Hirschsprung's Disease

5-Year-Old Boy with Obstructive Symptoms

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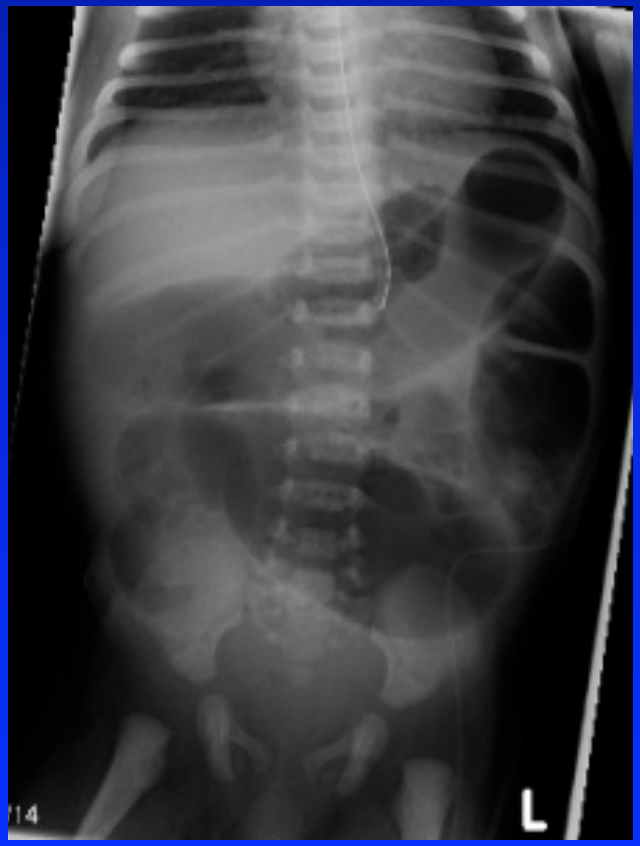
Abdominal Films

Day 2 of Life

Anteroposterior View



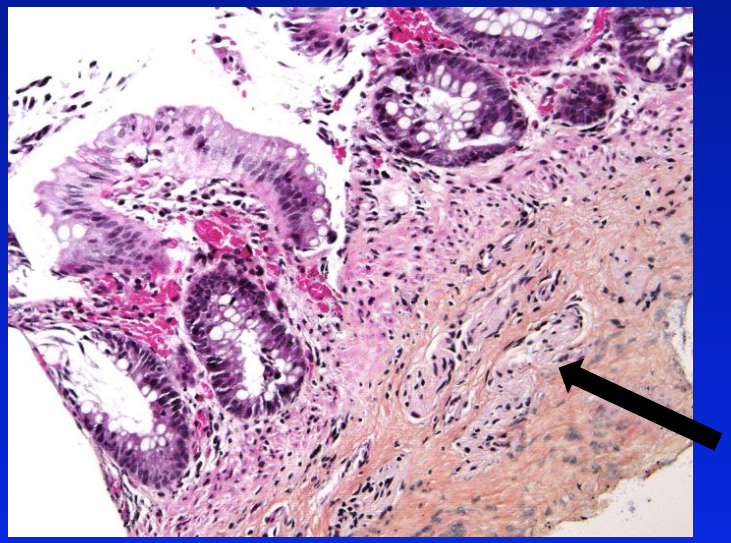
Lateral View

Contrast Enema

Anteroposterior View

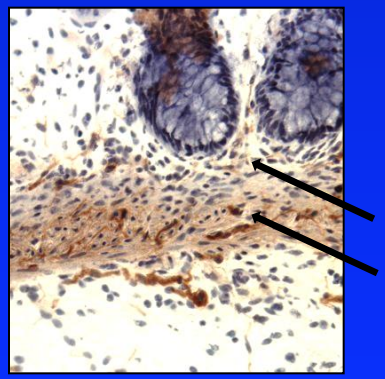
Lateral View

Biopsy Results



No ganglion cells on 80 examined levels. The nerves were mildly hypertrophic and the ACE staining pattern was abnormal.

