

بنام خدا



Esophageal Stenosis in Epidermolysis Bullosa

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Classification of classical epidermolysis bullosa

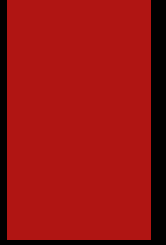
Epidermolysis bullosa simplex

Junctional epidermolysis bullosa

Dystrophic epidermolysis bullosa

Kindler epidermolysis bullosa

Gastrointestinal problems in EB patients



Malnutrition and anemia

Several factors contribute to severe nutritional compromise

- ❑ Feeding difficulties due to oropharyngeal involvement, esophageal narrowing, dysphagia, and poor swallowing coordination
- ❑ Moderate to severe malabsorption due to recurrent mucosal lesions of the small intestine
- ❑ High energy consumption from accelerated skin turnover, wound healing, and natural growth
- ❑ Hypercatabolic state from chronic inflammation and infection
- ❑ Transcutaneous loss of nutrients

Malnutrition may result

- ▶ Refractory anemia, largely caused by iron deficiency (resulting from reduced intake and absorption, blood loss from chronic skin and mucosal wounds, and loss of skin cells) and chronic inflammation
- ▶ Anemia may impact general well-being, fatigue, breathlessness, reduced exercise tolerance, poor wound healing, and anorexia
- ▶ Many have Hb<8g/dl and may need blood transfusion
- ▶ Hypoalbuminemia
- ▶ FTT
- ▶ Delayed puberty with secondary hypogonadism

Anemia

- ▶ Anemia has a significant Many patients with severe forms of EB maintain hemoglobin levels of <8 g/dL despite therapy with iron supplements. Blood transfusion may be necessary.

Oral cavity

- ▶ Intraoral blisters and erosions,
- ▶ Scarring and loss of normal architecture of intraoral soft tissues occur in most patients with JEB or DEB
- ▶ Dental Caries due to:
 - Enamel hypoplasia with pitting and furrowing
 - poor oral hygiene due to painful perioral and intraoral blisters or erosions and impaired manual dexterity
 - altered soft tissue architecture with contractures (microstomia, ankyloglossia), abnormal tongue mobility, and obliteration of oral vestibules resulting in decreased food clearance
 - frequent feeding with high-calorie, cariogenic soft diets
 - malnutrition


Gastrointestinal tract


any portion of the gastrointestinal tract may be injured

- Esophageal Strictures
- GERD
- Protein-losing enteropathy
- Rectal tears
- Anal fissures and stenosis
- Constipation

Esophageal Strictures

- ▶ Resulting from recurrent mucosal blistering and scarring
- ▶ The most frequent and, often, the most disabling complication
- ▶ Mostly occur in the upper third of the esophagus, but may arise anywhere,
- ▶ Leads to progressive dysphagia initially with hard or bulky foods, then with softer foods, and eventually with liquids
- ▶ In RDEB, develops in early childhood, and more than 50 percent of patients report symptoms by the age of 10

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- ▶ Dysphagia as the main symptom, food impaction, chest pain
 - ▶ May be present even in the absence of dysphagia
 - ▶ It affects various aspects of the patient's life.
 - ❑ Food intake
 - ❑ Malnutrition
 - ❑ Wound healing
 - ❑ Quality of life

- 
- ▶ May perform a barium esophagogram prior to endoscopic evaluation to establish the location, length, diameter, and number of strictures and to identify associated pathology such as esophageal diverticulum
 - ▶ These factors may help estimate the number of sessions that will be required to relieve symptoms and the risks associated with dilation

Patient Information

404-02-01-35905

ABDOLAHI, MILAD

M 010Y

Navigation and control panel with various icons and settings.



