Instituto Nacional de Pediatría

Star Médica

INTERNATIONAL COLORECTAL WEB MEETING

Karla Santos Jasso MD, PhD

National Institute of Pediatrics, Mexico City

June 2024

Reason for consultation was a second opinion because the number 10 hegar dilator does not pass

MEDICAL HISTORY

19-month-old male patient

Down Syndrome

38 weeks, 2200 grs, severe sepsis with enterocolitis after birth (abdominal distension, feeding intolerance, and evacuation with fresh blood)

- **Abdominal ultrasonogram**: showed large dilation of distal ileum

- **Contrast enema**: Stenosis of the terminal ileum with probable intestinal malrotation

MEDICAL HISTORY (2)

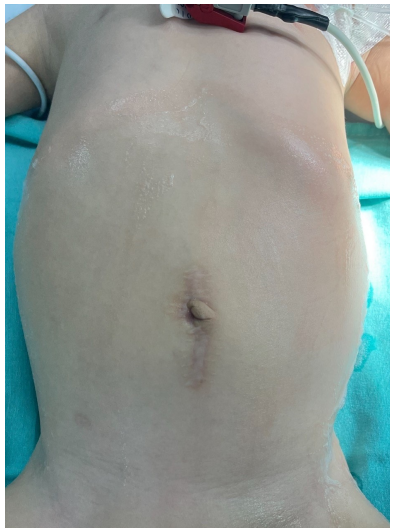
Rectal biopsy: aganglionic

23 days after birth

Transanal pull through (23 cm were resected)

Umbilical hernia with urgent procedure probably incarcerated, they found pus in the abdomen
They did a midline laparotomy and found apendicitis (1 yo)

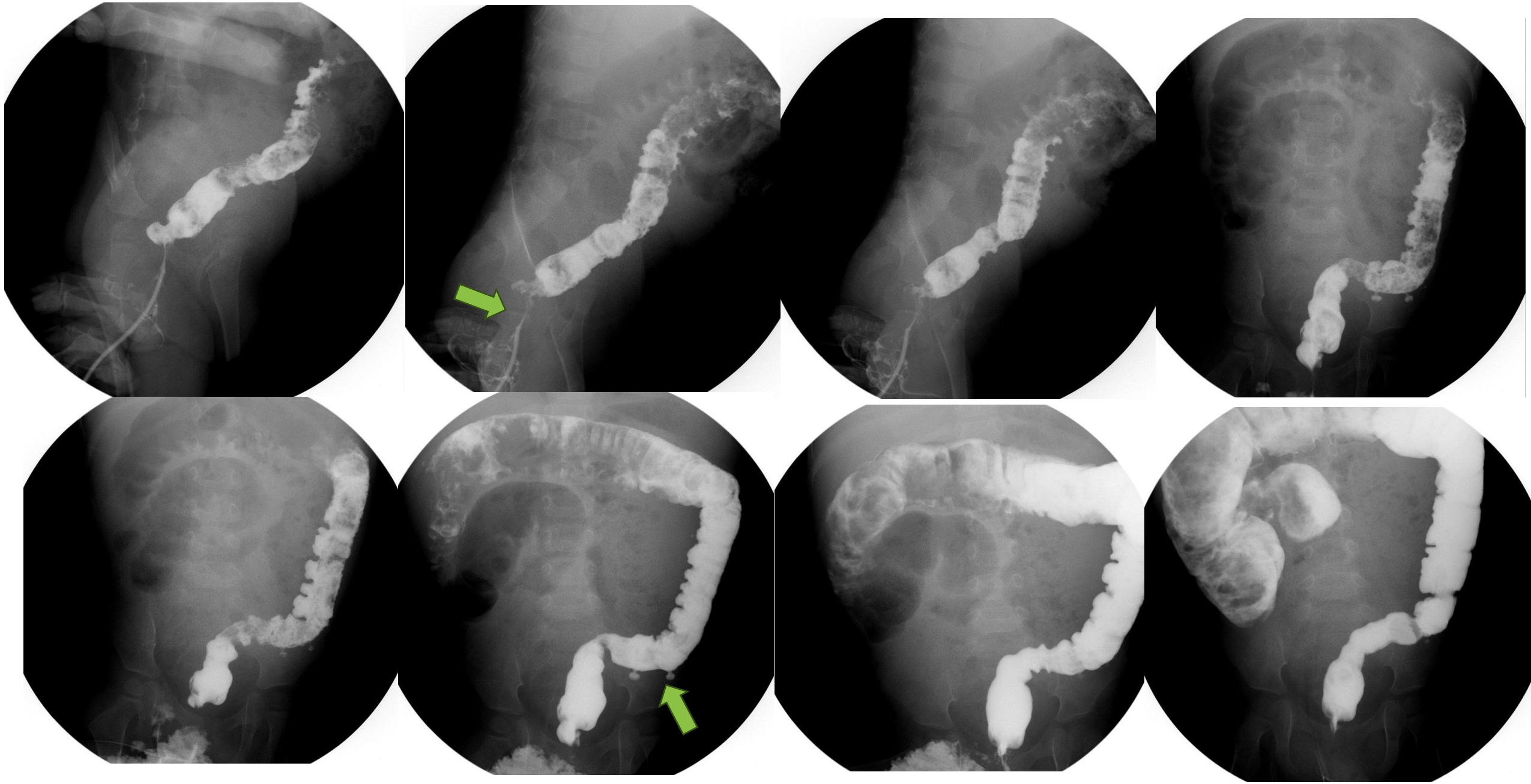
Clinical examination



- 19 months
- 6.6 kilograms
- Abdominal distention+++
- Only accepted Hegar dilator number 6 above anal margin
- There is a rectal stenosis approximately 3 cm above anal margin, fibrotic tissue was palpable on rectal examination

- We performed a rectal irrigation with two liters of saline solution and the adbomen improved considerably, the probe used was a 12fr .