nerves



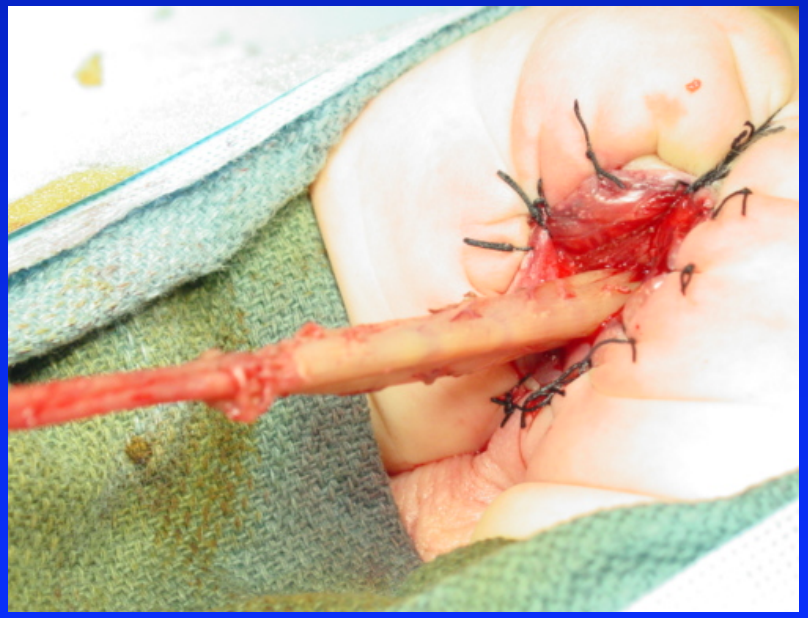
ACE staining in small nerve fibers from lamina propria (between glands) and in muscularis propria

Treatment

- Rectal irrigations continued 2-3 times per day.
- Excellent weight gain.
- No obstructive symptoms.
- Trans-anal Soave performed at 6 weeks of age.
- Discharged POD 2

