

PRACTICE GUIDELINES

Diagnosis and Management of Achalasia

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PREAMBLE

Guidelines for clinical practice are intended to suggest preferable approaches to particular medical problems as established by interpretation and collation of scientifically valid research, derived from extensive review of the published literature. When data are not available that will withstand objective scrutiny, a recommendation may be made based on a consensus of experts. Guidelines are intended to apply to the clinical situation for all physicians without regard to specialty. Guidelines are intended to be flexible, not necessarily indicating the only acceptable approach, and should be distinguished from standards of care that are inflexible and rarely violated. Given the wide range of choices in any health care problem, the physician should select the course best suited to the individual patient and the clinical situation presented. These guidelines are developed under the auspices of the American College of Gastroenterology and its Practice Parameters Committee. These guidelines are also approved by the governing boards of the American Gastroenterological Association and the American Society for Gastrointestinal Endoscopy. Expert opinion is solicited from the outset for the document. Guidelines are reviewed in depth by the committee, with participation from experienced clinicians and others in related fields. The final recommendations are based on the data available at the time of the production of the document and may be updated with pertinent scientific developments at a later time. The following guidelines are intended for adults and not for pediatric patients.

DEFINITION

Achalasia is a primary esophageal motor disorder of unknown etiology characterized manometrically by insufficient lower esophageal sphincter (LES) relaxation and loss of esophageal peristalsis, and radiographically by aperistalsis, esophageal dilation, minimal LES opening with a "bird-beak" appearance, and poor esophageal emptying of barium.

Achalasia is a well-recognized primary esophageal motor disorder of unknown etiology. Available data suggest he-

reditary, degenerative, autoimmune, and infectious factors as possible causes for achalasia, the latter two being the most commonly accepted possible etiologies (1, 2). Pathological changes found at autopsy or from myotomy specimens are in the esophageal myenteric (Auerbach's) plexus with a prominent but patchy inflammatory response consisting of T-lymphocytes and variable numbers of eosinophils and mast cells, loss of ganglion cells, and some degree of myenteric neural fibrosis (3). The end result of these inflammatory changes is the selective loss of postganglionic inhibitory neurons, which contain both nitric oxide and vasoactive intestinal polypeptide. The postganglionic cholinergic neurons of the myenteric plexus are spared leading to unopposed cholinergic stimulation (4). This produces high basal LES pressures, and the loss of inhibitory input results in insufficient LES relaxation. Aperistalsis is related to the loss of the latency gradient along the esophageal body—a process mediated by nitric oxide.

DIAGNOSIS

The diagnosis of achalasia should be suspected in anyone complaining of dysphagia for solids and liquids with regurgitation of food and saliva. The clinical suspicion should be confirmed by a barium esophagram showing smooth tapering of the lower esophagus leading to the closed LES, resembling a "bird's beak." Esophageal manometry establishes the diagnosis showing esophageal aperistalsis and insufficient LES relaxation. All patients should undergo upper endoscopy to exclude pseudoachalasia arising from a tumor at the gastroesophageal junction.

Most achalasia patients are symptomatic for years before seeking medical attention. The most common symptoms are dysphagia for solids and liquids, regurgitation, and chest pain. Although the dysphagia may initially be for solids only, as many as 70–97% of patients with achalasia have dysphagia for both solids and liquids at presentation (2). This contrasts with patients having strictures or ring whose dysphagia is limited to solids. Achalasia patients localize their dysphagia to the cervical or xiphoid areas. Over the years, patients learn to accommodate to their problem by using various maneuvers, including lifting the neck or drinking carbonated beverages to help empty the esophagus.

* See Appendix for list of members.

Regurgitation becomes a problem with progression of the disease, especially when the esophagus begins to dilate. Regurgitation of bland, undigested, retained food, or accumulated saliva occurs in about 75% of achalasia patients (5). It occurs most commonly in the recumbent position, awakening the patient from sleep because of coughing and choking. Chest pain or discomfort, located in the xiphoid area, is experienced by nearly 40% of patients with achalasia (5). It may mimic angina by location and character, but differs in not being aggravated by exercise or relieved by rest. About 60% of achalasia patients may have some degree of weight loss at presentation because of poor esophageal emptying and decreased or modified food intake (6). However, weight loss is usually minimal, and some patients are obese. Surprisingly, heartburn is reported by nearly 40% of achalasia patients. However, it is not related to the reflux of acidic gastric contents, but most likely to production of lactic acid from retained food or exogenous ingested acidic materials such as carbonated drinks.

