

# ACHALASIA

BIBLE CLASS 27/01/2021

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## ACHALASIA: DEFINITION

What is the definition of achalasia?

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Achalasia is a disorder characterised by insufficient LOS relaxation and absent peristalsis.

It is usually primary (idiopathic) but can be secondary to other conditions that affect oesophageal function.

In idiopathic achalasia, the enteric neurons controlling the LOS and oesophageal body musculature are affected by an unknown cause, most likely inflammatory.

Oude Nijhuis RAB, Zaninotto G, Roman S, et al. European Guideline on achalasia. UEG and ESNM recommendations. *United European Gastroenterol J* 2020.

# ACHALASIA: EPIDEMIOLOGY

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Reported global incidence from 0.03 to 1.63 per 100,000 persons per year

Prevalence ranging and 1.8 to 12.6 per 100,000 persons

It occurs equally in men and women, with no racial predilection.

The peak incidence occurs between 30 and 60 years of age.

Vaezi MF, Pandolfino JE, Yadlapati RH, Greer KB, Kavitt RT. ACG Clinical Guidelines: Diagnosis and Management of Achalasia. Am J Gastroenterol. 2020 Sep;115(9):1393-1411

WHEN DO YOU SUSPECT ACHALASIA?

## WHEN DO YOU SUSPECT ACHALASIA?

Dysphagia for both solids and liquids

Regurgitation of bland undigested food and saliva (mucus)

Substernal chest pain

Weight loss

Heartburn (27-42%, non-responsive to PPI)

# DIFFERENTIAL DIAGNOSIS



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Gastroesophageal reflux disease (PPI trial)

Eosinophilic esophagitis

Pseudo-achalasia

- Neoplastic infiltration of the cardia (adenocarcinoma of gastroesophageal junction, pancreatic, breast, lung, or hepatocellular cancer)
- Paraneoplastic (small cell bronchial cancer)

Previous surgery (overly tight fundoplication, gastric banding)

Chagas-Disease (Trypanosoma cruzi-infection)

- Endemic in Latin America, megacolon?, heart disease?, neurologic abnormalities?

# DIAGNOSTIC PROCEDURES

## DIAGNOSTIC PROCEDURES

High Resolution Manometry

Radiology: Timed barium swallow

Endoscopy

Impedance planimetry (EndoFLIP)

CT, EUS

# ENDOSCOPY

- Normal findings in 1/3 of patients
  - Passage of cardia against soft resistance
  - Food remnants
  - Dilated esophagus
  - Candidiasis
- (a) We suggest against making the diagnosis of achalasia solely based on endoscopy.  
Expert opinion – 100%
- (b) We suggest performing endoscopy in all patients with symptoms suggestive of achalasia to exclude other diseases.  
Expert opinion – 77.8%

Oude Nijhuis RAB, Zaninotto G, Roman S, et al. European Guideline on achalasia. UEG and ESNM recommendations. *United European Gastroenterol J* 2020

# HIGH RESOLUTION MANOMETRY (HRM)

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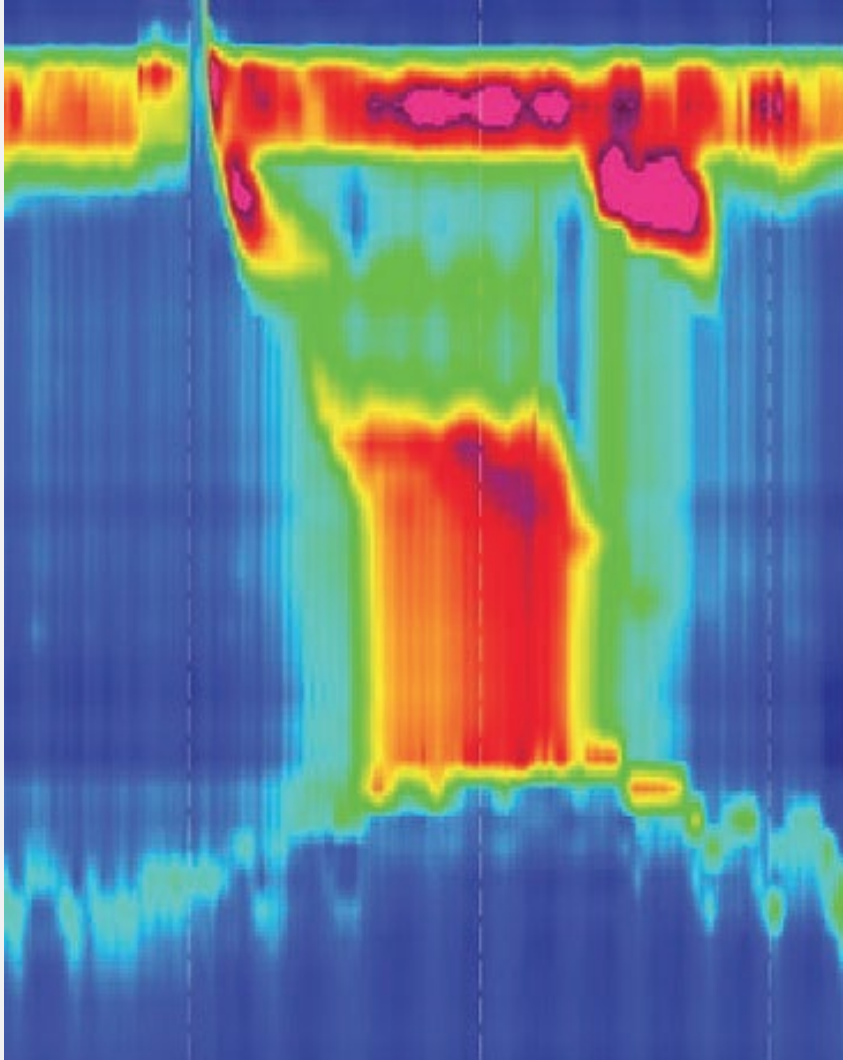
The diagnosis of achalasia requires not only impaired oesophago-gastric junction (OGJ) relaxation, but also absent or abnormal peristalsis.

