

Core Lecture: Esophageal Motility Disorders

John M. Wo, M.D.

Division of Gastroenterology/Hepatology

August 30, 2007

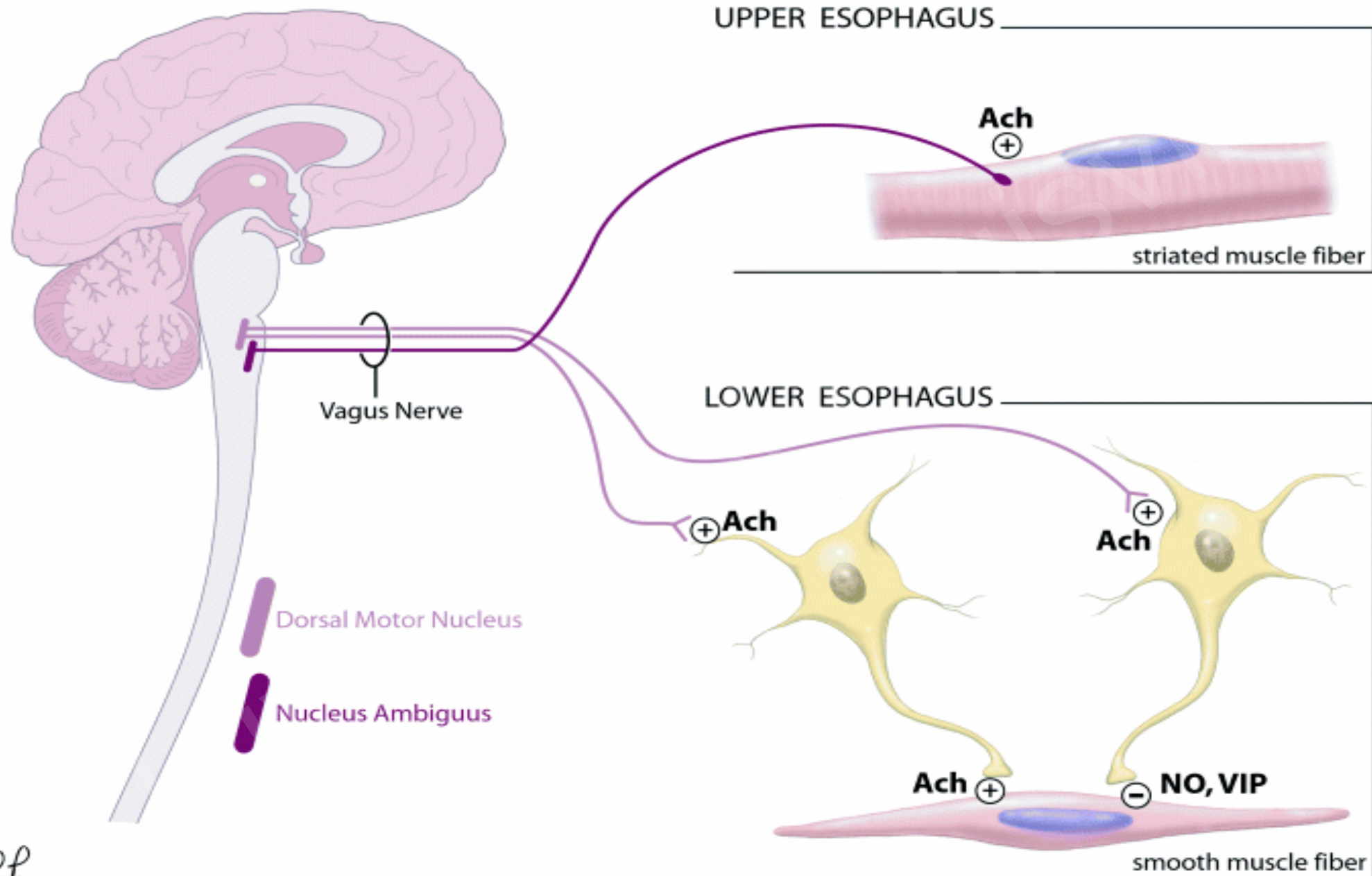
Core Lecture:

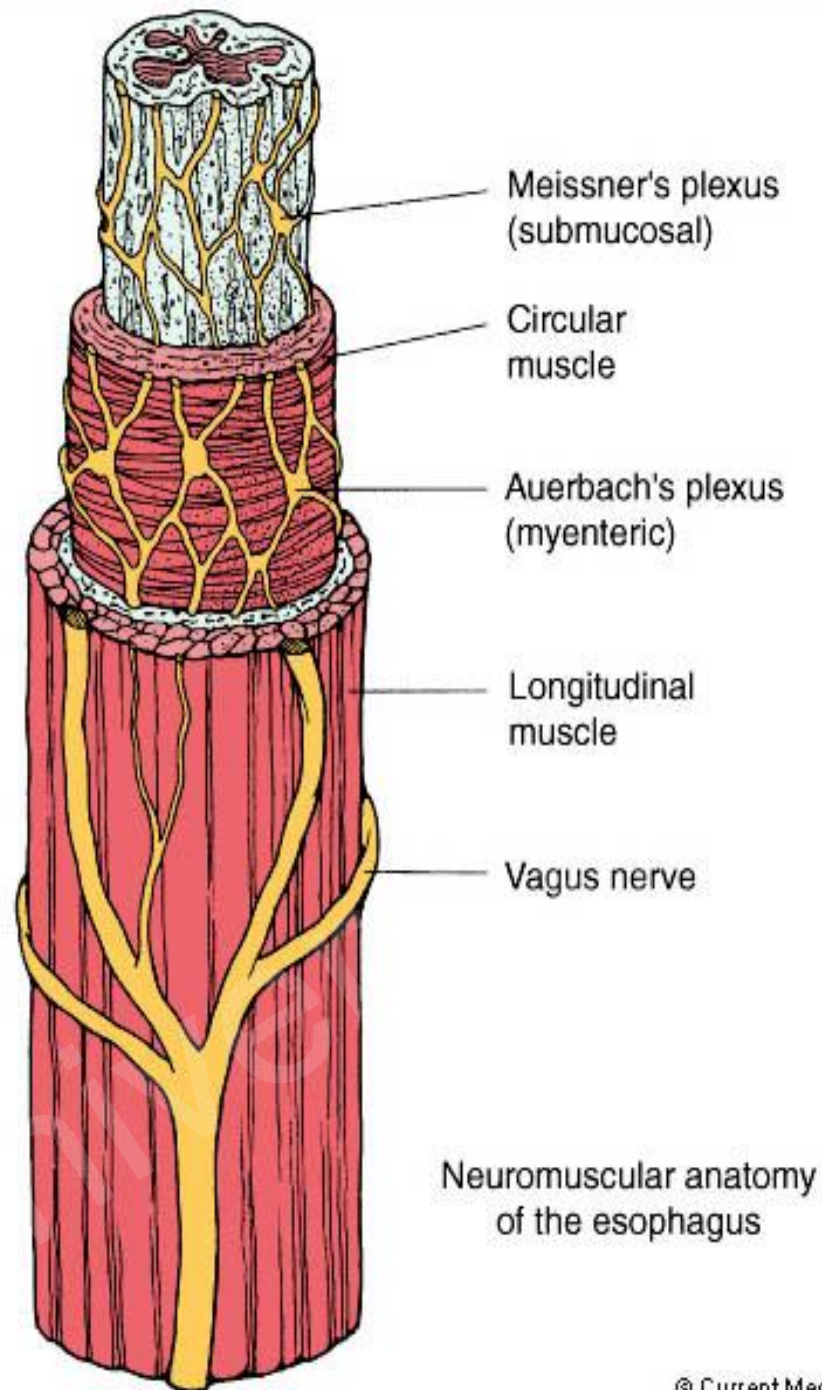
Esophageal Motility Disorders

- Normal esophageal anatomy and physiology
- Evaluation of esophageal function
- Classification of esophageal motility disorders
 - Hypercontracting and hypocontracting esophagus
- Specific esophageal motility disorders

Symptoms Suggesting Esophageal Origin

- Esophageal
 - Heartburn
 - Regurgitation
 - **Dysphagia**
 - Odynophagia
- Other
 - Atypical GERD (shortness of breath, cough, hoarseness, throat clearing, sore throat, globus, etc.)
 - **Chest pain**
 - Aspiration
 - Weight loss





- Any vagal or myenteric neuropathy may results in esophageal motility disturbance
 - Hypercontracting or Hypocontracting esophagus

Evaluation of the Esophagus

- Barium swallow (with barium tablet)
- Timed barium swallow (achalasia protocol)
- Upper endoscopy
- Esophageal manometry
- Ambulatory pH monitoring
 - Bravo and transnasal
- Esophageal provocation testing
 - Acid, tensilon, balloon distension
- Esophageal impedance

Clinical Utility of Esophageal Manometry

1. To accurately define esophageal motor function
2. To define abnormal motor function
3. To delineate a treatment plan based on motor abnormalities

