Functional Fecal Incontinence in Pediatrics

CDDW 2016
Montreal, Canada

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Faculty Financial Interest Disclosure

- None

Learning Objectives

- Identify the difference between retentive and non-retentive fecal incontinence in children
- Describe the initial steps in management of a child with fecal incontinence
- Assess the need for additional investigations & referral to other allied health members in children with refractory fecal incontinence
- Recognize the global impact of refractory fecal incontinence on the child and family's quality of life

Overview

- Definitions
- Pathophysiology
- Epidemiology
- Impact
- Management
- Second line investigations
- Summary

Definition

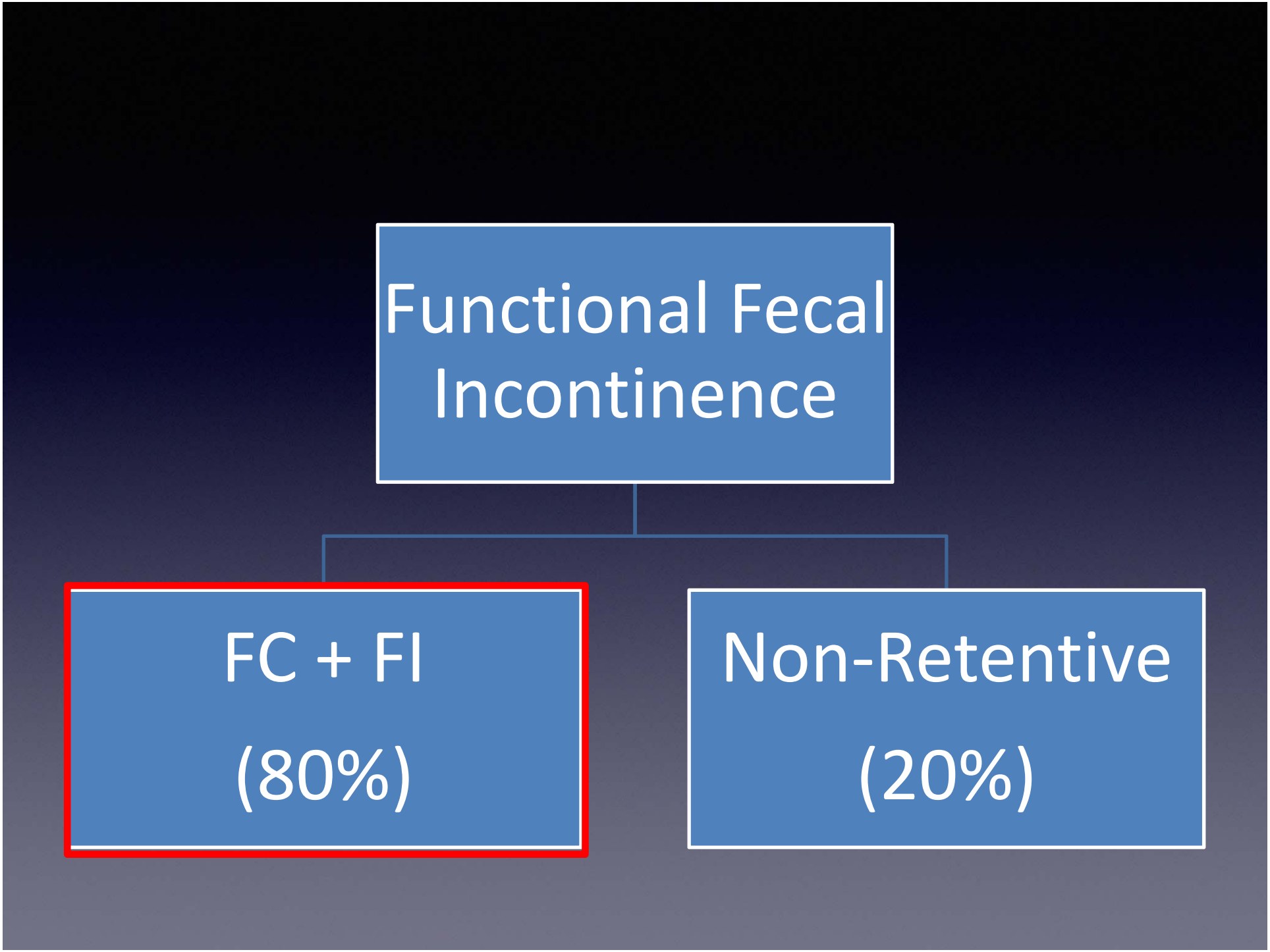
- Fecal incontinence
  - Involuntary passage of fecal material in the underwear
  - Occurring in a child with developmental age ≥ 4 years

Rasquin et al, Gastroenterology. 2006 Apr;130(5):1527-37

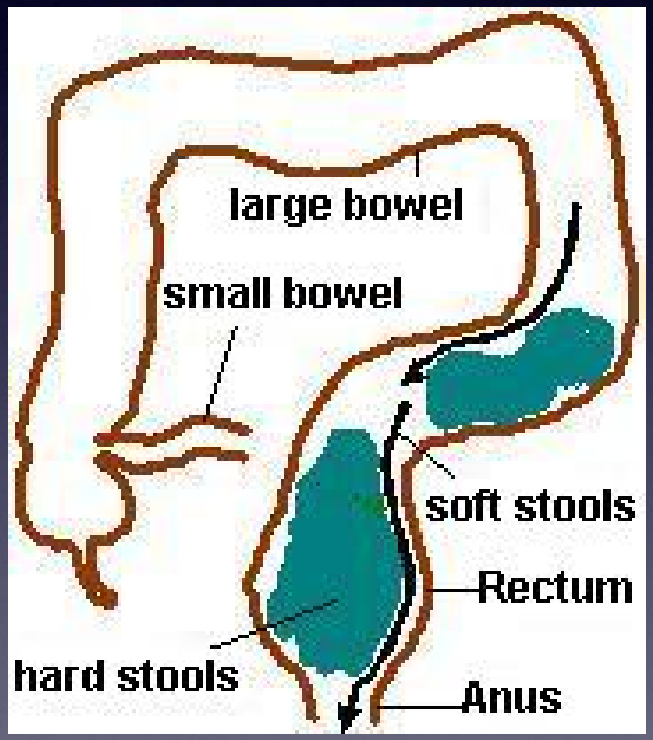
Definition

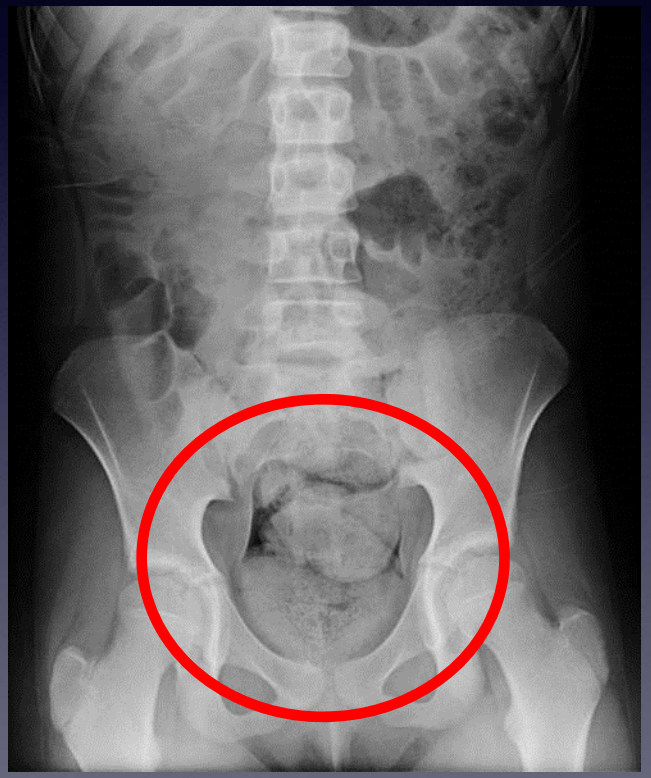
- Fecal incontinence
  - Found in 4 main groups of children:
    - Functional constipation
    - Non-retentive fecal incontinence
    - Children with anorectal malformations
    - Children with spinal abnormalities

Rasquin et al, Gastroenterology. 2006 Apr;130(5):1527-37

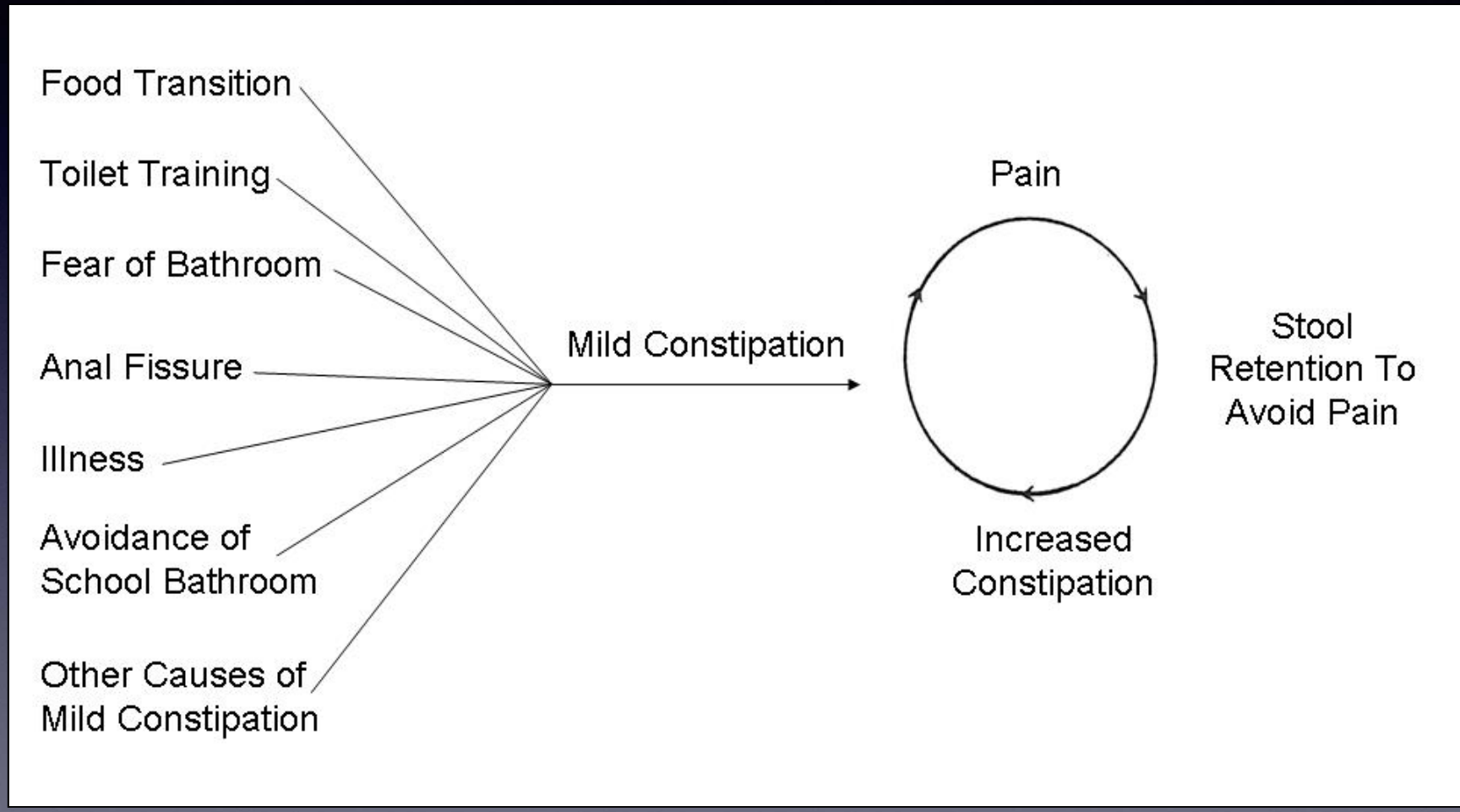

Functional Fecal Incontinence
FC + FI (80%)
Non-Retentive (20%)

