Endoscopic Myotomy for Non-achalasia Esophageal Disorders

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Non-achalasia disorders of the esophagus represent a wide variety of motor issues encountered in clinical practice. In this chapter, we will focus on those disorders that can best be described along a spectrum of abnormal contractility that may be amenable to myotomy to alleviate obstructive and/or painful symptoms. Before moving into a discussion of specific manometric abnormalities, it is important to understand the concept of why an endoscopic myotomy might be considered in the first place. The idea is that the area of abnormal contractility, by virtue of esophageal spasm or extreme contraction vigor, causes outflow obstruction and/or pain somewhere along the esophagus. It is simplest to understand the mechanism when the pathology is isolated to the lower esophageal sphincter. Formal "esophageal outflow obstruction" refers to a phenomenon unique to the lower esophageal sphincter and results from a failure in relaxation with the onset of a swallow leading to symptoms that can mimic formal achalasia or even heartburn. In this condition, relief of the obstruction by physically lysing the sphincter and rendering it non-functional makes inherent sense, especially given our understanding of achalasia and favorable results of myotomy. However, failure to propagate a normal peristaltic wave in the body of the esophagus as a result of spasm or hypercontractility can also lead to symptoms of pain and dysphagia from compartmentalization. This can occur throughout the esophageal body or in segments and represents similar pathophysiology as "esophageal outflow obstruction," but in a more proximal location. The compartmentalization in the spastic segment can also lead to feelings of regurgitation from retrograde flow depending on the size, consistency, and timing of the bolus. Theoretically, if there is excessive contractile strength in the segment, with or without official spasm and compartmentalization, the contraction could be perceived as painful. Or the pain could actually be from a trapped bolus itself and the resultant stretch on the esophageal wall.

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If the hypercontractility is isolated to the distal esophagus and lower esophageal sphincter, the symptoms of pain can be replicated, although the sensation of regurgitation may be less perceptible perhaps due to less overflow phenomena given the greater capacity of the esophagus to accommodate the bolus.

Despite the intellectualization of how compartmentalization and hypercontractility may cause symptoms, the treatment for such esophageal disorders is not uniform or agreed upon. We try muscle relaxants, tricyclic antidepressants, botulinum toxin injections, and dilation with varying degrees of success. When all else fails, finally, some interventionalists will consider myotomy. Given the unpredictable response rates of patients' symptoms with myotomy, most surgeons are not willing to put a person through a major operation to "see what happens." This is especially true when contemplating a long thoracic myotomy. However, with the advent of endoscopic myotomy, the idea of accessing the muscular wall without traversing the chest is appealing. As the experience grows, we, as an endoscopic surgical community, are enjoying quicker operating times with less and less morbidity afforded with endoscopic esophageal myotomy. Today, the risk–benefit ratio is shifting regarding endoscopic myotomy so that the thought of "trying" an esophageal myotomy when more conservative measures have failed is far more reasonable.

The classification of "non-achalasia" esophageal disorders itself represents a host of various manometric features that may not fit into a single category quite as neatly as achalasia does. Because of this, there are few good papers published specifically on the effectiveness of esophageal myotomy for these disorders and fewer still for endoscopic myotomy [1]. Furthermore, the manometric criterion for so-called spastic esophageal disorders is evolving rapidly such that published data may be quickly obsolete depending on the manometric technology used for the study acquisition and on how the manometric findings are interpreted and categorized. Lastly, the reported sample size for any specific manometric category treated by any means is low, making generalizations from the literature extremely difficult for an individual patient sitting in one's office. In 2014, we published our experience with endoscopic myotomy in 25 non-achalasia subtypes as part of a 100 POEM series [2]. Of the 25 patients, 12 were originally categorized as hyper-contractile, defined as DCI > 5000 (mmHg)(s)(cm) when able, five had diffuse esophageal spasm and eight had isolated lower esophageal sphincter dysfunction. Taken as a whole, the non-achalasia cohort had reasonable improvements in symptoms, although significantly less impressive than the achalasia group. Specifically, dysphagia and chest pain were relieved in 97 and 100% of the achalasia group compared with 70 and 75% of the non-achalasia group, respectively. Since then, we have continued to collect patient data and re-review the original manometry studies in an attempt to unify the diagnoses in line with the updated Chicago Classification V3 [3]. As our cohort grows, preliminary data suggests that the non-achalasia subtypes are doing better than expected after endoscopic myotomy with overall success rates approximately 85% (unpublished data).

