

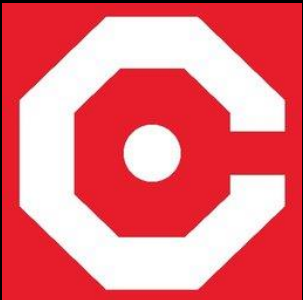
AMSER Case of the Month

July 2025

GF is an 84 y.o. male with history of enterocolitis, perforated sigmoid diverticulitis (s/p sigmoid resection with colostomy and colostomy reversal in 2016), severe malnutrition, and chronic diarrhea presenting with dysphagia, weight loss, and failure to thrive.

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Patient Presentation

- Presented to the Cooper ED on 4/4 with 50 lb weight loss since his hernia repair 18 months ago
- Recently admitted to OSH and workup included CT scans of the abdomen and pelvis which revealed enterocolitis
- He was discharged on Creon due to his history of pancreatic insufficiency, he also was found to have aspiration pneumonia and concern for connective tissue disorder
- He states that over the past month he has had new onset dysphagia to solids, especially “crusty” breads, and feels like he has a hard time swallowing pills and often spits them up
- He also rarely has dysphagia to liquids and can choke if he drinks too much too rapidly.
- He reports chronic diarrhea that waxes and wanes with occasional black stools attributed to pancreatic insufficiency, currently on Creon
- Otherwise, patient denies any fevers, chills, new respiratory complaints, cough or cold symptoms, focal weakness, or chest pain.
- He denies any current alcohol, tobacco or other recreational substance use, smoked tobacco from ages approximately 14-40, stopped drinking alcohol about 2 years ago per patient.


Pertinent Labs

Component	4/4/25 1327
Ref Range & Units (hover)	
 WBC	4.81
 RBC	3.94 ▼
 HEMOGLOBIN	12.8 ▼
 HEMATOCRIT	37.3 ▼
 MCV	94.7
 MCH	32.5 ▲
 MCHC	34.3
 RDW	15.8 ▲
 PLATELET COUNT	195
 MPV	11.5 ▲
 NRBC	0.0
 DIFFERENTIAL TYPE	AUTOMATED DIFFERENTIAL

Component	4/4/25 1327
Ref Range & Units (hover)	
 GLUCOSE	95
 BUN	25 ▲
 CREATININE	0.82
 SODIUM	145
 POTASSIUM	3.0 ▼
 CHLORIDE	112 ▲
 CO2	22
 CALCIUM	9.0
 ANION GAP	11
 eGFRu	87

Component	4/4/25 1457
Ref Range & Units (hover)	
 PHOSPHORUS	3.3

Component	4/4/25 1457
Ref Range & Units (hover)	
 LIPASE	12 ▼

Component	4/4/25 1457
Ref Range & Units (hover)	
 MAGNESIUM	2.1

What Imaging Should We Order?

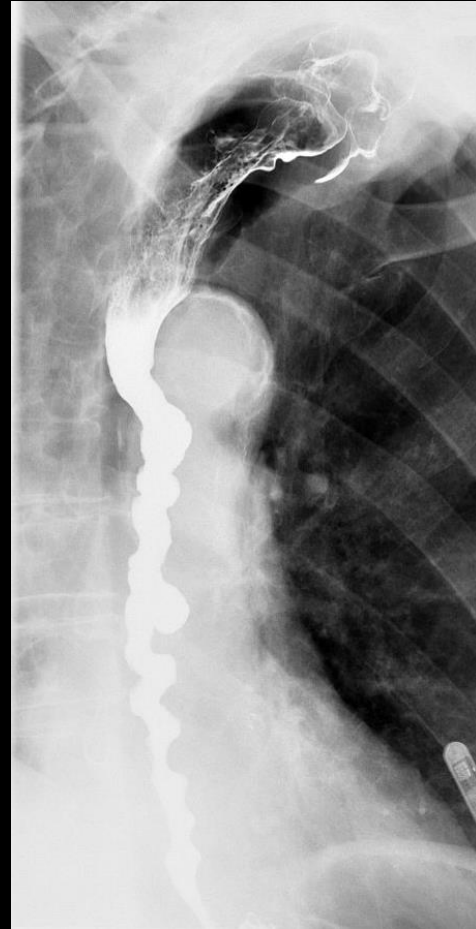
Select the applicable ACR Appropriateness Criteria

