Imaging in Hirschsprung Disease
International Hirschsprung Course 2021

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Children's Hospital Colorado

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Department
UNIVERSITY OF CO

No disclosures

Objectives

- 1. Build a **teamwork** approach between the *surgeon*, *gastroenterologist*, *radiologist* and *bowel management team*.
- 2. Explain the technique and nuances of contrast imaging exams in Hirschsprung Disease
- 3. Review the sensitivity and specificity of the contrast enema

Step One-Find someone with Passion

- WHY:
- Work *without* purpose
- Punishment

- Work *with* Purpose
- Passion

- **Passion-** is a feeling of intense enthusiasm towards or compelling desire for someone or something.
  - Improves focus, innovation, perfection, sati
    creativity

Question

- T/F: I have a wonderful radiologist that my team works well with in order to image my patients.

- A. True
- B. False

Hx: Newborn (2day) with constipation
Order: Barium Enema

1. *Clinical* exam

   Presence and location of anus

2. Creation of order for "*contrast enema*"
3. History

   Prenatal

   Passag

   mecon

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http://emedicine.medscape.co
overview#a2

What is important information to provide to your Radiologist History

1. Age (gestational and chronological)
2. Is the patient vomiting?

   Bilious or non-bilious.
3. Is the patient stooling?

   Did they pass meconium?

   Are the stools bloody?

Clinical Exam –

- Infant with delayed passage of meconium
- Vomiting? Bilious or Non-Bilious
- Abdominal Distension
- Location of Anus

Neonatal Bowel Obstruction

- Most common abdominal emergency in neonate
- Classified as High or Low
- Distinction on Abdominal radiograph
  - High: proximal to mid ileum
    - esophagus, duodenum, jejunum, prox ileum
    - Exception: esophageal atresia- bowel gas pattern dependent on if TEF present
  - Low: distal to mid ileum
    - distal ileum, colon
    - Many dilated air filled loops
    - Larger than vertebral body

Imaging Exam: High versus Low

Proximal obstruction

Duodenal atresia

Distal obstructio

Small left colon

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Neonatal Bowel Obstruction

- Low (Distal)
  - Hirschsprung disease **
  - Small left colon
  - Meconium ileus
  - Ileal atresia
  - Colonic atresia

Question

Should the radiology contrast enema be postponed or delayed if the patient has had a recent rectal exam or rectal irrigation because of concern of obscuring a transition zone?

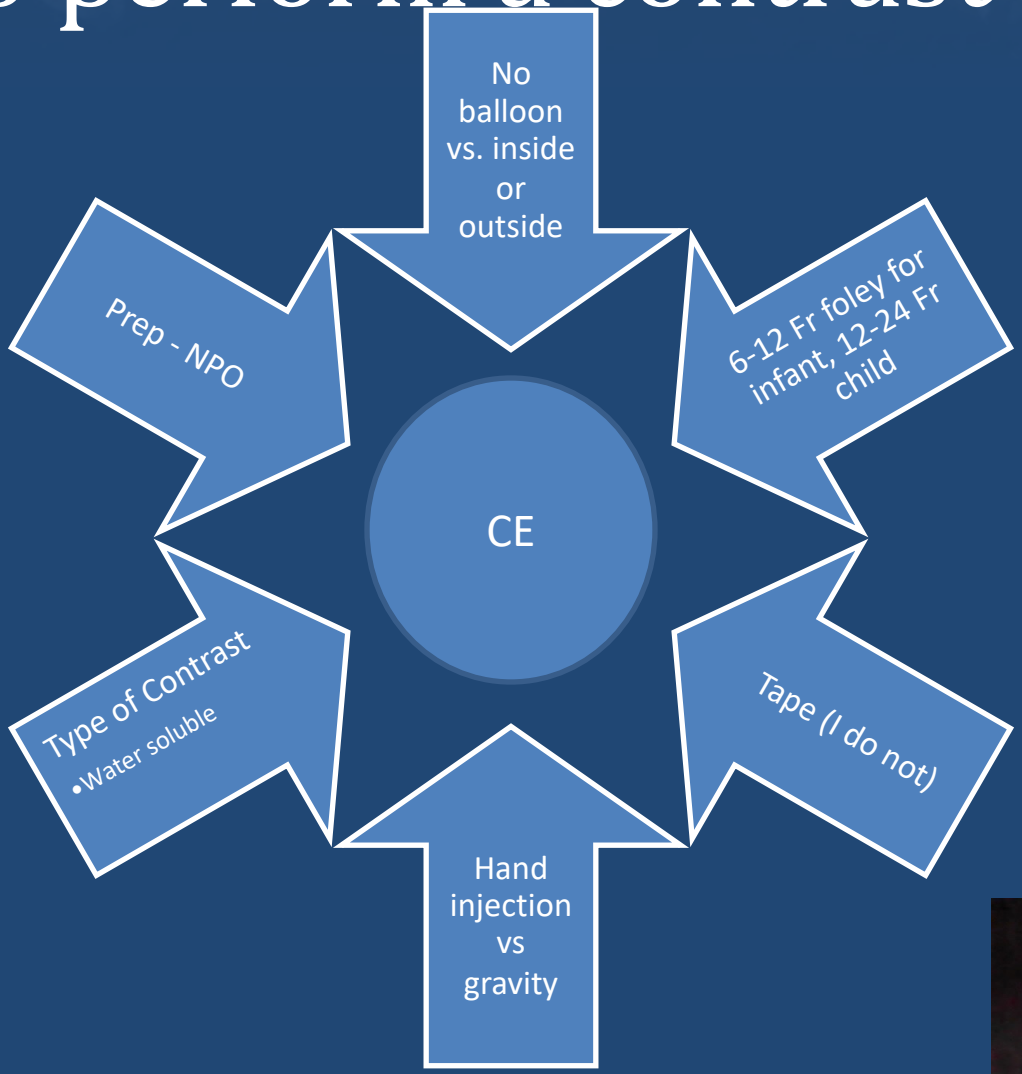
- A. Yes
- B. No

Question

- In your practice, is it considered safe to hand inject the contrast enema into the colon an infant instead of using a gravity bag using passive retrograde filling of the colon?

- A. Yes
- B. No

How to perform a contrast enema



Preparation of Patient

- No bowel prep

- Fasting – not required in neonate
  - I prefer 2-4 hours fasting in older children to prevent vomiting

Contrast selection

Contrast Agent	Osmolality (mosm/kg water)	Iodine (mg/ml)	Cost per 10 mL
E-Z-Paque Barium	0	0	$0.25
Visipaque 320	290	320	$13.76
Cysto-Conray 2	400	81	$1.10
Optiray 320	702	320	$15.75
Gastrografin	1940	367	$6.34

Cysto conray 2 or isovue 1:1 for newborn

Gastrografin 3:1 in 2 years and over

https://www.acr.org/-/media/ACR/Files/Clinical-Resources/Contrast_Media.pdf

Product	Generic name (concentration in mg contrast/ml)	Ionicity	Iodine+ (mg/ml)	Viscosity+ 25° C (cp or mPa.s)	Viscosity+ 37° C (cp or mPa.s)	Osmolality (mOsm/kg H2O)
INTRAVASCULAR						
Omnipaque™ 140 (GE Healthcare)	Iohexol 302	Nonionic	140	2.3*	1.5	322
Conray™ 30 (Covidien)	iothalamate (300)	Ionic	141	2	1.5	600
Ultravist® 150 (Bayer HealthCare)	iopromide	Nonionic	150	2.3*	1.5	328
Omnipaque™ 180 (GE Healthcare)	iohexol (388)	Nonionic	180	3.1*	2	408
Isovue®-200 (Bracco)	iopamidol (408)	Nonionic	200	3.3*	2.0	413
Conray™ 43 (Covidien)	iothalamate (430)	Ionic	202	3	2	1000
Omnipaque™ 240 (GE Healthcare)	iohexol (518)	Nonionic	240	5.8*	3.4	520
Optiray™ 240 (Guerbet)	ioversol (509)	Nonionic	240	4.6	3.0	502
Ultravist® 240 (Bayer Healthcare)	iopromide	Nonionic	240	4.9*	2.8	483
Isovue® 250 (Bracco)	iopamidol (510)	Nonionic	250	5.1*	3.0	524
Visipaque™ 270 (GE Healthcare)	iodixanol (550)	Nonionic	270	12.7*	6.3	290
Conray™ (Covidien)	iothalamate (600)	Ionic	282	6	4	1400
Isovue® 300 (Bracco)	iopamidol (612)	Nonionic	300	8.8*	4.7	616
Omnipaque™ -300 (GE Health-care)	iohexol (647)	Nonionic	300	11.8*	6.3	672
Optiray™ 300 (Guerbet)	ioversol (640)	Nonionic	300	8.2	5.5	651
Oxilan® 300 (Guerbet)	ioxilan (623)	Nonionic	300	9.4*	5.1	610
Ultravist® 300 (Bayer Healthcare)	iopromide	Nonionic	300	9.2*	4.9	607
Hexabrix™*** (Guerbet)	ioxaglate meglumine/ sodium (589)	Ionic	320	15.7*	7.5	≈600
Optiray™320 (Guerbet)	ioversol (680)	Nonionic	320	9.9	5.8	702
Visipaque™ 320 (GE Healthcare)	iodixanol (652)	Nonionic	320	26.6	11.8	290
Optiray™ 350 (Guerbet)	ioversol (740)	Nonionic	350	14.3	9.0	792
Omnipaque™ 350 (GE Healthcare)	iohexol (755)	Nonionic	350	20.4*	10.4	844
Oxilan® 350 (Guerbet)	ioxilan (727)	Nonionic	350	16.3*	8.1	721
Isovue® 370 (Bracco)	iopamidol (755)	Nonionic	370	20.9*	9.4	796
MD-76™ R (Guerbet)	diatrizoate/ meglumine/ sodium (760)	Ionic	370	16.4	10.5	1551
Ultravist® 370 (Bayer Healthcare)	Ioprokoi98mide	Nonionic	370	22.0*	10.0	774

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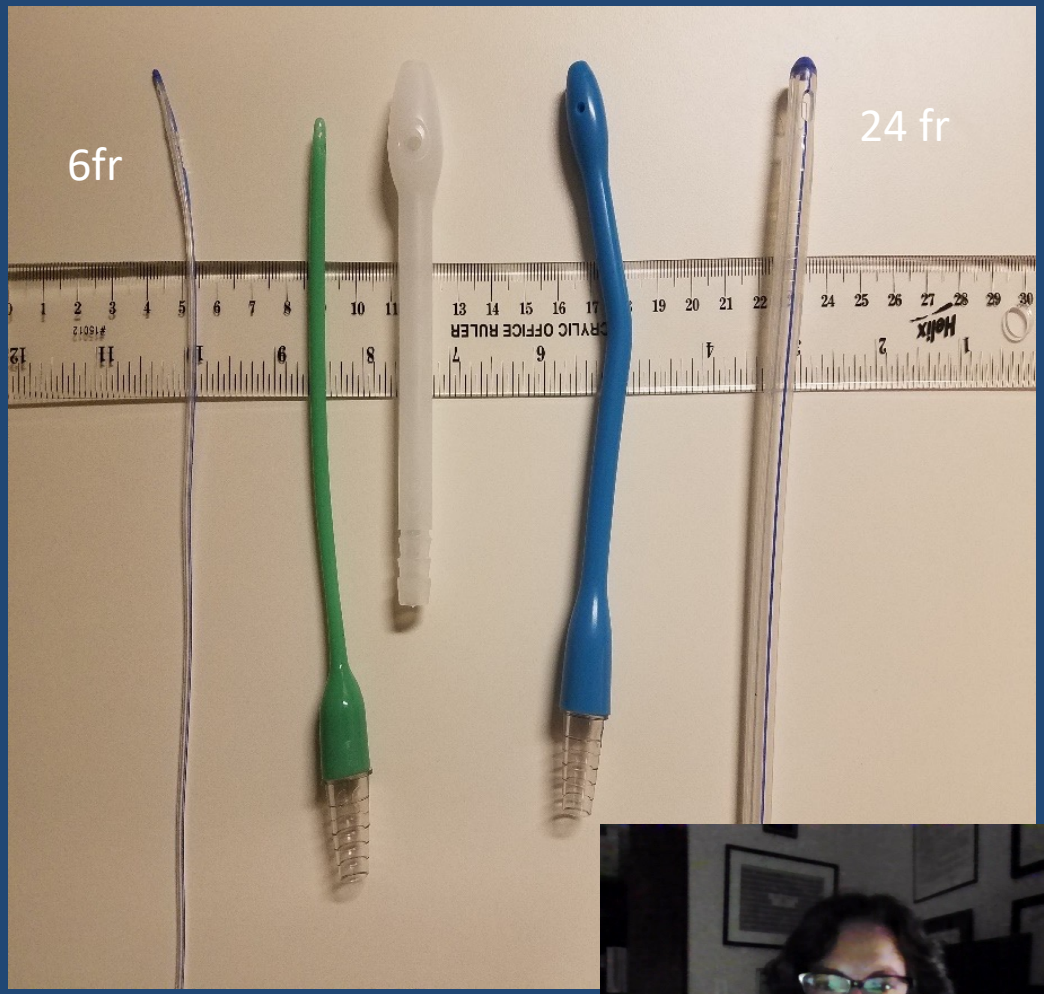
Tube Selection

