Esophageal Motility Disorders

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Definition based on x-ray or motility testing

Clinical context:  
Chest pain  
Dysphagia  
Absence of obstruction

Esophageal motility disorders, are defined really based on x-ray or motility testing. But, the clinical context is one of chest pain, dysphagia, in the absence of any obstruction.
There is a case history to start us off. This is a 36 year old woman who presents with chest pain occurring occasionally at night, heart burn, regurgitation of food and acid, and a rare trouble swallowing.
Management

• PPI
• Elevate the head of the bed
• Avoid late meals

• No improvement after 2 months

So, she is managed with PPI, told to elevate the head of the bed, and some of the recommendations we talked about when we discussed management of GERD, told to avoid late meals, and she returns two months later and says, “I’m not any better.”
What to do next? Is it an endoscopy, an x-ray, or should we double the PPI dose?
None of the above. What to do next is to take a more detailed history, because this is not your classic GERD. There is not even partial improvement. So now, you take a more detailed history, and what you find out. is that the dysphagia that was in your history initially as rare has actually occurred to solids and liquids equally. It has led to regurgitated food, and when you ask the patient, it looks undigested, therefore it has sat in the esophagus and come up without reaching the stomach. And, when she eats she occasionally reports chest pain. So now, you are on the right track because you suspect a motor disorder.
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<th>Key Questions for Diagnosis of Dysphagia</th>
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<td>• Is it truly dysphagia?</td>
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<td>• Is dysphagia for solids, liquids or both?</td>
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<td>• Is dysphagia intermittent or progressive?</td>
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<td>• At what level is the obstacle?</td>
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So, there are key questions to be asked when you’re trying to diagnose dysphagia. The first is, is it truly dysphagia? Patient will say, “I eat and it sits there. And, you translate that as food sticking, and in fact they are describing post prandial fullness. And, what I ask patient, I don’t say, “Do you have trouble swallowing?” Because most patients swallowing is a voluntary phase. And, they will say no, because they don’t perceive that the food sticking below in the retro sternal area is actually trouble swallowing. So I say, I ask patient, “Do you have food sticking?” And, I always get the answer I’m looking for.

The second question is, is it dysphagia for solids only, liquids or both? Because if it’s solids only, it’s mostly mechanical cause. Is it intermittent or progressive? Again, a stricture will cause sticking of a steak from time to time whereas achalasia will cause dysphagia on a regular basis and in increasing frequency. And, finally, at what level is the obstacle? Now, this is strictly again because for oropharyngeal dysphasia it is very easy clinically to recognize that the trouble is in the throat. But, beyond that, patients will tell you that the food stick in their throat when it’s actually stuck at the GE junction because of the referred discomfort.
Dysphagia
EGD or X-Ray First

Solids only
Interritten → EGD + dilation

Solids and liquids
Progressive → Videosophagram

So, the other debate is should we obtain EGD or x-ray first? And, those debates go on and on between surgeon, the gastroenterologist, and from a practical standpoint the answer to my, to me is very simple. If the patient is complaining of solids food dysphagia only, and it’s intermittent, the diagnosis is going to be either a stricture or a cancer, or a Schatzki ring And, I go for EGD first because not only will I make the diagnosis, but I can apply treatment at the same time and the patient consent can be obtained beforehand. If, on the other hand, patients are complaining now of solids and liquid dysphagia, and it is getting worse with time, I would start with an x-ray because it’s going to give me an idea of where the obstacle is, and if it’s a motor disorder, what the esophagus looks like.
How do we classify esophageal motor disorders? There are various ways to do it, and the simplest to my mind is to follow the anatomy and the physiology. The esophagus is divided in an upper third, which is striated muscle, under our own control, and lower two thirds, out of our hands, smooth muscle and it transitions on between the two. So, if you separate them this way, you will end up then in two sets of disease.

