



## Complete Summary

---

### GUIDELINE TITLE

Esophageal achalasia.

### BIBLIOGRAPHIC SOURCE(S)

Society for Surgery of the Alimentary Tract (SSAT). Esophageal achalasia. Manchester (MA): Society for Surgery of the Alimentary Tract (SSAT); 2003. 3 p.

### GUIDELINE STATUS

This is the current release of the guideline.

This guideline updates the previously issued version: Society for Surgery of the Alimentary Tract. Achalasia. Manchester (MA): Society for Surgery of the Alimentary Tract (SSAT); 1996-2000. 4 p.

## COMPLETE SUMMARY CONTENT

SCOPE  
METHODOLOGY - including Rating Scheme and Cost Analysis  
RECOMMENDATIONS  
EVIDENCE SUPPORTING THE RECOMMENDATIONS  
BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS  
QUALIFYING STATEMENTS  
IMPLEMENTATION OF THE GUIDELINE  
INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES  
IDENTIFYING INFORMATION AND AVAILABILITY  
DISCLAIMER

## SCOPE

### DISEASE/CONDITION(S)

Esophageal achalasia

### GUIDELINE CATEGORY

Diagnosis  
Risk Assessment  
Treatment

### CLINICAL SPECIALTY

Family Practice  
Gastroenterology  
Internal Medicine  
Surgery

## **INTENDED USERS**

Physicians

## **GUIDELINE OBJECTIVE(S)**

To guide primary care physicians to the appropriate utilization of surgical procedures on the alimentary tract or related organs

## **TARGET POPULATION**

Patients with symptomatic esophageal achalasia

## **INTERVENTIONS AND PRACTICES CONSIDERED**

### **Diagnosis**

1. Barium swallow
2. Endoscopy
3. Esophageal manometry
4. Prolonged pH monitoring
5. Endoscopic ultrasound
6. Computed tomography

### **Treatment**

1. Pneumatic dilatation
2. Intrasphincteric injection of botulinum toxin (Botox)
3. Laparoscopic Heller myotomy and partial fundoplication
4. Esophagectomy (reserved for failures after myotomy)
5. Surveillance endoscopy as follow-up

## **MAJOR OUTCOMES CONSIDERED**

- Symptom relief
- Symptom recurrence, gastroesophageal reflux following interventions
- Mortality rates associated with esophageal myotomy

## **METHODOLOGY**

### **METHODS USED TO COLLECT/SELECT EVIDENCE**

Searches of Electronic Databases

### **DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE**

Not stated

**NUMBER OF SOURCE DOCUMENTS**

Not stated

**METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE**

Not stated

**RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE**

Not applicable

**METHODS USED TO ANALYZE THE EVIDENCE**

Review

**DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE**

Not applicable

**METHODS USED TO FORMULATE THE RECOMMENDATIONS**

Expert Consensus

**DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS**

The Society for Surgery of the Alimentary Tract (SSAT) guidelines are based on statements and recommendations that were overwhelmingly supported by clinical evidence. Each represents a consensus of opinion and is considered a reasonable plan for a specific clinical condition.

(See companion document Gadacz TR, Traverso LW, Fried GM, Stabile B, Levine BA. Practice guidelines for patients with gastrointestinal surgical diseases. J Gastrointest Surg 1998;2:483-484.)

**RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS**

Not applicable

**COST ANALYSIS**

A formal cost analysis was not performed and published cost analyses were not reviewed.

**METHOD OF GUIDELINE VALIDATION**

## DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

The guidelines were reviewed by several committee members and then by the entire committee on several occasions. Each guideline was then sent back to the original author for final comment and reviewed again by the committee. Each guideline was approved by the Board of Trustees of the Society for Surgery of the Alimentary Tract and final comments were reviewed by the committee.

(See companion document Gadacz TR, Traverso LW, Fried GM, Stabile B, Levine BA. Practice guidelines for patients with gastrointestinal surgical diseases. J Gastrointest Surg 1998;2:483-484.)

## RECOMMENDATIONS

### MAJOR RECOMMENDATIONS

#### Diagnosis

In addition to careful symptomatic evaluation, the following tests should be routinely performed: Barium swallow usually shows narrowing at the level of the gastroesophageal junction ("bird beak") and various degrees of esophageal dilatation. Endoscopy is important to rule out the presence of a peptic stricture or cancer and gastroduodenal pathology. Esophageal manometry is the key test for establishing the diagnosis. The classic manometric findings are: (a) absence of esophageal peristalsis and (b) hypertensive or normotensive lower esophageal sphincter (LES) that fails to relax completely in response to swallowing.

