

Case #4

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INTERNATIONAL CENTER FOR
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39-days-old female patient with rectal biopsy diagnosis of Hirschsprung disease





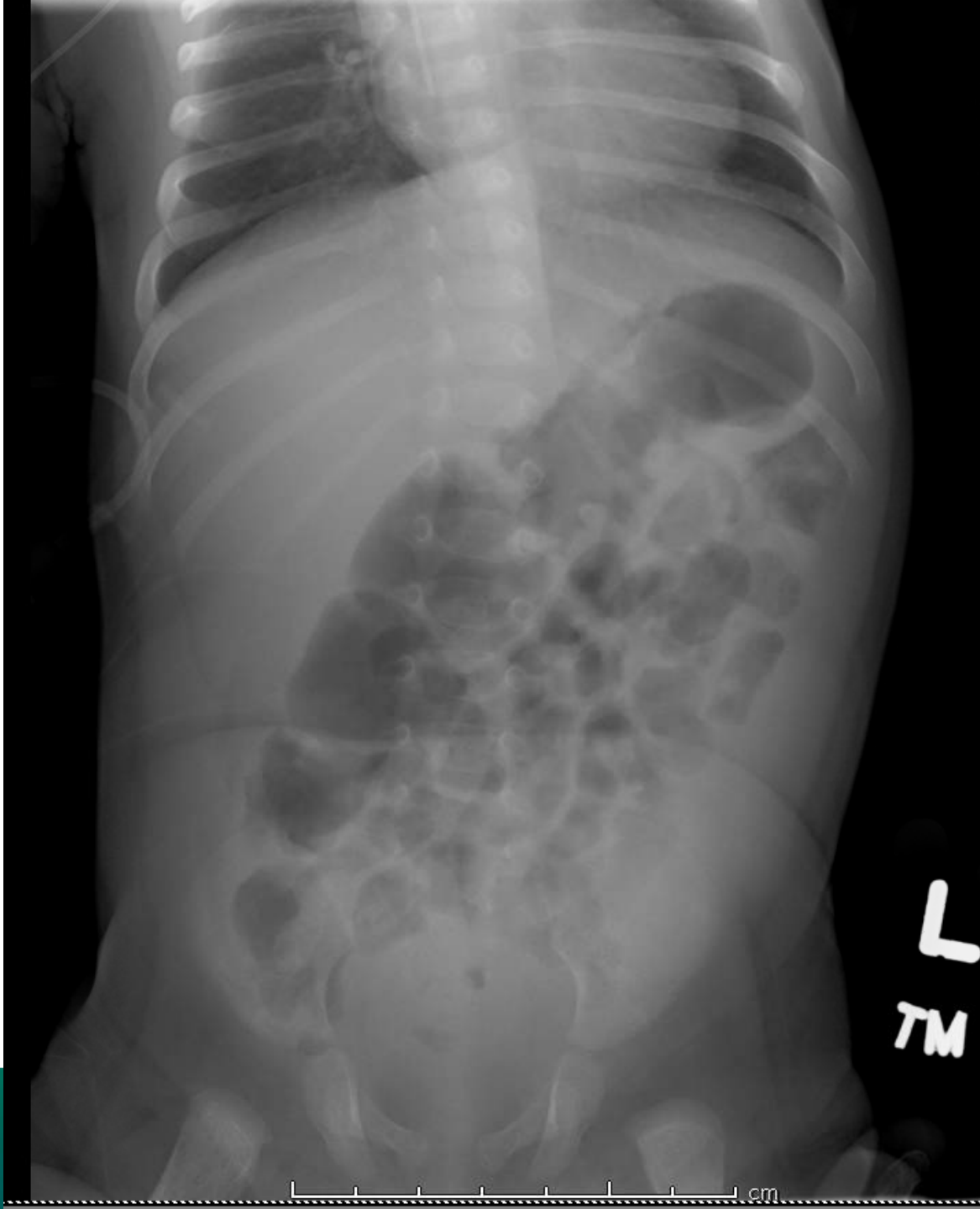