Functional Constipation with FI

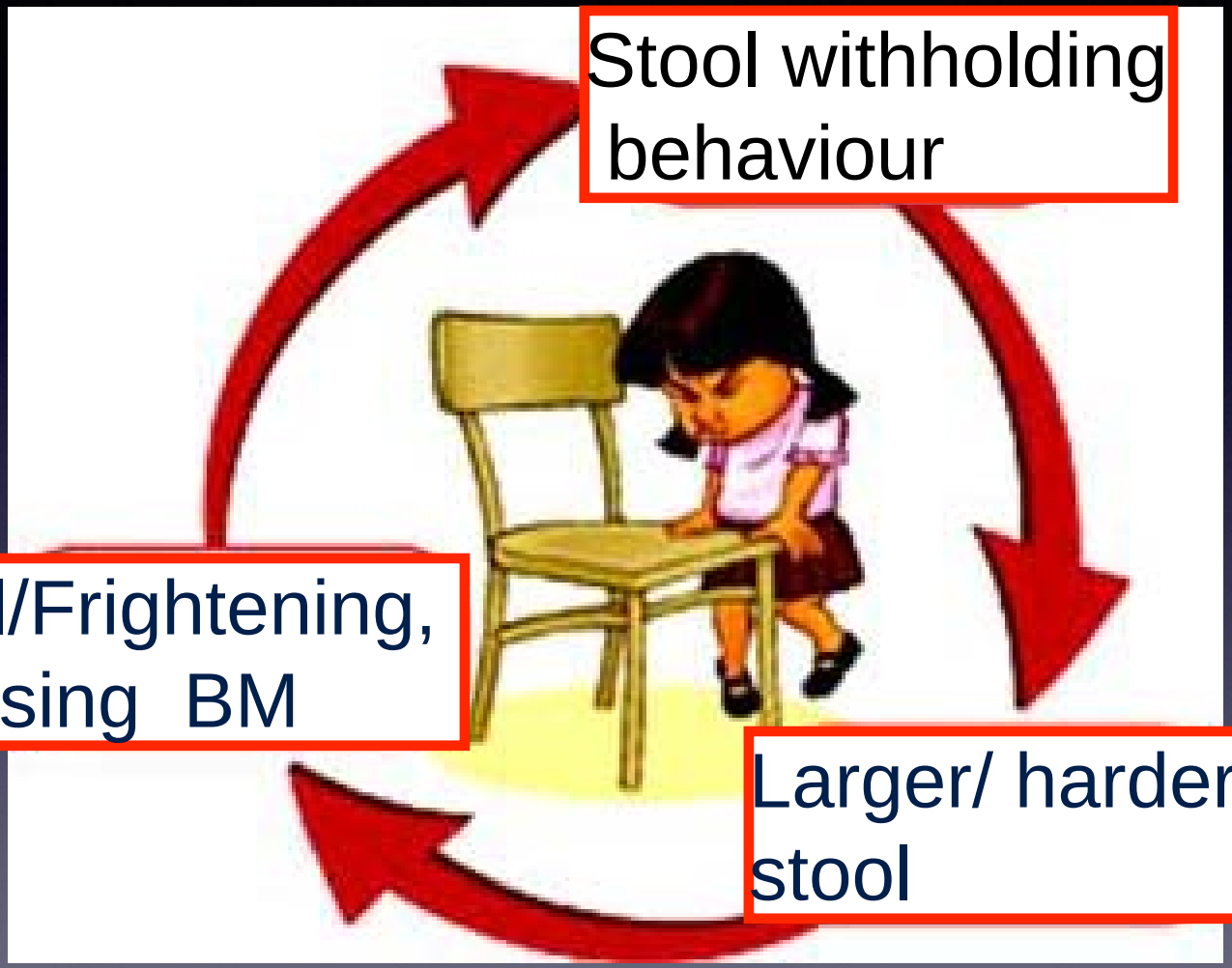


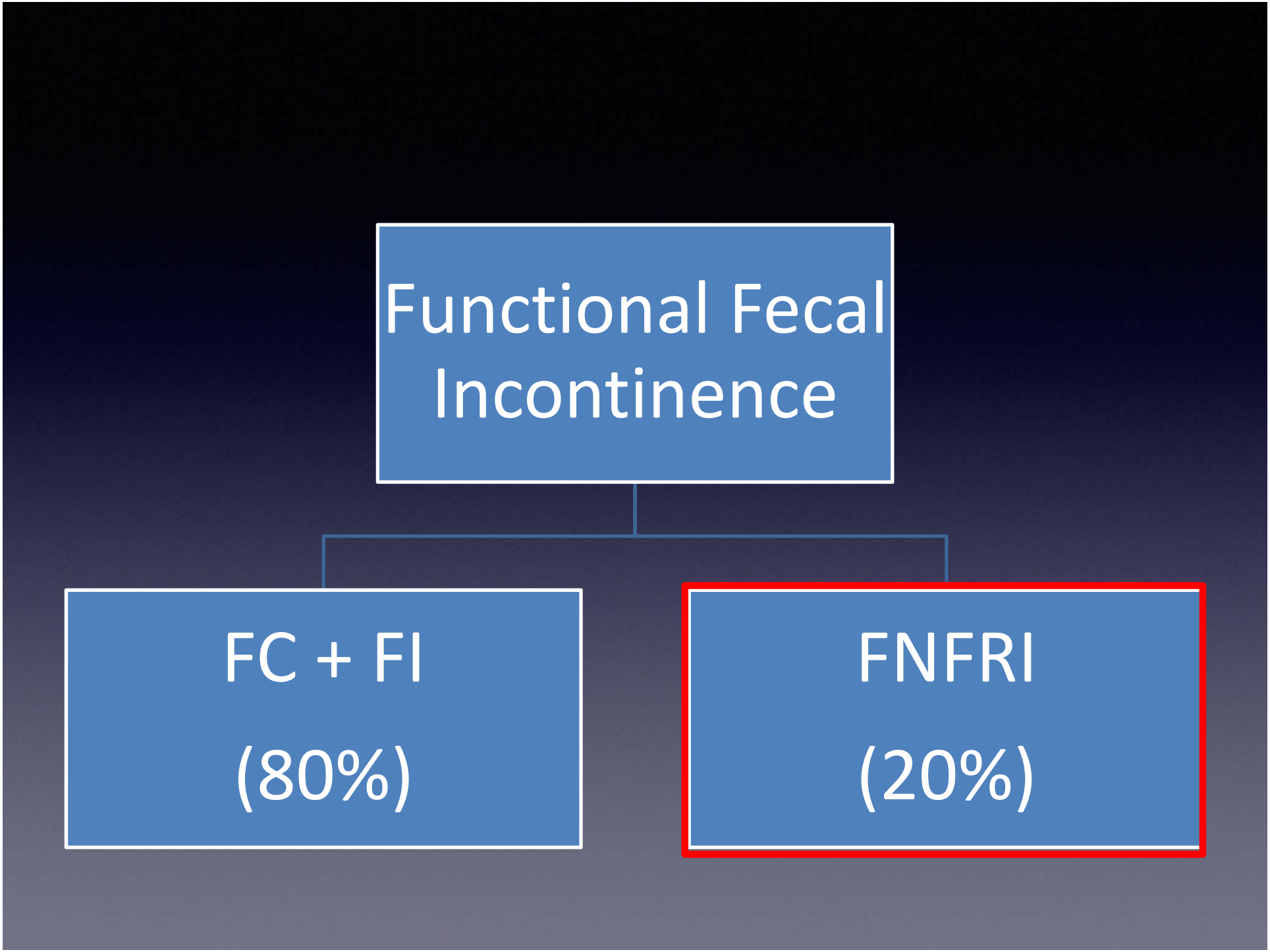


Pain-Retention-Pain Cycle



Rappaport & Levine, Pediatr Clin North Am 1986; 33: 859–69


Stool withholding behaviour
Painful/Frightening, distressing BM
Larger/ harder stool


Functional Fecal Incontinence
FC + FI
(80%)
FNFRI
(20%)

Functional Non-Retentive FI

H3b. Diagnostic Criteria* for Nonretentive Fecal Incontinence

Must include *all* of the following in a child with a developmental age at least 4 years:

1. Defecation into places inappropriate to the social context at least once per month
2. No evidence of an inflammatory, anatomic, metabolic, or neoplastic process that explains the subject's symptoms
3. No evidence of fecal retention

**Criteria fulfilled for at least 2 months before diagnosis*

Rasquin et al, Gastroenterology. 2006 Apr;130(5):1527-37

Functional Non-Retentive FI

- Unknown pathophysiology
  - ? Stress
  - ? Behavioural disorders
    - e.g. ADHD, ASD, Affective disorders

Epidemiology

- Significant problem
  - 3 – 4.4% of children attending general pediatric clinics
  - 21% seeking tertiary care pediatric GI care

Koppen IJ et al, J Pediatr Urol. 2015 Oct 21. pii: S1477-5131(15)00384

Epidemiology

- Age-related
  - Higher rates in younger children
    - Sweden & Netherlands:
      - 4-5 years: 4.1 – 9.8%
      - 11-12 years: 1.6 – 5.6%
    - Sri Lanka:
      - 10 years: 5.4%
      - 16 years: < 1%
- Gender influence
  - Male : female ratio 3:1 – 6:1

Rajindrajith et al, Aliment Pharmacol Ther. 2013 Jan;37(1):37-48
Burgers & Benninga,J Pediatr Gastroenterol Nutr. 2009 Apr;48 Suppl 2:S98-S100

Risk Factors for FI

- Low SES
- Toilet facilities
  - Inadequate
  - Unclean or unhygenic toilets
- Delay in consult
- Urban areas
- War zones
- Hospitalization
- Abuse
  - Emotional, physical

Rajindrajith et al, Aliment Pharmacol Ther. 2013 Jan;37(1):37-48
Rajindrajith J Pediatr Gastroenterol Nutr. 2015 Oct 16

Impact of FI

- Lack of control
- Lower self-worth
- Family stress and dysfunction
- Stigmatization
- Abuse
- Significantly lower HRQoL scores
- Can lead to low self-esteem and social withdrawal if symptoms persist into adulthood

Rajindrajith et al, Aliment Pharmacol Ther. 2013 Jan;37(1):37-48
Kovacic et al, J Pediatr. 2015 Jun;166(6):1482-7
Wald & Sigurdsson, Best Pract Res Clin Gastroenterol. 2011 Feb;25(1):19-27
Landman et al, J Dev Behav Pediatr. 1986 Apr;7(2):111-3

Initial management?

