

BIBLE CLASS: NEUROMUSCULAR ESOPHAGEAL DISORDERS

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Esophageal Motility Disorders: Current Approach to Diagnostics and Therapeutics



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Symptoms associated with neuromuscular esophageal disorders?

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Dysphagia

- Solids only: suggestive of mechanical obstruction
- Liquids +/- solids: suggestive of motility disorder

Chest pain

Regurgitation

Diagnostic tests

Diagnostic tests

Esophagogastroduodenoscopy

High resolution (impedance) manometry

Impedance planimetry (EndoFLIP)

Barium esophagogram

Endoscopy: what to look for?

- peptic stricture
- hiatal hernia
- Schatzki's ring
- eosinophilic esophagitis
- altered foregut anatomy

Diagnostic test	Protocol	Supportive evidence
Endoscopy	Careful attention to esophageal diameter, LES tone, retention of liquid/saliva on insertion	Puckered gastroesophageal junction with resistance to passage of scope with retained liquid in the esophagus is supportive of obstructive process such as achalasia Presence of "foam" in the esophagus suggest dysmotility but is not specific for a manometric diagnosis.

High resolution manometry

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Integrated Relaxation Pressure (IRP)

assesses the adequacy of swallow-induced LES relaxation defined by the median 4-second nadir of EGJ pressure in the 10-second postswallow period. Dependent on software/pressure transducer type

Normal: <15 mmHg supine, <12 mmHg upright (Medtronic)

<22 mmHg supine, <15 mmHg upright (Laborie/Diversatek)

Distal Contractile Integral (DCI)

DCI is a three-dimensional metric to assess the contraction vigor of esophageal smooth muscle contraction, integrating length, amplitude and duration of contraction