Scenario	Scenario ID	Procedure	Adult RRL	Peds RRL	Appropriateness Category
Dysphagia, retrosternal, not immunocompromised	3074198	● Fluoroscopy biphasic esophagram	1-10 mSv ⊕⊕⊕		Usually appropriate
		● Fluoroscopy barium swallow modified	1-10 mSv ⊕⊕⊕		May be appropriate
		● Fluoroscopy single contrast esophagram	1-10 mSv ⊕⊕⊕		May be appropriate
		● Esophageal transit nuclear medicine scan	1-10 mSv ⊕⊕⊕		May be appropriate
		● Fluoroscopy pharynx dynamic and static imaging	1-10 mSv ⊕⊕⊕		Usually not appropriate
		● CT neck and chest with IV contrast	10-30 mSv ⊕⊕⊕⊕	3-10 mSv [ped] ⊕⊕⊕⊕	Usually not appropriate
		● CT neck and chest without and with IV contrast	10-30 mSv ⊕⊕⊕⊕	3-10 mSv [ped] ⊕⊕⊕⊕	Usually not appropriate
		● CT neck and chest without IV contrast	10-30 mSv ⊕⊕⊕⊕	3-10 mSv [ped] ⊕⊕⊕⊕	Usually not appropriate



This imaging modality was ordered by the ER physician

Findings (unlabeled)

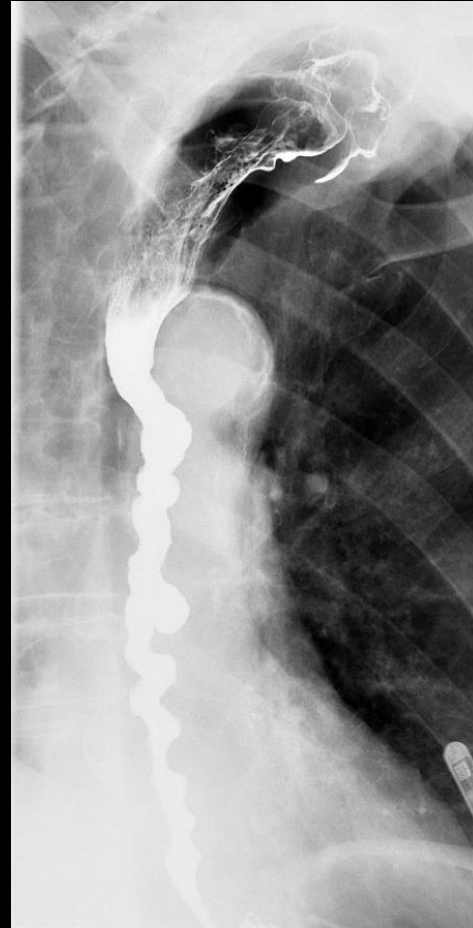
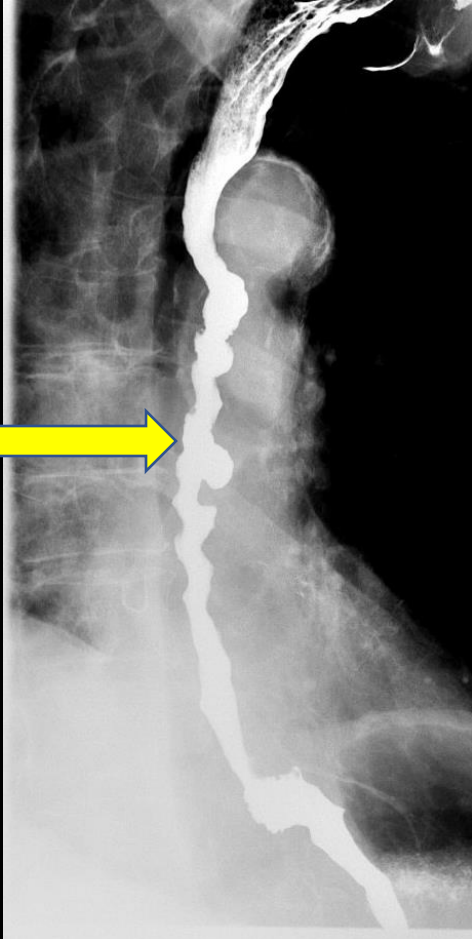


