PART 1

Anatomy and development

CHAPTER 1

Esophagus: anatomy and structural anomalies

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An understanding of the normal and abnormal histology and structure is essential to the clinical care of patients with esophageal disorders. Esophageal biopsies obtained during endoscopy sample the squamous mucosa and less commonly the lamina propria of the esophageal wall. The histologic evaluation of submucosal glands, Meissner's and Myenteric ganglia, and the muscularis propria depicted in Figure 1.1 typically requires surgical biopsy. Methods of endoscopic mucosal resection have allowed sampling of the esophageal submucosa and muscularis mucosa. Per oral esophageal myotomy may allow for histologic evaluation of deeper mural structures. Endoscopic ultrasonography can evaluate the structural integrity and anomalies of deeper structures including the muscularis propria. Extrinsic compression of the esophagus by adjacent mediastinal structures as shown in Figure 1.2 is better appreciated on radiographic barium examination, or cross sectional imaging than endoscopy. Feline esophagus, so-called eosinophilic esophagitis, is depicted in Figure 1.3 and can be mistaken for esophageal rings. The feline pattern is a transient phenomenon visualized with retching and esophageal shortening and may represent contraction of the muscularis mucosa. Upon relaxation of the esophageal musculature and distension with air insufflation, the plications disappear.

Esophageal developmental anomalies include vascular lesions, duplications and heterotopic gastric mucosa. Kartagener's syndrome leads to right-sided rather than left-sided aortic arch esophageal compression (Figure 1.4). Patients with dysphagia lusoria present with swallowing difficulties arising from extrinsic compression of the thoracic esophagus by congenital anomalies of the aortic arch, most commonly by an aberrant take-off of the right subclavian artery from the left side of the aortic arch (Figure 1.5). Congenital venous malformations illustrated in Figure 1.6 represent another vascular anomaly and are distinct from esophageal varices as vascular obstruction or portal hypertension is not present in the former. Congenital esophageal duplications assume both tubular (Figure 1.7) and cystic (Figure 1.8 and 1.9) forms. While most are apparent before the age of 1 year, 25% can present in adults with symptoms of dysphagia. Heterotopic gastric mucosa (inlet patch) shown in Figure 1.10 is a common congenital anomaly, with a prevalence of 4% based on an autopsy series. Infrequently, this anomaly is associated with cervical esophageal stricture (Figure 1.11) and web formation (Figure 1.12). Other uncommon developmental anomalies include esophageal atresia, congenital esophageal stenosis, and bronchopulmonary foregut malformations.

Structural esophageal anomalies include esophageal rings and webs, cricopharyngeal bar, pharyngoesophageal diverticula and diffuse idiopathic skeletal hyperostosis of the cervical spine. The most widely recognized structural anomaly is the lower esophageal mucosal or Schatzki ring that is found in about 10% of adults. It is one of the most common causes of dysphagia and food impaction, although the majority of Schatzki rings are asymptomatic. The inner diameter of the ring is a critical determinant for dysphagia and can be assessed on endoscopic retroflexed view (Figure 1.13), or ingestion of a barium tablet of known diameter. A cricopharyngeal bar is found in 5%–19% of radiographic studies of the pharynx. The majority are not associated with dysphagia. Pathologic and physiologic studies support shared features between symptomatic cricopharyngeal bars and Zenker's diverticula. The patient in Figure 1.14 has both a cricopharyngeal bar and small diverticulum. Therapeutic options of symptomatic cricopharyngeal bars and Zenker's diverticula include both endoscopic and surgical approaches. Epiphrenic diverticula arise from the distal esophagus and are often associated with an underlying spastic esophageal motility disorder (Figure 1.15a and b). With time, the diverticula can increase in size resulting in food retention, bezoar formation and symptoms of regurgitation (Figure 1.16). Treatment for large or symptomatic epiphrenic diverticula is most commonly surgical and includes not only a diverticulectomy but also treatment of the underlying motility disorder. Intramural

Yamada's Atlas of Gastroenterology, Fifth Edition. Edited by Daniel K. Podolsky, Michael Camilleri, J. Gregory Fitz, Anthony N. Kalloo, Fergus Shanahan, and Timothy C. Wang.