Normal: 450-8000 mmHg.s.cm

Weak: 100-449 mmHg.s.cm

Failed: <100 mmHg.s.cm

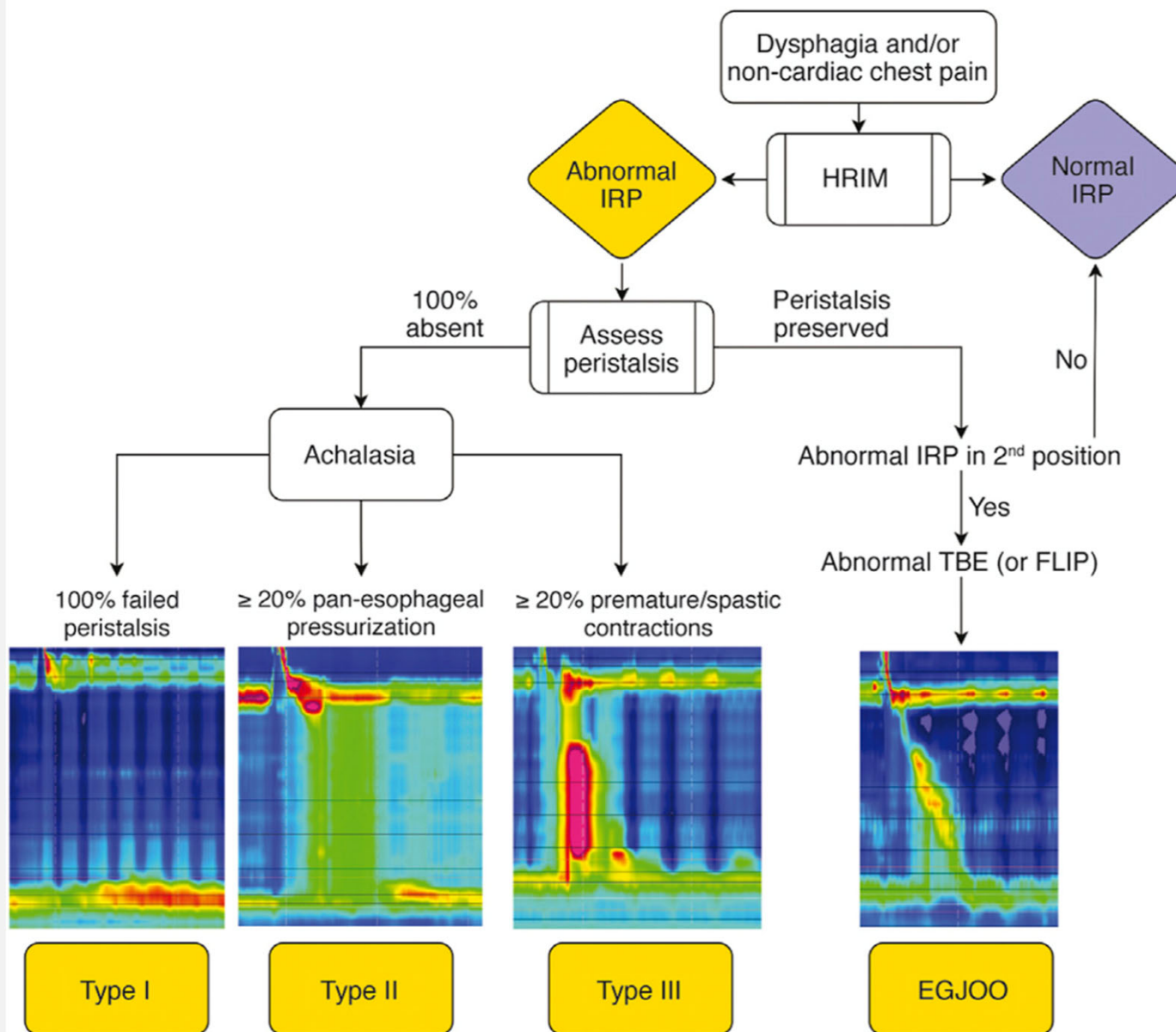