Although the concept is simple: if the abnormal area of the esophagus is causing obstructive symptoms, manifest primarily as dysphagia and perhaps chest pain and regurgitation, then preventing the contraction should be helpful to alleviate such symptoms, putting the concept to action is far more complex. It is not the surgery itself that is challenging, in most cases, but the patient selection.

As with all esophageal surgery, a comprehensive diagnostic evaluation is imperative prior to endoscopic myotomy. In brief, this includes cardiac evaluation to assess for cardiac sources of chest pain, upper endoscopy with biopsy to assess for malignancy, pseudoachalasia, hernia, etc., radiographic studies to evaluate for anatomic abnormalities, and quantitate emptying, manometry, and selective pH testing to rule out pathologic gastroesophageal reflux. Isolated endoscopic myotomy should not be performed in patients with abnormal acid exposure or hiatal hernias due to the inherent "refluxogenic" nature of the procedure. Accompanying chapters in this text cover the details on preoperative evaluation prior to endoscopic myotomy.

After the alternative diagnoses have been eliminated, it is reasonable to consider an operation for a non-achalasia esophageal motility disorder. First and foremost, there needs to be a symptom profile that reasonably lends itself to the concept that a myotomy would be helpful to relieve such symptoms and that relief would have a positive impact on the patient's quality of life. The primary symptoms that fit these criteria are dysphagia, chest pain, and regurgitation in the setting of hypercontractility or esophageal spasm. Even if the person has the most impressive manometry one has ever seen: No symptoms? No surgery! This is particularly relevant when considering variations in manometric technology, techniques, and normative values across diagnostic laboratories. One must review the raw data/pressure topography when planning an endoscopic myotomy for non-achalasia disorders.

Many people create diagnoses such as "achalasia variant" or "evolving achalasia" to describe the subtypes of manometric features that do not fit neatly into a named disorder category, but have elements of obstruction/compartmentalization either in the esophageal body or gastroesophageal junction. These terms are imprecise and are not encouraged. With the latest version of the Chicago Classification of Esophageal Motility Disorders V3 [3], it should be very rare that a recognized interpretation cannot be identified that fits all findings seen on pressure topography (Table 11.1). However, until the adoption of the Chicago Classification becomes universal in all testing laboratories, it is important to clarify some areas of change between the conventional and new high-resolution terminology that frequently lead to the confusion. These key points are particularly relevant to determine if a patient

Table 11.1 Manometric features of esophageal disorders possibly amenable to endoscopic myotomy

	IRP	% Normal peristalsis	% Premature contractions (spasm) with normal DCI	DCI (mmHg)(s)(cm)
Type I/II achalasia	High	0%	0%	<100
Type III (spastic achalasia)	High	0%	20%	>450
DES	Norm	30–80%	>20%	>450
Hyper-contractile (jackhammer)	Norm or high	30–80%	<80%	>8000 (in at least 20%)
EGJ outflow obstruction	High	>20%	n/a	>450

Adapted from The Chicago Classification of esophageal motility disorders, v3.0 [3]. *IRP* integrated relaxation pressure, *DCI* distal contraction integral

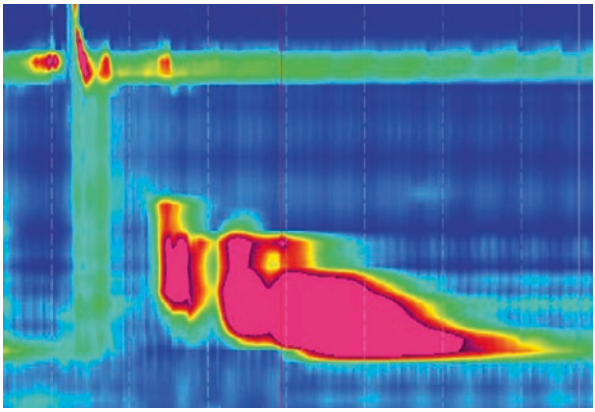

Fig. 11.1 HRM demonstrating typical hyper-contractile esophagus or Jackhammer pattern with a DCI of >1000 mmHg s cm without esophageal outflow obstruction

