

Complicated Hirschsprung's disease

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Present History

- 3-year-old child with proximal transverse colostomy
- Status post redo-pullthrough
- waiting for colostomy closure

PAST HISTORY

- Modified Duhamel procedure (single stage) on May 2019 (1 year of age)
- Exploratory laparotomy with transverse colostomy on June 2019
(site of perforation could not be found)
- Distal cologram on January 2020 was suggestive of stricture at the coloanal anastomosis

PAST HISTORY

- Colonoscopy done under general anesthesia on January 2021 showed the presence of stricture 1 cm above the dentate line . Balloon dilatation was done
- Child was kept on serial rectal dilatations
- Colostomy closure was done on March 2021
- On postoperative day 2 child developed features of intestinal obstruction
- Re exploration was done on March 2021 after which a redo colostomy, lysis of adhesions and rectal stricturoplasty was done .

Past History

- Child was lost to follow up for 2 years and then was taken up for diagnostic colonoscopy followed by rectal biopsy in January 2022
- As rectal biopsy showed absence of ganglion cells child was planned for a redo pull through

Colonoscopy

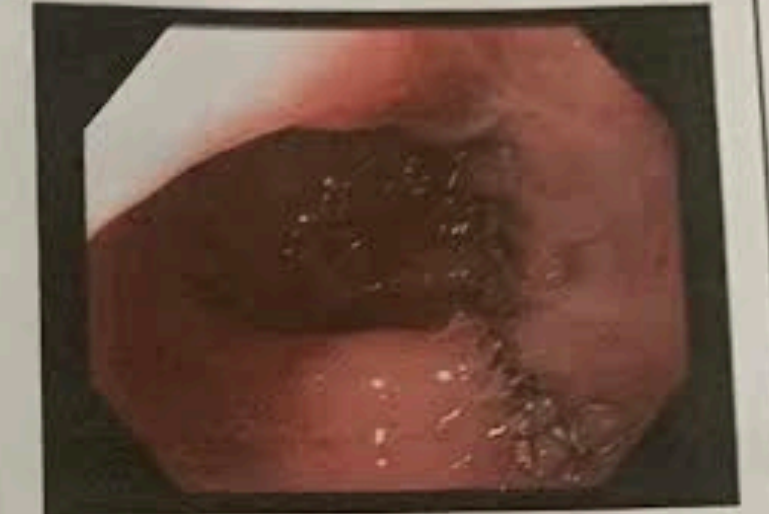
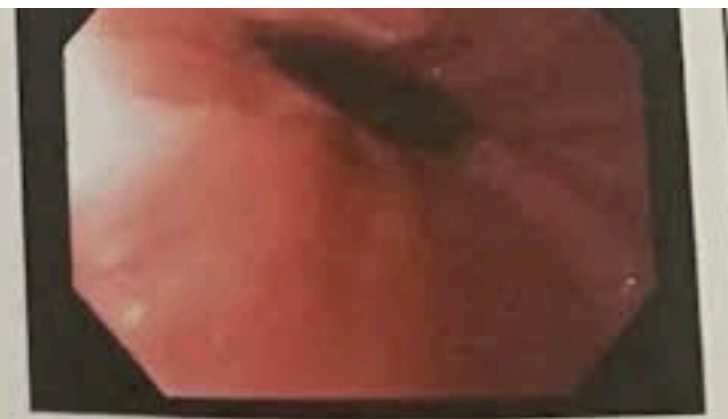
Instrument : Olympus CF H180AL
Sedation : General anaesthesia
Indication : F/U Hirschsprungs disease , status
coloanal anastomosis / post stricturoplasty / post
transverse colostomy , for assessment

Findings :
Scope passed through the distal stoma and visualised upto dentate line , coloanal anastomosis was noted just above the dentate line (~ 1 cm) , minimal luminal narrowing noted at the site of anastomosis but the scope was negotiated without any resistance. Colonic mucosa appeared edematous.