The mother learned to do rectal irrigations with this amount of solution, she did them for 3 weeks, where she observed an increase in weight to 7.6 kilos and an increase in appetite



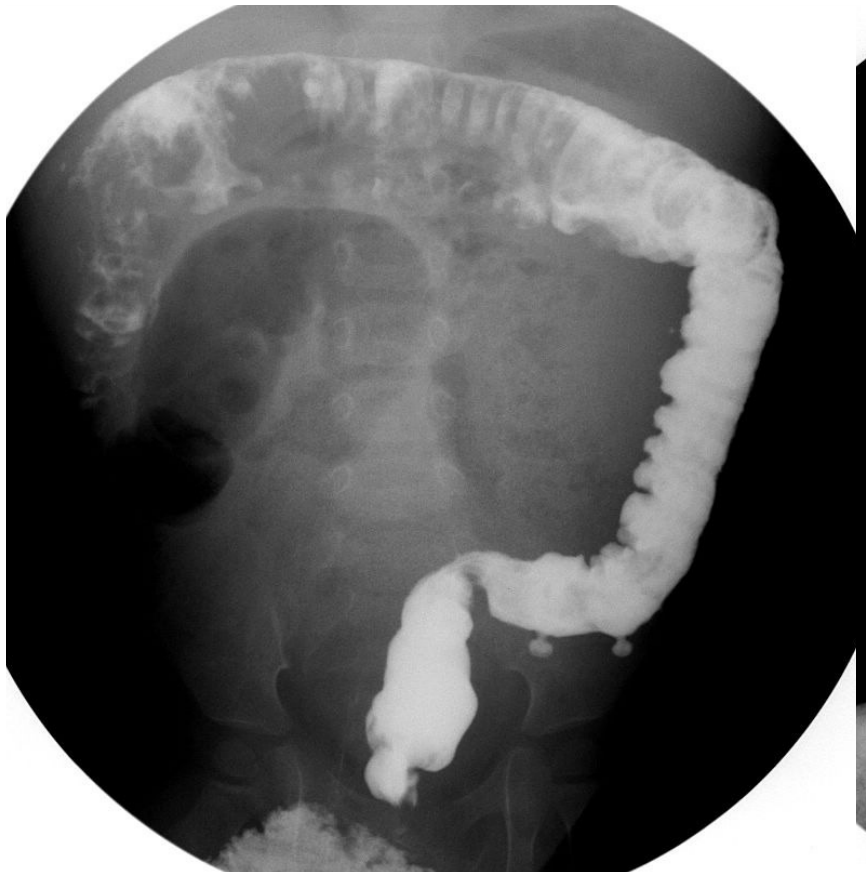
23 days after birth

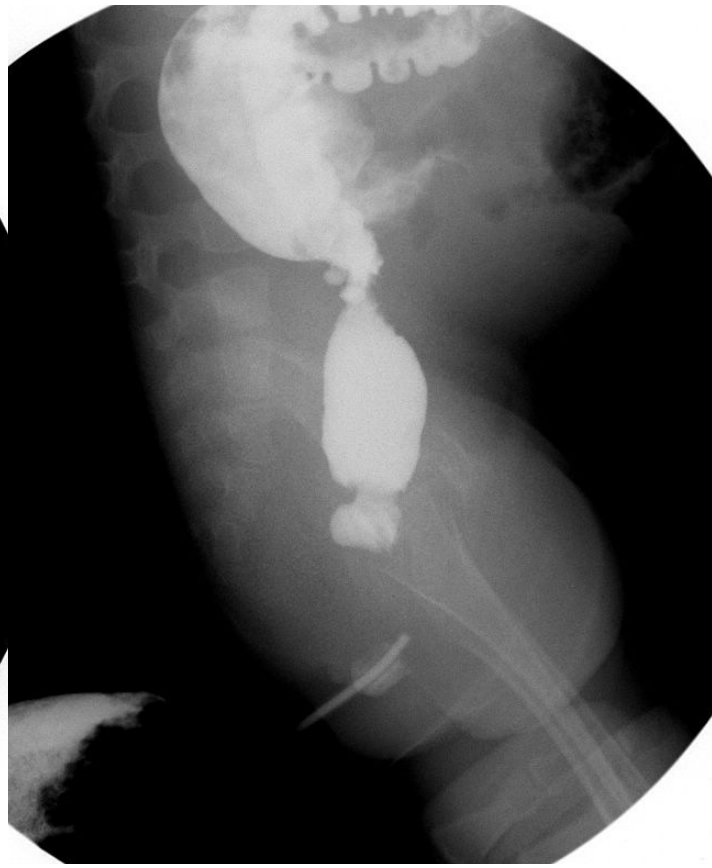
Transanal pull through (23 cm were resected)

