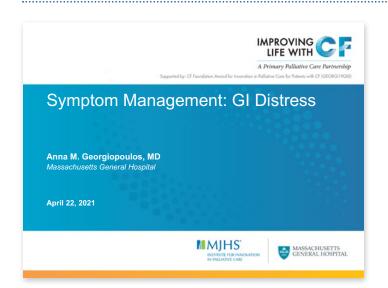
MODULE 3 -SYMPTOM MANAGEMENT: GASTROINTESTINAL DISTRESS

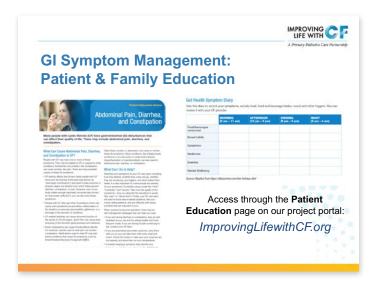
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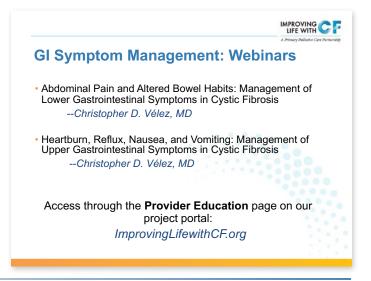
Anna Georgiopoulos, MD

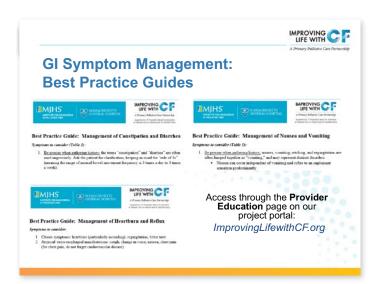
Massachusetts General Hospital Yawkey 6900, 55 Fruit Street, Boston MA 02114 ageorgiopoulos@partners.org













GI Symptom Management: Objectives

- Review selected topics as presented in two of the three GI Best Practice Guides
 - Constipation & diarrhea
 - Heartburn & reflux
 - Nausea & vomiting
- · Illustrate their practical application with case descriptions from two of three GI Best Practice Guides
 - Lower GI pediatric case: Constipation & diarrhea
 - Upper GI adult case: GERD
 - CF programs can use one or both cases as appropriate to their needs



GI Symptom Management: Key Content in the Best Practice Guides

- The three GI Best Practice Guides cover key content areas for primary palliative care of people with CF
 - Constipation & diarrhea
 - Heartburn & reflux
 - Nausea & vomiting
- Each guide covers three topics
 - Symptoms to consider
 - Diagnostic testing
 - Management



- **Key Content in All Best Practice Guides** · Physical symptom management is one component of
 - Goal to reduce distress, improve quality of life
- Management section of GI Best Practice Guides includes range of primary palliative care interventions to consider in co-developing a comprehensive treatment plan
 - Lifestyle modification

palliative care

- Psychosocial interventions
- Integrative interventions
- Medical management
- Endoscopic/surgical interventions
- When to refer to a specialist



GI Symptom Management: Assessment

- · Principles introduced in the Pain Best Practice Guide apply to GI symptom management as well
- · Competent treatment planning requires a competent assessment, guided by its objectives: The 3 "C's"
 - Character: Characterize the symptom to guide work-up and selection of therapies
 - Cause: Identify the etiology, if possible, to clarify the potential for disease-modifying therapy
 - Context: Understand the impact and context to integrate symptom management into the broader palliative plan of care



Multidimensional Symptom Description

- "P-Q-R-S-T"
 - "P" = Provocative and palliative factors
 - "Q" = Quality of the symptom
 - "R" = Region → Localized (where?), multifocal, or generalized
 - "S" = Severity → Verbal rating scale or a numeric scale "on average during the past week" and "at its worst during the past
 - "T" = Temporal features → Onset, course, and fluctuation



