The Esophagus

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Contents

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1.1 Macroscopic Anatomy

1.1.1 Shape and Structure

The digestive tract begins, we often forget, with the oral cavity. The teeth grind the food before the tongue and the striated muscles of the pharynx push it into the esophagus.

(a) Esophagus. The esophagus is a tube with an approximate diameter of 2.5 cm, which allows the passage of food from the oral cavity through the thoracic cavity into the stomach. The esophagus starts at the cricopharyngeal muscle (or upper esophageal sphincter) located at about 15 cm of the incisors, anterior to the C6 vertebra, and continues until the lower esophageal sphincter (positioned at the cardia, approximately 40 cm from the incisors at the level of T11 vertebra) before its entry in the abdominal cavity and the stomach. The esophagus is classically divided into three relatively identical segments identified as the upper, middle, and lower thirds of the esophagus (Fig. 1.1a).

At the cervical level, the esophagus is located in front of the vertebral column and behind the trachea. On each side, we find the carotid arteries and the recurrent laryngeal nerves, branches of the vagus nerve controlling motility of the pharynx and upper esophagus.

The thoracic esophagus then passes into the posterior mediastinum in front of the spine, behind the trachea, the carina, the heart, the aortic arch, then to the right of the aorta, and bordered by the pulmonary pleura (Fig. 1.1b).

The abdominal esophagus is made up of a short segment (1–2 cm long) between the diaphragm and the stomach. The two branches of the vagus nerve descend from the central nervous system by running on both sides of the esophagus; at the level of the diaphragmatic hiatus, the left and right branches of the vagus nerve are found on the anterior and posterior walls, respectively, of the esophagus (due to the rotation of the stomach during its fetal development; see ► Sect. 2.3 in ► Chap. 2). The surgical procedure of truncal vagotomy, once used to reduce gastric acid secretion and treat peptic ulcer, consisted of sectioning both of these vagal branches in the lower esophagus.

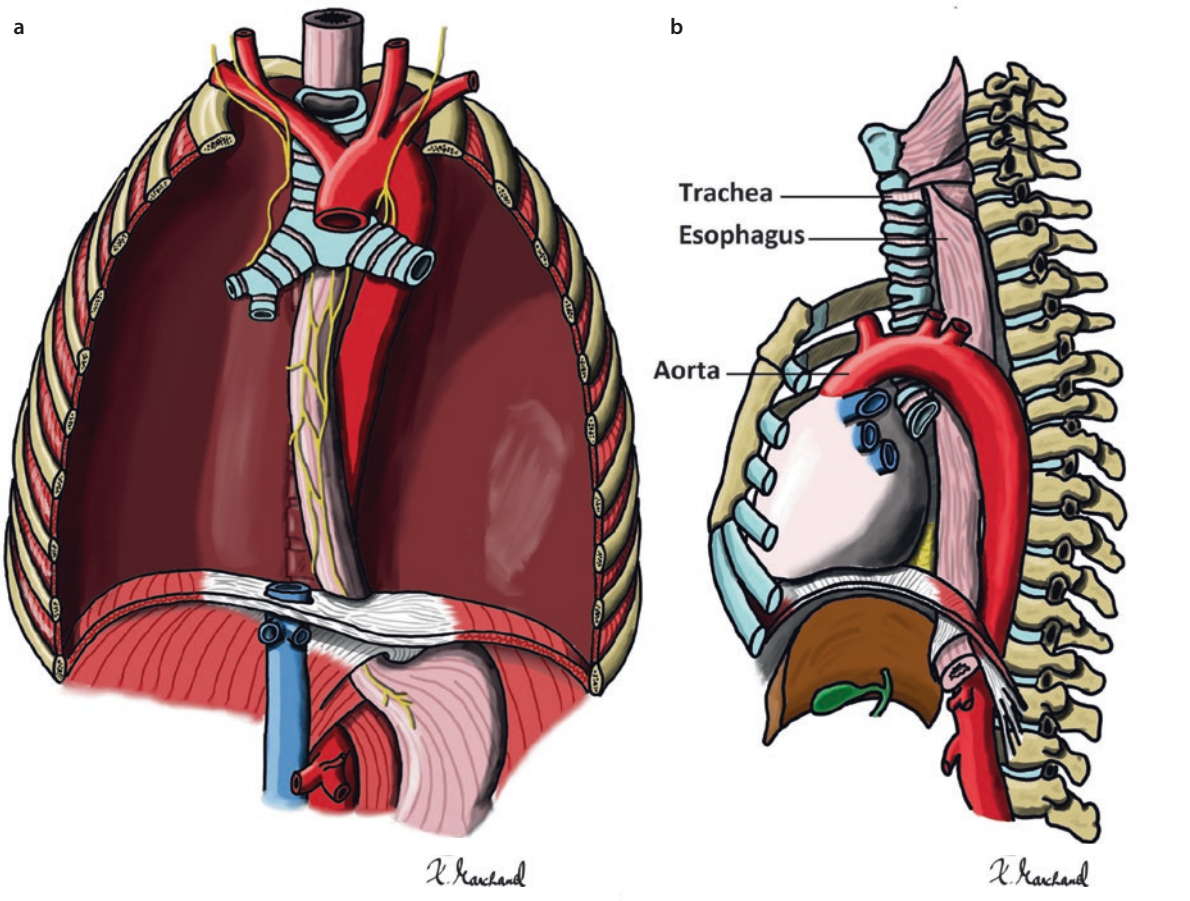

Fig. 1.1 **a** (left figure). Esophagus anatomy: front view. **b** (right figure). Esophagus anatomy: side view

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(b) Sphincters. At both ends of the esophageal tube, we find sphincters, areas of high pressure designed to close the tube entrance and acting as a one-way valve.

The upper esophageal sphincter (UES) or cricopharyngeal muscle is composed of fibers from pharyngeal muscles aligned transversally and of transverse fibers from the esophagus that encircle the esophageal tube and form a high-pressure zone probably intended to protect the tracheal airway from possible gastroesophageal regurgitations. Between the oblique muscle fibers from the pharynx and the transverse fibers from the upper esophageal sphincter, we find an area of potential weakness, Killian's triangle, that may give rise to Zenker's diverticulum (discussed later in this chapter), especially in the presence of a high pressure within the UES that generates an obstacle to food swallowing.

The lower esophageal sphincter (LES) is an area of high pressure, approximately 2 cm long, located at the esophagogastric junction that prevents regurgitation of gastric contents into the esophagus. It is normally located at the thoracoabdominal junction, at the level of the diaphragm, mainly on its abdominal side. This sphincter area is made up of muscle fibers from the lower esophagus ("internal" sphincter), and muscle fibers from the gastric fundus as well as the diaphragm pillars ("external" sphincter). Its role in gastroesophageal reflux disease (GERD) will be discussed subsequently.

1.1.2 Vascular Supply

(a) Arteries. The esophagus is supplied in its upper segment by four to six small arteries derived from the thyroid arteries, by arteries originating directly from the aorta or derived from intercostal or bronchial arteries in the middle segment, and by gastric arteries in the distal segment (Fig. 1.2a). The arteries form a dense irrigation network which protects the esophagus from an ischemic process.

(b) Veins. An extensive network of small veins drains the esophagus via the thyroid veins at its upper part and toward the azygos and intercostal veins in its middle

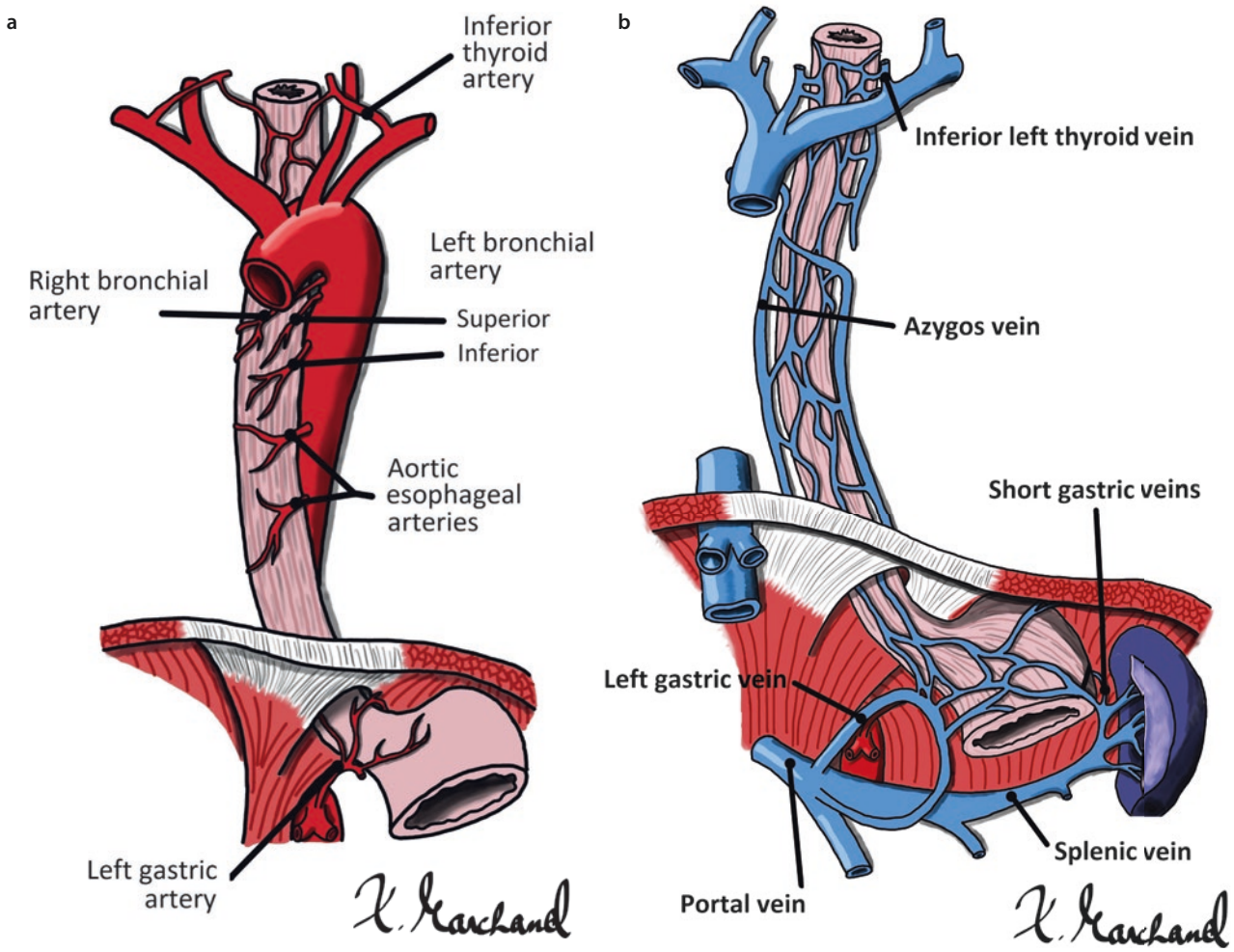


Fig. 1.2 **a** (left figure). Arteries of the esophagus. **b** (right figure). Veins of the esophagus

part. At the lower portion, esophageal veins may drain toward the abdomen to the short gastric veins or to the left gastric vein and thus toward the portal circulation; this explains then the possible formation of venous dilatations in the lower esophagus (called esophageal varices) in the setting of portal hypertension due to liver cirrhosis (◘ Fig. 1.2b).

(c) Lymphatics. Lymphatic channels arise in the mucosa and muscular region of the esophagus and drain to paraesophageal lymph nodes distributed all along the organ. In case of neoplasia, the lymphatic flow from the upper 2/3 of the esophagus moves upward (cervical lymph nodes, paratracheal lymph nodes of the upper mediastinum, paraesophageal lymph nodes of the middle or inferior mediastinum, etc.), while the lower esophagus may drain into the abdomen to celiac and perigastric lymph nodes.

The abdominal lymphatic secretions are channeled, via the thoracic duct running up along the esophagus, to join the left subclavian vein near by the jugular vein. Trauma, surgical or otherwise induced, of the cervical esophagus, may therefore damage this structure and lead to chylothorax and even intestinal lymphangiectasias.

1.1.3 Innervation

Intrinsic innervation of the esophagus is provided by the enteric nervous system (ENS) with a relatively sparse Meissner's submucosal plexus (controlling microcirculation and secretion) and Auerbach's myenteric plexus (regulating muscle contractions for motility), as elsewhere in the gastrointestinal tract (see ▶ Chap. 3).

Extrinsic innervation depends on sympathetic and parasympathetic systems. Cervical and thoracic sympathetic ganglia provide motor and sensitive innervation to the entire esophagus. The vagus nerve exerts a parasympathetic motor innervation to the upper esophagus as well as in the pharynx. Afferent fibers of the vagus nerve are also probably important in esophageal sensory transmission.

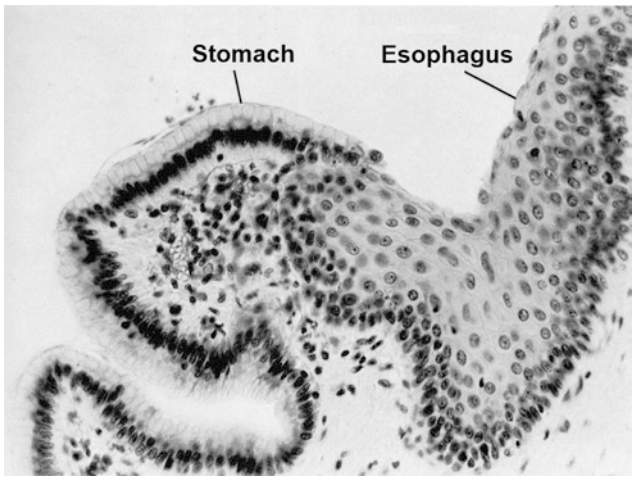
At the buccopharyngeal level, cranial nerves IX and XII are responsible, respectively, for sensitive and motor innervation. A damage of these nerves, after a vascular stroke of the brain stem, for example, will create swallowing dysfunction.