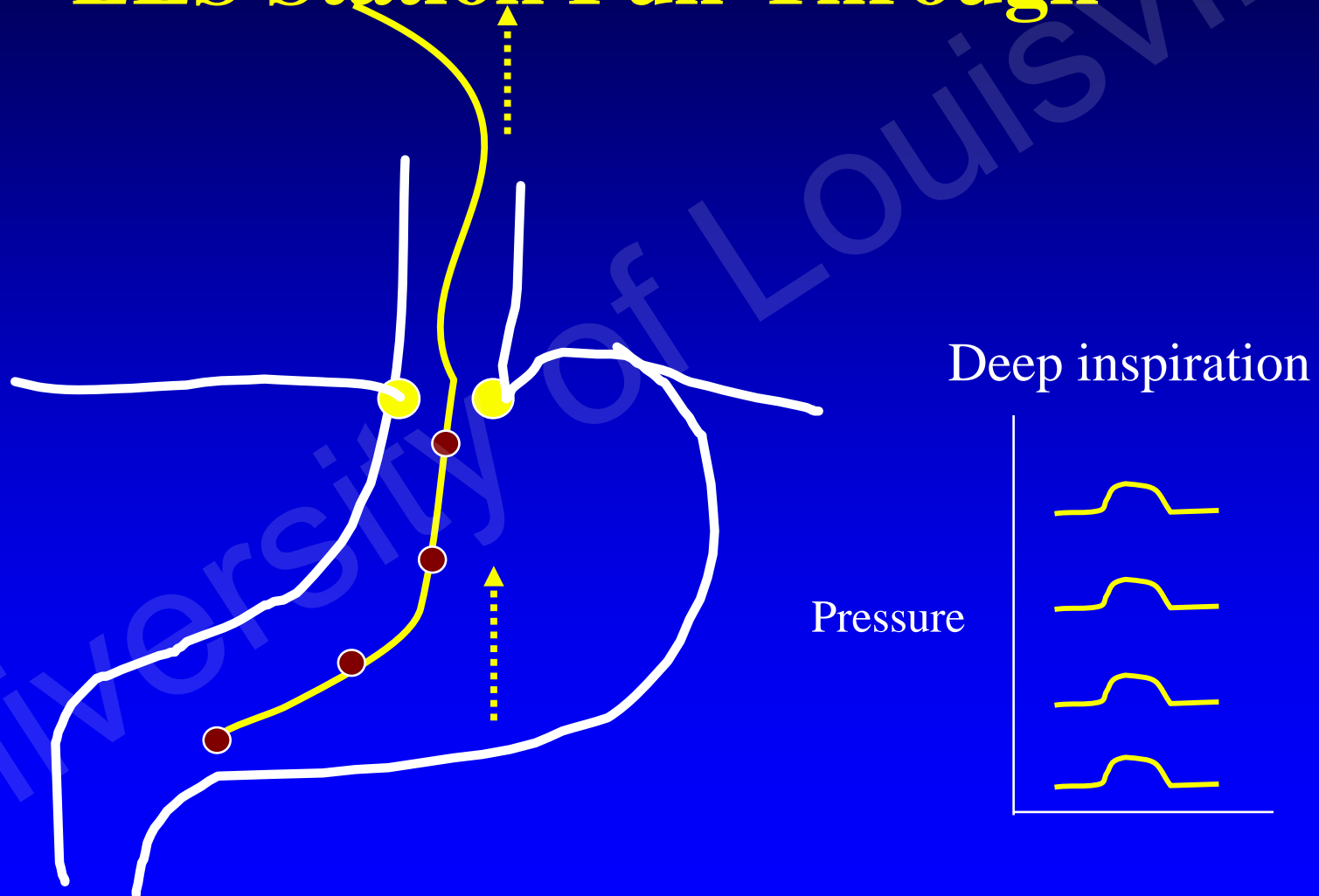
Indications for Esophageal Manometry

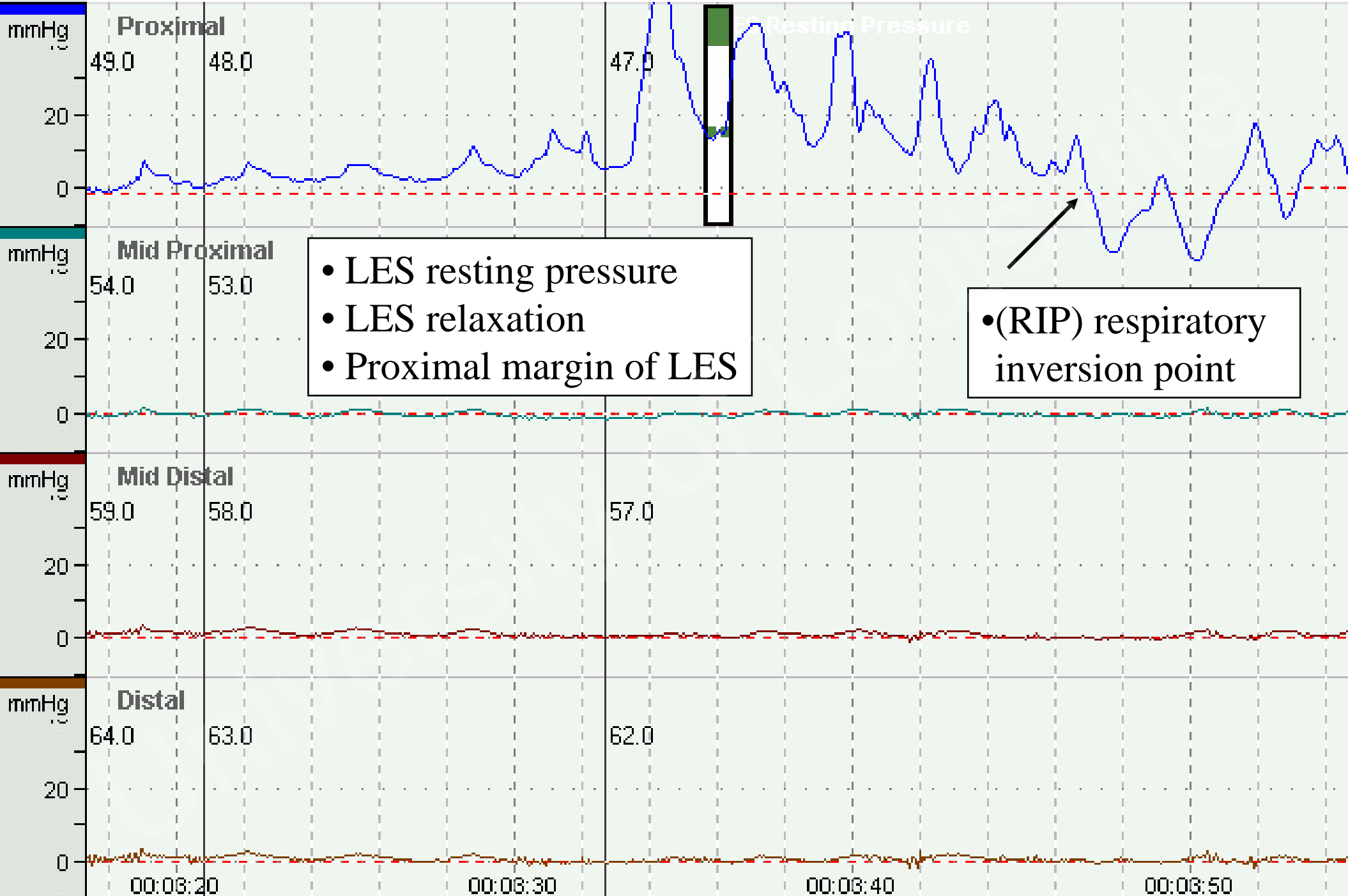
- Diagnose achalasia
- Suspect impaired esophageal motility
- Dysphagia of unclear etiology
- Pre-op evaluation for fundoplication
- Post-fundoplication evaluation
- Suspect diffuse UGI dysmotility

Esophageal Manometry Methods

- Water perfusion manometry
- Solid state manometry
 - Standard (every 5 cm)
 - High resolution (every 1 cm)

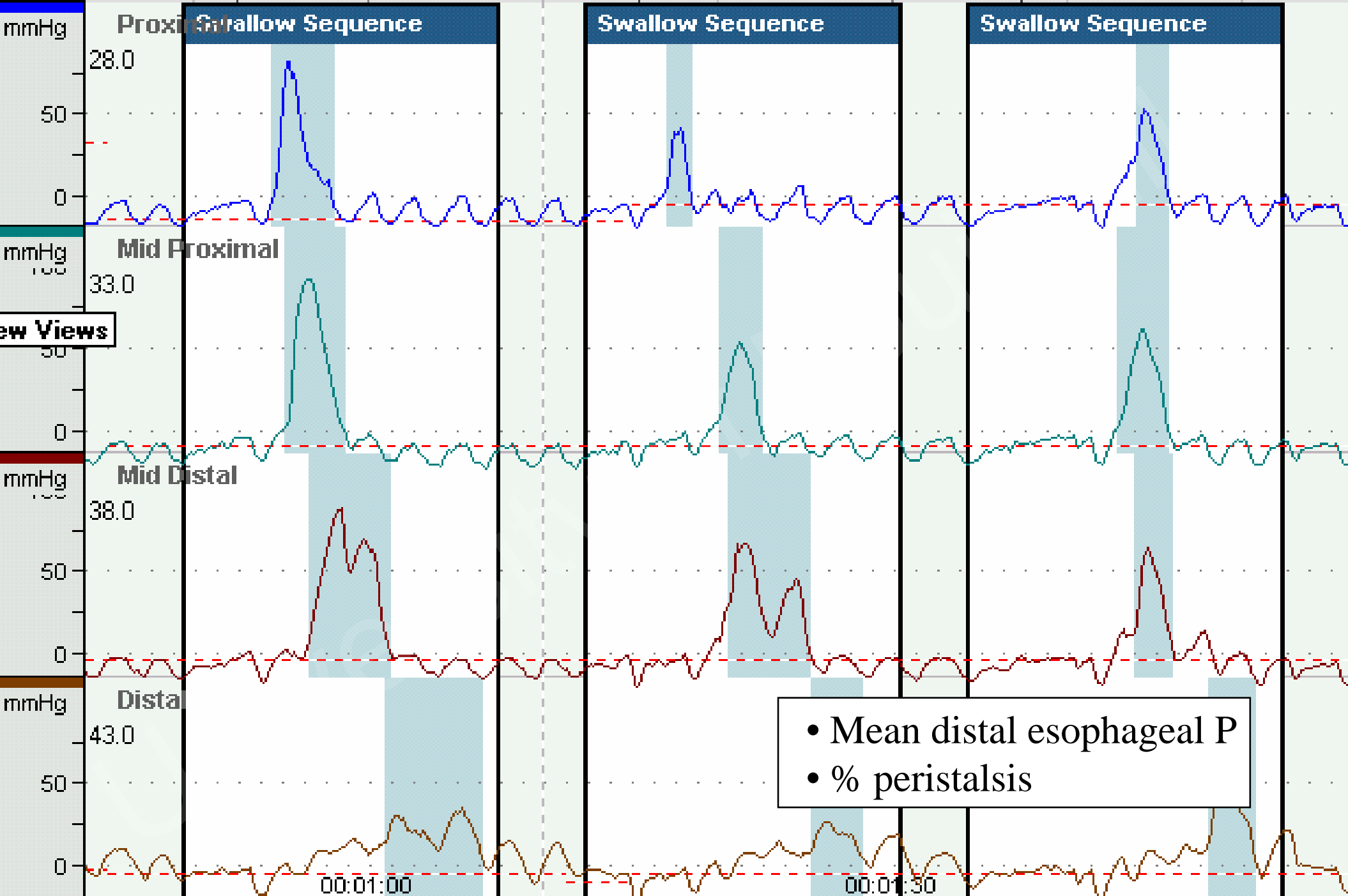
Esophageal Manometry: LES Station Pull-Through





Esophageal Manometry: Esophageal Body Measurements





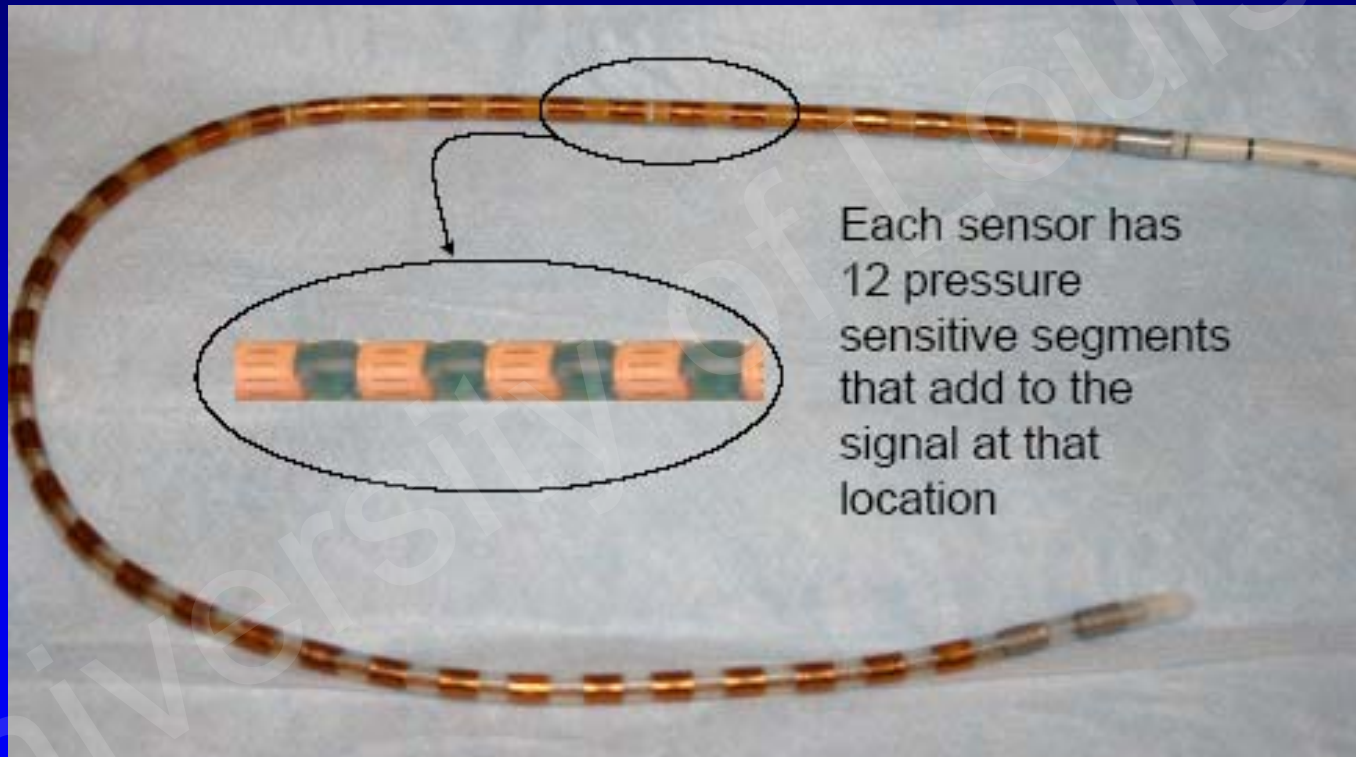
Normal Esophageal Manometry

| <u>Pressure</u> | <u>mmHg (SD)</u> | <u>Normal</u> |
|------------------|------------------|---------------|
| LES | 15.2 (10.1) | 15 - 45 |
| Mean distal P | 99 (40) | 40 – 180 |
| % of peristalsis | ---- | > 60% |

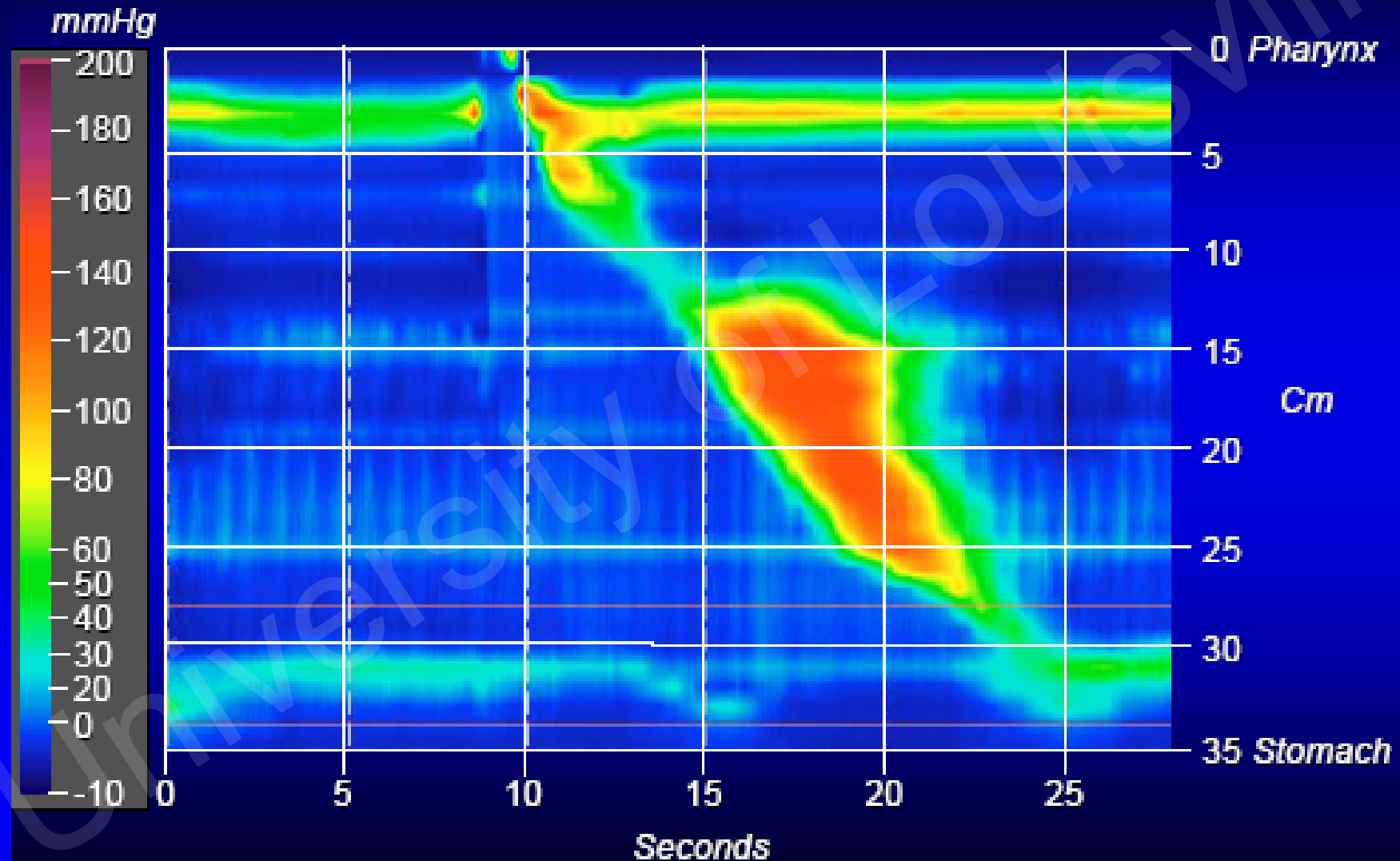
Richter et al. Dig Dis Sci 1987;32:583-592

Waring and Wo. Am J Gastroenterol 1995;90:35-37.

