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Management of Achalasia

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Case Presentation

- 28 yo male p/w dysphagia & vomiting
- Diagnosed with GERD; +*H.pylori*
- Prescribed ABx & PPI, did not tolerate
- Unable to swallow solids & liquids
- Retrosternal burning sensation
- Feels like “food is stuck”

Clinical History

- Weight loss ~20lbs over 2 months
- 8 yr hx of tobacco smoking: 1PPD
- Upper endoscopy
  - Esophageal furrows
  - Erythematous stomach
  - Path: mid esophageal bx: no eosinophils

Physical Examination

- Afebrile, Vitals stable
- Thin, well-appearing young man in NAD
- S1/S2 RR, Equal breath sounds
- Abdomen soft, NT/ND

- CBC: 4.9>12.1/39.9<133
- BMP: 142/4.1/109/24/17/1.08<90

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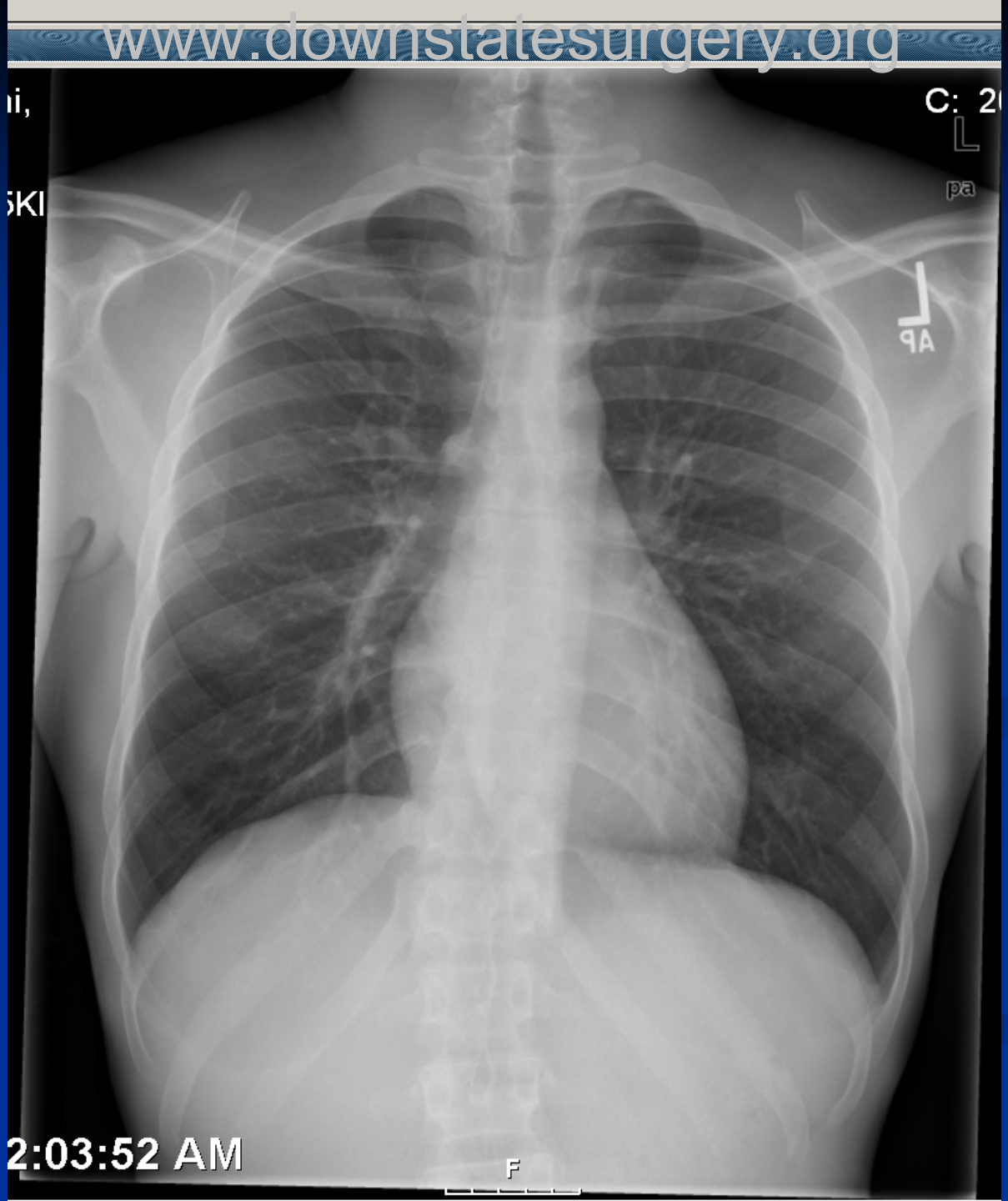

