Dysphagia

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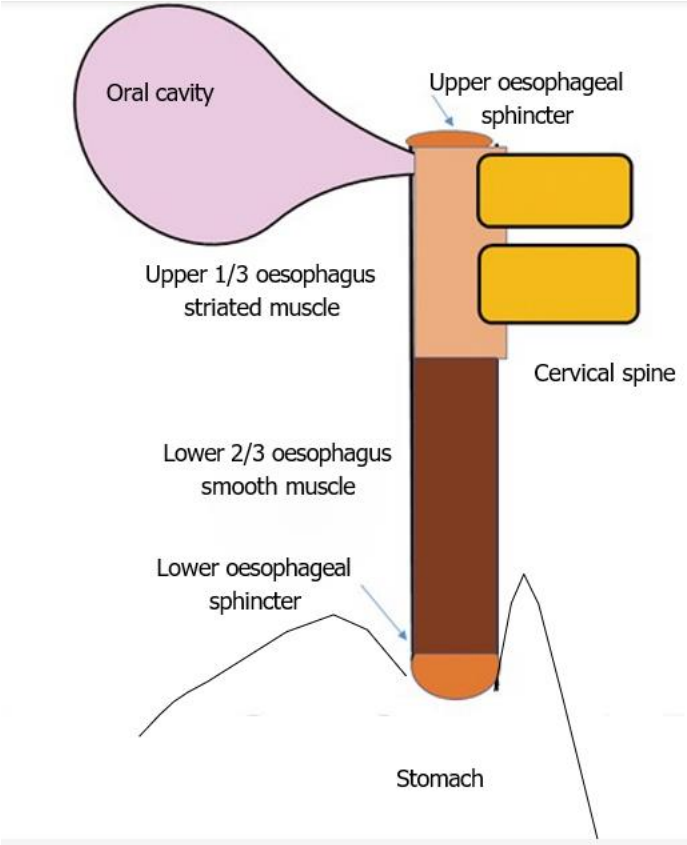
Definition

- Difficulty or discomfort in swallowing, as a symptom of disease.
- Divided into oropharyngeal and oesophageal dysphagia

Anatomy of swallowing



Oropharyngeal dysphagia

- History is key
- Symptom onset is usually within seconds after swallowing
- Drooling
- Coughing
- Nasal regurgitation
- Aspiration
- Choking.

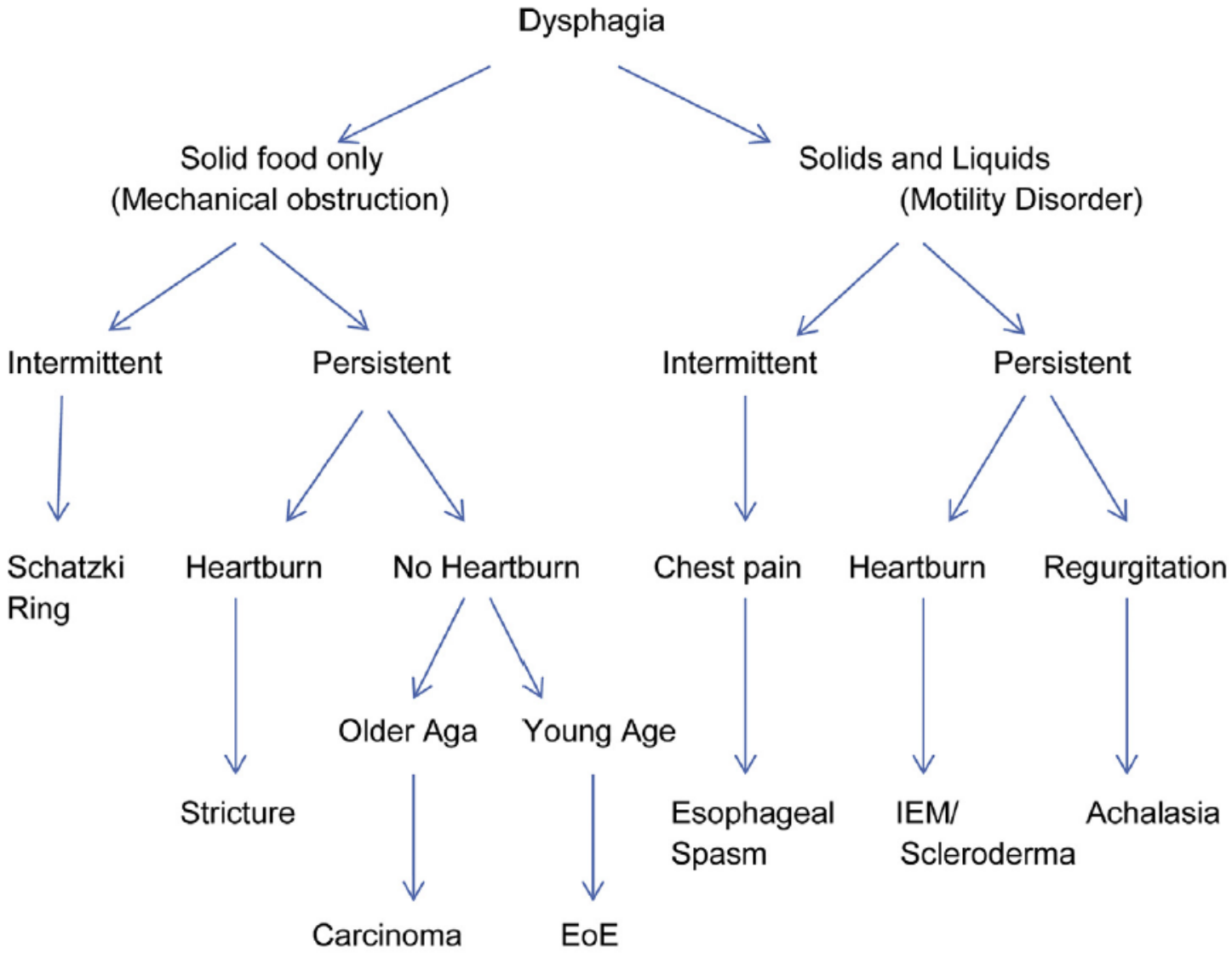
Oesophageal dysphagia

- Patients often present with a history of “food stuck in my throat or chest” after swallowing
- Symptoms may be specific to solids or liquids or both
- Often provided a clue to the underlying diagnosis

Red Flags

- Short history
- Progressive symptoms
- Age over 50 years old
- Persistent coughing or choking
- Aspiration pneumonia

