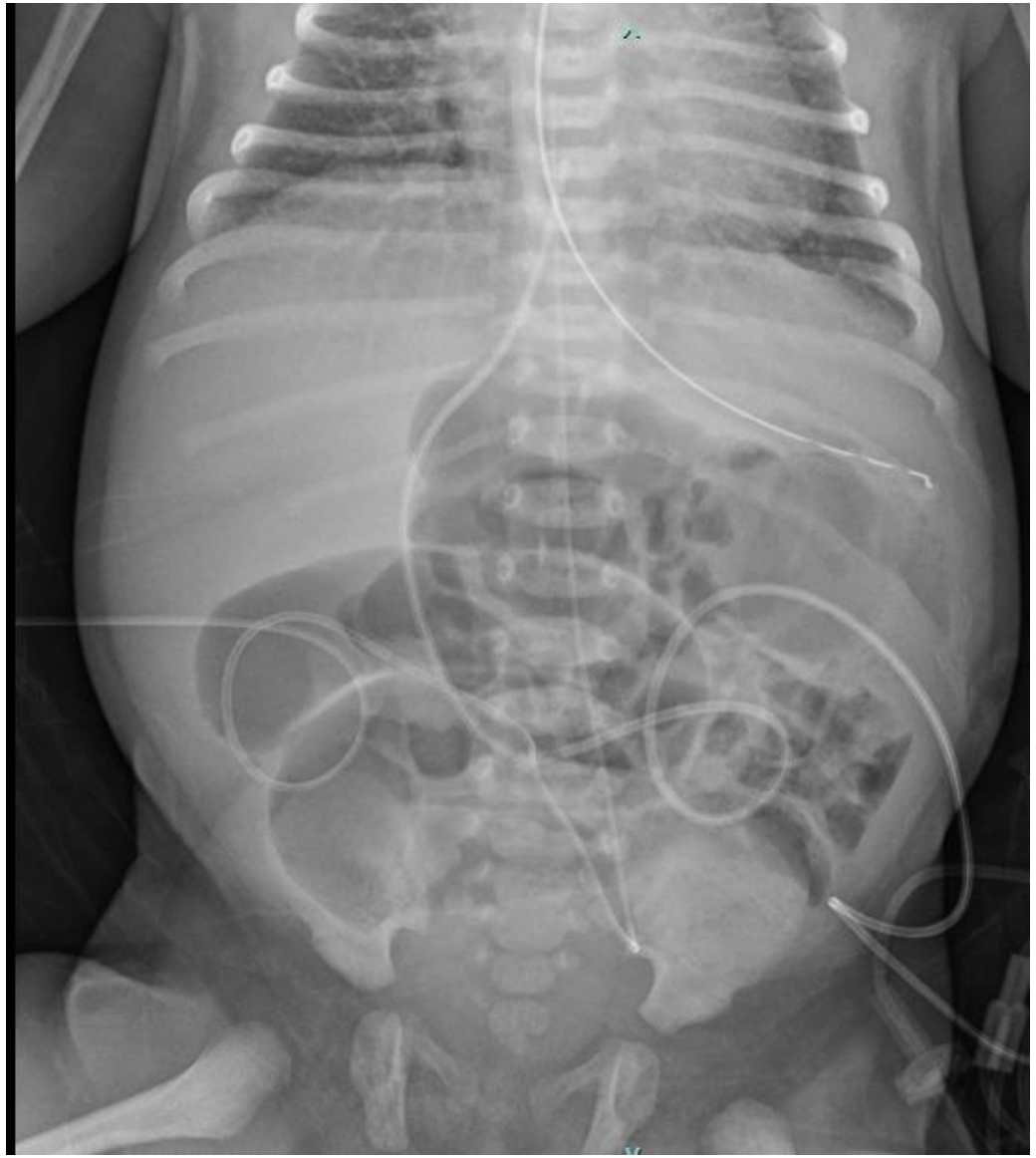
LONG SPINE

Echocardiogram – small patent ductus arteriosus

24 hours



Is she a good candidate for a primary PSARP? What additional work-up can aid in decision between a primary PSARP versus divided colostomy?

1.Yes - Proceed with PSARP. She is full term, otherwise healthy and 4 kg. No additional work-up needed.