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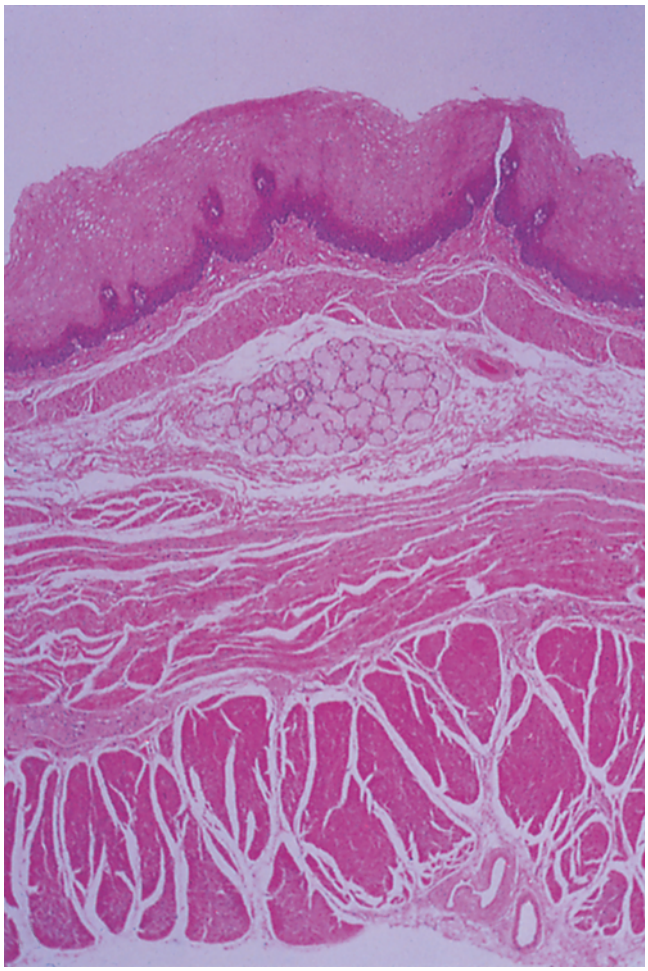

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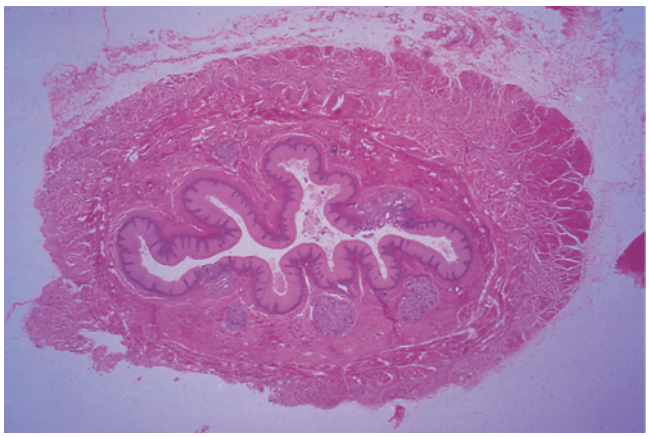


(b)

Figure 1.1 **(a)** This cross section (×2.5) from the middle third of the esophagus has a mixture of skeletal and predominantly smooth muscle in the muscularis propria. The submucosal glands are clearly shown. An esophageal cardiac gland in which a small focus of glandular epithelium interrupts the squamous mucosa is a normal finding, seen in at least 1% of all esophagi. **(b)** Longitudinal section of esophageal wall (×10). Source: Courtesy of Rodger C. Haggitt, M.D., Seattle, WA.

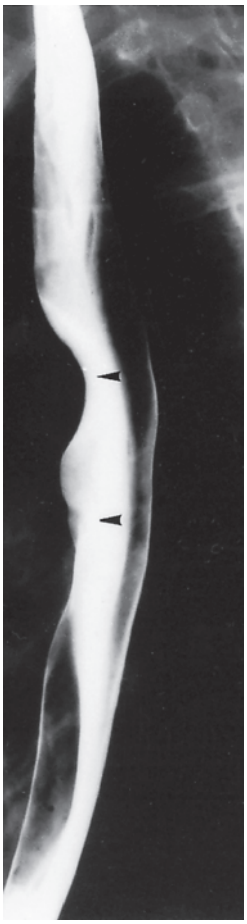
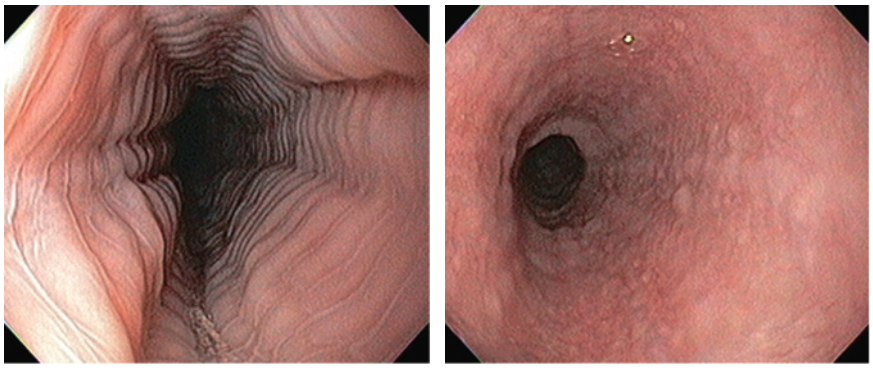


Figure 1.2 Barium esophagram shows normal indentation of the esophageal lumen but the aorta (top arrowhead) and left mainstem bronchus (bottom arrowhead).

pseudodiverticulosis is a rare finding best appreciated on barium esophagram rather than upper endoscopy. The disorder results from dilation on excretory ducts of submucosal esophageal glands and is associated with proximal esophageal strictures and esophageal candidiasis. Diffuse idiopathic skeletal hyperostosis (DISH) of the cervical spine leads to ossification of the anterolateral ligaments and enthuses. Dysphagia may result from extrinsic compression of the cervical esophagus (Figure 1.17).