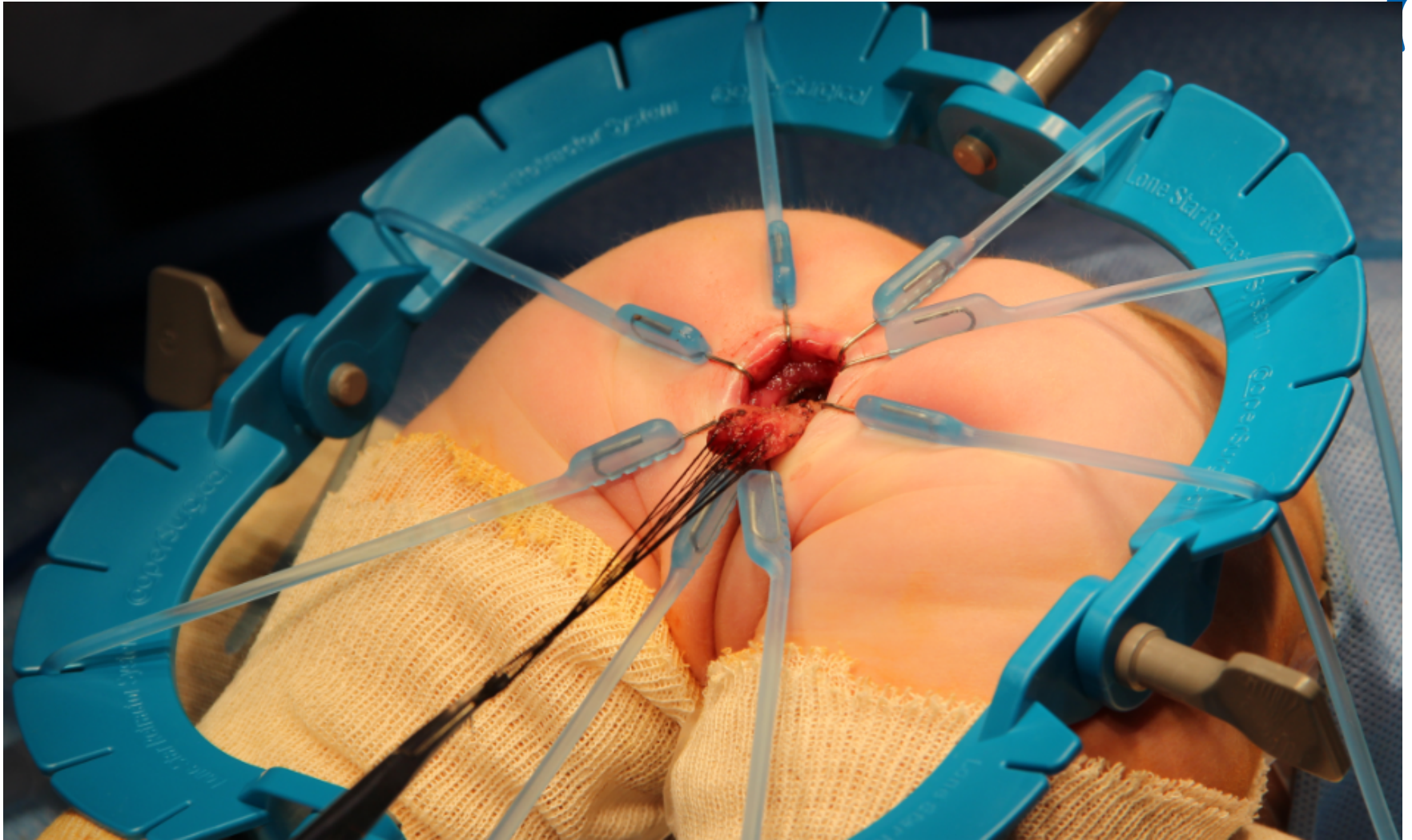




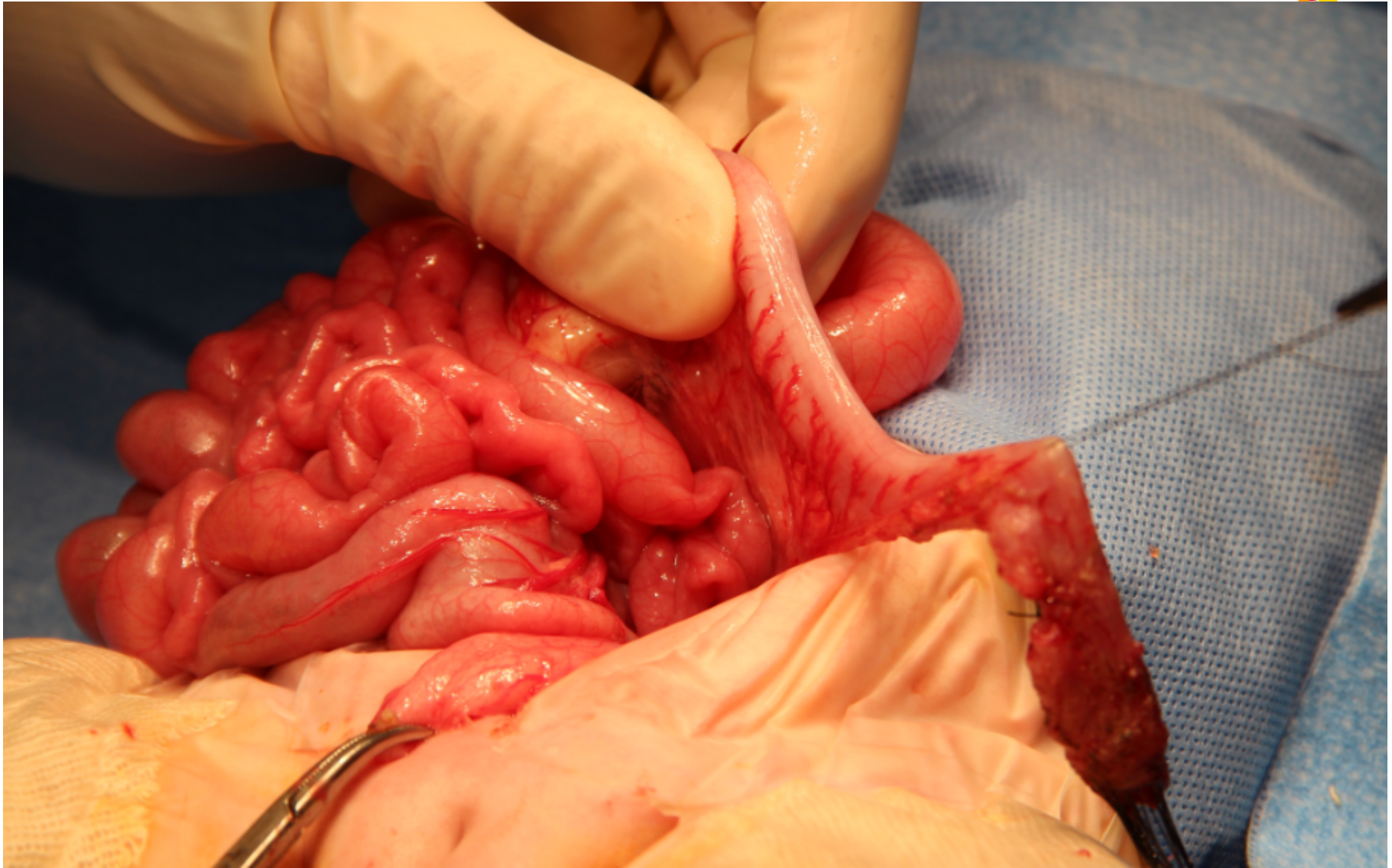
With these images, what would you do?