A penetration-aspiration scale. Dysphagia. 1996;11(2):93-98



The American Journal of Medicine, Vol 128, No 10, October 2015

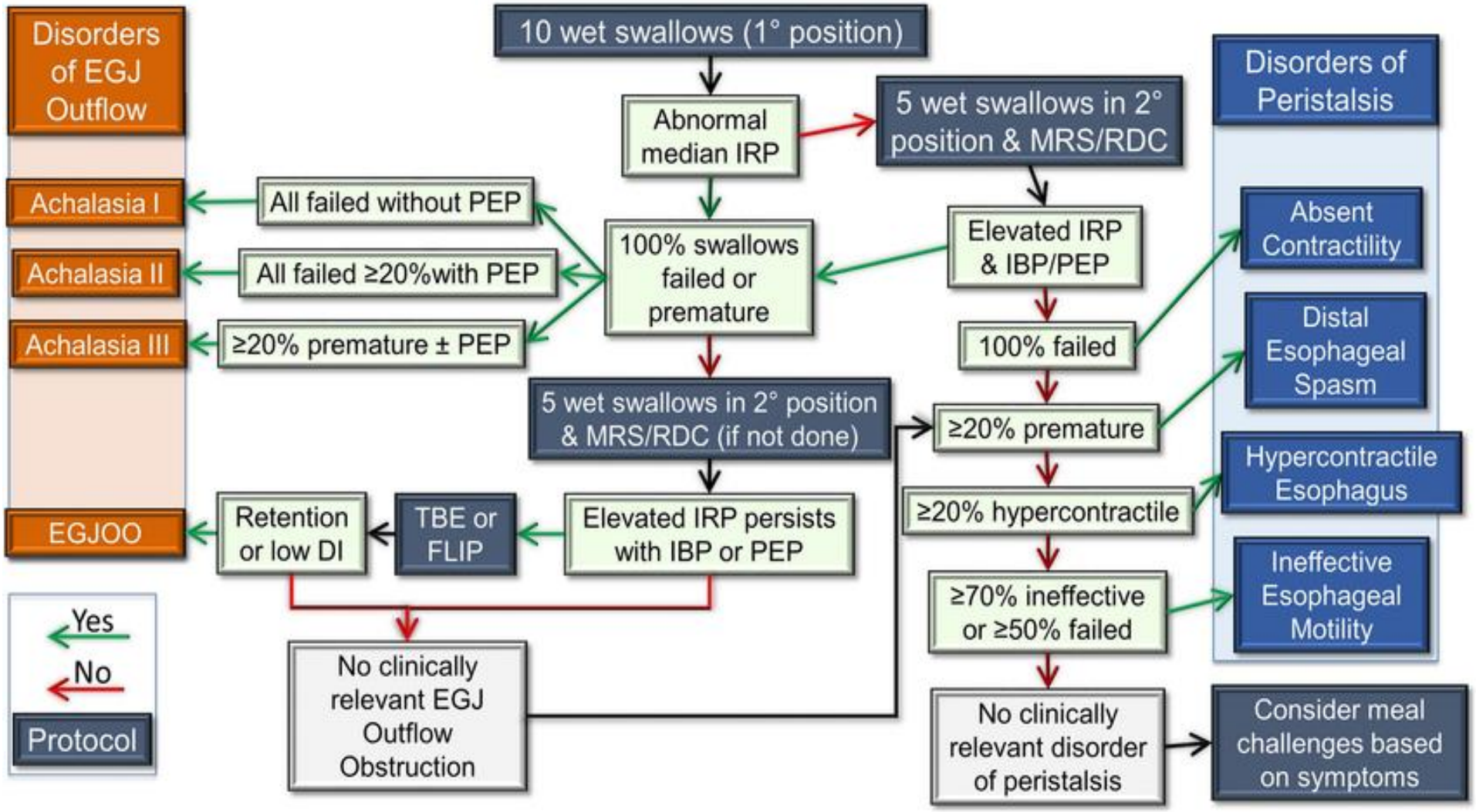
Diagnostic Tools

- Endoscopy
- Barium Swallow
- HRM

HRM

- Diagnostically Pathology divided into:
- Disorders of OGJ outflow:
  - Achalasia types 1-3
- Disorders of peristalsis:
  - Absent contractility
  - Distal oesophageal spasm
  - Hypercontractile oesophagus
  - Ineffective oesophageal spasm

The Chicago Classification v4.0: *Protocol and analysis algorithm*



Achalasia

ACG Clinical Guidelines: Diagnosis and Management of Achalasia

Michael F. Vaezi, MD, PhD, MSc, FACG[1], John E. Pandolfino, MD, MS, FACG[2], Rena H. Yadlapati, MD, MHS (GRADE Methodologist)[3], Katarina B. Greer, MD, MS[4] and Robert T. Kavitt, MD, MPH[5]

Achalasia is an esophageal motility disorder characterized by aberrant peristalsis and insufficient relaxation of the lower esophageal sphincter. Patients most commonly present with dysphagia to solids and liquids, regurgitation, and occasional chest pain with or without weight loss. High-resolution manometry has identified 3 subtypes of achalasia distinguished by pressurization and contraction patterns. Endoscopic findings of retained saliva with puckering of the gastroesophageal junction or esophagram findings of a dilated esophagus with bird beaking are important diagnostic clues. In this American College of Gastroenterology guideline, we used the Grading of Recommendations Assessment, Development and Evaluation process to provide clinical guidance on how best to diagnose and treat patients with achalasia.

Am J Gastroenterol 2020;115:1393–1411. https://doi.org/10.14309/ajg.0000000000000731; published online August 10, 2020

Background

- Achalasia is a primary oesophageal motor disorder of unknown etiology
- Peak incidence 30-60 years old
- Equally common among men and women
- 2-5 in 100,000 incidence per year. USA 1.6 per 100 000

Presenting Symptoms

- Dysphagia (most common)
- Regurgitation- ACG recommends that the patient with regurgitation symptoms despite PPI, should consider work up for Achalasia
- Non-cardiac chest pain
- Weight loss
- Heart burn

Pseudoachalasia

- Paraneoplastic syndrome eg, small cell lung Ca
- Pseudoachalasia from extrinsic processes such as prior tight fundoplication or laparoscopic adjustable gastric banding or from tumours infiltrating the OGJ causing obstruction
- Chagas disease- other features of diffuse enteric myenteric destruction as well a travel history

Investigations

1.Upper endoscopy:

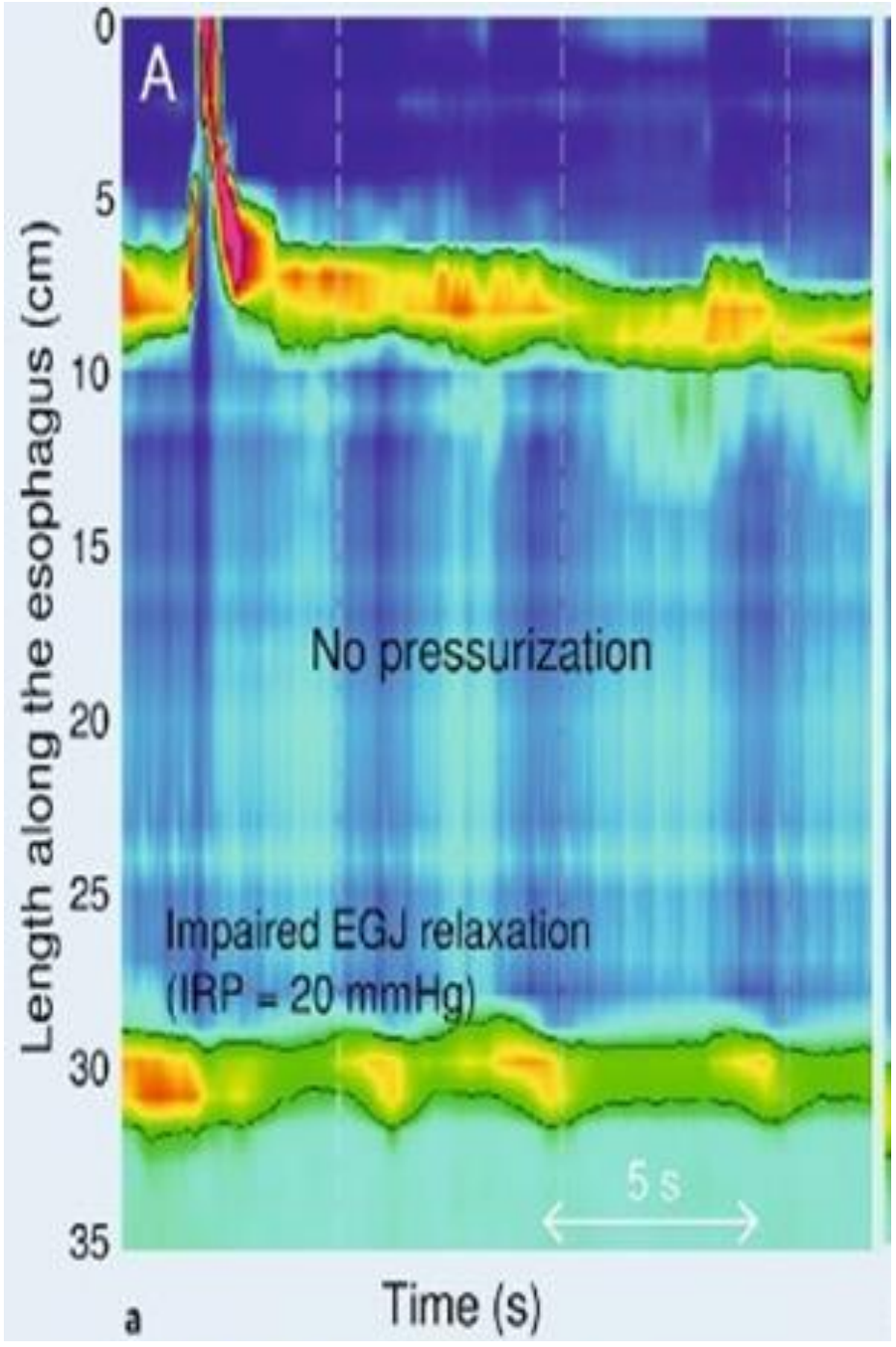
- Grossly normal, Tight lower oesophageal sphincter (LOS) characteristic POP when endoscope passed
- Dilated sigmoid oesophagus with retained food and saliva
- Part of work up of ruling out pseudoachalasia