When the diagnosis of achalasia is suspected, a barium esophagram with fluoroscopy is the single best diagnostic study. This test will reveal loss of primary peristalsis in the distal two-thirds of the esophagus with to-and-fro movement in the supine position. In the upright position, there will be poor emptying with retained food and saliva often producing a heterogeneous air-fluid level at the top of the barium column. Early in the disease, the esophagus may be minimally dilated, but more chronic disease is associated with sigmoid-like tortuosity and sometimes massive dilation of the esophageal body. There is a smooth tapering of the lower esophagus leading to the closed LES, resembling a "bird's beak." When the esophagus is minimally dilated, this may be misinterpreted as a peptic stricture. The presence of an epiphrenic diverticulum suggests the diagnosis of achalasia (7). Hiatal hernias are infrequent findings in patients with achalasia with reported prevalence of 1–14% compared with 20–50% found in the general population (8). The presence of a hiatal hernia on barium esophagram may make the diagnosis of achalasia less likely, but it does not rule it out, and does not change the management of these patients.

Esophageal manometry is the key test for establishing the diagnosis of achalasia (9) (Table 1). Because achalasia involves the smooth muscle portion of the esophagus, the manometric abnormalities are always confined to the distal two-thirds of the esophagus. In the body of the esophagus, aperistalsis is always present. This means that all wet or dry swallows are followed by simultaneous contractions that are classically identical to each other (isobaric or mirror images). The contraction amplitudes are typically low (10–40 mm Hg) and may be repetitive (2). The term "vigorous achalasia" is sometimes used when there is aperistalsis with normal or even high amplitude contractions in the esophageal body (10). Patients with vigorous achalasia usually have normal esophageal diameter on barium esophagram, but otherwise do not differ from patients with classic achalasia.

Table 1. Radiographic and Manometric Features of Achalasia

Barium esophagram	
Essential features:	<ul style="list-style-type: none"> ● "bird's beak" appearance of the LES with incomplete opening ● loss of primary peristalsis ● delayed esophageal emptying
Supportive features:	<ul style="list-style-type: none"> ● dilated or sigmoid-like esophagus ● epiphrenic diverticula
Manometry	
Essential features:	<ul style="list-style-type: none"> ● aperistalsis in distal 2/3 of the esophagus ● abnormal LES relaxation
Supportive features:	<ul style="list-style-type: none"> ● hypertensive LES pressure ● low amplitude esophageal contractions

Some manometric abnormality of the LES is always present in patients with achalasia. The LES pressure is usually elevated but may be normal (10–45 mm Hg) in up to 45% of patients; however, a low LES pressure is never seen in patients with untreated achalasia. Abnormal LES relaxation is seen in all achalasia patients. About 70–80% of patients with achalasia have absent or incomplete LES relaxation with wet swallows. In the remaining 20–30%, the relaxations are complete to the gastric baseline but are of short duration (usually <6 s) and functionally inadequate as assessed by barium and nuclear emptying studies (11).

Pseudoachalasia results from a tumor at the esophago-gastric junction or in an adjacent area. These patients mimic classic achalasia clinically and manometrically. The diagnosis should be suspected in patients with advanced age, shorter duration of symptoms, and marked weight loss (5, 12). However, the predictive accuracy of this triad of symptoms and signs is only 18% (13), possibly related to the low prevalence of the disease. Although the gastric cardia may be assessed radiographically, its sensitivity is poor in detecting tumors of the gastroesophageal junction causing pseudoachalasia. Therefore, all patients with suspected achalasia should undergo upper gastrointestinal endoscopy with close examination of the cardia and gastroesophageal junction. At endoscopy, the esophageal body usually appears dilated, atonic, and often tortuous with normal appearing mucosa. Sometimes, the mucosa is reddened, friable, thickened, or even superficially ulcerated secondary to chronic stasis, pills, or *Candida* esophagitis. Retained secretions, usually saliva, liquids, or sometimes food debris may be encountered. Patients with a markedly dilated esophagus may need esophageal lavage or a clear liquid diet for several days before endoscopy to avoid aspiration and to allow adequate visualization of the esophagus. The LES region usually has a "rosette" appearance and remains closed with air insufflation; however, the endoscope will easily traverse this area with gentle pressure allowing examination of the stomach. If excess pressure is required, the presence of pseudoachalasia should be highly suspected, the gastro-

esophageal junction and cardia closely examined, and biopsies taken. Tumors of the gastroesophageal junction may be missed endoscopically in up to 60% of patients with pseudoachalasia (5, 14). Endoscopic ultrasonography may prove useful in patients with a nondiagnostic endoscopy and a high degree of clinical suspicion for pseudoachalasia, but it is not recommended as a routine test in achalasia (15). The role of computed tomography scans is limited in the diagnosis of pseudoachalasia (14).

