

Diagnosis and Medical Management of Hirschsprung in the Newborn

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Hirschsprung Course, August 12-13, 2021.



Children's Hospital Colorado

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- Abdominal distension
- Lack of meconium during the first 24 – 48 hours of life





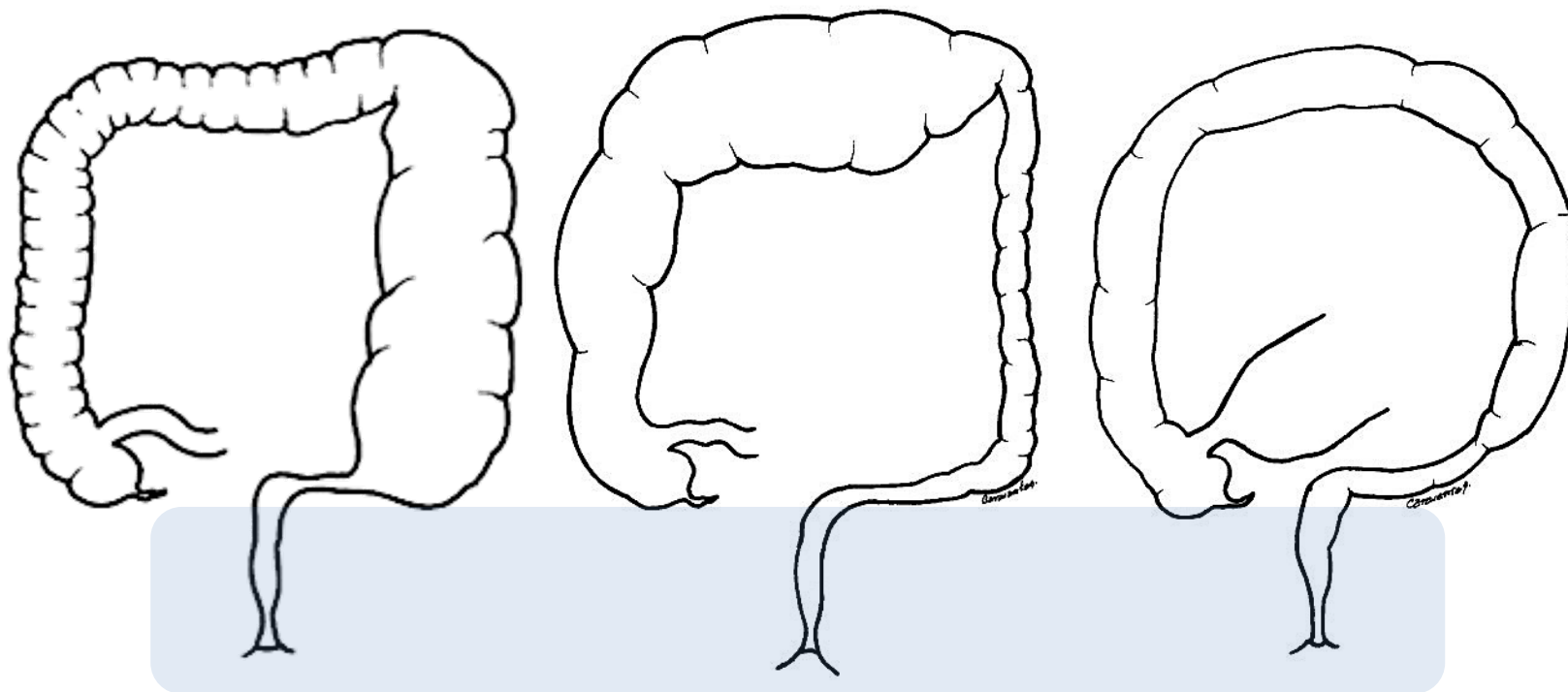
- Start Rectal Irrigation, NPO, start flagyl (metronidazole)



- It may be Hirschsprung, it may not be Hirschsprung...

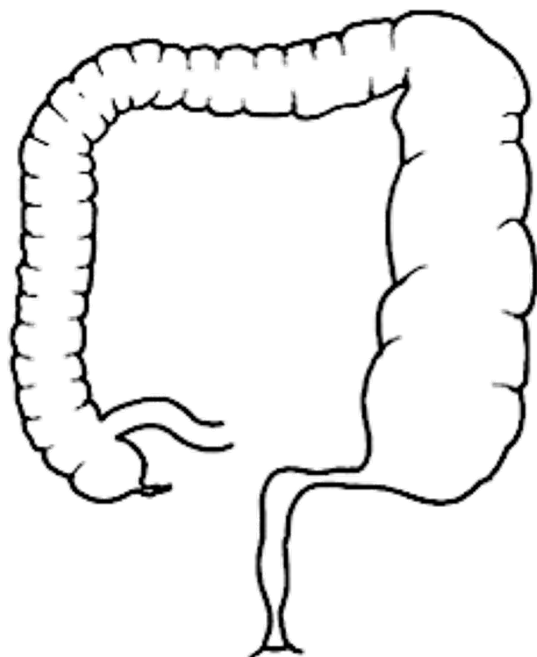


100% RECTAL DISEASE

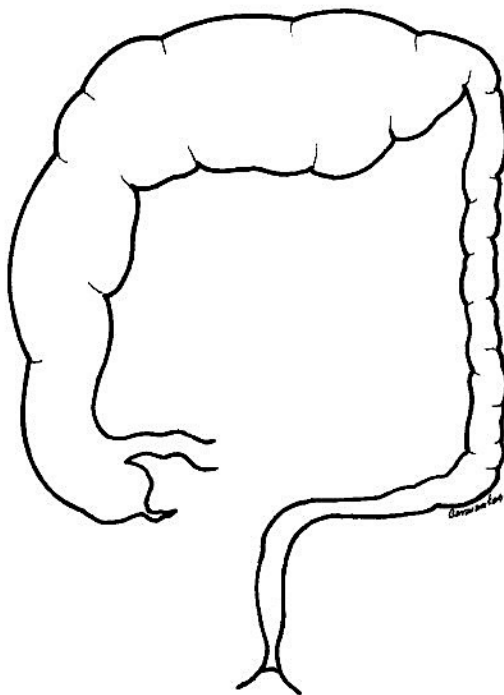




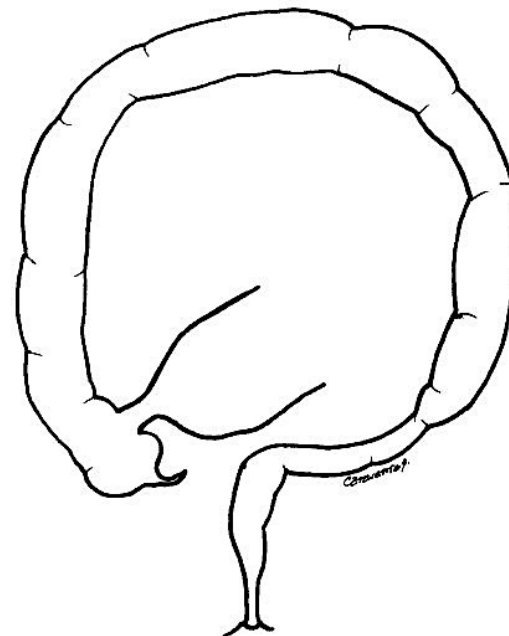
Recto-sigmoid



Long segment



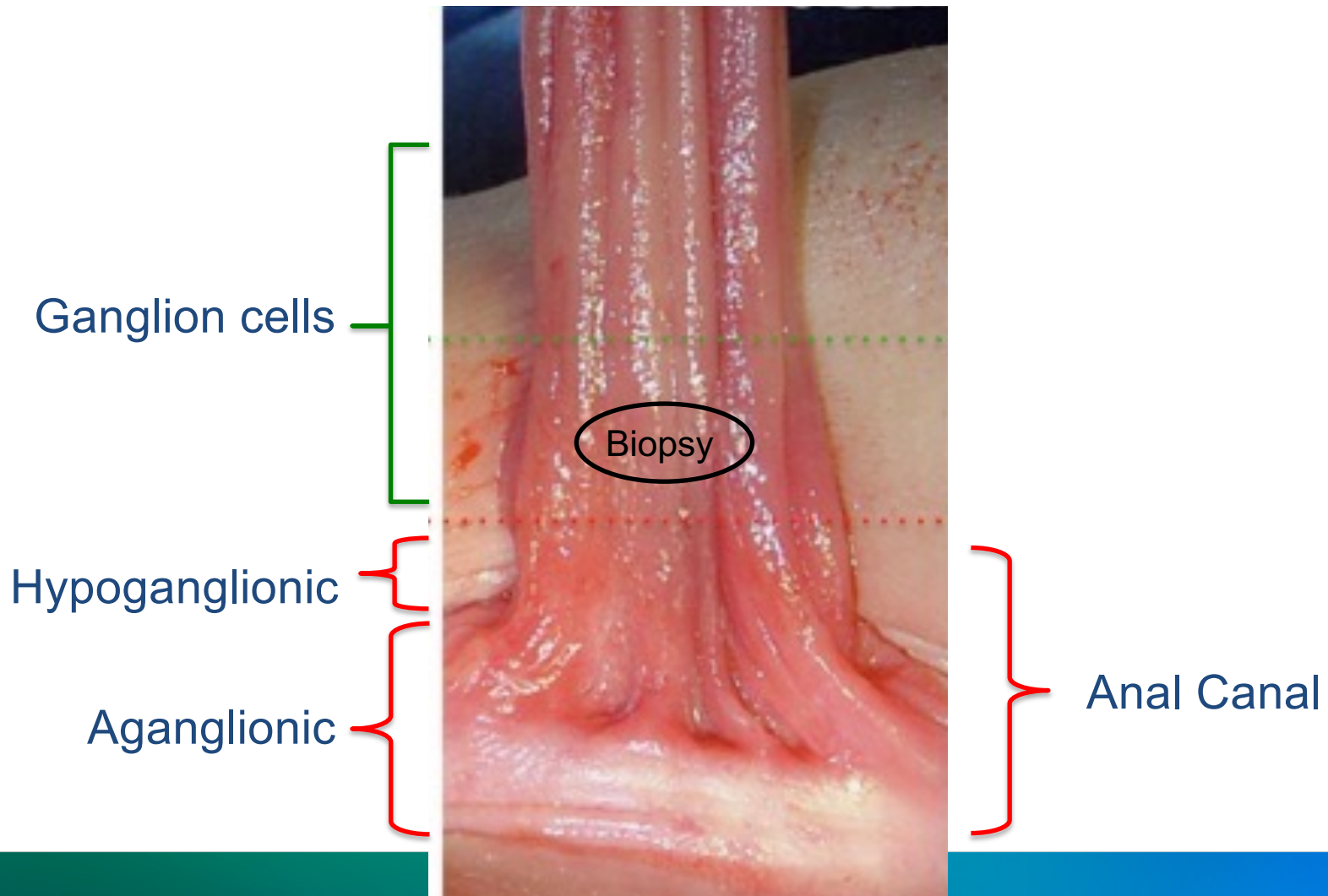
Total Colonic
Aganglionosis





- Rectal biopsy (full thickness vs. suction biopsy)
- Contrast enema

Where to obtain the biopsy?