1.2 Microscopic Anatomy

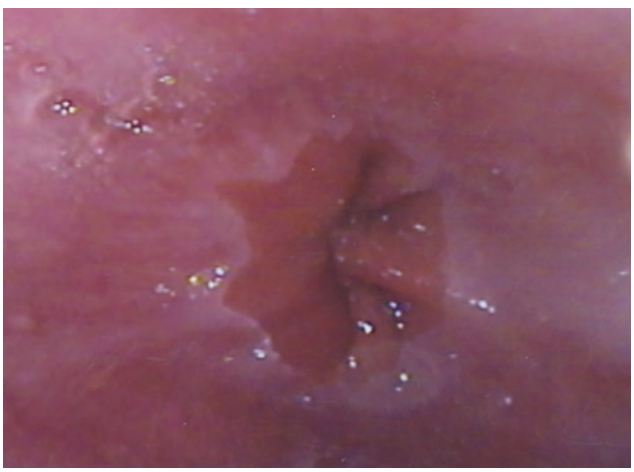
Like the rest of the digestive tract, the esophagus is made up of an internal mucosa layer resting on a muscular structure. The peculiarities of the esophagus are as follows:

1.2.1 Esophageal Mucosa

The esophageal *mucosa* is made of a stratified squamous epithelium (◘ Fig. 1.3). This squamous mucous membrane is also found at the very distal end of the digestive tract, at the anus, while the columnar (glandular) epithelium is the normal histology of the entire gastrointestinal tract from the stomach to the rectum. At approximately 40 cm from the incisor teeth, at the gastric cardia level, we find the Z line that marks the transition, clearly visible macroscopically (e.g., in endoscopy; ◘ Fig. 1.4), between the whitish squamous mucosa of



◘ **Fig. 1.3** Esophagus normal histology: squamous mucosa of the esophagus (on the right part of the figure) and columnar mucosa of the stomach (on the left part) as seen at the gastroesophageal junction. (Photo from W. Bloom and D.W. Fawcett, Textbook of Histology, 1968)



◘ **Fig. 1.4** In endoscopy, the meeting junction between the gastric reddish mucosa (at the center of the image) and of the esophageal whitish mucosa is obvious. (Photo by P.Poitras)

the esophagus and the reddish columnar epithelium of the stomach. The Z line corresponds to the lower portion of the lower esophageal sphincter.

The epithelial structure of the esophagus explains the relative paucity of absorptive or secretory phenomena so common to the rest of the digestive tract. This squamous mucous membrane also explains the presence of squamous cell neoplasia at this site level of the digestive tract. In Barrett's esophagus (secondary to GE reflux as discussed later), the squamous mucosa is replaced by a columnar mucosa allowing then the development of adenocarcinomas.

In the *submucosa*, we find blood vessels, lymphatics, some nerves of Meissner's plexus, as well as glandular cells secreting mucus and bicarbonate involved in the defense of mucosal integrity.

1.2.2 Muscularis

The esophagus, as most of the digestive tract, is made up mainly of smooth muscles organized in a circular inner layer and a longitudinal outer layer. The upper esophagus, however, contains striated muscles that are in fact an extension of the pharyngeal muscles. Diseases of the smooth muscles can thus affect the middle and distal esophagus, while pathologies of the striated muscles can affect the proximal region of the esophagus.

As elsewhere in the gastrointestinal tract, the intrinsic enteric nervous system is made up of the myenteric and submucous plexuses, respectively, located between the longitudinal and circular muscle layers and between the circular muscles and the mucous membrane.

1.2.3 Serosa

The serosa, usually lining the external muscle layer of digestive cavities, is absent in the esophagus. It is replaced by a thin adventitia (thin connective tissue layer).

1.3 Embryology/Development

1.3.1 Normal Development

The oropharynx, trachea, lungs, and esophagus develop from a common tube: the endoderm. In the 4th week of fetal life, a bud forms at the ventral part of the tube to become the respiratory system. The dorsal part of the tube will turn into the esophagus and a foregut from which the stomach will arise. The separation of the two tubes in respiratory and digestive organs is carried out around the 6th week. Esophageal lumen is formed around week 10 and will be epithelialized with squamous cells from the 16th week. From week 18 to 20, the fetus can swallow up to 500 ml amniotic fluid per day; the impossibility to perform this function (e.g., due, to esophageal atresia) may lead to an increase in this fluid and of uterine volume. Mechanisms of suction-deglutition and of esophageal peristalsis are mature enough at 34 weeks to allow bottle-feeding from that point onward.

We will now describe different malformations that may arise during development.

1.3.2 Atresias of the Esophagus

Atresias of the esophagus are often accompanied by a tracheoesophageal fistula (Fig. 1.5). They affect 1/3000 to 1/4500 births; 50% of the children carriers of these manifestations also have other abnormalities such as imperforate anus and heart defects.

Atresia of the esophagus results from an impaired canalization of the esophageal lumen, while broncho-esophageal fistulas are more likely the result of a separation failure of the two tubes.

These malformations will be suspected in a newborn baby that regurgitates saliva and liquids after ingestion. Passage through the bronchial tubes will cause coughing

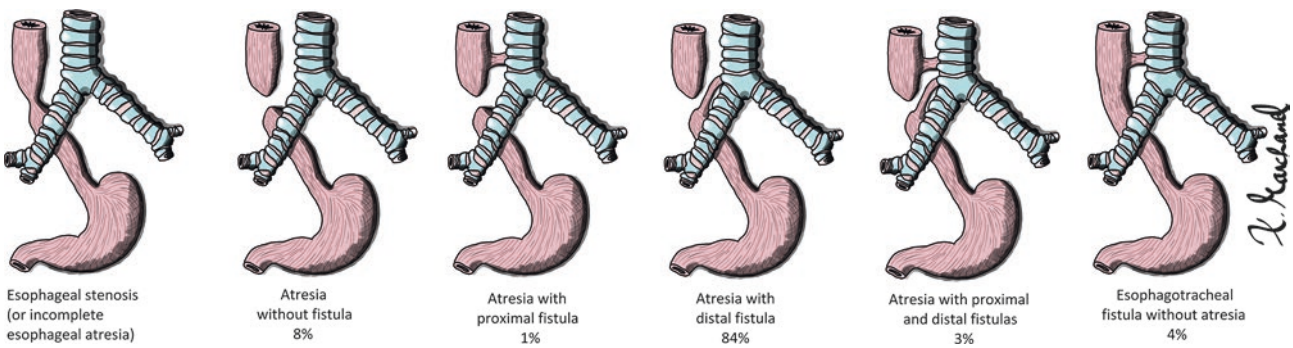


Fig. 1.5 Congenital atresia of the esophagus: various malformations are possible

and choking. Surgical correction will be required promptly.

1.3.3 Esophageal Stenosis/Strictures

These are rare congenital malformations that occur in 1/50,000 births. Stenoses are most often short and of fibrous or cartilaginous in nature (tracheobronchial remnant). They are often tolerated in the very young age when feeding only liquid food, but they will be revealed subsequently when dysphagia occurs when taking solid food.

1.3.4 Duplications and Cysts

Duplications and cysts are found in 1/8000 births. Double esophageal tubes are rare; most of the time, it consists in "cystic" structures located in the paraoesophageal region and without communication with the real esophagus. Patients are often asymptomatic (the condition being discovered accidentally during a radiological examination) or may suffer from symptoms due to compression by these additional structures. Surgical treatment will be required to control symptoms.

1.3.5 Rings and Webs

The most common ring is Schatzki's ring, an annular fibrous ring located at the esophagogastric junction. However, the congenital nature of Schatzki's ring is being questioned; there is increasing evidence that this band is the consequence of gastroesophageal reflux disease (GERD). See ▶ Sect. 1.9, point 6.

The membrane, or web, is a noncircumferential band, found mostly in the upper esophagus or middle esophagus. It can be solitary, sometimes associated with iron deficiency anemia, then called Plummer-Vinson's syndrome (or Paterson-Kelly's syndrome), or they can be multiple.

Esophageal dilatation per endoscopy is the usual treatment of symptomatic rings and webs.

1.4 Secretion/Absorption

Although some secretion of bicarbonates and mucus by submucosal glands occurs, overall the esophagus has minimal absorptive or secretory function.

Swallowing, however, benefits from saliva secretion. The parotid, submaxillary, and sublingual glands produce 1–2 liters of liquid daily, often in response to chewing, which will help the swallowing by lubricating the food. Saliva also contains enzymes, such as amylase and lipase, which initiate the digestion process. Bicarbonate-rich saliva plays an important role in the esophagus's ability to neutralize the gastric acid that could backflow into it.

1.5 Motility/Sensitivity

The role of the esophagus is to transport food from the mouth to the stomach. The act of swallowing (deglutition) involves a transfer of the food bolus from the oral cavity to the esophageal cavity and then a transport of the bolus through the esophageal body toward the stomach.

1.5.1 Oropharyngeal (Transfer) Motility

The food undergoes a first transformation in our plate (shredding by our utensils) before its form is again altered by dental chewing. This is how the food bolus is formed. The transfer of this bolus into the esophageal cavity initially involves a voluntary action of the striated muscles of the tongue and oropharynx, controlled mainly by the cranial nerve XII (hypoglossal nerve), followed by involuntary movements of the palate, the pharynx, the larynx, as well as the upper esophagus, all regulated by the vagus nerve (CN X) and glossopharyngeal nerve (CN IX). The stages of swallowing are represented in ◘ Fig. 1.6.

Any damage to neural structures (brain stem damage by vascular stroke, amyotrophic lateral sclerosis, Parkinson's, etc.) or striated muscles (oculopharyngeal dystrophy, myasthenia gravis, etc.) of this region may disrupt the physiology of swallowing and lead to a proximal dysphagia with possible false routes to the upper (nasal regurgitations) or lower (cough, choking) airways.

1.5.2 Esophageal (Transport) Motility

Once the voluntary swallowing of the bolus provided by the striated muscles of the oropharynx has pushed the food bolus into the esophageal cavity, the smooth muscles of the esophagus will ensure the progression of the

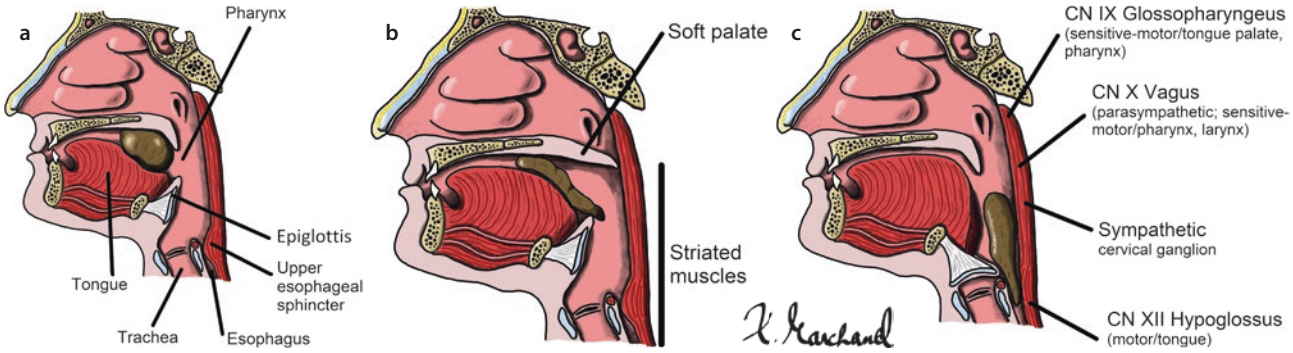


Fig. 1.6 Swallowing: **a** The bolus is pushed by the tongue towards the pharynx. **b** The soft palate rises to block the nasopharynx (and avoid regurgitation of food through the nose). **c** The larynx moves upward and anteriorly, while the epiglottis goes down to block the entrance to the trachea (and avoid intrabronchial aspiration of food resulting in acute airway obstruction, or complicated by aspiration pneumonia); the upper esophageal sphincter, normally closed and under tension, relaxes to let the food bolus get into the esophageal cavity where transport motility will take over. Swallowing is controlled by cranial nerves IX, X, and XII and cervical sympathetic ganglion

bolus along the organ. As explained extensively in ▶ Chap. 3, this peristalsis is mainly involuntary and relies on the enteric nervous system of Auerbach's plexus. The following steps are involved:

- Detection of the intraluminal food bolus with activation of sensitive afferent neural fibers [usually CGRP (calcitonin gene-related peptide)] in mucosa and muscle layers of the intestinal wall.
- The information is transmitted to the efferent motor neurons that will activate, via acetylcholine and neurokinins, the contraction of the circular muscles of the esophagus upstream of the bolus.
- At the same time, the efferent neural fibers will cause the relaxation, via VIP (vasointestinal polypeptide) and NO (nitric oxide), of the circular muscles localized downstream of the bolus to allow a well-synchronized propulsion forward.

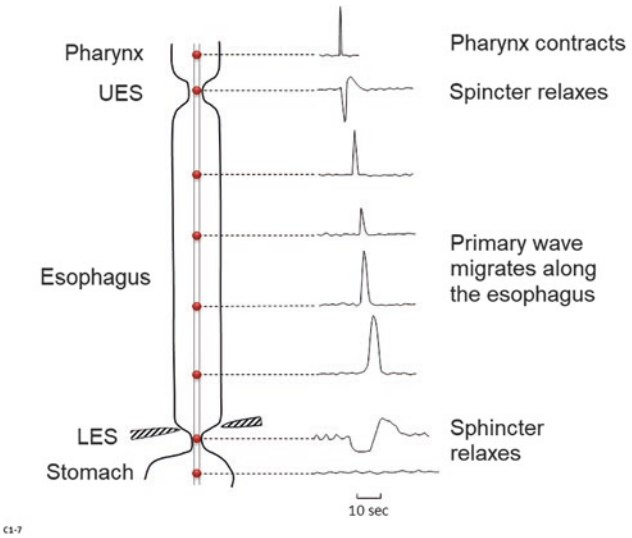


Fig. 1.7 Normal motility (on standard classical manometry) of the esophagus recorded using a probe inserted through the nose into the esophagus and the stomach to measure, at various levels of the organ, the intraluminal pressure variations secondary to contractions of the esophageal wall during swallowing a sip of water

Esophageal motility can be easily analyzed in clinical or research settings using manometry with the help of pressure sensors (installed on a thin tube or a wire introduced, usually nasally, into the esophagus to the stomach) that record the contractions of the esophagus at different points along the organ. Various contractile waves are identified (Fig. 1.7):

- The *primary wave* is a peristaltic contraction normally initiated in response to the ingestion of the food bolus. It migrates, at a rate of 3–4 cm/second, all along the esophagus, from top to bottom, pushing the bolus in front of it. The primary wave thus allows eating in a lying down position ("Roman" style), or even upside down as the opossum. In case of disappearance of this contractile wave, usually by damage to the smooth muscles of the esophagus (e.g., scleroderma), gravity becomes the only factor allowing the descent of food along the esophagus toward the stomach (and thus forcing a vertical position when eating).
- The *secondary wave* is identical to the primary wave above described, but not induced by swallowing. Initiated by local distension, it can start at any level of the esophagus and migrate downward. It can be triggered experimentally by the infusion of a liquid or by local distension (with a balloon) of an esophageal segment. It is generated strictly by the sensory and motor functions of the peristaltic reflex described before. In practice, it will occur during episodes of reflux to "clean" the esophagus of material refluxed by the stomach that could irritate or inflame the esophagus.