THE MANAGEMENT OF PATIENTS WITH ACHALASIA

Although there is no cure for achalasia, the goal of treatment should be relief of patient symptoms and improved esophageal emptying. The two most effective treatment options are graded pneumatic dilation and surgical myotomy. For patients who are at high risk for pneumatic dilation or surgery, endoscopic injection of the LES with botulinum toxin or pharmacological treatment with nitrates or calcium channel blockers may be acceptable alternatives.

No treatment can restore the muscular activity to the denervated achalasiac esophagus. Esophageal aperistalsis and impaired LES relaxation are rarely, if ever, reversed by any mode of therapy. Therefore, all the current treatment options for achalasia are limited to reducing the pressure gradient across the LES, thus facilitating esophageal emptying by gravity. This can be accomplished most effectively by pneumatic dilation and surgical myotomy or less effectively by pharmacological agents injected endoscopically into the LES (botulinum toxin) or taken orally (calcium channel blockers and nitrates).

Pneumatic dilation is the most effective nonsurgical treatment option for patients with achalasia. All patients considered for pneumatic dilation should be surgical candidates, since esophageal perforation may result from the procedure. Pneumatic dilation uses air pressure to intraluminally dilate and disrupt the circular muscle fibers of the LES. A variety of dilators were used in the past to treat patients with achalasia, including the Rider-Moeller, Sippy, Mosher, and Brown-McHardy dilators (1). Today, the most commonly used achalasia balloon dilators in the United States are the nonradiopaque graded size polyethylene balloons (Microvasive Rigiflex dilators). A less frequently used balloon is the over-the-endoscope Witzel dilator. Table 2 lists the recommended technique for performing pneumatic dilation using these graded balloons. Pneumatic dilation should always be carried out with sedation and under fluoroscopy. These dilators come in three different balloon diameters (3, 3.5, and 4 cm), and are positioned over a guidewire usually placed at endoscopy. The most important aspect of an effective pneumatic dilation is accurate positioning of the balloon across the LES and effective obliteration of the balloon waist visualized under fluoroscopy. The effectiveness of dilation does not depend on balloon distention time so long as the balloon waist is appropriately positioned and

Table 2. Recommended Technique for Pneumatic Dilation Using the Graded Balloons*

1. Fasting for at least 12 h before procedure.
2. Esophageal lavage with a large-bore tube (if needed).
3. Sedation and endoscopy in left lateral position.
4. Guidewire positioned in stomach and balloon passed over the guidewire.
5. Initial dilation with 3-cm diameter balloon; subsequent progression to 3.5-cm and 4-cm balloons may be required at separate sessions.
6. Accurate placement of balloon across gastroesophageal junction fluoroscopically.
7. Balloon distention to obliterate the waist, which usually requires 7–10 psi (this is the key to a successful dilation).
8. Gastrograffin study followed by barium swallow to exclude esophageal perforation.
9. Observation for 4 h for chest pain and fever.
10. Discharge with follow-up in 1 mo.

* Before proceeding with pneumatic dilation, it is important to ensure that a cardiothoracic surgeon is available in case of an esophageal perforation.

fully distended (17). After pneumatic dilation, all patients should undergo a gastrograffin study followed by barium swallow to exclude esophageal perforation (18). This procedure is usually performed as an outpatient with patients observed postprocedure for 4–6 h for chest pain and fever.

Studies to date indicate that by using the graded dilators, good-to-excellent relief of symptoms occurs in 50–93% of patients (Table 3) (1). The clinical response improves in a graded fashion with increasing size of the balloon diameter. Cumulatively, dilation with 3-, 3.5-, and 4-cm balloon diameters results in good-to-excellent symptomatic relief in 74%, 86%, and 90% of 359 treated patients, respectively (19–31) with an average follow-up of 1.6 yr (range 0.1–6 yr). Additionally, studies show that the rate of perforation may be lower with the serial balloon dilation approach (25); therefore, most experts start with the smallest, 3-cm balloon, except in patients who have had prior pneumatic dilations. The need for further dilation is based upon the persistence of symptoms usually assessed 4 weeks postprocedure or the recurrence of symptoms overtime.

