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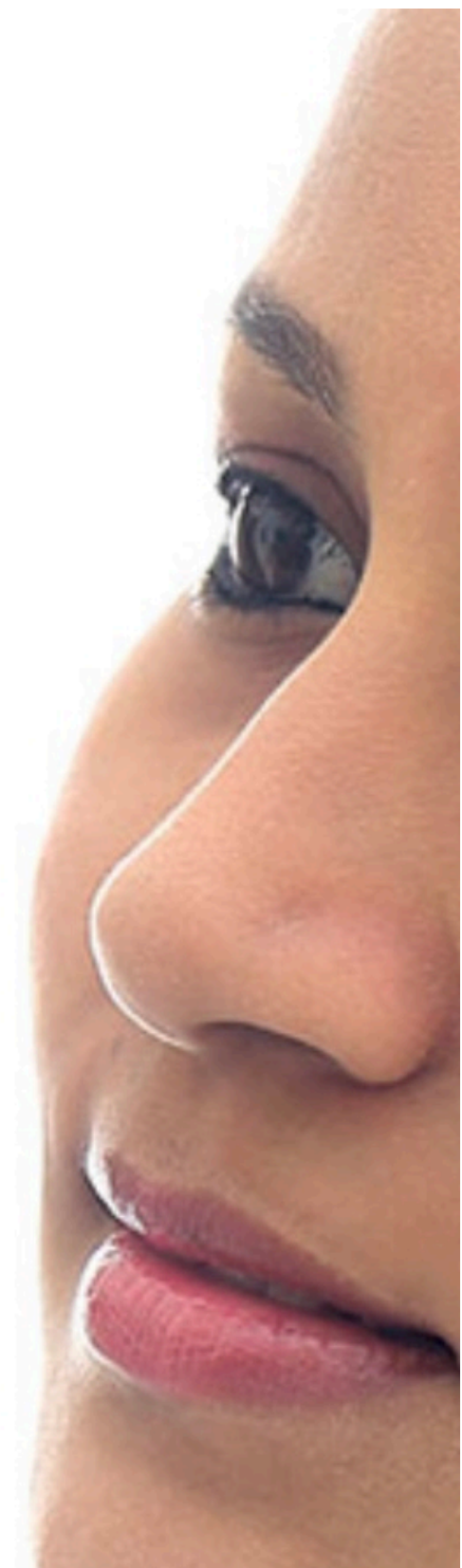
Management of Esophageal Stenosis





Definition of Esophageal Stricture

- *Esophageal stricture is a narrowing of the esophageal lumen causing dysphagia.*
- *Can be congenital or acquired*
- *Luminal (Intrinsic) vs Extrinsic (Extramural)*





Congenital Esophageal Strictures

- *Types: Tracheobronchial Remnants (TBR), Fibromuscular Stenosis (FMS), Membranous Webs*
- *Often present in neonates or infants with feeding difficulties.*

Acquired Esophageal Strictures

- *GERD-related strictures*
- *Caustic ingestion injuries*
- *Postsurgical or radiation-induced strictures*
- *Eosinophilic esophagitis*

Classification of Esophageal Strictures



Luminal (Intrinsic) Stricture:

Narrowing inside the esophageal lumen.

Common causes: GERD, caustic injury, surgery, congenital defects.

Extrinsic (Non-Luminal) Stricture:

Compression of the esophagus from outside.

Common causes: Vascular rings, mediastinal masses, enlarged lymph nodes or thyroid.

Management: Treat the underlying cause, surgery if needed.

Clinical Aspects of Esophageal Developmental Anomalies



ANOMALY	AGE AT PRESENTATION	PREDOMINANT SYMPTOMS	DIAGNOSIS	TREATMENT
Isolated atresia	Newborns	Regurgitation of feedings Aspiration	Esophagogram * Plain film: gasless abdomen	Surgery
Atresia + distal TEF	Newborns	Regurgitation of feedings Aspiration	Esophagogram * Plain film: gas-filled abdomen	Surgery
H-type TEF	Infants to adults	Recurrent pneumonia Bronchiectasis	Esophagogram * Bronchoscopy †	Surgery
Esophageal stenosis	Infants to adults	Dysphagia Food impaction	Esophagogram * Endoscopy †	Dilation ‡ Surgery §
Duplication cyst	Infants to adults	Dyspnea, stridor, cough (infants) Dysphagia, chest pain (adults)	EUS * MRI/CT †	Surgery
Vascular anomaly	Infants to adults	Dyspnea, stridor, cough (infants) Dysphagia (adults)	Esophagogram * Angiography † MRI/CT/EUS	Dietary modification ‡ Surgery §
Esophageal ring	Children to adults	Dysphagia	Esophagogram * Endoscopy †	Dilation ‡ Endoscopic incision §
Esophageal web	Children to adults	Dysphagia	Esophagogram * Endoscopy †	

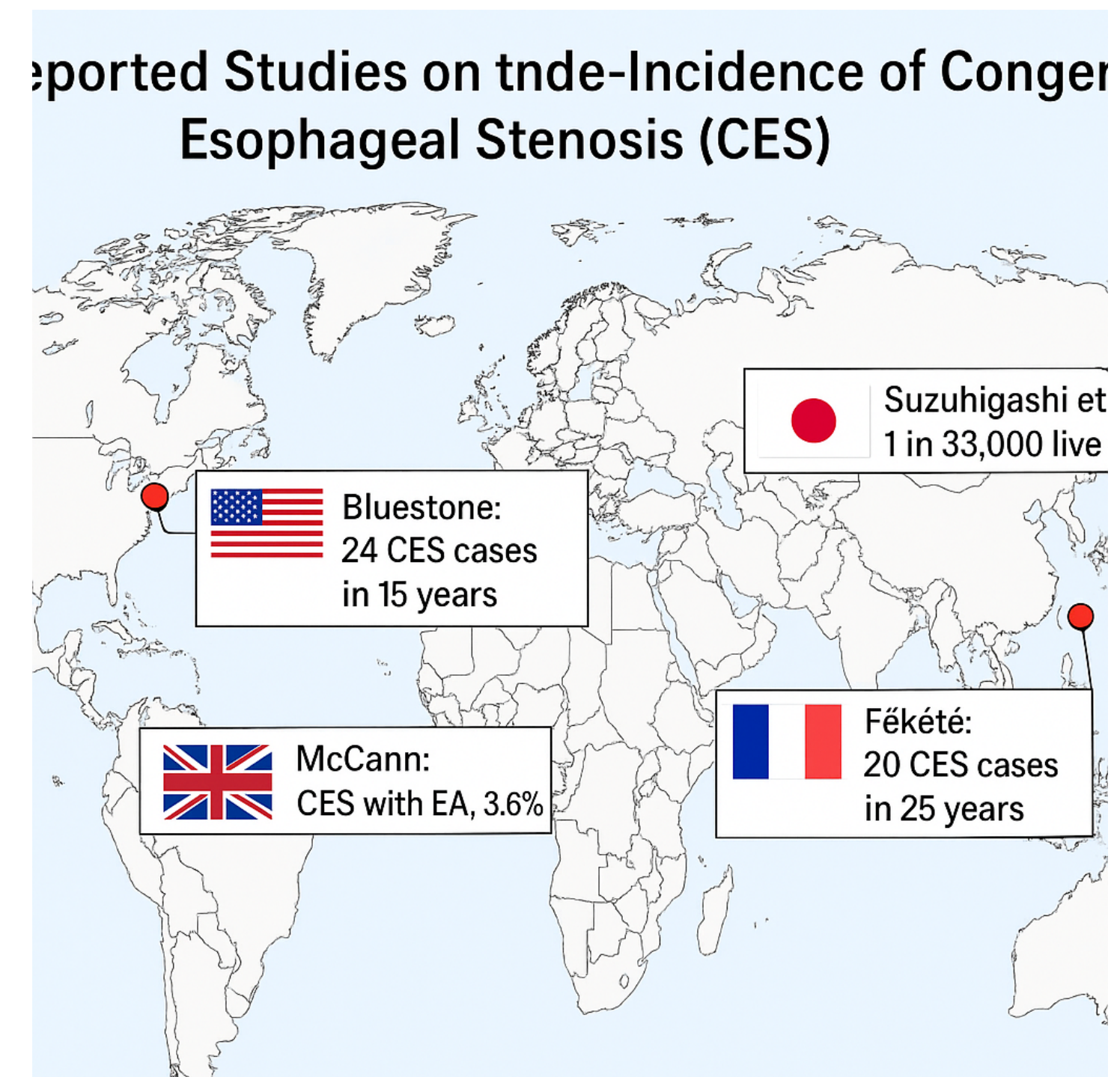
What is Congenital Esophageal Stenosis (CES)?