Therefore, oesophageal manometry is considered as being the gold standard for the diagnosis of achalasia, as it evaluates both pressures of the LOS and contractility of the oesophageal body.

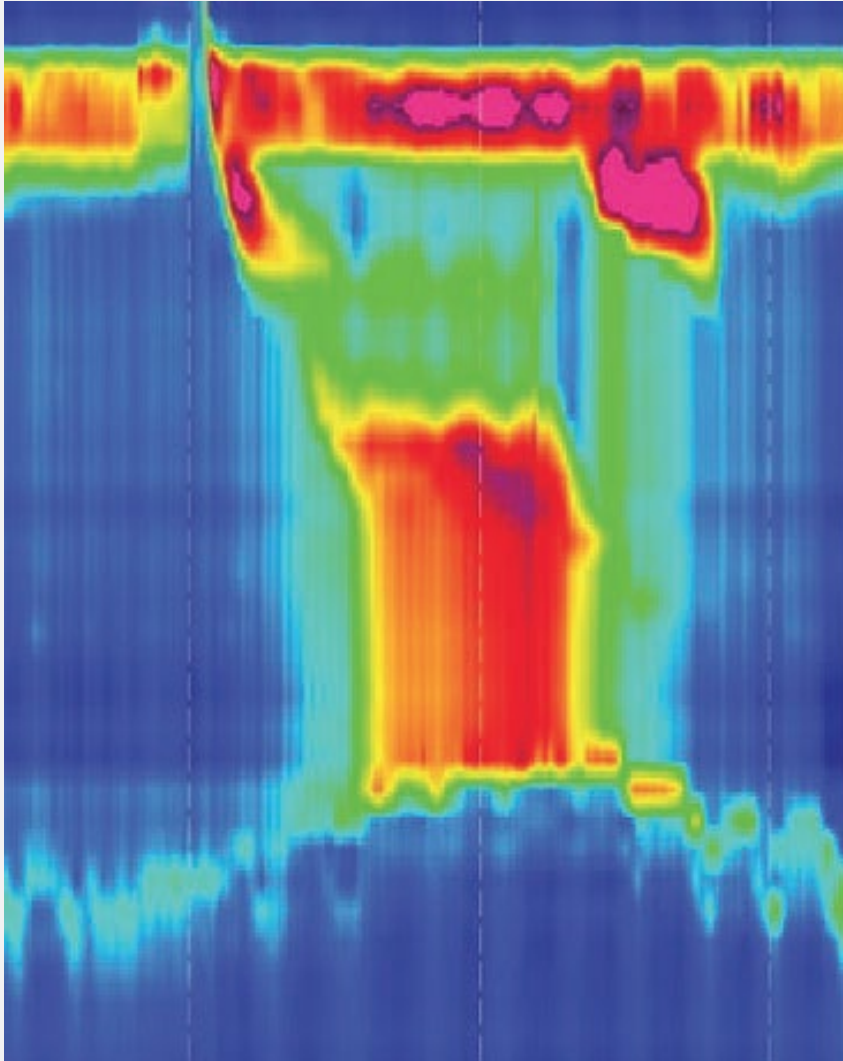
HRM (>21 pressure sensors at 1 cm intervals) vs. Conventional manometry (pressure sensors at intervals 3-5 cm):

Sensitivity (93% vs. 78%), specificities of both tests equal (100%)

