

Colorectal Web Meeting



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4-year-old male patient

This case is the evolution of 4 years
of treatment in our institution

Birth 02-25-2017



Referred to us (50 d after birth) because of
abdominal distention, gastric vomiting, and the need of
suppositories to pass stool

On hospital admission...



- After rectal irrigations



- 2nd day after rectal irrigations



- 3rd day after initial rectal irrigations.
- He was started on oral feeds



What would
you
recommend
now?

1. Hospital discharge
2. Continue rectal irrigations
3. Contrast enema
4. Rectal biopsy
5. I don't know

Rectal biopsy

Aganglionic in
the submucosal
plexus

Calretinin –

Doesn't have
hypertrophic
nerve trunks



Contrast enema
April 23, 2017

Which operation would you perform?

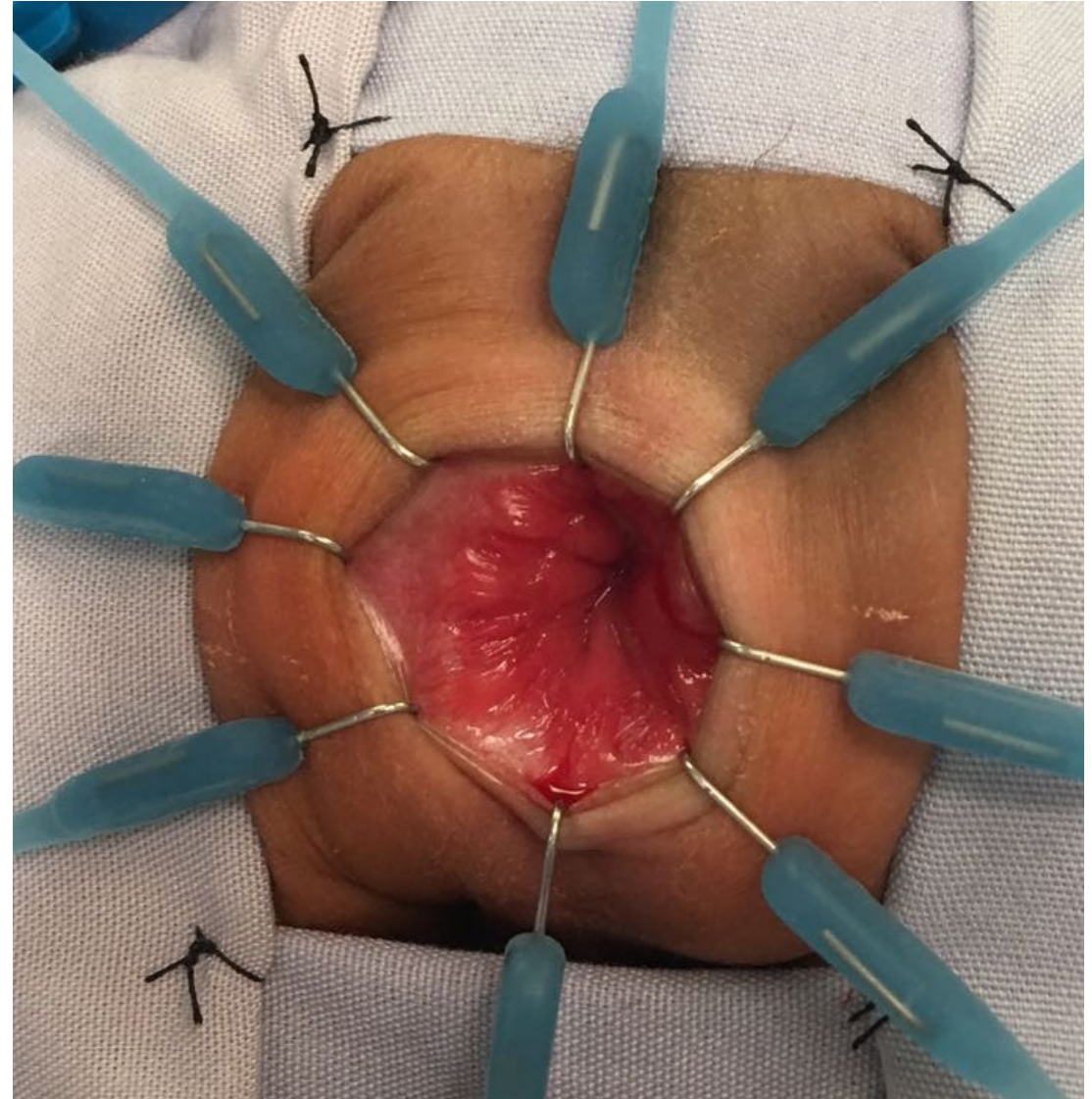
1. Ileostomy
2. Transverse colostomy
3. Transanal proctectomy and pull-through of the left colon
4. Sigmoidectomy with intraoperative frozen section biopsy proximal to the apparent transition zone
5. I don't know