CES is a congenital narrowing of the esophagus present at birth, but not necessarily symptomatic



Incidence of Congenital Esophageal Stenosis (CES)

- *Estimated incidence: 1 in 25,000 to 50,000 live births*
- *Suzuhigashi study (Japan): 1 in 33,000 live birth*
- *CES occurs at a rate of about 1 per 20–25 EA cases*





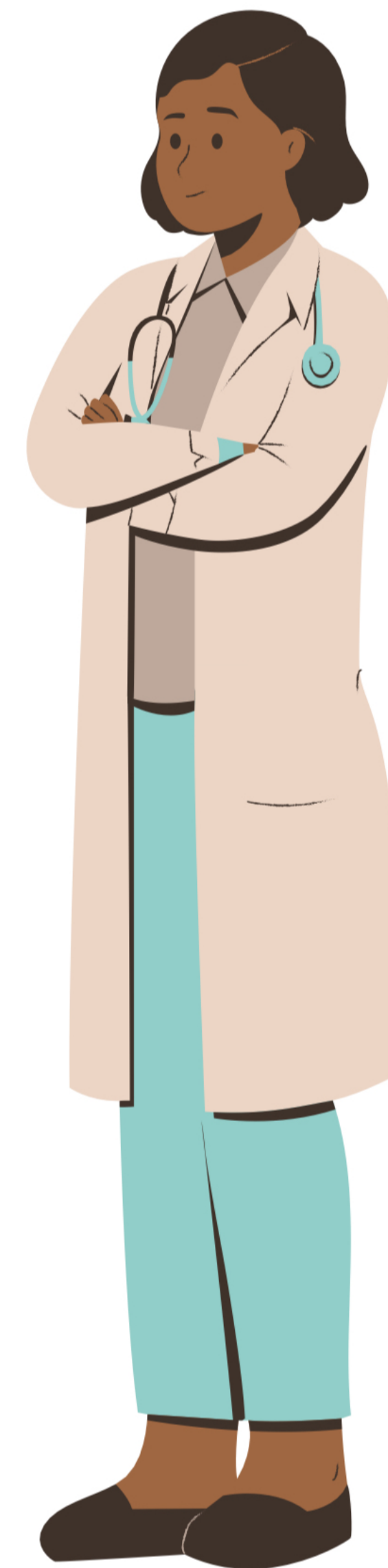
Types of Congenital Esophageal Stenosis (CES)?

1. *Membranous Webs*
 - *Thin mucosal diaphragm*
 - *Usually in the upper or mid-esophagus*
 - *May be asymptomatic or cause intermittent dysphagia*
2. *Fibromuscular Thickening*
 - *Most common type*
 - *Segmental hypertrophy of submucosa and muscularis propria*
 - *Typically located in the distal esophagus*
3. *Tracheobronchial Remnants (TBR)*
 - *Presence of cartilage or respiratory epithelium within the esophageal wall*
 - *Often associated with a rigid, non-dilatable stricture*
 - *May mimic acquired strictures; diagnosis confirmed histologically*

Initial Diagnostic Workup

1. Clinical History & Symptoms

- *Dysphagia (especially after starting solid food)*
- *Regurgitation or vomiting after meals*
- *Recurrent respiratory infections (aspiration)*
- *Poor weight gain or failure to thrive*



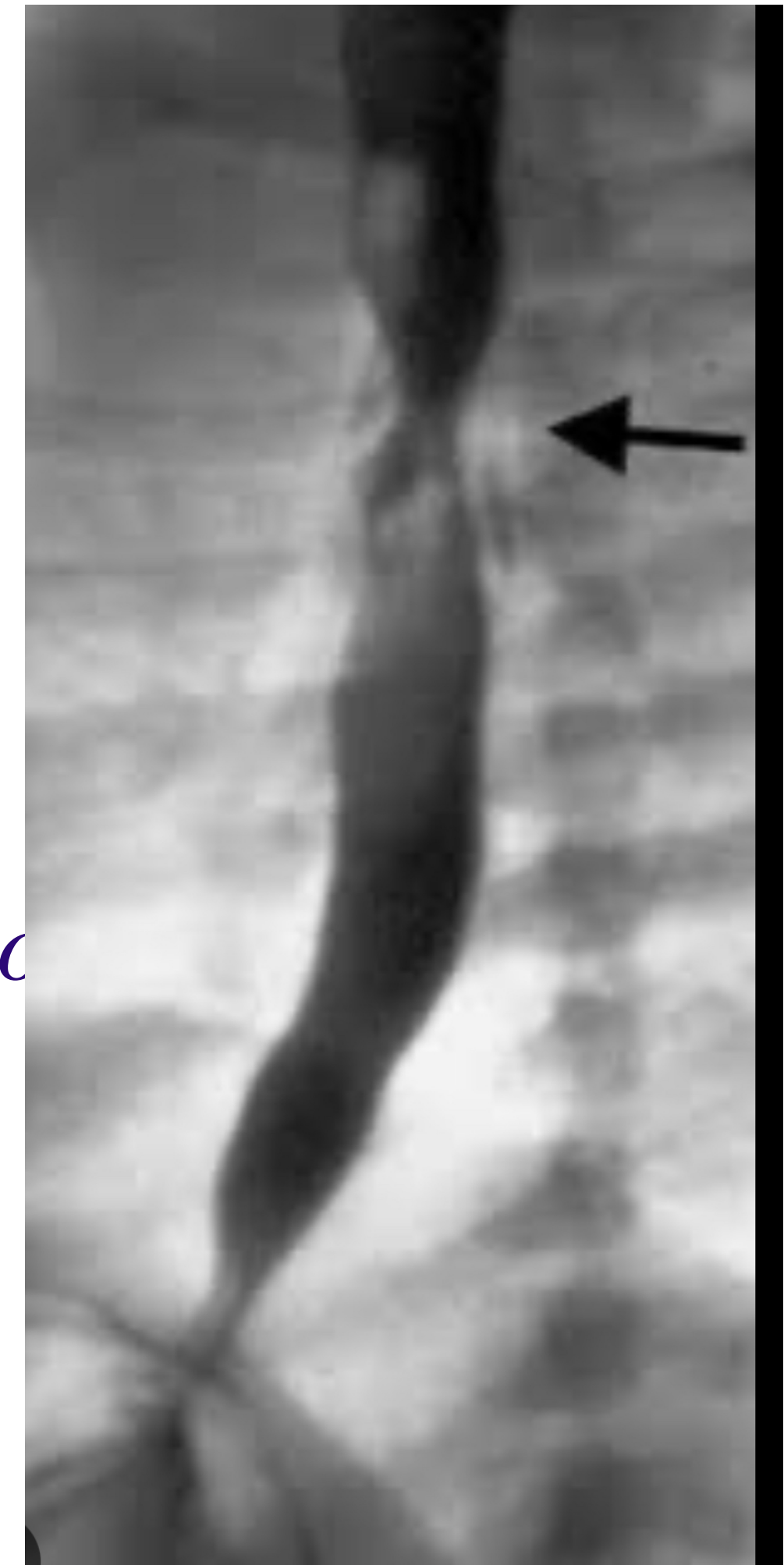
Initial Diagnostic Workup



2. Barium Swallow (Esophagogram) First-line imaging study

3. Upper GI Endoscopy

- *Endoscope meets resistance at the stenotic area*
- *Useful to rule out reflux-induced strictures or inflammation*



4. Endoscopic Ultrasonography (EUS)

- *Mini-probe or EUS can help distinguish subtypes*
- *Critical for deciding between dilation and surgery*

