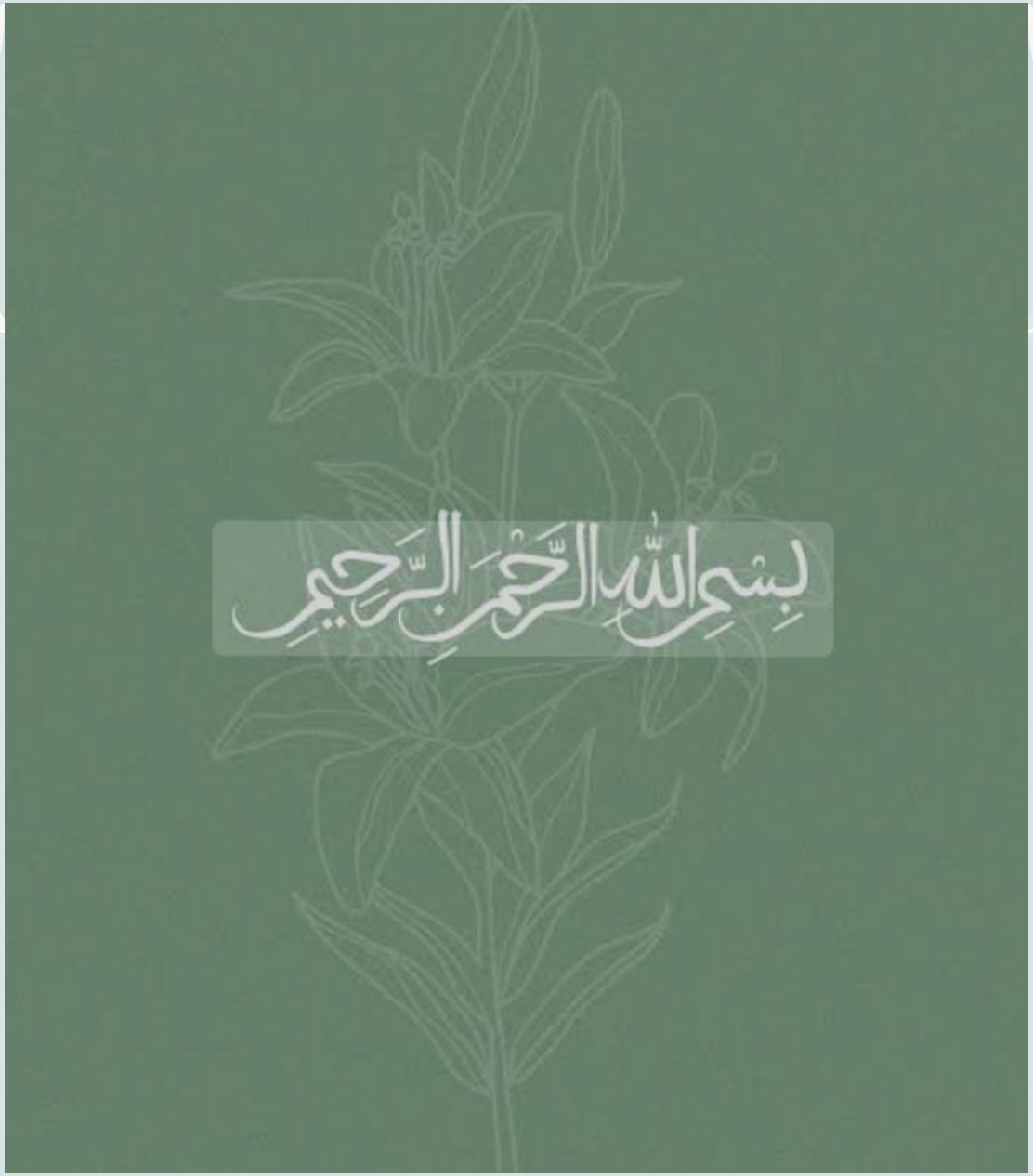
بسم الله الرحمن الرحيم

Therapeutic management of esophageal problems after Esophageal Atresia repair

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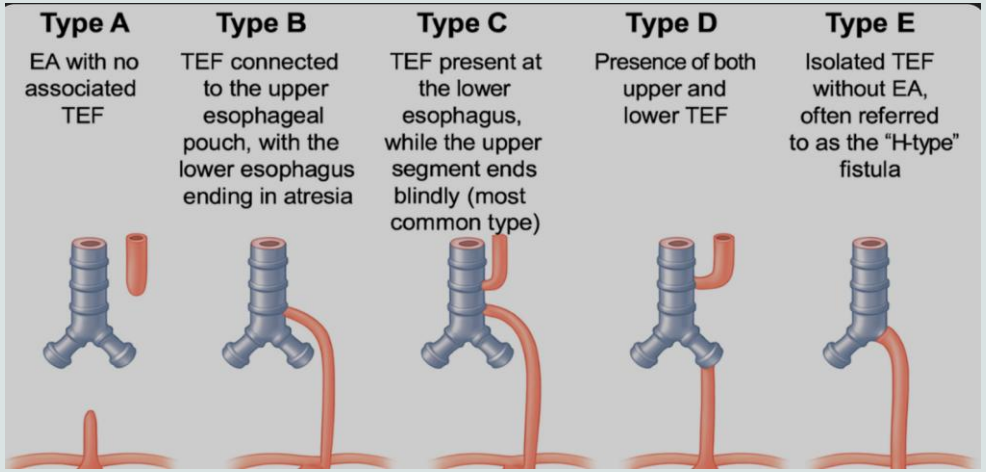
Esophageal Atresia



- Defect of the esophageal continuity, with or without a fistula
- Most common, life-threatening congenital malformations
- Survival rates: 87% overall, 72% syndromic
- Mortality : extreme prematurity or severe associated malformations (cardiac)
- The incidence of an EA:1 in 3500- 1 in 4500 live births
  - ✓ 84% proximal pouch and distal TEF
  - ✓ 4% H-type

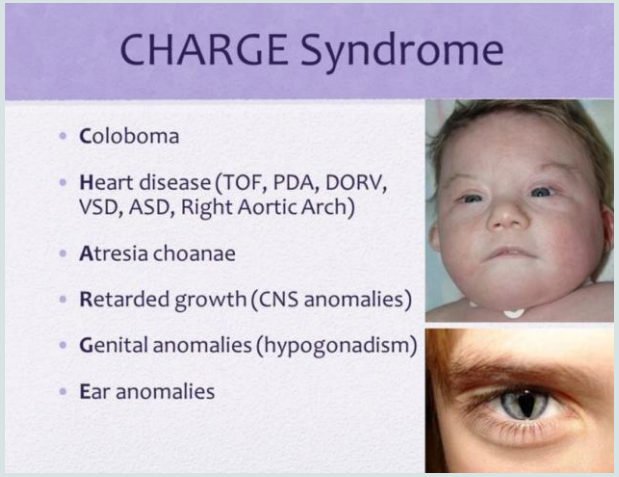
Risk factors:

- Exposure to thalidomide, statins, alcohol, smoking, contraceptive pills, hormones
- Endocrine diseases of the mother, such as diabetes
- Higher maternal age

The recurrence risk:

1. one affected child : 0.5–2%
2. an affected parent :3–4%

The most frequent associated anomalies as a part of



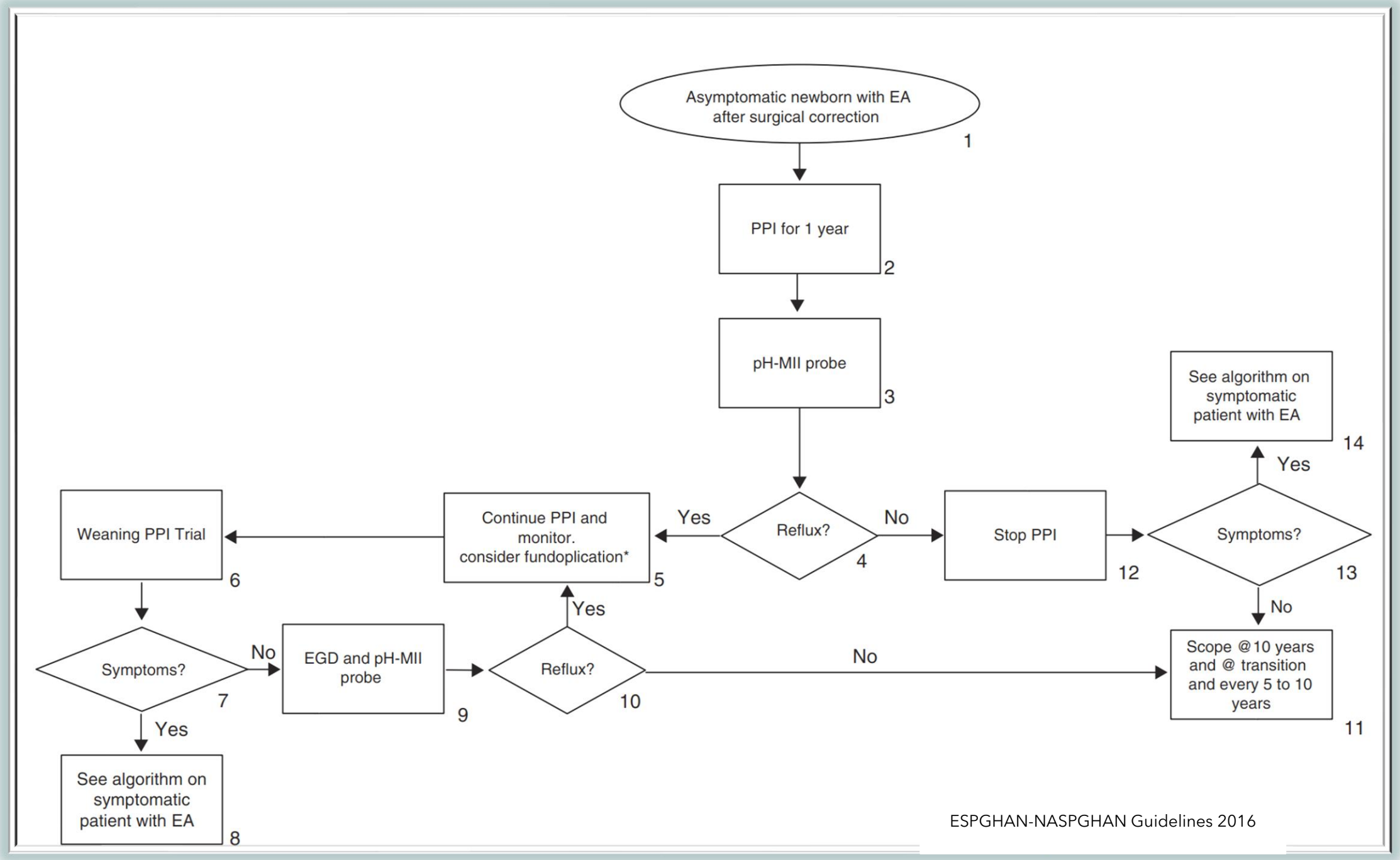
GER in EA patients

- The most frequent GI tract complication with a reported prevalence of 22% to 45%
- GER as a major factor for recurrent anastomotic strictures: ?

- **RECOMMENDATION:** Acid suppression in all EA patients in the neonatal period

How long GER should be treated?

- No prospective controlled studies
- Complications due to GER occur mostly during the first year of life:
  - Anastomotic strictures
  - Esophagitis
  - Cyanotic spells
  - Pulmonary problems
  - Failure to thrive

