



History

Male

3.7 kg

39 weeks

C-section

Mom history: gestational diabetes

Transferred to Children's: Day of life 5

- Delayed passage of meconium
- Bilious emesis
- Abdominal distention



With this history, what would you do?

- A. Abdominal X-ray
- B. Abdominal ultrasound
- C. Upper GI
- D. Contrast enema
- E. CT scan
- F. Nasogastric tube
- G. All of the above





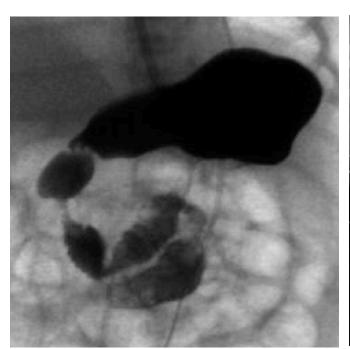


Based on this image, what would your treatment be?

- A. Irrigations
- B. Rectal biopsy
- C. Upper GI
- D. Contrast enema
- E. Option A and B
- F. All of the above



Upper GI Was Performed







Is this UGI normal?



Suction Rectal Biopsy

FINAL DIAGNOSIS:

Suction rectal biopsy, 1 cm and 2 cm
NERVE HYPERTROPHY AND AGANGLIONIC NERVE PLEXUS
ACETYLCHOLINESTERASE STAINING PATTERN IS ABNORMAL.



DAY OF LIFE 13

TRANSANAL ENDORECTAL PULL-THROUGH

"...the dissection was continued proximally until we identified a transition zone based upon the muscular hypertrophy. The biopsy was taken at this point, approximately 15-cm from the dentate line, and the frozen section report returned the diagnosis of ganglion cells present"...

Patient was discharged on Day 3 Post-op.



PATHOLOGY

Rectosigmoid colon, seromuscular biopsy (#1):

GANGLION CELLS PRESENT IN A MYENTERIC PLEXUS.

Colon/rectum, pull through (14 cm) (#2):

HIRSCHSPRUNG DISEASE, 4.0 CM AGANGLIONIC DISTAL SEGMENT PARTIALLY GANGLIONATED TRANSITION ZONE (1.5 CM).

PROXIMAL SEGMENT (8.5 CM) WITH GANGLION CELLS IN A MYENTERIC PLEXUS.



Colorectal and Hirschsprung Center Initial evaluation

- 6 years
- 14 to 20 bowel movements/day first 4 years of life
- 3 to 4 bowel movements day 4 6 years of life
- Most of them are involuntary bowel movements
- MTx Miralax® prescribed by pediatric surgeon
- Never had colitis
- Physicians have affirmed that is because he is lazy

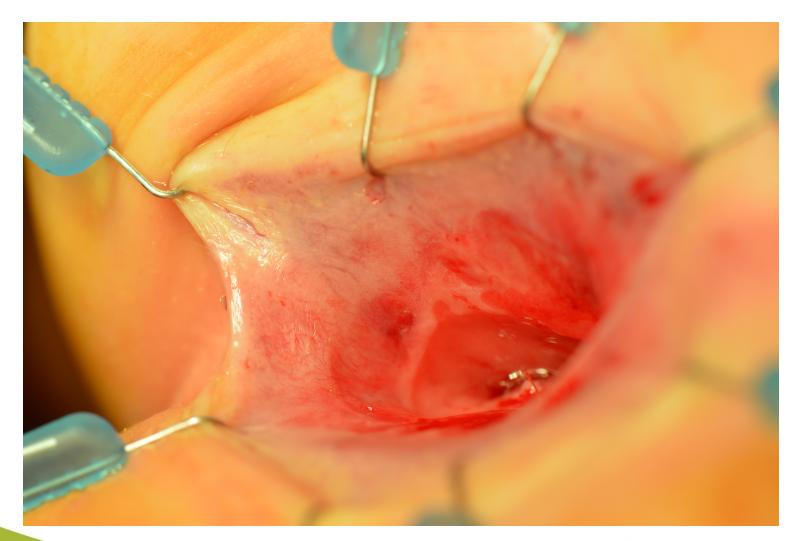


With this information, what would you do?

- A. Abdominal X-ray
- B. Rectal biopsy
- C. Contrast enema
- D. Start laxatives
- E. Enemas
- F. Anorectal examination under anesthesia
- G. Option A, B, C, F



Damage of the anal canal in 70% of its circumference





CE: Pre- and Postevacuation







Damage of the anal canal in 70% of its circumference

We placed a LoneStar retractor with 8 hooks in a symmetrical way. We were able to identify the anal canal.

- 12 to 3 o'clock-wise sewn The anastomosis was done in the anoderm and over the pectinate line
- 3 and 6, anastomosis was done in the anoderm
- 6 and 9, anastomosis was done in the anoderm
- 9 and 12, anastomosis was done in the anoderm and pectinate line.

Then, we obtained a full-thickness rectal biopsy from the lateral wall.



With this information, what would your management be?

- A. Laxative trial
- **B.** Imodium
- C.Redo pull through
- D.Colostomy
- E.Appendicostomy



Successful Bowel Management

- Family attended Bowel Management Workshop
- Colorectal enema 150-10-0-0 (4pm)
- Loperamide 8mg (Q12 am dosing)
- Lomotil 10mg (Q12 pm dosing)
- He was "accidents free for few weeks"



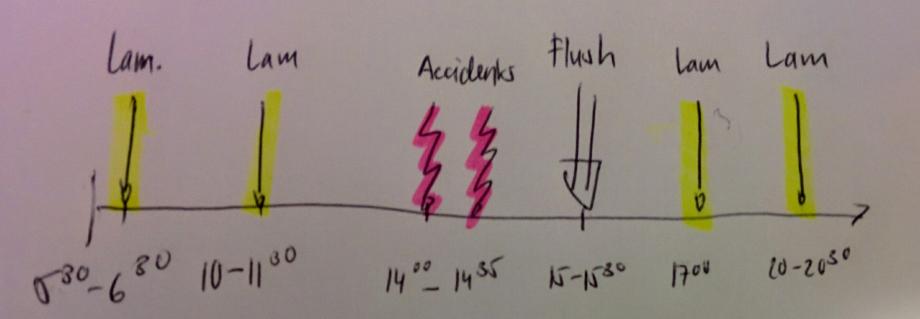
Successful Bowel Management

Because mom noticed better response with Lomotil

- Lomotil 5tabs Q6 (50mg)
- Enema: 150-10-0
- "Miserable" diet: rice, chicken, banana, apple, toast, meat, no milk.

Patient accident free (30 days) but unhappy with food choices





Flush 150/10 Lam (2.5mg Tss) 5Ths 4x/day

Unsuccessful Bowel Management

- Additional foods added Q48 hours
- Patient started with accidents
- Slowly decreased food/fluid intake to starvation
- Admitted to hospital for dehydration
- Loss of 7 kg
- Poor body image
- Angers easily

*Restarted enemas, miserable diet, behavioral health consult Lomotil 40 mg Q6 and added Loperamide 2 mg every day.