Trans-anal Soave Pullthrough

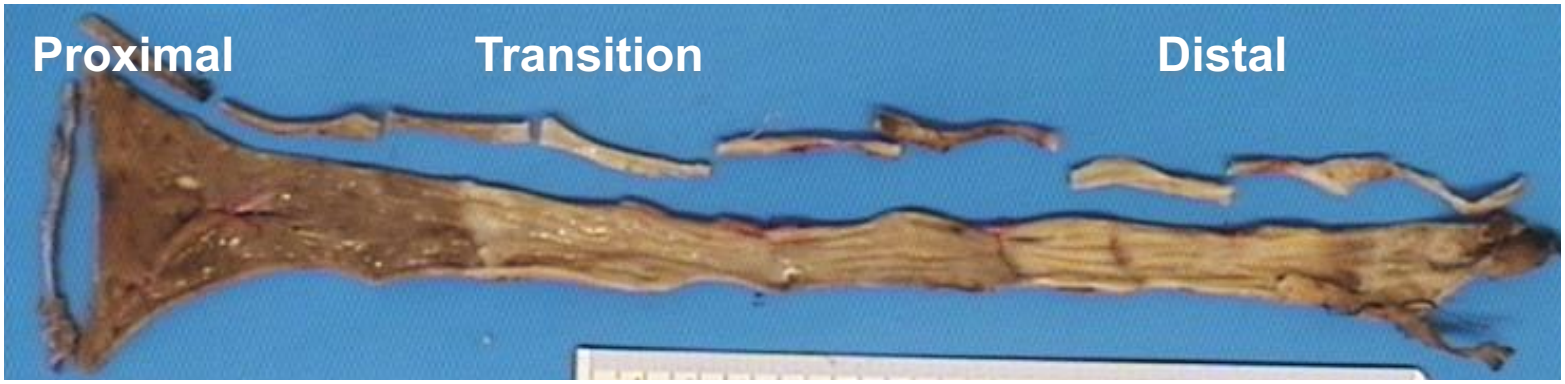
Trans-anal Dissection



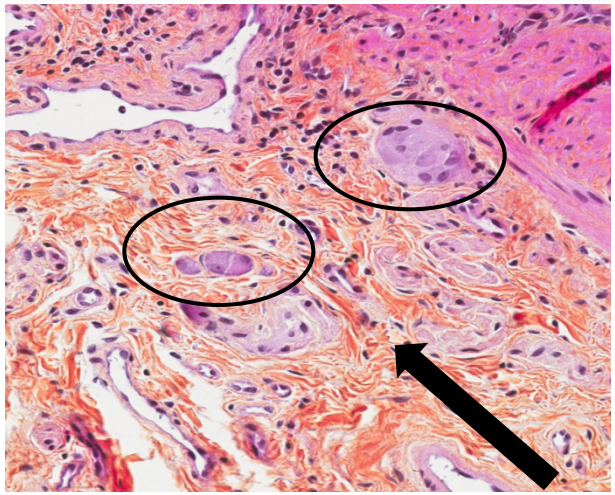
Completed Anastomosis

Pathology

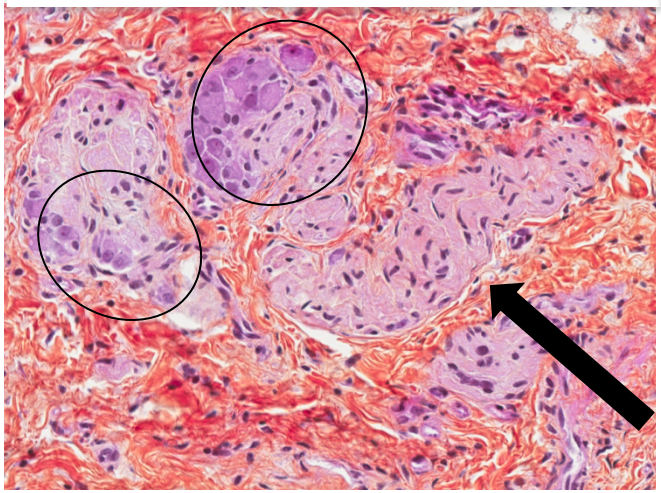
The resected recto sigmoid segment shows increased diameter proximally, and narrower diameter distally with a relatively ill-defined transition zone in between. The pathologist evaluates the presence of ganglion cells and the appearance of the nerves throughout the entire length of the resection with serial sections.



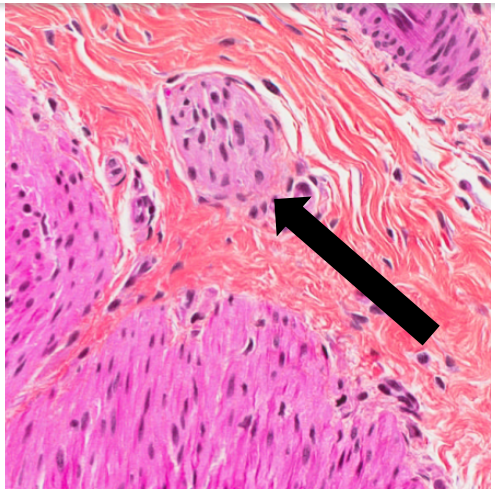
Proximal: normal ganglion cells (ovals) and small nerves (arrow)



Transition: ganglion cells present (circles) with large nerves (arrow)



Distal: large nerves (arrow) with no ganglion cells (aganglionosis)



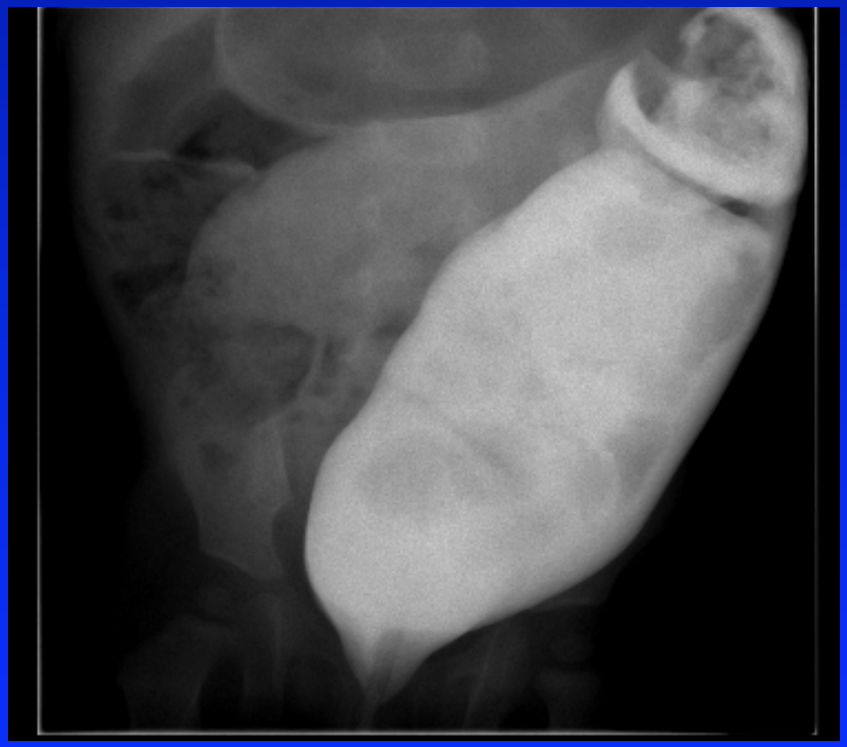
Progress

- No dilations needed.
- Excellent progress @ 2 weeks, 4 weeks, 3 months, 6 months, 1 year.
- @18 months –
  - Problems passing flatus.
  - Intermittent abdominal distention.
  - Chronic leakage of soft stool X 3 months.
- Exam:
  - Abdomen moderately distended, but soft and non-tender.
  - Rectal exam without stenosis or palpable cuff. Semi solid stool