**Esophageal Motor Disorders Classification**

- Striated muscle disorders
- Smooth muscle disorders
Striated motor disorders are basically what was called in the good old days transfer
dysphagia, meaning the transfer of the bolus from the mouth to the esophagus, or what we
call today oropharyngeal dysphagia. So, the trouble is in the upper part. Patient cannot
form a bolus, or cannot propel the bolus, or chokes on his food. And, the causes are
multiple. Any neurological disease can do it, trauma can do it, but the major cause is going
to be a stroke. And, you will see this in patients in nursing homes, or in hospitals after a
stroke.
On the other hand, smooth muscle disorders, there are four major categories which we will address. Achalasia, which by far is more frequent, diffuse spasm, nutcracker esophagus, and scleroderma. So, I will say a word about each one of those.
Achalasia

- Aperistalsis
- Poor LES relaxation
- High or normal LES resting pressure

Let’s start with achalasia: what is it? A-chalasia—no movement. It’s aperistalsis of the esophagus by definition, not a single peristaltic contraction, poor relaxation of the lower esophageal sphincter, but the lower esophageal sphincter can be high, pressure can be high, or normal. In the old definition of the disease, we insisted on having a high pressure. It does not matter what the pressure you start with, it’s the fact that the pressure gradient does not break when you eat.
What’s the pathogenesis of the disease? All we know is that there is a patchy inflammation of the myenteric plexus of the esophagus with decreased ganglion cells. Therefore, there is a loss of latency gradient across the esophagus, because what the myenteric plexus does is make the esophagus when we swallow and we initiate a swallow, the esophagus follows in sequence, number one, two, three as you go down the esophagus. And, when you lose that progression, then the bolus is stagnates in the esophagus and leads to aperistalsis. And, the defect of inhibitory neurons in the LES prevent it from relaxing. In other words, when we swallow, we inhibit the tonic contraction of the LES, and when that doesn’t occur, the LES creates now an obstacle to the passage of the bolus.
Achalasia: Symptoms

All age groups
- Dysphagia to solids and liquids
- Regurgitation
- Chest pain (40-50%)
- Respiratory symptoms
- Weight loss, mild

What about symptoms? Well, achalasia affects all age groups, although there is a higher preponderance in young people in their 20’s and 30’s and then a second peak in later years in their 70’s. But, it affects all age groups, it’s typically dysphagia to both solids and liquids, with food regurgitation, chest pain when we looked at our patient at the Cleveland Clinic it’s present in 40 – 50%. Respiratory symptoms have been reported but are fortunately rare, and weight loss, interestingly, is mild. You can actually have obese patients with achalasia. And, this is a testimony to how patients can really work around that problem to continue to eat.
This is a typical x-ray of a patient with achalasia. The esophagus looks atonic. Why? Because you have barium up to the throat, and this is hard to achieve when patient is a normal patient is swallowing barium. This esophagus is dilated, and the typical finding is the deformity at the lower esophagus. If you look carefully you will see the esophagus tapering to what is referred to as the “bird beak like deformity” because it’s reminiscent of a bird’s beak.
As time goes by, however, the esophagus becomes more and more dilated, diverticulus start to form, and we start referring to this as a sigmoid esophagus.
Now, there is a manometric, manometry tracing this happens to be a normal tracing. And as we look at the one, two, three, four, five, six, seven leads here, ignoring the first, which really is the answer for swallowing, we see as the patient swallows the proximal lead in blue, the pressure decreases. This is the upper sphincter. And then, a wave progresses from proximal to distal with the lower, the lowest one, the red one, red and green, the lower esophageal sphincter relaxing with swallowing.
And, this is a more modern tracing using high resolution manometry where we have 35 sensors throughout the esophagus and it’s expressed in colors showing you here the intensity of pressure melded by the color and the progression in time. So, these are two normal swallows they progress nicely down the esophagus.
In contrast, this is a patient with achalasia. Now, unlike two slides away, you see now that the progression is not existent, the lower esophageal sphincter is not relaxing as nicely, and this is shown better in the next slide.
where you have now a straight line coming down, no progression, the esophagus contracting at the same time. And, the lower esophageal sphincter pressure is not dropping.
Now, what’s the treatment in achalasia? We unfortunately cannot restore the myenteric plexus; we cannot regenerate the nerves. Our goal is limited really to reducing the gradient of pressure represented by the LES. And, this can be achieved with one of few things.
We can use a pharmacologic agent such as nitrates or calcium channel blockers to do it. We can forcefully dilate the esophagus endoscopically or surgically by myotomy, and I will say a word about botulinum toxin.
Now, this is a slide showing the effect of nitrate and calcium channel blocker on the LES pressure. Once you administer either a calcium channel blocker in solid, in broken line, or a nitrate in solid line, you will see that LES pressure decreases significantly and the effect lasts for 40 minutes. So, presumably, the patient could take a nitrate under the tongue before eating, and for the next half hour the LES will be open enough to allow the presence of food.
Does this work? Yes, it works briefly; unfortunately there are common side effects which particularly in the elderly, namely hypotension, make this not a practical solution. The symptom relief is partial and in time they stop working. So, this, the pharmacotherapy of achalasia really should be reserved for patient who are ineligible or refuse any other treatment. And frankly, we have not used it much lately.
Well, how about pneumatic dilation? Pneumatic dilation is the forceful breakup actually of the LES; you can not achieve that with a maloney or savary dilator, although some patient do report, and there are series reporting benefit even from savary dilation. But, for more lasting effect you have to forcefully break the fibers of the esophagus.
And, this is the most available dilator today. As you can see there is a balloon, there are markers inside that balloon that will show up fleuroscopically, there are four white spots there, and you can remotely control the pressure of this balloon while looking fleuroscopically to make sure that your balloon is in the right place.
So, it’s an outpatient procedure; it’s combined with endoscopy. You need fluoroscopic assistance, and while the technique is variable there are all kinds of recommendation as to the time and the pressure of the balloon. I think the most prudent thing is to, while looking under fluoroscopy as the balloon creates a waist at the LES and the LES gets effaced, you stop, you hold it for 30 seconds, and you let down