- 6-8 Fr Premie
- 10-12 Fr newborn
- 12-16 2 mo -2 yr
- 16-**24** FR 2 yr +

- Latex free

- Use balloon only if I need to prevent leakage



To Tape or Not to Tape

- Do what works for you
- Technologist is holding legs and catheter
- I inflate balloon to match rectal size if leaking

https://www.sparefoot.c
storage/blog/17251-best

Hand vs Gravity Filling

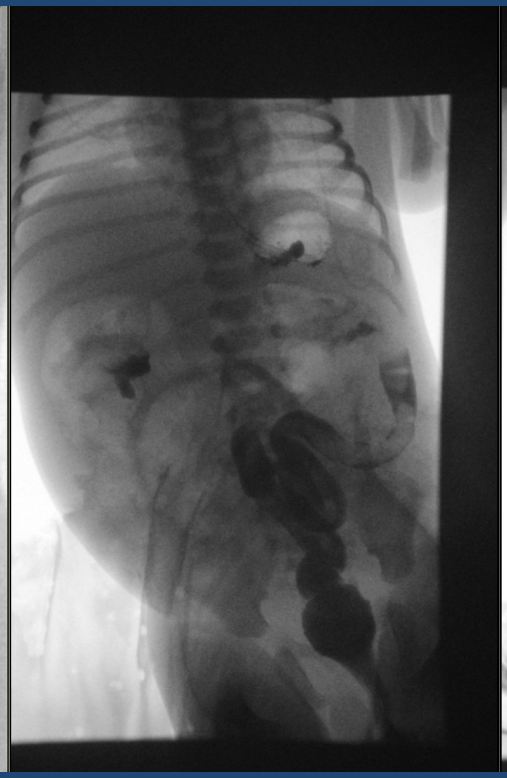
- Experienced fluoroscopist
  - I use had filling in infants because the volume is small (less than 60 ml)
  - 50 – 60 ml syringe to decrease pressure
  - I watch under fluoroscopy to slowly back fill the colon
  - Gravity bag for larger volumes, 6 mo and older
  - Some radiologists do not feel comfor hand injection or balloon inflation

Question

- Is it considered safe to inflate a foley balloon during contrast enema in an infant

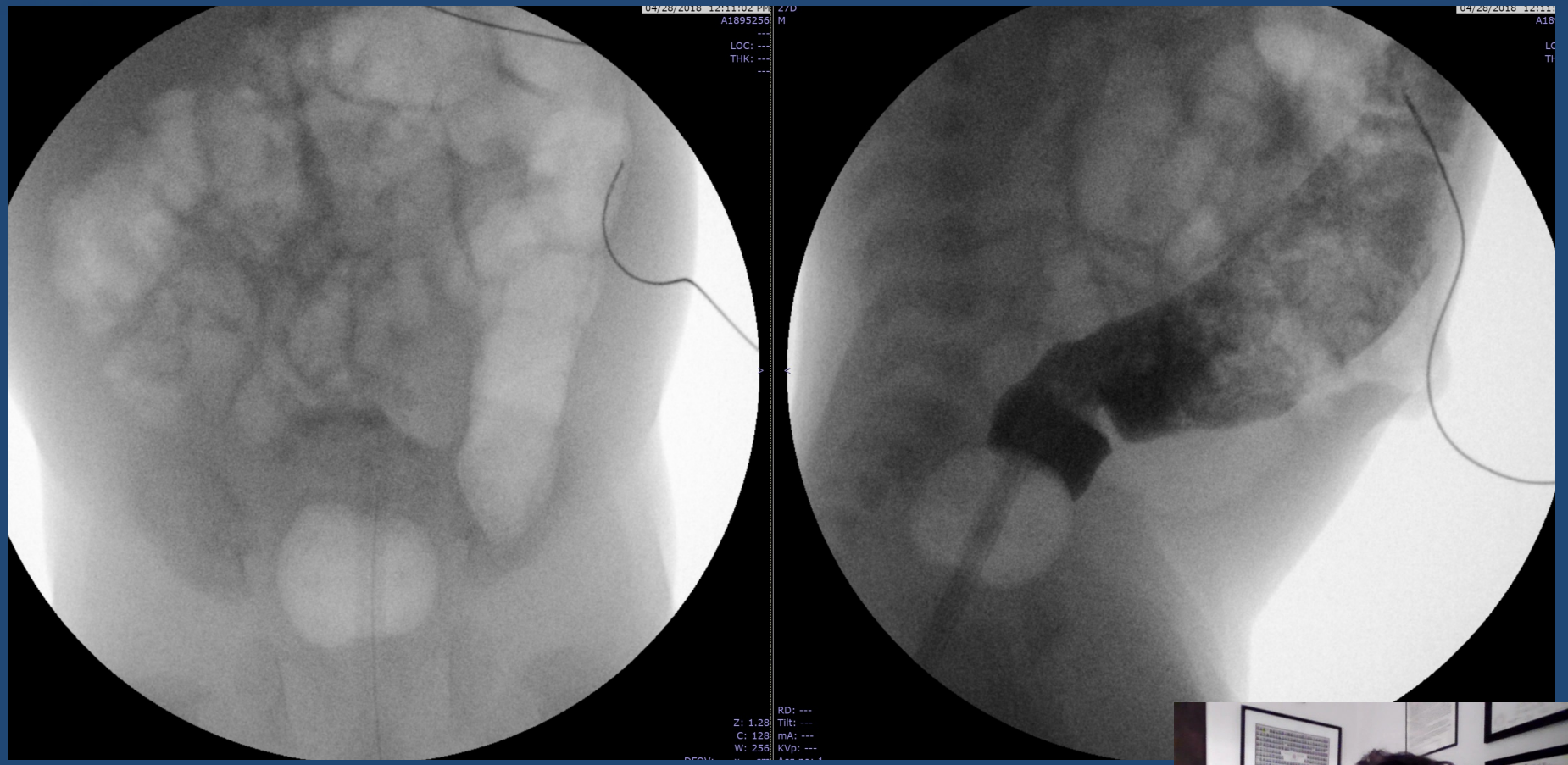
- Yes
- No

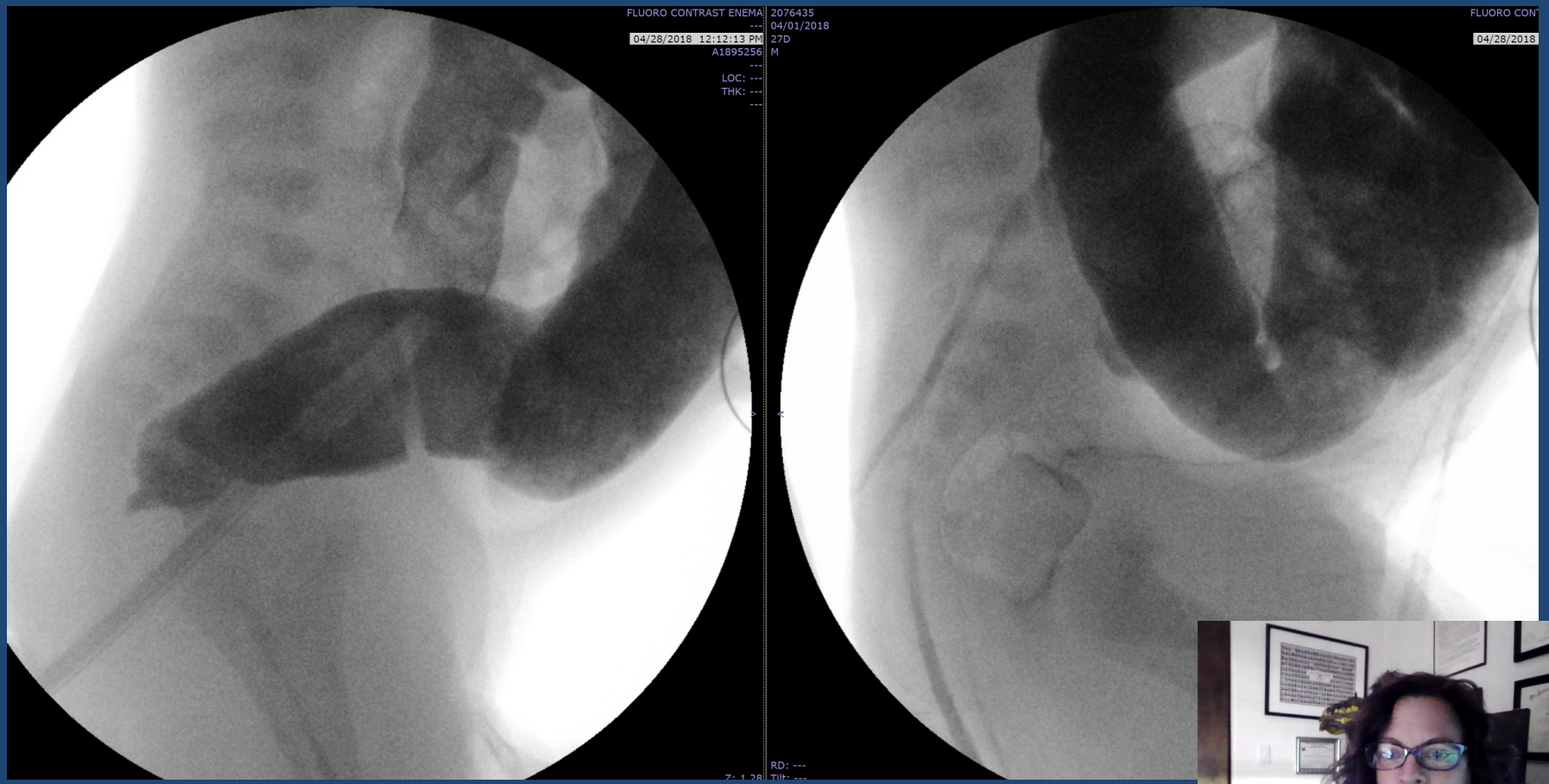
2 day old male transferred from outside hospital with bilious emesis



Balloon on inside only under fluoro guidance

27 day male with constipation





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Re presents with constipation at 28 months

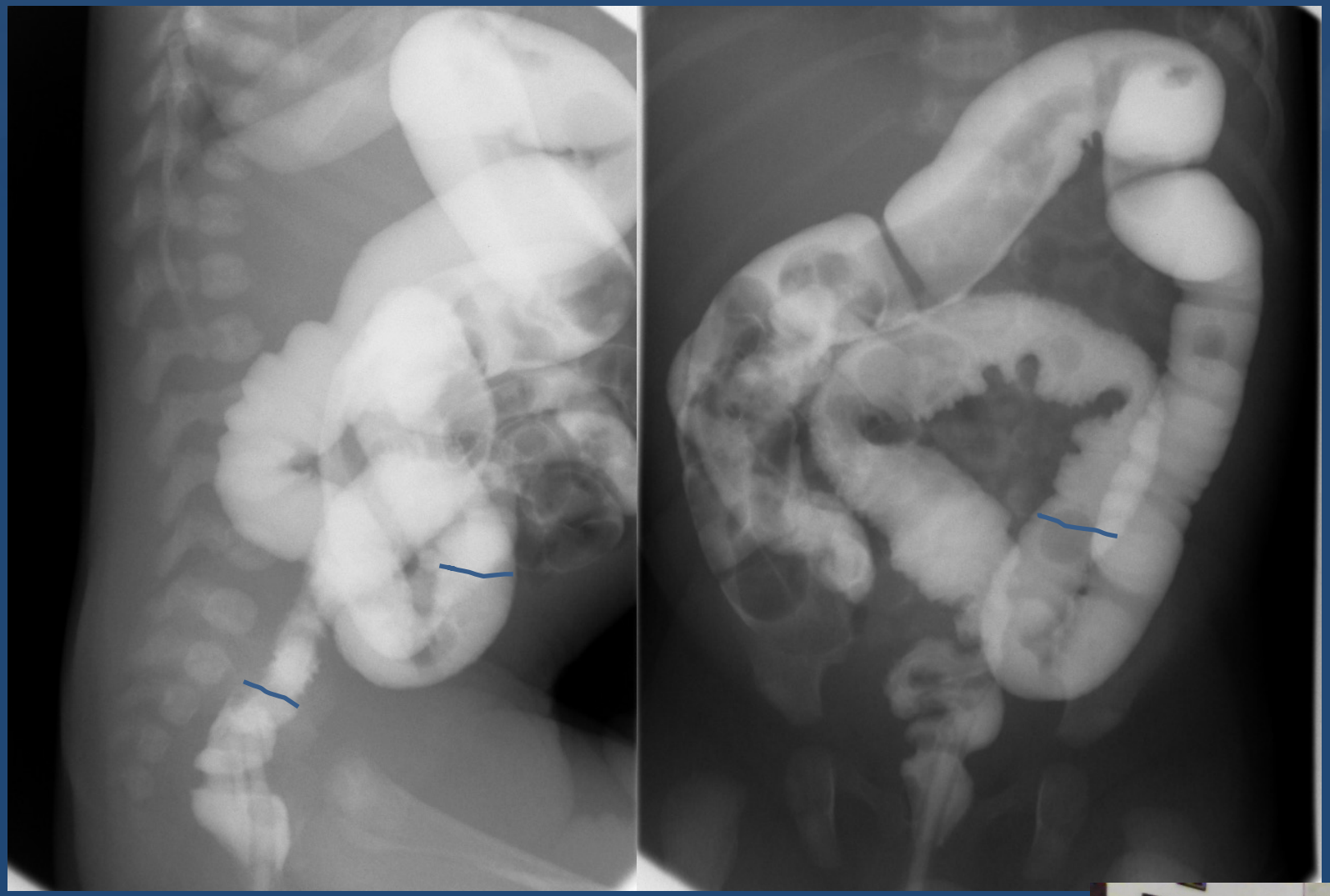
Re Bx

- Absent ganglion cells
- Botox

HD

- Features of a contrast enema that suggest HD:
  - presence of a radiological Transition Zone
  - irregular colonic contractions
  - irregular mucosa
  - an abnormal recto sigmoid index (1/1)

  - Harald Hirschsprung – Danish pediatrician who first described HD in 1886 following death of 2 infants who died of constipation.