Suction Rectal Biopsy

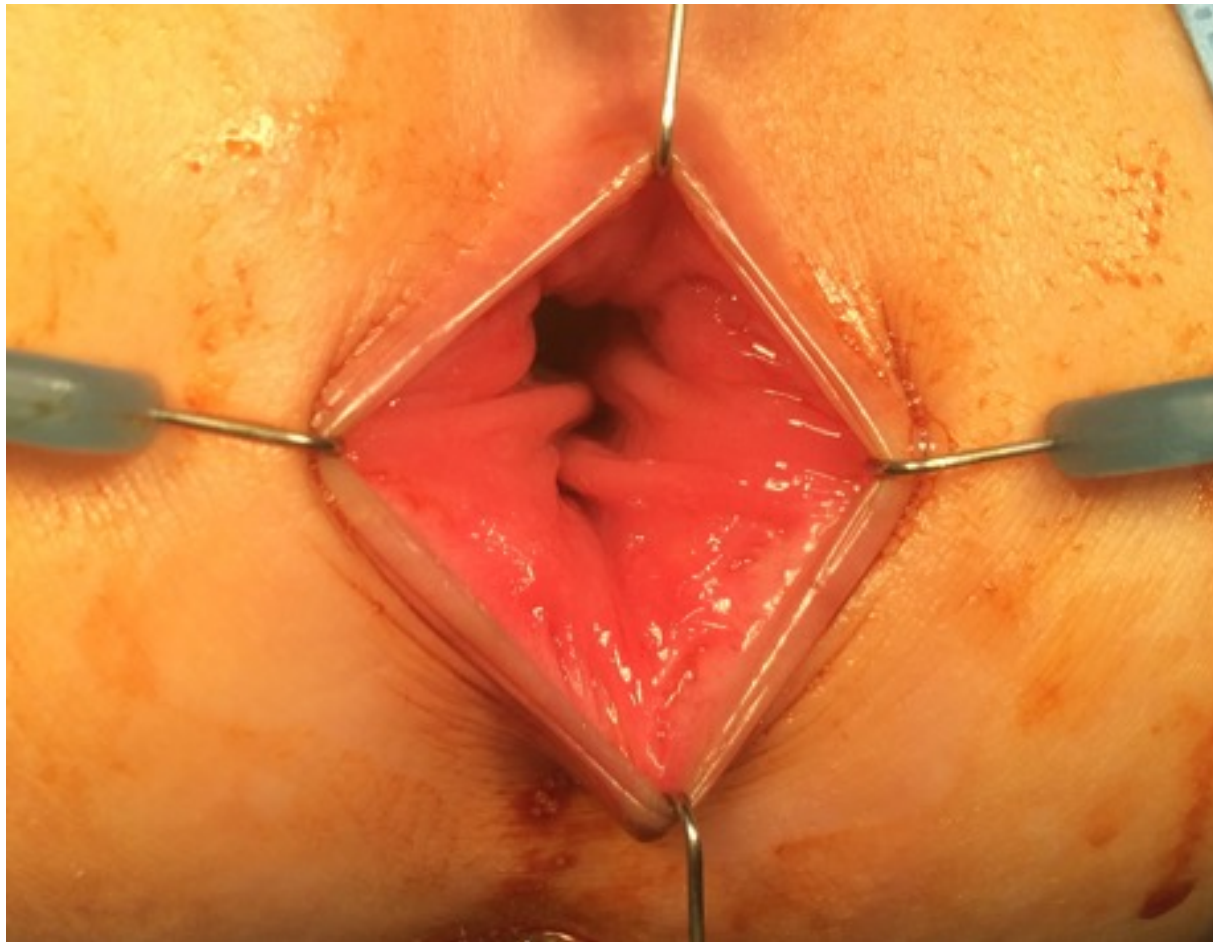


Noblett

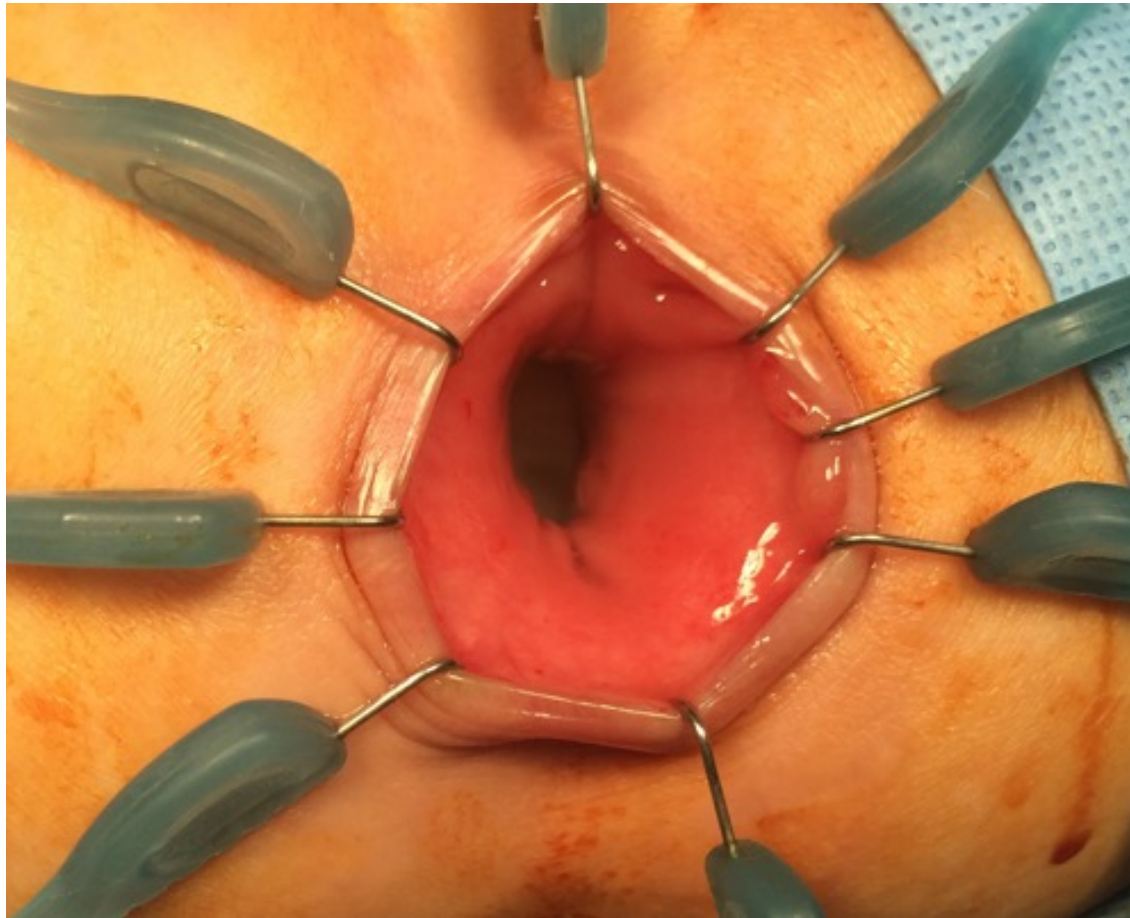


rbi2

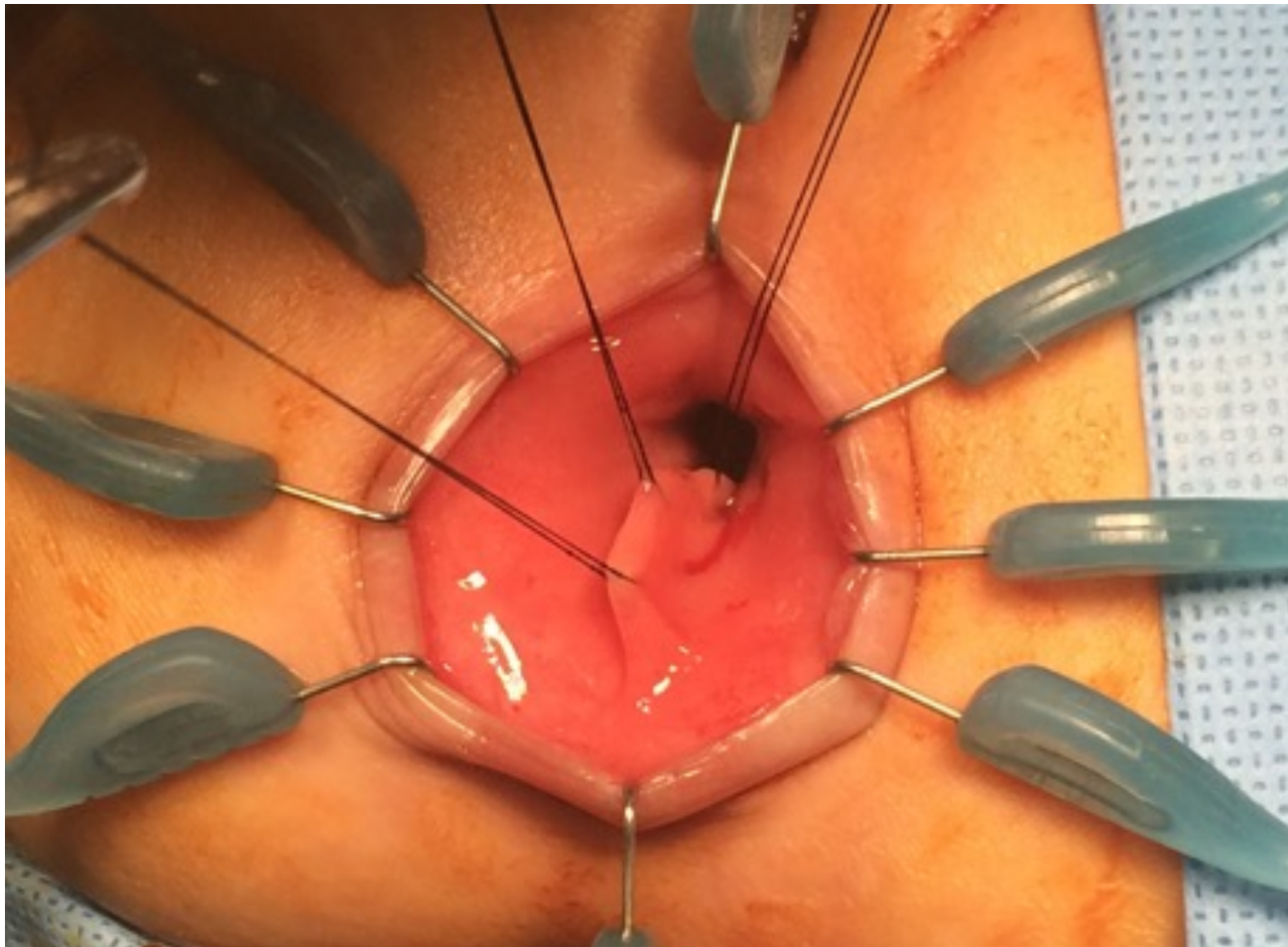
Full Thickness Rectal Biopsy



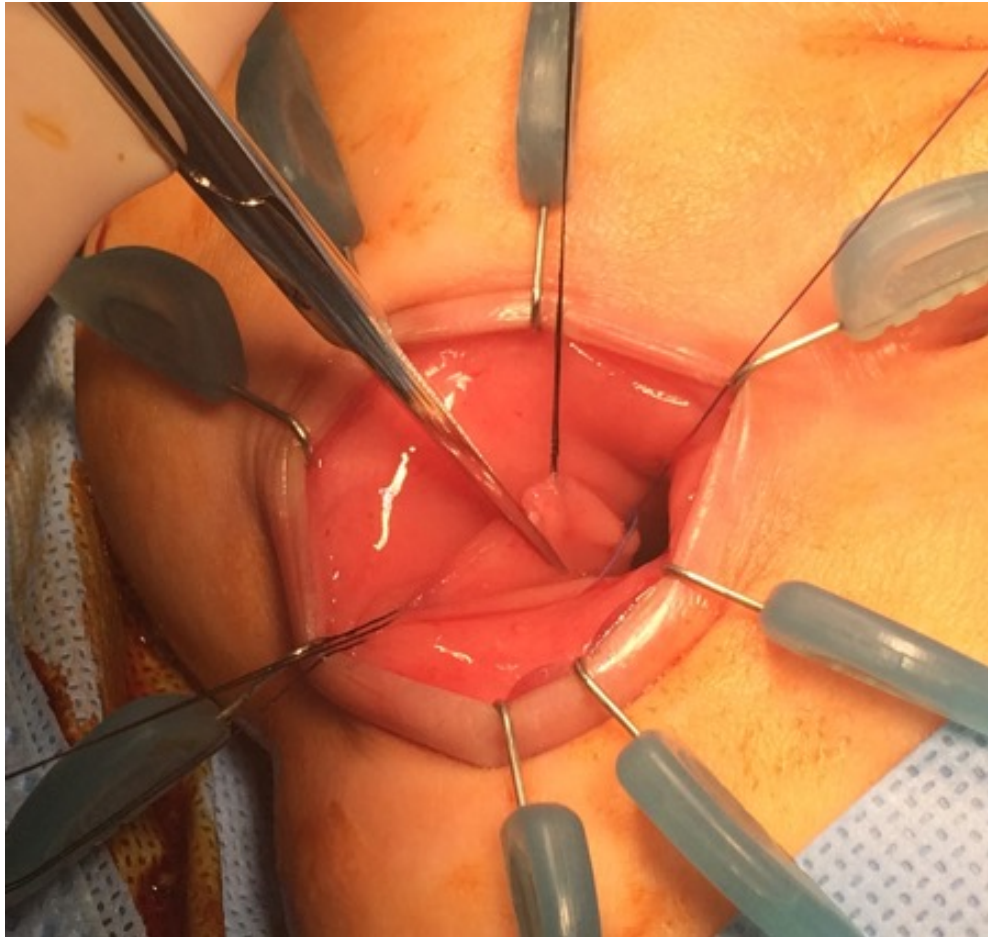
Full Thickness Rectal Biopsy



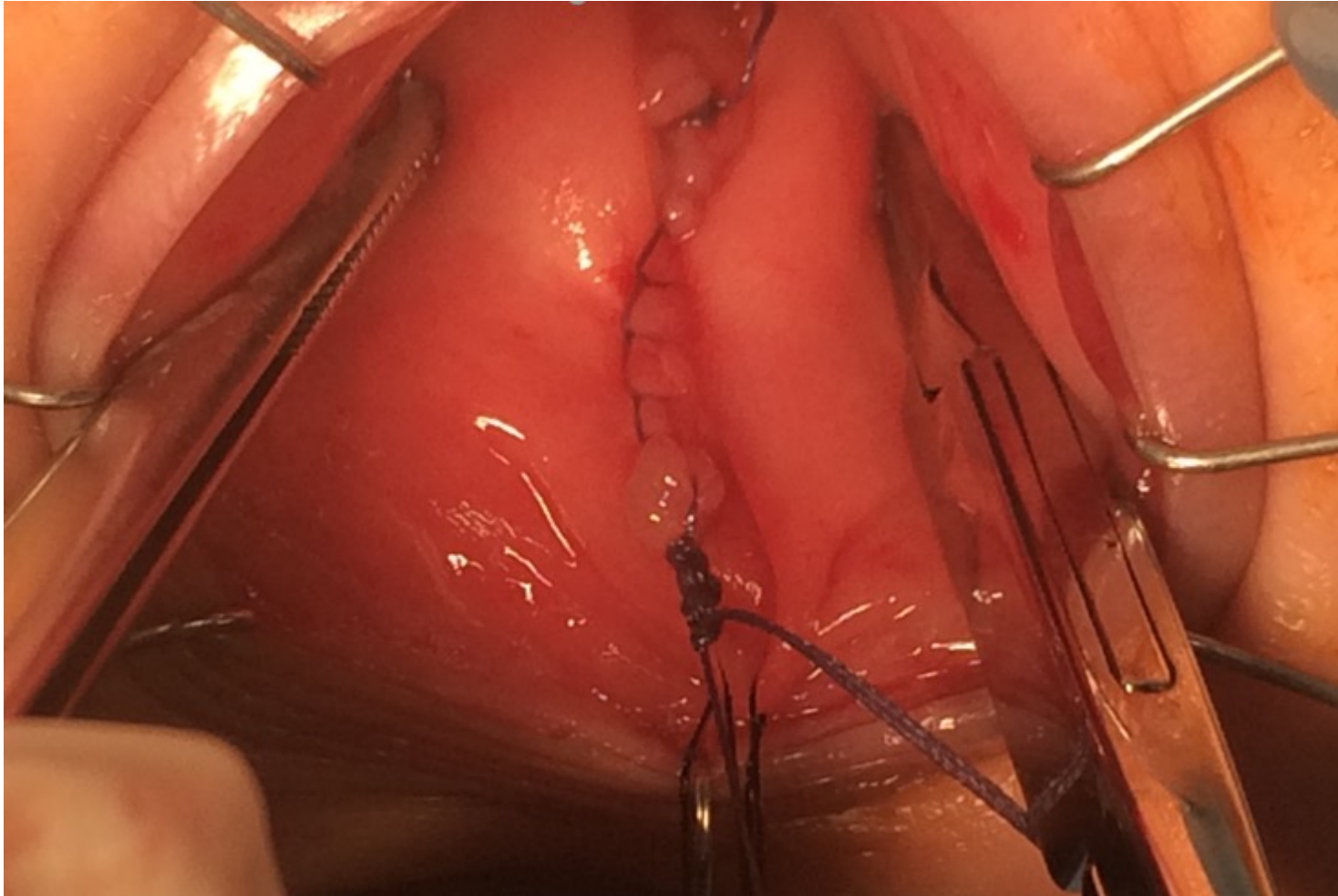
Full Thickness Rectal Biopsy



Full Thickness Rectal Biopsy



Full Thickness Rectal Biopsy





- Colostomy vs.
Primary delayed
repair



- How is the baby?
- How is the baby responding to rectal irrigations?
- How far from the Hospital do parents live?
- What is your surgical experience in transanal neonatal surgery?