High-Resolution Esophageal Manometry



Normal Peristalsis and Sphincter Relaxation



Classifications of Esophageal Motility Disorders

Hypercontracting esophagus

(Esophageal spastic disorders)

- Diffuse esophageal spasm
- “Nutcracker”
- Hypertensive LES

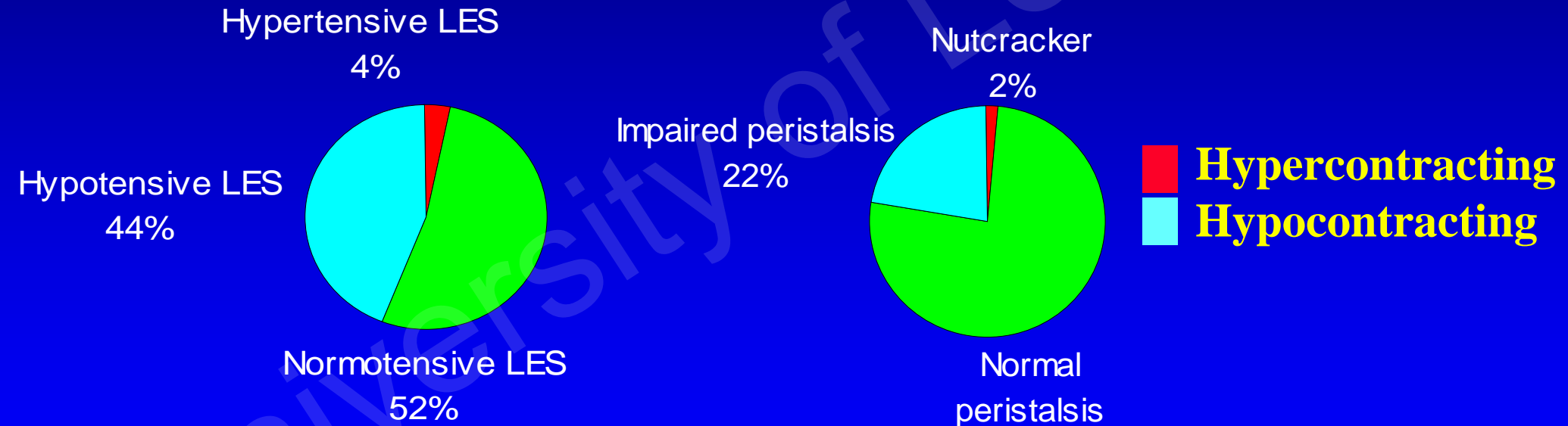
Hypocontracting esophagus

- Primary achalasia
- Secondary achalasia or impaired esophageal motility
 - Connective tissue diseases
 - Systemic sclerosis
 - Mixed connective tissue disease
 - Idiopathic inflammatory myopathy
 - Endocrine diseases
 - Diabetes
 - Neuromuscular diseases
 - Chagas disease
 - Amyloidosis
 - Paraneoplastic syndrome

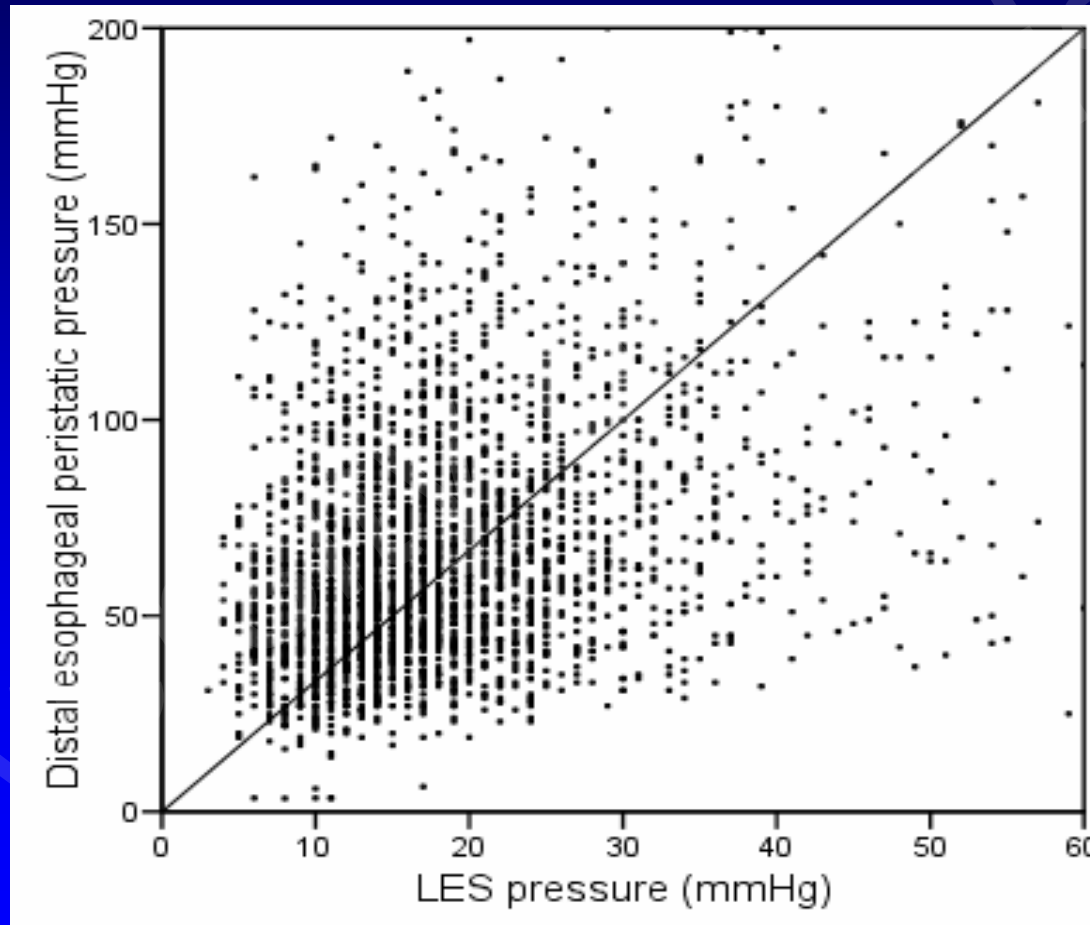
Results of Esophageal Manometry at UofL

Lower esophageal sphincter

Esophageal body



Scatter Plots Comparing Esophagus Body and LES



Kindig et al. Presented at DDW 2007 (n=2,796 manometries, achalasia excluded).

Hypercontracting Esophagus (Esophageal Spastic Disorders)

Hypercontracting Esophagus

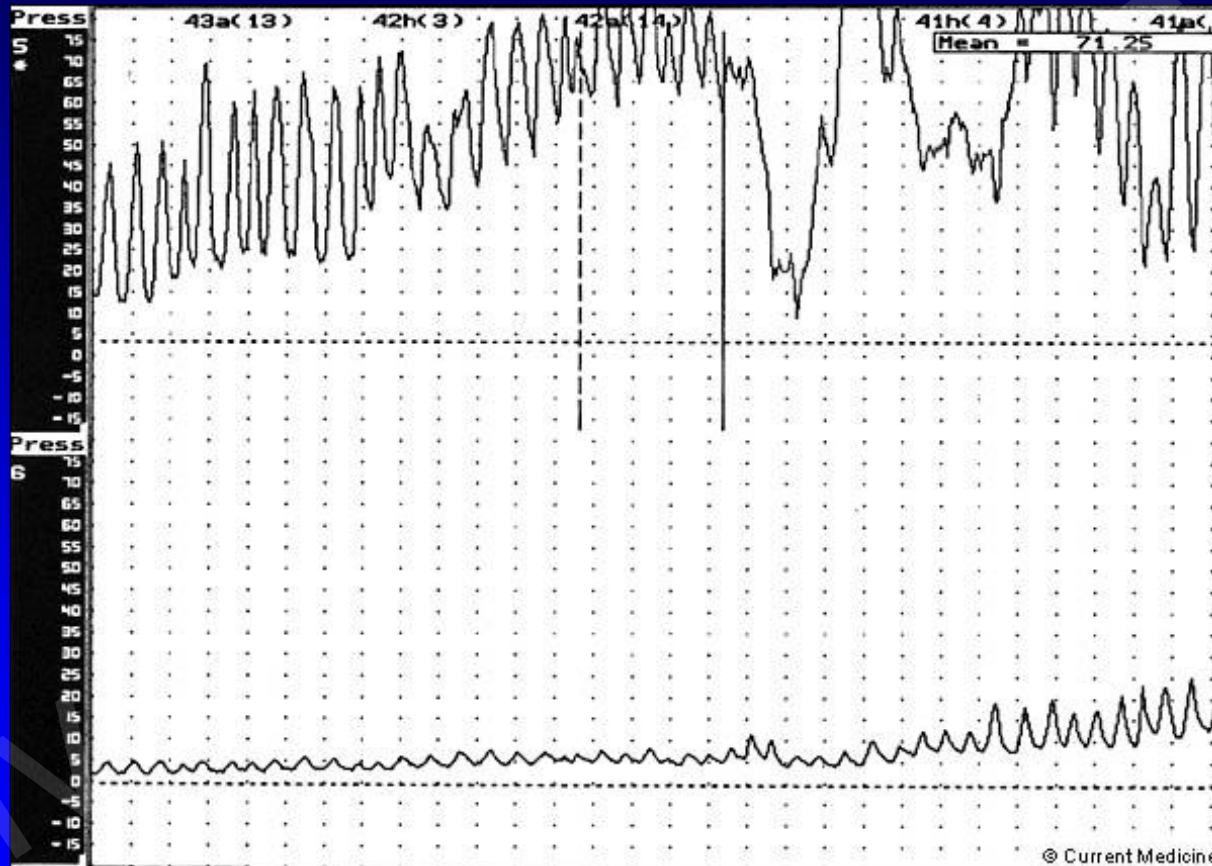
- Diffuse esophageal spasm
- Hypertensive LES
- Hypertensive esophagus (“Nutcracker”)

Diffuse Esophageal Spasm



>20 % simultaneous contraction

Hypertensive LES



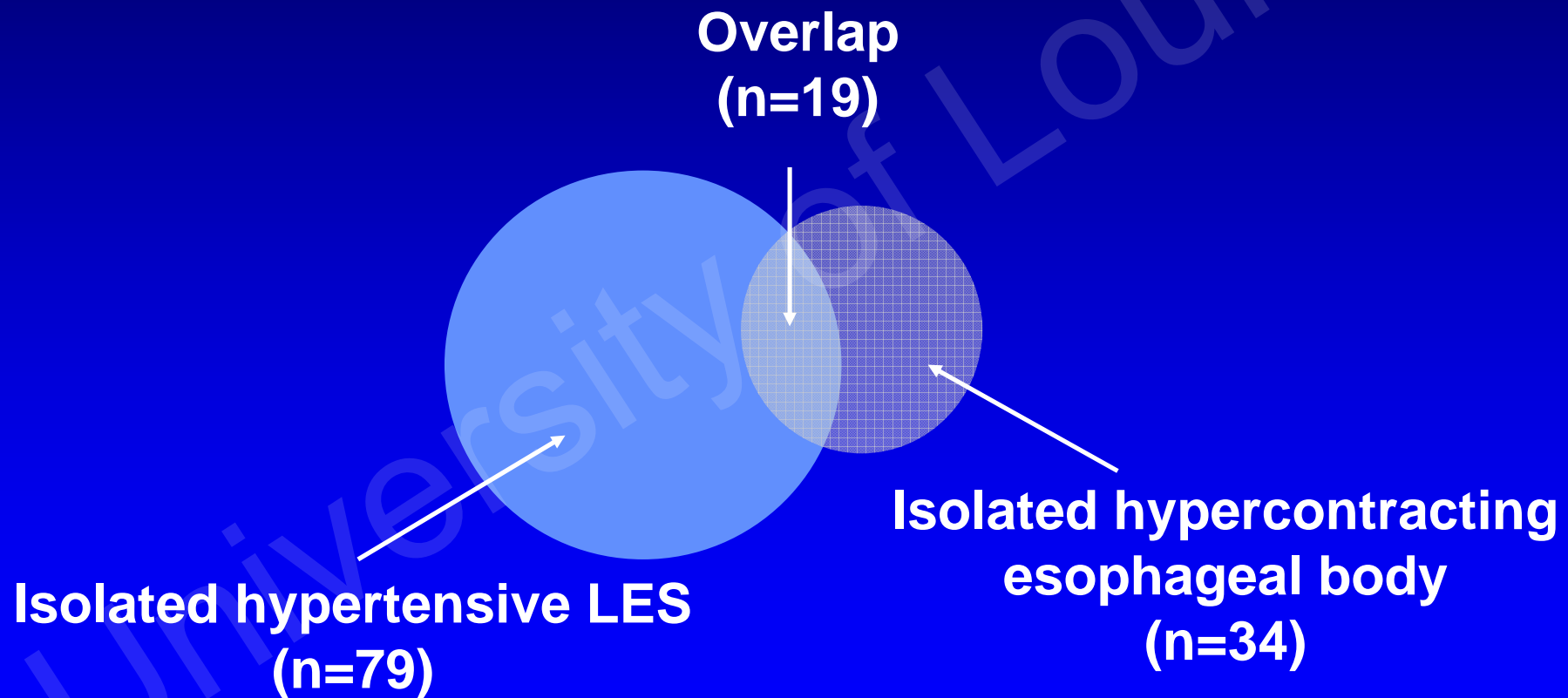
LESP >45 mmHg

Hypertensive Esophagus ("Nutcracker" Esophagus)



- Esophageal body P > 180 mmHg
- Normal peristalsis

Overlap is Uncommon between Hypertensive LES and Hypercontracting Esophageal Body (“Nutcracker”)



Underlying Causes of Esophageal Spastic Disorders

- GERD
- Esophageal obstruction
 - Stricture
 - Fundoplication
 - Food impaction
- Distension
 - Aerophagia
- Mucosal injury
 - Esophagitis
 - Bravo probe
- Idiopathic
- Secondary esophageal motility disorders
 - Diabetes
 - Pseudoobstruction
 - Amyloidosis
 - Paraneoplastic