Patient Information

404-03-21-38262

NAJAR, GHASEM

M 029Y





334221
MOKHTARI ALI, 1403.9.21.A
M, 14Y
12.11.2024



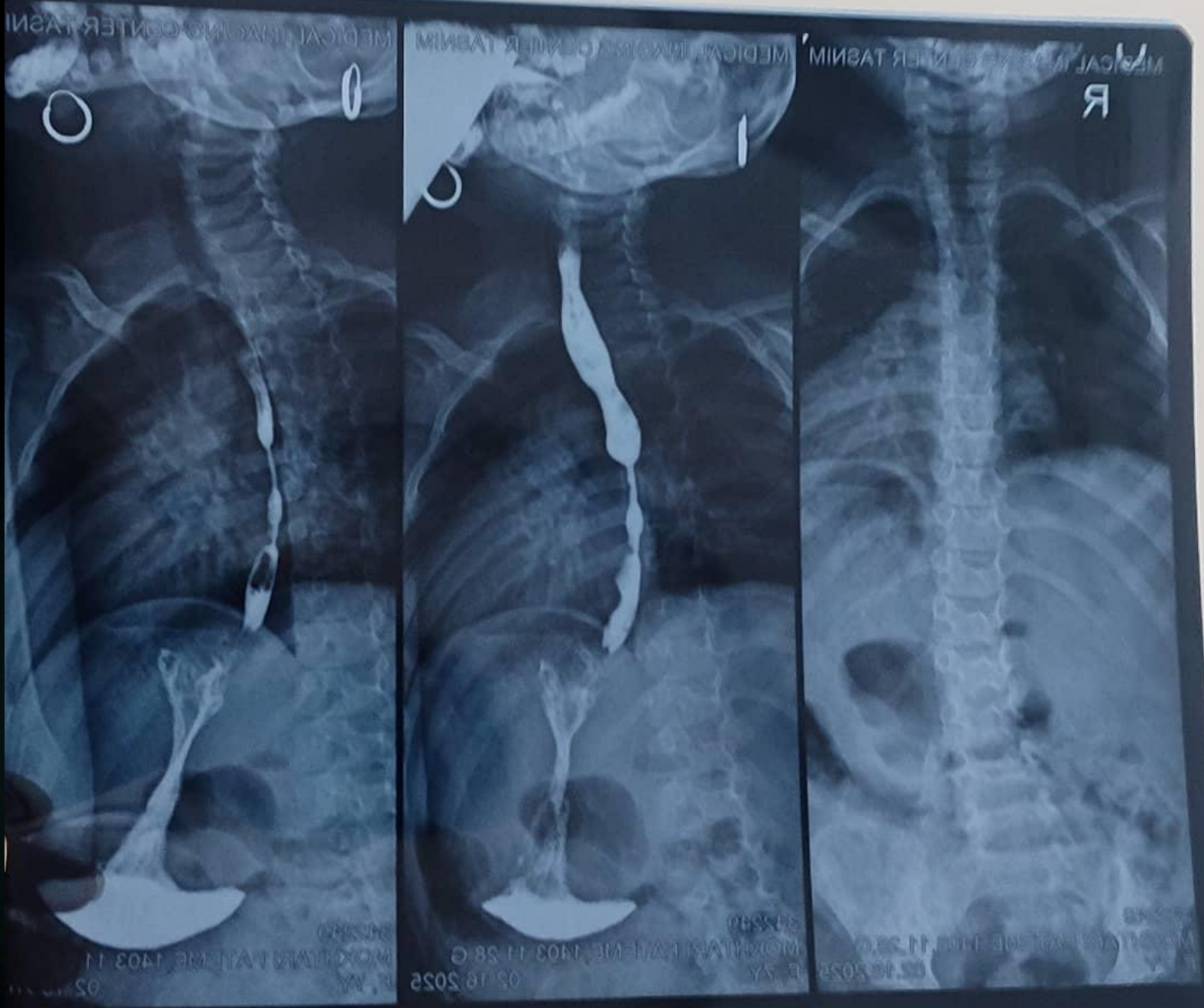
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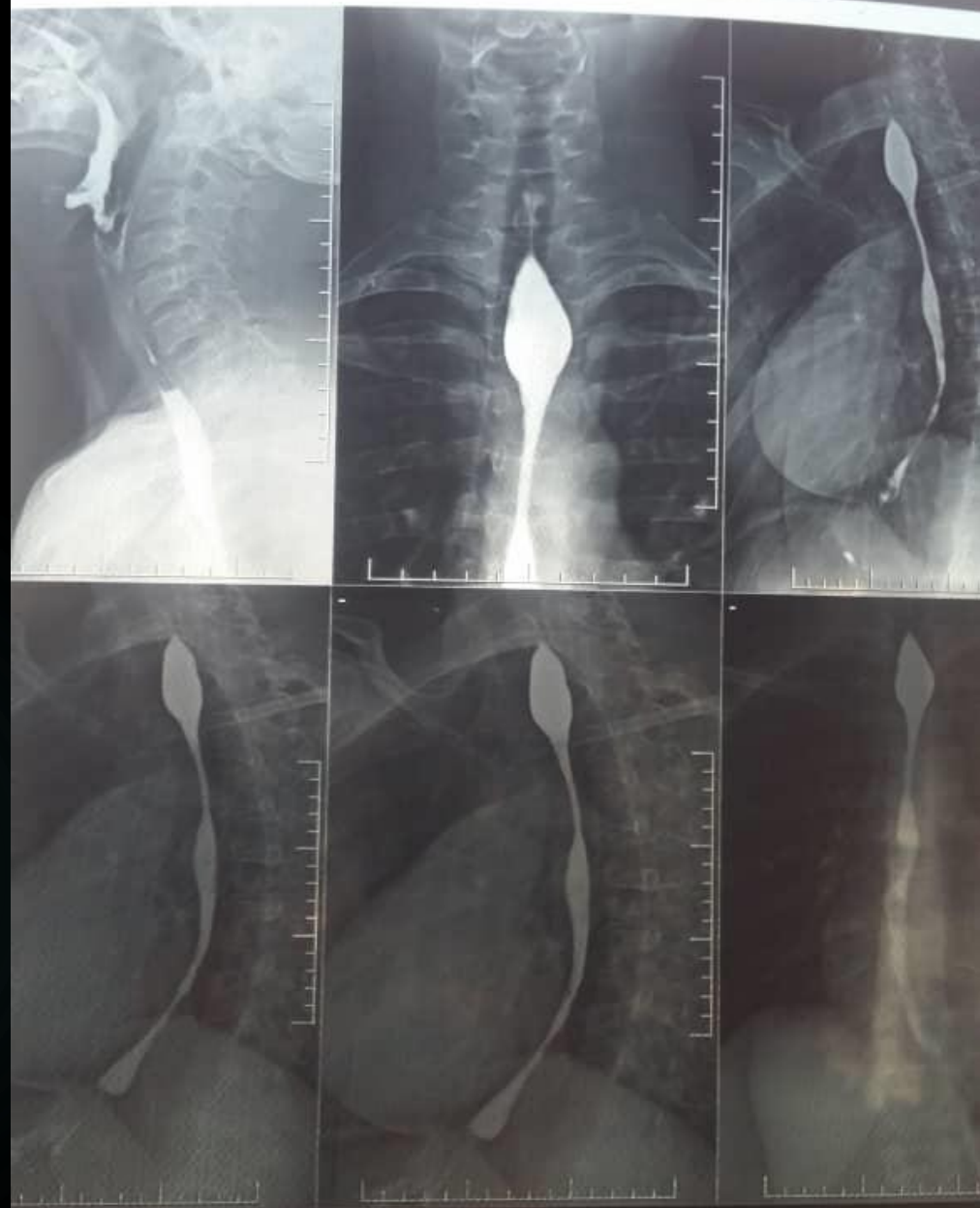