(a) (b)

Figure 1.3 **(a)** Feline esophagus demonstrating rippling or plications of the esophageal mucosa. This is a transient occurrence and disappears with continued observation. **(b)** Eosinophilic esophagitis can present with a similar appearance but the rings persist with air insufflation and less tightly spaced apart.

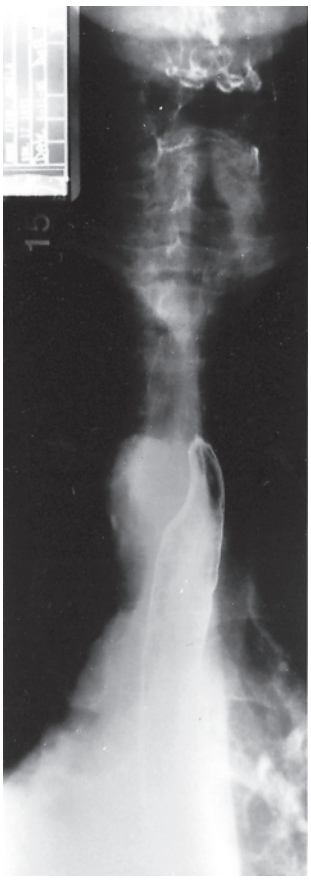
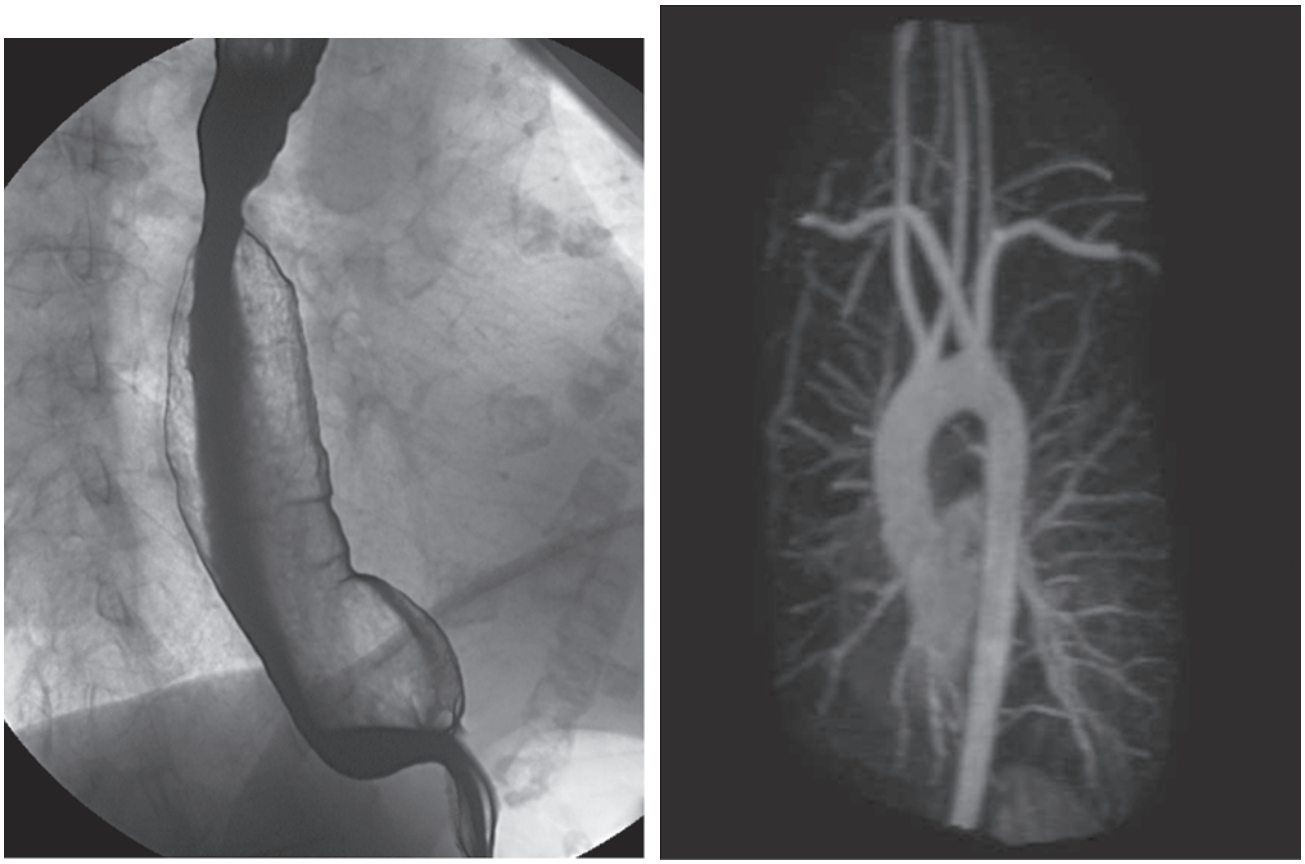


Figure 1.4 Barium esophagram of a patient with Kartagener's syndrome showing esophageal compression by the right sided aortic arch and dextrocardia.