Hypercontractile: >8000 mmHg.s.cm

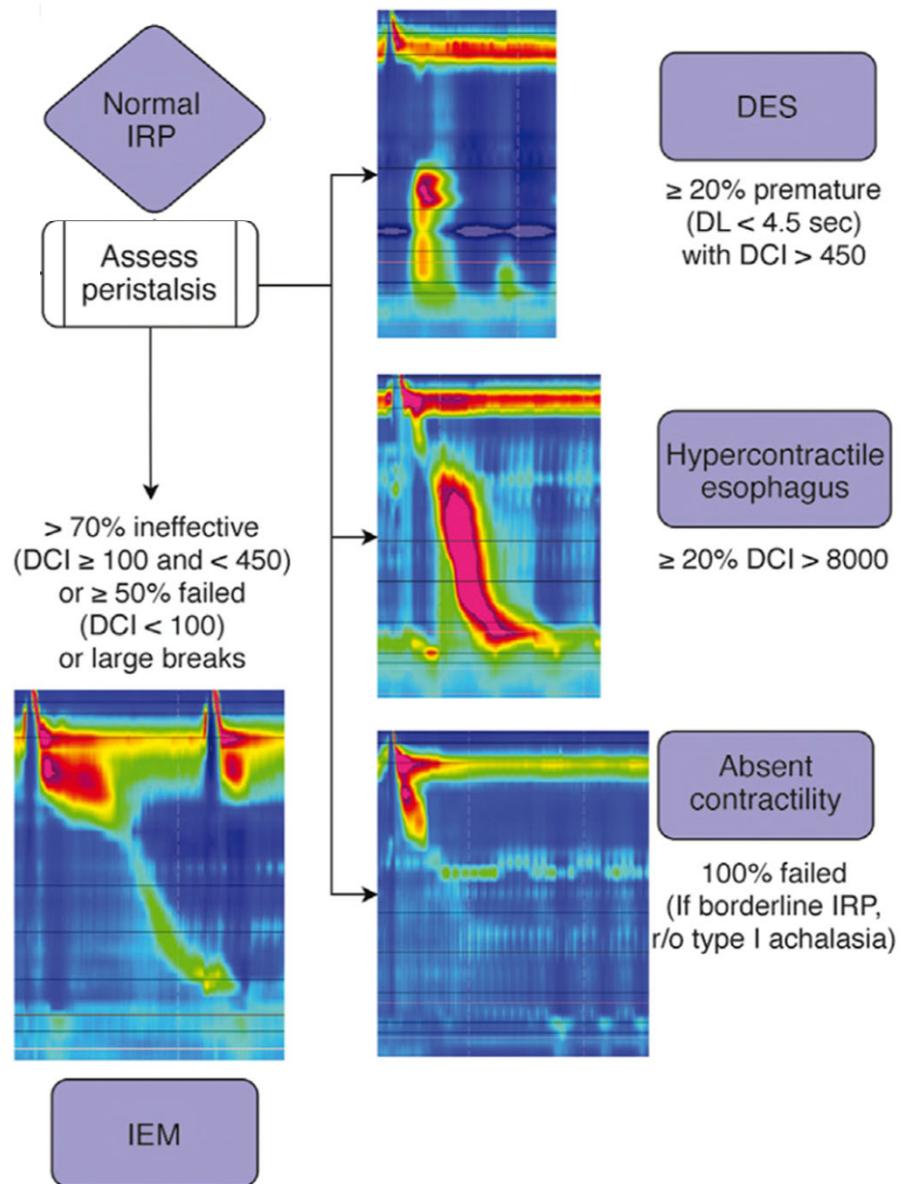
Distal Latency (DL)