- *Tertiary waves* are nonperistaltic simultaneous contractions. They are independent of swallowing and have no obvious motor function. They are encountered during pathological states such as achalasia or diffuse spasm.

1.5.3 Sphincters: Upper and Lower

Sphincters are areas of high intraluminal pressure that act as barriers functioning like unidirectional valves being closed and contracted in the resting state to prevent reflux, but getting relaxed and opened during swallowing to allow the passage of the bolus across.

(a) Upper esophageal sphincter or cricopharyngeal muscle. The upper esophageal sphincter (UES), which is composed of striated muscles, forms a barrier between the esophageal and oropharyngeal cavities, thus serving to protect the trachea of unexpected reflux from the esophagus.

When the bolus arrives from the oropharynx, the sphincter relaxes and opens to let the bolus enter the esophageal cavity. The cranial nerves IX, X, and XII play an important role in this coordination. Impaired relaxation or opening of the UES will result in upper (oropharyngeal/ transfer) dysphagia, and possibly the creation of a diverticular pouch (Zenker's diverticulum; see ► Sect. 1.9.1) in the area of wall weakness (Killian's triangle) between the oblique pharyngeal muscle fibers and the transverse fibers of the cricopharyngeus.

(b) Lower esophageal sphincter. LES is a high-pressure area, created by tonic contraction of specialized circular smooth muscle, that separates the stomach from the esophagus to prevent reflux of gastric material to the esophagus.

Hypotension or defective contraction of this sphincter will result in gastroesophageal reflux and possibly secondary esophagitis.

The LES normally contracted during the "resting state" must relax to open in a coordinated manner at the arrival of the esophageal bolus for its passage into the stomach. This highly coordinated relaxation relies on the NO and VIP inhibitory neurons of the enteric nervous system. Insufficient relaxation of the LES (as encountered in achalasia discussed later) will compromise esophageal transit, and this will result in lower (esophageal/transport) dysphagia.

Transient inappropriate lower sphincter relaxation (TILSR) not triggered by swallowing may occur "spontaneously" (or due to distension of the gastric fundus; 2–3/hr in normal individuals) and allow the retrograde passage of air (belching) or, unfortunately, of gastric fluid (gastroesophageal reflux). Their increased frequency is responsible for most cases of GE reflux disease (GERD) as discussed later.

1.5.4 Sensory Function

Sensory function is essential to detect the presence of the intraluminal food bolus (or refluxed gastric material) which then triggers peristaltic contractions that will harmoniously propel esophageal content toward the stomach.

In normal situations, we are not consciously aware of the passage of food or other sensations within the esophagus. Painful sensations can be perceived in abnormal situations, such as strong spastic contractions, food blockage, or prolonged acid exposure to the esophageal mucosa. Sensory or painful sensation can also be facilitated by visceral hypersensitivity as documented in various functional digestive disorders.

1.6 Inflammation Disorders

Inflammation of the esophagus can appear under different forms in response to different causal factors.

1.6.1 Peptic Esophagitis

Peptic esophagitis is secondary to the reflux of acidic gastric content into the esophagus [GERD (gastroesophageal reflux disease) extensively discussed in this chapter in the Sect. ► 1.8]. Peptic esophagitis is the most common form of esophagitis met in practice.

(a) Symptoms of peptic esophagitis. Esophagitis is clinically revealed by sensations of chest discomfort. Regurgitation of food or acidic-bilious liquid, retrosternal burning, and pyrosis (burning sensation along the esophagus from the bottom to the top) suggest GERD (with or without esophagitis); odynophagia (pain during the passage of food bolus) or dysphagia (feeling of food getting stuck during passage to the stomach) suggests esophagitis (secondary to acid GE reflux).

(b) Diagnosis of esophagitis. Esophageal endoscopy is the best way to diagnose esophagitis. The macroscopic appearance of esophageal mucosa seen at endoscopy (◘ Fig. 1.8) and/or its microscopic aspect on mucosal biopsies obtained during endoscopy will establish the diagnosis.

(c) Pathophysiology of peptic esophagitis. Acid-peptic reflux from the stomach is the dominant cause of peptic esophagitis. However, all individuals with GERD are

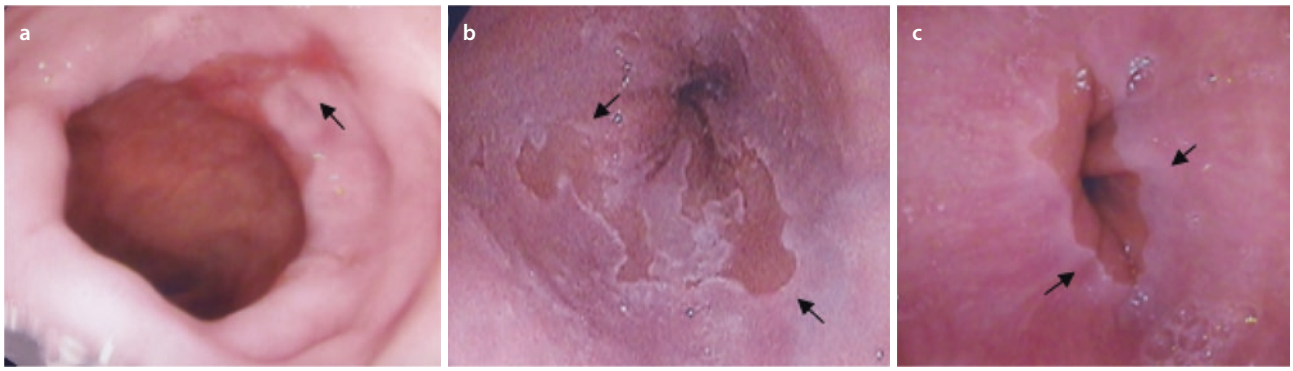


Fig. 1.8 Peptic esophagitis seen in endoscopy: **a** re-epithelialized ulceration (arrow) at the gastroesophageal junction (the gastric columnar and esophageal squamous mucosa are, respectively, of reddish and whitish coloration); **b** Barrett's esophagitis with columnar-type mucosa extending up above the GE junction into the esophagus; **c** normal GE junction. (Photos by P. Poitras)

not victims of esophagitis. A balance between aggression factors and esophageal defense probably explains this situation.

Aggression Factors:

- *Quantity of reflux*: Quantification of gastroesophageal reflux, by continuous recording of esophageal pH, reveals a certain relationship between the magnitude of the refluxed acid material into the esophagus and the severity of the esophagitis.
- *Quality of reflux*: Although the degree of acidity is paramount, bile, under experimental conditions, acts as a potentiating agent to acid in inducing esophageal inflammation, so it may contribute to the severity of esophagitis. Hypersecretion of gastric acid could be, in rare cases (such as a gastrinoma that causes a major hypersecretion of gastric acid; see Zollinger–Ellison syndrome in ▶ Chap. 2), a factor promoting mucosal damage to the esophagus.

Defense of the Esophagus:

- *Mucosal resistance* of the esophagus to aggression, for example, by secreting local mucus or bicarbonates, could be a protective factor. The concept of "mucosal barrier" so important in gastric pathophysiology (see ▶ Chap. 2) is however not as well developed in esophageal pathology.
- *Esophageal peristalsis* contributes to the defense of the esophagus by promoting the flow of refluxed gastric material back into the stomach (esophageal clearance). The loss of peristaltic secondary contractions, normally initiated during a backflow, probably contributes to the severe esophagitis seen in patients with esophageal muscle disease due to scleroderma.
- *Saliva,* rich in bicarbonate and with a neutral or alkaline pH, is a factor favoring the neutralization of gastric acid refluxed into the esophagus. However, no clinical situation (e.g., during hyposalivation by Sjogren's disease or after maxillary gland radiotherapy treatment) can confirm its clinical importance in the genesis of esophagitis.

(d) Complications of esophagitis. *Acute bleeding* from an esophagitis lesion is rare. More severely ulcerated areas or the use of anticoagulants may be contributing factors. Chronic occult bleeding from ulcerated esophagitis may explain some cases of iron deficiency anemia.

Stenosis is a narrowing of esophageal lumen due to local inflammation or scar from healed ulceration. It will be suspected in the presence of dysphagia, typically more with solid than with liquid foods, and it will be confirmed by endoscopy or radiology. The treatment of stenosis requires control of esophagitis and acid reflux, usually by proton pump inhibitors (PPIs). In some cases, mechanical dilatation with bougies or balloons will be required.

Barrett's esophagus: The squamous esophageal mucosa, in this condition, is replaced by an intestinal-type columnar mucosa.

The diagnosis is suspected during endoscopy which reveals a reddish, gastric-looking mucosa above the cardia, in esophageal territory. The esophagus may thus appear short (hence the name endo-brachy-esophagus) since the junction of the esophageal and gastric mucosa is higher than the usual endoscopic Z line classically located at 40 cm from the incisor teeth. The differential diagnosis will be made with a hiatal hernia raising the cardia (and thus the Z line) into the thorax above the usual diaphragmatic location. Endoscopic biopsies will confirm histological presence of intestinal metaplasia, in an anatomical segment otherwise identified as the esophagus.

Barrett's esophagus is important since it is considered preneoplastic, precursor to the development of a local adenocarcinoma. The risk of malignant evolution is estimated at 1/200 patients.

Treatment of Barrett's esophagus is disappointing since the suppression of reflux by medication or surgery does not seem to induce regression of the re-epithelialized glandular mucosa of the esophagus. Follow-up of this condition (with endoscopy and biopsies every 3–5 years) is often recommended hoping for early detection of high-grade dysplasia or early adenocarcinoma lesions allowing treatment of an early-stage cancer disease. The benefit of this monitoring strategy is however unclear.

(e) Treatment of peptic esophagitis. Treatment of peptic esophagitis is based on the reduction of acid exposure in the esophagus. This is usually achieved satisfactorily by inhibiting the activity of the proton pump located on gastric parietal cells secreting HCl in the stomach cavity and abnormally refluxed in the esophagus (see ▶ Chap. 2). Healing of inflammation is seen in 80–90% of cases of peptic esophagitis after 4–8 weeks of treatment with proton pump inhibitors (PPIs).

In case of failure of the usual therapy with a PPI administered at least 30 min before breakfast for optimal bioefficacy, various pharmacological strategies are possible: (a) doubling the PPI dose (in a single morning dose or, preferably, in divided doses, 30 min before breakfast and dinner); (b) addition of an H2 blocker (e.g., ranitidine 150–300 mg at bedtime) to optimize nocturnal acid suppression; and (c) addition of prokinetics (e.g., domperidone 10–20 mg before meals and at bedtime) to facilitate gastric emptying and possibly increase the tone of the lower esophageal sphincter can sometimes be tried, although evidence for efficacy is limited. Medication failure is usually not an indication for surgery since the results are often disappointing, except in cases where the control of incapacitating regurgitation is sought.

Discontinuation of PPIs results in clinical and/or endoscopic recurrence in 2/3 of patients, unless a predisposing condition (e.g., obesity, drugs reducing sphincter tone) was present and can be corrected. Continuation of PPI therapy appears to be the optimal solution (at the present time); PPIs are considered as safe drugs for chronic prolonged use (even for years). However, long-term disadvantages to chronic PPI therapy (based on retrospective case-control and cohort studies) are present, such as an increased risk of enteric infections (*Salmonella* RR 3, *Clostridium difficile* RR 2, etc.) or pneumonia (RR 4), as well as a possible risk of hip fracture; malabsorption of iron, calcium, and vitamin B12; or interaction with certain drugs including the antiplatelet agent clopidogrel (interaction on P450 cytochrome) or drugs requiring gastric acid milieu for optimal absorption (e.g., ketoconazole).

Anti-reflux surgery does not offer medical or economic benefits at the present time (aside for patients with predominant regurgitation). The role of novel endoscopic anti-reflux procedures remains to be defined.

1.6.2 Infectious Esophagitis

Infectious esophagitis is usually of viral or mycotic origin, more rarely bacterial.

(a) **Symptoms of infectious esophagitis**. Odynophagia as well as dysphagia or chest pain is found in infectious esophagitis. Heartburn or pyrosis is rare.

(b) **Viral esophagitis.**

- *Herpes simplex 1* can cause esophagitis in immunosuppressed patients, often by chemotherapeutic agents. It can also be seen during a primary infection to herpes in healthy subjects.

  Endoscopy may reveal along the esophagus vesicular lesions similar to herpes skin lesions, or small ulcers surrounded by healthy mucosa. Biopsies will reveal typical intranuclear inclusions on histological analysis, and culture can confirm the diagnosis of the virus (but is rarely done).

  Esophagitis usually resolves spontaneously in 1 to 2 weeks. Acyclovir (400 mg po 5 times/day or IV 5 mg/kg q 8 h for 7–14 days) will accelerate resolution of the infection, as well as foscarnet or other antiviral agents.
- *Cytomegalovirus:* CMV esophagitis is encountered mainly in patients immunosuppressed by HIV. It is often a source of severe esophageal pain.

  Endoscopy may reveal long, penetrating ulcers of esophageal mucosa, and biopsies will show characteristic intracytoplasmic viral inclusions.

  Treatment requires the use of ganciclovir or foscarnet.
- *Human Immunodeficiency Virus*: The HIV has been implicated in some cases of viral-looking esophagitis in HIV-positive patients when it was not possible to identify CMV or herpes. Transient symptoms of esophagitis may accompany the primary HIV infection.

(c) **Mycotic esophagitis.**

- *Candida albicans* is responsible for esophagitis in patients with AIDS or immunosuppressed by undernutrition, systemic diseases, chemotherapy, etc., but also in subjects whose immunity is only partially or locally compromised, such as asthmatics using corticosteroids administered by bronchial inhalation or people treated with antibiotics.

