International Journal of Medical Science and Health Research

Vol. 4, No. 05; 2020

ISSN: 2581-3366

Achalasia: Review and Assessment of Treatment Options.

Charles J. Haddad, M.D.
University of Florida Department of Community Health and Family Medicine, Jacksonville, Florida

Judella Haddad-Lacle', M.D.
University of Florida Department of Community Health and Family Medicine, Jacksonville. Florida.

Sofia Muenyi, M.D.
University of Florida Department of Community Health and Family Medicine, Jacksonville. Florida

Valery Muenyi, M.D.
University of Florida Department of Gastroenterology, Jacksonville, Florida.

Abstract
Achalasia is the failure of relaxation of the lower esophageal sphincter causing decrease or inability to pass ingested material from the esophagus to the stomach. It is thought to be caused from progressive degeneration of ganglion cells in the myenteric plexus in the esophageal wall. Achalasia can be associated with esophageal spasm or loss of peristalsis in the distal esophagus.

Symptoms of achalasia may include chest pain, vomiting, inability to pass ingested material, weight loss and heartburn. There are several options of treatment of achalasia, all of which attempt to decrease the resting pressure in the LES, decrease muscular spasm and to allow ingested material to pass into the stomach.

Diagnosis is made via esophageal manometry, esophagram, or Endoluminal functioning lumen imaging probe (EndoFlip). Current treatments of achalasia include pneumatic dilation, botulinum injections, Heller Myotomy, Per Oral Esophageal Myotomy (POEM). Medications to relax the LES may provide temporary symptom relief, but their effectiveness is variable.

Keywords: Achalasia, ganglion cells degeneration, lower esophageal spincter pressure, esophageal manometry.

Achalasia

Definition:
Achalasia is the failure of relaxation of the lower esophageal sphincter causing decrease or inability to pass ingested material from the esophagus to the stomach. It is thought to be caused from progressive degeneration of ganglion cells in the myenteric plexus in the esophageal wall. Achalasia is also called cardiospasm; esophageal a peristalsis or megaesophagus. There is an impairment of esophageal peristalsis and inability of the circular muscle and lower esophageal sphincter to relax during swallowing. Achalasia can be classified as primary when the cause is idiopathic or secondary, when the cause is a consequence of another systemic disease including

Vol. 4, No. 05; 2020

ISSN: 2581-3366

immunological disorders including lupus, hypothyroidism and diabetes.[1.] The disorder affects 1 in 100,000 individuals.[2]

Types of Achalasia:

There are three subtypes of achalasia based on high-resolution esophageal manometry studies. Type I: Classic, with minimal contractility in the esophageal body. Type II: with intermittent periods of paneshophageal pressurization. Type III: Spastic, with premature or spastic distal esophageal contraction.[3,4.] Symptoms are very similar between the three subtypes.

Achalasia Types	Manometry/contractility	symptoms	treatment
Type I	Minimal quiescent contractility	Vomiting more common. Weight loss	Pneumatic dilation (PD) Laparoscopic Heller myotomy (LHM) Per-oral endoscopic myotomy (POEM) Botulinum injection (BI) limited effect.
Type II	Intermittent periods or isobaric pressurization	Dysphagia Regurgitation	PD (first line) POEM/LHM (second line) BI (limited effect)
Type III	Premature or spastic simultaneous distal contractions with non-isobaric pressurization	Chest pain Dysphagia	POEM (first line) BI (limited effect)

Diagnosis

The esophageal dysmotility of achalasia ultimately leads to patient presenting with symptoms of dysphagia. The dysphagia is usually of solid and liquids without oropharyngeal transfer difficulties in 90% of patients, regurgitation in 75%, weight loss in 60%, chest pain in 50% and heartburn in 40% [5]. In patients presenting with achalasia type symptoms an upper endoscopy should be performed to exclude pseudoachalasia or other forms of esophageal gastric junction (EGJ) obstruction. Upper esophageal findings include a “puckered” EGJ (see photo 1), dilation of the esophagus as well as retain food and secretions, although these findings are nonspecific and the endoscopy can be normal especially in the early stages of the disease. A barium esophagram can show the classic “bird beak” appearance of the EGJ (see photo 2) with upstream esophageal dilation. High resolution manometry (HRM) is the test of choice for diagnosis of achalasia. The classic manometry finding include incomplete relaxation of the EGJ as well as the absence of organized peristalsis. Classic manometric findings include elevated Intergraded relaxation pressure (IRP) of >15mmHg in the presence of abnormal peristalsis or no peristalsis and is based on the Chicago classification. Three subtypes have been defined on HRM based on the Chicago classification: type I or classic achalasia with elevated IRP and low intraesophageal pressure, type II with panesophageal pressurization and type III with high-amplitude spastic

