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Esophageal rings and webs

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INTRODUCTION

Esophageal rings and webs are thin structures that partially occlude the esophageal lumen. Patients with esophageal rings or webs are usually asymptomatic. Symptomatic patients typically present with intermittent dysphagia to solids. This topic will review the pathogenesis, clinical manifestations, diagnosis, and treatment of esophageal rings and webs. The approach to patients with dysphagia and esophageal strictures is discussed elsewhere. (See "Approach to the evaluation of dysphagia in adults" and "Endoscopic interventions for nonmalignant esophageal strictures in adults".)

ANATOMY

Nonpathologic indentation of the esophagus may be caused by esophageal A and B rings and the crural diaphragm.

- A ring The A ring is caused by normal smooth muscle contraction in the esophagus. It is located in the distal esophagus just proximal to the squamocolumnar junction corresponding to the strongest part of the lower esophageal sphincter.
- **B ring** The B ring is a mucosal structure precisely at the squamocolumnar junction that is smooth and thin (≤5 mm in axial length). Owing to its location, a B ring is covered with squamous mucosa proximally and columnar epithelium distally [1].

• **Diaphragmatic impression** – Indentations in the most distal esophagus or proximal stomach that are the result of extrinsic compression by the crural diaphragm [2]. The size of a sliding hiatal hernia is measured from the diaphragmatic impression to the A ring; this is considered abnormal when it exceeds 2 to 3 cm. When less than 2 to 3 cm, it is a normal physiologic structure known as the phrenic ampulla.

TERMINOLOGY

- **Esophageal web** An esophageal web is a thin (<2 mm) eccentric membrane that protrudes into the esophageal lumen. Esophageal webs are covered with squamous epithelium and most commonly occur anteriorly in the cervical esophagus, causing focal narrowing in the postcricoid area (image 1).
- **Esophageal ring** The term esophageal ring refers to a concentric (2 to 5 mm) diaphragm of tissue that protrudes into the esophageal lumen. They are typically located in distal esophagus but can occur in other parts of the esophagus. Esophageal rings are usually mucosal but in rare cases may be muscular due to hypertrophy of an A ring. Schatzki ring, the most common type of esophageal ring, is a narrow mucosal B ring that is less than 12.5 mm in diameter [1]. (See 'Anatomy' above.)

EPIDEMIOLOGY

The true prevalence of esophageal rings and webs is unclear as most patients are asymptomatic. Esophageal B rings are found in 6 to 14 percent of routine barium esophagrams for dysphagia (image 2) [1,3,4]. Schatzki rings have been identified in up to 13 percent of patients undergoing upper endoscopy for dysphagia [5]. The prevalence of muscular esophageal rings is unclear, however, they appear to be rare and are usually identified in children.

Esophageal webs have been reported in 5 to 15 percent of patients undergoing barium esophagram for dysphagia [2].

PATHOGENESIS

The pathogenesis of esophageal webs is unknown. It is hypothesized that esophageal rings are secondary to chronic damage from gastroesophageal reflux. However, a congenital or developmental origin has also been proposed. Evidence for esophageal acid exposure causing

esophageal rings is supported by studies showing a reduced risk of recurrence following dilation if treated with acid suppression, as well as a longitudinal radiographic study that demonstrated the evolution of esophageal rings into peptic strictures due to reflux esophagitis [6]. (See 'Post-procedure acid control' below.)

ASSOCIATED CONDITIONS

Esophageal rings — Several conditions have been associated with esophageal rings.

Hiatus hernia — Schatzki rings are almost always associated with a hiatal hernia. In a study of 167 patients with a Schatzki ring, a hiatal hernia was detected in 97 percent [7].

Eosinophilic esophagitis — Schatzki rings have also been associated with eosinophilic esophagitis [8]. However, more often patients with eosinophilic esophagitis have multiple proximal esophageal rings, long strictured segments, linear furrows in the mucosa, and eosinophilic abscesses that appear as white papules. (See "Clinical manifestations and diagnosis of eosinophilic esophagitis (EoE)", section on 'Endoscopy'.)

