Agenda May 3

- 11:45 -12pm Lunch
- 12pm- 1pm Speaker Dr. Frederick Rescorla
- 1pm -1:15pm Questions

Hirschsprung's Disease

Fred Rescorla, MD

Section of Pediatric Surgery

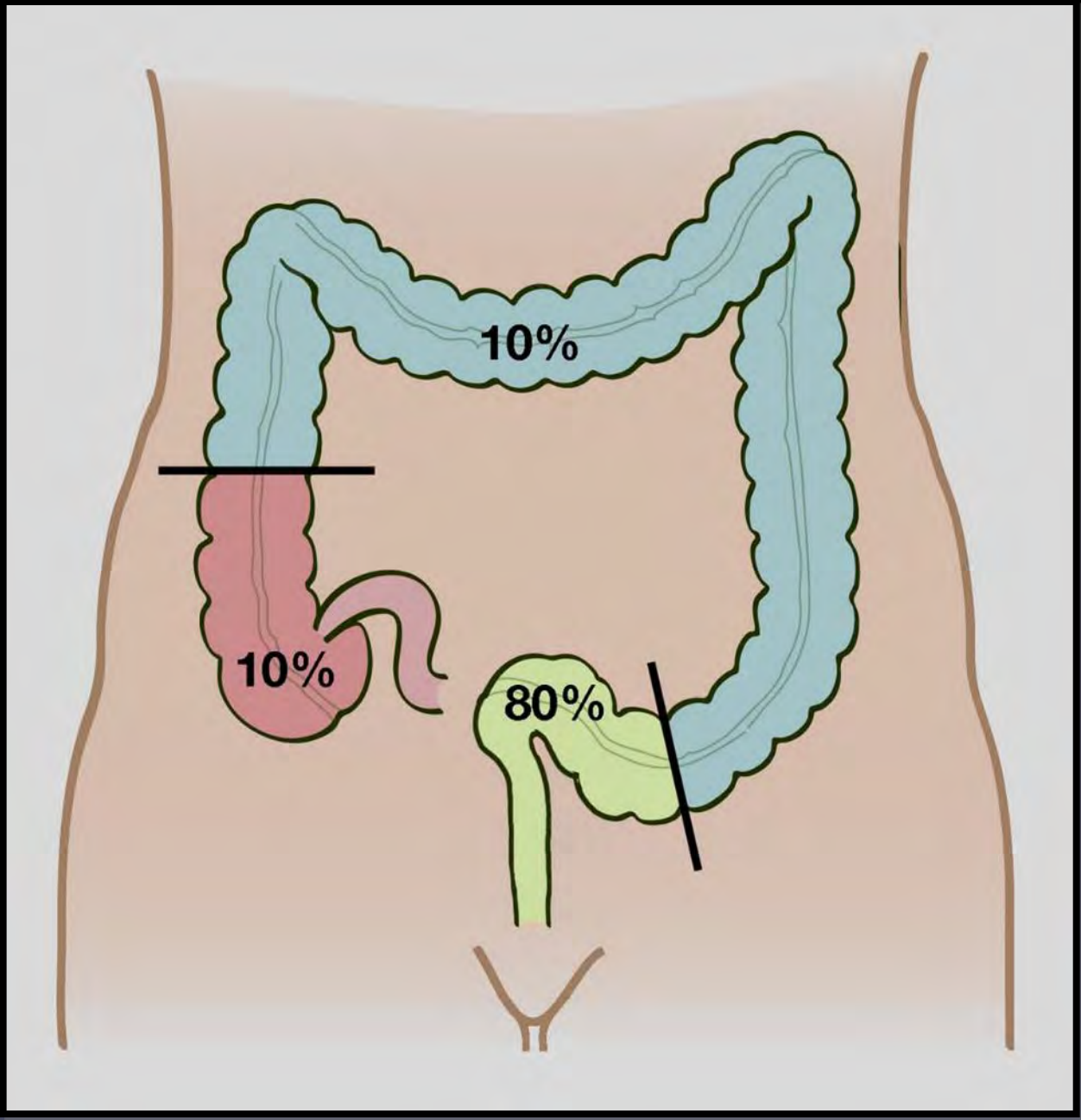
Indiana University School of Medicine

Indiana University Health

Riley Hospital for Children
Indiana University Health

Hirschsprung's Disease

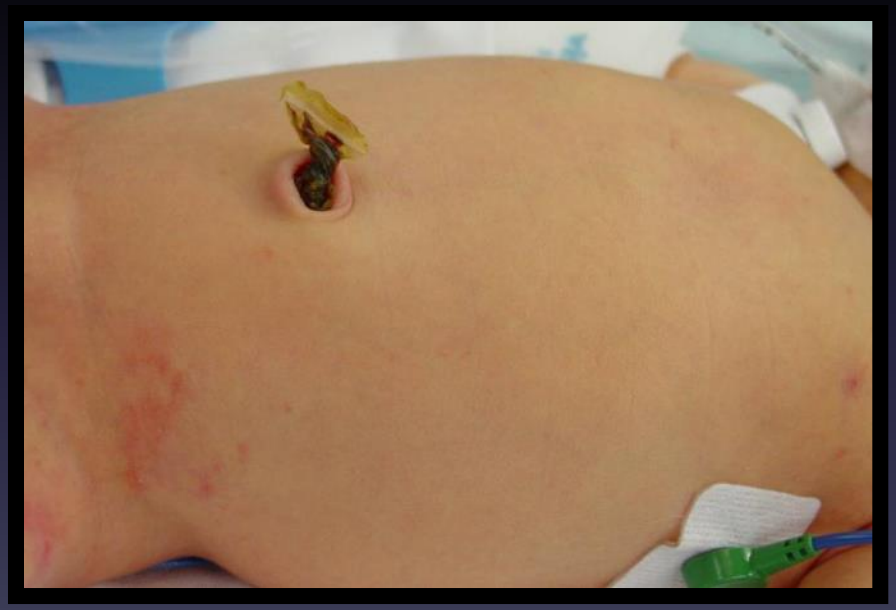
- Absence of ganglion cells – myenteric and submucosal plexuses of distal bowel
- Results in functional intestinal obstruction at level of aganglionosis


10%
10%
80%

Hirschsprung's - Etiology

- Ganglion cells from neural crest – migrate from proximal to distal by 13wks of fetal life
- Two theories: 1. failure of migration; or 2. neural cells migrate but fail to survive or proliferate

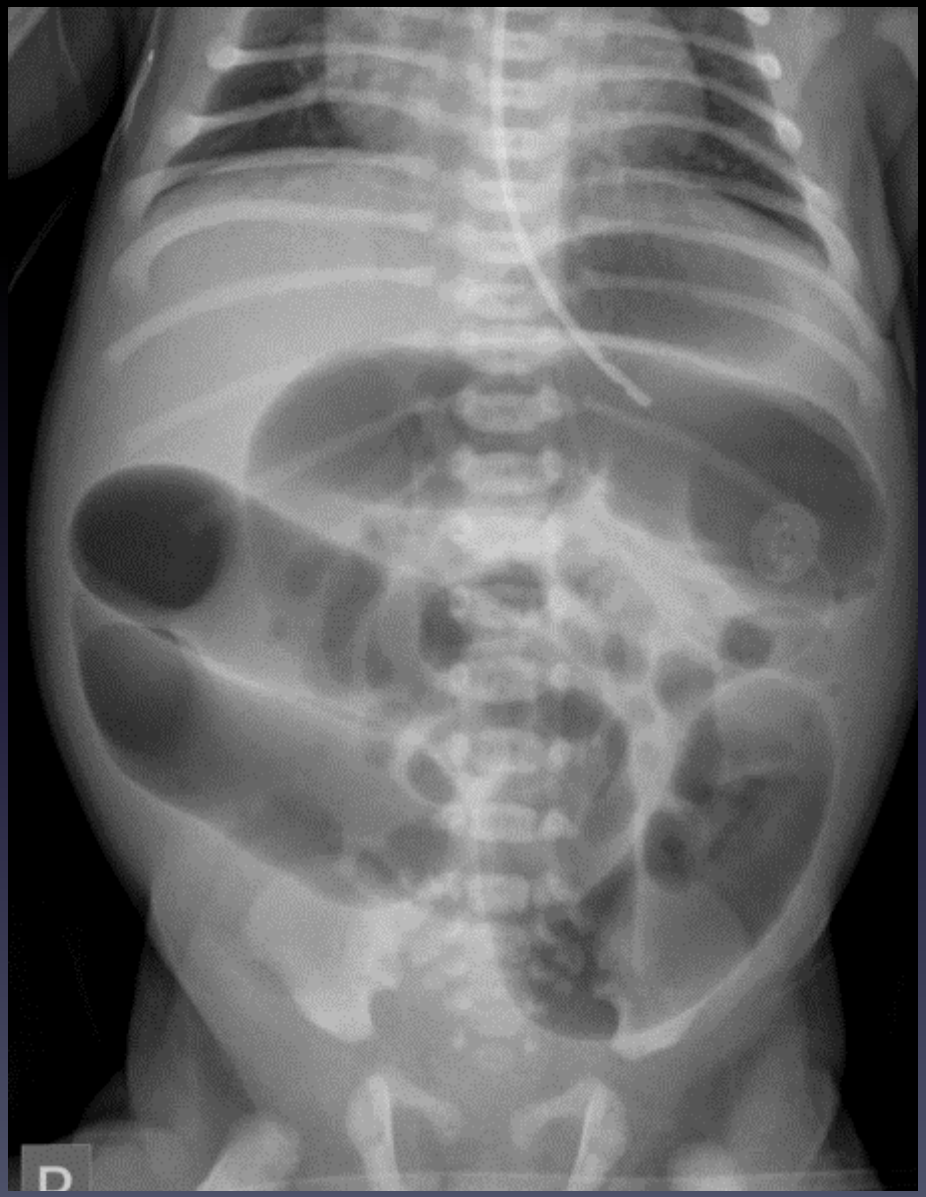
Hirschsprung's - Presentation



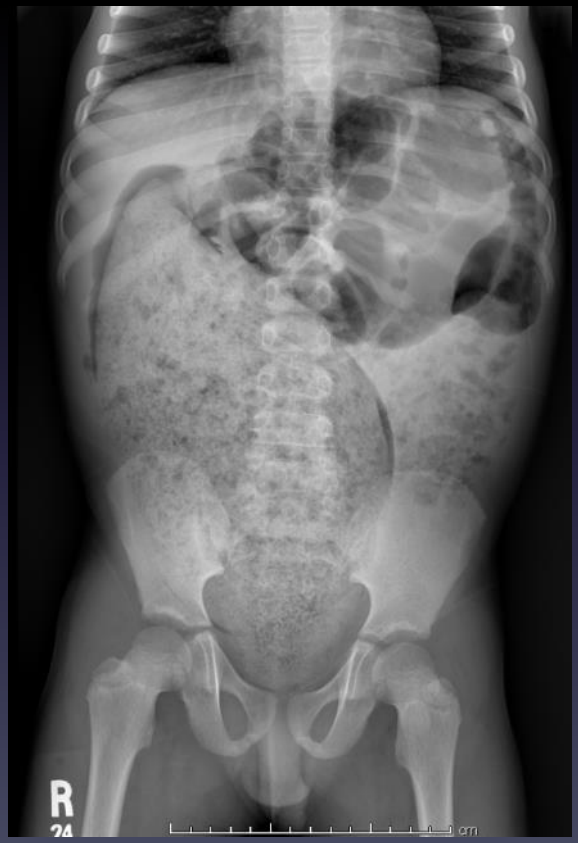
Newborns

- 50-90% as newborns: abdominal distention; feeding intolerance; bilious emesis
- 90%, delayed passage of meconium

2 day old boy, Down syndrome, no passage of meconium



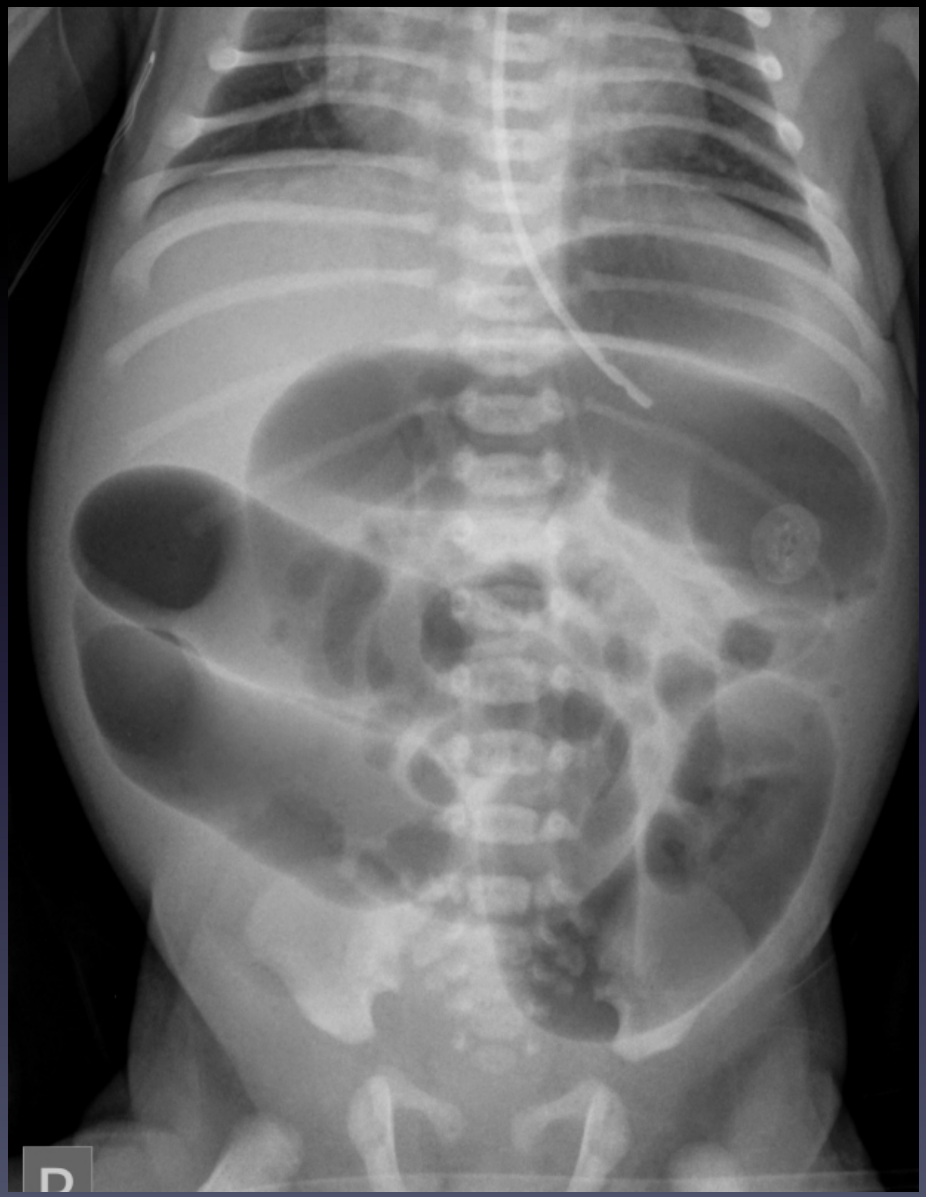
Hirschsprung's - Presentation



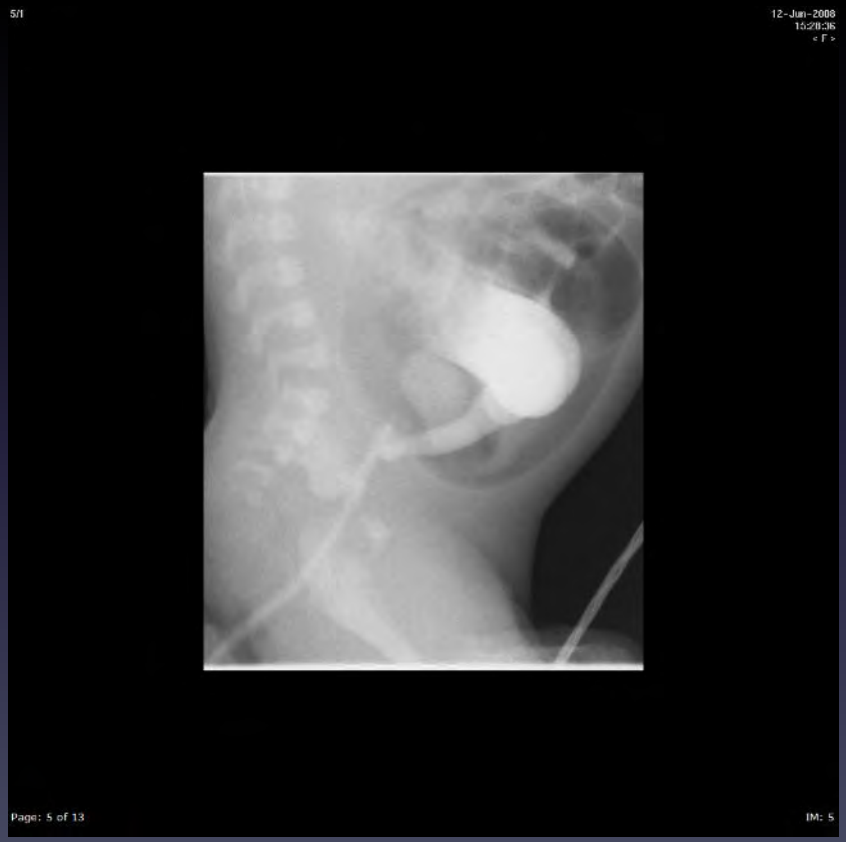
Older child

- Constipation
- Hx of failure to pass meconium in first 48hrs; failure to thrive; abdominal distention; dependence on enemas without encopresis

2 day old boy, Down syndrome, no passage of meconium



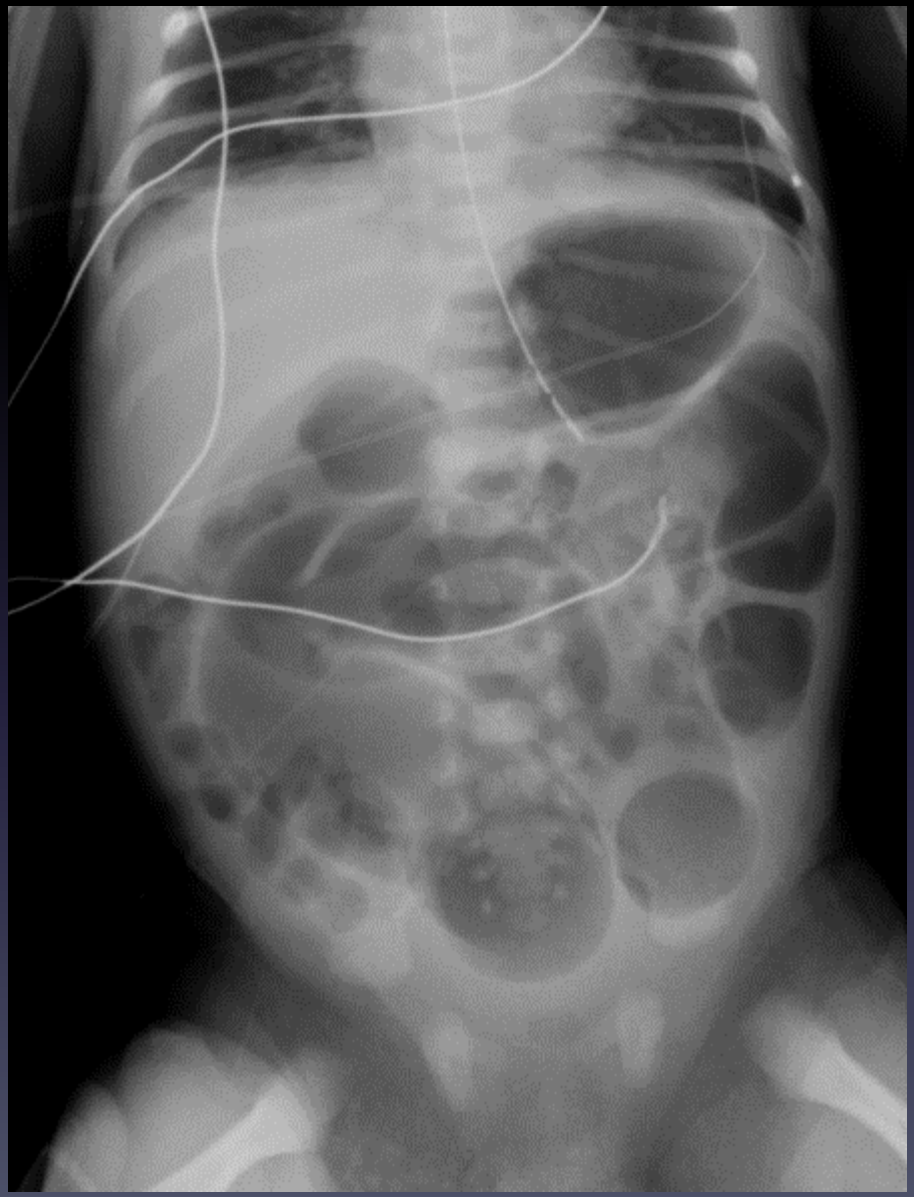
Contrast enema

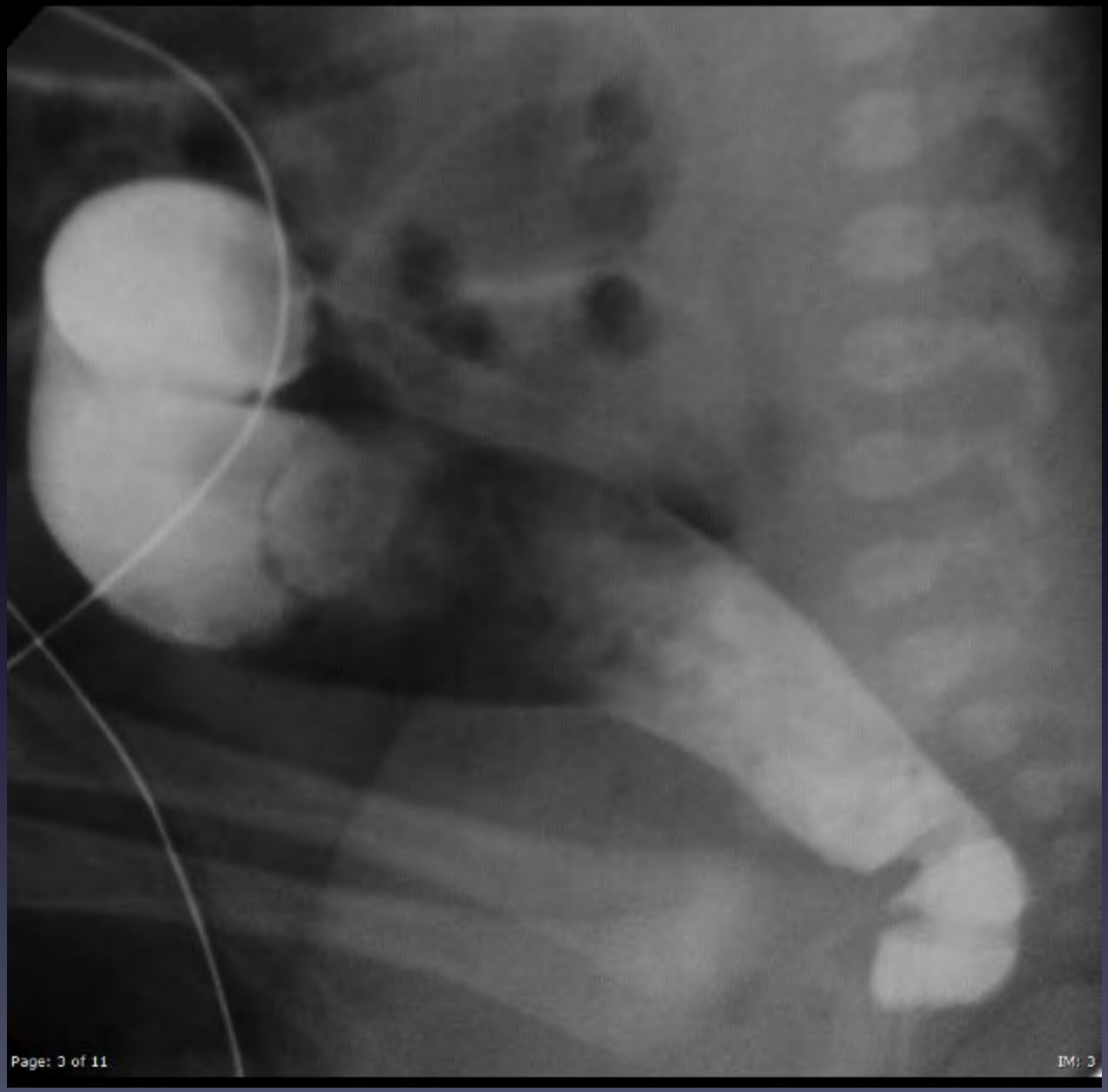


Transition zone

- Unprepped
- Lateral view
- Abnormal Rectal:colon ratio(<1)

1 day old boy

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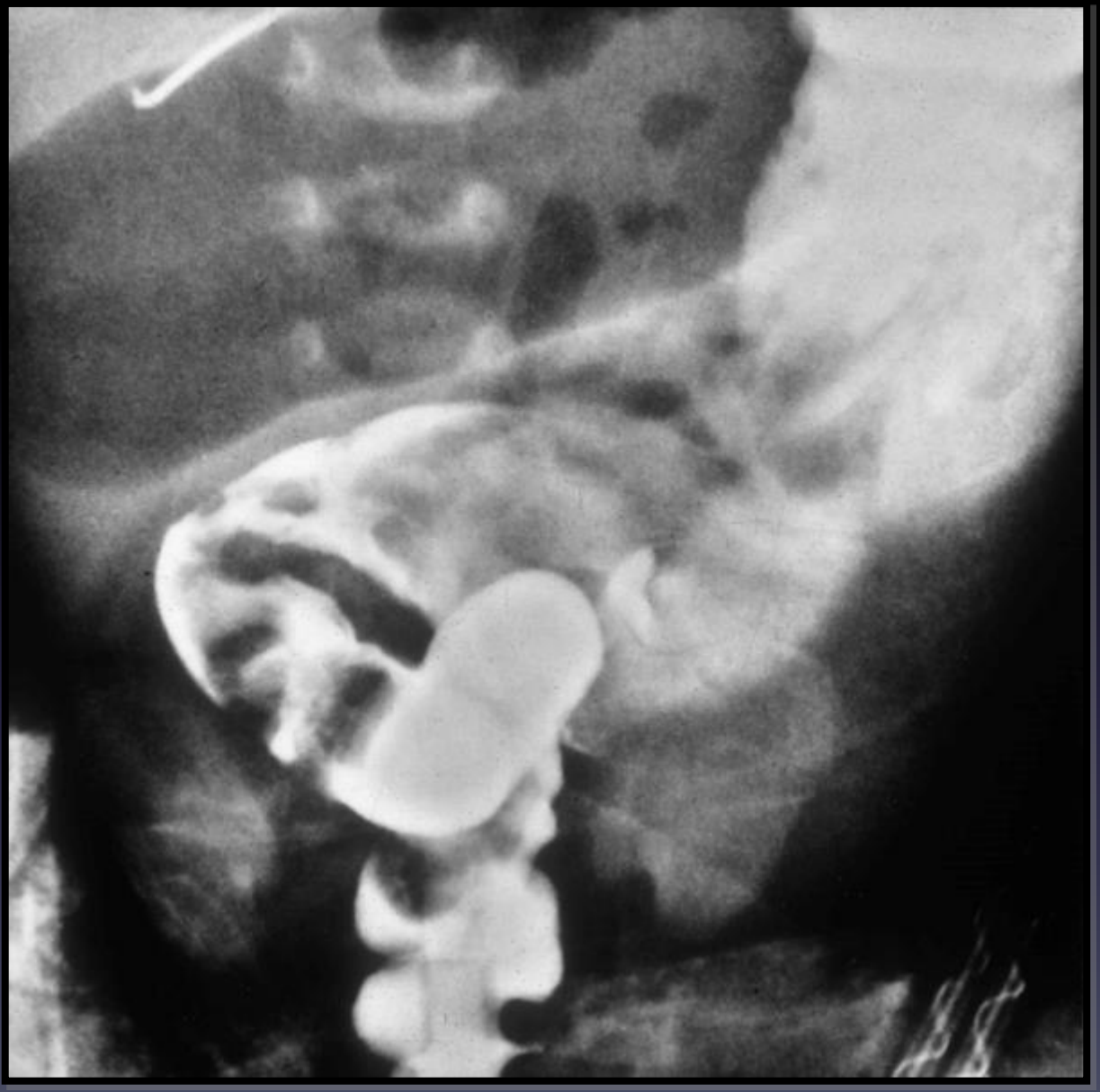
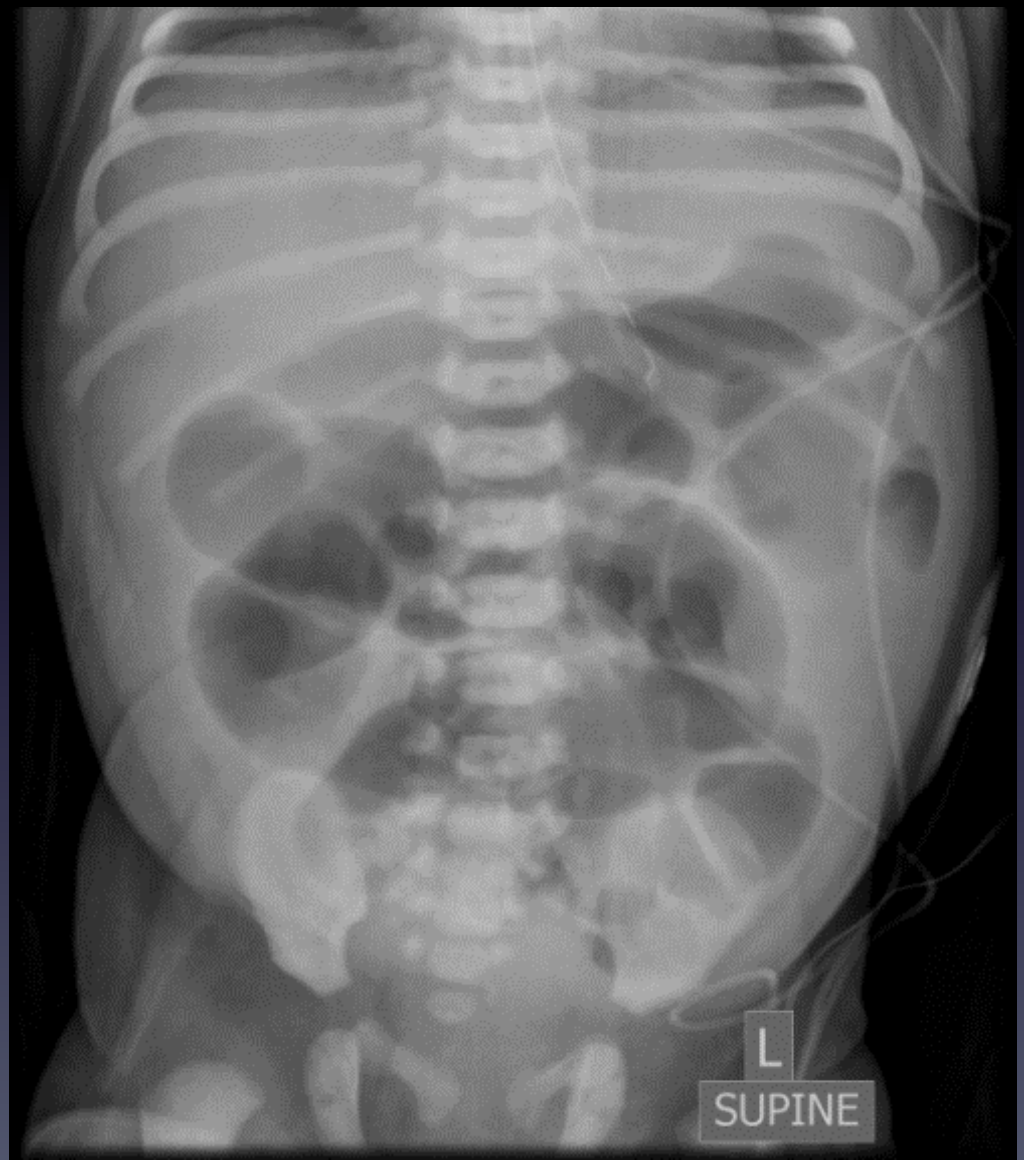
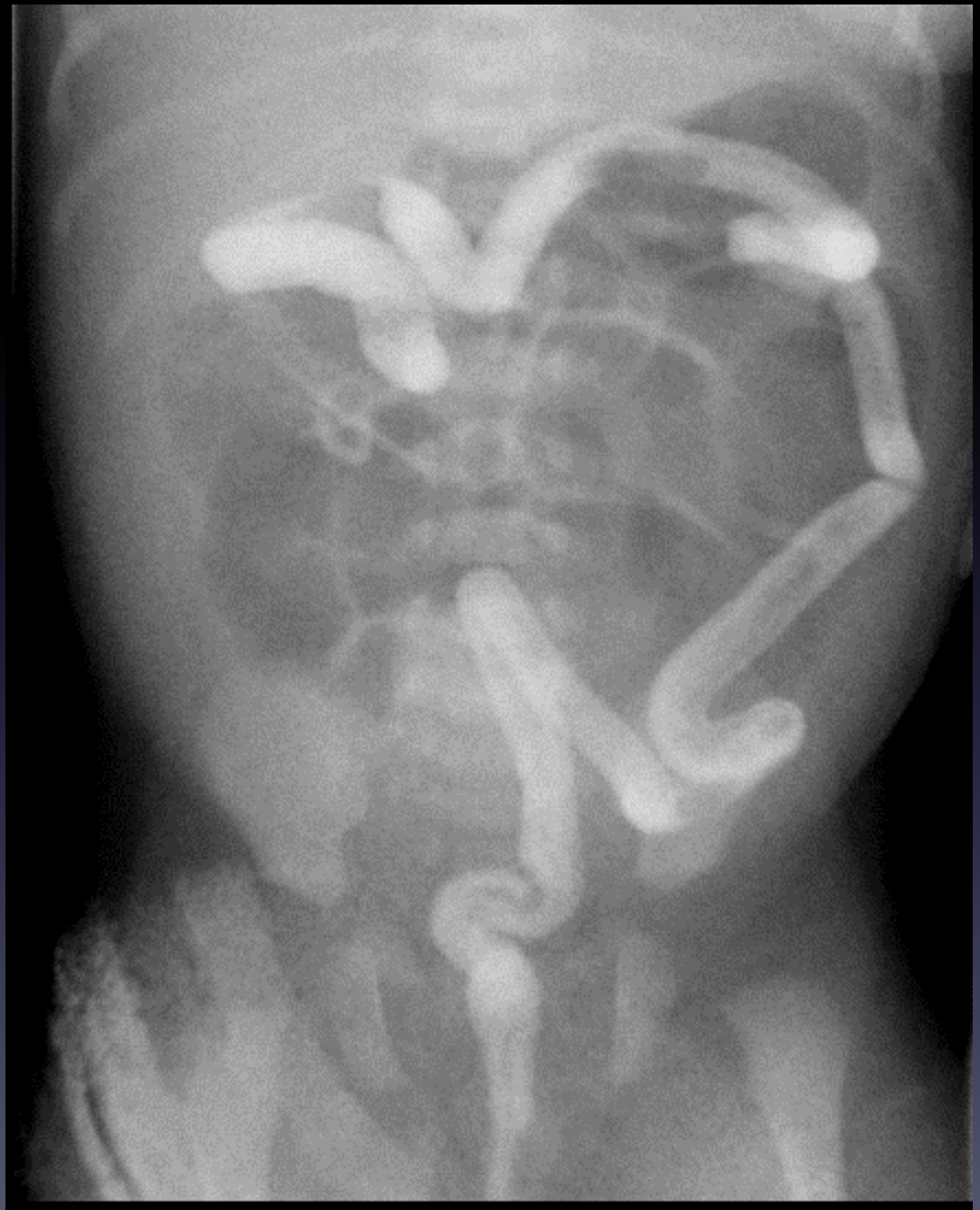
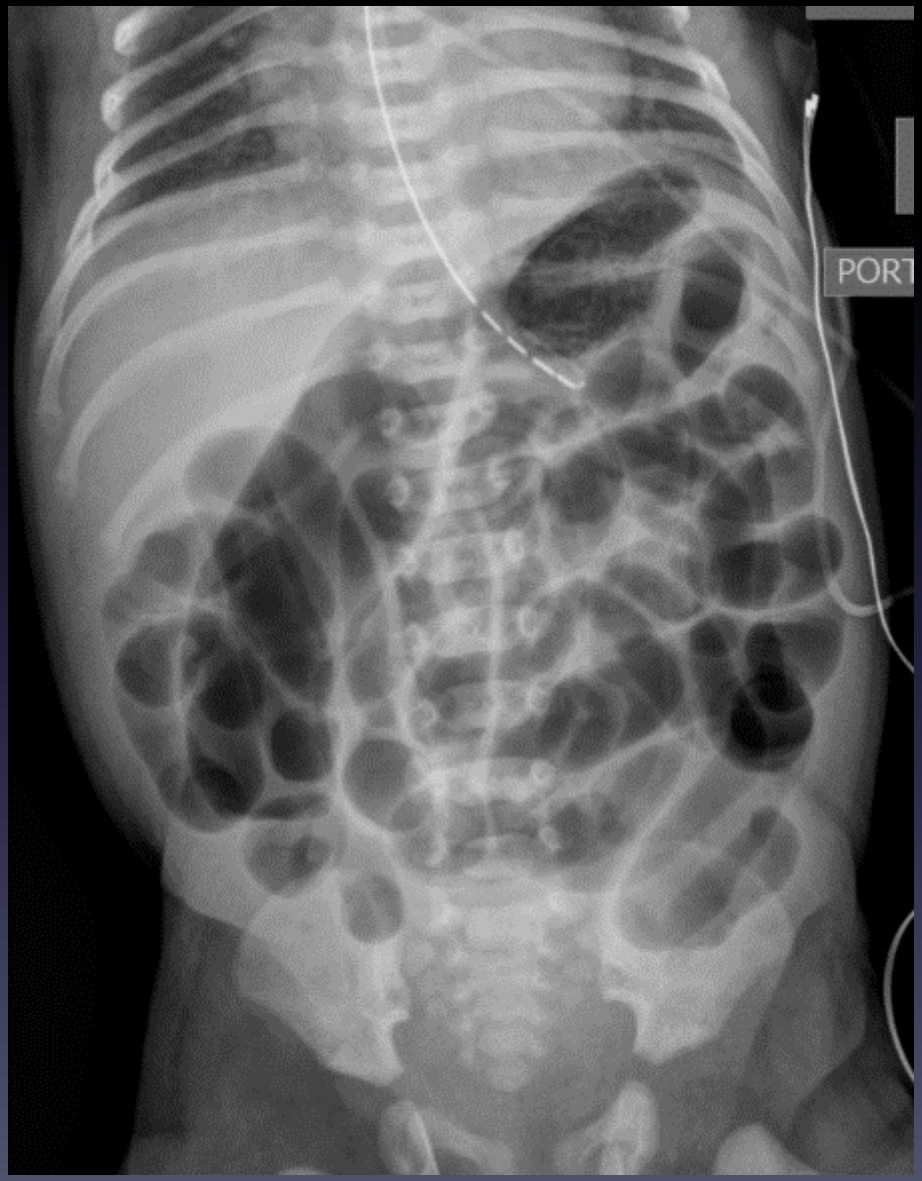


