Management of Spastic Disorders of the Esophagus

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KEYWORDS

• Distal esophageal spasm • Achalasia • Jackhammer esophagus
• High-resolution manometry • Esophageal motility

KEY POINTS

- Largely as a consequence of refined classification made possible with high-resolution manometry (HRM) and esophageal pressure topography (EPT), the current concept of esophageal spastic disorders has evolved to encompass spastic achalasia, distal esophageal spasm (DES), and jackhammer esophagus.
- These esophageal spastic disorders are conceptually distinct in that spastic achalasia and DES are characterized by a loss of neural inhibition, whereas jackhammer esophagus is associated with hypercontractility, presumably by activation of the cholinergic pathway.
- Because the defining endoscopic features may also occur in the setting of esophagogastric junction (EGJ) obstruction, endoscopic examination is required when esophageal spastic disorders are suspected to evaluate for mechanical obstruction.
- Therapeutic management depends on the presence of EGJ outflow obstruction.
- Extensive myotomy using the POEM technique might have a role in cases of treatment failure.

INTRODUCTION

Although reflux disease is the most common cause of esophageal chest pain, esophageal manometry is often done in the course of its evaluation, and manometric abnormalities indicative of DES are often reported. The identified abnormalities, however, are rarely the cause of chest pain and most investigators would agree that the clinical diagnosis of DES is overused. It was that observation that led to a classic reappraisal of DES by Richter and Castell,[1] conceived during the renaissance of esophageal

This work was supported by Grant No. R01DK56033 from the National Institutes of Health.
Conflict of interest: SR has served as consultant for Given Imaging.
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Gastroenterol Clin N Am 42 (2013) 27–43
http://dx.doi.org/10.1016/j.gtc.2012.11.002 **gastro.theclinics.com**
0889-8553/13/$ – see front matter

manometry in the early 1980s. Arguing for a more restrictive use of the diagnosis, those investigators proposed 2 required manometric criteria for DES: (1) simultaneous contractions in greater than 10% of wet swallows and (2) intermittent normal peristalsis. Other associated features were also described and some minor modifications were subsequently made, but it was these 2 criteria that became part of the lore of (conventional) manometry.

A lot has changed with respect to esophageal motility testing since 1984. Clinical studies are now commonly done with high-resolution systems using in excess of 30 closely spaced pressure transducers, and esophageal contractile patterns are displayed and analyzed in terms of pressure topography rather than as line tracings. Merging these concepts, current motility studies are more accurately termed, *HRM imaged with EPT*. Although these innovations had their roots in the early 1990s with the pioneering studies of Clouse and colleagues,[2–4] it was not until recently that commercial units became available, facilitating widespread adoption of EPT by the clinical community. The advantages of EPT compared with conventional manometry are several: (1) high-quality studies can be obtained that simultaneously image the entire esophagus, (2) standardized objective metrics have been developed for interpretation,[5–8] and (3) topographic patterns of contractility are easily learned and recognized with great reproducibility.[9,10] EPT also presented challenges, however, not the least of which was the need to reconsider the classification of esophageal motility developed for conventional manometric systems.[11] That classification effort led to improved understanding of achalasia subtypes[12] and hypomotility patterns.[13] Headway has also been made in the domain of hypercontractile conditions, including DES.[14,15] This work led to a conclusion, however, that the 2 essential criteria identified by Richter and Castell were suboptimal for defining DES as imaged in EPT and identified a heterogeneous group of patients, most of whom did not have DES.[14] Hence, the aims of this synopsis are to update the understanding of esophageal spastic disorders in the era of EPT.

WHAT ARE THE SPASTIC DISORDERS OF THE ESOPHAGUS?

Spastic disorders of the esophagus might be conceived of as hyperactive conditions of the esophagus due to contractions of either abnormal propagation (premature contractions) or extreme vigor. In the current iteration of the Chicago classification of esophageal motility disorders,[16] the relevant diagnoses are spastic (type III) achalasia, DES, and hypercontractile (jackhammer) esophagus. Despite differences in pathophysiology, which are discussed, these disorders share many similarities, including their clinical presentation: dysphagia, chest pain, regurgitation, and/or heartburn. The identification of these spastic disorders is based on the contractile pattern observed using HRM with EPT. The current Chicago classification criteria for identification of the spastic disorders are summarized in **Tables 1** and **2**.[16]

Distal Esophageal Spasm and Spastic Achalasia

Definition

DES is an uncommon disorder characterized by an impairment of ganglionic inhibition in the distal esophagus. Using conventional manometry, DES was defined by the presence of simultaneous contractions.[11] Using HRM with EPT, however, the higher-resolution recordings demonstrated that propagation velocity normally varies greatly along the length of the esophagus and finding regions of rapid propagation is common. A consequence of this finding is that the finding of rapidly propagated contractions is nonspecific for esophageal spasm.[14] Alternatively, premature contractions, defined by