A positive enema findings increase the likelihood of HD from clinical probability 13 % to 82%

The utility of the contrast enema in neonates with suspected Hirschsprung disease. 2015 Journal Ped Surg

- 158 CEs were reviewed.
- Common indications for CE were similar between non-HD and HD groups
- Interrater agreement of TZ was 89%

- posttest probabilities
  - positive 83%
  - inconclusive 32%
  - negative 2.5% (total colonic aganglionosis)

- https://doi.org/10.1016/j.jpedsurg.2015.03.019

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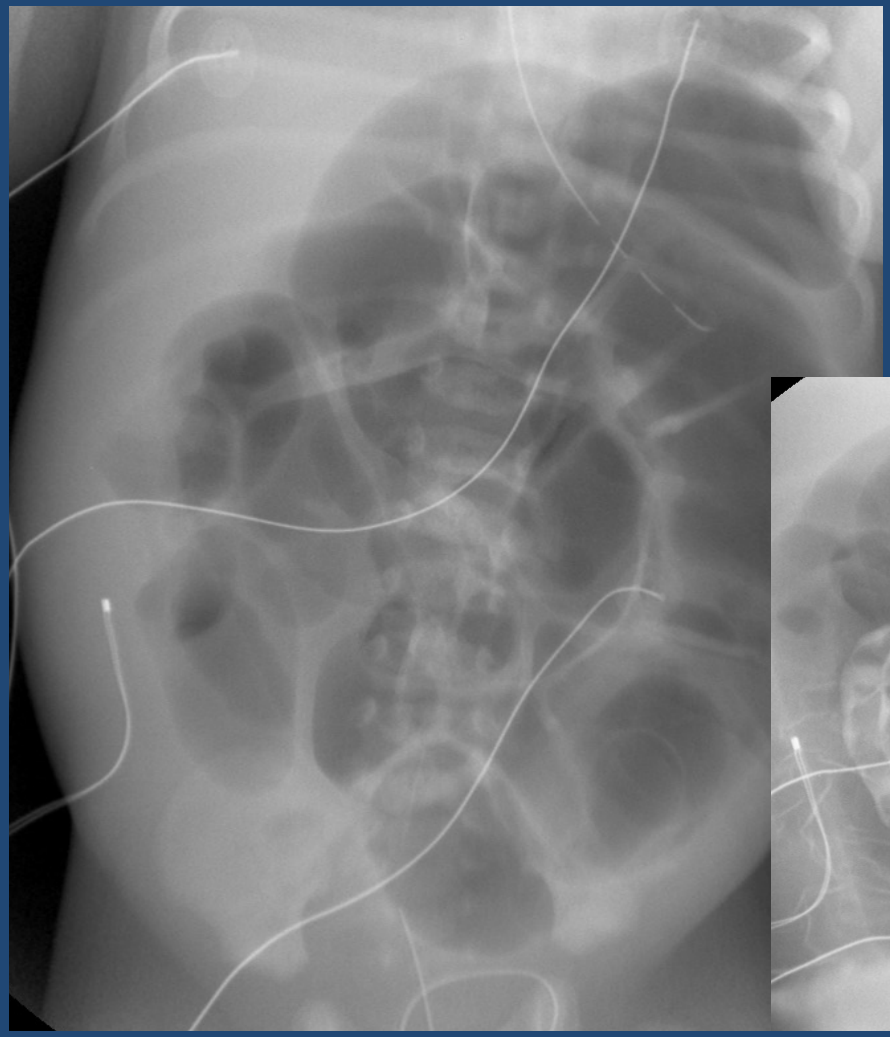
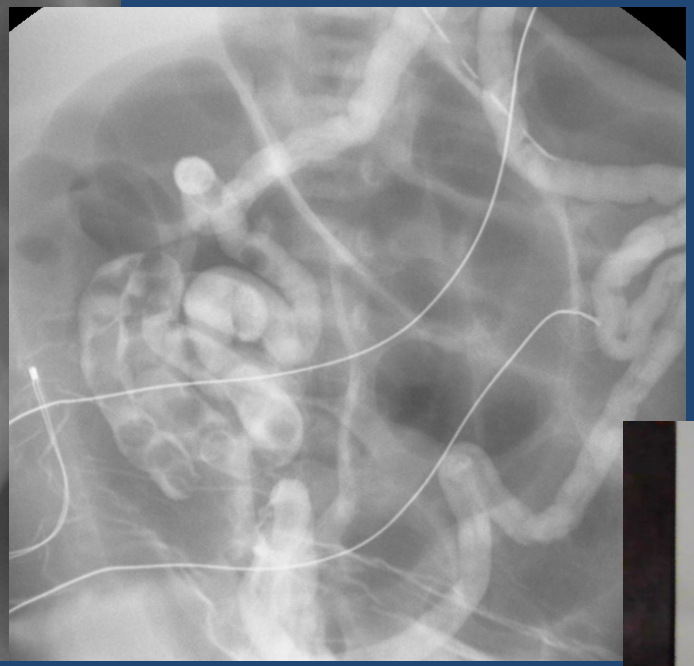
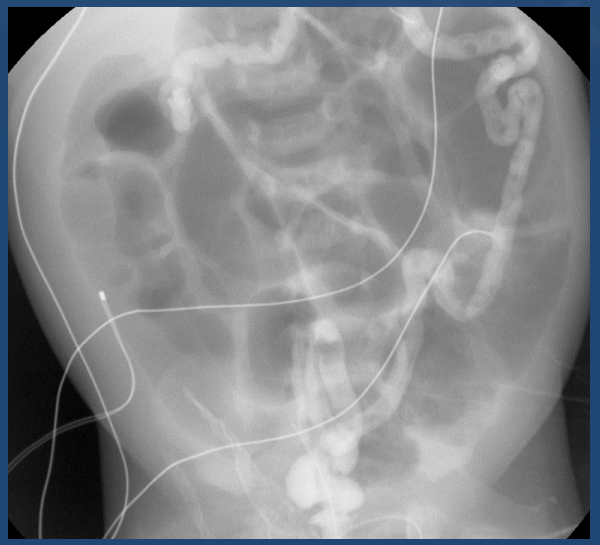
Neonate with distension

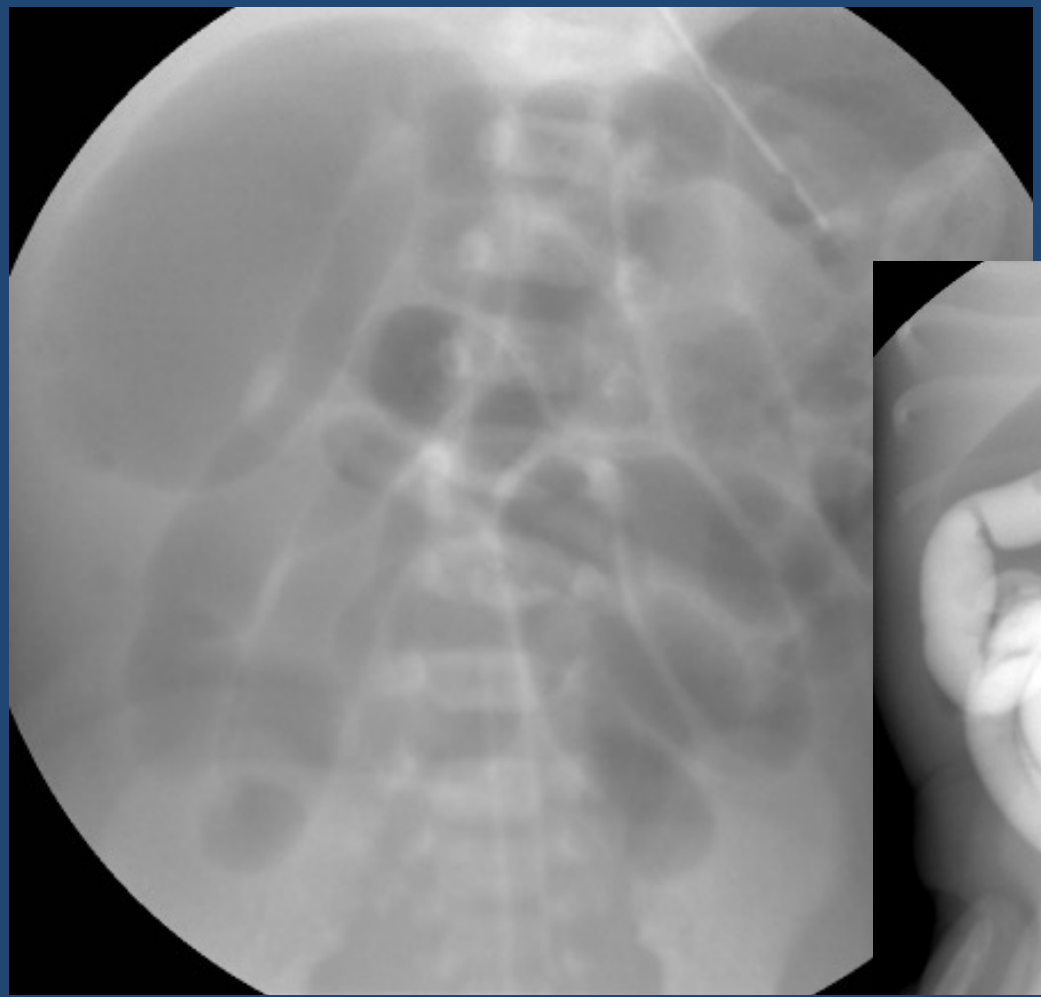
Microcolon with meconium ileus

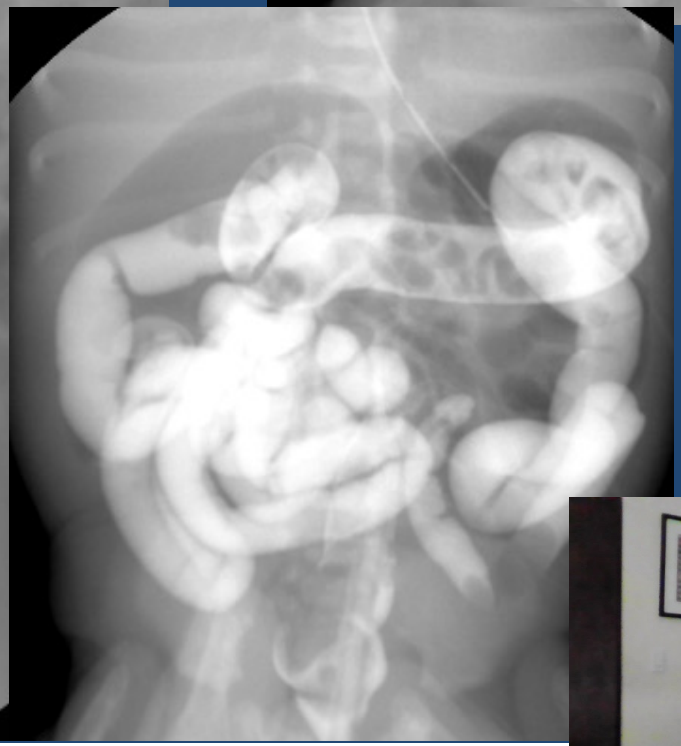


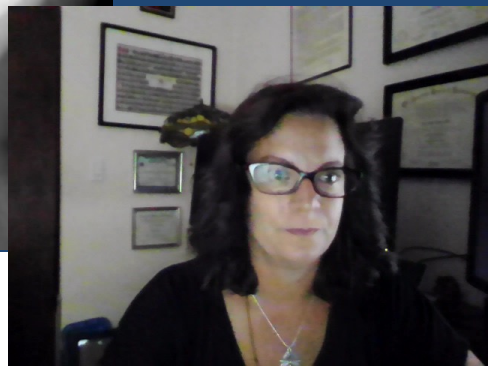




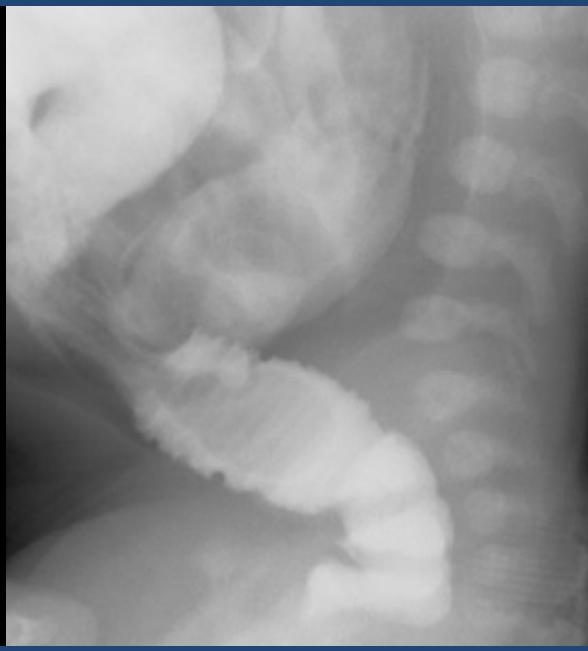
“Normal” size colon: Ileal atresia - (proximal)