Esophageal webs — Esophageal webs have been associated with Zenker's diverticulum, dermatologic and immunologic disorders, and iron deficiency anemia.

Plummer-Vinson syndrome — The triad of iron deficiency anemia, dysphagia, and a cervical esophageal web is known as Plummer-Vinson or Paterson-Brown-Kelly syndrome (image 3) [9-15]. Patients with Plummer-Vinson syndrome are typically White females in their fourth to seventh decade of life, but it has also been described in children and adolescents [11]. While in the past Plummer-Vinson syndrome was thought to be common in White individuals in northern countries, it is now extremely rare, possibly due to a fall in the prevalence of iron deficiency anemia in these countries over time. Recognition of Plummer-Vinson syndrome is important because it is a risk factor for developing esophageal or pharyngeal squamous cell carcinoma.

Other findings that may be present in patients with Plummer-Vinson syndrome include glossitis (picture 1), angular cheilitis, koilonychia (picture 2), splenomegaly, an enlarged thyroid, and symptoms of iron deficiency anemia (eg, fatigue and weakness) [11]. It is diagnosed based on the finding of iron deficiency anemia on laboratory testing and by demonstration of an esophageal web on barium esophagram, videofluoroscopy, or upper endoscopy. (See 'Clinical presentation' below.)

In patients with Plummer-Vinson syndrome, iron repletion may lead to rapid resolution of the dysphagia before hematologic abnormalities are corrected. However, in patients with significant obstruction of the esophageal lumen, esophageal dilation may also be required. Annual surveillance upper endoscopy for esophageal squamous cell carcinoma has also been suggested by some experts, although this has not been shown to improve patient outcomes [16]. (See 'Esophageal webs' above.)

Other associated conditions

- **Zenker's diverticulum** Esophageal webs have been associated with Zenker's diverticulum [17]. However, the potential role of esophageal webs in the pathogenesis of cricopharyngeal muscle hypertrophy and Zenker's diverticulum and their prevalence in patients with Zenker's diverticulum are not known.
- **Dermatologic diseases** Esophageal webs can be seen as extracutaneous manifestation of the following dermatologic conditions (table 1):
 - Epidermolysis bullosa Esophageal webs occur in up to 14 percent of cases [18]. (See "Diagnosis of epidermolysis bullosa".)
 - Bullous pemphigoid Esophageal webs can occur in patients with mucous membrane involvement which accounts for approximately one-third of patients with bullous pemphigoid [19]. (See "Clinical features and diagnosis of bullous pemphigoid and mucous membrane pemphigoid".)
 - Pemphigus vulgaris Esophageal webs can occur in association with oral mucosal lesions [20,21]. (See "Pathogenesis, clinical manifestations, and diagnosis of pemphigus".)
- Immunologic disorders In patients with chronic graft-versus-host disease following bone marrow transplantation, esophageal webs can result from the accretions of desquamative esophagitis [22]. (See "Clinical manifestations and diagnosis of chronic graft-versus-host disease".)

CLINICAL PRESENTATION

Most esophageal rings and webs are asymptomatic. Symptomatic patients usually present with dysphagia to solids that is particularly evident with hard solids (eg, meat or bread). The dysphagia is often intermittent, and patients will occasionally report modifying how they eat (eg, chewing food more thoroughly). The presence and severity of symptoms depends on the

internal diameter of the esophageal lumen. Patients with an esophageal lumen less than 13 mm (39 French) will usually experience solid food dysphagia [1]. However, esophageal rings with an internal 13 to 20 mm diameter can produce dysphagia, particularly when associated with a sliding hiatus hernia. (See "Approach to the evaluation of dysphagia in adults" and "Hiatus hernia", section on 'Type I: Sliding hernia' and 'Hiatus hernia' above.)