# HIGH RESOLUTION MANOMETRY (HRM)



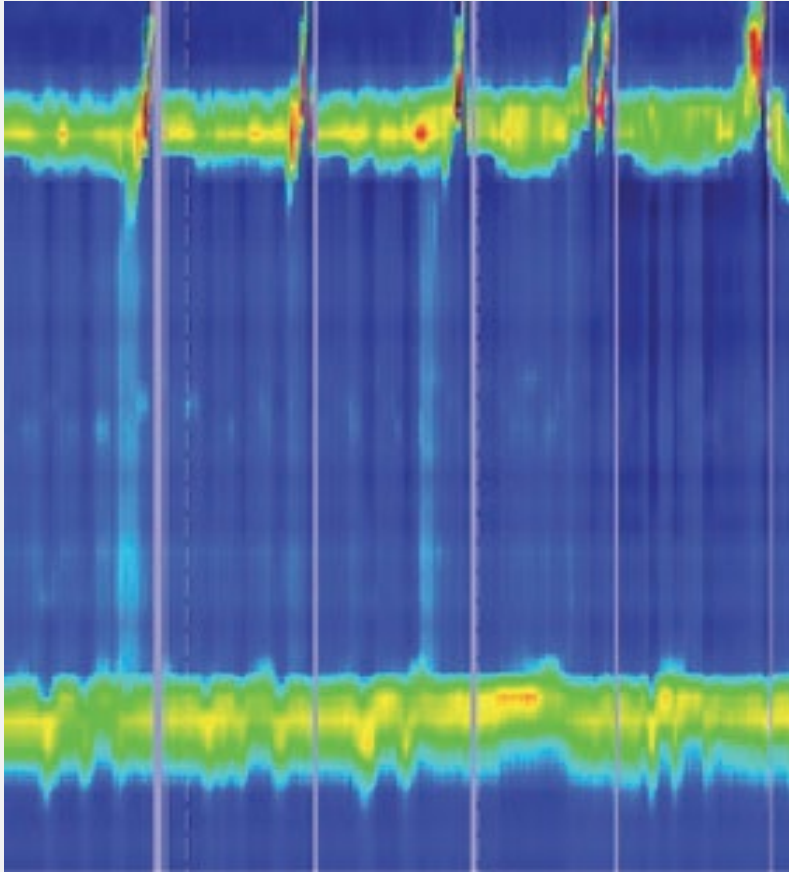
# HIGH RESOLUTION MANOMETRY (HRM)



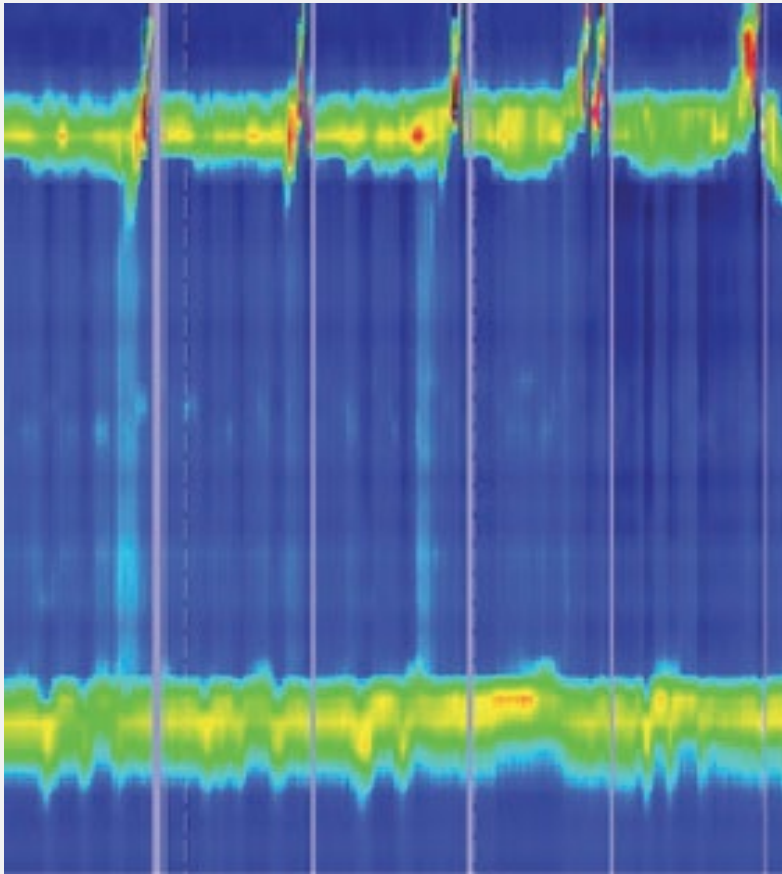
Median IRP > cut-off  
No normal peristalsis  
20% premature contraction with DCI > 450  
Type III: Spastic achalasia