  The infection is often localized more in the proximal esophagus and may include the oral

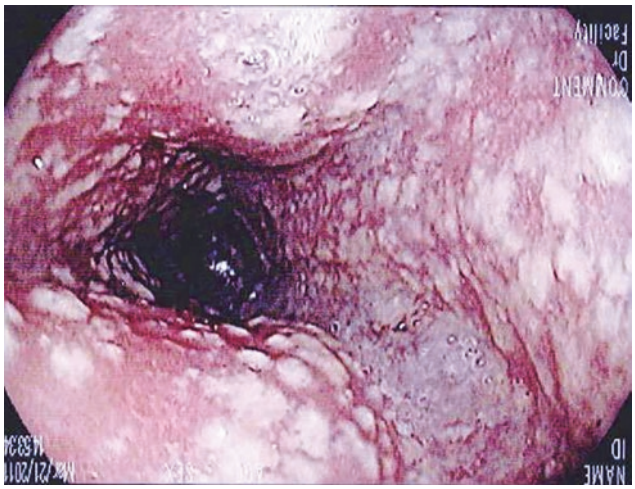


Fig. 1.9 Candida esophagitis: endoscopic view, white and flaky membranes attached to the esophageal mucosa. (Photo by P. Poitras)

cavity. The diagnosis is suspected at endoscopy in front of whitish and flaky membranes comparable to the thrush of oral candidiasis. Biopsies or cytological analyses reveal the presence of *Candida albicans* (Fig. 1.9).

The treatment can be done, in milder cases, with a topical solution of nystatin (500,000 U qid for 7–14 days). In severe cases or in HIV patients, fluconazole (100–200 mg per os id for 2 weeks) will be preferable.

- *Others:* Histoplasmosis, aspergillosis, cryptococcosis, and blastomycosis can occasionally affect the esophagus.

1.6.3 Eosinophilic Esophagitis

(a) **Definition of eosinophilic esophagitis.** Eosinophilic esophagitis refers to an infiltration of the esophagus by eosinophils as detected by microscopic analysis of endoscopic biopsies. It is characterized by massive eosinophilic infiltration (>15 eosinophils/high-powered field) distributed all along the esophagus. Eosinophil infiltration can also be found in gastroesophageal reflux, but it is usually moderate (less than five eosinophils/field) and restricted to the lower esophagus, as well as in eosinophilic gastroenteritis (with or without blood eosinophilia) damaging other digestive organs.

(b) **Clinic.** Eosinophilic esophagitis was first described in the 1970s, but its incidence has been rising steadily since then. Well known in pediatrics where it affects often atopic or allergic children, it is encountered now more and more frequently in the adult (often young men).

Dysphagia for solid foods and/or food impaction are cardinal symptoms of eosinophilic esophagitis.

(c) **Diagnosis of eosinophilic esophagitis.** Endoscopy may reveal, surprisingly considering the symptoms, an almost normal mucosa macroscopically (but microscopically infiltrated by eosinophils) or reveal suggestive changes such as longitudinal furrows, a trachea-like appearance with multiple thin rings distributed along the esophagus, or a diffuse narrowing of the esophagus. Biopsies are necessary and diagnostic.

(d) **Treatment of eosinophilic esophagitis.** The etiopathogenesis of eosinophilic esophagitis is poorly known, as is its treatment. PPIs appear to help in 40% of cases. Corticosteroid therapy is effective, preferably by topical application (fluticasone or budesonide) rather than oral in order to minimize steroid side effects. Exclusion diets (wheat, milk, eggs, peanuts, soy, seafood) have given very encouraging results (75–95% success in children, but diet compliance is difficult), thus supporting an allergic origin of the problem. Hypoallergenic elemental diet is also an option. Dilatation of stenosis by bougie or endoscopic balloon seems hazardous.

(e) **Evolution.** The long-term evolution is poorly known; some patients, unfortunately, seem to deteriorate, while many others seem to have a favorable spontaneous evolution.

1.6.4 Caustic Esophagitis

Ingestion of caustic acid or alkali often results in severe esophageal damage. It is usually accidental in children and linked to suicidal acts in adults. Corrosive household products, such as ammonia, laundry detergents, pipe cleaners, and pool acids, are frequently involved.

(a) **Symptoms.** In the acute initial period, dysphagia, chest pain, and/or sore mouth are often present. Esophagitis is often severe and can lead to perforation with secondary mediastinitis and its lethal complications if not treated. Perforation can also occur in the gastric area or intestine if these organs have been in contact with the corrosive agent.

Long-term symptoms include parietal healing with fibrosis and strictures of the esophageal lumen, which often requires mechanical dilatations, often repeated, or even surgical replacement of the esophagus. In the very long term, there is an increased risk developing squamous cell cancer.

(b) **Investigation.** Alkaline or acidic agents are equally toxic (even if their mechanisms of toxicity are different). Domestic bleach, fortunately, rarely leads to severe complications.

Endoscopy may be used to confirm the extent and severity of the caustic burns in order to assist in prognosis and care planning. Endoscopy (if one decides to do it) should be performed with great delicacy in order to avoid perforating necrotic and fragile organs.

(c) **Treatment of caustic esophagitis.** Careful monitoring of the patient is recommended so that early intervention can be initiated if complications ensue. Preventive maneuvers seem to be ineffective: forcing vomiting may make worse contact of the toxic agent with the esophagus; diluting or neutralizing solutions seem to be ineffective and may also promote harmful vomiting. Reducing the inflammatory reaction with systemic steroids (e.g., methylprednisolone 20–40 mg q 6 h IV) remains a debated therapeutic issue.

Esophageal and/or gastrointestinal perforations will require emergency, and often morbid, surgery. Depending on the condition of the tissue, drainage with suture of the perforation or more or less extensive removal of the affected organs will be required.

1.6.5 Drug-Induced Esophagitis

(a) **Pill esophagitis.** This is due to a medication pill "trapped" or stagnating in an esophageal segment that induces a local corrosive damage. It occurs most often in the upper esophagus, and it is characterized by localized chest pain, often with odynophagia and dysphagia, starting few hours after ingestion of the drug.

Endoscopy usually shows a well-circumscribed ulcer (or two "kissing" ulcers) surrounded by healthy mucosa, reflecting the "burn" by the stagnant pill. Aggravating factors are as follows:

- Some medications seem more irritating or corrosive; this includes potassium supplements, tetracycline and its derivatives, aspirin, and NSAIDs.
- Certain conditions are known to favor poor transport of the pill down the esophagus and "stagnation of the pill" with secondary corrosive damage. Any condition associated with abnormal esophageal transit is a risk factor.
- Most of the time, taking the medication in a lying down position, and with insufficient fluid bolus to promote trans-esophageal passage, is involved in pill-induced ulceration.

The ulcer usually heals spontaneously in a few days. Antacids, possibly with local anesthetics ("Pink Lady": 15–30 mL of antacid and 15–30 mL of viscous Xylocaine), can help relieve pain.

(b) **Bisphosphonates.** Frequently used bisphosphonates for the treatment of osteoporosis are associated with chemical esophagitis often more diffuse than those described above. The esophagitis mechanism remains unclear and implies, among other things, the reflux in the esophagus of the drug once dissolved in the stomach. To counteract this complication occurring mainly with alendronate, it is suggested to take the drug in a standing position (and stay in this position for at least 1 hr) and to ingest a large quantity of fluids to promote passage of the drug through the intestine and avoid its reflux in the esophagus or its stagnation in the stomach.

1.6.6 Radiation Esophagitis

Radiotherapy for the treatment of thoracic neoplasia (lymphoma, cancer of the lung/esophagus, etc.) can affect the esophagus. In the acute phase, during the treatment period, mucositis causing pain, odynophagia, and dysphagia can occur. Treatment with local analgesics and antacids helps relieve these symptoms. In some cases, dysphagia can be severe enough to compromise food ingestion, and temporary artificial tube feeding may be required. Delayed complications such as stenosis may require mechanical dilatations.

1.7 Tumor Disorders

1.7.1 Types of Esophageal Neoplasia

Squamous cell (epidermoid) carcinoma and adenocarcinoma are the two main malignant neoplasms that affect the esophagus (Fig. 1.10).

(a) **Squamous cell neoplasia** develops from the esophageal squamous epithelium. It is most commonly located to the middle and upper thirds of the organ. It preferentially affects males over 50. Its prevalence is high in some parts of the world including China, the northeast of France, or in the black population of North America. Predisposing factors involving chronic mucosal damage are well recognized: heavy alcohol ingestion, smoking, ingestion of hot tea, caustic esophagitis, etc. Previously established as the most important type of esophageal cancers, the frequency of the epidermoid carcinoma seems to have regressed in the recent decades (perhaps in response to a decreased exposition to predisposing factors).

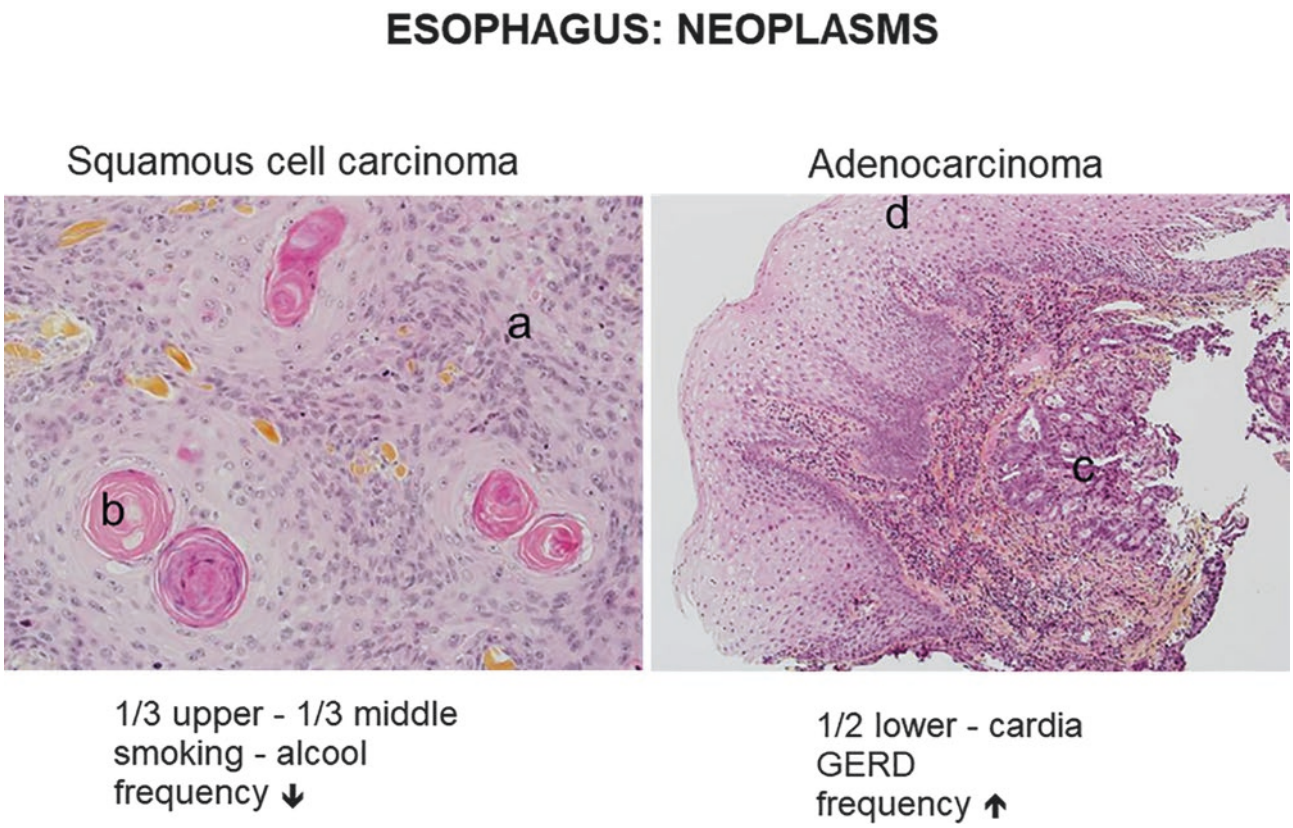


Fig. 1.10 Histology and characteristics of esophageal neoplasms: (1) The picture on the left shows a well-differentiated squamous cell carcinoma {(a); compared with the normal squamous cells (present on d)} with formation of keratin pearls (b); squamous cell cancer is most often located in the upper or middle esophagus. (2) the right picture shows adenocarcinoma cells (c) infiltrating the mucosa under a normal squamous epithelium (d). (Photos by G.Soucy)

(b) **Adenocarcinoma** was classically considered as a rare esophageal tumor, but it is now the most common esophageal cancer in the Western world. Adenocarcinoma develops from glandular tissue; it is therefore more readily seen during the re-epithelialization of the normal squamous esophageal epithelium by the glandular lining seen in Barrett's esophagitis. This type of cancer mainly affects the distal esophagus. It develops more readily in male subjects, of Caucasian origin, aged over 50 years and suffering from chronic gastroesophageal reflux.

1.7.2 Clinical

Esophageal neoplasia grows intra-luminally to obstruct the esophageal channel, thus causing progressive dysphagia, initially presenting with solid food and later on affecting also liquids. Weight loss, due to a deficient caloric intake, is common. Anemia caused by chronic bleeding from the ulcerated tumor may occur.

1.7.3 Diagnosis of Esophageal Cancer

X-rays of the esophagus (Fig. 1.11) can reveal the luminal obstruction by the tumor (although not as sensitive as endoscopy), but will not be able to differentiate the nature of the cancer.

Endoscopy is the best test to diagnose esophageal cancer; biopsies and/or cytology are required to confirm the diagnosis.

The progression of the disease is usually toward the adjacent lymph nodes and thus to the cervical, thoracic, or abdominal regions depending on the location of the primary tumor. Extent of the disease will be evaluated by axial tomography (CT scan), positron emission tomography (PET), or echo-endoscopy.

1.7.4 Treatment of Esophageal Cancer

Unfortunately, no curative treatment can be offered for esophageal cancer in a large proportion of patients. Survival after treatment is 30 to 40% at 5 years.