(a) (b)

Figure 1.5 Dysphagia lusoria represents symptomatic esophageal compression by a vascular anomaly of the aortic arch, most commonly by an aberrant right subclavian artery. **(a)** Barium esophagram in a patient reveals thoracic esophageal compression by an aberrant right subclavian artery posterior to the esophagus. **(b)** Magnetic resonance angiography reveals an aberrant right subclavian artery arising from the aortic arch.

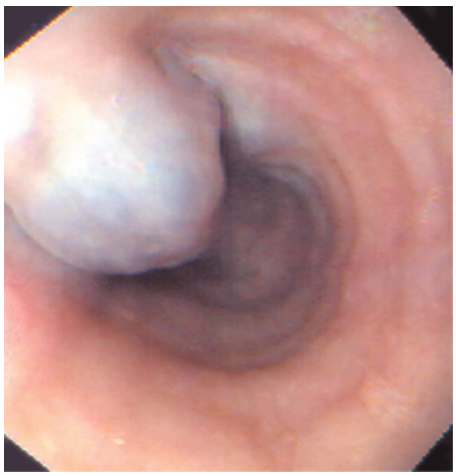
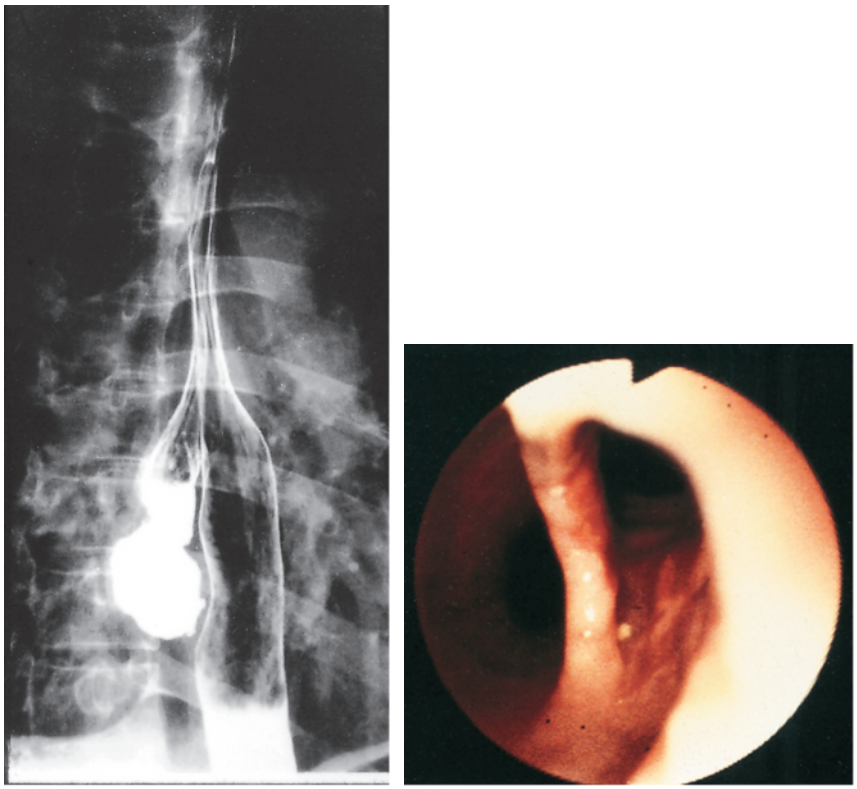
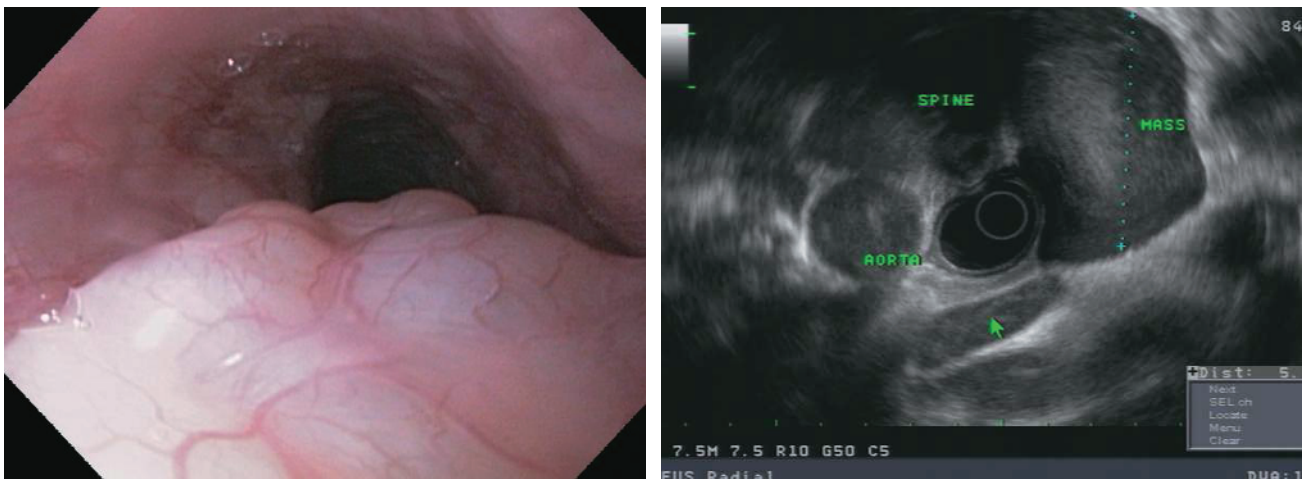