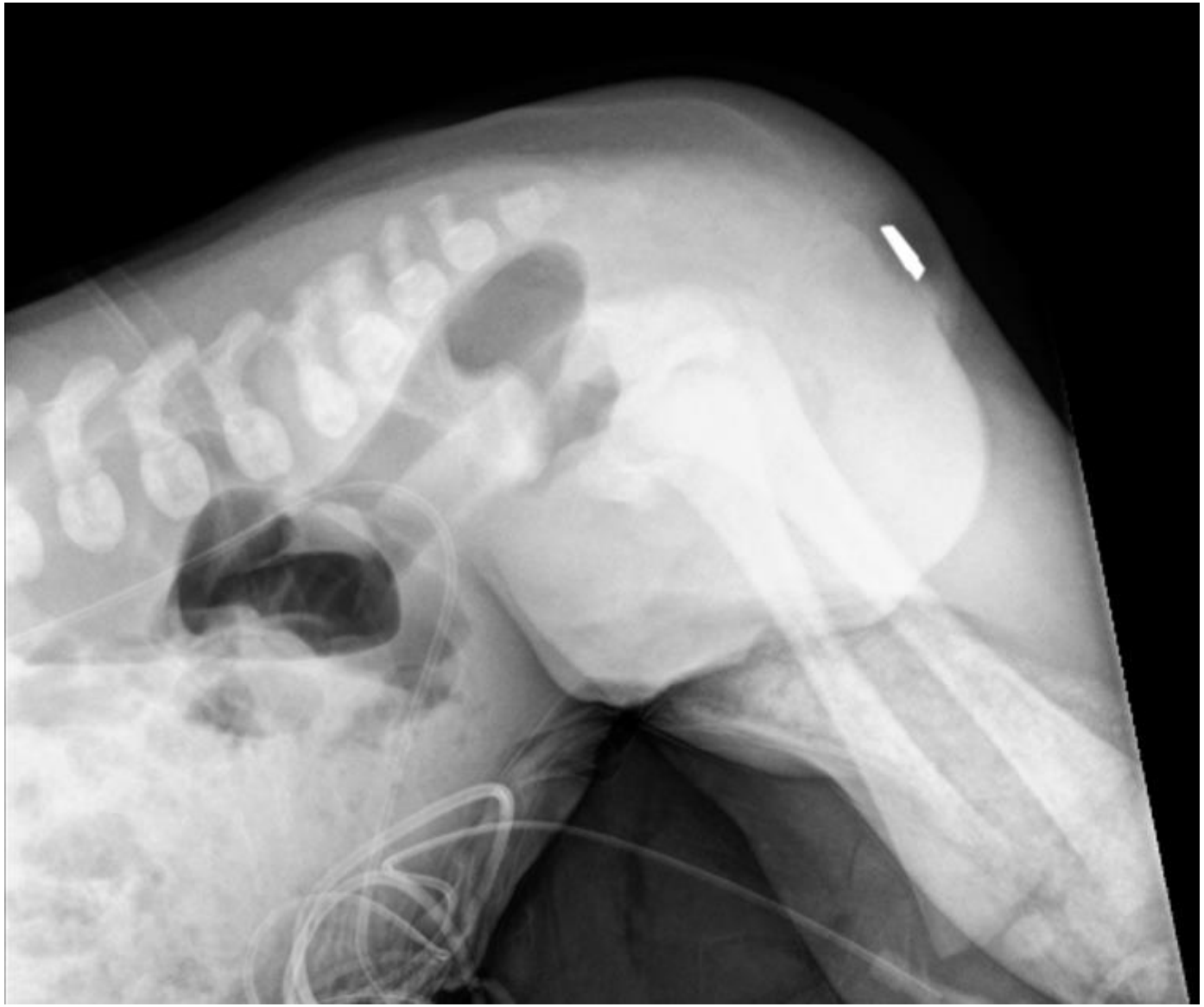
2.Yes – Proceed with PSARP. She is full term, otherwise healthy and 4 kg. Perineal ultrasound to evaluate level of the rectum.

3. No – Proceed with colostomy. The location of the fistula and distal rectum are unknown, and therefore we don't know the best operative approach (PSARP versus trans-abdominal). MRI pelvis to evaluate anatomy.

4. Unknown – we do not know the level of the fistula or distal rectum. Prone cross table lateral film will help in the decision.

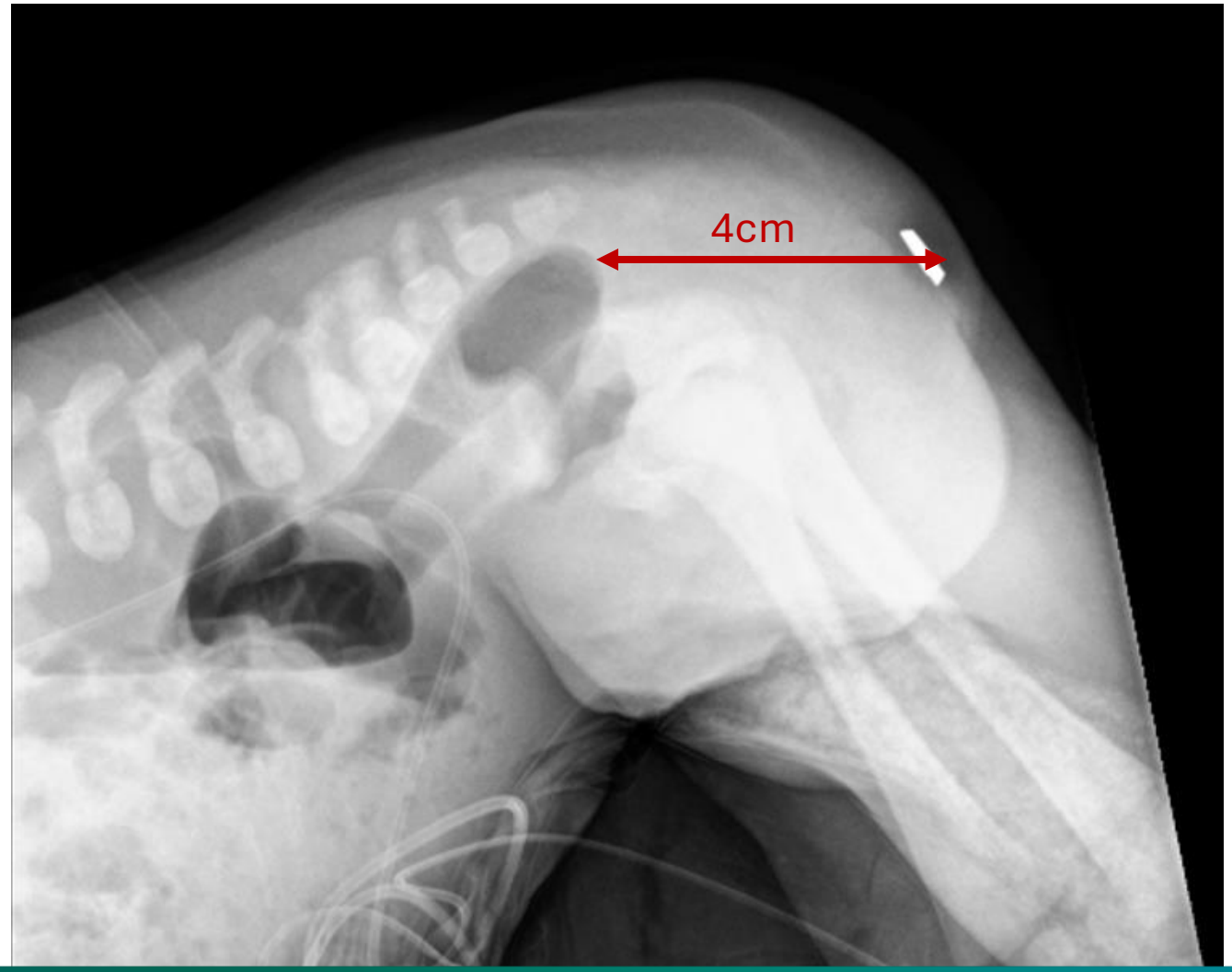
5. I always open a colostomy.