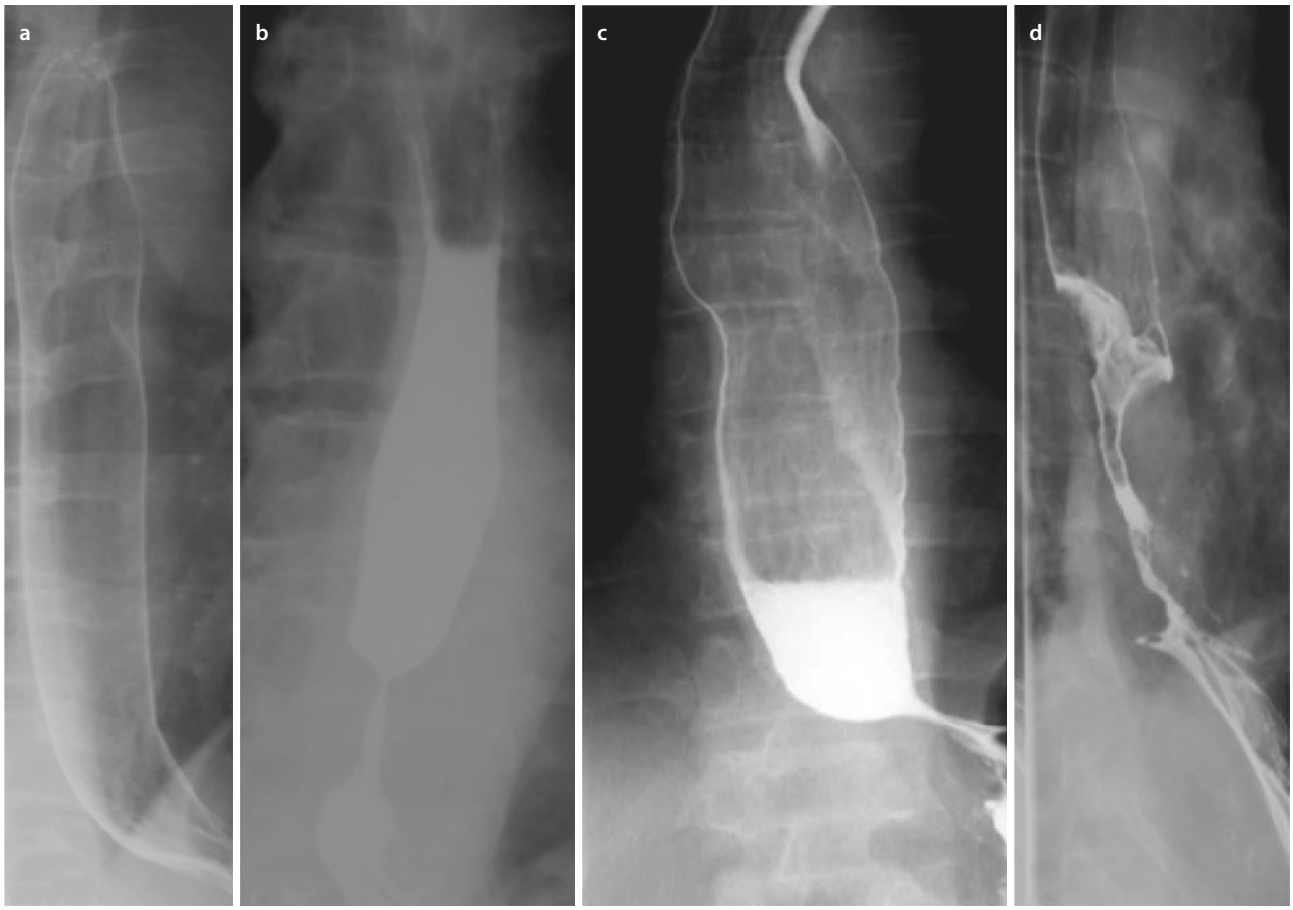


Fig. 1.11 **a** Radiography of a normal esophageal appearing as a cylinder about 25 cm long and 2.5 cm wide. **b** Peptic stricture is seen here in the distal esophagus with a typical smooth and symmetrical narrowing of the lumen. **c** Achalasia (1) the lumen of the esophagus is distended; (2) the cardia is slim and slender, appearing as a bird's beak. **d** Neoplasia (epidermoid here): the esophagus is narrowed by a proliferative mass protruding into the lumen of the organ and mimicking an "apple core." (Photos by R. Déry)

The overall therapeutic strategy is comparable for the epidermoid carcinoma or adenocarcinoma.

Faced with a disease that is believed to be limited and curable, the therapeutic procedures include radiation therapy, usually combined with chemotherapy (with cisplatin) frequently followed by excision surgery. Esophagectomy, with different reconstruction techniques (esophagogastric anastomosis with a pull-up of the stomach, interposition of a colonic or jejunal segment, etc.), is a complex and unfortunately often morbid procedure to be carried out in expert centers.

In the case of an advanced, unresectable disease, the strategies of palliation include chemotherapy ± radiotherapy to decrease the volume of the obstructive mass and secondary dysphagia, or stent placement, using endoscopic or radiological guidance, that re-establishes the esophageal lumen to allow an oral feeding.

1.8 Function Disorders

1.8.1 Gastroesophageal Reflux Disease (GERD)

(a) Definition. GERD is defined as the reflux of gastric material into the esophagus, resulting in troublesome symptoms.

(b) Symptoms of GERD. Regurgitation of food with a feeling of retrosternal discomfort or with bitter taste in the mouth, heartburn (retrosternal burning sensation due to the acidic pH of the refluxed material), and pyrosis (heartburn radiating upward, progressing from the bottom to the top of the esophagus) are indicative of GERD (with or without esophagitis). Odynophagia (retrosternal pain at the passage of food) or dysphagia (blockage sensation

at the passage of food) is suggestive of esophagitis (a complication of GERD).

GERD symptoms are common, being reported in 30 to 50% of the population, and their frequency appears to be increasing with age. GERD is considered as an illness when symptoms occur at least weekly and have an impact on the quality of life.

(c) Pathophysiology of GERD. Gastric content, made of ingested food and secretions produced by the stomach (HCl, pepsin) or refluxed from the intestine (e.g., bile), will be, under normal conditions, pushed to the duodenum by propulsive contractions of the stomach. The lower esophageal sphincter (LES) works as a unidirectional valve that relaxes to allow the swallowed food to enter the stomach cavity and then contracts back to its basal state in order to close the upper end of the stomach and thereby prevent reflux of gastric contents back up into the esophagus. The reflux from the stomach into the esophagus thus occurs when the gate capacity of the valve is insufficient to impede the passage of gastric material.

The LES measures 2 cm in length and is located at the diaphragm, mainly on the abdominal side of the thoracoabdominal junction. The sphincter can be seen as a two-component structure. The "internal" sphincter corresponds to an area made of specialized smooth muscles of the lower esophagus, which, although not identified histologically, generate a short high-pressure zone between the positive intra-abdominal and the negative intrathoracic pressures. The "external" sphincter is made of various anatomical factors of the lower esophagus that bolster this physiological high-pressure zone: the diaphragm (making a constrictive ring around the esophagus), the right crus of the diaphragm as well as the phreno-esophageal membrane (surrounding and fixing the lower esophagus to the thoracoabdominal junction), the size of the angle of His (allowing compression of the distal esophagus by the gastric fundus), and the intra-abdominal localization of the sphincter.

Circumstances favoring reflux are shown in Fig. 1.12 and discussed here:

- *Increased intragastric pressure* exceeding the lower esophageal sphincter retention capacity. The pressure is naturally positive in the abdomen and negative in the thorax. Pregnancy, abdominal obesity, and tight waist clothing are factors that increase intra-abdominal and, consequently, intragastric pressures. Gastric emptying failure by either distal obstruction or gastroparesis will result in stagnation of gastric contents and promote reflux into the esophagus. The recumbent position will shift the gastric content into the upper stomach, facilitating its reflux upward.

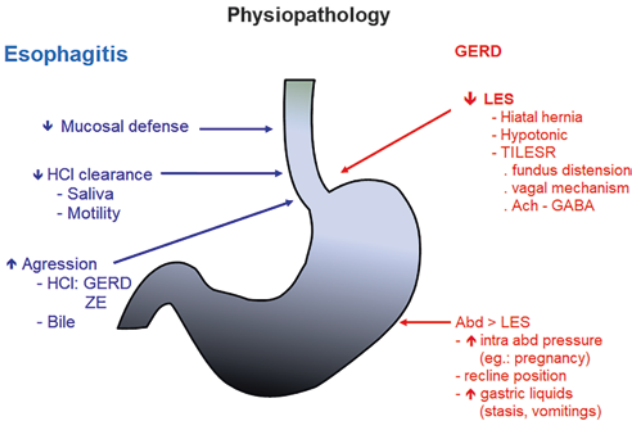


Fig. 1.12 Pathophysiology of gastroesophageal reflux disease (right of the picture in red characters) and of its most frequent complication, esophagitis (left of picture in blue)

- *Decreased barrier pressure*. The LES normally has a basal tone of 20–25 mmHg to impede reflux of gastric contents into the esophagus. Sphincter hypotonia (pressure below 10 mmHg) is a logical and obvious cause for reflux; it is found, however, in only a small percentage of cases. Smooth muscle esophageal diseases, such as scleroderma, can reduce the pressure of the lower esophageal sphincter (even to 0 mmHg) and result in marked gastroesophageal reflux. Some drugs (e.g., theophylline, calcium channel blockers, nicotine) as well as certain foods (e.g., chocolate, mint) can lower the pressure of the sphincter and may promote reflux.

  Hiatal hernia (intrathoracic displacement of the esophageal-gastric junction) seems to be a condition that alters the tension resistance of the LES, probably by compromising the anatomic factors that produce the "external sphincter."
- *Normotensive but incompetent barrier*. The lower esophageal sphincter normally relaxes during swallowing to let the food bolus migrate from the esophagus to the stomach. Transient and inappropriate relaxation, i.e., without being induced by swallowing, may occur. This phenomenon usually occurs in response to fundic distension, via a vagal reflex involving VIP and the GABA (gamma-aminobutyric acid). Studies have confirmed that the gastric content could use these transient and inappropriate relaxations to reflux into the esophagus. TILSR (transient inappropriate lower sphincter relaxation) which occurs 2–3 times/hr in normal subjects are more frequent in GERD (8–12/hr); increased number of TILSRs is now considered the main pathophysiological mechanism for GERD.
- The *acid pocket* is a concept born in 2001. It results from the simple observation that acid reflux occurs often after the meal, a paradoxical situation since the

gastric pH is then high due to the food present in the stomach. Studies have shown that acid secretions accumulate on top of food to form an acidic pocket in the upper part of the stomach. If the acid pouch is located in a hiatus hernia, chances for gastroesophageal reflux are clearly increased. PPIs raise the pH of the pocket secretions (from pH 1 to pH 4). Alginate foam (Gaviscon®, discussed below) works by forming a raft on this pocket (rather than mixing with the food chyme as other antacids) to "locally" neutralize the acid in the pocket.

(d) Complications of GERD. Esophageal as well as extra-esophageal complications are seen:

Esophageal complications:

- Peptic esophagitis is the main complication of gastroesophageal reflux, as previously discussed in the section of inflammatory diseases (► Sect. 1.6.1); stenosis or neoplasia is then possible.
- Nonerosive reflux disease (NERD) refers to GERD without esophagitis as established at endoscopy (endoscopy is positive in only 30% of patients suspected of GERD; among the patients with negative endoscopy, 50% have reflux documented on pH studies and are therefore identified as NERD). Since these patients seldom develop peptic complications (including esophagitis), even in the long run, NERD requires only a symptomatic treatment.

Extra-esophageal complications:

- ENT: Hoarse voice, pharyngeal pain, and cough may be symptoms of posterior laryngitis believed to be due to a reflux of irritating gastric material in some cases.
- Lungs: Asthma may possibly be exacerbated by gastroesophageal reflux. The mechanisms proposed are either intrabronchial aspiration of material refluxed into the hypopharynx or bronchiolar spasms triggered by a vagal reflex possibly induced by distension or irritation of the esophagus secondary to reflux. In rare cases, aspiration of the refluxed material may cause aspiration pneumonia.
- Chest pain: Retrosternal pain, sometimes even mimicking that of angina pectoris (including irradiation to the left arm or jaw), may be due to reflux. It is assumed that the pain is due to an esophageal spasm triggered by reflux. The possibility of coronary spasms in response to a vagal reflex triggered by GERD has also been proposed in some patients.

In presence of a clearcut gastroesophageal reflux, a good response of these extra-esophageal symptoms to treatment with acid suppression can be expected. When a reflux is suspected, a therapeutic test (using a twice daily PPI regimen for 4–8 weeks) could be useful in establishing the contribution of gastroesophageal reflux in the genesis of these extra-esophageal symptoms. The very limited effectiveness of PPIs (success of less than 40% and comparable to placebo in several studies) to relieve coughs, hoarse voice, or asthma raises doubts about the importance of GERD in these symptoms in a majority of patients.

(e) Investigation of GERD. Gastroesophageal reflux disease is a very common condition, but fortunately in most patients the diagnosis can be made based on clinical presentation, and further investigations are not required. Investigations need to be pursued in patients who do not respond appropriately to treatment, or in those with features suggesting complications (e.g., dysphagia, odynophagia, weight loss, anemia). Although of unproven value, many experts suggest screening endoscopy to rule out Barrett's esophagus be performed in certain higher risk patients (e.g., male Caucasians older than 50 years of age with chronic reflux symptoms for more than 5 years).

Modes of Investigation:

- *Endoscopy* is the most useful examination to confirm the diagnosis of GERD (if esophagitis is present in the esophagus), to guide management of esophagitis (in order to avoid long-term complications), to identify subjects at risk for adenocarcinoma (Barrett's esophagus), and to reassure the reflux patient without esophagitis (since these individuals usually continue to live without ever developing inflammation and its complications). In patients with heartburns, endoscopy reveals esophagitis in 30% of cases; others suffer from either NERD (nonerosive reflux disease) or esophageal hypersensitivity or functional heartburn.
- *Recording of esophageal reflux* can be done when the diagnosis is still unclear following endoscopy (e.g., normal endoscopy) or when other conditions justify to confirm the diagnosis (e.g., poor response to antireflux treatment). GERD can be confirmed by various methods that measure reflux of gastric material into the esophagus. Esophageal pHmetry, the most commonly used of these methods, continuously measures the esophageal pH using an electrode either installed on a catheter that is passed transnasally into the lower esophagus and connected to a device worn on a belt or shoulder strap, or clipped to the distal esophageal mucosa endoscopically. Esophageal pH is then monitored over a period of usually 24 hours or more. Detecting episodes of esophageal pH < 4 indicates reflux of gastric acid into the esophagus.

Other methods, such as using the concentration of bile in the esophagus or esophageal impedance, can also be used in select cases, particularly if non-acidic reflux is suspected.

- *Radiography* of the esophagus (or barium meal) can demonstrate reflux of the barium substance from the stomach into the esophagus. It can occur spontaneously (mainly in the recumbent position) or can be induced by different maneuvers increasing abdominal pressure (raising legs, etc.). However, the barium meal is neither specific nor sensitive for the diagnosis of GERD or esophagitis, and it rarely helps in the management of these diseases. It can be of some interest, however, in the study of dysphagia, as discussed subsequently.

(f) Treatment of GERD.