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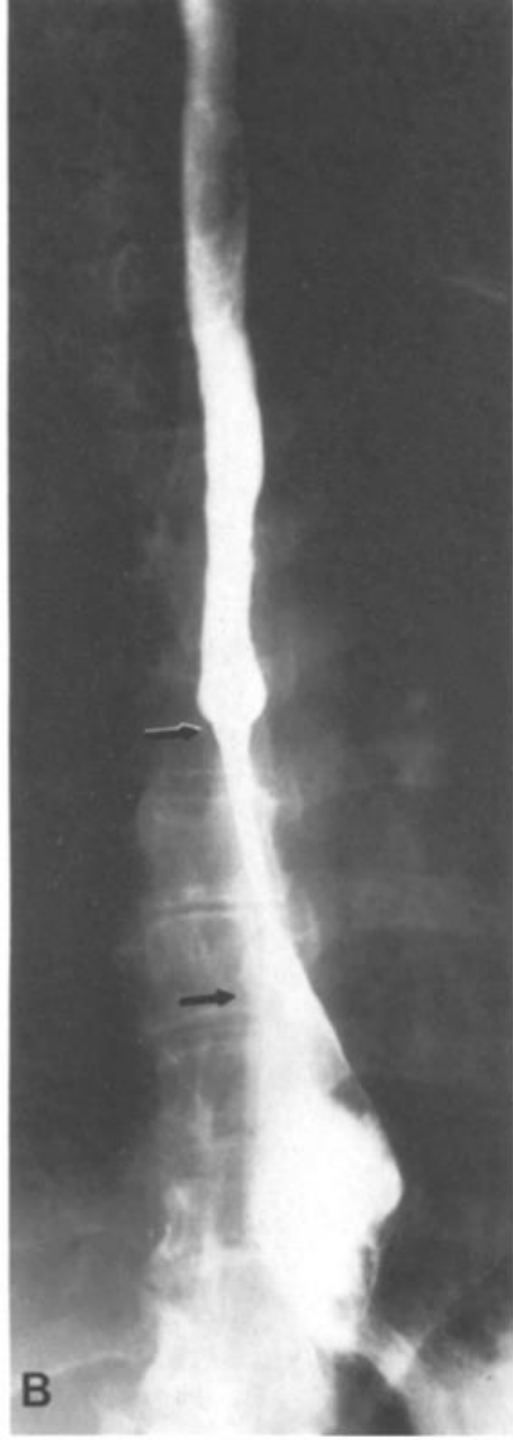
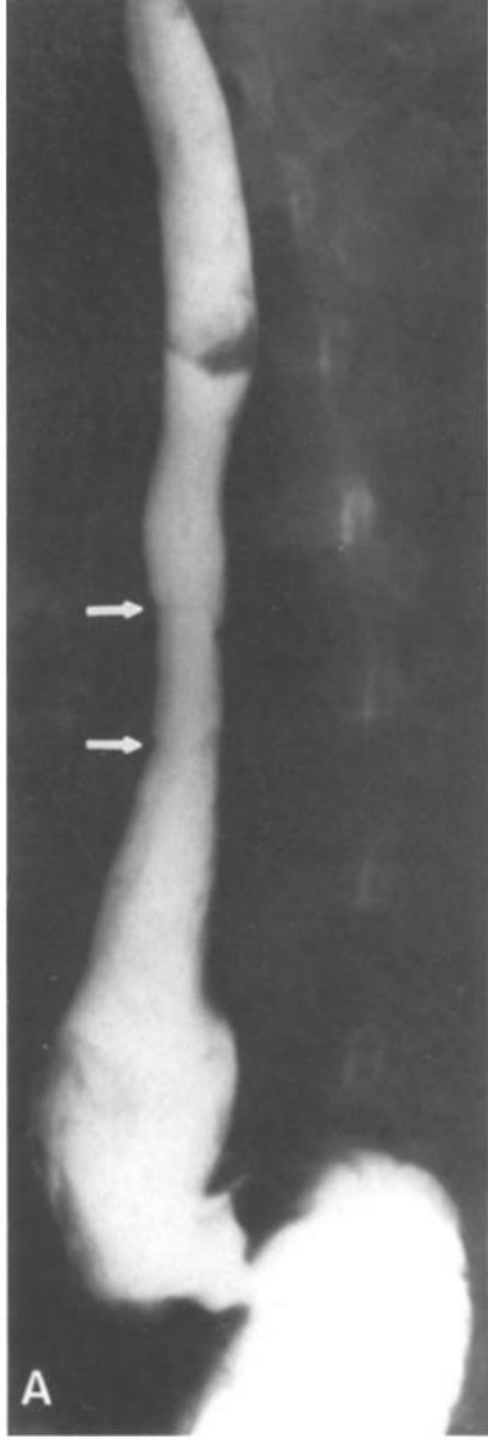