Overall, studies find a 2% cumulative perforation rate using the graded balloons, although some centers report higher perforation rates (1). Patients with prompt recognition of perforation and surgical repair have comparable outcomes to those undergoing elective surgery (16); however, surgery for perforation is via an open thoracotomy approach. It is important to note that the rate of perforation is variable and highly dependent on the skill of the endoscopist. Physicians who do not perform pneumatic dilations on a regular basis should consider referral to specialized centers with expertise in performing this procedure. Other less prevalent complications of pneumatic dilation include gastroesophageal reflux (0–9%), aspiration pneumonia, gastrointestinal hemorrhage, and esophageal hematoma (32). Patients with a dilated and tortuous esophagus, esophageal diverticula, or previous surgery at the gastroesophageal junction may be at an increased risk for esophageal perforation.

Table 3. Cumulative Effectiveness of the Graded Pneumatic Dilators in Achalasia

Reference	Number of Patients	Study Design	Dilator (Size/cm)	Objective Assessments	% Sx Improvement	Follow-up (yr) Mean (Range)	Perforation (%)
				% ↓ LES Pressure	Excellent/Good		
Cox (19)	7	Prospective	3		86	0.8 (0.5–1)	0
Gelfand (20)	24	Prospective	3, 4	60, 68	70, 93		0
Barkin (21)	50	Prospective	3.5		90	1.3 (0.1–3.4)	0
Stark (22)	10	Prospective	3.5		74	0.5	0
Makela (23)	17	Retrospective	3, 3.5, 4		50, 75, 75	0.5	5.9
Levine (24)	62	Retrospective	3, 3.5		85, 88		0
Kadokia (25)	29	Prospective	3, 3.5, 4	67	62, 79, 93	4 (0.3–6)	0
Kim (26)	14	Prospective	3, 3.5	39	75	0.3	
Lee (27)	28	Prospective	3, 3.5, 4				7
Abid (28)	36	Retrospective	3.5, 4		88, 89	2.3 (1–4)	6.6
Wehrmann (29)	40	Retrospective	3, 3.5	42	89	2–5	2.5
Lambroza (30)	27	Retrospective	3		67	1.8 (0.1–4.8)	0
Bhatnagar (31)	15	Prospective	3, 3.5		73, 93	1.2 (0.3–3)	0
Total	359			size 3	125/168 = 74%	1.6 (0.1–6) yr	7/345 = 2%
				size 3.5	184/214 = 86%		
				size 4	90/100 = 90%		

ration with balloon dilation and should be considered for surgical myotomy as the first treatment option.

Traditionally, symptom improvement is used to assess the success of pneumatic dilation. However, a recent study suggests that subjective and objective parameters of improvement are discordant in about 30% of patients post-pneumatic dilation (33), suggesting that subjective improvement alone may give a false sense of success in those with less than optimal relief of their distal esophageal obstruction. Objective tests to better assess improvement after pneumatic dilation include manometry (LES pressure <10 mm Hg), esophageal scintigraphy, and the timed barium esophagram (34–36). The adjunctive use of these tests may help to improve the long-term success of pneumatic dilation, but this premise is still speculative.

Surgical myotomy for achalasia involves performing an anterior myotomy across the LES (Heller's myotomy) usually associated with an antireflux procedure (loose Nissen, incomplete Toupet, or Dor fundoplication). In the past, the myotomy was done by an open procedure through a thoracic or abdominal incision. The hospital stay was 7–10 days with a substantial postoperative recovery period. The abdominal approach limits extension of the myotomy proximally, with usually only a 1- to 2-cm distal myotomy onto the stomach to decrease the frequency of postprocedure gastroesophageal reflux. The transthoracic approach allows a longer proximal extension of the myotomy to the level of the major pulmonary vessels, but the extension distally onto the stomach may be limited. The results from published studies, using either the abdominal or thoracic approaches, show good-to-excellent symptom improvement in 83% of 2660 patients undergoing myotomy through the abdominal approach and in 83% of 1210 patients who had a transthoracic esophagomyotomy with a mean follow-up of ~7 yr (1). The main late complication of a Heller's myotomy is gastro-

esophageal reflux disease. The cumulative rates of heartburn and reflux disease reported in the studies are 22% for the abdominal and 10% for the transthoracic approach (1). The operative mortality for both procedures is very low (0.2% vs 1%), with most studies reporting no deaths directly related to the operation.