48 hours



Would you proceed with primary PSARP?

1. Yes
2. No

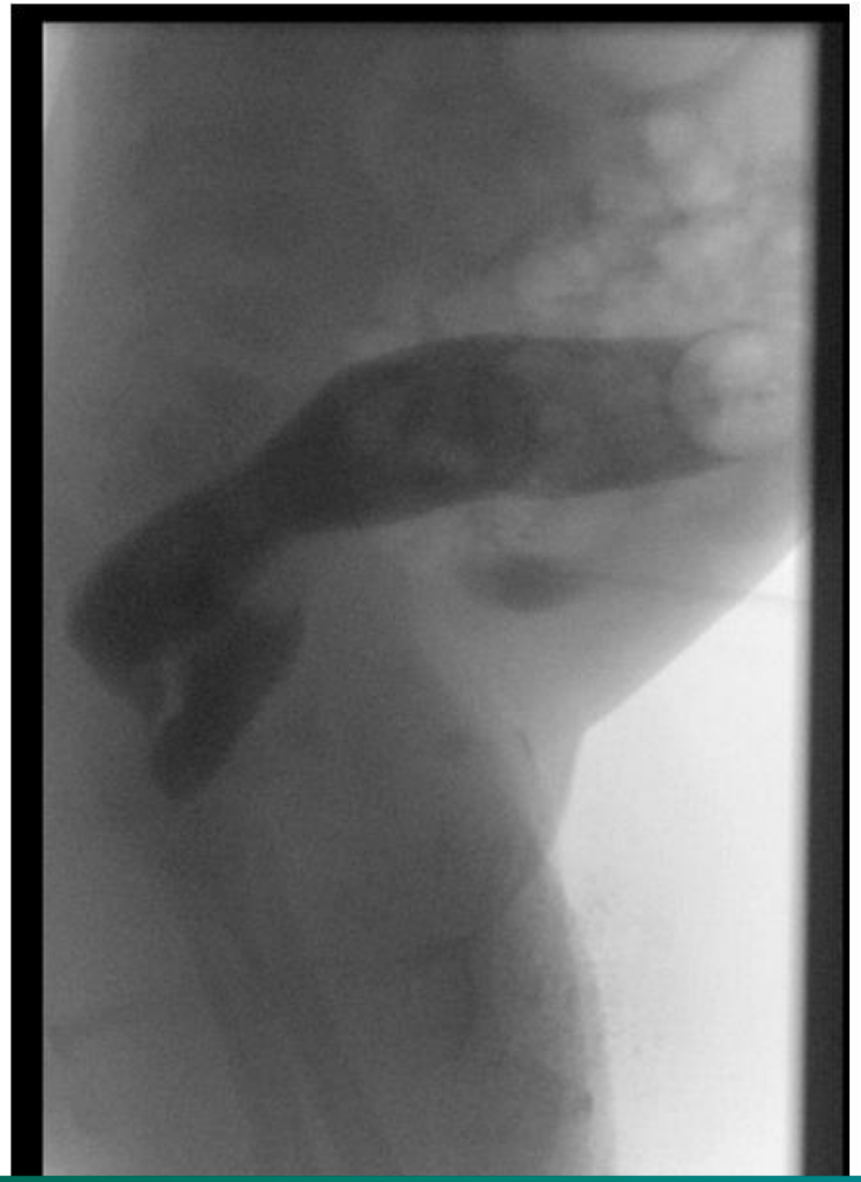


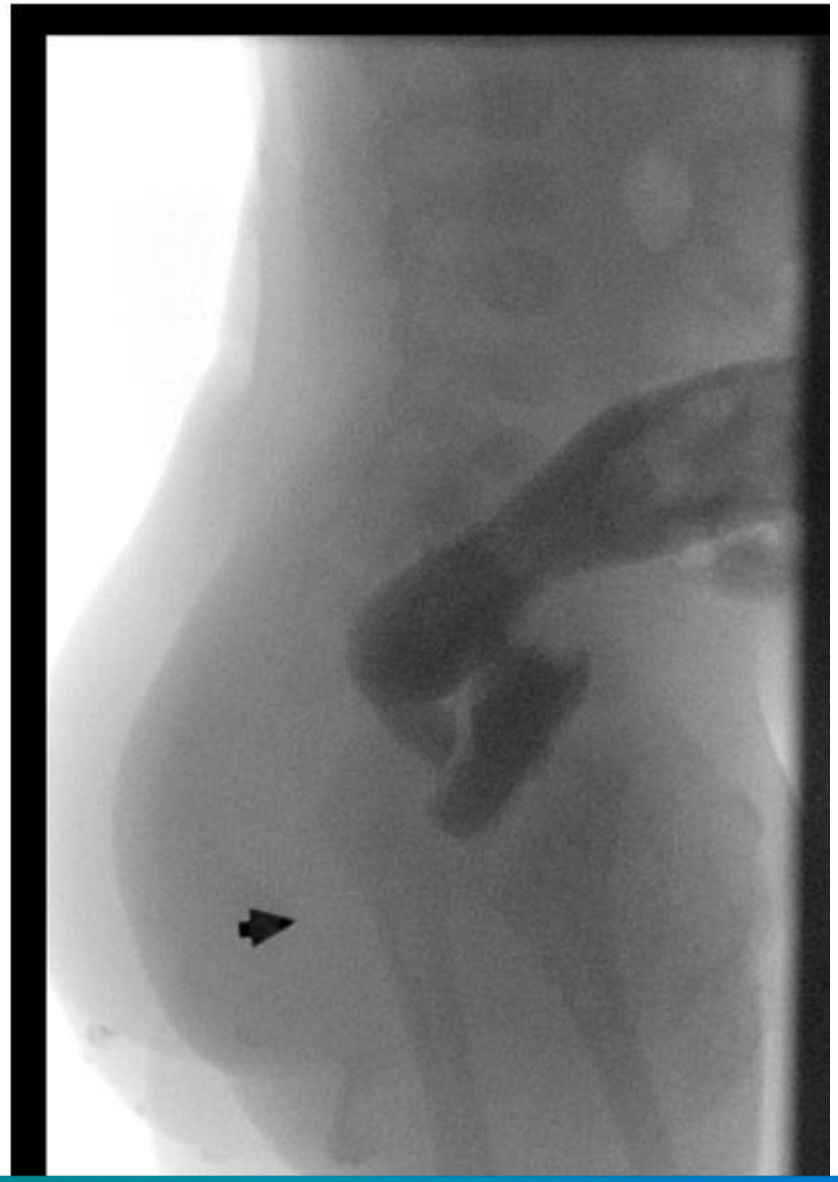
- DOL 2 –
  - Laparoscopic divided colostomy + mucus fistula at descending colon
  - Moderate meconium irrigated from mucus fistula