# HIGH RESOLUTION MANOMETRY (HRM)

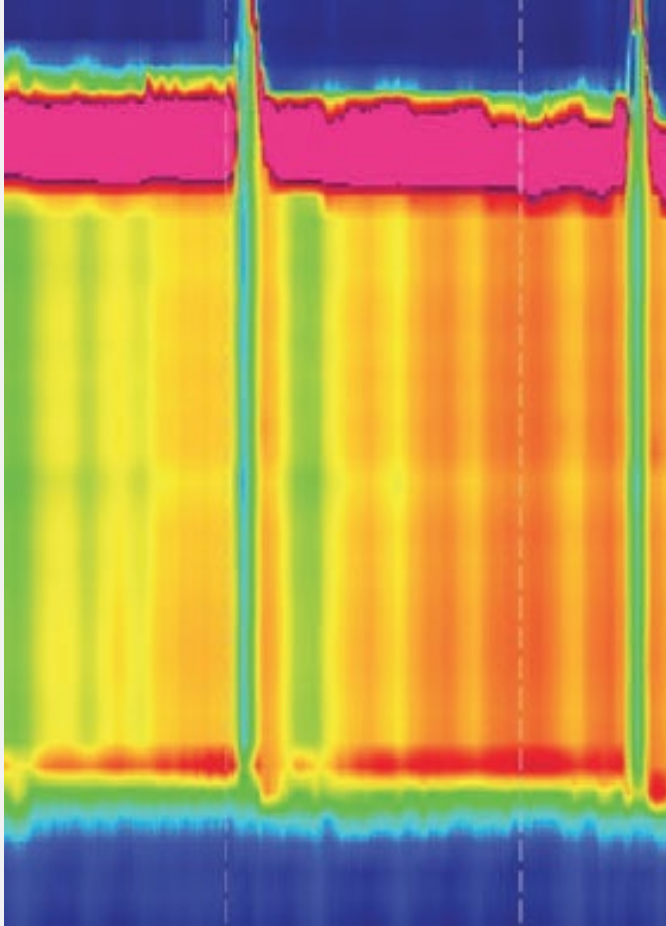


# HIGH RESOLUTION MANOMETRY (HRM)

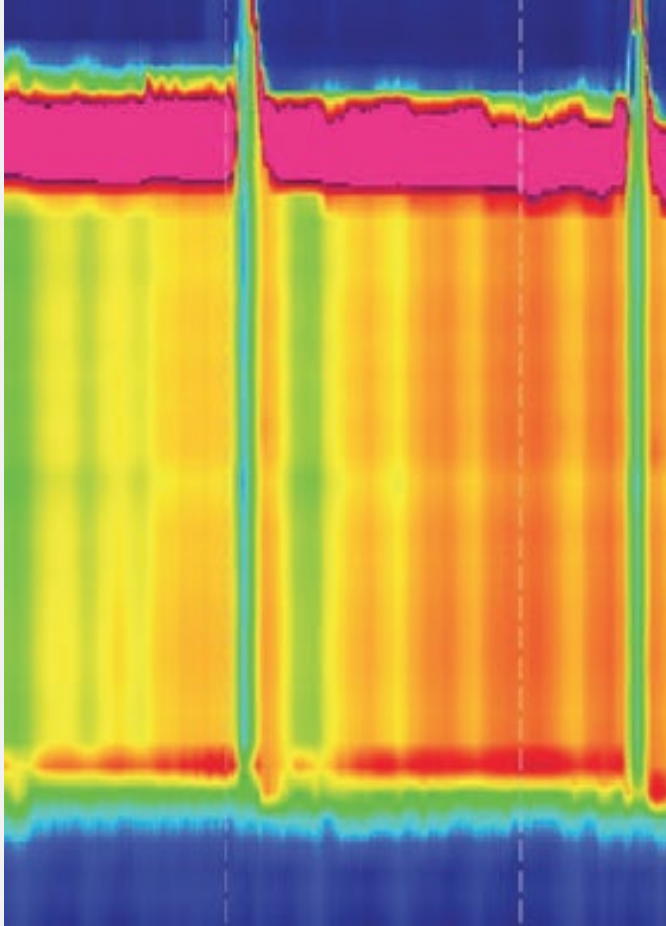


Median IRP > cut-off  
100% failed peristalsis  
Type I Classic achalasia

# HIGH RESOLUTION MANOMETRY (HRM)



# HIGH RESOLUTION MANOMETRY (HRM)



Median IRP > cut-off  
With oesophageal compression  
100% failed peristalsis  
20% pan-oesophageal pressurisation  
Type II Achalasia