Initial Management

- Education
  - Explain diagnosis, pathophysiology
  - Use simple language and allow time for parent questions
  - Review goals of treatment
  - Review medications, mechanism of action, and duration of treatment
  - Review natural history

Initial Management

- Potentially long road to recovery…
  - At 1 year follow-up, 41-67% of constipated children (with or without fecal incontinence) are not fully recovered
  - 31-52% of children remain symptomatic at 4-10 years after diagnosis and treatment

Yeung & Di Lorenzo (2013). Constipation in Children (Eds. Núñez & Fabbro). New York: Nova Science

Initial Management

- Disimpaction
  - Key step in treating fecal incontinence
  - Methods
    - Manual: immediate relief, unpleasant, +/- GA, +/- injury
    - Rectal: fast onset, may compound problem
    - Oral:
      - Route of choice
      - PEG3350 as effective as daily enemas; 1 – 1.5 g/kg PEG3350 x 3 days (75% disimpaction rate)
      - Other laxative types also have been successfully used in literature

Bekkali et al, Pediatrics. 2009 Dec;124(6):e1108-15
Youssef et al, J Pediatr. 2002 Sep;141(3):410-4

Maintenance

- Behavioural modification
  - Avoid ignoring body cues
  - Scheduled sit times
  - Address any punitive or abusive behaviour

Maintenance

- Maneuvers to facilitate pelvic floor relaxation
  - Step stool
  - Blowing bubbles

Maintenance

- School plan
  - Emergency kit
  - Address barriers to success

Maintenance

- Ongoing pharmacotherapy

TABLE 6. Dosages of most frequently used oral and rectal laxatives

Oral laxatives	Dosages
Osmotic laxatives	
Lactulose	1–2 g/kg, once or twice/day
PEG 3350	Maintenance: 0.2–0.8 g · kg^{-1} · day^{-1}
PEG 4000	Fecal disimpaction: 1–1.5 g · kg^{-1} · day^{-1} (with a maximum of 6 consecutive days)
Milk of magnesia (magnesium hydroxide)	2–5 y: 0.4–1.2 g/day, once or divided
	6–11 y: 1.2–2.4 g/day, once or divided
	12–18 y: 2.4–4.8 g/day, once or divided
Fecal softeners	
Mineral oil	1–18 y: 1–3 mL · kg^{-1} · day^{-1}, once or divided, max 90 mL/day
Stimulant laxatives	
Bisacodyl	3–10 y: 5 mg/day
	>10 y: 5–10 mg/day
Senna	2–6 y: 2.5–5 mg once or twice/day
	6–12 y: 7.5–10 mg/day
	>12 y: 15–20 mg /day
Sodium picosulfate	1 mo–4 y: 2.5–10 mg once/day
	4–18 y: 2.5–20 mg once/day
Rectal laxatives/enemas	
Bisacodyl	2–10 y: 5 mg once /day
	>10 y: 5–10 mg once /day
Sodium docusate	<6 y: 60 mL
	>6 y: 120 mL
Sodium phosphate	1–18 y: 2.5 mL/kg, max 133 mL/dose
NaCl	Neonate <1 kg: 5 mL, >1 kg: 10 mL
	>1 y: 6 mL/kg once or twice/day
Mineral oil	2–11 y: 30–60 mL once/day
	>11 y: 60–150 mL once/day

PEG = polyethylene glycol.

Tabbers et al, J Pediatr Gastroenterol Nutr. 2014 Feb;58(2):258-74

Maintenance

type 1		looks like: **rabbit droppings** Separate hard lumps, like nuts (hard to pass)
type 2		looks like: **bunch of grapes** Sausage-shaped but lumpy
type 3		looks like: **corn on cob** Like a sausage but with cracks on its surface
type 4		looks like: **sausage** Like a sausage or snake, smooth and soft
type 5		looks like: **chicken nuggets** Soft blobs with clear-cut edges (passed easily)
type 6		looks like: **porridge** Fluffy pieces with ragged edges, a mushy stool
type 7		looks like: **gravy** Watery, no solid pieces ENTIRELY LIQUID

Maintenance

- ?Dietary fiber
- ?Prebiotics
- ?Probiotics

Tabbers et al, J Pediatr Gastroenterol Nutr. 2014 Feb;58(2):258-74

Maintenance

- Follow-up!!
  - Monitor compliance
  - Medication adjustment
  - Identify obstacles to success
  - Provide reassurance and positive reinforcement

FNRFI - Management

- Similar approach to FC + FI except…
  - …AVOID LAXATIVES!!
- Behavioural treatment = cornerstone of therapy
- Often benefit from referral to Psychology
- Consider loperamide

What do you do with refractory FI?

Refractory FI

- Medications:
  - Inadequate?
  - Discontinued too soon?
  - Poor compliance?
- Are we being aggressive/rigorous enough?
- Is it the correct diagnosis?
- Do we need further investigations?
- Is it time for neurogastroenterology?
- Is it time for surgical intervention?

Refractory FI

Complimentary investigations

- TTG
- TSH
- Electrolytes
- Calcium
- Lead level
- Urine culture

Refractory FI
Medications

- Lubiprostone (Amitiza™)
- Linaclotide (Constella™)
- Prucalopride (RESOTRAN™)

Refractory FI
Botox

- DDW 2015 , poster, Su 1175
  - Anal Botulinum Toxin Injection Is Effective, Safe and Can Be Useful in Patients With Both Normotensive and Hypertensive Anal Pressure
    - Retrospective follow-up over 7 year period
      - 142 patients
      - Aged 8 mos -19 yrs
      - 70% response rate, >6 month duration in 33%
      - 17%>1 y

C. Zar-Kessler

Refractory FI
Complementary investigations
Colonic transit studies

- Functional studies that examine transit through the colon
- Techniques:
  - Radioopaque markers (aka “SITZMARKS®”)
  - Scintigraphy
  - Wireless motility capsule

Refractory FI
Complementary investigations
Radioopaque marker study

Slow Transit Constipation

Evacuation Disorder

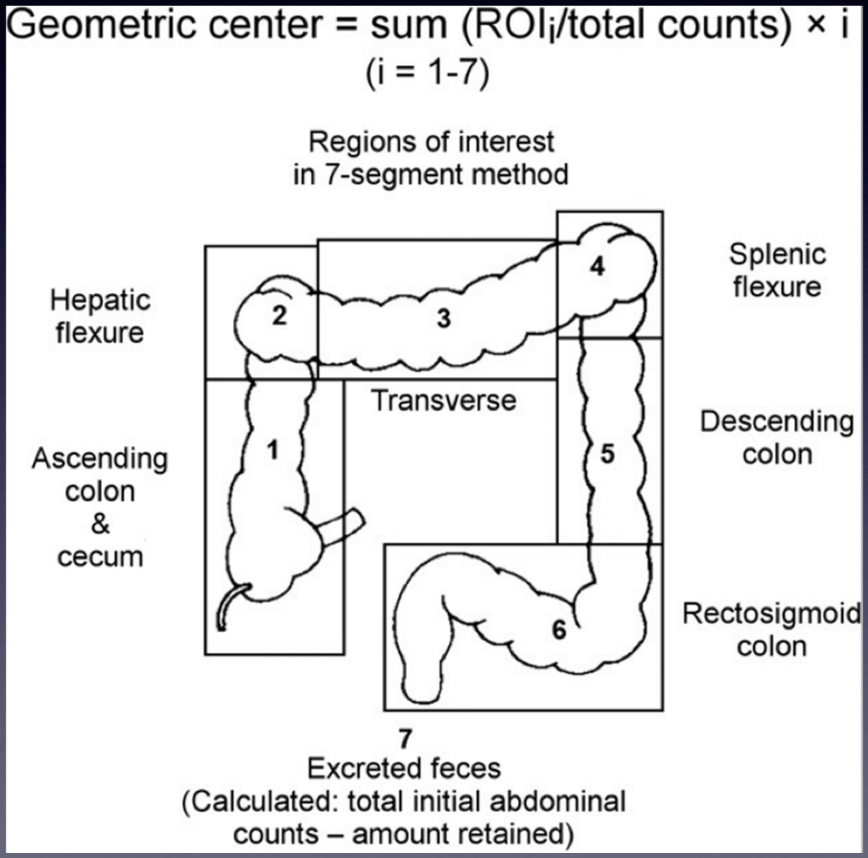
Refractory FI
Complementary investigations
Colonic scintigraphy

- Involves the ingestion of a radioactive isotope
- Progression followed with large-field view gamma camera
- Correlates with radioopaque marker transit studies
- Two delivery methods:
  - Liquid slurry
  - pH-sensitive polymer coated capsule

Maurer, J Nucl Med. 2015 Sep;56(9):1395-400

Refractory FI
Complementary investigations
Colonic scintigraphy

- Transit is assessed by calculating the geometric center
  - Weighted average of isotope distribution within the colon and stool