Clinical course

- DOL 5 – large meconium stool from vagina
  - Presumed diagnosis of rectovaginal versus rectovestibular fistula

- DOL 7 – Discharged home
  - Distal colostogram – 6 weeks old
  - Planned PSARP – 10 weeks old





What is the diagnosis?

1. Rectoperineal fistula
2. Rectovestibular fistula
3. Rectovaginal fistula
4. Cloaca
5. I don’t know

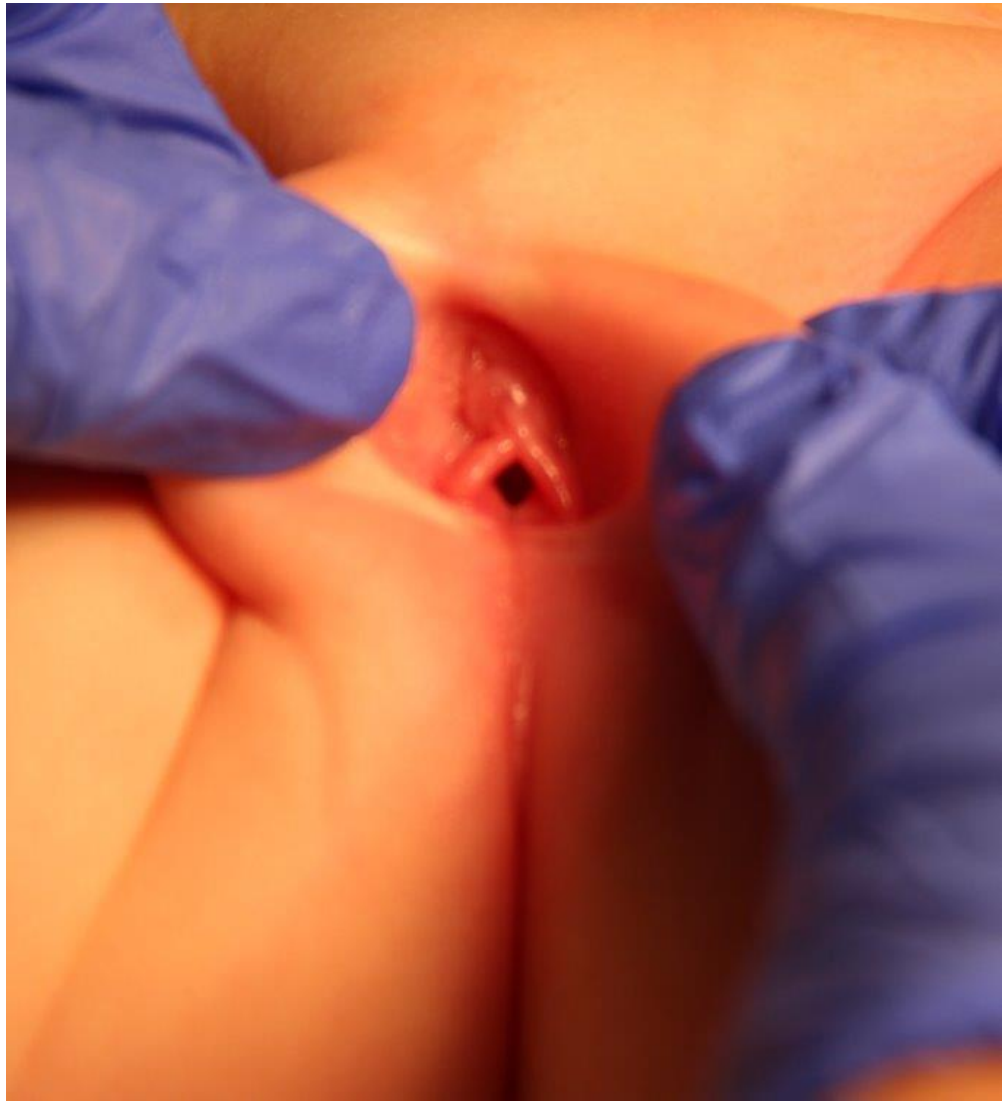
Initial OR

- 10 weeks old – OR for PSARP
  - Difficulty placing foley > 45 minutes
  - Colorectal intra-operative consultation

Hmmm… that's interesting. Any concerns before proceeding as planned?

