

ACG Clinical Guideline: Diagnosis and Management of Achalasia

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Abstract

Achalasia is a primary motor disorder of the esophagus characterized by insufficient lower esophageal sphincter relaxation and loss of esophageal peristalsis. This results in patients complaints of dysphagia to solids and liquids, regurgitation, and occasional chest pain with or without weight loss. Endoscopic finding of retained saliva with puckered gastroesophageal junction or barium swallow showing dilated esophagus with birds beaking in a symptomatic patient should prompt appropriate diagnostic and therapeutic strategies. In this ACG guideline the authors present an evidence-based approach in patients with achalasia based on a comprehensive review of the pertinent evidence and examination of relevant published data.

Introduction

In the development of this ACG Guideline, the central themes examined included definition, diagnosis, and current therapeutic options for patients with achalasia by interaction between the authors of the technical review and the ACG Practice Parameters Committee. Recommendations were made based on a comprehensive review of the pertinent evidence and examination of quality and relevant published data in the literature.

A search of MEDLINE via PubMed was made using the terms “achalasia” and limited to “clinical trials” and “reviews” for years 1970–2012, and language restriction to English was made for preparation of this document. The resultant conclusions were based on the best available evidence or, in the absence of quality evidence, expert opinion. The GRADE (Grading of Recommendations Assessment, Development, and Evaluation) system was used to grade the quality of evidence and strength of recommendations (Table 1) (1). The quality of evidence ranges from “high” (further research is very unlikely to change our confidence in the estimate of effect) to “moderate” (further research is likely to have an important impact on our confidence in the estimate of effect and may change the estimate) to “low” (further research is very likely to have important impact on our confidence in the estimate of effect and is likely to change the estimate), and “very low” (any estimate of effect is very uncertain). The strength of a recommendation is graded as strong when the desirable effects of an intervention clearly outweigh the undesirable effects and is graded as weak when uncertainty exists regarding the tradeoffs (Table 1).

Table 1. The GRADE (Grading of Recommendations Assessment, Development, and Evaluation) system for grading evidence and strength of recommendations
<i>Strength of recommendations</i>
<i>Strong:</i> The desirable effects of an intervention clearly outweigh the undesirable effects or clearly do not.
<i>Weak:</i> The tradeoffs are less certain between the desirable and undesirable effects of an intervention.
<i>Quality of evidence</i>
<i>High:</i> Further research is very unlikely to change our confidence in the estimate of effect.
<i>Moderate:</i> Further research is likely to have an important impact on our confidence in the estimate of effect and may change the estimate.
<i>Low:</i> Further research is very likely to have an important impact on our confidence in the estimate of effect and is likely to change the estimate.
<i>Very low:</i> Any estimate of effect is very uncertain.

Definition and Epidemiology of Achalasia

Recommendation

1. Achalasia must be suspected in those with dysphagia to solids and liquids and in those with regurgitation unresponsive to an adequate trial of proton pump inhibitor (PPI) therapy (strong recommendation, low-quality evidence).

Diagnosis of Achalasia

Recommendations

1. All patients with suspected achalasia who do not have evidence of a mechanical obstruction on endoscopy or esophagram should undergo esophageal motility testing before a diagnosis of achalasia can be confirmed (strong recommendation, low-quality evidence).
2. The diagnosis of achalasia is supported by esophagram findings including dilation of the esophagus, a narrow esophagogastric junction with “bird-beak” appearance, aperistalsis, and poor emptying of barium (strong recommendation, moderate-quality evidence).
3. Barium esophagram is recommended to assess esophageal emptying and esophagogastric junction morphology in those with equivocal motility testing (strong recommendation, low-quality evidence).
4. Endoscopic assessment of the gastroesophageal junction and gastric cardia is recommended in all patients with achalasia to rule out pseudoachalasia (strong recommendation, moderate-quality evidence).