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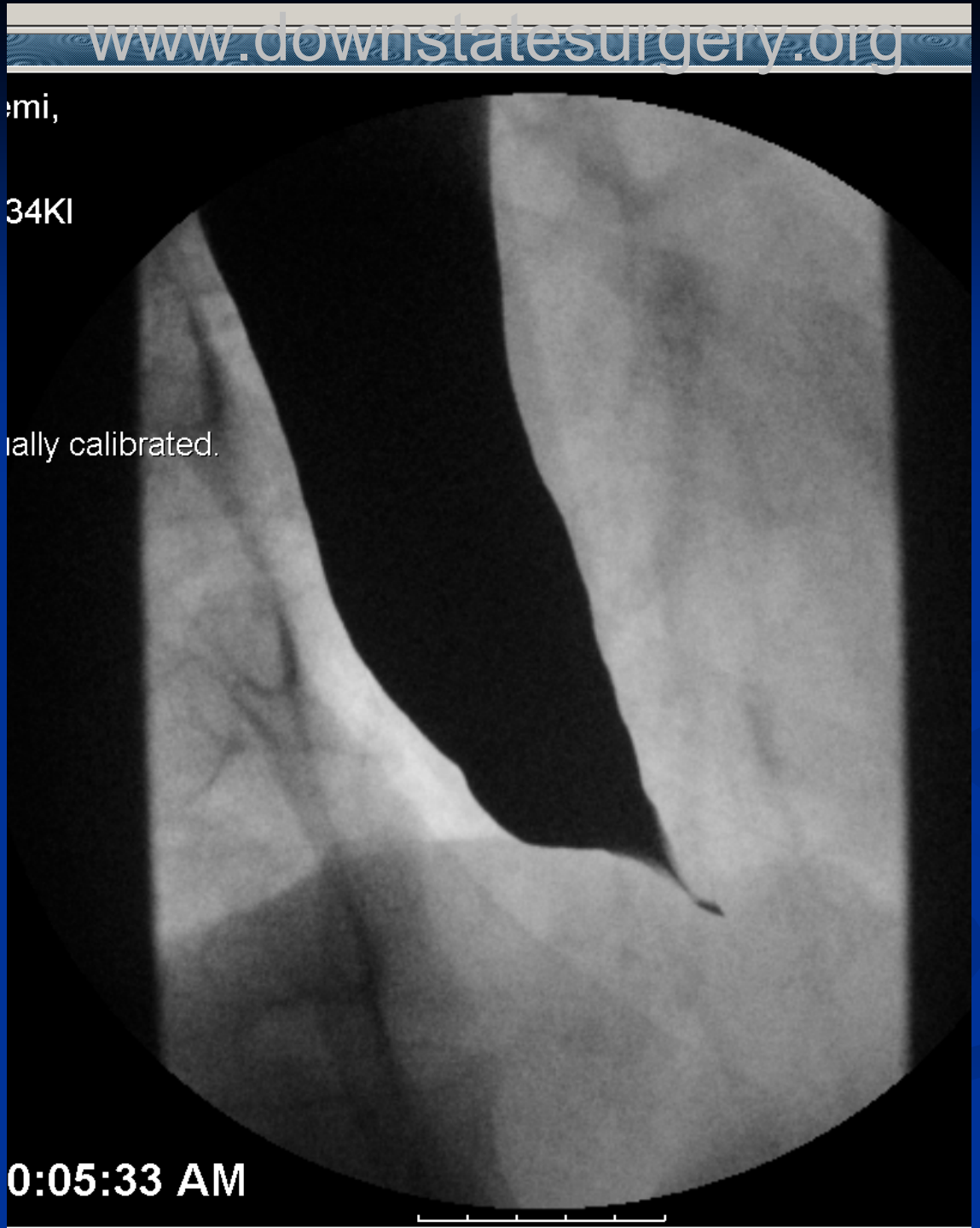

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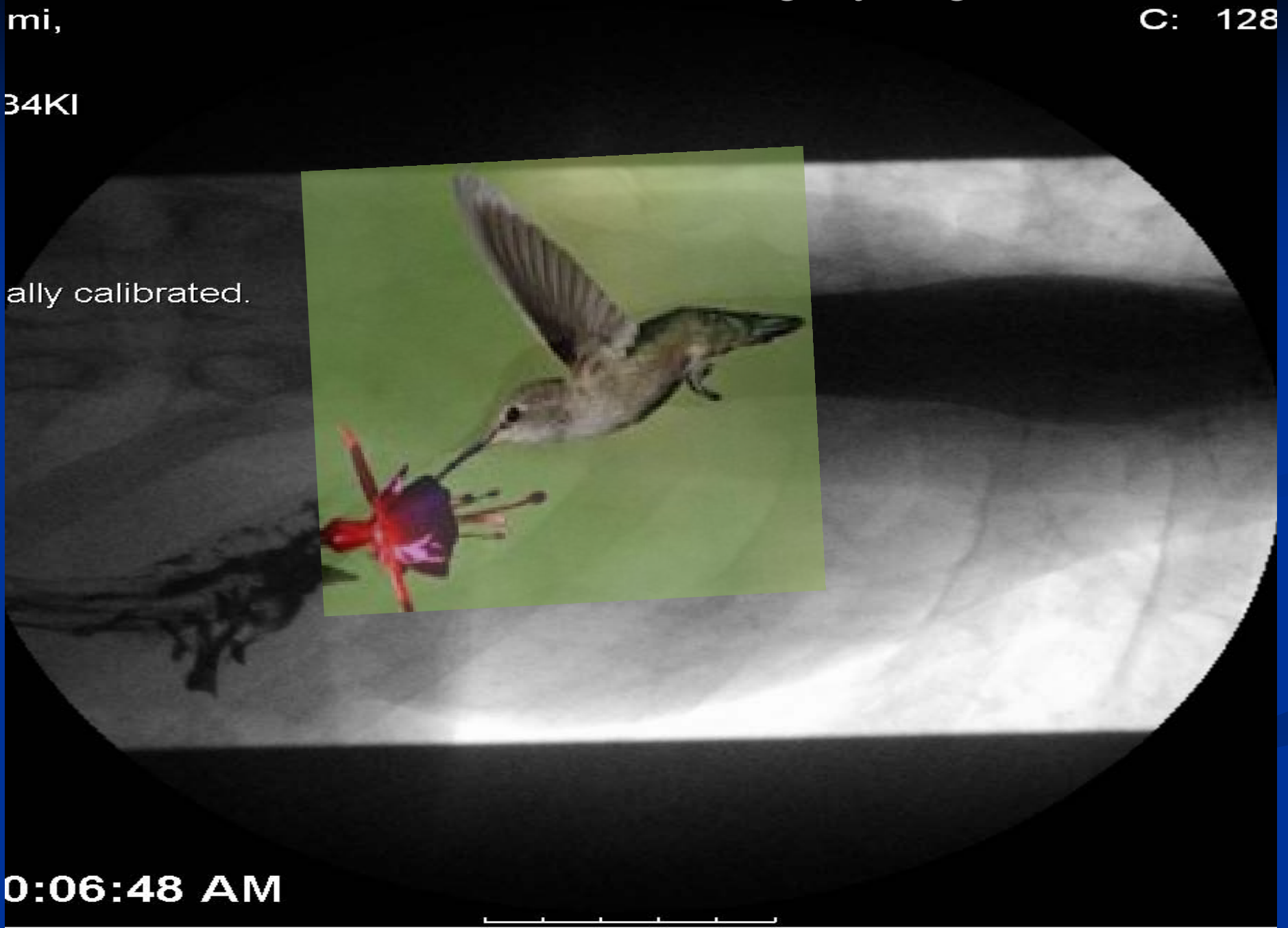

