Achalasia: Current Treatment

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Historical Points

• Sir Thomas Willis
  – Founding Member of The Royal Society
  – 1672, described a patient who was ‘unable to swallow liquids’
  – Initial treatment = esophageal dilatation using a carved whalebone with a sponge attached to its tip.

• Ernest Heller
  – Performed the first esophagomyotomy (~1914)

• An approach via thoracotomy was popularized in the 1950s/1960s by Ellis

• Laparoscopic Heller Myotomy gained popularity in the early 1990s
What is Achalasia?

• Origin of the word is Greek
  – “kalasis” meaning ‘loosening’
  – “khalan” meaning ‘relax’

• Epidemiology
  – Rare, 0.5 -1/100,000
  – Peak incidence is between 20 and 50 years of age

• Etiology
  – Idiopathic
  – Autoimmune/Infectious (?)

• Second only to GERD as the most common functional disorder of the esophagus requiring surgical intervention
Cellular Pathophysiology

- Progressive neuronal degeneration / damage in the mesenteric plexus, which results in a nonrelaxing, hypertensive lower esophageal sphincter (LES) and aperistalsis of the body of the esophagus.
How do we diagnose achalasia?

• History & Physical Examination
  – Progressive Dysphagia to Solids and Liquids
  – Regurgitation of undigested food
  – Reflux
  – Aspiration/aspiration pneumonia
  – Weight loss (late finding)
  – Chest pain (atypical finding)

• Diagnostic Testing...
  – Barium Esophagogram
    • To look for the presence of the “Bird’s Beak” Esophagus
    • 95% of patients will have a positive test
  – Esophageal Manometry
    • To demonstrate esophageal aperistalsis and insufficient LES relaxation with swallowing
  – Esophagogastroduodenoscopy
    • To Rule out “Pseudoachalasia” from mass/tumor at the lower esophageal junction
Two Main Types of Achalasia

- Classic Achalasia (4 Classic Manometric Findings)
  - Hypertensive LES, present in ~50% of patients
  - Incomplete / absent relaxation in the LES
  - Esophageal aperistalsis
  - Elevated LES baseline pressure
  - Aperistaltic esophagus in the distal smooth muscle segment of the esophageal body

- Vigorous Achalasia
  - Aperistaltic esophagus, pressures less than 60 mm Hg produced in the body of the esophagus
  - Simultaneous contraction waves of variable amplitudes, consistent with preserved muscle function
Radiographic Findings – Bird’s Beak
SAGES guidelines for the surgical treatment of esophageal achalasia

Dimitrios Stefanidis · William Richardson · Timothy M. Farrell · Geoffrey P. Kohn · Vedra Augenstein · Robert D. Fanelli
Treatment Modalities

• There are 4 major treatment modalities
  – Pharmacotherapy
  – Botox Injections
  – Pneumatic Dilation
  – Surgical

• All treatment modalities are palliative
Pharmacotherapy

• Calcium channel blockers, oral nitrates
  – Nifedipine, isosorbide dinitrate (sublingual)
• Reduce LES Pressure (~50%), but do not improve LES relaxation
  – Improvement in 53-87% (isosorbide); 0-75% (nifedipine)
  – “Sporadic, unreliable” at best
• Adverse / Side Effects
  – Hypotension, headache

• SAGES Guidelines:
  – Limited role in the treatment
  – Should be used in very early stages of the disease, temporizing measures until more definitive treatments
  – Patients who fail or are not candidates for other treatment modalities
**Botox Injections**

- **Endoscopic Injections, 4 quadrant pattern in LES**
- **Effectiveness**
  - 85% initially, 50% at 6 months, 30% at one year
  - As effective as pneumatic dilation, limited long term
  - Best for patients with vigorous achalasia and generally lower LES pressures (<50% of upper limit of normal)

- **SAGES Guidelines**
  - Botulinum toxin can be administered safely, but its effectiveness is limited especially in the long term.
  - Reserved for poor candidates for other more effective treatment options such as surgery or dilation
Pneumatic Dilation

- Most effective non-operative treatment
  - Success in 55-70% with single dilation; >90% multiple
- Stretches and ruptures the LES muscle fibers
- One dilation is not enough – most need repeat treatment
- Adverse / Side Effects
  - Rate of perforation ~1% (0.67 – 5.6%)
  - Overall complication rate 11%
    - Perforation, GERD, intramural hematoma
- Of note, pneumatic dilation + stenting not recommended (increased morbidity and mortality)

- SAGES Guidelines:
  - Of non-operative treatment techniques endoscopic dilation is the most effective for dysphagia relief in patients with achalasia
  - Associated with the highest risk of complications
  - To be considered in selected patients who refuse surgery or are poor operative candidates
**Operative Management - Heller Myotomy**

- The goal of myotomy is the destruction of the nonrelaxing LES
- Effectiveness – resolution of symptoms in >85% of patients
- Long term success in predicted by
  - High LES pressure
  - No prior therapy
  - Short symptom duration
  - Absence of sigmoidal esophagus
- Failure may be attributable to either incompletely relieving the obstructive achalasia or an obstructing antireflux mechanism
Fundoplication

• The type of fundoplication is important in order to avoid significant obstruction.

• In general...
  – 360-degree fundoplication is not used with myotomy for achalasia because it will be too obstructive in this setting.
  – Posterior Toupet and the anterior Dor partial fundoplications have been used effectively.
  – Heller myotomy without fundoplication is also effective in management

• Richards et al. 2004 Annals of Surgery
  – Lap Heller Myotomy w/wo Dor Fundoplication
  – Lap Heller + Dor was superior (with regard to reflux symptoms after operative management)
Heller Myotomy

- Myotomy begins on the esophagus, 5-6 cm proximally from the GE junction
- Extends distally 1.5 to 2 cm onto the cardia
- The myotomy is guided by intraoperative endoscopy to ascertain that it is carried at least 1.5 to 2 cm beyond the squamo-columnar junction.
Nissen Fundoplication

- Complete 360 degree wrap; GERD
- Not used as part of a Heller Myotomy
Toupet Fundoplication

- 270 degree POSTERIOR wrap
- Proponents of the Toupet claim it provides a better anti-reflux mechanism when the patient is in the supine position.
- Opponents state that a Toupet may be too obstructing

**Technical Considerations:** Requires extensive mobilization of the fundus, ligation of the short gastric vessels, and posterior dissection including disruption of the phrenoesophageal ligament
Dor Fundoplication

- Most popular partial fundoplication that is part of a Heller myotomy
- ANTERIOR 180-200 degree partial fundoplication
- Proponents prefer the added security of covering the exposed mucosa with the fundus
- Avoidance of complete mobilization of the abdominal esophagus (cf. Toupet)
- Technical Considerations:
  - (1) Does not require extensive mobilization of the fundus
  - (2) Short gastric vessels are left intact
  - (3) Posterior dissection unnecessary, thus eliminating the disruption of the phrenoesophageal ligament.
Summary

• Medical Management is not very good (long-term).
• Surgical Management is preferred and most effective short- and long-term.
• Laparoscopy has obviated the need for thoracoscopic techniques/approaches – there are exceptions.
• Most popular intervention for achalasia is a laparoscopic Heller myotomy (with or without a Dor fundoplication).
References

• Cameron’s Current Surgical Therapy
• ACS Surgery, Minimally Invasive Esophageal Procedures
• **Images pulled from references above and the public domain (www.google.com)