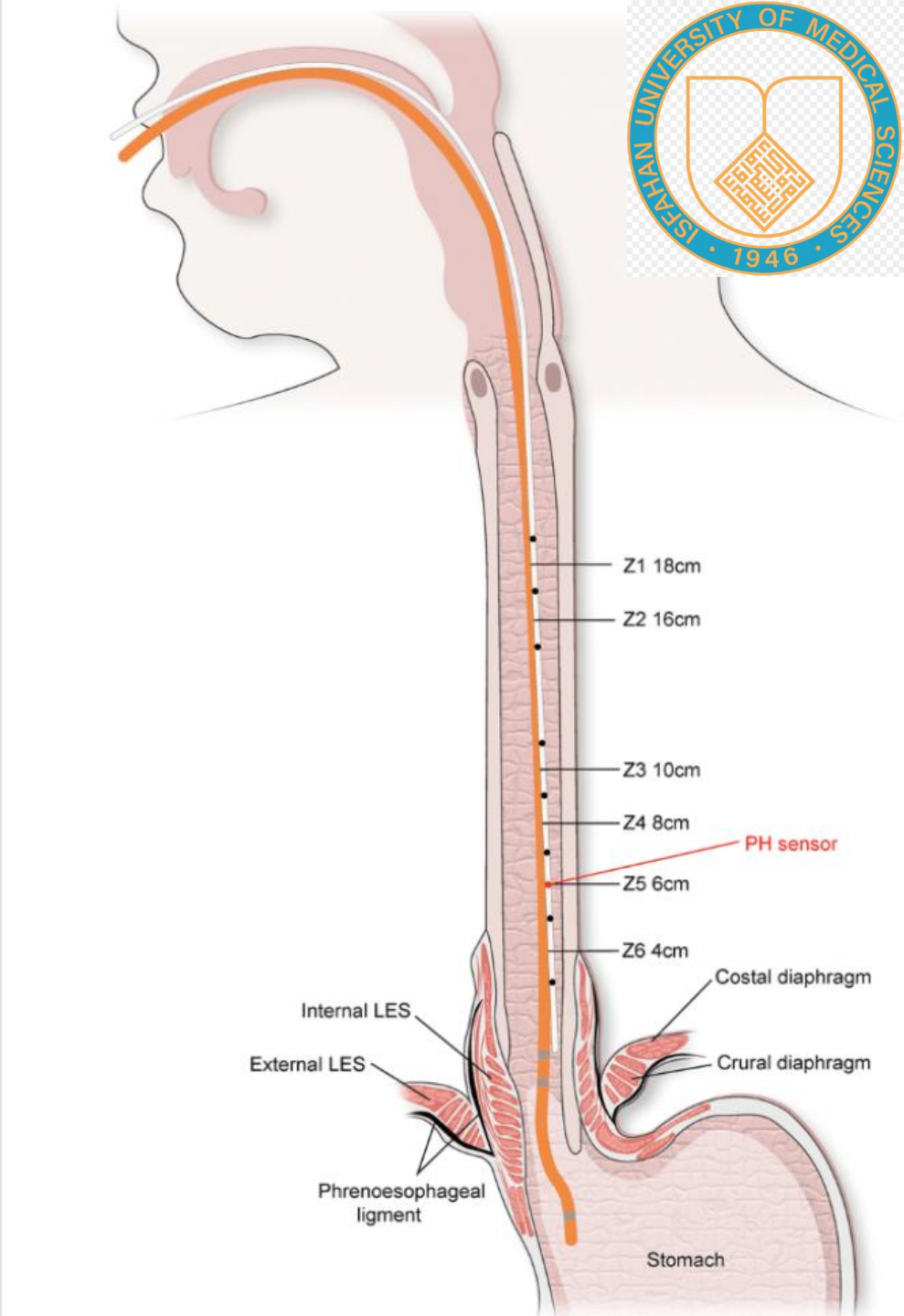


ESPGHAN-NASPGHAN Guidelines 2016

- The expert panel recommends 3 endoscopies throughout childhood:(Monitoring of GER in all EA patients (even asymptomatic))
  - After stopping PPI therapy
  - Before the age of 10 years
  - At transition to adulthood

pH-impedance monitoring

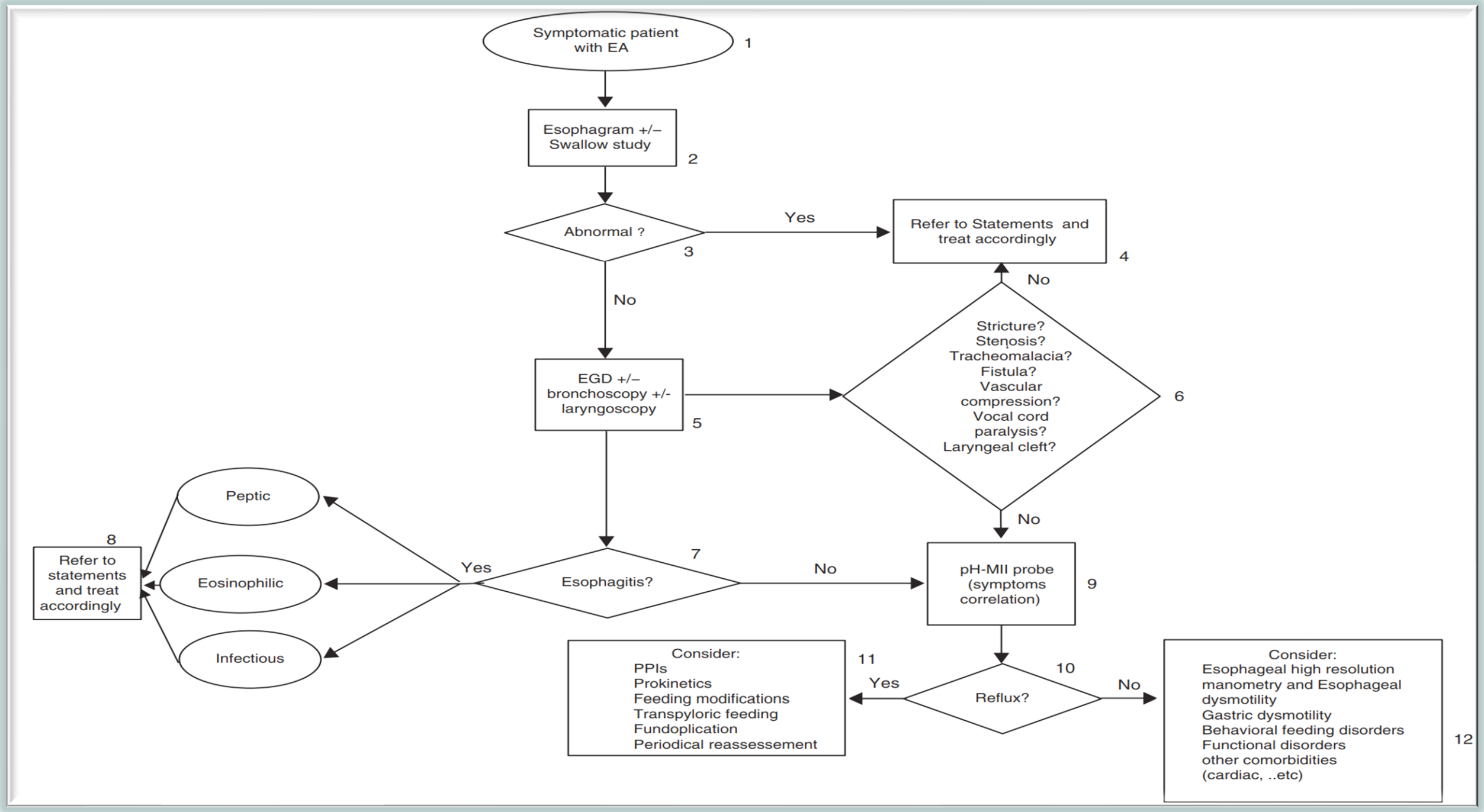
- Evaluate and correlate non-acid reflux with symptoms in selected patients
    - symptomatic on PPI
    - ALTE
    - on continuous feeding
    - with extra-digestive symptoms
    - GER symptoms with normal pH-probe and endoscopy



- **RECOMMENDATION:**
- GER be systematically treated for prevention of peptic complications and anastomotic stricture up to the first year of life or longer, depending on persistence of GERD.