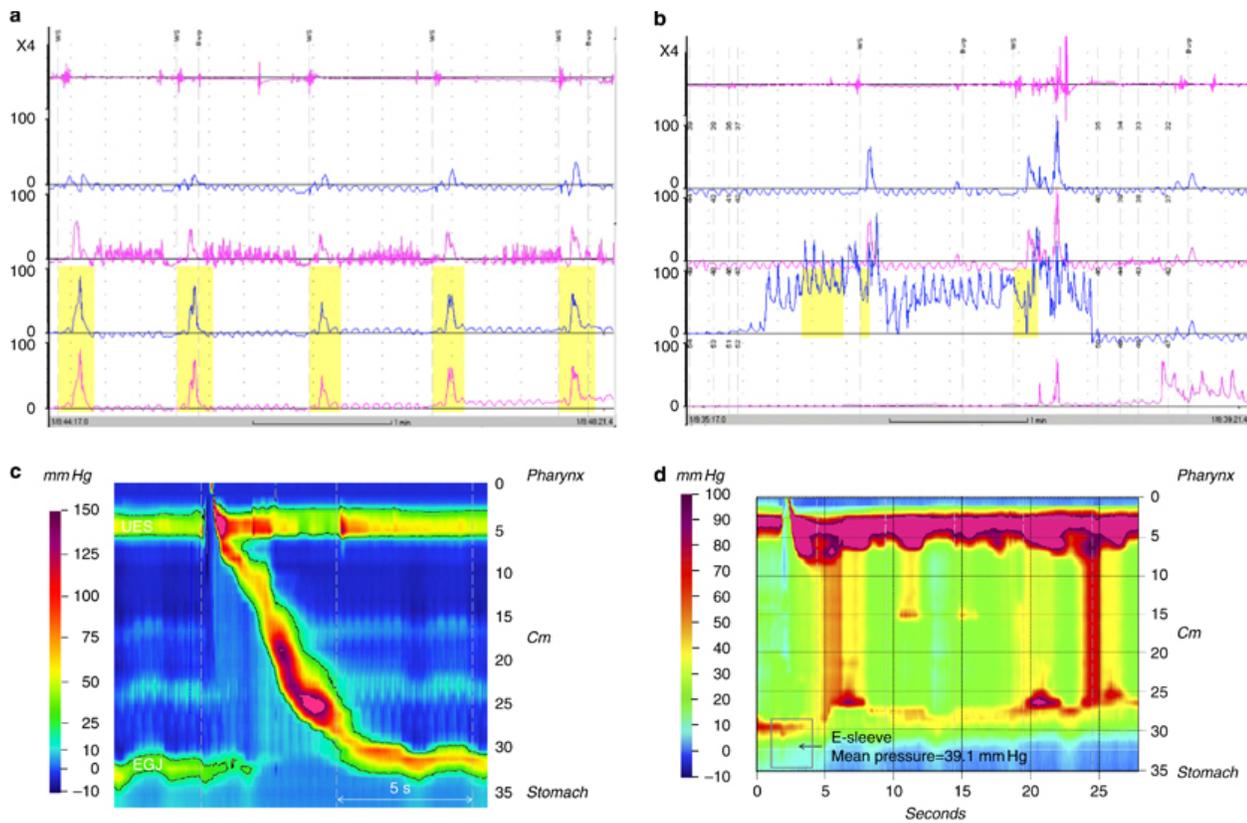


Figure 1. Manometric tracings of normal and achalasia. **(a)** Simultaneous esophageal contractions associated with high lower esophageal sphincter (LES) pressure and **(b)** incomplete relaxation noted during conventional water-perfused manometry. High-resolution manometry (HRM) tracings of **(c)** normal esophageal peristalsis and **(d)** achalasia showing simultaneous contractions along the esophagus with high E-sleeve LES pressure and incomplete relaxation. EGJ, esophagogastric junction; UES, upper esophageal sphincter.

Table 2. Comparison of manometric abnormalities in conventional and high-resolution manometry

Manometric features of achalasia	Conventional manometry Line tracing format	High-resolution manometry Esophageal pressure topography
<i>LES</i>		
	<p><i>Impaired LES relaxation^a</i></p> <ul style="list-style-type: none"> • Mean swallow induced fall in resting LES pressure to a nadir value of > 8 mm Hg above gastric pressure • Complete relaxation to gastric baseline with a short duration (< 6 s)^b <p><i>Basal pressure^b</i></p> <ul style="list-style-type: none"> • > 45 mm Hg 	<p><i>Impaired EGJ relaxation</i></p> <ul style="list-style-type: none"> • Mean 4 s IRP ≥ 10 mm Hg over test swallows^a
<i>Esophageal peristalsis</i>		
	<p><i>Aperistalsis in distal 2/3 of the esophagus</i></p> <ul style="list-style-type: none"> • No apparent contractions • Simultaneous contractions with amplitudes < 40 mm Hg 	<p><i>Aperistalsis</i></p> <ul style="list-style-type: none"> • Absent peristalsis (type I) • Pan-esophageal pressurization (type II)
<i>Atypical/variants</i>		
	<p><i>Vigorous</i></p> <ul style="list-style-type: none"> • Preserved peristalsis with esophageal contractions > 40 mm Hg • Simultaneous contractions > 40 mm Hg <ul style="list-style-type: none"> ○ Isobaric ○ Nonisobaric 	<ul style="list-style-type: none"> • Spastic achalasia (type III)

EGJ, esophagogastric junction; IRP, integrated relaxation pressure; LES, lower esophageal sphincter.

