Chapter 62

The oesophagus

Learning objectives

To understand:

- The anatomy and physiology of the oesophagus and their relationship to disease
- The clinical features, investigations and treatment of benign and malignant disease with particular reference to the common adult disorders

BACKGROUND

Surgical anatomy

The oesophagus is a muscular tube, approximately 25 cm long, mainly occupying the posterior mediastinum and extending from the upper oesophageal sphincter (the cricopharyngeus muscle) in the neck to the junction with the cardia of the stomach. The musculature of the upper oesophagus, including the upper sphincter, is striated. This is followed by a transitional zone of both striated and smooth muscle with the proportion of the latter progressively increasing so that, in the lower half of the oesophagus, there is only smooth muscle. It is lined throughout with squamous epithelium. The parasympathetic nerve supply is mediated by branches of the vagus nerve that has synaptic connections to the myenteric (Auerbach's) plexus. Meissner's submucosal plexus is sparse in the oesophagus.

The upper sphincter consists of powerful striated muscle. The lower sphincter is more subtle and created by the asymmetrical arrangement of muscle fibres in the distal oesophageal wall just above the oesophagogastric junction. It is helpful to remember the distances 15, 25 and 40 cm for anatomical location during endoscopy (Figure 62.1).

Physiology

The main function of the oesophagus is to transfer food from the mouth to the stomach in a coordinated fashion. The initial movement from the mouth is voluntary. The pharyngeal phase of swallowing involves sequential contraction of the oropharyngeal musculature, closure of the nasal and respiratory passages, cessation of breathing and opening of the upper oesophageal sphincter. Beyond this level, swallowing is involuntary. The body of the oesophagus propels the bolus through a relaxed lower oesophageal sphincter (LOS) into the stomach, taking air with it (Figure 62.2). This coordinated oesophageal wave which follows a conscious swallow is called primary peristalsis. It is under vagal control, although there are specific neurotransmitters that control the LOS.

The upper oesophageal sphincter is normally closed at rest and serves as a protective mechanism against regurgitation of oesophageal contents into the respiratory passages. It also serves to stop air entering the oesophagus other than the small amount that enters during swallowing.

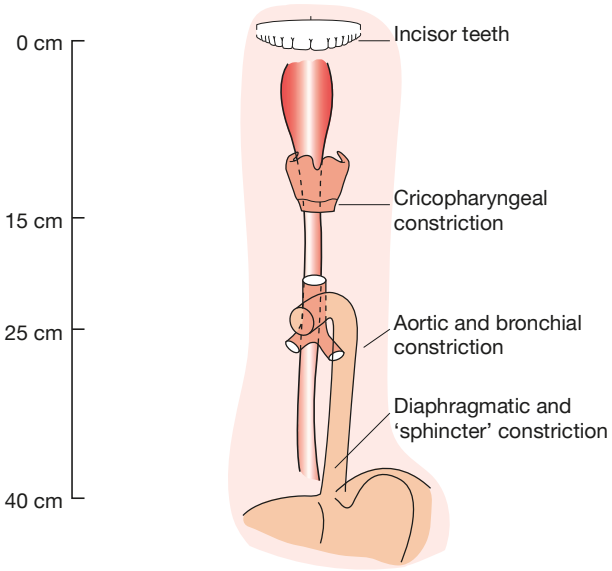

Figure 62.1 Endoscopic landmarks. Distances are given from the incisor teeth. They vary slightly with the build of the individual.

Leopold Auerbach, 1828–1897, Professor of Neuropathology, Breslau, Germany, (now Wroclaw, Poland), described the myenteric plexus in 1862.
Georg Meissner, 1829–1905, Professor of Physiology, Göttingen, Germany, described the submucosal plexus in 1852.

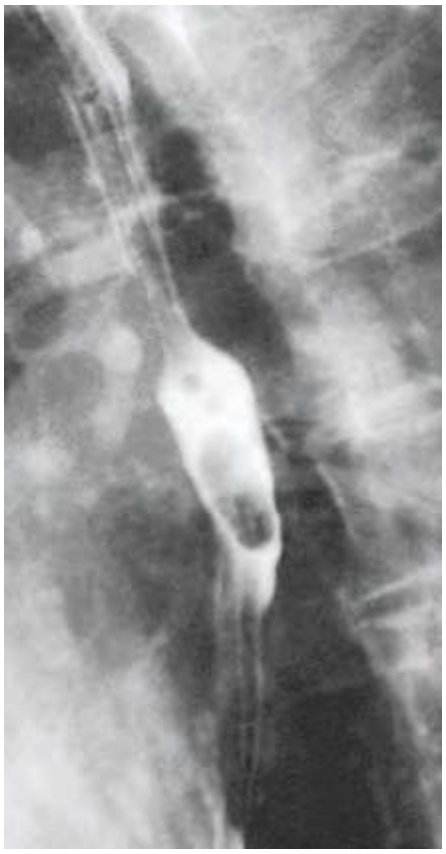

Figure 62.2 A bolus of barium or food usually takes air with it into the stomach.

The LOS is a zone of relatively high pressure that prevents gastric contents from refluxing into the lower oesophagus (**Figure 62.3**). In addition to opening in response to a primary peristaltic wave, the sphincter also relaxes to allow air to escape from the stomach and at the time of vomiting. A variety of factors influences sphincter tone, notably food, gastric distension, gastrointestinal (GI) hormones, drugs and smoking. The arrangement of muscle fibres, their differential responses to specific neurotransmitters and the relationship to diaphragmatic contraction all contribute to the action of the LOS. The presence of the physiological sphincter was first demonstrated by Code using manometry with small balloons. Until recently LOS pressure was measured by water-perfused tubes, but the introduction of catheters containing multiple microtransducers has meant that this approach has been superseded by high-resolution manometry. The normal LOS is 3–4 cm in length and has a pressure of 10–25 mmHg.

Manometry is also used to assess the speed and amplitude of oesophageal body contractions and ensure that peristalsis is propagated down the entire length of the oesophagus (**Figure 62.4**). Secondary peristalsis is the normal reflex preceded by a conscious swallow. It is worth remembering that most clearance swallows to neutralise refluxed gastric acid are, however, achieved by primary peristalsis, which carries saliva with its high bicarbonate content down to the lower oesophagus. Tertiary contractions are non-peristaltic waves that are infrequent (<10%) during laboratory-based manometry, although readily detected if manometry is undertaken while the patient eats a meal.

