CASE PRESENTATION

A COMPLICATED PULL THROUGH PROCEDURE

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CASE HISTORY

- I/O B.C
- 3 yrs. Female

- Referred by NICU- 2019
  - History of vomiting feeds and abdominal distension at Day 2 of life. AXR- showed dilated loops of bowel.

- Provisional diagnosis of Hirschsprung disease
  - Rectal washouts done- yielded temporary and inadequate decompression
  - surgical decompression therefore required

PROCEDURE

- day 7 of life- plan to do levelling colostomy vs. a primary procedure

- Intraop (laparotomy)- Frozen sections:-
  - no ganglion cells at the peritoneal reflection
  - ganglion cells at 10cm and 15cm above reflection

- Transition zone was noted up to 10cm level, with irregular tenia coli

POLL

- Would you attempt a primary pull through procedure at your institution?

  - Yes, my institution is well supported for this
  - No, I will not attempt

PROCEDURE

- Decision made to do SOAVE PROCEDURE

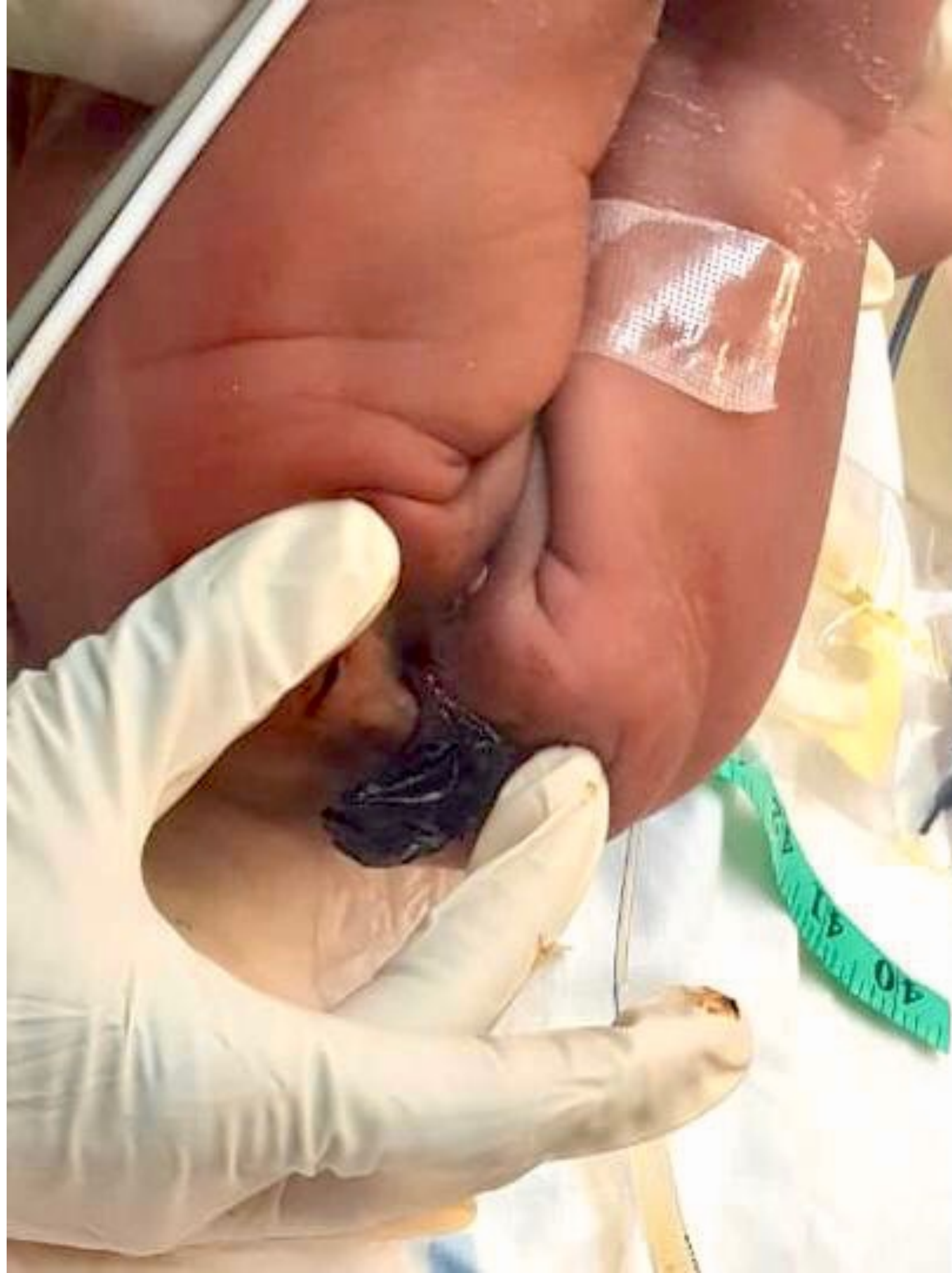
- Soave procedure performed
    - at 15cm mark
    - Distal resection at peritoneal reflection
    - Rectal seromuscular cuff
    - Tension free
    - Non-sutured- 4cm bowel external to anal verge