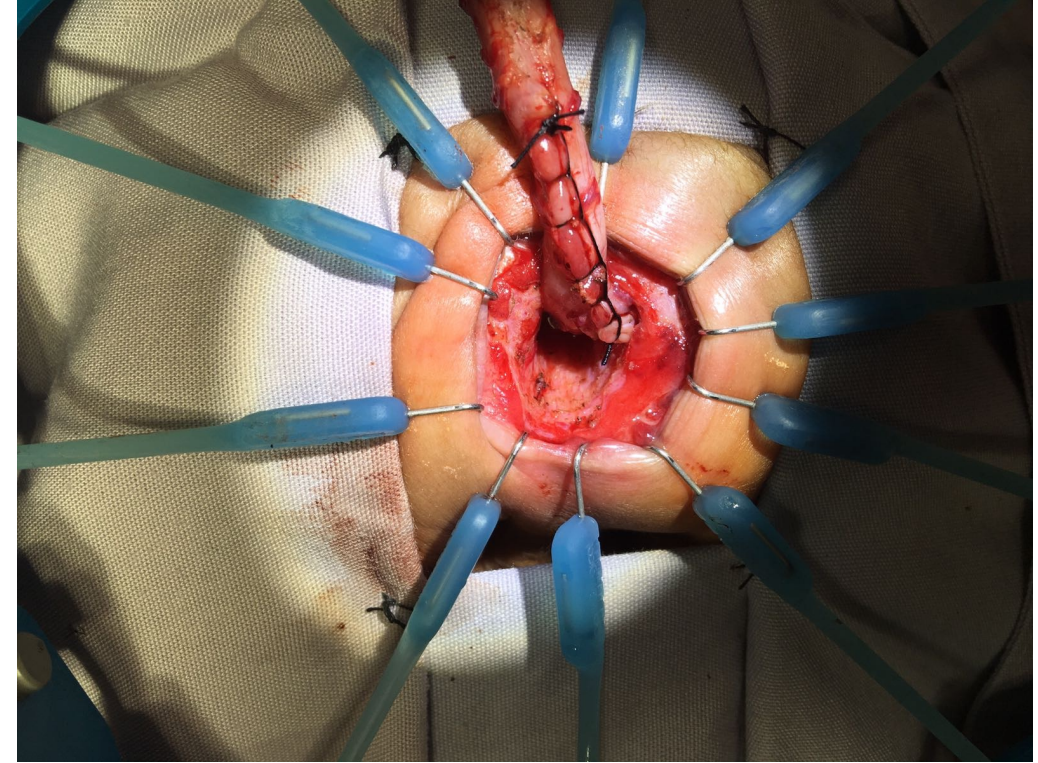
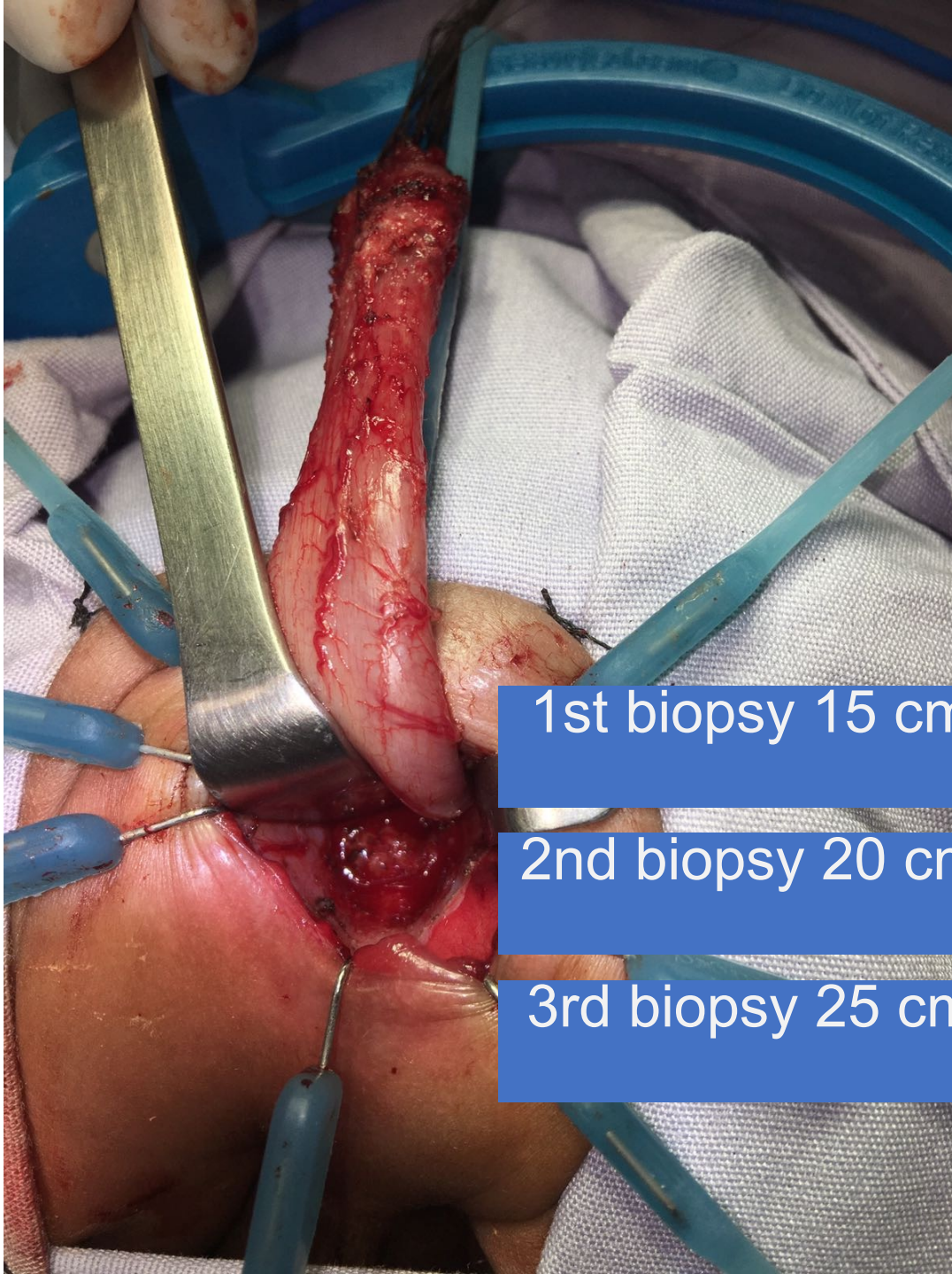
Colonic decompression with rectal irrigations + Contrast enema