- Do you have good and experienced pathologist for intra-operative frozen section diagnosis?



- Right transverse colostomy with mucous fistula and leveling biopsies



- 85 days old, female patient with Trisomy 21, presented to the emergency room with abdominal distension and emesis.

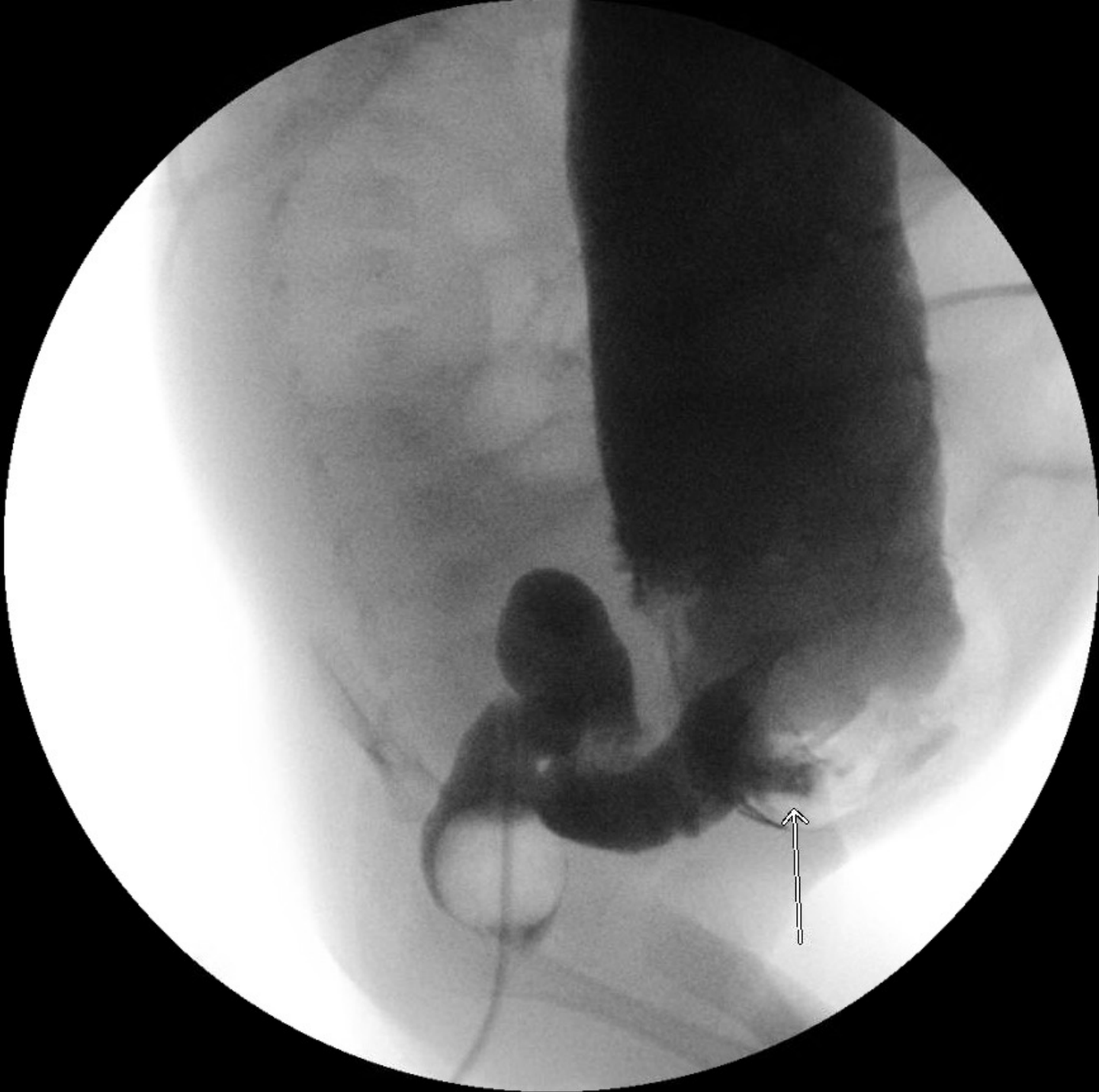


- First bowel movement happened at 7 days of life. Contrast enema at that time led to the diagnosis of meconium plug.



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- Due to malnourishment and severe bowel distension, a laparoscopic right transverse colostomy with mucous fistula was done.





- After 3 months, the contrast enema was repeated.

