

NL

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8-month-old infant previously followed in another hospital, recently moved to our region, is admitted to improve weight gain.

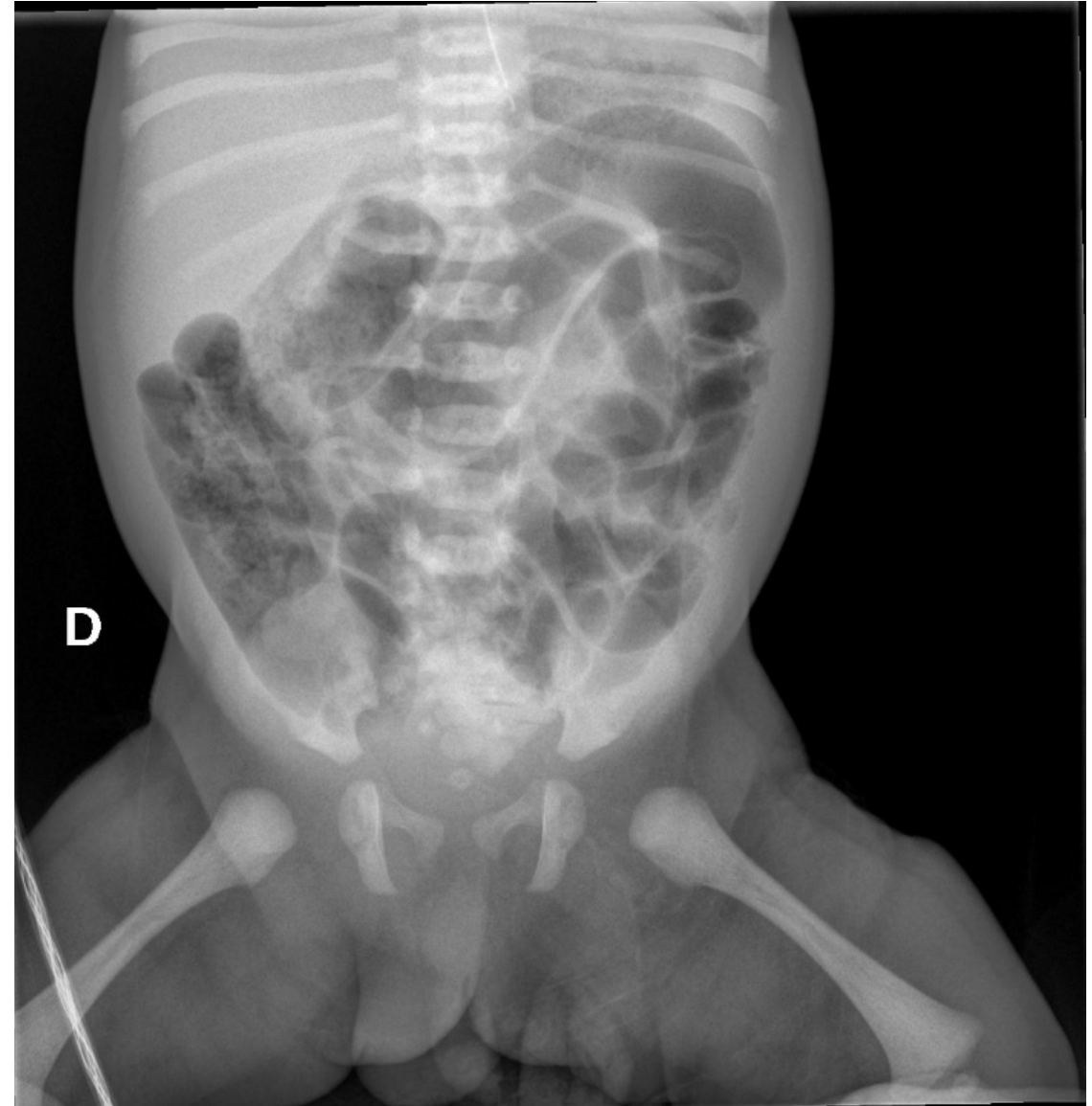
Parents reported multiples admissions due to “gastroenteritis”

- 5.9kg P1 OMS 60cm P<1

## PMH:

- FTB 37+2w + NWB  
3290kg
- Trisomy 21
- “Passed meconium in the first 24 hours of life, but scarce”

2<sup>nd</sup> DOL



## 4<sup>nd</sup> DOL

- “On 3rd day of life, abdominal distension was noted and question about meconium elimination. He was left NPO. Routine enemas by pediatric surgeon, but not effective.”