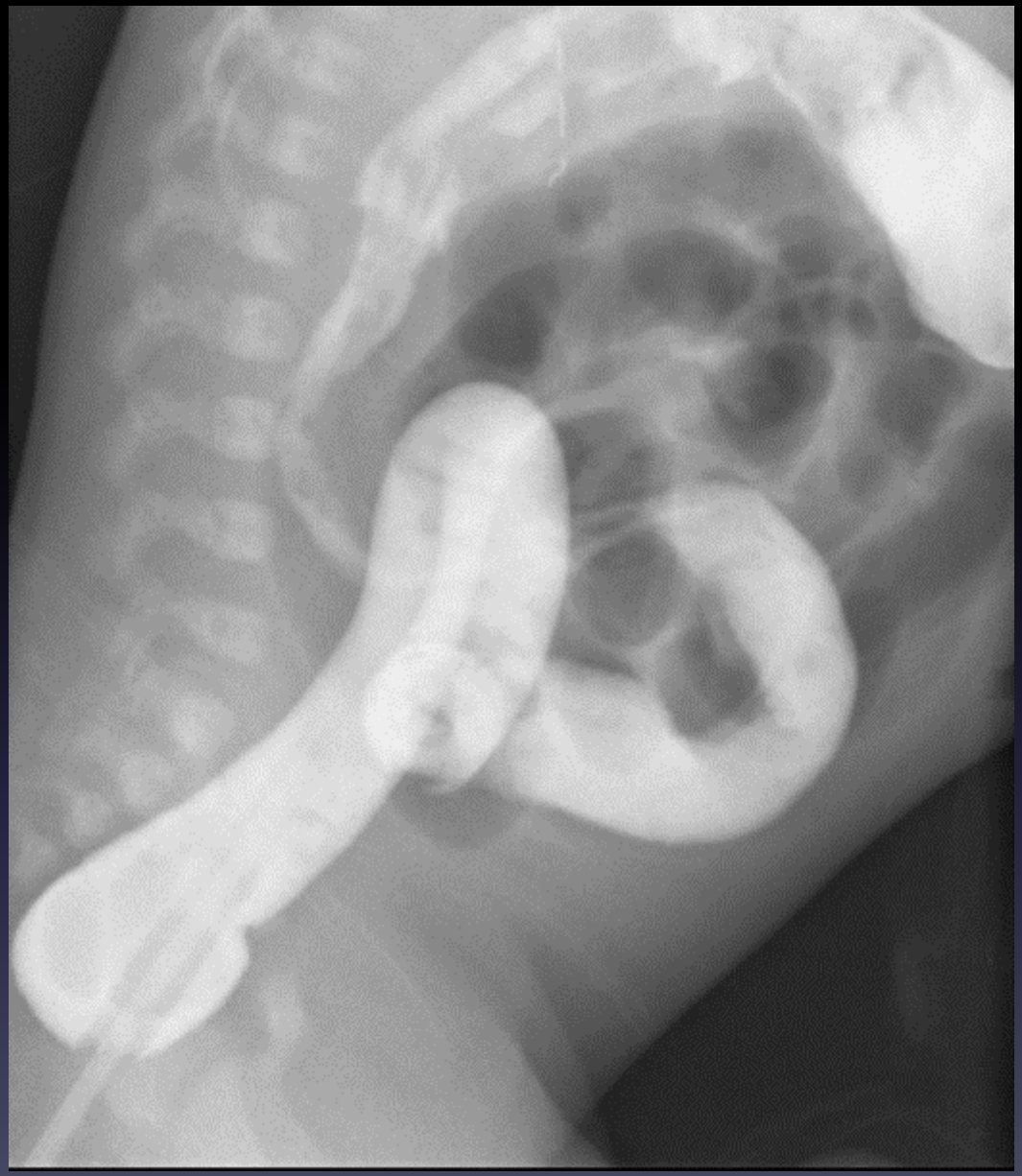
2 day old girl, total colonic Hirschprung's disease



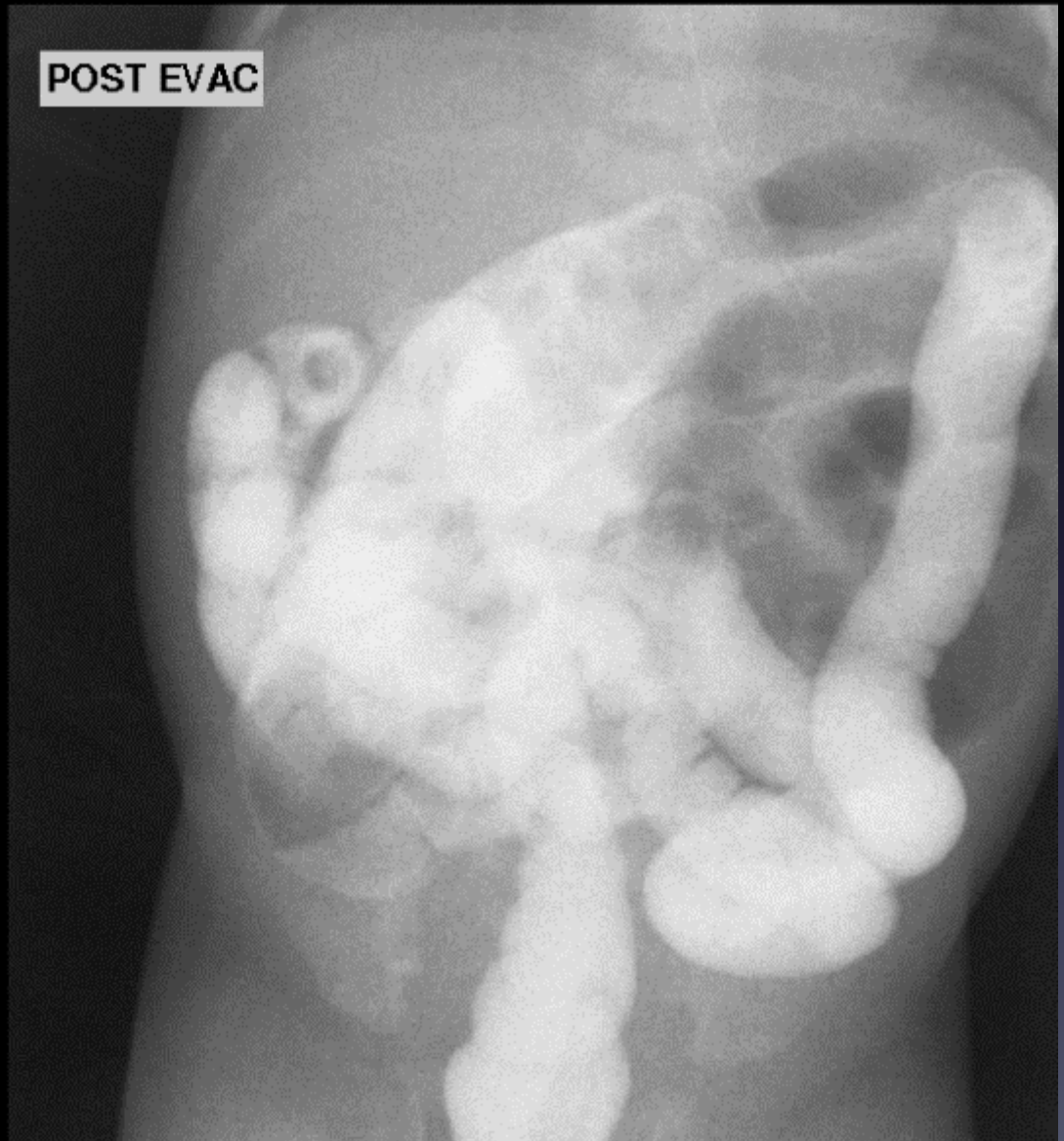


1 day old boy with total colonic Hirschsprung disease extending approximately to 10 cm of the terminal ilium.

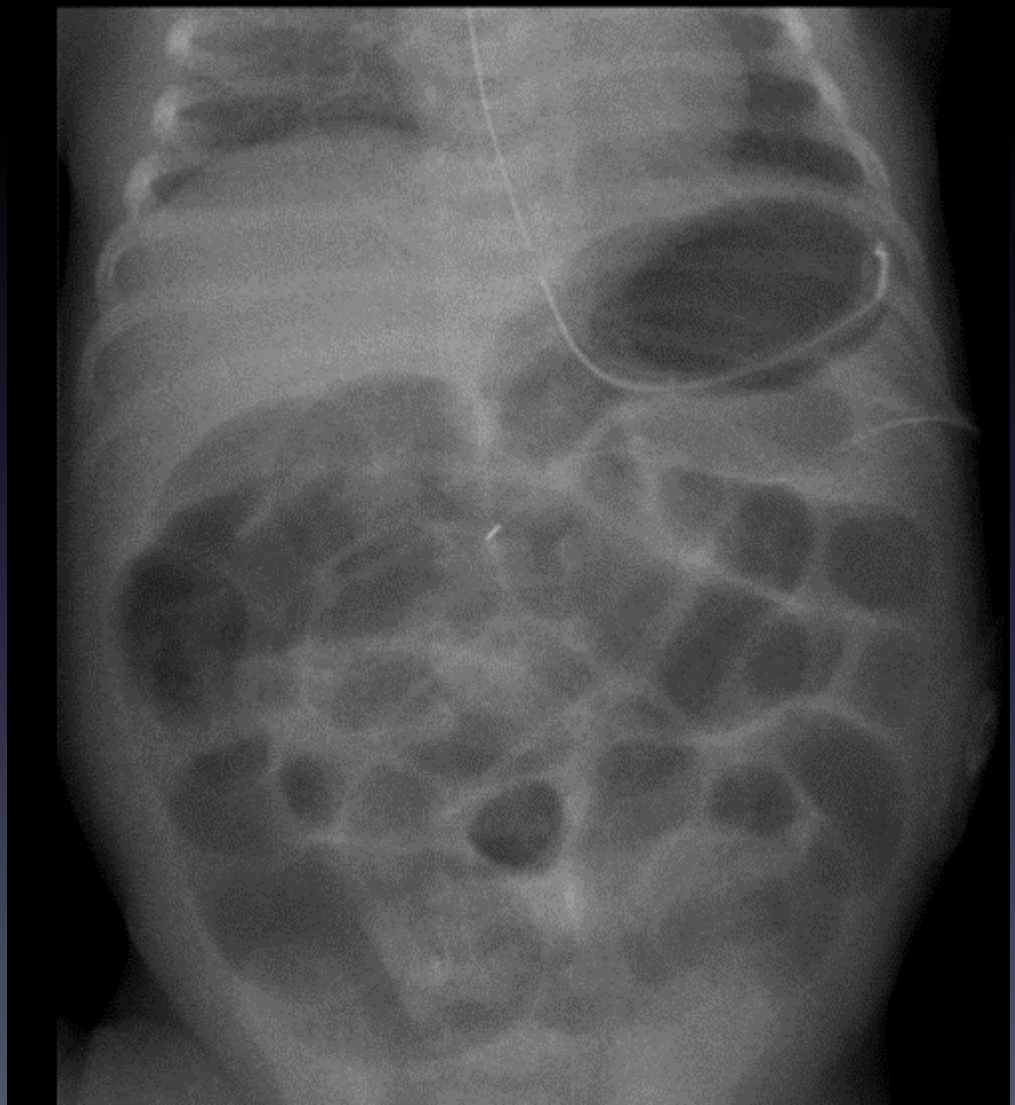


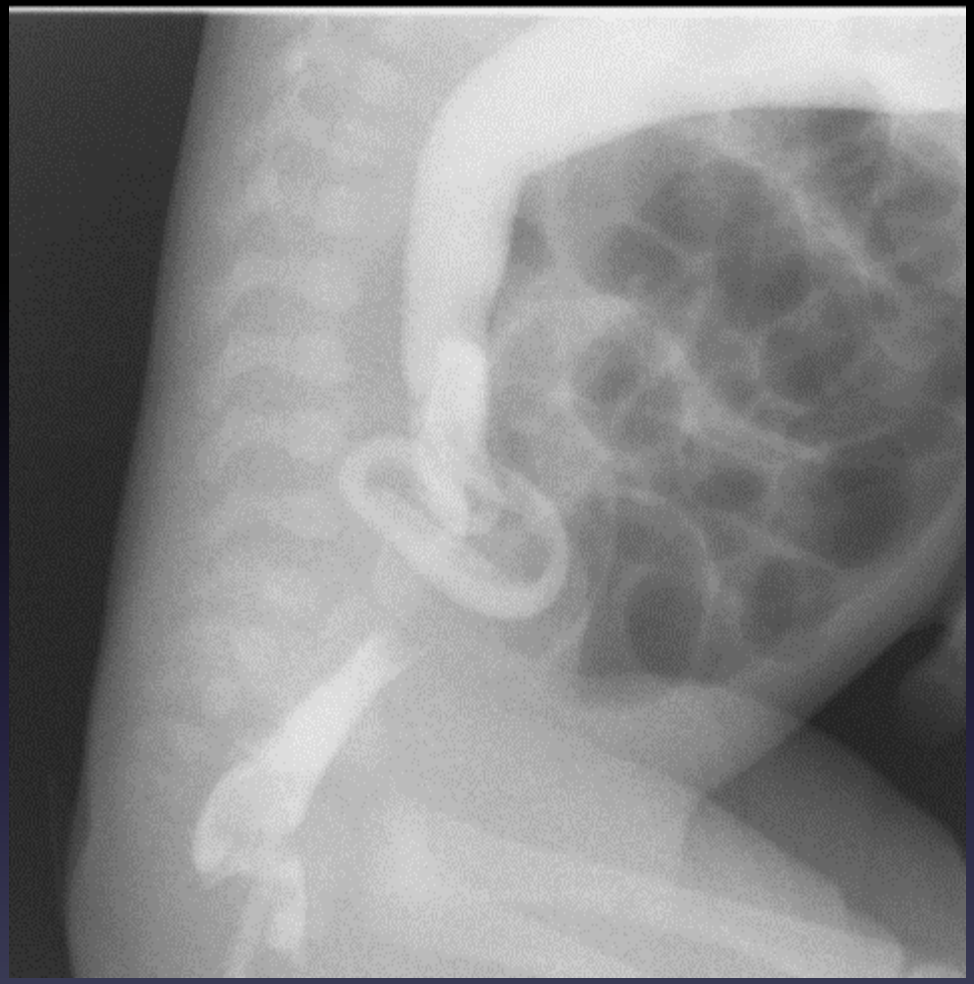


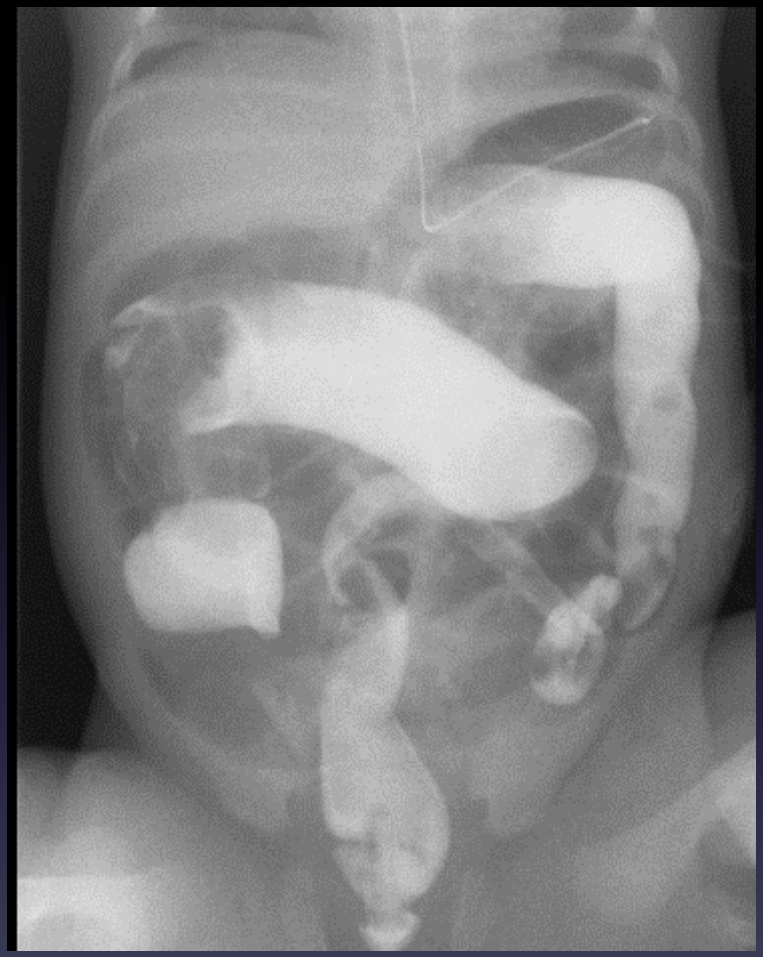
24hr film sometimes useful



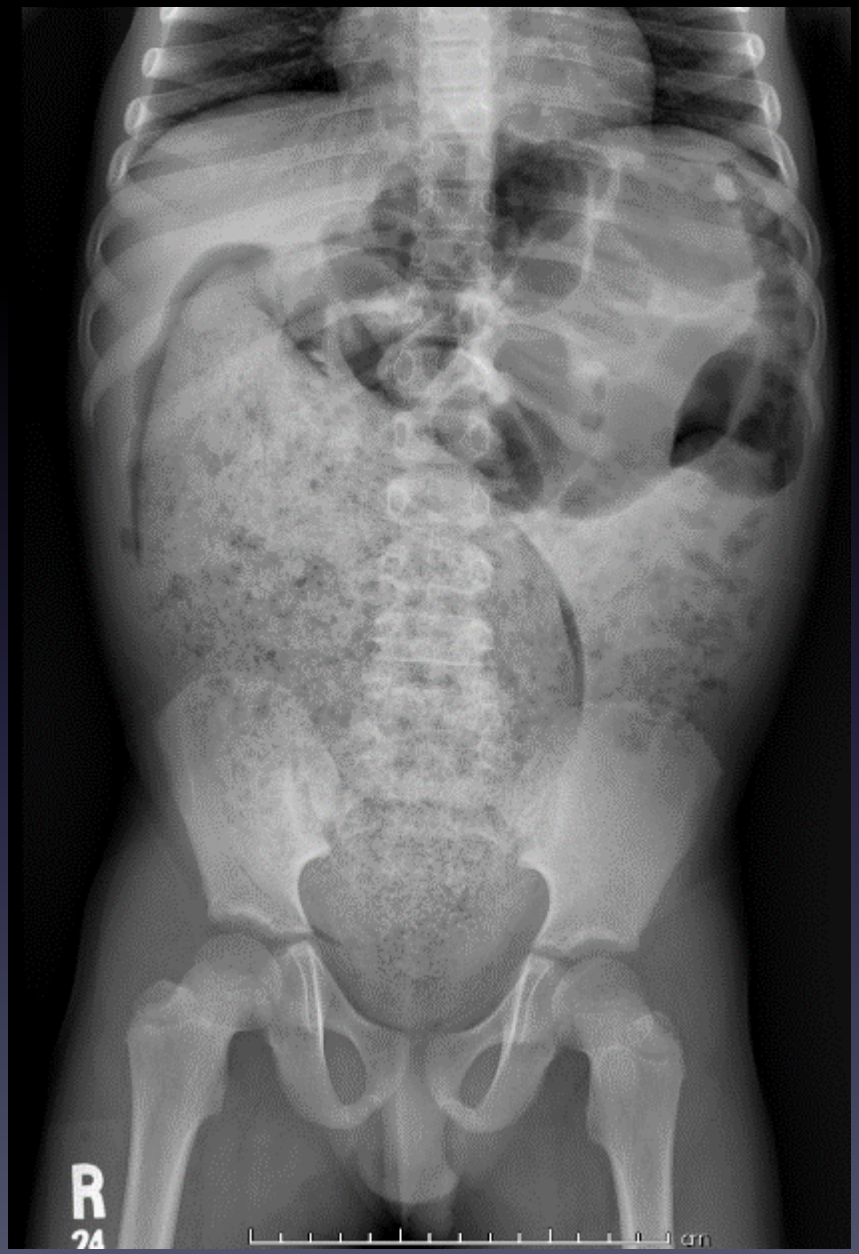
1 day old girl with small left colon syndrome, negative biopsy



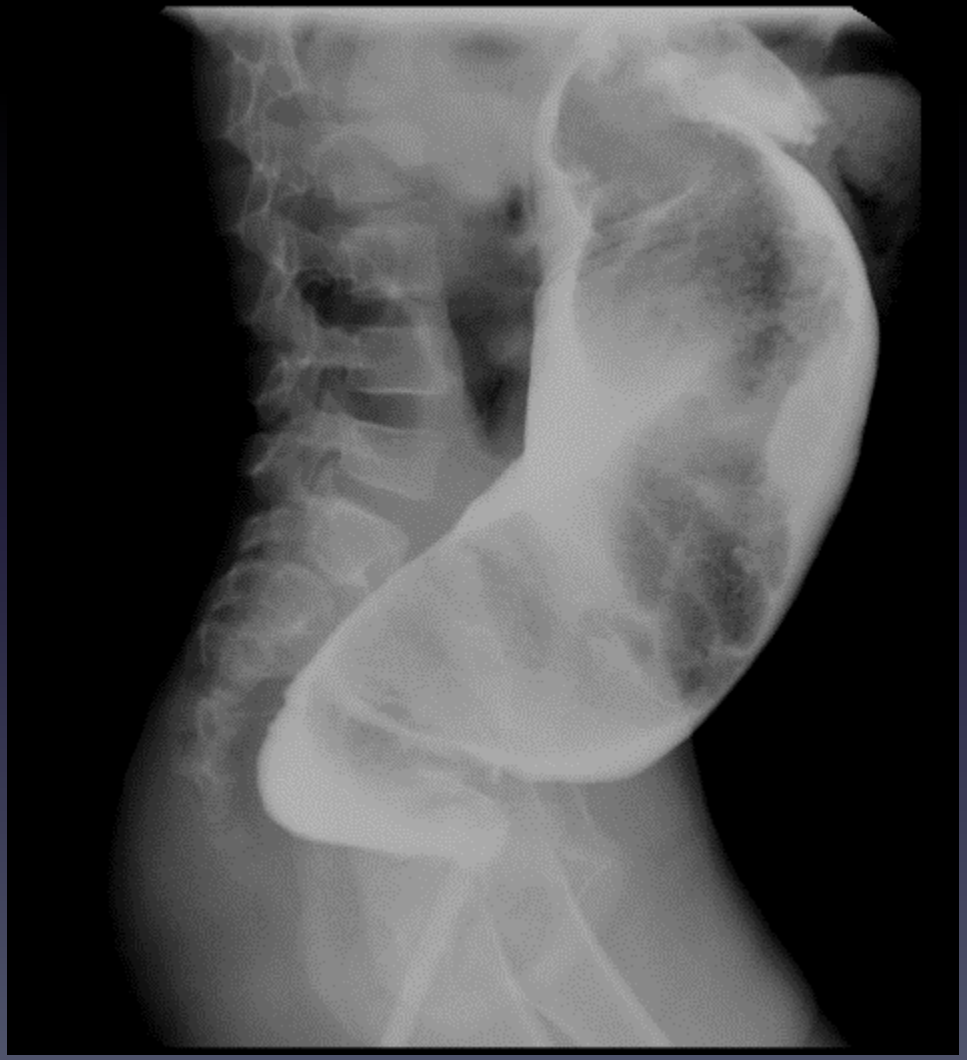




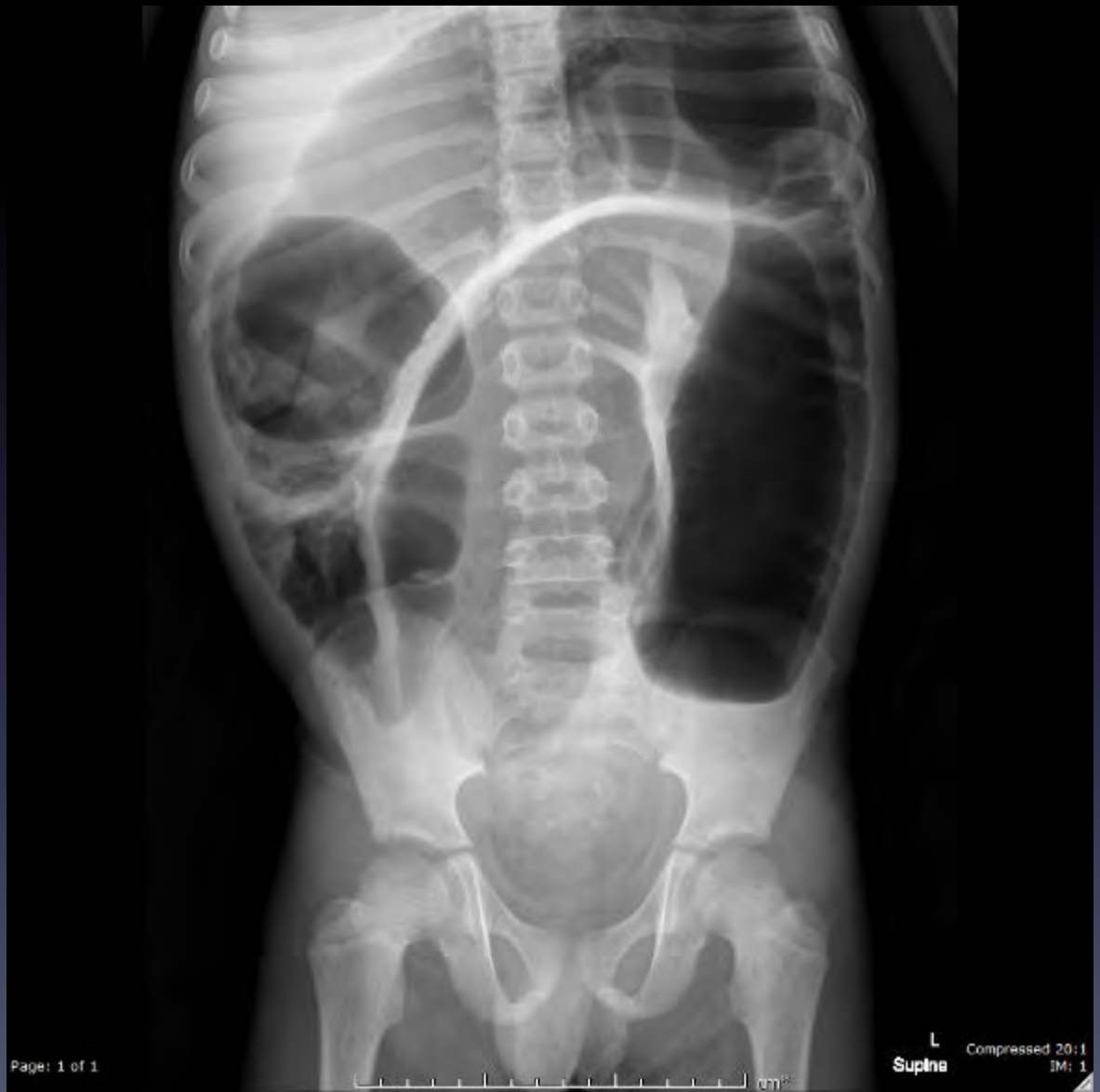
6 year old boy with constipation



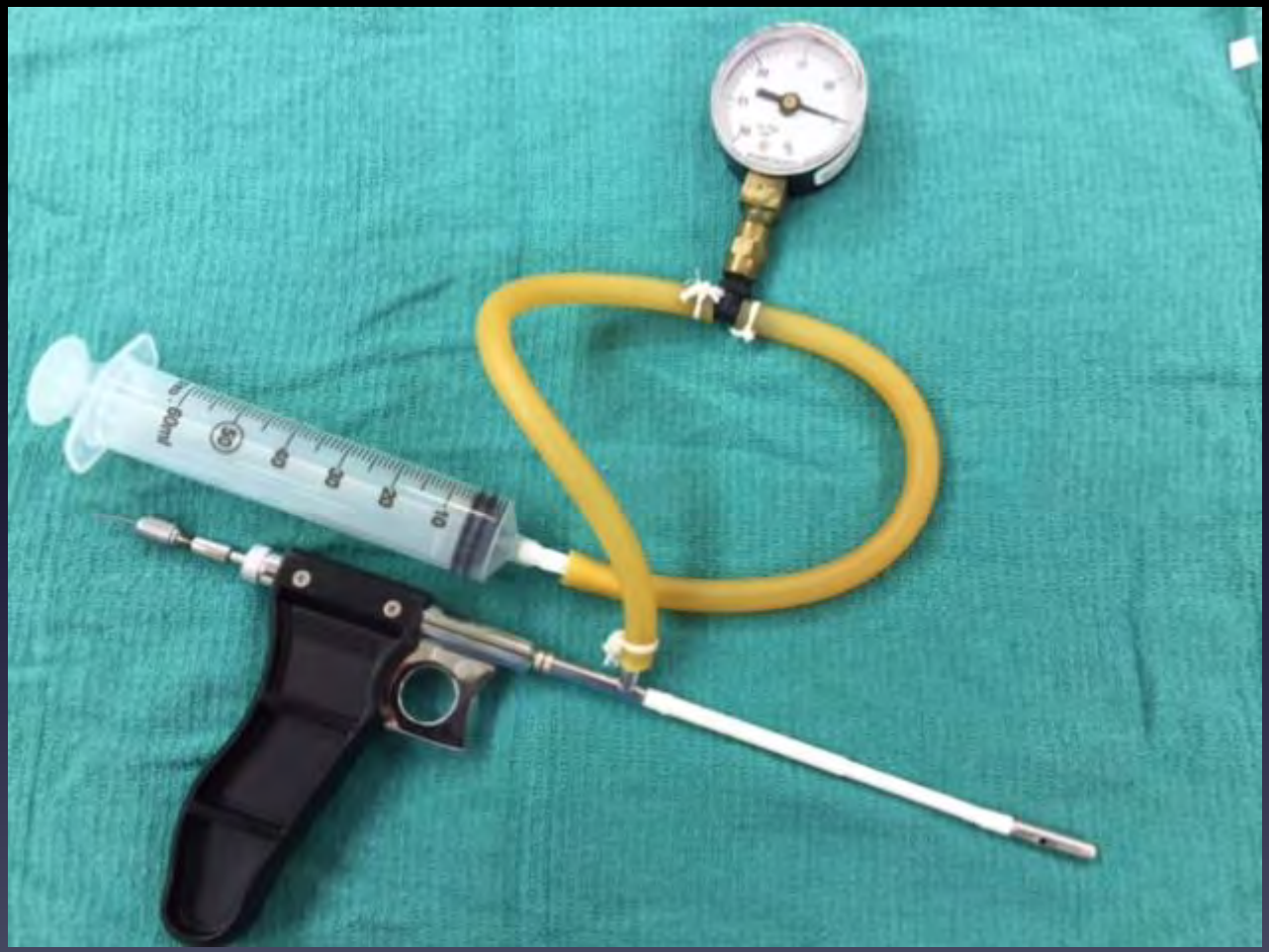
Few punch biopsies; some demonstrated ganglion cells and some agangliosis