Findings (unlabeled)



Findings: (labeled)

Corkscrew
Esophagus¹

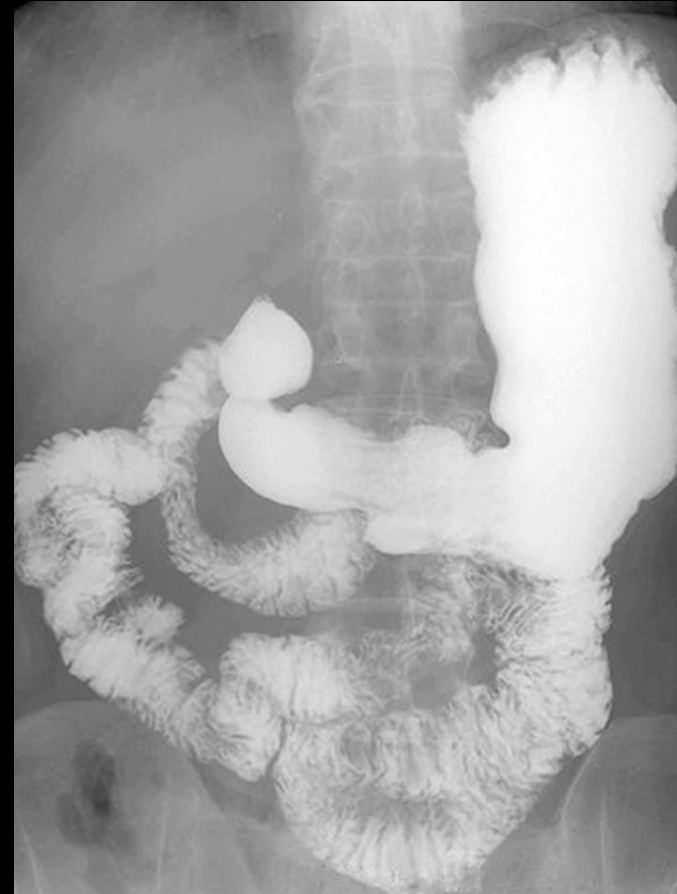


Findings: (labeled)

Organoaxial
rotation of the
stomach²



Normal
anatomy



Final Dx:

Distal Esophageal Spasm (Diffuse Esophageal Spasm)
Organoaxial rotation of the stomach without obstruction

Case Discussion:

Distal Esophageal Spasm

- **Presentation:**³ Dysphagia, regurgitation, and squeezing retrosternal chest pain during meals.
- **Pathophysiology:**³ Disorganized esophageal motility caused by Impaired inhibitory innervation that is characterized by repetitive, uncoordinated, nonprogressive contraction waves of the esophagus.
- **Approach**^{4,5}: Rule out life-threatening causes of chest pain if present (e.g. cardiac workup) followed by **upper endoscopy, esophageal barium swallow, and eventually manometry if diagnosis remains unclear**
 - **EGD:** Unremarkable/nonspecific findings
 - **Esophageal barium swallow:** Characteristic corkscrew or rosary beads appearance, which reflects abnormal contractions that leads to compartmentalization and curling of the esophagus
 - **Esophageal Manometry**
 1. Aperistalsis in greater than 30% of the wet swallows
 2. 20% simultaneous contractions
 3. The amplitude of the contractions in the distal three-fifths of the esophagus is greater than 30%.