Manifestation of Hypercontracting Esophagus

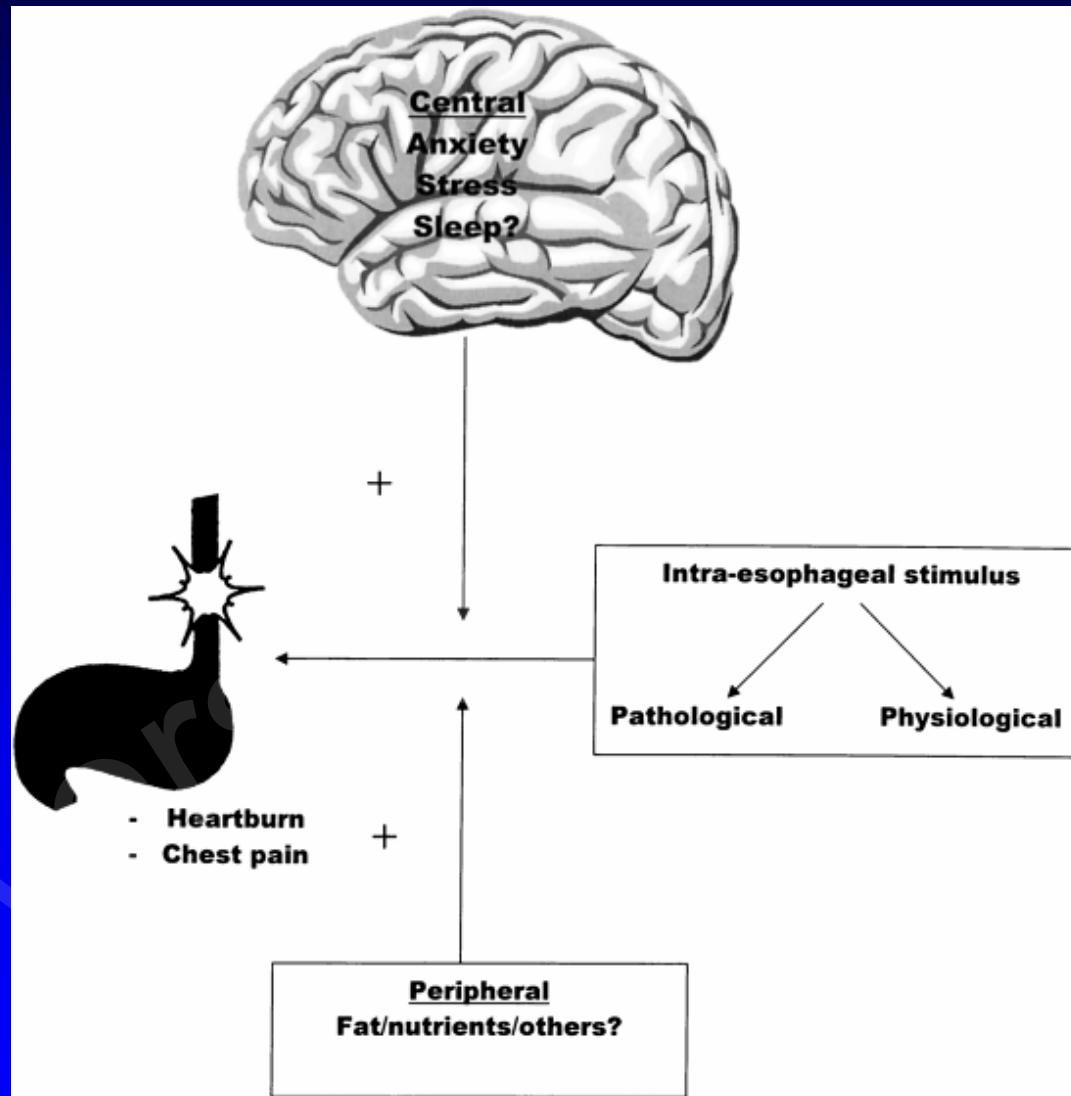
- Noncardiac chest pain
- Intermittent dysphagia
- Heartburn & regurgitation

Non-Cardiac Chest Pain

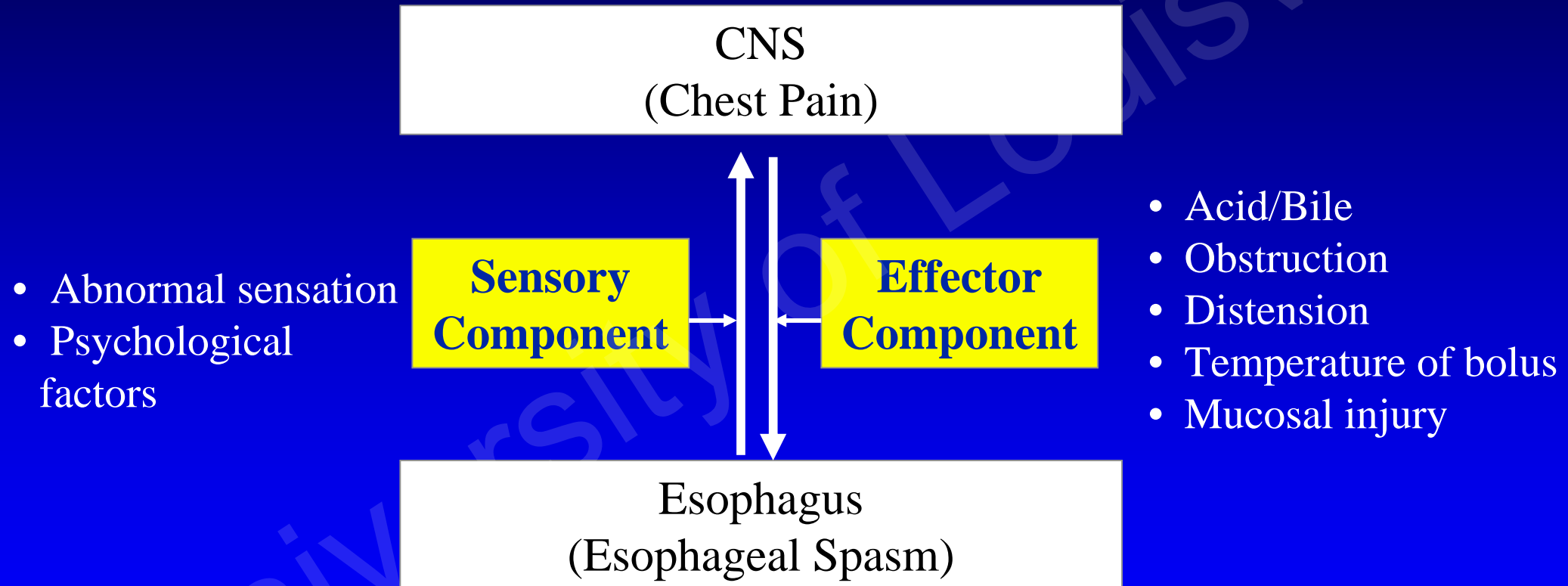
- Difficult to differentiate non-cardiac from cardiac chest pain.
- Patients may present with squeezing chest pain radiating to the back, left shoulder or jaw, mimicking myocardial ischemia.
- Chest pain can interrupt daily activity and increase work absenteeism.¹

¹Eslick et al. *Aliment Pharmacol Ther* 2004;20:909-15.

Brain-Gut Axis for Esophageal Chest Pain



Esophageal Origin for Noncardiac Chest Pain



Esophageal Spastic Disorders

- Lack of neuromuscular pathology
 - No loss of ganglion cells
 - Inconsistent changes by EM
 - No correlation with disease severity

Evaluation of Non-Cardiac Chest Pain

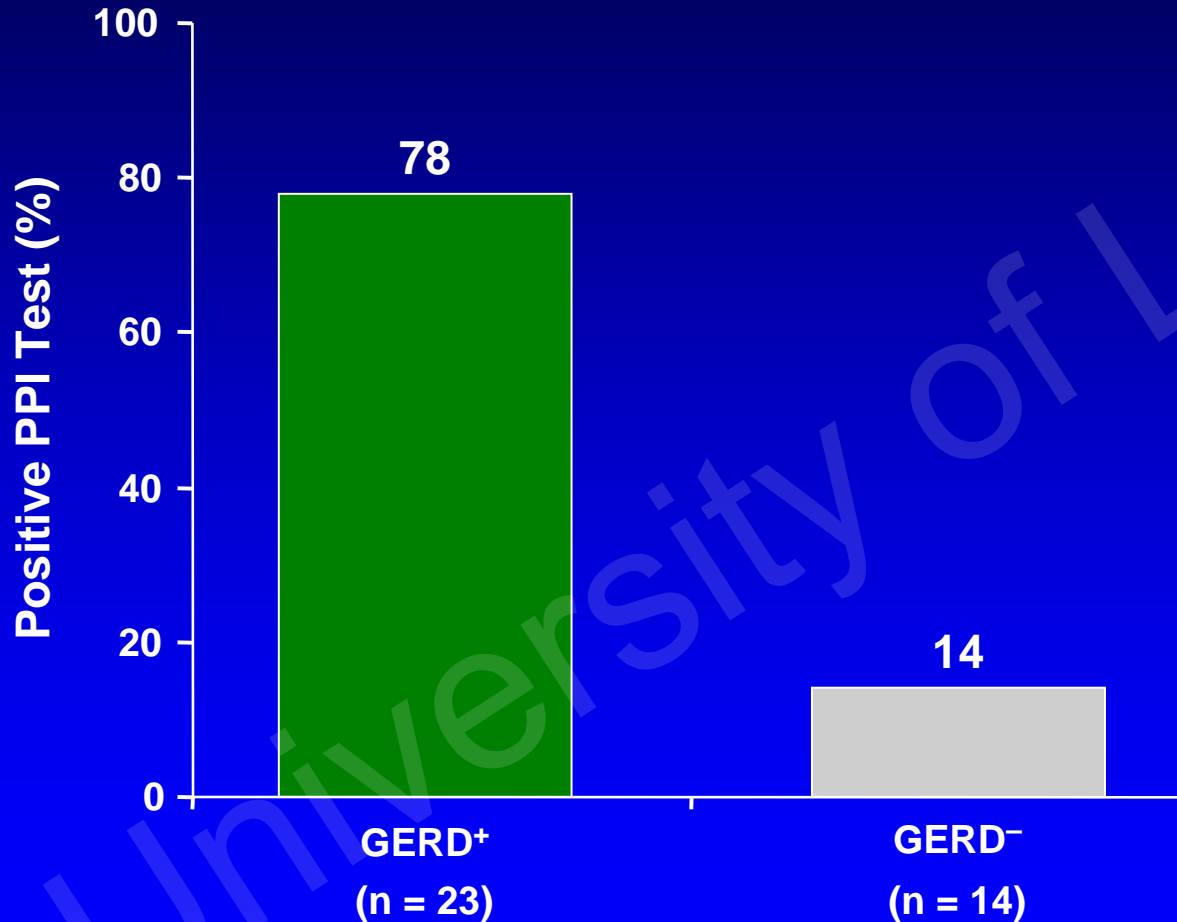
- Look for underlying cause
- Diagnostic testing
 - PPI test
 - Esophageal manometry
 - Upper endoscopy
 - Ambulatory pH monitoring
 - Ambulatory pH/impedance monitoring

Upper Endoscopy in Non-Cardiac Chest Pain

- Erosive esophagitis and Barrett's esophagus are found in only 10-25% of patients with non-cardiac chest pain.¹
- Given its low yield, upper endoscopy is not recommended as part of the initial workup.

¹Fang J et al. Am J Gastroenterol 2001;96:958-68.

PPI Test for Non-Cardiac Chest Pain



PPI test for GERD

- Sensitivity 78%
- Specificity 86%

Omeprazole 40 mg in the morning and 20 mg at night.

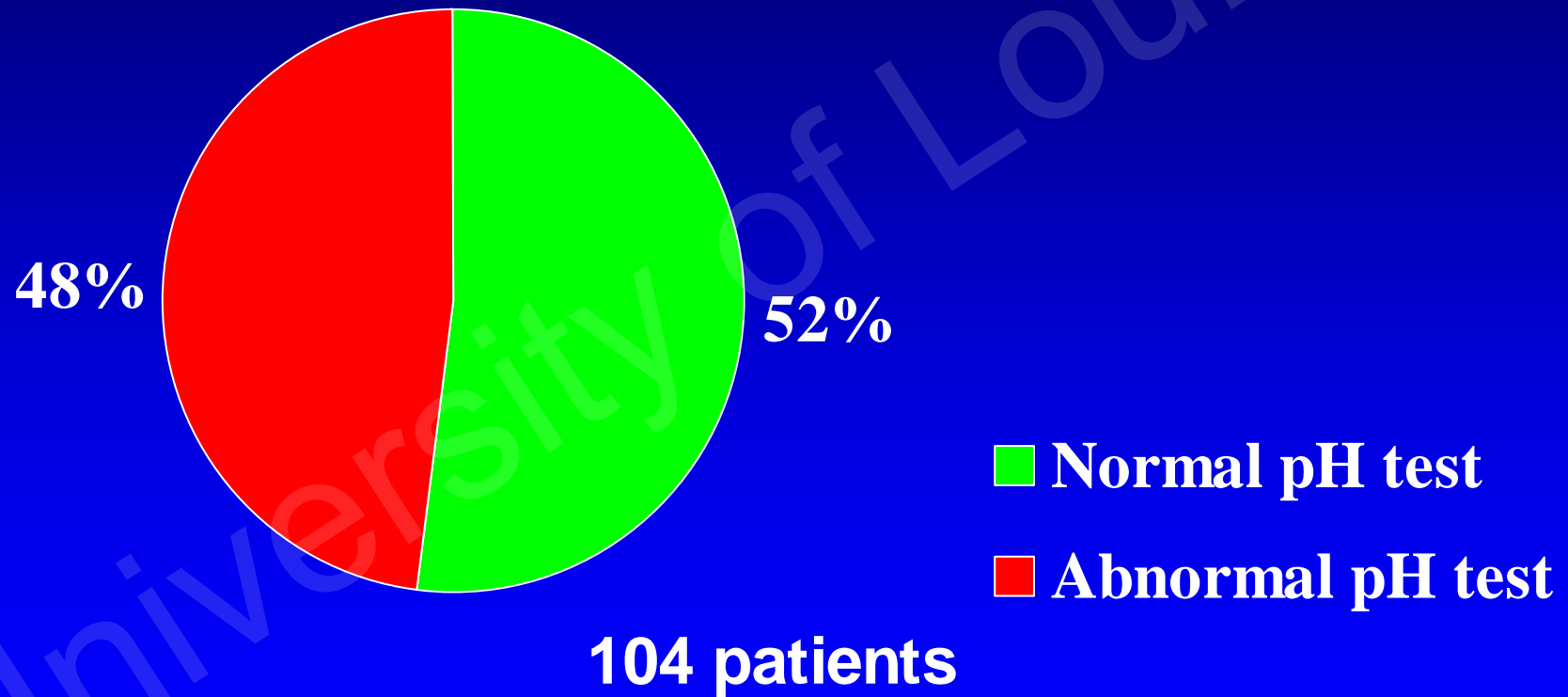
Fass et al. Gastroenterol 1998;115:42.