Occasionally, patients present with an acute onset of dysphagia or complete inability to swallow saliva due to a food impaction. In rare cases, patients with esophageal webs have Plummer-Vinson syndrome and associated clinical features of iron deficiency anemia. (See "Ingested foreign bodies and food impactions in adults", section on 'Clinical manifestations' and 'Plummer-Vinson syndrome' above.)

DIAGNOSIS

Esophageal rings and webs are usually diagnosed by barium swallow and/or upper endoscopy for evaluation of dysphagia or other upper gastrointestinal symptoms. In all patients with an esophageal ring, we perform an upper endoscopy to biopsy the esophagus for associated eosinophilic esophagitis. (See "Clinical manifestations and diagnosis of eosinophilic esophagitis (EoE)", section on 'Endoscopy' and "Clinical manifestations and diagnosis of eosinophilic esophagitis (EoE)", section on 'Histology' and "Treatment of eosinophilic esophagitis (EoE)", section on 'Esophageal dilation'.)

Barium esophagram — On barium esophagram, a lower esophageal mucosal ring appears as a thin (<3.0 mm) transverse circumferential ridge a few centimeters above the hiatus of the diaphragm [23]. Mucosal rings are smooth, symmetric narrowings that are 3 to 5 millimeters in axial extent with a luminal aperture that varies during the course of fluoroscopic examination. However, if the esophagus is not adequately distended, esophageal rings and webs give the appearance of subtle narrowing of the esophageal lumen and can easily be missed.

In order to adequately distend the esophagus, a barium esophagram should be performed using a full-column technique with the patient performing a Valsalva maneuver after swallowing a solid food bolus while in a prone (or prone oblique) position. The use of a barium tablet (12.8 mm) or marshmallow bolus can help determine if the esophageal ring is the cause of dysphagia if it reproduces the symptoms [24]. A properly performed examination will detect up to 100 percent of esophageal rings, compared with detection rates of only 17 to 49 percent for upright double-contrast and mucosal relief techniques [25].

Endoscopy and biopsy — On upper endoscopy, an esophageal web appears as a smooth, thin membrane that is usually noncircumferential [11]. Endoscopically, an esophageal ring appears as a thin membrane with a concentric smooth contour that projects into the lumen (picture 3). In the case of a Schatzki ring, an associated hiatal hernia is almost always present. Patients may have features of eosinophilic esophagitis (eg, multiple stacked circular rings, linear furrows, whitish papules, and small caliber esophagus) or an associated Zenker's diverticulum. (See "Clinical manifestations and diagnosis of eosinophilic esophagitis (EoE)", section on 'Endoscopy' and 'Associated conditions' above.)

Endoscopy is less sensitive as compared with a barium esophagram for the detection of esophageal rings and webs. Esophageal webs are often missed on upper endoscopy because of their proximity to the upper esophageal sphincter. The lower esophagus must be widely distended to identifying an esophageal ring [23]. Applying abdominal pressure can help visualize a Schatzki ring obscured in a reduced hiatal hernia [26]. (See 'Hiatus hernia' above.)

On histology, an esophageal rings are composed of mucosa and submucosa with basal cell hyperplasia, hyperkeratosis and, frequently, eosinophilic infiltration [27]. The presence of a large number of eosinophils (>15 eosinophils per high power field) is suggestive of eosinophilic esophagitis. (See "Clinical manifestations and diagnosis of eosinophilic esophagitis (EoE)", section on 'Histology'.)

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of an esophageal web or ring is broad and also includes other causes of dysphagia. These can be differentiated by history, barium esophagram, and upper endoscopy. (See "Approach to the evaluation of dysphagia in adults", section on 'Symptom-based differential diagnosis'.)