The advent of minimally invasive surgery and laparoscopic myotomy has resulted in shorter patient hospital stay (2 days), reduced morbidity, and quicker return to daily activity, making the procedure an attractive initial management option for healthy patients with achalasia. Studies show that laparoscopic cardiomyotomy has a cumulative good-to-excellent clinical response rate of 94% in 254 treated patients (Table 4) (37–48). However, long-term outcome of patients undergoing this procedure is unknown with current studies having a cumulative mean follow-up time of only 1 yr (range 0.1–4 yr). Before laparoscopic surgery, the most common indication for myotomy was the patient with recurrent symptoms after graded pneumatic dilations. However, laparoscopic surgery is increasingly performed as initial therapy for healthy patients, if a skillful surgeon is available. The cumulative rate of heartburn and reflux disease after laparoscopic myotomy is approximately 11% (1). Patients with megaesophagus (esophageal diameter >8 cm) or those with low LES pressure and persistent symptoms typically do not do well with either pneumatic dilation or surgical myotomy and may require an esophagectomy with a gastric pullup or colon interposition.

Endoscopic injection of botulinum toxin, type A, into the LES is the most recent treatment alternative for achalasia. Botulinum toxin acts by inhibiting the calcium-dependent release of acetylcholine from nerve terminals, thereby counterbalancing the effect of the selective loss of inhibitory neurotransmitters in achalasia (49, 50). It is commercially supplied as lyophilized powder (Oculinum; Allergan, Irwin, CA) in vials containing 100 units each. The powder must be

Table 4. Cumulative Effectiveness of the Laparoscopic Surgical Myotomy in Achalasia

Reference	Number of Patients	Study Design	Antireflux Procedure	Objective Assessments	% Sx Improvement	Follow-up (yr) Mean (Range)	% Complications	
				% ↓ LES Pressure	Excellent/Good		GERD	Mortality
Rosati (37)	25	Retrospective	yes	61	96	1 (1–2)		0
Ancona (38)	17	Retrospective	yes	72	100	0.7	6	0
Esposito (39)	8	Retrospective	yes		100	0.9 (0.8–1)		0
Raiser (40)	29	Prospective	yes		90	1.2 (1–2)	27	0
Morino (41)	18	Prospective	yes	60	100	0.7 (0.2–2)	6	0
Anselmino (42)	43	Prospective	yes	68	95	1 (0.3–4)	6	0
Delgado (43)	12	Prospective	yes	42	83	0.3 (0.1–1)	0	0
Slim (44)	8	Retrospective	yes	67	100	1		0
Bonovina (45)	33	Retrospective	yes	61	97	1 (0.3–2)		
Robertson (46)	9	Retrospective	no		88	1.1 (1–1.9)	13	0
Swanstrom (47)	12	Retrospective	yes	42	100	1.3	16	0
Hunter (48)	40	Retrospective	yes		90	1		0
Total	254			59%	240/254 = 94%	1 (0.1–4) yr	16/141 = 11%	0%

GERD = gastroesophageal reflux disease.

diluted with normal saline (5–10 ml) and used within 4 h of reconstitution without agitation of the solution because of the toxin's instability at room temperature. Botulinum toxin is injected endoscopically via a 5-mm sclerotherapy needle into the LES region as identified by a "puckered" appearance just above the gastroesophageal junction. Aliquots equaling 20–25 units of the toxin are injected into each of four quadrants for a total of 80–100 units.

Available data indicate that botulinum toxin is effective in relieving symptoms initially in about 85% of patients (1). However, symptoms recur in more than 50% of patients within 6 months possibly because of regeneration of the affected receptors (49). Older patients (>60 yr) and those with vigorous achalasia, defined as esophageal amplitude >40 mm Hg, are more likely to have a sustained response (up to 1.5 yr) to botulinum toxin injection (51). In those responding to the first injection, 76% will respond to a second botulinum toxin injection with decreasing response to further injections, usually from antibody formation to this foreign protein. Less than 20% of patients failing to respond to the first injection will respond to a second injection of botulinum toxin. Studies have shown that botulinum toxin is less effective than pneumatic dilation long term (52, 53). Additionally, some reports indicate that cardiomyotomy may be more difficult and less effective in patients who were previously treated with repeated botulinum toxin injections, possibly because of submucosal scar formation in the esophagus at the site of injection (54). Finally, the long-term safety of repeated injections of botulinum toxin in achalasia patients is unknown. Therefore, botulinum toxin injection should be reserved for elderly patients or patients who are at high surgical risk or refuse pneumatic dilation and surgical myotomy.