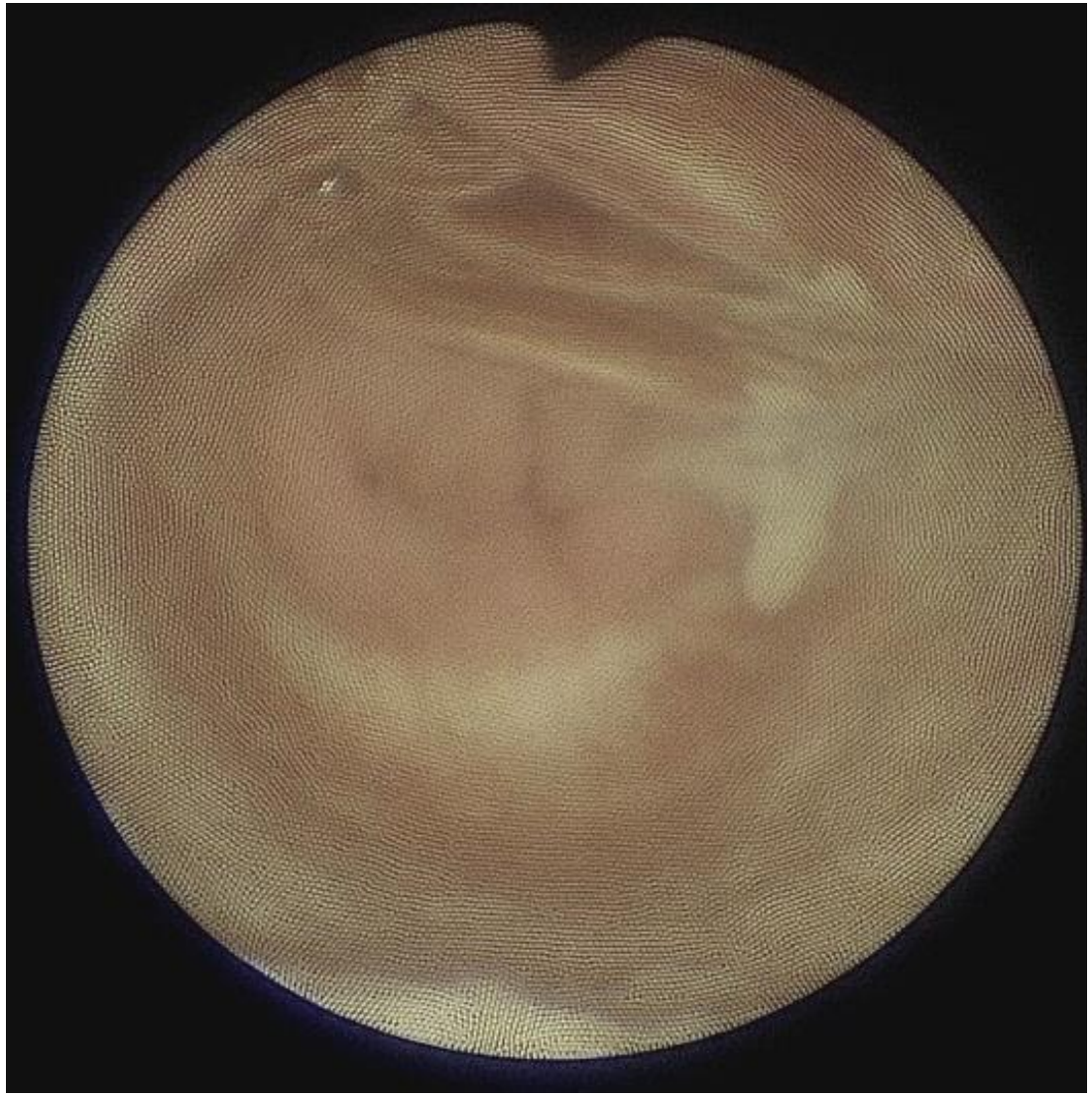
1. No – Proceed as planned. A difficult foley isn't out of the ordinary.
2. No – Proceed as planned but ask urology to evaluate patient on this admission and determine if she needs further work-up.
3. Yes – Rectovaginal fistulas are exceedingly rare. Can I see the perineum?

- Exam – cloaca
  - Short common channel – can see urethral orifice on exam
- Cysto/vaginoscopy -
  - 1cm common channel
  - Native vagina with single cervix
  - Normal urethra and bladder neck
- Case canceled
- Multidisciplinary follow-up in clinic > repair at later date



