Achalasia

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Kings County Hospital
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Achalasia

I. Anatomy
II. Historical Background
III. Achalasia
IV. Clinical presentation
V. Evaluation
VI. Treatment
Anatomy- esophagus

- Muscular tube - Conduit from the pharynx to the stomach

- Length is defined anatomically, from cricoid cartilage to the gastric orifice

- Distance from the incisor 40-45 cm (actual length: M 22-28 cm F 2 cm shorter)

- Passes behind aortic arch and left main bronchus.

- Enters abdomen through esophageal hiatus → 2-4 cm below the diaphragm
Anatomy - esophagus

Course of the esophagus:
- Neck and upper esophagus: left of midline
- Mid-esophagus: right of midline
- Lower esophagus: left of midline

Three areas of normal constrictions:
- Cricopharangeal
- Behind the aortic arch
- LES (thickening of the circular muscles ~ 4cm)
Anatomy - esophagus

- Fixed in position at two places:
  . Upper: firmly attached to the cricoid cartilage
  . Lower: Phreno-esophageal ligament to the esophagus which provides an air-tight seal between the thoracic and abdominal cavity.

(lack of fixation throughout its length allows both transverse and longitudinal mobility)
Vascular supply

**ARTERIAL SUPPLY**
- Upper → superior and inferior thyroid artery
- Middle → Bronchial arteries and esophageal branches directly from aorta
- Lower → L inferior phrenic and gastric

**VENOUS SUPPLY**
- Upper → esophageal venous plexus to azygos vein
- Lower → esophageal branches of the coronary vein, a tributary of the portal vein
Structure

- Consists of 3 layers: muscularis externa, submucosa, mucosa
Achalasia-historical note

- First described more than 300yrs ago
- Referred to as cardiospasm
- Thomas Willis (1621-1675)
  - Described a pt starving and unable to swallow
  - Conclusion was due to lower esophageal narrowing
  - Constructed the first dilator-made of whale bone and sponge
  - First successful treatment of achalasia
Achalasia-historical note

1914: Ernst Heller (1877-1964) -- First successful cardiomyotomy

- Anterior and posterior myotomies
- Extending 8cm or more into esophagus and stomach
Achalasia-historical note

- 1918: De Brune Groenveldt and Zaaijer – performed modified Heller myotomy
  - anterior only
  - Original technique was to excessive
Achalasia

- Uncommon (0.5-1 in 100,000)
- No sex predilection M=F
- Majority between ages 20-50s
- Ineffective relaxation of the LES combined with loss of esophageal peristalsis $\rightarrow$ impaired esophageal emptying and gradual dilatation
- Decrease or loss of myenteric ganglion cells
- Slight increase risk of esophageal carcinoma (approx. 10yrs earlier than the general population)
Achalasia - Presentation

- Dysphagia - delayed and progressive presentation (mean 2 years)
- Exacerbated by emotional stress or cold fluid
- 60-90% report spontaneous or forced regurgitation of undigested food
- 10% will have pulmonary complication
- Chest pain (≠ heartburn) - 30-50% resolves with Myotomy
Achalasia - Diagnosis

- **CXR**: air fluid levels
- **Barium swallow**: dilated esophagus with Bird's beak deformity. (pseudoachalasia from extrinsic mass may mimic the classic achalasia appearance)
- **Manometry**: gold standard
  - Elevated LES pressure (greater than 35mmHg)
  - Incomplete sphincter relaxation
  - Complete absence of peristalsis
- **Endoscopy**: dilated esophagus with tightly closed LES
  → gentle pressure will admit the scope with a "pop".
Achalasia
Achalasia
Achalasia - Treatment

Palliation of dysphagia is the key
→ relieve functional obstruction of distal esophagus

- pharmacotherapy
- botulinum toxin
- esophageal dilation
- operative myotomy
Achalasia- algorithm

Clinical symptoms
(dysphagia, regurgitation, weight loss)

Barium swallow

Manometry

Low-surgical-risk patient

Laparoscopic myotomy

Failure

Repeat myotomy or esophagectomy

Success

Pneumatic dilation

Failure

Repeat as needed

Success

Botox injection

No surgical-risk patient
(or refuses surgery)

High-surgical-risk patient

Laparoscopic myotomy

Failure

Repeat myotomy or esophagectomy

Success

Pneumatic dilation

Failure

Nifedipine or isosorbide

Success

Achalasia - Treatment

- **Pharmacotherapy:** (poorly absorbed and short lived, best reserved as adjunct to other therapies)
  - Nitrates
  - Ca++ channel blockers
  - Anticholinergics
  - Opioids
Botulinum Toxin Therapy

A

Dilated esophagus

B

Needle injects botulinum toxin into lower esophageal sphincter (stricture)

B

Lumen

Injection site (squamocolumnar junction)
Achalasia - Treatment

**Botox injection:**
- Bind to cholinergic nerves and irreversibly inhibit Acetyl Choline release
- 60-85% of patients get relief but 50% get recurrent symptoms within 6 months.
- Endoscopically injected
- For pt who are not candidates for other therapies
Achalasia - Treatment

Botox injection cont.

