

Total Colonic Aganglionosis – When and How to do the Pull-through

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Children's Hospital Colorado

INTERNATIONAL CENTER FOR
**COLORECTAL AND
UROGENITAL CARE**

Two Scenarios



- 1 (ideal) – you suspected Total Colonic Aganglionosis. Leveling biopsies and ileostomy.
- 2 - you did not suspect total colonic aganglionosis and this was an intra-operative finding. You have very good pathologist. Colectomy, ileo-anal anastomosis, protective ileostomy.

Our Protocol



- Ileostomy closure when:
 - Toilet trained for urine
 - Accepting rectal irrigations



Why don't we
recommend an ileo-anal
pull through in a baby?

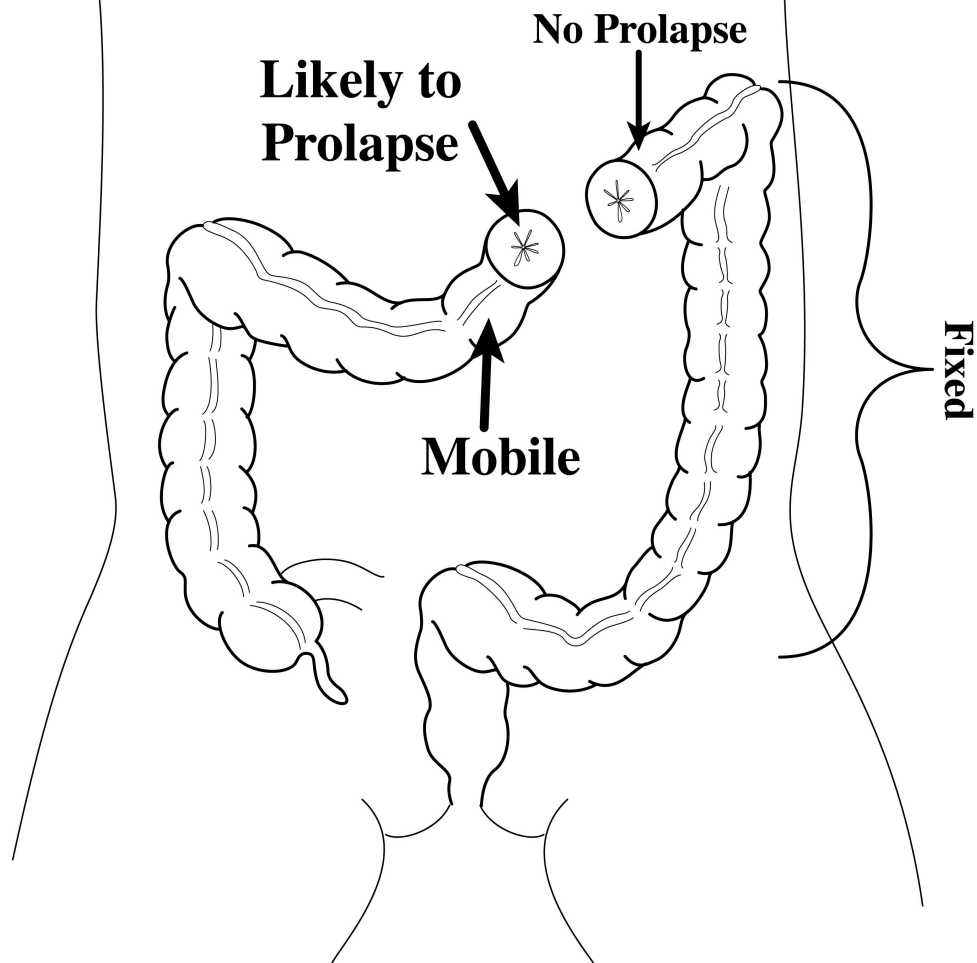
Ileostomy Prolapse

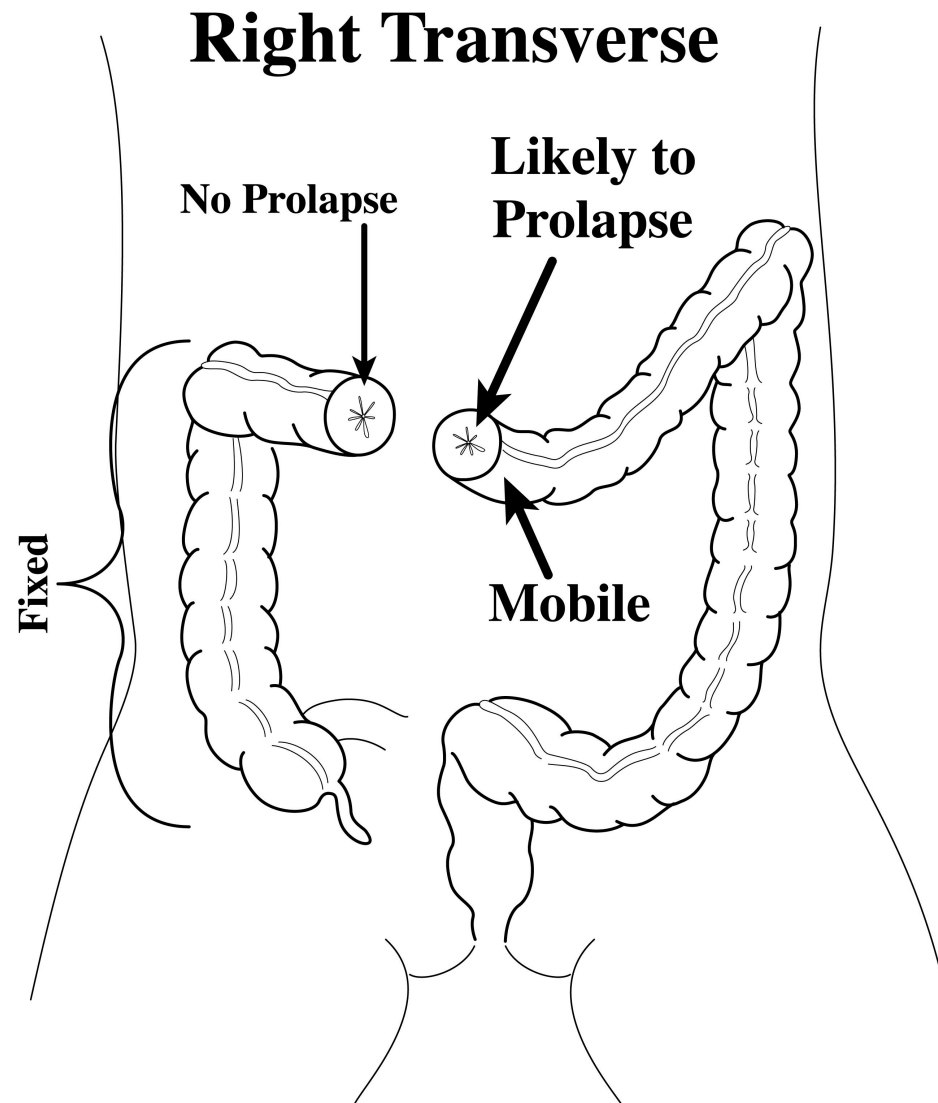


- Why do they happen?
- How can we avoid it?