(enemas or irrigation???)

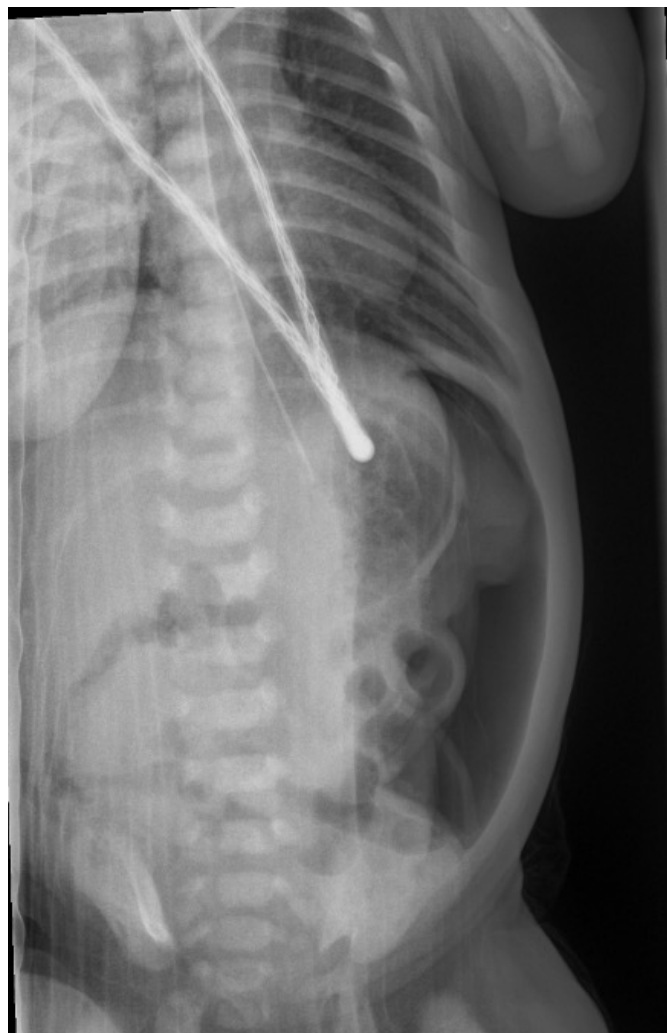


## 5<sup>th</sup> DOL

- Allegedly they performed a contrast enema, but there are no images
- Residual contrast seen iliac region