- Transanal proctectomy and pull-through of the left colon

- We initiated the transanal proctectomy with the placement of Lone Star retractor...





1st biopsy 15 cm above the distal level of the proctectomy

aganglionic

2nd biopsy 20 cm above the distal level of the proctectomy

aganglionic

3rd biopsy 25 cm above the distal level of the proctectomy

aganglionic

Laparotomy and transverse colon biopsy

aganglionic

cecal appendix

aganglionic

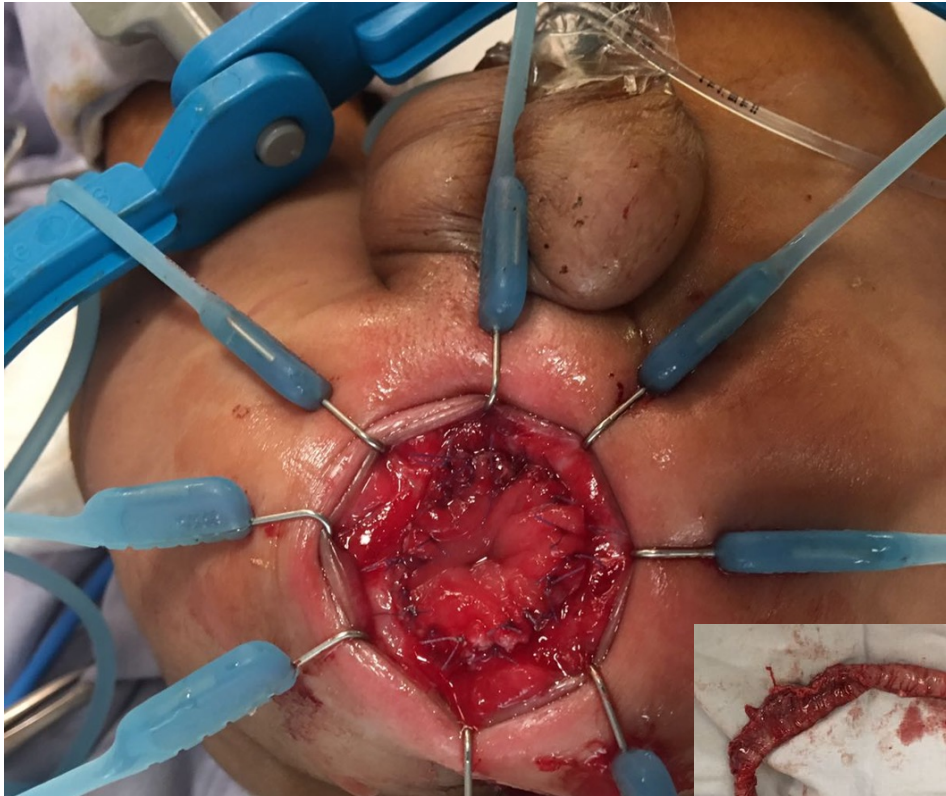


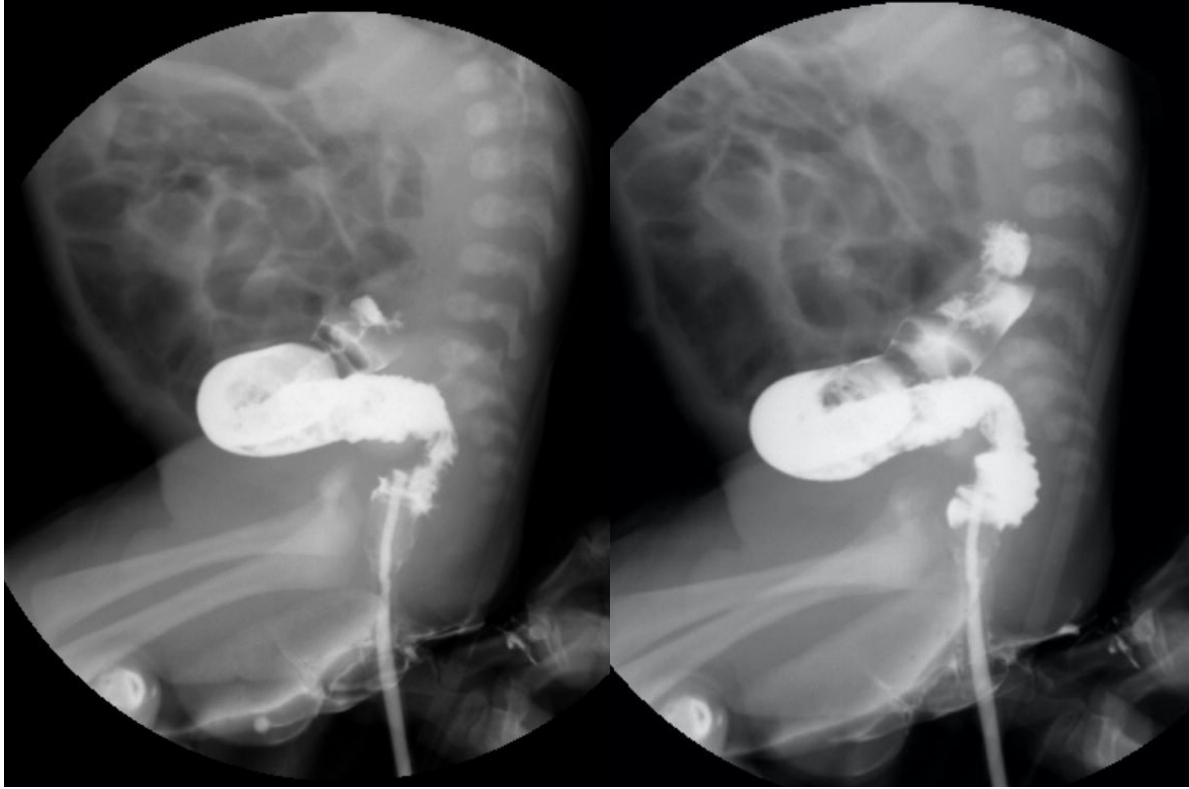
Total colonic aganglionosis without transition zone