Figure 1.6 Congenital venous malformations as depicted may also be referred to as primary esophageal varices when no secondary cause such as portal hypertension can be identified. These venous structures rarely bleed spontaneously. Endosonography confirmed a conglomerate of venous channels in this case.



(a) (b)

Figure 1.7 **(a)** Radiograph showing a large, congenital, tubular duplication of the esophagus. **(b)** Endoscopic view showing the opening to the tubular duplication (right) and esophageal lumen (left). Congenital esophageal duplications may be tubular or cystic.



(a) (b)

Figure 1.8 **(a)** Congenital esophageal duplication cysts may be present as submucosal lesions on upper endoscopy. **(b)** Endoscopic ultrasonographic image of a large duplication cyst. Duplication cysts are the second most common benign esophageal submucosal lesion with stromal tumors being more common. Source: Images courtesy of Sri Komanduri, MD.

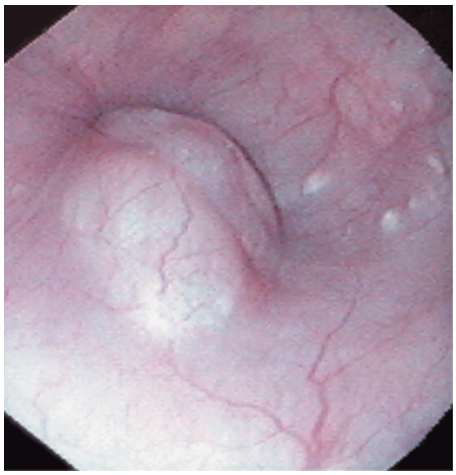


Figure 1.9 Small intramural cysts such as the bilobate type shown here are not symptomatic and are typically identified on barium esophagram or endoscopy for another indication. The cystic nature of the lesion can be confirmed using endoscopic ultrasonography. The differential diagnosis includes submucosal esophageal lesions and esophageal varices.

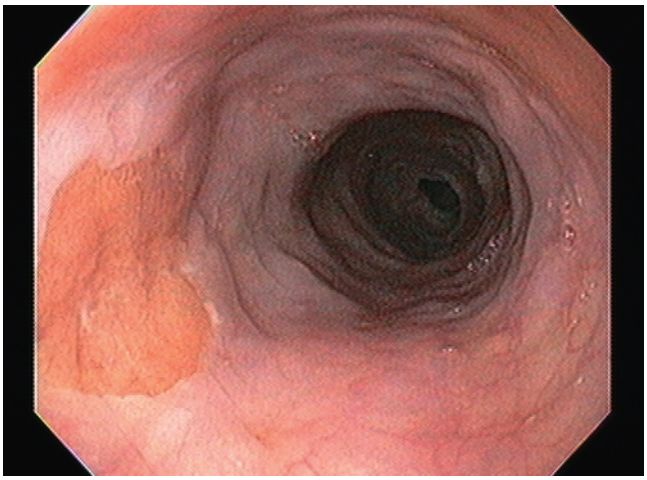


Figure 1.10 Heterotopic gastric mucosa (inlet patch) in the cervical esophagus. The reported prevalence approximates 4%. The lesions can be unifocal as in the case illustrated, multifocal or circumferential.

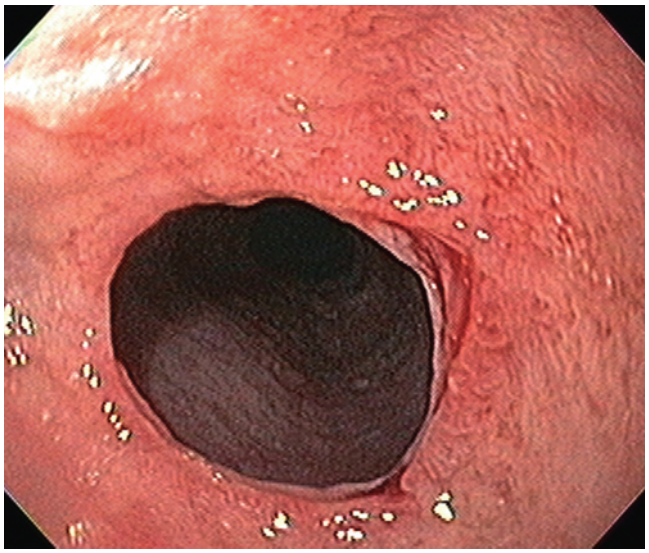
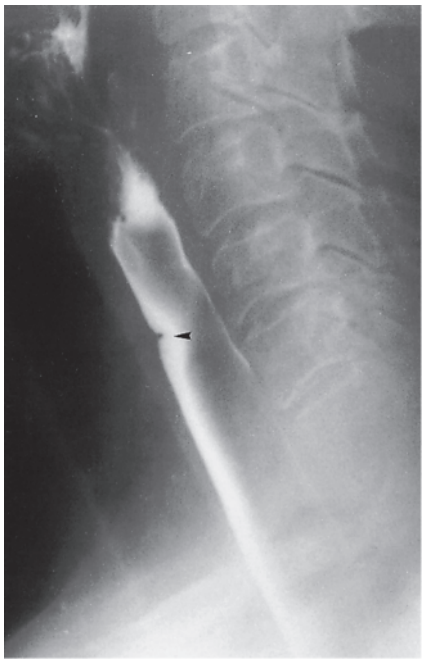