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Management

- ▶ Medical
- ▶ Dilatation

Medical

- ▶ Topical Corticosteroids
 - ❑ Efficacy remains questionable, No long-term study
 - ❑ Can be administered after dilatation or without dilatation
 - ❑ Budesonide
 - ❑ Complication: Mycotic Mucositis , Gastric fullness, Concern of systemic absorption



▶ Losartan

- ❑ Has an inhibitory effects on TGF- β 1, with a positive impact on the skin condition and quality of life in children with DEB
- ❑ Daily oral administration of losartan is well tolerated by children with RDEB, and provide preliminary evidence that it may reduce disease burden
- ❑ Further research is needed

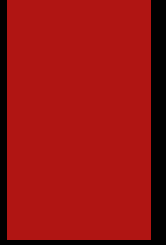
Esophageal dilation:

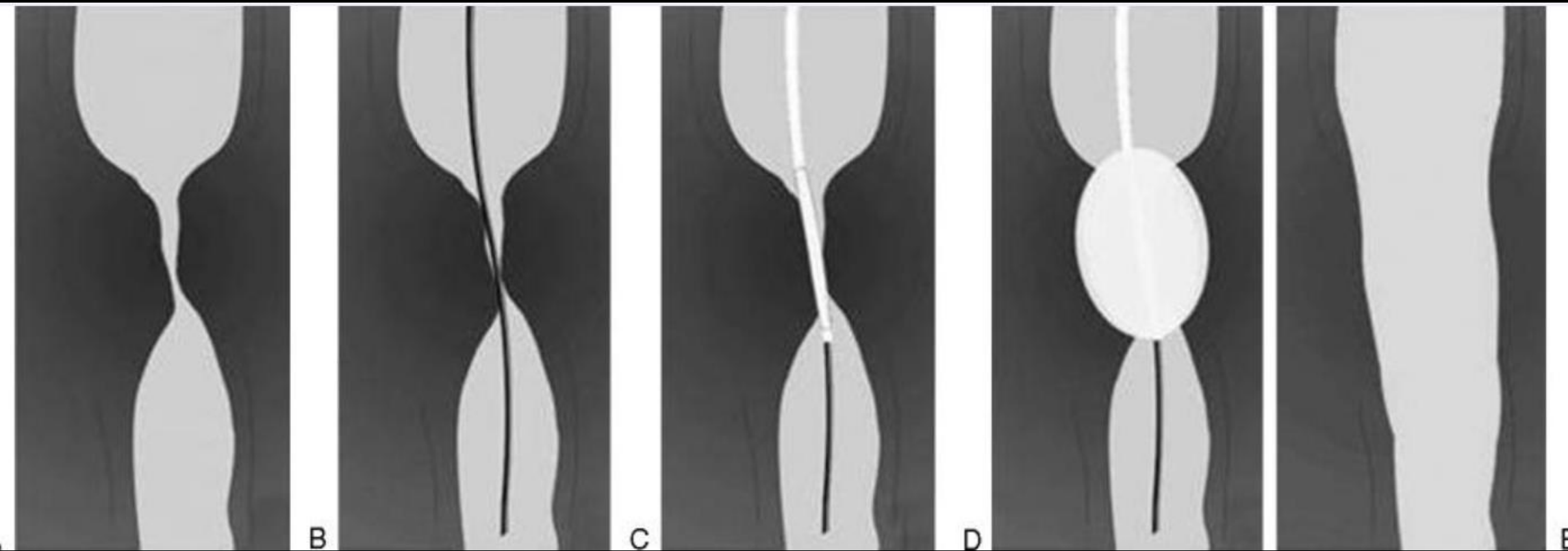
- ❑ Endoscopic balloon dilators
- ❑ Fluoroscopy-guided, over-the-guidewire hydrostatic balloon dilators
- ❑ Bougienage