DL measures timing of peristalsis: interval between upper esophageal sphincter relaxation to contractile deceleration point of the peristaltic wave in the distal esophagus.

Premature: DL <4.5 seconds

Peristaltic: DL \geq 4.5 seconds





High resolution manometry

Supportive evidence:

Multiple rapid swallows

Rapid drinking challenge

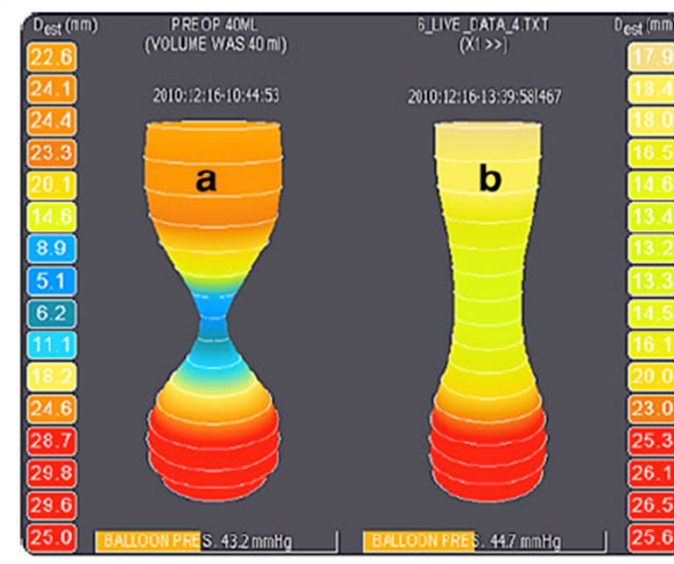
MRS (HRM)	5 swallows of 2-mL liquid at 2–3 s intervals	Absence of esophageal body contractility (DCI <100 mm Hg•s•cm) with complete deglutitive inhibition of LES during MRS Augmentation (peristaltic reserve) present if post-MRS esophageal body peristaltic contraction is normal (DCI >450 mm Hg•s•cm) and any of 3 post-contractions with increased contractile vigor (DCI >single swallow mean DCI)
RDC (HRM)	Rapid drink of 200 mL of liquid	Absence of esophageal body contractility (DCI <100 mm Hg•s•cm) with complete deglutitive inhibition of LES during RDC IRP >12 mm Hg (Medtronic software) and pan-esophageal pressurization (>20 mm Hg) are supportive of outflow obstruction

Impedance planimetry (FLIP)