GI Symptom Management: Objectives

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- · Rule of 3s: Normal bowel movement frequency is 3 times/day to 3 times/week
- · Be precise and ask for clarification when gathering history
 - "When you say you are constipated..."
 - Hard stool? Excessive straining?
 - Sense of incomplete evacuation?
 - Need to use fingers to remove/ promote stool passage?
 - Infrequent bowel movements?
 - Bloating with normal bowel movement frequency?
 - · Constipation with abdominal pain?
 - "When you say you have diarrhea ... "
 - Liquid stool? Frequent movements?
 - · Incontinence/accidents? Diarrhea in the setting of constipation?
 - · Alternating diarrhea with constipation? Diarrhea with abdominal pain?



Constipation & Diarrhea: Symptoms to Consider

- Two main distinctions in testing and management
 - Acuity
 - Acute
 - Chronic
 - Alarm signs: Signs and symptoms requiring immediate attention
 - Present
 - Absent



Constipation & Diarrhea: Symptoms to Consider

- Acute constipation/diarrhea
 - Emergent CF-related etiologies such as distal intestinal obstruction syndrome, antibiotic-associated diarrhea
 - Non-CF causes like C. difficile-associated diarrhea, irritable bowel syndrome
- · Chronic constipation/diarrhea
 - Benign CF-related etiologies such as pancreatic exocrine
 - Non-CF causes such as inflammatory bowel disease
 - "CF-triad" of dysmotility, dysbiosis, and inflammation



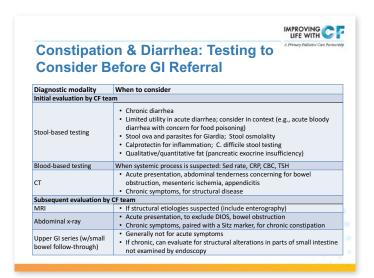
Constipation & Diarrhea: Symptoms to Consider

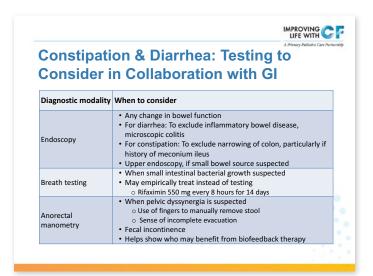
- · Alarm signs require prompt examination for organic, structural, or malabsorptive causes (insufficiency, malignancy)
 - Bleeding
 - Abdominal pain
 - Unexpected weight loss in adults
 - Failure to appropriately gain weight in children
- · Anxiety can increase severity or frequency
 - Functional bowel disorders like irritable bowel syndrome
 - History of sexual abuse/trauma

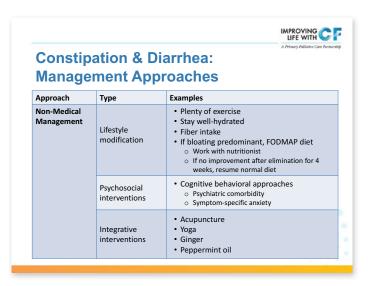
Focused Differential Diagnosis of

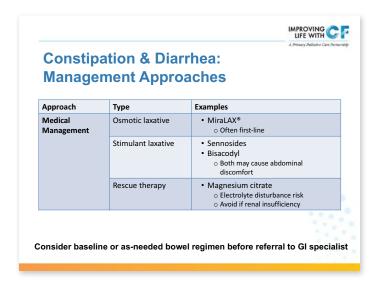


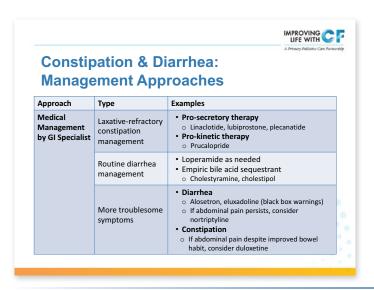
- **Constipation & Diarrhea in CF** · Slow-transit constipation CF-related causes of
- constipation
- Distal intestinal obstruction syndrome (DIOS)
- Small intestinal bacterial overgrowth
- CF-related causes of diarrhea
- · Pancreatic exocrine insufficiency • Small intestinal bacterial overgrowth
- Non-CF causes of altered bowel habits
- · Infectious enteritis
- · C. difficile-associated diarrhea
- · Antibiotic-associated diarrhea
- Irritable bowel syndrome · Pelvic dyssynergia
- Endocrinopathy (e.g., thyroid disturbance)
- · Medication-related