(a) ***Correction of risk factors and life style habits*** that promote GERD is important. Correcting an excessive intra-abdominal pressure (losing weight if obese, abandoning tight clothing, etc.), avoiding reclining positions favoring reflux (no lying down after meals, elevating the head of the bed, etc.), and minimizing LES hypotension (avoiding fatty foods, chocolate, peppermint, alcohol, tobacco, or drugs such as calcium channel blockers, nitrates, theophylline) can provide significant improvement in certain cases. Weight gain is often the trigger for GERD, and weight loss, although often difficult to achieve, is likely the most useful "lifestyle" intervention.

(b) ***Suppression of refluxed acid*** is currently the most effective pharmacological method to stop symptoms of reflux (and secondary esophagitis). Different approaches are used.

- Antacids: These drugs typically contain buffering agents such as bicarbonate, magnesium, and aluminum, to neutralize gastric HCl. They are available in liquid form (Maalox, Riopan, Amphogel, Gaviscon, 15–30 ml q 1–2 h prn) or tablets (Tums, Rolaids, etc.). Symptom relief is usually obtained rapidly within a few minutes, but it is short-lived. Available over the counter in pharmacies, these agents are useful for self-treatment of mild or occasional symptoms. High doses of antacids may cause side effects (especially diarrhea due to magnesium).
- H2 blockers: These (also called H2 receptor antagonists or H2RA)) suppress gastric H+ secretion by the parietal cells by blocking the histamine stimulation. More powerful than antacids, they revolutionized the treatment of acid-related diseases when introduced in 1977. Their ability to suppress acid is however limited by, among other things, tachyphylaxis due to downregulation of histamine receptors after a few days of pharmacological treatment. H2 blockers may be useful to control mild to moderate reflux symptoms (ranitidine 150–300 mg bid or famotidine 20–40 mg bid). They can also be used "on demand" for the relief of occasional reflux or peptic symptoms (in many countries, H2RA can be conveniently purchased over the counter without a prescription). Occasional use of these drugs will prevent tachyphylaxis, but because they require gut absorption and systemic circulation, it takes 30 to 60 minutes before they begin to inhibit acid production. Accordingly, they are not well suited for the prompt relief of symptoms when taken on a prn basis. In case of nocturnal symptoms uncontrolled by PPI, an H2RA taken at bedtime can help in some people.
- Proton pump inhibitors (PPIs): These are potent inhibitors of gastric acid secretion (see ▶ Chap. 2). Since their introduction in the 1980s, they have revolutionized the treatment of GERD and currently constitute the most effective treatment for reflux esophagitis. The usual doses of dexlansoprazole (30 mg die), esomeprazole (20 mg), lansoprazole (30 mg), omeprazole (20 mg), pantoprazole (30 mg), and rabeprazole (20 mg), taken in the morning at least 30 minutes before breakfast, will control the symptoms of reflux and heal reflux esophagitis in 80–90% of patients. Some individuals will benefit from a double dose (administered once in the morning or preferably in divided doses 30 minutes before breakfast and dinner). Recurrence of symptoms (or of esophagitis) when treatment is discontinued occurs in 2/3 of patients, and often a continuous therapy may be required. Maintenance therapy using PPI has been used for 20 years without significant side effects. It currently appears as the treatment of choice for most patients.

Reflux without esophagitis (NERD), given the absence of complications, can be treated symptomatically and on demand. On the other hand, reflux esophagitis should, even in the absence of symptoms, be managed, most of the time, by a continuous maintenance treatment to minimize the chance of peptic or neoplastic complications.

(c) ***Reducing reflux material*** appears as a logical therapeutic strategy. It could, in theory, be obtained by increasing the tone of the LES, by decreasing the numbers of TILSR (transient inappropriate lower sphincter relaxation), or by promoting gastric emptying of gastric contents to the intestine in order to

decrease the amount of material likely to backflow into the esophagus.
- However, few drugs are available that decrease the GE reflux. Domperidone (10 mg before meals and at bedtime), a gastrokinetic agent that increases gastric emptying and LES tone by acting as an antidopaminergic, may be useful in a small subset of patients, especially to decrease regurgitations poorly controlled by acid reduction agents.
- Reducing TILSRs appears as a logical pharmacological target, given their important role in the pathophysiology of GERD. Agonists of GABA (gamma-aminobutyric acid) are considered as therapeutic candidates; baclofen is sometimes used.
- Surgery can be used to decrease reflux (see below).

(d) ***Anti-reflux surgery*** was, before the demonstrated efficacy of PPIs, the only valid treatment for GERD. The Nissen fundoplication not only corrects the hiatal hernia but also wraps around the cardia a portion of the gastric fundus to restore the "external" sphincter (discussed in the anatomy ▶ Sect. 1.1.1 of this chapter). Fundoplication is usually performed laparoscopically; it may involve different technical variations such as the classical Nissen procedure or the partial Toupet fundoplication. Fundoplication decreases reflux by raising the basal tension of the LES while reducing TILSRs by incompletely understood mechanisms. Aside from the usual morbidity related to abdominal surgery, fundoplication can cause side effects in about 10% of subjects operated, including the following:
- Dysphagia: This may require dilatations to loosen up a too tight fundoplication.
- Gasbloat: in the majority of the subjects operated on, the new fundic valve will create a difficulty with vomiting or belching. In some patients, the inability to belch will cause abdominal discomfort by accumulation of gastric air.
- Dyspepsia: Functional dyspepsia is often due to a poor accommodation of the gastric fundus (see ▶ Chap. 2). This motor dysfunction can obviously be enhanced by a fundoplication procedure compromising fundic anatomy and function. The indication for fundoplication must therefore be considered very carefully in a patient with GERD and coexisting symptoms of functional dyspepsia.

In the case of "medical dependence" (i.e., patients who cannot stop their PPIs without a relapse of symptoms), surgical fundoplication does not seem, in the medium or long term, to provide results superior to those obtained by chronic PPI treatment. In fact, 5–10 years after fundoplication was performed to avoid the need of chronic PPI therapy, many patients will still require anti-reflux pharmacotherapy. In the setting of "medical resistance" (i.e., patients with persistent symptoms despite optimal twice daily PPI therapy), fundoplication often leads to disappointing results. Its place in GERD therapy today seems limited to patients where troublesome regurgitations resistant to medical treatment is the major issue, or for GERD patients who, for various reasons, prefer this intervention over chromic PPI therapy.

New techniques (e.g., magnetic sphincter augmentation device implanted at the cardia) could offer in a near future valid alternatives for GERD patients who desire to discontinue medical therapy, or are not compliant or are resistant to medical therapy.

(e) ***Endoscopic therapy***: A variety of endoscopic techniques aimed at correcting the sphincter barrier defect have been developed. These include intragastric plication of the fundus, longitudinal plication of the cardia, infiltration in the cardia with synthetic material(s), electro-fulguration of lower esophagus by radiofrequency (Stretta procedure), etc. The benefit of these endoscopic maneuvers over the current pharmacological or surgical treatments remains to be demonstrated.

(g) In Pediatrics:
- Gastroesophageal reflux disease (postprandial non-bilious food regurgitation) is physiological in infants up to 18 months of age.
- Only the occurrence of complications (peptic esophagitis, growth retardation, or respiratory problems secondary to the reflux) should trigger additional investigations and drug treatment.
- GE reflux is not the cause of the unexplained crying ("colic") in infants.
- Peptic esophagitis is rare in children.
- Children at risks for peptic esophagitis include those with chronic respiratory pathology (like cystic fibrosis), previous surgery for esophageal atresia, or neurological disability (encephalopathy of neonatal ischemic origin, of metabolic or genetic origin).
- There is no correlation between symptoms and the endoscopic aspect of the esophagus.
- Barium meal is not indicated to establish the diagnosis of GERD in children.
- The barium meal can document anatomical abnormalities that could generate regurgitations/vomiting in infants such as an intestinal malrotation, an obstacle to gastric evacuation (pyloric stenosis also seen in ultrasound), and a congenital stenosis of the esophagus.

1

1.8.2 Oropharyngeal (Transfer) Dysmotility

(a) **Symptoms**. Proximal dysphagia perceived in the oropharyngeal or cervical region is the predominant symptom. It may be associated with symptoms of nasal regurgitation, coughing, choking, and aspiration pneumonia, related to misdirection of the swallowed food or fluid into the nasopharynx, larynx, or trachea.

(b) **Pathophysiology.**

Damage to nerves (cranial nerves IX, X, and especially XII) controlling the transfer motor function:

- Brain stem nuclei damage by cerebral vascular stoke, tumors, Parkinson's, etc.
- Peripheral nerve damage by demyelinating disease (multiple sclerosis) or others (sclerosis, amyotrophic lateral sclerosis, etc.).

Damage to the striated muscles involved in transferring the food bolus from the oropharynx to the esophagus by diseases such as myasthenia gravis (dysphagia often worsening with fatigue), oculopharyngeal dystrophy (hereditary dystrophy common in Quebec and affecting pharyngeal and palpebral muscles), etc.

(c) **Investigation.** Radiology and endoscopy are both useful to evaluate oropharyngeal dysmotility.

Radiological assessment of the swallowing process, using fluoroscopy during barium ingestion, will be crucial to identify and characterize the motor disorder. Its diagnostic yield, however, is dependent upon the expertise of the examiner.

The radiological assessment will also be used to identify Zenker's diverticulum or an obstructive lesion of the hypopharynx or proximal esophagus that could give clinical symptoms of high dysphagia.

Endoscopy may be required to rule out a luminal obstructing lesion.

However, it must be remembered that the upper localization of dysphagia reported by the patient can be misleading since a middle or distal lesion may cause a sensation of upper dysphagia (on the other hand, the lower localization of dysphagia is more reliable to identify the obstructing site since a sensation of a blockade in the lower esophagus is never due to a proximal lesion).

(d) **Treatment of oropharyngeal dysmotility.** Various options are available:

- Treatment of the causal pathology if possible (e.g., myasthenia gravis).
- Diet adjustment and safe swallowing education by specialists (occupational therapist, dieticians, speech language pathologists, etc.).
- Surgical (or endoscopic) myotomy (or pharmacological relaxation by infiltration of botulinum toxin) of the UES can be done in some very selected cases to reduce sphincter resistance and then facilitate the passage of the bolus from the oropharynx toward the esophageal lumen.

1.8.3 Esophageal (Transport) Dysmotility

Transport dysmotility along the esophagus, which usually produces dysphagia or thoracic pain, can be due to hypo- or hypercontractility disorders of the smooth muscles of the esophagus or of the lower esophageal sphincter (LES). These disorders are recognized mainly by measuring contractile pressure waves of the esophagus via intraluminal manometry (see ◘ Fig. 1.13).

A. Hypomotility disorders. Absent contractility (as referred to in Chicago Classification) is most often due to *scleroderma,* a disease characterized by a collagenous fibrosing infiltration of skin tissues; it can also affect the digestive tract, with the esophagus being the most common region affected. It is characterized manometrically by esophageal contractile waves markedly decreased in amplitude (even not existing) and a lower esophageal sphincter with a very weak tone (even absent).

Esophageal hypomotility can also be seen with neuromuscular disorders affecting the GI tract (rare conditions often leading to pseudo-obstruction syndrome; see ▶ Chap. 3) or, sometimes, be due to an inflammatory response to esophagitis (peptic, caustic, etc.).

Symptoms Patients with scleroderma typically develop severe GERD symptoms (heartburn, regurgitation) because of their defective LES pressure barrier and poor esophageal clearance secondary to weak or absent peristalsis. Dysphagia is also common, and, although partly related to the absence of propulsive waves, is usually due to the development of reflux esophagitis, often complicated by peptic stricture formation. Overt or occult bleeding from esophageal ulceration may also occur.

Investigation ***Manometry*** of the esophagus is the determinant examination to identify the diagnosis. Hypomotility profile is mostly due to scleroderma, but it can be seen in rare infiltrative disorders such as amyloidosis or in chronic reflux esophagitis.

Endoscopy is essential to assess the severity of esophagitis.

Treatment GERD can have severe consequences in patients with scleroderma esophagus (due to the absence of LES to block reflux and of esophageal contractions for clearance of refluxed acid) and requires an aggressive treatment. Suppression of acid reflux is essential. PPIs can be given at high doses for optimal acid suppression.

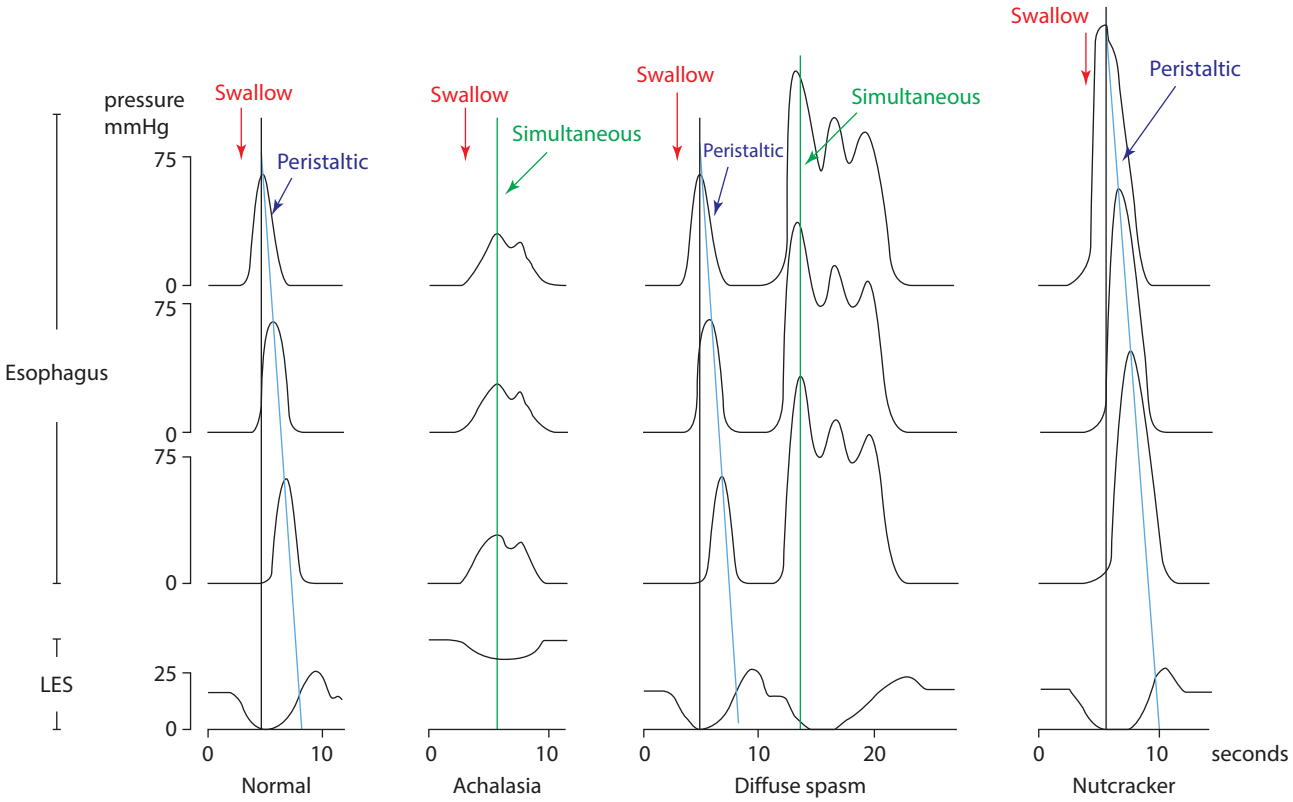


Fig. 1.13 Esophageal manometry contractions recorded in various conditions of hypermotility. (Modified from Smout 1992)

They can be coupled with prokinetics to promote gastric emptying (potentially affected by gastric hypomotility secondary to scleroderma infiltration of gastric smooth muscles).