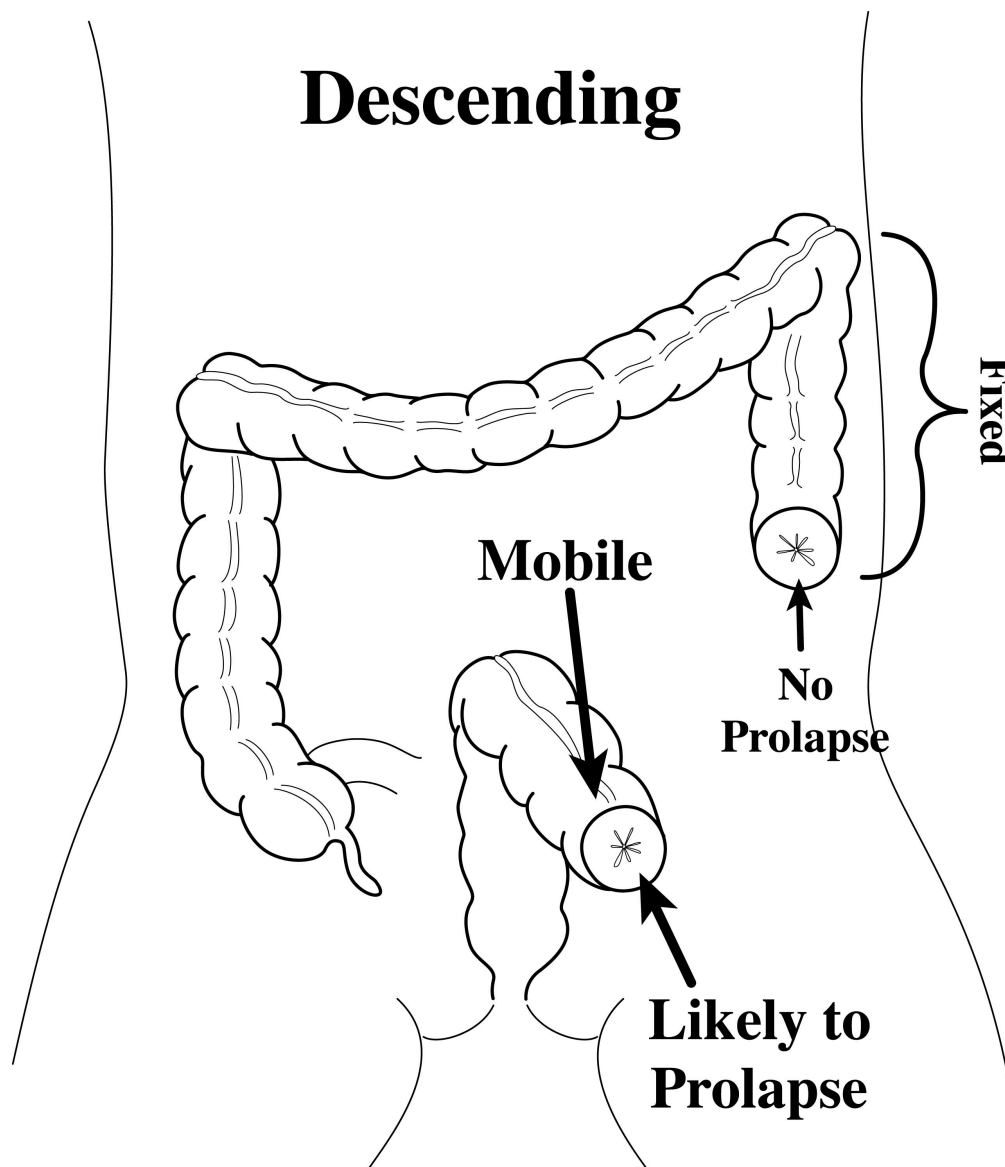
Left Transverse

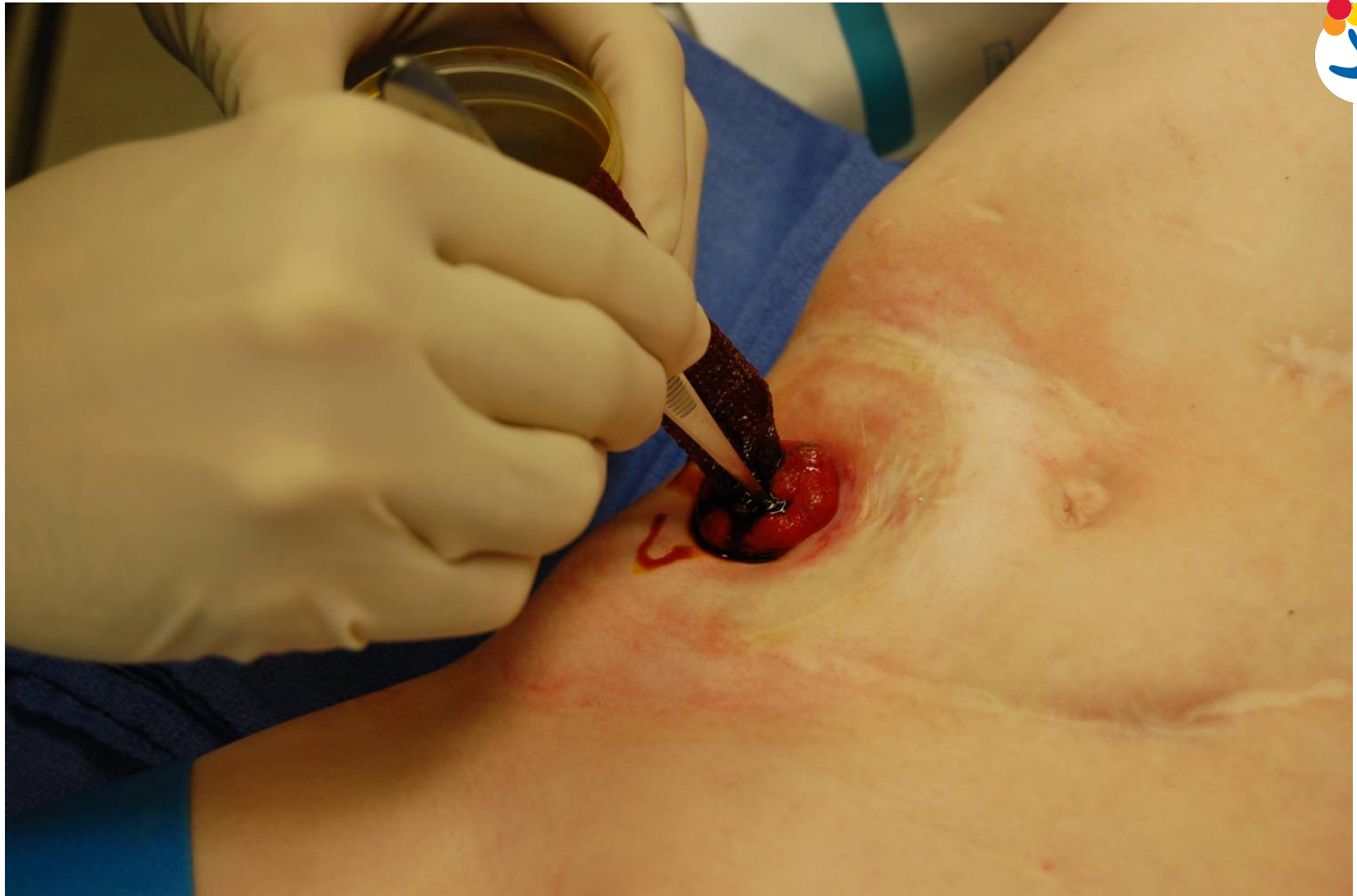




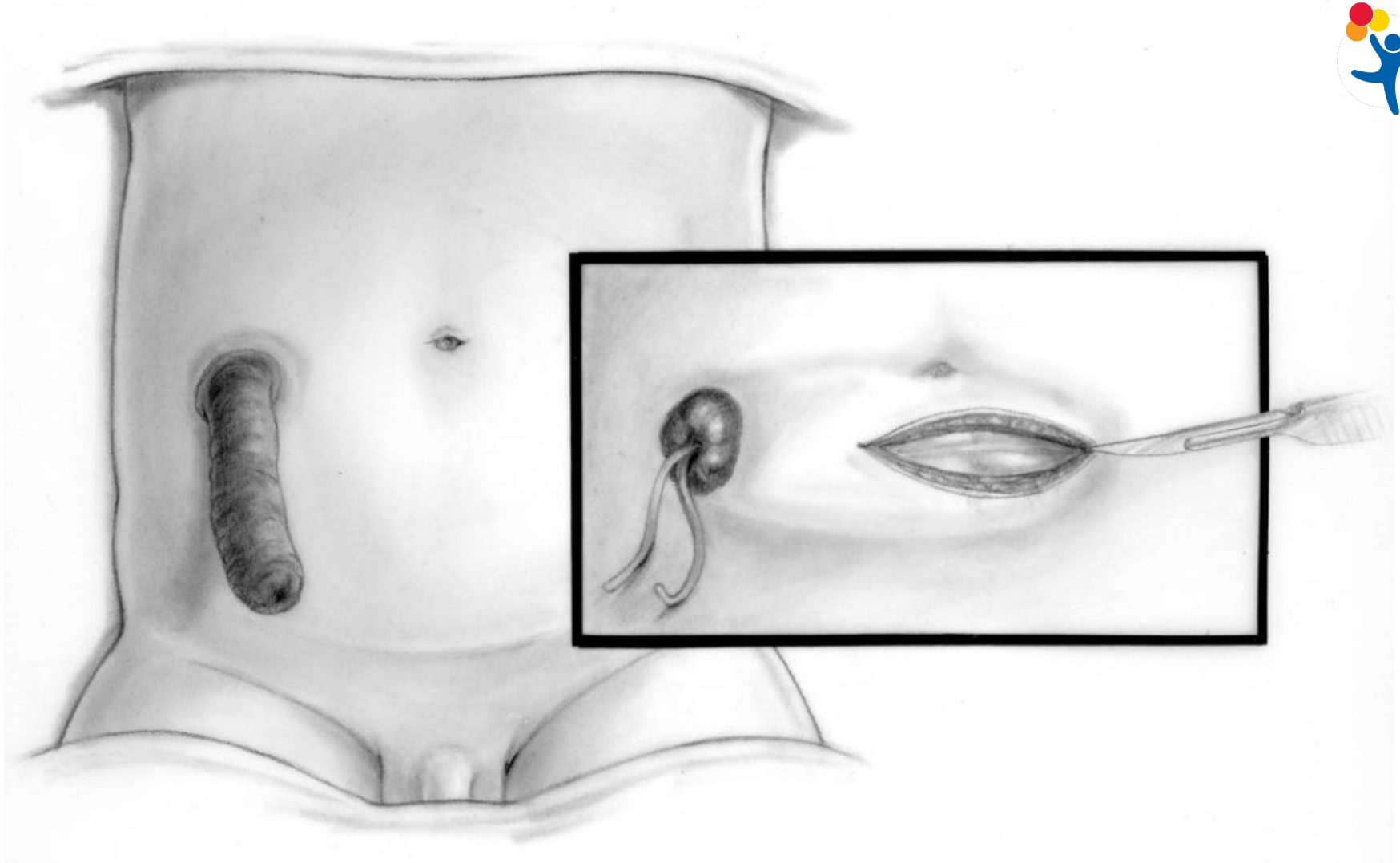


Descending









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Oral Sodium Supplementation



Request SOD CHLORIDE 4MEQ/ML INJ, 25X30ML Prescription

You can start with 1mEq/kg every 12 hours if the urinary sodium is lower than 20mmol/L.



In a patient with Total Colonic Aganglioneosis, a damaged anal canal means a permanent stoma.





You receive an email of an anxious mother saying that her 2 months old daughter received the diagnosis of total colonic aganglioneosis, underwent an ileostomy at the umbilicus, and the surgeon plans a pull-through in 6 months - she wants to know your opinion about the case.



Before giving your opinion, you should ask the following questions:

- Is the child eating and growing well?
- Is the defunctionalized colon still inside?
- Any signs of enterocolitis?
- Do you know how to do rectal irrigations if needed?