Ileum biopsy 12 cm proximal to the ileocecal valve

normoganglionic



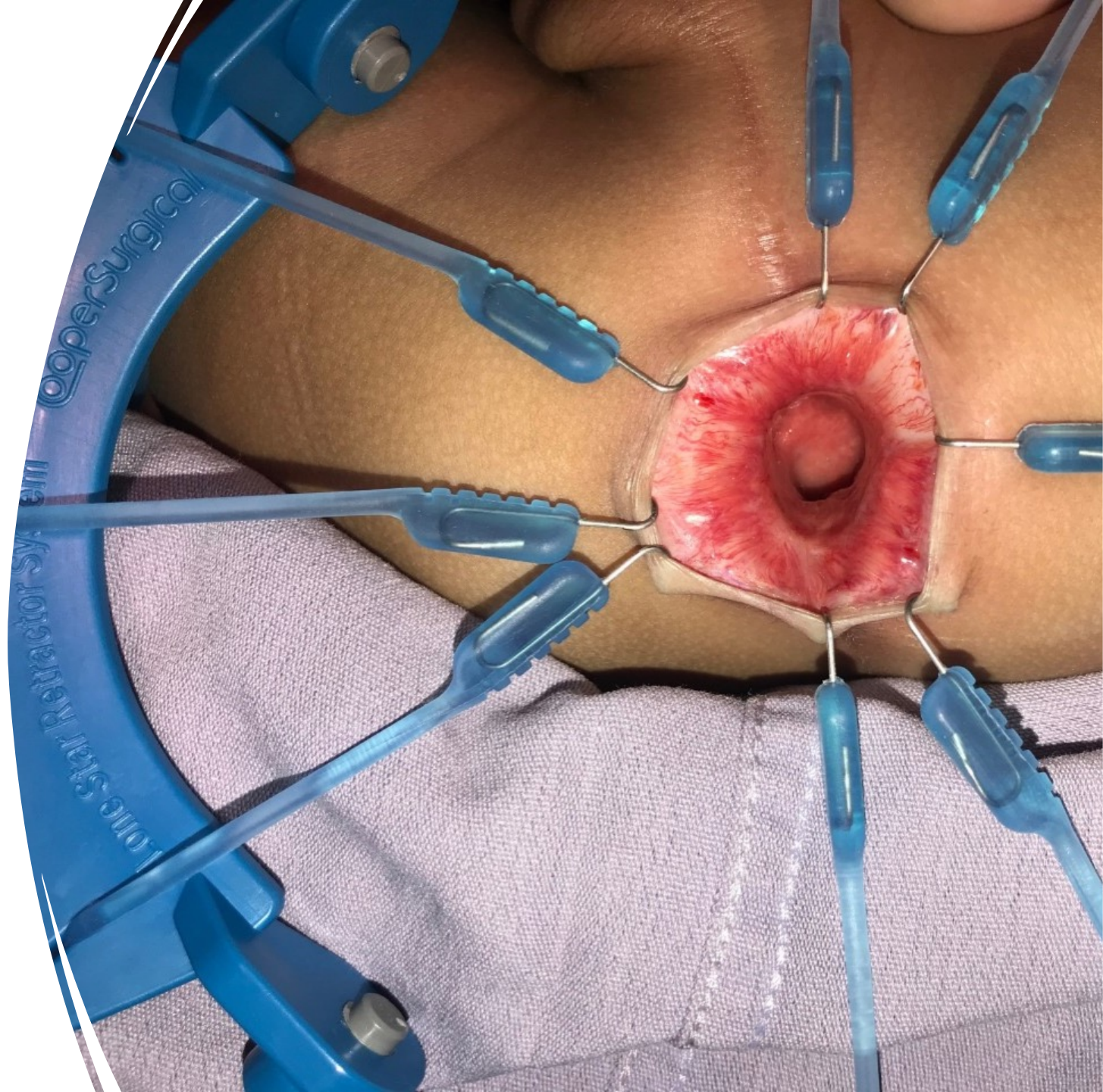


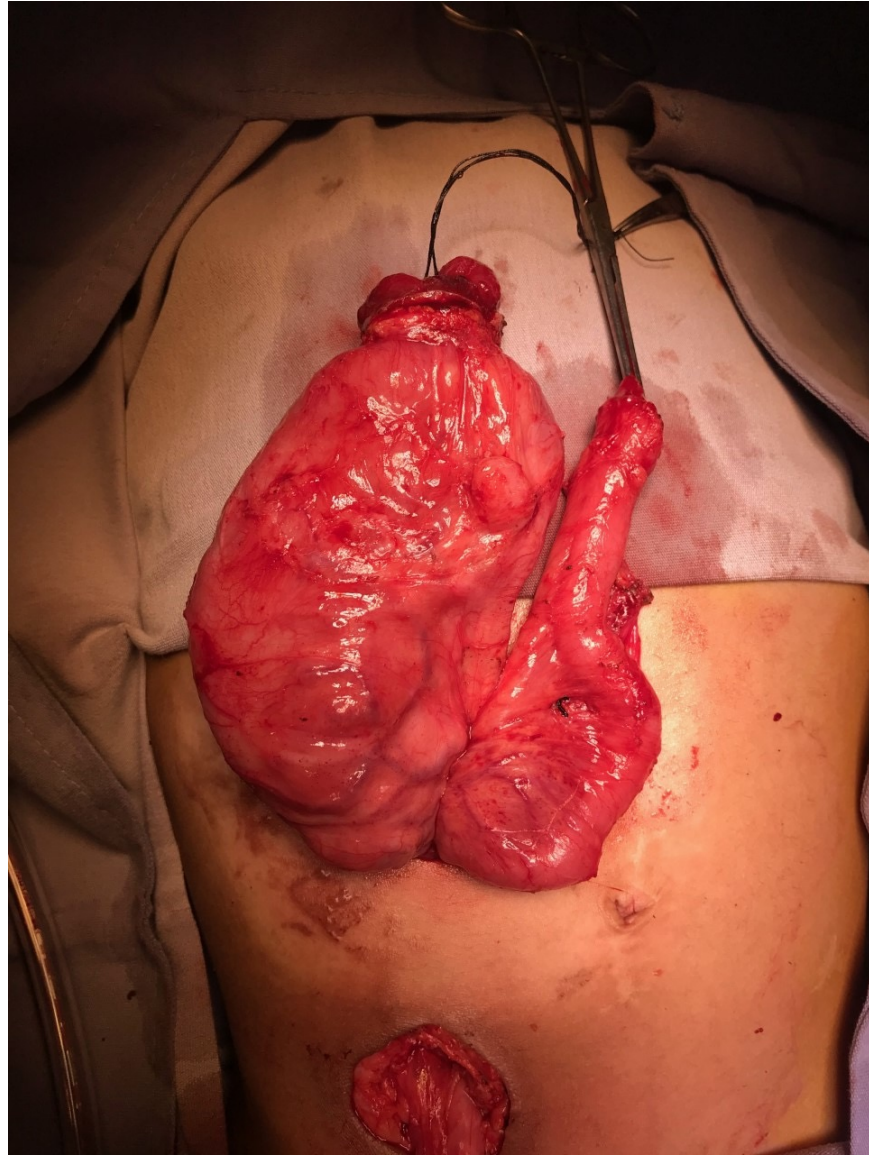


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- Dilatations program
 - At the age of 3 yr, this is the contrast enema
 - Has achieved urinary sphincter control



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- Rectal dilations 15-16 mm
 - Botox injection above the pectinate line