Case Discussion: Distal Esophageal Spasm

- **Management⁵**

- **Lifestyle modifications:** Taking smaller bites, chewing thoroughly, eating slowly
- **Pharmacologic intervention:**
 - Nitrates (isosorbide dinitrate)
 - PDE5 inhibitors (sildenafil)
 - Calcium channel blockers (nifedipine, diltiazem)

- **Future Directions**

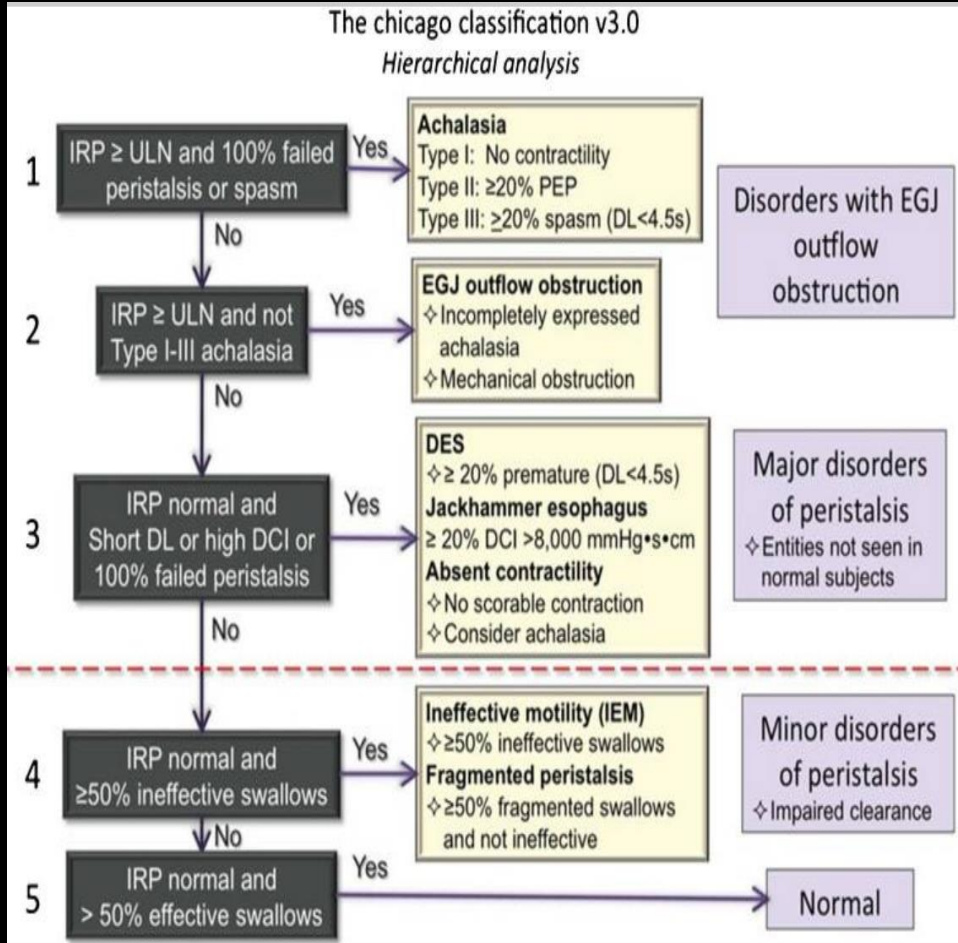
- **High-resolution manometry and High-resolution esophageal pressure topography^{6,7}**
 - Utilizes high-resolution catheter to transmit intraluminal pressure data that is subsequently converted into dynamic esophageal pressure topography plots
 - **Protocol:** Baseline phase and a series of ten wet swallows in the supine position.
 - **Chicago Classification v3.0**
 - $\geq 20\%$ premature contractions in a context of normal esophago-gastric junction relaxation
 - Presence of at least two premature contractions with a distal latency of under 4.5 seconds