Vol. 4, No. 05; 2020

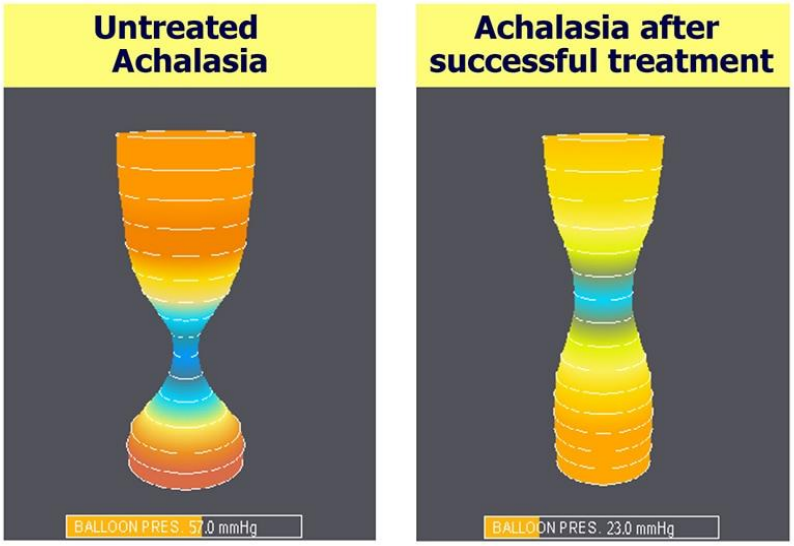
ISSN: 2581-3366

contractions [6] Endoluminal functioning lumen imaging probe (EndoFlip™) is a new technology that enables the assessment of the mechanical properties of the esophagus. EndoFlip uses a balloon mounted on a thin catheter placed transorally at the time of a sedated endoscopy. It provides real-time measurements of the pressure, dimensions and distensibility in the esophagus to enable identification of major motility disorders during the endoscopy. The technology uses impedance planimetry to estimate cross-sectional area. In achalasia patients, the EndoFLIP probe provides a dynamic assessment of distensibility of the esophagogastric junction (EGJ), which is valuable for evaluation and treatment. In patients in whom the diagnosis of achalasia is not clear by manometry, reduced distensibility of the EGJ appears to predict a good response to achalasia therapies. EGJ distensibility measurements performed during myotomy, either laparoscopic or endoscopic, can document whether distensibility has been appropriately increased by the treatment.

Table 1 The classification of achalasia defined by the Chicago Classification V3.0

Subtypes	Esophageal pressure topography features
Type I achalasia (classic achalasia)	Elevated median IRP (>15 mmHg*), 100% failed peristalsis (DCI <100 mmHg-s-cm)
Type I achalasia (with esophageal compression)	Elevated median IRP (>15 mmHg*), 100% failed peristalsis, panesophageal pressurization with ≥20% of swallows
Type III achalasia (spastic achalasia)	Elevated median IRP (>15 mmHg*), no normal peristalsis, premature (spastic) contractions with DCI >450 mmHg-s-cm with ≥20% of swallows
EGJ outflow obstruction	Elevated median IRP (>15 mmHg*), sufficient evidence of peristalsis such that criteria for types I–III achalasia are not met

*, cutoff value dependent on the manometric hardware. IRP, integrated relaxation pressure; DCI, distal contractile integral.