5. Histopathology (if surgical resection is done)

- *Confirms type of CES*

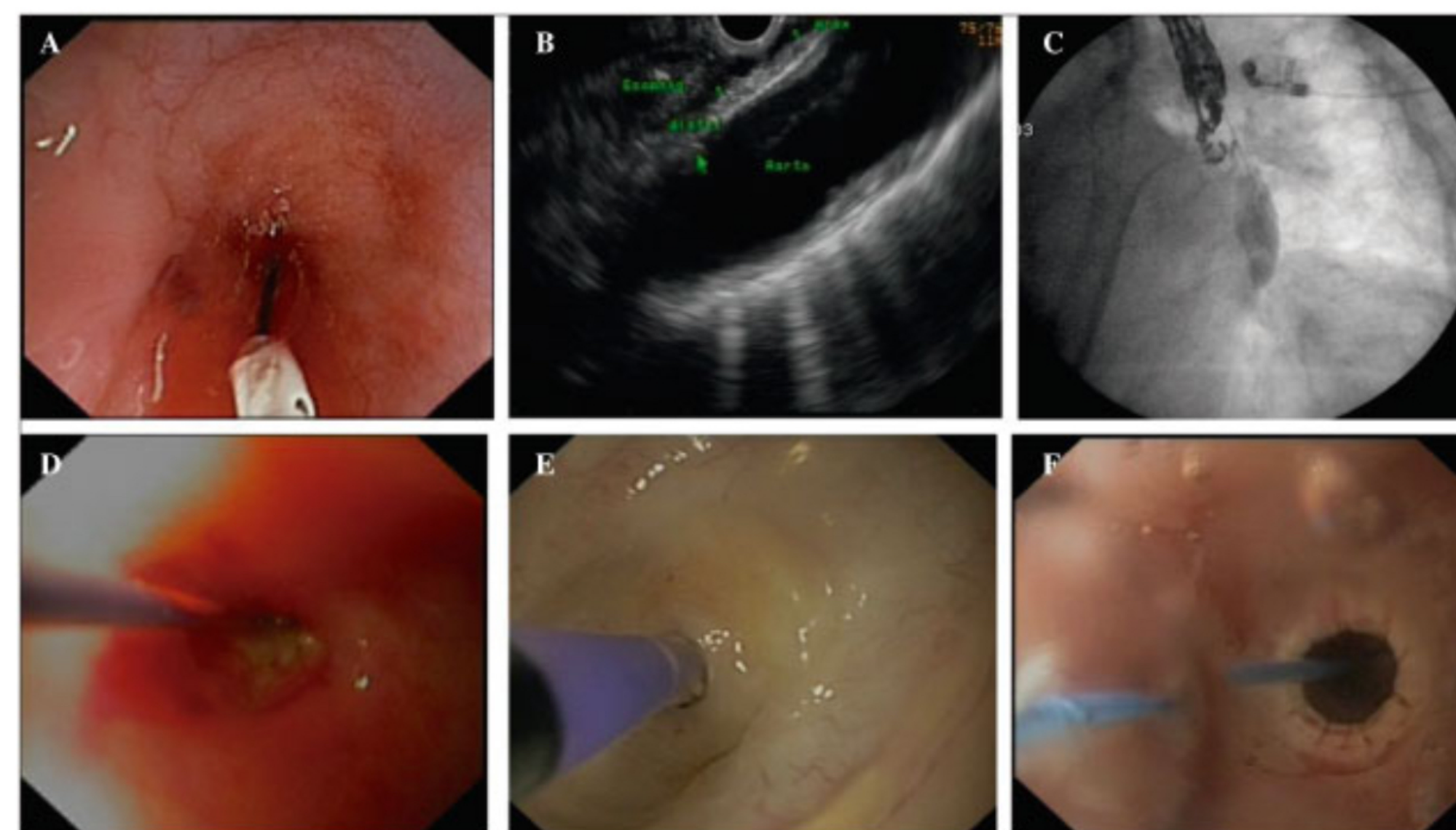
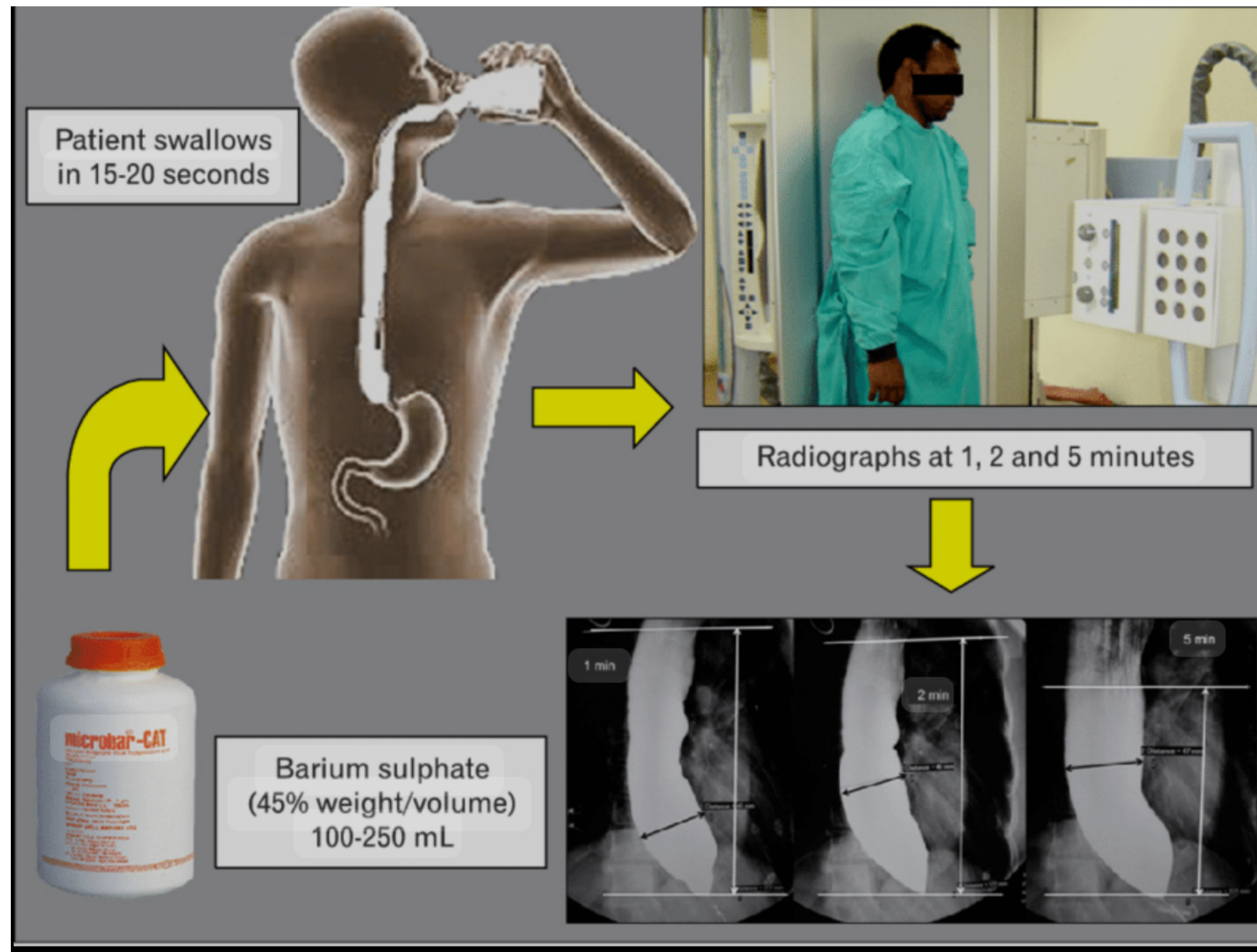


Fig. 1. Case #1. Complete esophageal stenosis successfully recanalized with EUS-guidance. A. Endoscopic view of a total esophageal stenosis. B. Endoscopic ultrasound (EUS) view of the esophageal stricture. C. Fluoroscopic image of contrast verification. D. 0.035-inch guide wire within the stenosis, and F. Balloon dilation up to 12 mm.

Barium Swallow Appearance





Barium Swallow Appearance in CES Subtypes

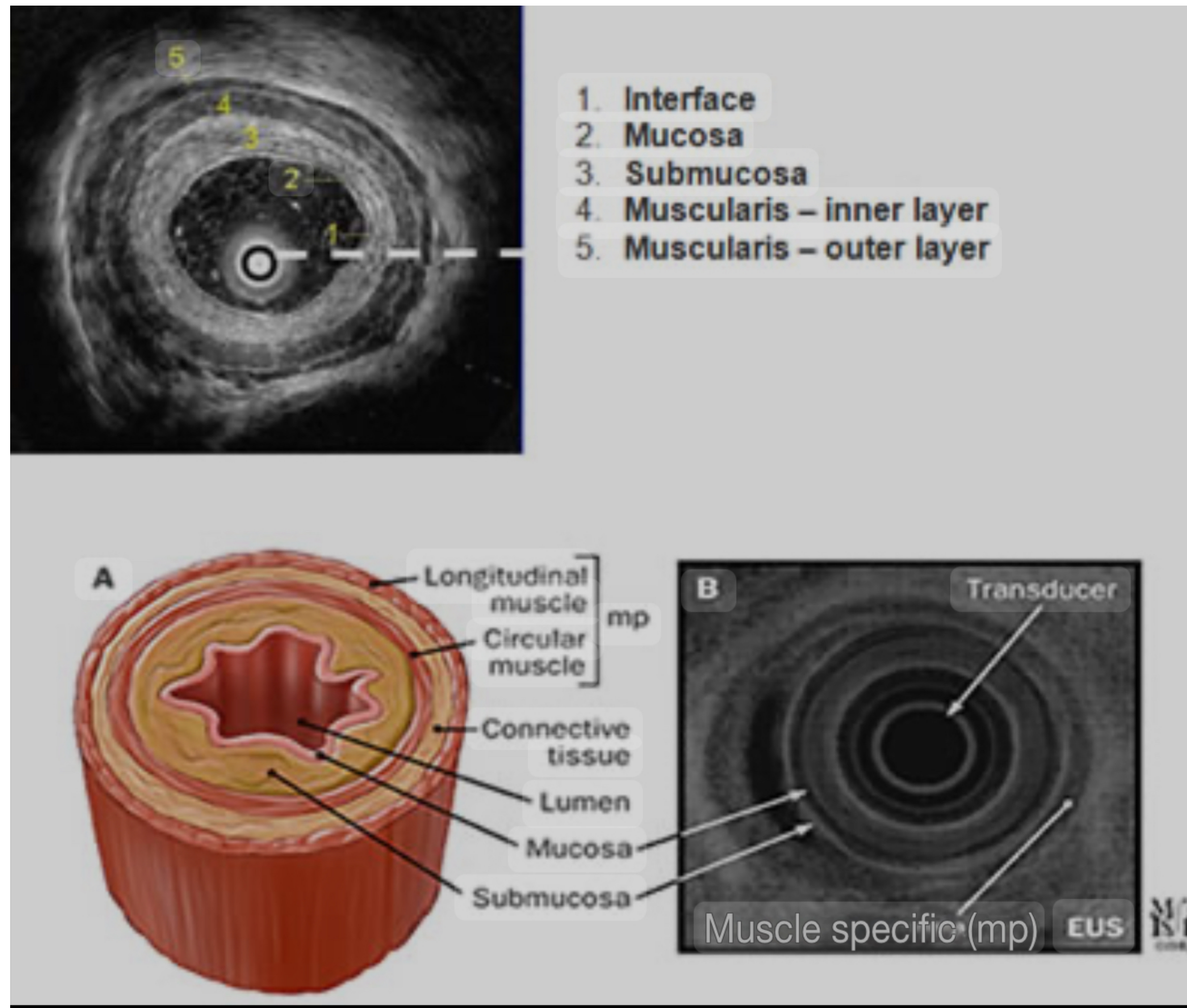
- **Fibromuscular Stenosis (FMS):**
 - Location: Distal esophagus
 - Barium Findings: Short, smooth, symmetric narrowing with gradual tapering
- **Tracheobronchial Remnants (TBR):**
 - Location: Distal esophagus
 - Barium Findings: Irregular, asymmetric narrowing; sometimes serrated appearance
- **Web (Membranous Diaphragm):**
 - Location: Mid or upper esophagus
 - Barium Findings: Thin, sharp ring-like filling defect; often transient and mobile

