Functional Fecal Incontinence in Pediatrics

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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 $\geq 4$ 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

Food Transition
Toilet Training
Fear of Bathroom
Anal Fissure
Illness
Avoidance of School Bathroom
Other Causes of Mild Constipation

Mild Constipation

Pain
Increased Constipation
Stool Retention To Avoid Pain

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

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

Burgers & Benninga, J Pediatr Gastroenterol Nutr. 2009 Apr;48 Suppl 2:S98-S100
Risk Factors for FI

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

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

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

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

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

PEG = polyethylene glycol.
## Maintenance

<table>
<thead>
<tr>
<th>Type</th>
<th>Looks Like</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>type 1</td>
<td>rabbit droppings</td>
<td>Separated hard lumps, like nuts (hard to pass)</td>
</tr>
<tr>
<td>type 2</td>
<td>bunch of grapes</td>
<td>Sausage-shaped but lumpy</td>
</tr>
<tr>
<td>type 3</td>
<td>corn on cob</td>
<td>Like a sausage but with cracks on its surface</td>
</tr>
<tr>
<td>type 4</td>
<td>sausage</td>
<td>Like a sausage or snake, smooth and soft</td>
</tr>
<tr>
<td>type 5</td>
<td>chicken nuggets</td>
<td>Soft blobs with clear-cut edges (passed easily)</td>
</tr>
<tr>
<td>type 6</td>
<td>porridge</td>
<td>Fluffy pieces with ragged edges, a mushy stool</td>
</tr>
<tr>
<td>type 7</td>
<td>gravy</td>
<td>Watery, no solid pieces: ENTIRELY LIQUID</td>
</tr>
</tbody>
</table>
Maintenance

- Dietary fiber
- Prebiotics
- Probiotics

Maintenance

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

• Similar approach to FC + Fl except...
  – ...AVOID LAXATIVES!!
• Behavioural treatment = cornerstone of therapy
• Often benefit from referral to Psychology
• Consider loperamide
What do you do with refractory FI?
Refactory 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 Fl

Complimentary investigations

- TTG
- TSH
- Electrolytes
- Calcium
- Lead level
- Urine culture
Refactory 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
Refractory FI
Complementary investigations

*Wireless motility capsule*

- Wireless motility capsule
  - Measures pressure, pH, temperature
  - Using all parameters, can estimate:
    - Gastric emptying 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”

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
Courtesy Dr. Alfred 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

• 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
  — “Successful” = normal bowel movement frequency and no/occasional fecal incontinence
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)
Resting Pressure
Balloon Insufflation
Deflate Balloon
RAIR
Baseline

Courtesy Dr. Alfred Yeung
Refactory 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

\textbf{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° spinal cord anomalies
  - Pelvic floor myopathy
Pelvic Floor Dyssynergia

Pelvic Floor Dyssynergia

Type I

Type II

Type III

Type IV

Pelvic Floor Dyssynergia

Type I

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

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

Malone et al, Lancet. 1990 Nov 17;336(8725):1217-8
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

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

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%)

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 with enema administration (0.7%)
  - Hyperphosphatemeia (0.7%)

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
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

<table>
<thead>
<tr>
<th>Author (reference no)</th>
<th>No. patients/procedures</th>
<th>Type of procedure</th>
<th>Outcome/Success rate (%)</th>
<th>Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Malone et al&lt;sup&gt;59&lt;/sup&gt;</td>
<td>31</td>
<td>MACE</td>
<td>61</td>
<td>Anorectal anomaly, neuropathic bowel, chronic constipation</td>
</tr>
<tr>
<td>Curry et al&lt;sup&gt;68&lt;/sup&gt;</td>
<td>300</td>
<td>MACE</td>
<td>79</td>
<td>Spina bifida, anorectal anomaly, Hirschsprung disease, constipation</td>
</tr>
<tr>
<td>Marshall et al&lt;sup&gt;64&lt;/sup&gt;</td>
<td>32</td>
<td>MACE</td>
<td>81</td>
<td>Slow transit constipation</td>
</tr>
<tr>
<td>Chait et al&lt;sup&gt;69&lt;/sup&gt;</td>
<td>163</td>
<td>Cecostomy</td>
<td>89</td>
<td>Spina bifida, imperforate anus, Klippel–Feil syndrome, cerebral palsy, Hirschsprung disease, paraplegia</td>
</tr>
<tr>
<td>Jaffray et al&lt;sup&gt;57&lt;/sup&gt;</td>
<td>49</td>
<td>37-MACE</td>
<td>81</td>
<td>Idiopathic constipation</td>
</tr>
<tr>
<td>Jaffray et al&lt;sup&gt;70&lt;/sup&gt;</td>
<td>42</td>
<td>MACE</td>
<td>76</td>
<td>Encopresis, inadequate stool evacuation</td>
</tr>
<tr>
<td>Yamout et al&lt;sup&gt;71&lt;/sup&gt;</td>
<td>29</td>
<td>Cecostomy</td>
<td>70</td>
<td>Idiopathic constipation</td>
</tr>
<tr>
<td>Wong et al&lt;sup&gt;72&lt;/sup&gt;</td>
<td>69</td>
<td>Cecostomy</td>
<td>95</td>
<td>Spina bifida, paraplegia, sacral agenesis and anorectal malformation</td>
</tr>
<tr>
<td>Donkol et al&lt;sup&gt;73&lt;/sup&gt;</td>
<td>21</td>
<td>Cecostomy</td>
<td>95</td>
<td>Fecal soiling</td>
</tr>
<tr>
<td>Siddiqui et al&lt;sup&gt;74&lt;/sup&gt;</td>
<td>105</td>
<td>MACE</td>
<td>69</td>
<td>Neurogenic fecal incontinence, anorectal malformations</td>
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<tr>
<td>Mugie et al&lt;sup&gt;75&lt;/sup&gt;</td>
<td>99</td>
<td>Cecostomy</td>
<td>Symptom free-71, Improved-20</td>
<td>Myelodyplasia, functional constipation, anorectal malformation, nonrelaxing internal anal sphincter, cerebral palsy</td>
</tr>
</tbody>
</table>

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

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>
<thead>
<tr>
<th>Table 3. Issues incompletely covered in perioperative counseling and teaching</th>
<th>No. Pts.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pain*</td>
<td>4</td>
</tr>
<tr>
<td>Need for bowel prep./nothing by mouth</td>
<td>2</td>
</tr>
<tr>
<td>Time to fine-tune regimen</td>
<td>3</td>
</tr>
<tr>
<td>Daily time commitment for irrigations</td>
<td>2</td>
</tr>
<tr>
<td>Colonic spasms</td>
<td>1</td>
</tr>
<tr>
<td>Character of rectal effluent</td>
<td>1</td>
</tr>
<tr>
<td>Felt procedure was minimized</td>
<td>1</td>
</tr>
</tbody>
</table>

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

*Emerging therapies*

- Artificial Bowel Sphincters
  - Acticon™
  - FENIX™

- Protocol for head to head trial comparing neuromodulation to FENIX™ just published

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

Behavioral consequences

Away

What do you do to move away from the unwanted stuff?

5-Senses Experiencing

Noticing the Difference

Who and what are important to you?

Mental Experiencing

Thoughts, feelings, sensations

Steps to move toward what you value

Toward

What could you do to move toward who is important to you?

Goals, values

www.drkevinpolk.com
General Approach to FI

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?