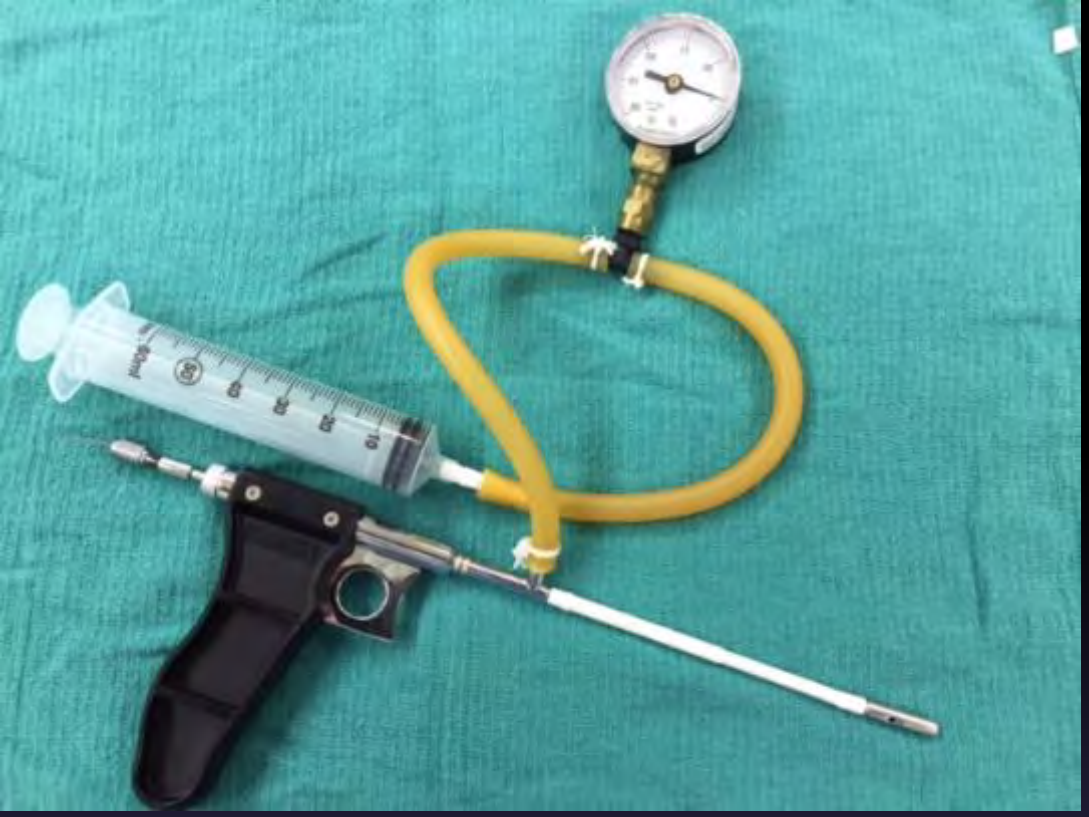


At the age of 7 year- colitis
Open biopsy- Low segment Hirschprung's disease

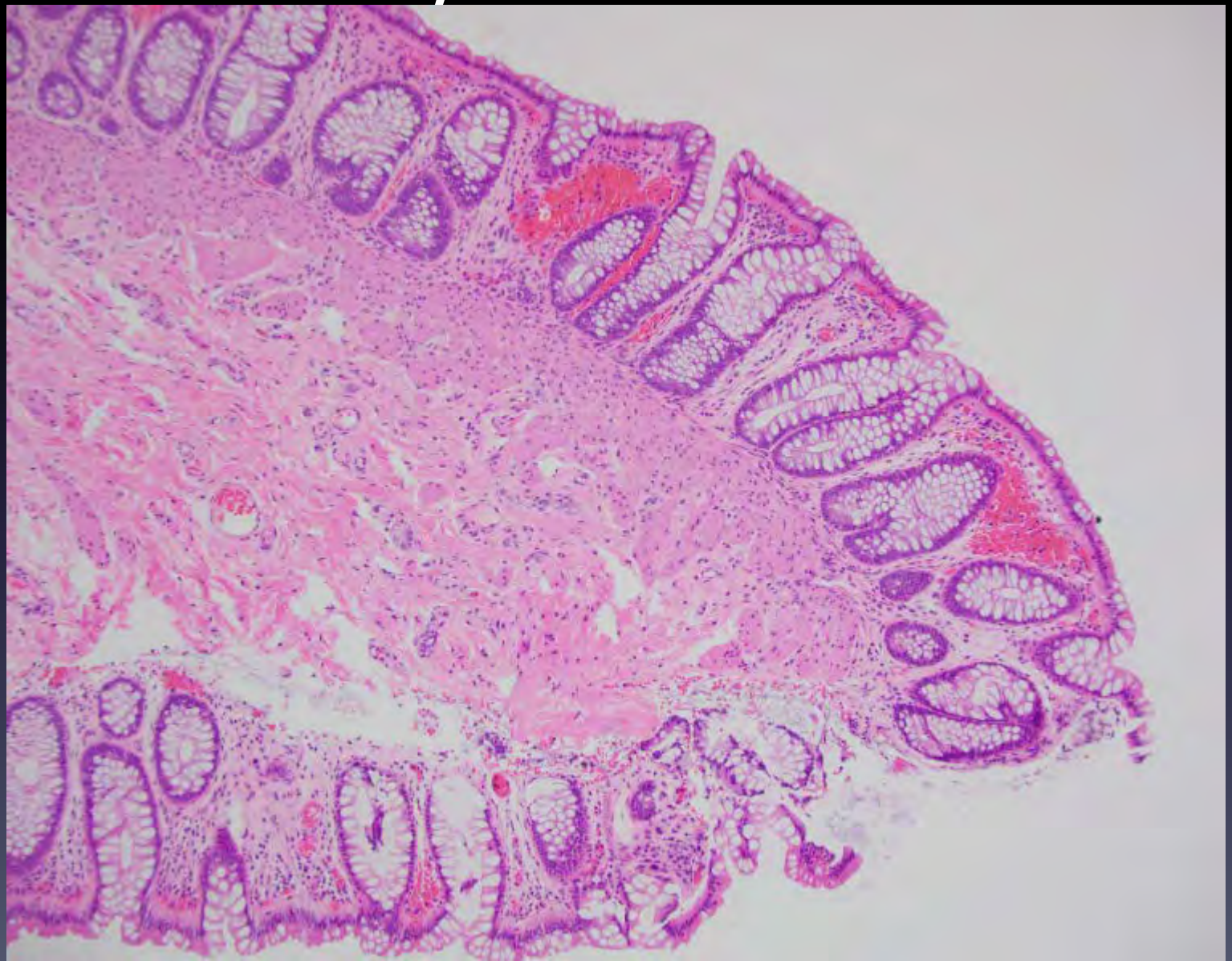


Suction Rectal biopsy

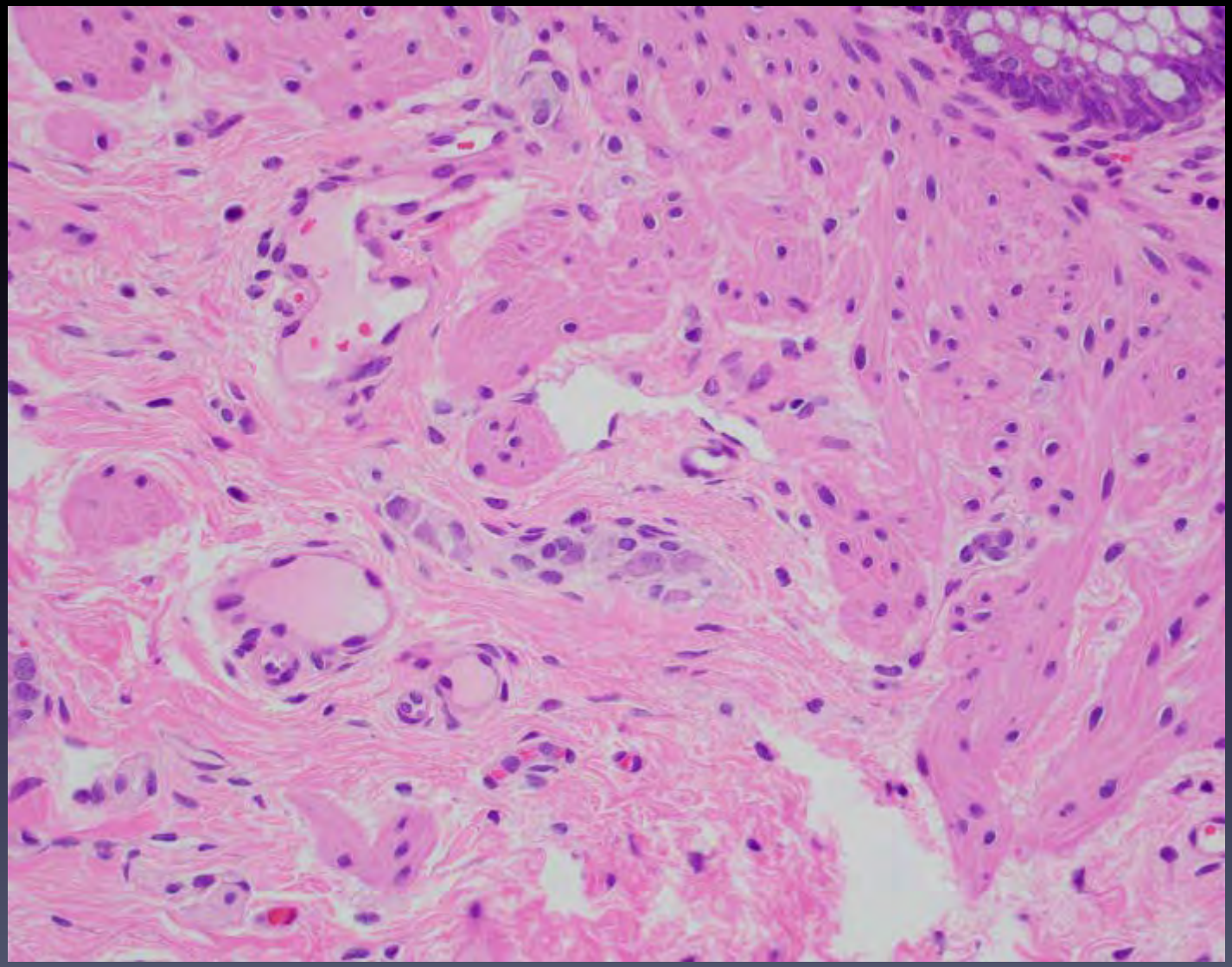




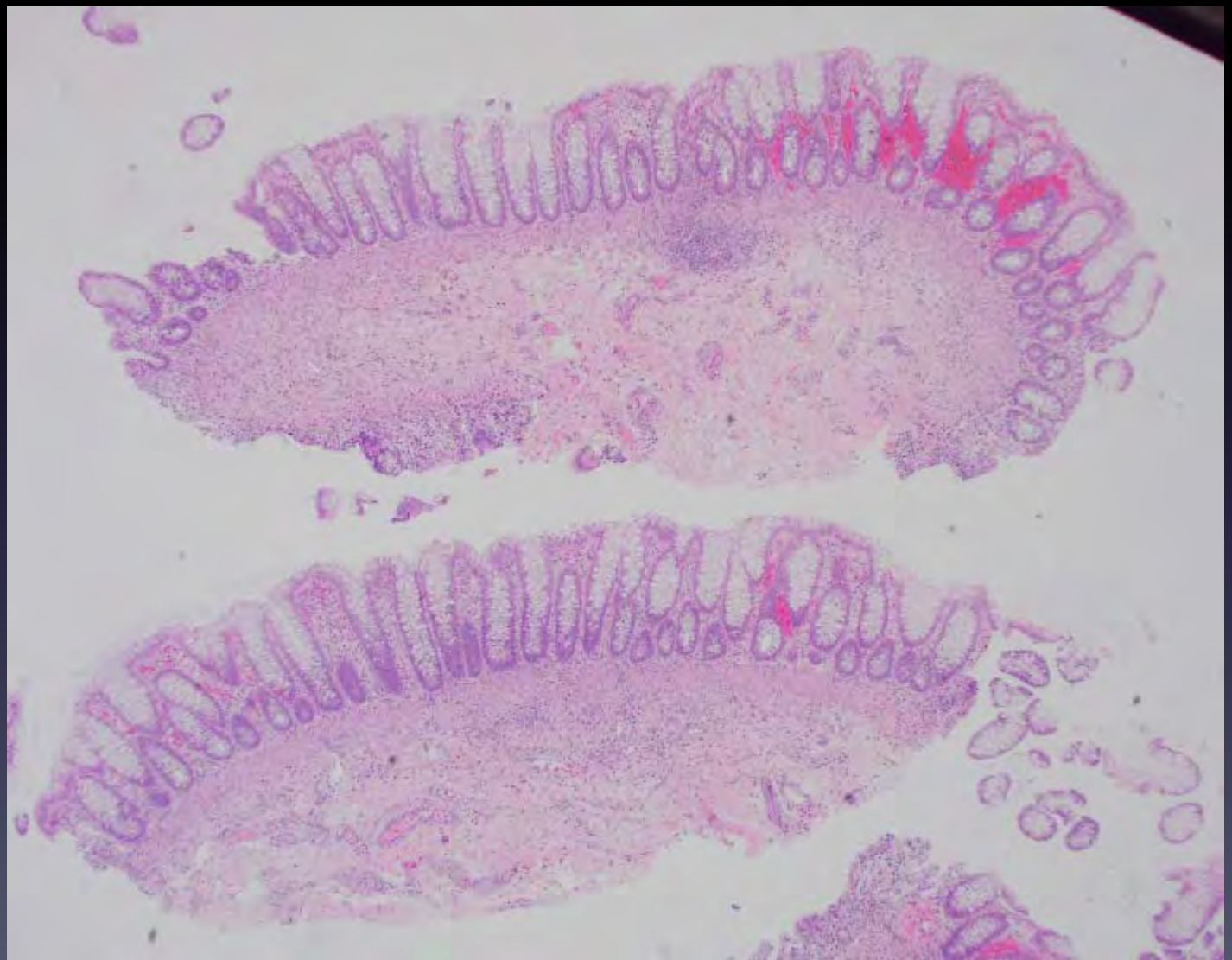
Suction Bx, H&E 10x- normal



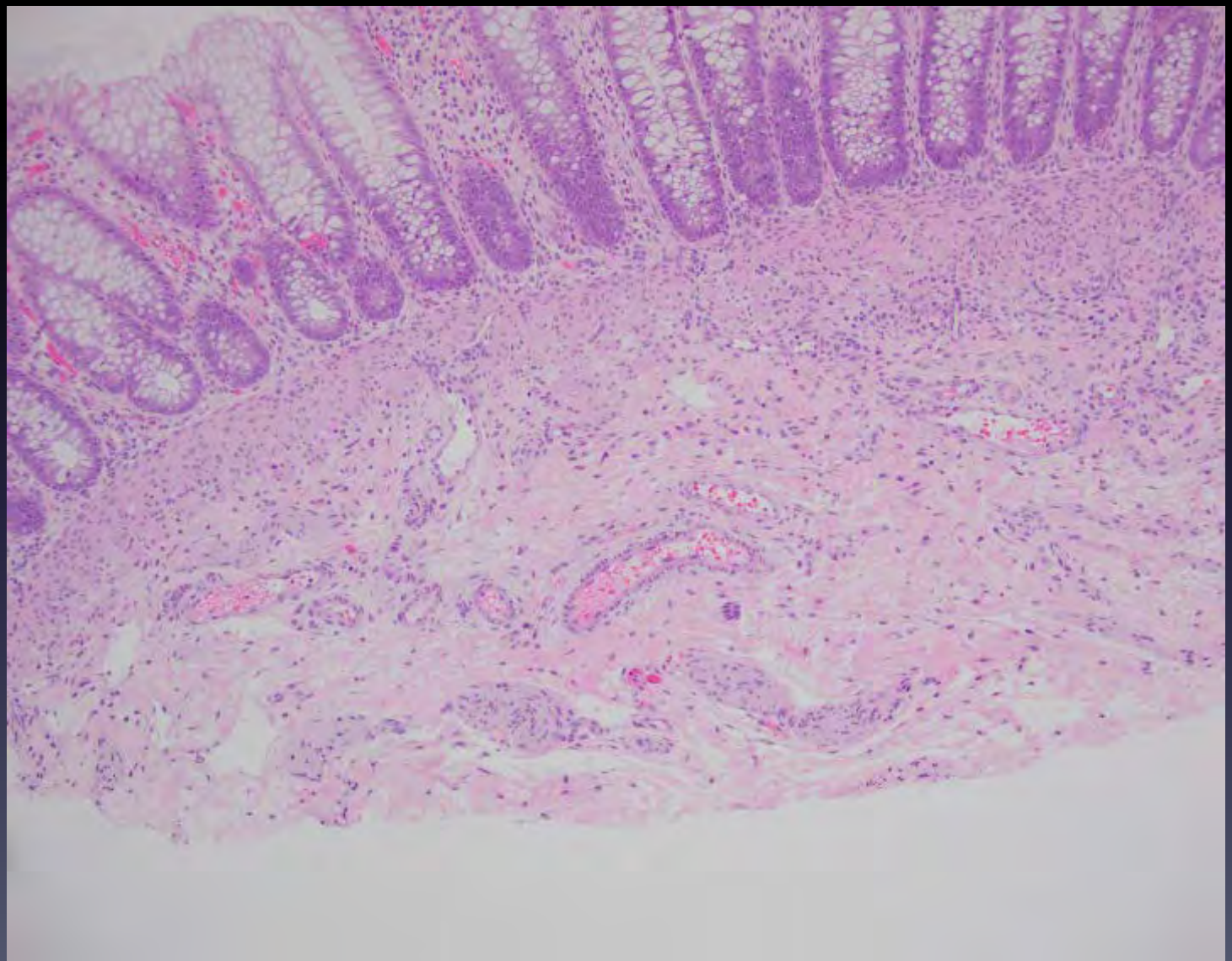
Suction Bx, H&E 40x- normal



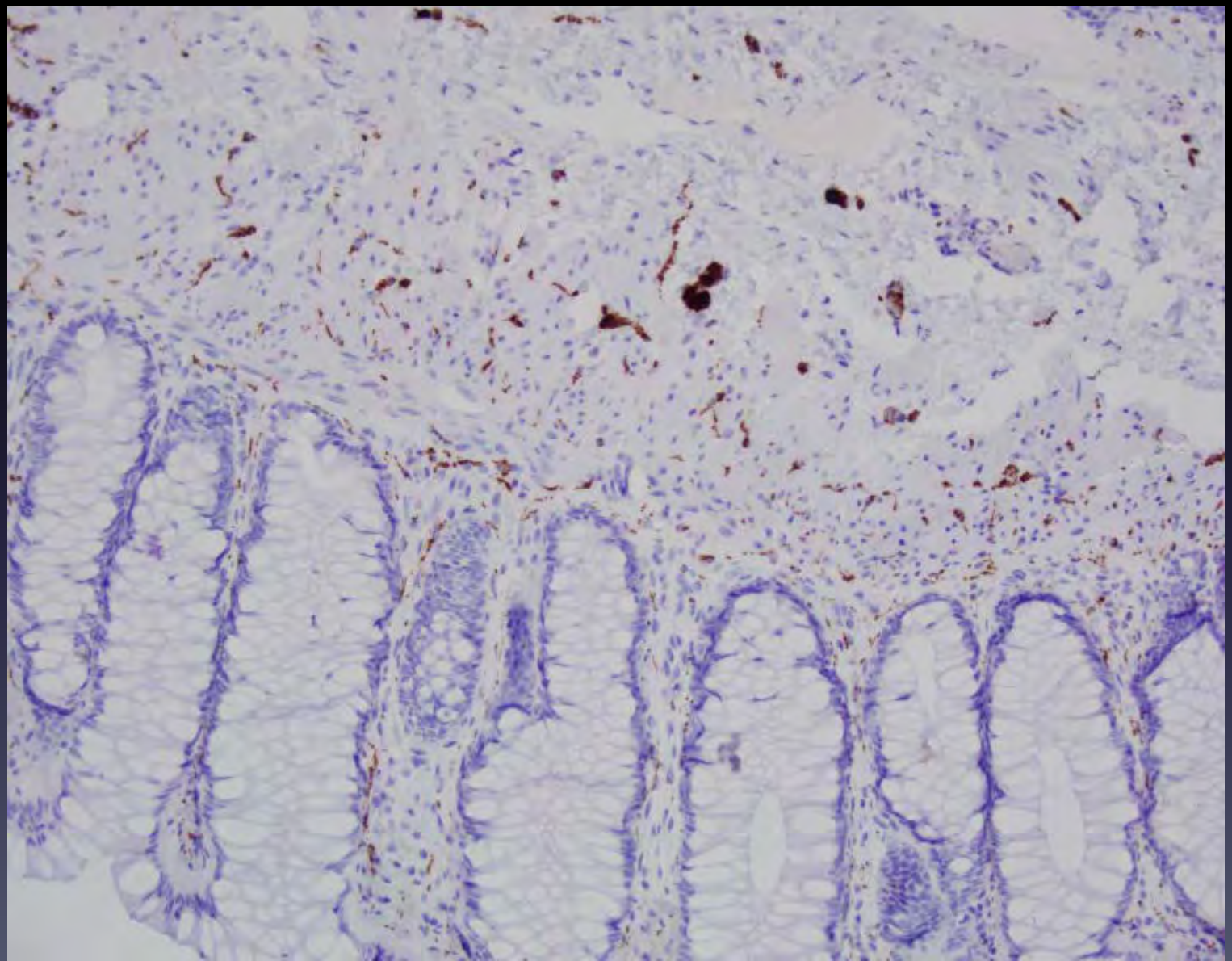
Suction Bx(2x) - Aganglionic



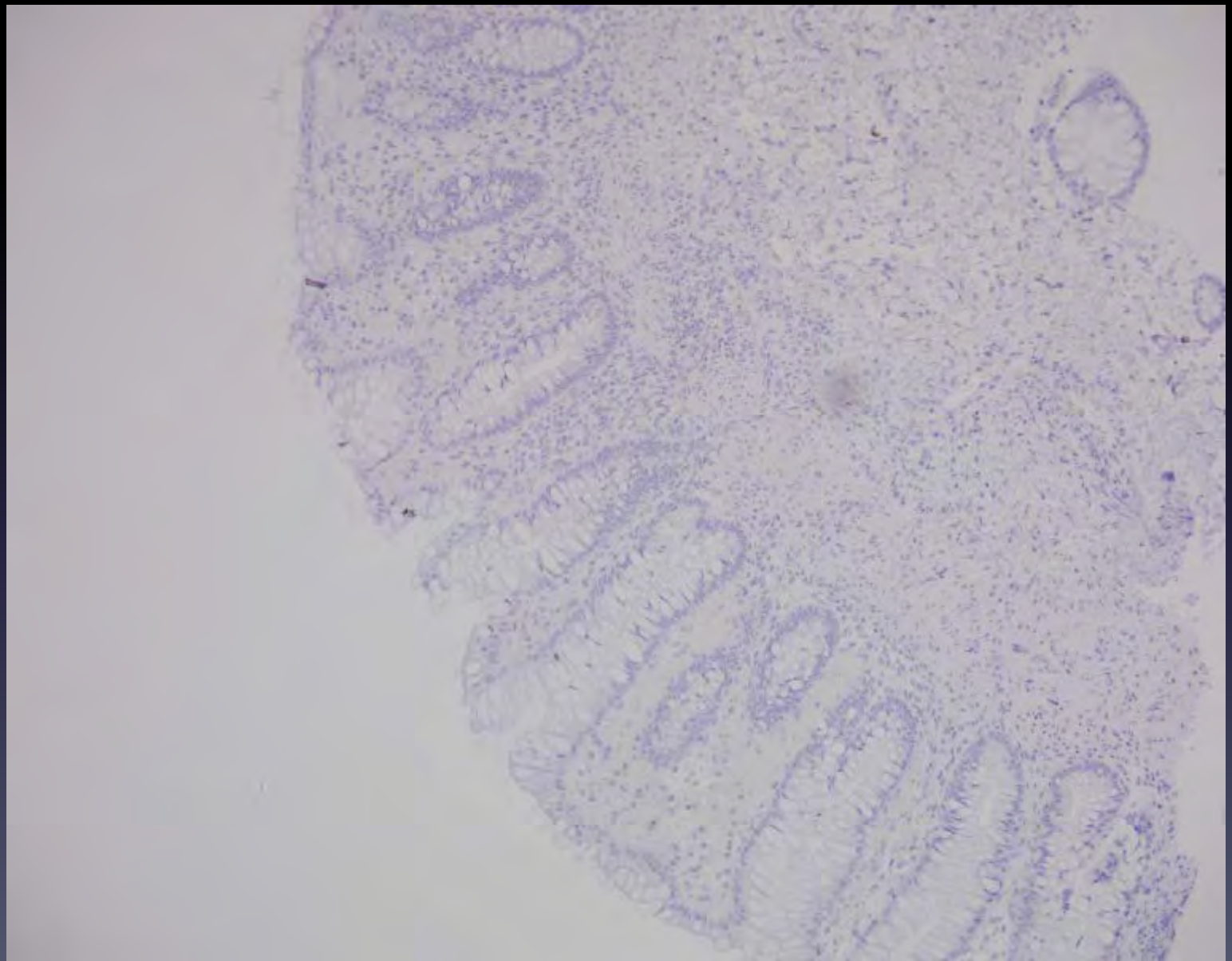
Suction Bx(4x) – Aganglionic – Hypertrophied Nerve Trunks



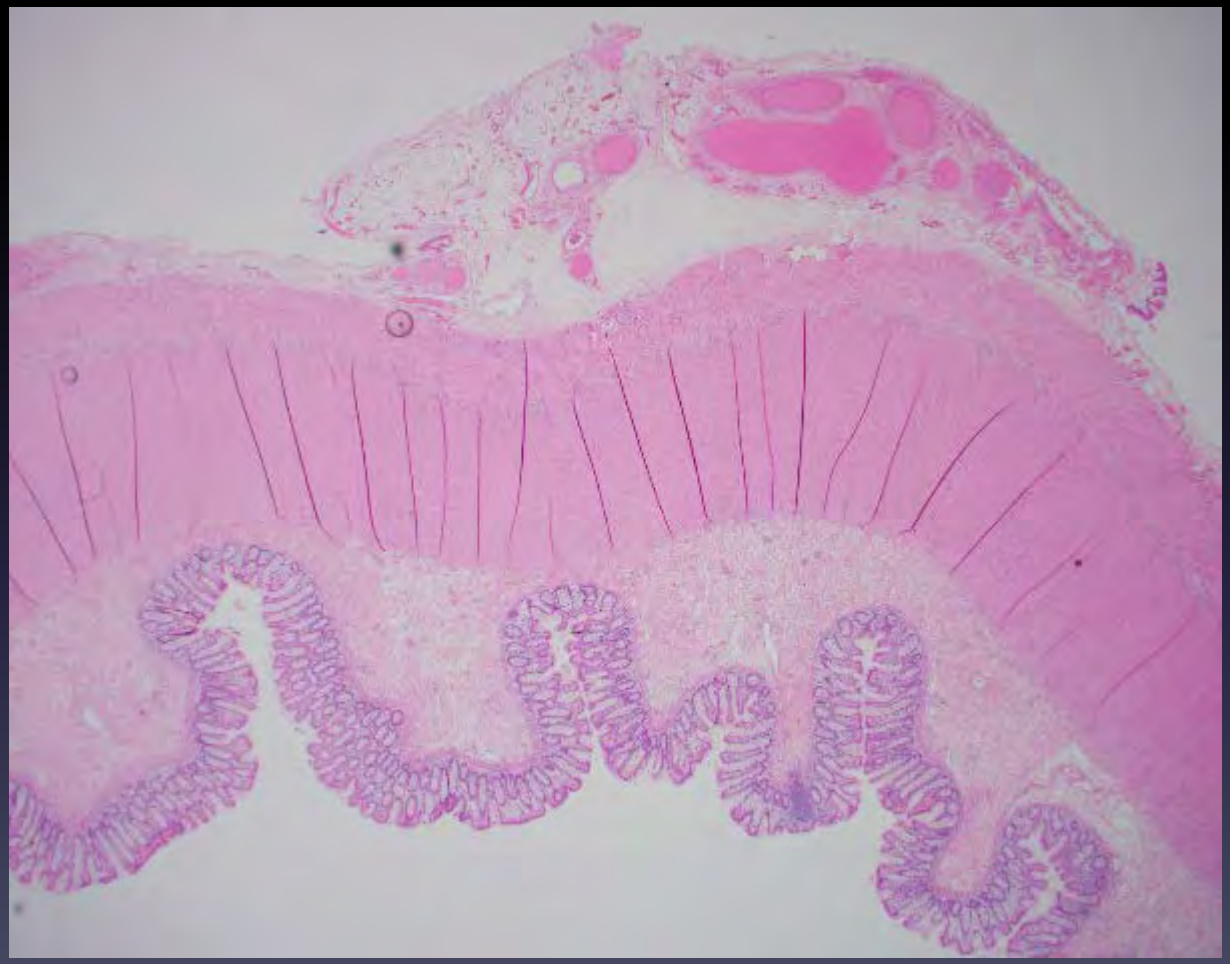
Suction Bx – Calretinin - +ganglion cells



Suction Bx – Calretinin – No ganglion cells

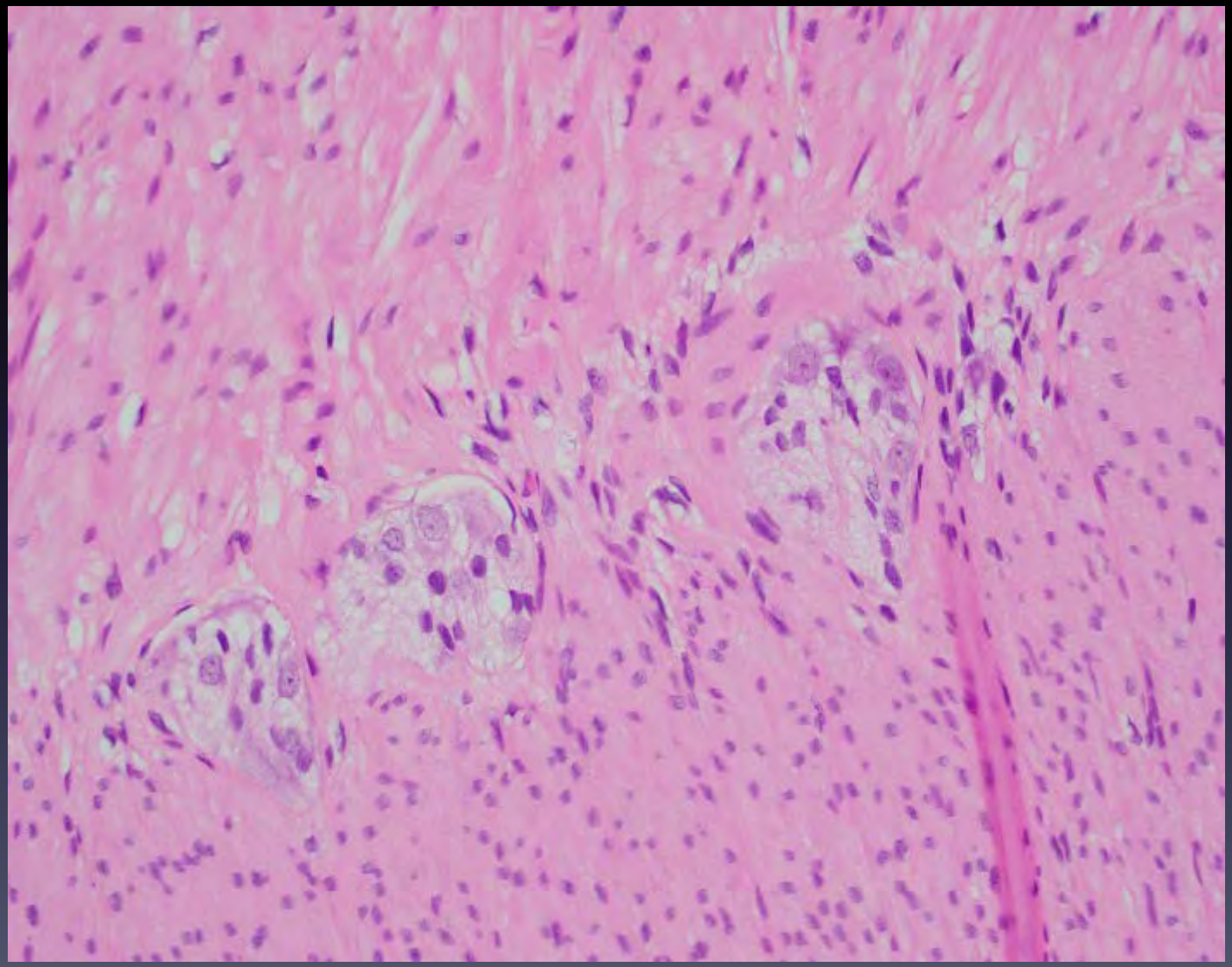


Normal colon (x2)

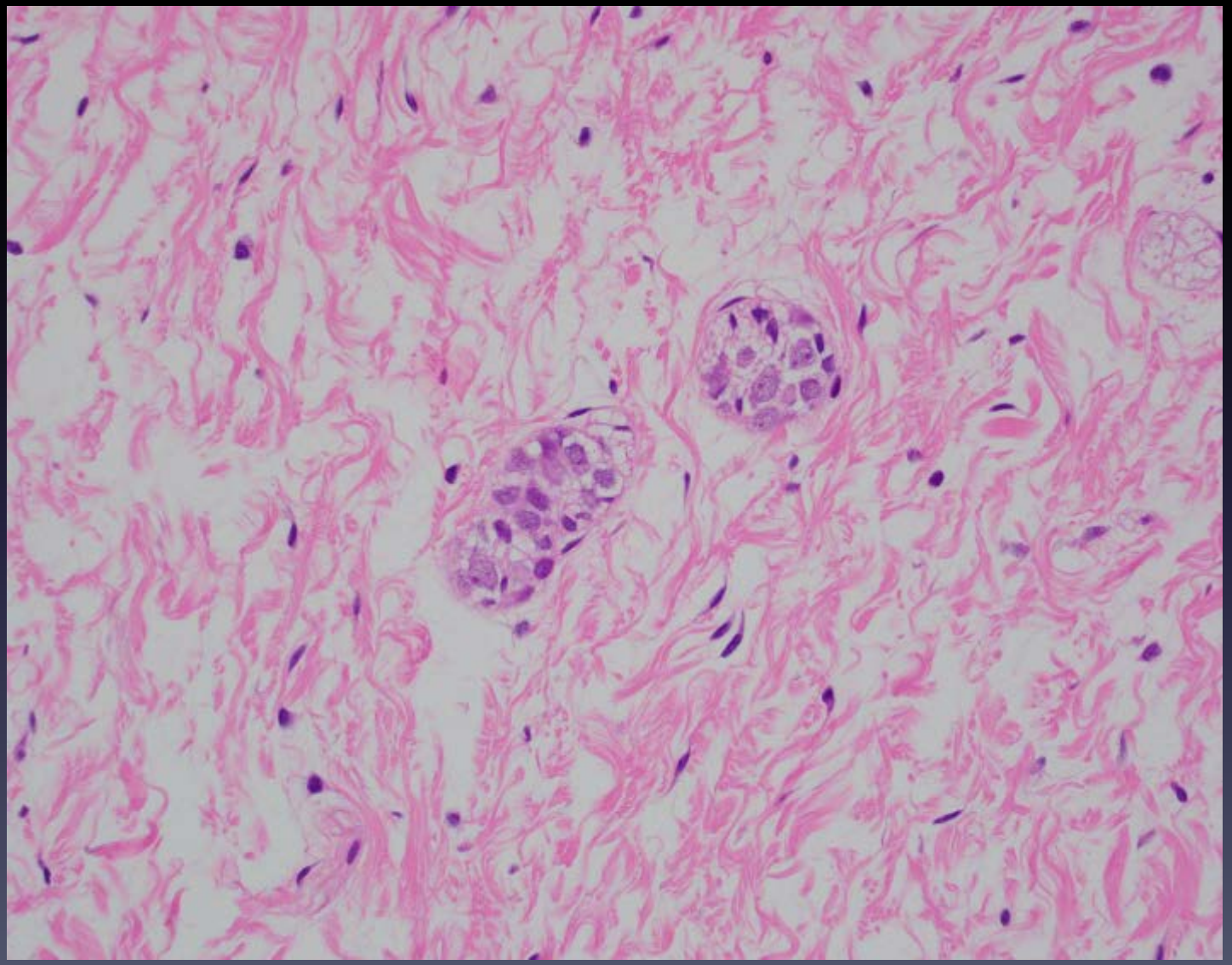


Normal Colon(4x)