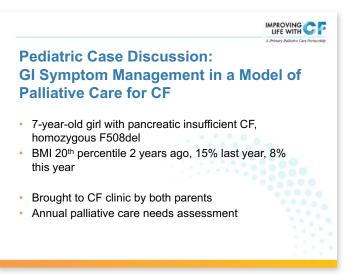














Integrated Palliative Care Outcome Scale (IPOS): GI Items for Ages ≥12 Years

How have the following symptoms affected you over the past week?	Not at all	Slightly	Moderately	Severely	Over- whelmingly
Nausea (feeling like you are going to be sick)	0	1	2	3	4
Vomiting (being sick)	0	1	2	3	4
Constipation	0	1	2	3	4
Poor appetite	0	1	2	3	4

Additional GI symptoms may be elicited by these IPOS items:

- What have been your main problems or concerns over the past week?
- Please list any other symptoms not mentioned above and tick one box to show how they have affected you over the past week

Murtagh FE, et al. Pall Med. 2019 Sep;33(8):1045-57



ADAPT-CF Communication Guide for Children <12 Years: GI Items

- Assessing for Distressing Symptoms and Palliative Care Needs through Targeted Communication (ADAPT-CF)
- · Use the ADAPT-CF, a pediatric communication guide for the
 - Ages 0-3: Direct symptom-specific questions to parents
 - · Ages 4-6: Direct questions to child
 - · Ages 7-11: Direct guestions to child
- · For children <12, CFF guidelines recommend annual assessment guided by the IPOS
 - · Items are not developmentally appropriate for young children



ADAPT-CF: Child Responses

7-year-old girl with pancreatic insufficient CF, declining BMI, at CF clinic with parents for annual needs assessment

How has your tummy been feeling?

- It feels sick when I eat.
- I have to go to the bathroom a lot. I hate when I have to go at school.

Are there times you don't want to eat when other people around you are eating, like at lunch or dinner?

· Dinner is gross. I don't like what they make.

Do you have any questions about your medicines or your treatment?

Why do I have to take them all the time?

How will you respond to the child's comments and question?



ADAPT-CF Family Conversation: Parent Concerns

What have been your family's main problems or concerns lately?

- Dad: "She doesn't eat well. She looks skinny."
- Mom: "She won't eat anything I cook. She just eats cereal. I don't know what to do."

What might be going on medically? What might be going on in the family system? At school? What else would you like to know?



Brief Assessment Scale for Caregivers (BASC) Responses from Parents:

Rate your distress during this past month:		A little distressed	Some distress	A lot of distress	Does not apply
Distress at seeing your relative or friend in so much pain or discomfort	1	2	3	4	0
Distress at not having enough time to do your job, other responsibilities and chores	1	2	3	4	0
Distress at having strained relationships with other family members over taking care of	1	2	3	4	0
your relative or friend					. 0 0

How might you interpret high vs. low scores? Do you think each parent would rate these items similarly?



Pediatric Case Discussion: GI Symptom Management

- 7-year-old girl with pancreatic insufficient CF, homozygous F508del
- Hard stools every 3-4 days
- Incontinence of liquid stool 1-2x/week
- Nausea & bloating after meals
- Low appetite, poor weight gain
 - Falling off former growth trajectory
- Parents feel probiotics are helping

What's the most likely explanation?

What are you most worried about?

What are your next steps in diagnostic assessment?

How urgent is further evaluation?



Pediatric Case Discussion: Diagnostic Assessment

- · Abdominal examination is not remarkable
- Abdominal X-ray full of stool
- LFTs are normal
- Vitamin D3 is low at 15

What does the diagnostic assessment indicate about the nature of the symptoms and next steps?