POLL

- What is your definitive procedure of choice?
  - Swenson
  - Soave
  - Duhamel
  - Transanal endorectal- if appropriate

CASE HISTORY

- Day 2 post op revealed non-viable bowel at anal verge
- Revision attempted
  - Revealed necrosed bowel exactly at puborectalis sling
  - The residual sigmoid and lower descending colon was mobilized and covered in moist sterile field to assess viability, however after 30 minutes, distal segment appeared dusky and threatened
  - Amputation to the level of viability
  - End stoma was fashioned at distal descending colon



Necrotic bowel seen at anal verge, Day 2 postop

CASE HISTORY

- Silastic Urinary catheter used as stent to keep distal rectum patent, and encourage epithelialisation

- Parent taught how to do DRE daily at home

- Referral made to International specialist centre via telehealth- unsuccessful attempt
    - Covid-19 travel restrictions for 2 years
    - No further financial benefits via local funding institutions

CURRENT EVENTS -2023

- Currently well
- Functional end stoma
- Gaining weight
- International referral made since 2019/2020- no longer viable option due to issue of funding
- Patient aware of her condition, Gets bullied at school about stoma
- Patient can be uncooperative and refuses to be examined PR

RECENT INVESTIGATION

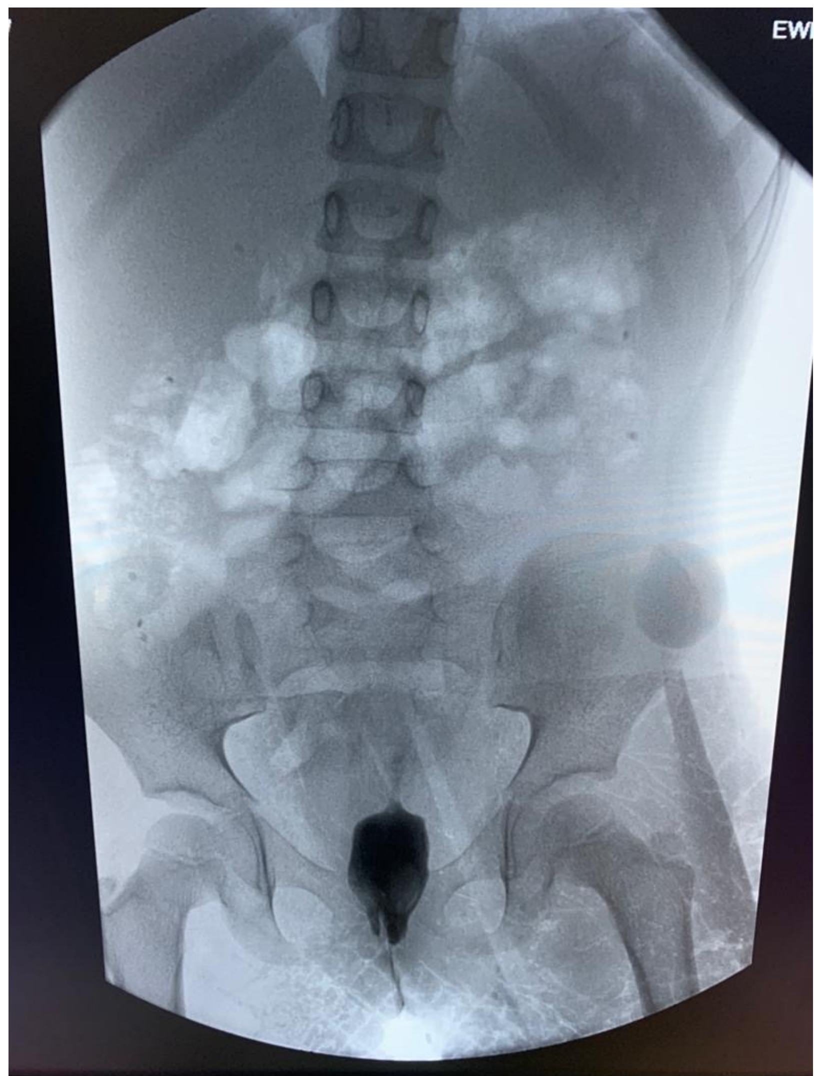
- <u>COLOSTOGRAM (31.05.2023)</u>
  - 8 Fr foley catheter inserted PR, balloon inflated 2ml