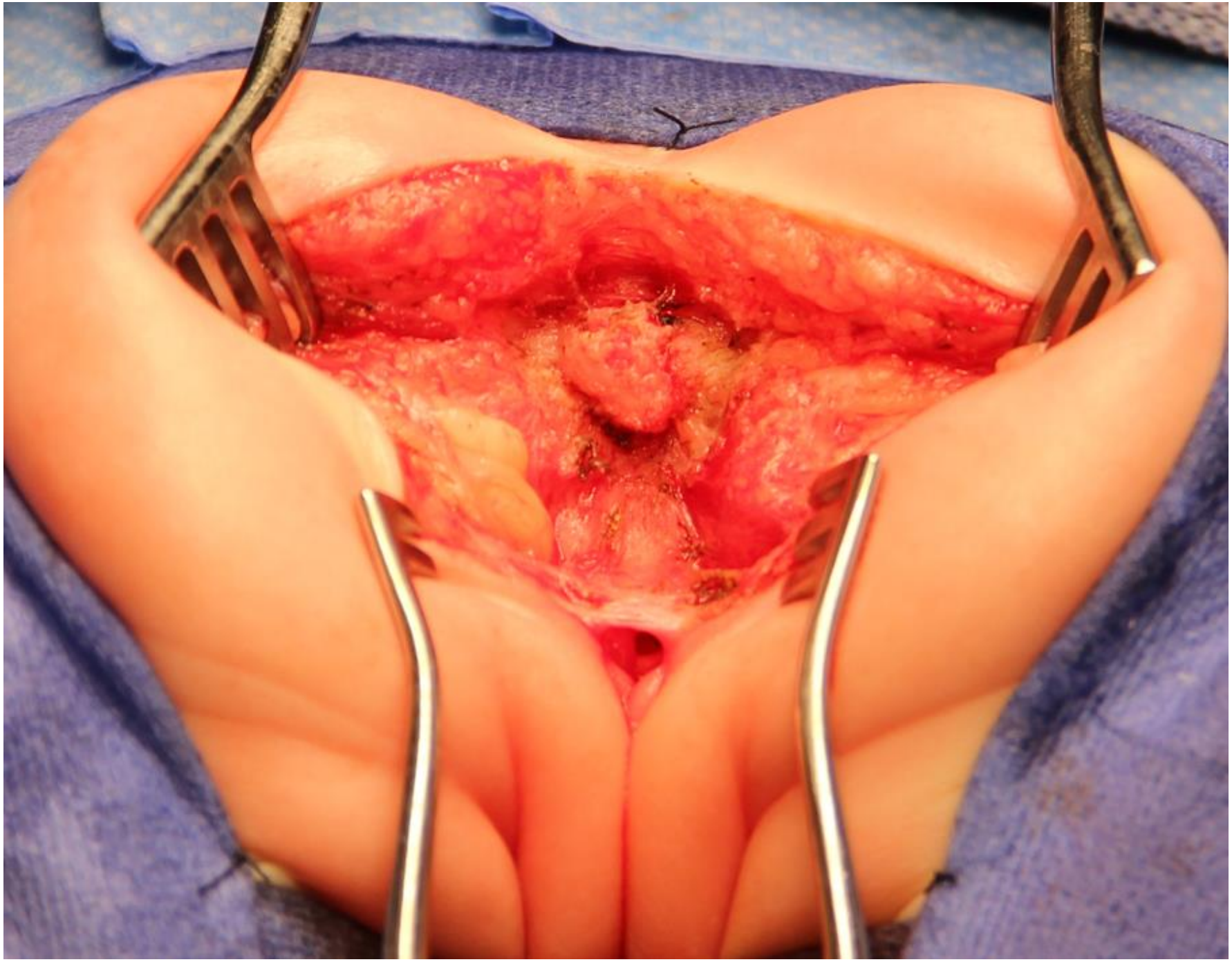
PSARVUP

- 4 months old
- Cysto/Vaginoscopy -
  - Common channel
    - 1cm – bifurcation urethra/vagina
    - 3 cm – rectum
  - Single vagina – 7 cm
    - Single cervix with midline septation
      - Suggests septated uterus
  - Normal urinary tract anatomy

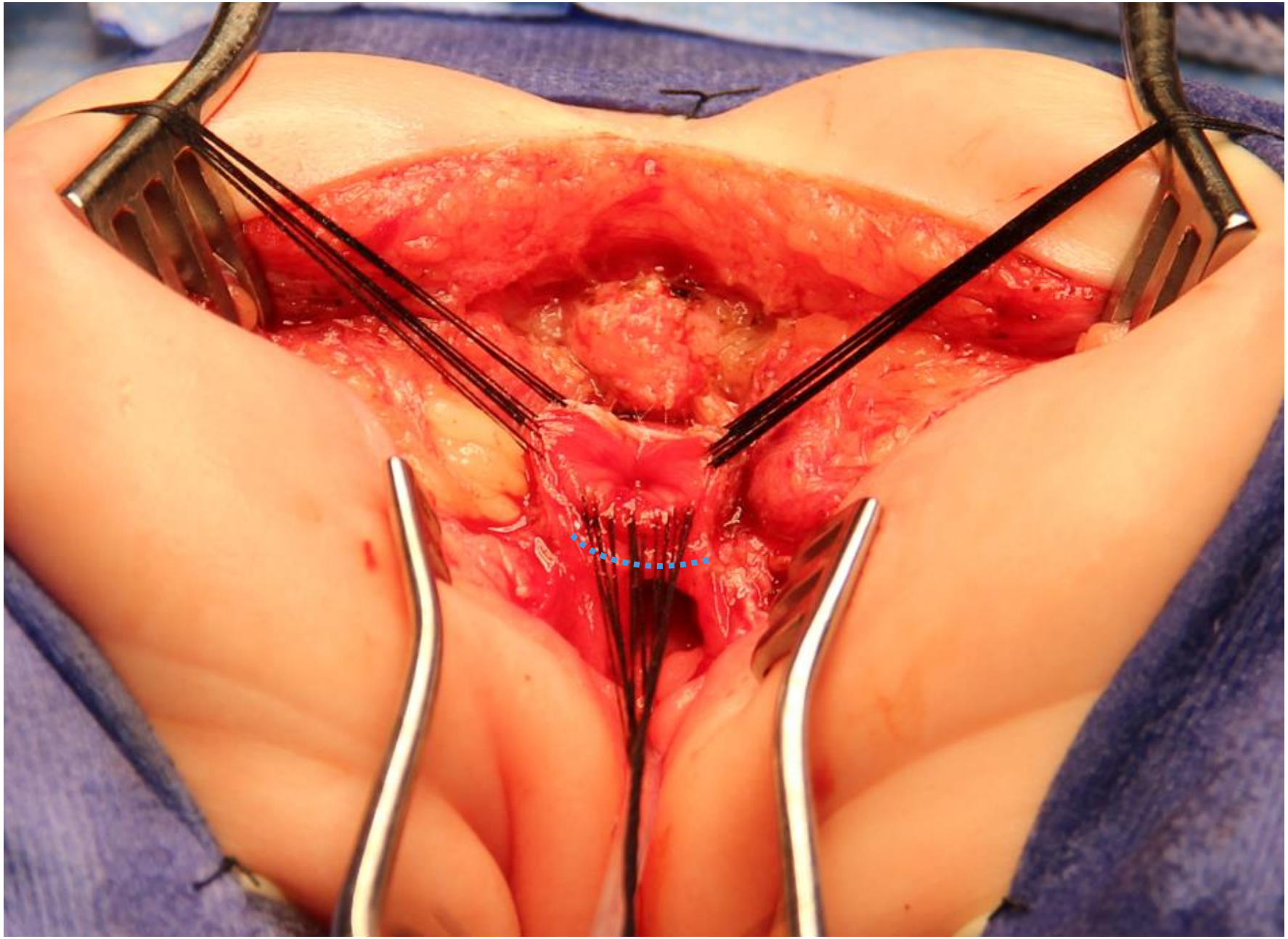


What is your operative approach based on these findings?