Further Workup

- Manometry ordered as outpatient
- Patient lost to follow up - 4 months
- Returned without manometry
- Evaluated by cardiothoracic surgery

Plan:

Transthoracic Heller myotomy, possible LHM

Surgery

- Underwent LHM with Dor fundoplication
- 5cm esophageal myotomy, 2cm gastric extension
- Intraoperative EGD by CT surgery

- Postoperative esophagram negative for leak

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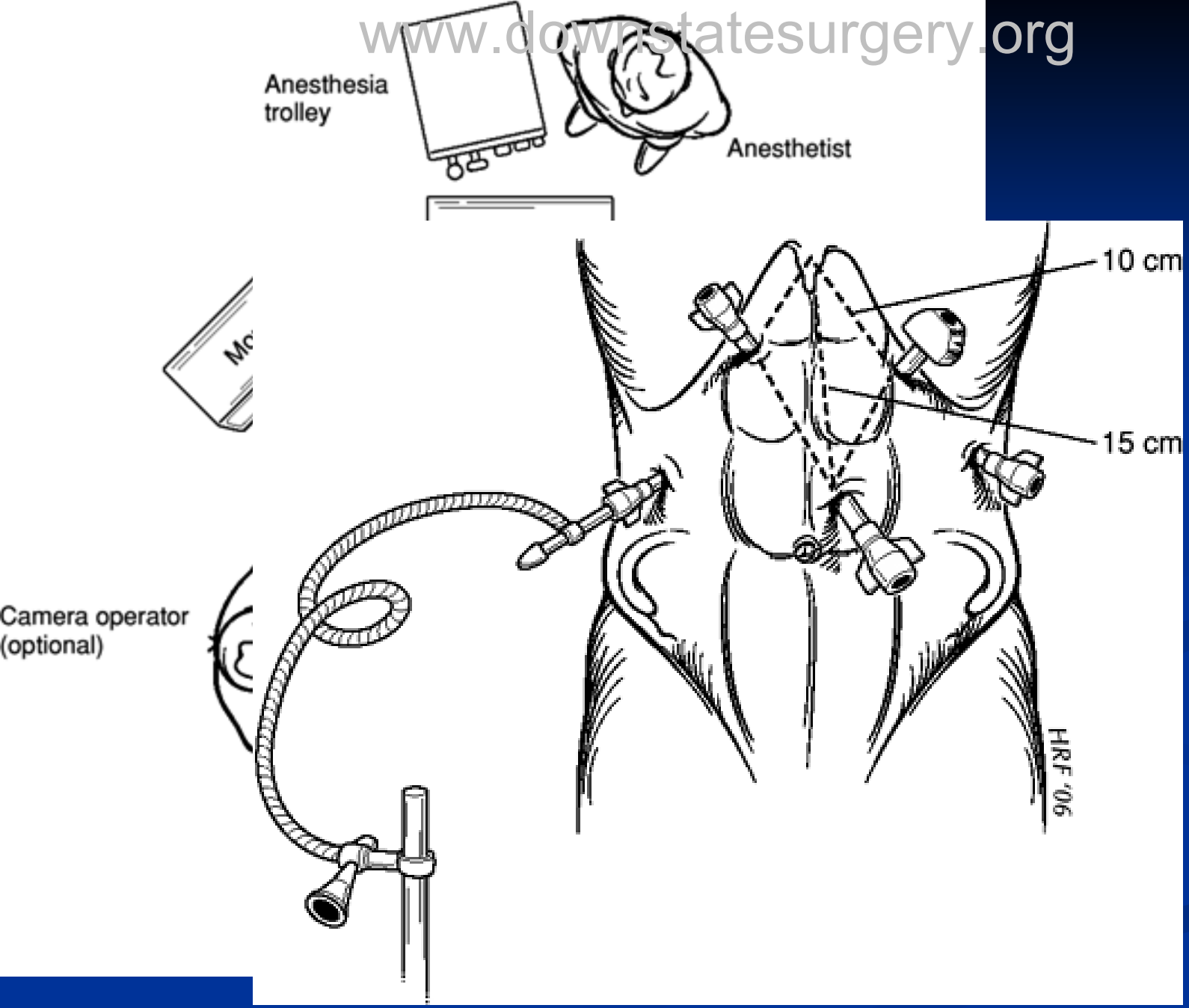