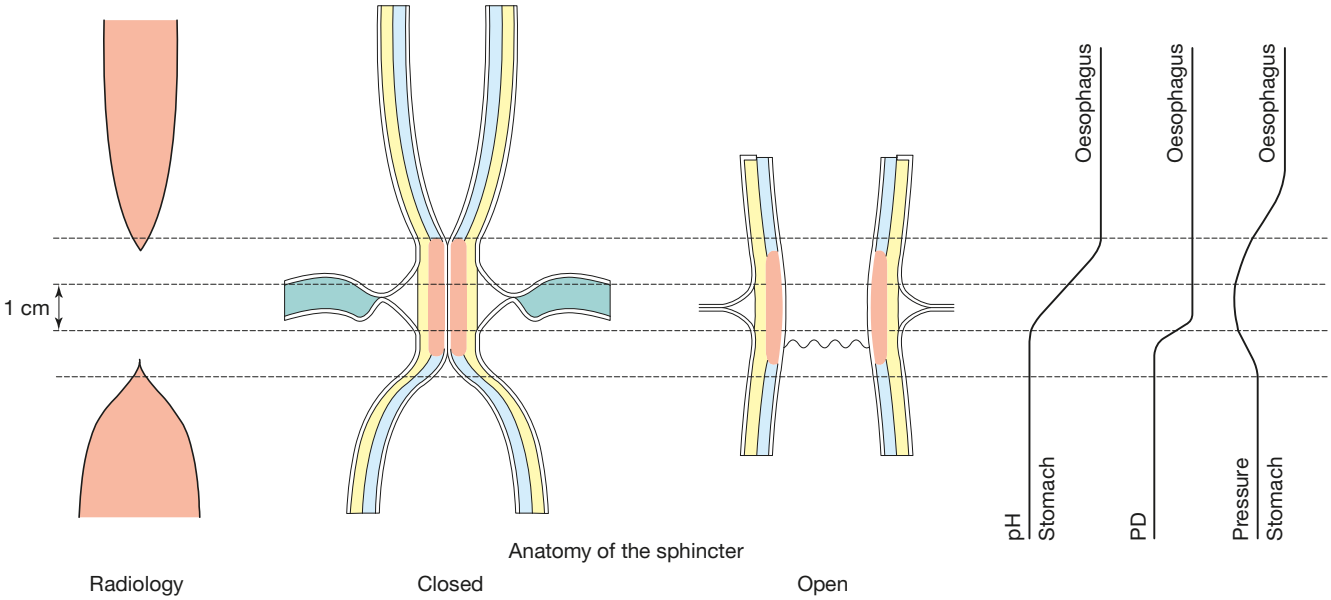

Figure 62.3 Correlation between the radiological appearances of a barium column and the lower oesophageal sphincter open and closed. The three curves on the right, set up vertically, show the pH gradient, the mucosal potential difference (PD), marking the junction of squamous and columnar epithelium, and the high-pressure zone of the sphincter.

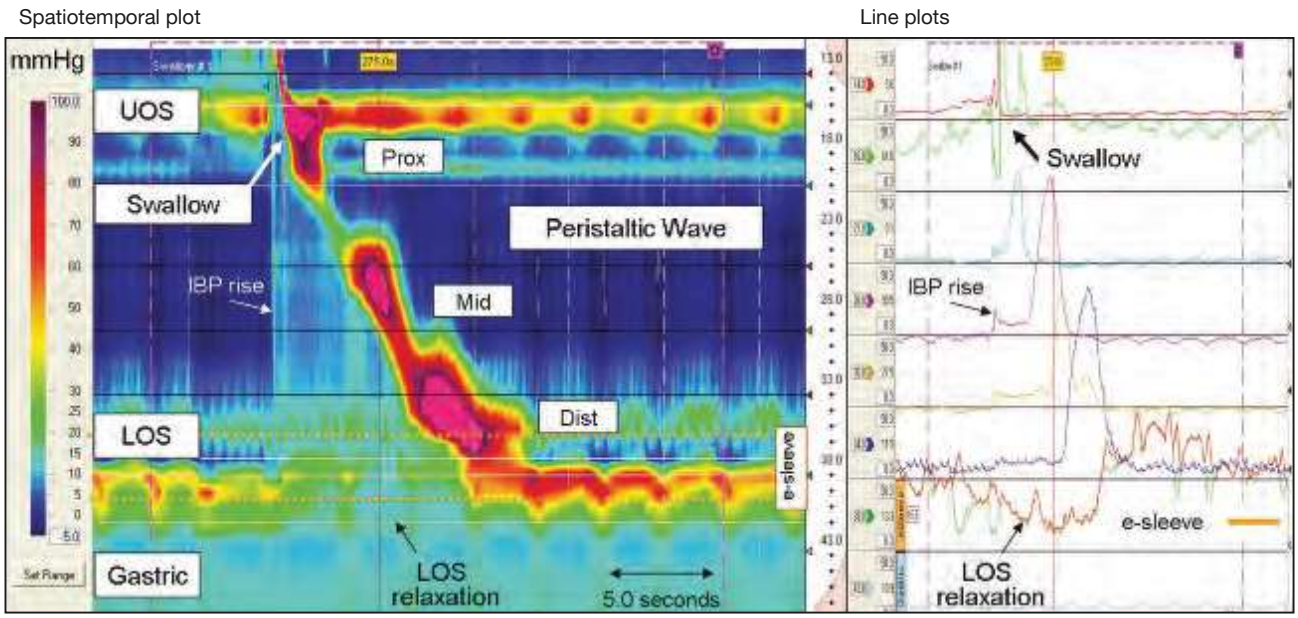

Figure 62.4 High-resolution manometry depicts oesophageal function from the pharynx to the stomach. The spatiotemporal plot presents the same information as presented in the 'conventional' line plots. Time is on the *x*-axis and distance from the nares is on the *y*-axis. Pressure is assigned a colour (legend right). The segmental anatomy of oesophagus is easy to appreciate (not seen on conventional manometry). The coordinated relaxation of the upper (UOS) and lower oesophageal sphincter (LOS) is obvious, as is the increasing pressure and duration of the peristaltic wave as it passes distally. The intrabolus pressure (IBP) between the peristaltic wave and oesophagogastric junction, and the pressure gradient across the gastro-oesophageal junction are visualized. These represent the force that drives bolus through the oesophagus and into the stomach.

Symptoms

Summary box 62.1

Symptoms of oesophageal disease

- Difficulty in swallowing described as food or fluid sticking (oesophageal dysphagia): must rule out malignancy
- Pain on swallowing (odynophagia): suggests inflammation and ulceration
- Regurgitation or reflux (heartburn): common in gastro-oesophageal reflux disease
- Chest pain: difficult to distinguish from cardiac pain

Dysphagia

Dysphagia is used to describe difficulty with swallowing. When there is a problem with swallowing in the voluntary (oral or pharyngeal) phases, patients will usually say that they cannot swallow properly, but they do not characteristically describe 'food sticking'. Instead, when they try to initiate a conscious swallow, food fails to enter the oesophagus, stays in the mouth or enters the airway, causing coughing or spluttering. Virtually all causes of this type of dysphagia are chronic neurological or muscular diseases. Oesophageal dysphagia occurs in the involuntary phase and is characterised by a sensation of food sticking. The nature of this type of dysphagia is often informative with regard to a likely diagnosis. Dysphagia may occur acutely or in a chronic fashion, can affect solids and/or fluids, and be intermittent or progressive. Although many patients point to a site of impaction, this is unreliable.

Odynophagia

Odynophagia refers to pain on swallowing. Patients with reflux oesophagitis often feel retrosternal discomfort within a few seconds of swallowing hot beverages, citrus drinks or alcohol. Odynophagia is also a feature of infective oesophagitis and may be particularly severe in chemical injury.

Regurgitation and reflux

Regurgitation and reflux are often used synonymously. It is helpful to differentiate between them, although it is not always possible. Regurgitation should strictly refer to the return of oesophageal contents from above a functional or mechanical obstruction. Reflux is the passive return of gastroduodenal contents to the mouth as part of the symptomatology of gastro-oesophageal reflux disease (GORD). Loss of weight, anaemia, cachexia, change of voice due to refluxed material irritating the vocal folds, and cough or dyspnoea due to tracheal aspiration may all accompany regurgitation and/or reflux.

Chest pain

Chest pain similar in character to angina pectoris may arise from an oesophageal cause, especially gastro-oesophageal reflux and motility disorders. Exercise-induced chest pain can be due to reflux.

Investigations

Radiography

Contrast radiography has been somewhat overshadowed by endoscopy but remains a useful investigation to demonstrate changes in oesophageal diameter, anatomical distortion or abnormal motility. An adequate barium swallow should be tailored to the problem under investigation. It may be helpful to give a solid bolus (bread or marshmallow) if a motility disorder is suspected. Video recording is useful to allow subsequent replay and detailed analysis. Barium radiology is, however, inaccurate in the diagnosis of gastro-oesophageal reflux, unless reflux is gross, and should not be used for this purpose. Plain radiographs will show some foreign bodies.