2. High resolution manometry (HRM) =gold standard

3. Barium oesophogram: bird’s beak appearance

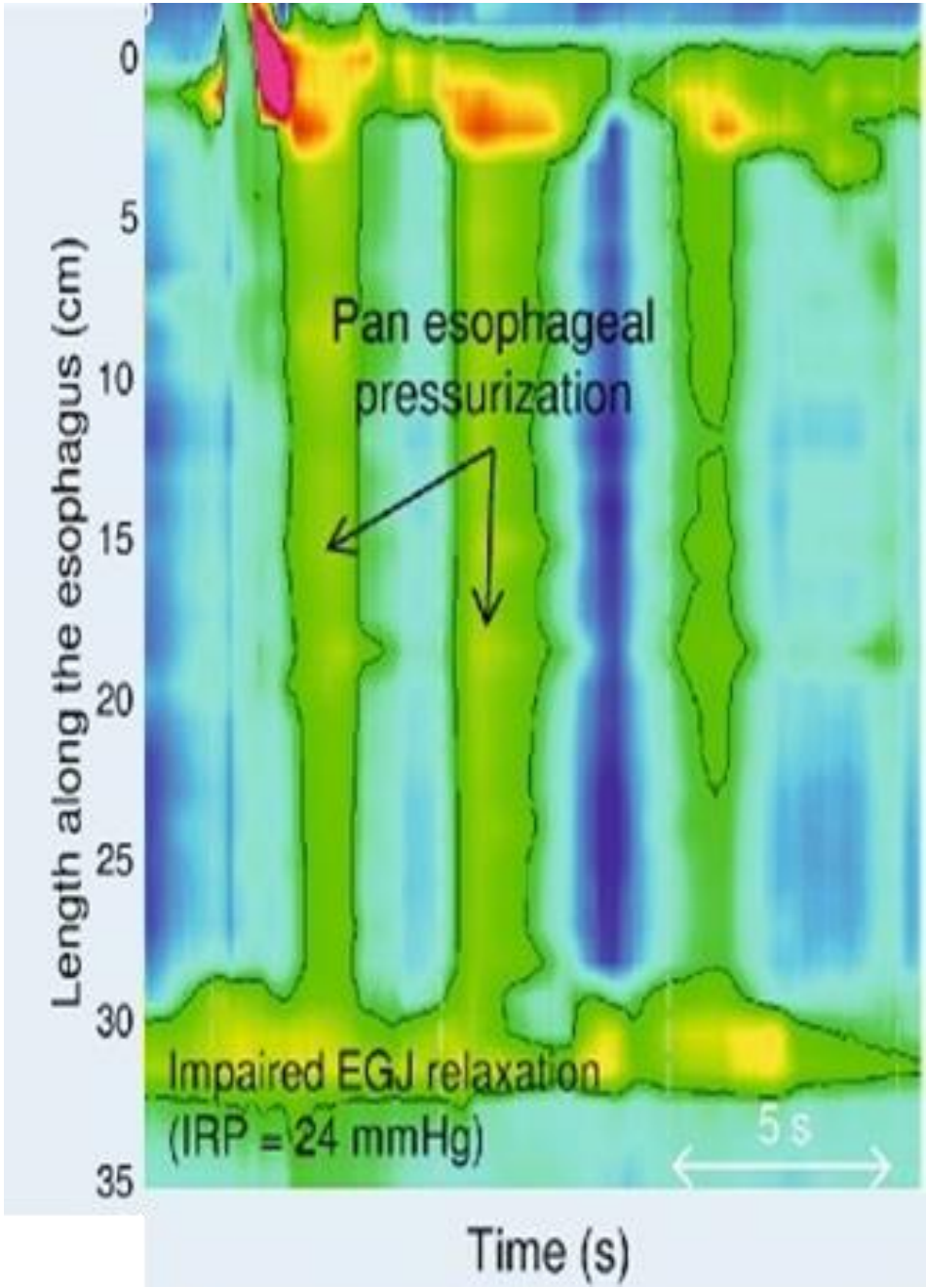
Achalasia type 1

- Type I (“classic achalasia”): 100% aperistalsis



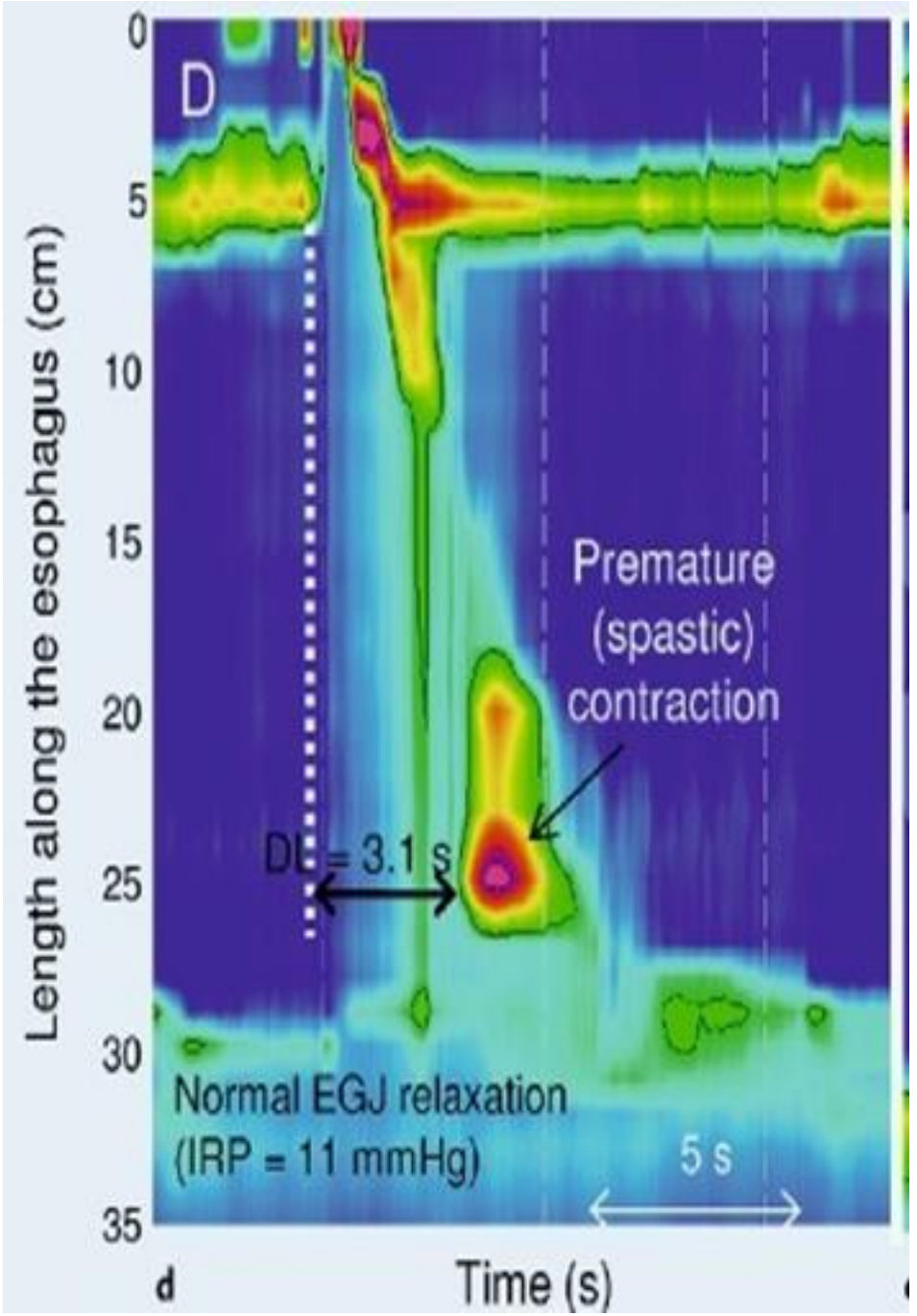
Achalasia type 2

- Type II: 100% aperistalasis with 20% or more pan-oesophageal pressurisation (Achalasia with compression)



Achalasia type 3

- Type III: Spastic type. Aperistalsis with 20% of the swallows revealing simultaneous contractions