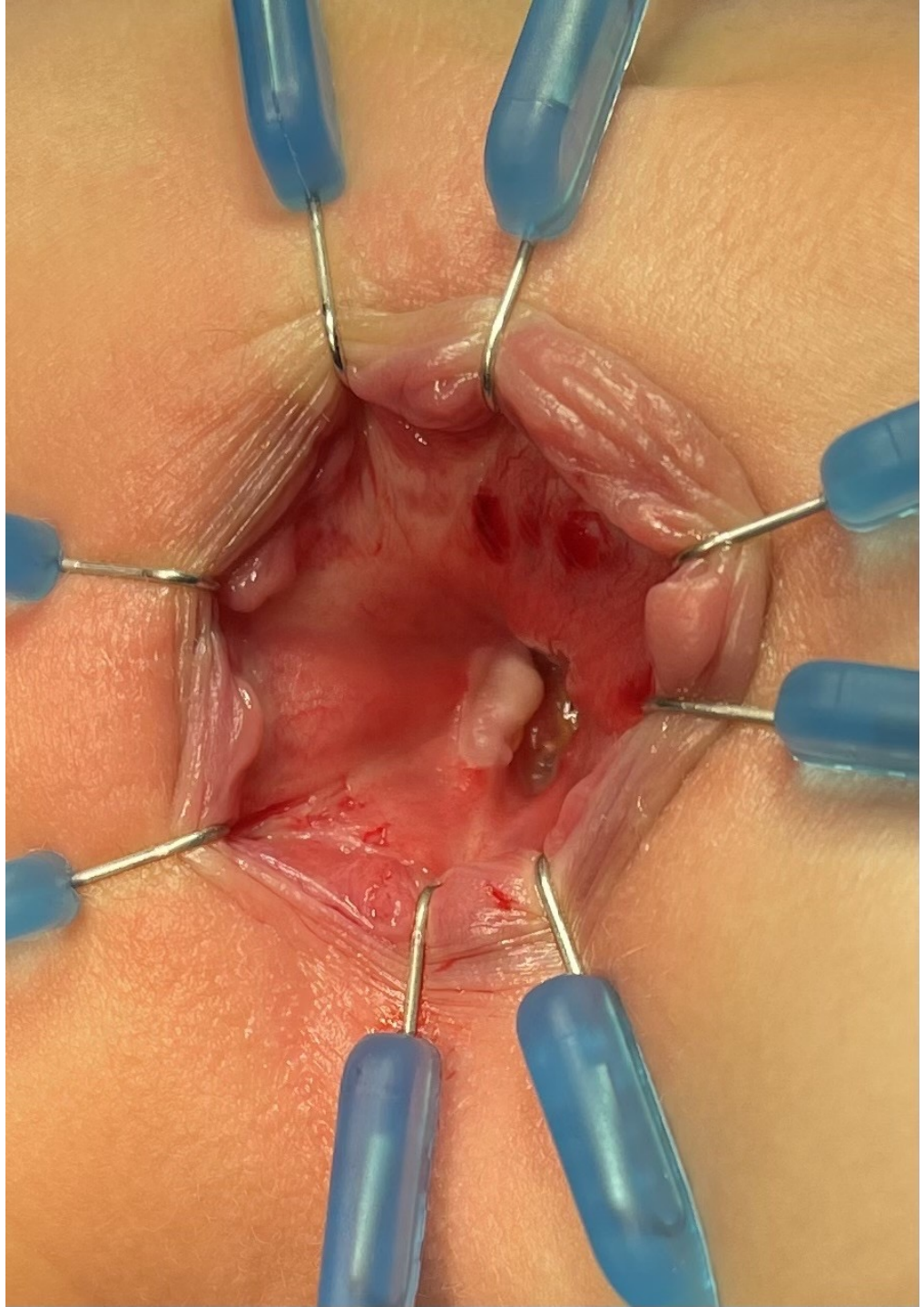
Definitive pathology report: Moderate chronic proctitis with transmural inflammation **hypertrophy of submucosal and myenteric plexuses with absence of ganglion cells no ganglion cells identified by immunostaining, calretinin negative** CD117 negative