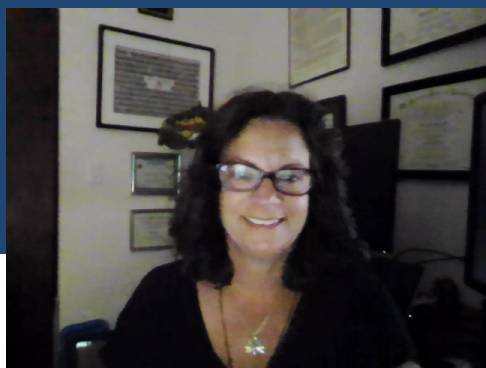




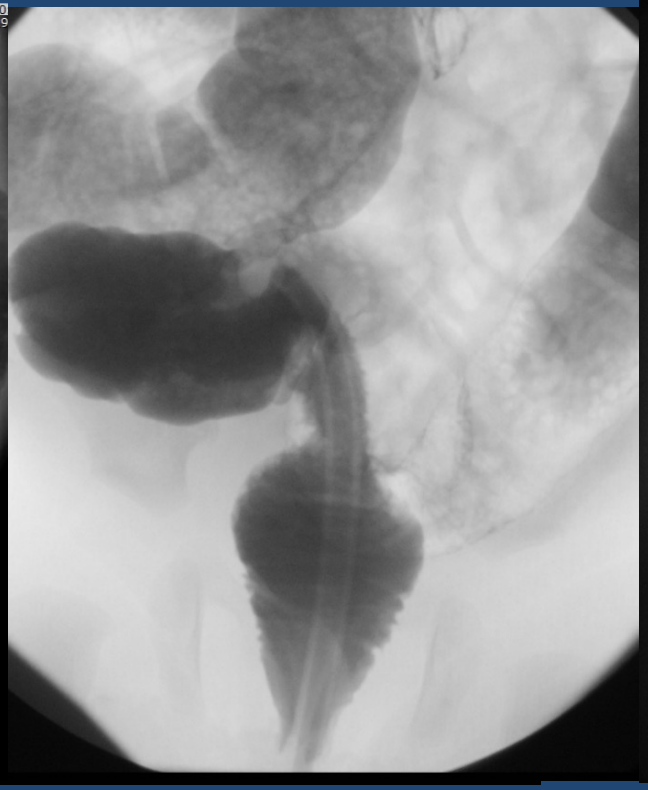


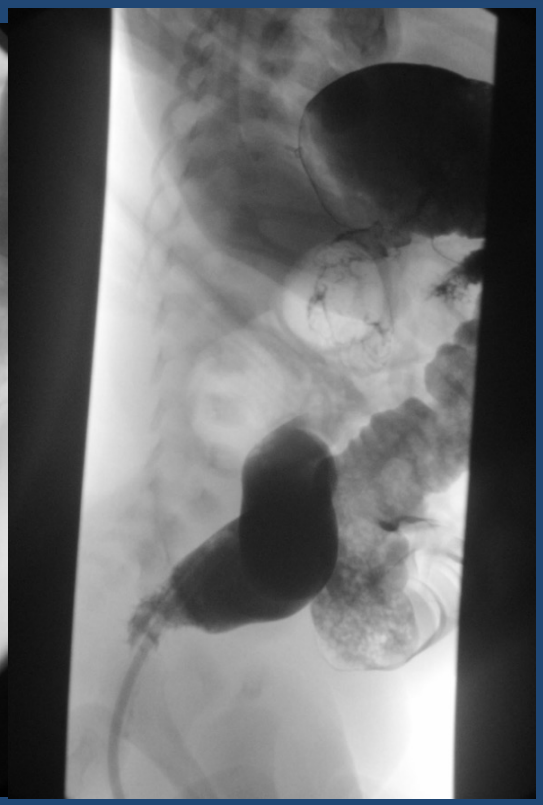
Proximal colon, enlarged: Irregular contracted rectosigmoid: Hirschsprung

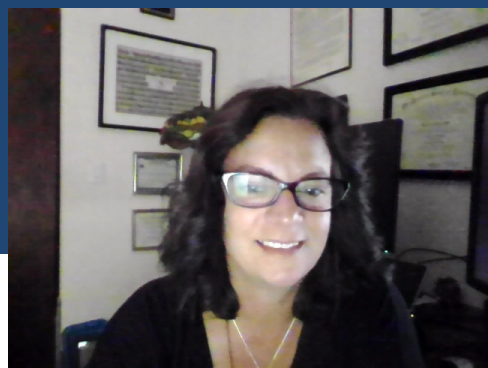




16 day







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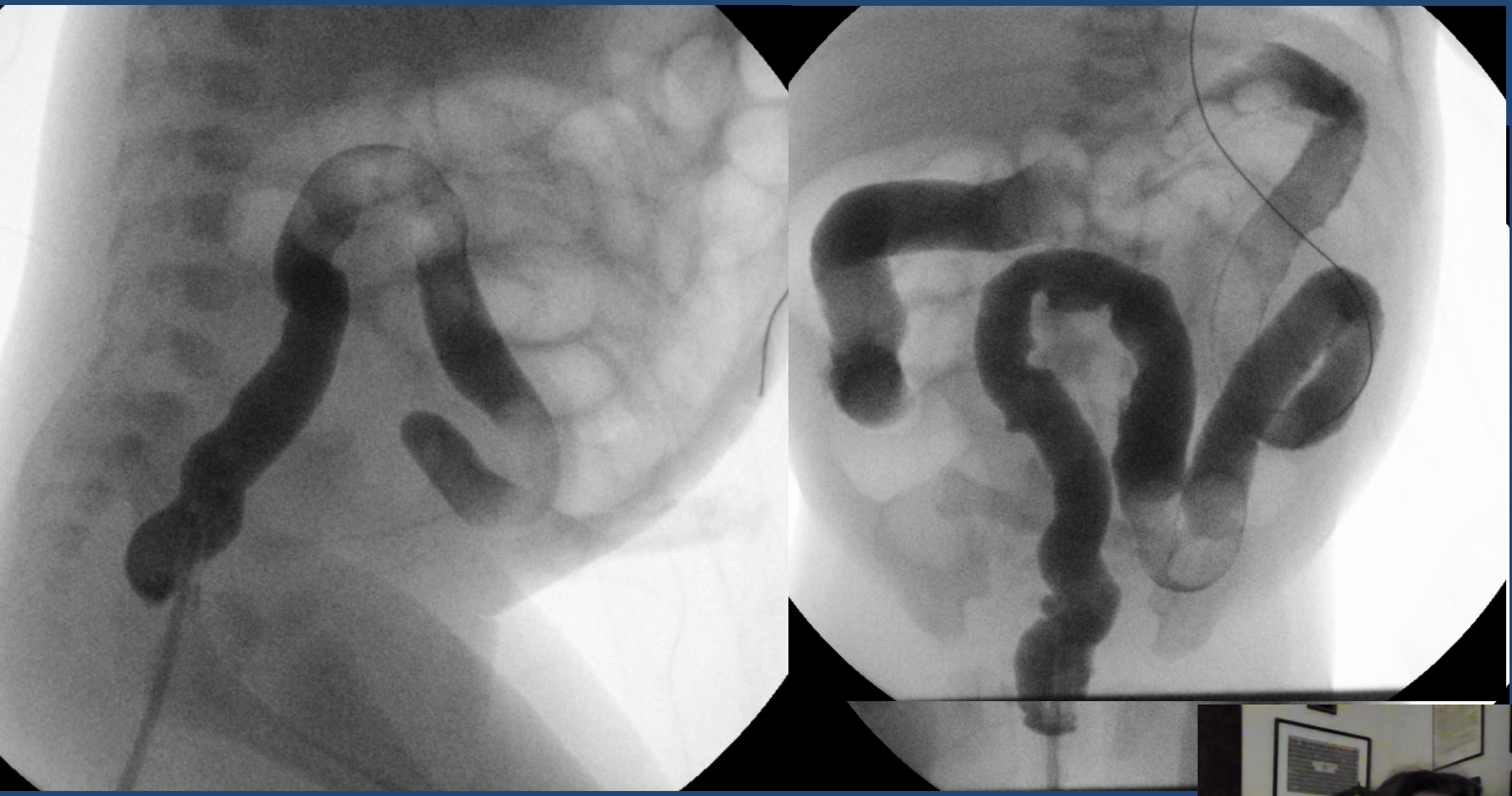
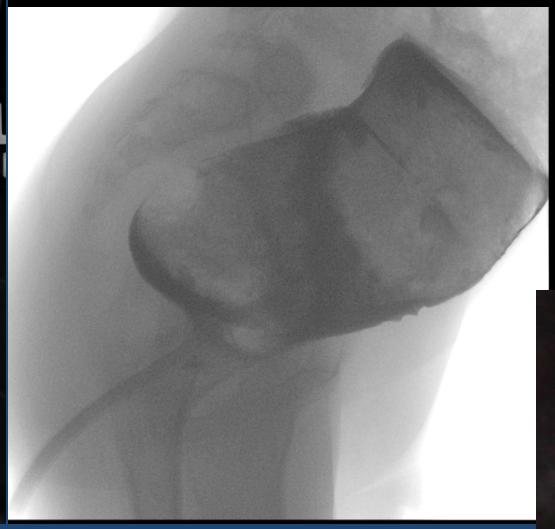
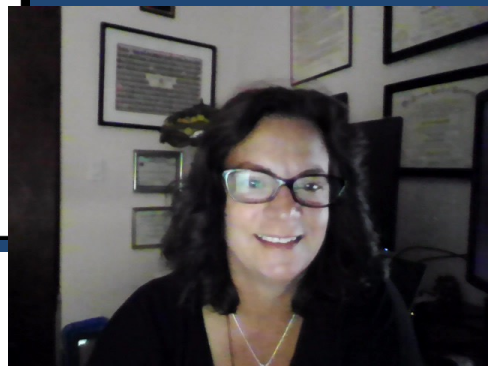
2 day old with vomiting

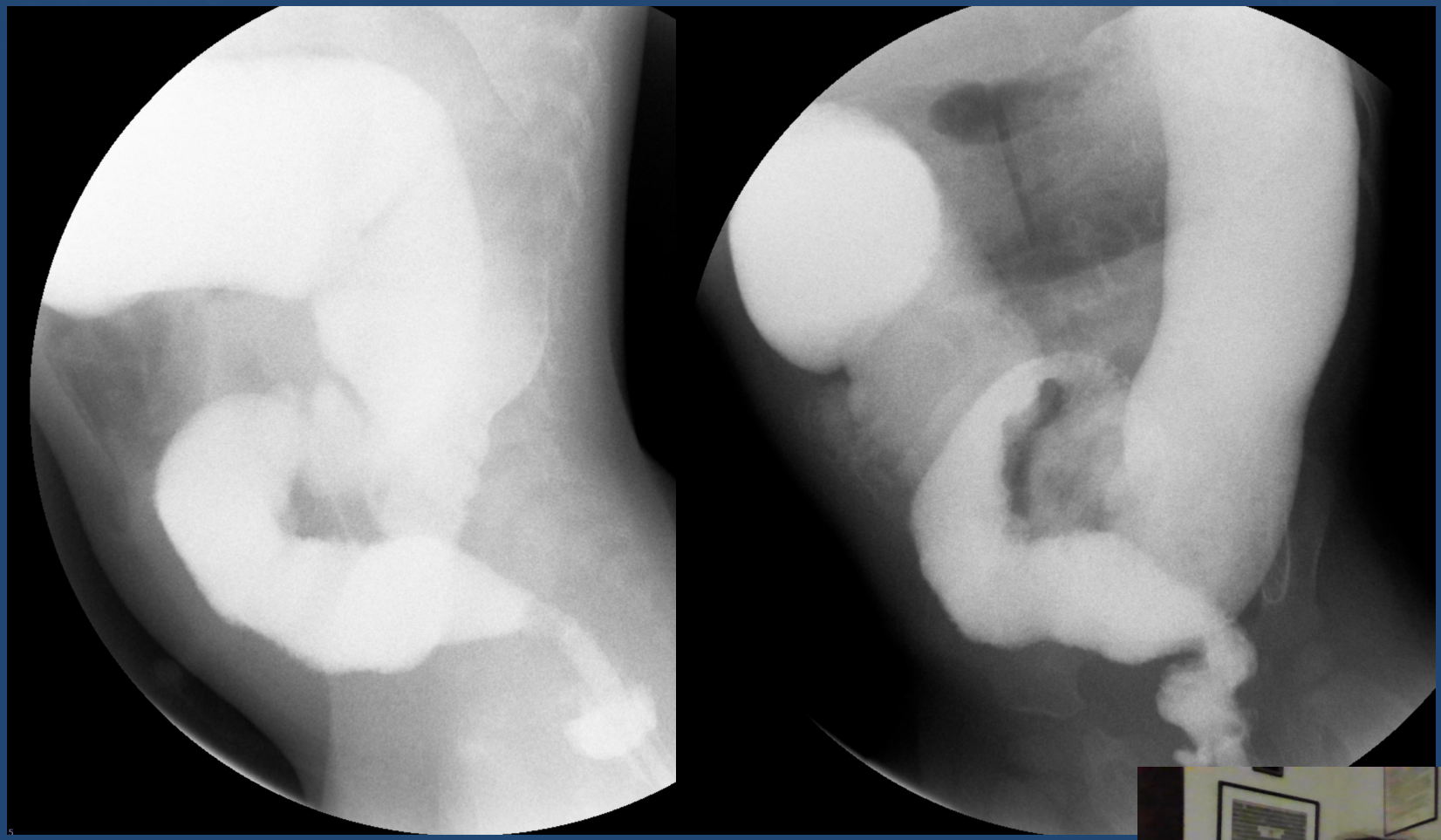


Total aganglionosis including ileum

12 yr old with encopresis







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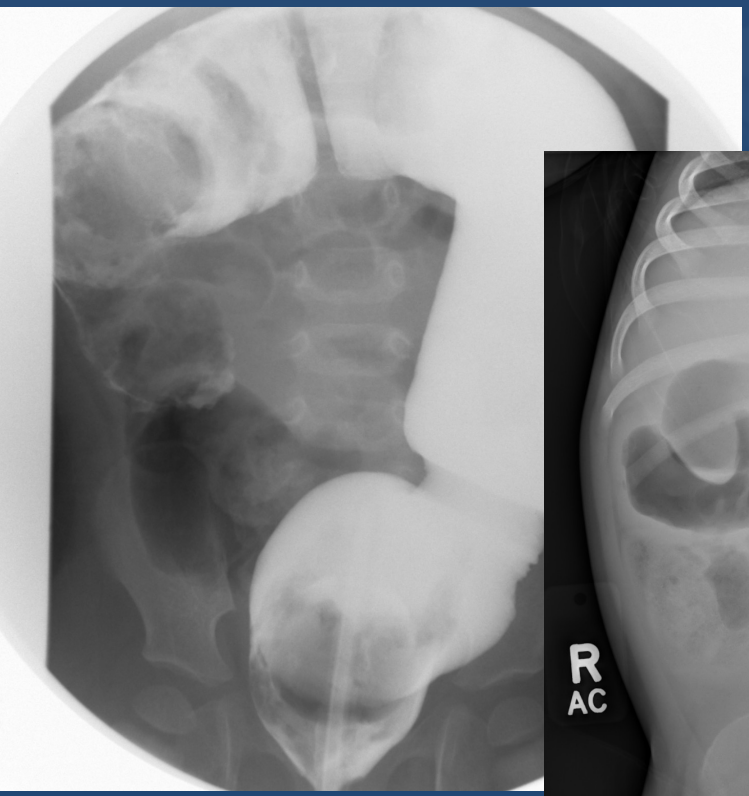