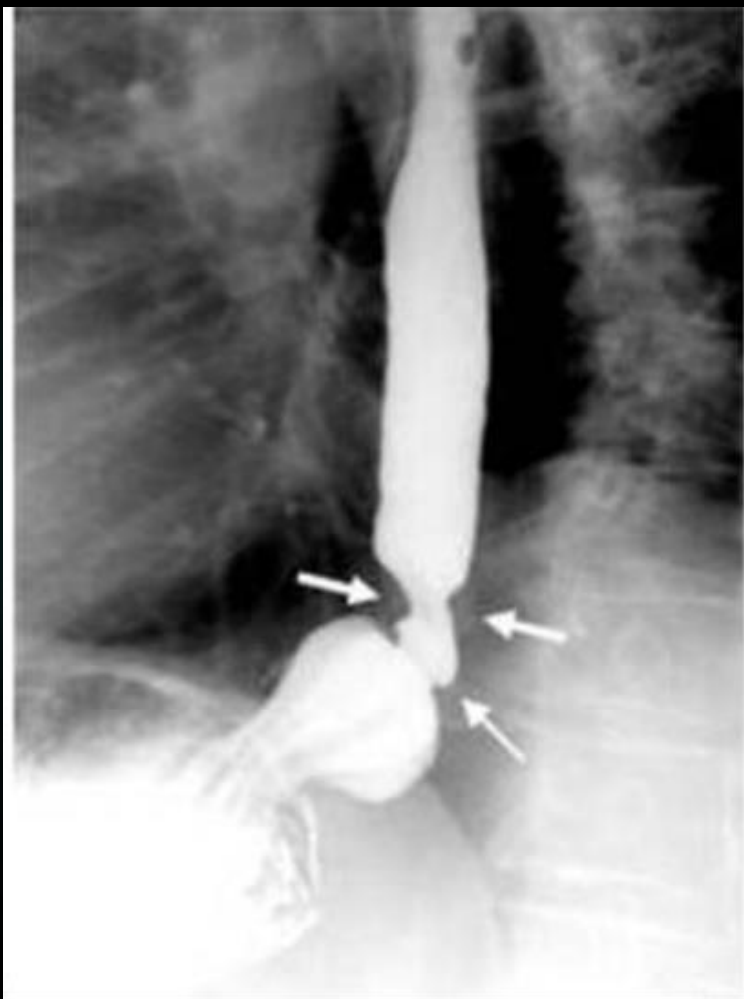
Bougienage

- ▶ Benefits: shorter operation duration and lower cost and low complication, a high success rate
- ▶ Bouginage treatment on 8 patients with EB in the form of two studies showed acceptable results after two dilation sessions, **but with a high mortality rate (1 out of 8)**.