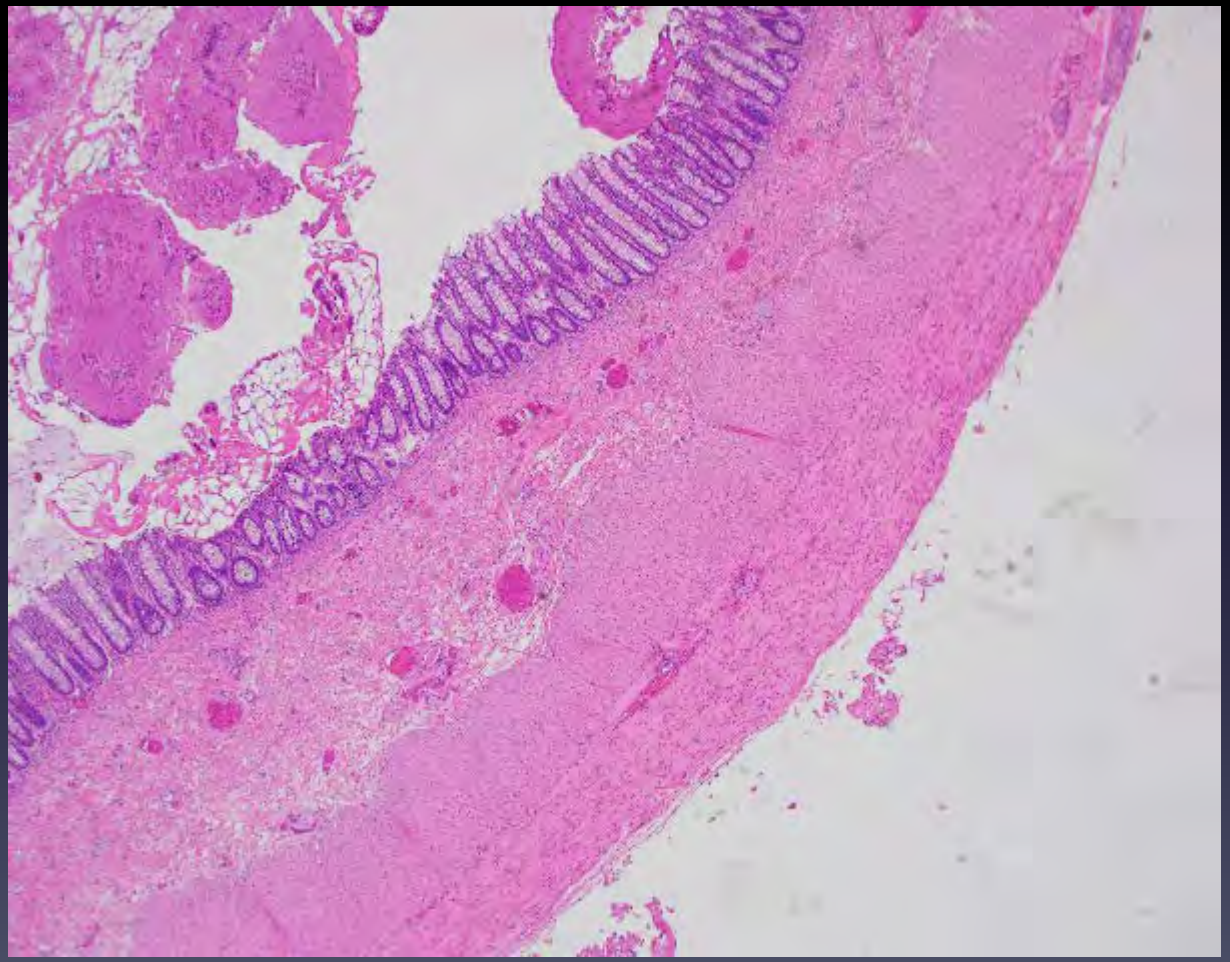
Normal Colon - Muscularis



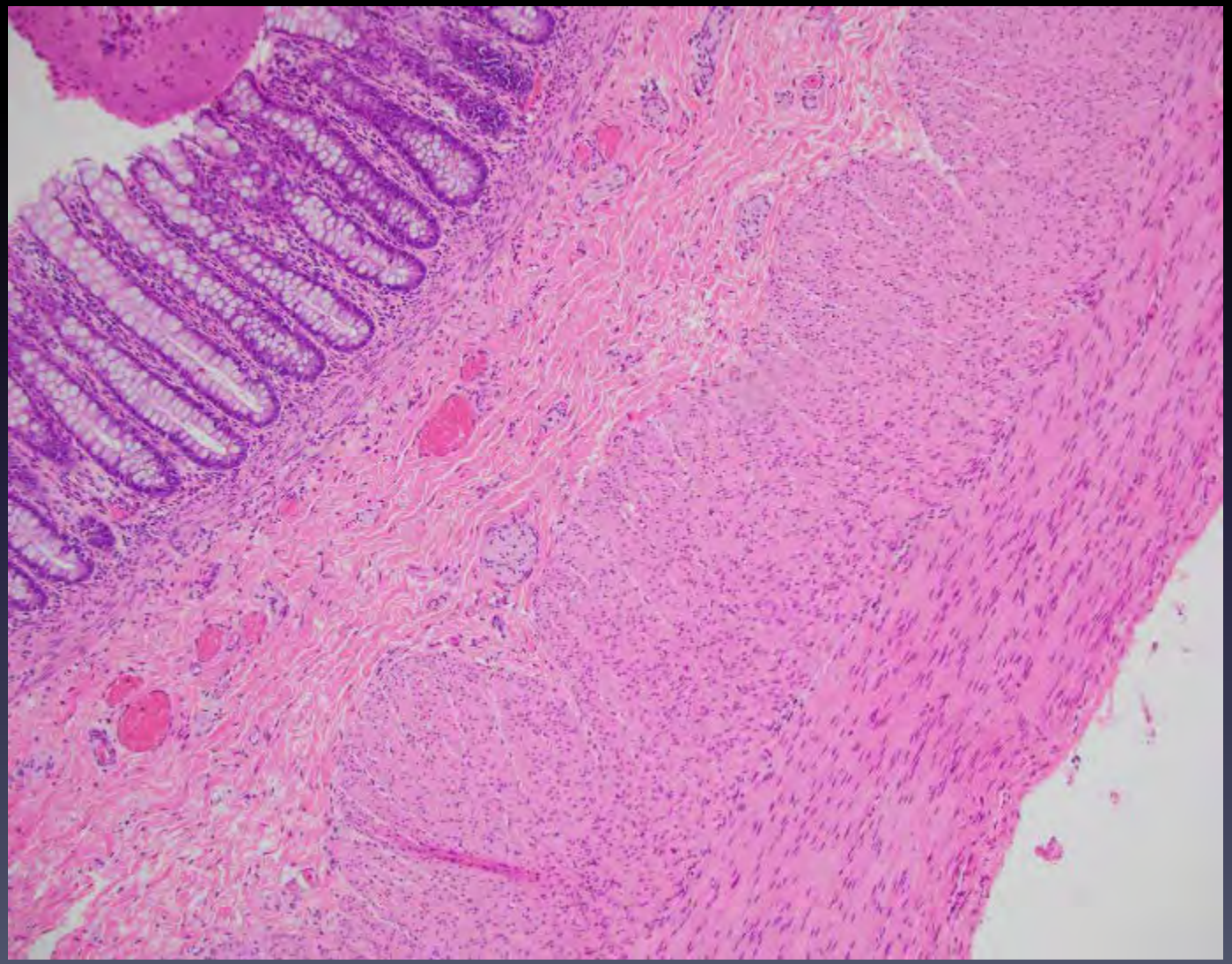
Normal Colon - Submucosal



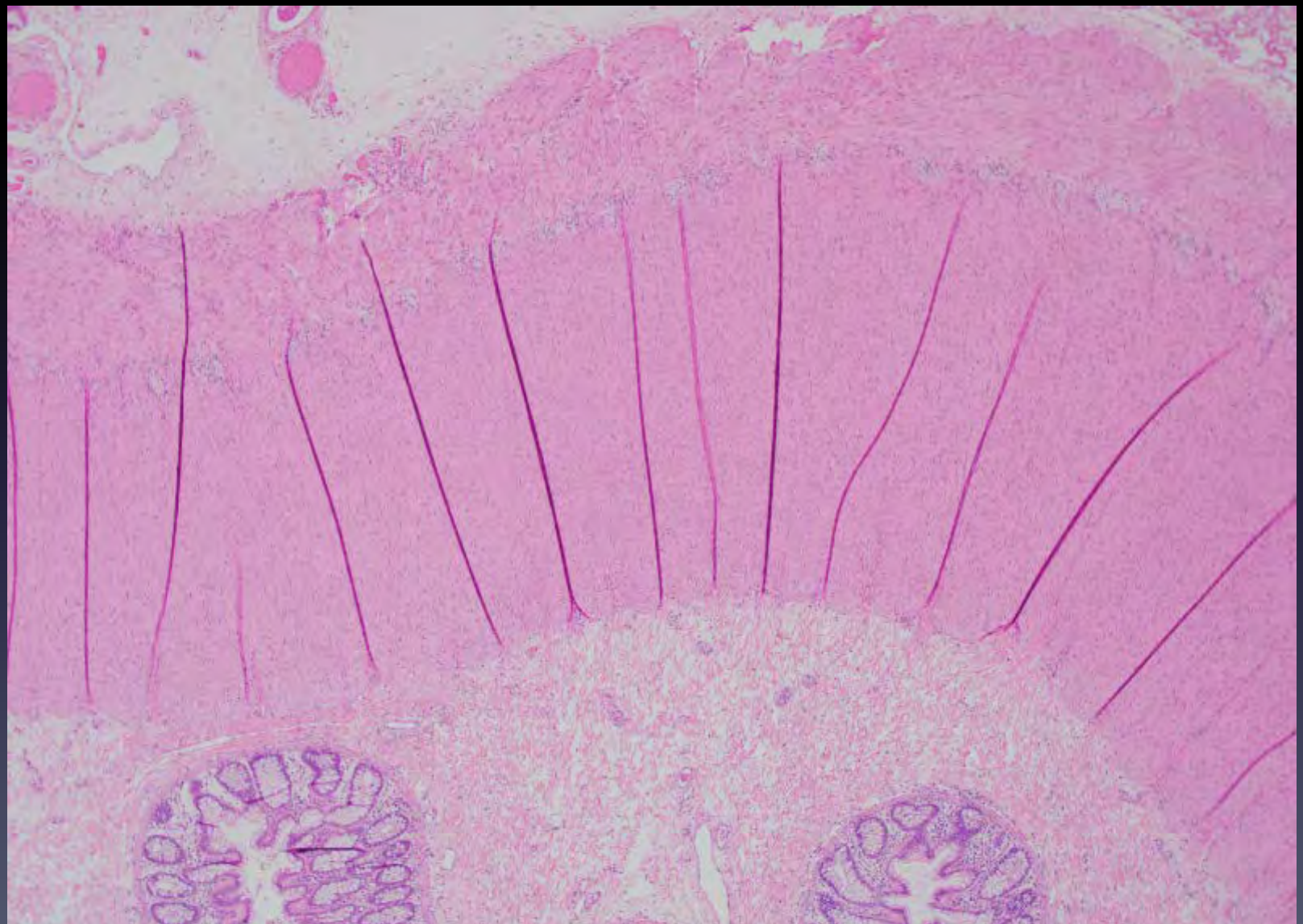
Hirschsprung's (2x)



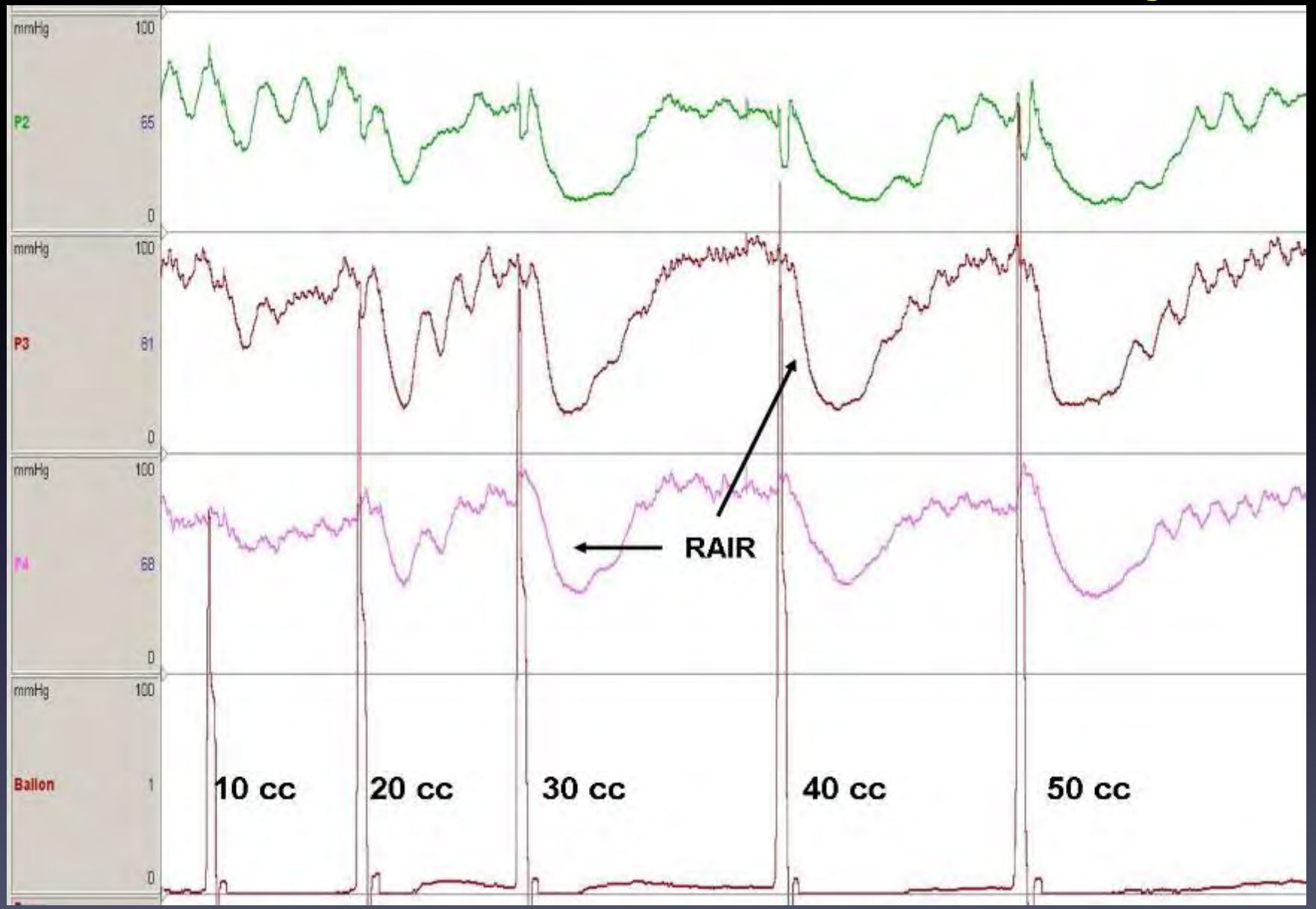
Hirschsprung's (4x)

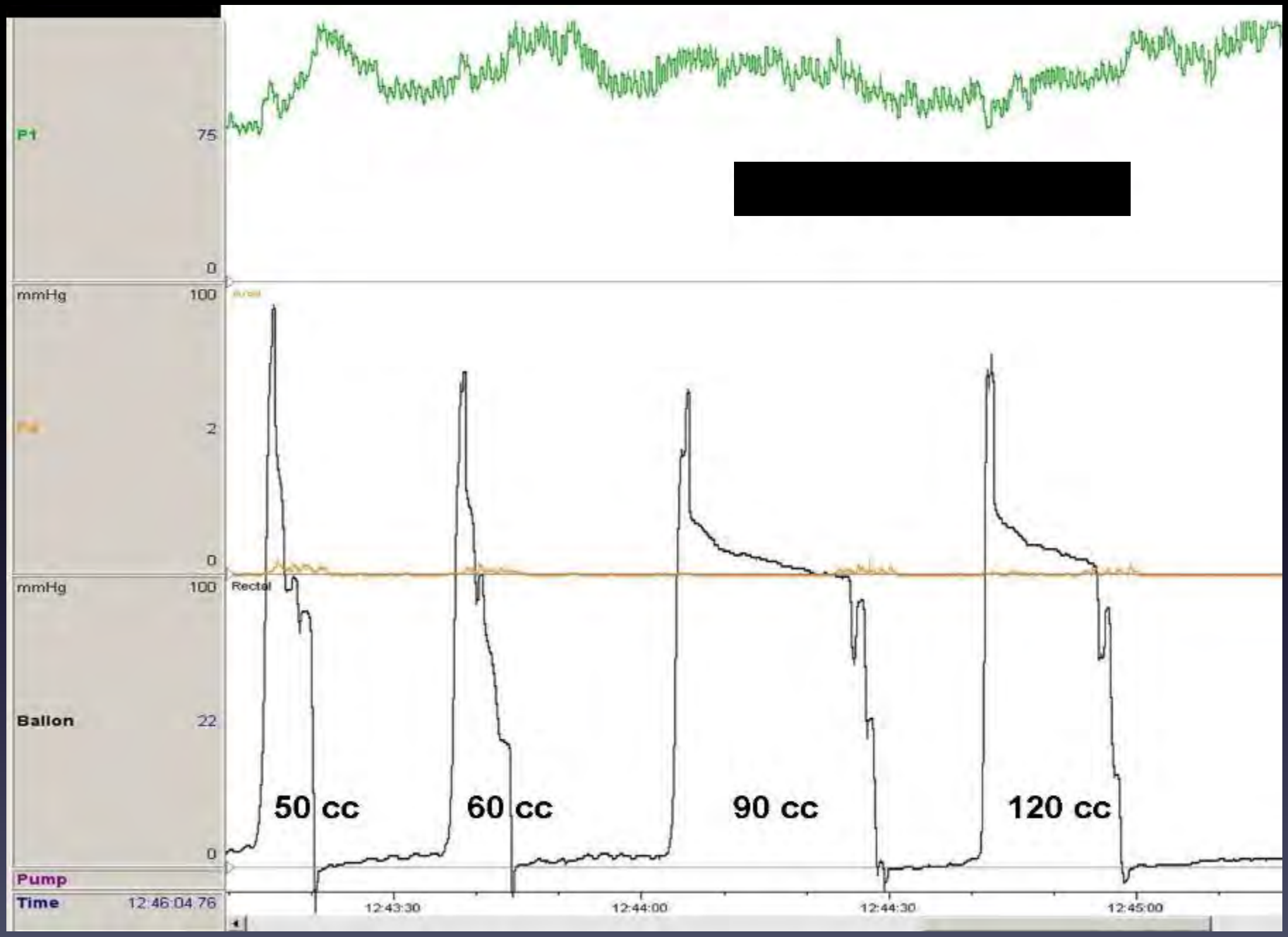


Normal Colon(4x)

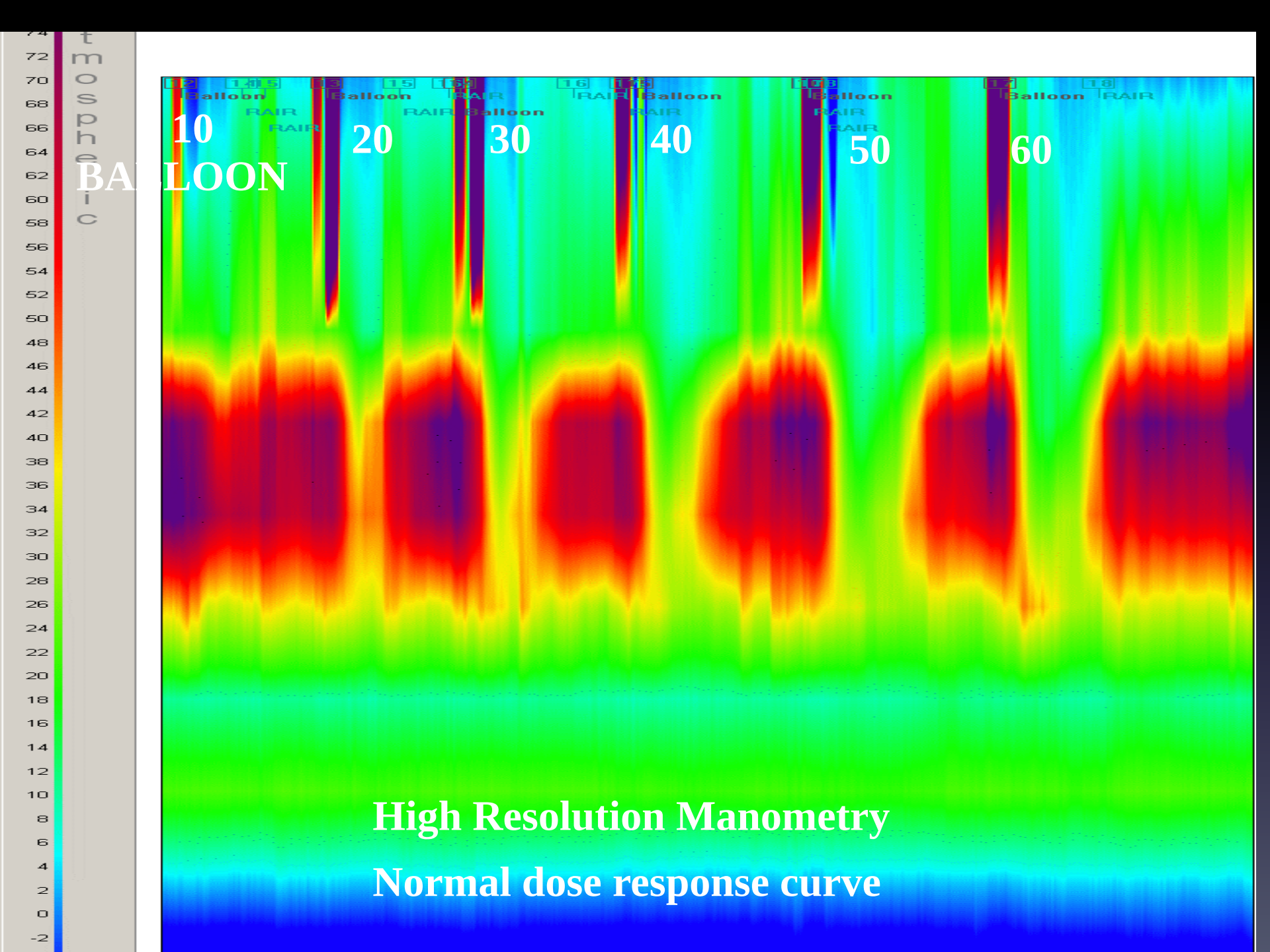


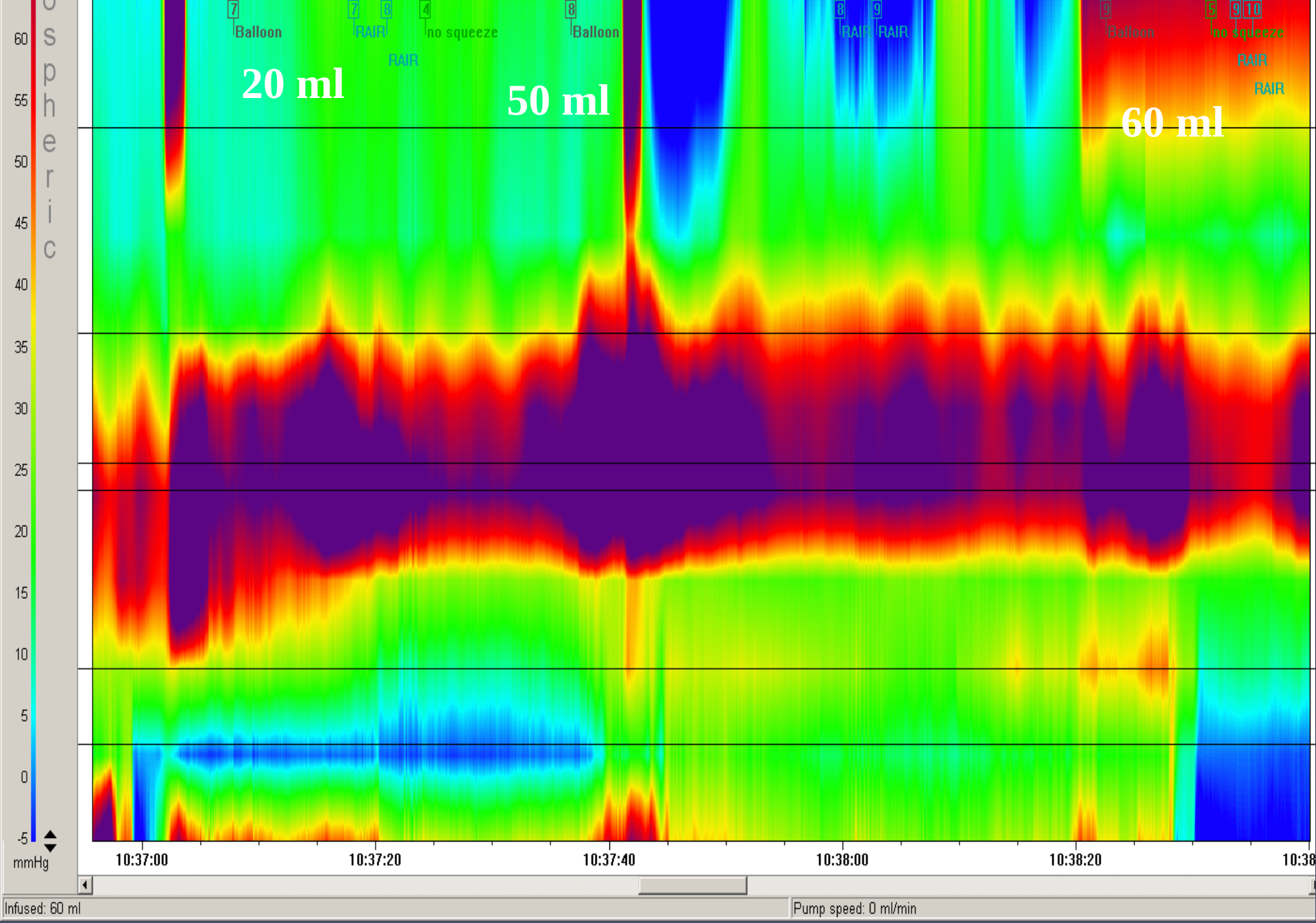
Normal Anorectal Manometry





Hirschsprung's disease


10
20
30
40
50
60
BALLOON
High Resolution Manometry
Normal dose response curve



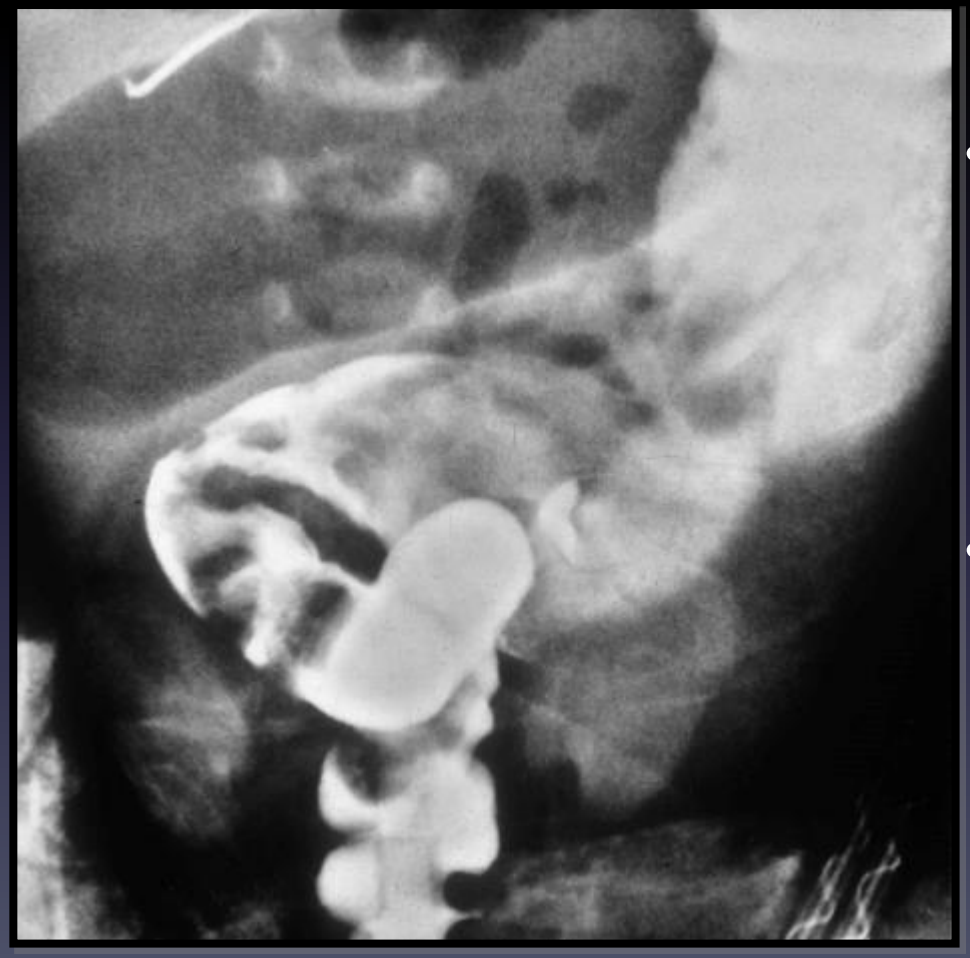
9 year old with intractable constipation. Absent RAIR

Surgical Reconstruction

- #1 Colostomy; Pullthrough – standard approach for years, "old school" but safe; still best for long segment disease
- #2 Rectal decompression; Primary perineal pullthrough, +/- laparoscopic mobilization of colon

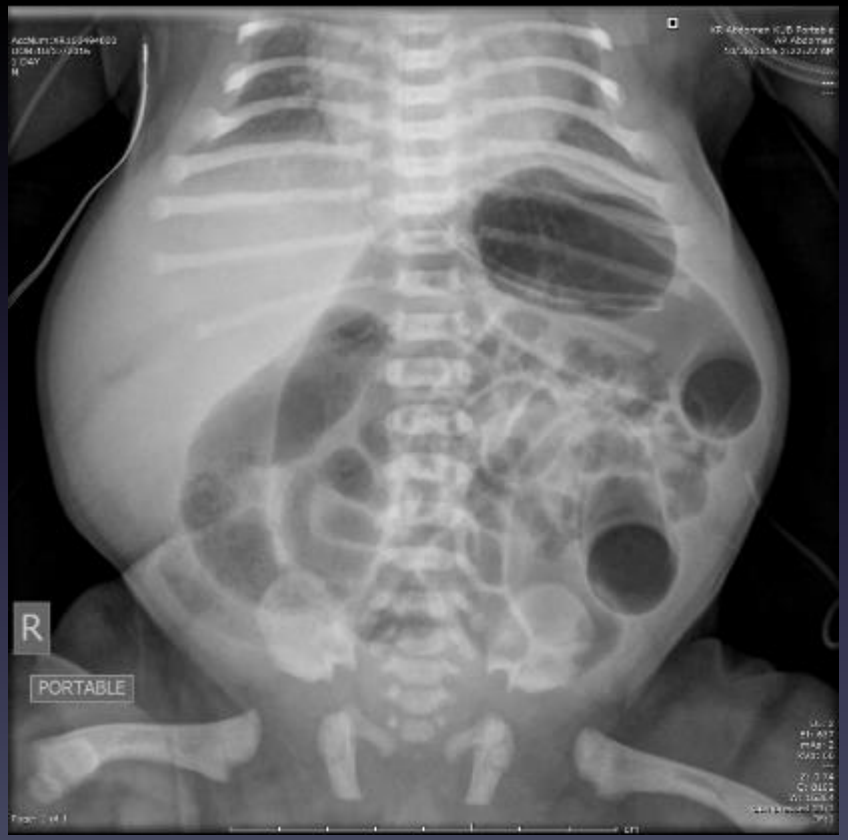
Rectal Decompression

Rectal Decompression

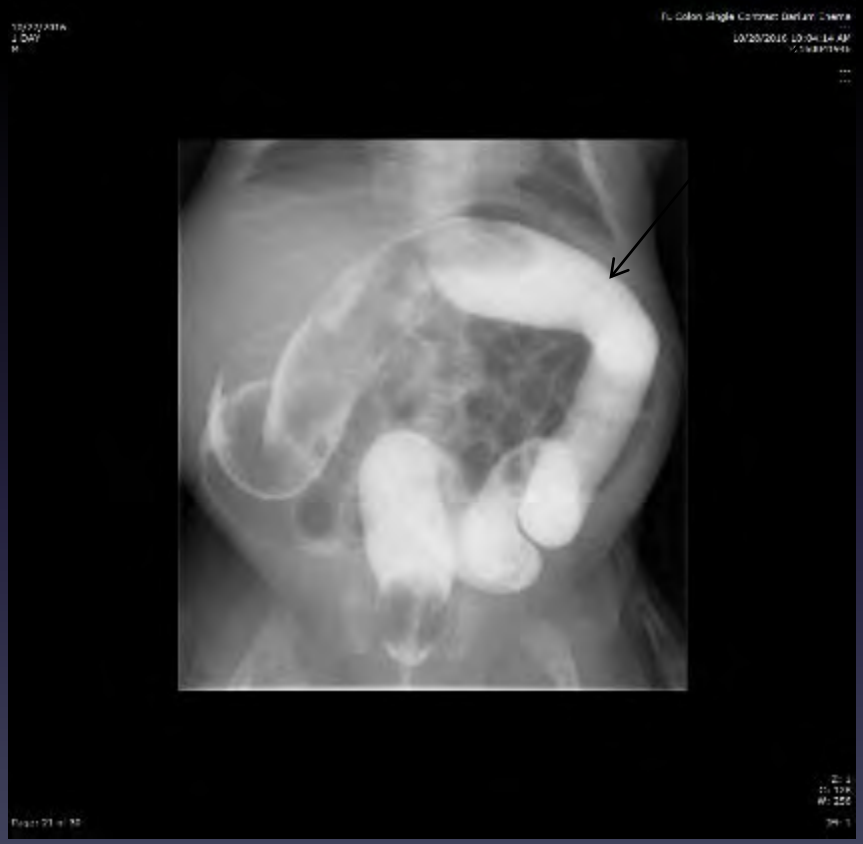


- Goal: decompress fecal load; relieve obstruction; allow proximal bowel to decrease in diameter – easier pullthrough
- Families: decompress tid-qid – often associated with spontaneous bowel movements