Comparison Table – Barium Swallow Findings in Esophageal Strictures



Feature	CES	Intraluminal Strictures	Extraluminal Compression
Appearance on Barium	Short, smooth, tapered stenosis	Irregular, long/multiple strictures	Smooth, rounded indentation
Mucosal Involvement	Normal mucosa	Mucosal irregularity present	No mucosal disruption
Common Causes	Congenital (e.g., TBR, FMS)	EoE, caustic injury, inflammation	Vascular ring, mediastinal mass
Diagnostic Tools	Barium swallow ± Endoscopy	Endoscopy with biopsy	CT, MRI, Echocardiography (if vascular)
Common Misdiagnoses	Achalasia, peptic stricture	GERD-related stricture, EoE	Tumor, vascular anomalies

EUS





Role of EUS in Diagnosing Congenital Esophageal Stenosis (CES)

- Endoscopic Ultrasound (EUS) offers high-resolution imaging of esophageal wall layers and adjacent structures — helpful in differentiating CES subtypes
- Non-invasive differentiation of subtypes before histologic confirmation.
- Particularly useful when:
 - Dilation fails or is incomplete.
 - Planning for surgical resection (e.g., TBR suspicion).
- Can help avoid unnecessary or ineffective dilation attempts.

EUS IN CES



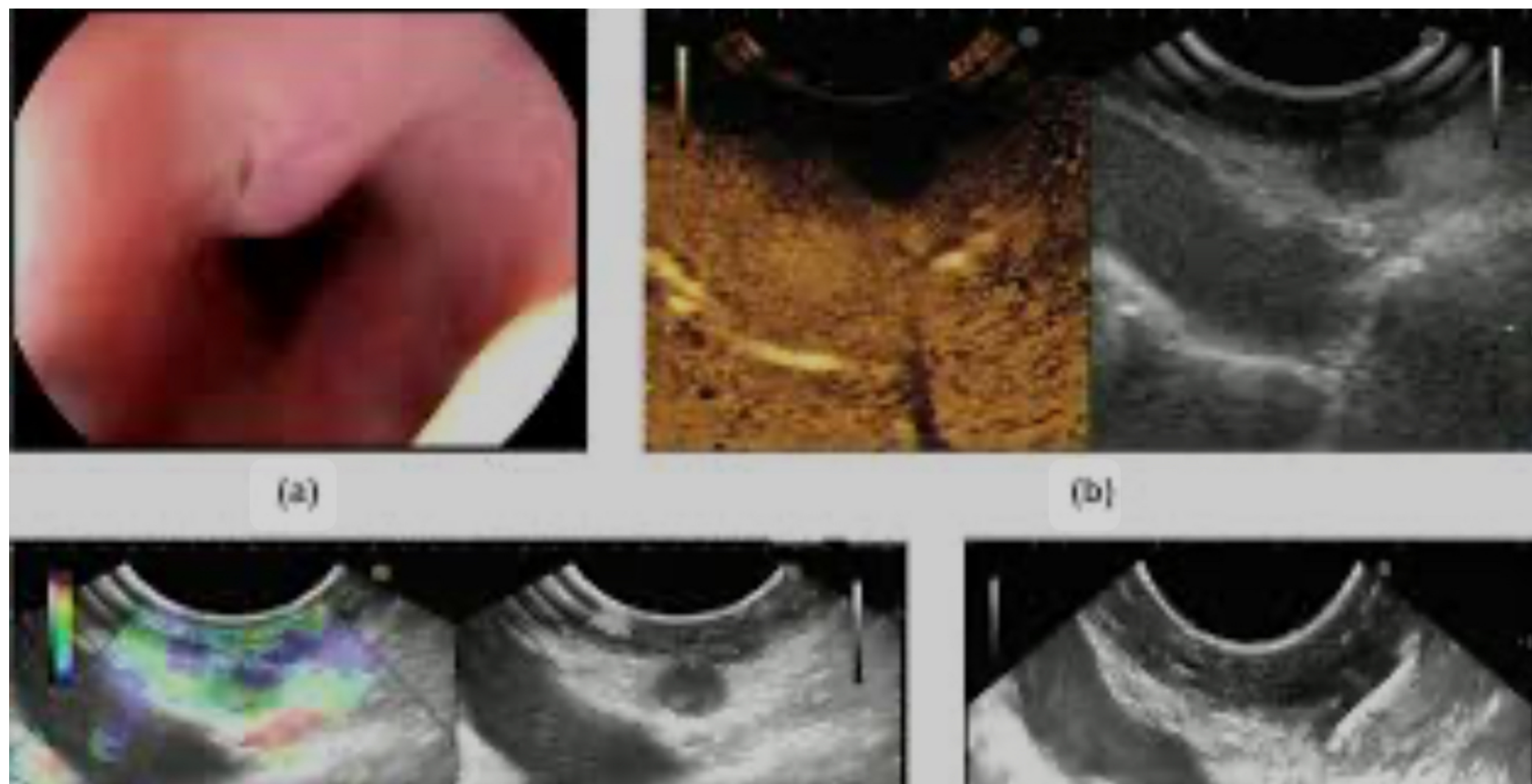
- **Fibromuscular Stenosis (FMS):**
Thickened muscularis propria; mucosa and submucosa remain normal.
- **Tracheobronchial Remnants (TBR):**
Hyperechoic foci or cartilage-like areas within the esophageal wall, possibly with acoustic shadowing.
- **Membranous Web:**
Thin intraluminal membrane involving mucosa or submucosa only; no thickening of deeper layers.



Comparison of Esophageal Stenosis Types on EUS

Type of Stenosis	EUS Findings	Diagnostic Clues
CES (Congenital)	Focal thickening of submucosa or muscularis propria; may show hyperechoic foci if cartilage is present (TBR type)	Often appears as localized wall thickening without mucosal disruption
Intraluminal (e.g. EoE, caustic)	Thickened mucosa $\hat{\pm}$ submucosa; layered structure may be disrupted; hypoechoic infiltration in EoE	Mucosal changes are dominant; EUS helps assess depth of involvement
Extraluminal (e.g. vascular ring, mass)	Normal esophageal wall layers; external compression visualized as extrinsic hypoechoic/anechoic mass or indentation	No intrinsic esophageal wall abnormality seen on EUS

- *“EUS is an advanced but underutilized tool in pediatric esophageal stenosis — it provides crucial structural insight, especially when endoscopy and imaging are inconclusive.”*



- *EUS is a valuable diagnostic tool that helps differentiate the origin of esophageal strictures—whether intrinsic (mucosal/submucosal), muscular, or extrinsic—guiding appropriate management.*

Endoscopic Clues





Endoscopic Clues for Differentiating CES Subtypes

- **Fibromuscular Stenosis (FMS):**
 - Location: Distal esophagus
 - Endoscopic Appearance: Smooth, short narrowing; mucosa intact
 - Response to Dilation: Partial or good response
- **Tracheobronchial Remnants (TBR):**
 - Location: Distal esophagus
 - Endoscopic Appearance: Irregular, firm, non-distensible wall; may feel rigid or cartilaginous
 - Response to Dilation: Poor or no response; suspect TBR if dilation fails
- **Membranous Web:**
 - Location: Mid or proximal esophagus
 - Endoscopic Appearance: Thin, diaphragm-like translucent membrane