• Achalasia – Patients with achalasia typically present with dysphagia to both solids and liquids and, unlike esophageal webs/rings, symptoms are not intermittent. Findings on barium swallow suggestive of achalasia include dilatation of the esophagus, narrow esophagogastric junction with "bird-beak" appearance, aperistalsis, and delayed emptying of barium. Upper endoscopy may reveal a dilated esophagus that contains residual food and fluid. The lower esophageal sphincter does not open spontaneously to allow effortless passage of the endoscope but can usually be traversed easily with gentle pressure on the endoscope. (See "Achalasia: Pathogenesis, clinical manifestations, and diagnosis", section on 'Barium esophagram'.)

• **Esophageal stricture** – Esophageal strictures are longer in axial length as compared with esophageal rings and webs and have tapered ends. The caliber of an esophageal stricture does not change with peristalsis in contrast to a muscular esophageal ring. (See "Endoscopic interventions for nonmalignant esophageal strictures in adults".)

MANAGEMENT

Overview — Esophageal webs usually rupture during diagnostic upper endoscopy as the endoscope traverses the web. However, in patients with a partial disruption of an esophageal web, we perform esophageal dilation. (See "Endoscopic interventions for nonmalignant esophageal strictures in adults", section on 'Procedure'.)

In patients with esophageal rings, biopsies should be obtained to rule out eosinophilic esophagitis. Whether or not to perform a dilation in cases of eosinophilic esophagitis is controversial; some experts reserve dilation for patients with who have not responded to medical therapy whereas others base the decision on the severity of dysphagia and the diameter of the rings. (See "Treatment of eosinophilic esophagitis (EoE)", section on 'Pharmacologic therapy'.)

Following esophageal dilation, we treat patients with an esophageal ring with a once-daily standard dose of a proton pump inhibitor (PPI) for six weeks. For patients with esophageal rings and evidence of gastroesophageal reflux (eg, patients who experience frequent heartburn, or erosive esophagitis on endoscopy), we continue PPI therapy indefinitely [28].

Initial therapy — The goals of therapy for symptomatic esophageal rings and webs are relief of dysphagia and the prevention of recurrent symptoms.

Esophageal dilation — The approach to dilation in a patient with an esophageal ring or web depends upon the presence of associated eosinophilic esophagitis. (See 'Overview' above.)

Technique — In patients without associated eosinophilic esophagitis, first line of therapy is a single dilation with a large bougie dilator (≥50 French) or balloon dilator (18 to 20 mm) [29]. The initial dilator used is large in order to disrupt the esophageal ring/web.

In patients with eosinophilic esophagitis, graded dilation is necessary, as forceful dilation has been associated with deep mucosal tears and esophageal perforation. Esophageal dilation in patients with eosinophilic esophagitis is discussed in detail separately. (See "Clinical manifestations and diagnosis of eosinophilic esophagitis (EoE)", section on 'Diagnosis' and

"Treatment of eosinophilic esophagitis (EoE)", section on 'Esophageal dilation' and "Endoscopic interventions for nonmalignant esophageal strictures in adults", section on 'Procedure'.)

Safety and efficacy — A single dilation with a large dilator is successful in alleviating dysphagia in most patients and has a low risk of complications [29,30]. However, recurrent dysphagia occurs in the majority of patients with an esophageal ring [30-33]. In a study that evaluated 33 patients after dilation of an esophageal ring to ≥50 French, recurrent dysphagia one, two, and five years post-dilation were reported in 32, 65, and 89 percent of patients, respectively [30]. Neither the initial ring size nor the presence or absence of esophagitis were associated with symptomatic recurrence. (See 'Recurrent and refractory symptoms' below.)