Treatment options

- Pharmacological
- Endoscopic
- Surgical

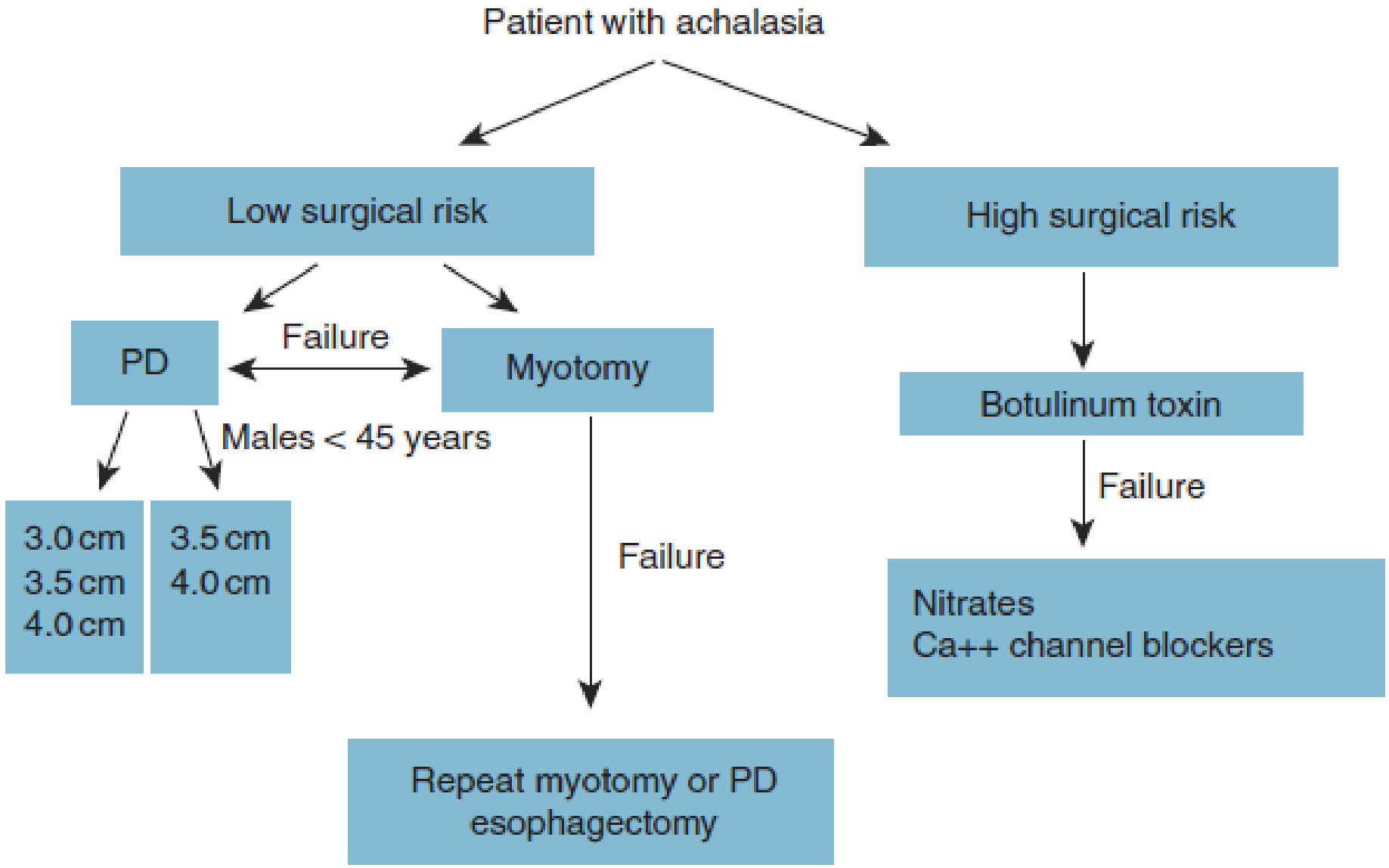


Figure 2. Recommended treatment algorithm for patients with achalasia. PD, pneumatic dilation.

Pharmacological options

- Calcium channel blockers
- Nitrates
- Least effective
- Symptomatic improvement 53 to 87%

Endoscopic

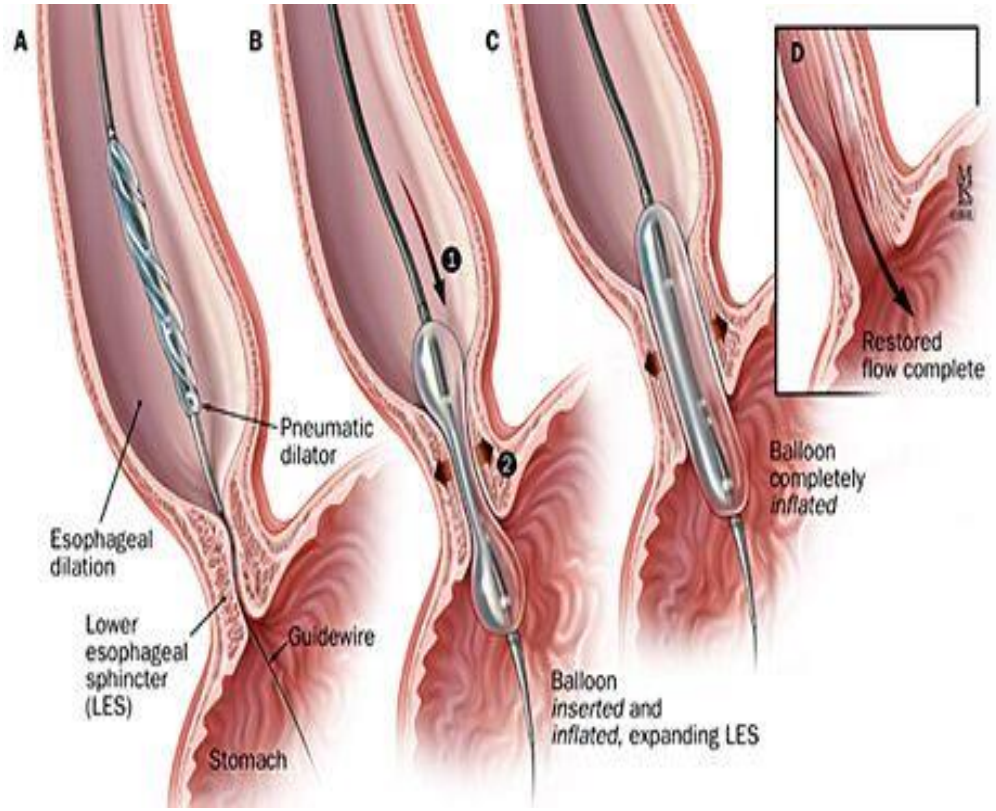
- **Botulinum toxin**
  - Durability **6-12months**
  - **Potent presynaptic inhibitor of acetylcholine release** from nerve endings
  - 1 month response rate >75%

- **Pneumatic dilatation**

- **Per-oral endoscopic myotomy (POEM)**
- If expertise available:
  - POEM should be considered as **primary therapy for type III achalasia**
  - POEM should be considered a treatment option **comparable** to laparoscopic Heller myotomy for any of the achalasia syndromes
  - AGA guidelines 2017

Pneumatic dilatation

- Endoscopic, graded dilatation
- Good short term results
- No GA
- Requires repeated dilatation
- Risk of perforation ~5%



Response to therapy

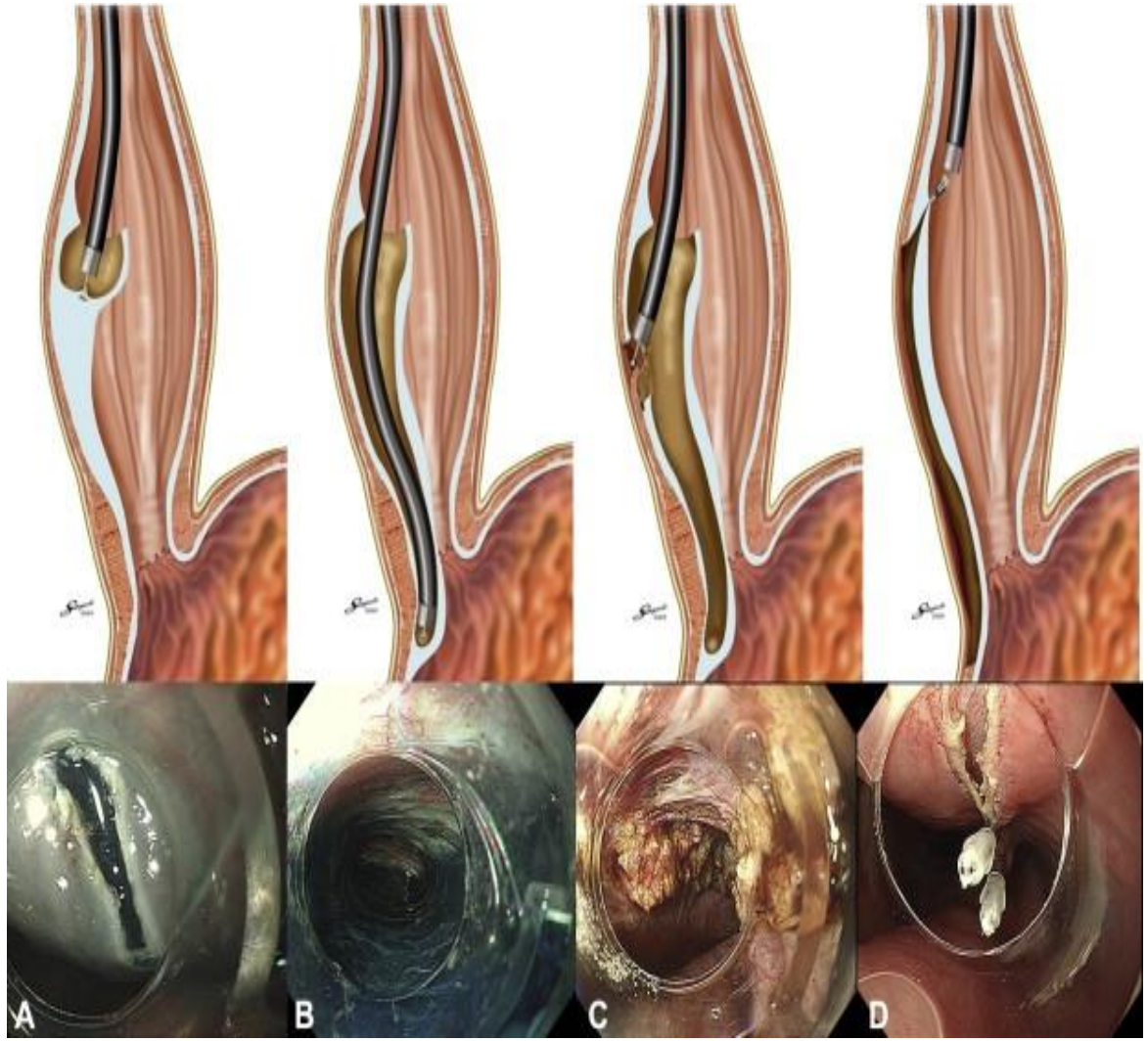
Graded approach results in better outcomes