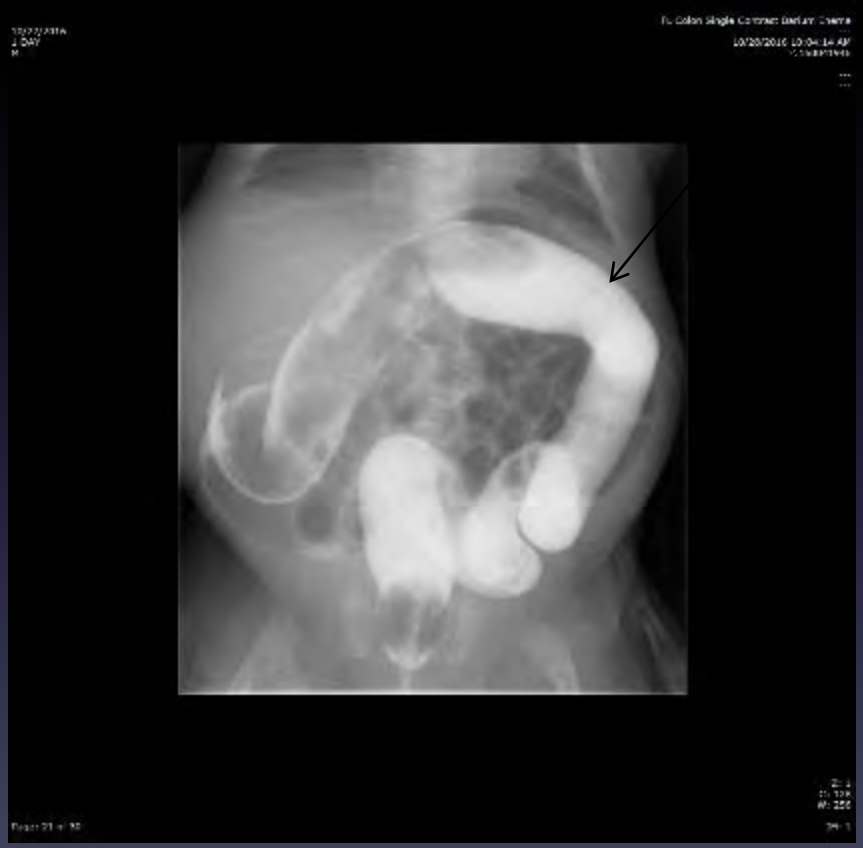
Recent Case



- Newborn male
- Cartilage hair hypoplasia dwarfism
- Abdominal distention
- Failure to pass meconium

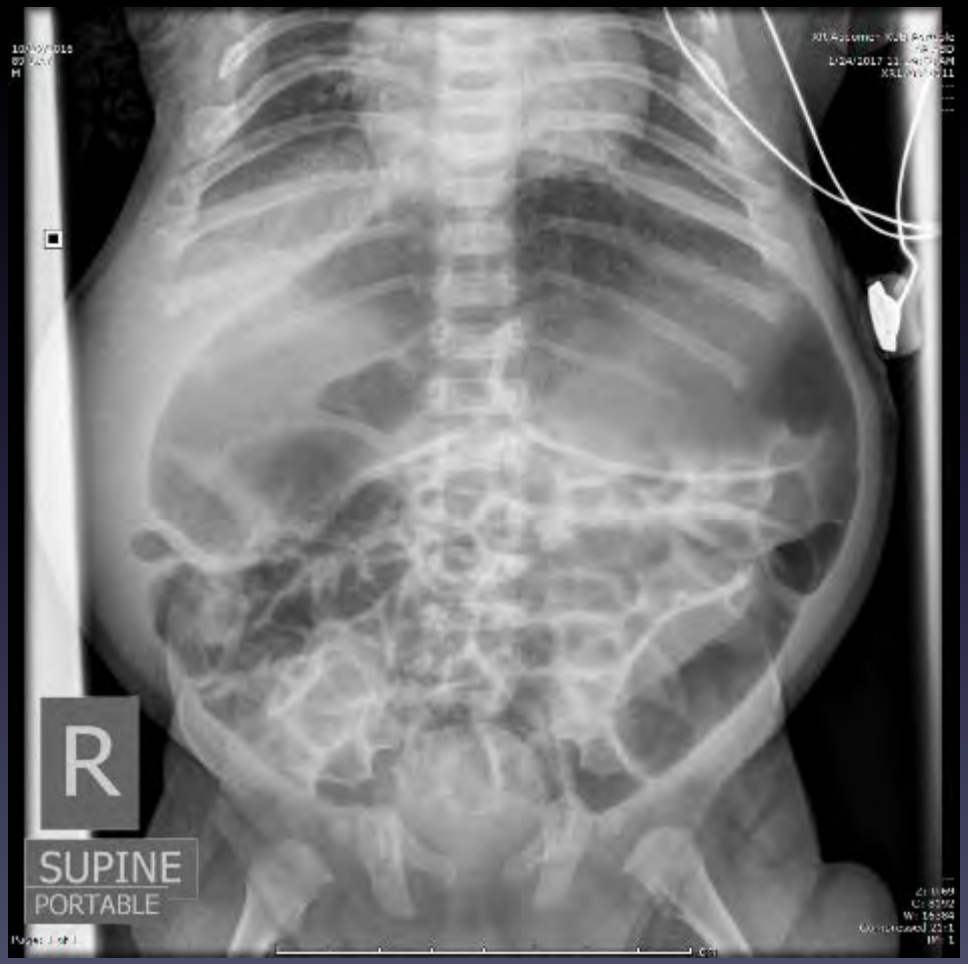


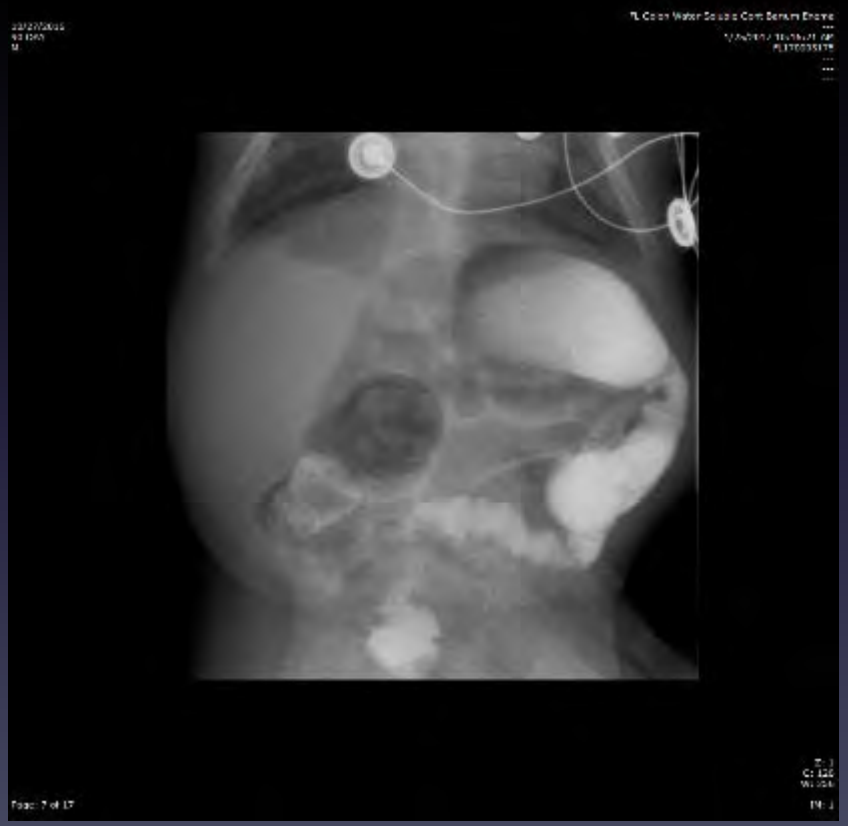
- Suggestion of transition at splenic flexure
- Suction biopsy – Hirschsprung's disease



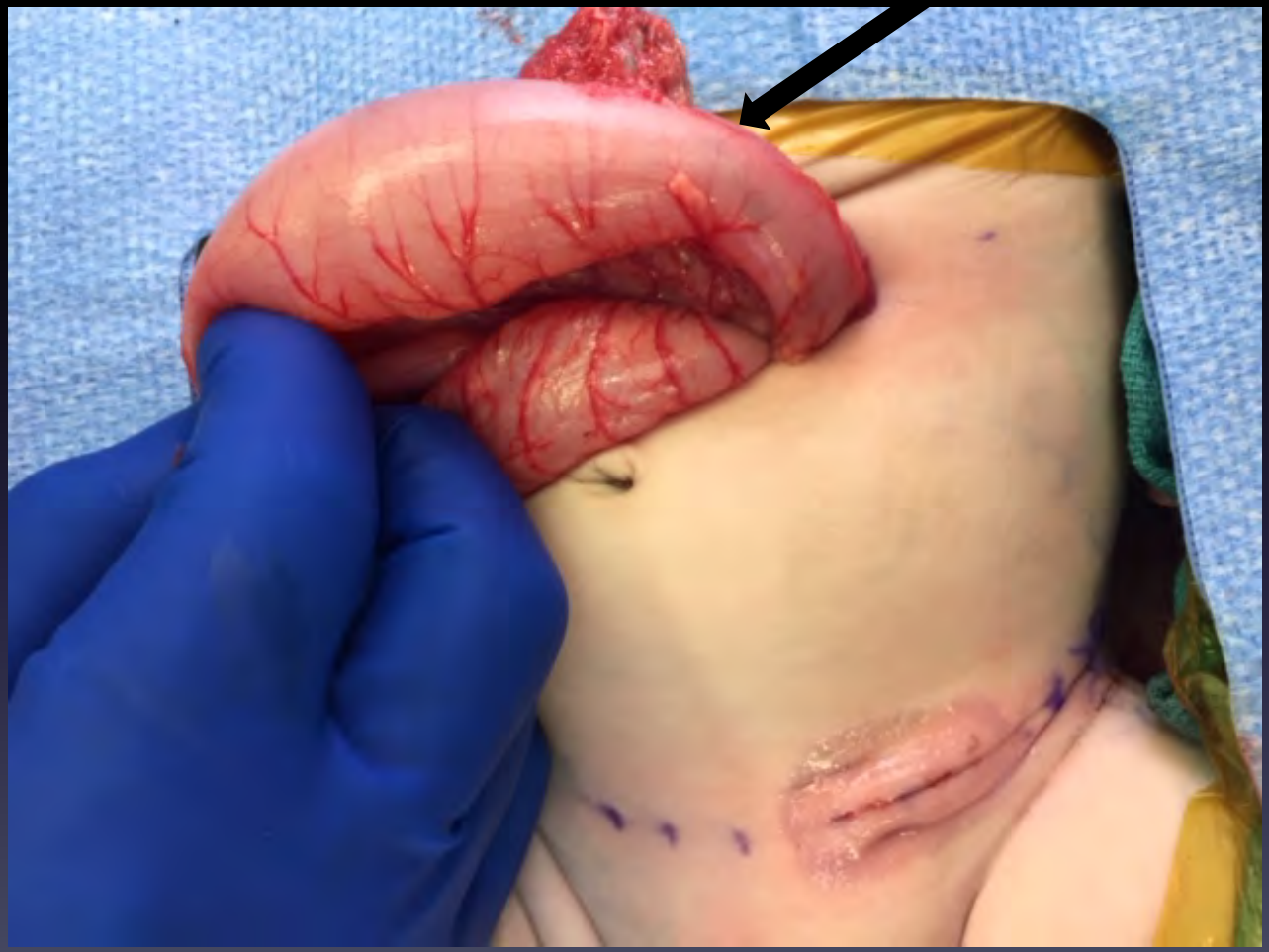
- Rectal decompressions and irrigations
- Family taught technique
- Home on full feeds; abdomen soft, nondistended

Two months of age - Returned – Distention despite good rectal output



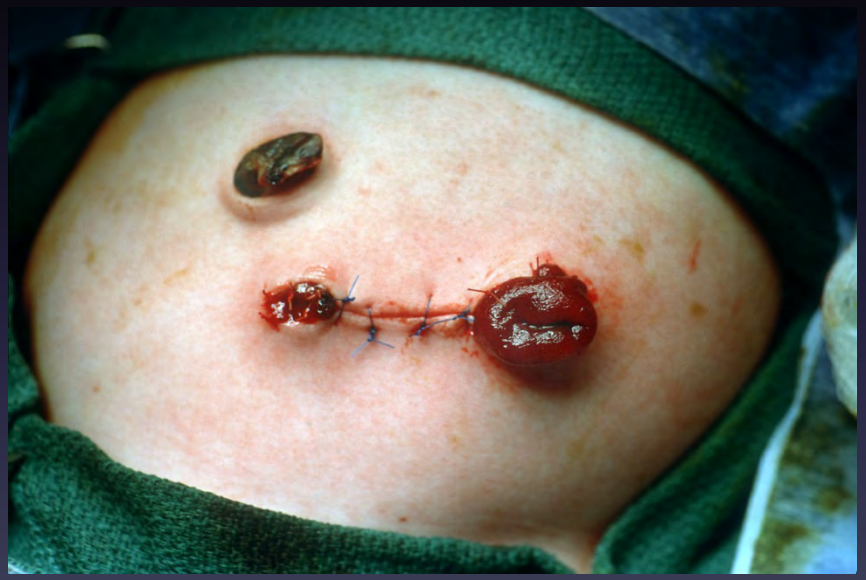


- Enema – more apparent transition at splenic flexure
- Failure of adequate transanal decompression
- Colostomy

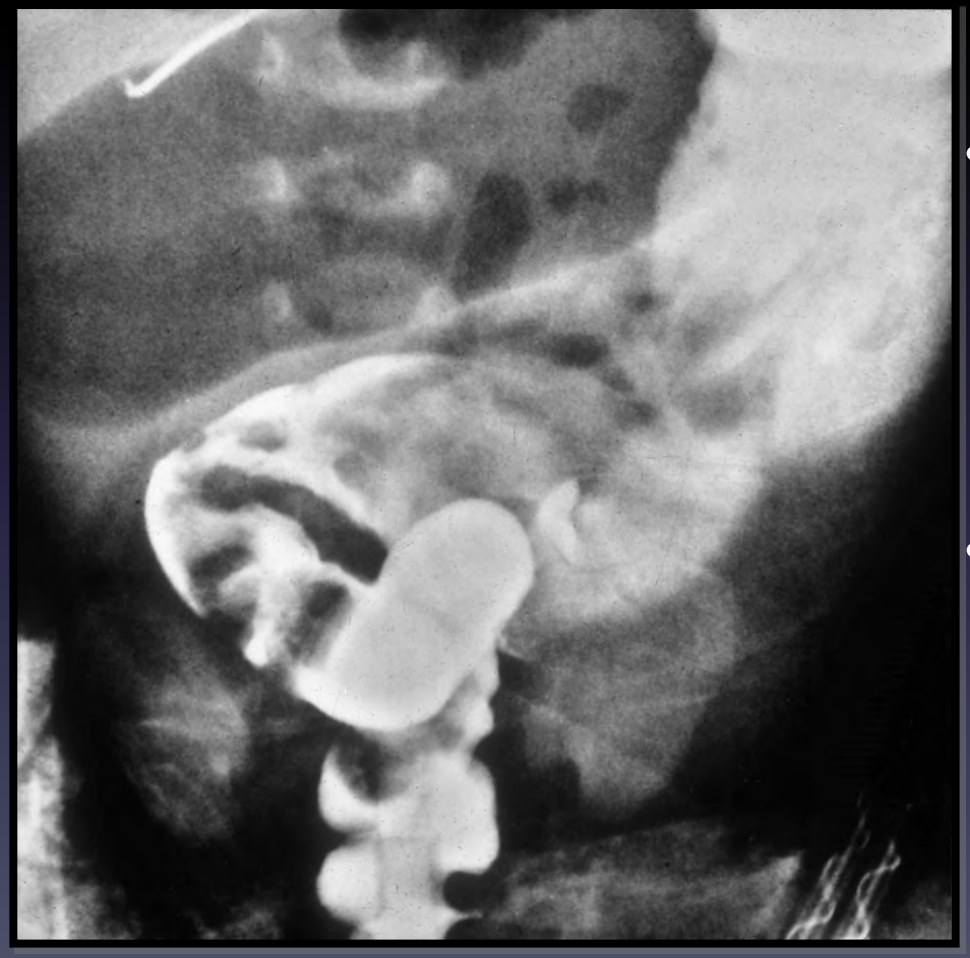


Staged approach

- Colostomy
- Time – bowel decreases in size with decompression
- Pullthrough, 2-4 months later



Rectal Decompression



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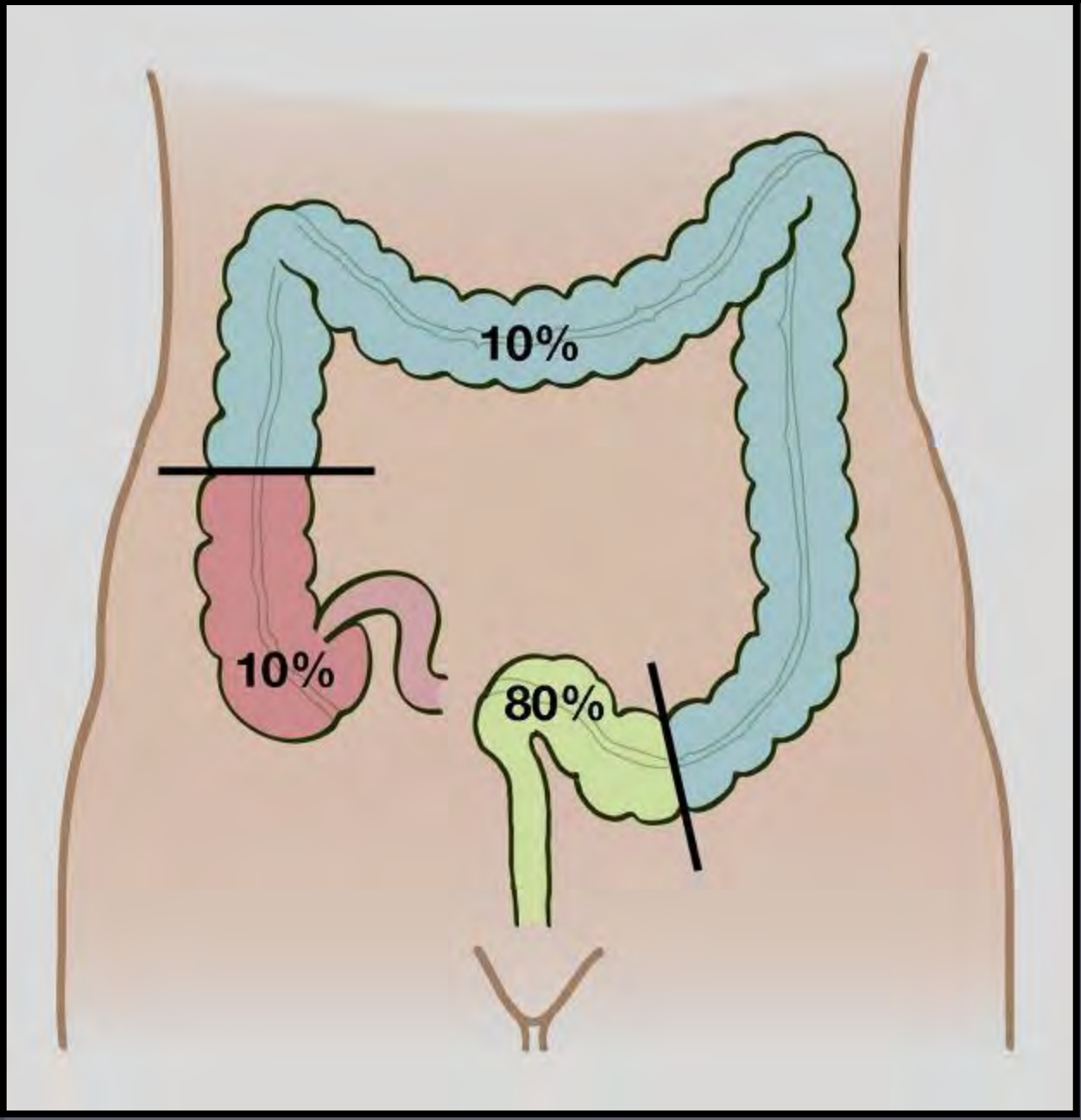
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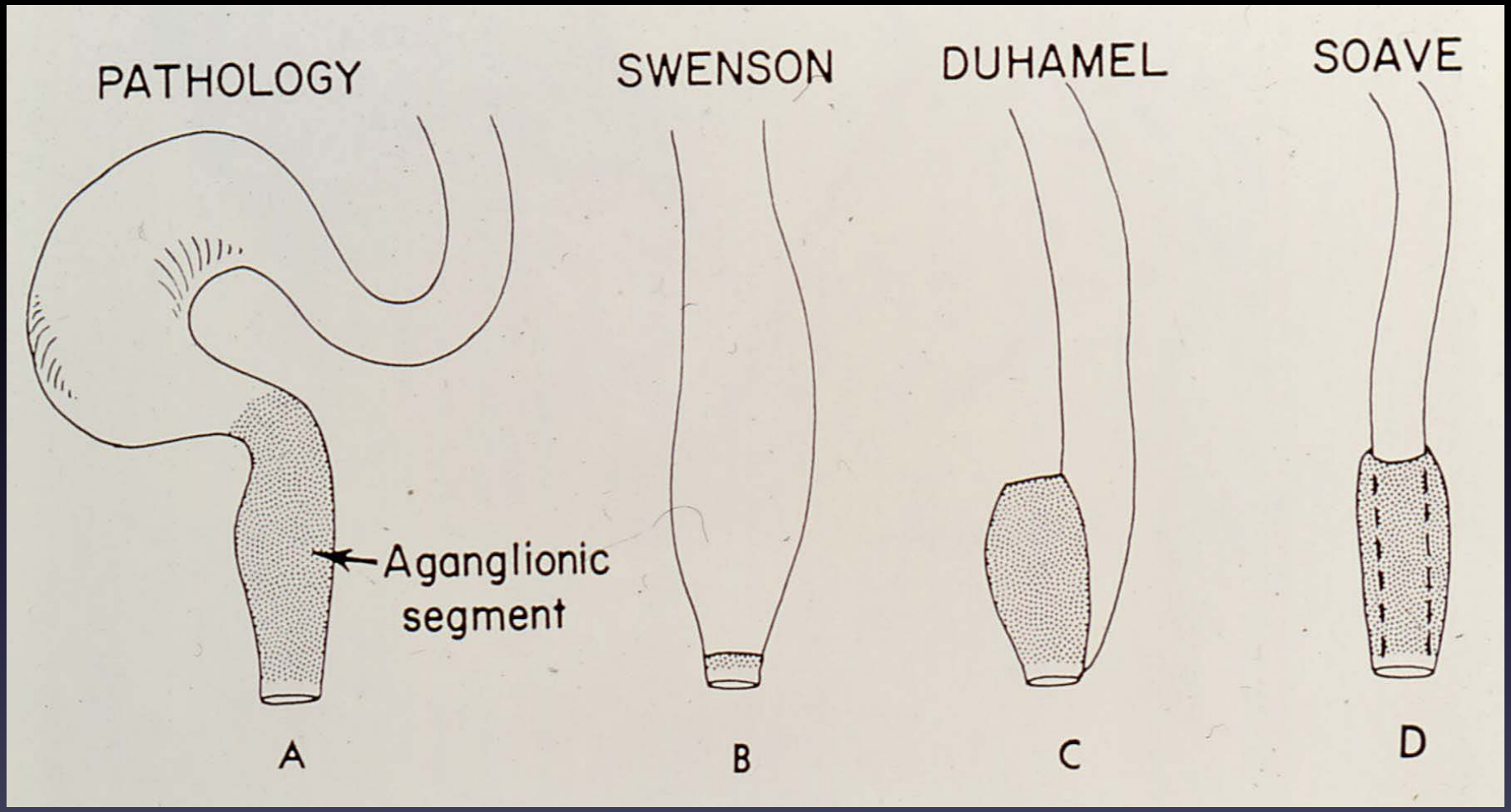
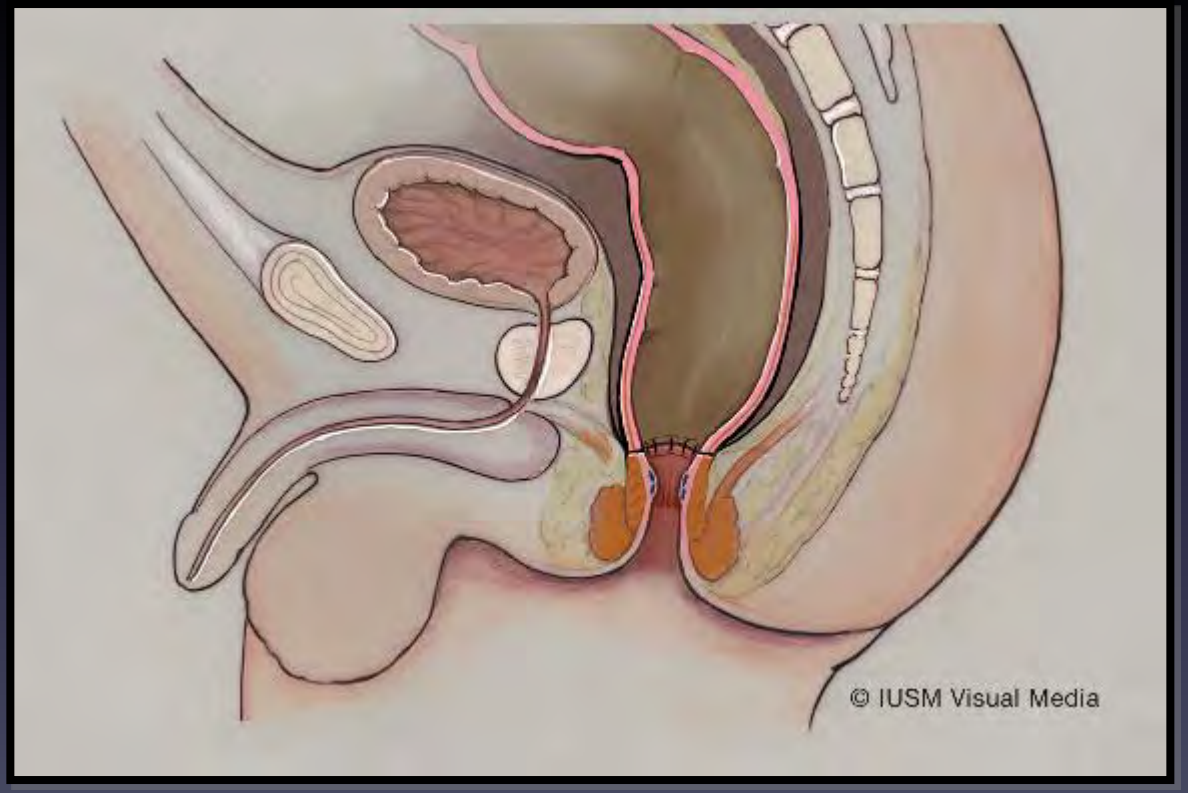
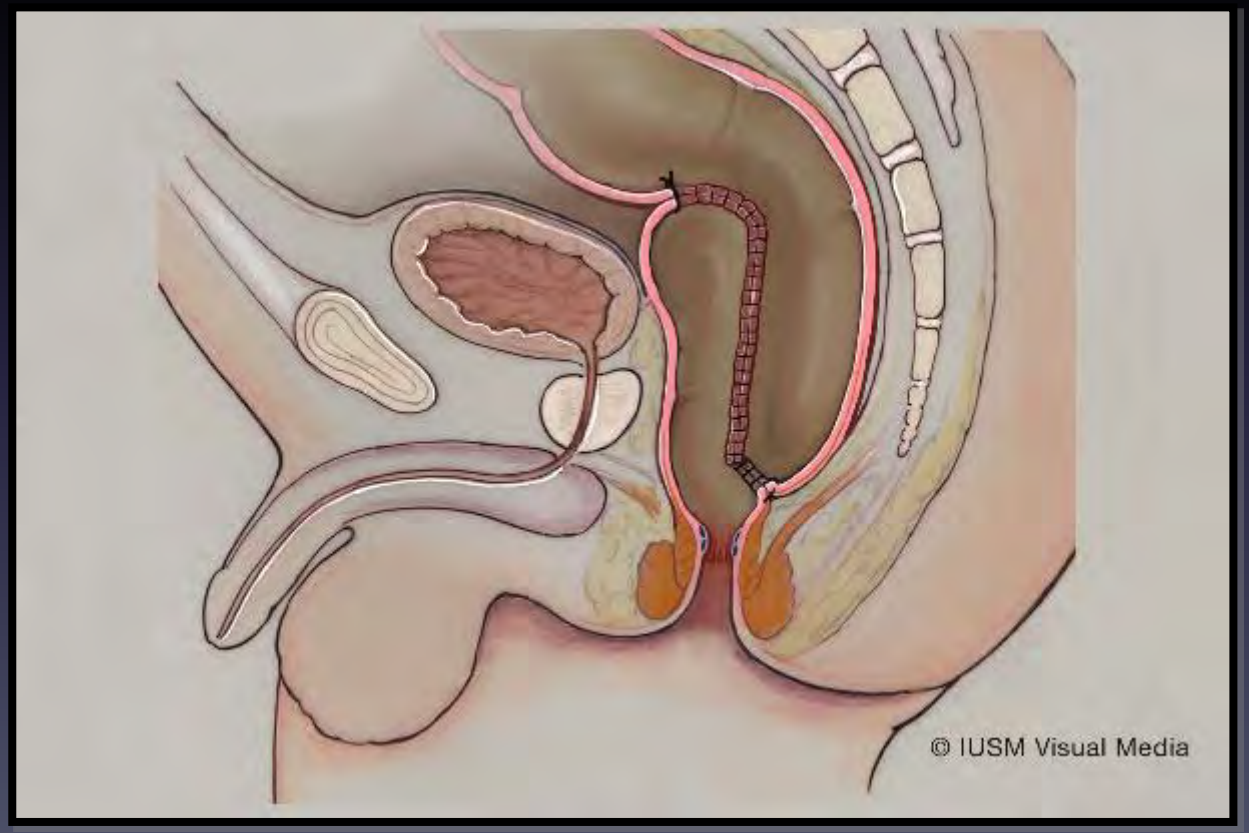