Treatment

Vol. 4, No. 05; 2020

ISSN: 2581-3366

The goal of treatment of achalasia is symptomatic relief. Unfortunately, 5% of patients will not improve and will need an esophagectomy [5]. Several studies have shown that treatment outcomes depend on the subtype and this can help guide the selection of treatment [7,8] Although studies suggest variability in the effectiveness of the various treatment modalities, Botulinum toxin, laparoscopic Heller myotomy (LHM), pneumatic dilation (PD) and perioral endoscopic myotomy (POEM) are the current options. Botulinum toxin injected in the lower esophageal sphincter (LES) reduces the LES pressure by inhibiting the release of acetylcholine from nerve endings [9]. It is considered fairly safe with rare side effects including mediastinitis and allergic reactions [5]. The main pitfall of this treatment is durability, which is limited to months. Currently there is limited evidence to support its use in patient less than fifty years old[10]. It should be reserved for patients who are not surgical candidates or as a bridge to more effective therapy. Repeat treatment is safe but less effective than initial treatment. The botulinum injections may cause scarring or fibrosis of the injection site potentially complicating future treatments such as POEM or surgical myomectomy.

Pneumatic dilation (PD) and LHM are both effective treatment options for achalasia PD works by disrupting the LES fibers through intraluminal dilation of a pressurized balloon thereby reducing the LES pressure and resistance of the bolus flow. It is effective as an initial treatment in terms of symptoms but effectiveness declines overtime and retreatment might be needed [11]. LHM is a surgical technique that disrupts the fibers of the LES. It has evolved from open surgery to minimally invasive with laparoscopic myotomy with partial fundoplication. Multiple studies have shown variable outcomes in terms of symptoms relief when comparing PD and LHM [11]. The international society for disease of the esophagus recommends graded pneumatic dilation as an effective treatment and LHM for patients wishing longer remission without further dilation.

The concept of natural orifice transluminal endoscopic surgery (NOTES) has inspired endoscopists and endoscopic surgeons to create less invasive treatment even for esophageal achalasia [12]. POEM is less invasive when compared to LHM with at least similar outcomes. The procedure works by endoscopically dissecting the circular muscles of the lower esophagus thereby reducing the LES pressure seen in achalasia (See photo 3). Available data suggest that LHM and POEM are both acceptable first line therapies in the management of achalasia patients, although the risk of GERD tends to be higher. POEM is typically performed in four consecutive steps: mucosal incision, creation of a submucosal tunnel, myotomy, and closure of mucosal incision.[13]

Contraindications for POEM include liver cirrhosis with portal hypertension and esophageal varices, coagulation or bleeding disorders, erosive esophagitis. Special caution should be used when considering treatment of patients with potential esophageal fibrosis caused from previous radiation, pneumatic endoscopic dilation or botox injections.

Vol. 4, No. 05; 2020

ISSN: 2581-3366

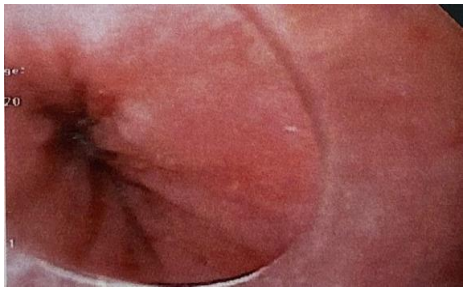
Treatments for Achalasia:

	Effectiveness	**Adverse effects**	**Comments**
Medications			
Isosorbide Mononitrate	Low	Headache, flushing, hypotension.	Short acting
Calcium channel blocker e.g. Nifedipine	Low	Hypotension, flushing, dizziness.	Short acting
Others: Sildenafil, atropine and theophylline.	Limited	variable	Data lacking
Botulinum toxin injection.	High	Scarring and fibrosis of LES	Injected via upper endoscopy into LES. Benefits lasts 6-12 weeks
Surgical			
Pneumatic dilation	Variable	Esophageal damage, bleeding.	Performed via upper endoscopy using inflatable balloon. Benefits last only weeks.
Heller myotomy	High	GERD and potential post-op complications.	Performed via laparoscopy disrupting the muscle fibers of the LES
P.O.E.M. (Per Oral Esophageal Myomectomy)	High	GERD, esophageal bleeding and rupture, mediastinitis.	Performed via upper endoscopy disrupting the muscle fibers of the LES