PPI Test for Non-Cardiac Chest Pain

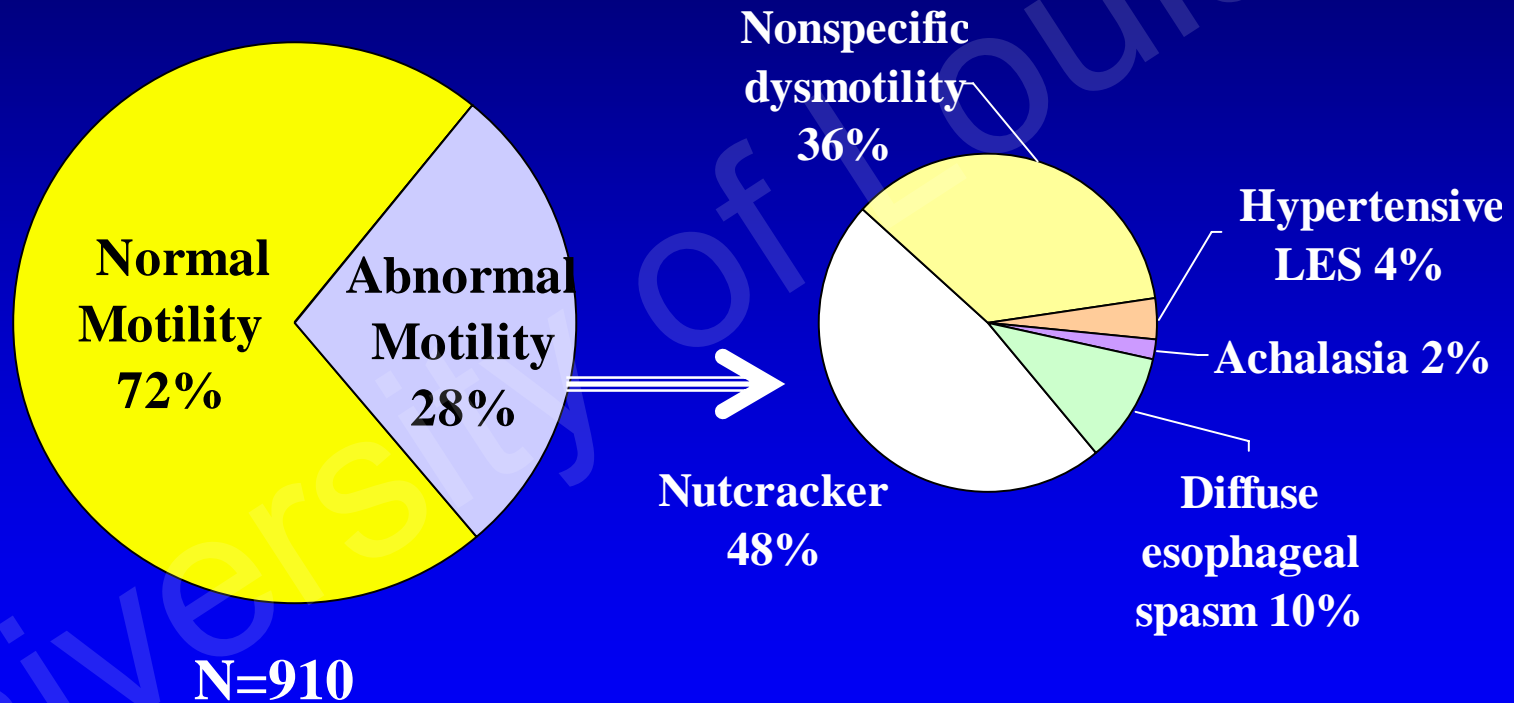
- Computer decision analysis models find that starting with the PPI test reduces the need for diagnostic procedures by 43% - 59%.¹⁻²
- Diagnostic testing should be reserved for non-responders to empiric PPI therapy.

1. Fass et al. Gastroenterol 1998;115:42.
2. Ofman et al. Am J Med 1999;107:219.

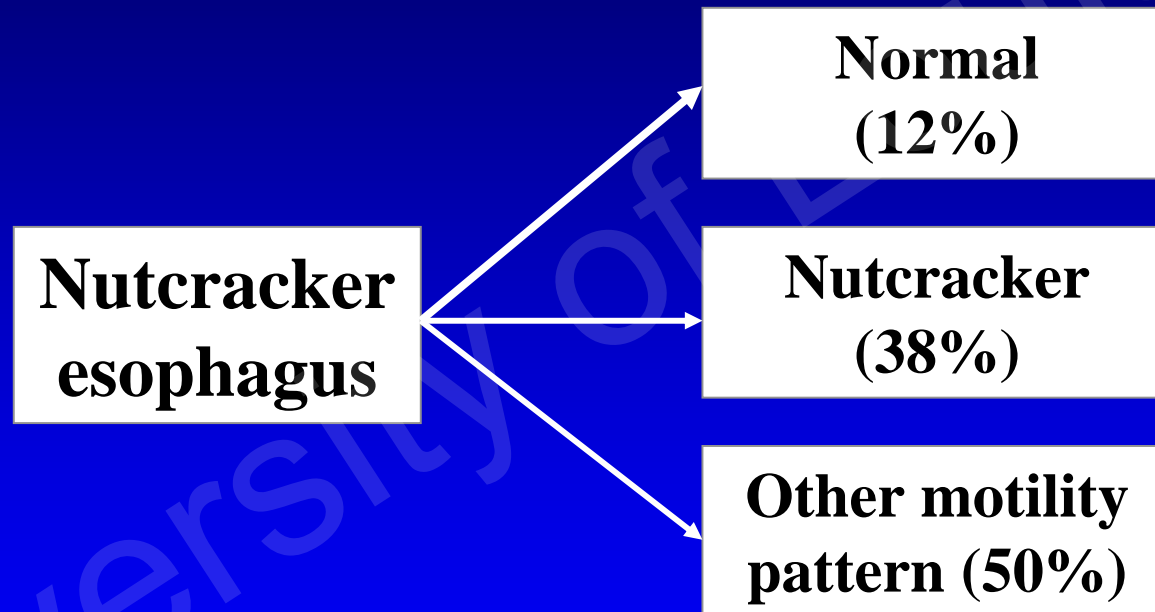
Results of Ambulatory pH Testing in Patients With Non-Cardiac Chest Pain



Esophageal Motility Abnormalities in Patients with Non-Cardiac Chest Pain



Esophageal Spastic Disorder is Intermittent



Dalton et al. Am J Gastroenterol 1988;83:623-28.

Narducci et al. Am J Gastroenterol 1985;80:242-85.

Achem et al. Am J Gastroenterol 1993;84:7-851

Smooth Muscle Relaxant is Ineffective for Esophageal Spastic Disorders

| Study* | Therapy | Motility | Outcome |
|-----------------------|------------|-------------|------------|
| Cattau '91 (n=14) | Diltiazem | ↓ Pressures | Improved |
| Drenth '90 (n=8) | Diltiazem | | No benefit |
| Richter '87 (n=20) | Nifedipine | ↓ Pressures | No benefit |
| Davies '87 (n=8) | Nifedipine | | No benefit |
| Nasarallah '85 (n=20) | Nifedipine | No benefit | Benefit |
| Davies '82 (n=10) | Nifedipine | | No benefit |

*Placebo-controlled cross-over studies

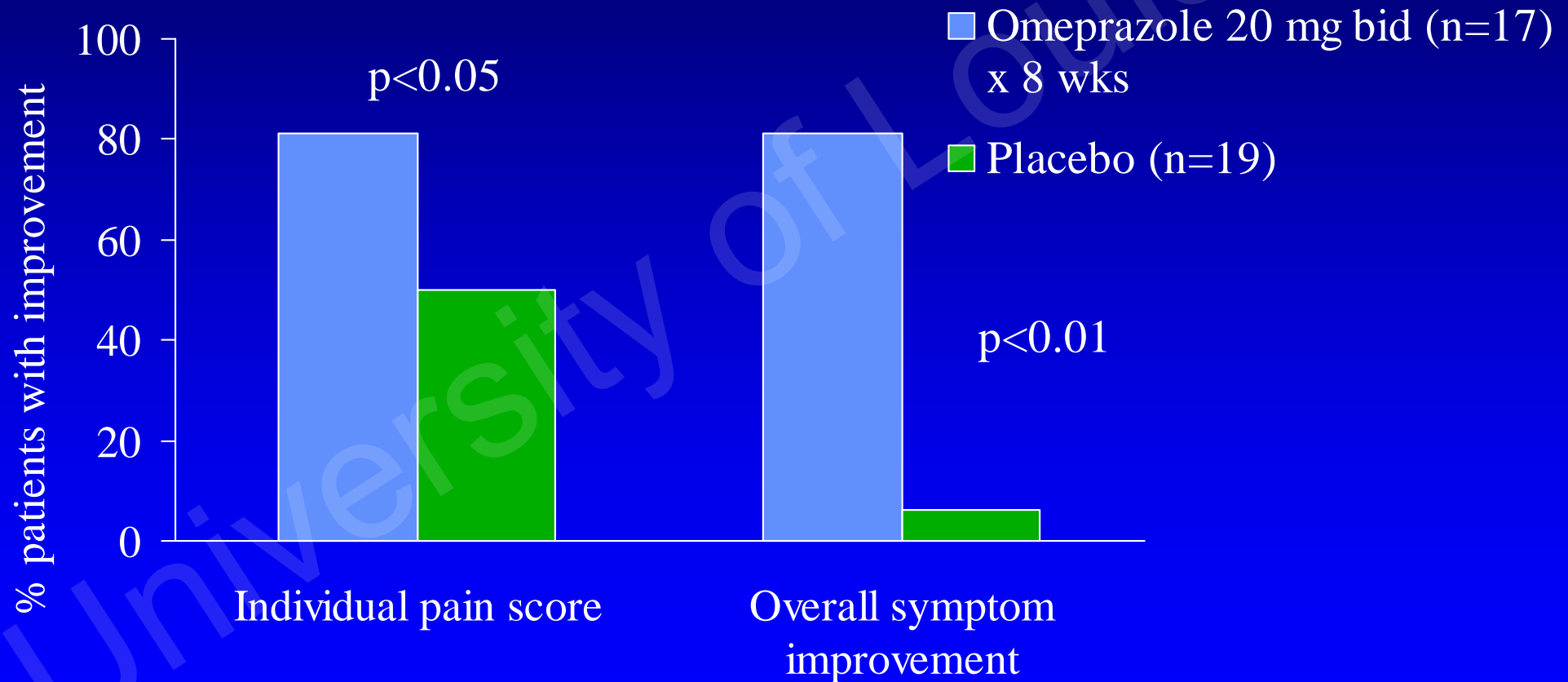
Esophageal Motility Abnormalities are Mostly Non-Specific Phenomena from External Stimuli

- Stress can alter esophageal pressures.¹
- Many patients with hypercontracting esophagus have GERD.²
- Look for secondary causes

¹Anderson KO et al. Dig Dis Sci 1989;34:83-91.

²Achem SR et al. Am J Gastroenterol 1993;88:187-92

PPI Treatment for Non-Cardiac Chest Pain



Patients with chest pain and +pH test

Achem et al. Dig Dis Sci 1997;42:2138.

PPI Treatment for Non-Cardiac Chest Pain

- Empiric treatment with a twice daily PPI for 2 to 3 months is a reasonable approach.
- PPI may also be effective in patients with hypercontracting dysmotility associated with GERD.¹

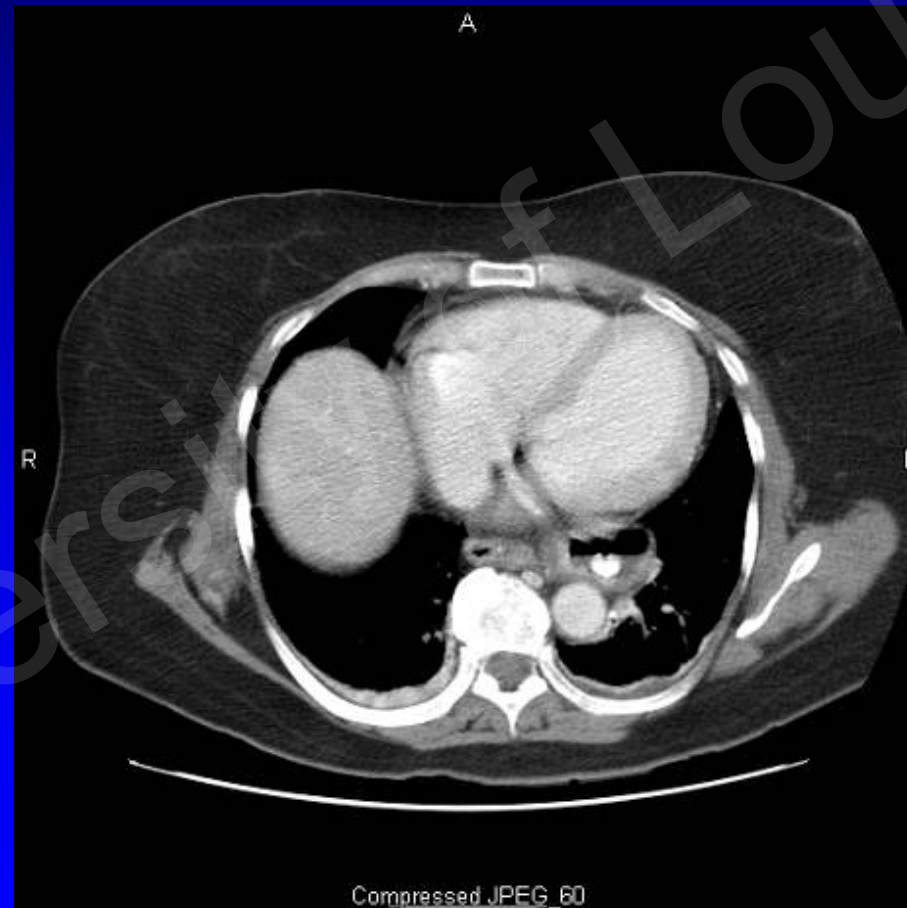
¹Borjesson M et al. Aliment Pharmacol Ther 2003;18:1129-35.