Role of esophagoscopy in EA patients

- Complications due to GERD can be observed during childhood, adolescence, and adulthood :
  - Late or recurrent anastomotic stenosis
  - Esophagitis, dysphagia
  - Barrett esophagus
  - Pulmonary complications
- Endoscopy with biopsies is mandatory for routine monitoring of GERD in patients with EA.
- Macroscopic abnormalities: at least 4 biopsies in each quadrant 1 cm above the Z line

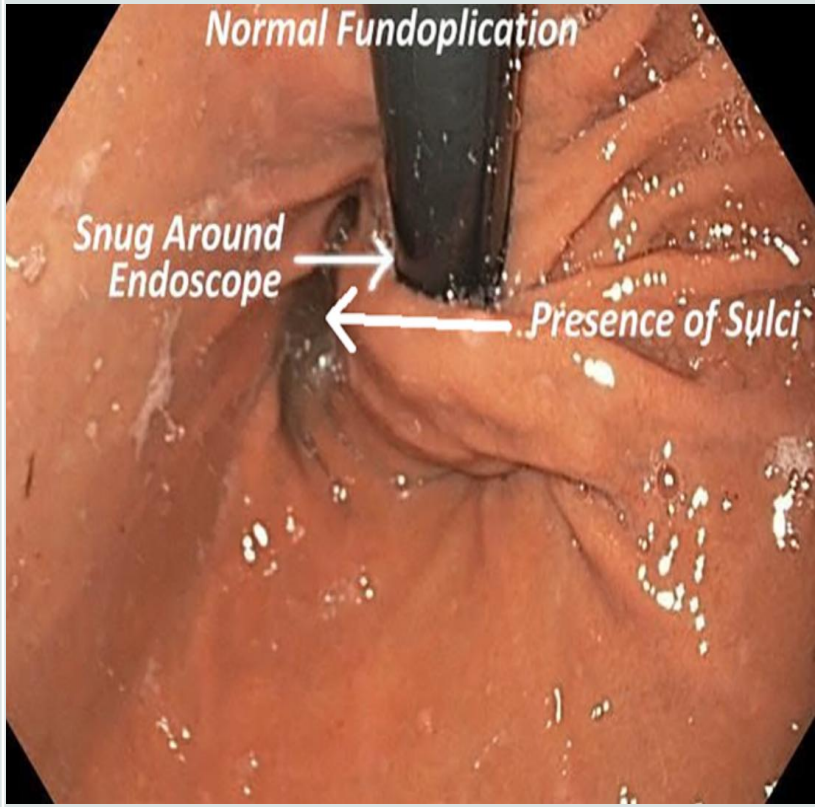

Symptomatic patient with EA
1
Esophagram +/– Swallow study
2
Abnormal ?
3
Yes
Refer to Statements and treat accordingly
4
No
EGD +/– bronchoscopy +/- laryngoscopy
5
Stricture?
Stenosis?
Tracheomalacia?
Fistula?
Vascular compression?
Vocal cord paralysis?
Laryngeal cleft?
6
No
Esophagitis?
7
Yes
Peptic
Eosinophilic
Infectious
8
Refer to statements and treat accordingly
No
pH-MII probe (symptoms correlation)
9
Reflux?
10
Yes
Consider:
PPIs
Prokinetics
Feeding modifications
Transpyloric feeding
Fundoplication
Periodical reassessement
11
No
Consider:
Esophageal high resolution manometry and Esophageal dysmotility
Gastric dysmotility
Behavioral feeding disorders
Functional disorders
other comorbidities (cardiac, ..etc)
12

Fundoplication in EA patients with GER

Severe esophageal dysmotility predisposes EA patients to post-fundoplication complications:

- preventing gravity-driven esophageal clearance
- worsen respiratory symptoms

- Anti-reflux surgery indications:
  - Failure of maximum conservative therapy for GER
  - Failure to thrive
  - Acute life-threatening event (ALTE)
  - Esophagitis
  - Recurrent anastomotic stenosis



What evaluations should be performed before fundoplication?

At least

- Barium-contrast study
- Endoscopy with biopsies
- pH-metry

Extra-esophageal manifestations in EA patients

GI causes of pulmonary symptoms:

- Aspiration due to mucus or food retention in the proximal pouch or distal esophagus
- Anastomosis Stricture
- Impaired esophageal motility
- Congenital esophageal stenosis
- Aspiration during swallowing
- GER
- Recurrent or missed fistulae
- Eosinophilic esophagitis
- Esophageal pooling over a fundoplication

Non-GI causes:

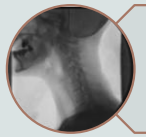
- Vocal cord paralysis
- Laryngeal clefts
- Vascular rings
- Aspiration during swallowing

EA patients with respiratory symptoms: Ruled out of anatomic abnormalities

- laryngeal cleft
- anastomotic stricture
- vocal cord paralysis
- vascular ring
- missed or recurrent fistulae
- congenital stenosis

How should clinicians investigate extra-esophageal manifestations in EA patients?

- Diagnostic testing:


Modified barium swallow (video fluoroscopic swallow studies)