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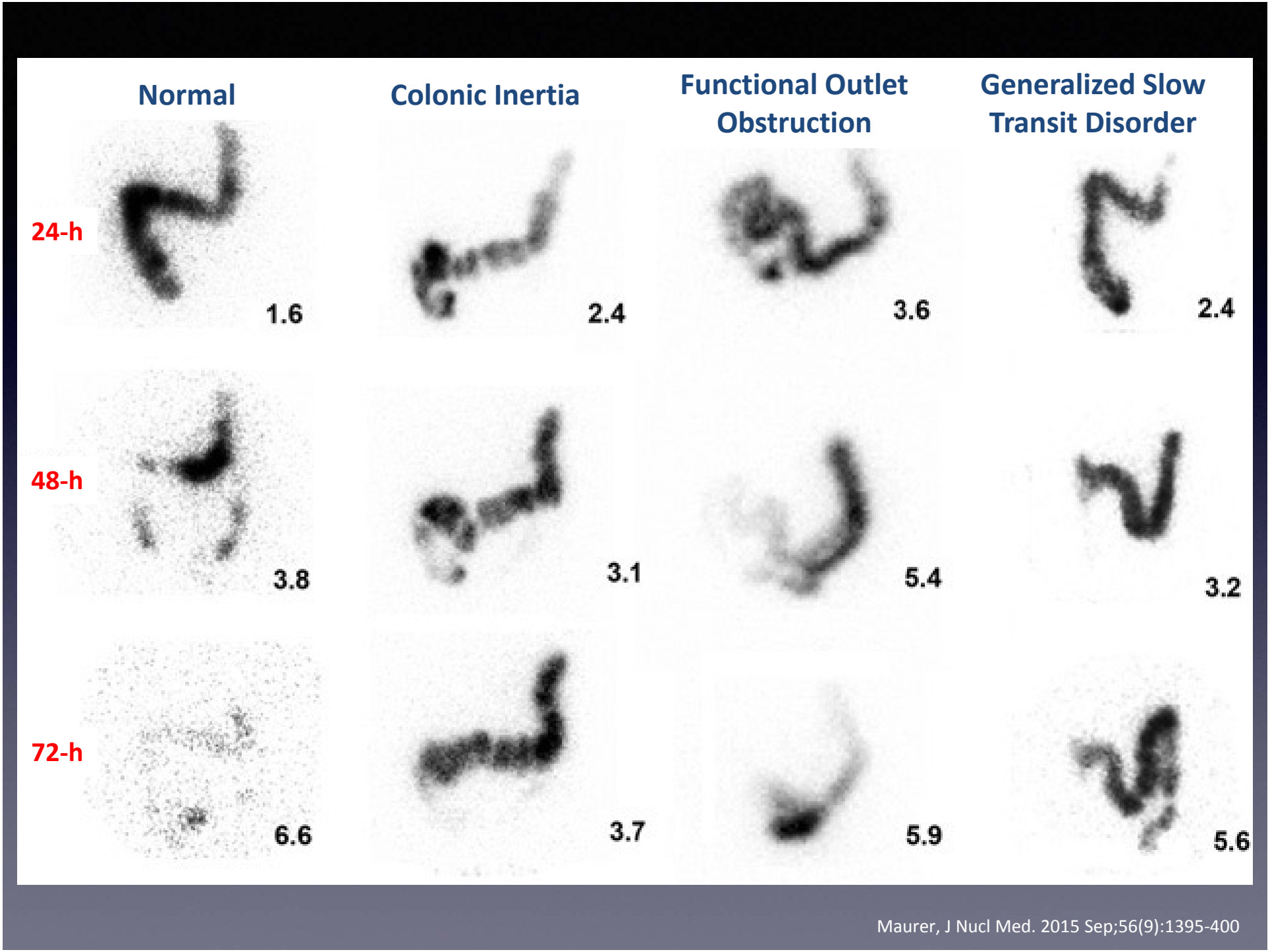


Maurer, J Nucl Med. 2015 Sep;56(9):1395-400

Refractory FI
Complementary investigations
Wireless motility capsule

- Wireless motility capsule
  - Measures pressure, pH, temperature
  - Using all parameters, can estimate:
    - Gastric empting time
    - Colonic transit time
    - Whole gut transit time

Refractory FI
Complementary investigations
Colonic transit studies

- Based on current guidelines, only radioopaque marker transit studies deemed useful
  - "If diagnosis is unclear, may help distinguish between FC + FI and FNRFI"

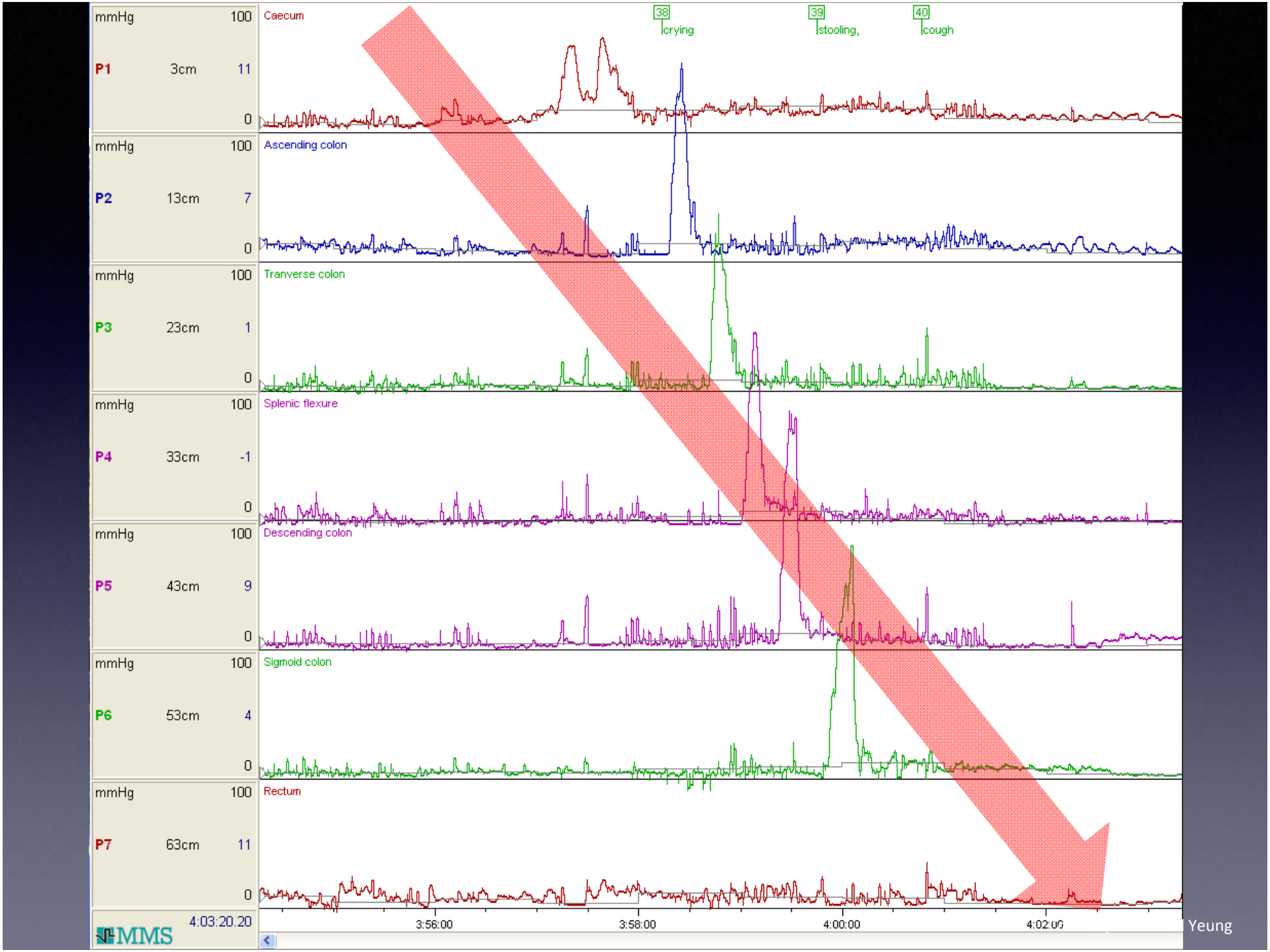
Tabbers et al, J Pediatr Gastroenterol Nutr. 2014 Feb;58(2):258-74

Refractory FI
Complementary investigations
Colonic manometry

- Measures luminal pressure changes over time
- Solid state versus water-perfused

Refractory FI
Complementary investigations
Colonic manometry

- Components of the study
  - Fasting phase
  - ± Stimulation
  - Response to caloric load
- Total duration: 4 – 6 hours

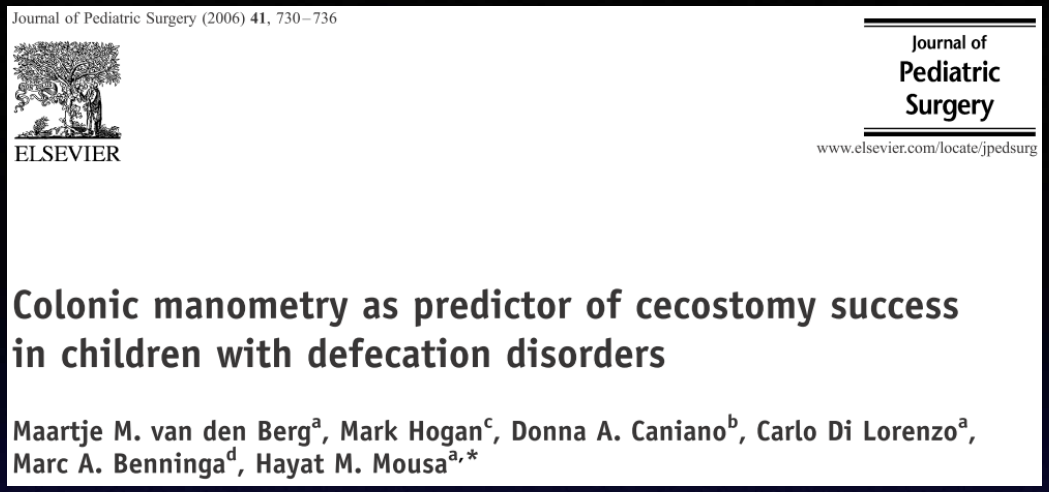

mmHg
100
Caecum
38
crying
39
stooling,
40
cough
P1
3cm
11
0
mmHg
100
Ascending colon
P2
13cm
7
0
mmHg
100
Tranverse colon
P3
23cm
1
0
mmHg
100
Splenic flexure
P4
33cm
-1
0
mmHg
100
Descending colon
P5
43cm
9
0
mmHg
100
Sigmoid colon
P6
53cm
4
0
mmHg
100
Rectum
P7
63cm
11
0
4:03:20.20
MMS
3:56:00
3:58:00
4:00:00
4:02:00

Yeung

Refractory FI
Complementary investigations
Colonic manometry

- Severe constipation, unresponsive to medical therapy and associated with slow transit without evidence of an evacuation disorder
- Clarify the pathophysiology of persistent symptoms after removal of aganglionic segment in Hirschsprung's disease
- Evaluation of diverted colon before possible closure of diverting ostomy
- Predict response to antegrade enemas via cecostomy

Camilleri et al, Neurogastroenterol Motil. 2008;20(12):1269-82.