Post-procedure acid control — Even in patients without reflux symptoms or evidence of gastroesophageal reflux disease (GERD) on pH testing, acid suppression may reduce the risk of recurrence. This was illustrated in a randomized trial with 44 consecutive patients who underwent dilation of a Schatzki ring [32]. All patients with objective evidence of GERD on esophageal 24 hour pH testing were treated with omeprazole. The remaining patients were randomly assigned to omeprazole or placebo. There were no recurrences of Schatzki rings in patients with GERD during a mean follow-up of 43 months. Among the patients without GERD, the probability of recurrence was significantly lower in patients assigned to omeprazole as compared with placebo (recurrence rates 7 percent at 37 months versus 47 percent at 20 months).

Recurrent and refractory symptoms — Recurrent dysphagia occurs in a majority of patients with esophageal rings after an initial response to esophageal dilation. A small subset of patients with lower esophageal rings have refractory symptoms that do not improve despite repeated attempts at esophageal dilation. (See 'Safety and efficacy' above.) Esophageal webs do not recur once disrupted and are rarely refractory to esophageal dilation.

Our approach — We treat recurrent symptomatic esophageal rings with repeat dilation and long-term acid suppression (eg, omeprazole 20 mg daily).

In patients with frequent recurrences (eg, more than one or two times per year) or refractory symptoms we obtain additional mucosal biopsies of the esophagus to rule out eosinophilic esophagitis as eosinophilic infiltration can be patchy. (See "Clinical manifestations and diagnosis of eosinophilic esophagitis (EoE)", section on 'Diagnosis' and "Treatment of eosinophilic esophagitis (EoE)".)

For refractory rings in patients who do not have eosinophilic esophagitis we use an alternative technique to disrupt the esophageal ring [31,34]. (See 'Therapeutic options' below.)

Therapeutic options — The choice of therapy depends on the prior technique used for dilation and the endoscopist's familiarity with the available systems. Therapeutic endoscopic options include:

- **Balloon dilation** Balloon dilation can be used if the initial treatment was with bougienage.
- Intralesional injection of corticosteroids prior to dilation It is hypothesized that injection of corticosteroids into the stricture may reduce stricture recurrence following dilation. However, the efficacy of this approach has not been established in patients with refractory esophageal rings/webs. (See "Endoscopic interventions for nonmalignant esophageal strictures in adults", section on 'Initial intervention'.)
- Incisional therapy Endoscopic incisional therapy using electrocautery can be used to dilate a refractory esophageal ring (figure 1 and picture 4). A randomized trial comparing electrosurgical incision to bougie dilation as primary therapy found that electrosurgical incision offered somewhat better improvement in dysphagia at one-month and a modestly longer symptom-free period (eight versus six months) [31]. Although data are limited, case series suggest that transendoscopic electrosurgical incision can be effective in patients with refractory esophageal rings [31,35-37].
- **Laser division** There are case reports of successful treatment of refractory esophageal webs with endoscopic laser division [38].
- **Biopsy obliteration** Feasibility of using biopsy forceps to rupture Schatzki's ring has been described in a small number of patients. This option should be reserved for cases refractory to standard therapies [39].

SOCIETY GUIDELINE LINKS

Links to society and government-sponsored guidelines from selected countries and regions around the world are provided separately. (See "Society guideline links: Esophageal strictures, foreign bodies, and caustic injury".)

SUMMARY AND RECOMMENDATIONS

• Esophageal rings and webs are thin structures that partially occlude the esophageal lumen. (See 'Introduction' above.)