Prolonged pH monitoring may be helpful preoperatively in patients who have previously failed treatment with pneumatic dilatation, Botulinum toxin (Botox), or surgical myotomy, for whom a myotomy is planned. Demonstration of reflux clearly indicates the need for a fundoplication in addition to the myotomy.

In patients older than 60 years of age with recent onset of dysphagia and excessive weight loss, secondary or pseudo-achalasia should be ruled out. Because a cancer of the gastroesophageal junction is the most common cause of pseudo-achalasia, an endoscopic ultrasound or a computed tomography (CT) scan of the gastroesophageal junction can help to establish the diagnosis.

#### Treatment

Treatment is palliative, and it is directed toward elimination of the outflow resistance at the level of the gastroesophageal junction. The following treatment modalities are available to achieve this goal:

Pneumatic dilatation has a success rate between 70 and 80%. Gastroesophageal reflux occurs after dilatation in 25 to 35% of patients. Up to 5% of patients may sustain a perforation at the time of a dilatation. These patients may require open surgery to close the perforation and perform a myotomy.

Intrasphincteric injection of botulinum toxin results in initial relief of symptoms in about 60% of patients, but this is transitory and symptoms will return in the majority of patients within a year. Subsequent injections are less effective and the benefit is of briefer duration. In addition, this treatment may cause an inflammatory reaction at the level of the gastroesophageal junction, which obliterates the anatomic planes. Consequently, a myotomy is more difficult, a mucosal perforation occurs more frequently, and the relief of dysphagia is less predictable. Because of these shortcomings, botulinum toxin should be reserved for elderly or high-risk patients who are poor candidates for dilatation or surgery.

Traditionally, pneumatic dilatation has been the first line of treatment for esophageal achalasia, while surgery was reserved for patients who had persistent dysphagia after multiple dilatations or who had suffered a perforation during dilatation.

Today, minimally invasive surgery has completely changed this treatment algorithm and a laparoscopic Heller myotomy and partial fundoplication is preferred by most gastroenterologists and surgeons as the primary treatment modality. Critical details of the operation include a generous myotomy of the lower esophagus, extending well onto the gastric wall. Because of the lack of esophageal peristalsis, a partial (Dor or Toupet), rather than a total fundoplication is frequently added to prevent reflux. Patients can usually eat the morning of the first postoperative day and can be discharged home after one or two days.

The need for esophagectomy for achalasia is very uncommon, even in the presence of a dilated esophagus, and should be reserved for failures after myotomy.

All patients undergoing treatment for achalasia should be followed by surveillance endoscopy, because they are at increased risk for development of both squamous and adenocarcinoma.

### **Expected Outcomes**

About 90% of patients have long-term relief of dysphagia after a myotomy, with a low incidence of symptomatic acid reflux. Patients should undergo 24-hour pH testing routinely after surgery, as reflux is often asymptomatic, and should be treated with proton pump inhibitors if abnormal acid reflux is present.

### **Qualifications for Performing Operations for Achalasia**

At a minimum, surgeons who are certified or eligible for certification by the American Board of Surgery, the Royal College of Physicians and Surgeons of Canada, or their equivalent should perform operations for achalasia. The qualifications of a surgeon performing any operative procedure should be based on training, experience, and outcomes.

### **CLINICAL ALGORITHM(S)**

None provided

## EVIDENCE SUPPORTING THE RECOMMENDATIONS

### TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of supporting evidence is not specifically stated for each recommendation.

## BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

### POTENTIAL BENEFITS

- *Pneumatic dilatation* has a success rate between 70 and 80%.
- *Intrasphincteric injection of botulinum toxin* results in initial relief of symptoms in about 60% patients, although relief is transitory and symptoms will return in the majority of patients within a year.
- About 90% of patients have long-term relief of dysphagia after a myotomy, with a low incidence of symptomatic acid reflux.

### POTENTIAL HARMS

- *Pneumatic dilatation*. Gastroesophageal reflux occurs after dilatation in 25 to 35% of patients. Up to 5% of patients may sustain a perforation at the time of a dilatation.
- *Intrasphincteric injection of botulinum toxin* results in initial relief of symptoms, but this relief is transitory and symptoms will return in the majority of patients within a year. Subsequent injections are less effective and the benefit is of briefer duration. In addition, this treatment may cause an inflammatory reaction at the level of the gastroesophageal junction, which obliterates the anatomic planes. Consequently, a myotomy is more difficult, a mucosal perforation occurs more frequently, and the relief of dysphagia is less predictable.
- Aspiration of retained food in the esophagus at the time of induction of anesthesia and perforation of the esophageal mucosa are the most common operative complications. Persistent or recurrent dysphagia occurs in 5 to 10% of patients.
- Up to 15% patients may experience gastroesophageal reflux after myotomy. The mortality rate after elective myotomy is less than 1%.