B. Hypermotility disorders. Hypermotility of the esophagus is described under the following categories:

1. ***Achalasia of the esophagus*** is a disease of the intrinsic innervation of the esophagus (infiltration of lymphocytes into the myenteric plexus leading to destruction of neurons controlling LES relaxation and esophageal peristalsis) and is classically characterized by the following contractile anomalies:
   - Inadequate relaxation of the LES at the passage of the food bolus.
   - Increased basal tone of the LES.
   - Absence of peristaltic waves within the esophageal body.
   - Possibility of tertiary spastic waves of the esophageal body.

Symptoms Lower (transport) dysphagia, often for both liquid and solid foods, and regurgitations of undigested stagnant food from the esophagus are cardinal symptoms of achalasia; chest pain is also reported by some patients. Complications such as weight loss and aspiration pneumonia (if nocturnal regurgitation is present) can occur.

Causes of achalasia Idiopathic primary achalasia is the most common.

Achalasia can also be secondary to the following:

- Chagas disease due to the *Trypanosoma cruzi* parasite, present mainly in South and Central America and capable of attacking esophageal innervation. This parasite also frequently affects the innervation of the heart resulting in fatal heart failure.
- A tumor of the gastric cardia infiltrating the enteric nervous system in the region of the LES.
- A paraneoplastic manifestation (most often from lung cancer) affecting the motor function of the esophagus as well as the stomach (probably by antibodies against the enteric nervous system).

Investigation of achalasia A *barium swallow* (see Fig. 1.11) can reveal a characteristic image of (1) esophageal dilation (mega esophagus), with possibly a fluid level and food retention, and (2) narrowing of the esophagogastric junction mimicking the beak of a bird. Plain films of the lungs and abdomen may suggest the diagnosis of achalasia by showing an enlargement of the mediastinum due to the esophagus filled with fluid, as well as an absence of the gastric air bubble.

During *endoscopy*, the LES is usually closed, and some difficulty or resistance can be felt in passing the endoscope through the cardia. Endoscopy is essential to eliminate a tumoral infiltration of the cardia.

Manometry is the definitive test to diagnose achalasia of the esophagus.

An example of conventional (or traditional) manometry is presented in Fig. 1.13 (characteristic motor abnormalities were described above). Since the year 2000, high-resolution manometry (HRM) has been used to more precisely study the motor activity of the esophagus. Done with a nasogastric manometry catheter carrying several electronic pressure sensors distributed every centimeter (at least) along the esophagus and combined with sophisticated computer analysis of the obtained data, HRM allows a graphic representation of the esophageal transit and pressures (Fig. 1.14), as well as an objective quantitative analysis of the motor function (with newly identified computerized parameters such as IRP (integrated relaxation pressure of the LES) or DIC (distal integrated contraction of the peristaltic distal wave). Observations and concepts obtained by HRM are grouped together in the "Chicago Classification," a tool in perpetual development (a bit like the DSM in psychiatry). Achalasia is now divided into three manometric types (I, II, III; Fig. 1.15), suggesting the presence of differing pathophysiology (and/or stage) of disease and, possibly, the need for targeted adapted therapies.

Treatment of achalasia. In order to alleviate dysphagia, the treatment aims to reduce the obstacle created by the nonrelaxing sphincter. The therapeutic possibilities are as follows:

- Surgical myotomy of the LES, usually done laparoscopically, and often with reconstruction by fundoplication to avoid GE reflux secondary to the sphincter weakness induced by myotomy.
- Myotomy can also be performed endoscopically (POEM: per oral endoscopic myotomy).

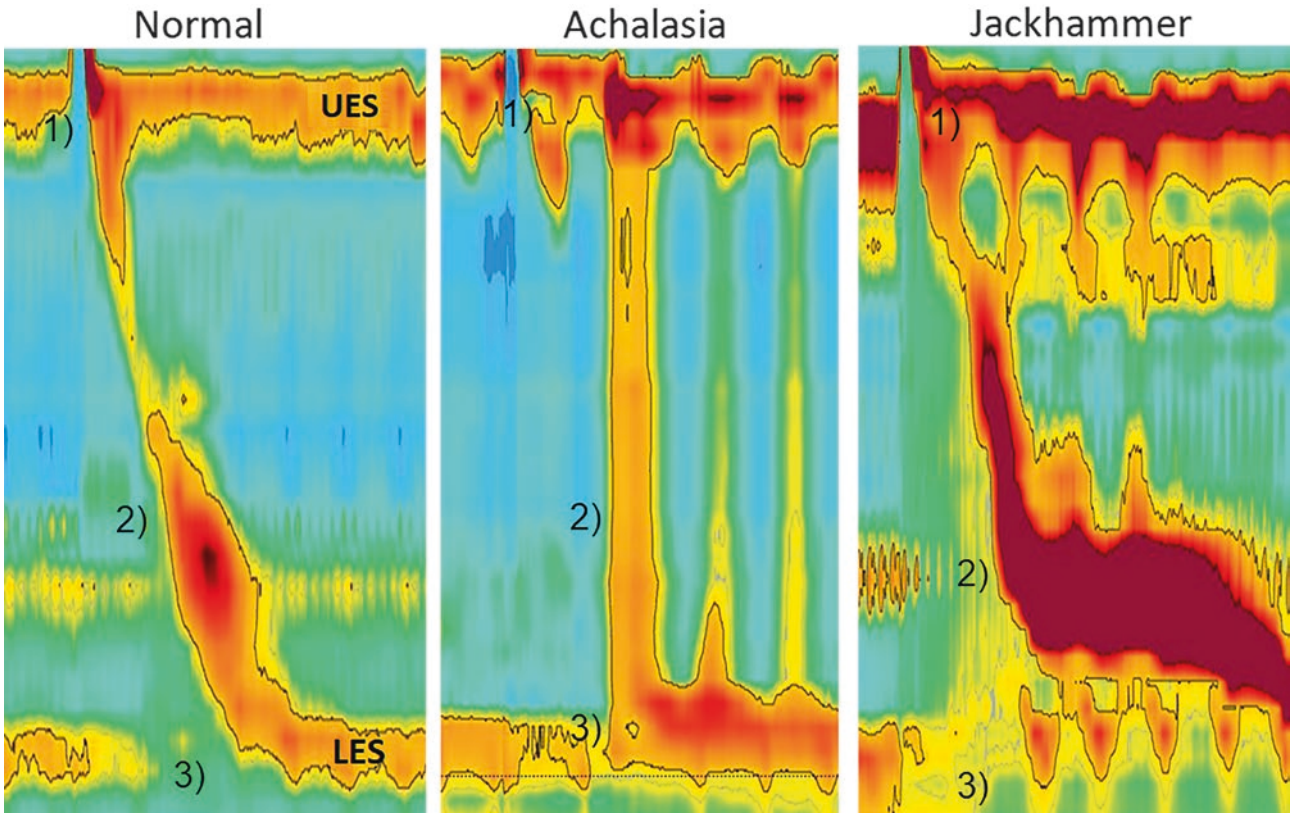


Fig. 1.14 High-resolution esophageal manometry: examples of tracings with colors that identify a pressure gradient (green < yellow < red). Left tracing: normal. (1) During swallowing, the upper esophageal sphincter (UES) relaxes; (2) a peristaltic contraction develops in the esophageal body (3) the lower esophageal sphincter (LES) relaxes allowing the bolus to pass into the stomach. - Central tracing: achalasia (type II). (1) After the relaxation of the UES, (2) no propulsive wave (here we have a diffuse luminal pressurization) occurs in the esophagus; (3) the LES does not relax. - Right tracing: jackhammer esophagus. (1) normal relaxation of the UES; (2) a propulsive esophageal wave is present, but it is of very large amplitude and prolonged duration in the distal region; (3) the LES relaxes normally. (Tracings from M. Bouin)

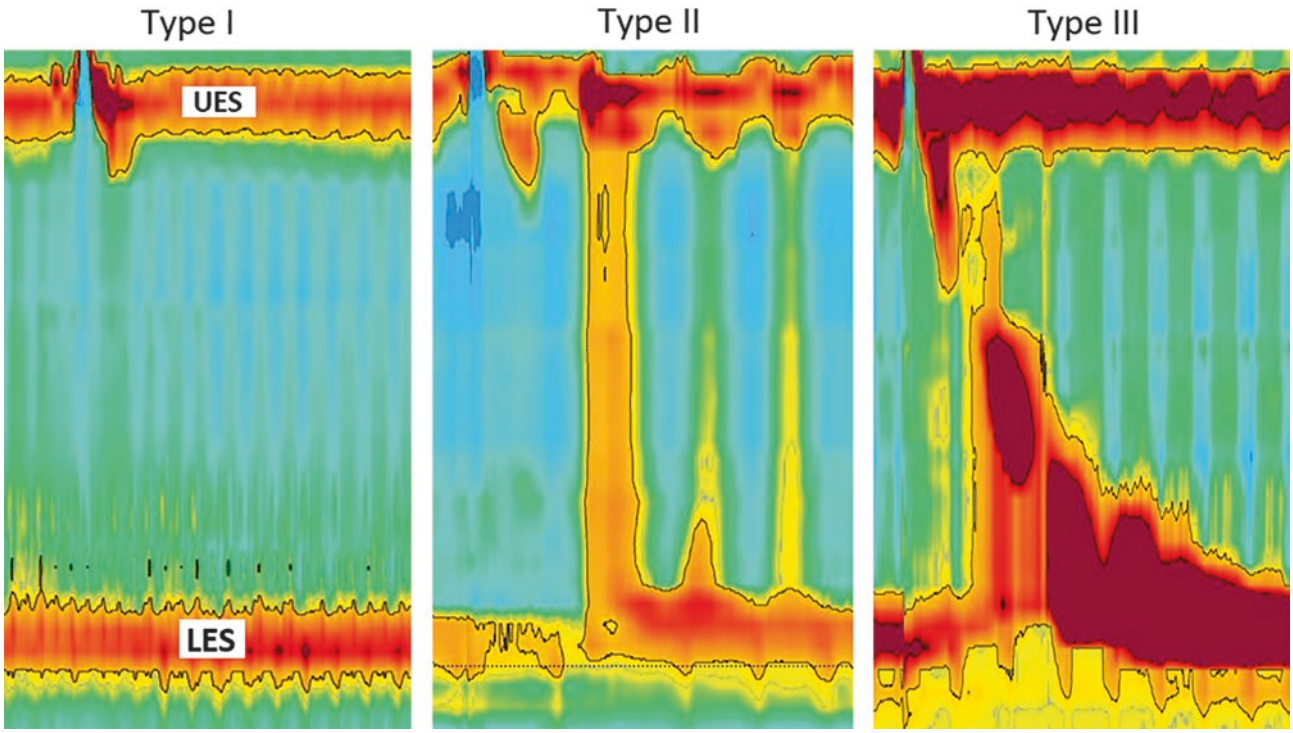


Fig. 1.15 Achalasia of the esophagus according to MOHR (Chicago Classification 3.0, 2015). -Type I: absence of measurable contractions-pressures in the esophageal lumen. -Type II (most common): no migratory contractions, but diffuse increase (pressurization) of intra-esophageal pressure. -Type III (most difficult to treat): achalasia with spasmodic contractions of the esophageal muscle. The lower esophageal sphincter (LES) is incapable of relaxation in all three types. (Tracings from M. Bouin)

- Pneumatic dilatation of the LES by per oral dilatation balloons. It is a simple technique, performed under endoscopic and/or fluoroscopic visualization, but having a significant risk of perforation (5–10% of procedures).
- Botulinum toxin endoscopic infiltration of the LES is a simple technique, however, with short-lasting results (3–12 months, and can be repeated). It can be very helpful for nonsurgical candidates.
- Pharmacological reduction of sphincter tone with calcium channel blocker drugs (nifedipine, etc.) or nitric derivatives (isosorbide, etc.) may help some patients.

These treatments, by alleviating the sphincter obstacle, usually allow an adequate caloric feeding, but the swallowing function may remain suboptimal due to the absence of esophageal contractions for propulsion of the food down within the esophagus (many patients need to eat in upright position relying upon gravity for food transit to the stomach).

2. ***Diffuse esophageal spasm*** (distal esophageal spasm, DES) is an abnormality characterized by nonperistaltic, multiphasic, prolonged contractile waves (tertiary waves), often of excessive amplitude (Fig. 1.13).

(a) Symptoms of DES. The primary symptoms are dysphagia and chest pain. The latter occurs sporadically, is usually of short duration (seconds to minutes), and can be qualitatively similar to the pain of cardiac ischemia.

(b) Causes of DES. Diffuse spasm is rarely seen during manometry tests (mainly due to its intermittent schedule). It may be idiopathic or more often associated with achalasia of the esophagus (and responsible for chest pains that may be experienced by patients suffering of achalasia).

The term "esophageal spasm" is frequently used in clinic to refer to constrictive chest pain of noncardiac origin and that is attributed to presumed esophageal spasm. These spastic phenomena can be caused by the ingestion of very cold or very hot food or occur during episodes of GE reflux. The manometric (or other) documentation of these episodes is difficult precisely because of their brief and intermittent nature.

3. ***Jackhammer esophagus*** is detected by HRM: LES relaxation is normal, but the esophageal waves are markedly hypercontractile (DIC > 8000 mmHg) and

often repetitive (such as the bursts of a jackhammer) (Fig. 1.14). It is seen in about 3% of the manometry tracings in a tertiary center. It is most often associated with dysphagia, but symptoms of reflux or chest pain are not rare (40%).