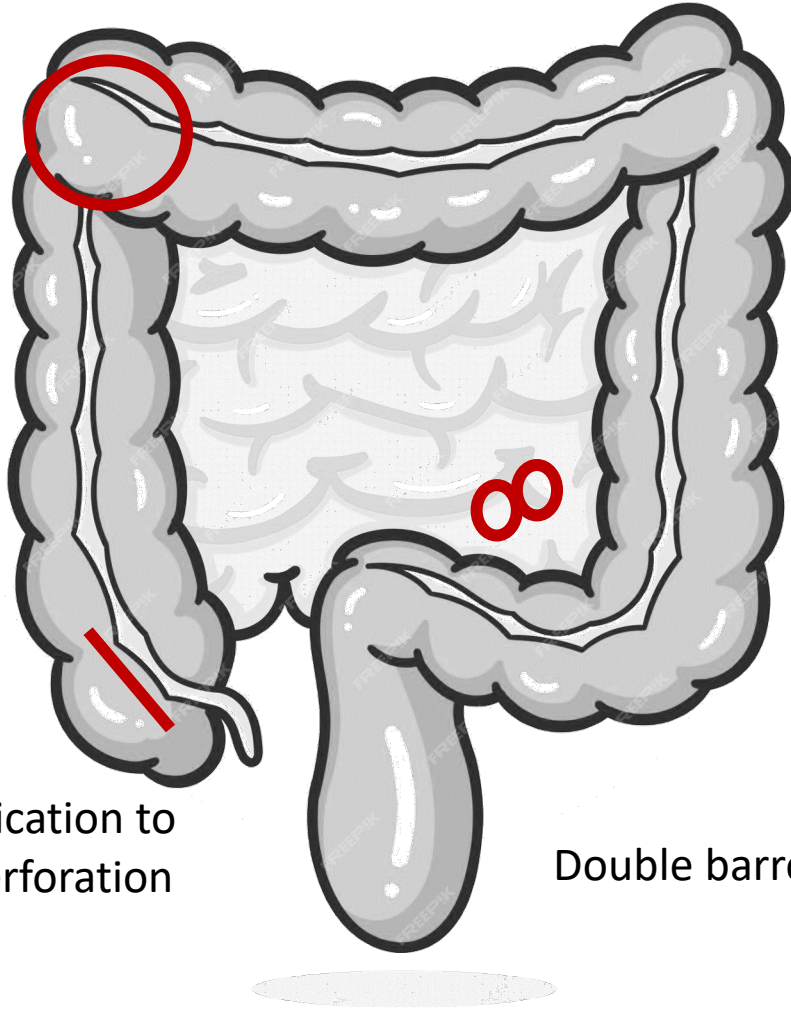
# 6<sup>th</sup> DOL



# 7<sup>th</sup> DOL: surgical intervention

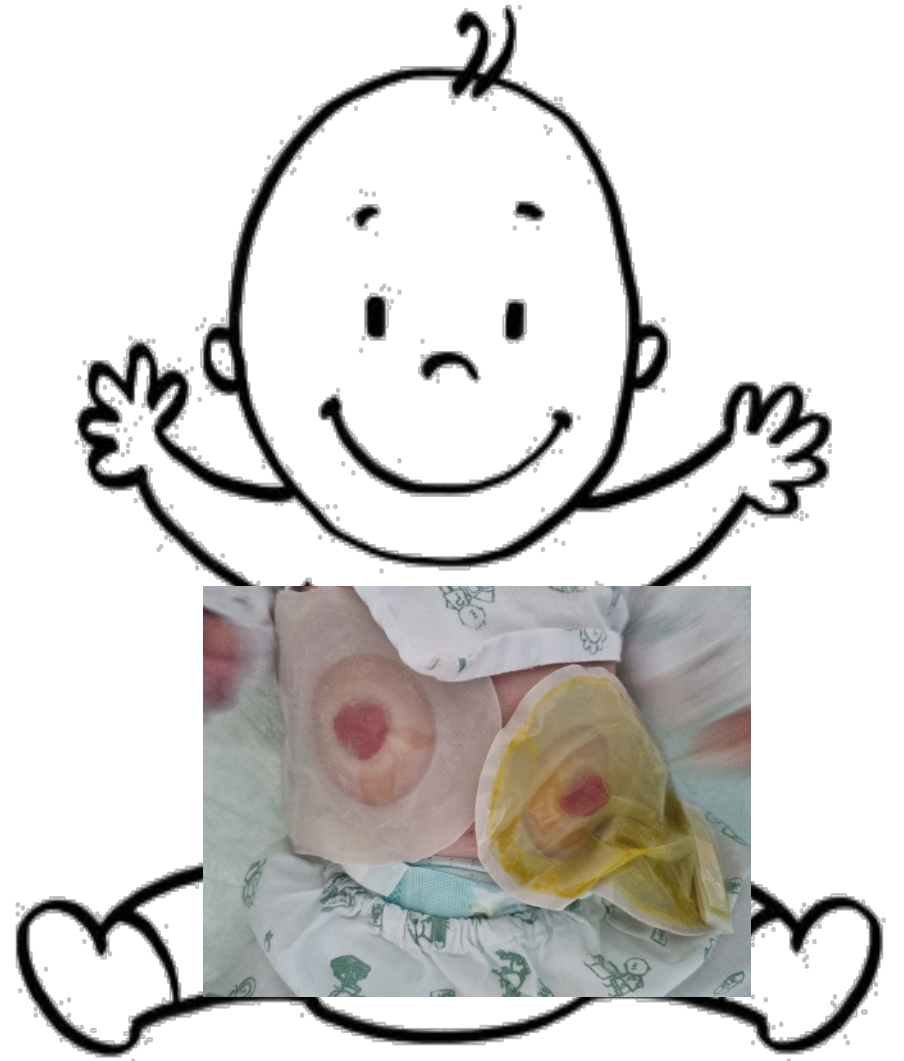
- Free intra-abdominal fluid and generalized intestinal inflammation
- Left colon caliber change with transition zone over the splenic flexure, 3cm funnel shape
- 2 irregular necrotic areas: cecum and hepatic flexure
  - Loop hepatic flexure colostomy
    - Biopsy from colostomy
    - Cecal plication to close perforation
- Double barrel ileostomy 15cm proximal to ICV - left iliac region