Cross-sectional imaging by computed tomography (CT) is now an essential investigation in the assessment of neoplasms of the oesophagus and can be used in place of a contrast swallow to demonstrate perforation. The role of CT and other cancer-specific tests is described later.

Endoscopy

Endoscopy is necessary for the investigation of most oesophageal conditions. It is required to view the inside of the oesophagus and the oesophagogastric junction, to obtain a biopsy or cytology specimen, for the removal of foreign bodies and to dilate strictures. Traditionally, there are two types of instrument available, the rigid oesophagoscope and the flexible video endoscope, but the rigid instrument is now virtually obsolete.

For flexible video gastroduodenoscopy, general anaesthesia is not required; most examinations can be done on an outpatient basis, and the quality of the magnified image is superb. The technology associated with video endoscopy continues to improve. Magnification is a standard feature of the modern endoscope and widely used in conjunction with agents that can be sprayed on to the mucosa, such as acetic acid to enhance mucosal detail. Novel techniques that rely on fluorescence and narrow band imaging to enhance visual contrast are becoming increasingly used for the identification of mucosal abnormalities that are not easily seen with white light, e.g. in patients with Barrett's oesophagus undergoing endoscopic surveillance.

As a matter of routine, the stomach and duodenum are examined as well as the oesophagus. If a stricture is encountered, it may be helpful to dilate it to allow a complete inspection of the upper GI tract, but this decision should be dictated by clinical circumstances and an appreciation of the perforation risk, especially if the visual appearances are thought to indicate neoplasia.

Endosonography

Endoscopic ultrasonography relies on a high-frequency (5–30 MHz) transducer located at the tip of the endoscope to provide highly detailed images of the layers of the oesophageal wall and mediastinal structures close to the oesophagus. Radial echoendoscopes have a rotating transducer that creates a circular image with the endoscope in the centre, and this type of scanner is widely used to create diagnostic transverse sectional images at right angles to the long axis of the oesophagus (**Figure 62.5**). Linear echoendoscopes produce a sectoral image in the line of the endoscope and are used to biopsy submucosal oesophageal lesions, mediastinal masses such as lymph nodes (**Figure 62.6**) or suspicious lesions that might lie outside a proposed surgical field. Radial scanners without optical components are available for passage through narrow strictures over a guidewire, and catheter probes are available that can be passed down the endoscope biopsy channel.

Oesophageal manometry

Manometry is now widely used to diagnose oesophageal motility disorders. Electronic microtransducers that are not influenced by changes in patient position during the test have gradually supplanted perfusion systems. High-resolution

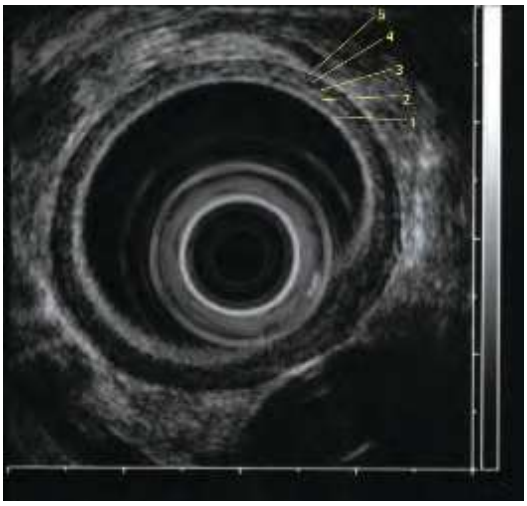

Figure 62.5 Radial endosonography indicating wall layers as alternating hyper- and hypoechoic bands.

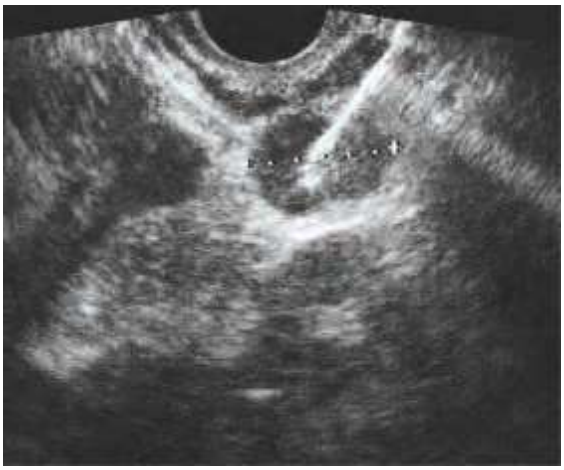

Figure 62.6 Linear endosonography with the needle traversing an ultrasound plane for guided biopsy.

manometry uses a multiple (up to 36) microtransducer catheter with the results displayed as spatiotemporal plots, and this has now largely supplanted earlier manometry systems. This system provides comprehensive information about oesophageal body function and associated behaviour of the LOS.

Twenty-four-hour pH and combined pH-impedance recording

Prolonged measurement of pH is now accepted as the most accurate method for the diagnosis of gastro-oesophageal reflux. It is particularly useful in patients with atypical reflux symptoms, those without endoscopic oesophagitis and when patients respond poorly to intensive medical therapy. A small pH probe is passed into the distal oesophagus and positioned 5 cm above the upper margin of the LOS, as defined by manometry. The probe is connected to a miniature digital recorder that is worn on a belt and allows most normal activities. Patients mark symptomatic events such as heartburn. A 24-hour recording period is usual, and the pH record is analysed by an automated computer program. An oesophageal pH <4 at the level of the pH electrode is conventionally considered the cut-off value and, in most oesophageal laboratories, the total time when pH is <4 in a 24-hour period does not exceed 4% in a healthy adult. Patterns of reflux and the correlation between symptoms and oesophageal pH <4 can be calculated. Most laboratories use a scoring system (Johnson–DeMeester) to create a numerical value, above which reflux is considered pathological. Radiotelemetry pH probes are also available that can be fixed to the oesophageal wall endoscopically without the need for a transnasal catheter. The introduction of catheter assemblies that incorporate multiple electrodes along the length of the catheter, as well as a pH probe, has resulted in combined measurement of pH and electrical impedance. Impedance measurement differentiates anterograde from retrograde bolus transit and gas from liquid, and provides a reliable measure of non-acidic or weakly acidic events that cause symptoms. This may be important in the identification of patients with non-erosive reflux disease on endoscopy.

Therapeutic procedures

Dilatation of strictures

Stricture dilatation is essentially undertaken for benign conditions and should be used with caution in the context of malignant disease. The risks associated with dilatation for malignant disease are discussed later. The advent of guidewire-directed dilatation of the oesophagus in the 1970s was a major advance over earlier blind dilatation systems. Their use is now considered standard practice. There are many different designs, but essentially they are solid dilators of increasing diameter or inflatable balloons with rigid walls. To restore normal swallowing, a stricture should be dilated to at least 16 mm in diameter, although this may need to be achieved over a series of procedures, depending on the perceived nature of the stricture. A guidewire is passed down the biopsy channel of an endoscope and through the stricture under vision. If the stricture is long or tortuous, this should be undertaken under radiological guidance to ensure that the guidewire passes easily into the stomach. The endoscope is withdrawn, leaving the guidewire in place, and graduated dilators or a balloon dilator are passed over the guidewire, sometimes with radiographic screening for safety purposes. The dilatation of reflux-induced strictures is usually straightforward. These strictures are nearly always short and at the oesophagogastric junction, so that the stomach is visible through the narrowed segment. Radiological control is rarely needed. Conversely, distal oesophageal adenocarcinomas extending into the stomach are often soft, friable and tortuous to negotiate. Caustic strictures often occur high in the thorax and may be very long.