FLIP = Functional Lumen Imaging Probe

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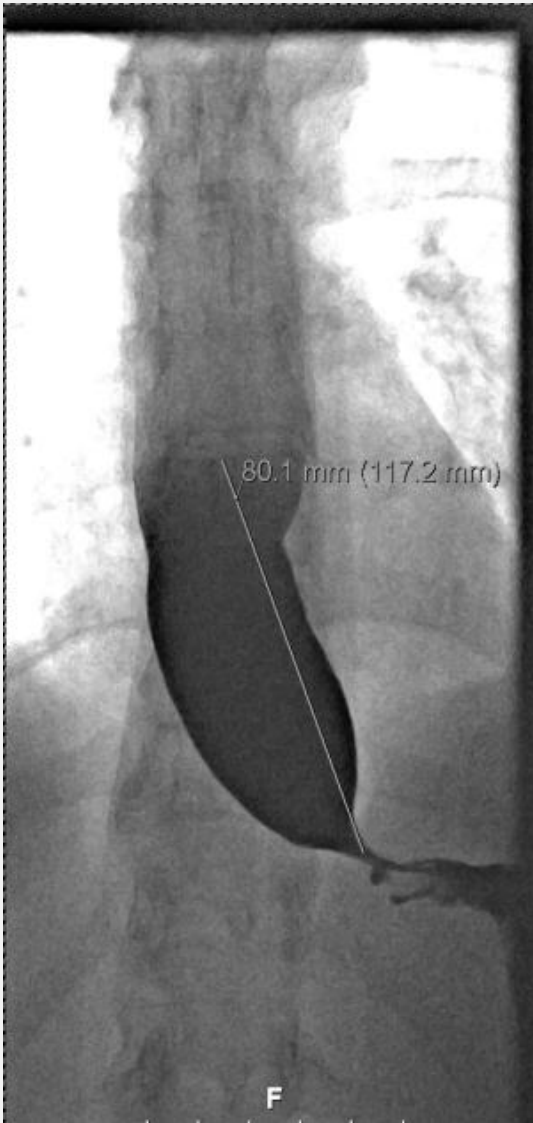


FLIP

Placed transorally with balloon
traversing the EGJ
Volumetric distention of the balloon
to predetermined volumes based
on catheter type

EGJ-DI of $<2.0 \text{ mm}^2/\text{mm Hg}$ is abnormal
EGJ diameter of $<13 \text{ mm}$ is likely abnormal
Role in various manometric diagnoses is evolving

Barium Esophagogram



Barium Esophagogram

Perform in upright position using 8 oz
or 236 mL of barium
Evaluate barium height at 1, 2, and
5 min

Barium column height to support outflow obstruction (such as
achalasia)
>5 cm at 1 min
>2 cm at 5 min
13-mm barium tablet retention supports obstructive process
3% improvement (decrease) in pre- to post-treatment barium height
at 5 min might predict long-term clinical remission in achalasia
Corkscrew appearance may be suggestive of distal esophageal
spasm

ACHALASIA

Elevated Integrated Relaxation Pressure (IRP),
no normal peristalsis

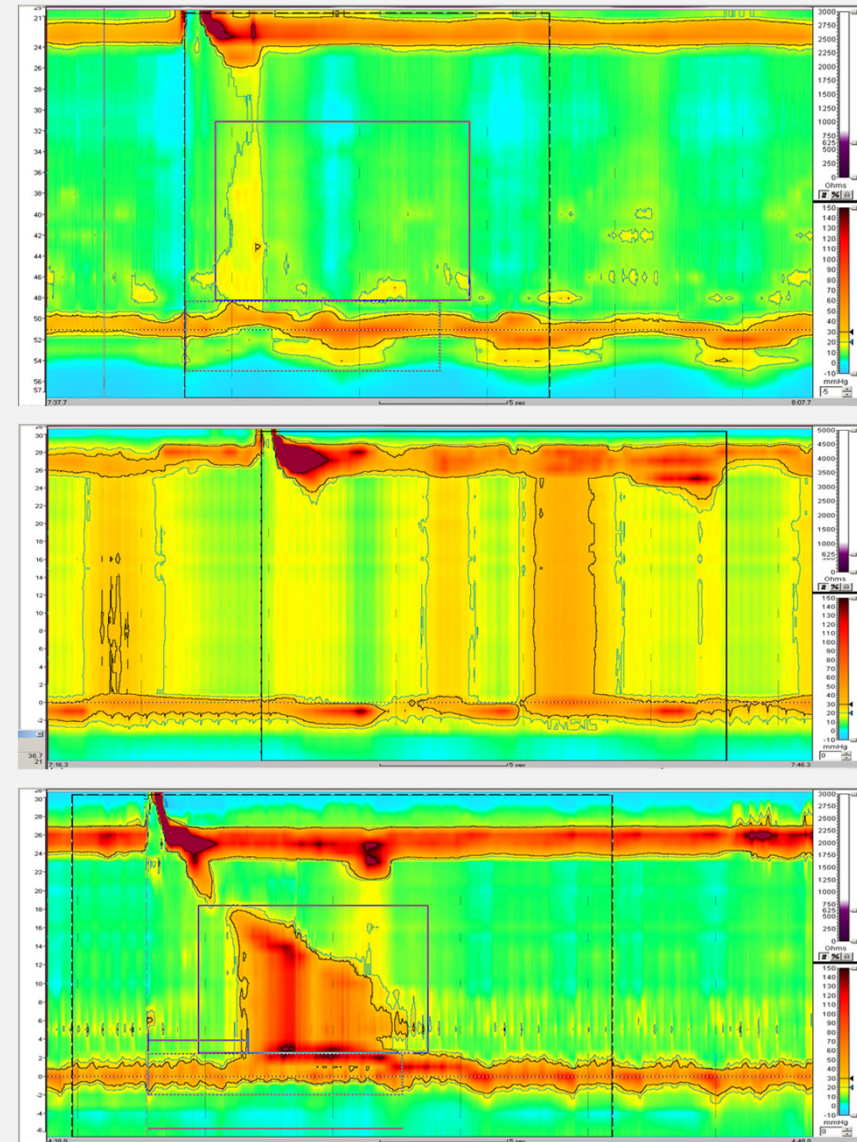
Elevated LES resting pressure, esophageal
pressure