Endoscopic Features in Esophageal Stenosis

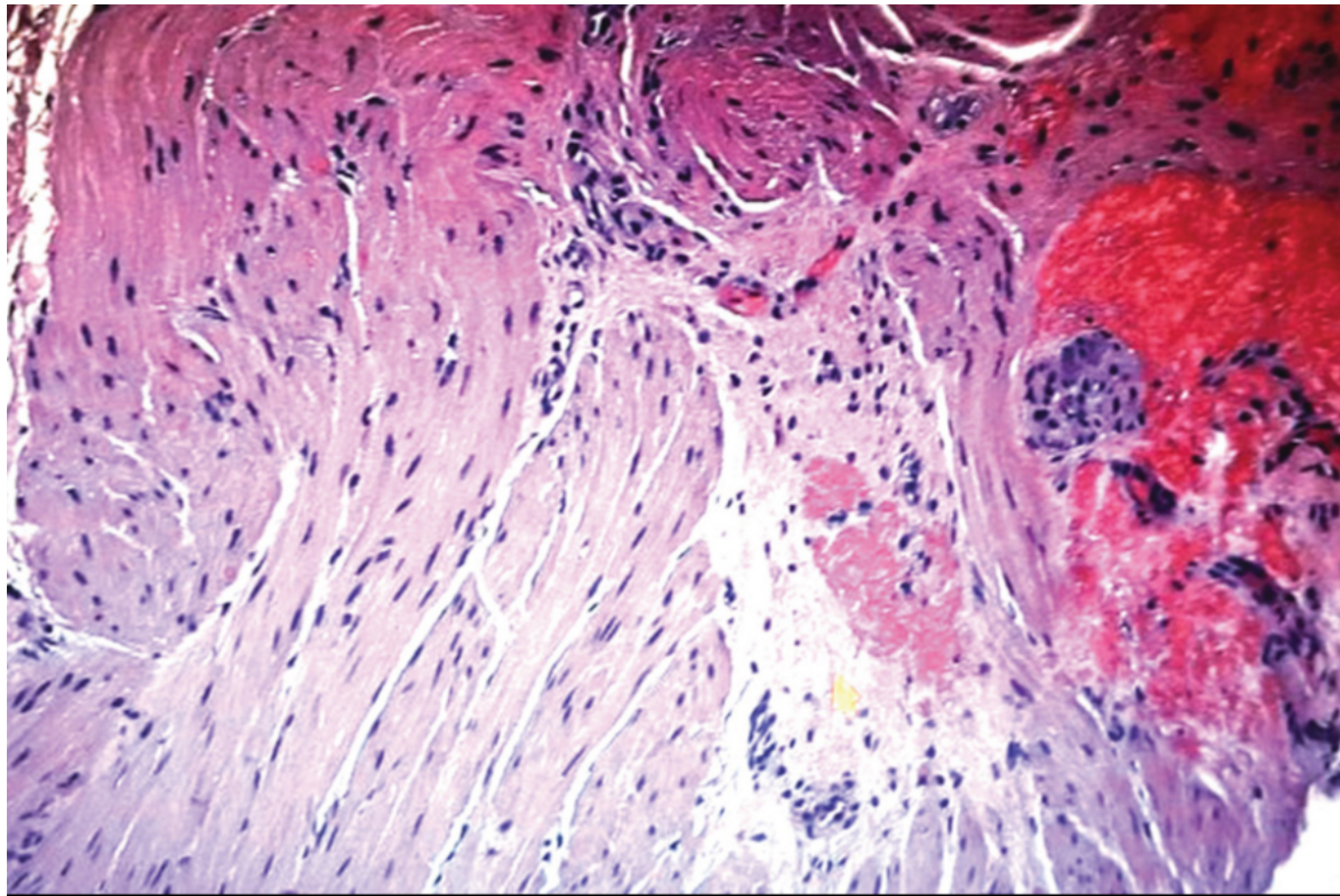


Cause	Endoscopic Appearance	Key Diagnostic Clues
Congenital Esophageal Stenosis (CES)	Narrowed lumen with normal overlying mucosa in many cases (especially FMS); may appear as short segment narrowing; sometimes not easily visible	Biopsy often non-diagnostic; submucosal nature requires deep tissue sampling or resection
Eosinophilic Esophagitis (EoE)	Linear furrows, white exudates, concentric rings (‘‘trachealization’’), mucosal edema	Often patchy; histology shows ≥ 15 eosinophils/HPF; requires multiple biopsies
Caustic Stricture	Long-segment narrowing, irregular or ulcerated mucosa, fibrosis	History of ingestion; strictures often multiple or diffuse; high recurrence risk
Post-Surgical/ Anastomotic Stricture	Circumferential narrowing at surgical site; smooth or fibrotic	History of esophageal atresia/TEF repair; short segment; often recurrent after dilation
Achalasia	Dilated esophagus with tight LES; food residue or retained secretions	Normal mucosa; ‘‘pinpoint’’ LES; confirmed by manometry
External Compression (e.g., vascular ring)	Normal mucosa and wall; impression or narrowing without internal lesion	Biopsy normal; further evaluation with imaging (CT/MRI, EUS)

- *CES often appears deceptively normal on endoscopy — superficial biopsies may miss the diagnosis.*
- *EoE and caustic injuries show clear mucosal abnormalities; biopsies are diagnostic.*
- *Extraluminal compressions show no mucosal change — require imaging beyond endoscopy.*



Histologic Comparison of Esophageal Strictures



Histologic Comparison of Esophageal Strictures

Cause	Key Histologic Findings
Congenital Esophageal Stenosis (CES)	Varies by subtype:
FMS (Fibromuscular):	Submucosal fibrosis and disorganized smooth muscle
TBR (Tracheobronchial remnants):	Presence of cartilage, respiratory glands, or ciliated epithelium
Membranous type:	Thin web-like structure with minimal muscular involvement
Eosinophilic Esophagitis (EoE)	≥15 eosinophils per high-power
Mucosal edema	
Eosinophilic microabscesses may be seen	
Caustic Injury Stricture	Mucosal necrosis, submucosal
Chronic inflammatory infiltrate, sometimes ulceration	
Post-surgical Stricture (e.g., EA repair)	Fibrosis at anastomosis site
“ Chronic inflammation	
“ Sometimes granulation tissue	
Achalasia	Normal mucosa in most cases
“ Minimal inflammation	
Histology not diagnostic; diagnosis relies on manometry	
Extrinsic Compression (e.g., vascular rings)	Normal mucosa and submucosa
“ No intrinsic esophageal changes	

Important Notes on Histology in CES

- Superficial biopsies usually non-diagnostic
- Deep tissue sampling or post-resection essential for accurate diagnosis
- Correct subtype identification prevents ineffective treatments (e.g., avoid dilation in TBR)





Early suspicion, correct imaging timing, and awareness of CES subtypes are essential for diagnosis and effective treatment planning.



CES – Associated Congenital Anomalies

Common Anomalies:

- *Esophageal Atresia (EA) ± TEF*
- *Cardiac defects: VSD, ASD, PDA*
- *Gastrointestinal: Duodenal atresia, malrotation, imperforate anus*
- *Genitourinary: Hypospadias, renal anomalies*
- *Skeletal anomalies: Hemivertebra, limb defects*
- *Chromosomal: Trisomy 21, 18, 13*
- *Syndromes:*
 - *VACTERL association*
 - *Fanconi anemia*

CES Associated with EA



- *Triad complicating management:*
 - *Stricture*
 - *Esophageal dysmotility*
 - *Gastroesophageal reflux (GER)*



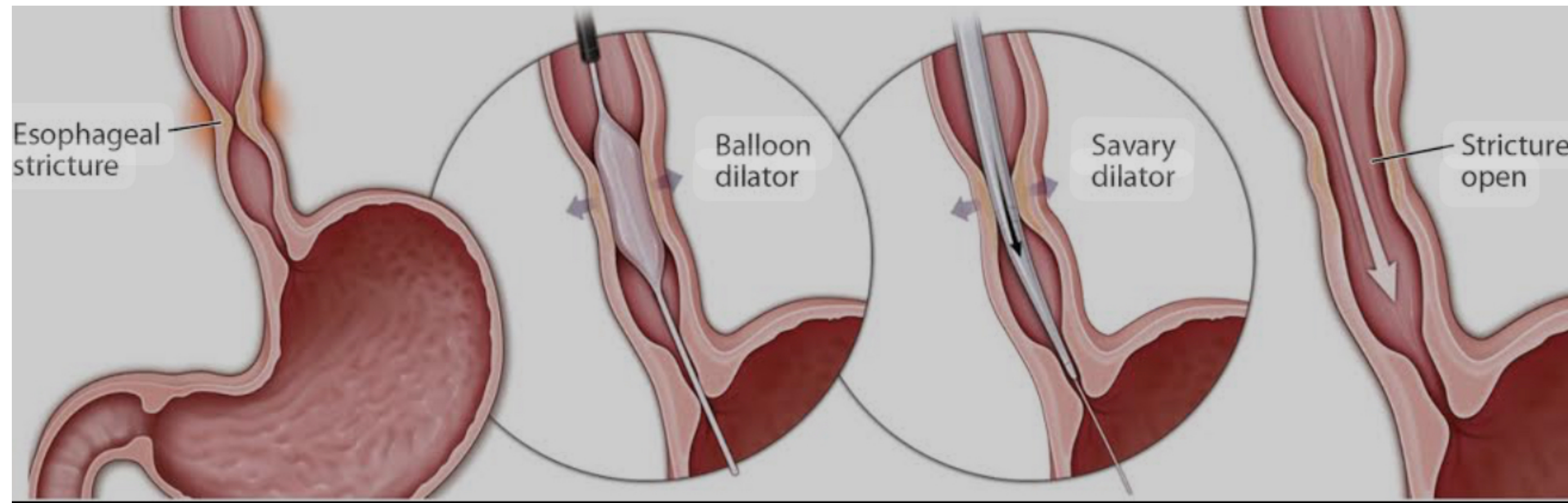


Main Treatment Approaches for CES

- *Two main treatment options:*
 1. *Dilatation (Conservative)*
 2. *Surgical Resection*
- Always evaluate for associated anomalies in CES, especially with EA
- Multisystem assessment is recommended
- **CES with EA is more challenging to treat**