PATHOLOGY
SWENSON
DUHAMEL
SOAVE
Aganglionic segment
A
B
C
D

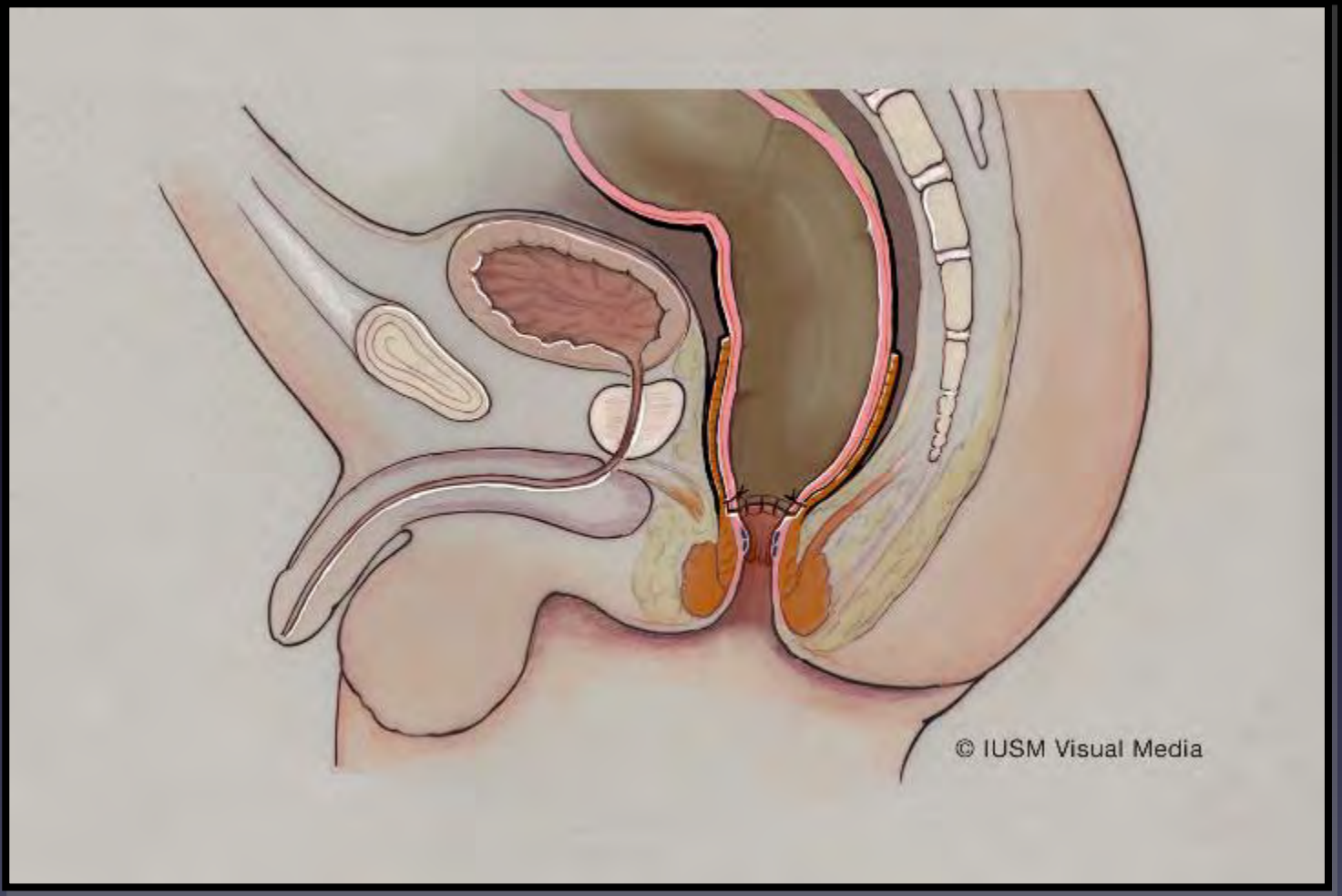
Swenson

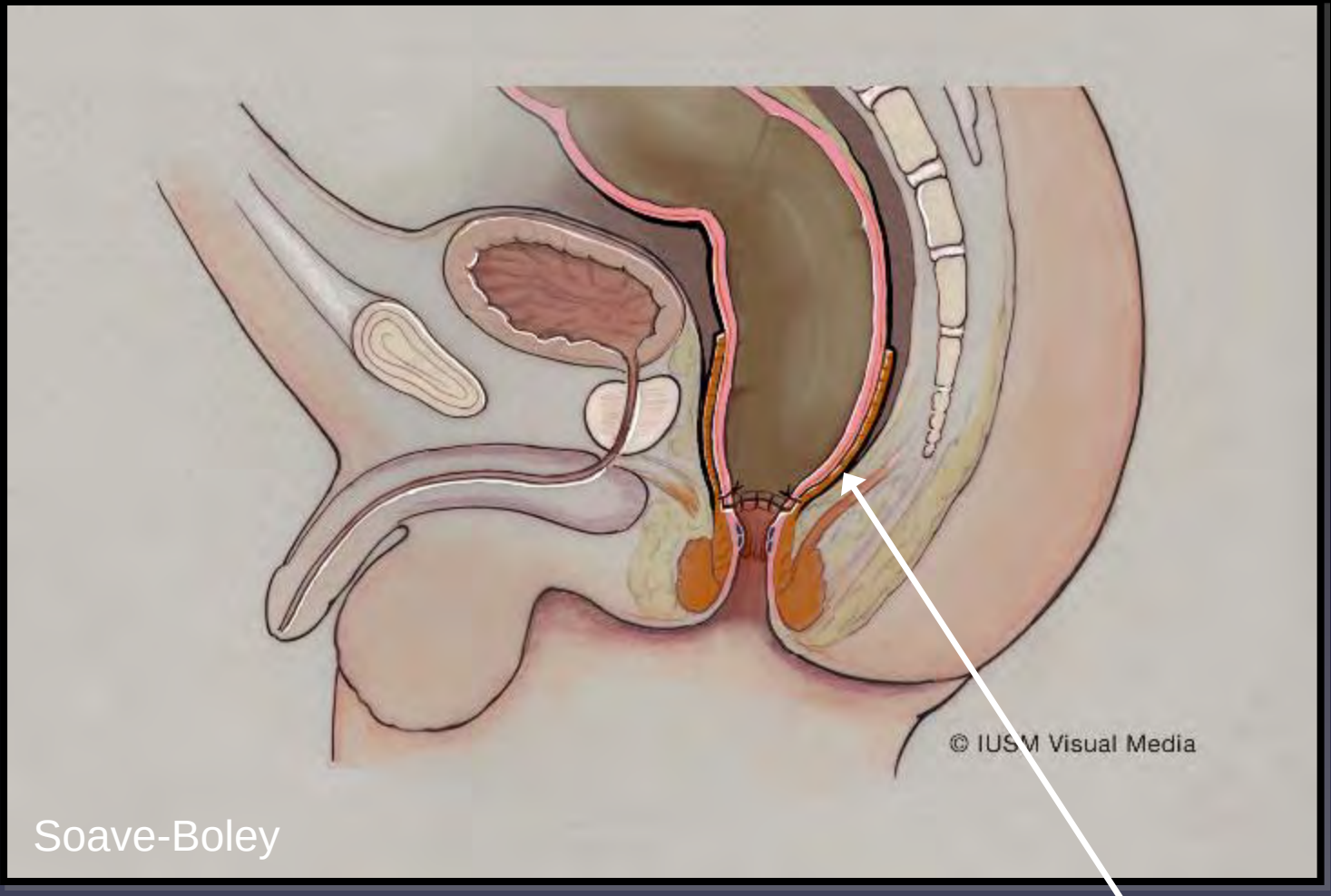


Duhamel



Soave-Boley Endorectal

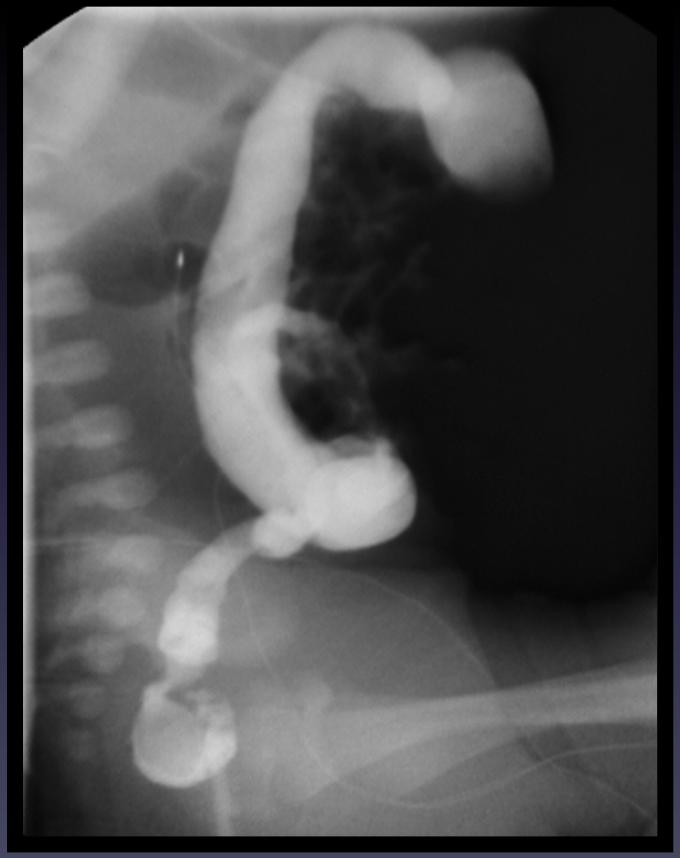




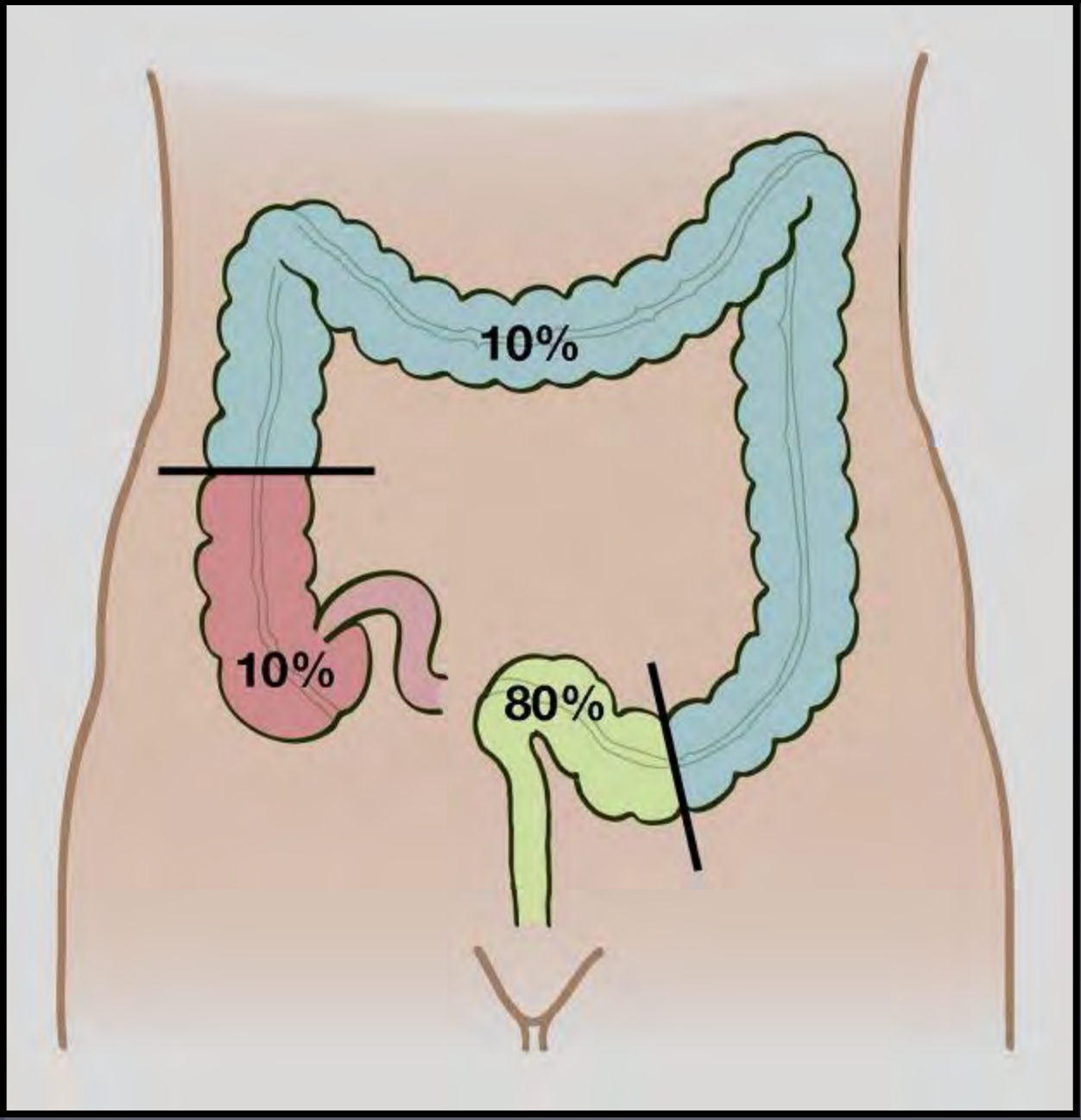
Soave-Boley

Rectal muscle cuff

Hirschsprung's Disease – Primary Perineal Pullthrough



- 8wks of rectal decompressions at home
- Good weight gain
- Resolution of abdominal and proximal colonic dilatation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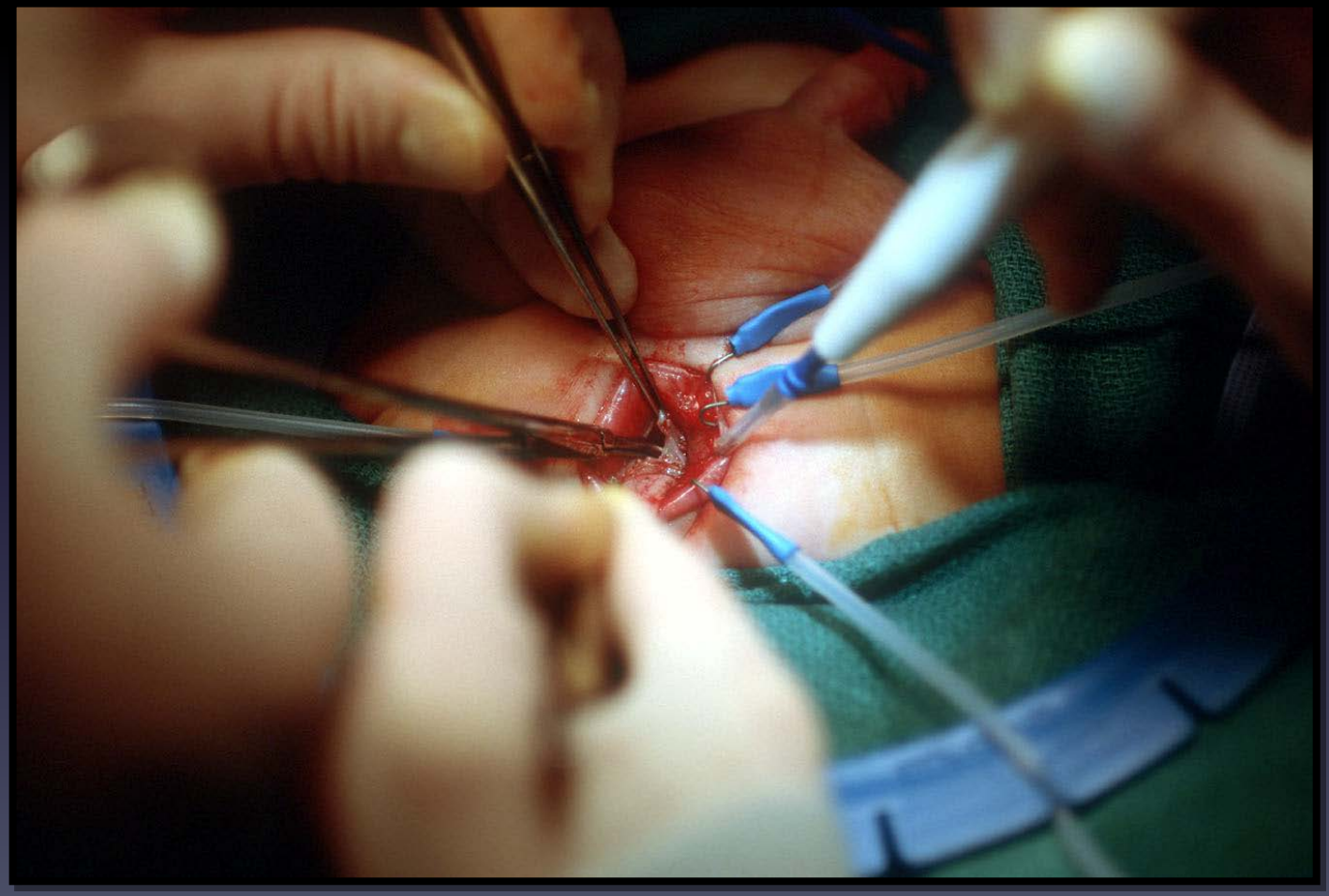
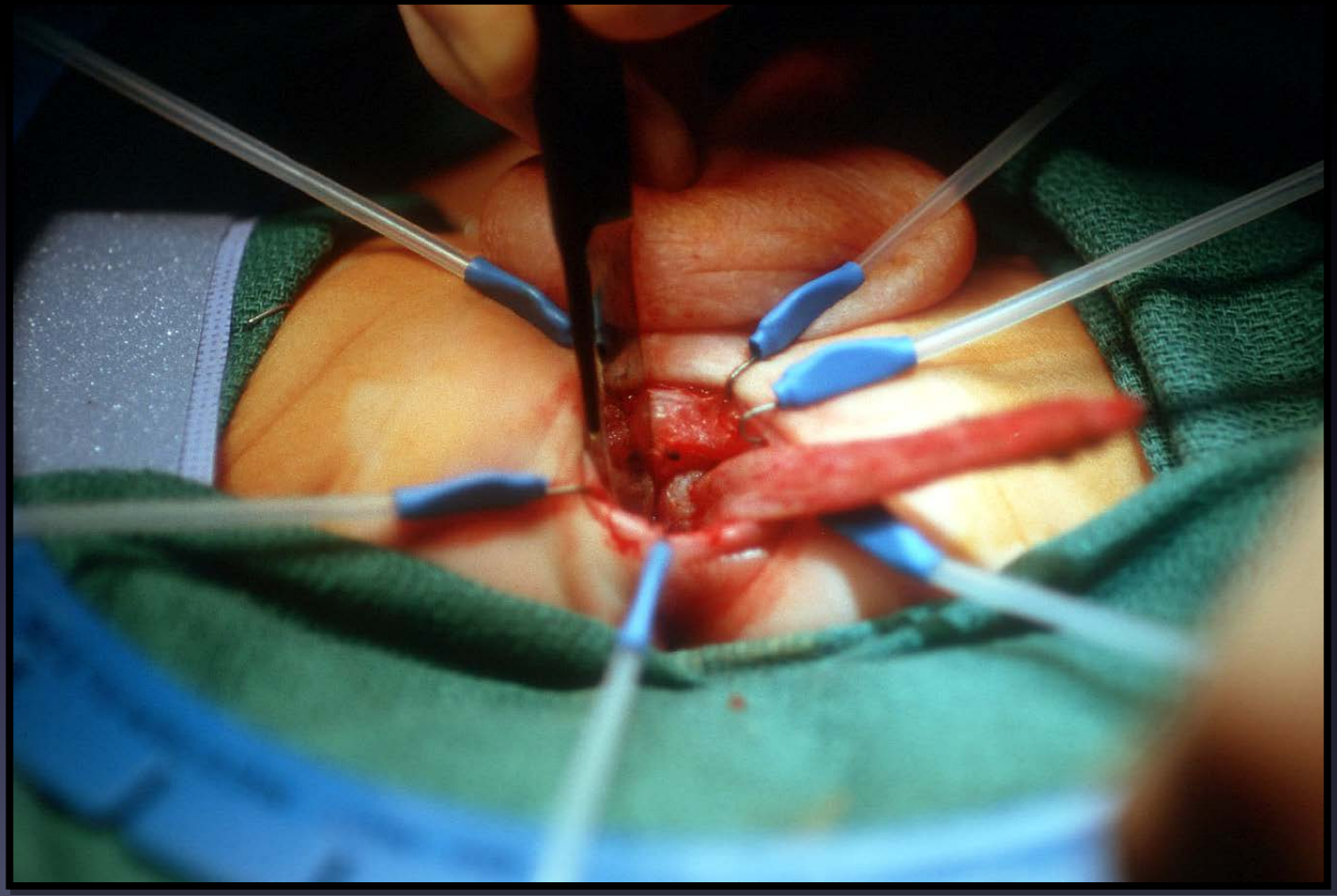
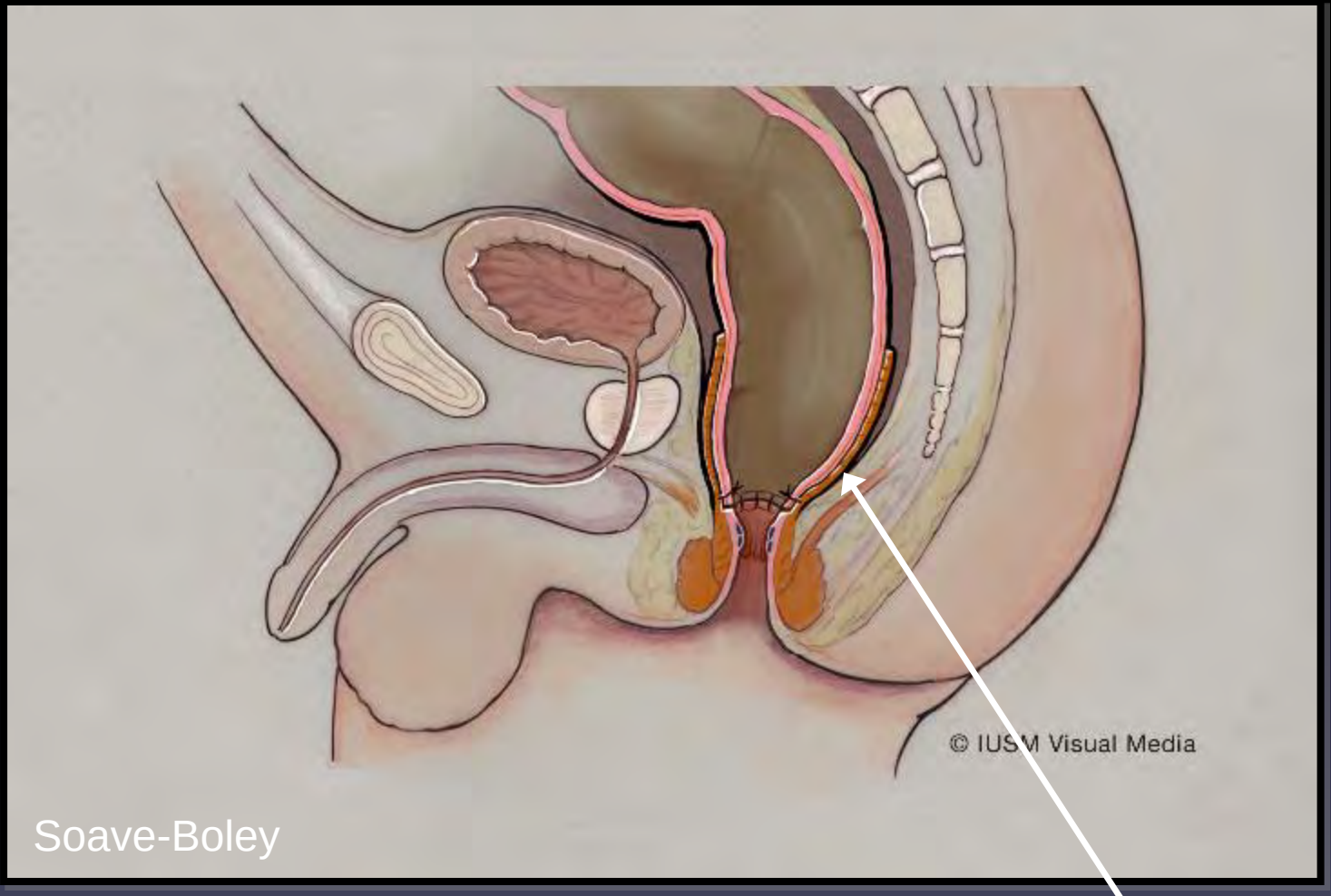
Hirschsprung's Disease – Primary Perineal Pullthrough



Hirschsprung's Disease – Primary Perineal Pullthrough





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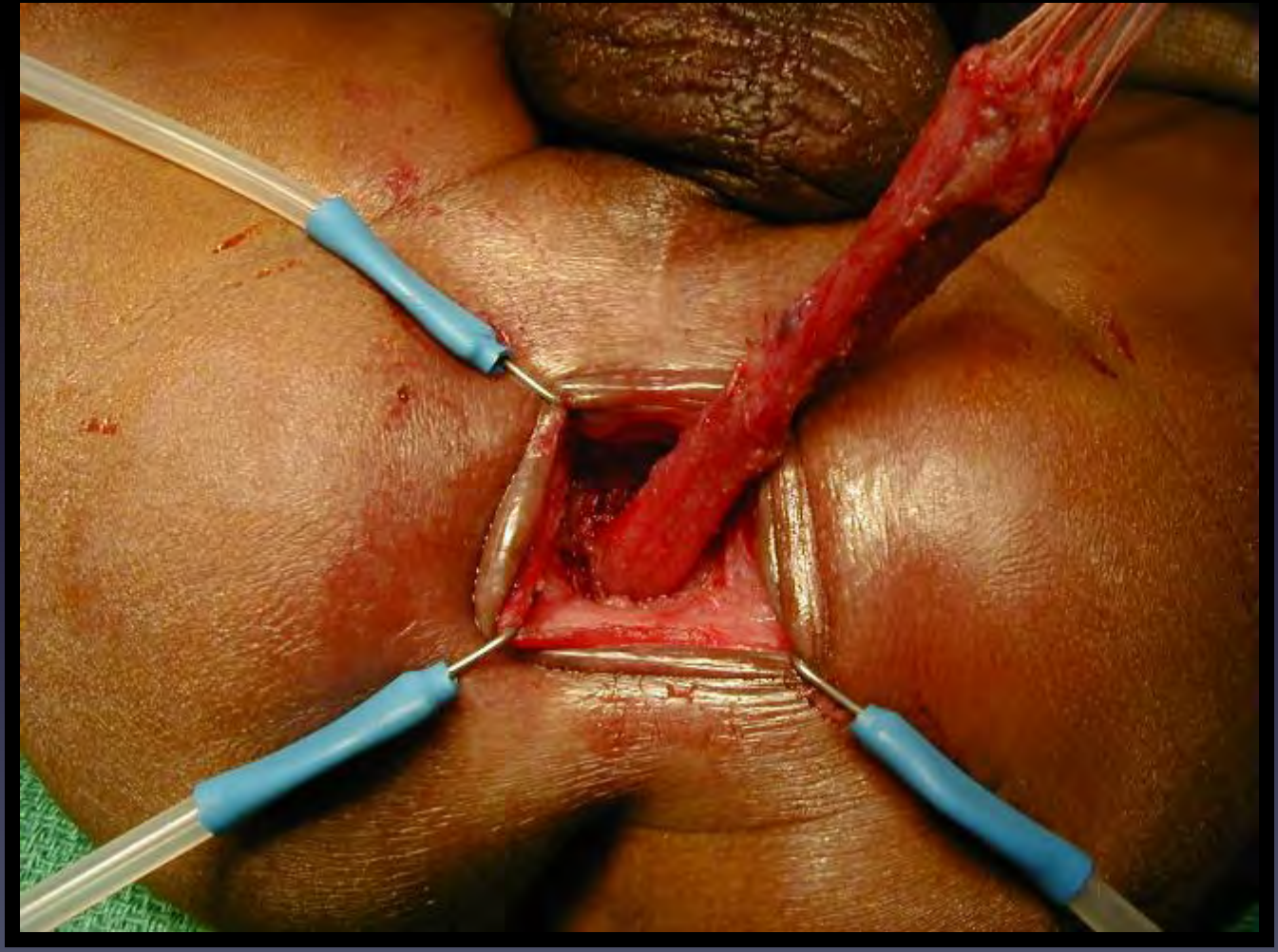
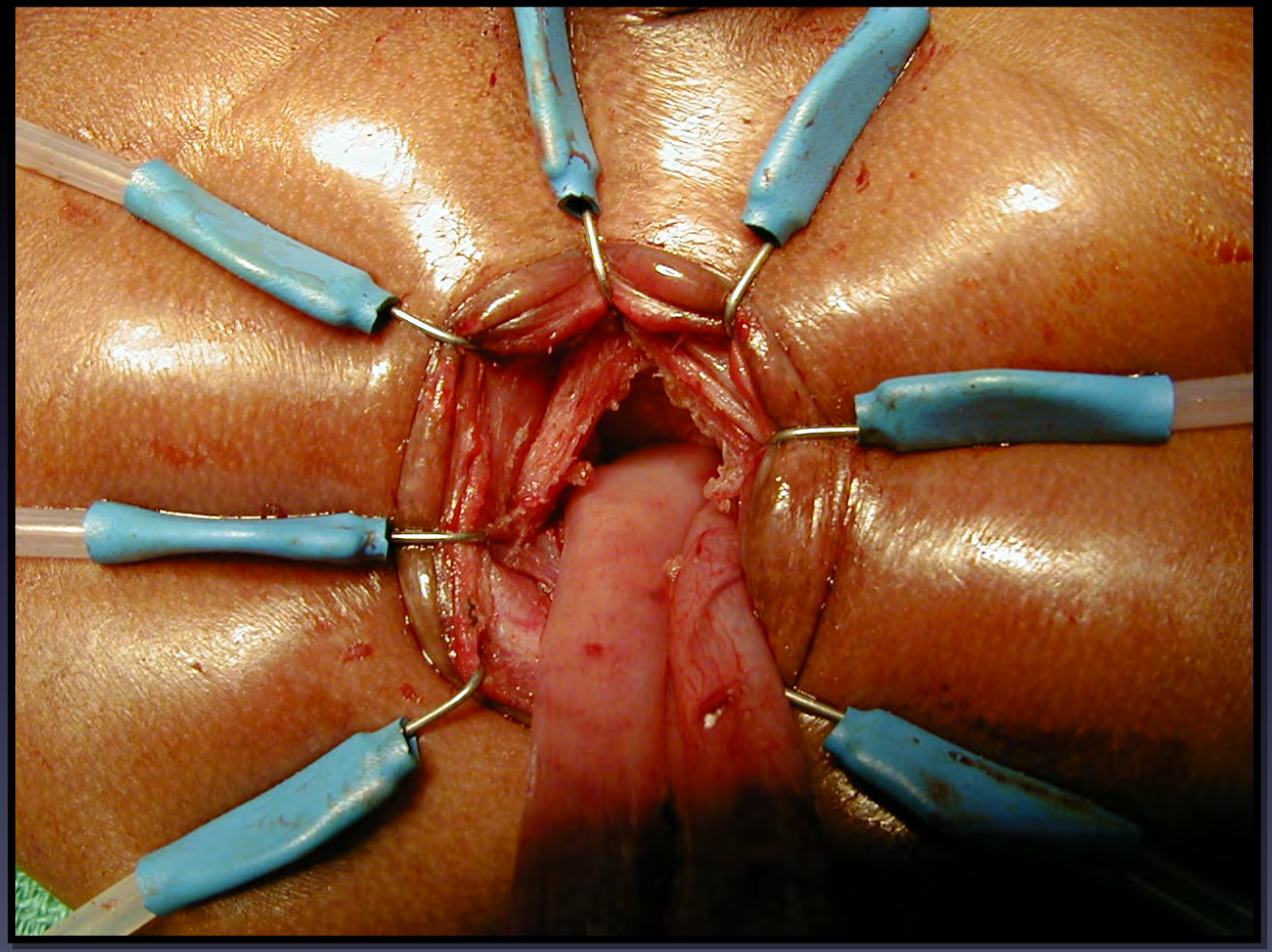
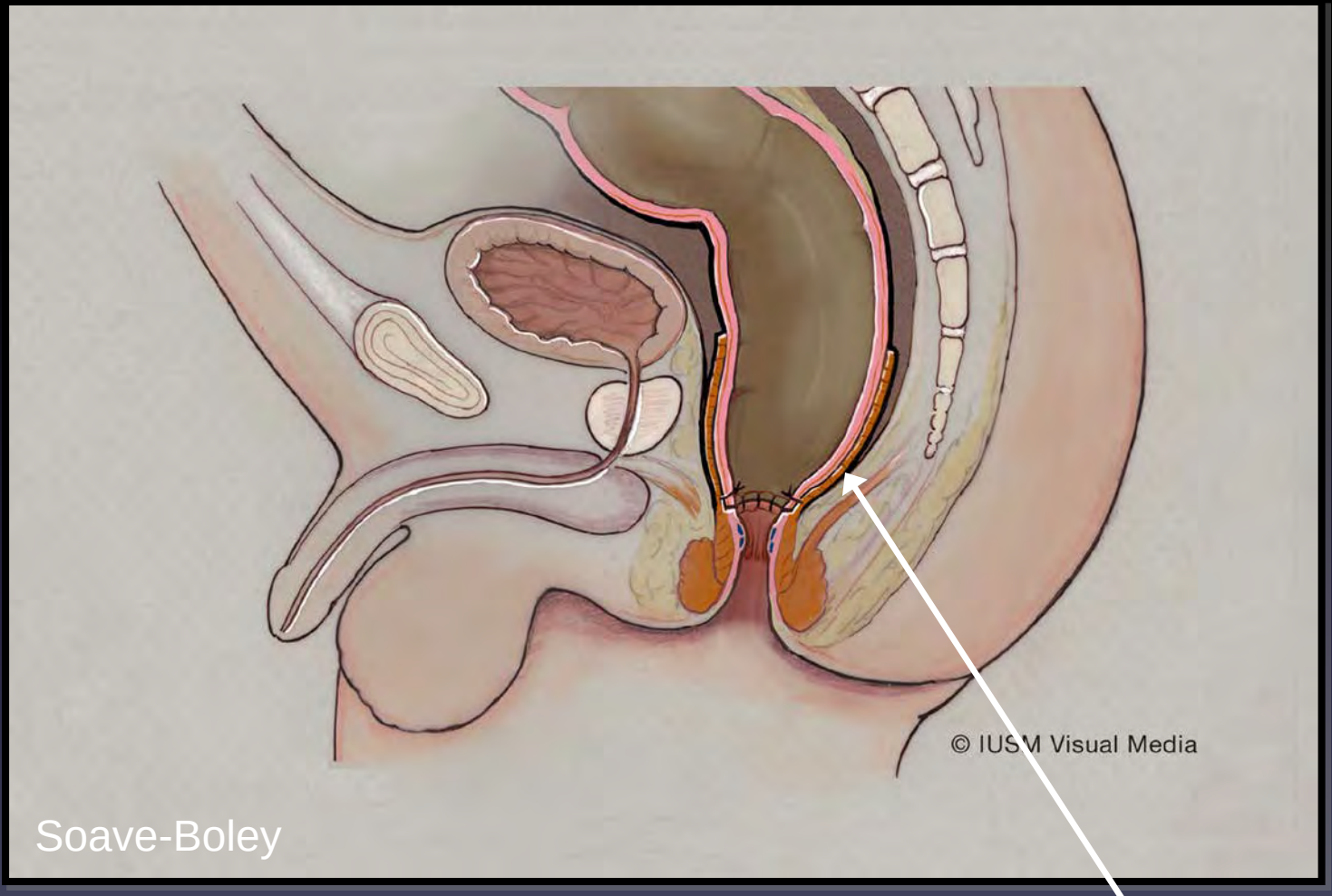


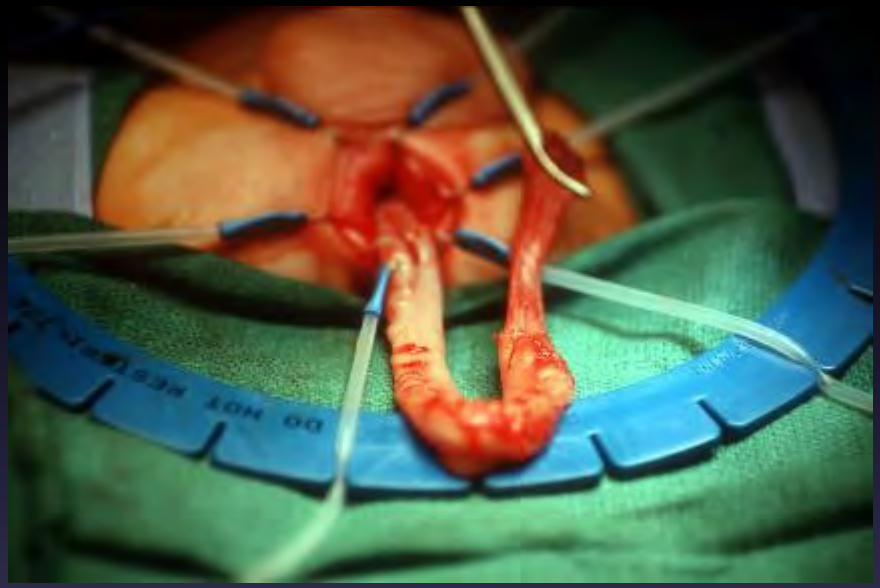
Hirschsprung's Disease – Primary Perineal Pullthrough