Fluoroscopically Guided Balloon Dilation of the Esophagus

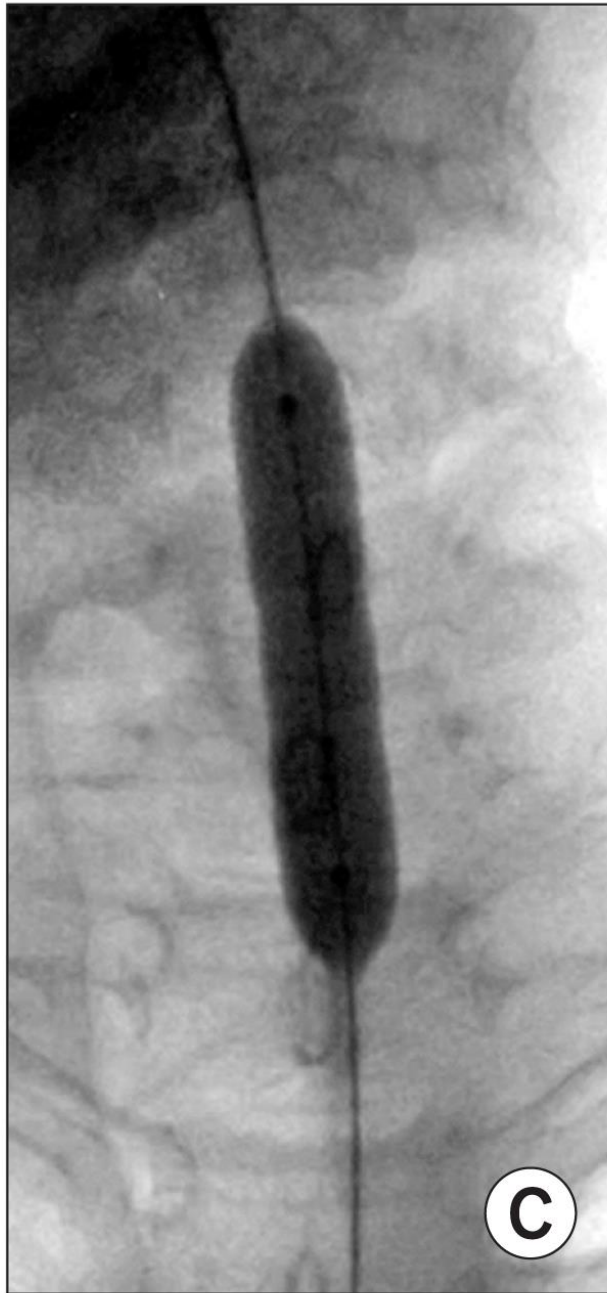
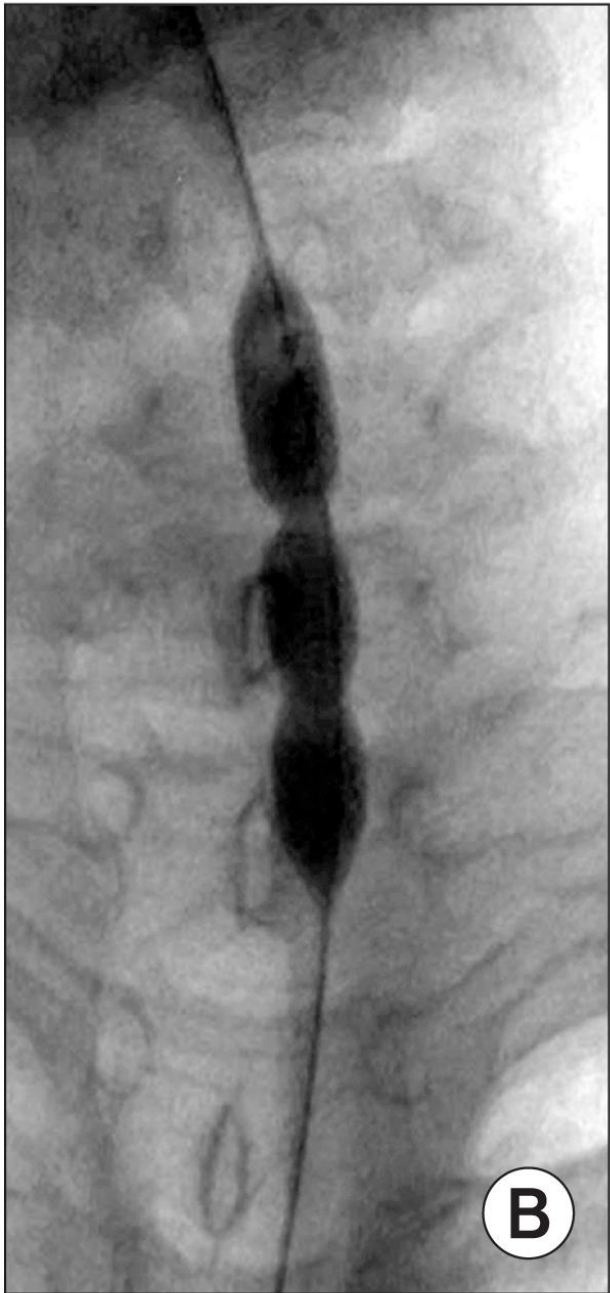
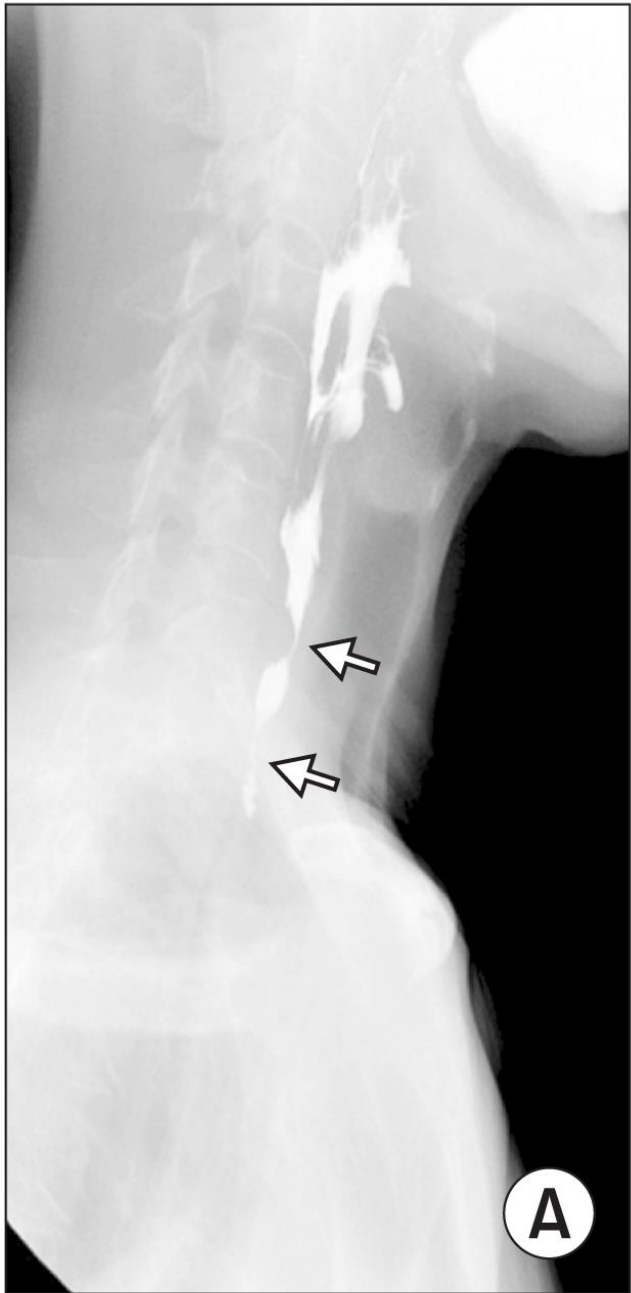