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**Pneumatic Dilation**

- Outpatient procedure
- Combined with endoscopy
- Fluoroscopic assistance
- Technique variable
  - Inflation to ablate waist (30 seconds)
Now, the perforation unfortunately is the main concern with the pneumatic dilation. And in the best of hands it’s still 2 to 3%. It’s independent of age, I’ve seen it in young people, I’ve seen it in 82 year old people. The role of the technique is questionable; I don’t think it’s what you do, I think it’s the thinness of the esophageal wall. Unfortunately, when it occurs it’s going to require thoracotomy in most patients. And so, that’s the concern of pneumatic dilation.

Pneumatic Dilation: Perforation

- Independent of age
- Role of technique?
- Frequency 2-3%
- Requires thoracotomy
Surgical myotomy comes up looking better than pneumatic dilation at least initially. And, the misconception is once that you have operated, pneumatic dilation cannot be done. That is not true; we have dilated patient after surgery as well as sent patient who have failed pneumatic dilation to surgery. It’s of affected and accompanied by a fundoplication. Today, it’s done laparoscopically which obviously improves the outcome and makes the hospital stay much shorter. But, however simple it is, it should be done by an experienced surgeon. This is not a surgery to be done by a thoracic surgeon who has done one before in their lifetime.

**Surgical Myotomy**

- Results slightly superior to P.D.
- P.D. may precede or follow surgery
- Laparoscopic with fundoplication (partial)
- Experienced surgeon
And, this is how it is done. This picture show the myotomy already done; you see the esophageal mucosa bulging out, and the surgeon is applying now a fundoplication with a stitch on the stomach going around the stomach and then to the esophagus to create what they refer to as loose fundoplication. Because, if you do too tightly fundoplication you have defeated the purpose, of course, of reducing the gradient of pressure.
And, this is now the stitches are in place and the surgeon is about to tie them up.
How about botulinum toxin? Fortunately, its ease of administration is so that it has become in the hands of many gastroenterologists the primary treatment, and this is absolutely wrong. How does it work? It inhibits the release of acetylcholine and therefore decreases the LES pressure significantly, up to 60% or more. And, it’s done by injecting 80 to 100 units endoscopically in four quadrant fashion around the LES or slightly above.