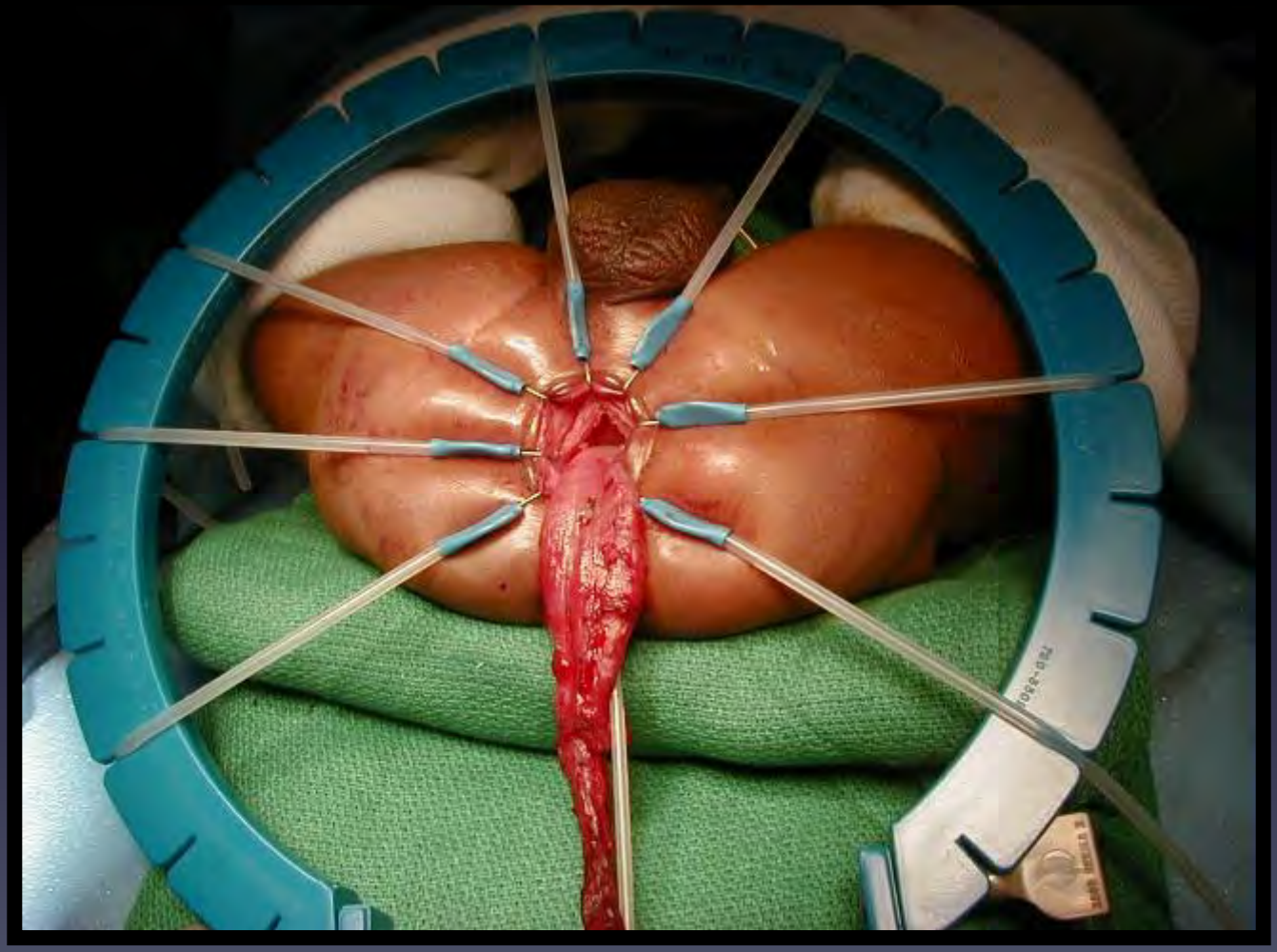


Soave-Boley

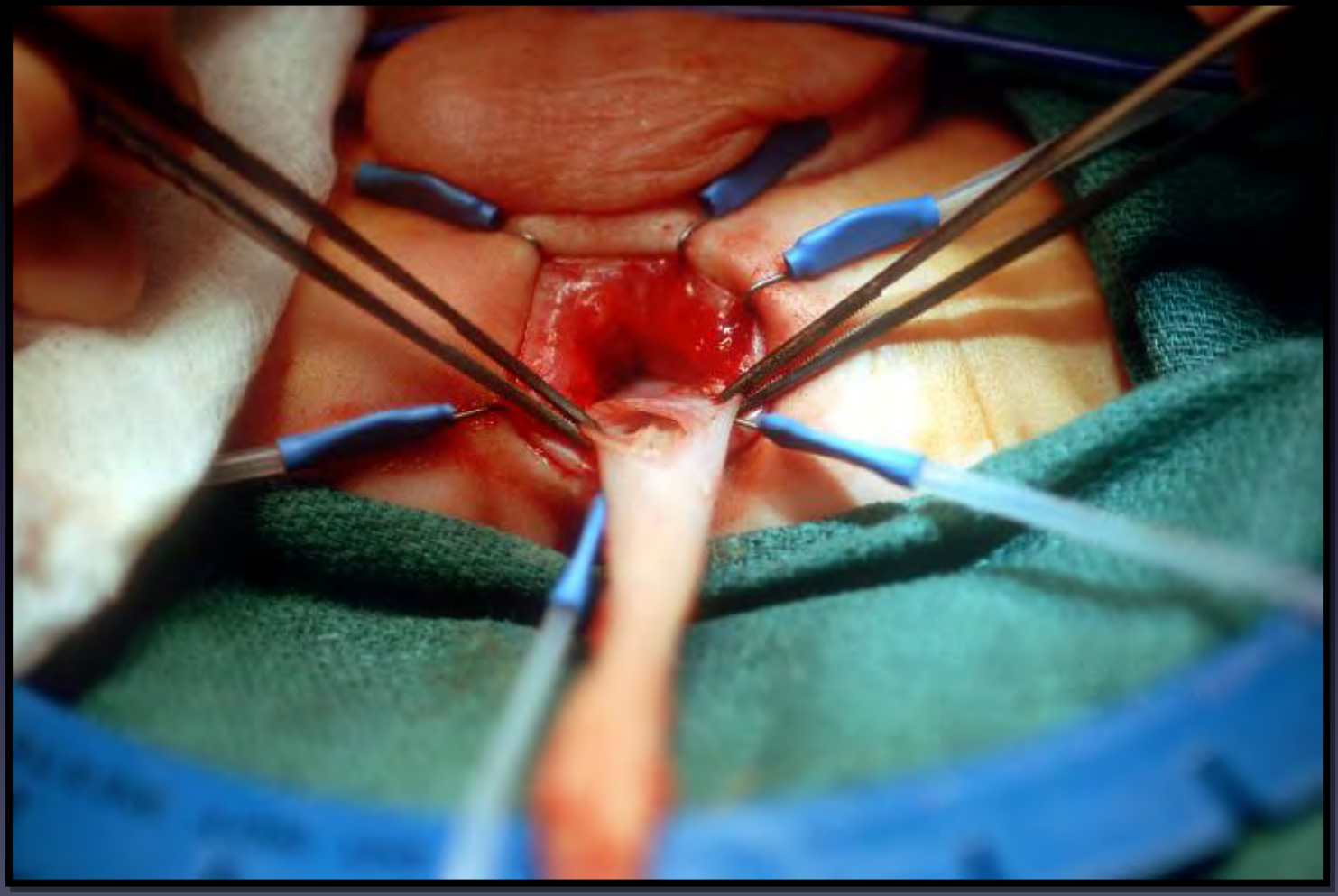
Rectal muscle cuff



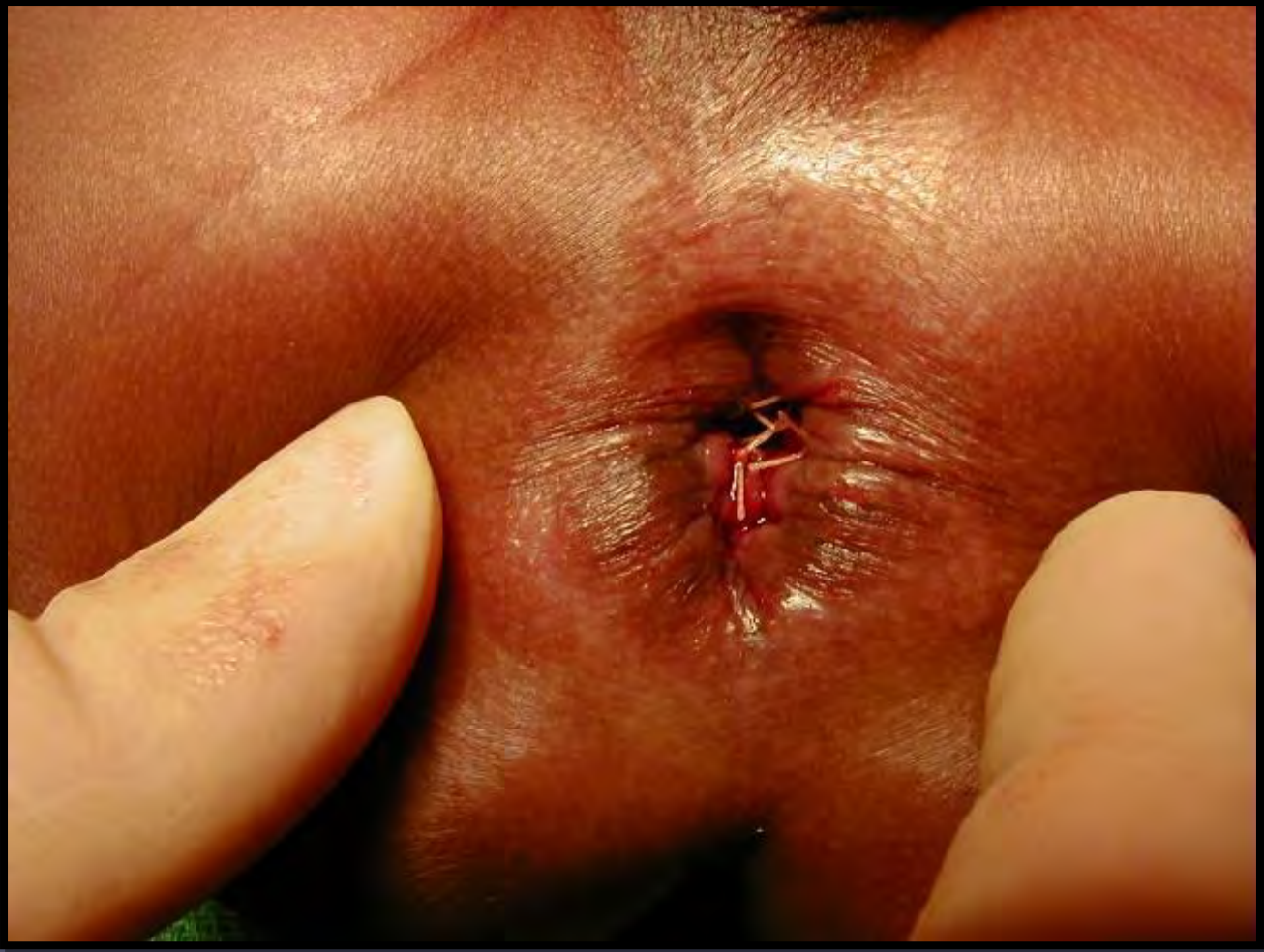
Hirschsprung's Disease – Primary Perineal Pullthrough



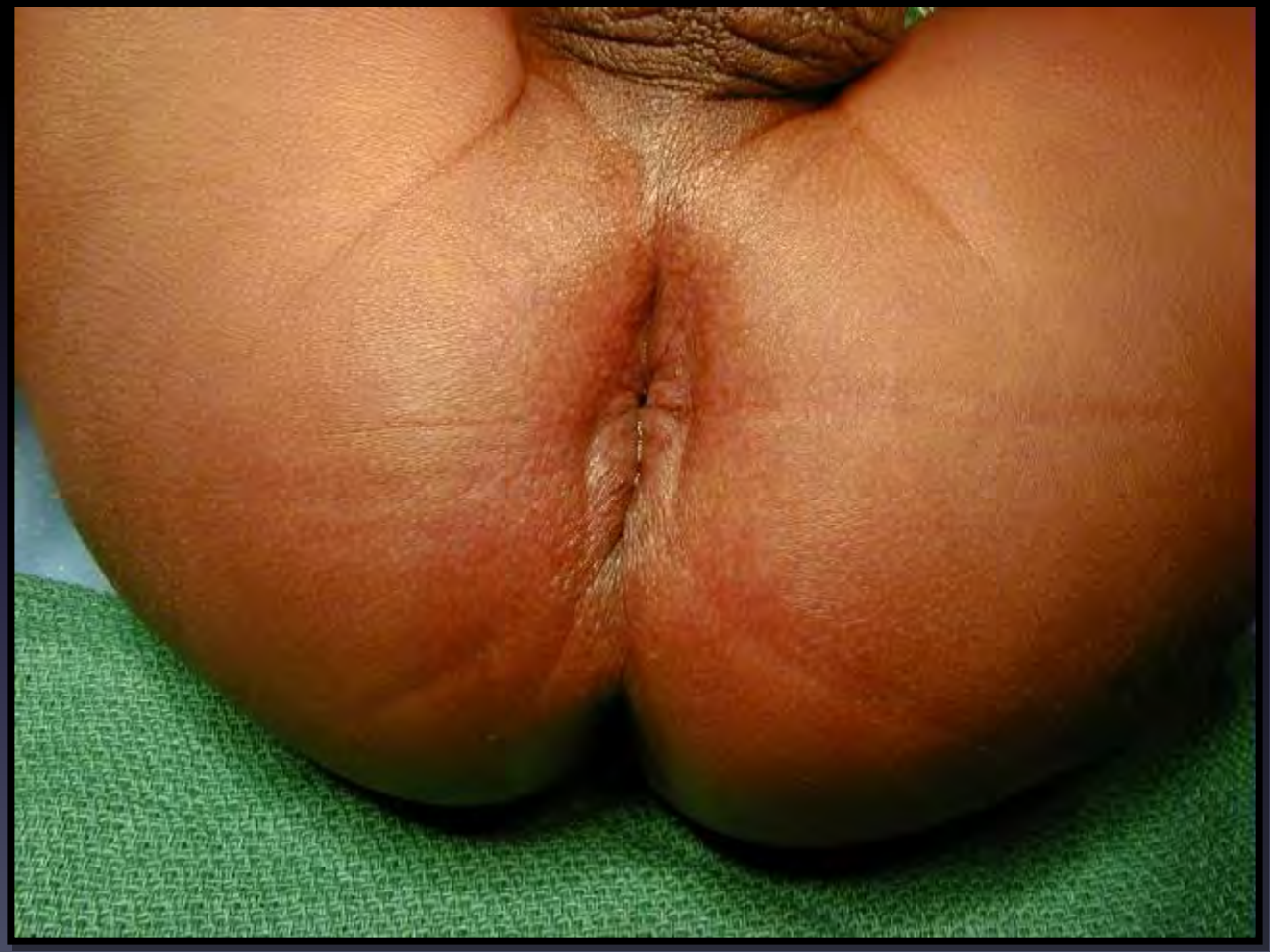
Hirschsprung's Disease – Primary Perineal Pullthrough



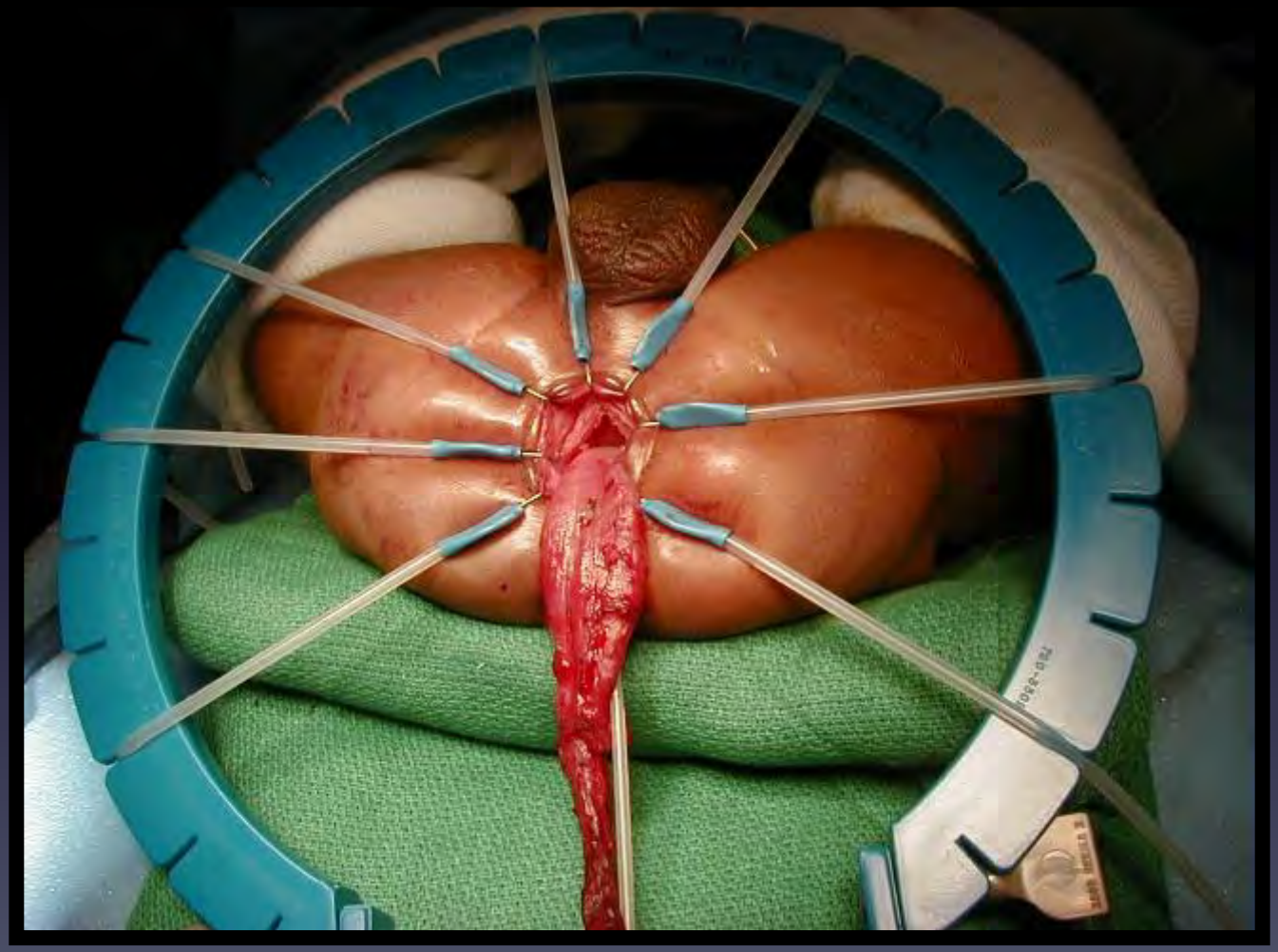
Hirschsprung's Disease – Primary Perineal Pullthrough

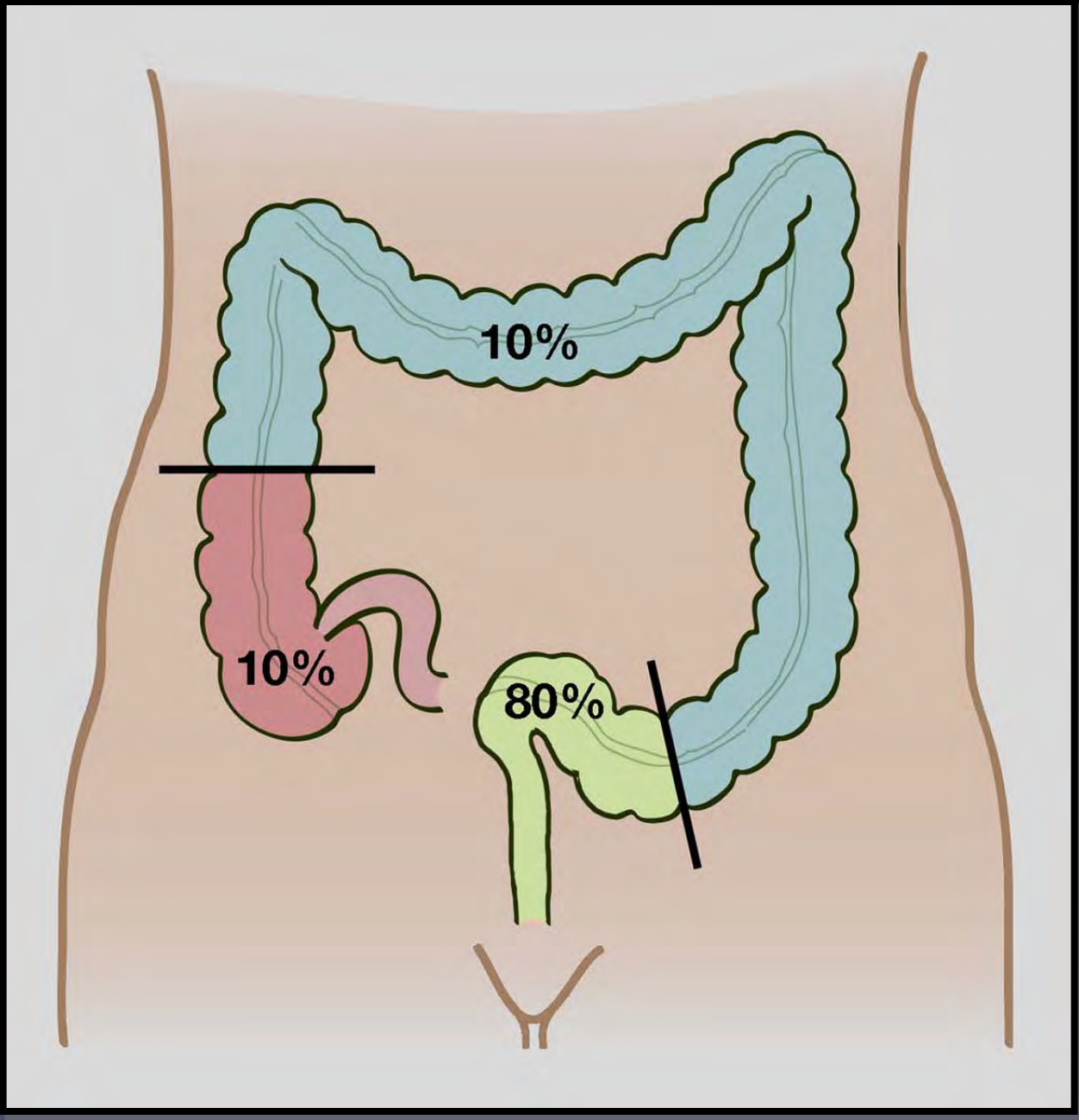


Hirschsprung's Disease – Primary Perineal Pullthrough



If known longer segment or fail to reach ganglion cell: laparoscopy or laparotomy

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- Very dilated proximal bowel – not amenable to primary pullthrough

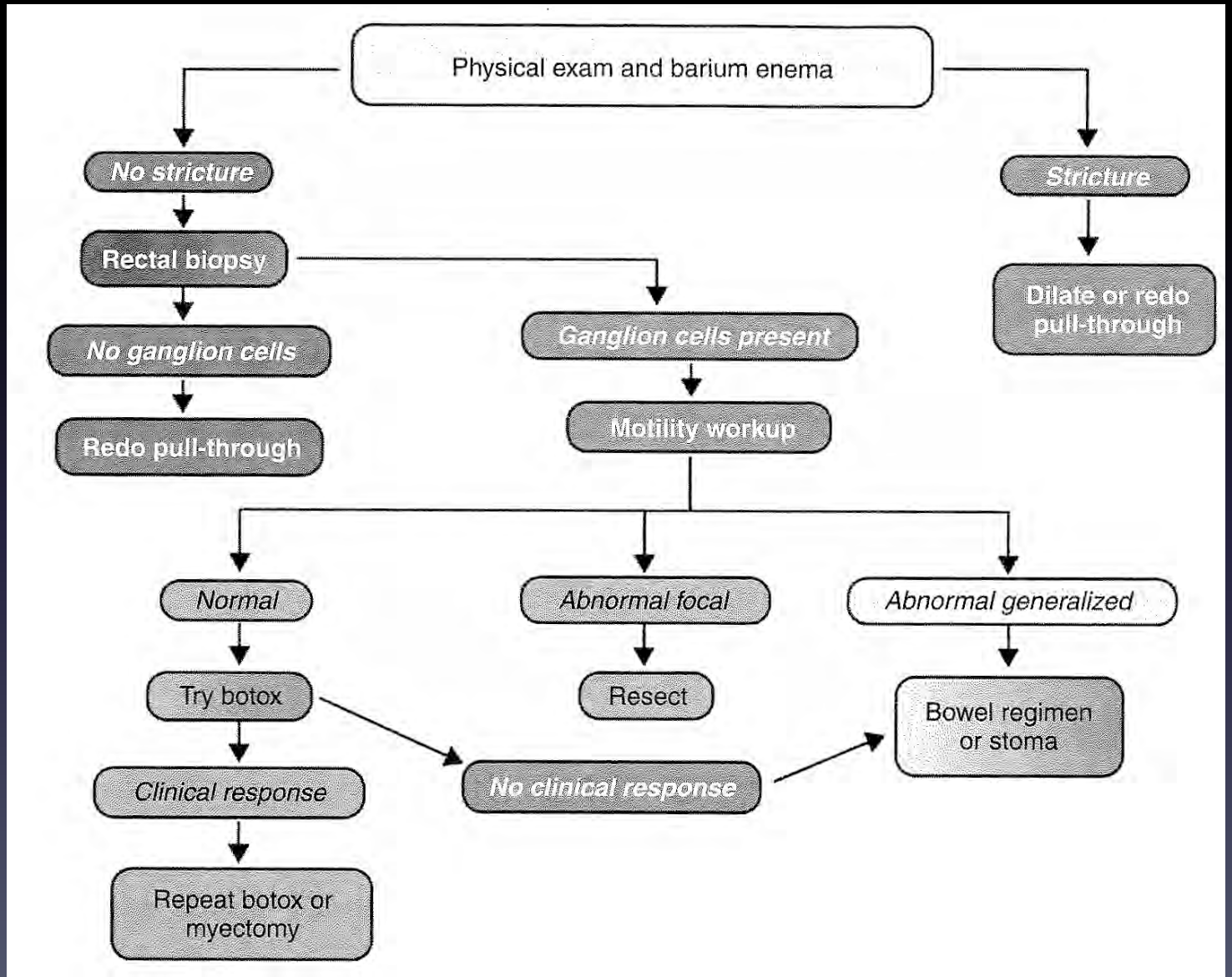
Early Postoperative Care

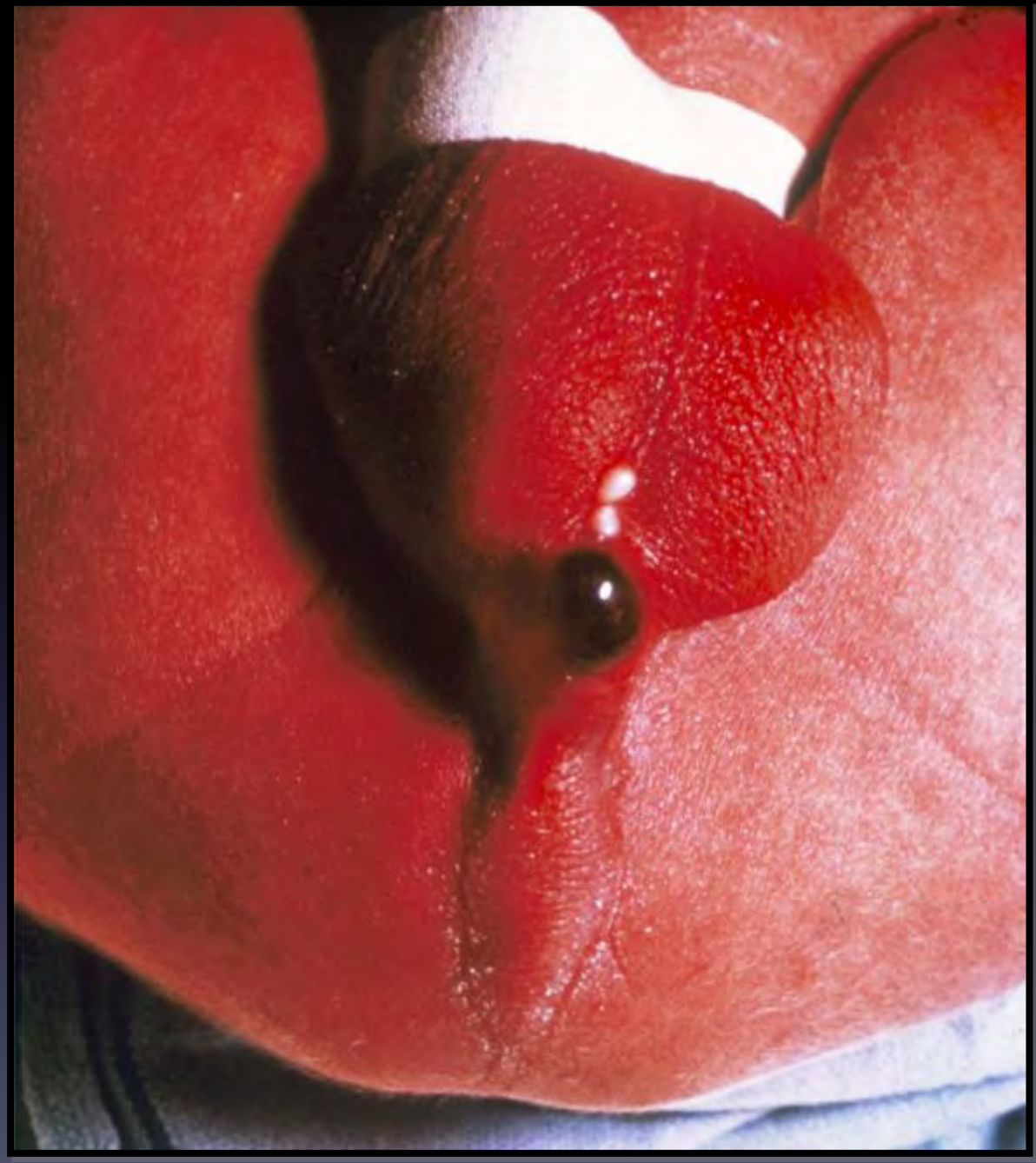
- Avoid stricture – anal dilatation/calibration – see frequently post op
- Internal sphincter achalasia – (They all have it), some obstructive symptoms – most "grow out of it"; can try intrasphincteric botulin toxin

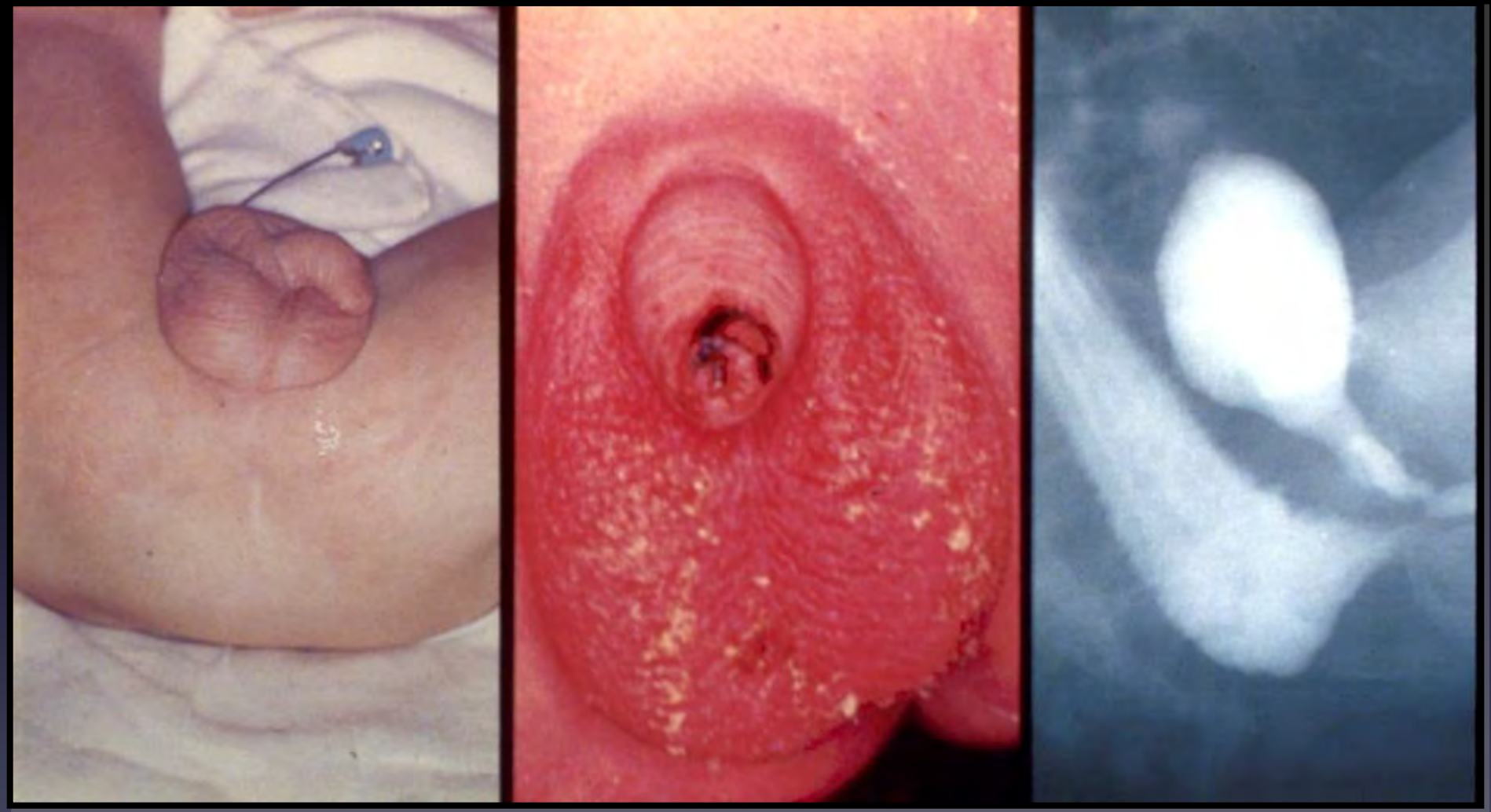
Long-Term Complications

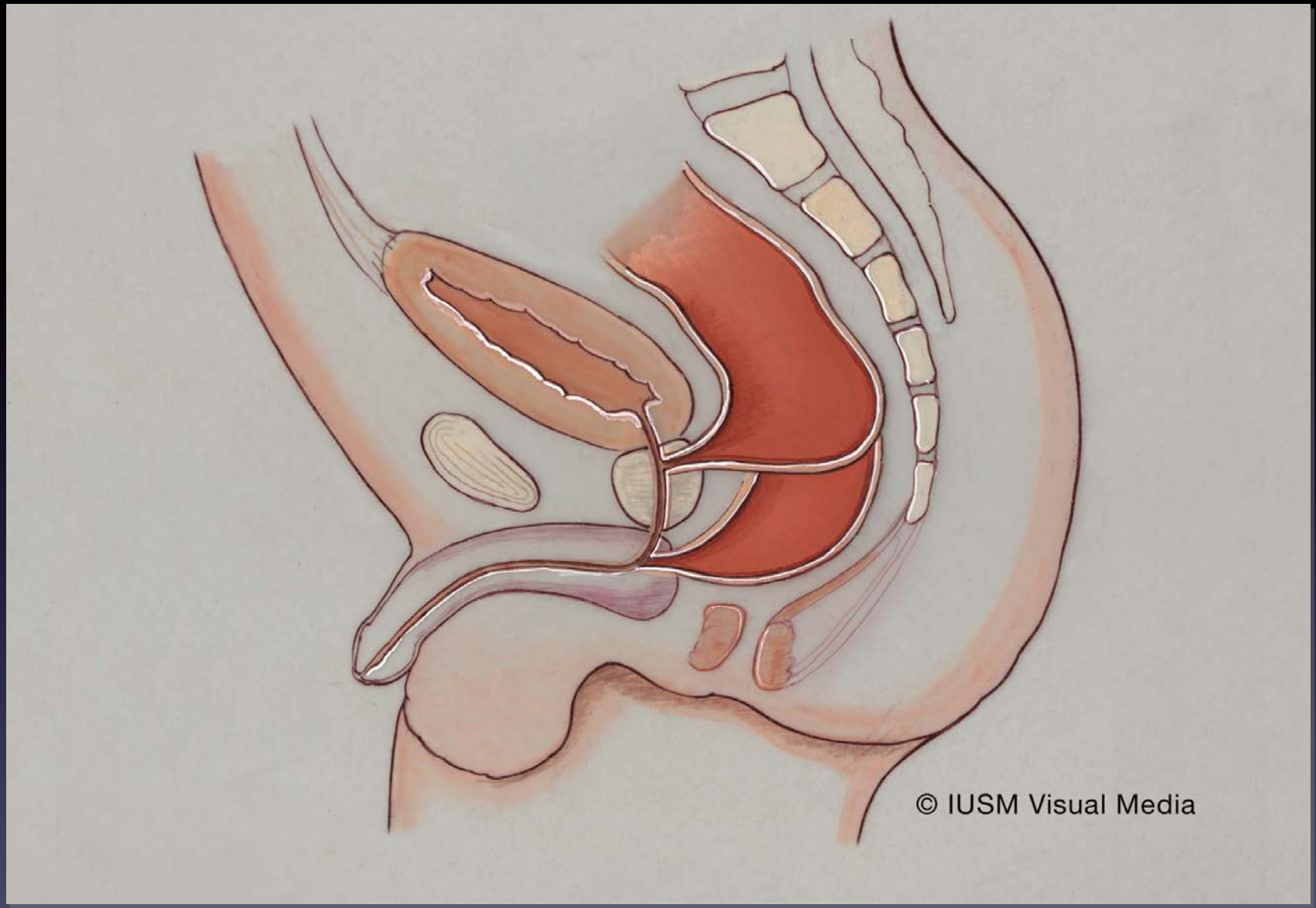
- Most symptoms: obstructive, soiling, enterocolitis – resolve in 1st five years
- Exceptions: Long segment – total colonic – enterocolitis; Down Syndrome – enterocolitis and incontinence