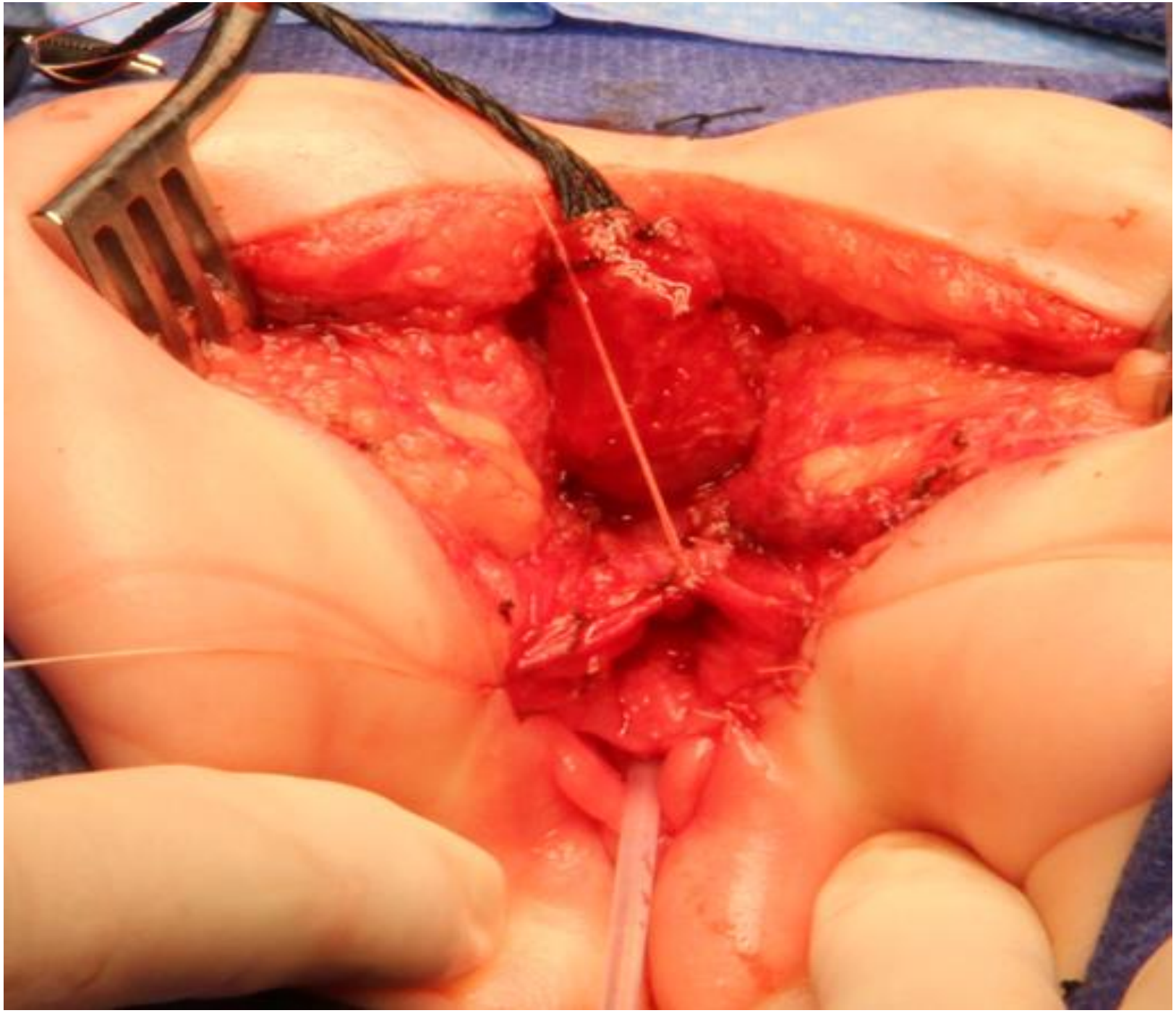
1. Posterior sagittal approach only
2. Total body preparation with plan for laparotomy and posterior sagittal approach



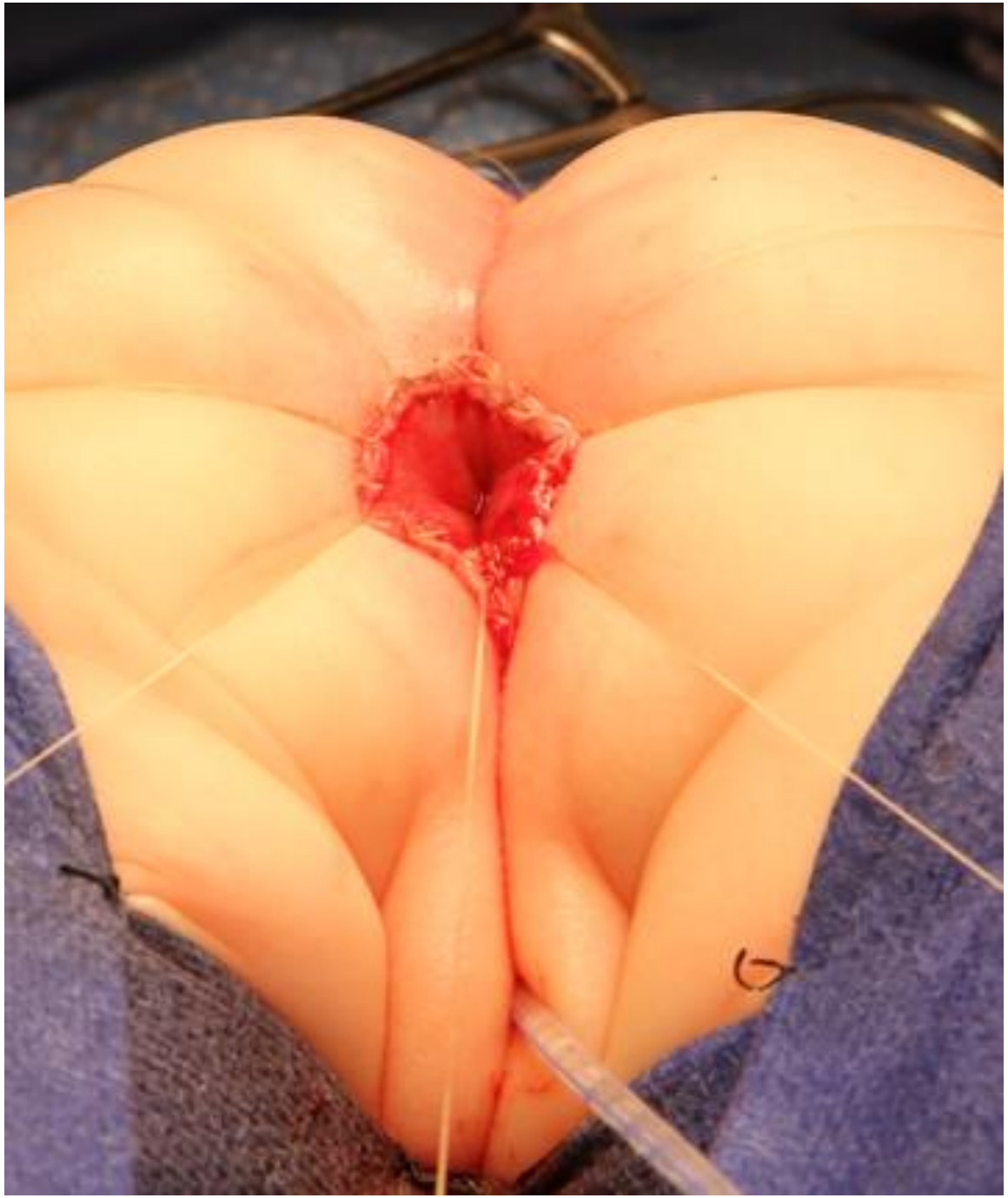
- Posterior sagittal approach
- Delineate –
  - Entire common channel
  - Clearly ID rectum, vagina, urethra
- Place foley