^a Required for diagnosis.

^b Supportive for the diagnosis.

Treating Achalasia

Recommendations

1. Either graded pneumatic dilation (PD) or laparoscopic surgical myotomy with a partial fundoplication are recommended as initial therapy for the treatment of achalasia in those fit and willing to undergo surgery (strong recommendation, moderate-quality evidence).
2. PD and surgical myotomy should be performed in high-volume centers of excellence (strong recommendation, low-quality evidence).
3. The choice of initial therapy should be guided by patients' age, gender, preference, and local institutional expertise (weak recommendation, low-quality evidence).
4. Botulinum toxin therapy is recommended in patients who are not good candidates for more definitive therapy with PD or surgical myotomy (strong recommendation, moderate-quality evidence).
5. Pharmacologic therapy for achalasia is recommended for patients who are unwilling or cannot undergo definitive treatment with either PD or surgical myotomy and have failed botulinum toxin therapy (strong recommendation, low-quality evidence).

Patient Follow-up

Recommendations

1. Patient follow-up after therapy may include assessment of both symptom relief and esophageal emptying by barium esophagram (strong recommendation, low-quality evidence).
2. Surveillance endoscopy for esophageal cancer is not recommended (strong recommendation, low-quality evidence).

Treatment Algorithm

A reasonable tailored treatment algorithm for patients with achalasia is outlined in Figure 2. Symptomatic patients with achalasia who are good surgical candidates should be offered information about the risks and benefits of the two equally effective treatment options of PD and surgical myotomy. The choice between the procedures should depend on patient preference and institutional expertise. However, to maximize patient outcome, both procedures should be performed in centers of excellence with adequate volume and expertise. PD should be performed in a graded manner, starting with the smallest balloon (3.0 cm), except in younger males (<45 years old) who may benefit with the initial balloon size of 3.5 cm or surgical myotomy. In patients unresponsive to PD, surgical myotomy should be performed. Repeat dilation can be performed in patients with recurrent symptoms after surgical myotomy. Poor surgical candidates should initially undergo injection of the LES with botulinum toxin and should be aware that repeat therapy is often needed. Other medical therapies with nitrates or calcium channel blockers may be offered if there is no clinical response to botulinum toxin injection. Esophagectomy may be needed in those with dilated esophagus (>8 cm) with poor response to an initial myotomy.

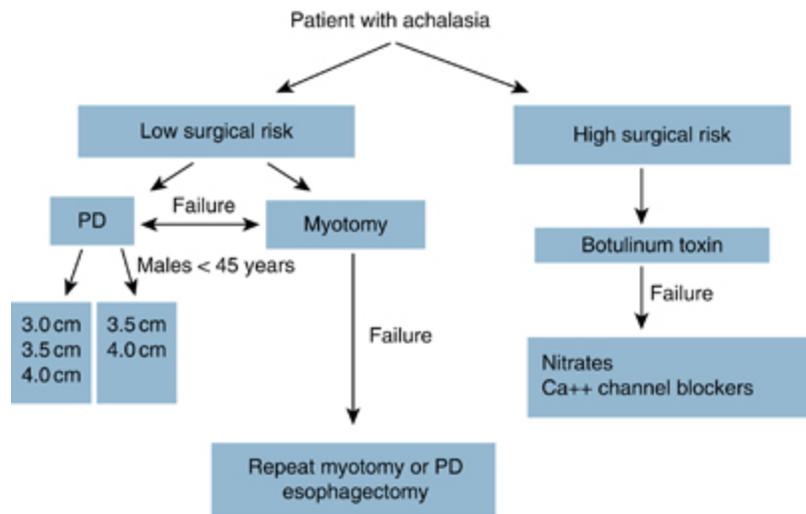


Figure 2. Recommended treatment algorithm for patients with achalasia. PD, pneumatic dilation.