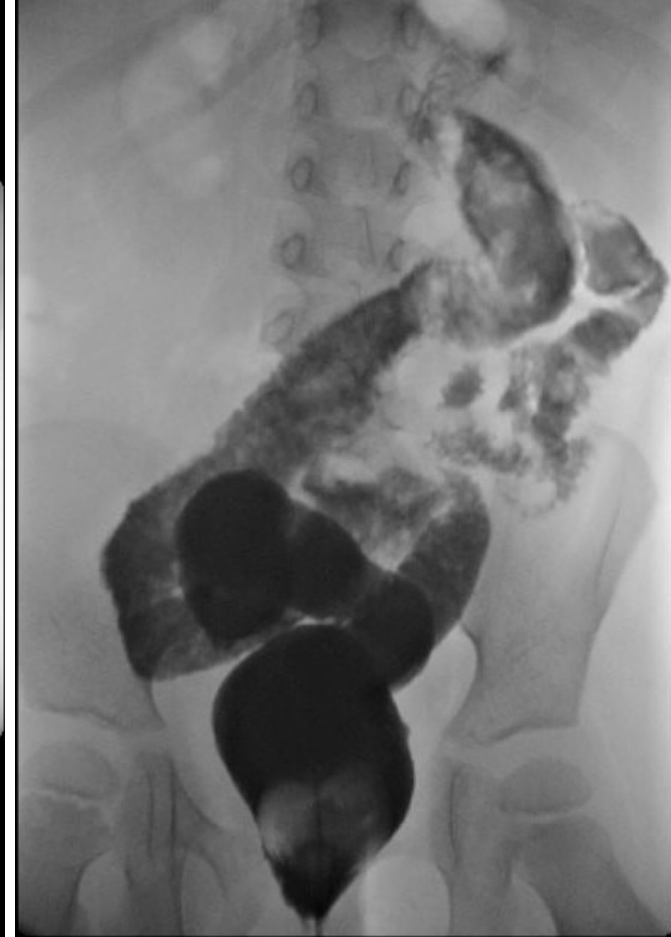


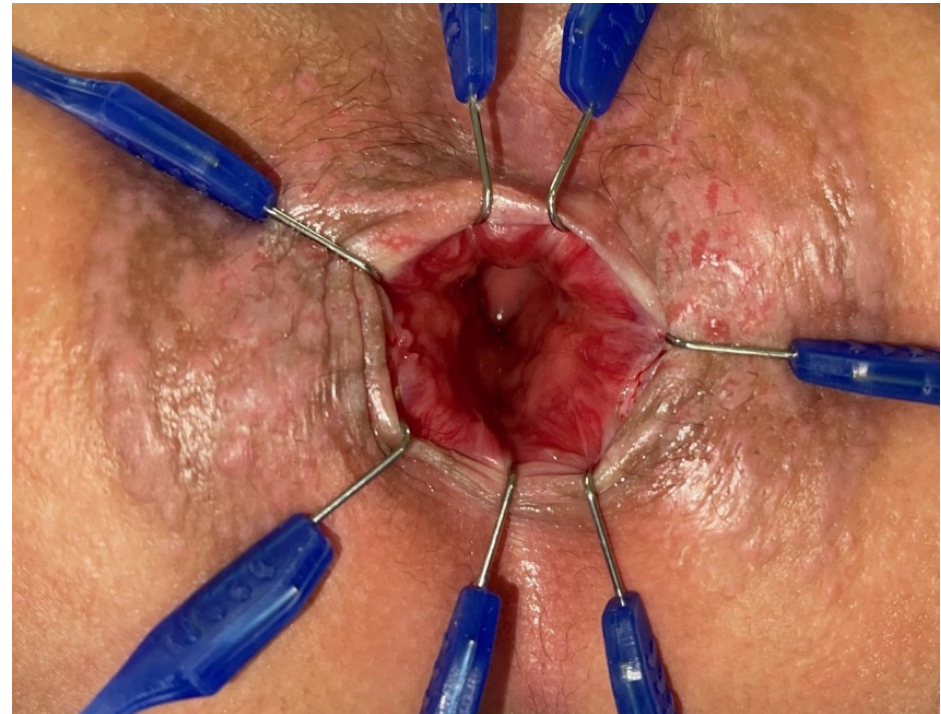
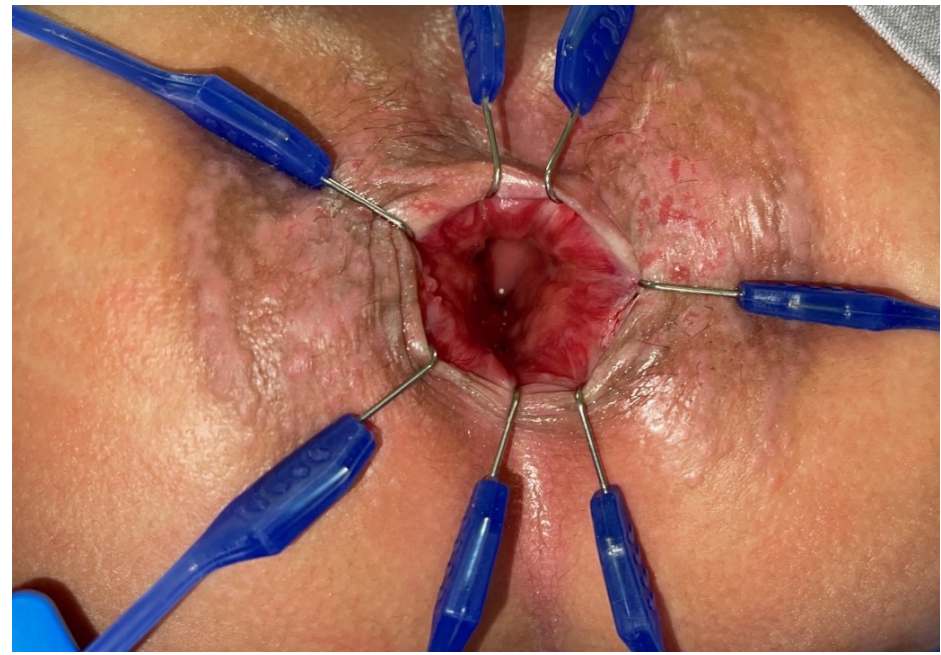
- Santulli ileostomy