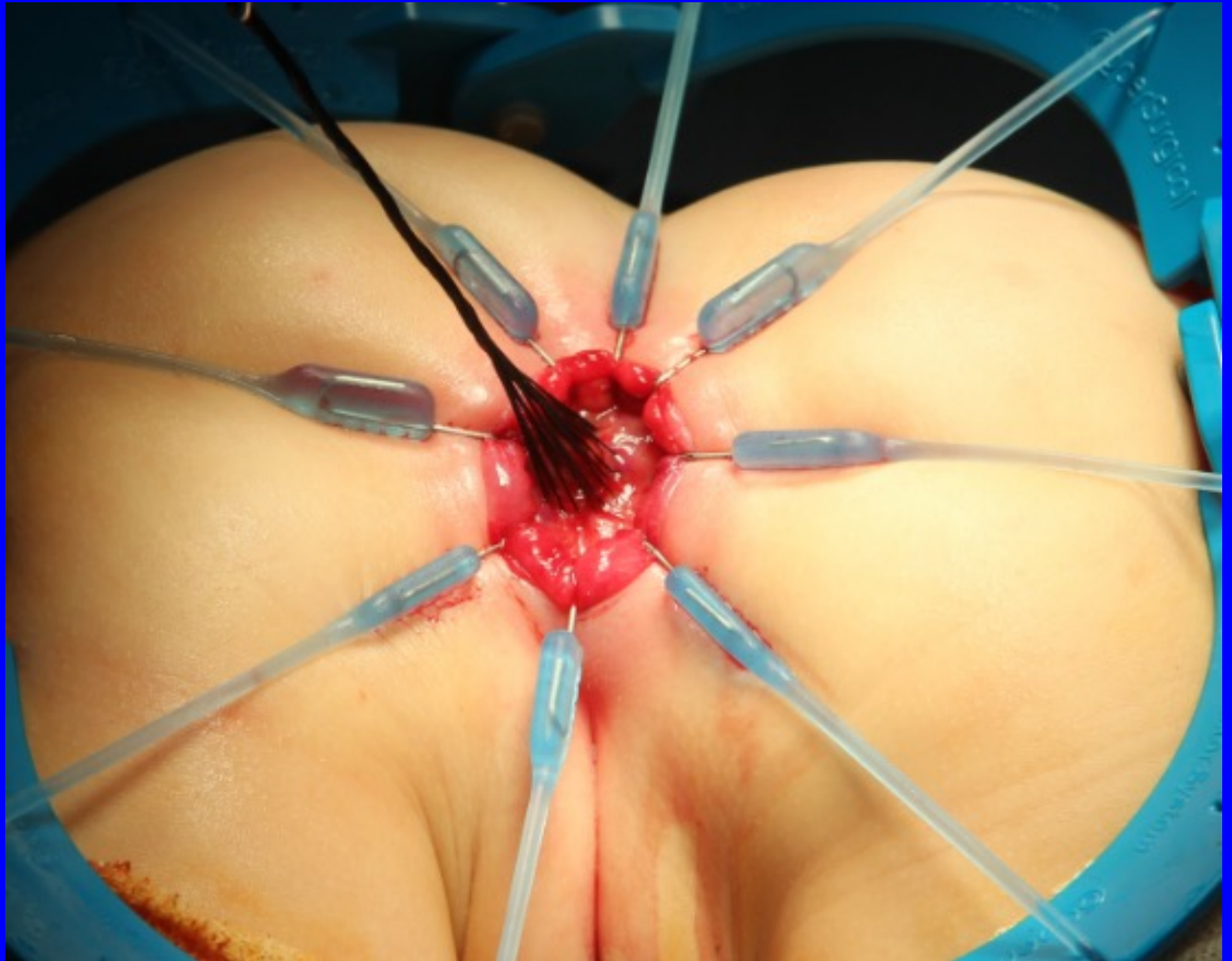


2 months after ileostomy revision



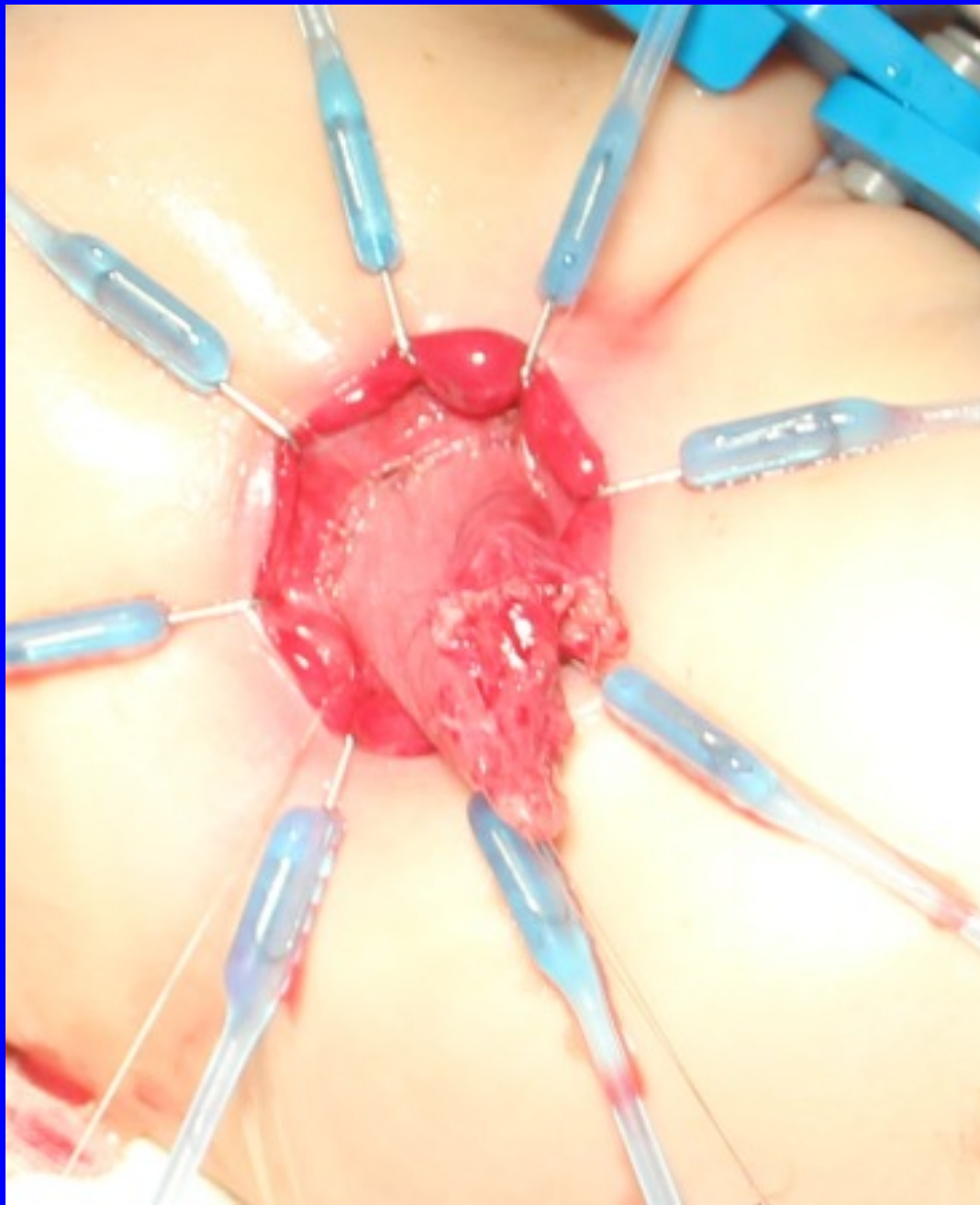
- Ileostomy was revised.
- Mother was taught how to do rectal irrigations.
- Patient is growing and eating well without any complains.