is suitable for endoscopic myotomy as manometric interpretation can be complex after achalasia has been ruled out. According to the most recent reiteration of the Chicago Classification, the term "hypercontractility" refers to increased contraction vigor confined to the esophageal body, extending into the lower esophageal sphincter, or confined to the sphincter alone. Such "jackhammer" patterns are conventionally known as nutcracker esophagus with or without a hypertensive sphincter (although the new criteria for "jackhammer" is more specific than "nutcracker," further discussion is beyond the scope of this chapter) (Fig. 11.1). Similarly, the new terminology "esophagogastric junction outflow obstruction" refers specifically to isolated elevations in integrated relaxation pressure, which would conventionally fall into the category of "non-relaxing lower esophageal sphincter." However, hypercontractile esophagus isolated to the sphincter in association with an elevated integrated relaxation pressure can occur (conventionally known as hypertensive non-relaxing sphincter).

The finding of esophageal spasm is considered separately from hypercontractility, although it can have similarities with outflow obstruction. Esophageal spasm is defined as premature contractions of normal contraction vigor in more than 20% of test swallows. A premature contraction is defined by the rapidity by which the wave front moves from the initiation of a swallow to the distal esophagus. More precisely, it is the time interval between the relaxation of the upper esophageal sphincter to the inflection point of the contractile front of the propagated swallow within 3 cm of the lower esophageal sphincter (contractile deceleration point) known as distal latency. A normal distal latency is >4.5 s. Anything less than that is considered premature, rapid, or spastic. Importantly, the contractile deceleration point needs to be measured along the pressure wave created from the esophageal contraction not to be confused with the potentially elevated intrabolus pressure that precedes the waveform. Many automated computer-generated interpretations make this mistake and over-call esophageal spasm when it really represents isolated gastroesophageal

outflow obstruction. Patients with esophageal spasm are generally differentiated from spastic achalasia by the presence of an elevated integrated relaxation pressure. However, on occasion, some gray areas will be encountered when patients exhibit characteristics across categories. For example, achalasia should still be considered in patients with normal integrated relaxation pressures but 100% failed peristalsis, particularly if there is evidence of esophageal body pressurization. The point is, there is not a specific category for which myotomy could be applicable. The precise name applied to the disorder is less important than understanding the underlying pathophysiology that may be causing the symptoms one is trying to alleviate (Fig. 11.2).

Once one has determined that the esophageal manometric findings of hypercontractility and/or esophageal spasm are present and may correlate with a convincing symptom profile and there are no contraindications to endoscopic myotomy, the next step is surgical planning. The pressure topography from the high-resolution manometry needs to be carefully reviewed, this time as a physical map of the esophagus—again reading a report is not adequate. Look for the location and extent of the high-pressure zone. Compare the manometric findings with the films from the contrast esophagram. Where exactly is the problem? Where is the target relative to the gastroesophageal junction? Is it confined to the junction or does it extend proximal into the esophagus? This will help you build a surgical diagram and provide information directing how long a potential myotomy would need to be. Pay close attention to correlating the patient's symptoms with the objective tests. Importantly, the myotomy needs to extend across the gastroesophageal junction regardless of specific manometric findings confined to the sphincter. In our experience, leaving the junction intact in patients who have a targeted esophageal body myotomy alone leads to relative outflow obstruction and esophageal dilation along the myotomy even if the sphincter area was manometrically normal to begin with. However, determining proximal extent of the myotomy in non-achalasia disorders is determined by a combination of manometric findings, contrast studies, intraoperative visualization of the extent of the high-pressure zone, and symptoms. For example, a long myotomy may be the best choice if there is primarily sub-sternal chest pain and correlating spasm into the middle or proximal esophageal body. Similarly, if the patient describes sub-sternal dysphagia and regurgitation, the body may also need to be addressed. However, if a patient describes primarily lower dysphagia correlating with lower esophageal sphincter findings, a standard length myotomy focusing on the junction may be adequate. When in doubt, we suggest extending the myotomy proximally to release all areas of potential concern. From a technical standpoint, there are a few unique considerations associated with a long myotomy. First, make sure that the entry point is proximal enough to allow for sufficient overlap between the mucosotomy and the myotomy. Consider a few extra centimeters of overlap to account for the longer operative time and higher chance of tearing the mucosotomy with instrumentation. Importantly, be mindful that patients with esophageal body disease often have significantly hypertrophic muscularis propria, which requires much more energy delivery to achieve myotomy. We recommend actively managing the energy in the tunnel to decrease the risk of injury to