Journal of Pediatric Surgery (2006) 41, 730–736

ELSEVIER

Journal of Pediatric Surgery

www.elsevier.com/locate/jpedsurg

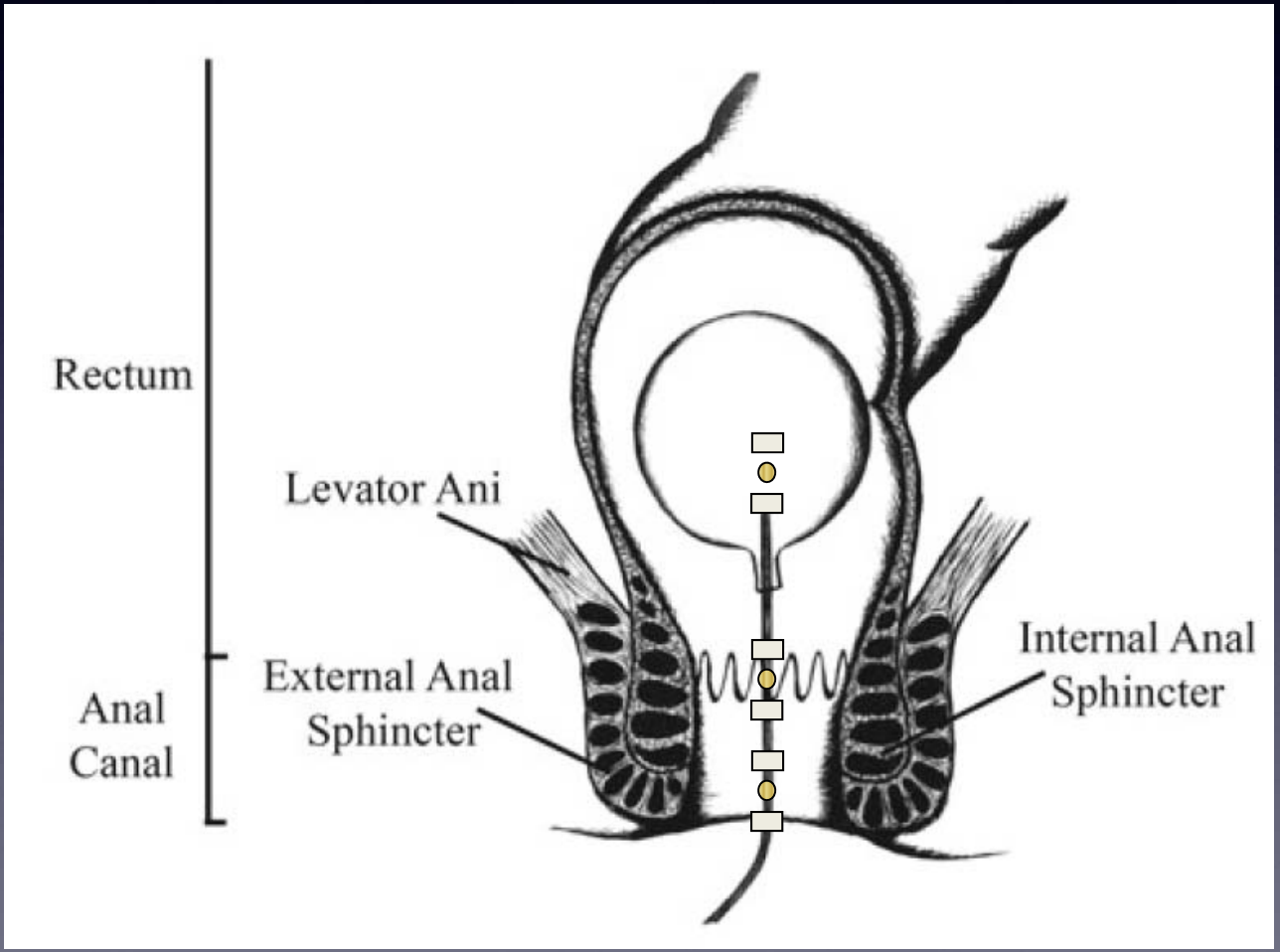
Colonic manometry as predictor of cecostomy success in children with defecation disorders

Maartje M. van den Berg[a], Mark Hogan[c], Donna A. Caniano[b], Carlo Di Lorenzo[a], Marc A. Benninga[d], Hayat M. Mousa[a,*]

- 32 children with chronic constipation
- Evaluated with colonic manometry and treated with cecostomy
- Patients with HAPCs present 11X more likely to have a successful outcome post-cecostomy
  - "Succesful" = normal bowel movement frequency and no/occasional fecal incontinence

van den Berg et al, J Pediatr Surg. 2006 Apr;41(4):730-6

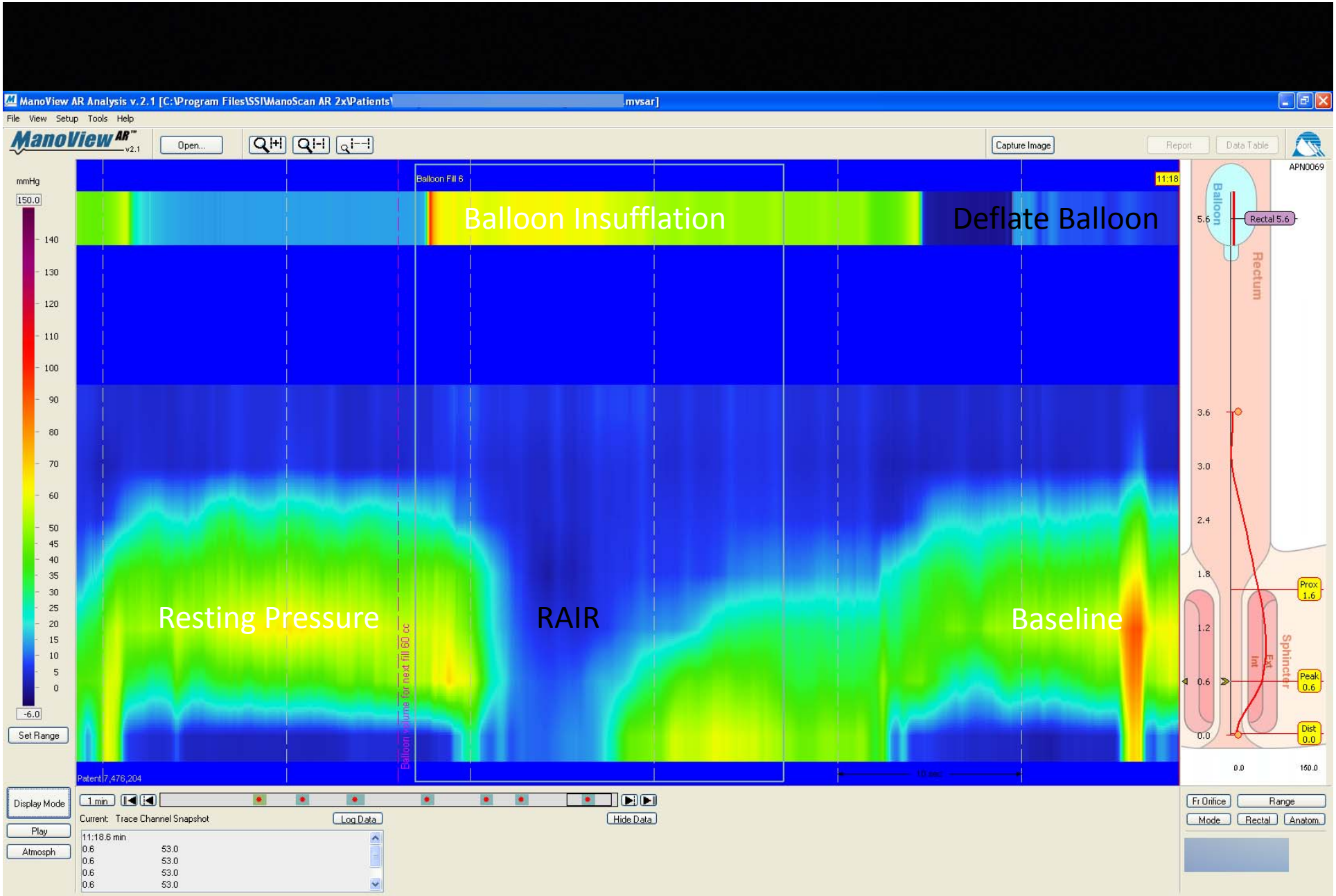
Refractory FI
Complementary investigations
Anorectal manometry



Faure et al, Pediatric Neurogastroenterology: Gastrointestinal Motility and Functional Disorders in Children 2013

Refractory FI
Complementary investigations
Anorectal manometry

- Components of study
  - Presence/absence of the rectoanal inhibitory reflex (RAIR)



Courtesy Dr. Alfred Yeung

Refractory FI
Complementary investigations
Anorectal manometry

- Components of study
  - Presence/absence of the rectoanal inhibitory reflex (RAIR)
  - Resting pressure
  - Rectal sensation
  - Pelvic floor dynamics

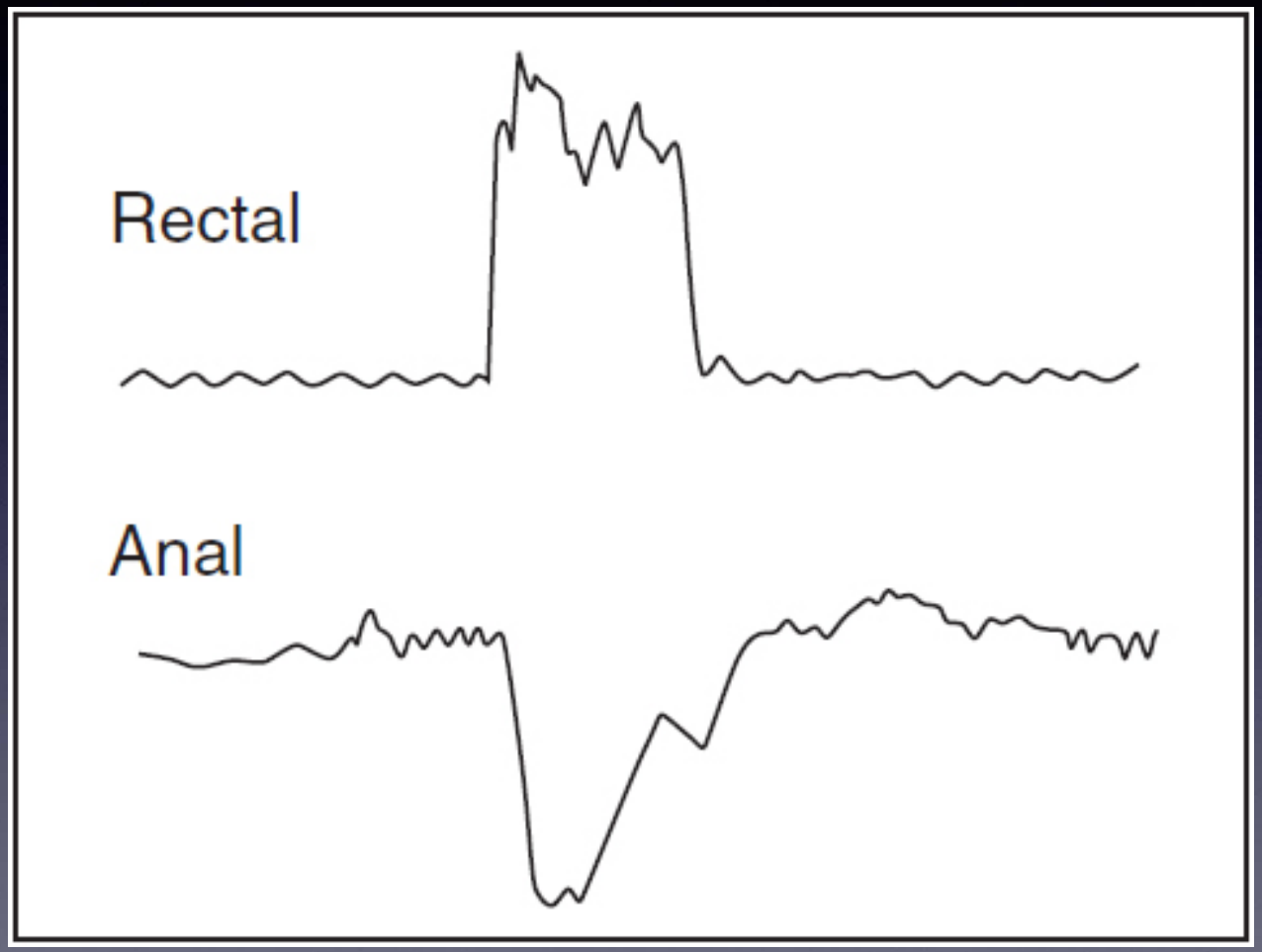
Refractory FI
Complementary investigations
Anorectal manometry