Typ I: - no panesophageal pressurisation

Typ II: - panesophageal pressurisation
>30 mmHg in $\geq 20\%$ of swallows

Typ III: - spastic, premature contractions
in $\geq 20\%$ of swallows

Kahrilas PJ et al. Neurogastroenterol Motil 2015;27:160–174



Achalasia: Treatment

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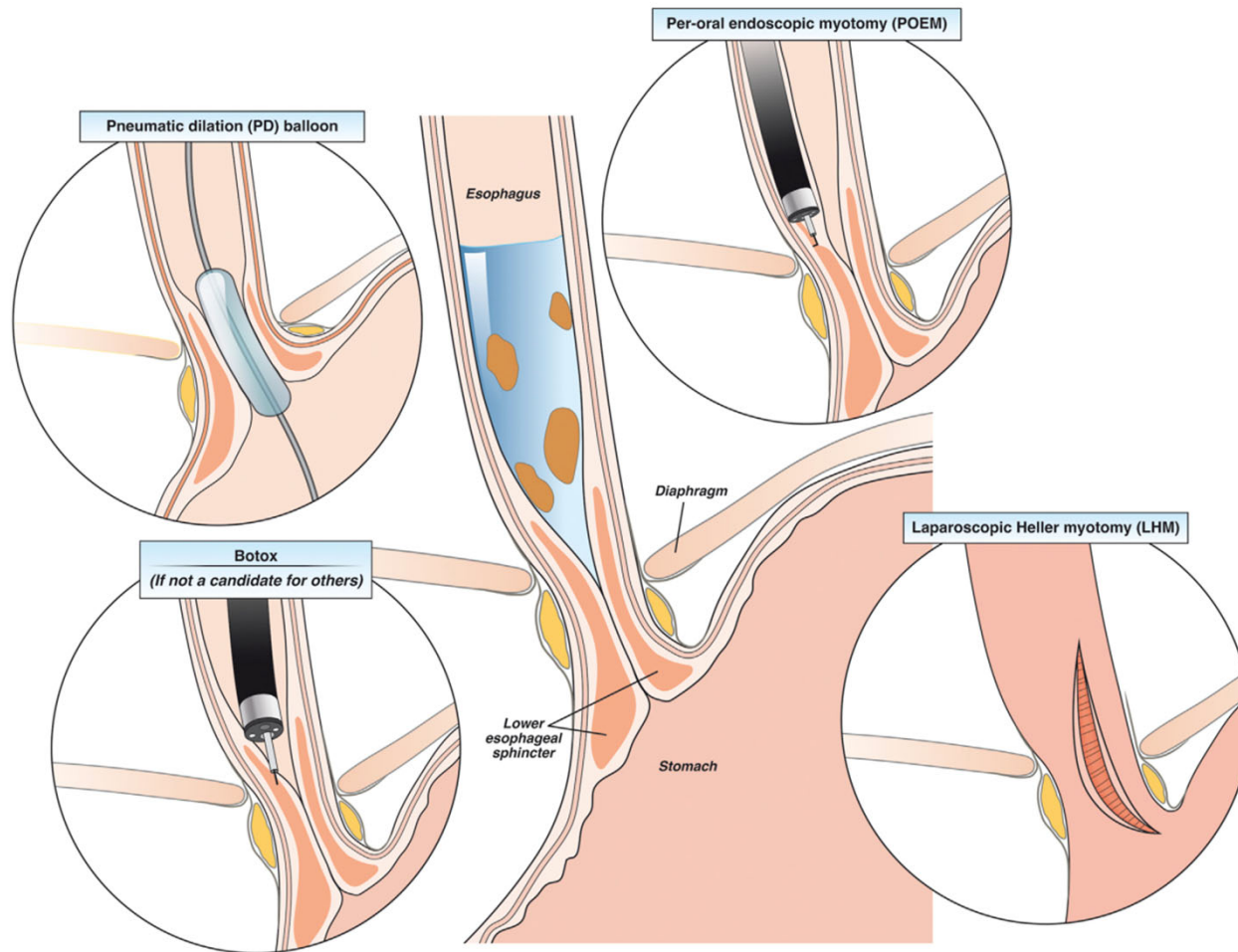