Balloons for oesophageal use tend to have inflation diameters of 25–40 mm and may also be used for dilatation. Pneumatic dilatation is widely used to disrupt the non-relaxing LOS in achalasia.

Thermal recanalisation

Various types of laser (mainly Nd–YAG), bipolar diathermy, injection of absolute alcohol or argon-beam plasma coagulation have all been used successfully to ablate tissue in order to recanalise the oesophagus.

CONGENITAL ABNORMALITIES

See Chapter 9.

FOREIGN BODIES IN THE OESOPHAGUS

All manner of foreign bodies have become arrested in the oesophagus (**Figure 62.7**). Button batteries may be a troublesome problem in children. The most common impacted

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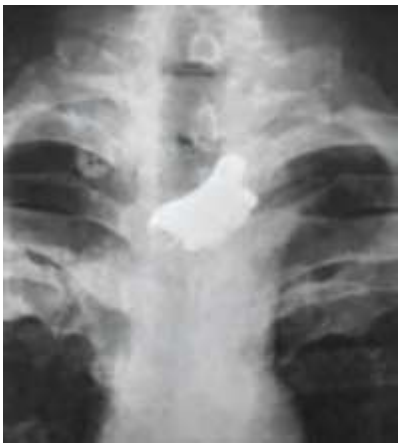


Figure 62.7 False teeth impacted in the oesophagus. (Note: modern dentures are usually radiolucent.)

material is food, and this usually occurs above a significant pathological lesion (**Figure 62.8**). Plain radiographs are often useful for foreign bodies, but modern denture materials are not always radiopaque. A contrast examination is not usually required and only makes endoscopy more difficult.

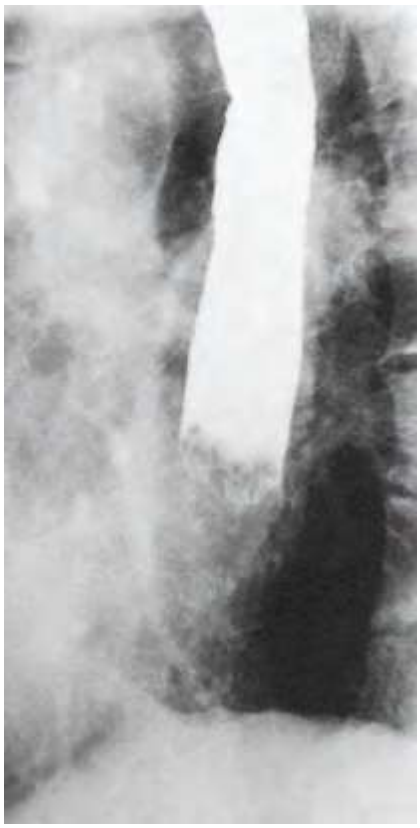


Figure 62.8 An impacted meat bolus at the lower end of the oesophagus. This may be the first presentation of a benign stricture or a malignant tumour.

Summary box 62.2

Foreign bodies

- The most common is a food bolus, which usually signifies underlying disease
- It is usually possible to remove foreign bodies by flexible endoscopy
- Beware of button batteries in the oesophagus

Foreign bodies that have become stuck in the oesophagus should be removed by flexible endoscopy using suitable grasping forceps, a snare or a basket. If the object may injure the oesophagus on withdrawal, an overtube can be used, and the endoscope and object can be withdrawn into the overtube before removal. Button batteries can be a particular worry because they are difficult to grasp, and it is tempting to push them on into the stomach. However, an exhausted battery may rapidly corrode in the GI tract and is best extracted. A multiwire basket of the type used for gallstone retrieval nearly always works. An impacted food bolus will often break up and pass on if the patient is given fizzy drinks and confined to fluids for a short time. The cause of the impaction must then be investigated. If symptoms are severe or the bolus does not pass, it can be extracted or broken up at endoscopy.

PERFORATION

Perforation of the oesophagus is usually iatrogenic (at therapeutic endoscopy) or due to 'barotrauma' (spontaneous perforation). Many instrumental perforations can be managed conservatively, but spontaneous perforation is often a life-threatening condition that regularly requires surgical intervention.

Summary box 62.3

Perforation of the oesophagus

- Potentially lethal complication due to mediastinitis and septic shock
- Numerous causes, but may be iatrogenic
- Surgical emphysema is virtually pathognomonic
- Treatment is urgent; it may be conservative or surgical, but requires specialised care

Barotrauma (spontaneous perforation, Boerhaave's syndrome)

This occurs classically when a person vomits against a closed glottis. The pressure in the oesophagus increases rapidly, and the oesophagus bursts at its weakest point in the lower third, sending a stream of material into the mediastinum and often the pleural cavity as well. The condition was first identified by Boerhaave, who reported the case of a grand admiral of

Hermann Boerhaave, 1668–1738, Professor of Medicine and Botany, the University of Leiden, the Netherlands, creator of the modern method of clinical teaching.

the Dutch fleet who was a glutton and practised autoemesis. Boerhaave's syndrome is the most serious type of perforation because of the large volume of material that is released under pressure. This causes rapid chemical irritation in the mediastinum and pleura followed by infection if untreated. Barotrauma has also been described in relation to other pressure events when the patient strains against a closed glottis (e.g. defecation, labour, weight-lifting).

Diagnosis of spontaneous perforation

The clinical history is usually of severe pain in the chest or upper abdomen after a meal or a bout of drinking. Associated shortness of breath is common. Many cases are misdiagnosed as myocardial infarction, perforated peptic ulcer or pancreatitis if the pain is confined to the upper abdomen. There may be a surprising amount of rigidity on examination of the upper abdomen, even in the absence of any peritoneal contamination.

The diagnosis can usually be suspected from the history and associated clinical features. A chest radiograph is often confirmatory with air in the mediastinum, pleura or peritoneum. Pleural effusion occurs rapidly either as a result of free communication with the pleural space or as a reaction to adjacent inflammation in the mediastinum. A contrast swallow or CT is nearly always required to guide management (**Figure 62.9**).

Pathological perforation

Free perforation of ulcers or tumours of the oesophagus into the pleural space is rare. Erosion into an adjacent structure with fistula formation is more common. Aerodigestive fistula is most common and usually encountered in primary malignant disease of the oesophagus or bronchus. Coughing on eating and signs of aspiration pneumonitis may allow the problem to be recognised at a time when intervention may be appropriate and feasible. Covering the communication with a self-expanding metal stent is the usual solution. Erosion into a major vascular structure is invariably fatal.

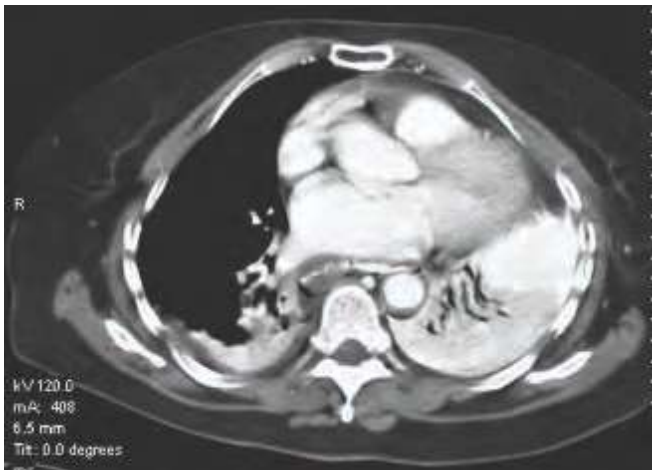


Figure 62.9 Computed tomography scan showing the site of perforation in the lower oesophagus.

Penetrating injury

Perforation by knives and bullets is uncommon, even in war, because the oesophagus is a relatively small target surrounded by other vital organs.