Case Discussion: Distal Esophageal Spasm

Hierarchical Approach

In the diagnosis of esophageal dysmotility

The Chicago classification v3.0
Hierarchical analysis



Step 1: Integrated Relaxation Pressure (IRP) is **greater** than the upper limit of normal (ULN), suggestive of insufficient lower esophageal sphincter relaxation

- Identify type of achalasia/obstruction from there using:
 - Premature Esophageal Pressurization (PEP):** Refers to pressurization of the esophageal body prior to the lower esophageal sphincter (LES) fully relaxes; elevation correlates to **Type II achalasia**
 - Distal Latency (DL):** Refers to the time interval from the start of a swallow (measured at the upper esophageal sphincter) to the contractile deceleration point in the distal esophagus; a shortened DL correlates to **Type III achalasia**
 - Absence of any peristalsis/contractility correlates to **Type I achalasia**
- If IRP is elevated but without any other findings and does not fit an achalasia category, consider a **mechanical obstruction**

Step 2: IRP is **normal** but...

- DL is shortened (<4.5s):** consider **Distal Esophageal Spasm (DES)**
- Distal Contractile Integral (DCI) is elevated:** consider **Jackhammer esophagus**
 - DCI:** Quantifies amplitude, duration, and length of the esophageal contraction in the distal esophagus.

Step 3: IRP is **normal** in the presence of ineffective swallows with on other findings, consider **Ineffective Motility (IEM)**

References:

1. Fonseca EK, Yamauchi FI, Tridente CF, Baroni RH. Corkscrew esophagus. *Abdom Radiol (NY)*. 2017 Mar;42(3):985-986.
2. Bauer K, Keller C. Organoaxial gastric volvulus: a rare cause of an acute abdomen. *GMS Interdiscip Plast Reconstr Surg DGPW*. 2019 Mar 25;8:Doc04. doi: 10.3205/iprs000130. PMID: 30984512; PMCID: PMC6441817.
3. Grübel C, Borovicka J, Schwizer W, Fox M, Hebbard G. Diffuse esophageal spasm. *Am J Gastroenterol*. 2008 Feb;103(2):450-7. doi: 10.1111/j.1572-0241.2007.01632.x. Epub 2007 Nov 15. PMID: 18005367.
4. Burmeister S. Review of current diagnosis and management of diffuse esophageal spasm, nutcracker esophagus/spastic nutcracker and hypertensive lower esophageal sphincter. *Curr Opin Otolaryngol Head Neck Surg*. 2013 Dec;21(6):543-7. doi: 10.1097/MOO.0000000000000002. PMID: 24157634.
5. Valdovinos MA, Zavala-Solares MR, Coss-Adame E. Esophageal hypomotility and spastic motor disorders: current diagnosis and treatment. *Curr Gastroenterol Rep*. 2014 Nov;16(11):421.
6. Kahrilas PJ, Bredenoord AJ, Fox M, Gyawali CP, Roman S, Smout AJ, Pandolfino JE; International High Resolution Manometry Working Group. The Chicago Classification of esophageal motility disorders, v3.0. *Neurogastroenterol Motil*. 2015 Feb;27(2):160-74. doi: 10.1111/nmo.12477. Epub 2014 Dec 3. PMID: 25469569; PMCID: PMC4308501.
7. Achem SR, Gerson LB. Distal esophageal spasm: an update. *Curr Gastroenterol Rep*. 2013 Sep;15(9):325. doi: 10.1007/s11894-013-0325-5. PMID: 23892829.