Vol. 4, No. 05; 2020

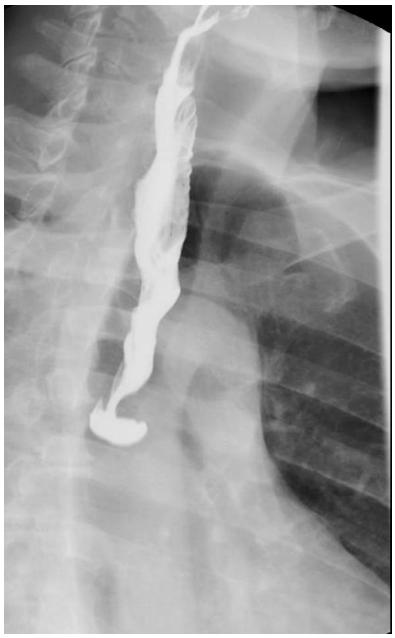
ISSN: 2581-3366

Photo 1



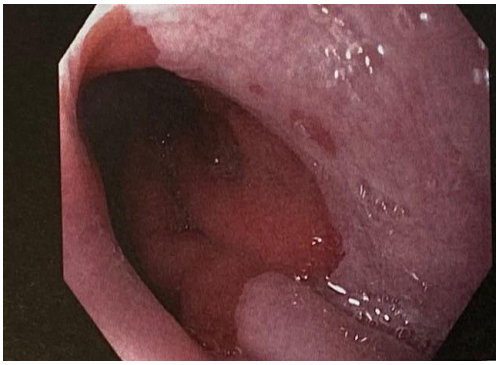
Showing “puckered appearance”. LES Pre POEM procedure

Photo 2



Showing Bird`s Beak on esophagram.

Photo 3



Showing open LES post POEM procedure

Vol. 4, No. 05; 2020

ISSN: 2581-3366

References:

1. Farias GFA, de Moura DTH, et al. Peroral endoscopic myotomy (POEM): a comparative study between Chagasic and idiopathic achalasia. Endosc Int Open.2020 Apr; 8(4): E506-E512.
2. Wilfong C, Ross S, et al. Doing more with less: our decade of experience with laparo-endoscopic single site Heller myotomy supports its application. Surg Endosc 2020Mar 16.
3. Patel D, Lappas B, Vaezi M. An overview of Achalasia and Its Subtypes.Gastroenterol Hepatol 2017; 13(7):411-421.
4. Meillier A, Midani D, et al. Difference of achalasia subtypes based on clinical symptoms, radiographic findings, and stasis scores.
5. Vela MF, Richter JE, Wachsberger D, et al. Complexities of managing achalasia at a tertiary referral center: use of pneumatic dilatation, Heller myotomy, and botulinum toxin injection. Am J Gastroenterol 2004; 99:1029-36.
6. Samo S, Carlson DA, Gregory DL, et al. Incidence and prevalence of achalasia in Central Chicago, 2004-2014, since the widespread use of high-resolution manometry. Clin Gastroenterol Hepatol 2017; 15: 366-73.
7. Salvador R, Costantini M, Zaninotto G, et al. The preoperative mano- metric pattern predicts the outcome of surgical treatment for esophageal achalasia. J Gastrointest Surg 2010; 14:1635-45.
8. Rohof WO, Salvador R, Annese V, et al. Outcomes of treatment for achalasia depend on manometric subtype. Gastroenterology 2013; 144:718-25; quiz e713-4.
9. Hoogerwerf WA, Pasricha PJ. Pharmacologic therapy in treating achalasia. Gastrointest Endosc Clin North Am 2001; 11:311-24.
10. Allescher H D, Storr M, Seige M et al. Treatment of achalasia: botulinum toxin injection vs. pneumatic balloon dilation. A prospective study with long-term follow-up. Endoscopy 2001; 33: 1007–17.
11. West R L, Hirsch D P, Bartelsman J F *et al.* Long term results of pneumatic dilation in achalasia followed for more than 5 years. Am J Gastroenterol 2002; 97: 1346–51.
12. Inoue H, Minami H, Kobayashi Y, et al. Peroral endoscopic myotomy (POEM) for esophageal achalasia. Endoscopy 2010; 42:265-71.
13. www.uptodate.com/contents/Overview of the treatment of achalasia