Dilatation as First-Line Treatment

- *First step treatment for all patients*
- *Balloons preferred over bougies due to safer radial force*
- *Bougies useful in long, tortuous, or severely fibrotic strictures*
- *Performed under general anesthesia with endoscopic or fluoroscopic guidance*
- *Use of guide wire to minimize perforation risk*





Normal Diameter by Age:

- *Newborns (0–1 month): around 5 mm*
- *Infants (1–12 months): 6–7 mm*
- *Toddlers (1–3 years): 8 mm*
- *Preschoolers (3–5 years): 10 mm*
- *School-age children (6–10 years): 12 mm*

How to Dilate – Diameter Guidelines:



1. Initial Diameter:

- Start with a dilator or balloon slightly smaller than the measured diameter of the stricture

2. Incremental Rule – “Rule of 3” or “Modified Rule of 5”:

- Traditionally: Do not increase more than 3 mm per session (“Rule of 3”).
- **Newer evidence suggests up to 5 mm is safe in most children (“Rule of 5”), with low risk (<1%) of perforation.**

Target Diameter:

- *Goal is to gradually reach a physiologic lumen*
- *Newborns: ~6–8 mm*
- *Toddlers: ~10 mm*
- *Older children (>5 years): 12–14 mm*





*Comparison of Esophageal Dilation in
Children: EoE vs. Caustic Injury vs.
Congenital Esophageal Stenosis
(CES)*



1. *Eosinophilic Esophagitis (EoE)*

- Esophageal dilation in EoE is reserved for patients with persistent dysphagia or strictures that do not respond to medical therapy (topical steroids or dietary therapy).
- The typical increment per session is 1–2 mm, with a maximum of 3 mm recommended to reduce perforation risk.
→ “It is recommended to perform no more than 3 dilations in 1-mm increments at each session.”

• Dellon ES, Liacouras CA, Molina-Infante J, et al. J Pediatr Gastroenterol Nutr. 2018; 66(1):82–98)

• UpToDate: “Esophageal dilation in eosinophilic esophagitis”)



2. Caustic Esophageal Injury

- *Children with esophageal burns (especially alkali ingestion) often develop long fibrotic strictures.*
- *Some studies allow up to 3 increments of 3 mm per session under fluoroscopic control.*
→ *“Balloon size was gradually increased by 3 mm per dilatation for a maximum of 3 increments.”*
- *In general, dilation should be conservative, with increments of 0.5–1 mm per session to minimize risk of perforation.*
- *The risk of perforation is significantly higher in caustic injuries (1–6%).*

• Rafeey M, et al. J Pediatr Surg. 2016; 51(9):1425–9)

• Merei JM. Pediatr Surg Int. 2003;19(6):442–6



3. *Congenital Esophageal Stenosis (CES)*

- 20–30% of children respond to 1–2 dilations, especially non-TBR types.
- *In TBR type, dilation is usually ineffective, and early surgical intervention is preferred.*
→ “Dilation is often ineffective in tracheobronchial remnants and surgery is usually required.”
- *For responsive types, dilation increment is typically $\leq 1-2$ sizes per session.*
→ “It is generally recommended not to dilate more than 2 French sizes per session.”
- *Risk of perforation is higher in TBR, lower in membranous/fibromuscular types.*

When to Stop Esophageal Dilation:



- *Clinical and Radiological Improvement:*
 - *Discontinue when the patient becomes asymptomatic (no dysphagia, regurgitation, or feeding difficulties).*
 - *Esophageal diameter has reached near-normal for age on imaging (e.g., contrast esophagogram or endoscopy).*
- *Lack of Response or Complications:*
 - *If no significant improvement after multiple dilation sessions.*
 - *Suspected or confirmed presence of:*
 - *Fibrosis*
 - *Cartilage remnants*
 - *Severe scarring or structural abnormality*

What Should Be Done in The Refractory and the Recurrent Esophageal Stricture?



Lewis Spitz was the first to demonstrate a congenital basis for the distal stenosis associated with EA by showing TBR in an esophagectomy specimen which was explained in an embryological basis

The Refractory and the Recurrent Esophageal Stricture

- *No standard definition exists.*
- *Refractory: Failure to achieve 14 mm lumen after 5 dilatations (2-week intervals).*
- *Recurrent: Inability to maintain 14 mm for >1 month after successful dilation.*
- *Not due to active inflammation or neuromuscular dysfunction.*



What Should Be Done if Dilatations with Adjuncts Fail?



1. *Re-evaluate Treatment:*

- *Confirm proper technique and adequate number of dilations.*
- *Rule out missed underlying causes (e.g. inflammation, infection, vascular anomalies).*

2. *Adjunct Therapies:*

- *Topical steroids or oral corticosteroids (e.g. dexamethasone).*
- *Proton pump inhibitors (e.g. omeprazole).*

3. *Endoscopic Options:*

- *Endoscopic Electrocautery Incisional Therapy (EIT):*
 - *Cuts the stricture ring to enhance dilation success*





4. *Esophageal Stenting:*

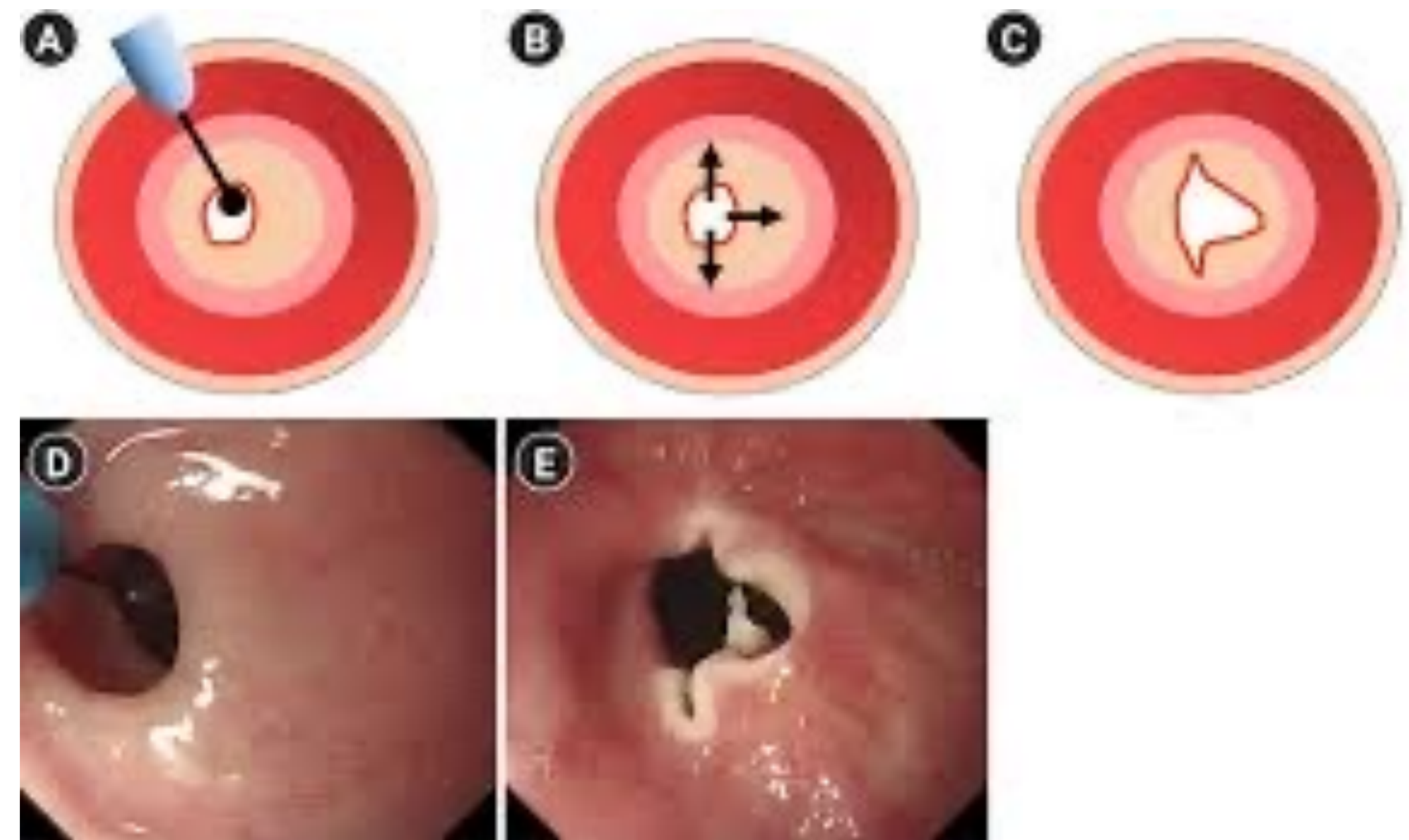
- *Removable covered stents for refractory strictures.*
- *Custom dynamic stents for improved motility and long-term patency.*
- *Indicated in:*
 - *Post-anastomotic strictures*
 - *Caustic injuries*
 - *Perforations and leaks*