1. Manage with laxatives, this is idiopathic constipation
2. Open a colostomy immediately
3. Rectal irrigations first, followed by transanal approach
4. Transanal approach immediately
5. I don't know

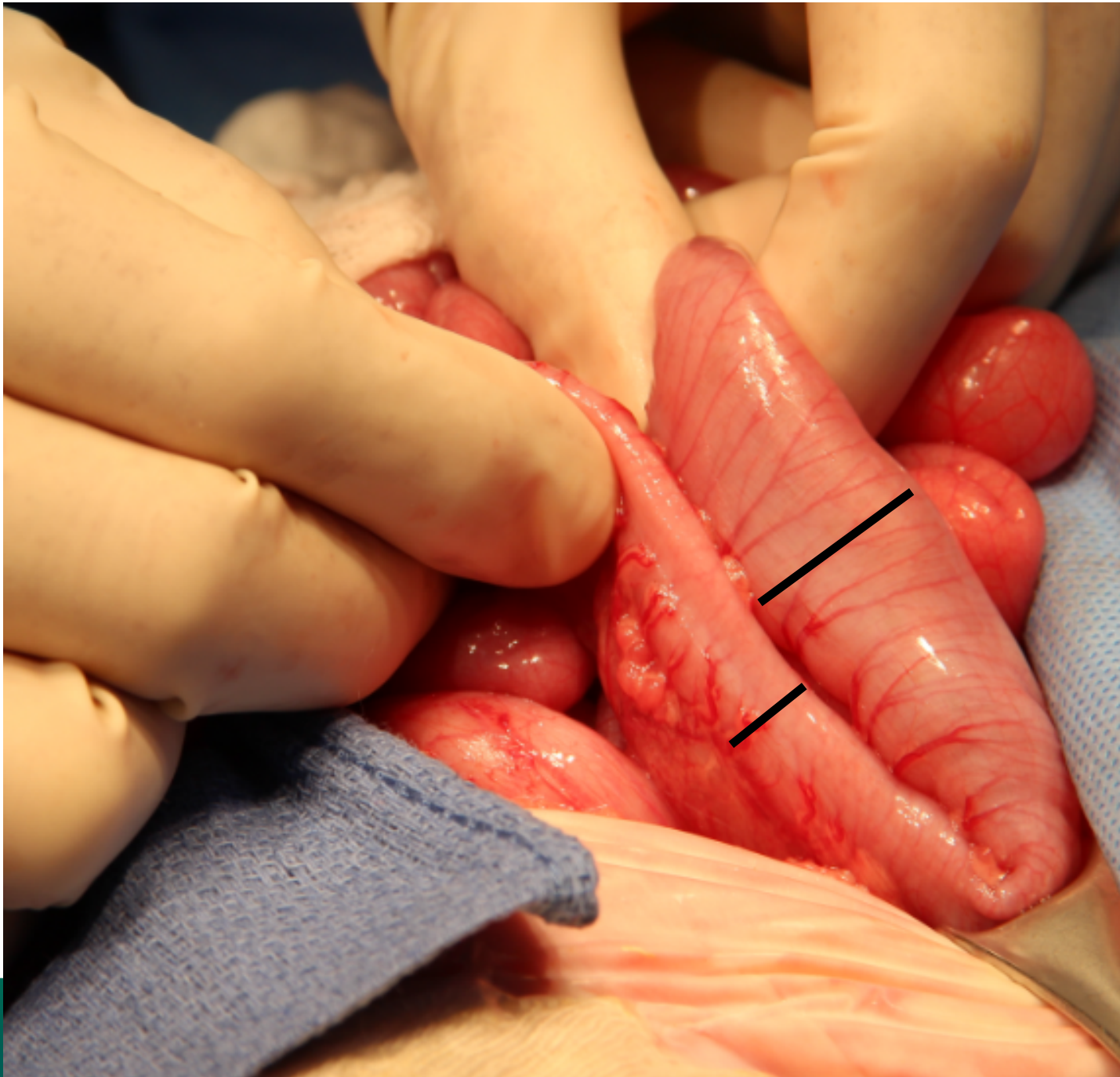


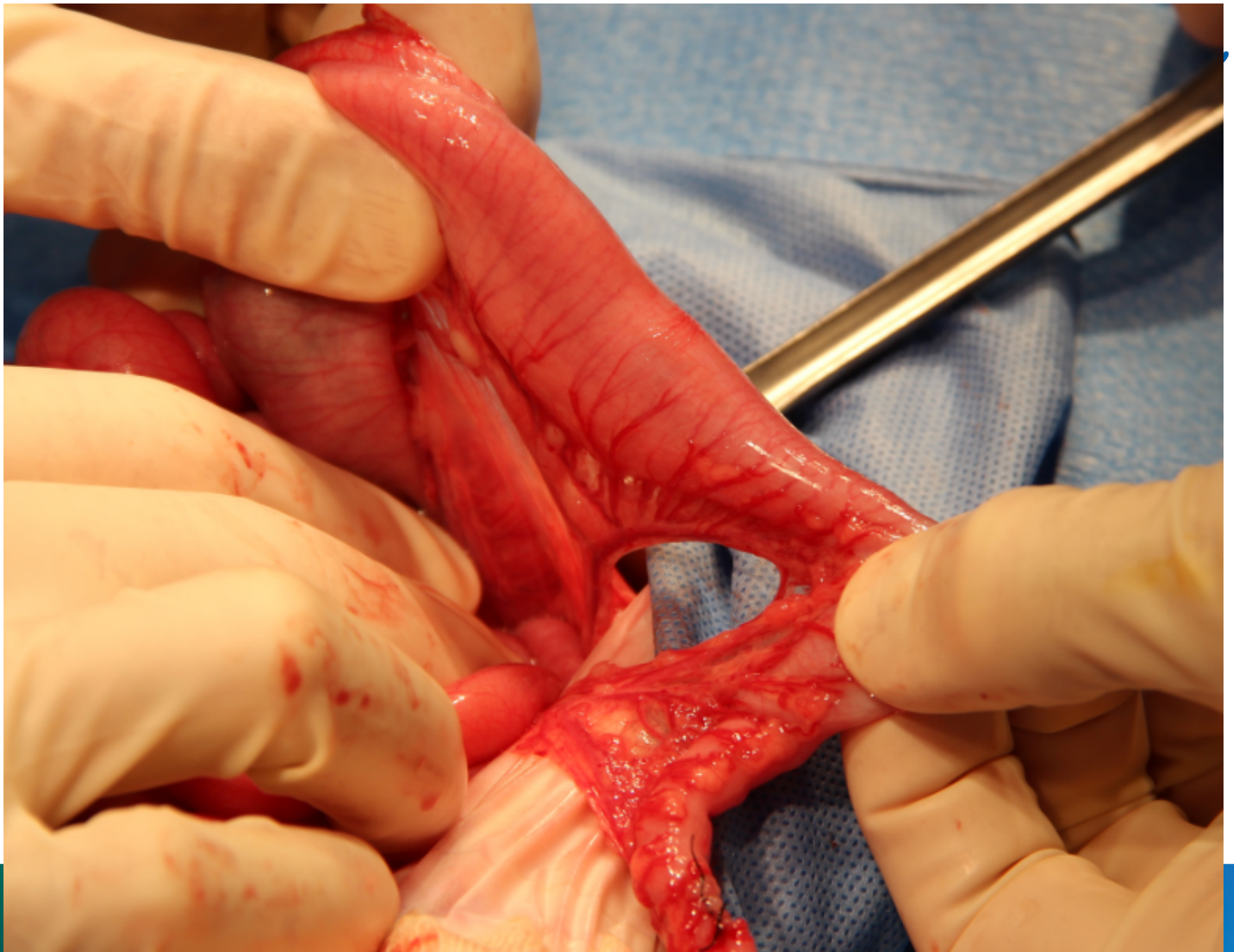