Pediatric Case Discussion: Constipation & Diarrhea

- Based on the characterization of symptoms and diagnostic assessment
- Symptoms are chronic and likely represent CF-related constipation with secondary overflow diarrhea
 - Intervention is indicated
 - Symptoms are distressing to both child and parents
 - Impact on appetite, weight gain, growth, nutrition
 - Concern for adequacy of pancreatic enzyme therapy
 - Impact on individual functioning at home and school
 - Impact on family functioning, especially at mealtimes



Pediatric Case Discussion: Chronic Constipation Management

The GI Best Practice Guides include a range of primary palliative care interventions to consider in co-developing a comprehensive treatment plan

What interventions might you suggest?

- Lifestyle modifications
- Psychosocial interventions

 - If family is coping well?

 If family is experiencing frequent criticism, conflict, overcontrol, oppositional behavior?
- Integrative interventions
- Medical management or endoscopic/surgical interventions
- · When to refer to a specialist

Consider simultaneous vs. sequential interventions. Some may not be appropriate or acceptable to the patient/family. Which CF team members might be helpful to engage?

Pediatric Case Chronic Cons	e: Sample Interventions for tipation
Intervention Type	Examples and Notes
Lifestyle modification	Increase hydration Work with CF team nutritionist around meal planning
Psychosocial intervention	Depending on family needs: • Supportive intervention, reinforcing strengths <u>during assessment</u> • Behavioral intervention around mealtime challenges • Parent child interaction therapy (PCIT) or family therapy
Integrative intervention	Ok to continue using probiotics
Medical management	Start polyethylene glycol

(e.g., acute pain, bleeding)

· Optimize enzyme dose, storage, adherence

Age-appropriate education for child (what enzymes do, when to take them)

Consult GI if signs & symptoms become refractory or urgent



GI Symptom Management: Objectives

- Review selected topics as presented in 2 of the 3 the GI Best Practice Guides
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 - Heartburn & reflux
 - Nausea & vomiting
- · Illustrate their practical application with case descriptions from 2 of the 3 GI Best Practice Guides
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- Classic symptoms of Gastroesophageal Reflux Disease (GERD)
 - Heartburn (particularly ascending)
 - Regurgitation
 - Bitter taste
- Atypical/extra-esophageal manifestations of GERD
 - Cough

Refer to a specialist

- Change in voice
- Nausea
- Chest pain



Heartburn & Reflux: Symptoms to Consider

Frequently missed mimickers of GERD

- Rumination syndrome
 - Effortless regurgitation of food that was just chewed and swallowed
 - Responds to diaphragmatic breathing
- Achalasia
 - Pathologic failure of lower esophageal relaxation associated with esophageal body peristaltic abnormalities
 - Can have regurgitation symptoms
 - · With difficulty swallowing, can be associated with aspiration



Heartburn & Reflux: **Symptoms to Consider**

Frequently missed mimickers of GERD (Cont'd)

- Gastroparesis
 - Delayed stomach emptying, most often idiopathic
 - May complain of regurgitation of stomach contents while supine
- Supragastric belching
 - Air is predominant complaint
 - Consider diaphragmatic breathing exercises



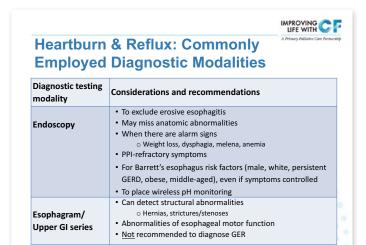
Heartburn & Reflux: Symptoms to Consider

- · Before attributing typical or atypical GER symptoms to GERD, consider disease of other organ systems and alarm signs
- · Do not forget cardiovascular disease, including myocardial infarction!