Foreign bodies

The oesophagus may be perforated during removal of a foreign body but, occasionally, an object that has been left in the oesophagus for several days will erode through the wall.

Instrumental perforation

Instrumentation is by far the most common cause of perforation. Modern instrumentation is remarkably safe, but perforation remains a risk that should never be forgotten.

Summary box 62.4

Instrumental perforation

- Prevention of perforation is better than cure

Perforation related to diagnostic upper GI endoscopy is unusual with an estimated frequency of about 1:4000 examinations. Perforation can occur in the pharynx or oesophagus, usually at sites of pathology or when the endoscope is passed blindly. A number of patient-related factors are associated with increased risk, including large anterior cervical osteophytes, the presence of a pharyngeal pouch and mechanical causes of obstruction. Perforation may follow biopsy of a malignant tumour.

Patients undergoing therapeutic endoscopy have a perforation risk that is at least 10 times greater than those undergoing diagnostic endoscopy. The oesophagus may be perforated by guidewires, graduated dilators or balloons, or during the placement of self-expanding stents. The risk is considerably higher in patients with malignancy.

Diagnosis of instrumental perforation

In most cases, a combination of technical difficulties and an interventional procedure should lead to a high index of suspicion. History and physical signs may be useful pointers to the site of perforation.

Cervical perforation may result in pain localised to the neck, hoarseness, painful neck movements and subcutaneous emphysema. Intrathoracic and intra-abdominal perforations, which are more common, can give rise to immediate symptoms and signs either during or at the end of the procedure, including chest pain, haemodynamic instability, oxygen desaturation or visual evidence of perforation. Within the first 24 hours, patients may additionally complain of abdominal pain or respiratory difficulties. There may be evidence of subcutaneous emphysema, pneumothorax or hydropneumothorax. In some patients, the diagnosis may be missed and recognised only at a late stage beyond 24 hours, as unexplained pyrexia, systemic sepsis or the development of a clinical fistula.

Prompt and thorough investigation is the key to management. Careful endoscopic assessment at the end of any procedure combined with a chest radiograph will identify many cases of perforation immediately. If not recognised immediately, then early and late suspected perforations should be assessed by CT. A pre-contrast scan easily demonstrates air outside the GI tract, while oral contrast localises the site of the oesophageal defect and accurately delineates specific fluid collections. Where CT is not available, a water-soluble contrast swallow should be performed although this is not always reliable in disclosing a small contained leak. If a water-soluble study is negative and clinical suspicion is high, a dilute barium swallow should be considered.

Treatment of oesophageal perforations

Perforation of the oesophagus usually leads to mediastinitis. The loose areolar tissues of the posterior mediastinum allow a rapid spread of GI contents. The aim of treatment is to limit mediastinal contamination and prevent or deal with infection. Operative repair deals with the injury directly, but imposes risks of its own; non-operative treatment aims to limit the effects of mediastinitis and provide an environment in which healing can take place.

The decision between surgical and non-surgical management rests on four factors. These are:

1. the site of the perforation (cervical vs thoracoabdominal oesophagus);
2. the event causing the perforation (spontaneous vs instrumental);
3. underlying pathology (benign or malignant);
4. the status of the oesophagus before the perforation (fasted and empty vs obstructed with a stagnant residue).

It follows that most perforations that can be managed non-surgically occur in the context of small instrumental perforations of a clean oesophagus without obstruction, where leakage is likely to be confined to the nearby mediastinum at worst *(Table 62.1)*.

Instrumental perforations in the cervical oesophagus are usually small and can nearly always be managed conservatively. The development of a local abscess is an indication for cervical drainage, preventing the extension of sepsis into the mediastinum.

TABLE 62.1 Management options in perforation of the oesophagus.

Factors that favour non-surgical management	Factors that favour surgical repair
Small septic load	Large septic load
Minimal cardiovascular upset	Septic shock
Perforation confined to mediastinum	Pleura breached
Perforation by flexible endoscope	Boerhaave's syndrome
Perforation of cervical oesophagus	Perforation of abdominal oesophagus

The conservative management of an instrumental perforation in the thoracoabdominal parts of the oesophagus can be undertaken when the perforation is detected early and before oral alimentation. General guidelines for non-surgical management include:

- pain that is readily controlled with opiates;
- absence of crepitus, diffuse mediastinal gas, hydropneumothorax or pneumoperitoneum;
- mediastinal containment of the perforation with no evidence of widespread extravasation of contrast material;
- no evidence of on-going luminal obstruction or a retained foreign body.

In addition, conservative management might be appropriate in patients who have remained clinically stable despite diagnostic delay. The principles of non-interventional management involve hyperalimentation, preferably by an enteral route, nasogastric suction and broad-spectrum intravenous antibiotics.

Surgical management is required whenever patients:

- are unstable with sepsis or shock;
- have evidence of a heavily contaminated mediastinum, pleural space or peritoneum;
- have widespread intrapleural or intraperitoneal extravasation of contrast material.

Ongoing luminal obstruction (often related to malignancy) in a frail patient considered unfit for major surgery can be dealt with by placement of a covered self-expanding stent. Expanding metal stents should be used with caution in patients with benign disease because they cause significant tissue reaction and some designs are impossible to remove at a later date. Biodegradable and removable stents may be used alone or as a bridge to later definitive treatment where perforation accompanies obstruction.

For patients requiring surgery, the choice is from direct repair, the deliberate creation of an external fistula or, rarely, oesophageal resection with a view to delayed reconstruction. Direct repair is preferred by many surgeons if the perforation is recognised early (within the first 4–6 hours), and the extent of mediastinal and pleural contamination is small. After 12 hours, the tissues become swollen and friable and less suitable for direct suture. The hole in the mucosa is always bigger than the hole in the muscle, and the muscle should be incised to see the mucosal edges clearly. It is essential that there should be no obstruction distal to the repair. A variety of local tissues (gastric fundus, pericardium, intercostal muscle) have been used to buttress such repairs.

Primary repair is inadvisable with late presentation and in the presence of widespread mediastinal and pleural contamination. These patients tend to be more ill as a result of the delay, and the aim of treatment should be to achieve wide drainage with the creation of a controlled fistula and distal enteral feeding. This can usually be achieved by placing a T-tube into the oesophagus along with appropriately located drains and a feeding jejunostomy. In unusual circumstances, e.g. with extensive necrosis after corrosive ingestion, emergency oesophagectomy may be necessary. Oesophagostomy and gastrostomy should be performed with a view to delayed reconstruction.

MALLORY–WEISS SYNDROME

Forceful vomiting may produce a mucosal tear at the cardia rather than a full perforation. The mechanism of injury is different. In Boerhaave's syndrome, vomiting occurs against a closed glottis and pressure builds up in the oesophagus. In Mallory–Weiss syndrome, vigorous vomiting produces a vertical split in the gastric mucosa, immediately below the squamocolumnar junction at the cardia in 90% of cases. In only 10% is the tear in the oesophagus (Figure 62.10). The condition presents with haematemesis. Usually, the bleeding is not severe, but endoscopic injection therapy may be required for the occasional, severe case. Surgery is rarely required. There are two other injuries to the oesophagus that lie within the spectrum of the mucosal tear of Mallory–Weiss and the full-thickness tear of Boerhaave. Intramural rupture produces a dissection within the oesophageal wall that causes severe chest pain, often with odynophagia. It is best diagnosed by contrast radiology. Intramural haematoma is seen most often in elderly patients on anticoagulants or patients with coagulation disorders, and usually follows an episode of vomiting. Large haematomas causing dysphagia can occur, extending from the cardia up to the carina. The diagnosis is readily made on endoscopy. Both intramural rupture and intramural haematoma can be managed conservatively. Symptoms usually resolve in 7–14 days, and oral intake can be reinstituted as soon as symptoms allow.