Scope was passed through the rectum and visualised upto splenic flexure , coloanal anastomosis was noted just above the dentate line (~ 1 cm) , minimal luminal narrowing noted at the site of anastomosis but the scope was negotiated without any resistance. Colonic mucosa appeared edematous.

Scope was passed through the proximal stoma and visualised upto terminal ileum , stools noted in the lumen and the visualised mucosa was normal , terminal ileal mucosa was normal and bile stained.

Impression :
F/U Hirschsprungs disease , status coloanal anastomosis /
post stricturoplasty / post transverse colostomy
Minimal narrowing at the site of anastomosis
? Diversion colitis



Full thickness rectal biopsy

Calretinin IHC done on A1 and A 2 block *:
Calretinin-immunoreactive mucosal nerves are completely absent
Ganglion cells are not identifiable

Impression

Full thickness rectal biopsy shows

- No identifiable ganglion cells
- Submucosal and myenteric nerve hypertrophy
- Absent calretinin immunoreactive mucosal innervations

HISTOLOGICAL FEATURES ARE CONSISTENT WITH HIRSCHSPRUNG DISEASE
(a known case of Hirschprung disease S/P colostomy with rectostricturoplasty)

COMMENTS

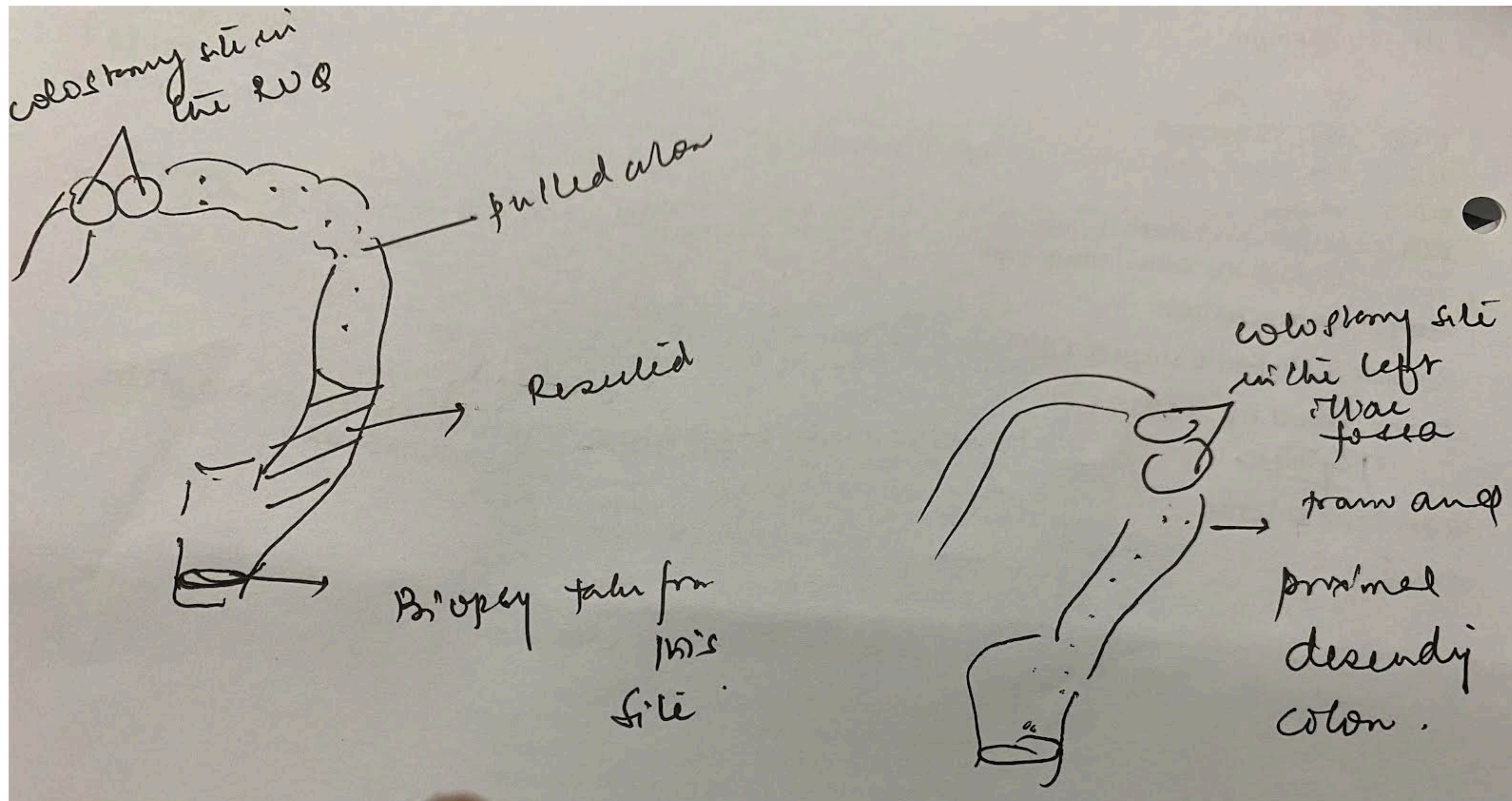
* Immunostaining done in Oncore Diagnostics, Bangalore and interpretation done in PIMS,
Puducherry
IHC not under NABL scope