Calcium channel blockers and long-acting nitrates are effective in reducing LES pressure and temporally relieving dysphagia, but do not improve LES relaxation or improve

peristalsis. Both agents are used sublingually by opening the capsule and placing the contents under the tongue 15–45 min before meals with doses ranging from 10–30 mg for nifedipine and 5–20 mg for sublingual isosorbide dinitrate (55, 56). These drugs decrease LES pressure by approximately 50% with the long-acting nitrates having a shorter time to maximum effect (3–27 min) compared to sublingual nifedipine (30–120 min). Overall, calcium channel blockers improve patient symptoms by 0–75%, whereas sublingual nitrates result in symptom improvement in 53–87% of patients with achalasia (1). The clinical response to these pharmacological agents is short acting; they usually do not provide complete symptom relief, and efficacy decreases with time. Side effects such as headache, hypotension, and pedal edema are common limiting problems. Given these limitations, calcium channel blockers and nitrates are recommended only for patients who are very early in their disease with a nondilated esophagus, for symptomatic patients who are not candidates for pneumatic dilation or surgical myotomy, or for those who refuse invasive therapy and fail botulinum toxin injections.

A suggested treatment algorithm for patients with achalasia is shown in Figure 1. Symptomatic patients with achalasia who are good surgical candidates should be given the option of either graded pneumatic dilation or laparoscopic cardiomyotomy. The choice between the two procedures depends on institutional preference and experience. In patients unresponsive to graded pneumatic dilation, laparoscopic myotomy should be performed. In myotomy failures, repeat pneumatic dilation can be attempted. In patients who are poor candidates for surgery, initial treatment with botulinum toxin is currently the preferred approach. Nifedipine or isordil may prove to be beneficial in those unresponsive to botulinum toxin. Those with a megaesophagus (sigmoid esophagus and diameter >8 cm), or those with low LES

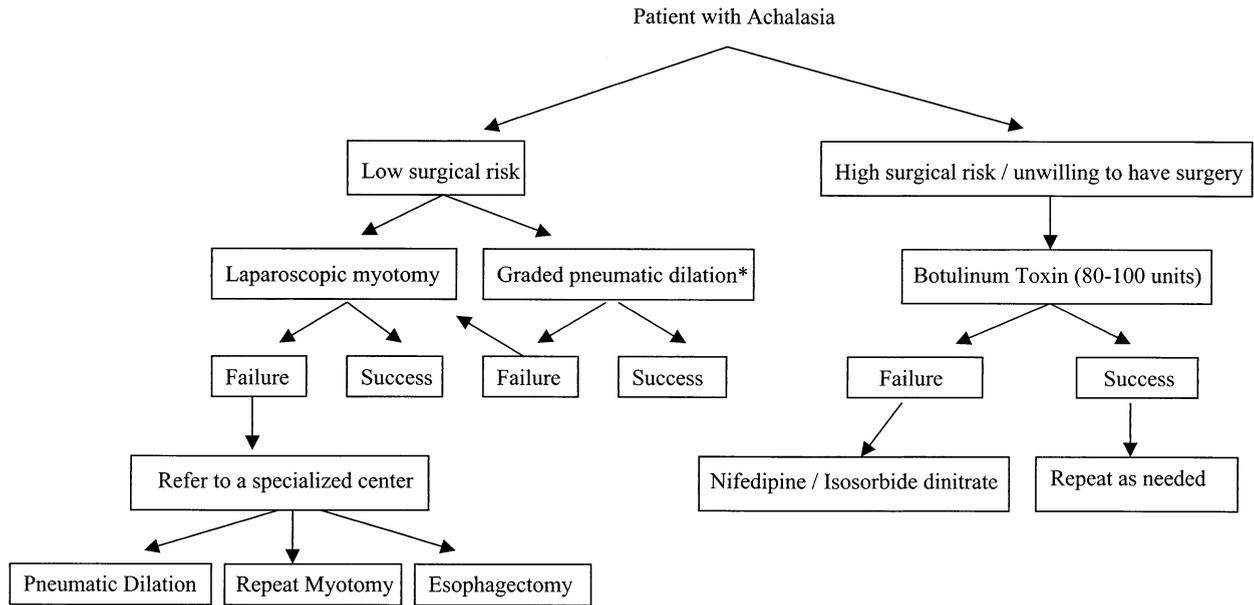


Figure 1. Suggested treatment algorithm for patients with achalasia.
 *Initial dilation with a 3-cm balloon followed by 3.5-cm and then 4-cm balloons in the nonresponders.

pressure with persistent symptoms may require esophagectomy.

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APPENDIX

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