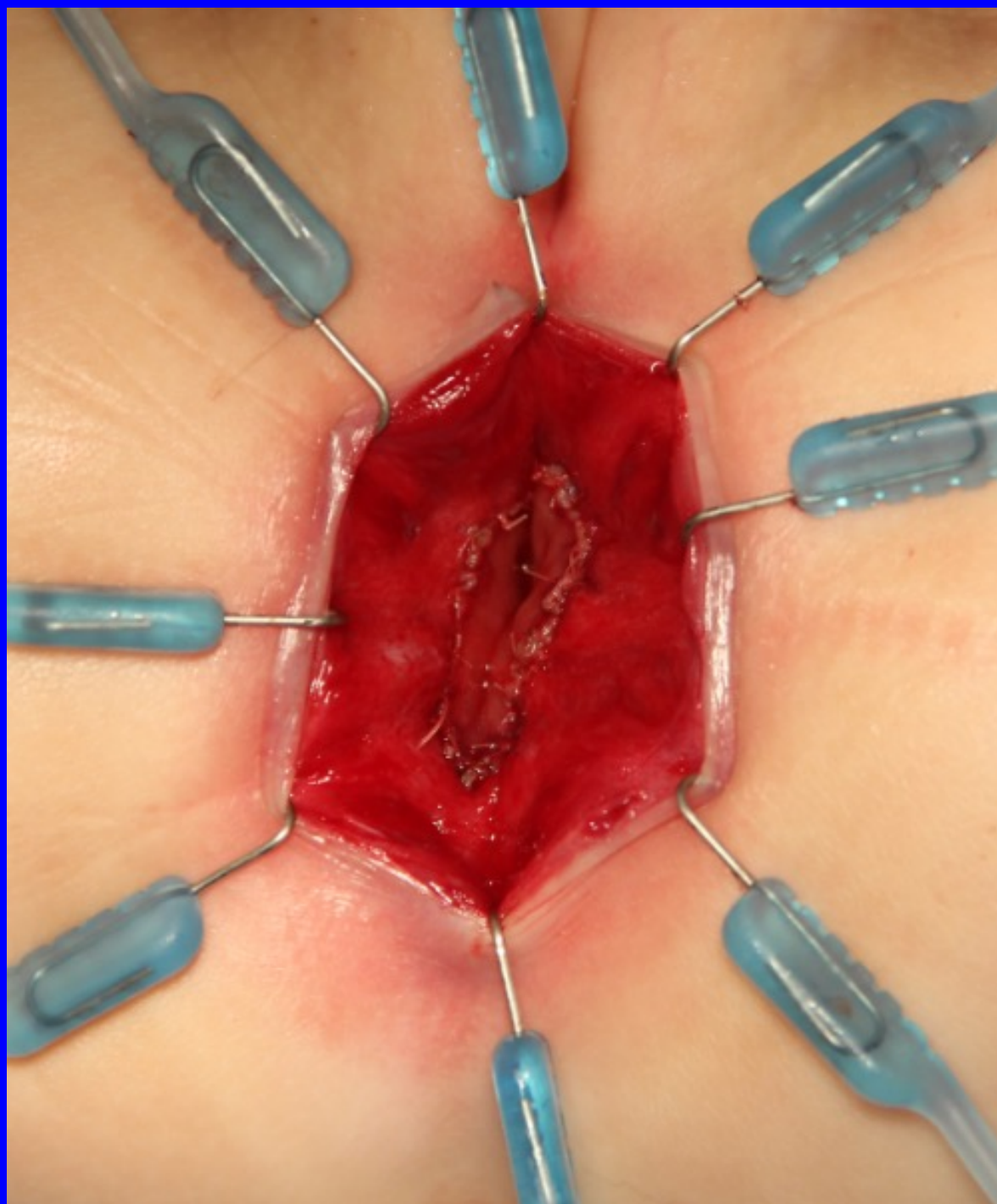
When would you
recommend the ileo-
anal pullthrough?