Loop hepatic flexure colostomy



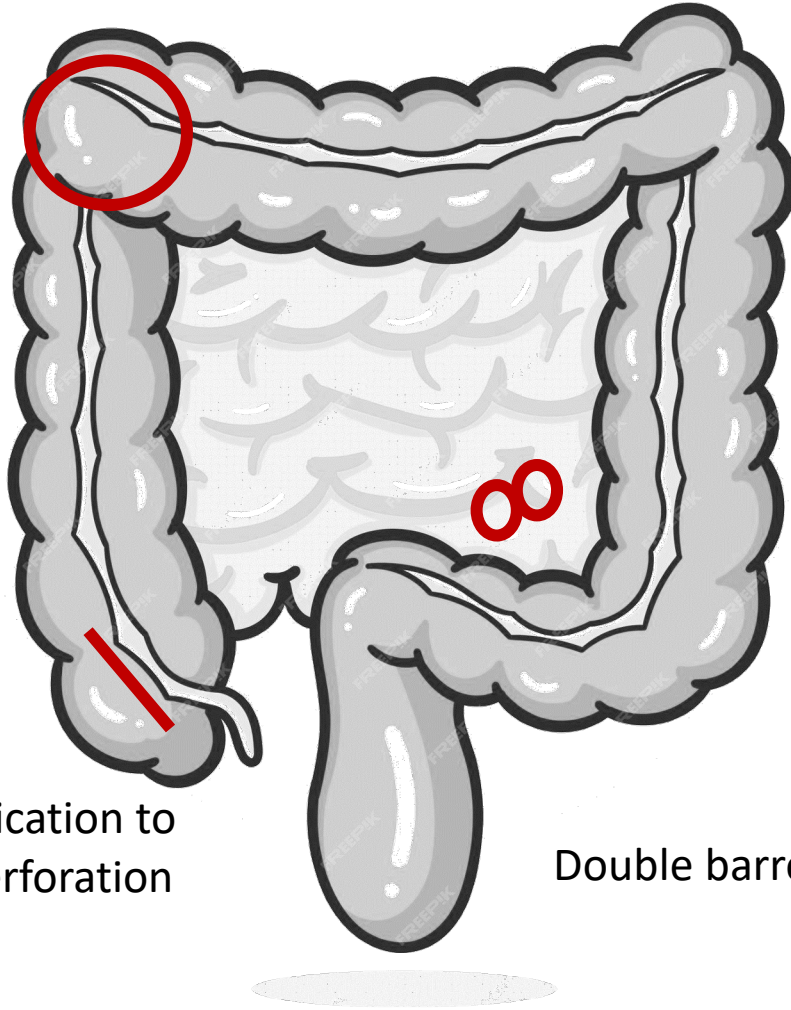
Cecal plication to close perforation

Double barrel ileostomy



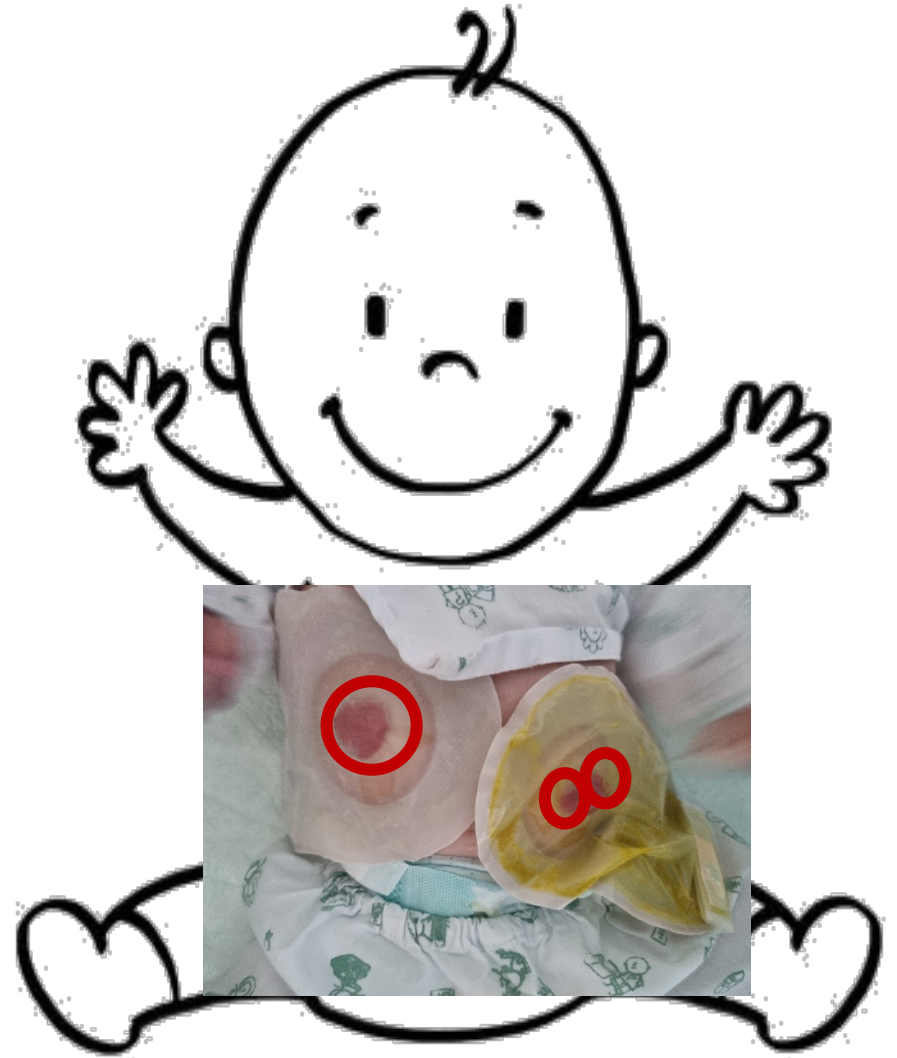


Loop hepatic flexure colostomy



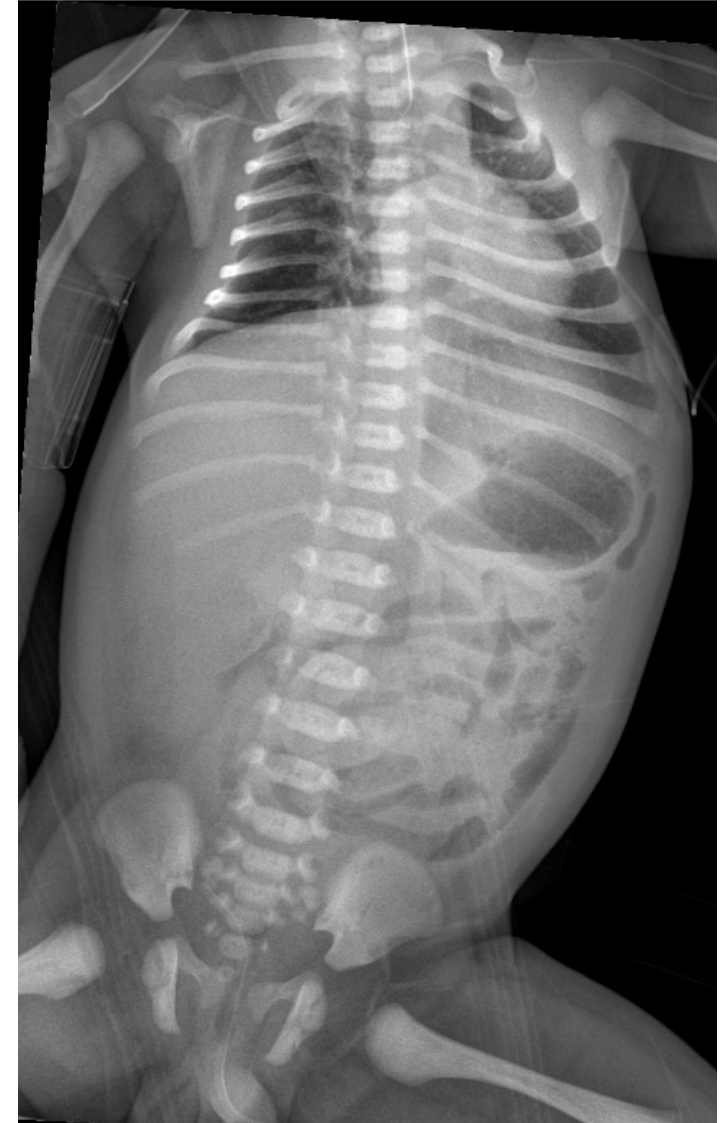
Cecal plication to close perforation

Double barrel ileostomy

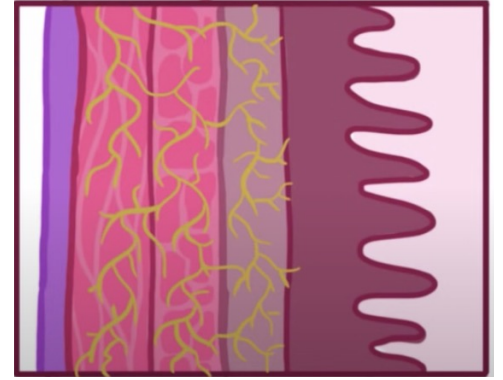


# Post-op admission

- PN for 9 days
- NPO 24 h, trophic to full feeding 2d
- After 24h initial bowel movement
- Mild laryngomalacia

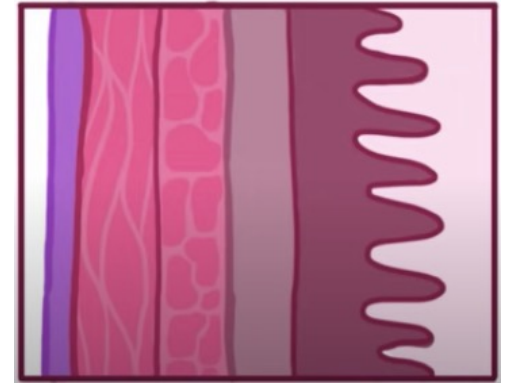


# Histopathology report



Large bowel. Mucosa with superficial ischemic necrosis and regenerative epithelial changes. Submucosal fibrosis. Transmural necrotic focus and signs of perforation. Presence of neuronal plexuses