Postoperative EGD


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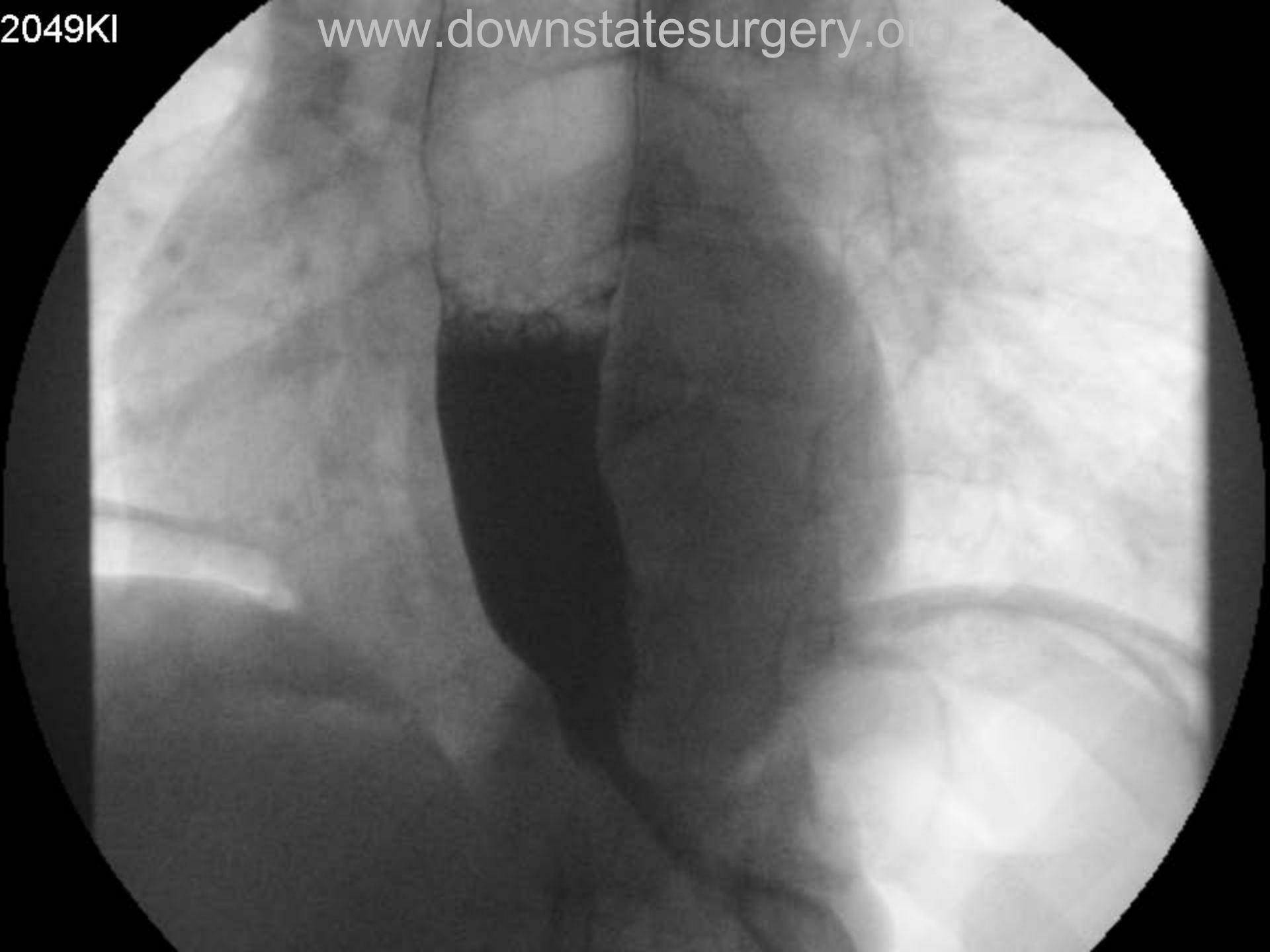
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