5. *Surgery (Last Resort):*

- *Only if all conservative and endoscopic measures fail.*
- *High morbidity, especially in children.*

Endoscopic Electrocautery Incisional Therapy (EIT):

- Radial incisions using needle-knife electrocautery followed by balloon dilatation
- Best for short strictures (<1 cm)
- Safe with minimal complications
- Limited pediatric data but promising results



Esophageal Stenting in Children (Indications & Types)



Indications

- Used when dilation fails in recurrent/refractory esophageal strictures
- Also for post-dilatation/perforation, anastomotic leaks, caustic injuries

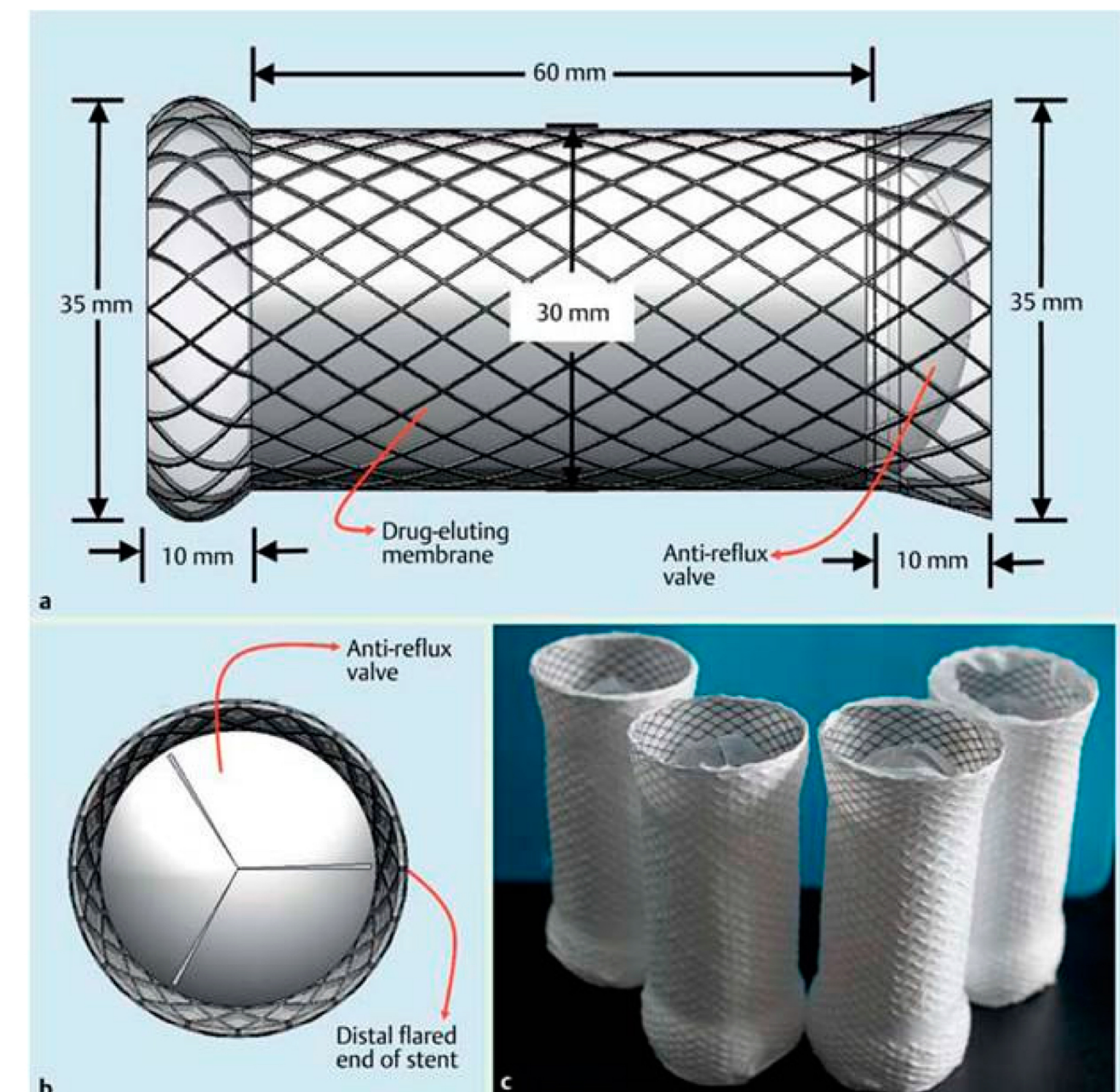


Self-Expanding Metal Stents (SEMS)

- Made of flexible metals (stainless steel, nitinol).
- Automatically expand to open strictures.
- Mainly used in adults with malignant strictures.
- Risks in children: migration, inflammation.

Plastic Stents

- Made of polyethylene or polyurethane.
- Larger diameter, stiffer than metal stents.
- Used selectively in children with fibrotic strictures.



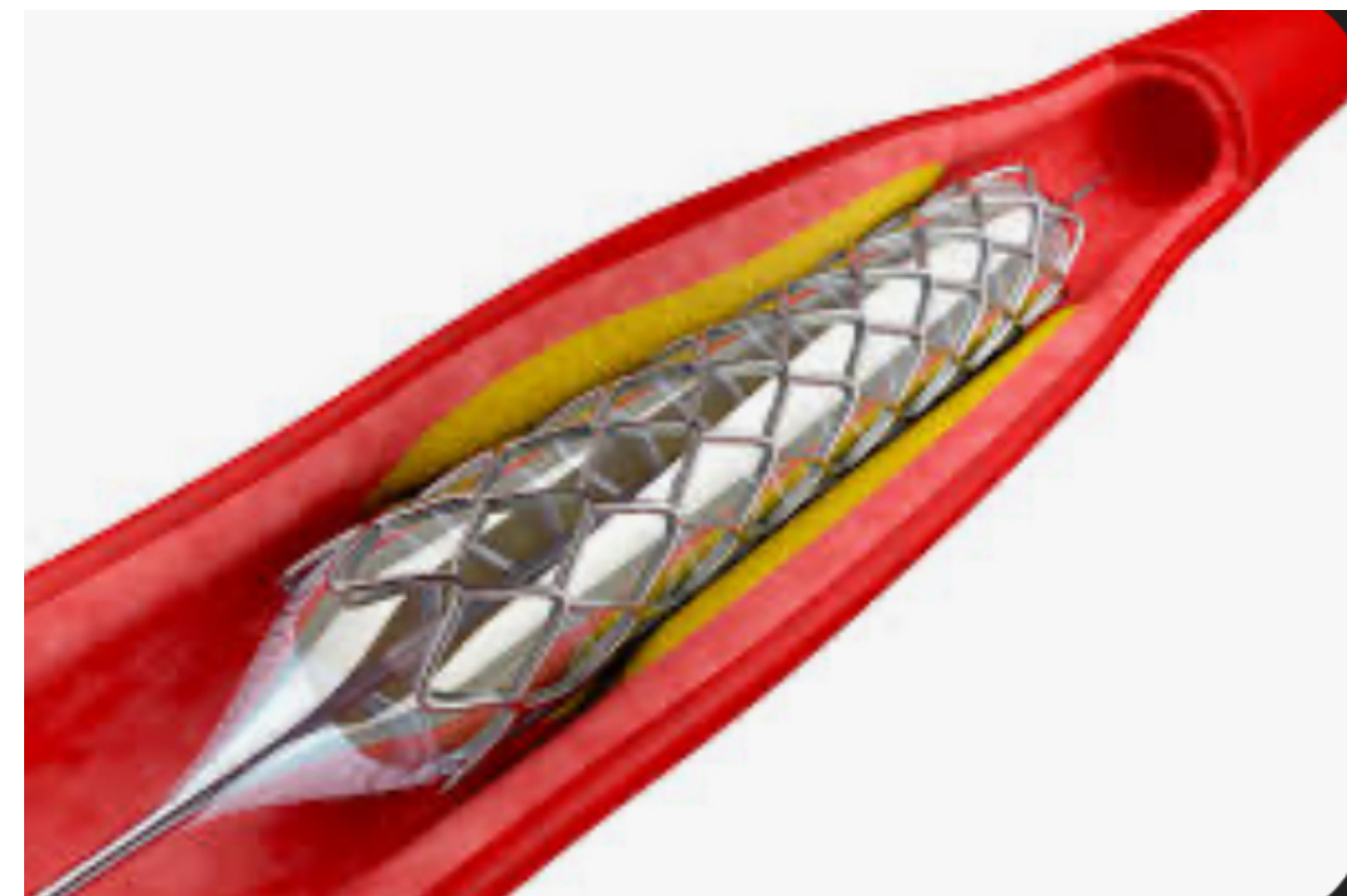
- Faulx AL et al., Gastrointest Endosc, 2013
- UpToDate: Esophageal stents in benign and malignant disease

Covered SEMS

- Metal stents coated with silicone/polyurethane to prevent tissue ingrowth.
- Reduces occlusion, but higher risk of migration.