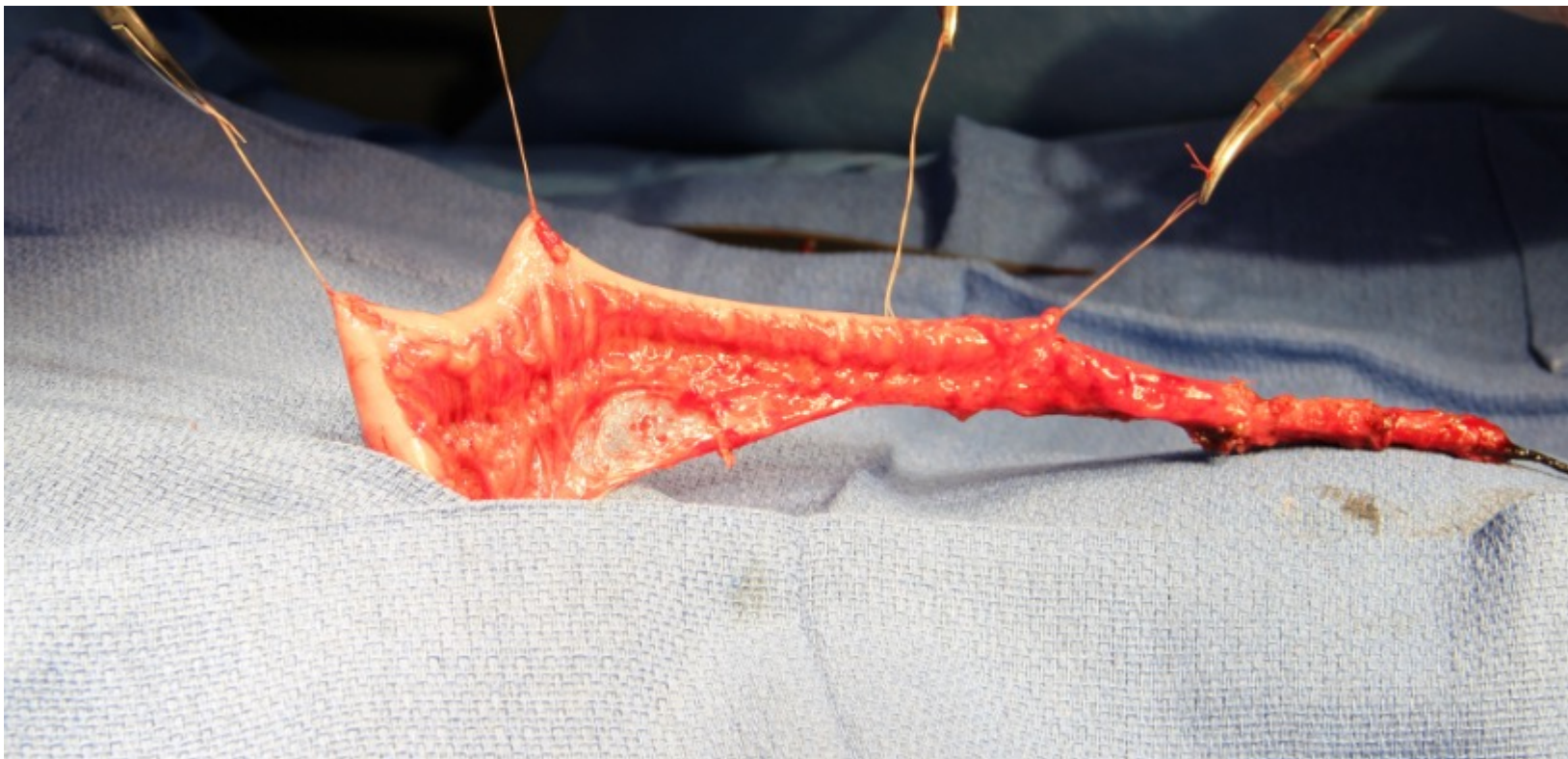


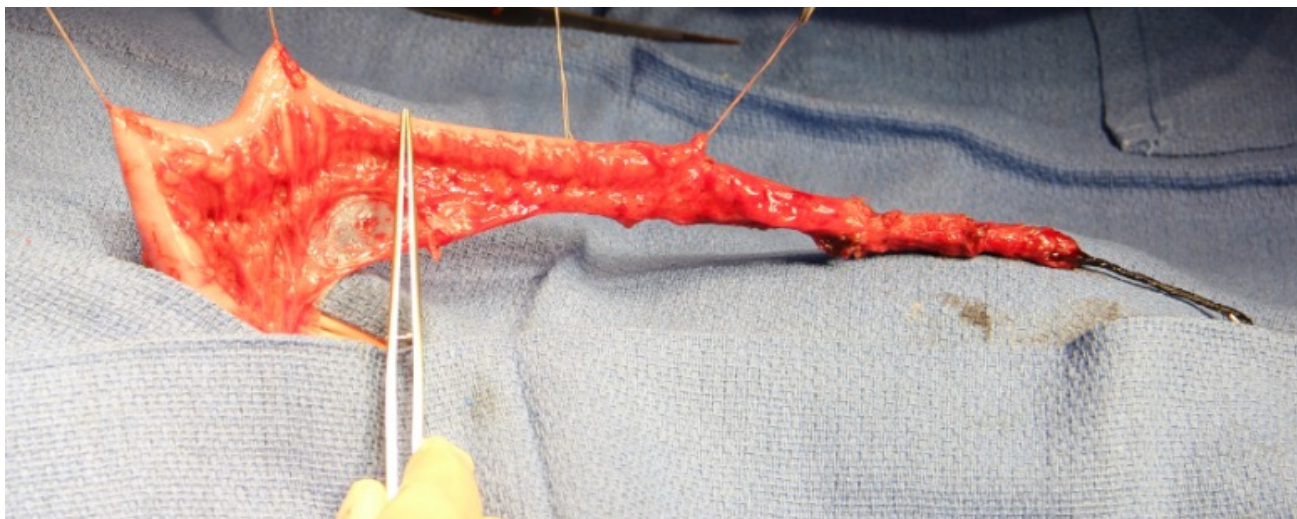
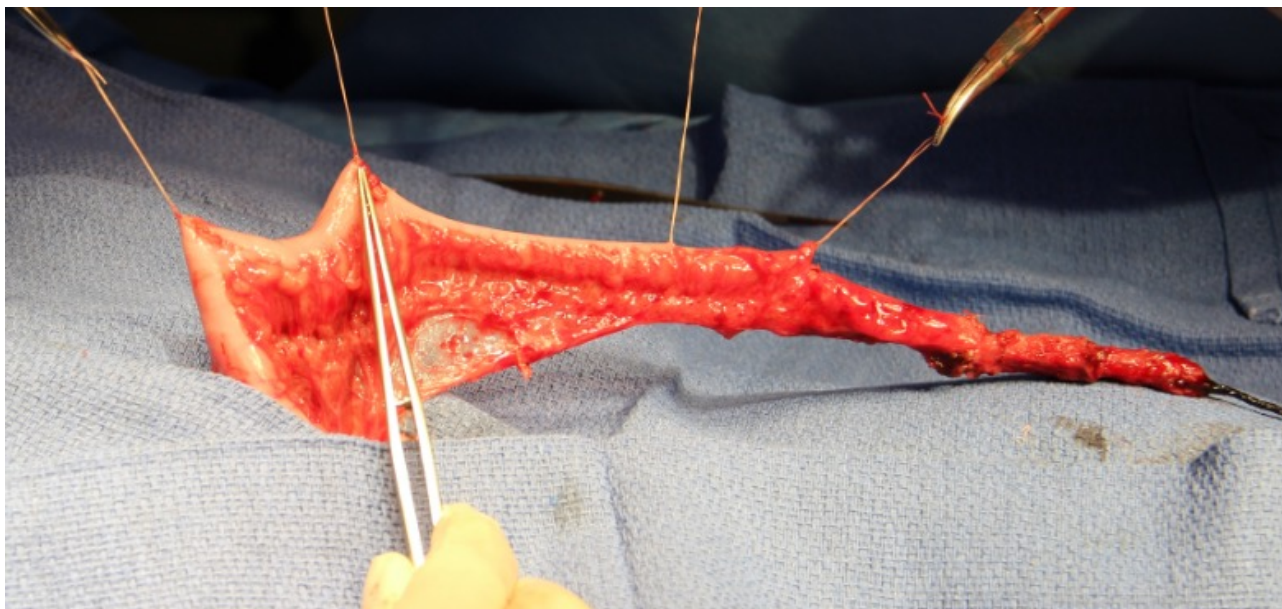
















- Patient is now 4 yo and is toilet trained for urine and stool.
- She is taking senna (30 mg) daily.



- Term male newborn, around 20 hours of life started refusing feeds and developed abdominal distension.
- Had not passed meconium yet.



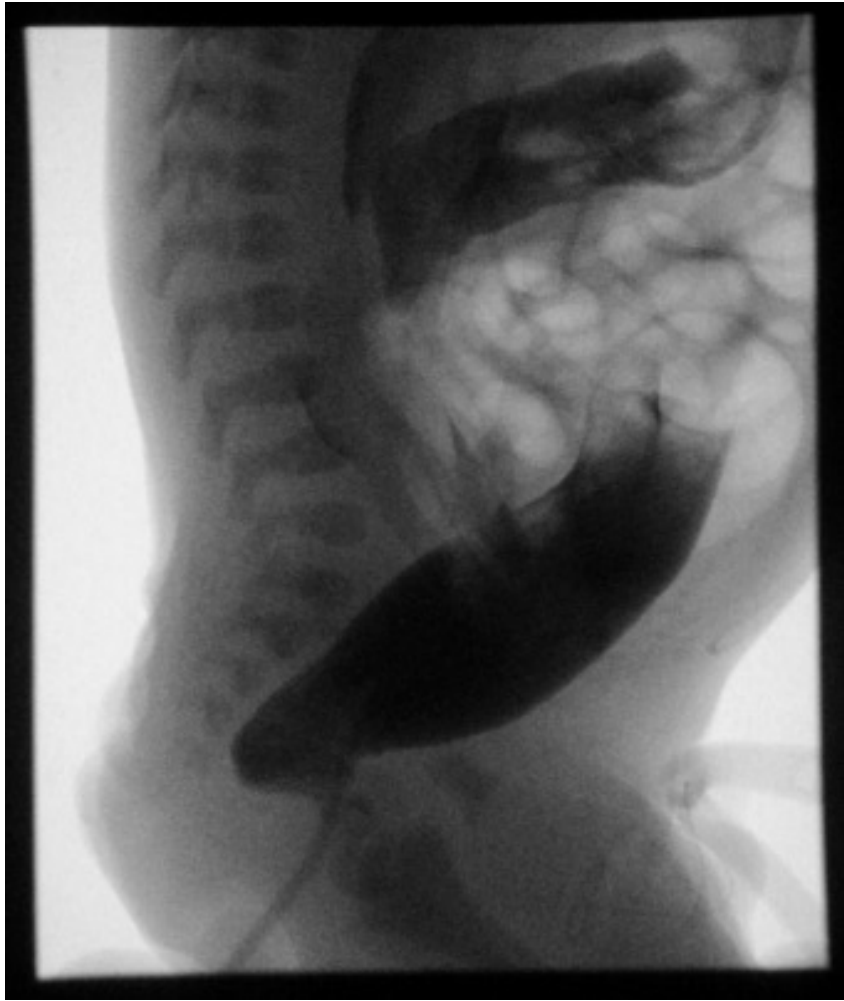
JV



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- NPO
- Rectal irrigations were started
- Flagyl was started
- Suction rectal biopsy and contrast enema done





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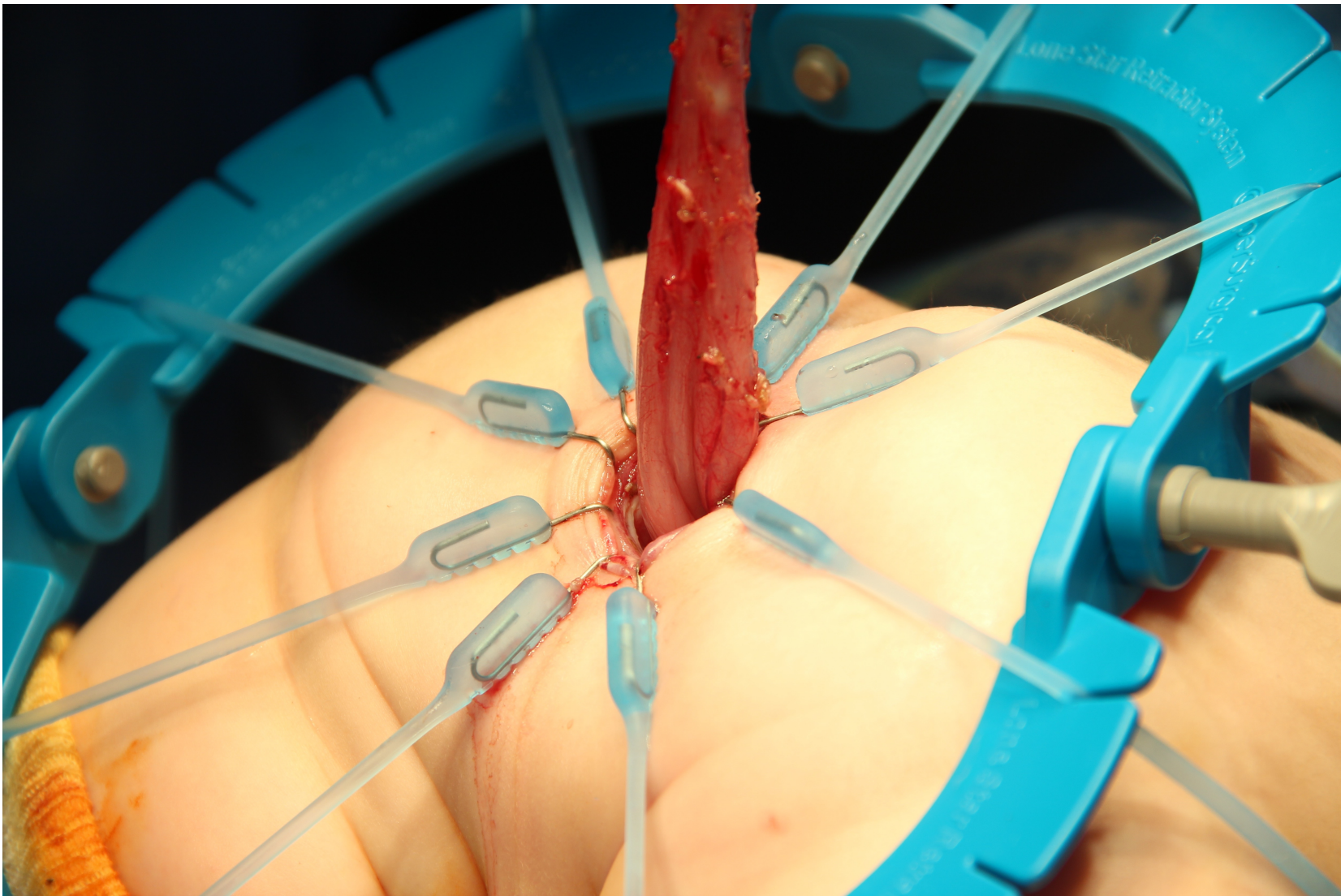
- Suction rectal biopsy with absent ganglion cells and hypertrophic nerves.
- Patient was discharged on rectal irrigations 3x/day.



- At 2 months of age he underwent pullthrough for Hirschsprung.