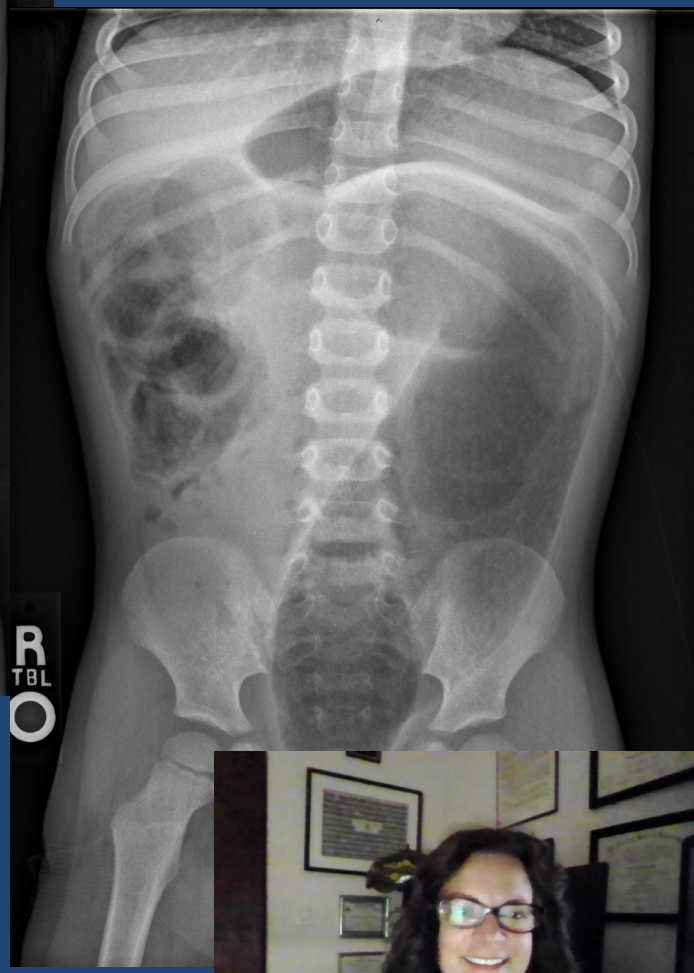

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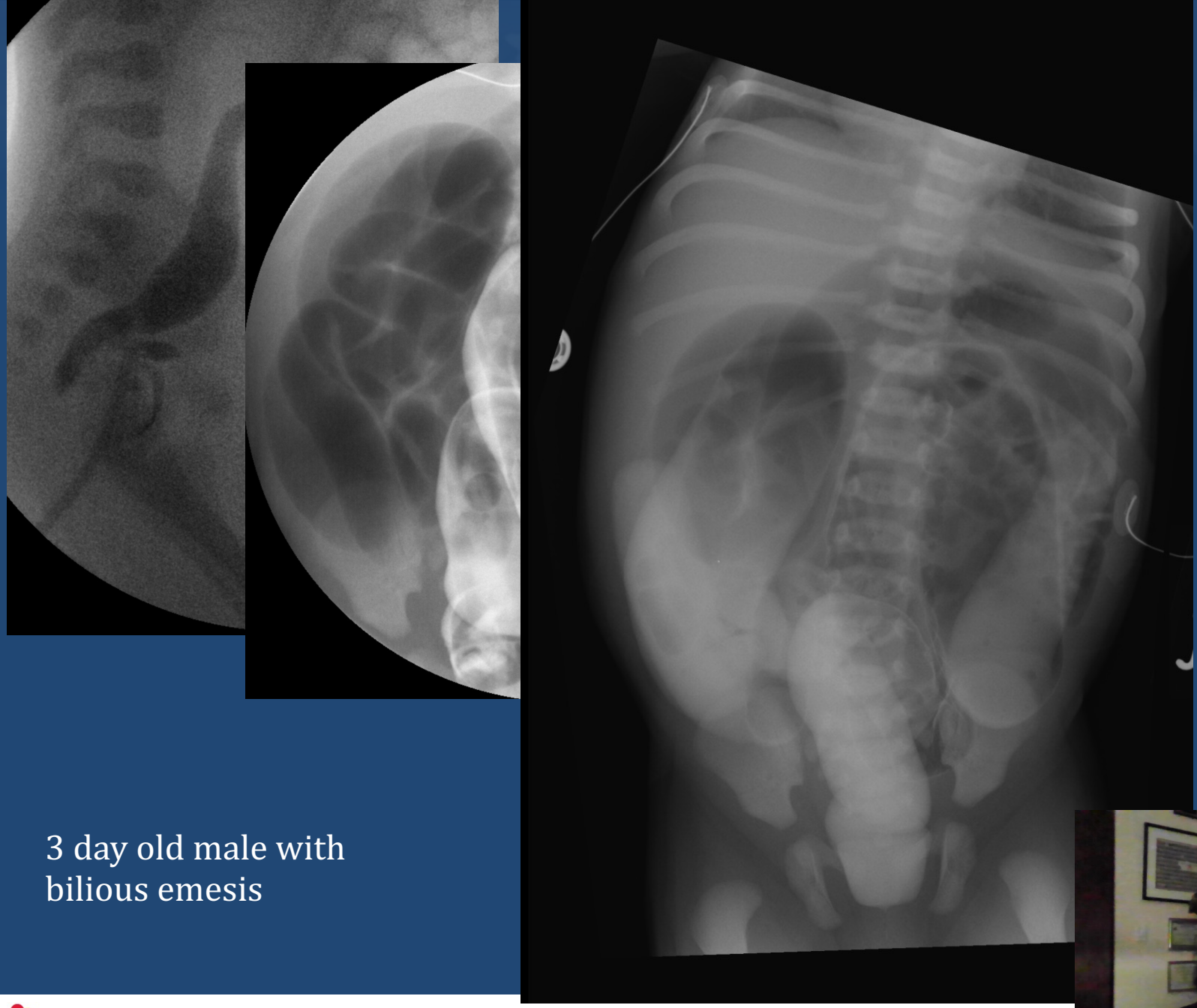

3 day old male with bilious emesis

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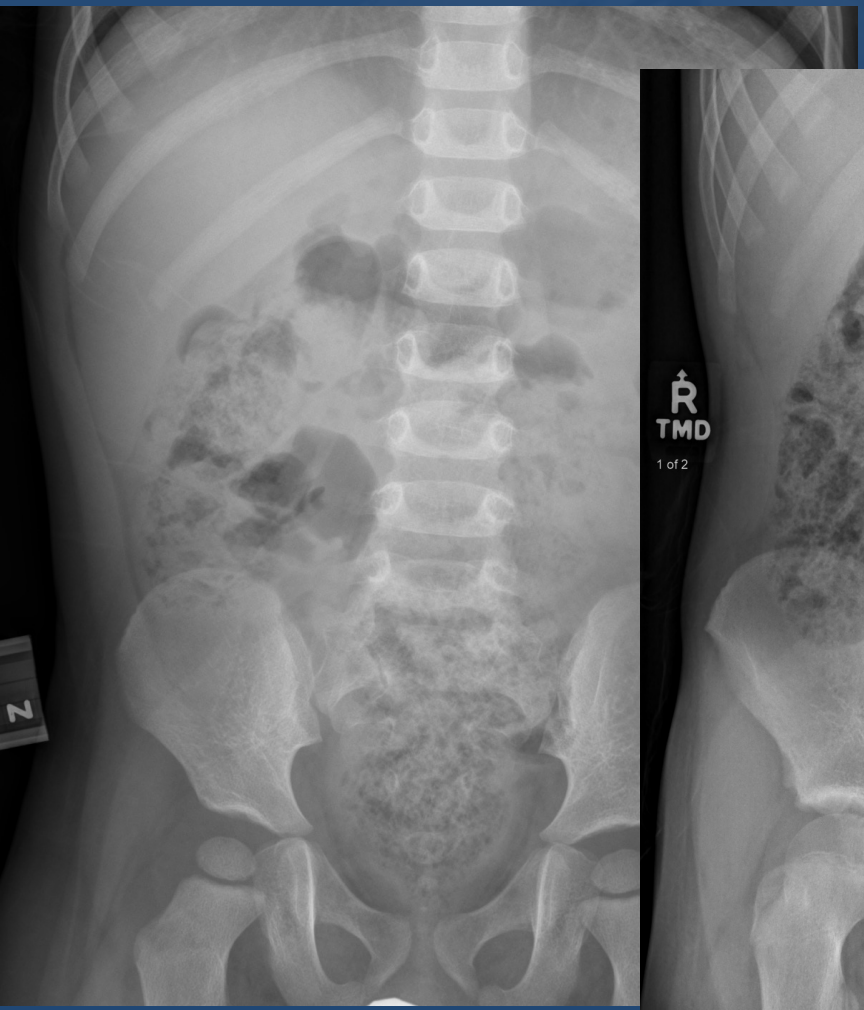

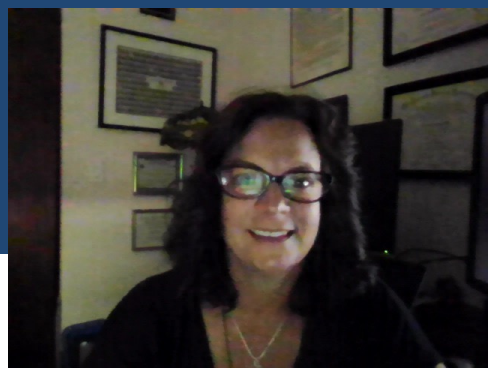

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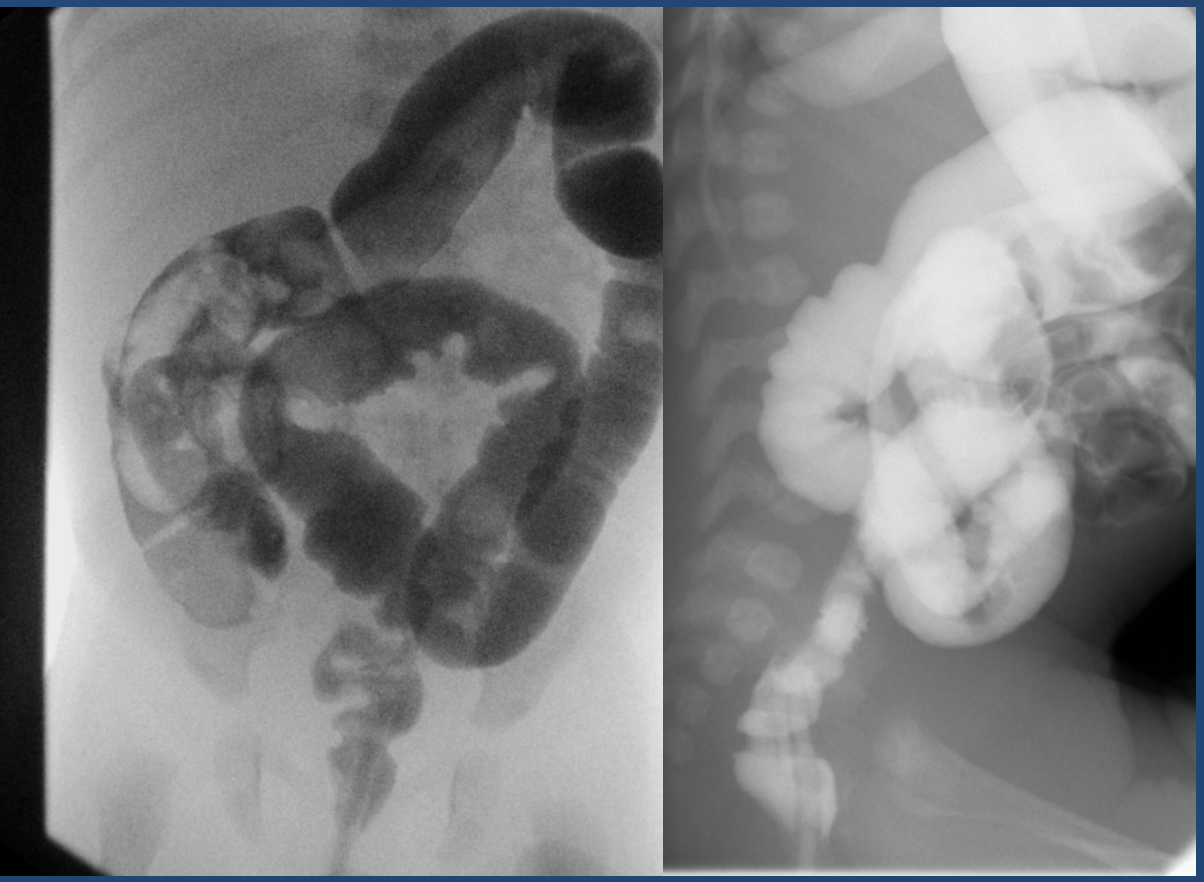
6 day old with Constipation



3 year later with multiple episode of colitis

Does the transition zone reliably delineate aganglionic bowel in Hirschsprung's disease?