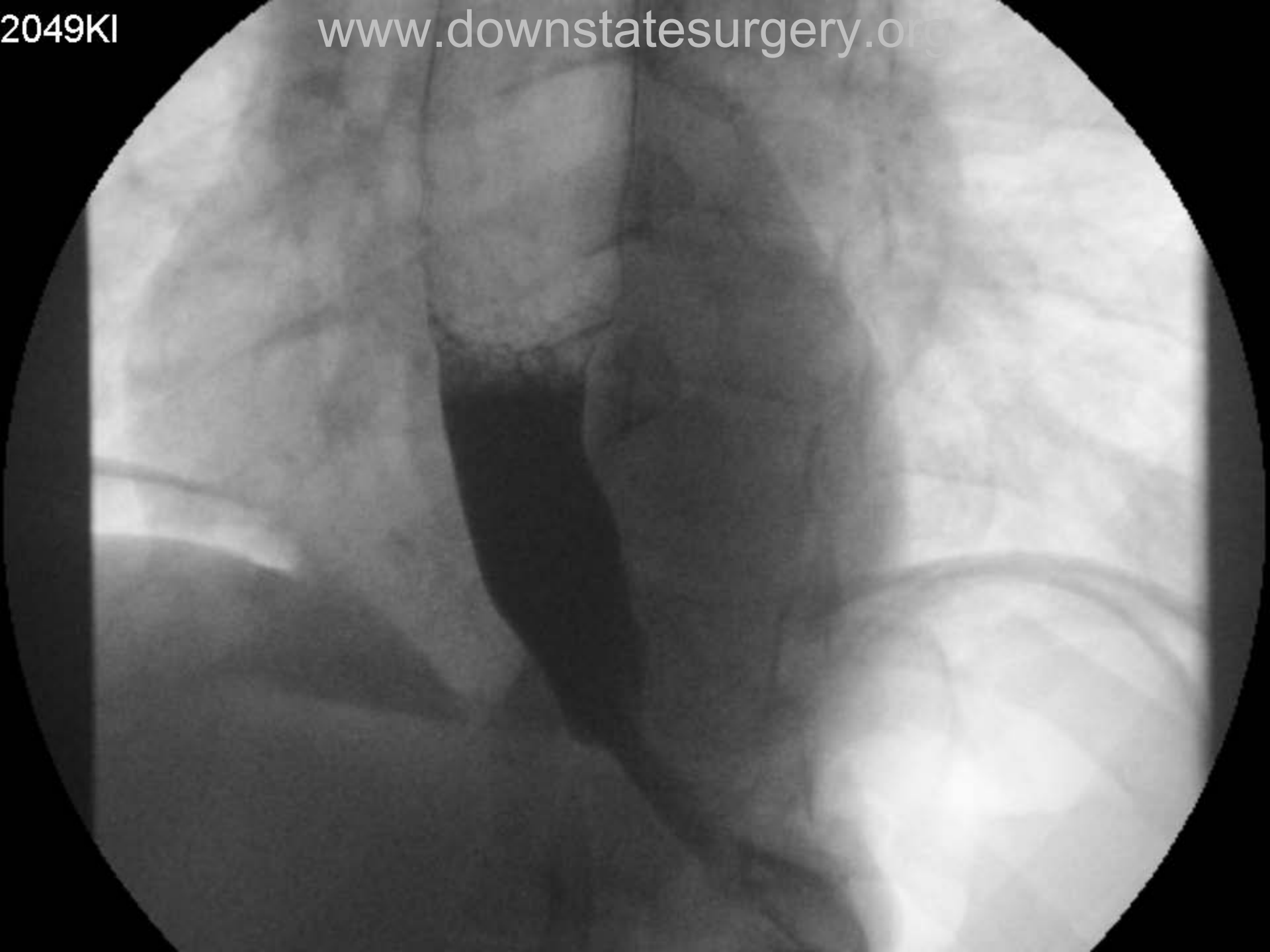
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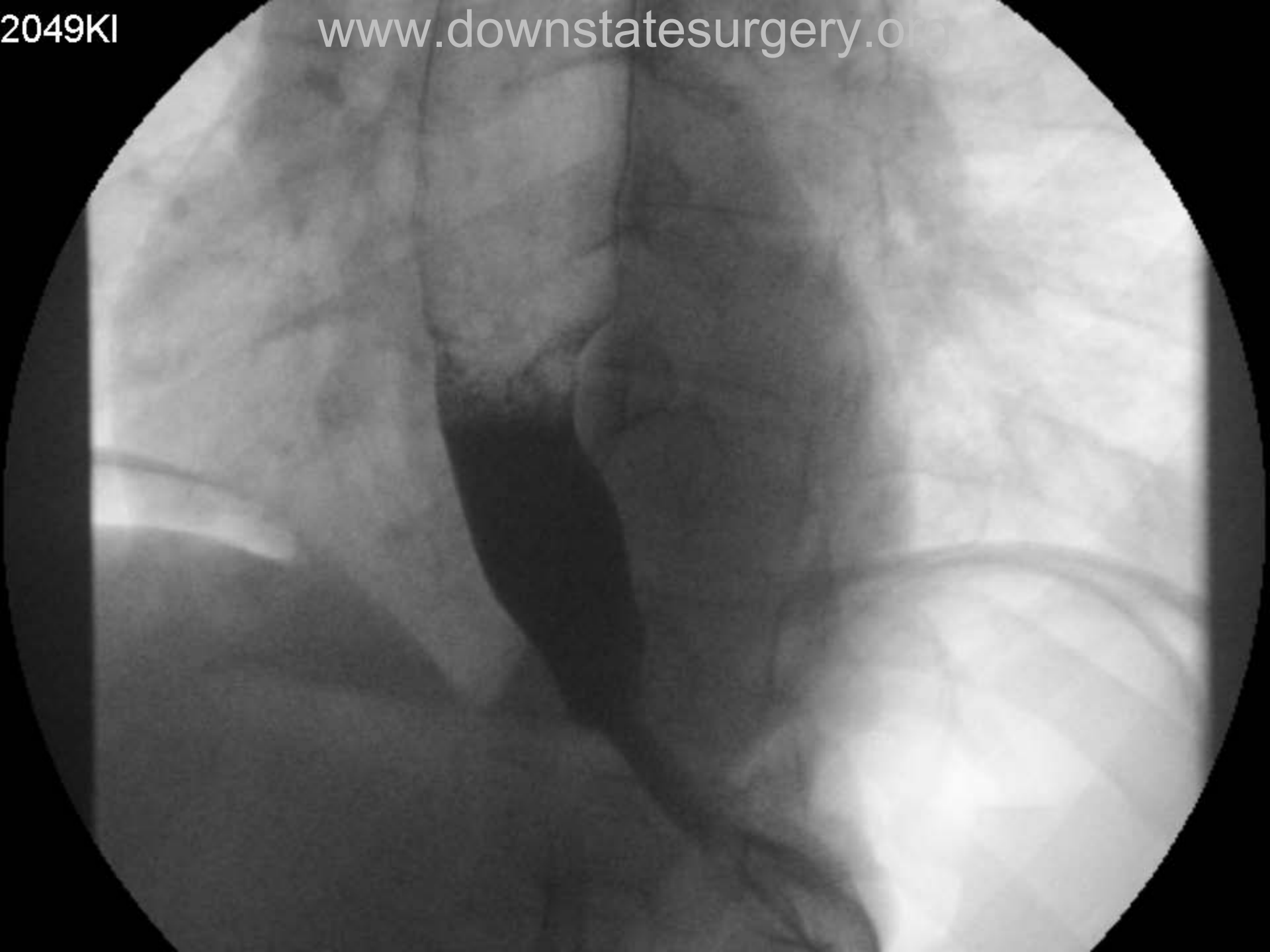