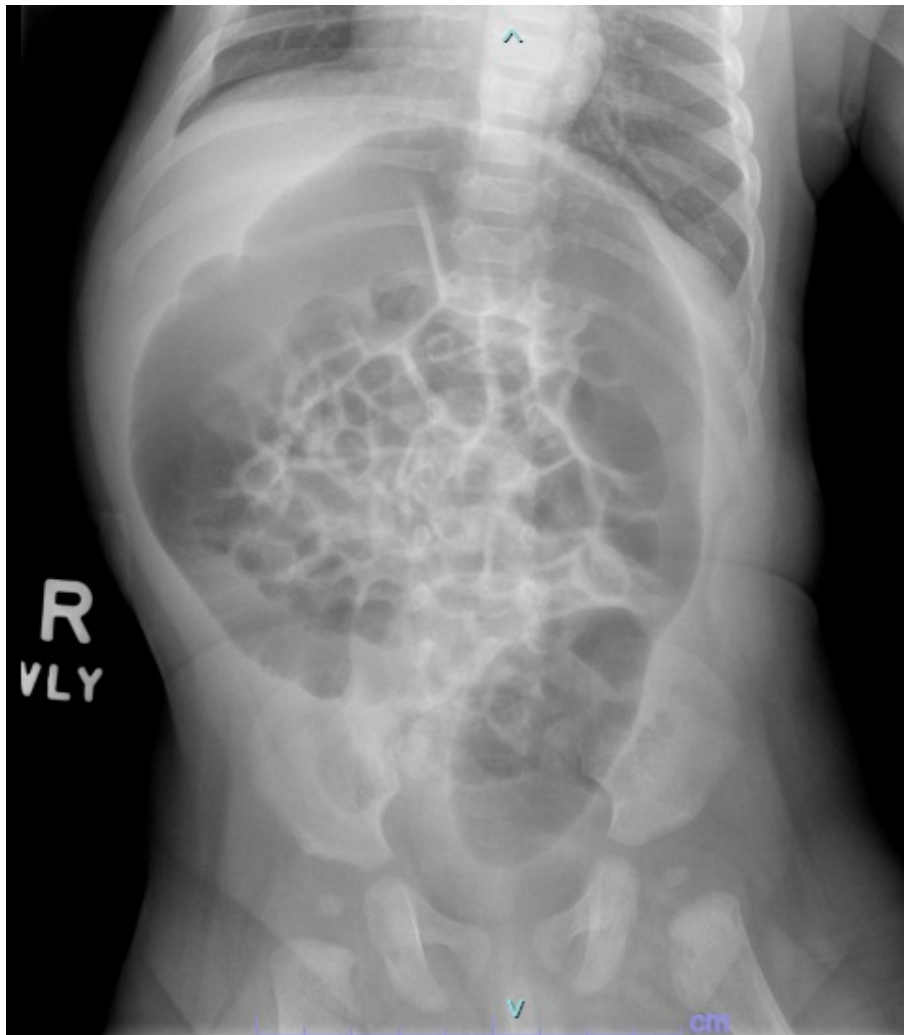
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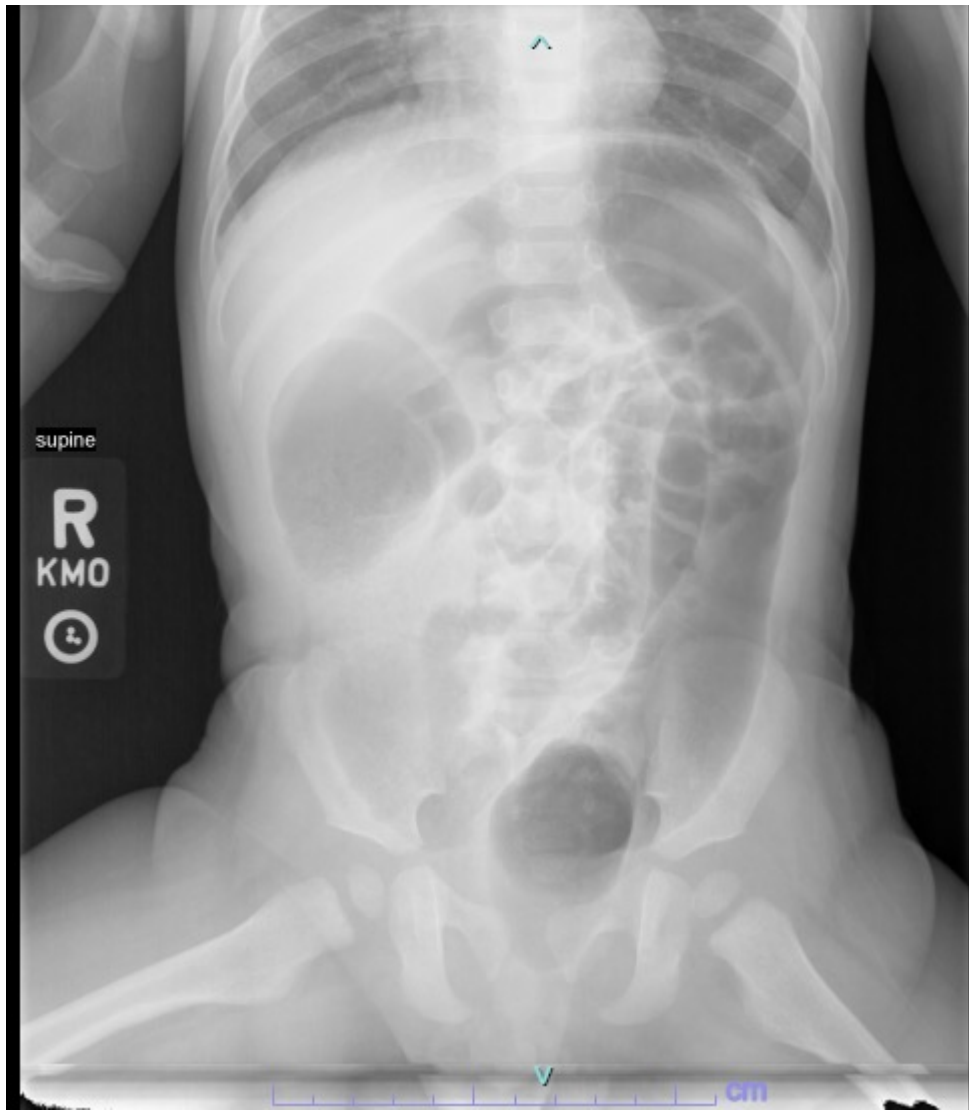
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- Several episodes of enterocolitis during the first year of life.









- Patient is now 5 yo and has one voluntary bowel movement a day.



- 8 months old, male patient.
- He did not pass meconium within the first 24 hours of life, received the diagnosis of meconium plug.



- Did relatively well with breast milk and glycerin suppositories.
- Once solid foods were started, stooling on every two weeks.