Fiber optic endoscopic evaluation of swallowing

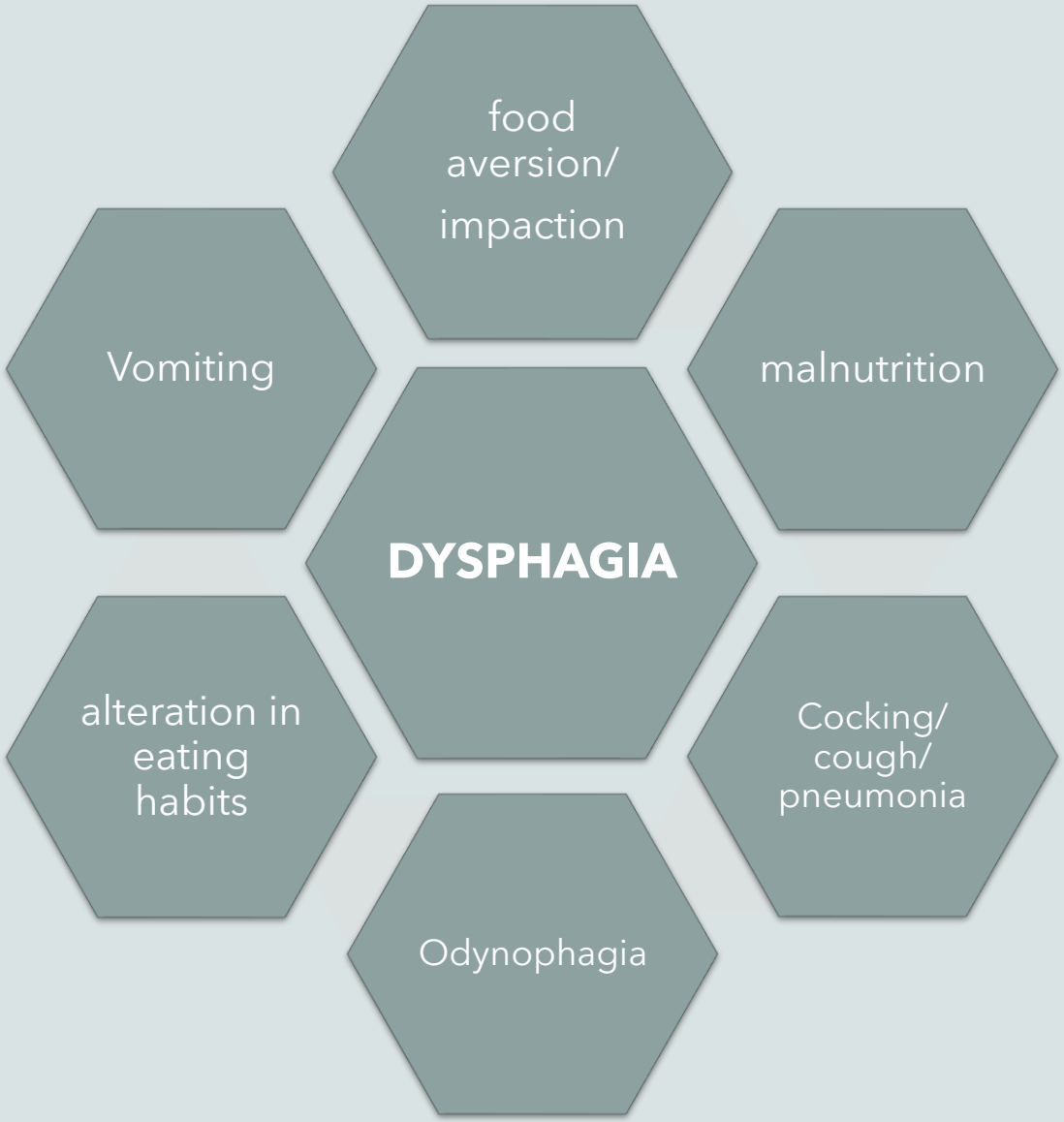

Rigid bronchoscopy

Dysphagia and esophageal function in EA

The etiology of dysphagia

- Inflammatory or anatomic causes such as peptic esophagitis
- Eosinophilic esophagitis
- Anatomic stenosis
- Congenital stenosis
- Post-fundoplication obstruction
- Vascular anomalies
- Anastomotic diverticulum
- Inlet patch

- Esophageal motility: contrast study, esophageal manometry or video fluoroscopy
- The pattern of esophageal dysmotility ≠ presence or severity of dysphagia
- The underlying cause of the dysmotility
    1. Intrinsic factors related to abnormal development of the esophagus
    2. Operative maneuvers responsible for a partial denervation
    3. Postoperative complications(local trauma and inflammation leading to further neuronal and muscular damage)


food aversion/ impaction
Vomiting
malnutrition
DYSPHAGIA
alteration in eating habits
Cocking/ cough/ pneumonia
Odynophagia

Investigation of dysphagia in EA patients

1. Esophagography: distal CES
2. Endoscopy with biopsies :
   - anastomosis (stricture, diverticulum)
   - esophageal mucosa (peptic, eosinophilic or infectious esophagitis)
   - diagnosis of other etiology
3. HR esophageal manometry: dysphagia in EA patients who have a normal esophagogram and endoscopy with biopsy

Dysphagia management in EA patients

Treatment options:

- Treatment of esophagitis (peptic, eosinophilic, or infectious) and inlet patch
- Prokinetics
- Treatment of stricture, stenosis, mucosal bridge, or anastomotic diverticulum
- Surgical repair of vascular anomaly
- Gastrostomy tube feeding
- Esophageal replacement
- Dilation of fundoplication
- Feeding adaptation

Feeding and nutrition in EA patients

- Feeding difficulties in EA are multifactorial:
  - Oropharyngeal disorders
  - Esophageal disorders
  - Behavioral disorders
- Fundoplication: create a functional esophageal outlet obstruction in the context of dysmotility
  - ➢ cause dysphagia and feeding difficulties in children with EA
- The most efficacious methods of avoiding feeding disorders: ?

Risk of malnutrition

1. Comorbidities (cardiac, genetic, neurologic), which may have a large impact on growth
2. A history of GER and low birth weight were both predictors of reduced growth

- Intensive early enteral and oral nutrition intervention and advances in neonatal care and surgery have reduced the risk of long-term malnutrition in children with EA

Eosinophilic esophagitis (EoE) in EA

- The largest reported number in studies: 17% incidence (greater than the reported incidence of EoE in the general pediatric population)
- Etiology:
  - possible genetic association
  - impairment of esophageal mucosal barrier function by acid refluxate
  - prolonged exposure to acid suppressive medication

- Misdiagnosis or delayed diagnosis due to presenting symptoms of EoE similar to GER
- Typical macroscopic findings of EoE may not be seen in all EA patients
- Management of EA patients with EoE should follow consensus recommendations for treatment of EoE in the general population

An anastomotic stenosis

Most frequent post-operative complication

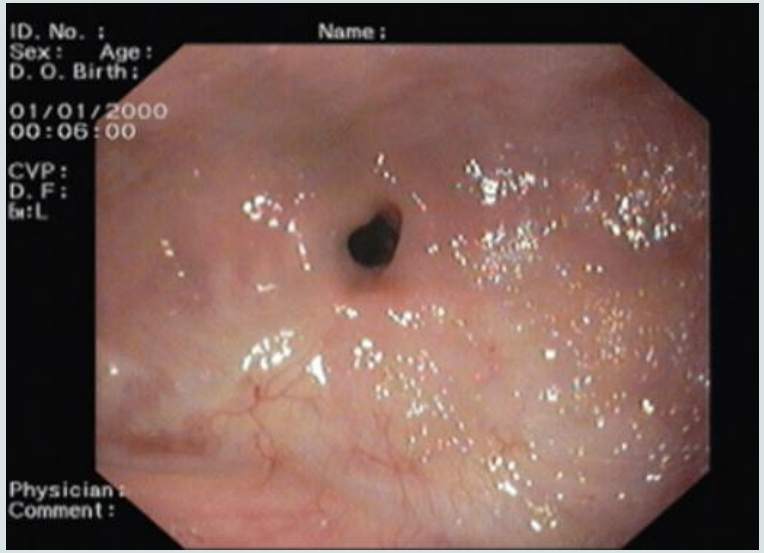
Possible risk factors:

1-Long gap

2-Delayed anastomosis

3-Postoperative anastomotic leak

4-GER



Symptoms

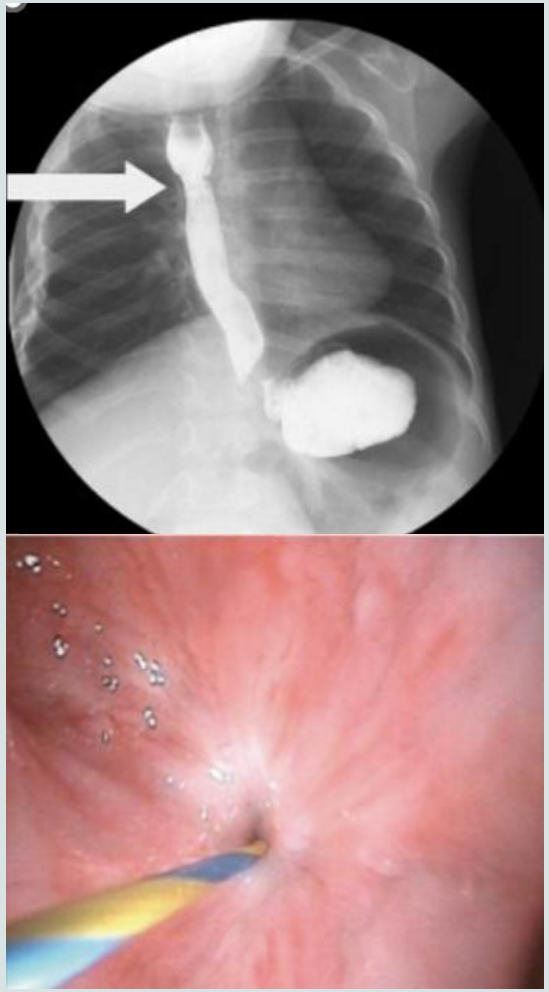
- Feeding and swallowing difficulties
- Regurgitation and vomiting
- Mucus or food retention in the proximal pouch
- Cough
- Drooling
- Recurrent respiratory infections
- Foreign body impaction
- Poor weight gain

➢ Close follow-up of all EA patients in the first 2 years of life with special attention to symptoms suggestive of AS

How should anastomotic stricture be diagnosed in EA?

- Contrast radiographs:
    1. Esophageal morphology
    2. Associated CES and planning a patient specific therapeutic strategy

- Endoscopic :combined diagnosis and treatment with dilation



Management of anastomotic stricture

- First line of therapy: Anastomotic dilation

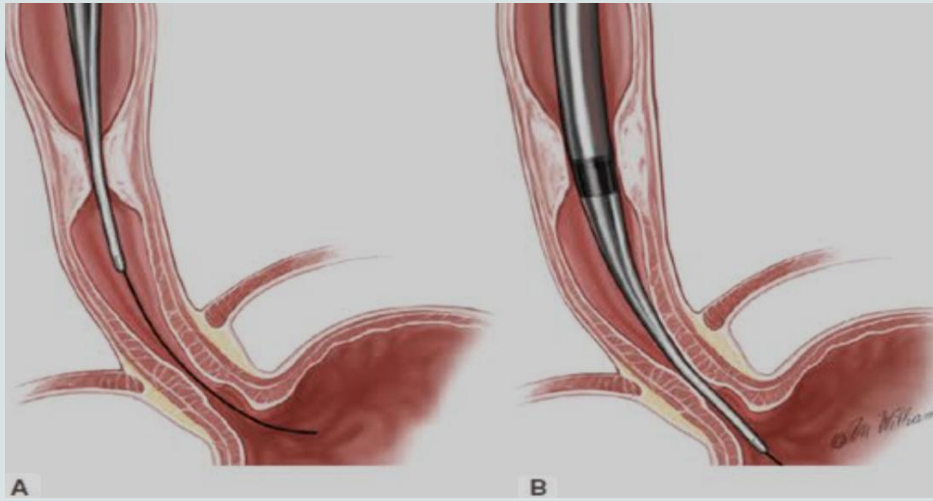
- Aim of dilation: obtain an esophageal diameter that allows a normal, age-appropriate capacity for oral feeding, without respiratory or digestive symptoms

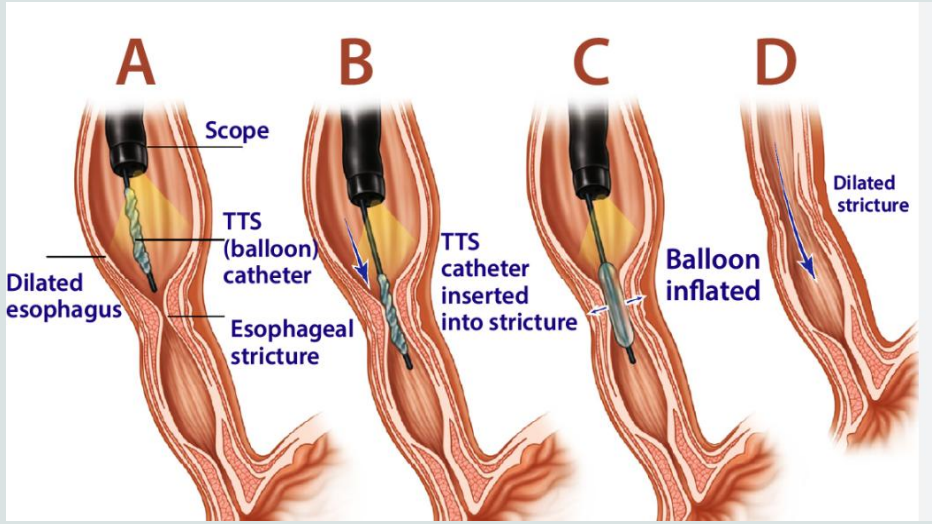
ENDOSCOPIC DILATION

- Patient preparation:
  1. Adjusting medications especially in whom with cardiac anomaly
  2. Antibiotic prophylaxis: not needed in Most patients