**Clostridium Botulinum Toxin**

- Inhibits release of acetylcholine
- LESP ↓ 60%
- 80-100 units injected endoscopically in 4 quadrants

How about botulinum toxin? Fortunately, its ease of administration is so that it has become in the hands of many gastroenterologists the primary treatment, and this is absolutely wrong. How does it work? It inhibits the release of acetylcholine and therefore decreases the LES pressure significantly, up to 60% or more. And, it’s done by injecting 80 to 100 units endoscopically in four quadrant fashion around the LES or slightly above.
So, you mix the solution, and by the way once you’ve mixed it the shelf life is so short that you do not mix it until the patient shows up because it is expensive.
And, this is a picture showing how the needle is being inserted through the scope, and the medication is being applied around the esophagus. So, it’s fairly simple to do.
Botulinum Toxin: Indications

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<th>Appropriate</th>
<th>Inappropriate</th>
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<td>Elderly, frail patients</td>
<td>Young patients</td>
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<tr>
<td>High surgical risk</td>
<td>Healthy patients</td>
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<tr>
<td>Patient refused other Rx</td>
<td>Diagnosis doubtful</td>
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<td>Pseudoachalasia</td>
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But, the indication really should be appropriately in elderly, frail patient in whom no other treatment is being considered, who are a high surgical risk, who refuse other treatment or who have pseudoachalasia, which is a condition created by cancer. And, in whom the longevity is going to be very short. It’s inappropriate in young patients. It’s inappropriate in healthy patients or when the diagnosis in doubt. Why? Because in long term, botulinum toxin stops working. And, if you are going to consider surgery, the surgeon tells us that botulinum toxin causes scarring in the area and apparently makes their surgery more difficult. So, the first line of treatment in young, healthy patient is not botox.
Okay, now how do the two procedures compare? There are, unfortunately, one may be controlled study, but most comparative studies are looking at patients who are over time and they’re very imperfect. So, what we did at the Cleveland Clinic some years ago is call our patients who have either been pneumatic dilation or myotomy. And, obtain an x-ray and an interview. And, we found out that even patients who thought were doing well had actually a failure rate of 67% for myotomy and 74% for pneumatic dilation. So, what you have to keep in mind is whatever you do, in time the LES either goes up together or some phenomenon, but the symptoms return.