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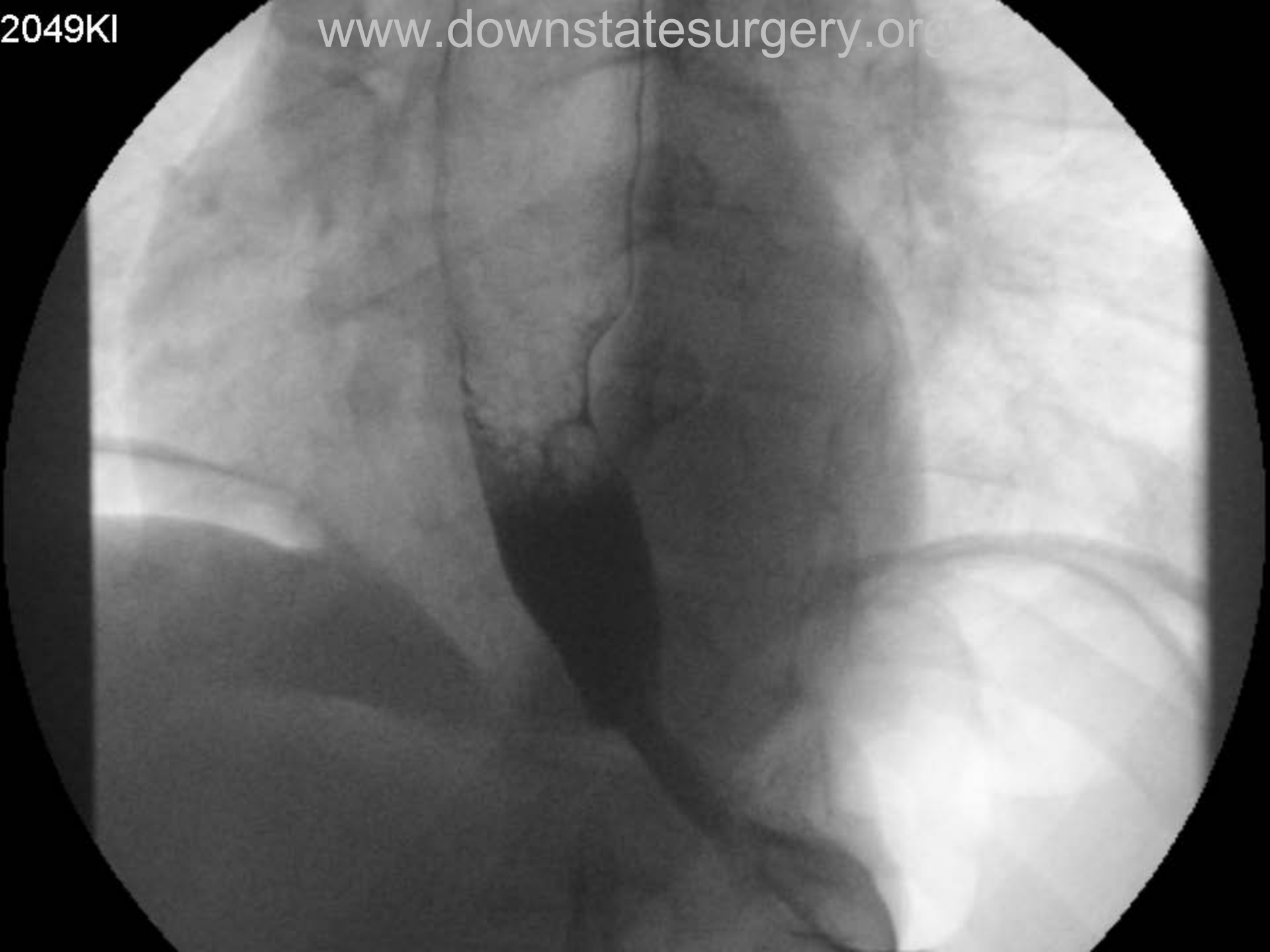
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Perioperative Course