Types of dilators

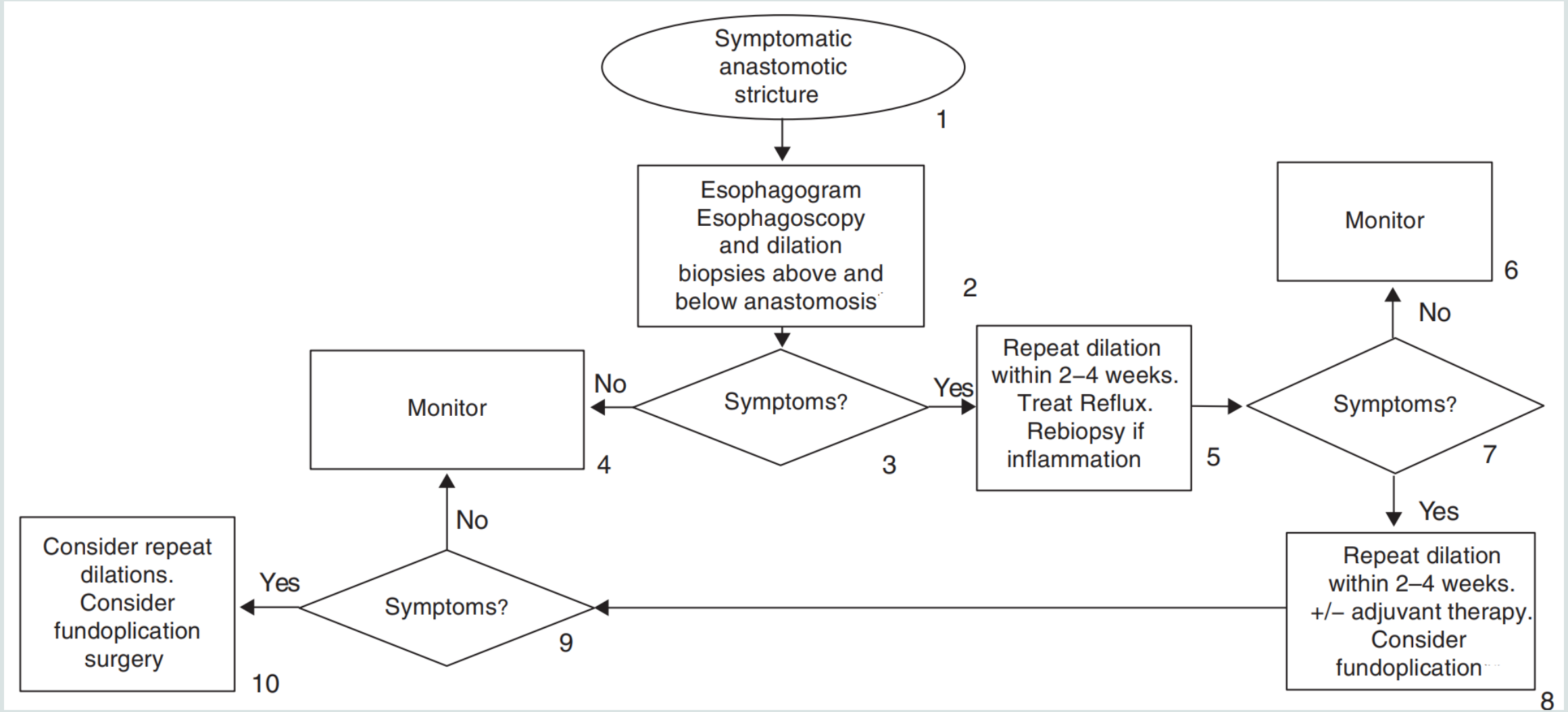
1. PUSH DILATORS:
    - Wire-guided or non-wire-guided dilators
    - Longitudinal and radial force from the proximal to the distal end of the stricture
2. BALLOON DILATORS:
    - Gradual dilation in 1 to 1.5 mm increments with three sizes per balloon

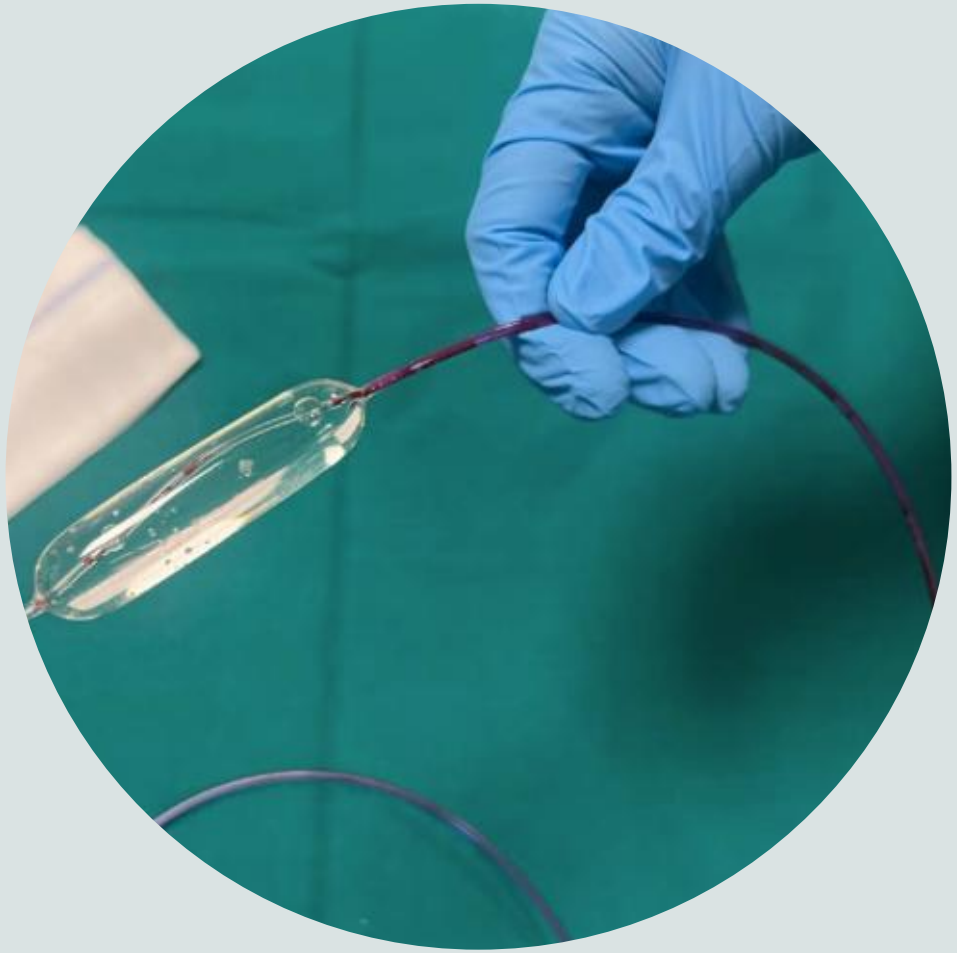




Parameters influencing the decision to choose between balloon or push dilator:

- Stenosis characteristics :
  - Stenosis length less than 1-2 cm, both methods are suitable, but in long stenosis, the balloon is more controllable.
  - shape and stiffness: push dilators work for softer stenosis
- History: Review of previous occurrences and presence of fibrosis


Symptomatic anastomotic stricture
1
Esophagogram Esophagoscopy and dilation biopsies above and below anastomosis
2
Symptoms?
3
No
Monitor
4
Yes
Repeat dilation within 2–4 weeks. Treat Reflux. Rebiopsy if inflammation
5
Symptoms?
7
No
Monitor
6
Yes
Repeat dilation within 2–4 weeks. +/– adjuvant therapy. Consider fundoplication
8
Symptoms?
9
No
Monitor
Yes
Consider repeat dilations. Consider fundoplication surgery
10



Through-the-scope balloon dilators

- Diagnostic endoscopy to determine the location, length, and extent of the stricture.
- Preventing proximal migration of the balloon during inflation
- Maintain insufflation for 30 to 60 seconds per dilation
- Next size : If the inflated balloon can be moved freely through the stricture

Determinants of success:

- Length and diameter of the stenosis
- Time of first dilation
- Absence of severe reflux and active inflammation

Frequency and total number of endoscopic sessions:

- Rule of 3:
  - Maximum 3 dilation per session
  - At least 3-week intervals
  - Increase in diameter by up to 3mm per session
  - 3-5 sessions before other intervention

REFRACTORY STRICTURES

Initial treatment

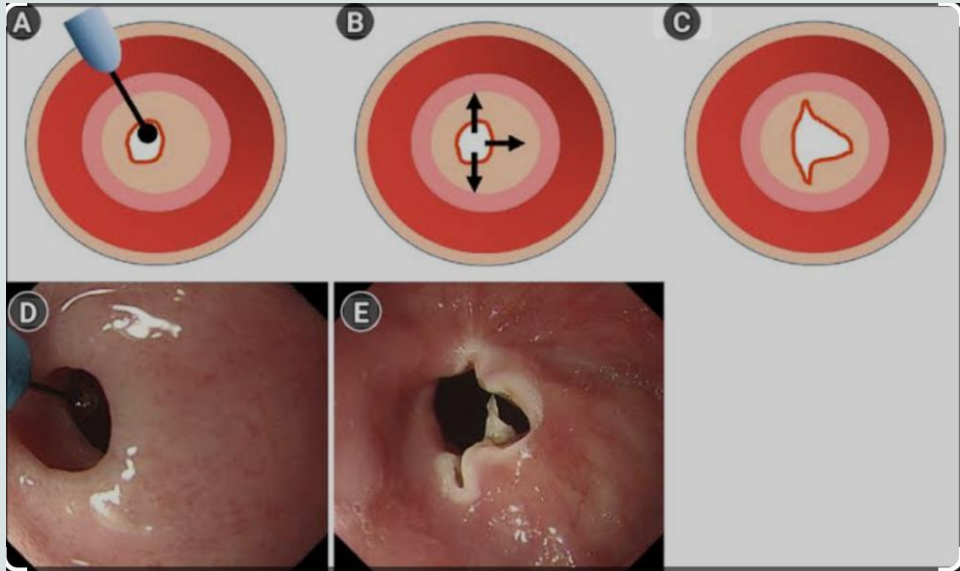
- Glucocorticoid injection into the stricture and endoscopic dilation during one endoscopic session
- Inhibiting collagen deposition and enhancing its breakdown locally, thereby reducing scar formation
- Lower risk of stricture recurrence and fewer endoscopic dilation sessions

Mitomycin c:

- Inhibit fibrosis
- Injection (or topical sponge application) at the stricture site after dilation
- Common concentration: 0.1 mg/ml(0.4 mg/ml in some literature)
- Volume used: 1-2 ml for the entire narrowed area
- Duration of contact with mucosa: 2-5 min
- Indication: resistant fibrosis after more than 3 dilation sessions

Endoscopic incisional therapy (EIT)

- For treating short (<1 cm) anastomotic strictures
- Fibrotic rim of the stricture is directly incised in a radial fashion using a needle-knife or snare tip
- Indication: Thick annular or high resistance strictures (more than 4 previous dilations) and no response to previous cases



Esophageal stent

FCSEMS:

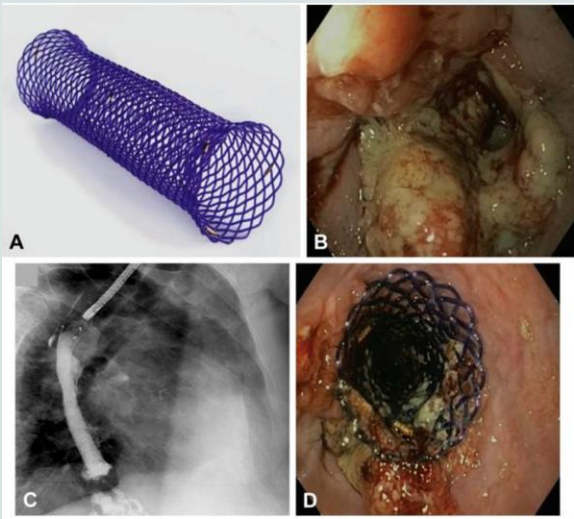
- Fully covered nature facilitates stent removal
- After placement: remain in place for at least six to eight weeks (but not to exceed 12 weeks) before endoscopic removal.

Plastic stents:

- Silicone coated self-expandable plastic stent
- After placement, self-expandable plastic stents typically remain in place for at least six to eight weeks (but not to exceed 12 weeks) before endoscopic removal

Biodegradable stents

- made from woven surgical suture material (polydioxanone) which is degraded by hydrolysis in 8 to 12 weeks.



Complication of advanced technique

- Esophageal leak/perforation: EIT or dilation
- Infection: mitomycin or steroid injection
- Local necrosis, migration: stent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