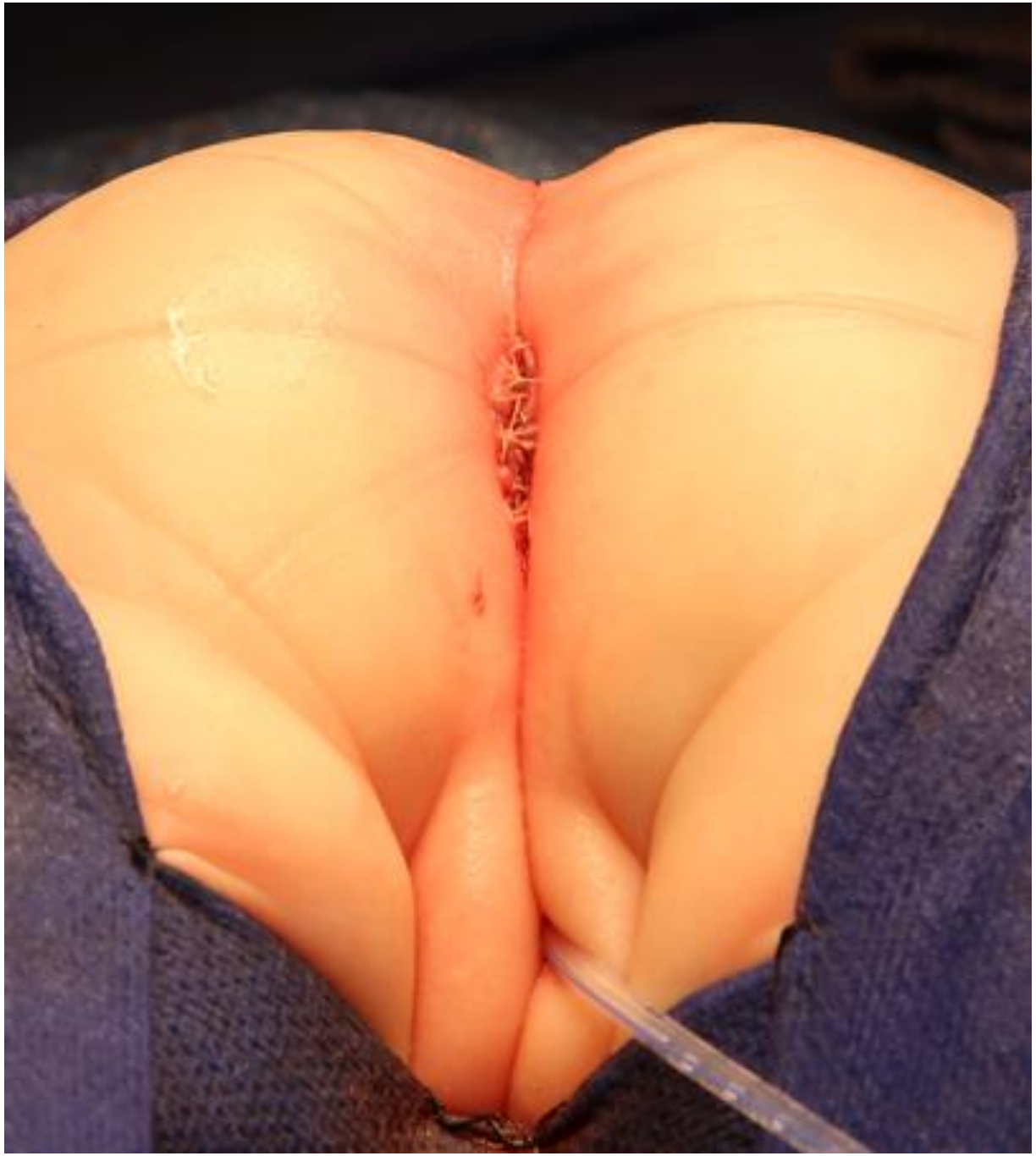
- Open distal rectum > 5-0 silk circumferentially for uniform traction
- Dissection of lateral rectal walls
- Anterior dissection > separate rectum from vagina



- Anterior dissection until rectum and vagina were distinct and separate structures
- Mobilization proximal rectum > ensure tension free anoplasty
- Minimal mobilization of posterior wall of vagina > vaginoplasty without TUG mobilization
- Mark limits of sphincter
- Perineal body reconstruction
- Vagina well perfused, no tension



- Sutures from posterior edge of muscle complex into back wall of the rectum > ensure rectum lay within sphincter mechanism
- Closure of posterior sagittal incision in layers
- Anoplasty in 2 layers
  - Deep layer x 4 quadrants
  - Trim distal rectum (3 mm)
  - Superficial layer



- Anoplasty performed under slight tension > anus retracted slightly after sutures cut, no evidence of prolapse

- Post-op care -
  - Bacitracin x 5 days
  - Double diaper x 1 month
  - Foley x 1 day
  - Follow-up appointment – 2 weeks post-op
    - Start dilations
      - BID, increase size qWeek
  - Colostomy closure once final dilator size reached (~ 4-6 weeks)