44% compared to 28% at 6 years i.t.o symptoms

Predictors of favorable clinical response to PD include:

- older age (> 45 years), female gender, narrow oesophagus predilation, LES pressure after dilation of < 10 mm Hg , and type II pattern on HRM

POEM



Submucosal tunneling

Tunneling beyond GE junction

Circular muscles divided

Closure of mucosal entry

Surveillance

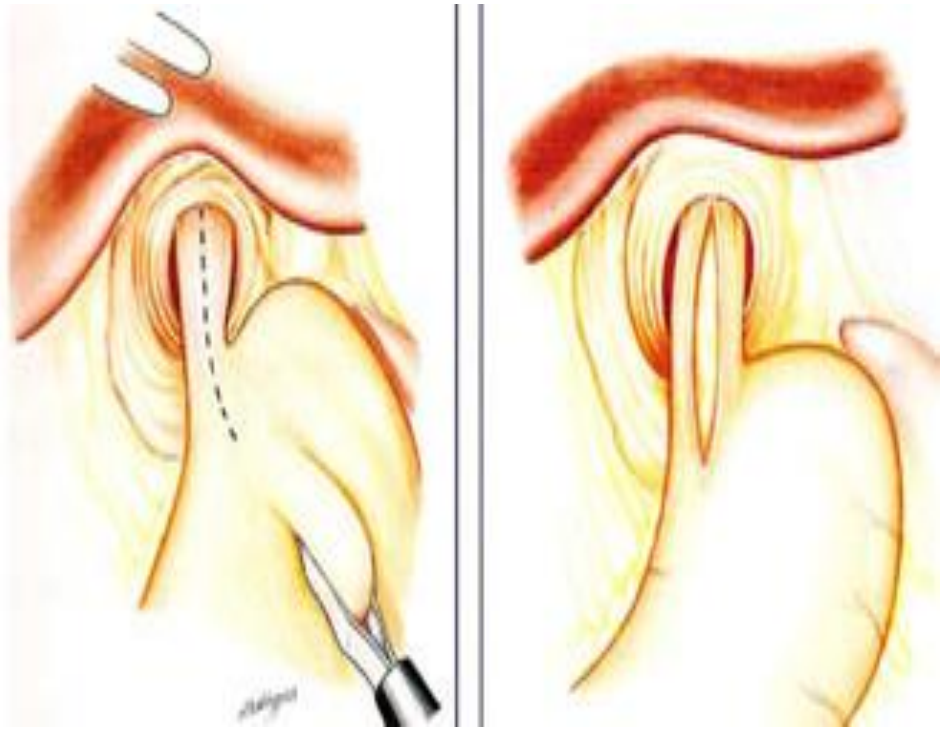
- Higher incidence of squamous cell ca.
- 1 cancer per 300 patients years
- Limited data to support routine cancer screening

Surgical options

- Laparoscopic Heller myotomy
- Oesophagectomy

Heller myotomy

- 87% success after 2 years



Oesophagectomy

- Some patients may develop “ end-stage ” achalasia characterized by megaoesophagus or sigmoid oesophagus and significant oesophageal dilation and tortuosity

EoE

CLINICAL PRACTICE GUIDELINES

AGA Institute and the Joint Task Force on Allergy-Immunology Practice Parameters Clinical Guidelines for the Management of Eosinophilic Esophagitis

Ikuo Hirano,[1] Edmond S. Chan,[2] Matthew A. Rank,[3] Rajiv N. Sharaf,[4] Neil H. Stollman,[5] David R. Stukus,[6] Kenneth Wang,[7] Matthew Greenhawt,[8] and Yngve T. Falck-Ytter,[9] on behalf of the AGA Institute Clinical Guidelines Committee and the Joint Task Force on Allergy-Immunology Practice Parameters

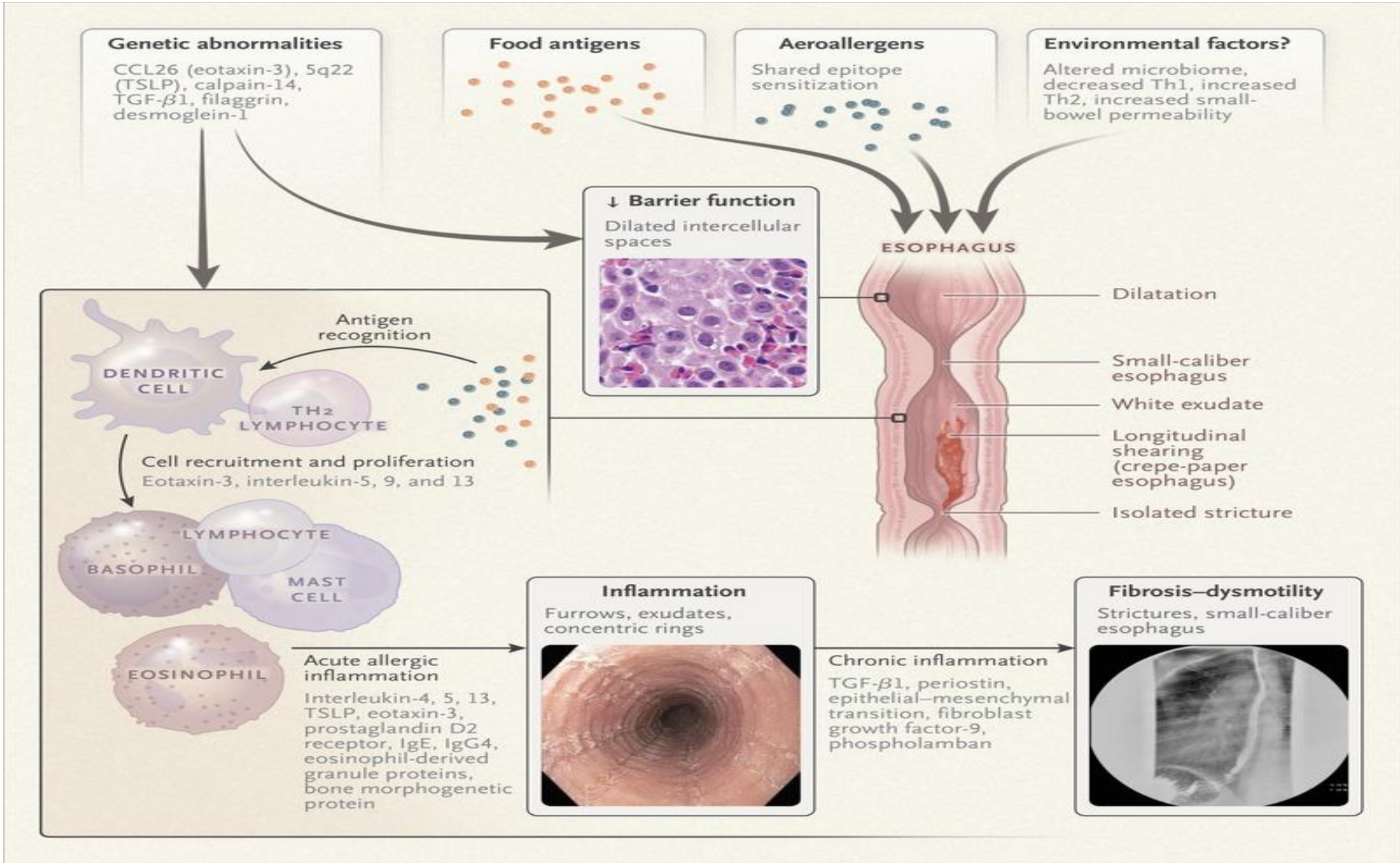
PREVALENCE

- Atopic male
- Caucasian most commonly
- Male: female 2-3:1
- US 52/100000
- Mean age of diagnosis 30-50 years (5-9 in kids)