Difficult Cases

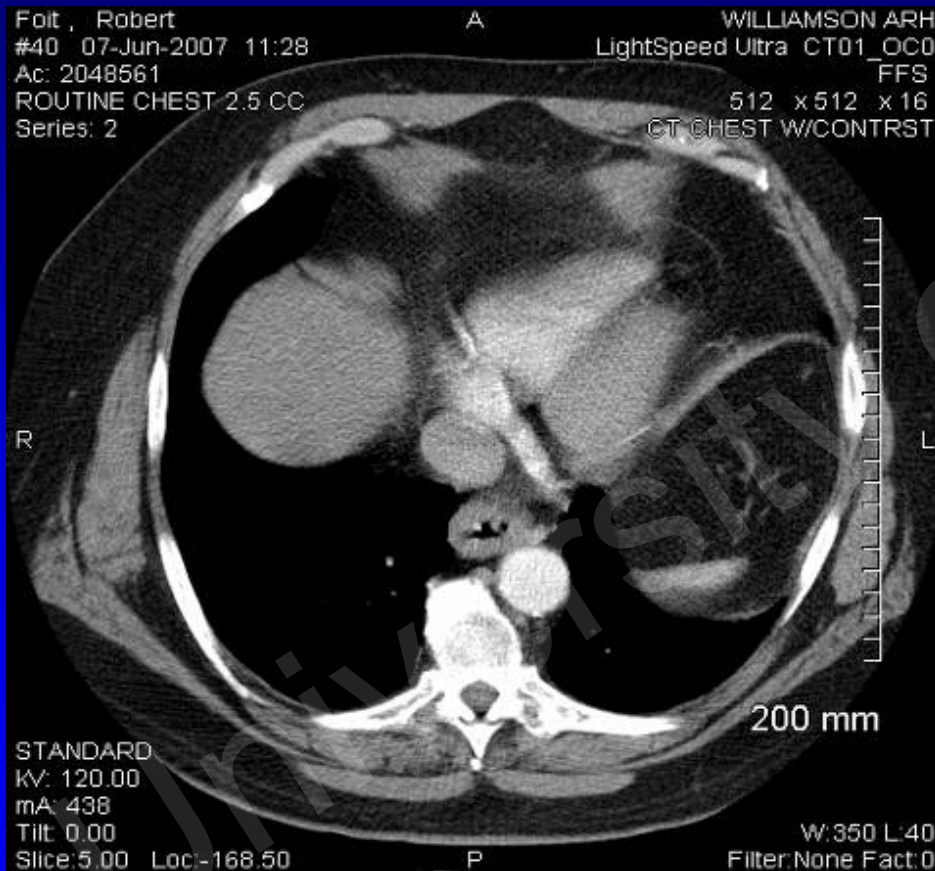
A Patient with Intermittent Dysphagia



A Patient with Intermittent Dysphagia (Cont.)



A Patient with Hypertensive LES

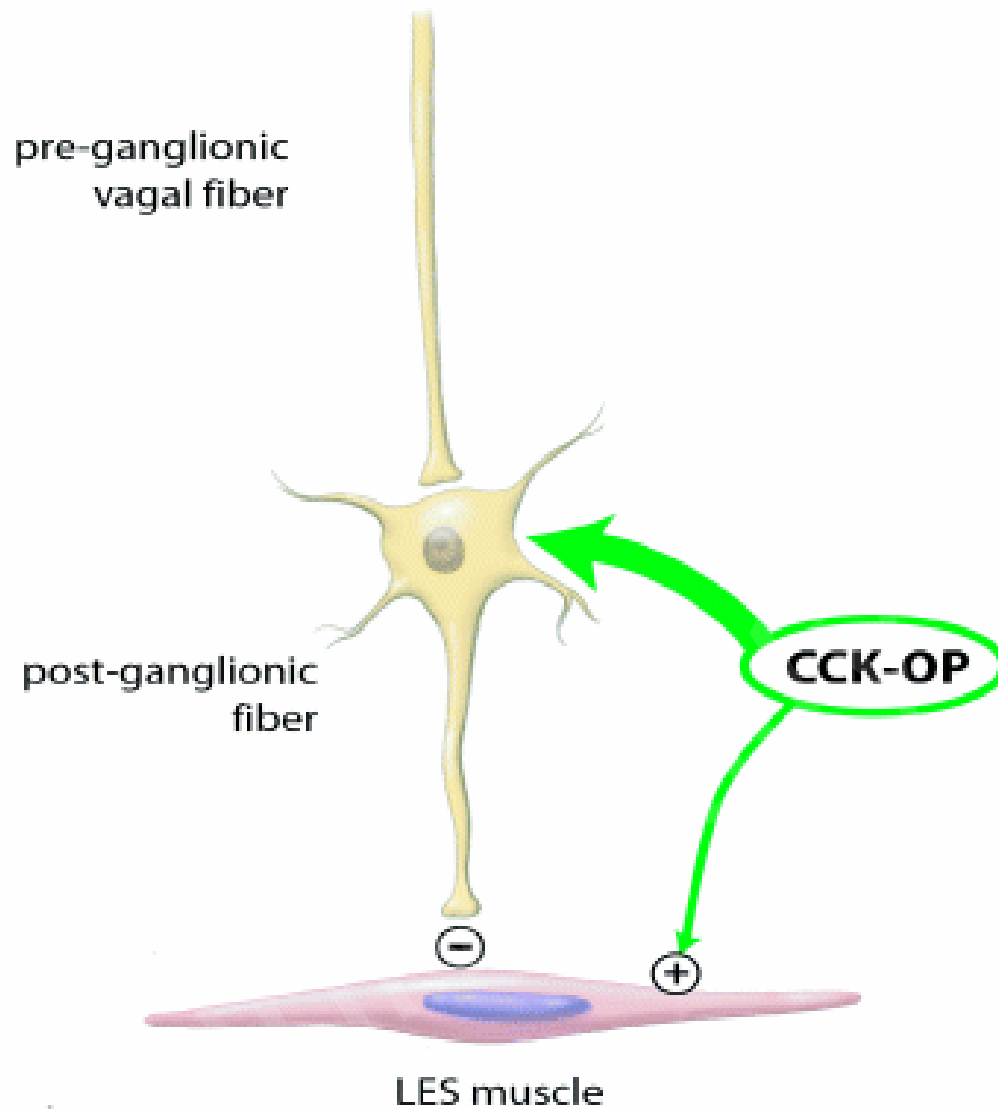


Hypocontracting Esophagus: Aperistalsis and Impaired Esophageal Peristalsis

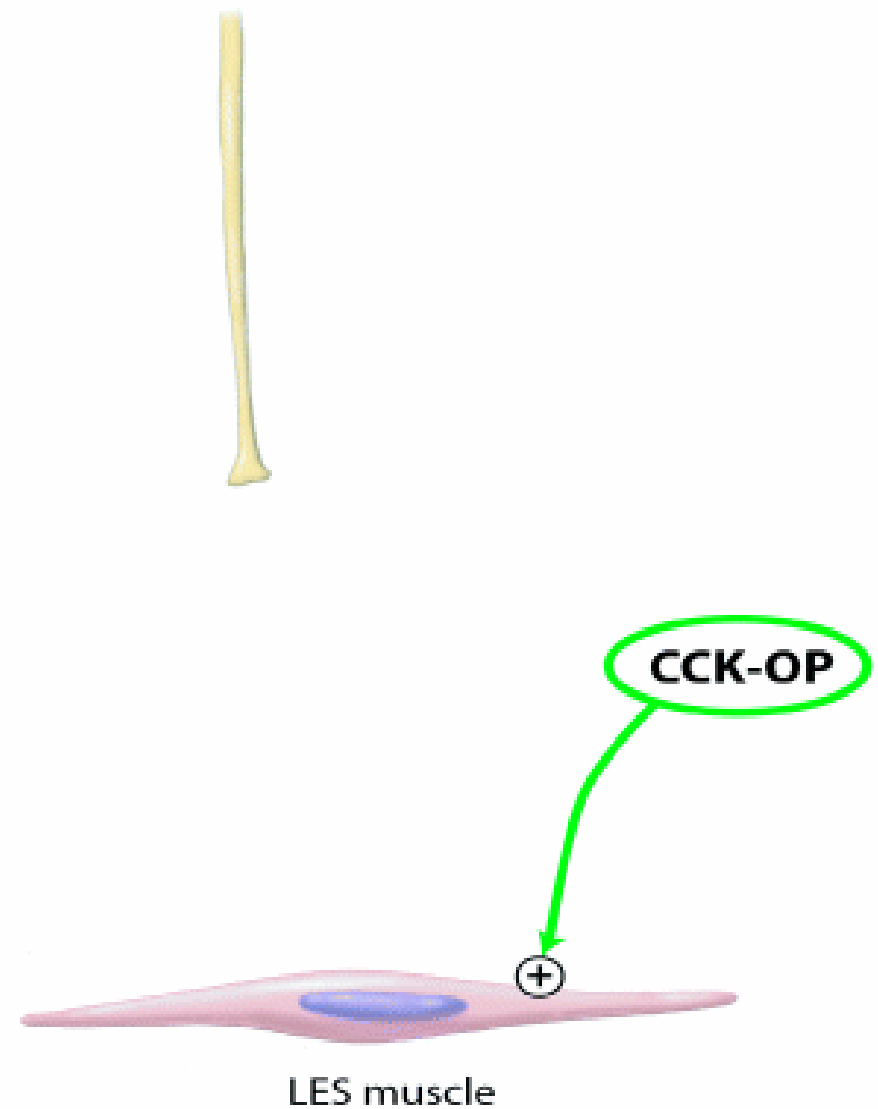
Esophageal Aperistalsis

- Primary aperistalsis (achalasia)
- Secondary aperistalsis
 - Connective tissue diseases
 - Chagas disease
 - Paraneoplastic syndrome
 - Post-fundoplication
 - Vagal trauma
 - Severe GERD

A. NORMAL PATIENT



B. ACHALASIA PATIENT



Primary Achalasia

- Decrease # of inhibiting neurons in the LES
- Patients can be young or old
- Etiology is still unclear
 - Inflammatory response and infection likely
- Chronic progression of symptoms
- Presentation can be subtle in early achalasia

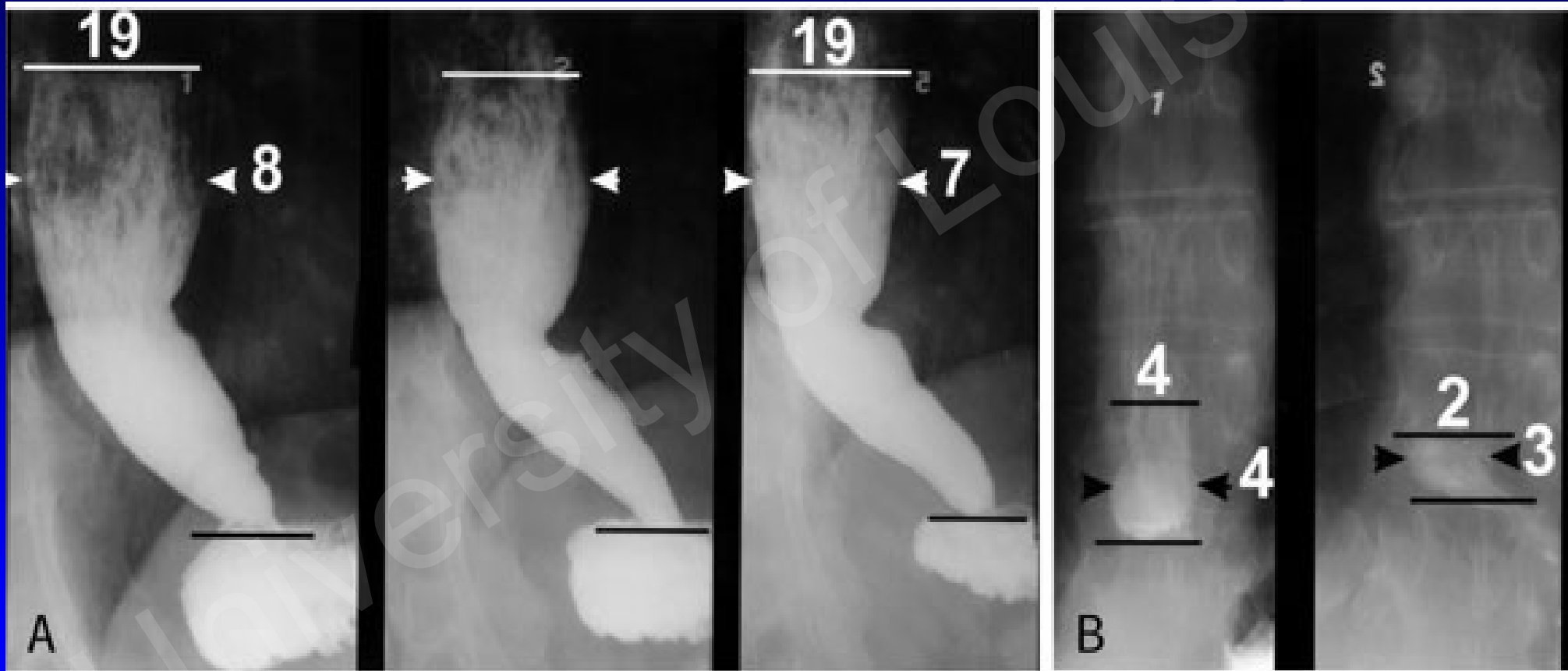
Symptoms of Achalasia Can be Diverse

- Chronic dysphagia to liquids and solids
- Nocturnal regurgitation
- Chest pain
- Heartburn
- Weight loss
- Aspiration/choking

Achalasia



Timed Barium Esophagram



Kostic et al. J Thorac CV Surg
200-;120:935.