*** End Of Report ***

Past History

- Child underwent redo-pullthrough on April 2022
- Frozen section was taken at multiple sites.
- Biopsy suggested -
 - Transition zone in the rectosigmoid region
- Presence of ganglion cells in the descending and transverse colon
- Mobilization of colostomy , ascending colon and cecum was done with resection of the sigmoid and descending colon was pulled through with coloanal anastomosis done just proximal to the dentate line .

Line diagram of procedure-



Biopsy of the resected specimen of sigmoid colon

submucosal plexuses. No evidence of nerve hypertrophy.

C1 - C8: Serial slices from distal to proximal end (with respect to anus) show wall of a colon with identifiable ganglion cells in both myenteric and submucosal plexuses at distal end (C1) and towards distal end. Significant nerve hypertrophy is not detected. Focal mucosal surface erosions are evident. Swelling of myenteric plexus with edema is noted in some sections (C3)

C9-C12 Sections show colonic wall with identifiable ganglion cells, however mild thickening of submucosal nerves is noted. The nerves appear to be increased in number. Mucosa shows focal erosions.

C13, C14 Sections from slices towards proximal end show thickening of submucosal nerves and increase in their number. Few myenteric ganglia show reduced number of ganglion cells. Serosal surface shows neutrophilic infiltrate.

C15- Sections from proximal most end (with respect to anus) shows cautery artifact. Many prominent thickened nerves are noted in submucosa. Mucosal erosions and focal ulcerations are noted. Occasional ganglion cells are noted in the myenteric and submucosal plexuses, however their number is markedly reduced.

Impression

Segment of sigmoid colon (Hirschsprung Disease)

- Proximal surgical end (with respect to anal end) - Markedly thickened and hyperplastic submucosal nerves along with reduction in the number of ganglion cells. Mucosal erosions and ulcerations seen (C15) ... Suggestive of Transition zone.

- Approximately 2cm proximal portion (C9 - C14) ... Features suggestive of transition zone.

Points to remember -

- Early development of enterocolitis in postoperative patients of Hirschsprung's disease
- Stricture of coloanal anastomosis vs transitional zone pull through.

Does the audience suggest an alternative way of management of this case ?

Present Examination

- Fibrotic ring at the anastomotic site approximately 4 cm from the anal verge
- Admits index finger easily and 15/16 dilator
- Child is on daily dilatations and distal stoma washes
- Awaiting colostomy closure .

Question .

- What is the next best step for this child ?
- Colostomy closure or permanent colostomy?
- When should the colostomy closure be done ?