- Radiologist agreement of the site of transition zone on contrast enema was 90.6%.
- The concordance between the radiographic transition zone and pathologic extent of aganglionic bowel was 62.5%.
- The subgroup of patients with long-segment HD revealed a concordance of only 25%.
- *Conclusion*: Contrast enema delineation of the transition zone in HD needs to be regarded with caution. This is especially true in long-segment disease, where knowledge of the extent of aganglionic bowel is most crucial to surgical planning.

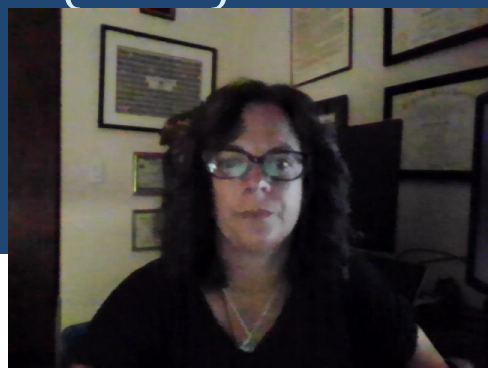
Jamieson, D.H., Dundas, S.E., Belushi, S.A. et al. Pediatr Radiol (2004) 34: 811. https://doi.org/10.1007/s00247-004-1292-7

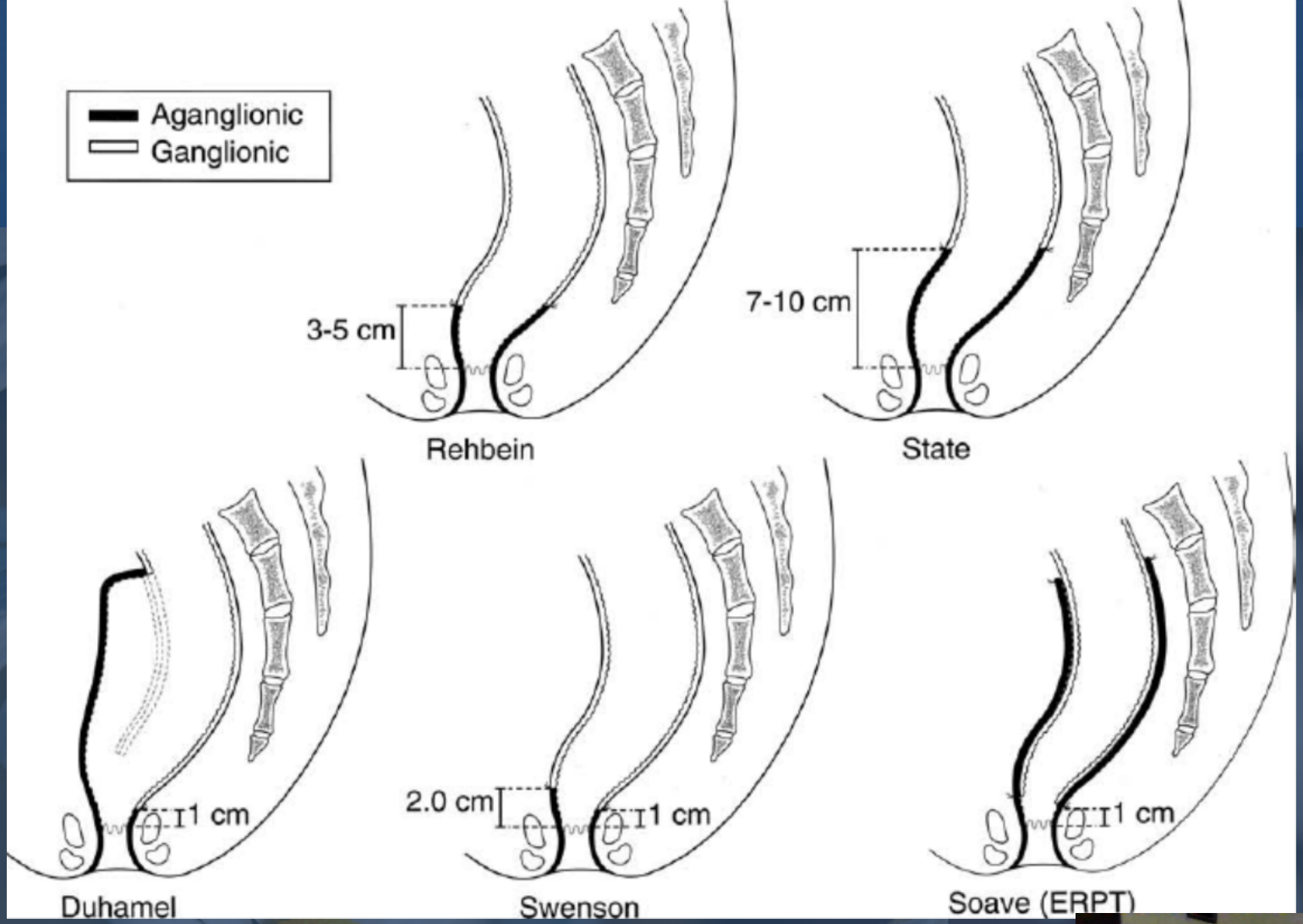
Diagnostic value of the preoperatively detected radiological transition zone in Hirschsprung's disease

- Regarding an association between the radiological TZ and pathological results in HD, namely the 3–5 cm above the TZ with ganglion cells and 3–5 cm below the TZ without ganglion cells.

- We found that the correlation rate between the radiological TZ and pathological results in the rectosigmoid was 88.5% .
- Correlation rate between the radiological TZ and pathological results in the descending colon was 44.4%.
- Correlation rate between a radiological TZ and the pathological results in infants was 69.0%, which is lower than that of older children whose correlation rate was 85.3%

- Chen, X., Xiaojuan, W., Zhang, H. et al. Pediatr Surg Int (2017) 33: 581. https://doi.org/10.1007/s00383-017-4064-9





Schematic illustrations of several operative pull-through techniques for the treatment of Hirschsprung disease.

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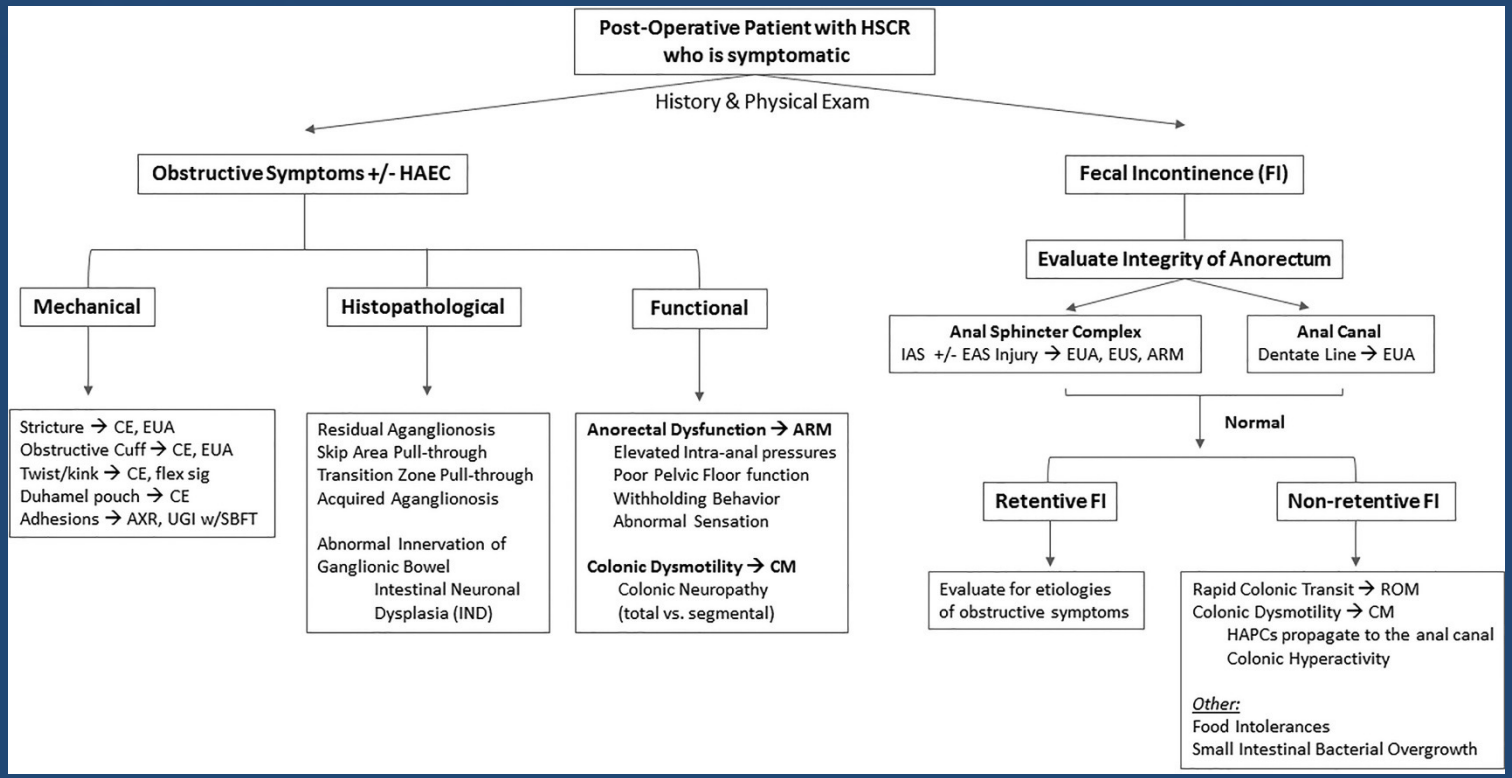
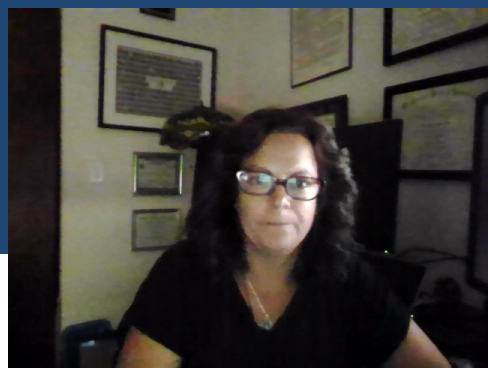
Imaging of the Post surgical patient with HD



Indications to reimage:

- Soiling
- Continued constipation
- Lack of spontaneous bowel movements



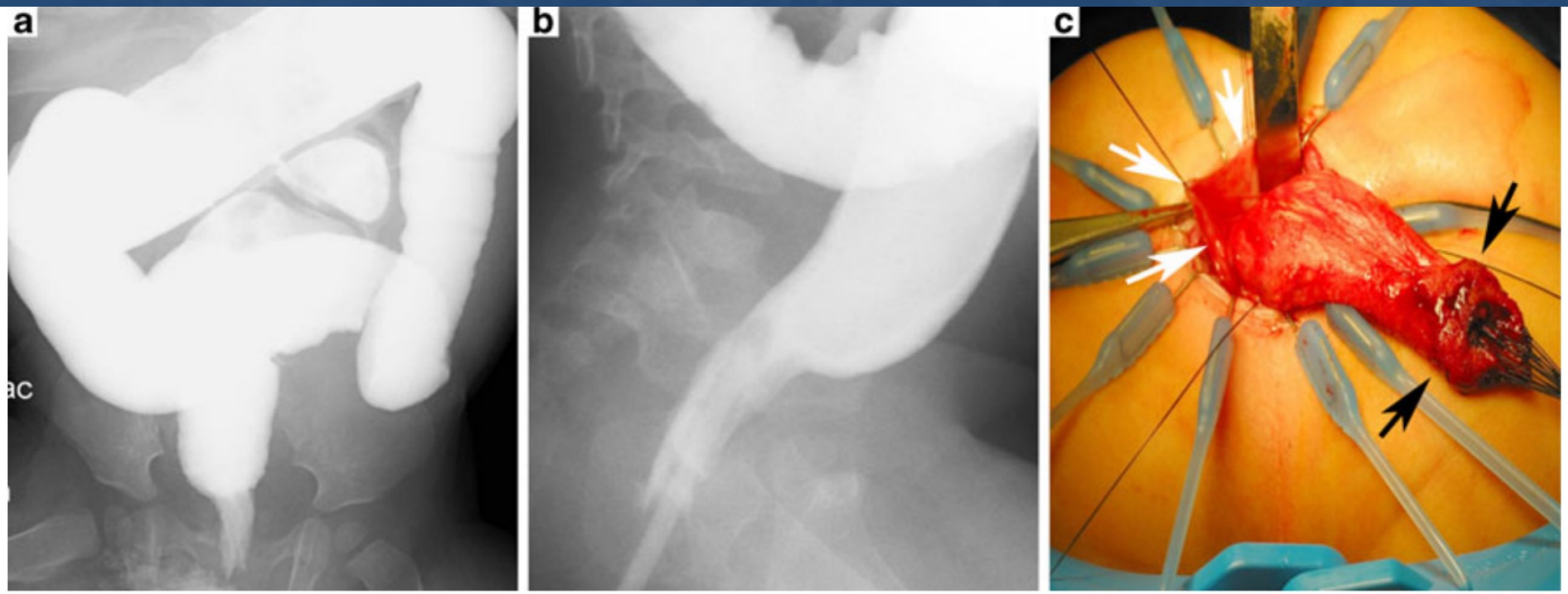
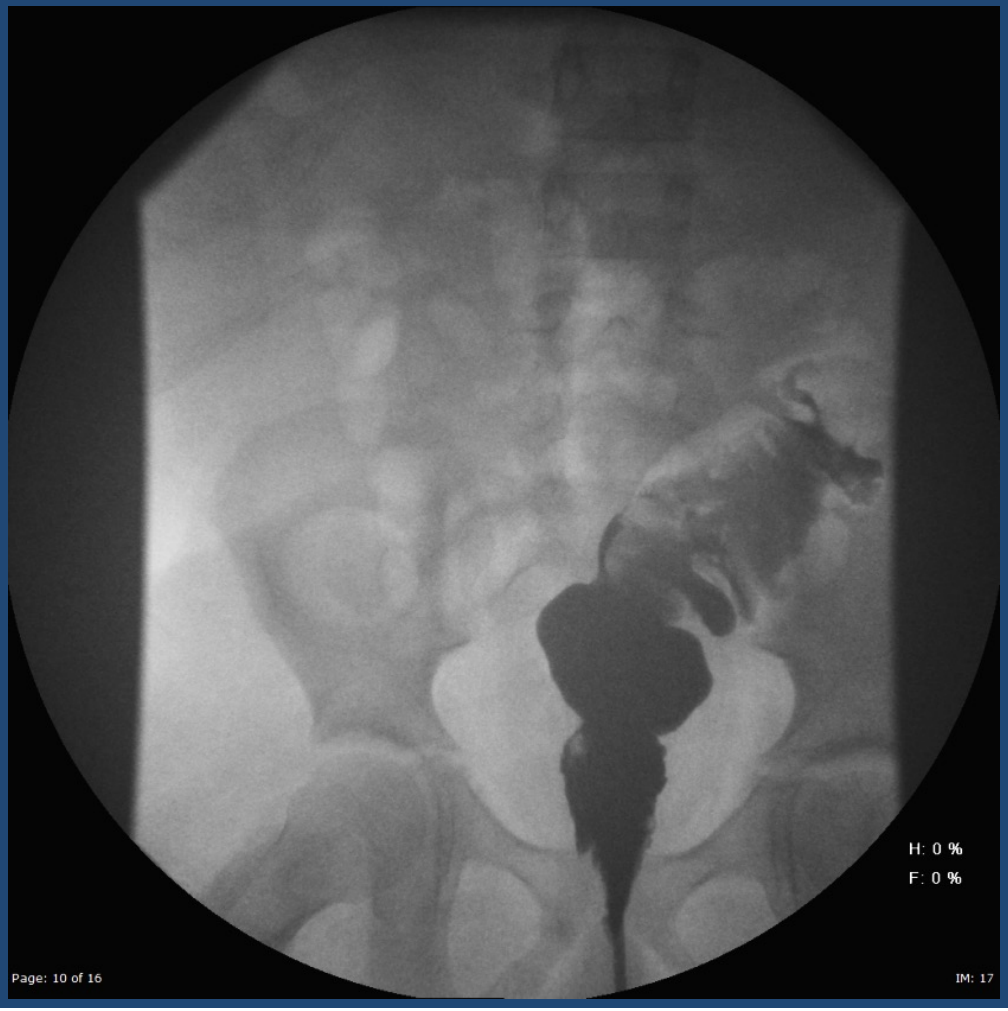
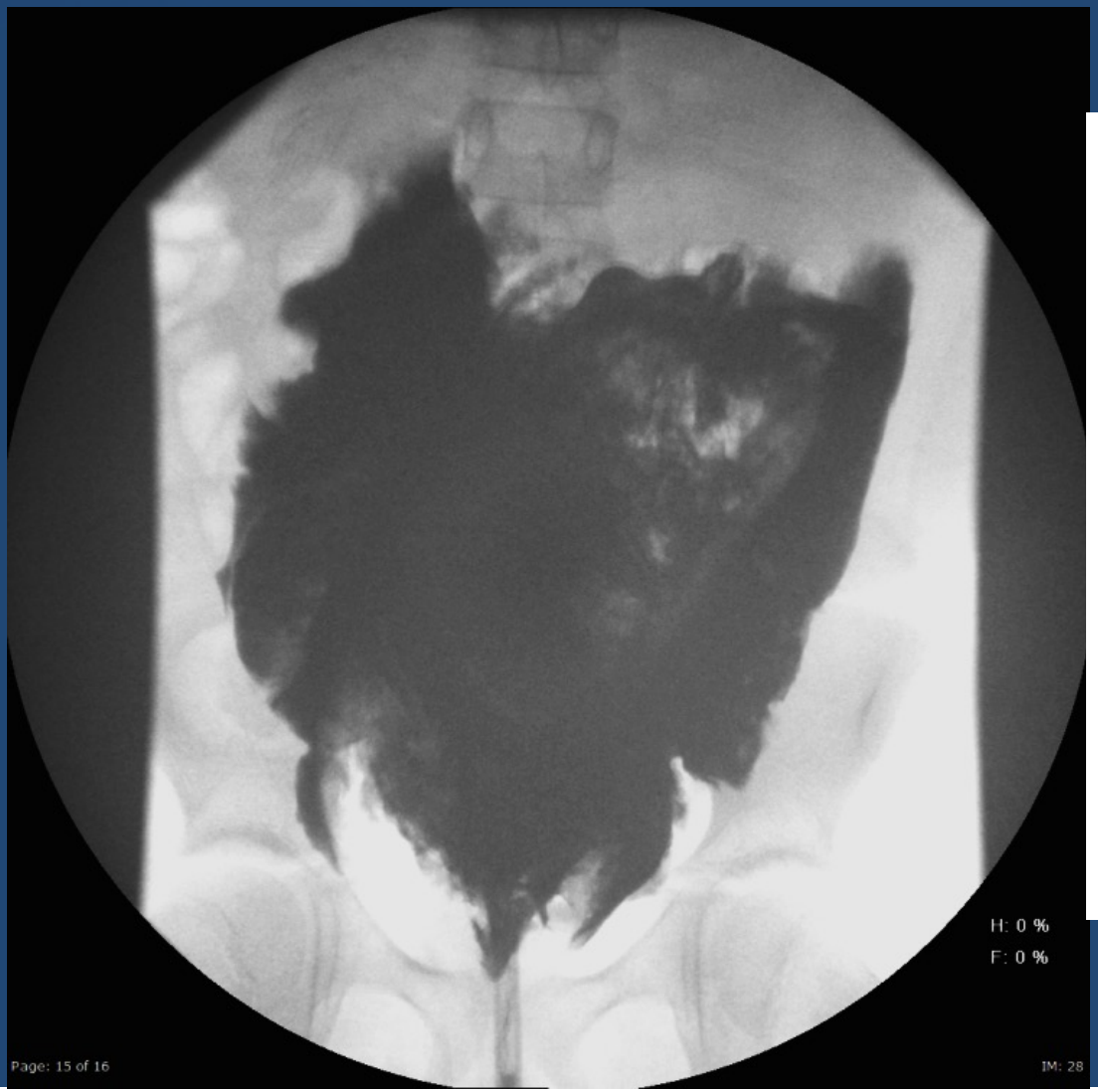


Fig. 4 Obstructing Soave cuff in a 1-year-old boy. **a** Frontal overhead view of full colon from contrast enema shows narrowed distal segment. **b** Lateral fluoroscopic spot view from contrast enema shows widening of the presacral space and narrowed distal segment. **c** Gross surgical photo during repair shows thickened rim of tissue at the distal end of the mobilized colon representing the area of narrowed pull-through (*black arrows*) due to the obstructing retained cuff (*white arrows*)

Garrett KM, Levitt MA, Peña A, Kraus SJ. Contrast enema findings in patients presenting with poor functional outcome after primary repair for Hirschsprung disease. Pediatr Radiol. 2012 Sep;42(9):1099-106. doi: 10.1007/s00247-012-2394-2. Epub 2012 Apr 19. PMID: 22526281.

10 yr male OSH with history of Hirchsprung disease, s/p colostomy, evaluate for stricture