- **Advantages:** safety, ease of administration, minimal side effects
- **Disadvantages:** expensive, need for multiple injections, and efficacy decreased with repeated injection
- **Cause obliteration of the dissection planes between submucosa and muscular layer which will make subsequent surgery more difficult and increase risk of perforation.

Pneumatic Dilator
Achalasia - Treatment

Esophageal dilation (under fluoroscopy)

- Standard nonoperative therapy
- Break the muscle fibers
- For pts with limited life expectancy
- Can have repeated dilatation
- 60-80% success rate, 5yr recurrence rate 50%
- Efficacy is decreased after second dilatation
- Perforation rate ~ 2%
- PPI reduces the need for repeat dilatation

Csendes A: Late results of a prospective randomised study comparing forceful dilation  Gut 30:299, 1989
Esophageal myotomy

A: Esophageal dilation

B: Myotomy incision

Lower esophageal sphincter

Stomach

Pouting mucosa through myotomy
Achalasia – Surgical treatment

- Excellent results in 90-95%
- Gold standard
- 1914 - Ernest Heller - double myotomy
- Modified by Zaaijer - single myotomy
- World’s largest experience
  - Brazil, Chagas’ disease-endemic
    - 1 in 8 inhabitants, in which 5% develops achalasia
- Traditionally trans-thoracic or trans-abdominal
- Now minimally invasive Laparoscopic / Thoracoscopic
- Robotic Heller myotomy

Pinotti HW: Surgical complications of Chagas’s disease  World J surg 1991
Achalasia – Surgical treatment

- **Indications:**
  - Younger than 40yrs old (group which PD is <50% effective)
  - High risk of perforation
    - Esophageal diverticula
    - Previous surgery of GE junction
    - Tortuous or dilated distal esophagus
  - Recurrent symptoms despite Botox or PD therapy
  - Personal choice of therapy
    - Lower risk of perforation
    - Better long term outcome
    - Decrease chance of re-intervention
Achalasia – Surgical treatment

- Expose mucosal surface
- Length of myotomy
  - Cephalad: 1-2 cm beyond the dilated esophagus
  - Caudal: 1-2 cm into the gastric musculature or when transverse veins are encountered
- Check for perforation
  - Meythlene blue
  - Air

Robotic heller myotomy
Complications

- **Intra-op**
  - Mucosa perforation

- **Post-op:**
  - Dysphagia - adhesion, inadequate myotomy
  - GERD - long myotomy, nerve damage
  - Delay perforation - inadequate myotomy
Achalasia – Surgical treatment

- Which esophageal technique should be used?
- Any role for anti-reflux procedure?
Trans-thoracic

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<th>Design</th>
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Totals 1,379 86 ± 5 6.6 ± 4 10 ± 15 0.01 ± 0.05

- Excellent result
- Less GERD* compare to trans-abdominal
- * Phreno-esophageal ligament is not disrupted and shorter myotomy
- No fundoplication is necessary

Farshad Abir: surgical treatments of achalasia: current status and controversies. Digestive surgery 2004
Trans-abdominal

- Excellent result – comparable to trans-thoracic
- More GERD*, less dysphagia
- *Longer myotomy onto stomach (3cm)

Farshad Abir: surgical treatments of achalasia: current status and controversies. Digestive surgery 2004

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Totals 2,680

83 ± 15 6 ± 3.9 12.3 ± 9 1.2 ± 3

P = Prospective; R = retrospective; F/U = follow-up; compl = complications; mean ± SDEV.
Laparoscopic

- Excellent result
- *Decrease hospital stay (average 42-48hrs post-op)
- Improve GERD by antireflux procedure

Farshad Abir: surgical treatments of achalasia: current status and controversies. Digestive surgery 2004

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P = Prospective; R = retrospective; F/U = follow-up; compl = complications; mean ± SDEV.
Achalasia – Surgical treatment

- Currently, no prospective randomized trials comparing the various approaches to myotomy
- Excellent results
- Technique used should depend on individual surgeon’s comfort and experience
- Anti-reflux should be performed with abdominal approach