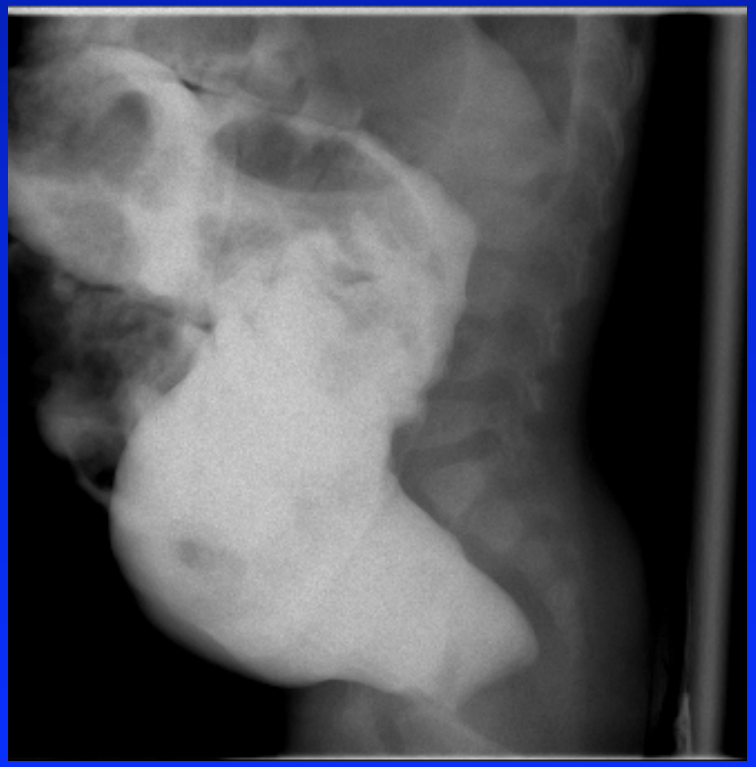
Contrast Enema

Age 18 months

Anteroposterior View



Lateral View



What next?

Review pathology &

A. Start laxatives
B. Obtain Barium Enema.
C. Perform REUA & biopsy.
D. Perform REUA & Biopsy & Inject Botox
E. Other

Work-Up

- Review Pathology
- Started senna.

Persistent Obstructive Symptoms After Surgery for Hirschsprung's Disease: Development of a Diagnostic and Therapeutic Algorithm

By Jacob C. Langer
Toronto, Ontario

Background: Although most children with Hirschsprung's disease (HD) do well after pull-through surgery, some continue to have persistent obstructive symptoms that may lead to significant morbidity. The author's goal was to develop an organized algorithm for the workup and ultimate management of these symptoms.

Methods: All children referred to the author with severe obstructive symptoms after a pull-through for HD were evaluated using an algorithm based on 5 potential etiologies, and appropriate therapeutic interventions were instituted.

Results: A total of 49 children were evaluated over 7 years. Mechanical obstruction was found using rectal examination and barium enema in 7, of which, 2 had resection of a Duhamel spur, and 5 had a repeat pull-through (after failed dilatation). Rectal biopsy results showed aganglionosis in 10, of whom, 8 underwent repeat pull-through, and 2 refused further surgery. Abnormal intestinal motility or intestinal neuronal dysplasia was found proximal to the aganglionic segment in 10 children using colonic and small bowel manometry or laparoscopic biopsies; 4 of these had additional colon resected and repeat pull-through, and 6 were treated with a bowel management routine, cecostomy, or stoma formation. Internal sphincter achalasia was felt to be the primary cause of symptoms in 14, and all were treated with intrasphincteric botulinum toxin. Eight were found not to fall into any of the above groups and were treated medically for presumed functional megacolon; 2 of these ultimately had a colostomy.

Conclusions: An organized approach to the diagnosis and management of obstructive symptoms in a child after a pull-through for HD permits accurate diagnosis and effective treatment in the majority of cases.