Khan et al, Digestion 2021

Table 3. Etiological factors associated with EoE

N	Etiologies	Associated factors
1	Inflammatory mediators	TH2-mediated IL-4, IL-5, IL-13, IL-33 and TSLP
2	Genetic susceptibility	TSLP and CAPN14
3	Environmental factors	Pollen seasons, cold or dry climate
4	Atopic conditions	Asthma, allergic rhinitis, and atopic dermatitis
5	Dietary products	Cow's milk, soy, egg, wheat, peanut/tree nuts, seafood, and legumes
6	Esophageal microbiome	*Haemophilus*, *Corynebacterium*, *Neisseria*, *Firmicutes* and *Bacteroides*
7	Oral immunotherapy	IgE-mediated food allergy (cow milk, egg, and shellfish)
8	Inversely associated conditions	*Helicobacter pylori* and the development of EoE, and EoE versus Barrett's esophagus
9	Acid suppressor	PPI → preventing peptic digestion of food allergens → alter microbial dysbiosis


Genetic abnormalities
CCL26 (eotaxin-3), 5q22 (TSLP), calpain-14, TGF-β1, filaggrin, desmoglein-1
Food antigens
Aeroallergens
Shared epitope sensitization
Environmental factors?
Altered microbiome, decreased Th1, increased Th2, increased small-bowel permeability
↓ Barrier function
Dilated intercellular spaces
ESOPHAGUS
Dilatation
Small-caliber esophagus
White exudate
Longitudinal shearing (crepe-paper esophagus)
Isolated stricture
Antigen recognition
DENDRITIC CELL
TH2 LYMPHOCYTE
Cell recruitment and proliferation
Eotaxin-3, interleukin-5, 9, and 13
LYMPHOCYTE
BASOPHIL
MAST CELL
EOSINOPHIL
Acute allergic inflammation
Interleukin-4, 5, 13, TSLP, eotaxin-3, prostaglandin D2 receptor, IgE, IgG4, eosinophil-derived granule proteins, bone morphogenetic protein
Inflammation
Furrows, exudates, concentric rings
Chronic inflammation
TGF-β1, periostin, epithelial–mesenchymal transition, fibroblast growth factor-9, phospholamban
Fibrosis–dysmotility
Strictures, small-caliber esophagus

DEFINITION

- Immune mediated clinicopathological condition
- Oesophageal infiltration with eosinophils→ inflammation and strictures

CLINICAL PRESENTATION

- Dysphagia
- Food impaction
- Heartburn, regurgitation
- Chest pain

Gastrointestinal symptoms	Atypical symptoms
Dysphagia (adolescents and adults)	Chest pain
Food impaction (adolescents and adults)	Rhinitis
Heartburn	Asthma
Regurgitations	Hoarseness
Abdominal pain	Croup, cough
Feeding disorders (paediatric < 2 years)	Rhinosinusitis
Failure to thrive (paediatric < 2 years)	Atopic dermatitis
	Sleep disorders breathing

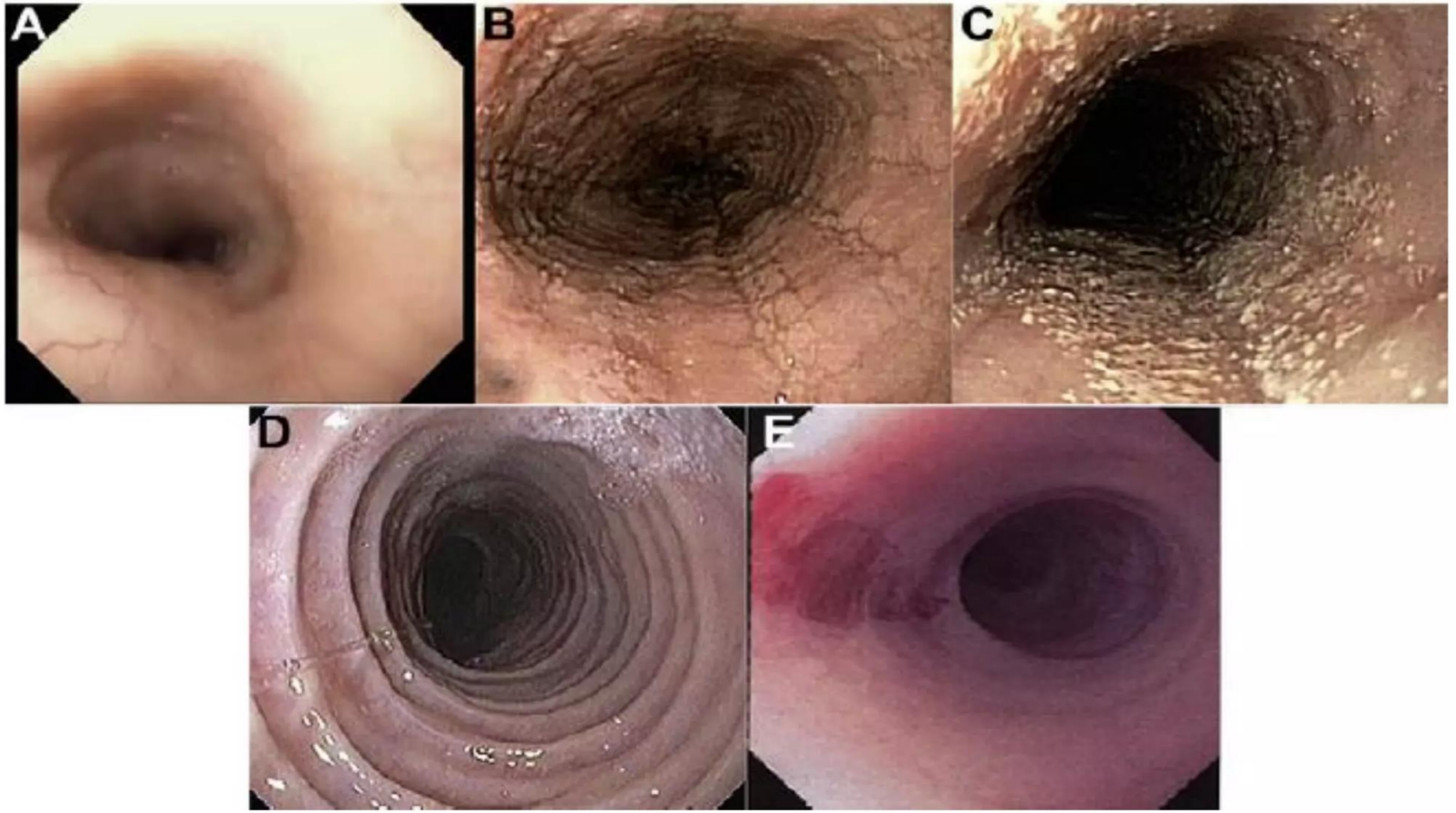
Diseases associated with esophageal eosinophilia
EoE
GERD
PPI-responsive esophageal eosinophilia
Eosinophilic gastrointestinal diseases
Esophageal infections (e.g., fungal, viral, parasitic)
Crohn's disease
Celiac disease
Achalasia
Hypereosinophilic syndrome
Pemphigus
Vasculitis
Drug hypersensitivity
Connective tissue diseases
Graft vs. host disease

DIAGNOSIS

- No pathognomic endoscopic findings
- Suggestive gastroscopy: 38% histological evidence of EoE
- Schatski’s ring: 20% of cases of EoE
- Normal gastroscopy: 9.8% evidence of EoE

Dhar A, *et al*. *Frontline Gastroenterology* 2022

ENDOSCOPIC



A : Normal, B : Esophageal furrowing, C : White mucosal plaques(eosinophilic microabscess),
D : Esophageal ring trachealization, E : Small-caliber esophagus with mucosal tearing after endoscopy.

Eosinophilic esophagitis: Updated consensus recommendations for children and adults, J Allergy Clin Immunol 2011;128:3-20

EoE **E**ndoscopic **R**e**F**erence **S**core (EREFS)

