



## **Esophageal Dysmotility and Achalasia**

Esophageal dysmotility, the result of abnormal peristaltic function of the esophagus is idiopathic and associated with aging. Diffuse esophageal spasm, a unique primary cause of esophageal dysmotility presents with non-cardiogenic chest pain that may or may not be related to active swallowing. If esophageal dysmotility occurs in the setting of poorly controlled diabetes, chronic reflux, or scleroderma, dysmotility secondary to these systemic disorders should be considered.

Achalasia is the poor relaxation of the gastroesophageal sphincter with associated dysmotility and diffuse dilation of the esophagus. Patients with long-standing achalasia are at greater risk of aspiration and malignancy in the dilated esophagus. Smooth beak-like tapering of the distal esophagus is the characteristic finding of this lesion on modified barium swallow.

### **Anatomy & Physiology**

- The esophagus is a dynamic tube, innervated by the vagus nerve, with unique neuromuscular anatomy and peristaltic mechanism to effectively propel food to the stomach. The inner circular layer of the muscularis propria of the esophagus is arranged in concentric circles and is responsible for the sequential peristaltic contractions that propel the food bolus toward the stomach.
- The lower esophageal sphincter (LES) consists of a thickening of the circular esophageal musculature at the entrance of the stomach, manometrically defined as a high-pressure zone located at the gastroesophageal junction.
- With each swallow a peristaltic wave starts above the pharynx and moves in a distal direction. This pushes the bolus through the relaxed UES. Primary peristalsis, the peristaltic wave initiated by the swallow moves further in a distal direction with a propagating velocity of 3 to 5 cm/s. If the esophagus is dilated in the absence of swallowing, for example, during inflation of air during endoscopy, secondary peristalsis will be triggered. It starts at the location of the distension. The volume and viscosity of the bolus influence the amplitude and velocity of propagation of the peristaltic wave via input of sensory nerves to the enteric nervous system and central nervous system. During swallowing the LES relaxes so the bolus can pass the sphincter and reach the stomach. After relaxation the sphincter returns to contracted resting state.



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### **Assessment**

- The gold standard for the workup of suspected esophageal dysmotility is high resolution manometry (HRM). However, modified barium swallow (MBSS) and esophagram can also be helpful.
- In the setting of distal esophageal spasm patients may have symptoms very infrequently, which makes it difficult to diagnose. The primary means of diagnosis is esophageal manometry. Sometimes spastic contractions are also seen during endoscopy and on radiographic images (MBSS or esophagram).
- Achalasia is diagnosed by a combination of barium swallow and esophageal manometry. The two pathognomonic findings on manometry are aperistalsis of the esophageal body and impaired relaxation of the LES during swallowing. Esophageal contractions that occur are usually low in amplitude and simultaneous throughout the esophagus. A functional obstruction of the esophagus that leads to dilation and stasis. High resolution manometry has revealed several achalasia subtypes. Type I is classic achalasia, Type II is characterized by pan-esophageal pressurization, and Type III is characterized by spastic contractions.
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### **Pathophysiology**



- Esophageal dysmotility occurs when there is an aberration of esophageal peristalsis. This can be caused by distal esophageal spasm, hypercontractile esophagus, or absent contractility. In distal esophageal spasm premature rapid propagating contractions are triggered by emotions or by eating foods or by gastroesophageal reflux.
- Hypercontractile esophagus is also called nutcracker esophagus or jackhammer esophagus. In this condition the esophageal contraction amplitude is abnormally high or the contraction duration long, while there is normal LES relaxation.
- Loss of contraction, or aperistalsis is also a cause of esophageal dysmotility. In absent contractility there are no signs of peristaltic contractions, but the LES relaxation is normal. In some cases, it's secondary to systemic sclerosis. Ineffective esophageal motility and fragmented peristalsis are causes of esophageal dysmotility. Ineffective esophageal motility is characterized by a very low amplitude of peristaltic contractions. Fragmented peristalsis is characterized by the presence of large pressure breaks in the peristaltic contractions in the esophagus. Weak contractions impair clearance of refluxed gastric contents and chronic excessive acid exposure leads to weakening of peristaltic function.
- Esophageal dysmotility is the result of another disease. In patients with scleroderma, CREST (calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, telangiectasia), and mixed connective tissue disease, the smooth muscles of the esophagus can be involved, causing very weak to absent peristalsis in the distal esophagus. There is a progressive fibrosis of smooth muscle resulting in decreased peristalsis to aperistalsis. The pressure in the LES is usually very low. In the case of polymyositis and dermatomyositis, the striated muscles of the esophagus can be involved.
- Achalasia is a primary esophageal motility disorder, characterized by the absence of esophageal peristalsis and impaired relaxation of the lower esophageal sphincter. These abnormalities stem from the impairment of the inhibitory innervation to the esophageal smooth muscle and the lower esophageal sphincter. The cause of achalasia is unknown, but the disease may be autoimmune, secondary to viral infection or neurodegenerative.
- Secondary achalasia, or pseudoachalasia refers to achalasia occurring secondary to malignancy or other entities (Chagas disease or bariatric surgery). Chagas disease is



caused by infection with the parasite *Trypanosoma cruzi* and is characterized by widespread destruction of the myenteric plexus

- The pathophysiology of achalasia involves impairment of the inhibitory innervation to the esophageal smooth muscle and LES. An inflammatory process leads to degeneration of ganglion cells in the myenteric plexus of the esophageal body and the LES, which results primarily in the loss of the inhibitory neurotransmitters nitric oxide and vasoactive intestinal polypeptide. The inflammatory reaction is associated with infiltration of T cells, which leads to slow destruction of ganglion cells. The underlying cause is unknown. .
  - Stavropoulos S, Friedel D, Modayil R et al. Diagnosis and Management of esophageal achalasia. *BMJ*. 2016;353: i2785.
  - Bredenoord A; Smout A, Tack J. Ed. (2016). *The Esophagus In: A Guide to Gastrointestinal Motility Disorders*. New York: Springer, p39-47.

### **Treatment**

- The treatment for esophageal dysmotility depends upon the underlying cause. For distal esophageal spasm medications that induce smooth muscle relaxation, such as nitroglycerine and calcium channel blockers are moderately effective. In severely symptomatic cases endoscopic botulinum toxin injection or even myotomy may be considered. For hypercontractile esophagus the treatments are the same.
- For ineffective motility and fragmented contractions treatment with acid-suppression mediations will sometimes lead to improvement in esophageal motility. In this group prokinetic drugs have no proven efficacy in the improvement of esophageal hypocontractility.
- Achalasia, while not effectively treated medically, it can be effectively treated in most patients by pneumatic dilation, Heller esophagomyotomy or peroral endoscopic myotomy (POEM)
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  - Stavropoulos S, Friedel D, Modayil R et al. Diagnosis and Management of esophageal achalasia. *BMJ*. 2016;353: i2785.