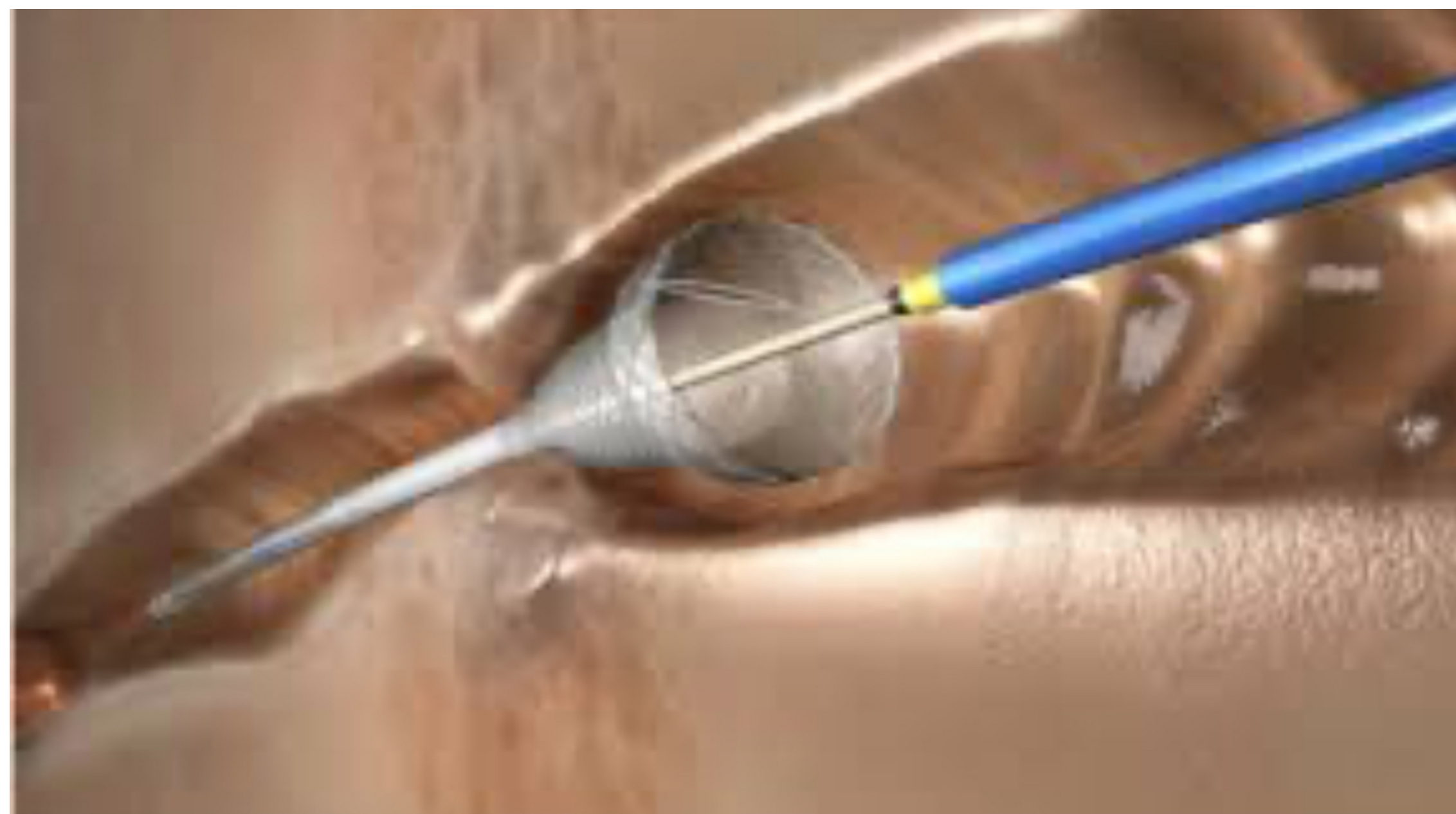
Biodegradable Stents

- *Made of absorbable materials.*
- *Potentially ideal for temporary pediatric use.*
- *Requires further safety and efficacy studies.*



Key Pediatric Considerations

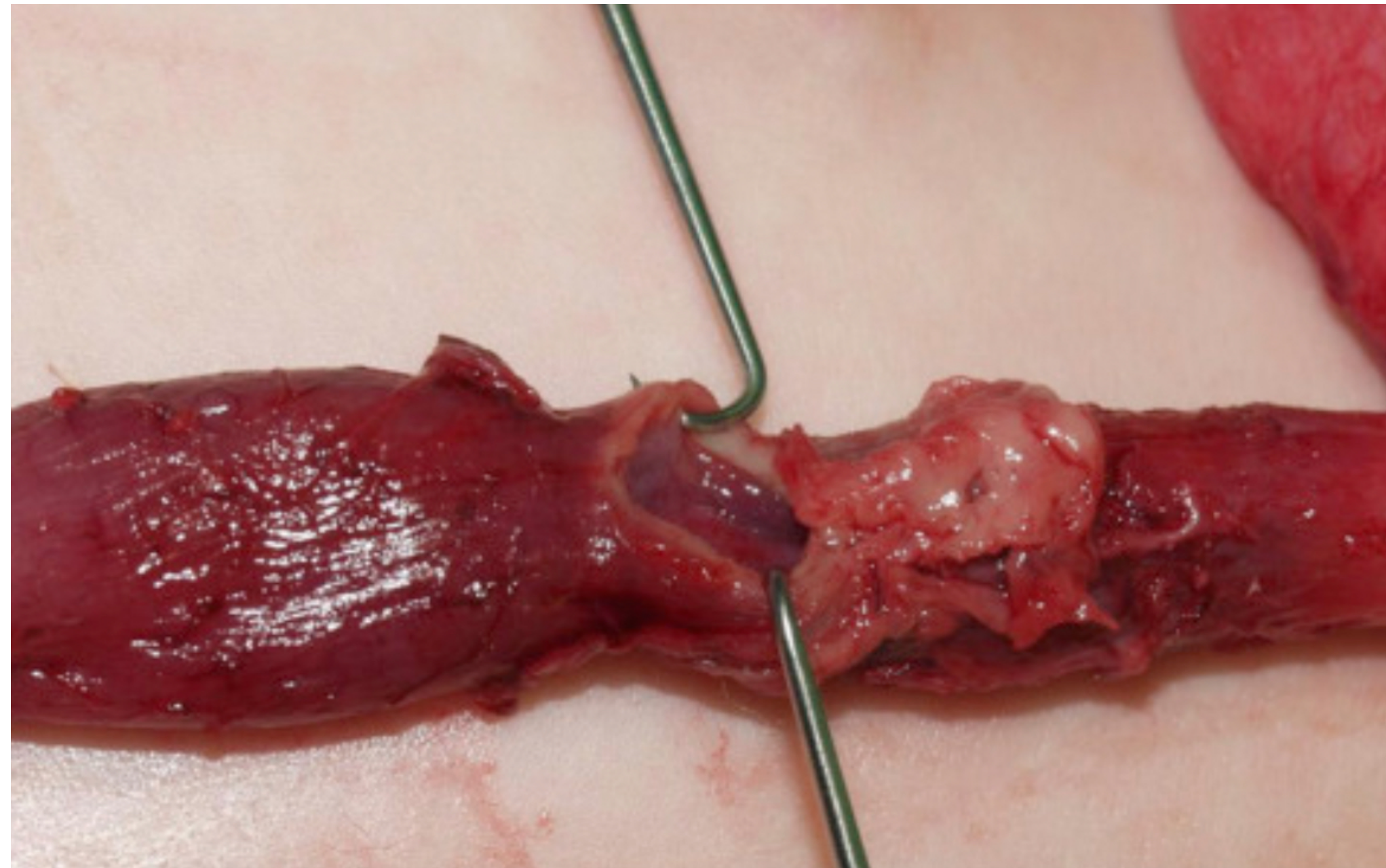
- Stent choice depends on patient size, stricture type, treatment duration.
- Long-term metal stents not recommended in growing children.
- Biodegradable stents are promising but need more research.



Surgical Treatment of CES

Indications for Surgery

- *TBR subtype with cartilage if balloon dilatation fails after 3–5 sessions*
- *Insufficient initial dilatation or rapid symptom recurrence*
- *Severe complications reported with repeated dilatations in some cases*



Surgical Approach

- *Fundoplication recommended if gastroesophageal junction is disturbed to prevent GER and hiatus hernia*
- *Esophageal replacement avoided unless absolutely necessary*



Postoperative Considerations



- *Risk of recurrent strictures requiring repeated dilatations*
- *FMD subtype usually responds well to balloon dilatation but surgery may be needed if dilatations fail*





Thank you

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