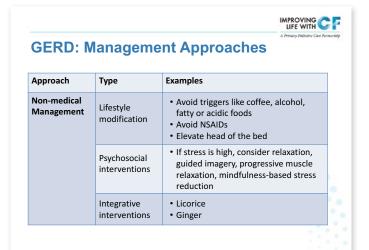


Heartburn & Reflux: Symptoms to Consider

- Alarm signs should generally prompt endoscopy
 - Painful swallowing (odynophagia)
 - Difficulty swallowing (dysphagia)
 - Proton pump inhibitor (PPI) refractory symptoms
 - Weight loss
 - Melena, particularly with history of condition predisposing to malignancy
 - · Smoking or Barrett's esophagus



	& Reflux: Less-Commonly Diagnostic Modalities
Diagnostic testing modality	Considerations and recommendations
High resolution	Identify esophageal motor abnormalities of pathologic GER
manometry	Guide catheter-based ambulatory reflux and impedance
	monitoring
	Indicated prior to lung transplantation or anti-reflux
	procedures
Catheter-based	Diagnoses reflux hypersensitivity or functional heartburn,
ambulatory reflux/	which may respond to neuromodulation
impedance monitoring	 Tricyclic antidepressants, SSRIs, SNRIs, trazodone
Wireless-based	Only provides information on acid exposure time, association
ambulatory reflux	of acidic reflux w/ symptoms
monitoring	Requires endoscopic placement
	Capsule can become dislodged, especially during placement,
	and require bronchoscopy to remove

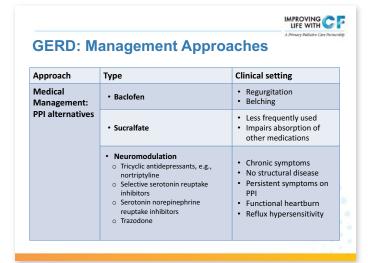


Approach	Clinical setting	Approaches Examples and comments
Medical Management: First-line pharmacotherapy	PPI naïve without alarm signs, infrequent symptoms	Can use histamine-2 antagonists before pursuing further testing
	PPI naïve without alarm signs, frequent/ bothersome symptoms	Before refer to GI can start PPI Start once daily dosing 30 minutes before breakfast; increase to twice daily 30 minutes before breakfast and before dinner Re-evaluate in 8 weeks; consider additional PP doses, medications from other classes, and if diagnostic testing is indicated
	Stopping treatment	Likely best to taper off PPI If taking daily, reduce to every-other-day dosing for 2-4 weeks and as-needed histamine 2 antagonism then stop If taking twice per day, reduce to daily dosing for 2-4 weeks before every-other-day dosing



GERD: Factors to Consider When Using PPIs in CF

- PPIs may reduce acid-related pulmonary damage or improve response to enzyme replacement in people with CF
- · Risk of enteric infections, increased pulmonary exacerbations
 - Stomach acid is an important barrier to infectious disease
- · Limited data of other adverse events such as bone density/fracture risk
- · Inappropriate PPI usage is common!
 - May take it as needed, miss doses, or time inappropriately in relation to meals
 - Ask before considering someone treatment-refractory





GERD: Management Approaches

Approach	Туре	Examples and comments
Endoscopic & Surgical	Traditional fundoplication	Surgical
Interventions	Transoral incisionless fundoplication (TIF)	• Endoscopic
	Sphincter augmentation with devices	Linx: Surgically placed Stretta: Endoscopically- performed induced muscular hypertrophy (controversial)



- 43-year-old man with pancreatic sufficient CF
- FEV1%pred 80
- CF first diagnosed 5 years ago after years of recurrent pneumonia and sinusitis
- · Annual palliative care needs assessment



The patient completes the Integrated Palliative Care Outcome Scale (IPOS) as part of the annual needs assessment

What have been your main problems or concerns over the past week? Stomach pain

How have the following symptoms affected you over the past week?	Not at all	Slightly	Moderately	Severely	Over- whelmingly
Nausea (feeling like you are going to be sick)	0	1	(2)	3	4
Poor appetite	0		2	3	4
Pain	0	1	(2)	3	4

What else would you like to know? What are your next steps in assessment?