  - Fluoroscopy performed- ultravist (50:50), 20ml inserted. Able to visualize rectal pouch, with adequate distension

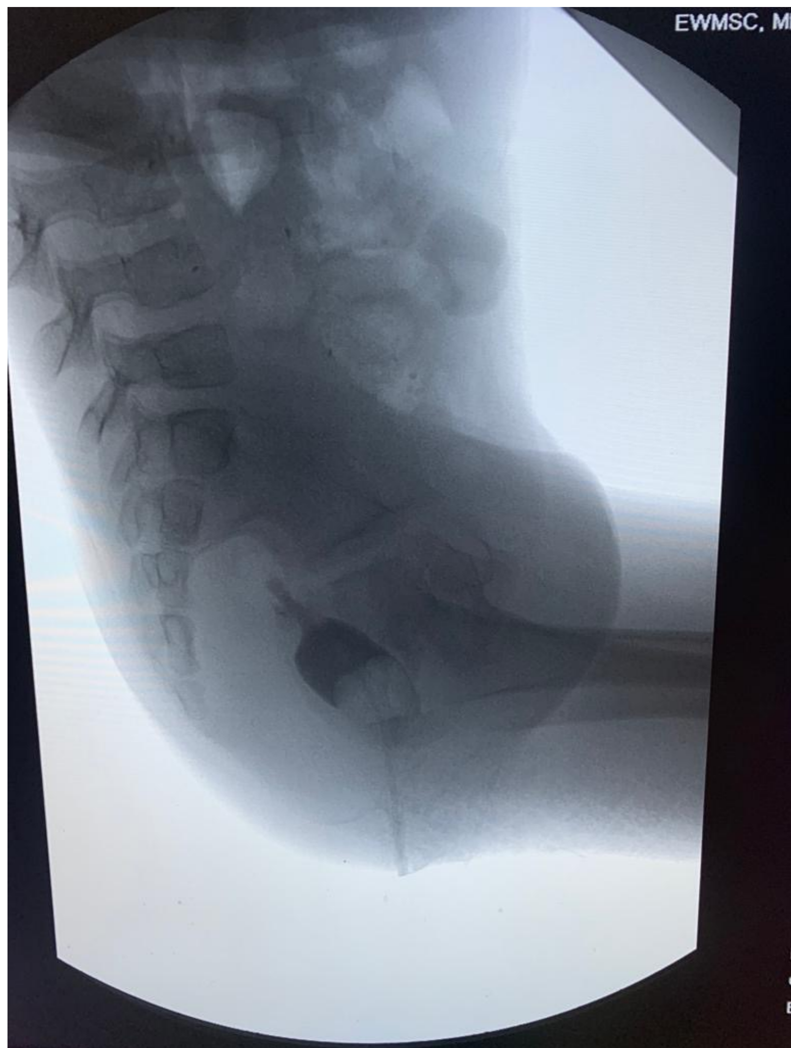
  - Catheter bulb deflated and 40 ml ultravist solution inserted while catheter was subsequently removed-revealed possible stenotic segment distally