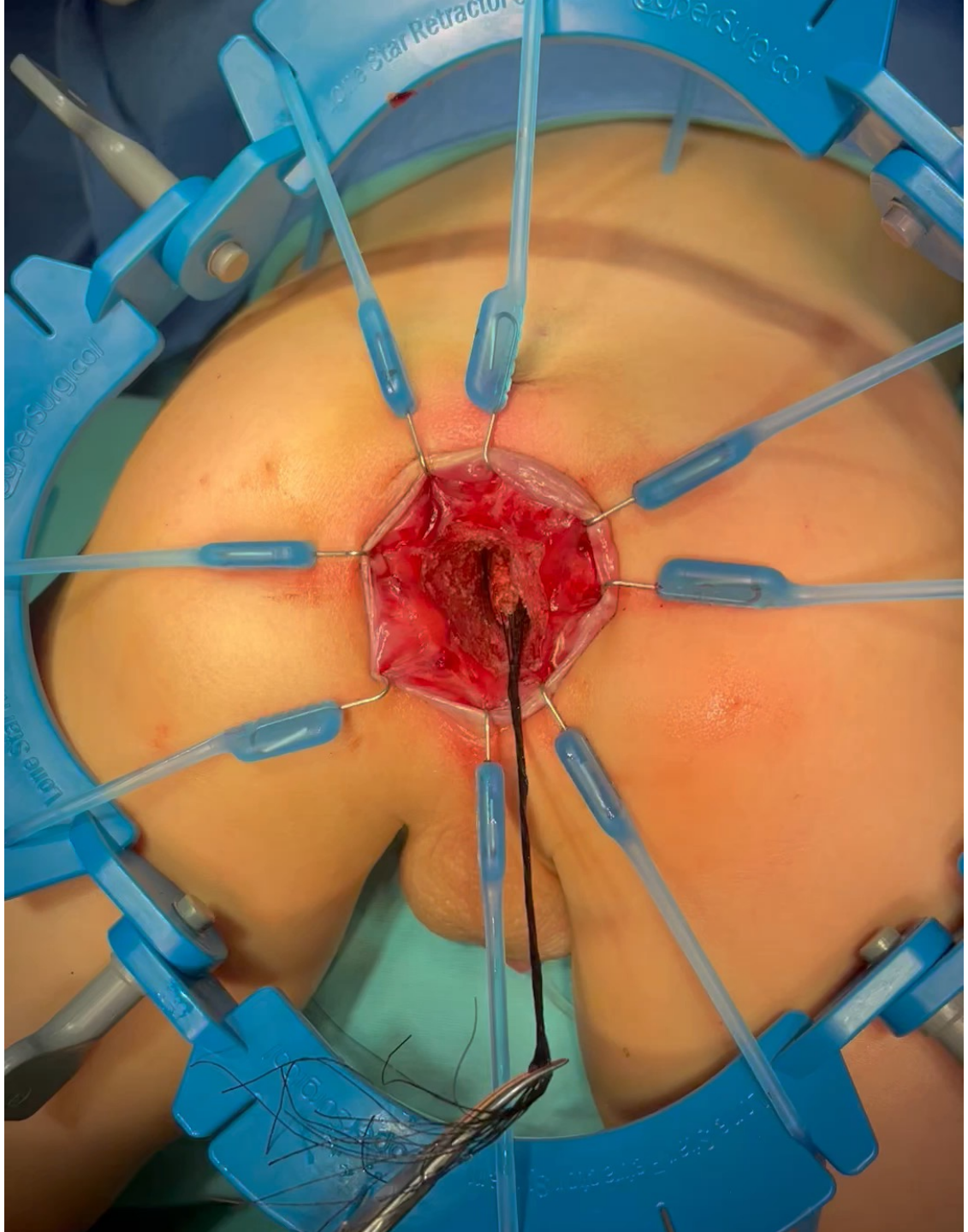
Which operation would you perform?

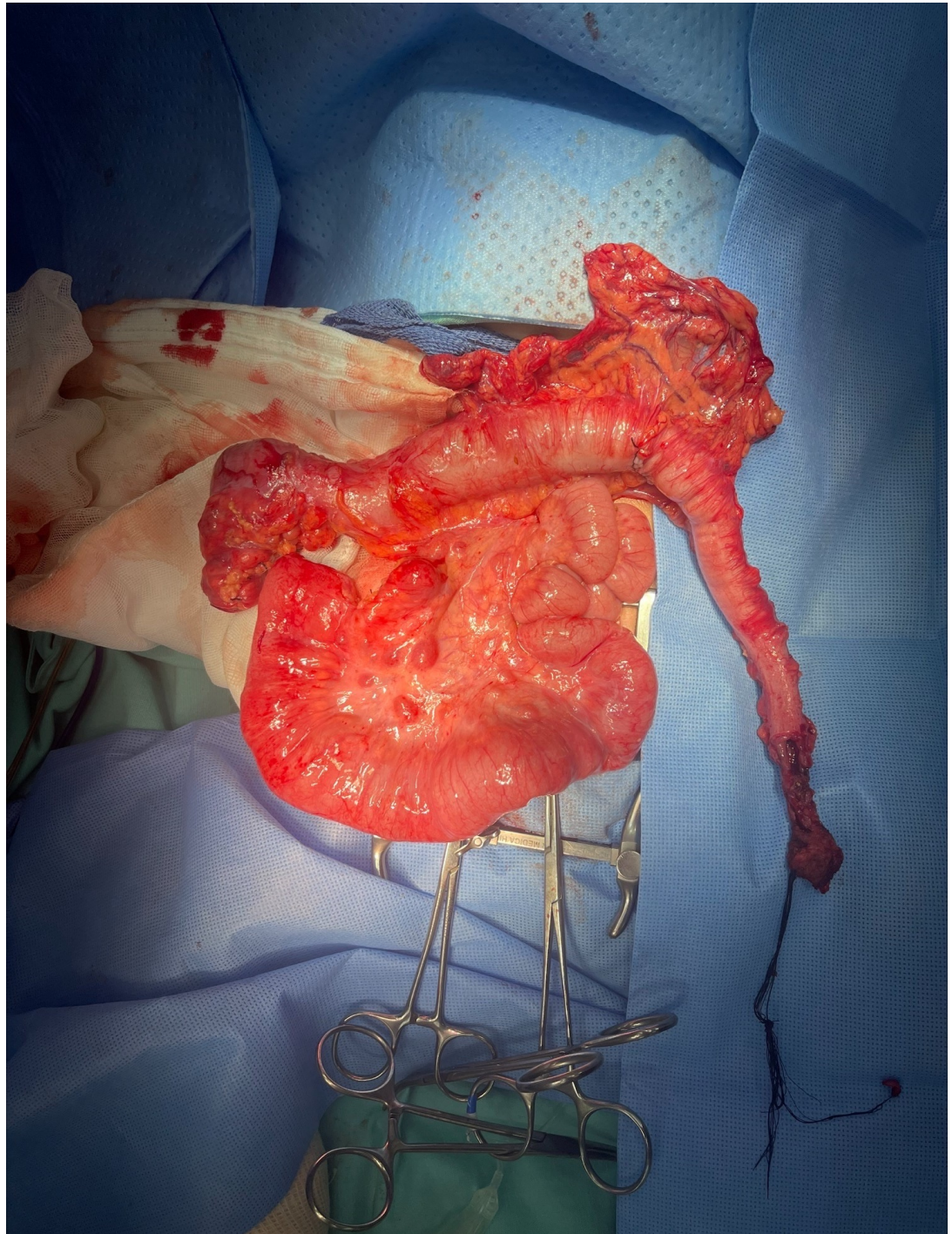
a) Ileostomy
b) Transverse colostomy
c) Transanal with colonic stenosis resection and pull-through of colon or ileum normoganglionic
d) Transanal with colonic stenosis resection and pull-through of colon or ileum normoganglionic with protective ileostomy
e) I don’t know