CORROSIVE INJURY

Corrosives such as sodium hydroxide (lye, caustic soda) or sulphuric acid may be taken in an attempted suicide. Accidental ingestion occurs in children and when corrosives are stored in bottles labelled as beverages. All can cause severe damage to the mouth, pharynx, larynx, oesophagus and stomach. The type of agent, its concentration and the volume ingested largely determine the extent of damage. In general, alkalis are relatively odourless and tasteless, making them more likely to be ingested in large volume. Alkalis cause liquefaction, saponification of fats, dehydration and thrombosis of blood vessels which usually leads to fibrous scarring. Acids cause coagulative necrosis with eschar formation, and this coagulant may limit penetration to deeper layers of the oesophageal wall. Acids also cause more gastric damage than alkalis because of the induction of intense pylorospasm with pooling in the antrum.

Symptoms and signs are notoriously unreliable in predicting the severity of injury. The key to management is early endoscopy by an experienced endoscopist to inspect the whole of the oesophagus and stomach (Figure 62.11). Deep ulcers and the recognition of a grey or black eschar signify the most severe lesions with the greatest risk of perforation. Minor injuries with only oedema of the mucosa resolve rapidly with no late sequelae. These patients can safely be fed. With more severe injuries, a feeding jejunostomy may be appropriate until the patient can swallow saliva satisfactorily. The widespread use of broad-spectrum antibiotics and steroids is not supported by evidence.

Regular endoscopic examinations are the best way to assess stricture development (Figure 62.12). Significant stricture formation occurs in about 50% of patients with extensive mucosal damage (Figure 62.13). The role and timing of repeat endoscopies with or without dilatation in such patients

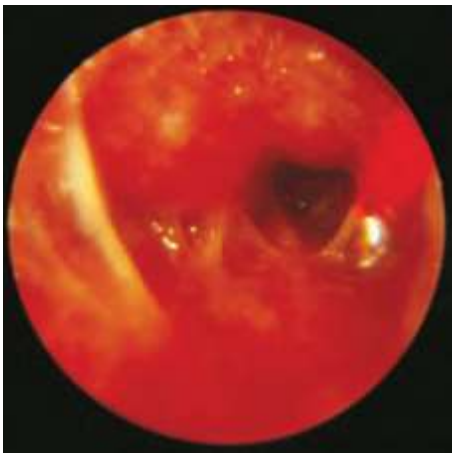


Figure 62.11 Acute caustic burn in the haemorrhagic phase.

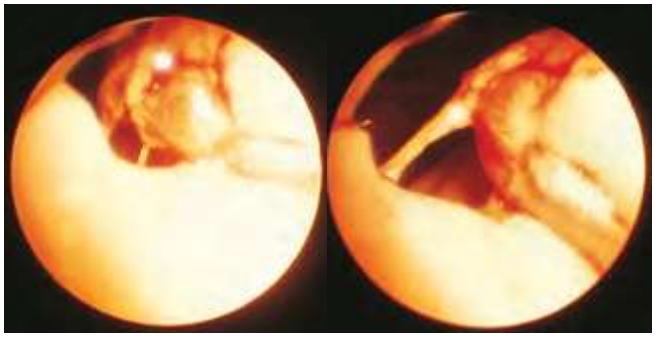


Figure 62.10 The endoscopic appearance of a mucosal tear at the cardia (Mallory–Weiss).

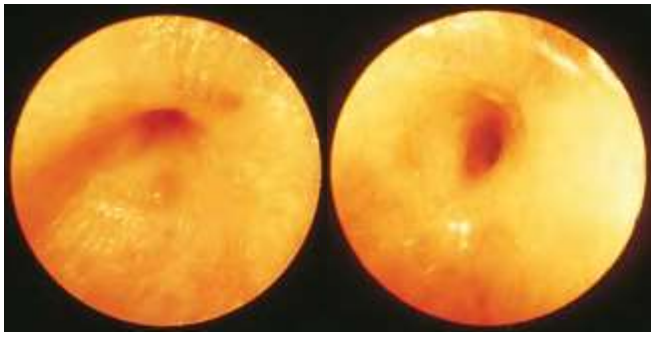


Figure 62.12 The late result of a caustic alkali burn with a high oesophageal stricture.

George Kenneth Mallory, b.1926, Professor of Pathology, Boston University, Boston, MA, USA.
Soma Weiss, 1898–1942, Professor of Medicine, Harvard University Medical School, Boston, MA, USA.

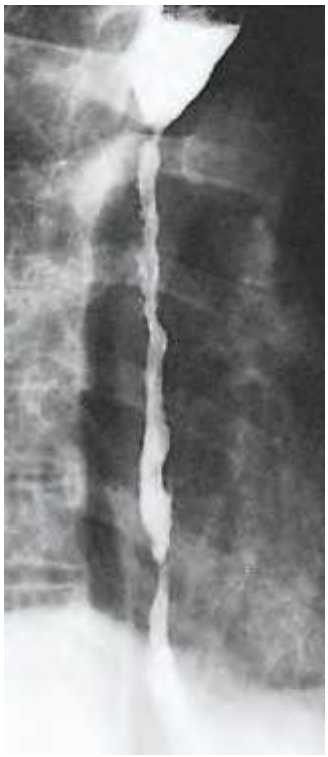


Figure 62.13 Caustic or lye stricture with marked stenosis high in the body of the oesophagus. The strictures are frequently multiple and difficult to dilate unless treated energetically at an early stage.

remain controversial. Other than the need for emergency surgery for bleeding or perforation, elective oesophageal resection should be deferred for at least 3 months until the fibrotic phase has been established. Oesophageal replacement is usually required for very long or multiple strictures. Resection can be difficult because of perioesophageal inflammation in these patients. Because of associated gastric damage, colon may have to be used as the replacement conduit.

There is also controversy with regard to the risk of developing carcinoma in the damaged oesophagus and stomach and how this might influence management. The lifetime risk is certainly less than 5%. Some surgeons advocate resection and replacement, whereas others believe that oesophageal bypass and endoscopic surveillance are preferable, because removal of the badly damaged oesophagus from a scarred mediastinum can be hazardous.

Summary box 62.5

Corrosive injury

- Skilled early endoscopy is mandatory

DRUG-INDUCED INJURY

Many medications, such as antibiotics and potassium preparations, are potentially damaging to the oesophagus, because tablets may remain for a long time, especially if taken without an adequate drink. Acute injury presents with dysphagia and odynophagia, which may be severe. The inflammation usually resolves within 2–3 weeks, and no specific treatment is required apart from appropriate nutritional support. A stricture may follow.

GASTRO-OESOPHAGEAL REFLUX DISEASE

Aetiology

Normal competence of the gastro-oesophageal junction is maintained by the LOS. This is influenced by both its physiological function and its anatomical location relative to the diaphragm and the oesophageal hiatus. In normal circumstances, the LOS transiently relaxes as a coordinated part of swallowing, as a means of allowing vomiting to occur and in response to stretching of the gastric fundus, particularly after a meal to allow swallowed air to be vented. Most episodes of physiological reflux occur during postprandial transient LOS relaxations (TLOSRs). In the early stages of GORD, most pathological reflux occurs as a result of an increased number of TLOSRs rather than a persistent fall in overall sphincter pressure. In more severe GORD, LOS pressure tends to be generally low, and this loss of sphincter function seems to be made worse if there is loss of an adequate length of intra-abdominal oesophagus.