INDEX WORDS: Hirschsprung's disease, post–pull-through complications, treatment algorithm for failed pull-through.

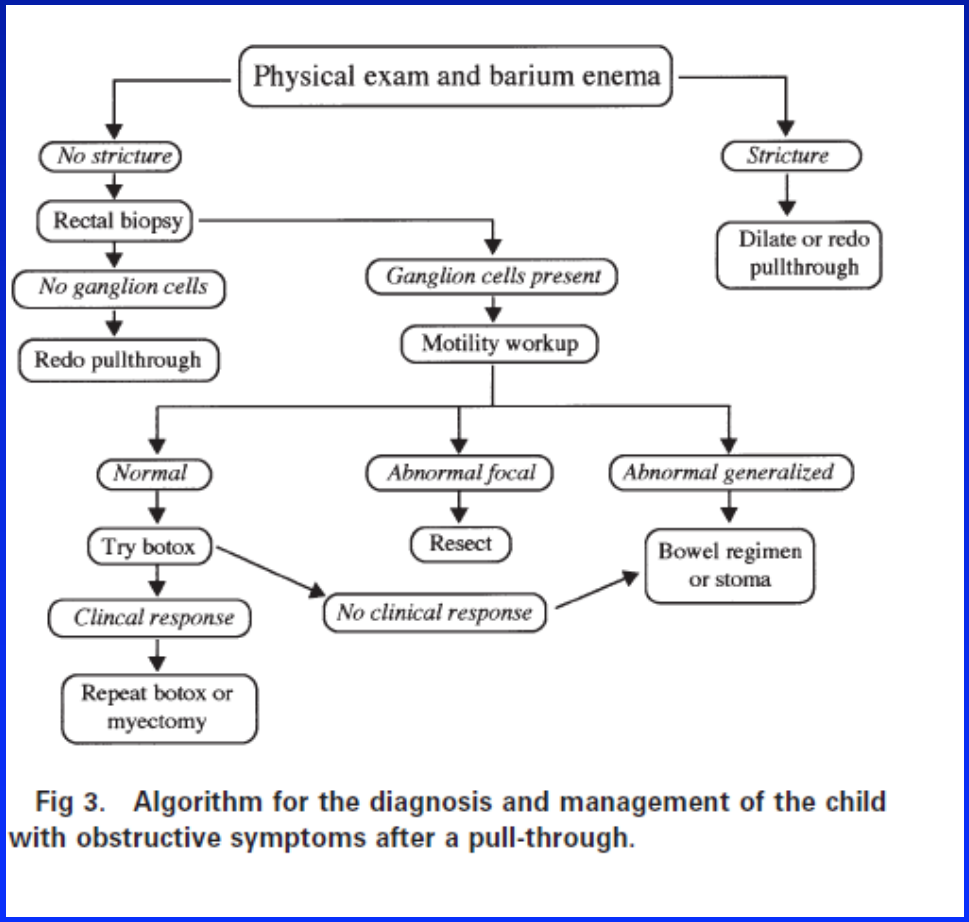


Fig 3. Algorithm for the diagnosis and management of the child with obstructive symptoms after a pull-through.

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Procedure

- Anorectal Exam Under Anesthesia.
- Deep rectal biopsy.
- US-guided injection of botulinum toxin into internal sphincter.

Procedure Results

- REUA
  - **No stenosis**
  - **No palpable cuff**
- Deep rectal biopsy.
  - **Normal ganglion cells. Normal nerves.**
- US-guided injection of botulinum toxin into internal sphincter.
  - **Transient improvement lasting 2 months.**

Now What?

A. Increase laxative dose
B. Retrograde Enemas / Bowel Management
C. Further Botox Injections
D. Myomectomy
E. Redo pull-through
F. Other

Additional Procedures

- A course of 3 Botox Injections 3 months apart
  - Diminishing return with each injection.

Progress

- Continent of urine and stool at 4 years.
- Soiling 2-3 X per week
- Persistent difficulty passing gas with intermittent abdominal distention.
- Stool always semi-solid.
- Decreased appetite and poor weight gain during previous year.

Why is the stool semi-solid?

A. Poor colonic water absorption.
B. Poor colonic motility.
C. Bacterial overgrowth.
D. Other malabsorptive disorder.

???

- What is the etiology?
- Motility studies?
- Probiotics?
- Fecal transplant?
- Redo pullthrough?
- Time?

Current Status

- **Significant improvement during COVID-19 crisis!!!**