- Discharged home POD#2 after tolerating diet
- Seen in clinic postoperatively, no symptoms

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QUESTIONS?

Achalasia

- Literally means “failure to relax”
- First described in 1672 by Sir Thomas Willis
- Affects 1 in 100,000 in North America
- Peak incidence between ages 20 and 50
- Slight male predominance

Pathophysiology

- Etiology remains unclear, two theories exist:
  - Progressive degeneration of neurons
  - Viral infection of neurons
- Auerbach's plexus is affected
- Results in aperistalsis and hypertrophic LES
- Increased risk of cancer
  - Squamous cell carcinoma from irritation by food
  - Adenocarcinoma from reflux after dilatation

Clinical Presentation

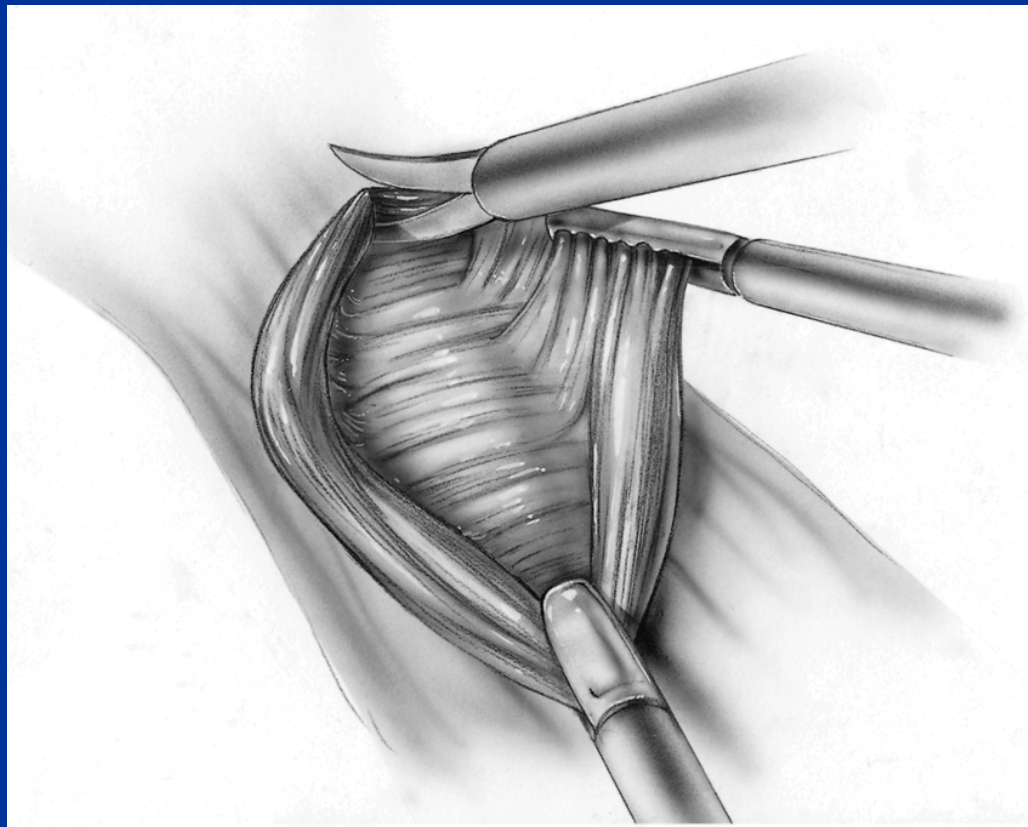
- Progressive dysphagia to liquids then solids
- Often misdiagnosed as GERD in early stages
- Aspiration pneumonia and WL in later stages
- Clinical triad: dysphagia, regurgitation, WL

Initial Evaluation

- Barium Swallow
- EGD is required to rule out pseudoachalasia
- Esophageal Manometry
  - Failure of LES to relax
  - Aperistalsis of the esophagus

Treatment Modalities

- Medical Treatment
- Endoscopic
  - Botulinum Toxin
  - Balloon Dilatation
  - POEM
- Surgery



Medical Treatment

- Nitrates & Calcium CB relax smooth muscle, LES ↓
- Temporarily relieve dysphagia
- Decreased efficacy in long term use
- Studies show minimal clinical improvement
- Reserved as temporizing tx or in high risk patients

Botulinum Toxin

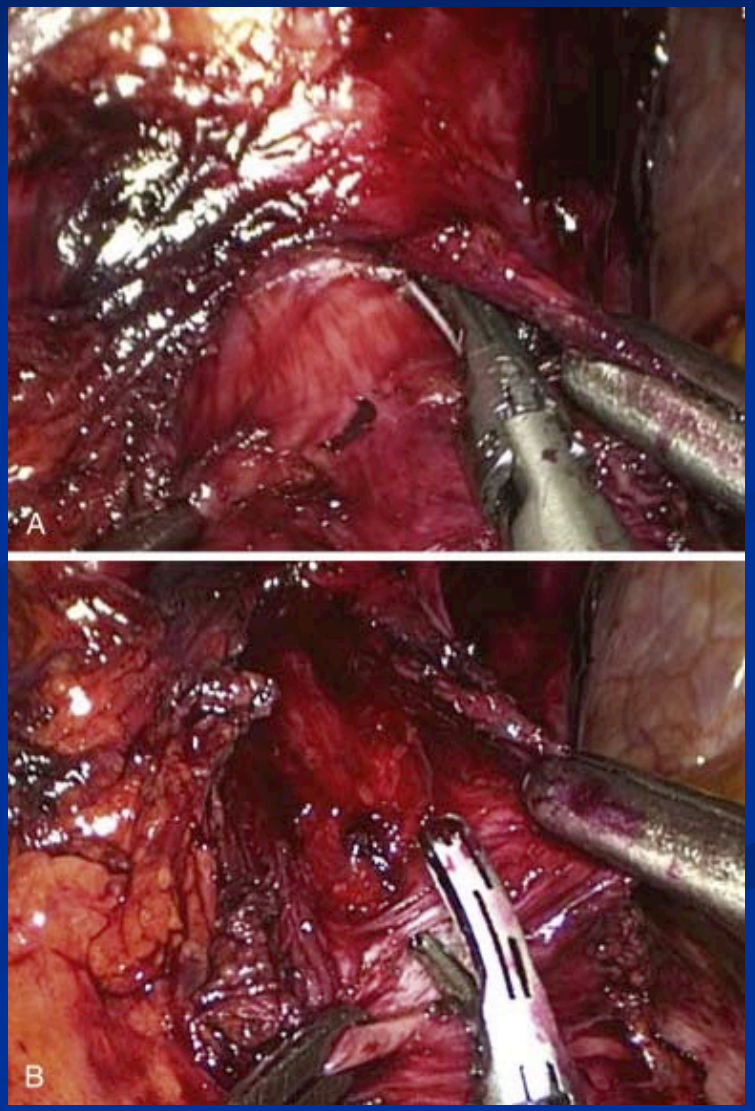
- Potent inhibitor of Ach at presynaptic terminal
- Lasts 3-12 months in 50-60% of cases
- Repeated injections are often required
- Can sclerose gastroesophageal junction
- Reserved for patients who are poor candidates
- Some benefit noted in older pts, vigorous achalasia