B





Benefits

- ▶ Provides superior visual monitoring during balloon expansion
- ▶ Less mucosal trauma of oropharynx (compared to endoscopic method balloon dilation)

Endoscopic balloon dilators

- ▶ The main procedure

Benefits


- Direct visualization of the esophagus
- Assessment of the balloon's caliber and pressure
- Prompt detection of perforations or bleeding during procedure
- Identification of additional strictures and potential pathologies

Disadvantages

- ❑ More mucosal trauma of oropharynx
- ❑ A small risk of perforation, independently of the balloon expansion effect

Complications

- ▶ Minor complications: pain, fever, vomiting, nausea, transient odynophagia, and dysphagia (no need for prolong hospitalization and manageable with supportive treatment)
- ▶ Major complications such as esophageal perforation (all had been managed with conservative treatment)

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- ▶ Often multiple procedures are required because of recurrence
 - ▶ Despite esophageal dilation, some patients present with significant failure to thrive and require the placement of a gastrostomy tube for supplemental enteral feeding

Prediction of Stricture Reoccurrence

- ▶ Long (>1 cm) segment involvement
- ▶ Multiple locations

General Consideration In Endoscopic Balloon Dilation

- ▶ IV Line
- ▶ Sampling

Lab

- CBC
- Iron Storage Status (Ferritin, SI, TIBC)
- Vit D
- Folate
- Vit B12
- Albumin
- Cr
- AST, ALT, Bilirubin
- Vit C
- Na/K, Ca/P
- Selenium, Zinc

Blood transfusion

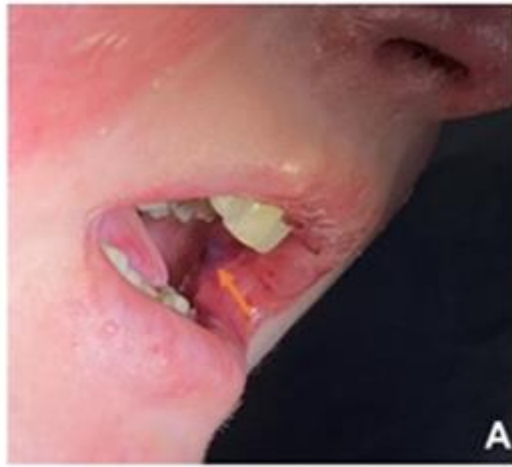
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- ▶ Care and handling
 - ▶ Elastic Band and Nasal Oxygen