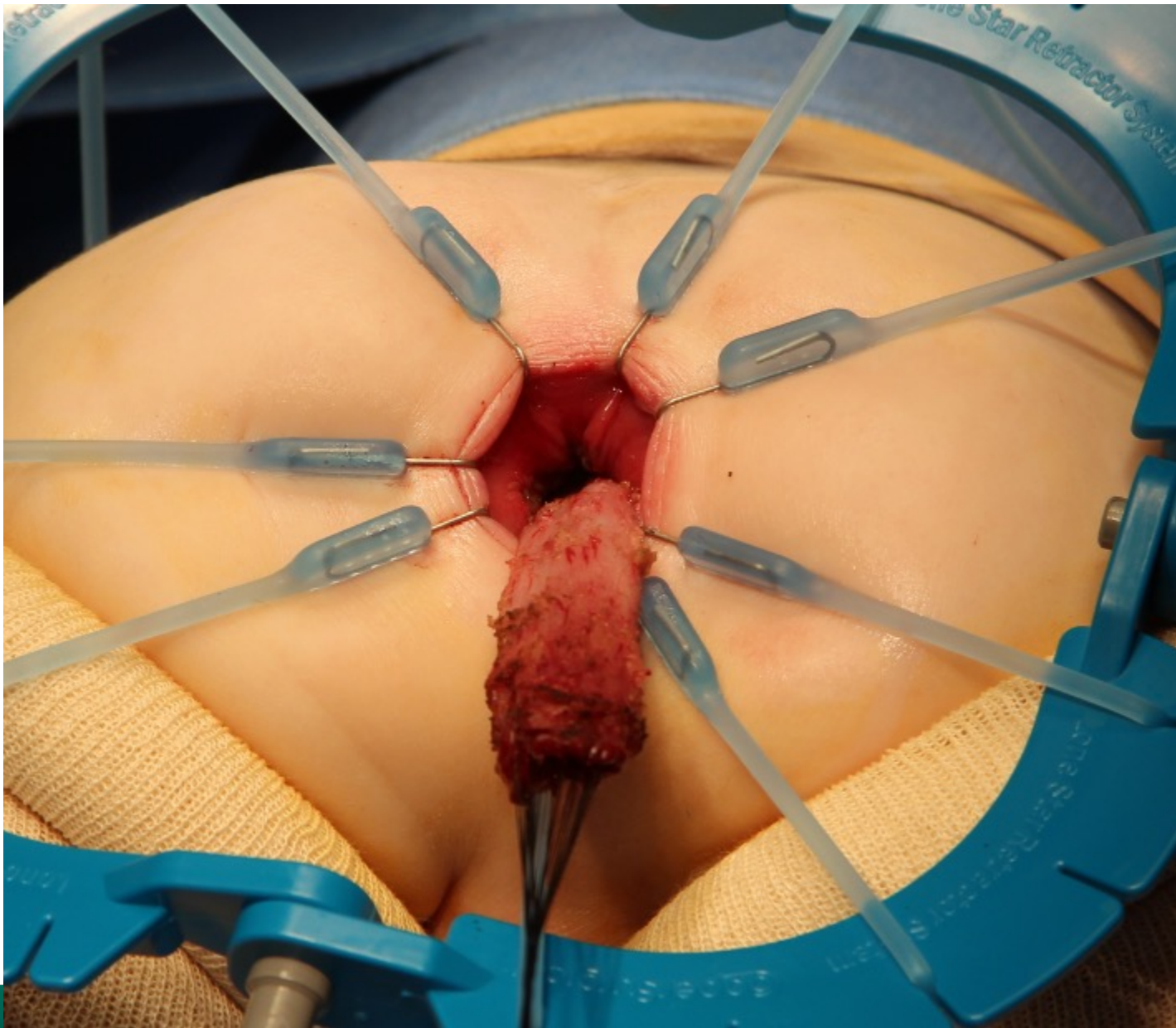


- The power of mothers

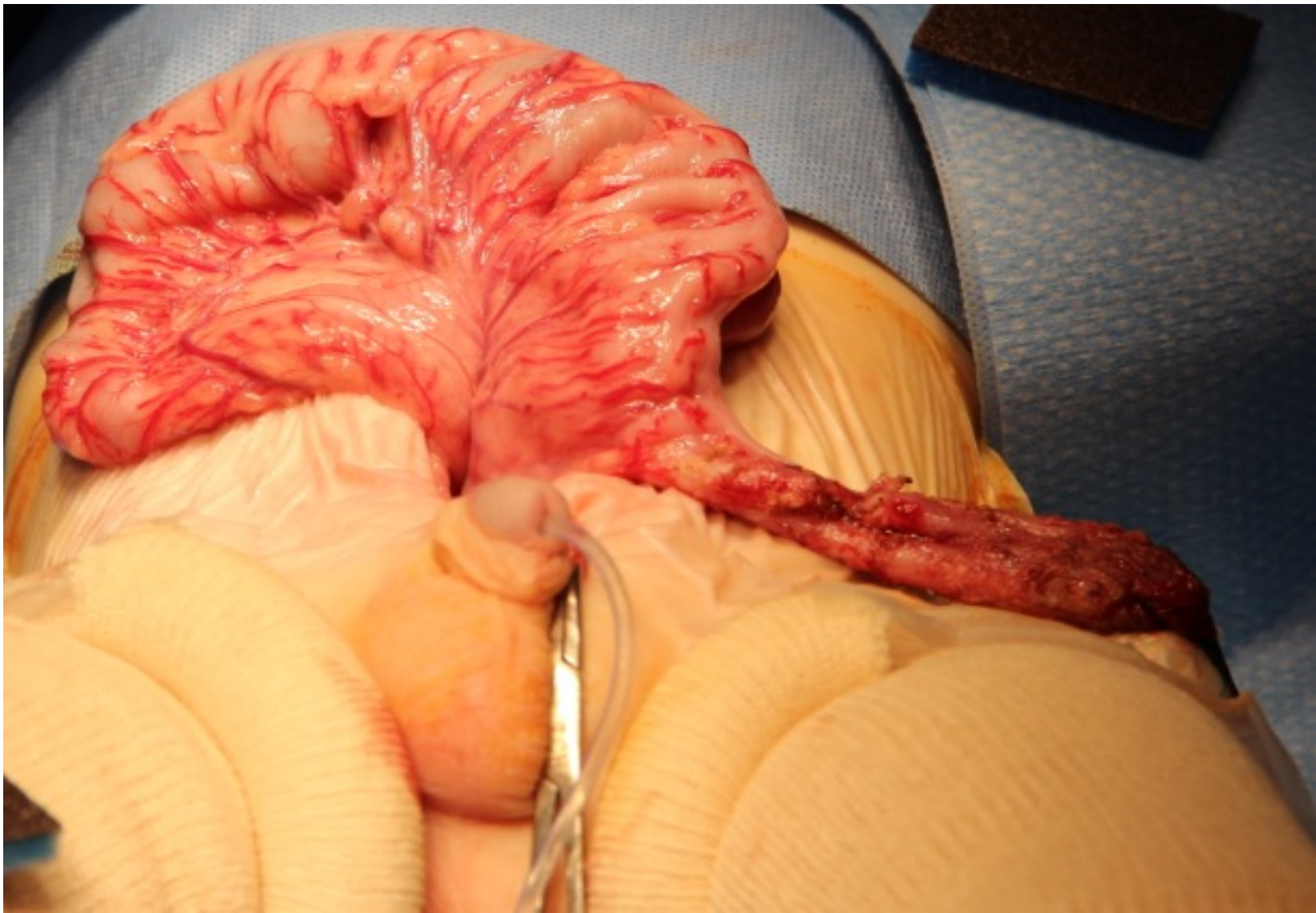


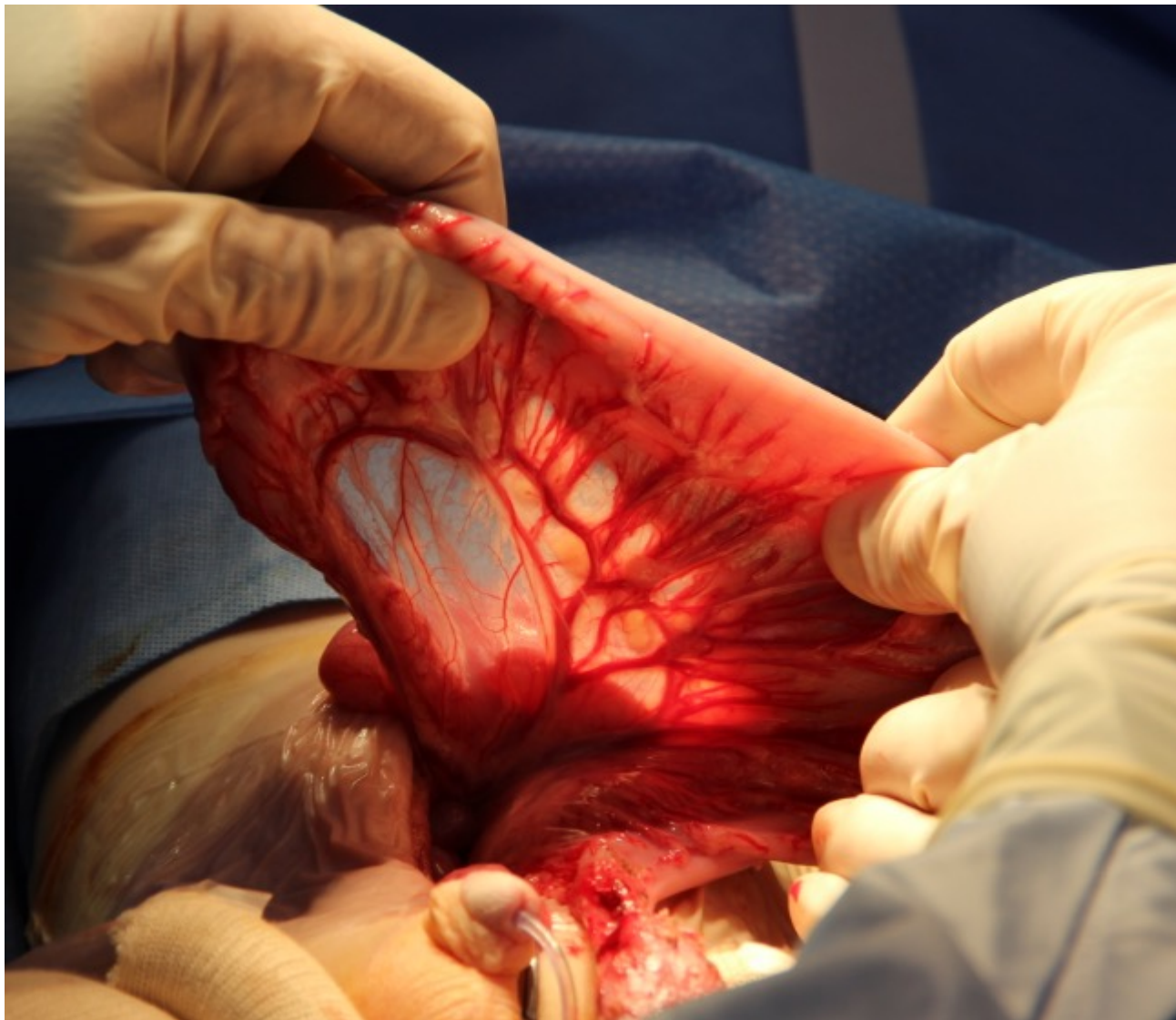
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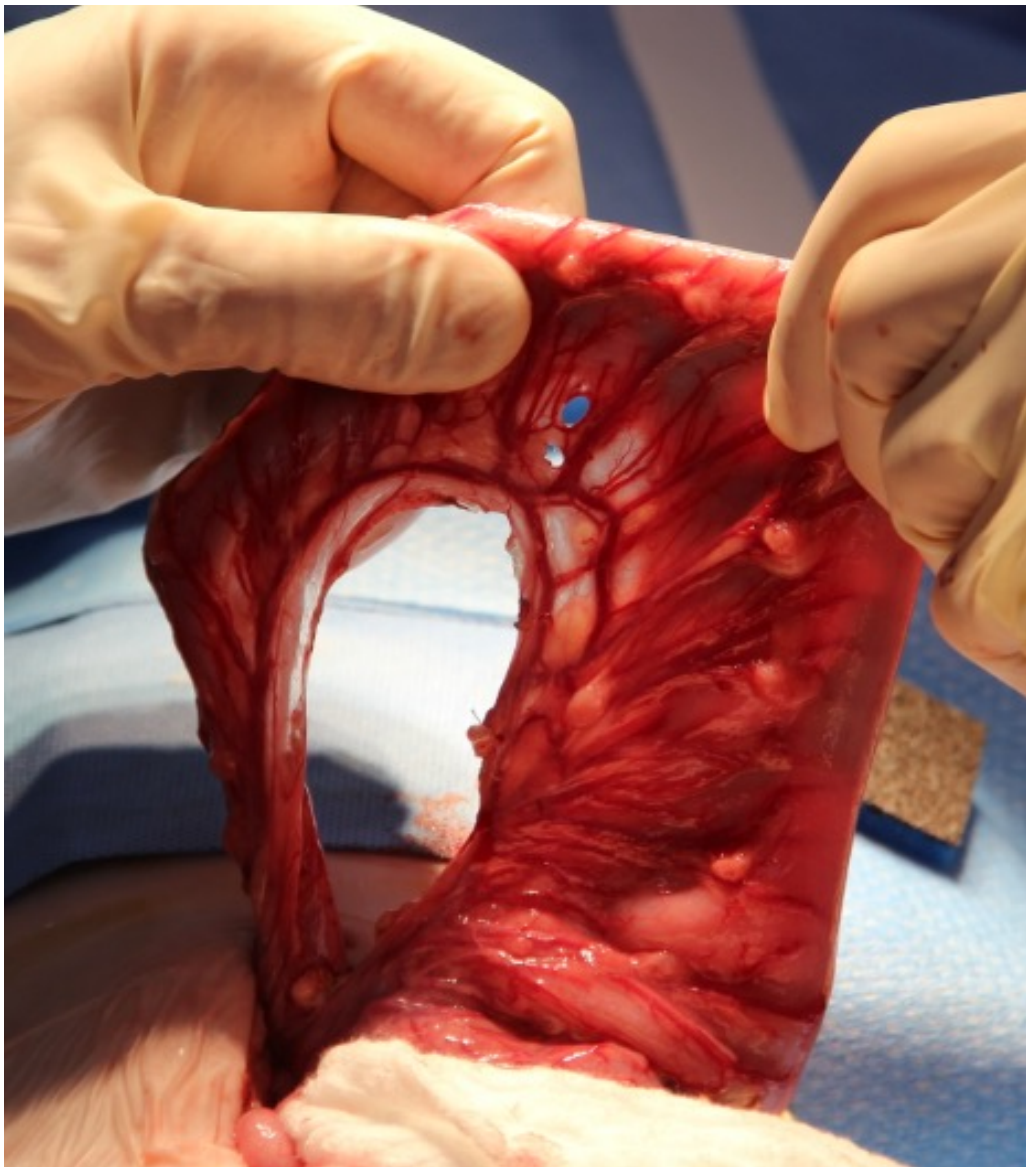


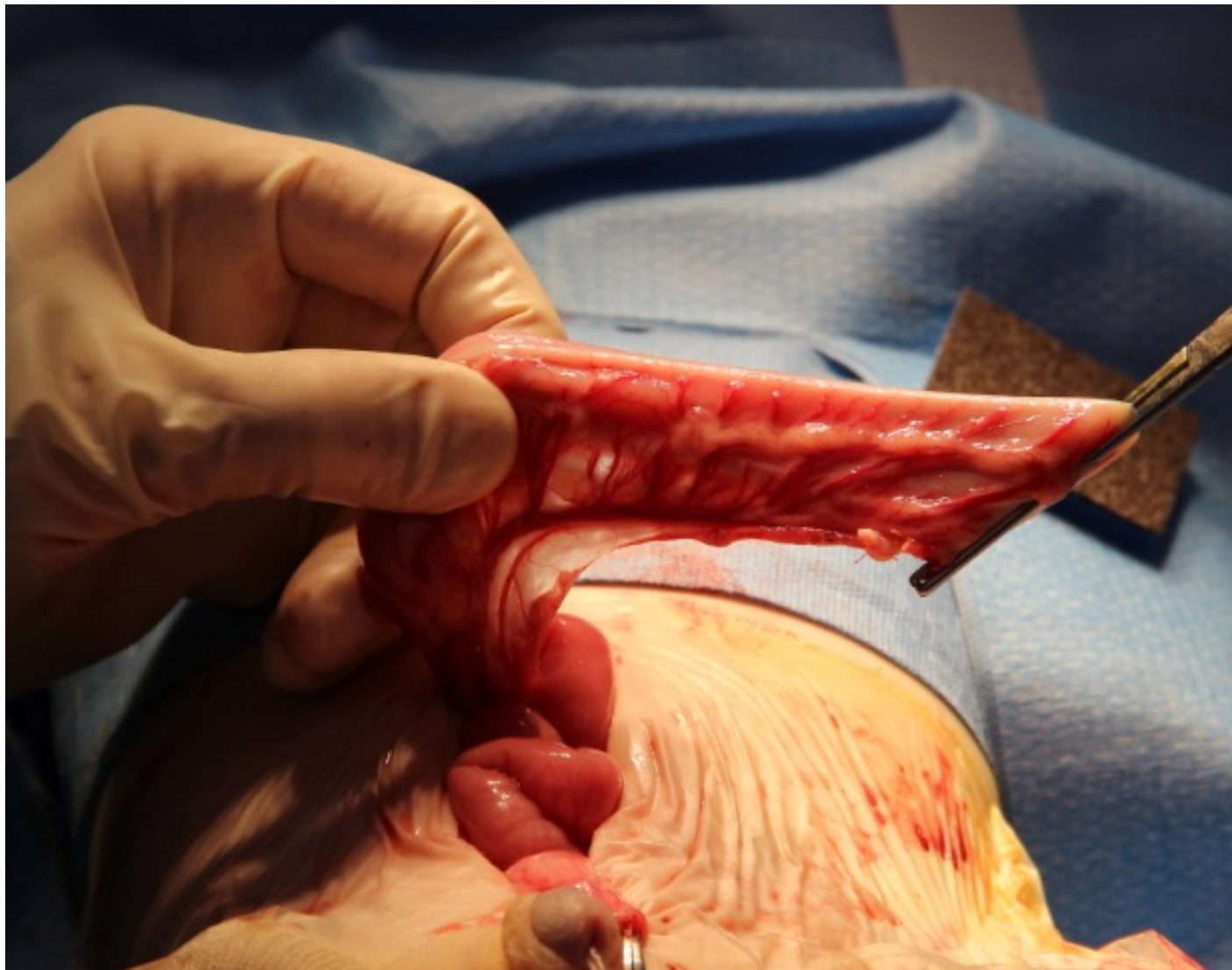
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- 39 days old female patient with rectal biopsy during the first week of life diagnosing her with Hirschsprung disease