# HR-MANOMETRY

Elevated Integrated Relaxation Pressure (IRP),

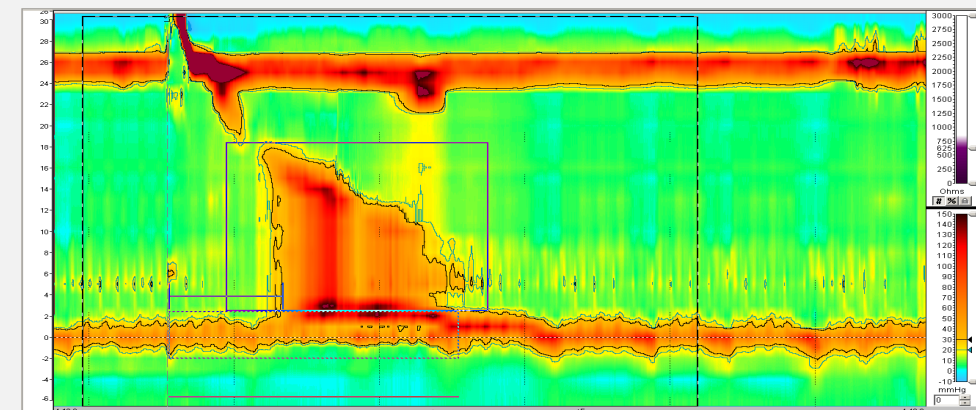
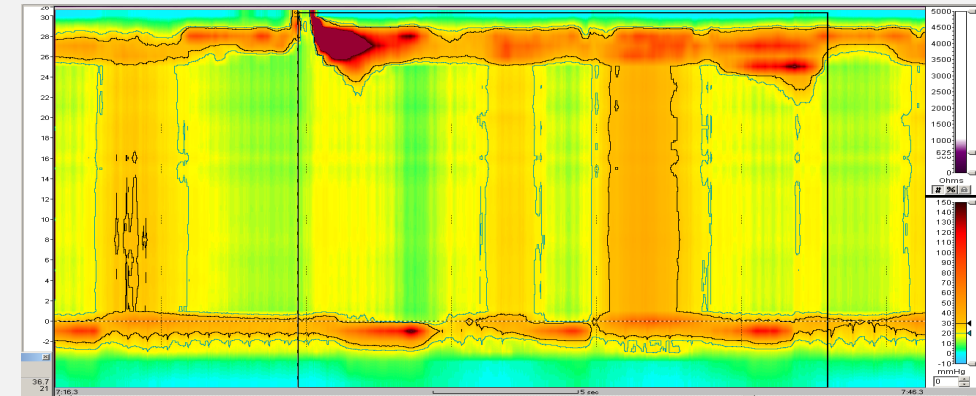
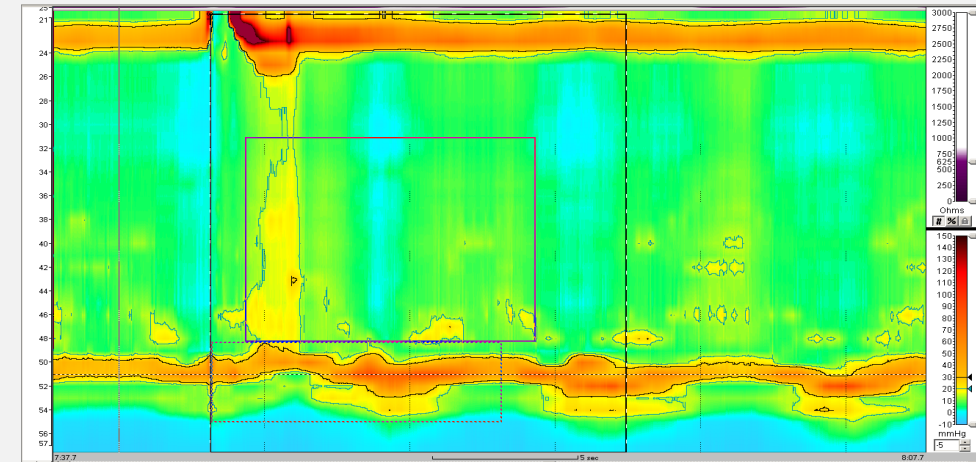
no normal peristalsis

Elevated LES resting pressure, esophageal pressure

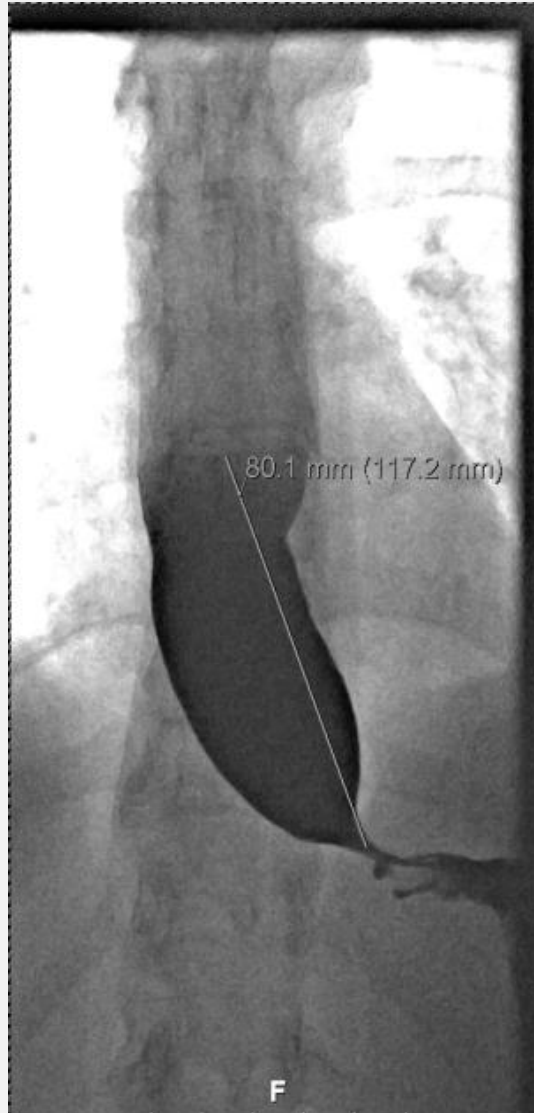
Typ I: - no panesophageal pressurisation

