TABLE 35-2 ESOPHAGEAL MOTOR DISORDERS

FEATURE	ACHALASIA	SCLERODERMA	DIFFUSE ESOPHAGEAL SPASM
Symptoms	Dysphagia Regurgitation of nonacidic material	GERD Dysphagia	Substernal chest pain (angina-like) Dysphagia with pain
Radiographic appearance	Dilated, fluid-filled esophagus Distal "bird-beak" stricture	Aperistaltic esophagus Free reflux Peptic stricture	Simultaneous noncoordinated contractions
CONVENTIONAL MANOMETRIC FINDINGS			
LES	High resting pressure Incomplete or abnormal relaxation with swallow	Low resting pressure	Normal pressure
Body	Low-amplitude, simultaneous contractions after swallowing	Low-amplitude peristaltic contractions or no peristalsis	Some peristalsis Diffuse and simultaneous nonperistaltic contractions, occasionally high amplitude

GERD, Gastroesophageal reflux disease; LES, lower esophageal sphincter.

carefully interviewed. Weight loss may be encountered as well. Achalasia may result in pulmonary symptoms, a complication suggested by the presence of cough resulting from aspiration of esophageal contents. Neuronal denervation is the underlying mechanism of achalasia. This results in failure of esophageal body peristalsis and failure of the LES to relax in response to swallowing.

High-resolution manometry has been used to define three subtypes of achalasia. Each is associated with incomplete LES relaxation. Type 1 (classic achalasia) is devoid of any significant decreases in esophageal pressurization. Type 2 is associated with simultaneous pressurization throughout the esophagus after a swallow. Type 3 (spastic achalasia) is associated with luminal obliterating contractions after a swallow.

When achalasia is identified, its treatment can progress through several stages: (1) medical therapy with nitrates and calcium channel blockers to lower sphincter pressures and with PPIs; (2) botulinum toxin injection to lower LES pressures; (3) pneumatic dilation to facilitate luminal opening; and (4) esophageal myotomy, performed either laparoscopically or in the traditional transthoracic approach (Heller myotomy).

Secondary Achalasia

A number of conditions may result in a syndrome resembling achalasia. The most common and important of these causes is malignancy involving the region of the gastroesophageal junction (e.g., adenocarcinoma of the cardia, gastric lymphoma) or, less commonly, metastatic involvement from a distant primary malignancy (e.g., small cell lung cancer, Hodgkin's lymphoma, hepatocellular carcinoma). The result is a syndrome that radiographically and manometrically resembles primary achalasia. Such involvement is typically seen in older patients in whom the development of dysphagia is rapid. Most of these cases are identified by endoscopic examination. Other diseases that may mimic achalasia include Chagas' disease (South American trypanosomiasis) and intestinal pseudo-obstruction, which may be caused by a host of conditions ranging from familial neuropathies and myopathies to disorders such as muscular dystrophy.

Rings and Webs

Nosology is critical when discussing rings and webs. An esophageal *ring* is a distal esophageal structure that occurs at the esophagogastric junction; it is covered on one side by squamous mucosa and on the other by columnar mucosa. One exception to this rule is Schatzki's ring, which is typically located radiographically about 2 cm proximal to the esophagogastric mucosal junction, as described later. An esophageal *web* is a ring-like structure along the entire length of the esophagus that is covered entirely by squamous mucosa. Cervical esophageal webs are typically diaphragm-like structures found in the immediate postcricoid area. They may be identified incidentally on radiographic or endoscopic imaging, although they can easily be missed if care is not taken to carefully examine this area of the esophagus. The pathogenesis of cervical webs is unknown. Pathologically, they are composed of normal squamous tissue overlying connective tissue; this bland histology separates these typical cervical webs from those changes found in the proximal esophagus associated with dermatologic diseases (e.g., epidermolysis bullosa, pemphigoid, chronic graft-vs-host disease). Symptomatic patients are more likely to be female. Typically, the symptom complex is intermittent solid food dysphagia. The association between iron deficiency anemia and esophageal webs is referred to as Paterson-Kelly or Plummer-Vinson syndrome.

Treatment of the typical cervical web is usually simple. Most can be disrupted during the performance of endoscopy. Esophageal bougienage may be required in some patients.

A lower esophageal ring (Schatzki's ring) is thought to be the most common cause of dysphagia (E-Fig. 35-3). The history tends to be characteristic. Patients are typically older than 40 years of age and report intermittent, solid food dysphagia occurring early in a meal. The dysphagia is usually transient, and if the bolus passes, the remainder of the meal can be consumed. Patients may go for many weeks without symptoms. The dysphagia is not progressive, and constitutional symptoms such as bleeding or weight loss are absent. Occasionally, patients with Schatzki's ring may have food impaction at presentation.

Rings can be visualized either radiographically or endoscopically, and they cause varying degrees of luminal narrowing. Rings with a luminal diameter of 13 mm or less are more likely to be symptomatic, although other factors, including bolus size and effectiveness of esophageal peristalsis, contribute to the intermittent episodes of dysphagia.

The pathogenesis of esophageal rings is not established. Current research has focused on the likely contribution of acid reflux. Evidence suggests that many patients respond to acid suppression with PPIs, in terms of both increased luminal size of the