Adult Case Discussion: GI Symptom Management

- 43-year-old man with pancreatic sufficient CF
 - "Feels like acidy stuff comes up after I eat, especially at night. I've never had that before. Is it heartburn?'
 - "Doesn't happen every day but when it does, it's no good. This week my wife made chili and I barely slept."
- Work has been more stressful lately

What's the most likely explanation?

What are you most worried about?

What are your next steps in diagnostic assessment?

How urgent is further evaluation?



IMPROVING C

Adult Case Discussion: Diagnostic Assessment

- Based on the characterization of symptoms
 - Moderate intermittent symptoms of heartburn and reflux
 - Symptoms are distressing, but no alarm signs
 - No prior treatment
- Decision made to defer endoscopy and treat empirically as Gastroesophageal Reflux Disease (GERD)
- Check liver tests and GGT to screen for CF-Related Liver Disease as part of routine CF care
- Review recommendations for earlier start to colonoscopy screening in people with CF



Adult Case Discussion: GERD Management

The GI Best Practice Guides include a range of primary palliative care interventions to consider in co-developing a comprehensive treatment

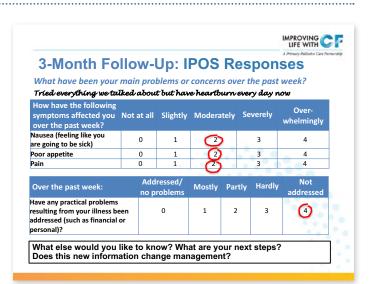
What interventions might you suggest?

- · Lifestyle modifications
- Psychosocial interventions
- · Integrative interventions
- Medical management or endoscopic/surgical interventions
- · When to refer to a specialist

Consider simultaneous vs. sequential interventions. Some may not be appropriate or acceptable to the patient. Which CF team members might be helpful to engage?

Adult Case: Initial Interventions for GERD

Examples
Avoid eating close to bedtime Reduce triggers (spicy food) Elevate head of the bed
Problem solve recent job-related stressors that seem to exacerbate symptoms
Prefers not to use medications Wants to try caffeine-free ginger tea first
Agrees will try OTC histamine-2 blocker (famotidine) if ginger ineffective
Consult GI if signs & symptoms become refractory or urgent





Adult Case Discussion: GERD Management Next Steps

- · 43-year-old man with pancreatic sufficient CF, persistent heartburn; pain and nausea impacting appetite
- Called between clinic visits: Famotidine not helping
- Started on PPI, instructed to increase to twice daily, no benefit
- Recently started smoking again
 - Attributes to losing job, with resulting financial stress
- Assessment
 - Moderate GERD with no response to lifestyle modification, H2 blocker, and PPI in context of increased psychosocial stressors

What additional diagnostic evaluations or interventions might you suggest?

Adult Case: Persistent G	Next Step Interventions for ERD
Intervention Type	Examples
Lifestyle modification	Smoking cessation for GERD and pulmonary health
Psychosocial intervention	Help access unemployment benefits Teach guided imagery for relaxation
Integrative intervention	Try chamomile tea after dinner to help sleep and settle stomach
Medical management	Ensure taking PPI correctly (30 minutes before meals) and consistently Schedule endoscopy
Refer to a specialist	Consult GI specialist if remains treatment refractory, endoscopy abnormal, or signs & symptoms become urgent



Alternate Case Scenarios: GI Symptom Management—GERD

- What signs, symptoms, or diagnostic findings would you be especially alert for in this patient?
 - What are you most concerned to rule out before continuing symptomatic treatment?
- Would this case be managed differently if the patient had recently initiated highly effective modulator therapy? How?
- How might a case of worsening GERD present differently in an infant, toddler, or school-aged child?
 - Would evaluation and management differ? How?



Adult Case Discussion: GERD

43-year-old man with late-diagnosed, pancreatic sufficient CF, moderate GERD, recent job loss

What about the caregiver?

- Identifies wife as primary support
- Offer Brief Assessment Scale for Caregivers (BASC)