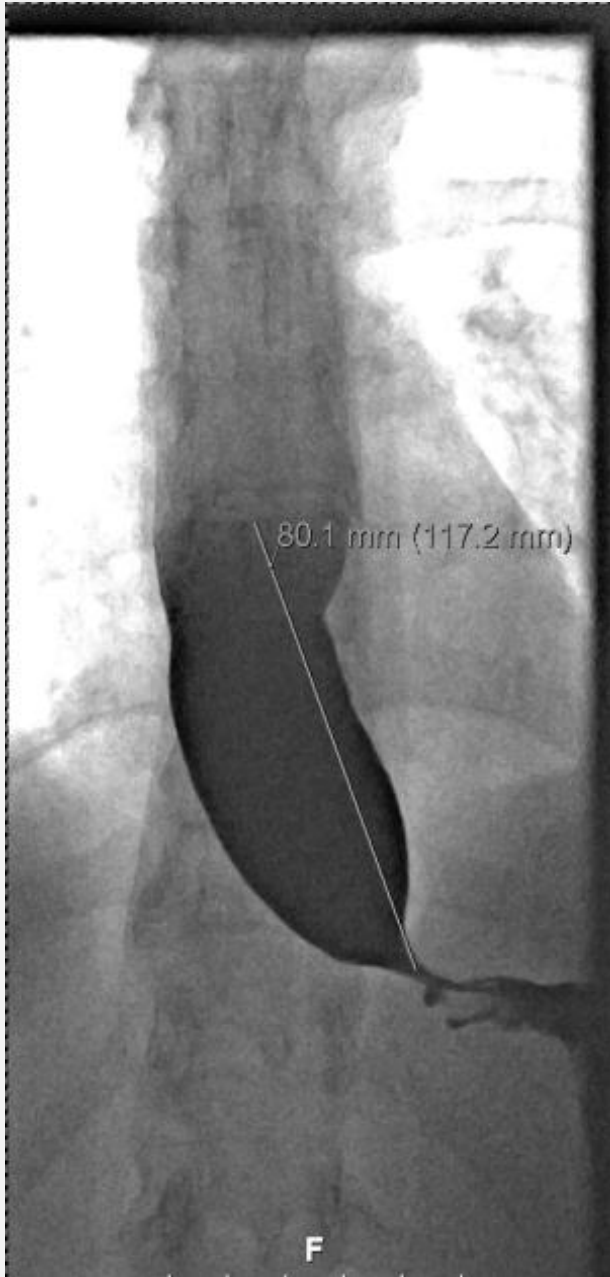
Typ II: - panesophageal pressurisation  
>30 mmHg in  $\geq 20\%$  of swallows

Typ III: - spastic, premature contractions  
in  $\geq 20\%$  of swallows



# RADIOLOGY: TIMED BARIUM ESOPHAGRAM





## TIMED BARIUM SWALLOW

- „Bird beak“
- Dilated esophagus
- Retention of contrast media

(200 ml diluted barium suspension, height and width of contrast media column after 1, 2 and 5 minutes)

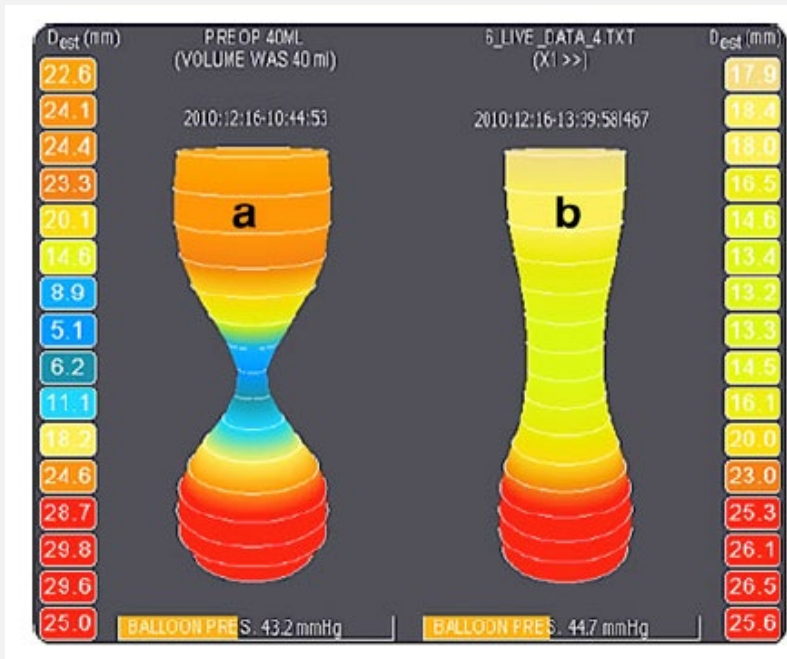
We suggest using a barium oesophagram to diagnose achalasia if manometry is unavailable, although it is less sensitive than oesophageal manometry.

The working group suggests using TBO, if available, over standard barium oesophagram

# IMPEDANCE PLANIMETRY



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Oesophageal impedance planimetry (EndoFLIP) is a technique in which the impedance measurements of cross-sectional area and intrabag pressure is simultaneously measured to evaluate distensibility.

OGJ distensibility in untreated achalasia patients is lower in untreated achalasia than in healthy controls ( $<1.6 \text{ mm}^2/\text{mmHg}$  vs.  $>2.7 \text{ mm}^2/\text{mmHg}$  at 40 ml balloon filling volume)

Limited information about esophageal peristalsis

We suggest against making the diagnosis of achalasia solely based on impaired OGJ distensibility as measured with impedance planimetry.

## ADDITIONAL TESTING

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We suggest additional testing using CT or endoscopic ultrasound only in those achalasia patients suspected of malignant pseudoachalasia.

Multiple recognised risk factors for malignant pseudoachalasia, for example >55 years of age, duration of symptoms, <12 months, weight loss >10 kg, severe difficulty passing the LOS with a scope, may prompt further imaging.