Obstructive Symptoms





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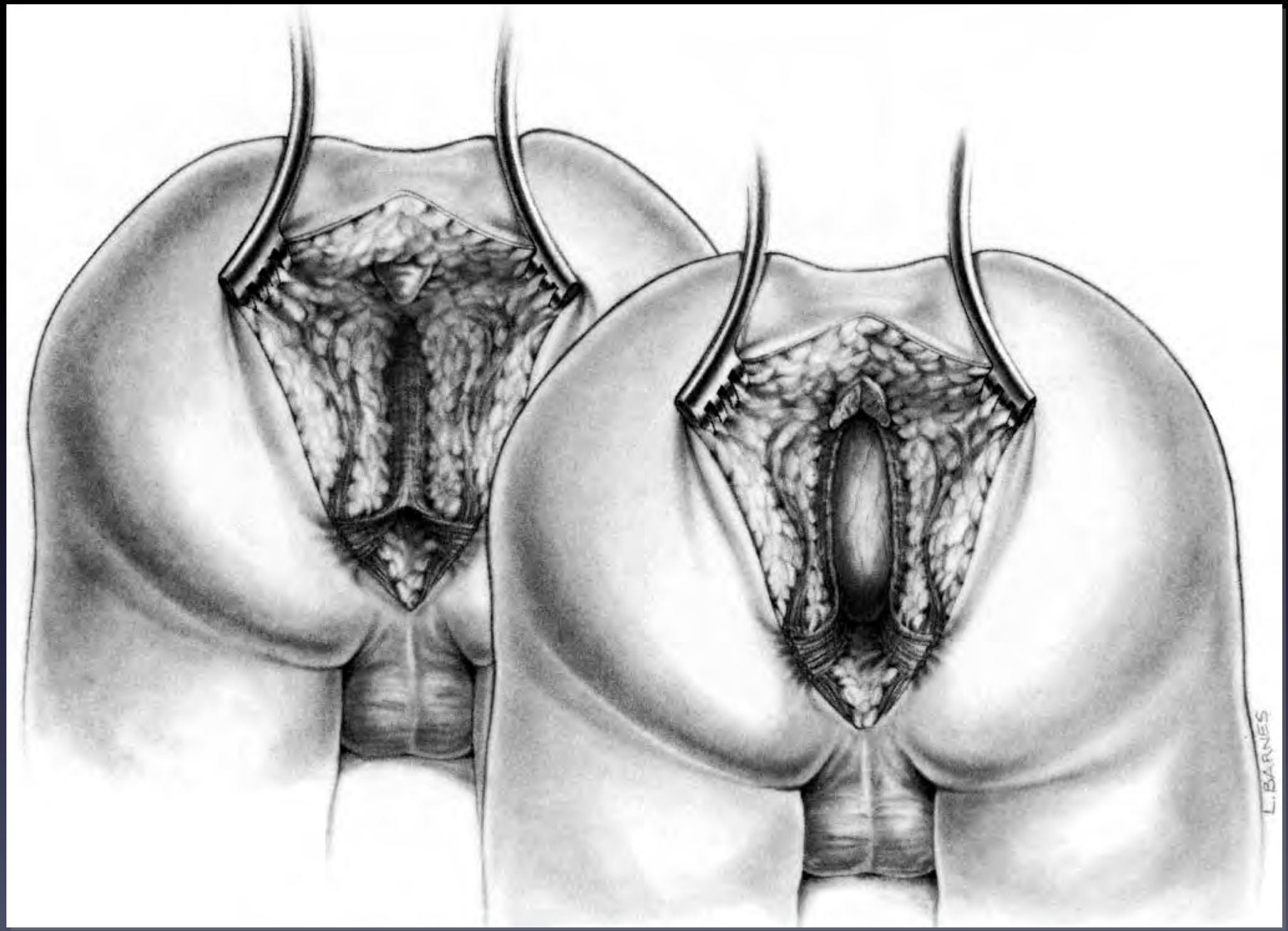
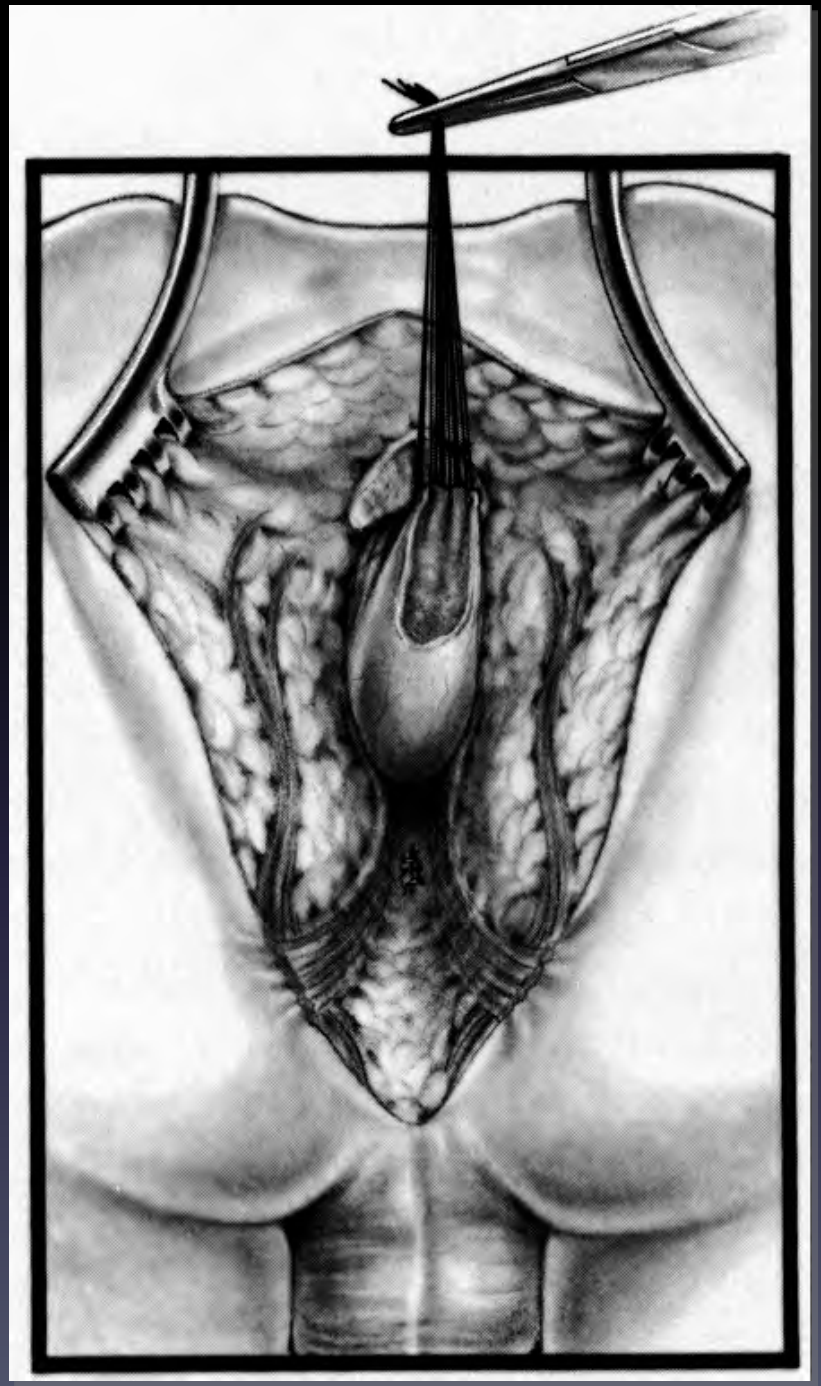
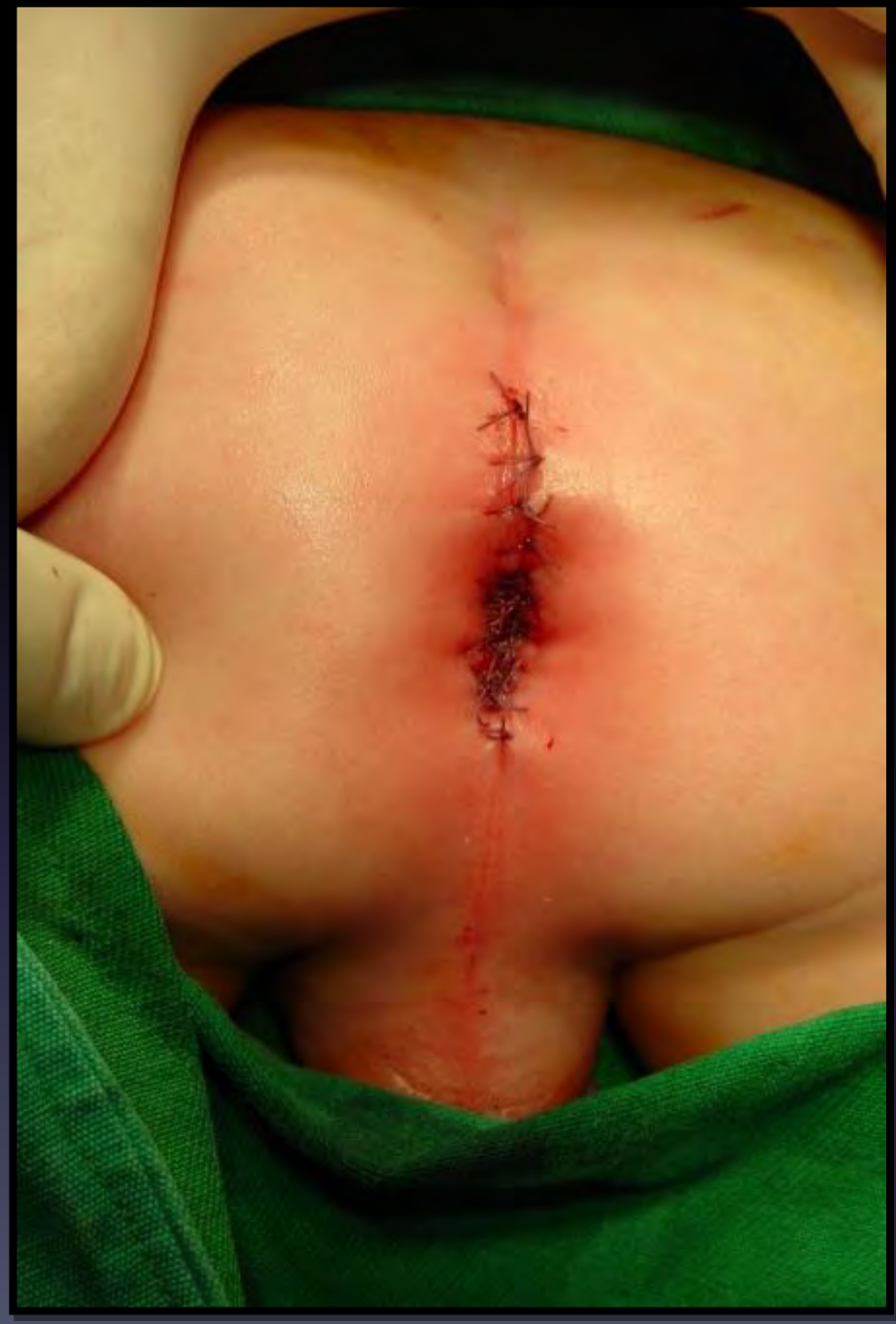

L. BARNES





Social quality of life for adult patients with anorectal malformations

1/3 of high or intermediate anomalies → fecal soiling

Iwai, et al. J Pediatr Surg 2007

Preliminary report: the antegrade continence enema

P. S. MALONE P. G. RANSLEY E. M. KIELY

The principles of antegrade colonic washout and the Mitrofanoff non-refluxing catheterisable channel were combined to produce a continent catheterisable colonic stoma. The intention was that antegrade washouts delivered by this route would produce complete colonic emptying and thereby prevent soiling. The procedure has been successfully carried out in five patients with intractable faecal incontinence.

Lancet 1990; **336**: 1217–18.

Introduction

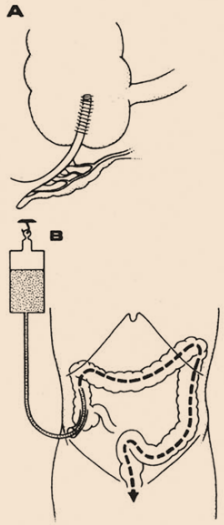
Congenital abnormalities, such as spina bifida, imperforate anus, and Hirschsprung's disease, and abnormalities that follow spinal injury, repeated pelvic surgery, or any operation on the rectum or anus have the potential to produce faecal incontinence. Treatment consists of training to establish a regular bowel habit, control of stool consistency by diet and drugs, and the use of purgatives or enemas to produce regular colonic emptying. Operative measures, such as sphincter reinforcement with muscle slings, are also used but they are of little benefit for incontinence of neuropathic origin. Shandling and Gilmour[1] reported high success rates with an enema continence catheter which administers large-volume saline enemas retrogradely. A small number of patients remain incontinent despite all attempted treatments and opt for a permanent colostomy.

We describe here a new operative technique that facilitates the administration of antegrade washouts to empty the colon and thus prevent soiling—the antegrade continence enema (ACE). The principle of the procedure is that the appendix is reimplanted in a non-refluxing manner into the caecum and the other end is brought out on the abdominal wall as a continent stoma. This procedure provides a catheterisable channel through which antegrade washouts are given to produce colonic emptying (see figure).

Patients and methods

Since October, 1989, ACE has been used in five patients, who have been followed up for 2–8 months (table 1). In four patients (1, 2, 4, 5) the procedure was carried out in conjunction with bladder reconstructive surgery for urinary incontinence. Every effort had been made to control faecal incontinence, including repeat pull-through procedures and pelvic floor surgery for the two patients with anorectal malformation. The new technique was used as the last resort in patients who would otherwise have undergone colostomy formation.

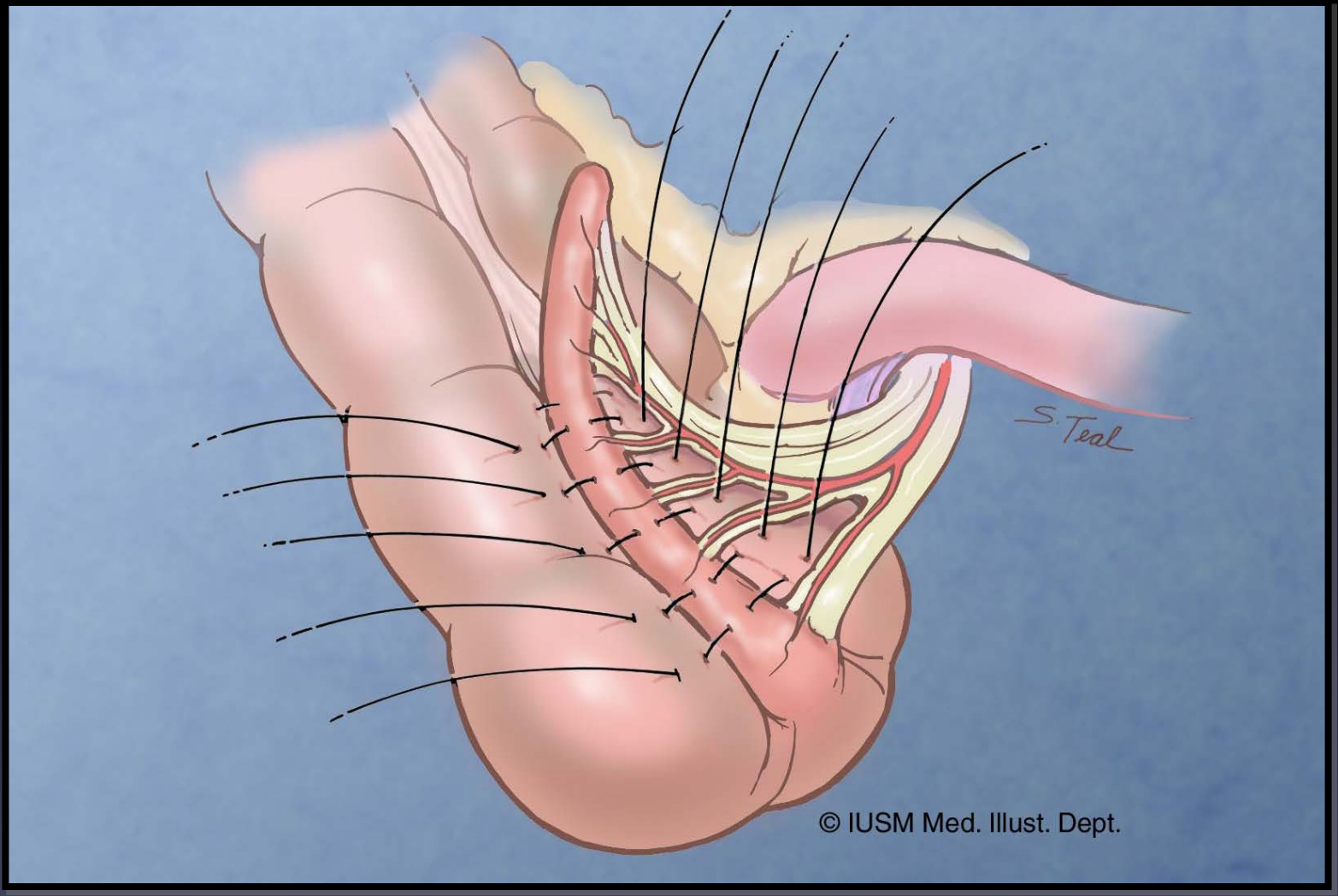
Preoperative bowel preparation and prophylactic antibiotics are recommended. A right iliac fossa gridiron incision is used, but if additional procedures are to be done, a laparotomy is required. The caecum and appendix are delivered from the wound, and the blood supply to the appendix is carefully preserved. The appendix is then resected on its vascular pedicle, with a generous cuff of caecum left at its base. The caecal defect is closed and the distal end of the appendix is amputated. At this stage it is important to check the patency of the appendix lumen by passage of a catheter through it. A 5 cm long submucosal tunnel is fashioned by sharp dissection through one of the taenia on the caecum, and the mucosa of the bowel is opened at its distal end. The appendix is reversed and its distal end is anastomosed to the mucosal opening in the caecum by means of absorbable sutures. The appendix is placed in the submucosal tunnel and the seromuscular layers of the caecum are closed over it, thus creating a non-refluxing channel (figure, A)—the Mitrofanoff principle.[2] The site of the stoma is marked on the abdominal wall before operation and the appendix is now delivered through this site. The caecum is anchored to the anterior abdominal wall, with no kinking of the appendix. The stoma is fashioned by means of a broad-based lateral skin flap that is made into a tube and anastomosed to the cuff of caecum on the base of the appendix. This flap serves to bury the appendix and minimise discharge on the abdominal wall. The resulting non-refluxing continent stoma provides access to the proximal colon by way of a narrow catheterisable channel. A catheter is left in situ for 2–3 weeks to prevent kinking of the channel, but washouts can be started after 10 days without removal of the catheter (figure, B).



Non-refluxing appendicocaecostomy (A) and ACE principle (B).

For washout a bag of irrigation saline is hung above the toilet and is delivered by way of an intravenous giving set. It is also possible to use tap water with one (5 ml) teaspoonful of salt per pint (0·5 l)

ADDRESSES: **Wessex Regional Centre for Paediatric Surgery, Southampton General Hospital** (P. S. Malone, FRCSI); and Departments of Urology (P. G. Ransley, FRCS) **and Paediatric Surgery** (E. M. Kiely, FRCS), **Hospitals for Sick Children, Great Ormond Street, London, UK.** Correspondence to Mr P. S. Malone, Wessex Regional Centre for Paediatric Surgery, General Hospital, Tremona Road, Southampton SO9 4XY, UK.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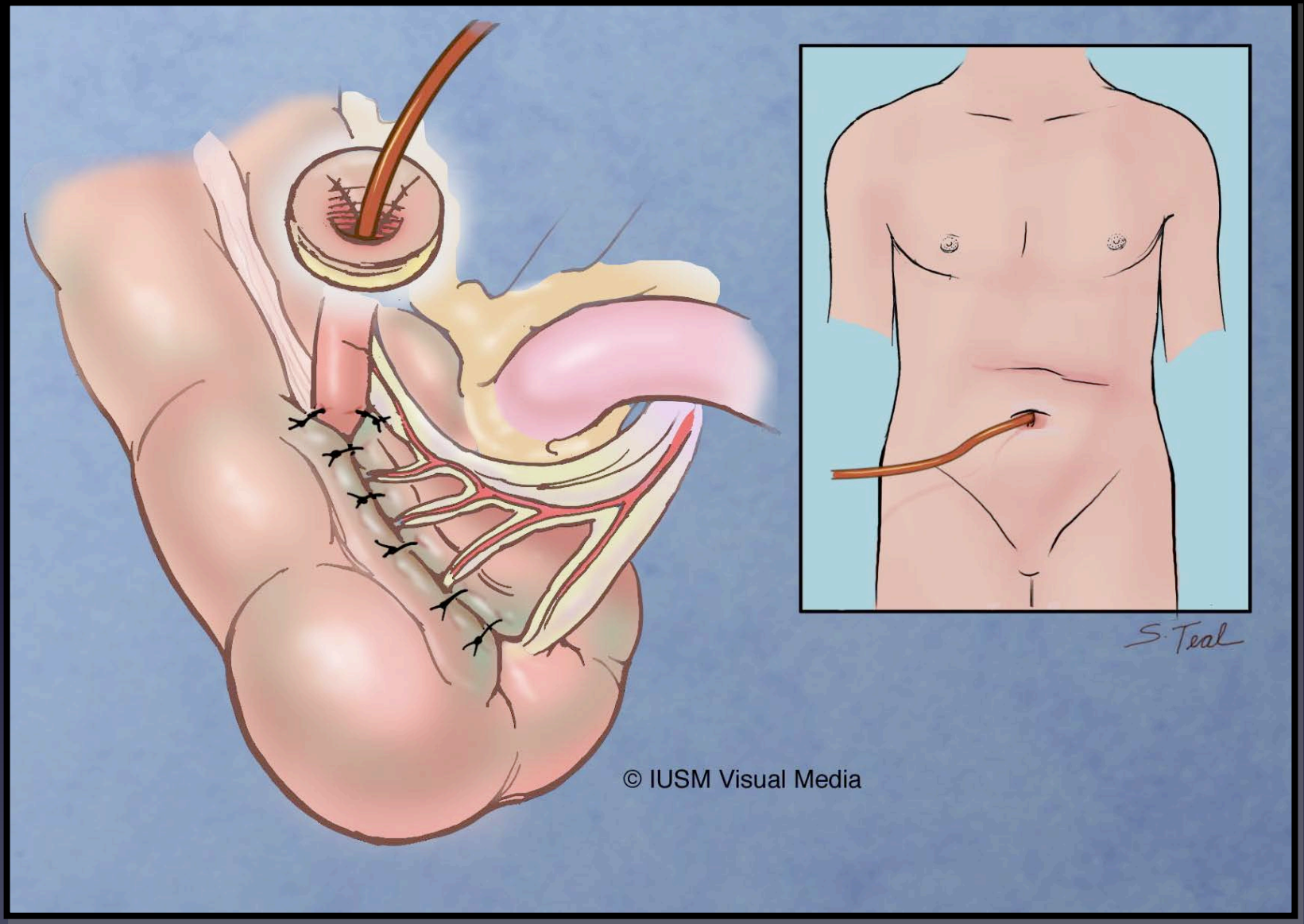
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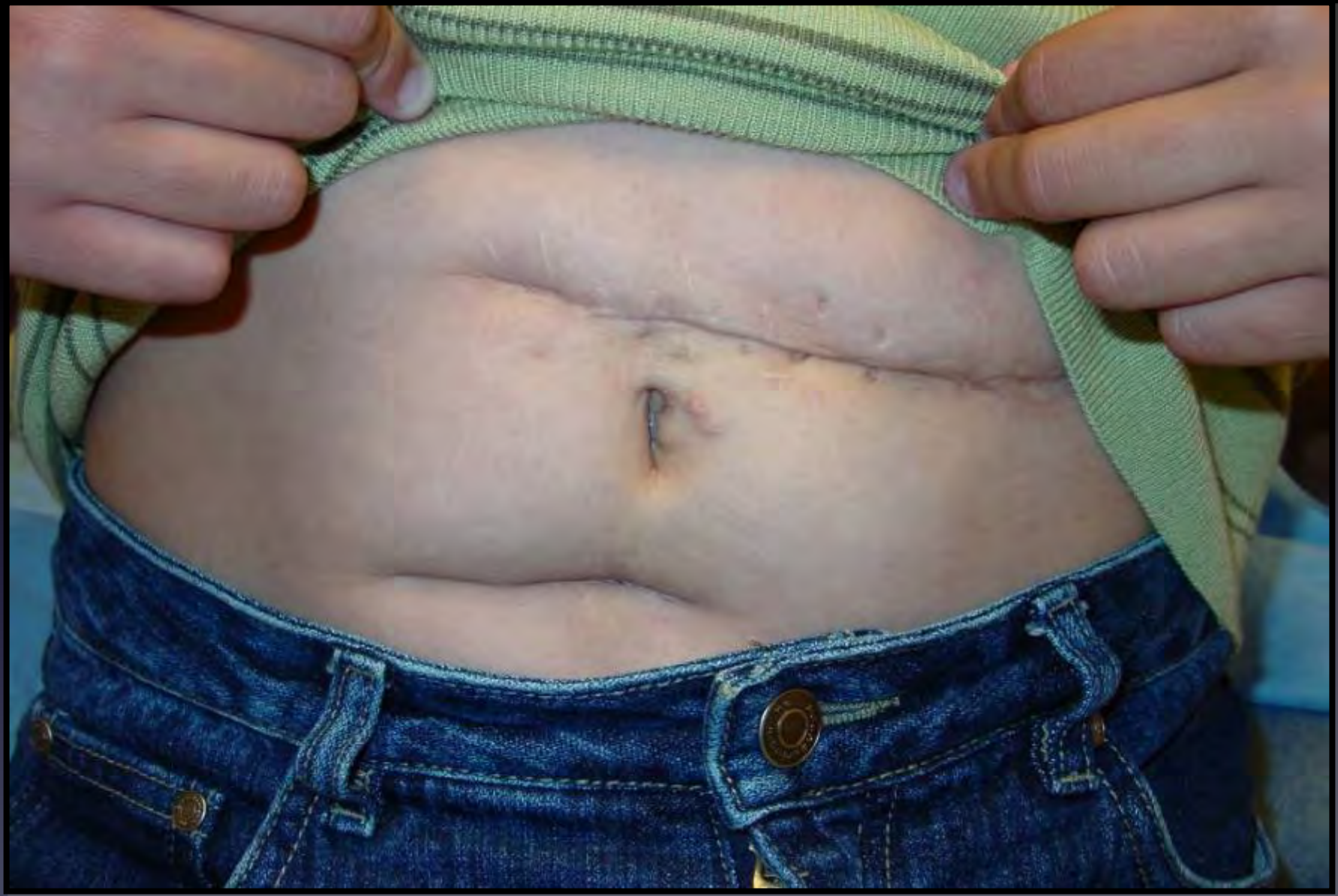

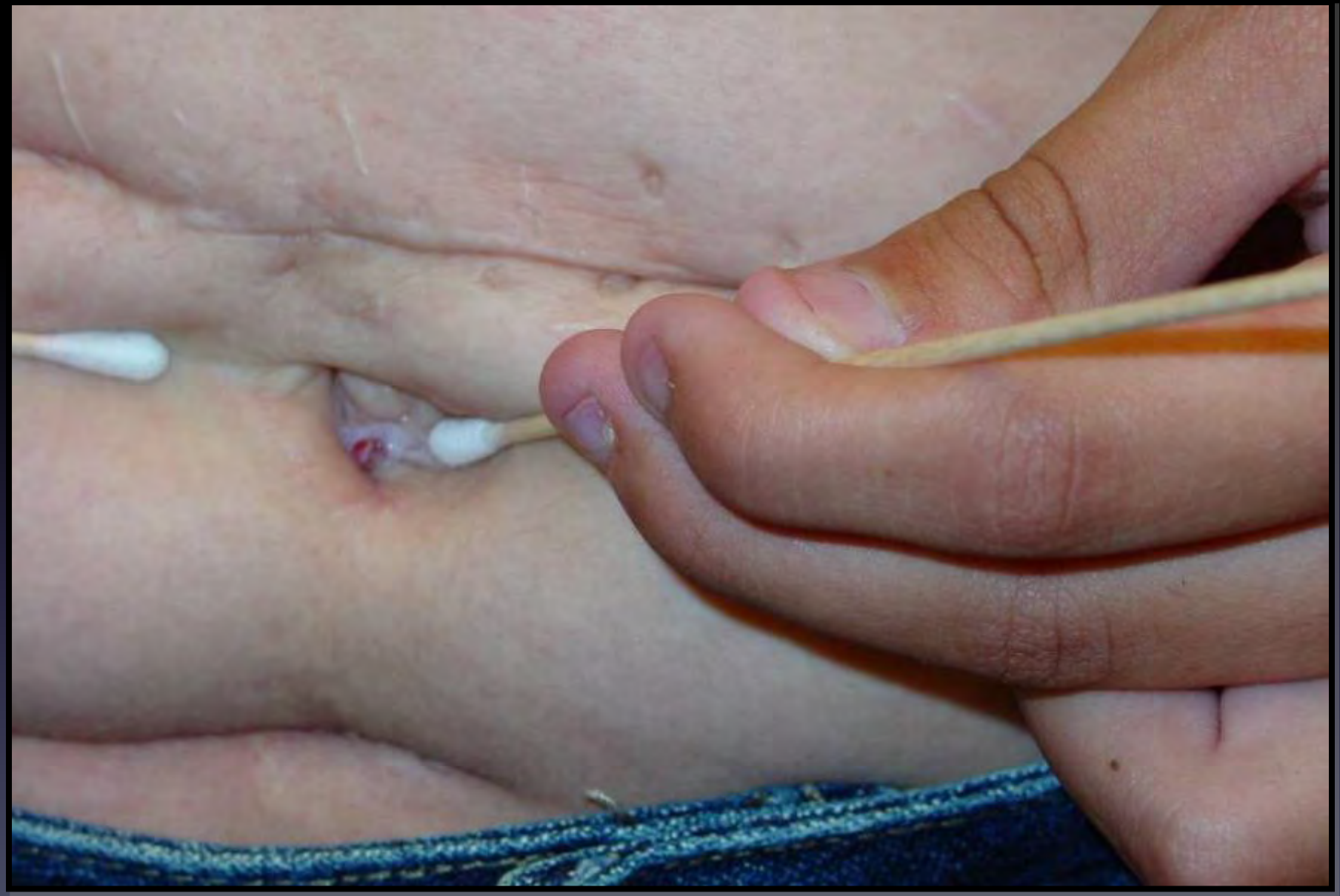

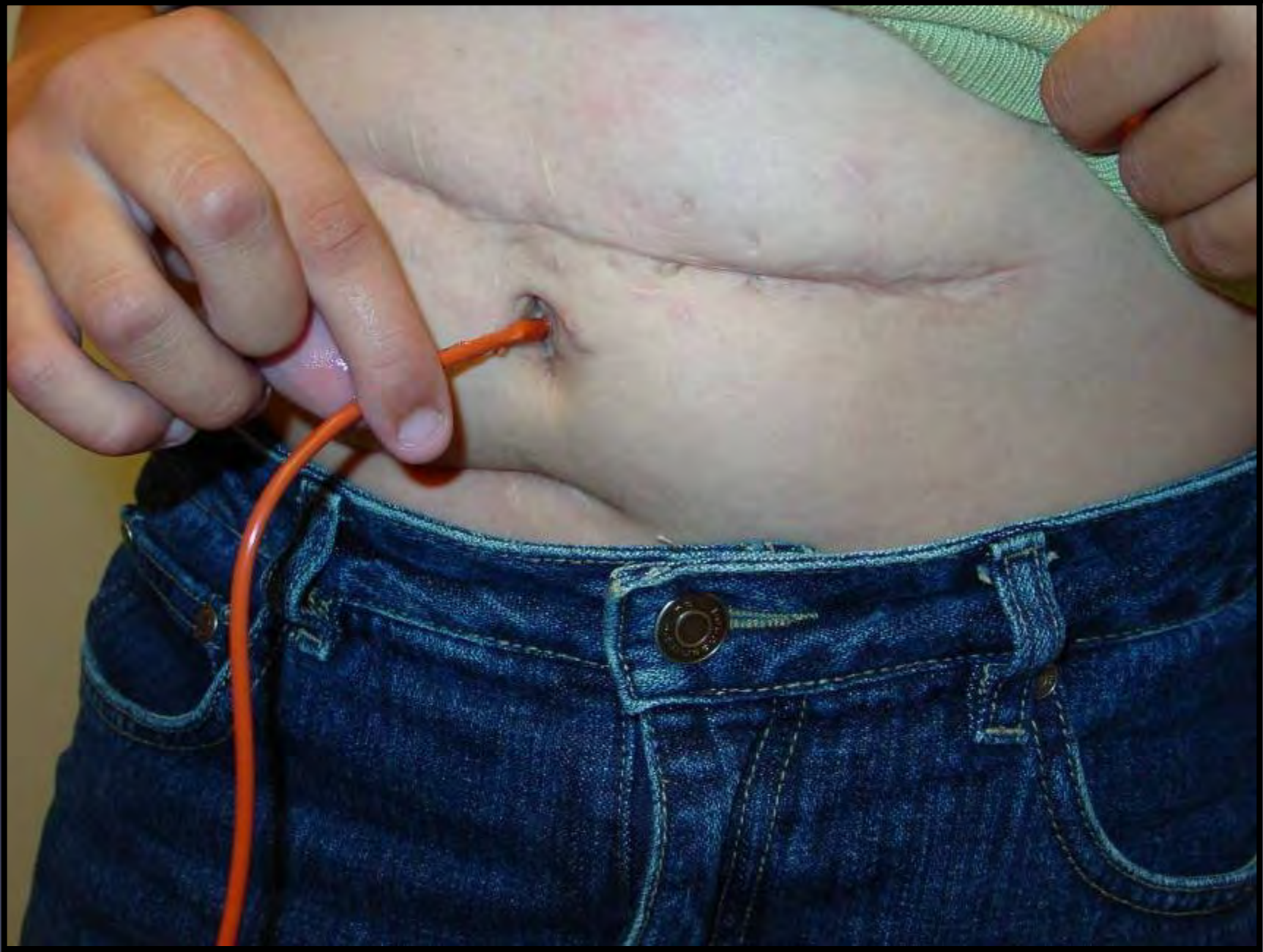

Long-Term Complications

- Most symptoms: obstructive, soiling, enterocolitis – resolve in 1st five years
- Exceptions: Long segment – total colonic – enterocolitis; Down Syndrome – enterocolitis and incontinence
- MACE, rarely as a salvage for intractable constipation and incontinence