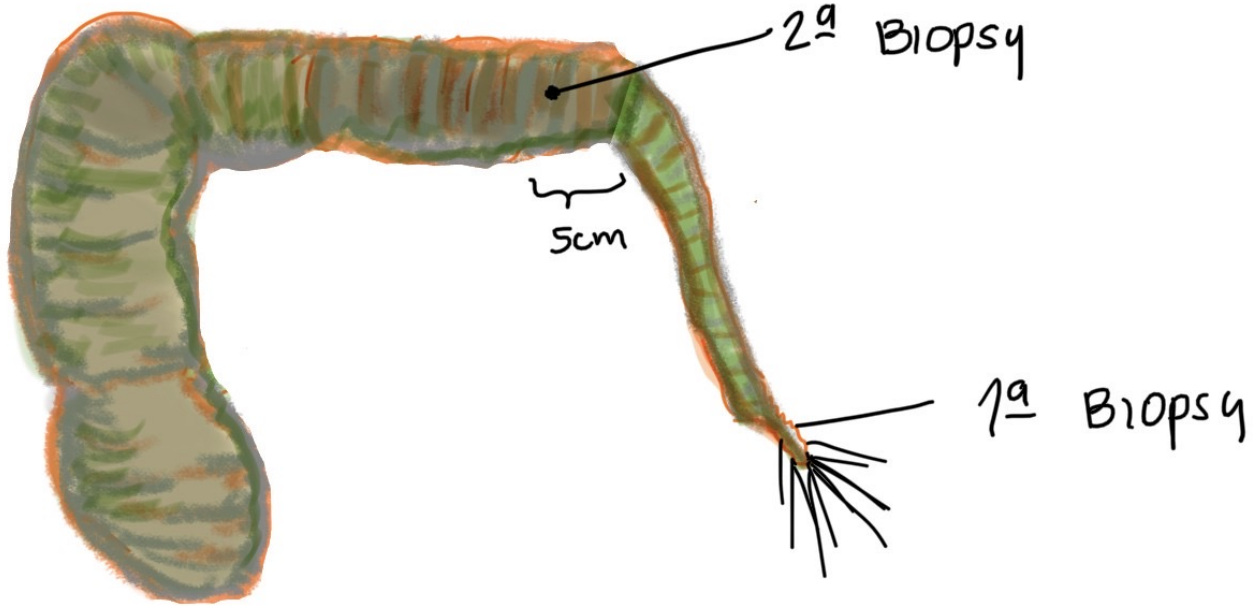


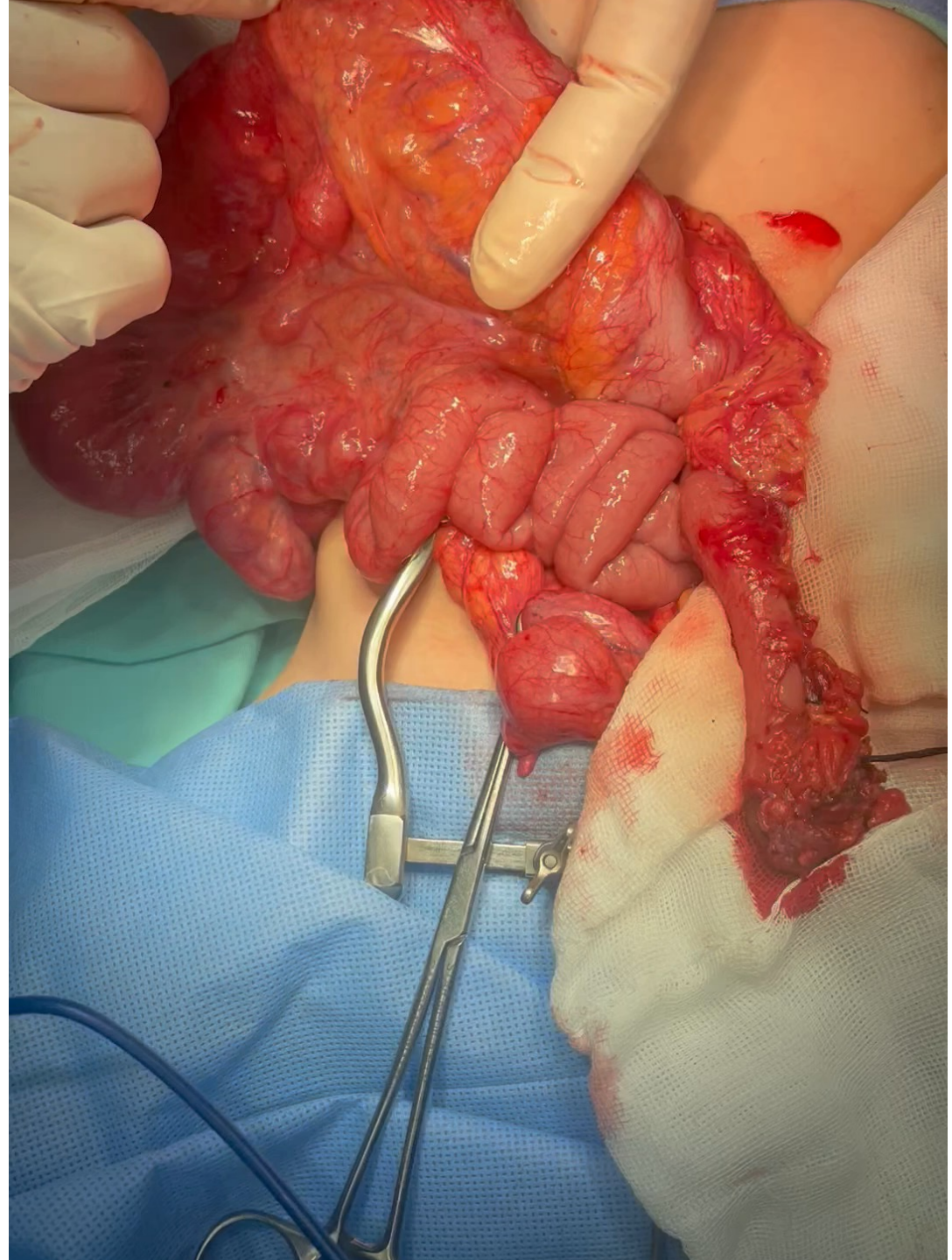




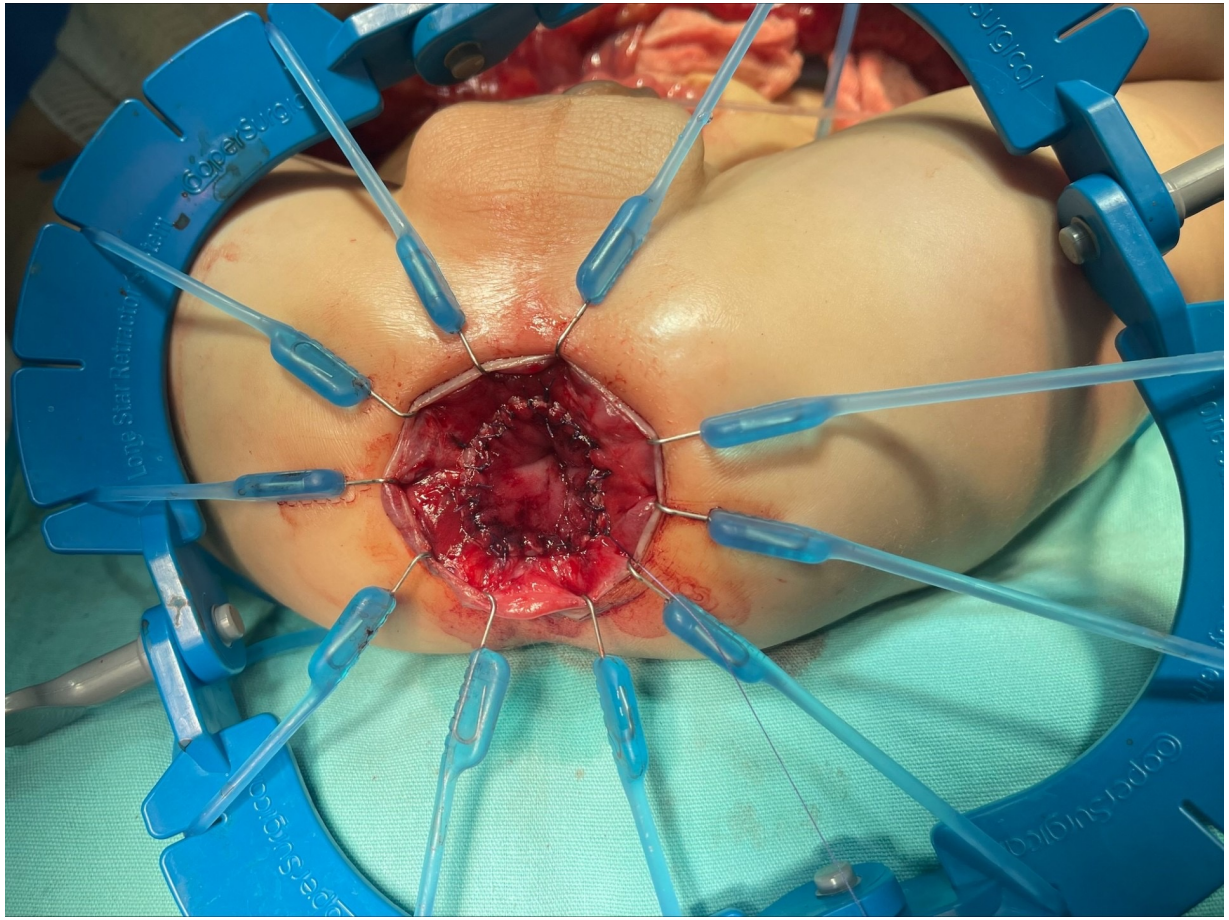