Grade 0 Grade 1 Grade 2 Grade 3

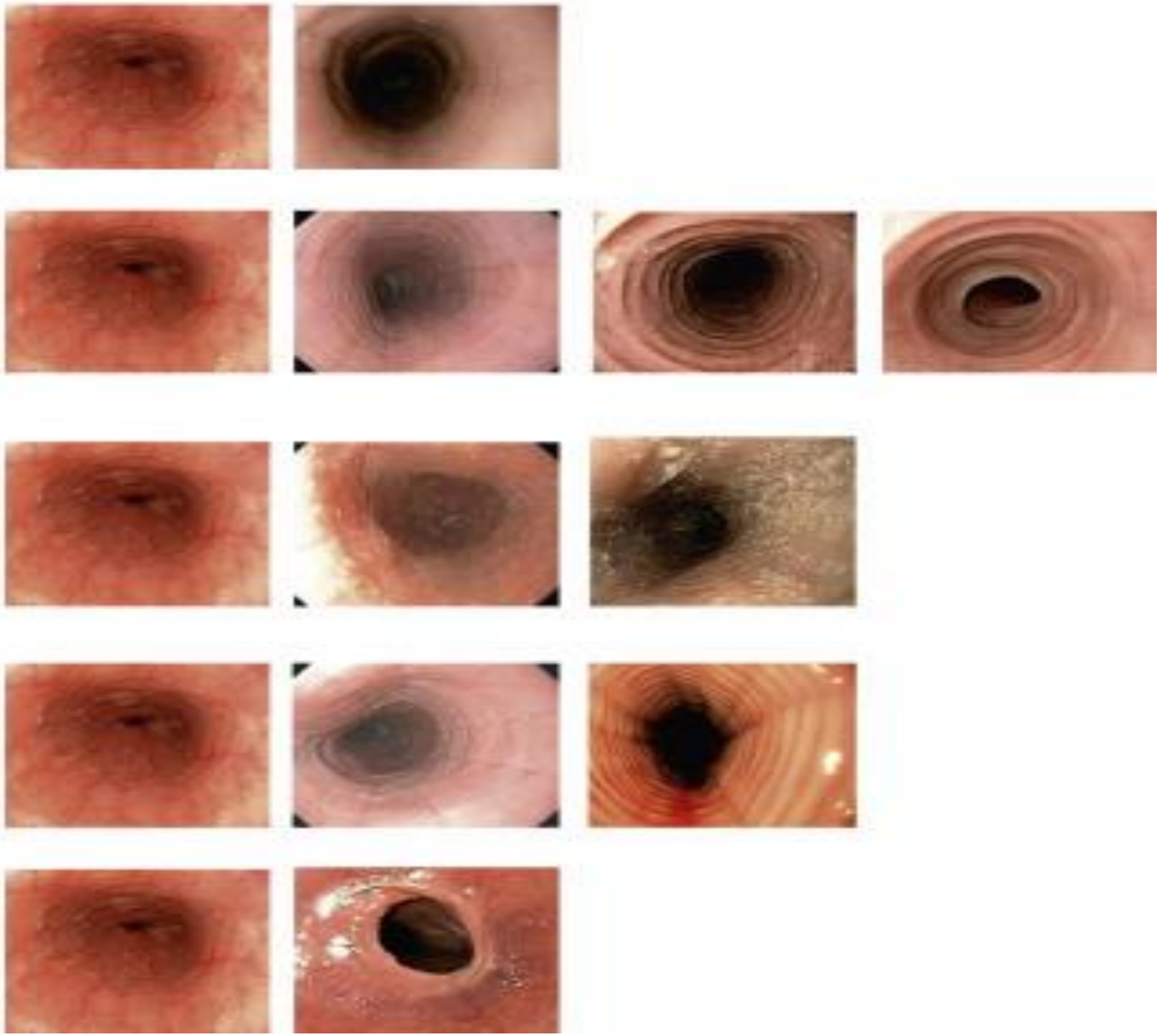
Edema (loss of vascular markings)
- Grade 0: Distinct vascularity
- Grade 1: Absent or decreased

Rings (trachealization)
- Grade 0: None
- Grade 1: Mild (ridges)
- Grade 2: Moderate (distinct rings)
- Grade 3: Severe (scope will not pass)

Exudate (white plaques)
- Grade 0: None
- Grade 1: Mild (≤ 10% surface area)
- Grade 2: Severe (> 10% surface area)

Furrows (vertical lines)
- Grade 0: None
- Grade 1: Mild
- Grade 2: Severe (depth)

Stricture
- Grade 0: Absent
- Grade 1: Present



BIOPSY

- 2-4 mucosal biopsies from mid and distal oesophagus (at least 6 from at least 2 areas)
- If suspecting another disease process- biopsies of antrum/duodenum

HISTOLOGIC

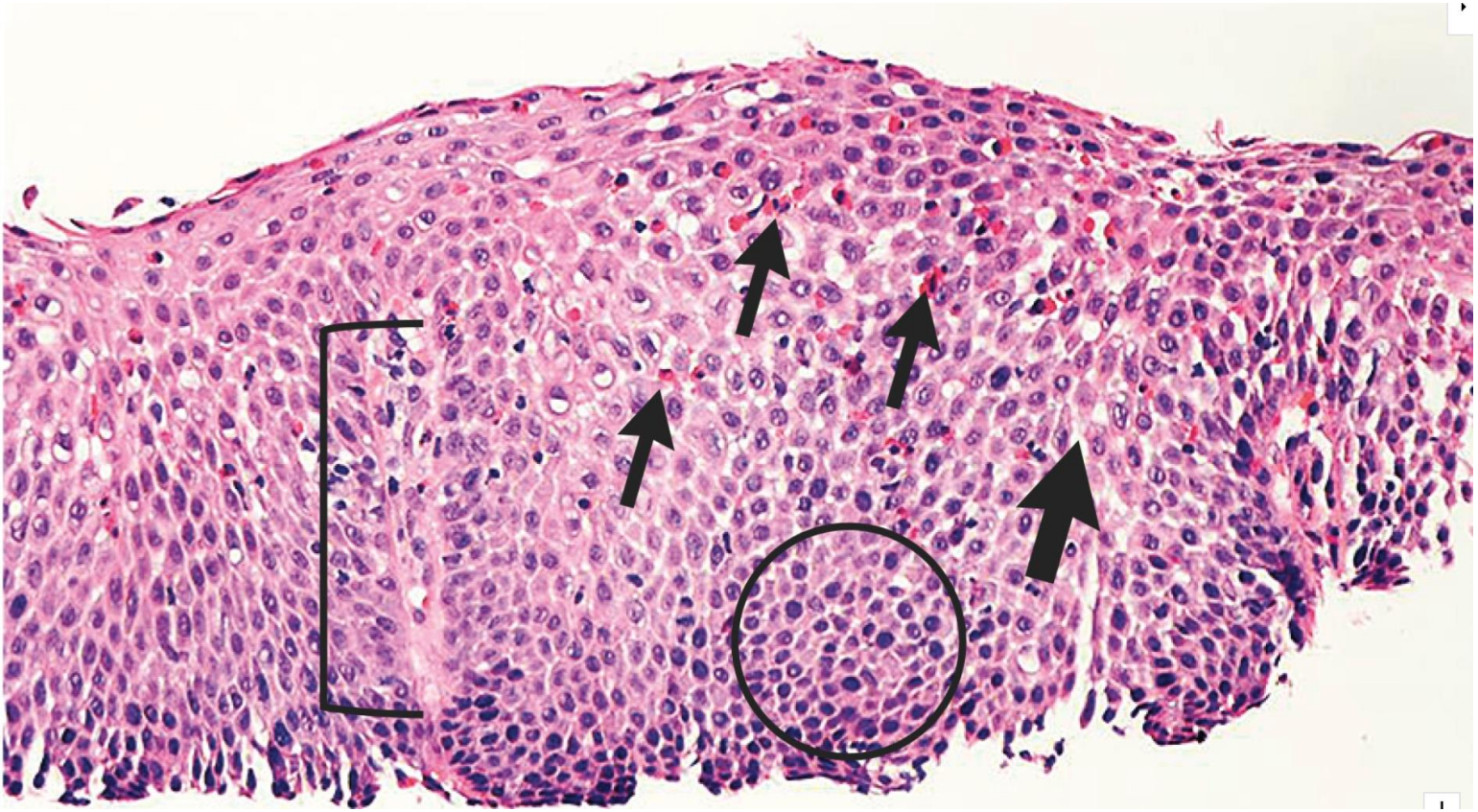


Figure 2. Histologic Characteristics of Eosinophilic Esophagitis.

Routine staining with hematoxylin and eosin reveals numerous eosinophils (thin arrows), dilated intercellular spaces (thick arrow), basal zone hyperplasia (circle), and papillary elongation (bracket).

HISTOLOGIC

- >15 eosinophils per hpf
- NOT specific to EoE

TABLE E3. Histologic features of EoE

Mucosal eosinophilia
Eosinophil microabscess formation
Superficial layering of eosinophils
Extracellular eosinophil granules
Epithelial desquamation
Basal zone hyperplasia
Rete peg elongation
Dilated intercellular spaces
Subepithelial fibrosis/sclerosis–lamina propria fibrosis
Mastocytosis and mast cell degranulation
$CD8^+$ lymphocytes and B cells

EoE vs PPI-REE

- PPI-REE is diagnosed when patients have oesophageal symptoms and histologic findings of eosinophilia
- Have a response both clinically and histologically to PPI
- Distinct entity from EoE

Table 5. Complications associated with EoE

Conditions	Characteristics
Inflammatory conditions	Furrows, white exudates, edema, esophageal rings, and stenosis
Esophageal perforation	Boerhaave syndrome, fibrostenotic condition, and esophageal dilatation
Hepatic portal venous gas	Intraluminal gas enters the portal venous circulation due to endoscopic dilation for benign esophageal strictures with EoE
Intramucosal dissection of the esophagus	Inflammatory conditions → separation of mucosa and/or submucosa → false lumen
Esophageal dysmotility	Esophageal mucosa infiltration by eosinophils and their interactions with the microenvironment and inflammatory cytokines
Achalasia-like changes	Esophageal muscularis propria → abnormally buildup of eosinophils → myoactive and neuroactive eosinophilic secretory products → cytotoxic eosinophil secretory products
Adrenal insufficiency	Low morning serum cortisol levels