Conditional recommendation, low certainty of evidence

Consensus: 100% agree [Vote: A.., 66.7%; A22.2%; A, 11.1%; D 0%; D., 0%; D.., 0%]

# THERAPY OF ACHALASIA

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Goal: Symptom relief

Reduction of outflow obstruction at the level of the LES

Pharmakologic

Nitrates, Ca<sup>++</sup>-channel-inhibitors (Nifedipine)

Botulinum-toxin

Endoscopic

Dilatation

Temporary stenting

POEM = PerOral Endoscopic Myotomy

Surgical

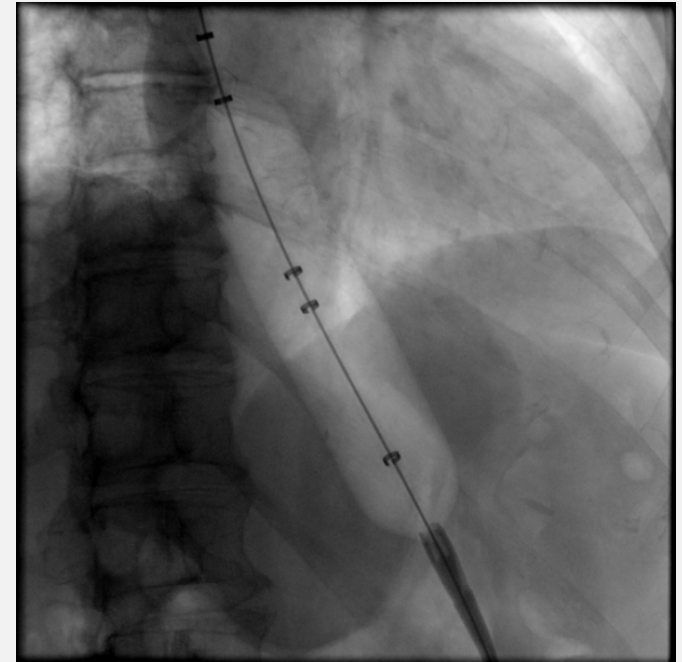
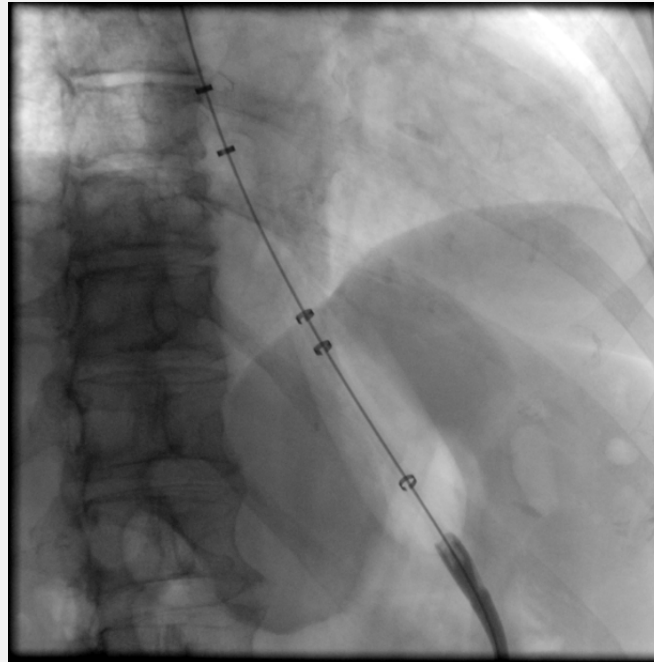
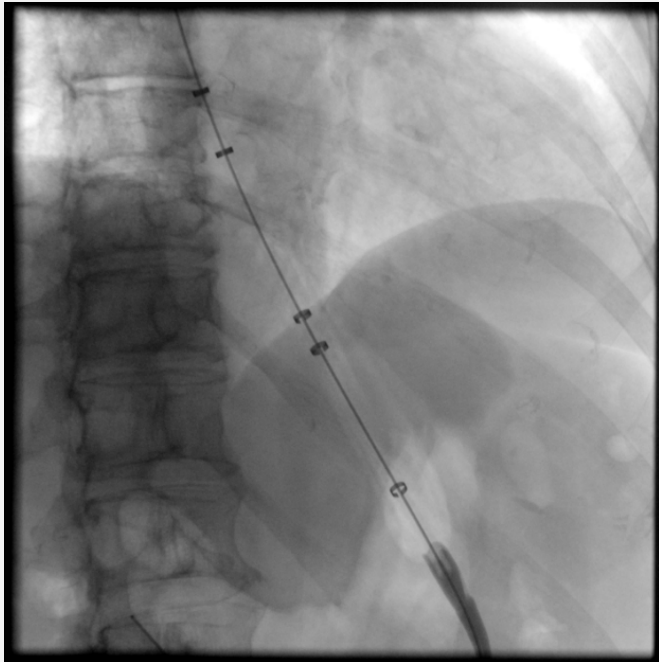
Laparoscopic cardiomyotomy

± fundoplication (Dor vs. „floppy Nissen“)

# PNEUMATIC DILATATION

Performed under sedation and under fluoroscopy

Initial dilatation with 30 mm balloon, 8-15 psi for 15-60 seconds



## PNEUMATIC DILATATION

Performed under sedation and under fluoroscopy

Initial dilatation with 3.0 cm balloon, 8-15 psi for 15-60 seconds

Graded dilatation approach: Cumulatively, dilation with 3.0, 3.5, and 4.0 cm balloon diameters results in good-to-excellent symptomatic relief in 74, 86, and 90 % of patients with an average follow-up of 1.6 years (range 0.1 – 6 years).