- The term esophageal ring refers to a concentric (2 to 5 mm) diaphragm of tissue that protrudes into the esophageal lumen. They are typically located in the distal esophagus but can occur in other parts of the esophagus. Esophageal rings are usually mucosal but in rare cases may be muscular due to hypertrophy of an A ring. Schatzki ring, the most common type of esophageal ring, is a narrow mucosal B ring that is less than 12.5 mm in diameter. (See 'Anatomy' above and 'Terminology' above.)
- An esophageal web is a thin (<2 mm) eccentric membrane that protrudes into the
 esophageal lumen. Esophageal webs are covered with squamous epithelium and most
 commonly occur anteriorly in the cervical esophagus, causing focal narrowing in the
 postcricoid area (image 1). (See 'Terminology' above.)
- Most esophageal rings and webs are asymptomatic. Symptomatic patients usually present with intermittent dysphagia to solids. The presence and severity of symptoms depends on the internal diameter of the esophageal lumen. Patients with an esophageal lumen less than 13 mm (39 French) will usually experience solid food dysphagia. Occasionally, patients present with an acute onset of dysphagia or complete inability to swallow saliva due to a food impaction. (See 'Clinical presentation' above.)
- Esophageal rings and webs are usually diagnosed by barium swallow and/or upper endoscopy for evaluation of dysphagia or other upper gastrointestinal symptoms. In all patients with an esophageal ring, we perform an upper endoscopy to biopsy the esophagus for associated eosinophilic esophagitis. Dilation in patients with eosinophilic esophagitis is usually reserved for patients with rings who have not responded to medical therapy. (See 'Diagnosis' above and "Treatment of eosinophilic esophagitis (EoE)", section on 'Esophageal dilation'.)
- Esophageal webs are usually ruptured during diagnostic endoscopy by passage of the endoscope through the web. However, in patients with a partial disruption of an esophageal web, we perform esophageal dilation.
 - For patients with an esophageal ring without associated eosinophilic esophagitis, we suggest dilation with a large caliber bougie (≥50 French) or balloon dilator (18 to 20 mm), rather than graded dilations starting with smaller dilators (**Grade 2C**). However, patients with eosinophilic esophagitis require more cautious graded dilation because of an increased risk of deep mucosal tears and esophageal perforation. (See 'Overview' above and 'Esophageal dilation' above and "Treatment of eosinophilic esophagitis (EoE)", section on 'Esophageal dilation'.)

- For patients with recurrent esophageal rings or with esophageal rings and evidence of gastroesophageal reflux, we suggest indefinite proton pump inhibitor (PPI) therapy rather than short-term PPI treatment following dilation (**Grade 2B**). For all other patients, we suggest PPI treatment for six weeks following dilation, rather than indefinite treatment (**Grade 2C**). (See 'Post-procedure acid control' above.)
- In patients with frequent recurrences (eg, more than one or two times per year) or refractory symptoms we obtain additional biopsies of the esophagus to rule out eosinophilic esophagitis. We treat recurrent symptomatic esophageal rings with repeat dilation and long term acid suppression. For refractory rings in patients who do not have eosinophilic esophagitis we use an alternative technique to disrupt the esophageal ring. (See 'Recurrent and refractory symptoms' above.)

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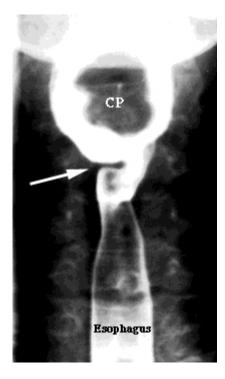
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GRAPHICS

Esophageal web on barium swallow

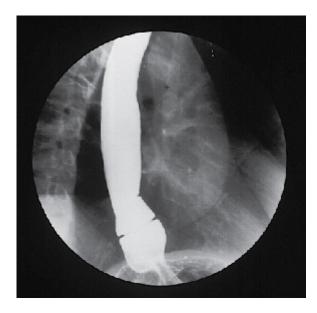


This modified barium swallow, obtained in a 45-year-old man with dysphagia, demonstrates an asymmetric esophageal web arising from the right side of the upper esophagus (arrow).

Courtesy of Jonathan Kruskal, MD, PhD.

Graphic 76514 Version 4.0

Esophageal (Schatzki) ring seen on barium swallow

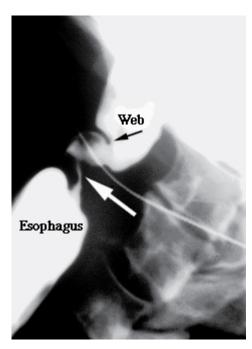


Esophageal (Schatzki) ring at the gastroesophageal junction visualized on a barium swallow.