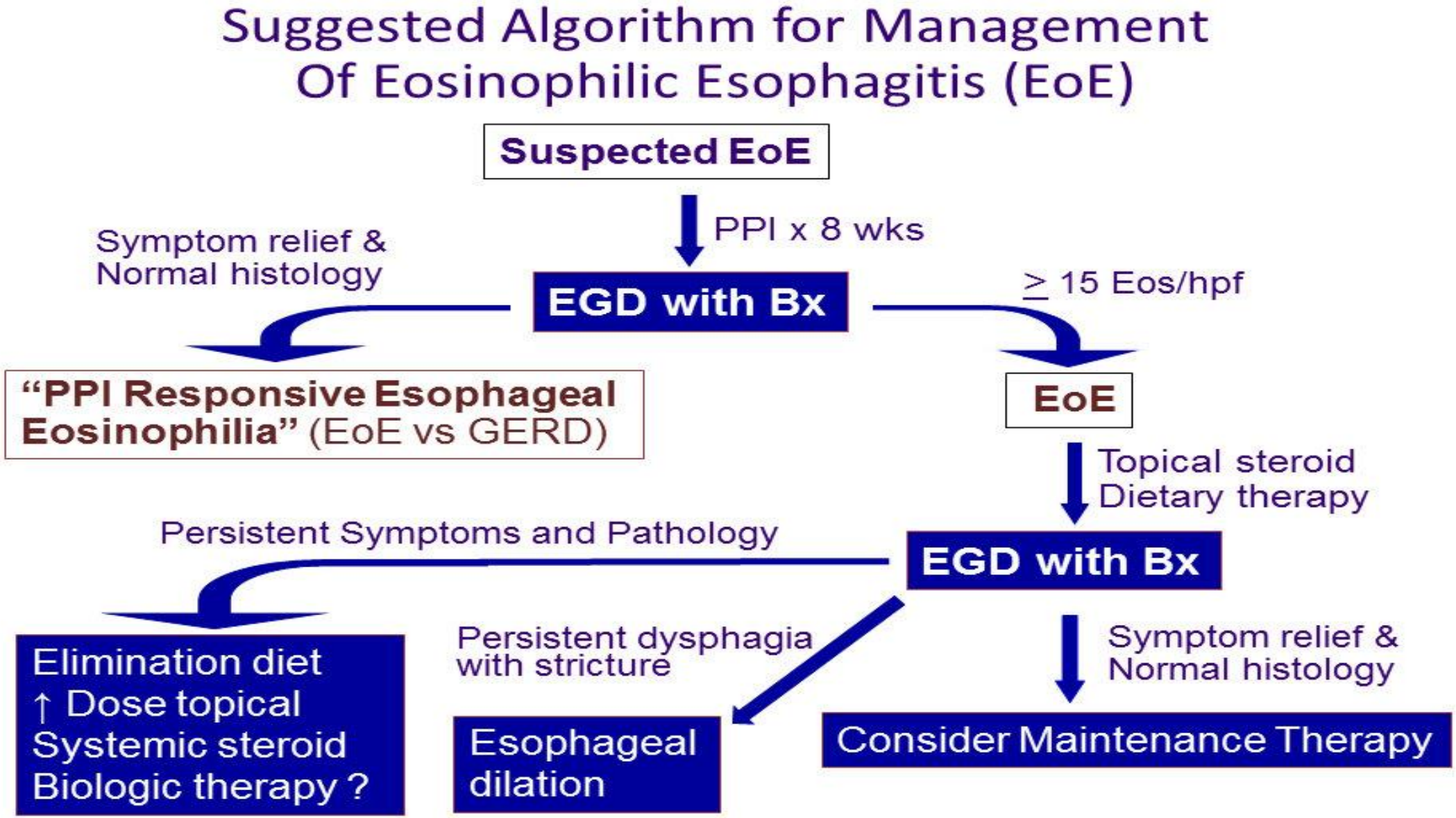
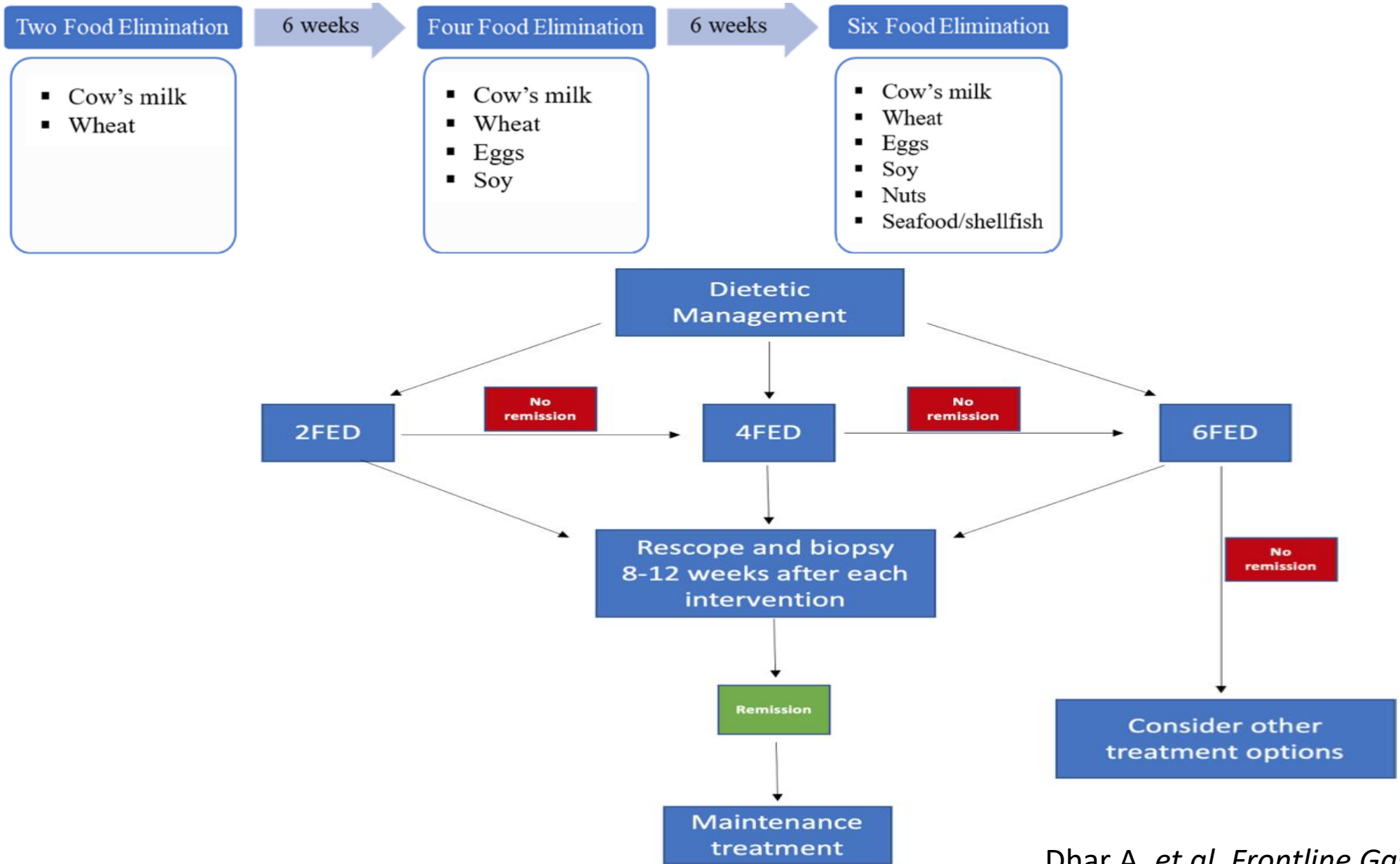


Figure 3: Management of eosinophilic esophagitis

DIET

- **Elemental diet:** Amino acid, carbohydrate, lipid, vitamin/mineral based formula. Induces histo and clinical resolution in >95% of patients
- **SFED:** remission in 72%, maintain in 45%
- **Test directed elimination diet**- children (48%) Adults (32%)



Dhar A, *et al*. *Frontline Gastroenterology* 2022

STEROIDS

- Topical: swallowed
  - Fluticasone 220mcg 2-4 puffs BD
  - Budesonide 0.5-1 mg BD
- Liquid formulations mixed with something to increase viscosity (sucralose/honey/syrup)
- Fast for 30 min after

OTHER AGENTS

- LRTA
- Cromolyn
- Immune modulators
- Biologics
  - **DUPILUMAB:** Approved this year by FDA for EoE
  - Inhibits IL-4, IL-13

ENDOSCOPIC Mx

- Endoscopic dilatation of strictures