Treatment by endoscopic injections of botulinum toxin or by surgical (or endoscopic) long myotomy of the esophagus can be proposed.

4. ***Nutcracker esophagus*** [called so because of the spiral spastic contraction of the esophagus mimicking a nutcracker (or corkscrew) seen during barium X-ray in some of these patients] identified in traditional manometry is probably close to the jackhammer esophagus seen in high resolution manometry. It is characterized by peristaltic waves of exaggerated amplitude and duration. Its clinical significance has always been debated.
5. ***Opioid-induced esophageal dysmotility***. Chronic use opiates can affect the esophagus just as it affects the colon (constipation) or stomach (gastroparesis). It is now recognized as a cause of esophageal dysmotility (often with dysphagia and syndrome of obstruction of the GE junction in manometry).

In 2021, Chicago Classification v. 4.0 was proposed to identify contractile abnormalities detected during high resolution manometry testing of the esophagus (see Table 1.1).

Table 1.1 Esophageal manometric abnormalities on HRM: Chicago Classification v. 4.0

Disorders of esophagogastric junction outflow
Achalasia type I (no esophageal contractility)
Achalasia type II (with esophageal lumen hyperpressure)
Achalasia type III (with esophageal spasm)
Esophagogastric junction outflow obstruction (mechanical obstruction)
Disorders of peristalsis
Absent contractility (scleroderma)
Distal esophageal spasm
Hypercontractile esophagus (jackhammer, opiates)
Ineffective esophageal motility

1.8.4 Sensitivity Disorders

Some patients will experience esophageal symptoms (pyrosis, dysphagia, acute retrosternal pain, etc.) unexplained by any lesional or recognized motor abnormality of the esophagus.

- Globus (previously called globus hystericus) refers to a sensation of a lump or tightness in the throat (without laryngeal or upper esophageal lesion to explain it). It is usually clearly related to stress or anxiety and treated by reassurance.
- Esophageal hypersensitivity and functional heartburn refer to patients with heartburns without esophagitis (only 30% of endoscopies for reflux symptoms indicate erosive esophagitis) or reflux [only 50% of symptomatic patients with a negative endoscopy have reflux documented on pH studies and are identified as NERD (nonerosive reflux disease)].

In many of these patients, an exaggerated sensitivity to various provocative stimuli such as intraesophageal infusion of acid (Bernstein test) or balloon dilation can be demonstrated. Hypersensitivity is manifested either by an abnormally low pain threshold that results in patients feeling stimuli at undetectable levels in a normal subject (allodynia)or by a painful sensation of exaggerated intensity (hyperalgesia).

As with other digestive pathologies involving visceral hypersensitivity (irritable bowel syndrome, functional dyspepsia, etc.), the pathophysiology and treatment of these conditions remain poorly defined. Tricyclic agents (amitriptyline), serotoninreuptake inhibitors (SSRI: citalopram, venlafaxine, etc.), or pregabalin are often prescribed to alleviate symptoms. Psychological interventions may also help.

1.9 Miscellaneous

1.9.1 Hiatal Hernia

(a) **Sliding hernia**. In this hernia, the cardia (and upper stomach) abandons its intra-abdominal place to slip up into the chest cavity (see Fig. 1.16). This is the most common hiatal hernia. It is frequently observed in radiological or endoscopic examinations, and it is often wrongly accused as the source of the patient symptoms. Hiatal hernia was long considered as a

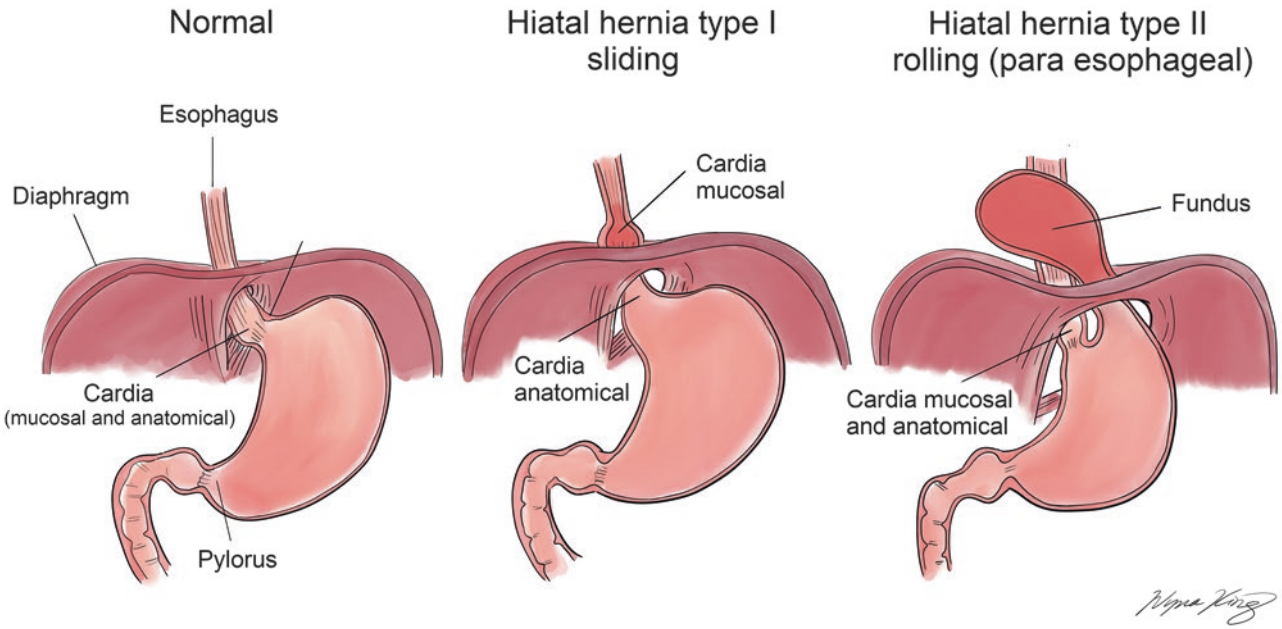


Fig. 1.16 Representation of the most frequent forms of hiatal hernias

synonym for gastroesophageal reflux [since the only treatment for esophagitis or reflux available then involved the surgical correction of the hernia by a fundoplication procedure (Nissen, Belsey, etc.)]. It is now recognized that (1) large hernias are certainly involved in reflux or regurgitation and may require surgical correction, (2) hiatal hernia may be a risk factor for GERD by modifying the anatomy of the cardia and disrupting the barrier function normally carried out by the internal and so-called external sphincters as previously discussed, but (3) hiatal hernia is not a disease in itself, and can be found in many asymptomatic individuals.

(b) **Paraesophageal (or diaphragmatic or rolling) hernia.** This is frequently considered as a serious condition. In this form of hernia, the cardia (usually) remains in its normal anatomical position, but a portion of the fundus (or gastric body) is herniated (rolls) into the chest cavity through a paraesophageal hiatus or diaphragmatic tear (see Fig. 1.16). This hernia does not cause acid reflux, but may lead to dysphagia (compression of the distal esophagus by the dilated herniated fundic pouch) or pain (fundic distension of the pouch after meal). Given the associated risk of gastric volvulus, corrective surgery is usually recommended.

The following hernia classification is also used:

- Type I: sliding hernia (90% of cases).
- Type II: paraesophageal hernia (rolling).
- Type III: mixed hernia where Types I and II coexist.
- Type IV: hernia of the whole stomach [often with other viscera (small bowel, colon, etc.)].

1.9.2 Diverticulum

(a) **Traction diverticulum** is due to attraction of the esophageal wall by its adhesion to a surrounding lesion (e.g., tuberculous granuloma). The entire esophageal wall is then present in these diverticula that remain, in most cases, asymptomatic.

(b) **Pulsion diverticulum** is obtained by the thrust of the mucosa through a weakened area of the muscular layers:

- *Epiphrenic diverticulum* is located above the lower esophageal sphincter. It develops most of the time in conjunction with esophageal achalasia or diffuse esophageal spasm where the exaggerated intraluminal pressure in the distal esophagus forces herniation of the mucosa through the muscle layers of the esophageal wall. The diverticulum itself is rarely symptomatic.
- *Zenker's diverticulum* is found in the posterior pharynx, in front of the vertebral column, just above the upper esophageal sphincter (UES), and, as for the epiphrenic diverticulum described above, is usually related to a sphincter defect. It develops in Killian's triangle (where the oblique fibers of the pharynx meet the transverse fibers of the UES) that constitutes a zone of relative weakness where the mucosa can protrude through the muscle layers in response to pathologically elevated intraluminal pressure caused by a poorly compliant and relaxing UES.

Zenker's diverticulum is manifested by the following phenomena:

- Upper dysphagia (obstruction to food passage by a poorly relaxing UES or due to external compression by a large diverticulum).
- Cervical mass (rare), created by the diverticulum (filled with secretions and food).
- Regurgitation, often nocturnal and spontaneous, of undigested food.
- Halitosis, due to stagnation of food and debris in the diverticulum.

Barium X-ray will demonstrate the diverticulum localized at the posterior part of the pharynx, in front of the vertebral column, and above an often hypertrophic-appearing cricopharyngeal muscle.

Endoscopy must be carried out with great caution since the instrument may inadvertently enter and perforate the diverticulum. Investigation of upper dysphagia is often better served by a preliminary radiological study to guide the endoscopic maneuvers.

Treatment of Zenker's diverticulum is realized by a myotomy of the cricopharyngeal muscle, surgically or endoscopically, to release the sphincter obstruction.

1.9.3 Esophageal Rupture

Rupture of the esophagus is a catastrophic condition, due to the ensuing mediastinitis, and it must be identified and treated promptly. Patients present with severe chest pain, fever ± septic state, and subcutaneous emphysema (feeling of crackling of the subcutaneous tissue of the upper chest or neck due to infiltration of swallowed air through an esophageal perforation).

Radiological examination, preferably by CT scanning of the thorax, will reveal the localization and size of the perforation as well as possible fluid collections or abscesses of the mediastinum.

Most perforations are iatrogenic (e.g., during endoscopy and/or dilatation by balloon or bougie). It can complicate severe caustic esophagitis, but can also occur spontaneously (Boerhaave's syndrome), in which case it is often associated with a chest trauma (car accident, violent vomiting or coughing, etc.) resulting in a strong and acute increase in intraesophageal pressure.

Small esophageal perforations may spontaneously close fairly quickly. These cases may be treated with antibiotics and cessation of oral feeds. Other cases require surgical drainage, ± suture of the perforation or even removal of the esophagus.

1.9.4 Esophageal Bleeding

Bleeding of esophageal origin will present with hematemesis of bright red blood and/or melena (see ▶ Chap. 11 on upper digestive hemorrhage).

Endoscopy may reveal the following:

- An esophagitis with or without esophageal ulcer (a rare cause of massive acute bleeding).
- An ulcerated neoplasia (chronic bleeding is more frequent).
- Esophageal varices secondary to portal hypertension (a common cause of massive upper GI hemorrhage in patients with cirrhosis of the liver).
- A Mallory-Weiss tear (see next point).

1.9.5 Mallory-Weiss

The Mallory-Weiss tear is a common cause of digestive hemorrhage. It is a longitudinal mucosal tear (1–2 cm long) of the cardia. It is a mechanical tear secondary to vomiting efforts with up and down movements of the cardia. In most cases, the bleeding of a Mallory-Weiss will be moderate and will cease spontaneously with rapid tear re-epithelialization in 24–48 hours.

1.9.6 Schatzki's Ring

It was classically identified on radiological barium swallow as a circumferential ring at the gastroesophageal junction that caused intermittent dysphagia to solid foods. It is now recognized mainly by endoscopy (◘ Fig. 1.17) and treated by per-endoscopic dilatation (if dysphagia is present). Initially considered as a birth defect, it is increasingly seen as a subtle form of peptic stricture associated with GE reflux and requiring treatment with PPIs to prevent its recurrence.

PS: For complementary readings on the esophagus, see ▶ Chaps. 9 and ▶ 29.

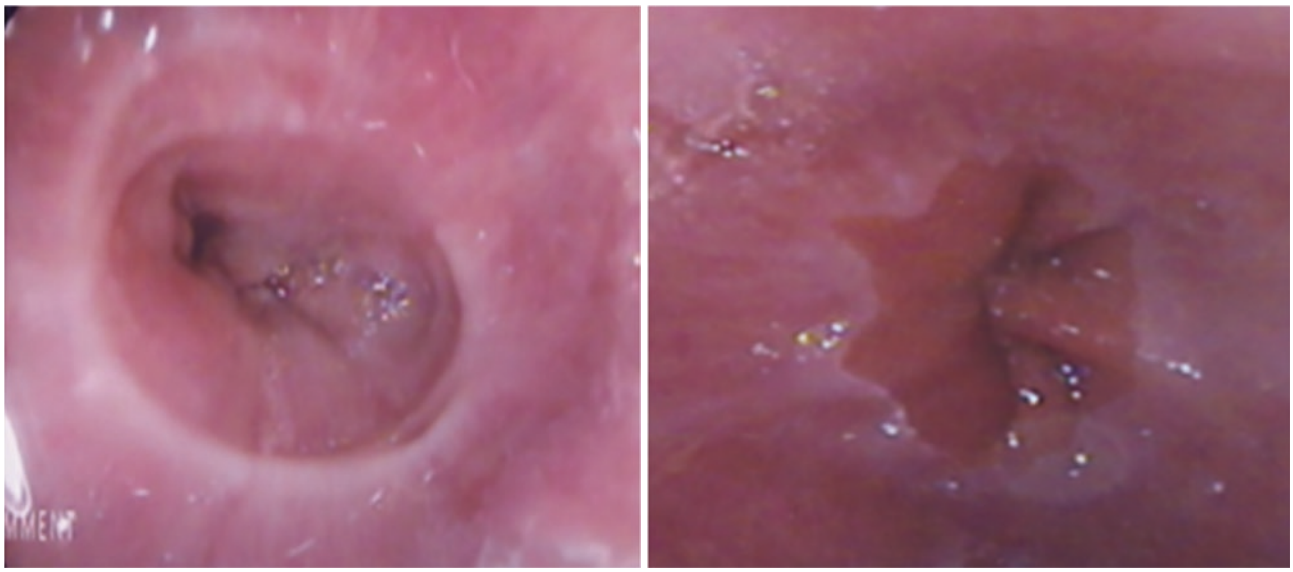


Fig. 1.17 Left: Schatzki's ring seen in endoscopy as a fibrous narrowing of the esophagogastric junction and that may block the transport of solid foods; figure on the right: normal GE junction. (Photos by P. Poitras)