Figure 1.11 A large, circumferential focus of heterotopic gastric mucosa in the cervical esophagus associated with a circumferential mucosal web immediately distally. The web in this case likely represents a form of peptic stricture related to acid secretion from parietal cells within the inlet patch.



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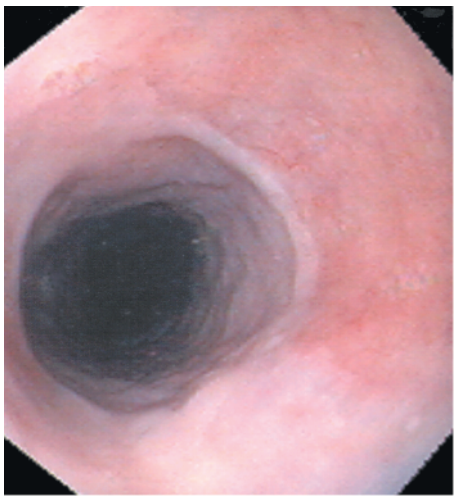
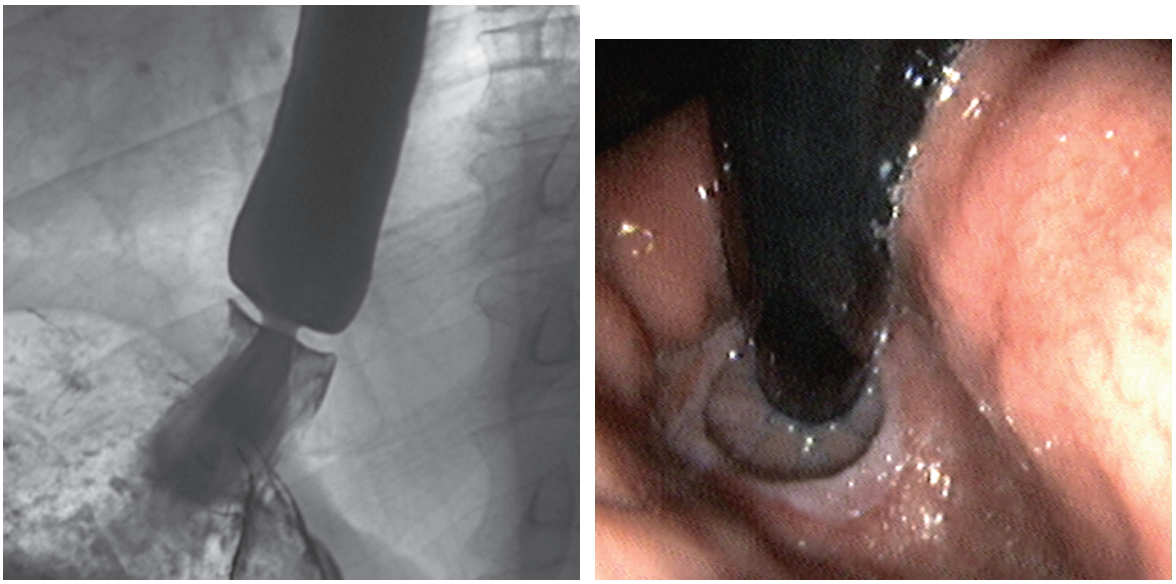