Pulseoximetry



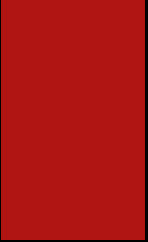
mitten deformity

Mouthpiece



Dental Caries



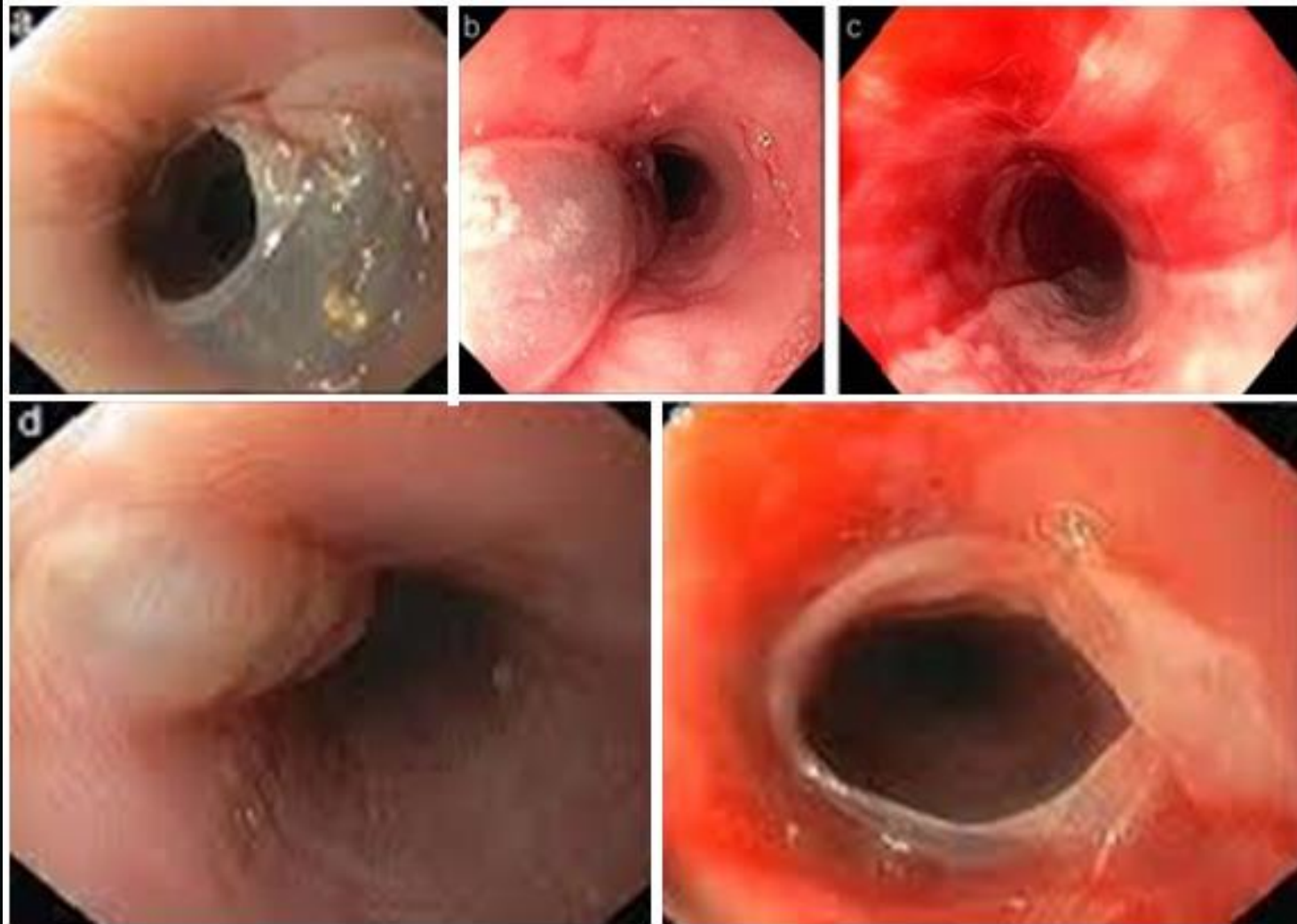


Technical

▶ Bullas

The least number of times to insert
the endoscope





Acute Sloughing

- ▶ Clinical manifestations: frequent vomiting, coughing, and the expulsion of a substantial tissue string, hematemesis, aspiration pneumonia, choking, and expulsion of a large string of tissue, progressive retching, ...
- ▶ Dexamethasone, IV antibiotics, IV fluids, increased PPI dose, no oral intake, and pain medication
- ▶ May need to gastrostomy or balloon dilation
- ▶ Could occur in varying lengths and is an emergency
- ▶ The lining coughed up needs to be cut at the mouth not pulled and the emergency services called immediately for urgent assessment and management. Expert multidisciplinary care is needed to manage this rare but serious condition

شاد وتندرست باشید