- Does not:
  - Diagnose constipation
  - Distinguish between FC + FI and NRFI

Refractory FI
Complementary investigations
Anorectal manometry

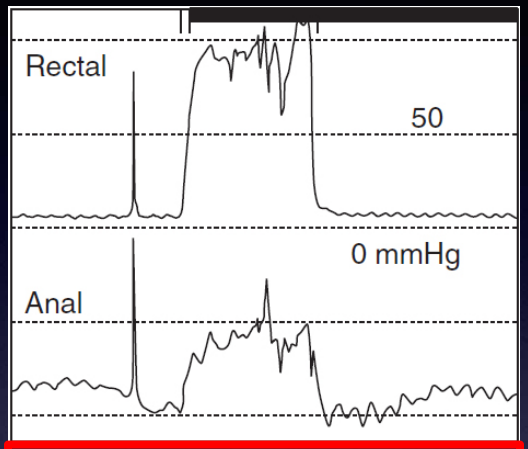
- Useful to diagnose:
  - Diagnose non-relaxing internal anal sphincter (RAIR)
  - Pelvic floor dyssynergia
  - Neurodysfunction 2^{o} spinal cord anomalies
  - Pelvic floor myopathy

Pelvic Floor Dyssynergia



Rao and Singh, J Clin Gastroenterol 2010;44:597-609.

Pelvic Floor Dyssynergia



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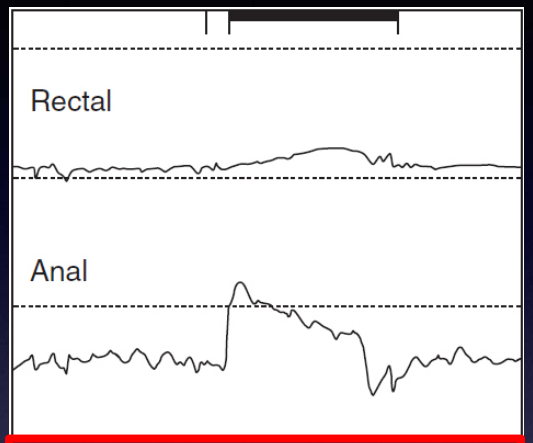

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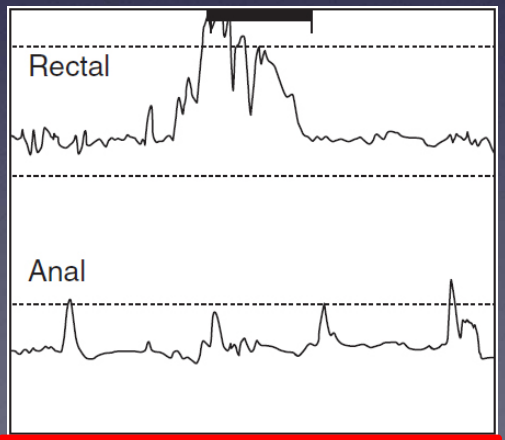
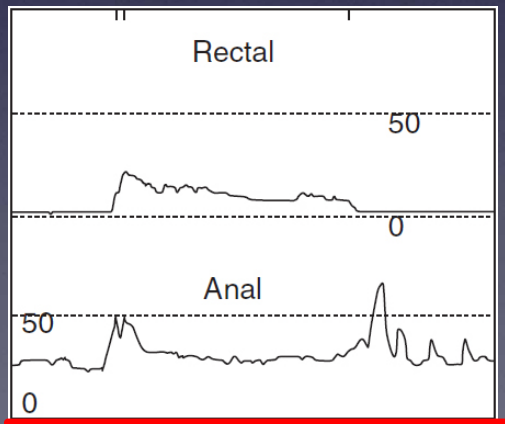


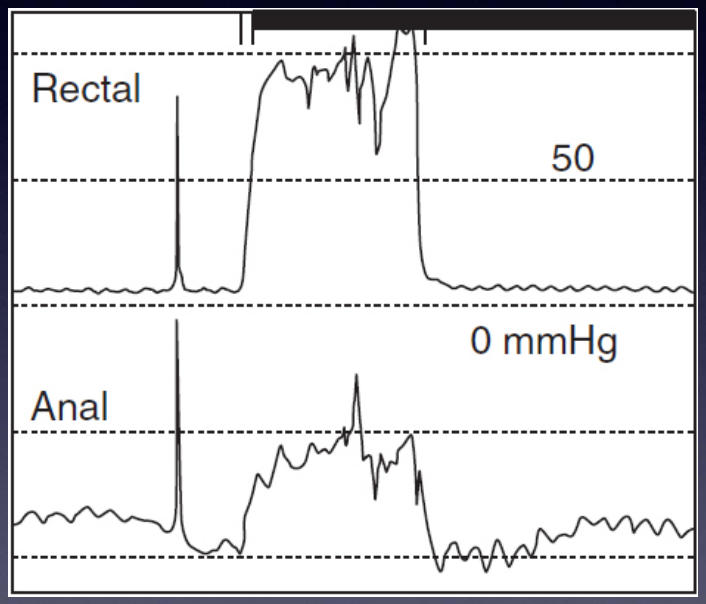
Type III



Type IV

Rao and Singh, J Clin Gastroenterol 2010;44:597-609.

Pelvic Floor Dyssynergia



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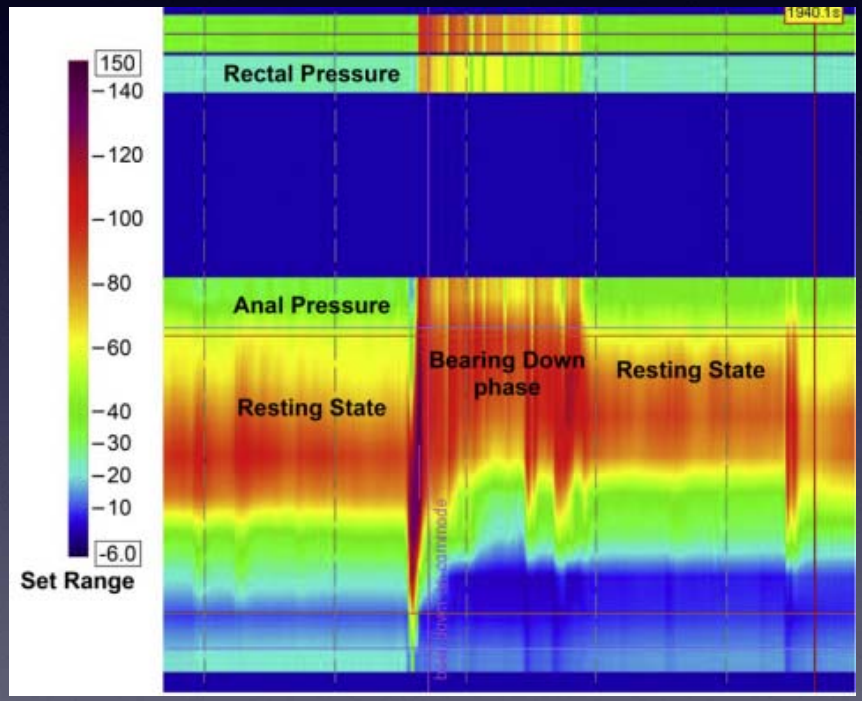


Rao and Singh, J Clin Gastroenterol 2010;44:597-609.
Rao and Meduri, Best Pract Res Clin Gastroenterol 2011;25(1):127-40.

Refractory FI
Biofeedback

- ↑ rectal sensation
- Strengthens external anal sphincter
- ↑ muscle coordination
- Improves dynamics of defecation

Refractory FI
Biofeedback

- Need highly motivated patients
- Expensive
- Lack of service providers
  - Particularly for children
- No supportive evidence in pediatric FC + FI or FNRFI
  - Contrasts with adult studies
    - Recommended if pelvic floor dyssynergia is diagnosed

Tabbers et al, J Pediatr Gastroenterol Nutr. 2014 Feb;58(2):258-74
Rao et al, Neurogastroenterol Motil. 2015 May;27(5):594-609

Refractory FI

ACE

Antegrade continence enemas

- May consider if medically refractory FI
- Allows for antegrade irrigation of the colon
- Goal is complete bowel evacuation and continence
- Several techniques described

Refractory FI

ACE

Antegrade continence enemas

- Malone antegrade continence enema (MACE)
  - Variation on Mitrofanoff
  - First described in 1990
  - Appendix used to create a non-refluxing enteral conduit
  - Multiple modifications subsequent to original paper

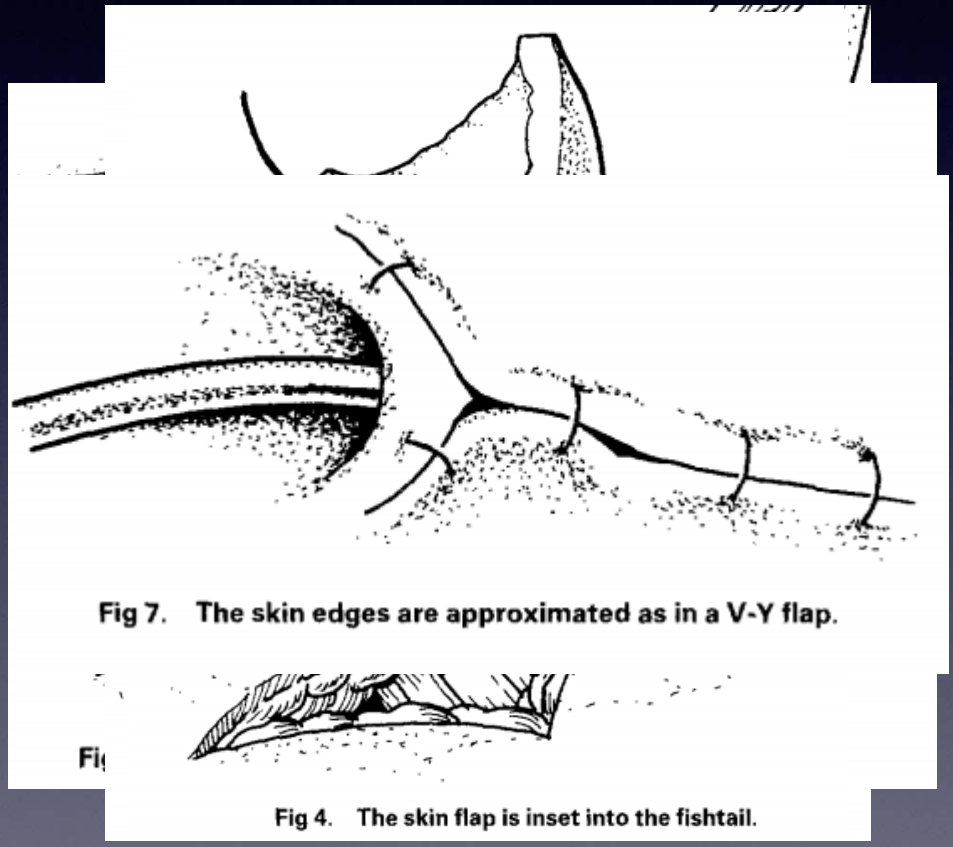


Fig 7. The skin edges are approximated as in a V-Y flap.