https://s-media-cache-ak0.pinimg.com/736x/99/d559beb9acf3b.jpg

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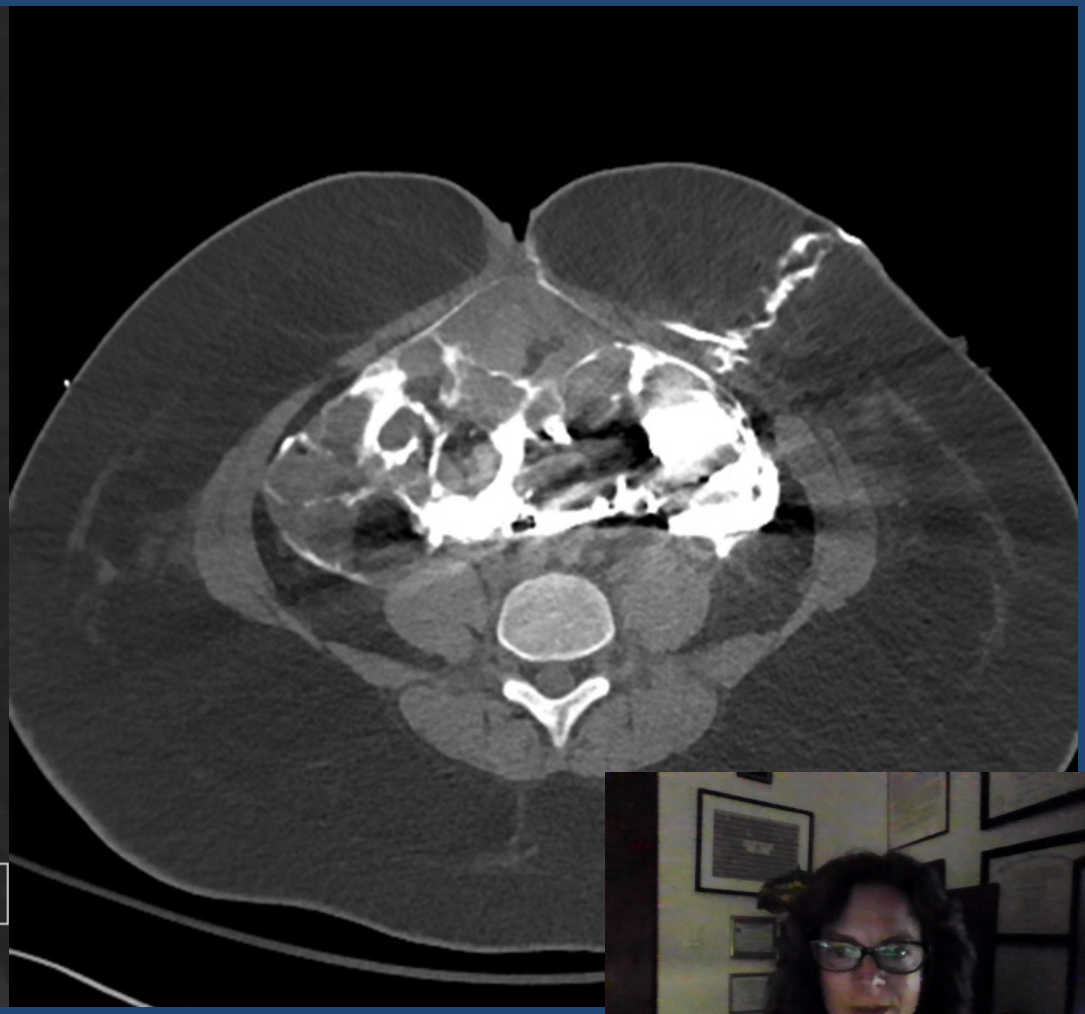
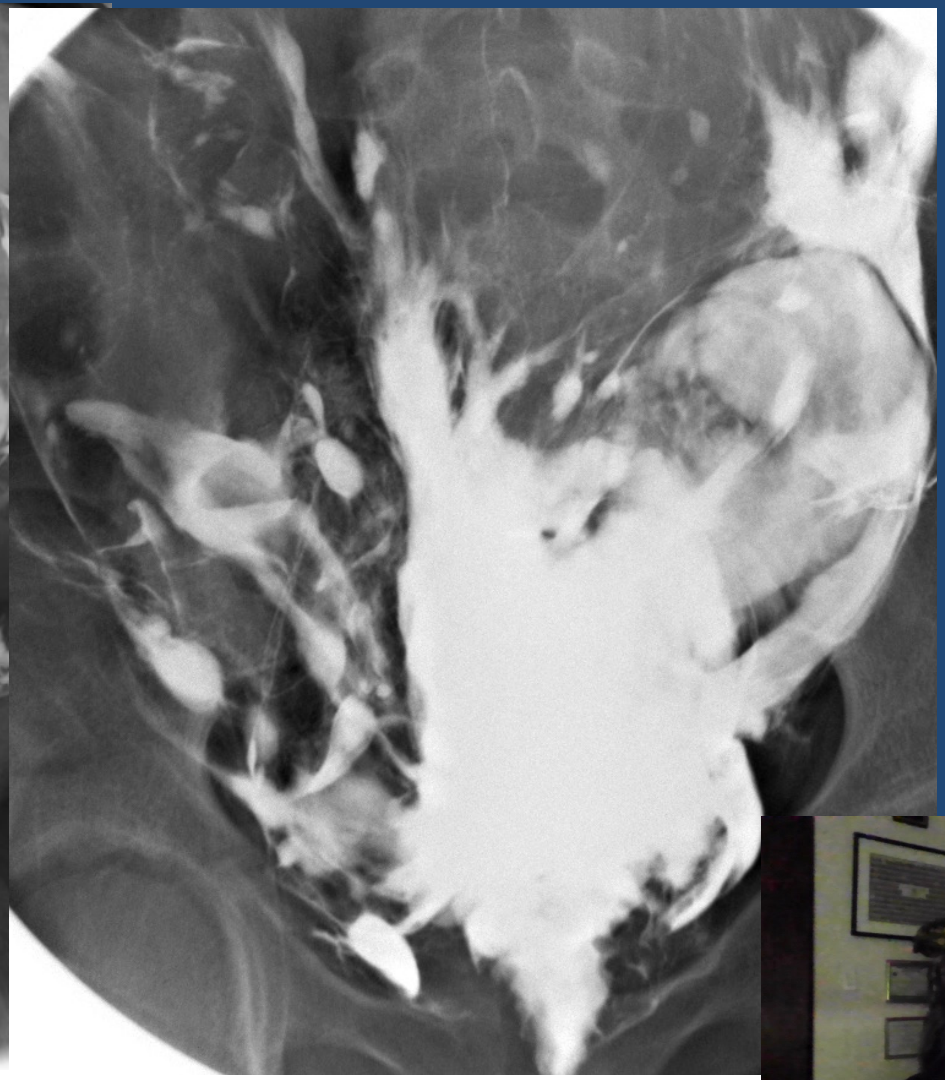
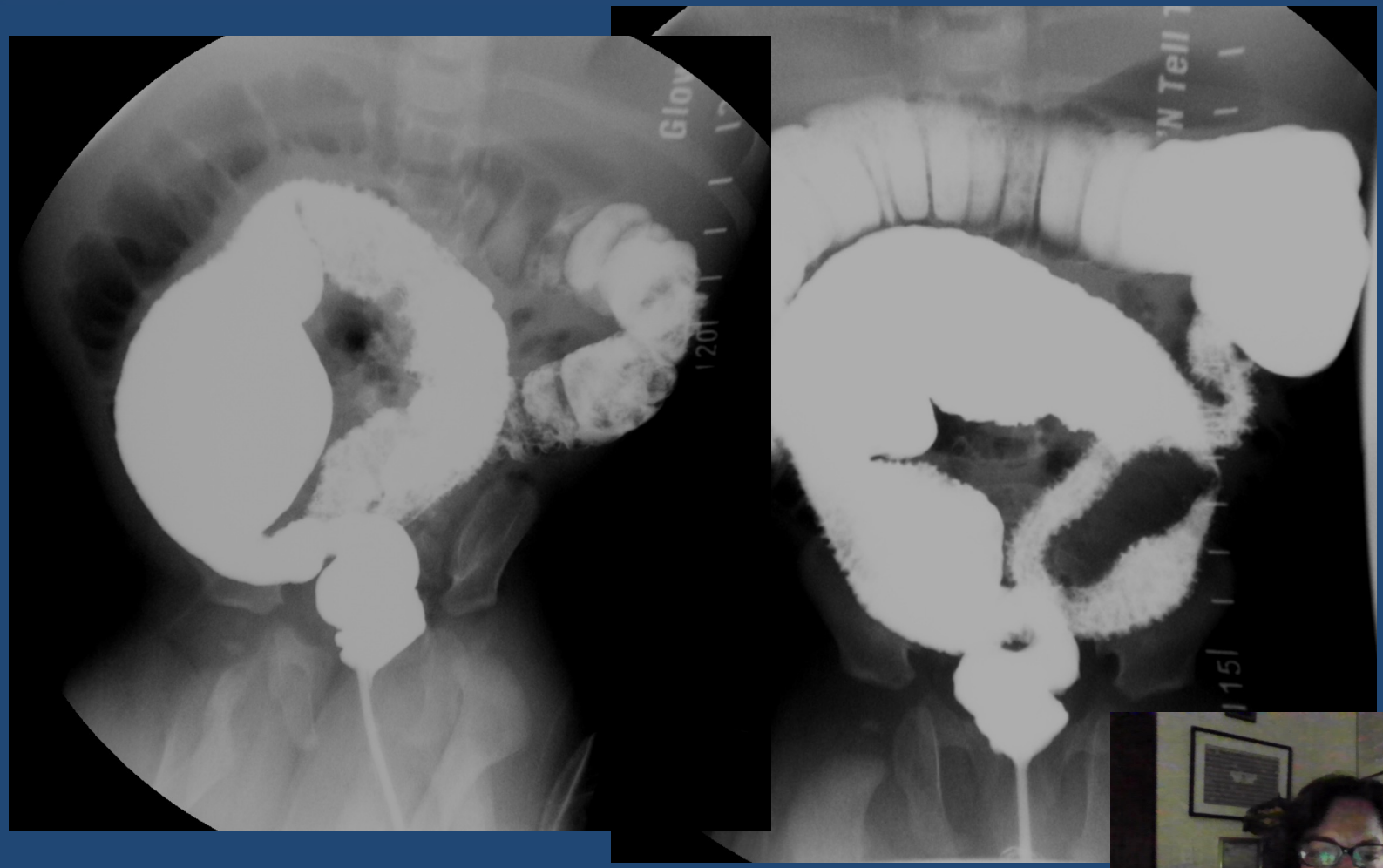
Days later, CT attempted



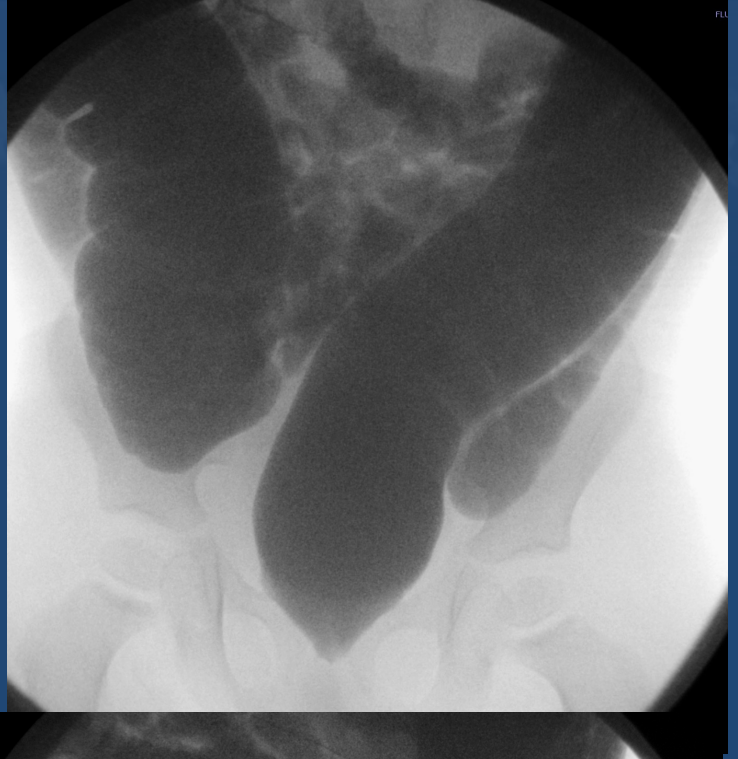
2 years later, complains of continuous ostomy leakage, eval colon anatomy

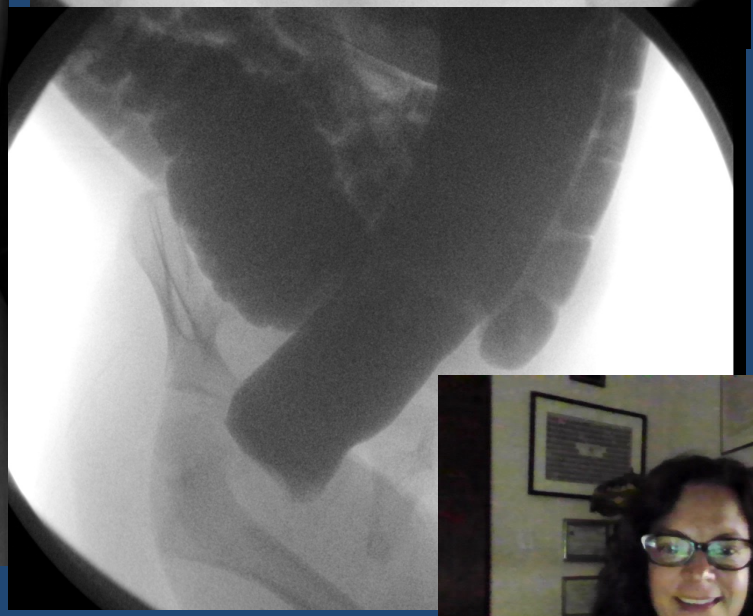


4 mo male




R
BG





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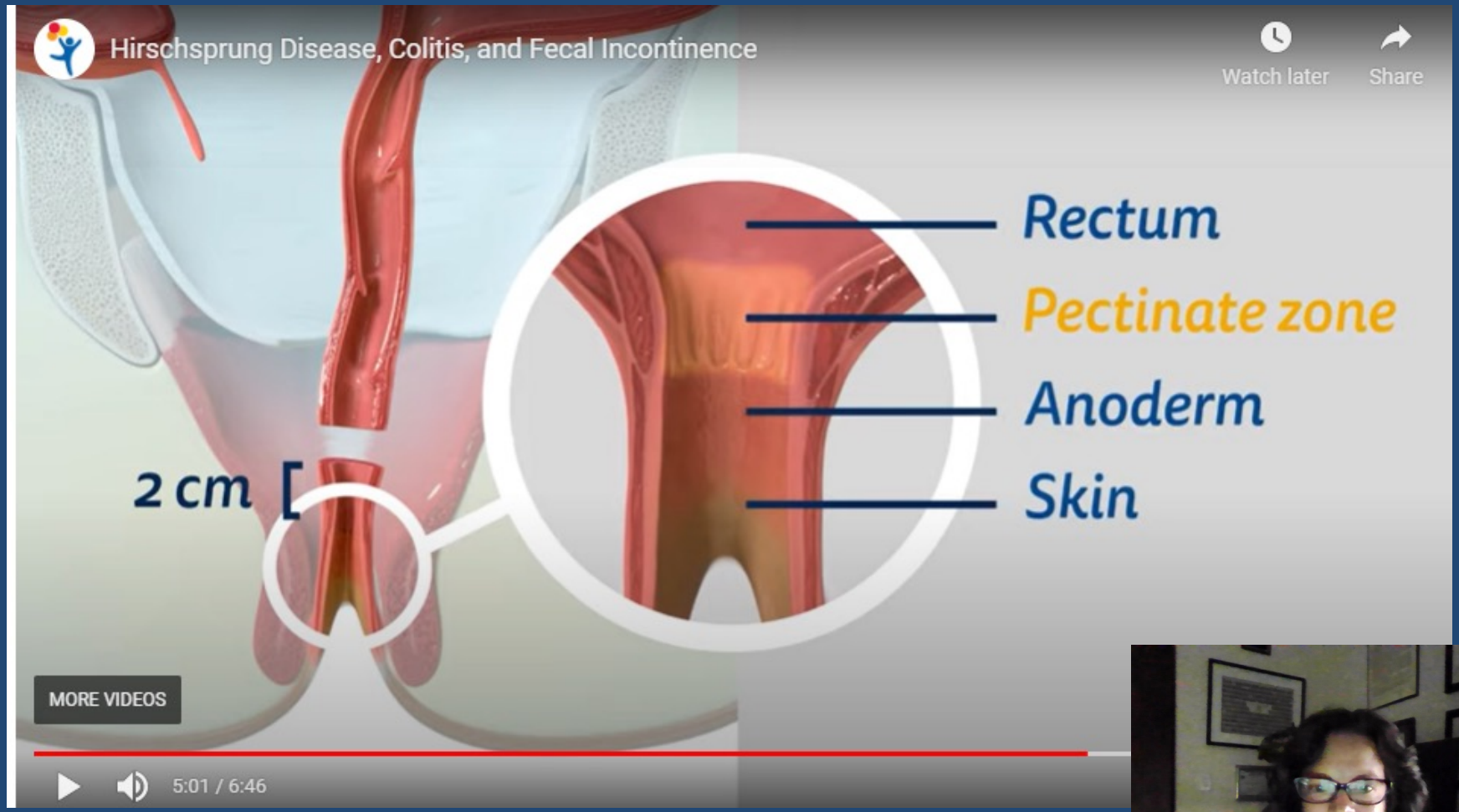

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Radiology Conclusions

1. History and Physical Exam
2. Form a good working relationship with your surgeons, GI team and Radiologists
3. Contrast enema TZ is most sensitive in rectal or rectal sigmoid regions.
4. Contrast enema is not reliable in identifying the pathologic TZ in long segment HD
5. Do not use BARIUM in pediatric colo

Childrens Hospital Colorado HD Video

https://youtube/oSjaG8eVYNs

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