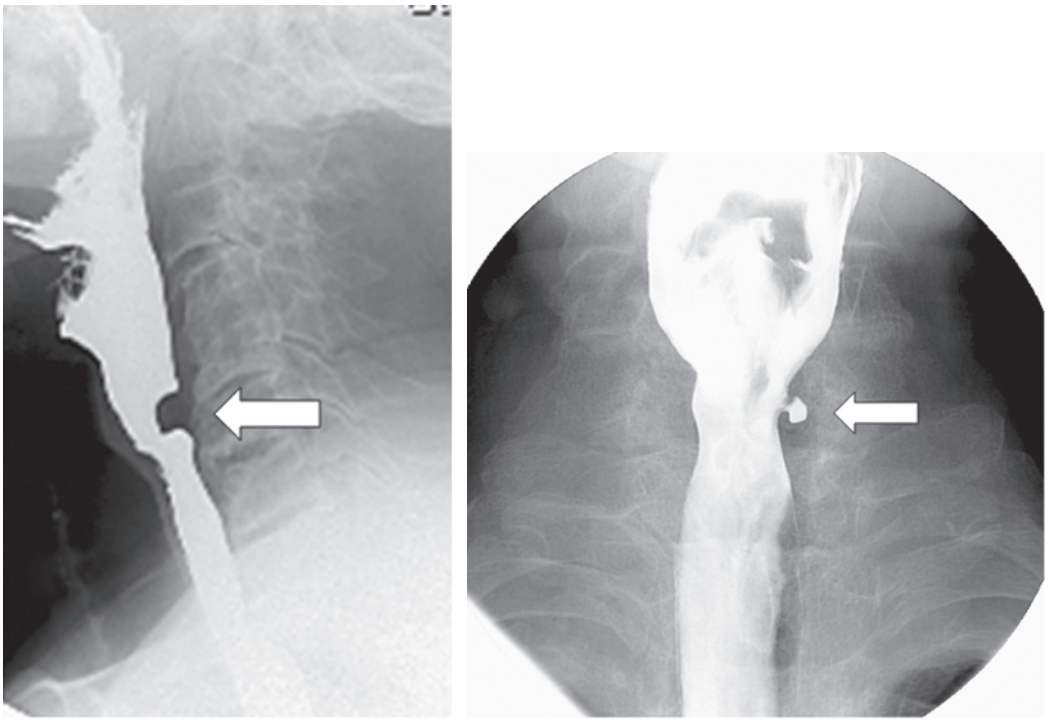
(b)

Figure 1.12 **(a)** Barium contrast radiograph showing a mucosal web in the cervical esophagus, often an incidental finding. **(b)** Corresponding endoscopic view of the cervical web from A demonstrates a proximal gastric inlet patch with web creating a shelf or lip at the distal aspect of the heterotopic gastric mucosa.



(a) (b)

Figure 1.13 **(a)** A high grade stenosis from a Schatzki ring located at the esophagogastric junction on a barium esophagram. **(b)** Retroflexed endoscopic view of a Schatzki ring. Schatzki's rings are almost invariably seen in association with hiatal hernia as is the cases here. The inner ring diameter of a Schatzki ring is an important determinant of whether the ring is associated with dysphagia.



(a) (b)

Figure 1.14 **(a)** Barium esophagram depicting a cricopharyngeal bar in an elderly patient presenting with dysphagia. The bar is a posterior indentation (arrow) arising from the cricopharyngeus muscle. **(b)** A small Zenker's diverticulum (arrow) is seen in the same patient originating from the left lateral aspect of the posterior pharynx in this anterior-posterior view. Physiologic data links the pathogenesis of Zenker's diverticula with increased intraluminal pressure that develops as a result of limited opening of the upper esophageal sphincter.