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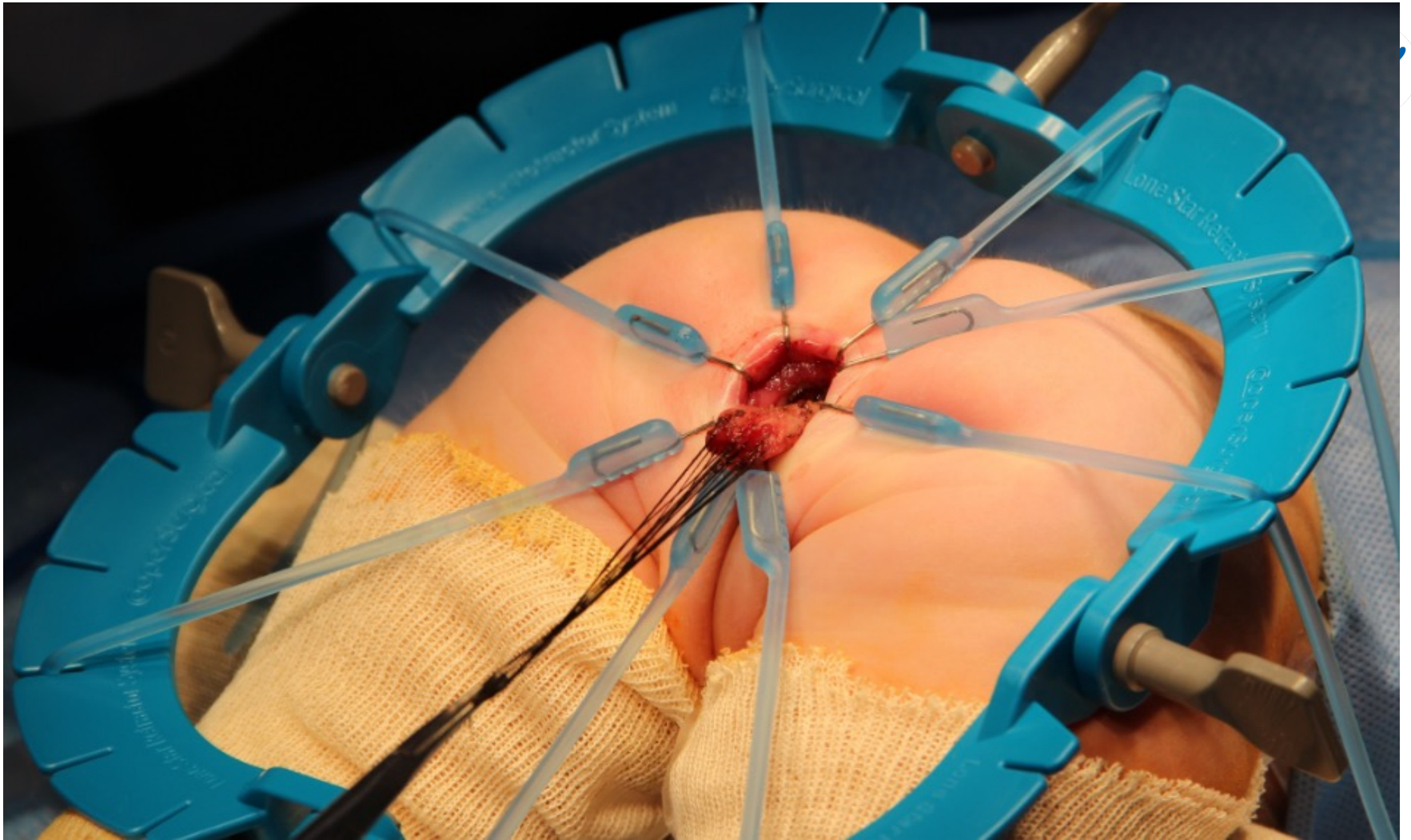
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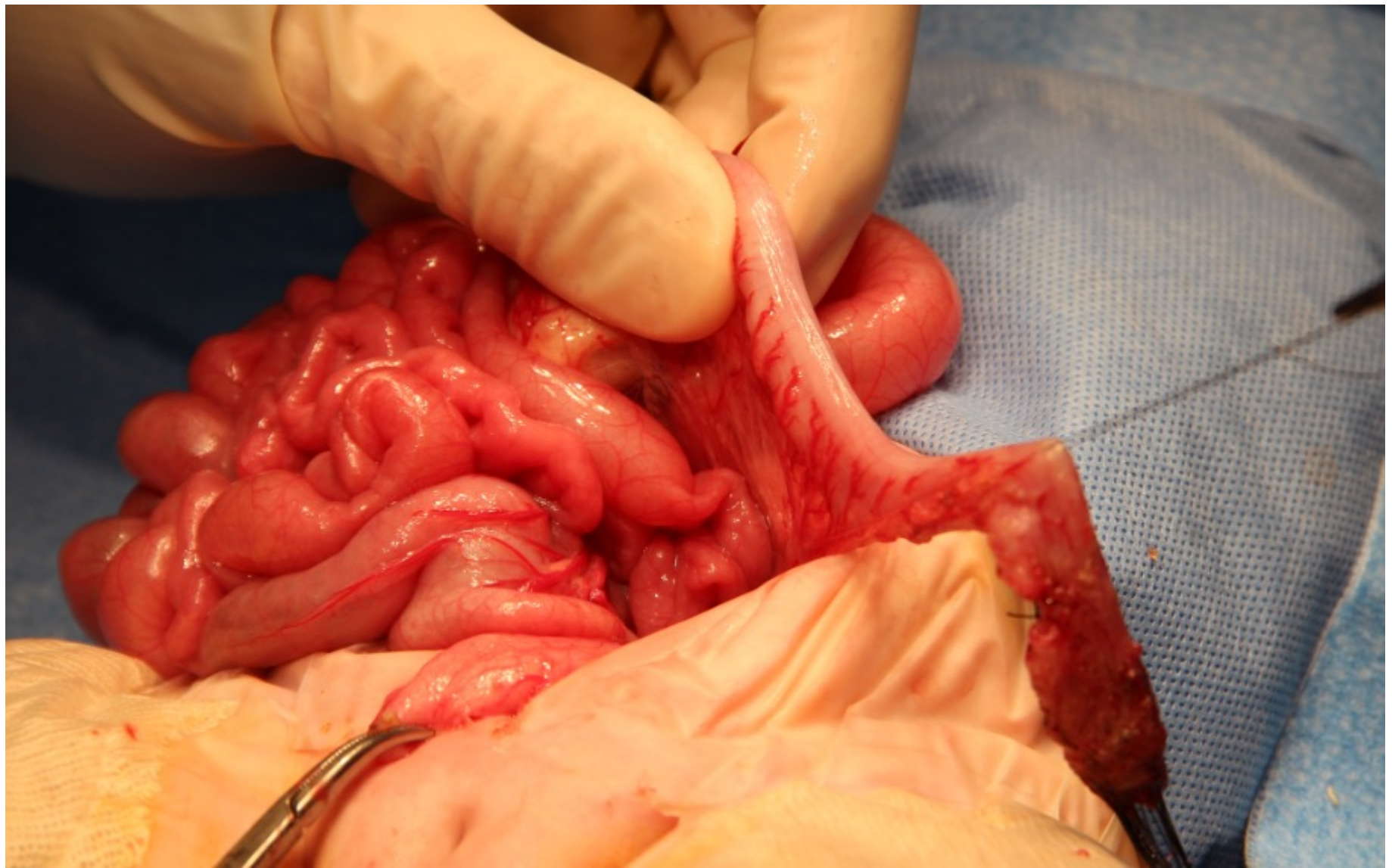


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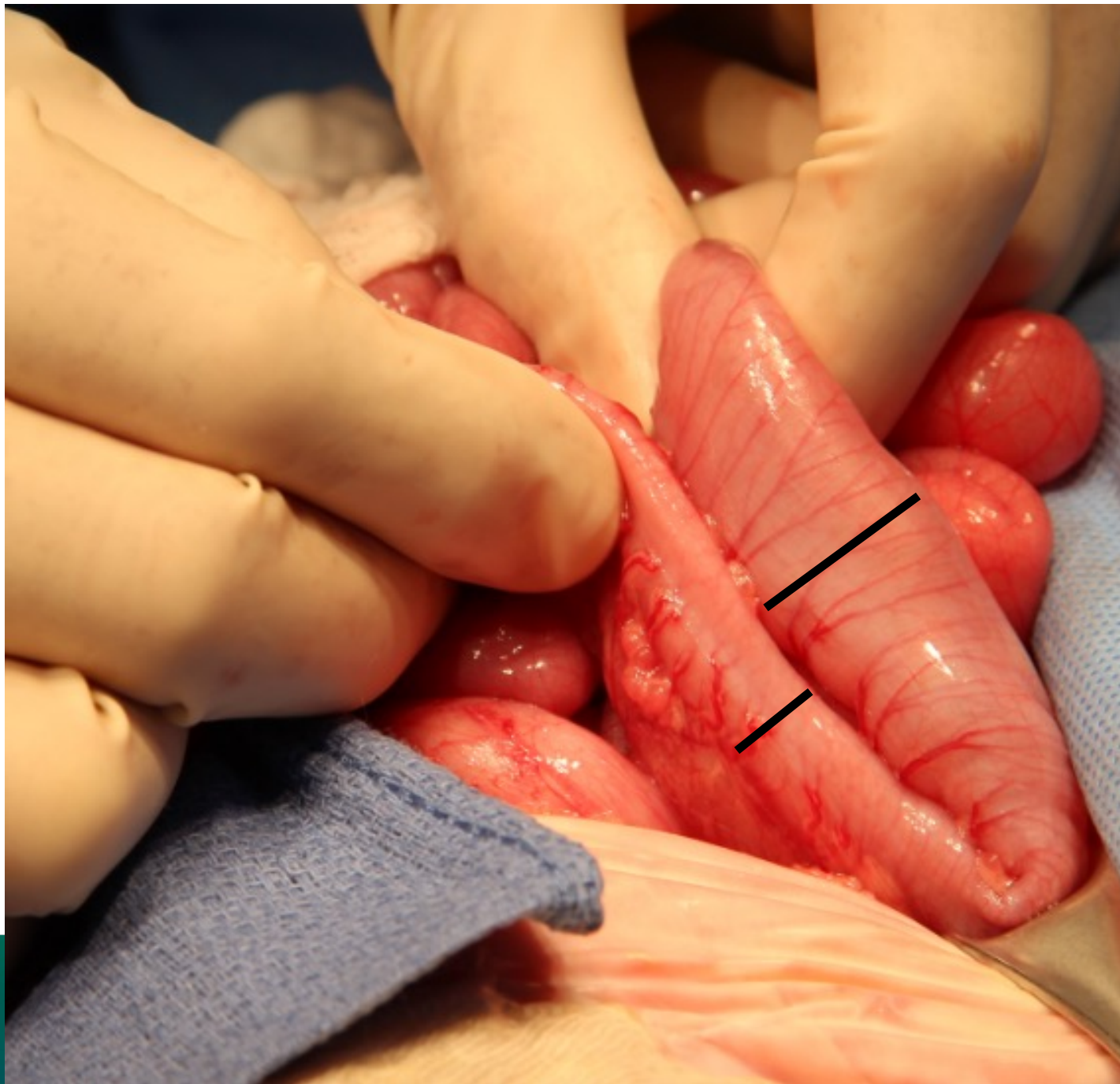