Courtesy of Peter J Kahrilas, MD.

Graphic 68185 Version 4.0

Barium swallow in a patient with Plummer Vinson syndrome

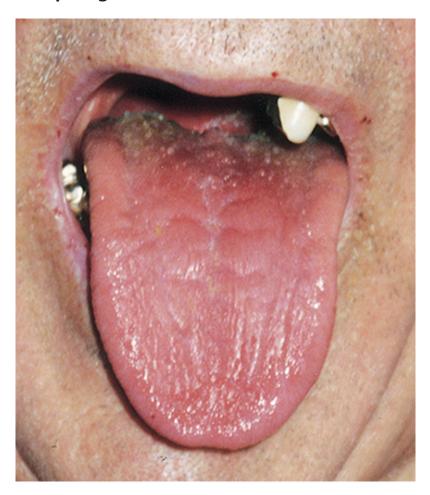


This barium swallow study obtained in a 53-year-old female with dysphagia and anemia demonstrates an upper esophageal web (black arrow) immediately above a tight stricture of the esophagus (white arrow).

Courtesy of Jonathan Kruskal, MD, PhD.

Graphic 81131 Version 3.0

Atrophic glossitis



A smooth tongue that has lost its papillae and is often sore suggests a deficiency in riboflavin, niacin, folic acid, vitamin B12, or iron. This patient had vitamin B12 deficiency.

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Graphic 54472 Version 6.0

Koilonychia (spoon nail) associated with iron deficiency



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Graphic 86419 Version 2.0

Conditions associated with esophageal webs

Dermatologic diseases
Epidermolysis bullosa (EB)
EB dystrophica
EB junctional
EB simplex
EB acquisita
Bullous pemphigoid
Pemphigus vulgaris
Stevens-Johnson syndrome (toxic epidermal necrolysis)
Psoriasis
Immunologic disorders
Graft-versus-host disease

Graphic 68674 Version 2.0

Esophageal (Schatzki) ring

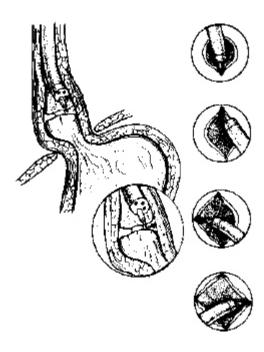


Endoscopic view of an esophageal (Schatzki) ring, which often cannot be well visualized unless the lower esophagus is widely distended. The ring appears as a thin membrane with a concentric smooth contour that projects into the lumen.

Courtesy of James B McGee, MD.

Graphic 55092 Version 3.0

Electrocautery incision of a lower esophageal ring

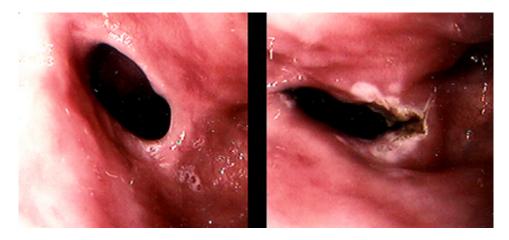


Electrocautery incision of lower esophageal ring using a needle knife papillotome. The ring is splayed by incising in four quadrants starting from the luminal edge of the ring toward the esophageal wall for seven-eighths of the ring thickness.

Adapted from Burdick, JS, Venu, RP, Hogan, WJ. Gastrointest Endosc 1993; 39:616.

Graphic 58915 Version 2.0

Transendoscopic electrocautery incision of an esophageal ring



Endoscopy demonstrating a mucosal ring in the distal esophagus (left panel), which has been incised using a needle-knife papillotome (right panel).

Courtesy of Moises Guelrud, MD.

Graphic 59650 Version 2.0

Contributor Disclosures

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