### Long-Term Results

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<th>Pneumatic Dilation</th>
<th>Myotomy</th>
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<tr>
<td>No. patients</td>
<td>47</td>
<td>15</td>
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<tr>
<td>Median months post Rx</td>
<td>85 (54-111)</td>
<td>73 (53-108)</td>
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<tr>
<td>Failure</td>
<td>35 (74%)</td>
<td>10 (67%)</td>
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- Failure defined by symptoms and X-ray
And, so what we do with our patient is obtain a timed barium swallow, and I will explain the technique in a minute briefly, and we ask them to come periodically, namely once a year to have this done, and we ask them how they’re doing and we look at the x-ray as well in order to stay ahead of the deterioration that occurs over time. And, (slide 34) the timed barium swallow is to give a certain amount of barium, the patient is asked to swallow it quickly over 30 seconds and upright left posterior oblique position. X-rays at one, two, five minutes we measured the width and the height of the barium column. And, this use is a simple way of judging the esophageal emptying over time by comparing x-ray to the other year after year.
So, this is an x-ray where at one, two, and five minutes you can see that the barium hasn’t changed. So, this is not a good result, because at five minutes there is still a column of barium hanging up.
So, how do we approach the treatment of achalasia? I think you have to ask your question, the first question, is the patient a good surgical risk or a poor surgical risk?
If it’s a good surgical risk, I go to laparoscopic myotomy because it is a great improvement on thoracic myotomy. It is, although there are complications, they are controllable. So, if the patient fail, we can go to pneumatic dilation, if they fail unfortunately only esophagectomy is the last recourse. If on the other hand, pneumatic dilation is chosen and it fails, you can always go to surgery. And, if it succeeds, you can repeat it two or three years from now.
If it’s a poor surgical risk, on the other hand, this is when I start with botox. If it succeeds, you repeat it as needed, and most patients will need it every six months to a year. If it fails, on the other hand, you can repeat it again, and if it fails again then you are left to either palliation or reconsidering again intervention and assessing the risk and seeing if the patient can submit to surgery or to pneumatic dilation.
A word about diffuse esophageal spasm. It’s a clinical entity expressed by dysphagia, chest pain, spastic contraction, but in the presence of peristaltic contraction. And, this is a main distinction with achalasia, where there is absolutely no peristalsis.
So, this is a tracing of a patient with diffuse spasm, and the first swallow you see on the left is normal. The second one in contrast, as you can see, the contraction is not peristaltic, it is much stronger and it lasts much longer.
And, this is expressed by high resolution manometry. Again, you can see the stark contrast between the first and the second swallow.
Now, on x-ray you will see the tertiary contraction and there are various names for it. Corkscrew esophagus, rosary bead esophagus, etc. etc. What I want to emphasize is seeing this is not uncommon in elderly patients, but they are not symptomatic. So, you cannot make the diagnosis of diffuse esophageal spasm on x-ray alone. You need the clinical context. And you cannot either tell elderly patients that it’s okay to have dysphagia, the work up of dysphagia in elderly patient is just as important.
Now, diffuse non cardiac chest pain has been touted as a major cause of esophageal problem. In fact, it’s not true. This is an old study taking 100 patients with non cardiac chest pain, they’ve already seen a cardiologist, come to your clinic and you suspect the esophagus. You do a motility testing and as you can see in blue, about 72% have a normal motility. Only 28% have abnormal motility. Of these 28%, 48% have nutcracker esophagus, which I will talk about shortly. Non esophageal, non specific motor disorder about a third, 36% in purple, only 10%, the orange quadrant, 10% have diffuse spasm. So, it’s 10% of 28%. So, if you take again 100 patients with non cardiac chest pain, only 2 – 3% have diffuse spasm. So, it’s not such a common condition.
The nutcracker esophagus was recognized about 20 years ago, and it’s a manometric entity. It’s as if I’m by high amplitude, higher than 180 mg of mercury on average in the distal esophagus, with a duration of more than 6 seconds. The contraction by definition are peristaltic; there is no diffuse spasm here. And, it’s been one of the differential diagnosis of chest pain.
It turns out with time, and this is an example of nutcracker esophagus the contractions look wild, but they progress. The food is progressing. There may be dysphagia rarely, but usually there isn’t.
And, this is a depiction in high resolution manometry of this weird contraction, but it moves down..
I think we all agree today that nutcracker esophagus is just an manometric variant, probably an epiphenomenon seen in patients with reflux, but frankly as a standing entity by itself clinically at least I don’t think it’s terribly important
Scleroderma, on the other hand, is obviously quite a distressing disorder, and it affects the esophagus by creating an inadequate LES with a distal loss of peristalsis.
And, this is an image of an esophagus with scleroderma. As you can see, it absolutely eight on esophagus, the lower esophageal sphincter is almost non existent down in orange and green.
And if you depict that in high resolution manometry it looks like nothing is happening.
So, what is my approach to dysphagia and esophageal motor disorder? If the patient is complaining of what sounds typically as oropharyngeal transfer dysphagia, we should start with a video esophagram. If you find that you were wrong and there is a structural abnormality, then you refer the patient to ENT. If, on the other hand, it’s a neuromuscular disorder, a physiologic one, then you obtain a neurologic evaluation and at the end of the day, your speech/swallowing therapies is going to be the most helpful in treating the disorder.
If on the other hand, it’s an esophageal dysphagia, and it’s solids alone, you proceed to endoscopy. If it’s solids and liquids, obtain an esophagram, it’s normal, proceed to manometry. It’s abnormal, it’s a structural disorder, you go to EGD. If it’s neuromuscular, you proceed to manometry.
Dysphagia

- Deserves attention at any age
- X-ray and EGD reliable and effective in most cases
- Carefully obtained history essential

So, my take home message is that dysphagia deserves attention at any age, x-ray and EGD reliably and effectively predict most cases, and a carefully obtained history is absolutely essential. Thank you.
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