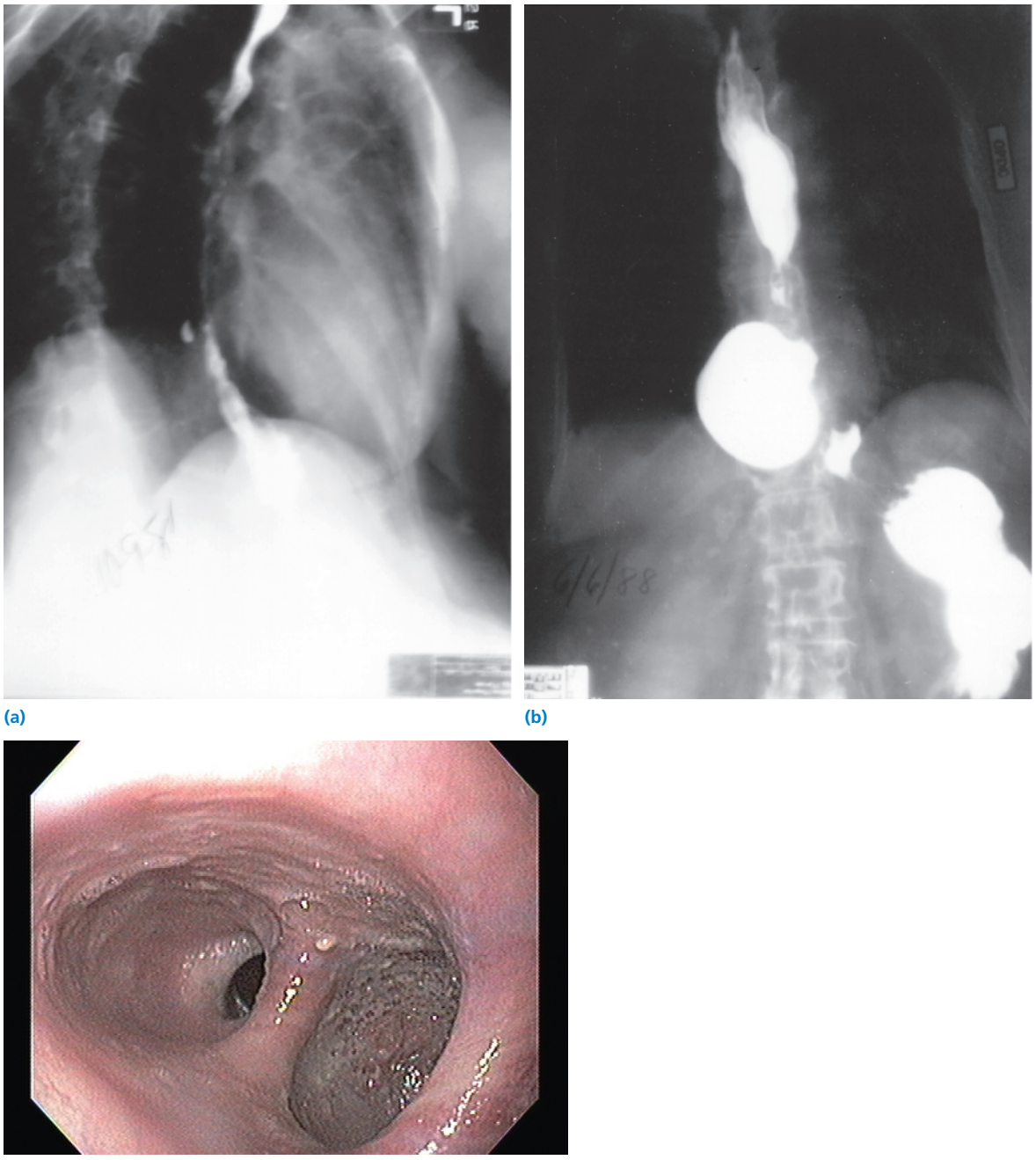


Figure 1.15 **(a)** Esophagram of a 75 year old woman shows a tiny epiphrenic diverticulum projecting to the right side in the distal esophagus. **(b)** Eight years later, there was a marked increase in the size of the diverticulum and the patient developed symptoms of dysphagia and chest pain. **(c)** In another patient, a moderate sized, wide mouthed diverticulum originates to the right of the esophageal lumen.

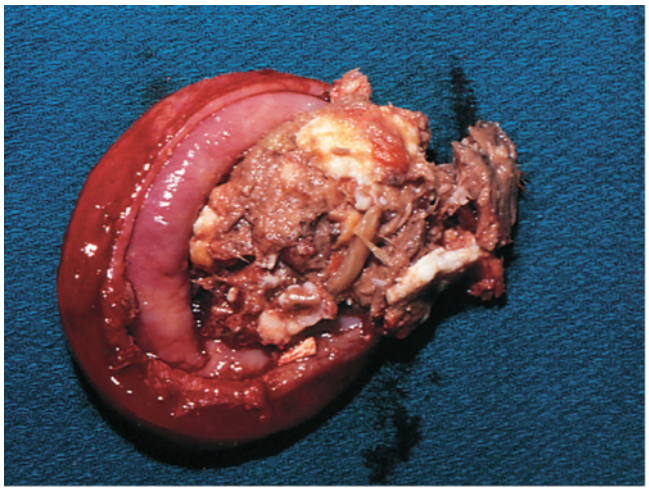


Figure 1.16 Surgical specimen of a resected esophageal diverticulum which contained a large bezoar. Source: Courtesy of Thomas W. Rice, MD.

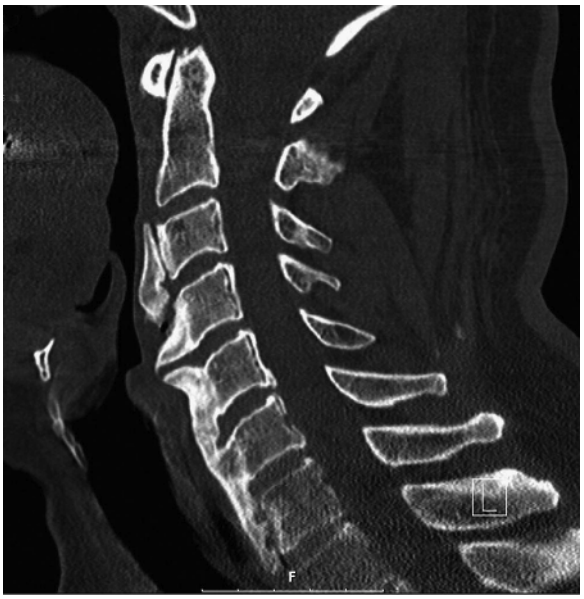


Figure 1.17 Sagittal computed tomography view of the cervical spine of a 62 year old man with diffuse idiopathic skeletal hyperostosis and moderate dysphagia. Anterior ossification of C3–C7 produces extrinsic compression of the esophageal inlet and cervical esophagus. Source: Verlaan J-J, Boswijk PFE, de Ru JA, Dhert WJA, Oner FC. The Spine Journal 2011(11);1058–1067. Reproduced with permission from Elsevier.