# Follow up

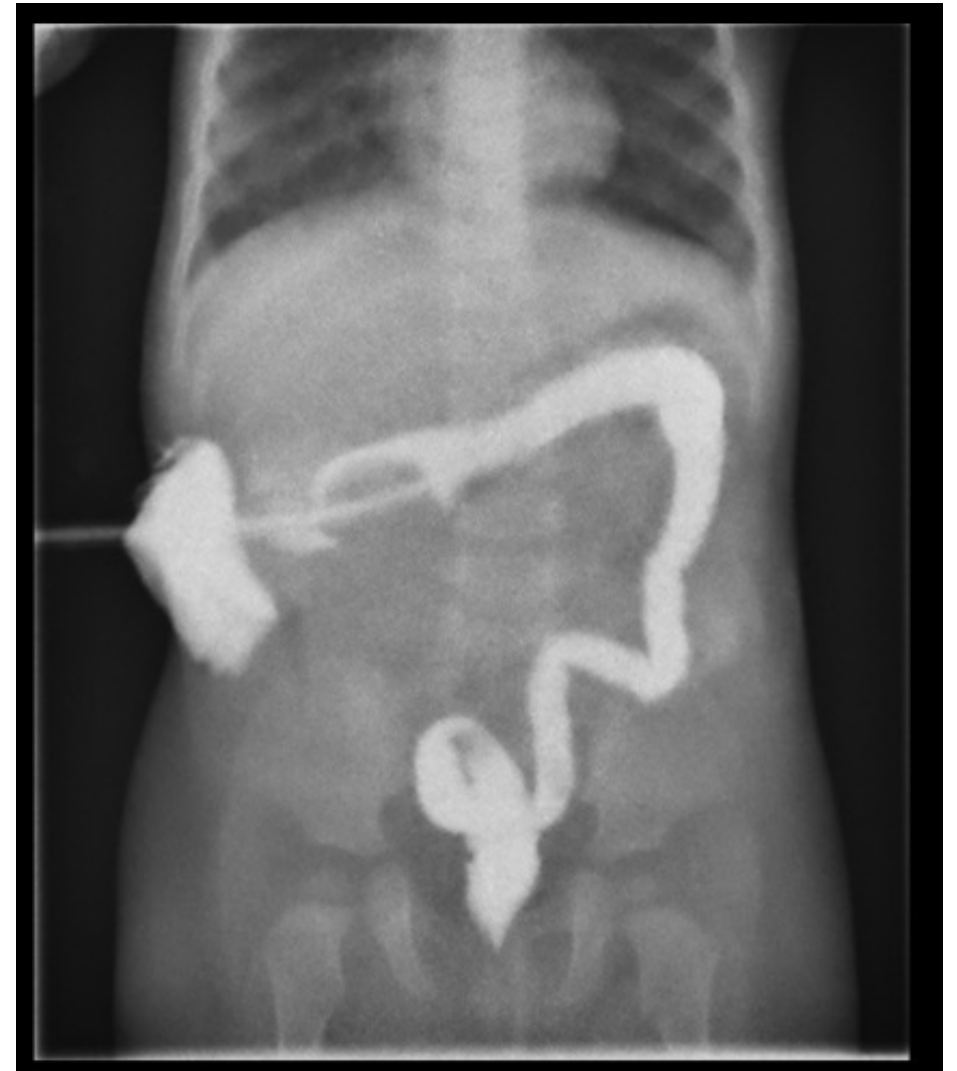


- Rectal biopsy: Fibrous fragment of 0.5 x 0.4 cm with suture thread. Anorectal mucosa with a minimal focus of intestinal mucosa. Inadequate biopsy (distal. Possible physiological hypoganglionism. Negative calretinin).

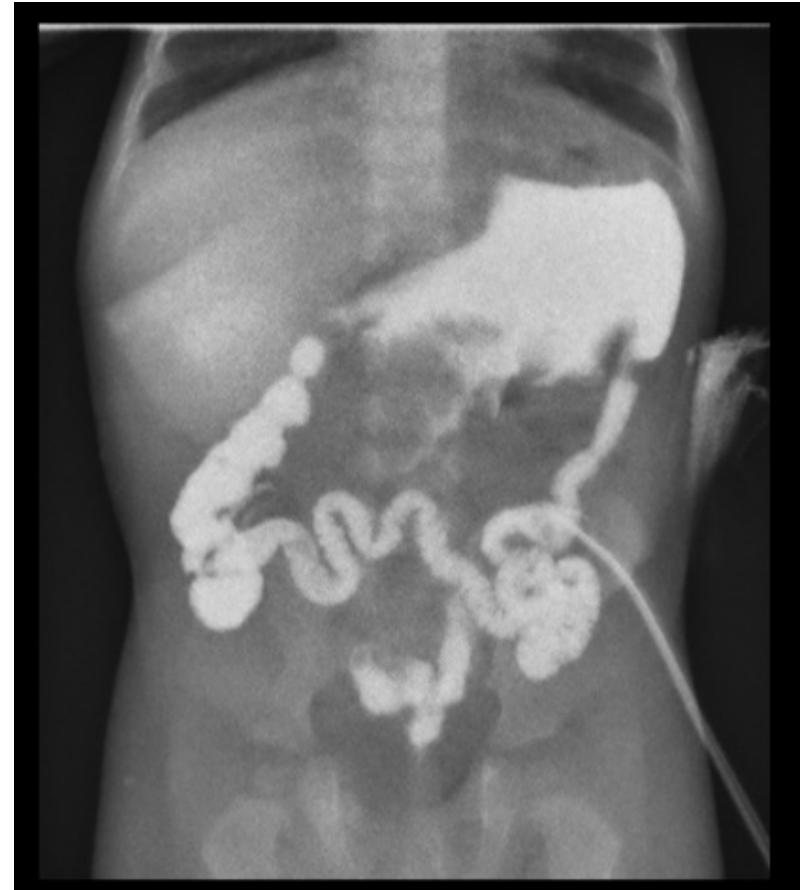
# Contrast enema



# From colostomy



# From ileostomy



- Clinical sepsis: pain, fever and prostration
- Admission to the ICU 2 days





- Is it really Hirschsprung Disease? → full-thickness open biopsy
- Is it a long-segment HD? → Colon mapping (laparoscopic-assisted vs only stomas)
- Failure to thrive +/- multiple HAEC → ileostomy closure + end colostomy vs leaving ileostomy (expecting long-segment)
- + left non-palpable testicle → Laparoscopic 1<sup>st</sup> stage Fowler-Stephens procedure