Figure 2. Treatment options in patients with achalasia.

Esophagogastric Junction outflow obstruction(EGJOO):Treatment

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EGJOO

- may be an artifact of HRM testing,
- therapy should be based on predominant symptom and severity of symptoms
- 52%–92% of patients with mild symptoms might have spontaneous resolution.
- Standard endoscopic dilation:response rate of 69.6%
- botulinum toxin injection: response rate 63.6%

Disorders of peristalsis: Treatment

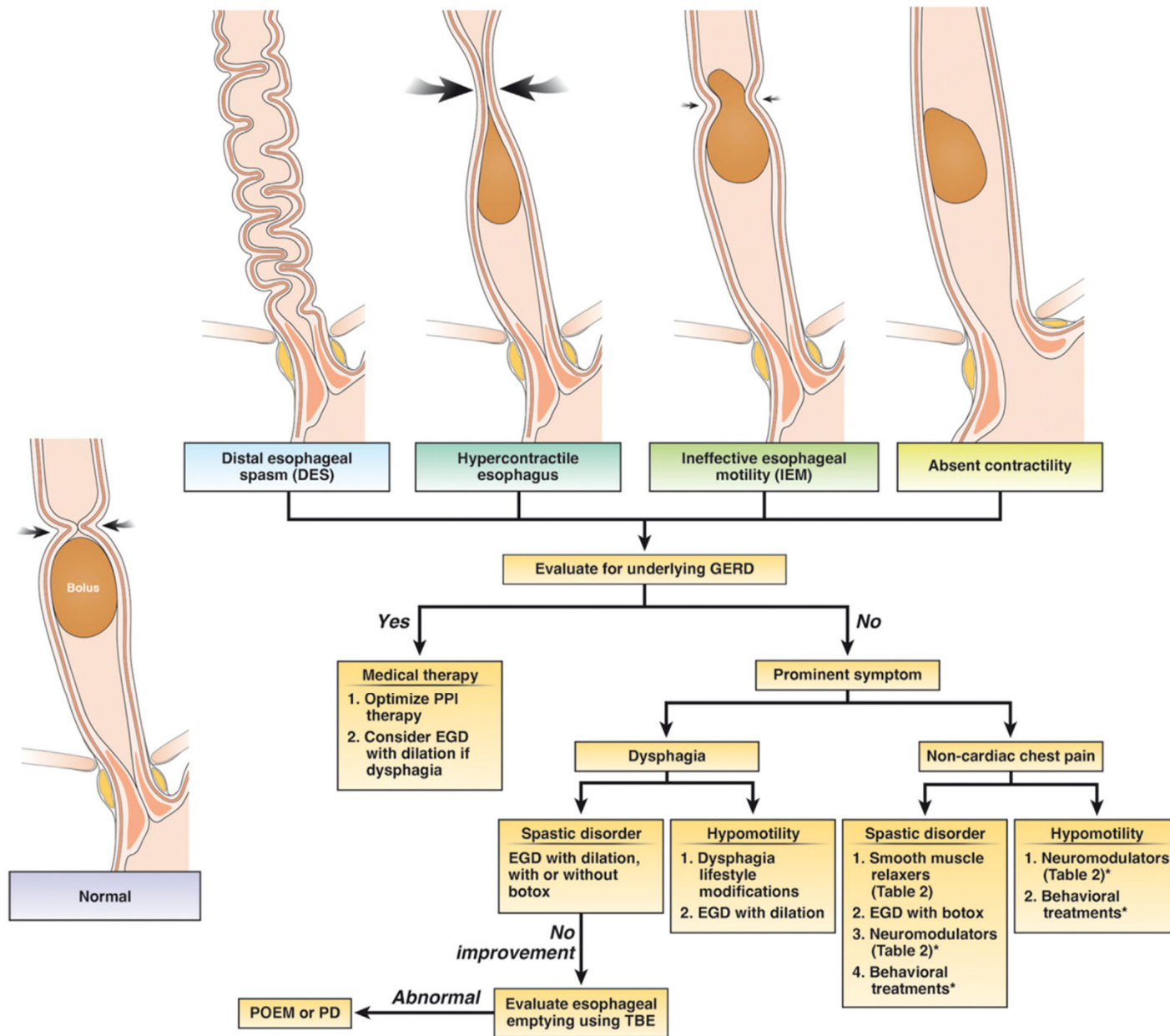


Table 2. Smooth Muscle Relaxers and Neuromodulators That Might Be Used in Patients With Disorder of Esophageal Peristalsis and NCCP as the Primary Symptom

		Dosage	Level of evidence	Side effects	Special considerations
Smooth muscle relaxers	Peppermint oil	5 drops in 10 mL of water (or 2 Altoid mints sublingual before every meal)	Case series ¹⁴⁷	Might worsen reflux symptoms	Decreases simultaneous contractions and amplitude, but less effect on pain
	Calcium channel blockers	Diltiazem 60–90 mg 4 times a day Nifedipine 10 mg 30 min before meals	RCT in nutcracker esophagus ¹⁰⁵	Hypotension, peripheral edema, headaches, dizziness, and tachyphylaxis	Improves chest pain and decreases esophageal amplitude
	Phosphodiesterase-5 inhibitor	Sildenafil 25–50 mg twice a day	RCT in spastic motor disorder ¹⁴⁸	Headache, hypotension, and dizziness	Might improve manometric findings, but less effect on symptoms
TCAs	Imipramine Amitriptyline	50 mg daily 10–25 mg at night	RCT for NCCP ^{111,149} RCT for NCCP ¹⁵⁰	QT prolongation, dry mouth, excessive sleeping, dizziness, and constipation	Might use nortriptyline (less anticholinergic effects in hypomotility disorder and NCCP)
SNRI	Venlafaxine	75 mg daily	RCT for NCCP ¹⁵¹	Sleep disturbances, nausea, hypertension	Less constipation compared with TCA, but similar pain relief
SSRI	Sertraline	50–200 mg daily	RCT for NCCP ^{152,153}	Nausea, restlessness, dry mouth, diarrhea	Might be beneficial if concomitant anxiety disorder or symptom hypervigilance
	Paroxetine	10–50 mg daily	RCT for NCCP ¹⁵⁴		More drug-drug interactions due to inhibition of P450 isoenzymes Short half-life, risk of discontinuation syndrome if stopped abruptly Fluoxetine preferred in patients who may miss doses (due to longer half-life)
Miscellaneous	Trazadone	100–150 mg daily	RCT in spastic motor disorder ¹¹²	Nausea, dizziness, and drowsiness	Less effect on manometric abnormalities, but significant improvement in chest pain