SuperSurgical




2ª Biopsy
5cm
1ª Biopsy



ileocolic artery ?
right colic artery ?

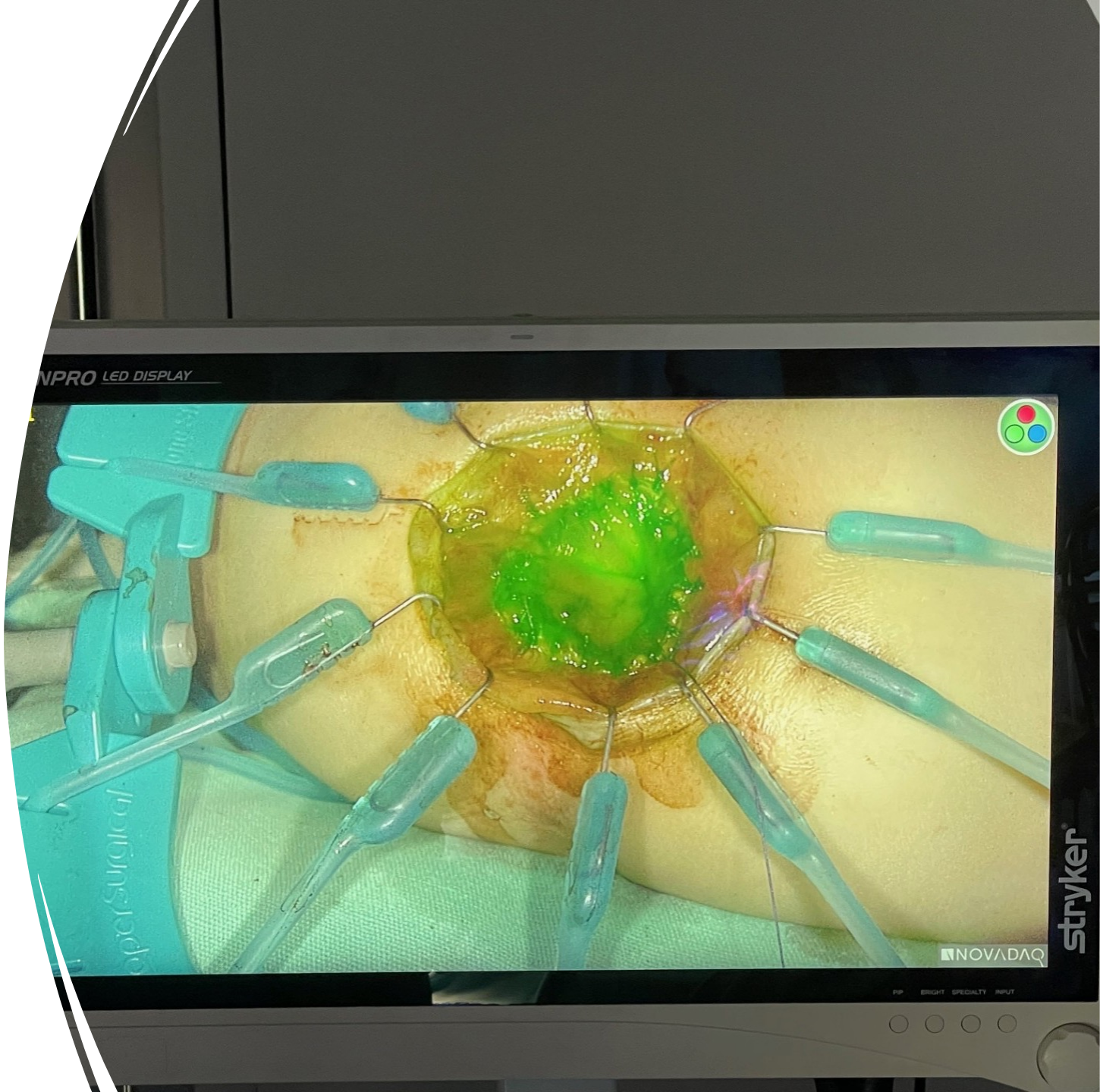


Discussion 1

Appendicitis (neonatal or infant)

Two major theories have been proposed to explain intra-abdominal neonatal appendicitis:

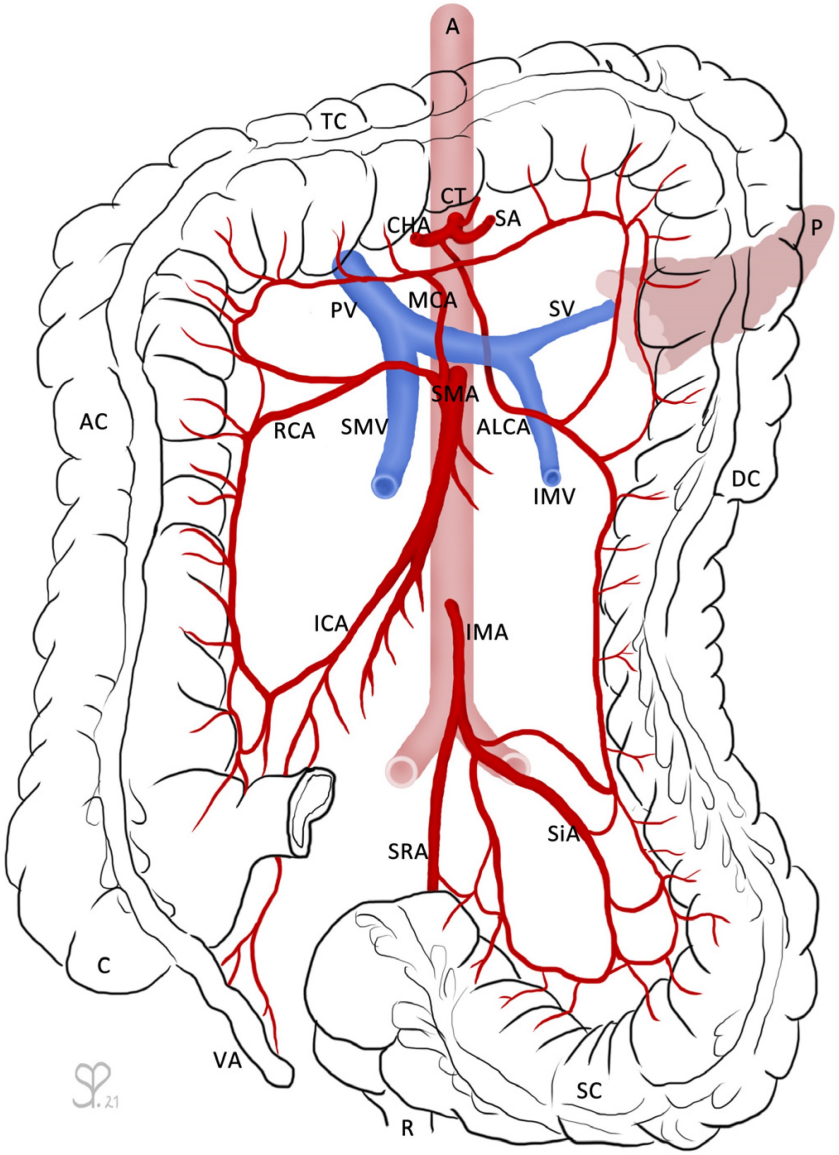
- neonatal appendicitis is a localized form of necrotizing enterocolitis
- obstructive caecal distension due to underlying Hirschsprung's disease,
- It is paramount to have a high index of suspicion and it should be included in the differential diagnosis of an infant presenting with an acute abdomen with vague symptoms and signs.



Discussion 2

- The right colic artery:
- The weighted mean incidence with which the right colic artery arose from other parent vessels was calculated at 36.8% for the superior mesenteric artery, 31.9% for the ileocolic artery, 27.7% for the root of the middle colic artery and 2.5% for the right branch of the middle colic artery.

- The lack of the iatrogenic right colic artery or as an anatomical variant may have an implication in the loss of the right colon.



Discussion 3

THANK YOU
KARLA SANTOS JASSO MD
santosjasso@hotmail.com

Pediatric Surgery International (2022) 38:25–30
https://doi.org/10.1007/s00383-021-05023-9

ORIGINAL ARTICLE

Is the appendix a good organ to diagnose total colonic aganglionosis?

Marina L. Reppucci[1] · Michael A. Arnold[2,3] · Mark Lovell[2,3] · Karla Santos-Jasso[4] · Jill Ketzer[1] · Alberto Pena[1] · Luis de la Torre[1] · Andrea Bischoff[1]

Accepted: 1 September 2021 / Published online: 23 September 2021

Abstract

Purpose The use of the appendix for diagnosis of Total Colonic Aganglionosis (TCA) remains controversial. This study aimed to categorize the presence of ganglion cells in the appendix and determine its reliability as a histological specimen for the diagnosis of TCA.

Methods This was a combined retrospective and prospective study. Permanent histological specimens of normal appendices removed during appendectomy for malrotation or falsely presumed appendicitis, and from patients with short segment Hirschsprung's disease (HD) or TCA were included. Permanent specimens of the appendix tip from Malone procedures were prospectively collected. All specimens were independently reviewed by two experienced pathologists in a standardized manner to assess for the presence of ganglion cells.

Results A total of 112 appendices were evaluated. Nine came from patients with a diagnosis of TCA, and five from patients with HD. Ganglion cells were present in all specimens from patients with diagnoses other than TCA and were absent in all specimens from patients with TCA.

Conclusion In the correct clinical setting, the absence of ganglion cells in the appendix can allow for a reliable diagnosis of TCA.

Keywords Total Colonic Anganglionosis · Hirschsprung's disease · Appendix