Endoscopic Dilatation

- Most effective nonsurgical treatment
- Pneumatic dilators preferred, rupture LES muscle
- Younger patients and males less likely to benefit
- Pulmonary symptoms & 2 failed attempts => TF
- Complications: perforation, reflux, hematoma

Surgical Approaches

- Laparotomy
- Transthoracic
- Laparoscopy



Transthoracic approach

- Do not disrupt the phrenoesophageal ligament
- Facilitates longer esophageal myotomy
- Drawbacks:
  - Chest tube drainage
  - Dual lumen intubation
  - Limited options of fundoplication
  - Inability to carry out myotomy onto stomach

Transabdominal Approach

- Three options: open, laparoscopic, robotic
- Superior to transthoracic, lower reflux rates
- LHM: shorter stay, less pain, lower morbidity
- Robotic assistance may decrease mucosal tears
- Myotomy > 4 cm (esophagus) 1-2 cm (gastric)

Predictors of Surgical Failure

- Severe preoperative dysphagia
- Low preop LES pressure < 30-35 mm Hg
- Stage IV disease (sigmoid esophagus)
- Prior endoscopic therapy

Fundoplication

- Protective against reflux post myotomy
- Nissen fundoplication is counter-productive
- Dor/Toupet fundoplication is preferred
- No evidence as to which is best

Controversies

- Since LHM, drawing comparisons with endoscopy
- Over past 15 years, numbers have favored LHM
- GI physicians among the first to see patient
  - Invasive surgery vs outpatient procedure ?
- Refined techniques improved endoscopic statistics
- Change in clinical algorithm?

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Pneumatic Dilation versus Laparoscopic Heller's Myotomy for Idiopathic Achalasia

Guy E. Boeckxstaens, M.D., Ph.D., Vito Annese, M.D., Ph.D., Stanislas Bruley des Varannes, M.D., Ph.D., Stanislas Chaussade, M.D., Ph.D., Mario Costantini, M.D., Ph.D., Antonello Cuttitta, M.D., J. Ignasi Elizalde, M.D., Uberto Fumagalli, M.D., Ph.D., Marianne Gaudric, M.D., Ph.D., Wout O. Rohof, M.D., André J. Smout, M.D., Ph.D., Jan Tack, M.D., Ph.D., Aeilko H. Zwinderman, Ph.D., Giovanni Zaninotto, M.D., Ph.D., and Olivier R. Busch, M.D., Ph.D., for the European Achalasia Trial Investigators*

Study

- 201 patients randomly assigned to PD or LHM
  - Pneumatic dilatation 30 mm, 35 mm, 40 mm
  - Laparoscopic Heller myotomy, Dor fundoplication
- Primary outcome drop in Eckhardt score to ≤ 3
- Secondary outcomes
  - Need for retreatment
  - Esophageal emptying
  - Rate of complications

Results

- Patients followed for two years
- PD group (N=95): 27 with recurrent symptoms, 17 required re-dilatation; 5 failed & had surgery. 4 suffered perforation, 2 repaired operatively
- LHM group (N=106): 16 failed & underwent dilatation. Mucosal tears in 16, repaired intraop

Authors' Conclusions

- PD and LHM are both effective treatments
- Rate of treatment success is similar
- LHM with Dor is not superior to PD
- Choice of procedure based on available expertise
- Age and gender should also be considered

Criticisms

- Esophageal dilatation requires multiple tx
- Rate of mucosal tears & reflux in LHM are high
- 1-1.5 cm gastric extension is insufficient
- Follow up is short: success dwindles after 10yrs

New Directions

- POEM

Summary

- Achalasia: MC esophageal primary motor disorder
- LHM & partial fundoplication best option ≤ 40y
- Need to determine value of subtypes & tx options
- New goals:
  - Individualized treatment based on subtype
  - Vaccine/restoring function to denervated esophagus

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THANK YOU

References

- Fischer, Mastery of Surgery, 6th Edition
- Cameron, Current Surgical Therapy 10th Edition
- Velasco, Rush University Review of Surgery
- SAGES Guidelines for Surgical Treatment of Achalasia

QUESTION 1

- **A 35-year-old woman has complaints of dysphagia, regurgitation, and weight loss. Esophagography shows narrowing of the distal end of the esophagus, and manometry studies show significant tertiary waveforms. The LES has high residual pressure on swallowing. Which of the following has not been implicated as a possible cause of her disease?**

A *Helicobacter pylori* infection
B Severe emotional stress
C A parasitic infection
D Drastic weight reduction
E Degeneration of the Auerbach plexus

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A LES pressure of 40 mm Hg
B LES pressure of 10 mm Hg with deglutition
C Esophageal body pressure above baseline
D Significant aperistalsis
E High-amplitude waveforms

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QUESTION 3

- **A patient arrives at the emergency department 8 hours after balloon dilation of her esophagus with complaints of dysphagia and chest pain. She was found to be febrile, tachycardic, and normotensive. Esophagography showed “bird's beak” narrowing and a leak at the distal end of the esophagus with contrast material in the left side of the chest. After fluid resuscitation and antibiotics, which of the following is the most appropriate management?**

A Nasogastric tube decompression and observation

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C Left thoracotomy, primary repair, myotomy, and drain placement

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