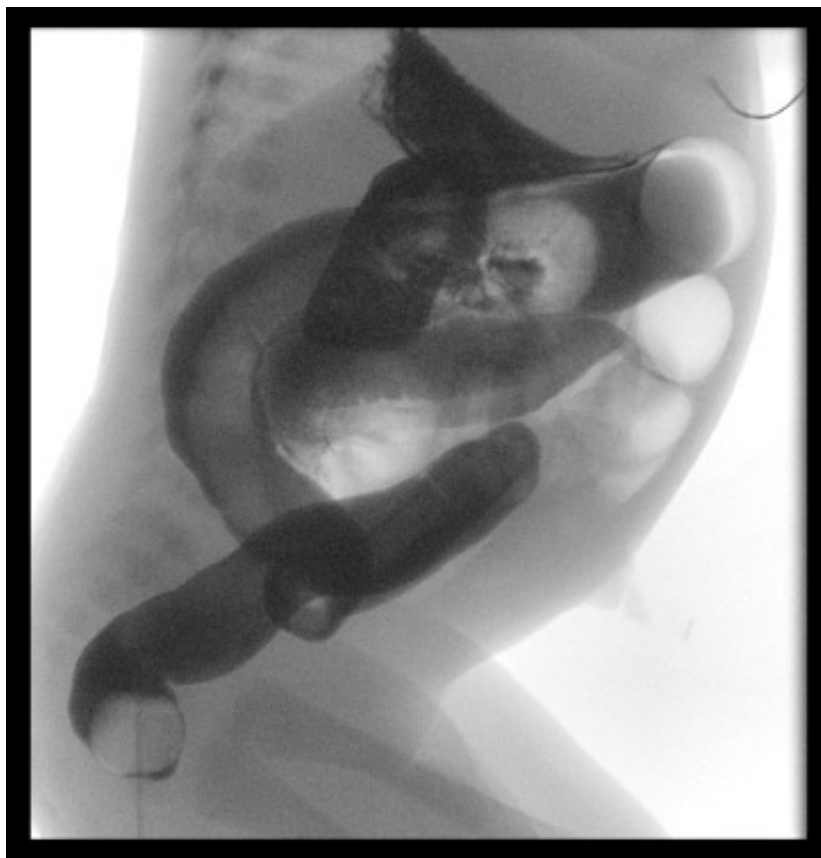
- Patient is 9 yo and has 4 to 6 voluntary bowel movements per day.





3 days old male patient
presented to the hospital
with bilious emesis and
abdominal distension







Underwent leveling
biopsies and ileostomy.

Currently waiting to meet
criteria for ileo-anal pull-
through.

Patches and Pouches (Kimura, Lester-Martin/Duhamel, J pouch)



Idea/Desire

- Absorb water
- Form solid stool
- Decrease number of bowel movements

Pouches and Patches Reality



- Fecal stasis
- Bacterial proliferation
- Inflammation
- Secretory diarrhea



Questions?