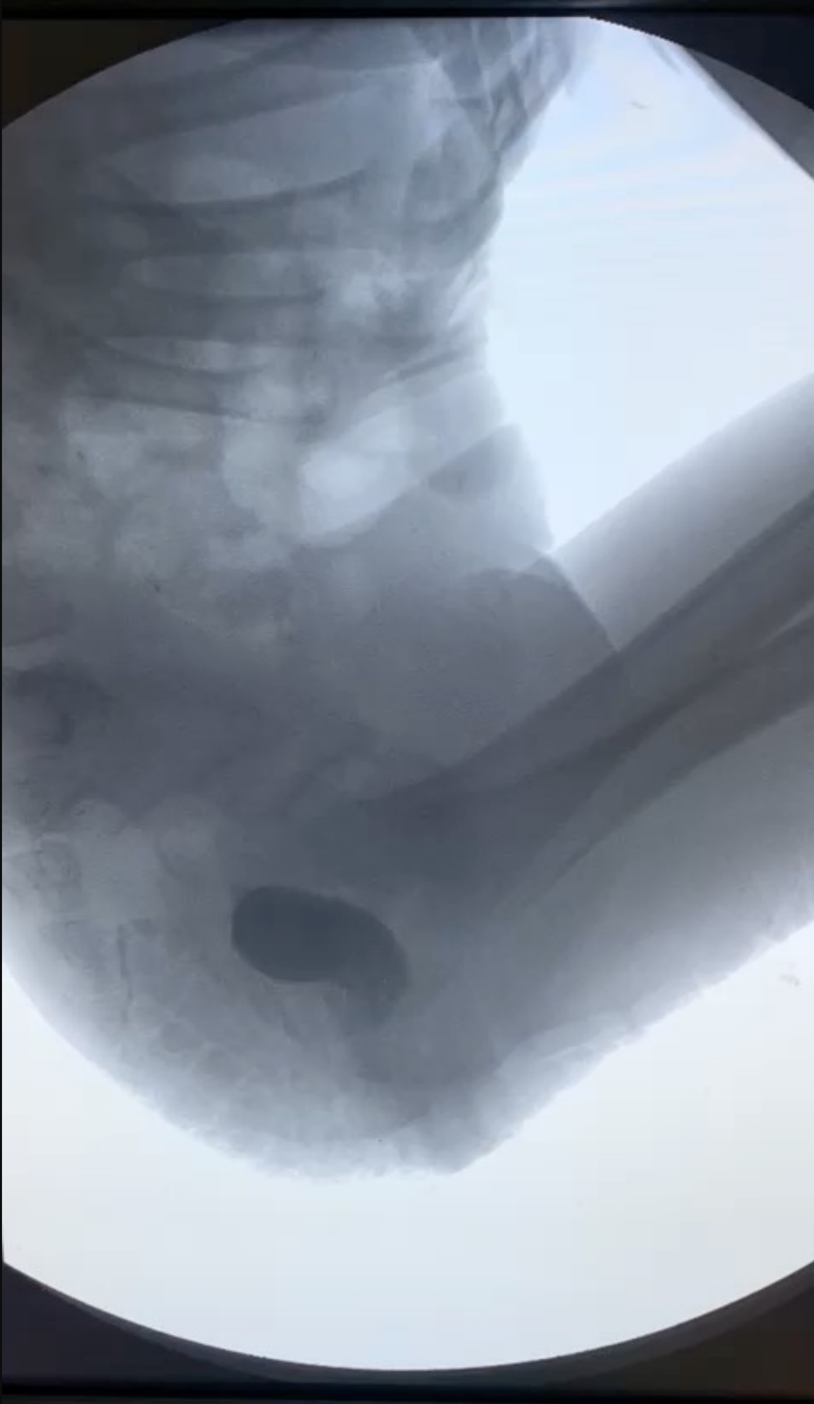
  - Proximally, 20 ml ultravist solution inserted through stoma, large bowel appear relatively normal/adequate

