ESOPHAGUS

The esophagus develops from the cranial portion of the foregut and is recognizable by the third week of gestation. It is a hollow, highly distensible muscular tube that extends from the epiglottis in the pharynx to the gastroesophageal junction. Acquired diseases of the esophagus run the gamut from highly lethal cancers to the persistent "heartburn" of gastroesophageal reflux that may be chronic and incapacitating or merely an occasional annoyance.

Esophageal Obstruction

The esophagus is, essentially, a tube that delivers ingested solid food and fluids to the stomach. This can be impeded by structural, i.e. (mechanical) obstruction or functional obstruction. The latter results from disruption of the coordinated waves of peristaltic contractions that follow swallowing. Esophageal manometry allows separation of esophageal dysmotility into three principal forms, termed nutcracker esophagus, diffuse esophageal spasm, and hypertensive lower esophageal sphincter.

- *Nutcracker esophagus* describes patients with high-amplitude contractions of the distal esophagus that are, in part, due to loss of the normal coordination of inner circular layer and outer longitudinal layer smooth muscle contractions.
- *Diffuse esophageal spasm* is characterized by repetitive, simultaneous contractions of the distal esophageal smooth muscle.
- Lower esophageal sphincter dysfunction, such as high resting pressure or incomplete relaxation, are present in many patients with nutcracker esophagus or diffuse esophageal spasm. In the absence altered patterns of esophageal contraction, these sphincter abnormalities are termed *hypertensive lower esophageal sphincter*. As discussed below, these can be distinguished from achalasia in that the latter includes reduced esophageal peristaltic contractions.

Because wall stress is increased, esophageal dysmotility may result in development of small diverticulae, primarily the epiphrenic diverticulum located immediately above the lower esophageal sphincter. Similarly, impaired relaxation and spasm of the cricopharyngeus muscle after swallowing can result in increased pressure within the distal pharynx and development of a Zenker diverticulum (pharyngoesophageal diverticulum), which is located immediately above the upper esophageal sphincter. Zenker diverticulae are uncommon, but typically develop after age 50 and may reach several centimeters in size. When small they may be asymptomatic, but larger Zenker diverticulae may accumulate significant amounts of food, producing a mass and symptoms that include regurgitation and halitosis.

In contrast to functional obstruction, mechanical obstruction, which can be caused by strictures or cancer, presents as progressive dysphagia that begins with inability to swallow solids. With progression ingestion of liquids is also affected. Because obstruction develops slowly, patients may subconsciously modify their diet to favor soft foods and liquids and be unaware of their condition until the obstruction is nearly complete.

Benign *esophageal stenosis*, or narrowing of the lumen, is generally caused by fibrous thickening of the submucosa and is associated with atrophy of the muscularis propria as well as secondary epithelial damage. Although occasionally congenital, stenosis is most often due to inflammation and scarring that may be caused by chronic gastroesophageal reflux, irradiation, or caustic injury. In general, patients with functional obstruction or benign strictures maintain their appetite and weight, while, as discussed later, malignant strictures are often associated with weight loss.

Esophageal mucosal webs are idiopathic ledge-like protrusions of mucosa that may cause obstruction. These uncommon lesions typically occur in women older than age 40 and can be associated with gastroesophageal reflux, chronic graft-versus-host disease, or blistering skin diseases. In the upper esophagus, webs may be accompanied by iron-deficiency anemia, glossitis, and cheilosis as part of the *Paterson-Brown-Kelly* or *Plummer-Vinson syndrome*. In general, esophageal webs are semi-circumferential lesions that protrude less than 5 mm, have a thickness of 2 to 4 mm, and are composed of a fibrovascular connective tissue and overlying epithelium. The main symptom of webs is nonprogressive dysphagia associated with incompletely chewed food.

Esophageal rings, or *Schatzki rings*, are similar to webs, but are circumferential, thicker, and include mucosa, submucosa, and, occasionally, hypertrophic muscularis propria. When present in the distal esophagus, above the gastroesophageal junction, they are termed *A rings* and are covered by squamous mucosa; in contrast, those located at the squamocolumnar junction of the lower esophagus are designated *B rings* and may have gastric cardia-type mucosa on their undersurface.

Achalasia

Increased tone of the lower esophageal sphincter (LES), as a result of impaired smooth muscle relaxation, is an important cause of esophageal obstruction. Normally, release of nitric oxide and vasoactive intestinal polypeptide from inhibitory neurons, along with interruption of normal cholinergic signaling, allows the LES to relax during swallowing. *Achalasia is characterized by the triad of incomplete LES relaxation, increased LES tone, and aperistalsis of the esophagus*. Symptoms include dysphagia for solids and liquids, difficulty in belching, and chest pain. Although there is some increased risk for esophageal cancer, it is not considered great enough to warrant surveillance endoscopy.

Primary achalasia is the result of distal esophageal inhibitory neuronal, that is, ganglion cell, degeneration. This leads to increased tone, an inability to relax of the lower esophageal sphincter, and esophageal aperistalsis. Degenerative changes in the extraesophageal vagus nerve or the dorsal motor nucleus of the vagus may also occur. The cause is unknown; rare familial cases have been described.