The absence of an intra-abdominal length of oesophagus results in a sliding hiatus hernia. The normal condensation of peritoneal fascia over the lower oesophagus (the phreno-oesophageal ligament) is weak, and the crural opening widens, allowing the upper stomach to slide up through the hiatus. The loss of the normal anatomical configuration exacerbates reflux, although sliding hiatus hernia alone should not be viewed as the cause of reflux. Sliding hiatus hernia is associated with GORD and may make it worse but, as long as the LOS remains competent, pathological GORD does not occur. Many GORD sufferers do not have a hernia, and many of those with a hernia do not have GORD. It should be noted that rolling or paraoesophageal hiatus hernia is a quite different and potentially dangerous condition (see below). A proportion of patients have a rolling hernia and symptomatic GORD or a mixed hernia with both sliding and rolling components. Reflux oesophagitis that is visible endoscopically is a complication of GORD and occurs in a minority of sufferers overall, but in around 40% of patients referred to hospital.

In western societies, GORD is the most common condition affecting the upper GI tract. This is partly due to the declining incidence of peptic ulcer as the incidence of infection with *Helicobacter pylori* has reduced as a result of improved socioeconomic conditions, along with a rising incidence of GORD in the last 30 years. The cause of the increase is unclear, but may be due in part to increasing obesity. The strong association between GORD, obesity and the parallel rise in the incidence of adenocarcinoma of the oesophagus represents a major health challenge for most western countries.

Clinical features

The classic triad of symptoms is retrosternal burning pain (heartburn), epigastric pain (sometimes radiating through to the back) and regurgitation. Most patients do not experience all three. Symptoms are often provoked by food, particularly those that delay gastric emptying (e.g. fats, spicy foods). As the condition becomes more severe, gastric juice may reflux to the mouth and produce an unpleasant taste, often described as 'acid' or 'bitter'. Heartburn and regurgitation can be brought on by stooping or exercise. A proportion of patients have odynophagia with hot beverages, citrus drinks or alcohol. Patients with nocturnal reflux and those who reflux food to the mouth nearly always have severe GORD. Some patients present with less typical symptoms such as angina-like chest pain, pulmonary or laryngeal symptoms. Dysphagia is usually a sign that a stricture has occurred, but may be caused by an associated motility disorder.

As GORD is such a common disorder, it should always be the first thought when a patient presents with oesophageal symptoms that are unusual or that defy diagnosis after a series of investigations.

Diagnosis

In most cases, the diagnosis is assumed rather than proven, and treatment is empirical. Investigation is required only when the diagnosis is in doubt, when the patient does not respond to a proton pump inhibitor (PPI) or if dysphagia is present. The most appropriate examination is endoscopy with biopsy. If the typical appearance of reflux oesophagitis, peptic stricture or Barrett's oesophagus is seen, the diagnosis is clinched, but visible oesophagitis is not always present, even in patients selected as above. This is compounded in clinical practice by the widespread use of PPIs, which cause rapid healing of early mucosal lesions. Many patients will have received such treatment before referral. The endoscopic appearances of the normal oesophagus, hiatus hernia, oesophagitis and stricture are shown in **Figures 62.14–62.20**. It is worth remembering that the correlation between symptoms and endoscopic appearances is poor. On the other hand, there is a strong correlation between worsening endoscopic appearances and the duration of oesophageal acidification on pH testing.

In patients with atypical or persistent symptoms despite therapy, oesophageal manometry and 24-hour oesophageal pH recording (ideally with impedance measurement) may be justified to establish the diagnosis and guide management.

Summary box 62.6

Diagnostic measurement in GORD

- 24-hour pH recording is the 'gold standard' for diagnosis of GORD
- Length and pressure of the LOS are important

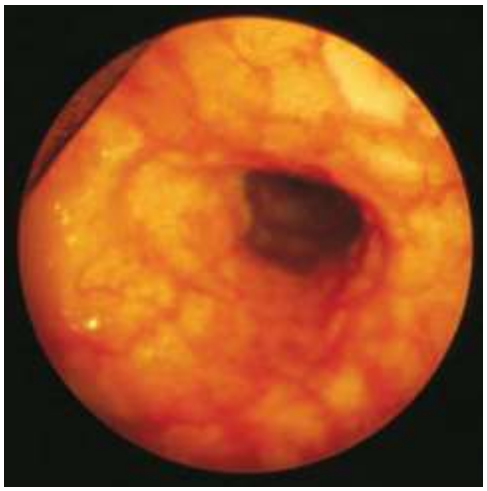


Figure 62.14 The endoscopic appearance of the normal squamous mucosa in the body of the oesophagus.

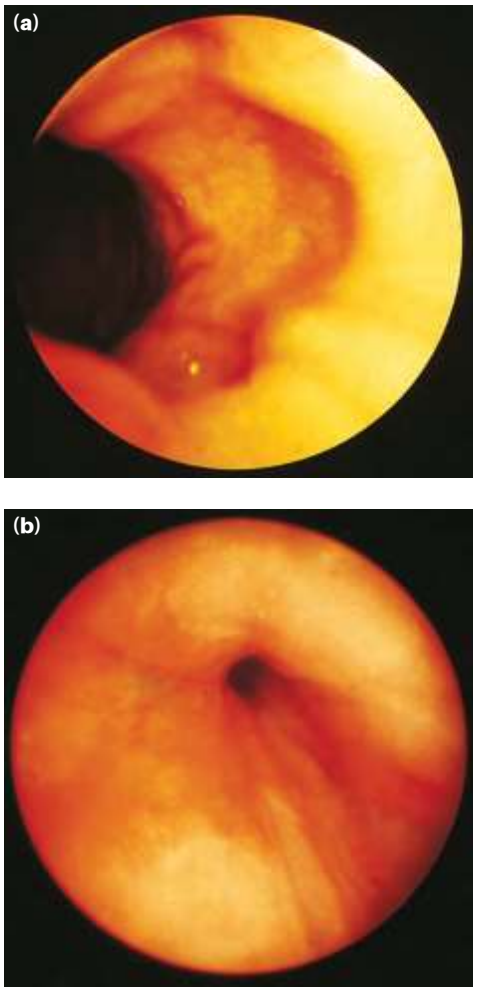


Figure 62.15 The normal lower oesophageal sphincter: **(a)** open; **(b)** closed.

Norman Rupert Barrett, 1903–1979, surgeon, St Thomas' Hospital, London, UK.

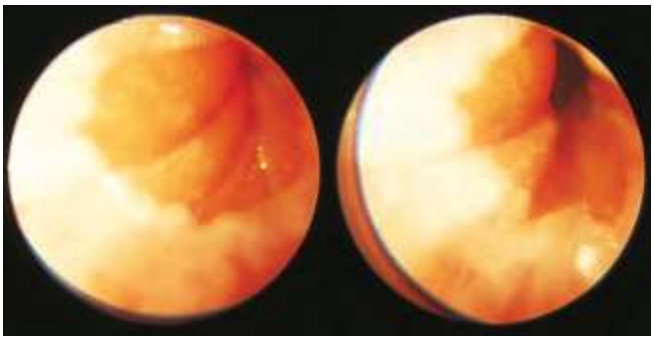


Figure 62.16 The squamocolumnar junction is clearly seen in the lower oesophagus with a normal sharp demarcation.

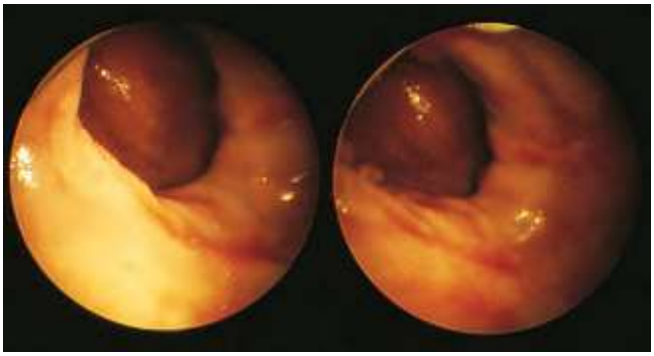


Figure 62.17 Sliding hiatus hernia. The diaphragm can be seen constricting the upper stomach.