COLOSTOGRAM



COLOSTOGRAM



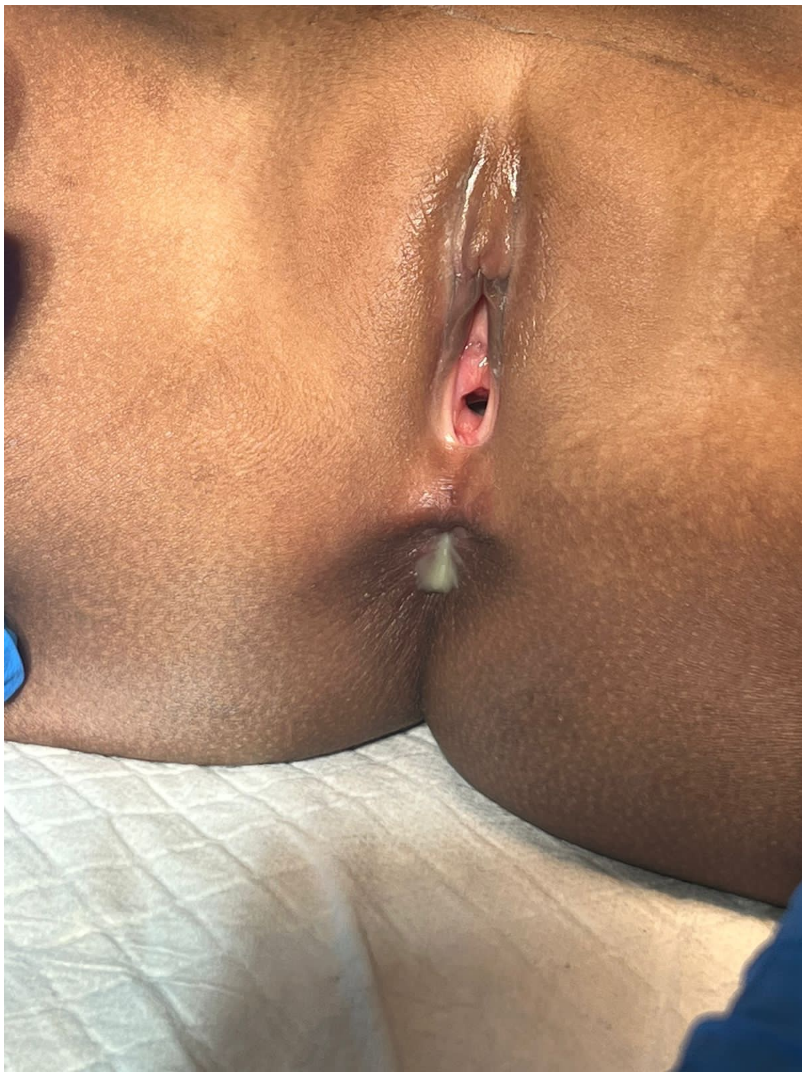


COLOSTOGRAM

CURRENT EVENTS- 2023

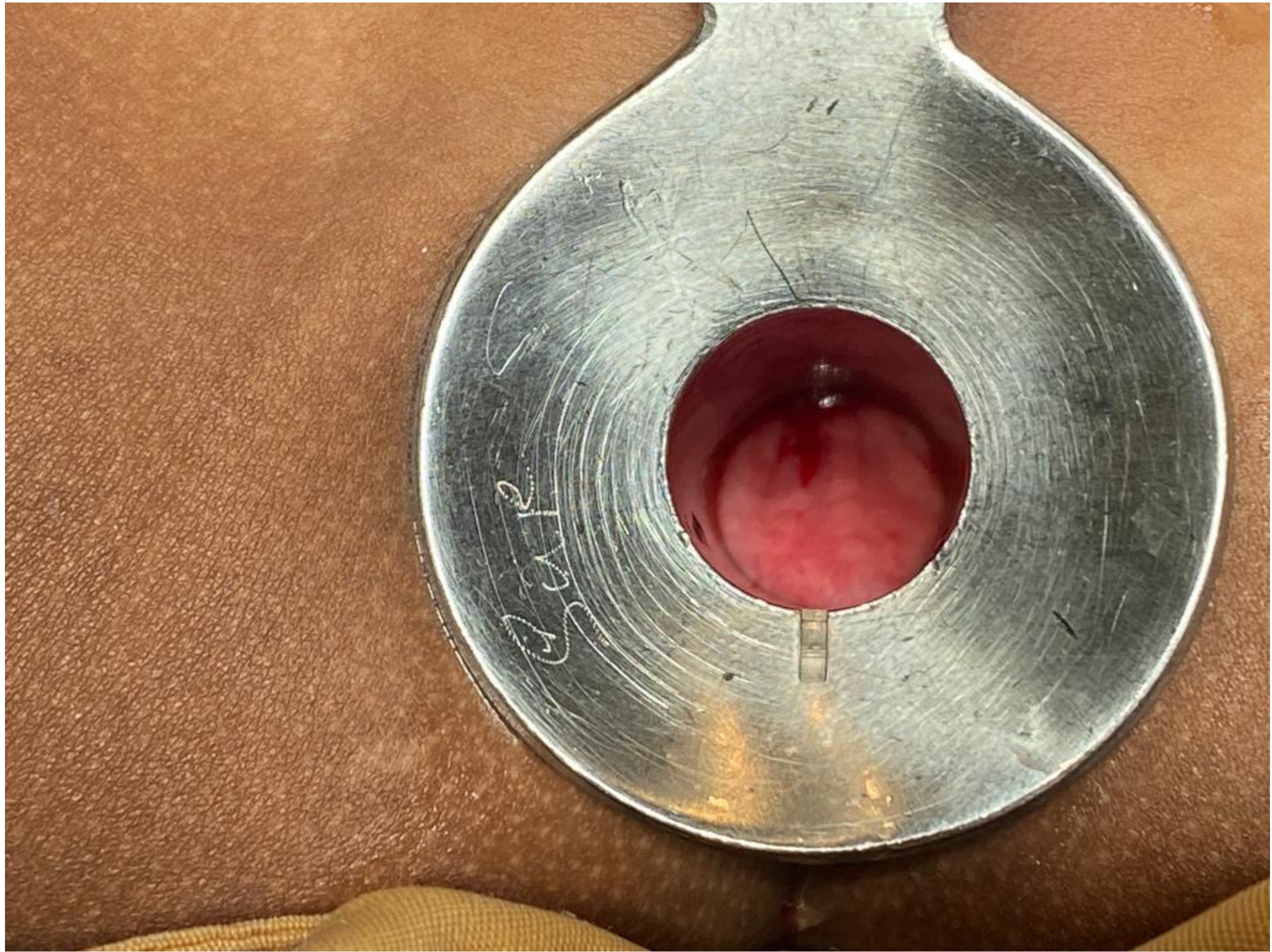
EXAMINATION

- Healthy, pink, functional end stoma
- Mucous PR
- EUA (12.06.23)
  - Pouchitis
  - DRE easily performed, can admit hegar 16
  - Anorectum not stenosed or fibrotic
  - Rectal pouch 5.5cm long (up to anal verge), anal sphincter 2cm
  - Pink healthy appearance of mucosa on proctoscopy, anal columns seen



Pouchitis on DRE

Proctoscopy findings



CHALLENGES

- Urgent need to make a decision about the next step
- Minimal surgical experience in salvage procedures
- Minimal surgical experience in transanal approach
- No local funding for referral to special centre
- Social issues as patient gets older

QUESTIONS

- What are the surgical salvage options in this patient?
- Would a Duhamel procedure be a suitable option now in this case?
- Is there any other information and/or investigations that may be required?
- With this recent clinical information, can the definitive surgery be performed locally with specialist guidance?
- OR, would international assistance at a specialist centre be more appropriate?

THANK YOU