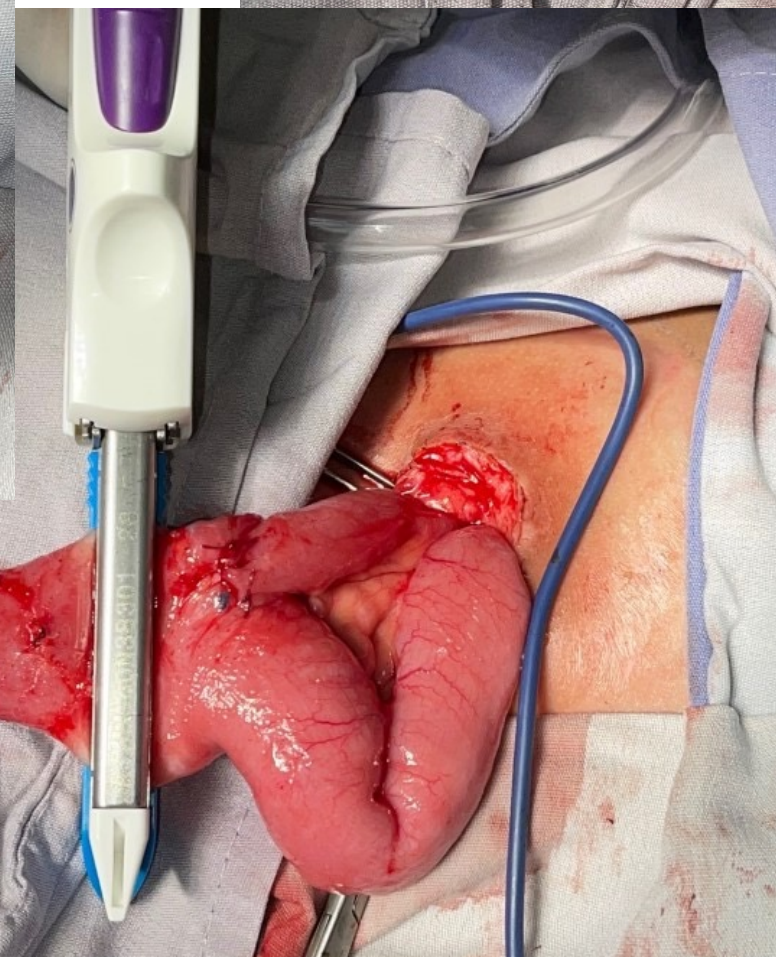
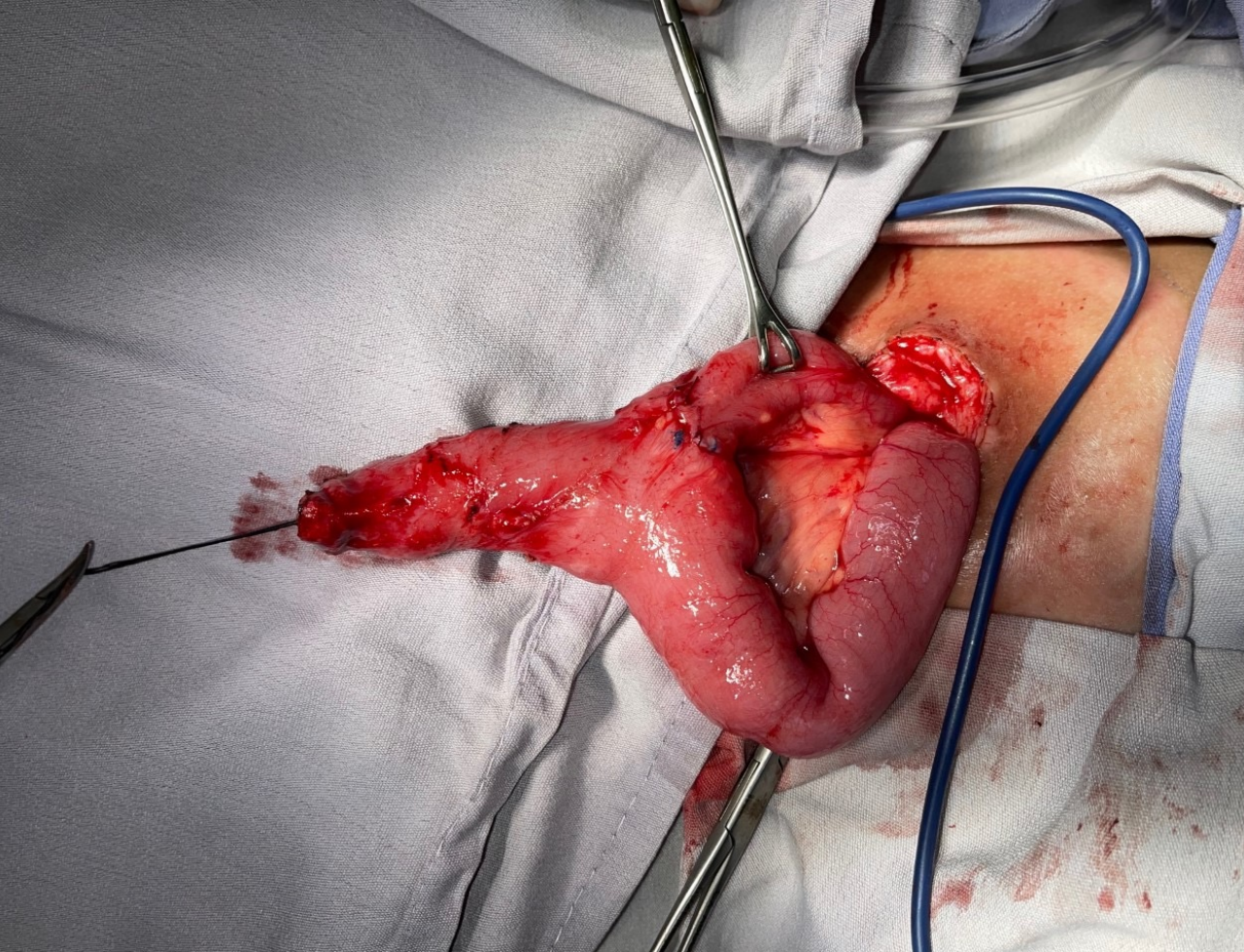
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- This is the final aspect



5 mo after the
Santulli
procedure, the
distal ileon
showed some
enlargement







Ileostomy (Santulli) closure with a stapler



What we have learned with this case...

- In total colonic aganglionosis, the transition zone can be identified in a contrast enema in just 30% of patients.
Currently, I review the studies more thoroughly
- **DO NOT DESPAIR**
- Staged surgical treatment is possible
- Important follow up is a must

Our little patient now evacuates through the rectum, he was discharged from the hospital one day after the Santulli closure

He felt some sort of separation anxiety when he noticed he did not have an ostomy bag, and although he does not need it anymore, he sticks a colostomy bag on his abdomen all the time



Thank you

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