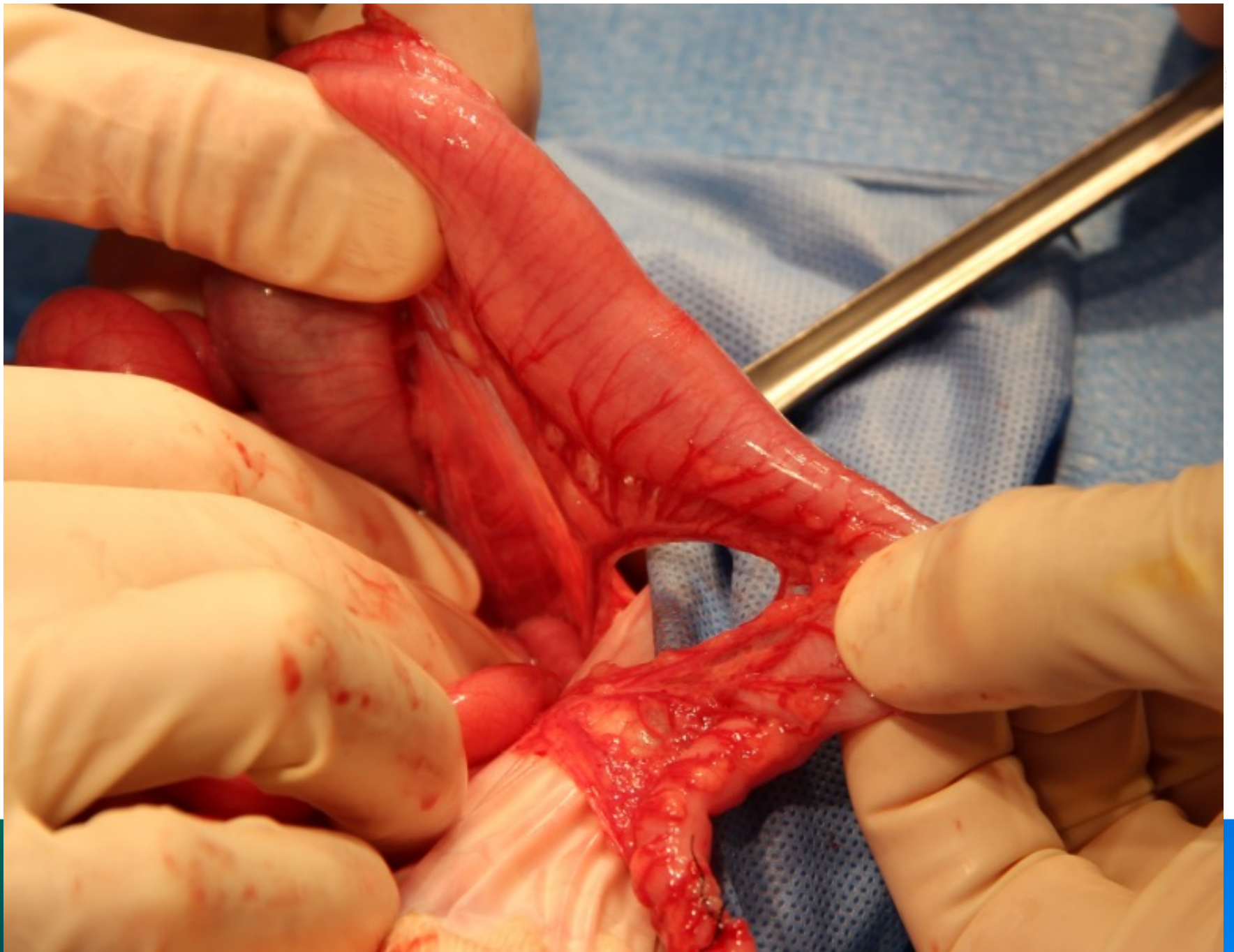


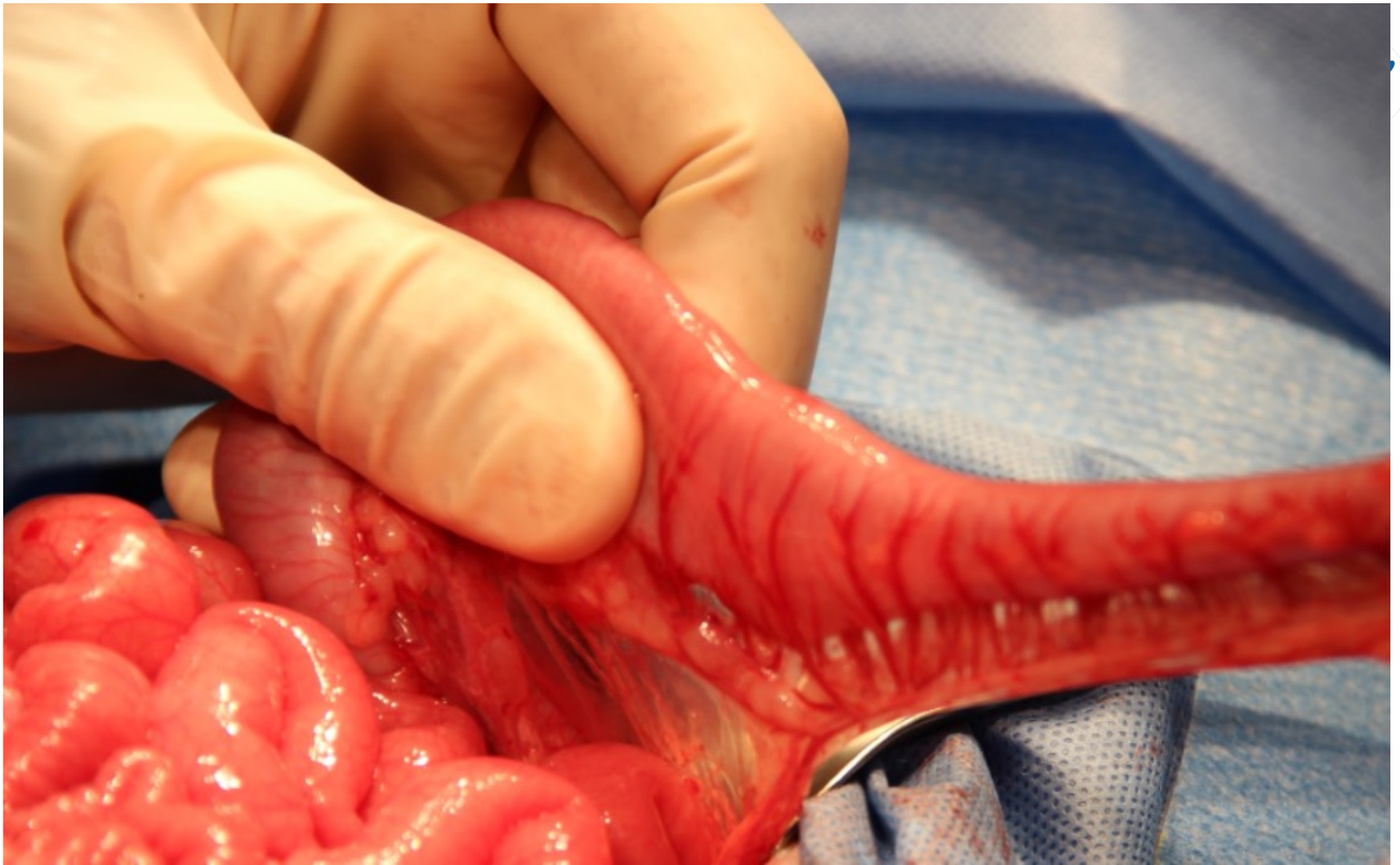
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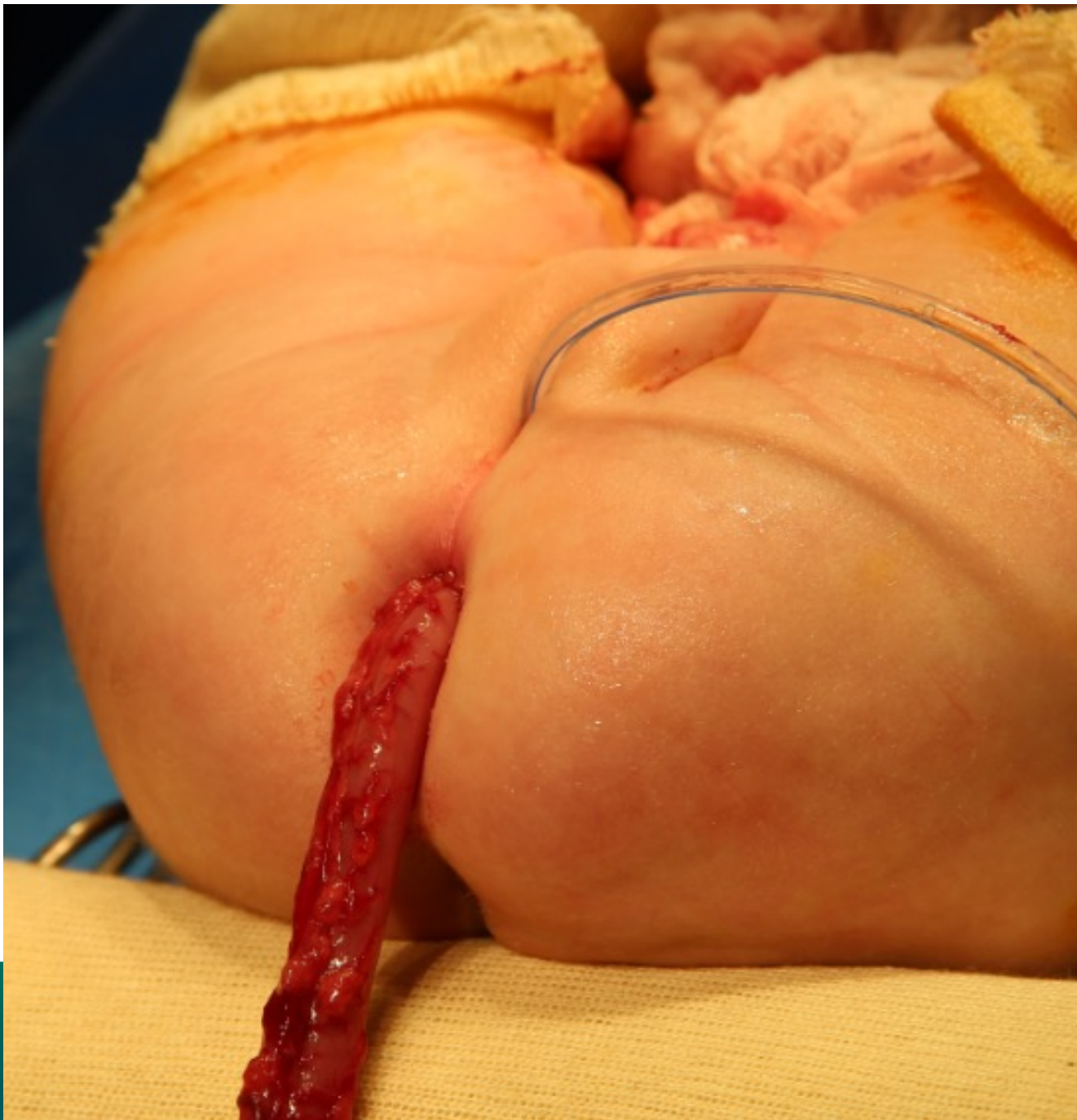
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7 days post op





- What are the chances of future siblings having Hirschsprung or offsprings?



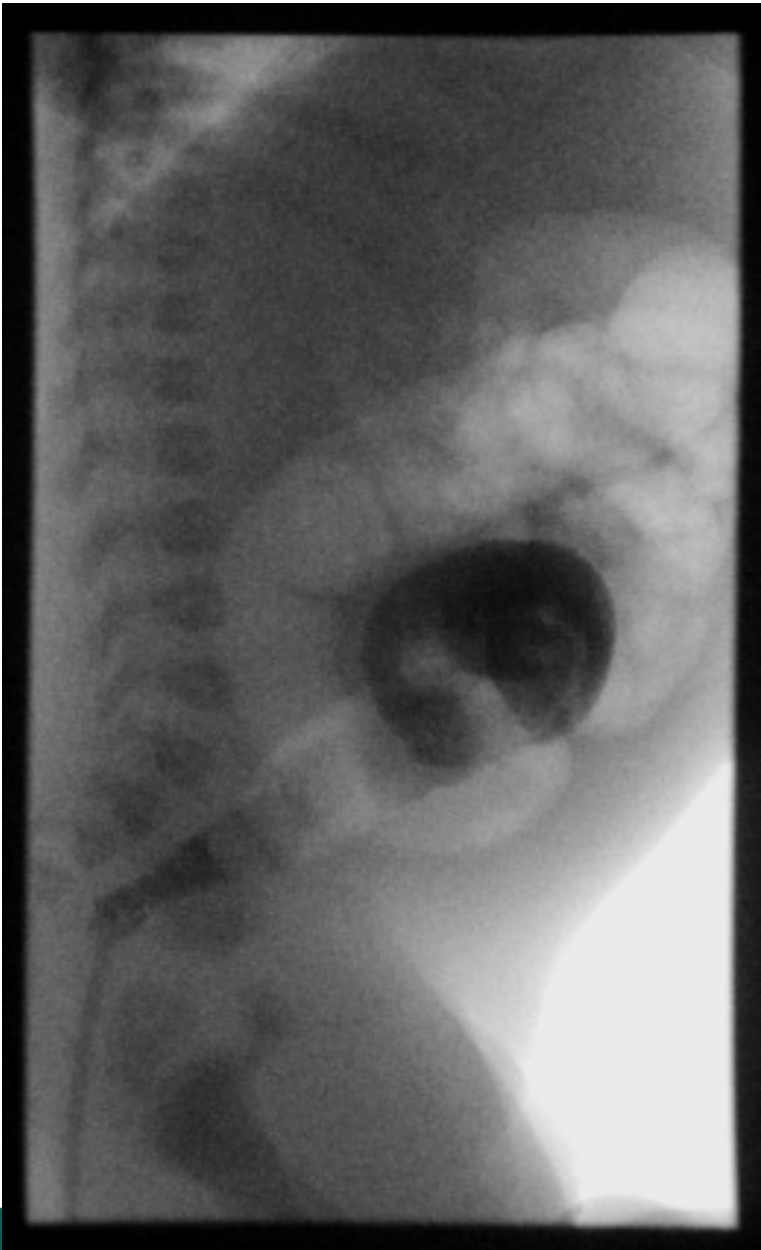
To demonstrate how to read this table, here is an example: if you have a son with long-segment HSCR, and your next child is a girl, her chance to have HSCR is 7-9%. Your son's chance to have a son with HSCR is 16-19%.

% Risk to relatives	Rectosigmoid (short segment)		Colonic-segment(descending colon)		Long segment & total colonic aganglionosis	
	Male	Female	Male	Female	Male	Female
Siblings of affected males	4-5	1	9-10	7	9-12	7-9
Siblings of affected females	5-6	1-2	12-13	10	21-24	17-19
Offspring of affected males	0-1	<1	10-11	8-9	16-19	12-14
Offspring of affected females	0-1	<1	14-15	11	27-29	21-22

Adapted from Badner et al. Am.J.Hum.Genet. 46: 569-580 (1990).



- Female patient, one day of life



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Thank you



Doctors Pena Bischoff



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