Fig 4. The skin flap is inset into the fishtail.

Malone et al, Lancet. 1990 Nov 17;336(8725):1217-8
Griffiths & Malone, J Pediatr Surg. 1995 Jan;30(1):68-71

Refractory FI

ACE

Antegrade continence enemas

- Chait cecostomy
  - Avoids another operation in population with extensive surgical history
  - Self-retaining pigtailed catheter
  - Inserted percutaneously under fluoroscopic guidance

Shandling et al, J Pediatr Surg. 1996 Apr;31(4):534-7
Chait et al, Radiology. 1997 Jun;203(3):621-4

Refractory FI

ACE

Antegrade continence enemas

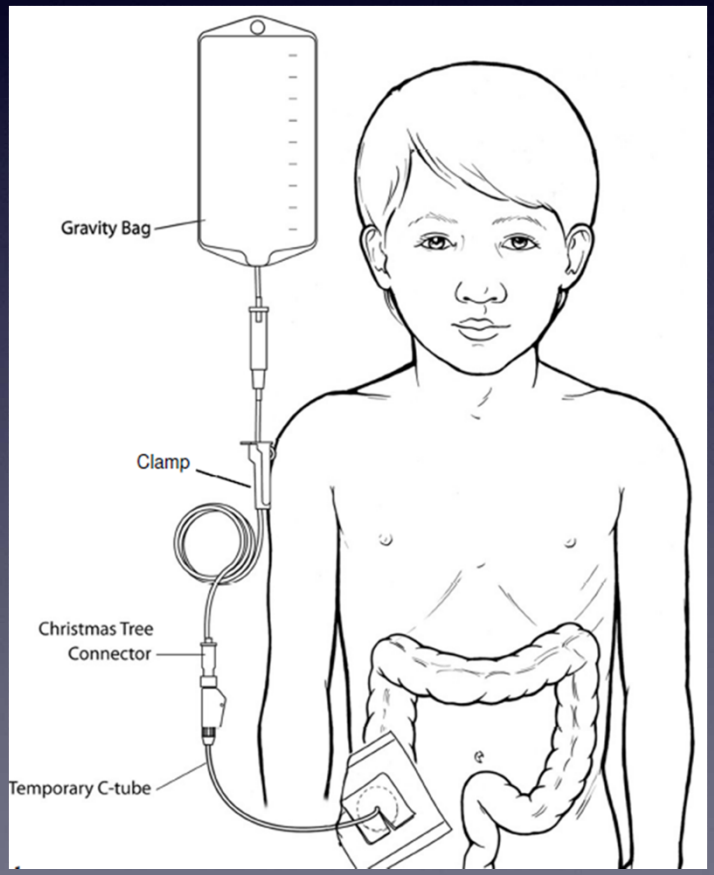
- Chait cecostomy
  - Low profile
  - Reversible
  - May not be possible in patients with interposed bowel

Shandling et al, J Pediatr Surg. 1996 Apr;31(4):534-7
Chait et al, Radiology. 1997 Jun;203(3):621-4

Refractory FI

ACE

Antegrade continence enemas



Courtesy of Nationwide Children's Hospital

Refractory FI

ACE

Antegrade continence enemas

- Complications s/p ACE:
  - Stoma stenosis/necrosis (27%)
  - Stoma leak (6.6%)
  - Difficulty catheterizing stoma (3.7%)
  - Pain w/ enema administration (3%)
  - Wound infection (2.9%)
  - Adhesive bowel obstruction (1.5%)

Graf et al, J Pediatr Surg. 1998 Aug;33(8):1294-6.

Refractory FI

ACE

Antegrade continence enemas

- Complications s/p ACE:
  - Appendiceal necrosis (0.7%)
  - Hypertrophic stomal granulation tissue (0.7%)
  - Mucus discharge and peristomal dermatitis (0.7%)
  - Cecal volvulus (0.7%)
  - Nausea/dizziness w/ enema administration (0.7%)
  - Hyperphosphatemeia (0.7%)

Graf et al, J Pediatr Surg. 1998 Aug;33(8):1294-6.

Refractory FI

ACE

Antegrade continence enemas

- Complications s/p tube cecostomy:
  - Tube dislodgement
  - Granulation tissue
  - Site infection
  - Leakage
  - Tube breakage
  - Tract stenosis

Chait et al, Radiology. 1997 Jun;203(3):621-4
Hoy et al, J Urol. 2013 Jun;189(6):2293-7

Refractory FI

ACE

Antegrade continence enemas

- Heterogeneity in the literature in terms of:
  - Outcomes
    - Complications, Measures of success , QOL
  - When to administer
  - What to administer
  - When to wean
  - How to wean

Refractory FI

ACE

Antegrade continence enemas

Author (reference no)	No. patients/ procedures	Type of procedure	Outcome/Success rate (%)	Diagnosis
Malone et al[59]	31	MACE	61	Anorectal anomaly, neuropathic bowel, chronic constipation
Curry et al[68]	300	MACE	79	Spina bifida, anorectal anomaly, Hirschsprung disease, constipation
Marshall et al[64]	32	MACE	81	Slow transit constipation
Chait et al[69]	163	Cecostomy	89	Spina bifida, imperforate anus, Klippel–Feil syndrome, cerebral palsy, Hirschsprung disease, paraplegia
Jaffray et el[57]	49	37-MACE 12-Cecostomy	81 75	Idiopathic constipation
King et al[58]	42	MACE	76	Encopresis, inadequate stool evacuation
Jaffray et al[70]	80	MACE	70	Idiopathic constipation
Yamout et al[71]	29	Cecostomy		Spina bifida, paraplegia, sacral agenesis and aanorectal malformation
Wong et al[72]	69	Cecostomy		Fecal soiling
Donkol et al[73]	21	Cecostomy	95	Neurogenic fecal incontinence, anorectal malformations
Siddiqui et al[74]	105	MACE	69	Myelodysplasia, functional constipation, anorectal malformation, nonrelaxing internal anal sphincter, cerebral palsy
Mugie et al[75]	99	Cecostomy	Symptom free-71 Improved-20	Spinal abnormality, cerebral palsy, imperforate anus, Hirschsprung disease, urological disorder, behavior problems

Arya et al, Am J Ther. 2016 Jan 21

Refractory FI

ACE

Antegrade colonic enemas

- MACE vs Chait cecostomy
  - No significant difference
    - Successful outcome
    - Rate of complications
      - MACE associated with 3X more leakage
      - CC associated with granulation tissue
    - Changing type of cecostomy

Masadeh et al, J Pediatr Surg. 2013 Oct;48(10):2128-33
Hoy et al, J Urol. 2013 Jun;189(6):2293-7

Refractory FI

ACE

Antegrade colonic enemas

- MACE vs Chait cecostomy
  - Depends on center-specific expertise and resources
    - Surgeon, IR
    - Available resources post-transition
  - Family's preference

Refractory FI

ACE

Antegrade colonic enemas

- Pre-operative assessment:
  - Barium enema
    - Interposed bowel, colonic dilatation
  - Colonic manometry
    - Colonic motility
    - HAPCs
  - +/- anorectal manometry

Refractory FI

ACE

Antegrade colonic enemas

TABLE 3. *Issues incompletely covered in perioperative counseling and teaching*

	No. Pts.
Pain*	4
Need for bowel prep./nothing by mouth	2
Time to fine-tune regimen	3
Daily time commitment for irrigations	2
Colonic spasms	1
Character of rectal effluent	1
Felt procedure was minimized	1

* Responders did not state whether pain was postoperative, related to catheter insertion or related to irrigations.

Yerkes et al, J Urol. 2003 Jan;169(1):320-3

Refractory FI

ACE

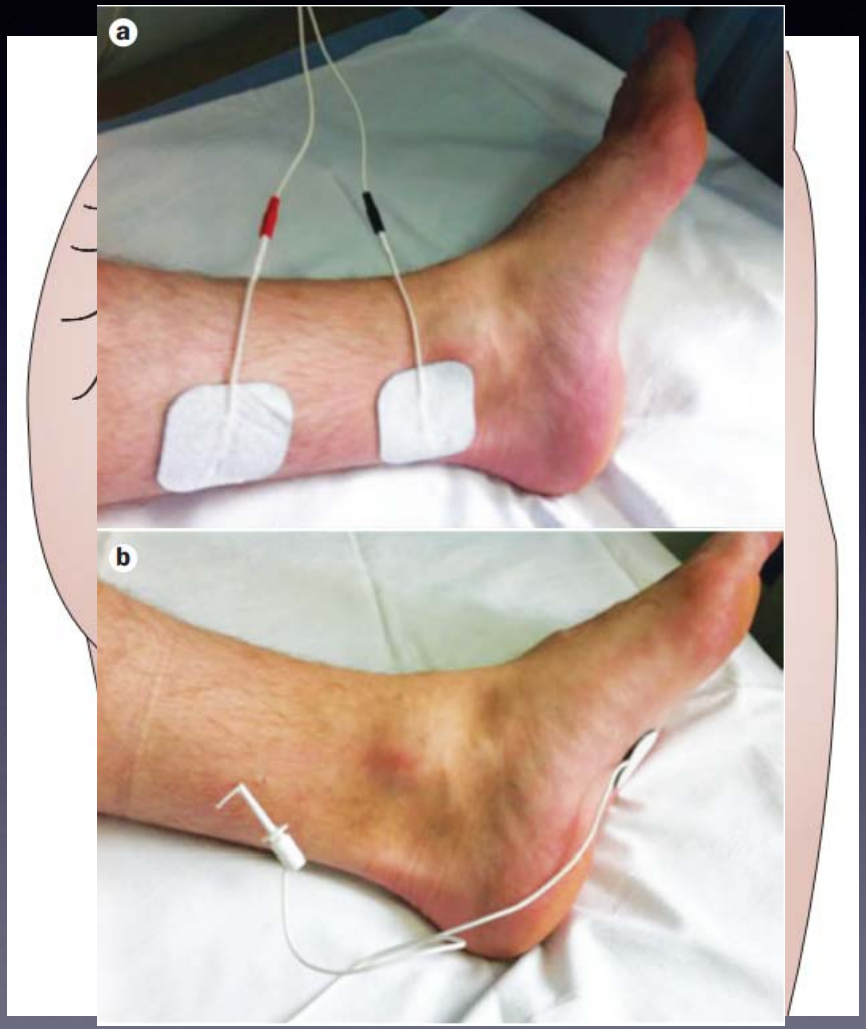
Antegrade colonic enemas

- Don’t forget to prepare your patient for transition to adult care
  - Require annual tube changes
  - Ongoing support for individual maintenance regimens
  - Cecostomy site skin care