The most serious complication is esophageal perforation with an overall median rate in experienced hands of 1.9 % (range 0 – 16 % )

All patients considered for Pneumatic Dilatation must also be candidates for surgical intervention in the event of esophageal perforation needing repair.

# PNEUMATIC DILATATION

## **1. Achalasia dilatation**

- 1.1 Perform dilatation with pneumatic balloons 30–40 mm in diameter starting at 30 mm in the first session to reduce the risk of complications (GRADE of evidence: high; strength of recommendation: strong).
- 1.2 Perform a second dilatation session 2–28 days later with a larger size balloon of 35 mm (GRADE of evidence: high; strength of recommendation: strong).
- 1.3 Consider repeat dilatation (after the initial series) during follow-up to maintain symptom response (GRADE of evidence: high; strength of recommendation: strong).
- 1.4 Perform dilatation under endoscopic or fluoroscopic control based on clinician's preference and local expertise (GRADE of evidence: moderate; strength of recommendation: strong).
- 1.5 Consider proton pump inhibitor (PPI) therapy after dilatation as the technique has 10–40% rate of symptomatic gastro-oesophageal reflux disease (GORD) or ulcerative oesophagitis after treatment (GRADE of evidence: high; strength of recommendation: strong).
- 1.6 Consider performing a water-soluble contrast swallow after dilatation to screen for perforation, but it is not essential (GRADE of evidence: moderate; strength of recommendation: weak).



## DILATATION VS. MYOTOMY

201 patients randomised, follow-up time 43 mo (95% CI 40-47 mo)

	<b>Pneumatic Dilatation</b>	<b>Laparoskop. Heller Myotomy + Dor Hemipl.</b>	<b>p</b>
n	95	106	
1 yr Eckardt Score $\leq 3$	90%	93%	
2 yr Eckardt Score $\leq 3$	86%	90%	0.46
2 yr LES-Pressure	12 mmHg [95% CI 9.7-14]	10 mmHg [95% CI 8.7-12]	0.27
2 yr barium column	3.7 cm [95% CI 0-8.8]	1.9 cm [95% CI 0-6.8]	0.21
Perforation	4%	12%	
Abnormal acid exposure	15%	23%	0.28

No significant differences, but:

Single surgical procedure vs. multiple pneumatic dilatations (max. 3 series with 30, 35, 40 mm)

## POEM VS. LAPAROSCOP. MYOTOMY

	<b>POEM</b>	<b>Laparoskop. Heller Myotomie</b>	<b>p</b>
Reports	21	53	
n	1958	5834	
Follow-up time	16.2 mo	41.5 mo	0.001
1 yr Dysphagia improved	93.5%	91.0%	0.01
2 yr Dysphagia improved	92.7%	90.0%	0.01
GERD symptoms	OR 1.69 (95% CI 1.33–2.14)		0.0001
Abnormal acid exposure	OR 4.30 (95% CI 2.96–6.27)		<0.0001
Erosive Esophagitis	OR 9.31 (95% CI 4.71–18.85)		<0.0001

POEM results in lower rates of dysphagia than laparoscopic cardiomyotomy, but significantly higher rates of reflux

## RECOMMENDATIONS

1. Either graded pneumatic dilation (PD) or laparoscopic surgical myotomy with a partial fundoplication are recommended as initial therapy for the treatment of achalasia in those fit and willing to undergo surgery (strong recommendation, moderate-quality evidence).
2. PD and surgical myotomy should be performed in high-volume centers of excellence (strong recommendation, low-quality evidence).
3. The choice of initial therapy should be guided by patients' age, gender, preference, and local institutional expertise (weak recommendation, low-quality evidence).
4. Botulinum toxin therapy is recommended in patients who are not good candidates for more definitive therapy with PD or surgical myotomy (strong recommendation, moderate quality evidence).
5. Pharmacologic therapy for achalasia is recommended for patients who are unwilling or cannot undergo definitive treatment with either PD or surgical myotomy and have failed botulinum toxin therapy (strong recommendation, low-quality evidence).

## RECOMMENDATIONS

(a) Treatment decisions in achalasia should be made based on patient-specific characteristics, the patient's preference, possible side effects and/or complications and a centre's expertise.

Overall, graded repetitive PD, LHM and POEM have comparable efficacy.

Strong recommendation, moderate certainty of evidence

Consensus: 100% agree [Vote: A., 55.6%; A., 44.4%; A, 0%; D 0%; D., 0%; D., 0%]

(b) BTX therapy should be reserved for patients who are too unfit for more invasive treatments, or in whom a more definite treatment needs to be deferred.

Conditional recommendation, moderate certainty of evidence

Consensus: 100% agree [Vote: A., 100%; A., 0%; A, 0%; D 0%; D., 0%; D., 0%]