## QUALIFYING STATEMENTS

### QUALIFYING STATEMENTS

These guidelines have been written by the Patient Care Committee of the Society for Surgery of the Alimentary Tract (SSAT). Their goal is to guide primary care physicians to the appropriate utilization of surgical procedures on the alimentary tract or related organs and they are based on critical review of the literature and expert opinion. Both of the latter sources of information result in a consensus that is recorded in the form of these Guidelines. The consensus addresses the **range** of acceptable clinical practice and should not be construed as a standard of care. These Guidelines require periodic revision to ensure that clinicians utilize

procedures appropriately, but the reader must realize that clinical judgment may justify a course of action outside of the recommendations contained herein.

## IMPLEMENTATION OF THE GUIDELINE

### DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

## INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

### IOM CARE NEED

Getting Better  
Living with Illness

### IOM DOMAIN

Effectiveness

## IDENTIFYING INFORMATION AND AVAILABILITY

### BIBLIOGRAPHIC SOURCE(S)

Society for Surgery of the Alimentary Tract (SSAT). Esophageal achalasia. Manchester (MA): Society for Surgery of the Alimentary Tract (SSAT); 2003. 3 p.

### ADAPTATION

Not applicable: The guideline was not adapted from another source.

### DATE RELEASED

1996 (revised 2003 Feb 1)

### GUIDELINE DEVELOPER(S)

Society for Surgery of the Alimentary Tract, Inc - Medical Specialty Society

### SOURCE(S) OF FUNDING

Society of Surgery of the Alimentary Tract, Inc.

### GUIDELINE COMMITTEE

Patient Care Committee

## **COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE**

Not stated

## **FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST**

Not stated

## **GUIDELINE STATUS**

This is the current release of the guideline.

This guideline updates the previously issued version: Society for Surgery of the Alimentary Tract. Achalasia. Manchester (MA): Society for Surgery of the Alimentary Tract (SSAT); 1996-2000. 4 p.

## **GUIDELINE AVAILABILITY**

Electronic copies: Available from the [Society for Surgery of the Alimentary Tract, Inc. Web site](#).

Print copies: Available from the Society for Surgery of the Alimentary Tract, Inc., 900 Cummings Center, Suite 221-U, Beverly, MA 01915; Telephone: (978) 927-8330; Fax: (978) 524-0461.

## **AVAILABILITY OF COMPANION DOCUMENTS**

The following is available:

- Gadacz TR, Traverso LW, Fried GM, Stabile B, Levine BA. Practice guidelines for patients with gastrointestinal surgical diseases. J Gastrointest Surg 1998;2:483-484.

Electronic copies: Not available at this time.

Print copies: Available from the Society for Surgery of the Alimentary Tract, Inc., 900 Cummings Center, Suite 221-0, Beverly, MA 01915; Telephone: (978) 927-8330; Fax: (978) 524-8890.

## **PATIENT RESOURCES**

None available

## **NGC STATUS**

This summary was completed by ECRI on March 28, 2000. The information was verified by the guideline developer as of May 30, 2000. This summary was updated by ECRI on September 8, 2004.

## **COPYRIGHT STATEMENT**

For terms governing downloading, use, and reproduction of these guidelines, please contact: [ssat@prri.com](mailto:ssat@prri.com).

## DISCLAIMER

### NGC DISCLAIMER

The National Guideline Clearinghouse™ (NGC) does not develop, produce, approve, or endorse the guidelines represented on this site.

All guidelines summarized by NGC and hosted on our site are produced under the auspices of medical specialty societies, relevant professional associations, public or private organizations, other government agencies, health care organizations or plans, and similar entities.

Guidelines represented on the NGC Web site are submitted by guideline developers, and are screened solely to determine that they meet the NGC Inclusion Criteria which may be found at <http://www.guideline.gov/about/inclusion.aspx>.

NGC, AHRQ, and its contractor ECRI Institute make no warranties concerning the content or clinical efficacy or effectiveness of the clinical practice guidelines and related materials represented on this site. Moreover, the views and opinions of developers or authors of guidelines represented on this site do not necessarily state or reflect those of NGC, AHRQ, or its contractor ECRI Institute, and inclusion or hosting of guidelines in NGC may not be used for advertising or commercial endorsement purposes.

Readers with questions regarding guideline content are directed to contact the guideline developer.

© 1998-2008 National Guideline Clearinghouse

Date Modified: 9/15/2008