Swallow 100-250cc of 45% barium over 30-45
seconds. Take pictures at 1, 2 and 5 minutes

STUDY DETAILS

EQUIPMENT

CAPTURE

REVIEW

LES

BODY

UES

REPORT

PROTOCOL

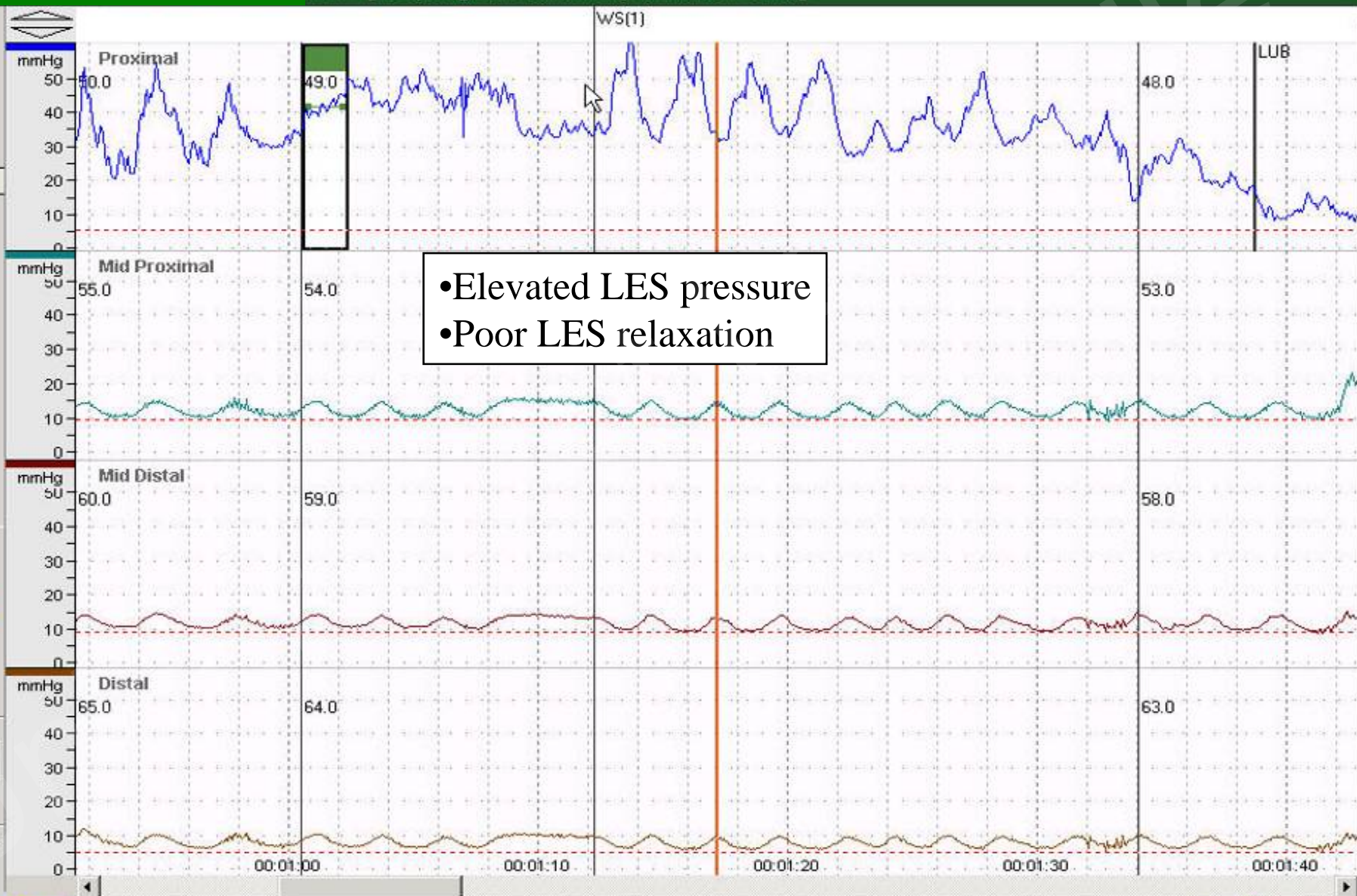
Markers

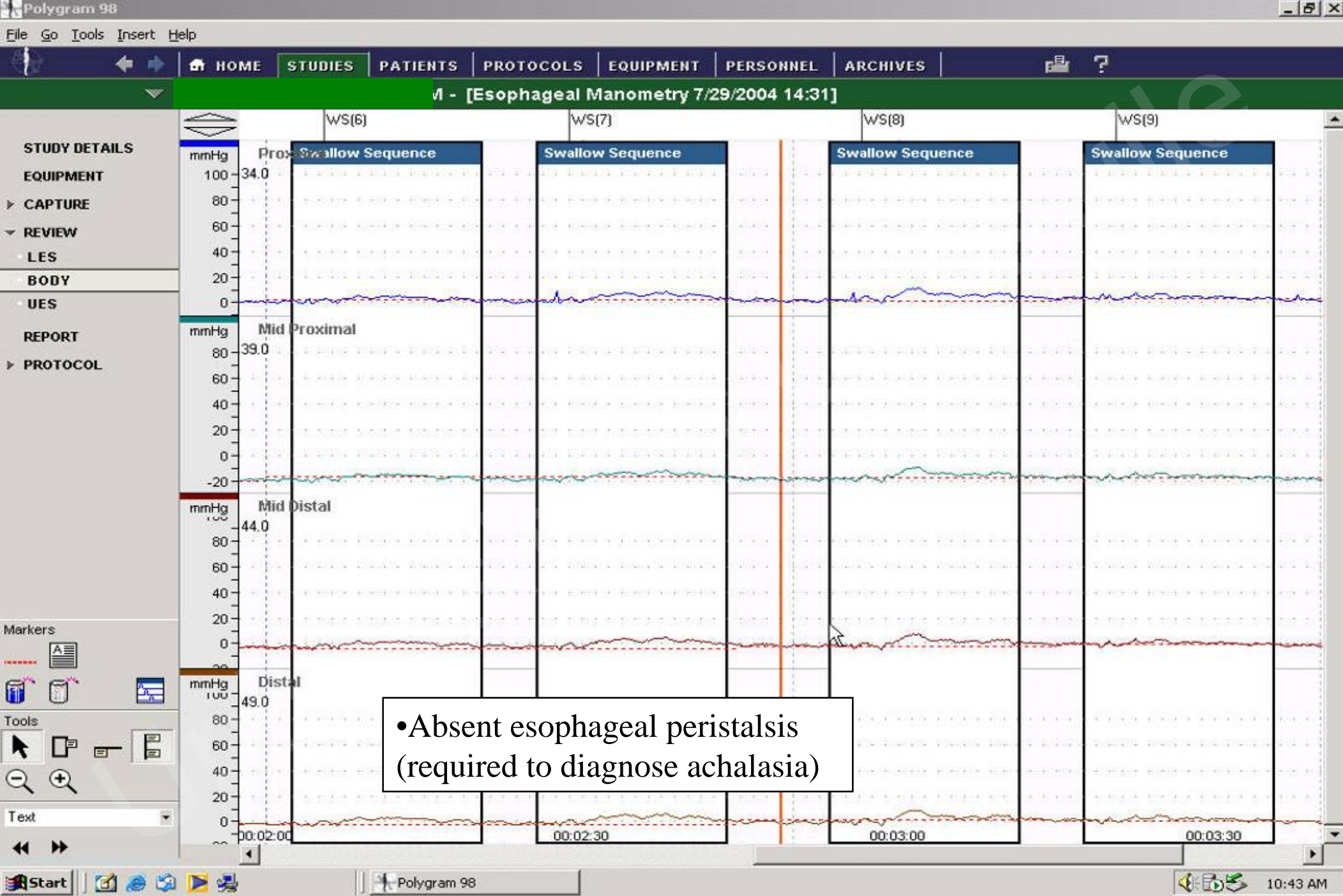


Tools

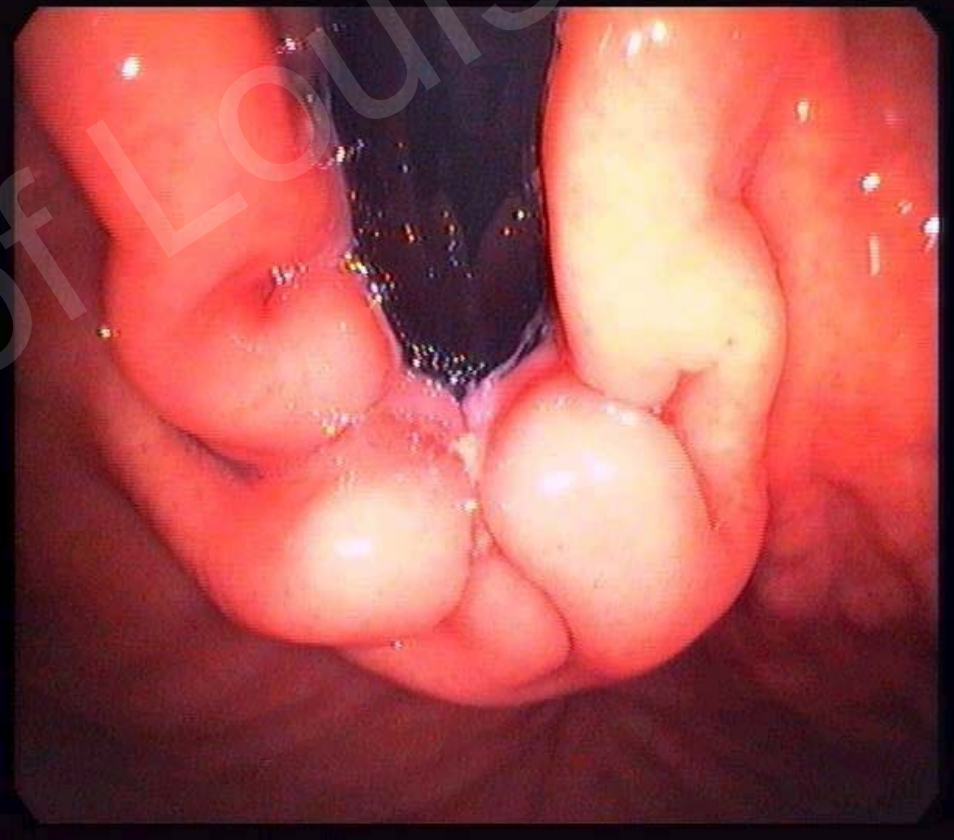


Text





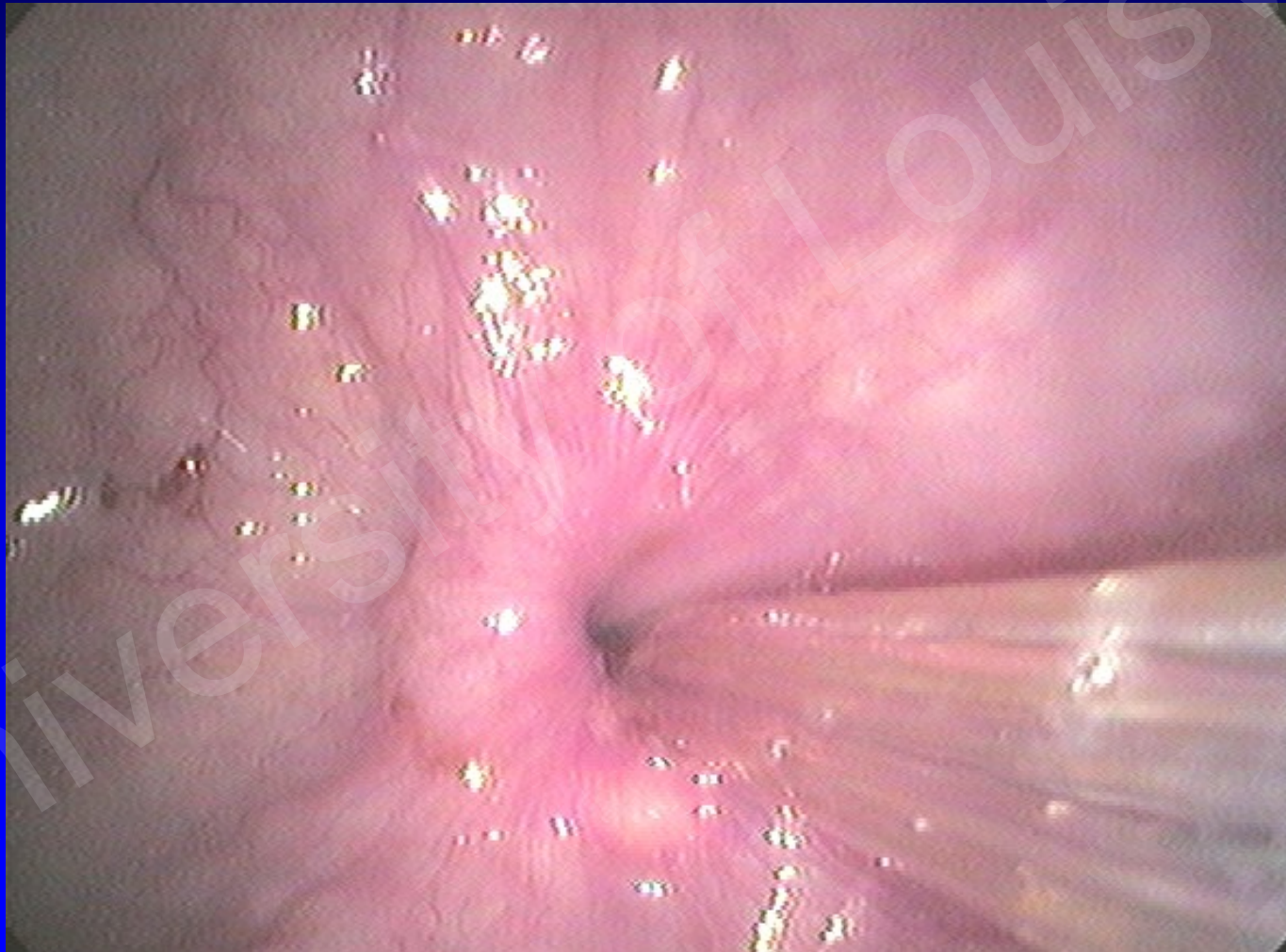
Achalasia



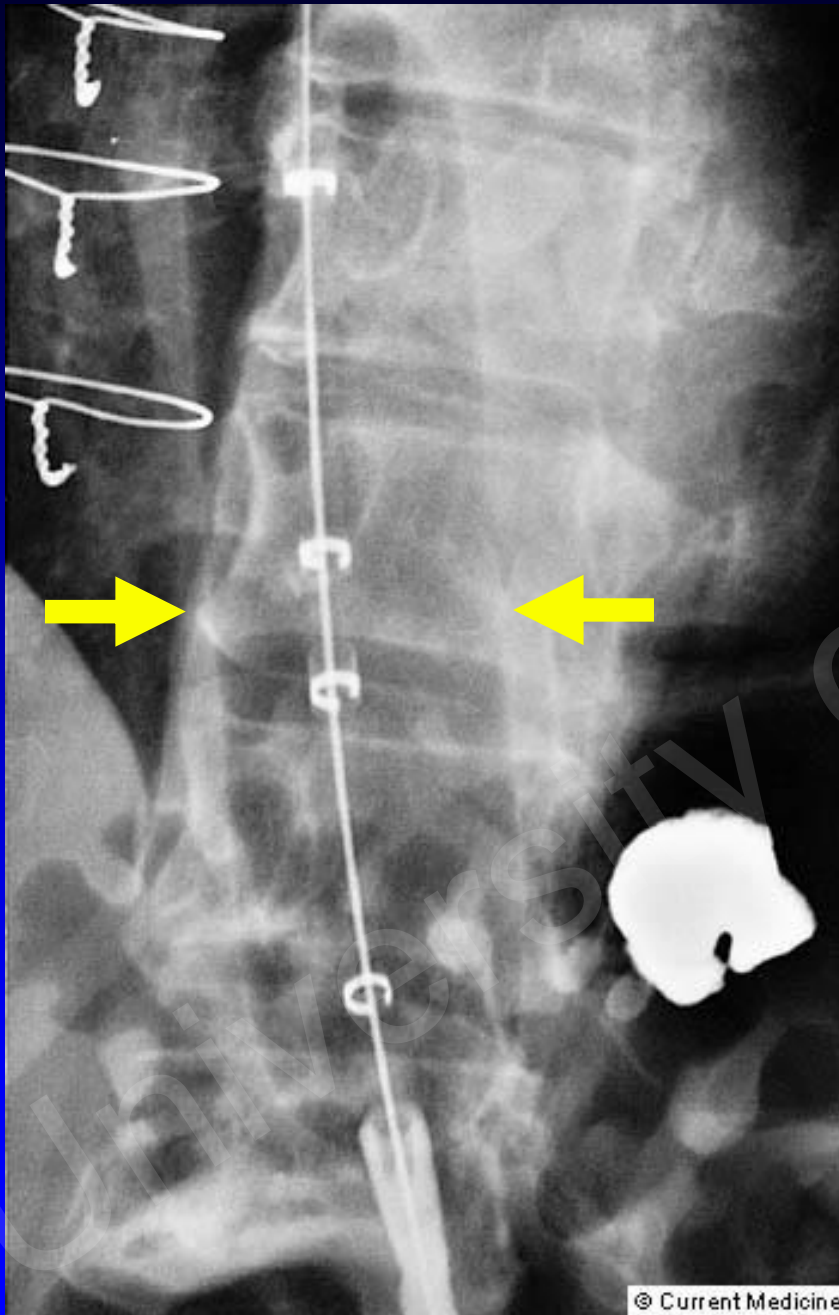
Treatment Should be Individualized in Primary Achalasia

- Nitrates and calcium channel blockers
 - Benefit is short term
- Endoscopic botox injection
 - Symptoms always recur
 - Useful for elderly patients or poor surgical candidates
- Endoscopic pneumatic dilation
- Laparoscopic Heller myotomy

Achalasia: Pneumatic Balloon Dilation



Achalasia: Pneumatic Balloon Dilation



Obliterate the “waist”
created by the LES

Pneumatic Dilation in Achalasia

- Goal: rupture the LES
 - Gastrograffin & barium swallow after dilation
- Success
 - 65-80%
- Perforation
 - 2 to 15% (depends on balloon size)

Impaired Esophageal Peristalsis

| | |
|---------------------------|-----------|
| Mean distal peristaltic P | < 30 mmHg |
| or | |
| Peristaltic waves | < 60% |

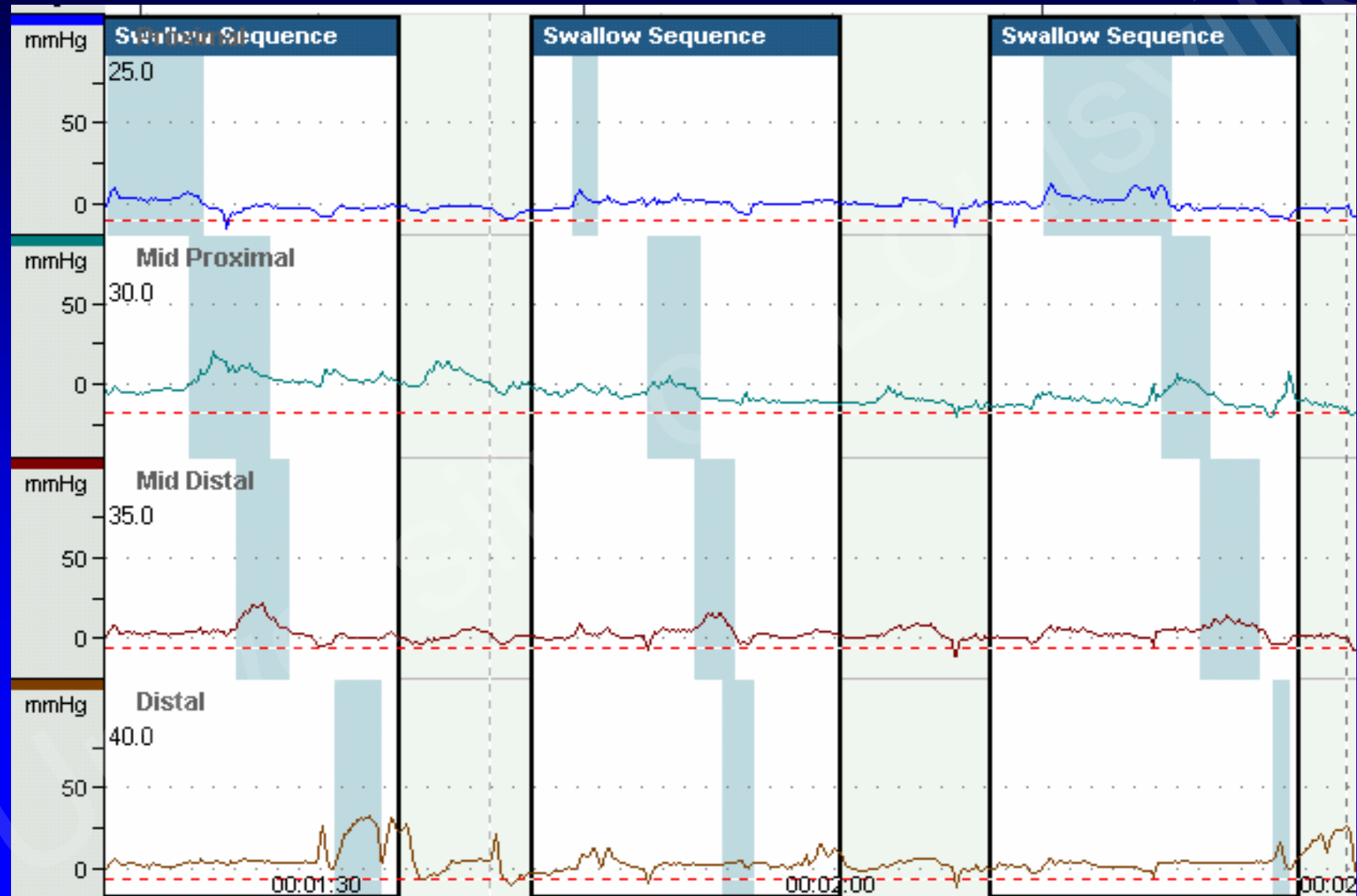
Underlying Causes of Secondary Achalasia and Hypocontracting Esophagus

- GERD