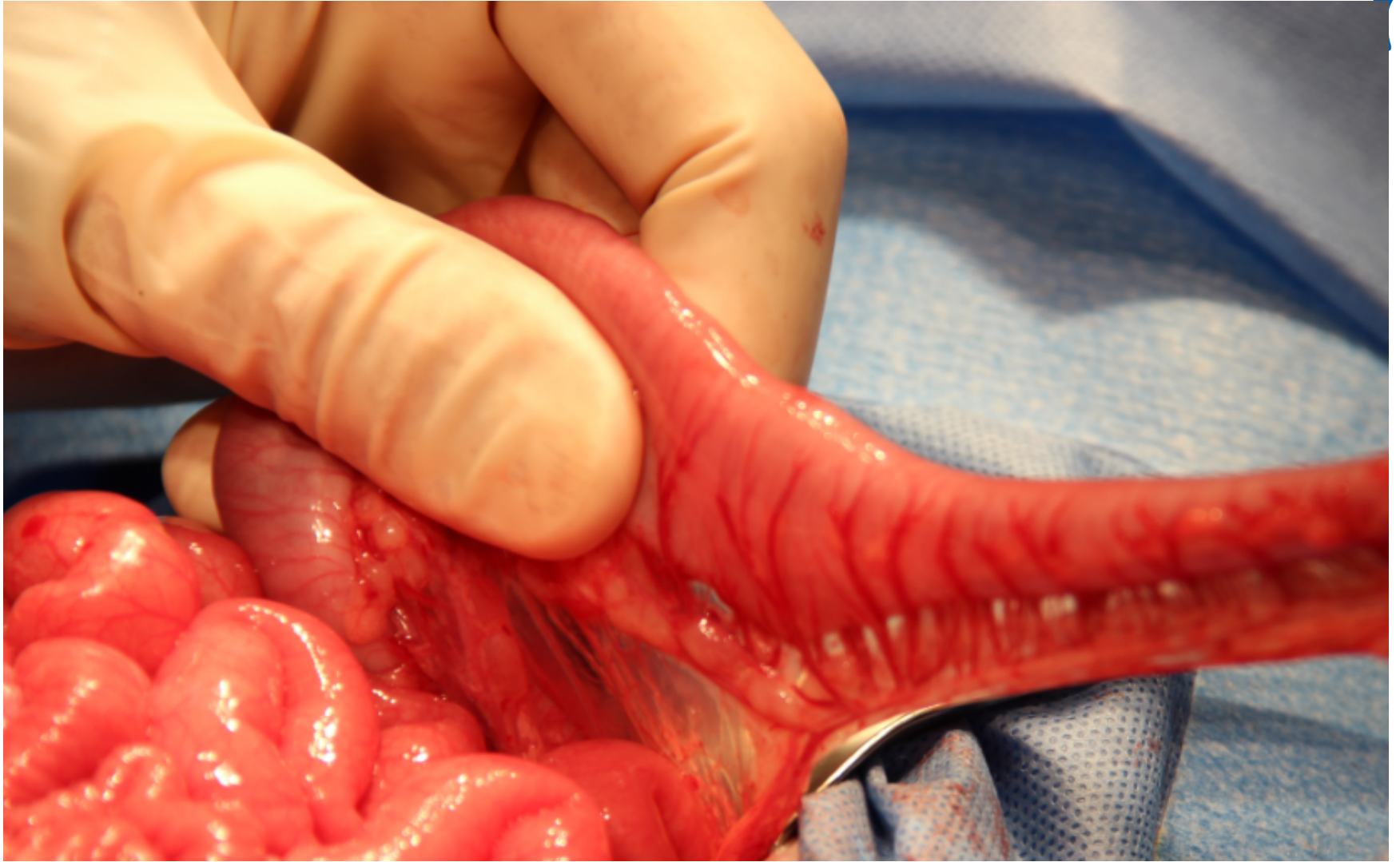


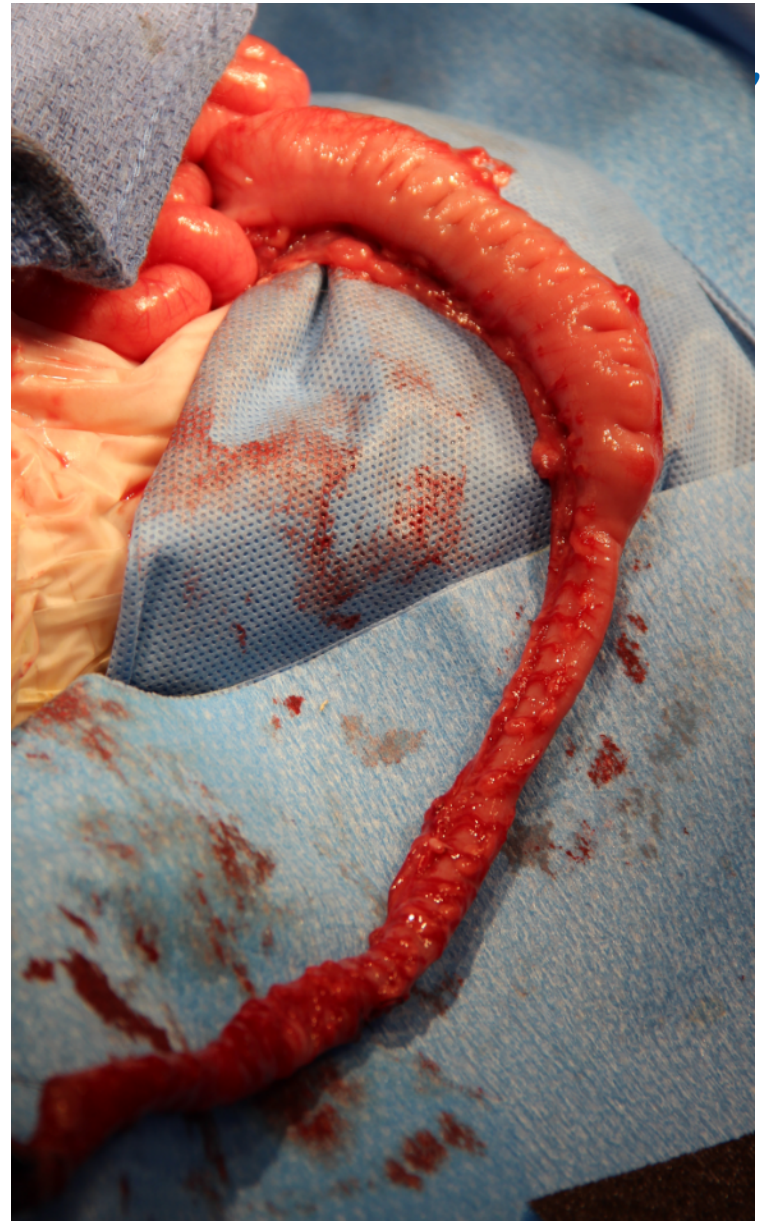




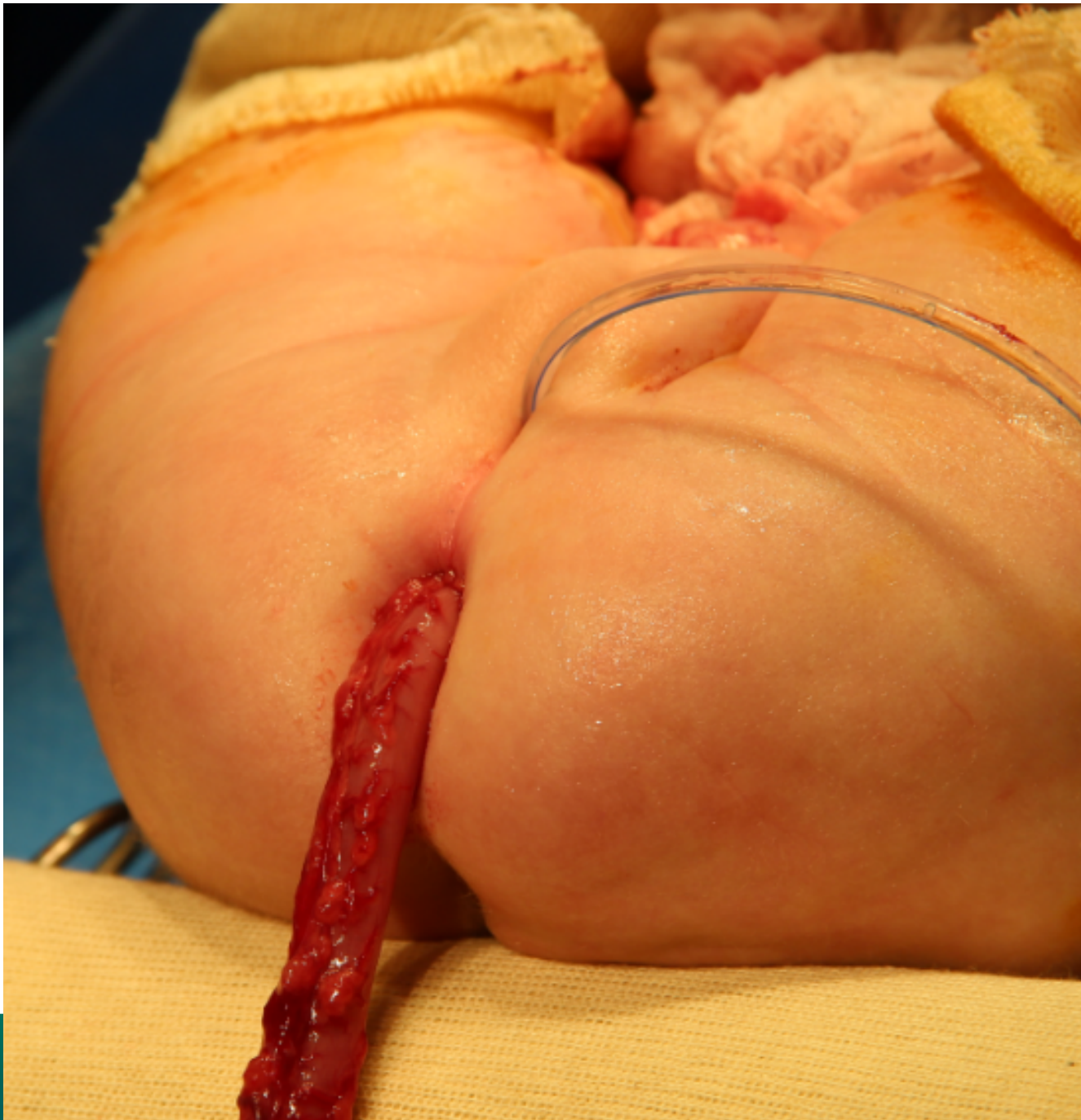












7 days post op

What would you do?

1. Colostomy
2. Exploratory laparotomy
3. Rectal irrigation
4. I don't know



Family asked about chances for another child to also have HD



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).

Thank You!