Refractory FI

Emerging therapies

- Neuromodulation
  - Transcutaneous sacral nerve stimulation
  - Percutaneous tibial nerve stimulation
  - Sacral nerve stimulation

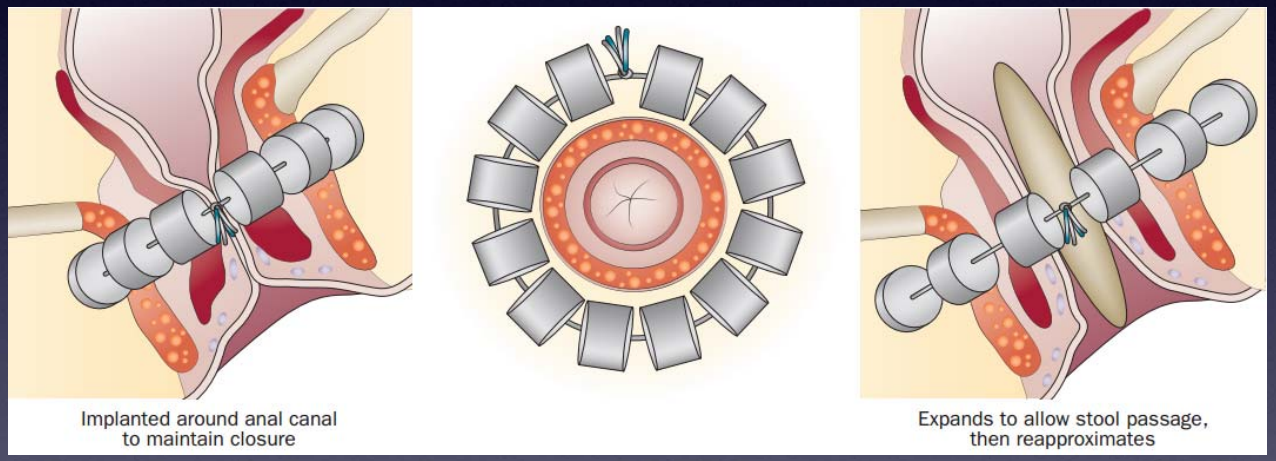


Mitchell & Sagar, Nat Rev Gastroenterol Hepatol. 2014 May;11(5):279-86

Refractory FI

Emerging therapies

- Artificial Bowel Sphincters
  - Acticon™
  - FENIX™



  - Protocol for head to head trial comparing neuromodulation to FENIX™ just published
    - Williams et al, Int J Colorectal Dis. 2016 Feb;31(2):465-72

Mitchell & Sagar, Nat Rev Gastroenterol Hepatol. 2014 May;11(5):279-86

Refractory FI

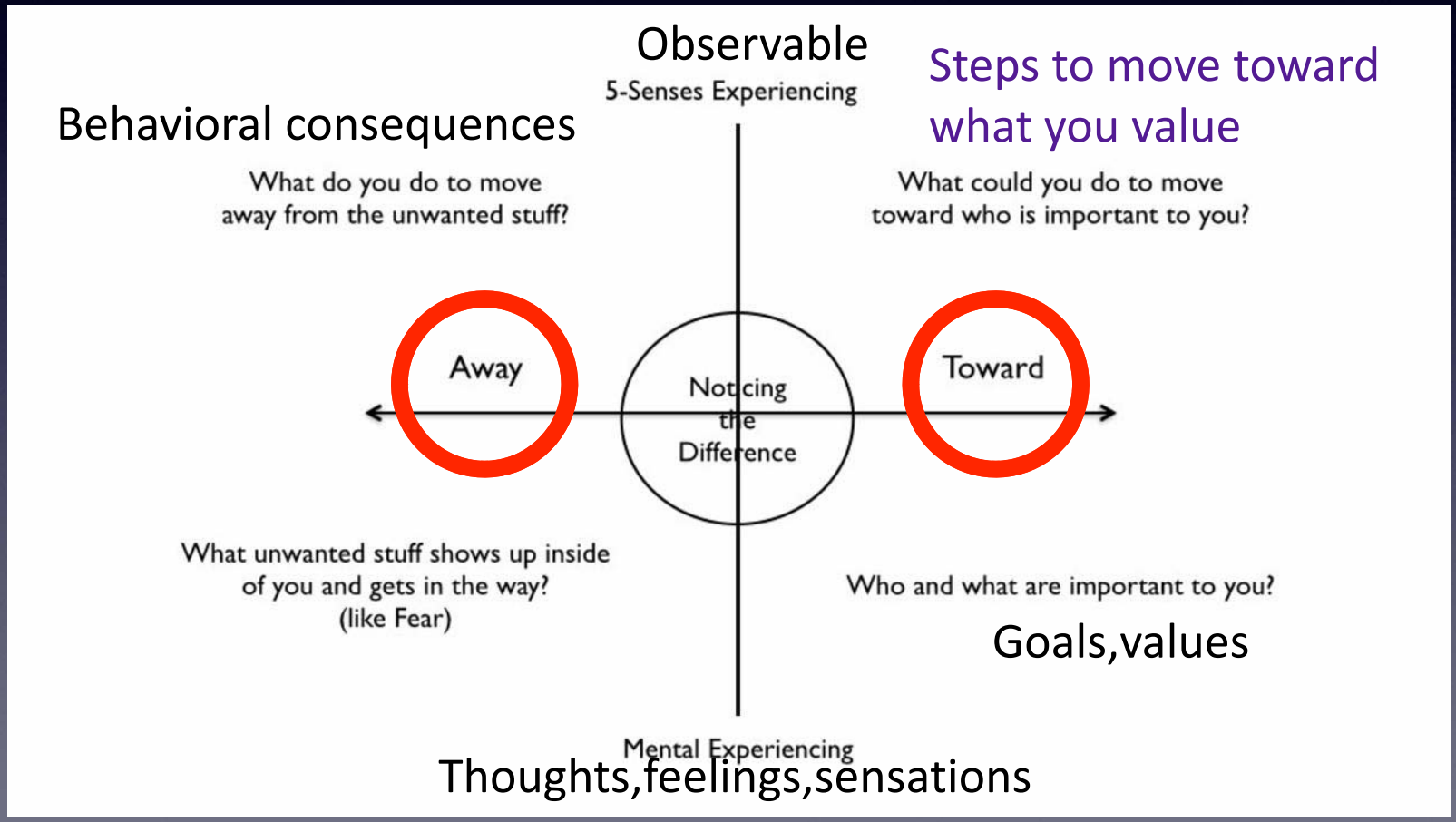
Behavioral Modification

- Individual therapy
- Group therapy
- Family therapy
- What does it include?
- Behavioural interventions could be effective when combined with intensive medical management

Refractory FI

Behavioral Modification

The ACT Matrix



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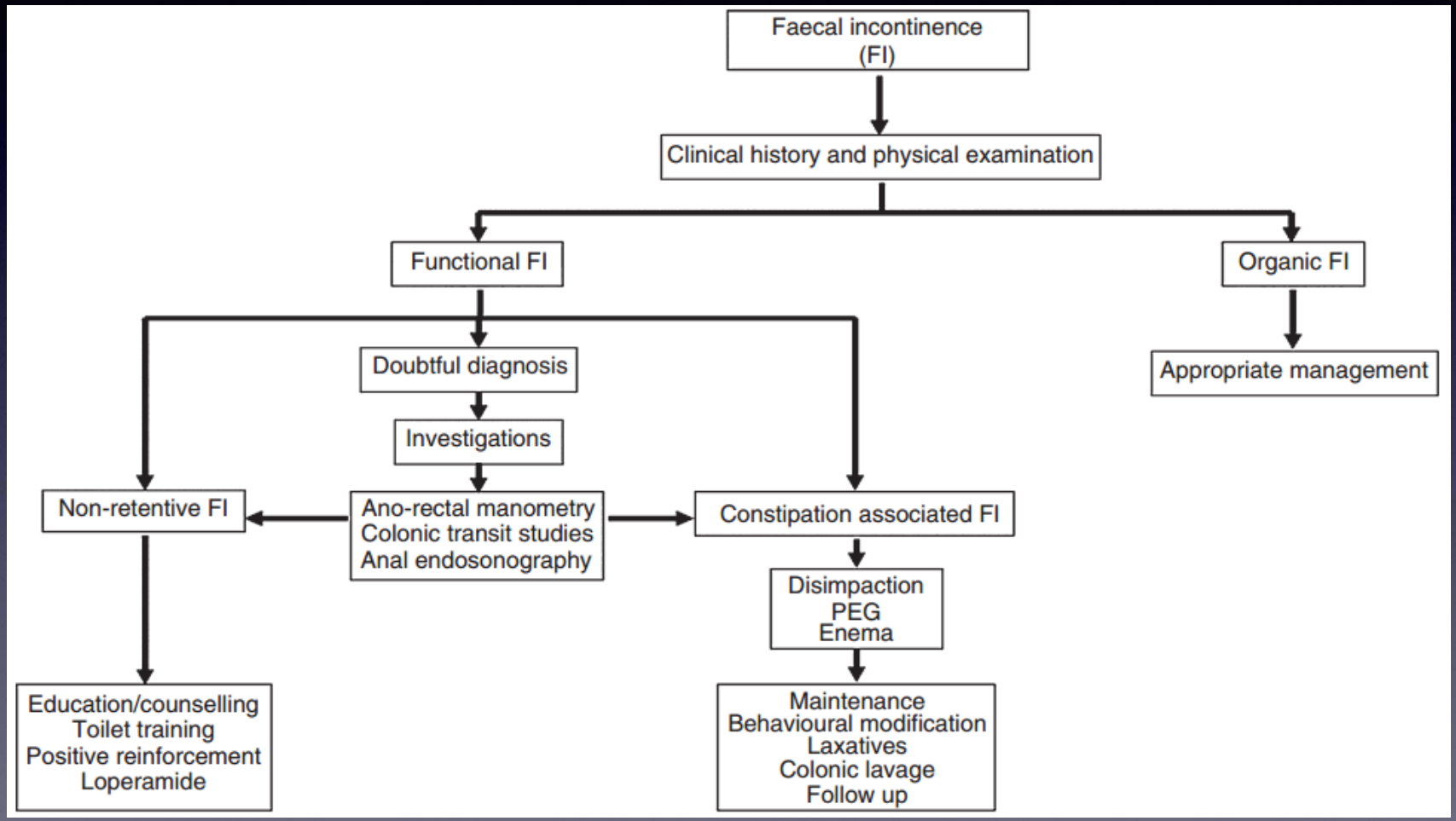
General Approach to FI



Rajindrajith et al, Aliment Pharmacol Ther. 2013 Jan;37(1):37-48

Summary

- Functional fecal incontinence is a worldwide problem
- Negatively impacts both children and their families with long-lasting effects
- Important to rule out organic causes and differentiate between FC + FI and NRFI
- Mainstays of treatment involve a multipronged approach with positive reinforcement and support for the family
- Novel emerging treatments on the horizon

Questions?