- Connective tissue diseases
 - Systemic sclerosis
 - Mixed connective tissue disease
 - Idiopathic inflammatory myopathy, lupus, Sjogren's
- Endocrine diseases
 - Diabetes
- Neuromuscular diseases
 - Chagas disease
 - Amyloidosis
 - Paraneoplastic syndrome
 - Autonomic neuropathy

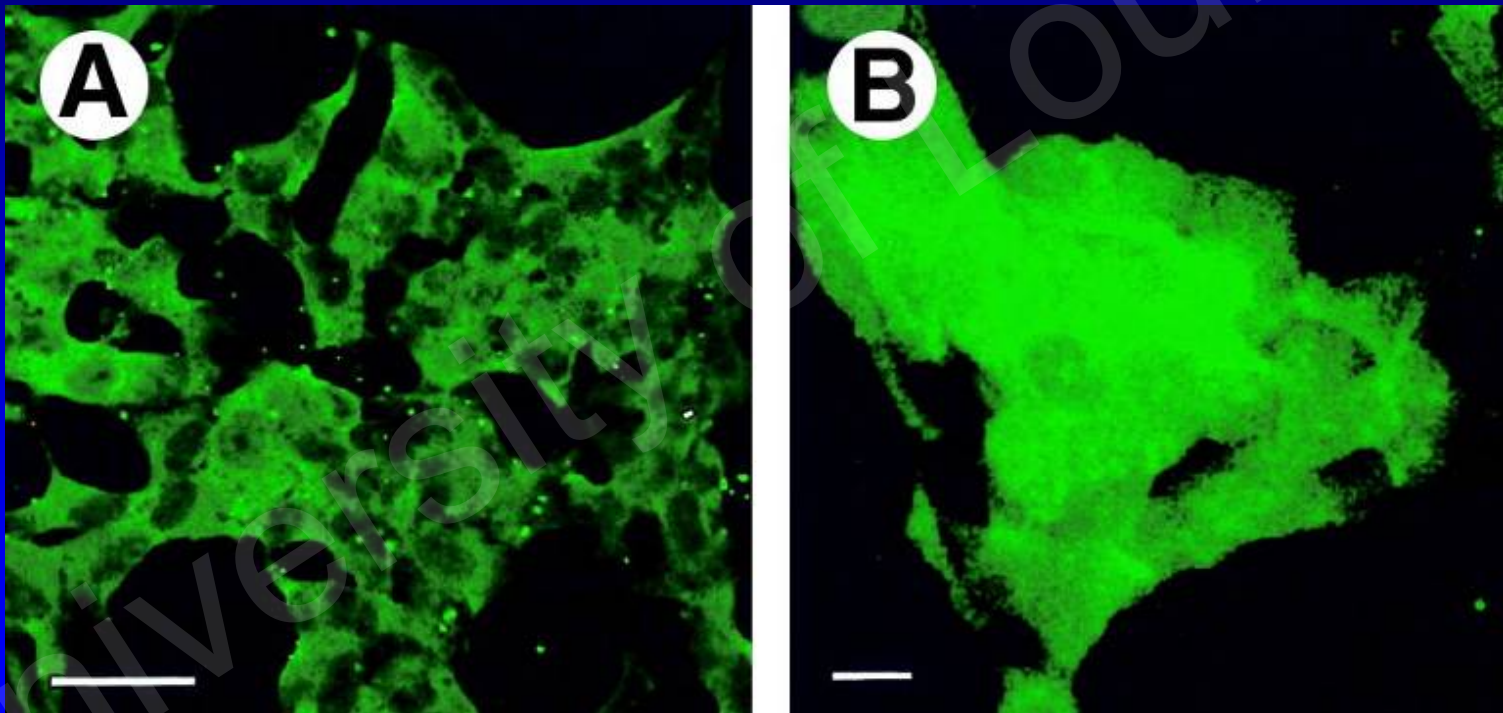
Systemic Sclerosis

- Early stage
 - Neural dysfunction, ?Vascular insufficiency
 - Esophagus response to edrophonium
- Late stage
 - Neural and muscular dysfunction
 - Smooth muscle fibrosis
 - Poor response to methacholine
- Acid reflux is associated with impaired esophageal motility

Systemic Sclerosis



Paraneoplastic GI Motility Syndrome: Anti-Hu Antibody* Against Enteric Neurons



*Antinuclear neuronal antibodies (ANNA)

Paraneoplastic GI Motility Syndrome

- Cancer antigens mimicking neuronal tissues.
- Myenteric plexus infiltrated by lymphocytes and plasma cells.
- Cancers
 - Small cell lung cancer (80%), breast, ovarian, multiple myeloma, Hodgkin's lymphoma.
- GI symptoms can precede diagnosis of cancer.

Summary:

Esophageal Motility Disorders

- Hyper vs. Hypocontracting esophagus
- Hypercontracting (esophageal spastic) disorders represent a dysfunction rather than the cause
- Look for underlying cause
 - GERD, systemic diseases, diffuse motility disorder, paraneoplastic, etc.