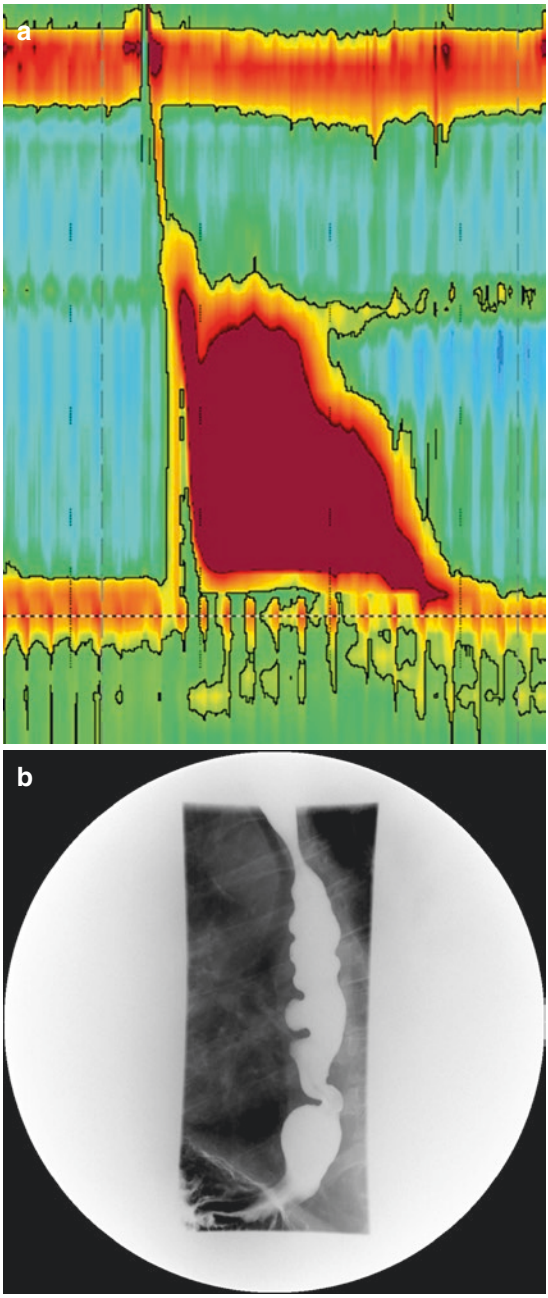

Fig. 11.2 (**a**) This swallow demonstrates a premature contraction (spasm) with elevated contraction vigor (Jackhammer) without esophageal outflow obstruction. There were 30% of swallows in this study demonstrating normal distal latency. (**b**) The esophagram from this same patient demonstrates a typical spastic pattern. Note in both studies the abnormal segment extends to just below the aortic arch (proximal indentation on esophagram and vascular artifact on manometric topography). This patient had a long endoscopic myotomy, which eliminated the dysphagia and improved but did not eliminate the chest pain

surrounding structures due to inadvertent conduction. Specifically, use the lowest energy settings possible to achieve a hemostatic myotomy, usually endocut modes, and switch to higher voltage or coagulation setting only when needed.

In summary, endoscopic myotomy for non-achalasia esophageal motility disorders is more complex both in terms of preoperative evaluation and surgical technique. The most frequent manometric classifications lending themselves to myotomy are hyper-contractile esophagus, esophageal spasm, and esophagogastric junction outflow obstruction. Despite a relative paucity of data, it seems that myotomy certainly can be performed with good results in carefully selected patients in whom the symptoms of dysphagia and chest pain correlate to manometric hypercontractility and spasm.

References

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