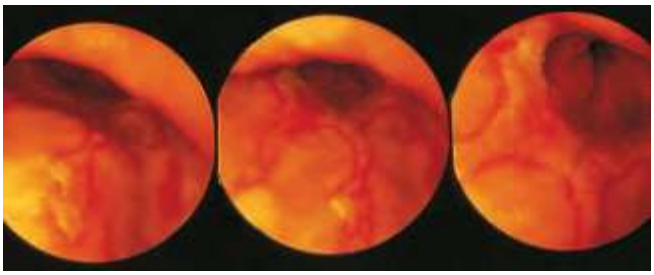


Figure 62.18 Reflux oesophagitis.

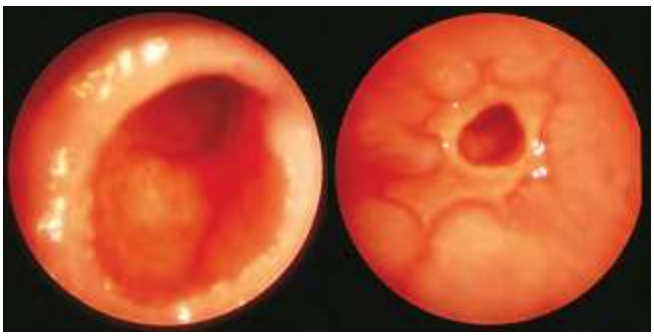


Figure 62.19 Benign stricture with active oesophagitis (left) and healed with columnar epithelium (right).

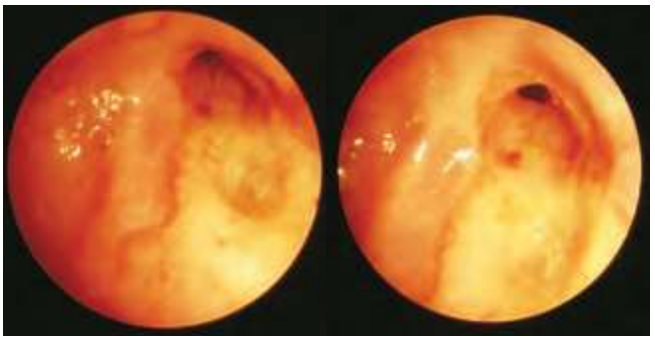


Figure 62.20 Ulceration associated with a benign peptic stricture.

As a matter of routine, proton pump inhibitors (PPIs) are stopped 1 week before oesophageal pH recording, but acid secretion is sometimes reduced for 2 weeks or more, and this can necessitate repeat examination after a prolonged interval without a PPI. Manometry and pH recording are essential in patients being considered for antireflux surgery. Although the main purpose of the test is objectively to quantify the extent of reflux disease, it is also used to rule out a diagnosis of achalasia. In the early stages of achalasia, chest pain can dominate the clinical picture and, when associated with intermittent swallowing problems and non-specific symptoms, it is easy to see how a clinical diagnosis of GORD might be made. Patients with achalasia can also have an abnormal pH study as a result of fermentation of food residue in a dilated oesophagus. Usually, the form of the pH trace is different from that of GORD, with slow undulations of pH rather than rapid bursts of reflux, but the complete absence of peristalsis on manometry is pathognomonic of achalasia.

A CT scan gives the best appreciation of gastro-oesophageal anatomy. This may be important in the context of surgery for rolling or mixed hiatus hernias, but it is unimportant in most patients with GORD.

Management of uncomplicated GORD

Medical management

Most sufferers from GORD do not consult a doctor and do not need to do so. They self-medicate with over-the-counter medicines such as simple antacids, antacid–alginate preparations, H_2-receptor antagonists or PPIs. Consultation is more likely when symptoms are severe, prolonged and unresponsive to the above treatments. Simple measures that are often neglected include advice about weight loss, smoking, excessive consumption of alcohol, tea or coffee, the avoidance of large meals late at night and a modest degree of head-up tilt of the bed. Tilting the bed has been shown to have an effect that is similar to taking an H_2-receptor antagonist. The common practice of using additional pillows has no significant effect.

PPIs are the most effective drug treatment for GORD. Indeed, they are so effective that, once started, patients are

very reluctant to stop taking them. Given an adequate dose for 8 weeks, most patients have a rapid improvement in symptoms (within a few days), and more than 90% can expect full mucosal healing at the end of this time. For this reason, a policy of 'step-down' medical treatment is advocated based on the general advice outlined above and a standard dose of a PPI given for 8 weeks. At the end of that time, the dose of PPI is reduced to that which keeps the patient free of symptoms, and this might even mean the cessation of PPI treatment. As most patients do not make major lifestyle changes and as PPIs are so effective, many remain on long-term treatment. For the minority who do not respond adequately to a standard dose, a trial at an increased dose or the addition of an H_2-receptor antagonist is recommended. If unsuccessful, these patients should be formally investigated.

PPI therapy is also important in patients with reflux-induced strictures, resulting in significant prolongation of the intervals between endoscopic dilatation. As yet, fears that chronic acid suppression might have serious long-term side effects including the risk of gastric cancer seem unwarranted.

Surgery

Strictly speaking, the need for surgery should have been reduced as medication has improved so much. Paradoxically, the number of antireflux operations has remained relatively constant and may even be increasing. This is probably due partly to increased patient expectations and partly to the advent of minimal access surgery, which has improved the acceptability of procedures.

Endoscopic treatments

A number of endoscopic treatments have been tried in the last 10 years that attempt to augment a failing LOS. These involve endoscopic suturing devices that plicate gastric mucosa just below the cardia to accentuate the angle of His, radiofrequency ablation (RFA) to the level of the sphincter and the injection of submucosal polymers into the lower oesophagus. The procedures have generally been applied to patients with only small hiatus hernias or none at all, so only a small proportion of patients who present to hospitals are suitable. Although most methods produce some temporary improvement in symptoms and objective assessments of reflux, failure rates at 1 year are high and can be over 50%. There are few large series that have reported long-term outcomes. Only RFA has been shown to be effective for as long as 10 years when around two-thirds patients remain off PPI medication.

Surgical treatments

The indication for surgery in uncomplicated GORD is essentially patient choice. The risks and possible benefits need to be discussed in detail. Risks include a small mortality rate (0.1–0.5%, depending on patient selection), failed operation (5–10%) and side effects such as dysphagia, gas bloat or abdominal discomfort (10%). With current surgical techniques, 85–90% of patients should be satisfied with the result of an antireflux operation. Patients who are asymptomatic on a PPI need a careful discussion of the risk side of the equation. Those who are symptomatic on a PPI need a careful clinical review to make sure that they will benefit from an operation. Reasons for failure on a PPI include 'volume' reflux (a good indication for surgery), a 'hermit' lifestyle in which the least deviation from lifestyle rules leads to symptoms (a good indication), psychological distress with intolerance of minor symptoms (a poor indication; these patients are likely to be dissatisfied with surgery), poor compliance (a good indication if the reason for poor compliance is the side effects of treatment, otherwise a bad indication) and misdiagnosis of GORD.

WHICH SURGERY?

There are many operations for GORD, but they are virtually all based on the creation of an intra-abdominal segment of oesophagus, crural repair and some form of wrap of the upper stomach (fundoplication) around the intra-abdominal oesophagus. The contribution of each component to operative success is widely debated, but it is clear that operations that fail to address all three components have inferior success rates. The major types of anti-reflux operation were all developed in the 1950s (**Figure 62.21**). When performed correctly, nearly all are effective. Most randomised trials do not show a clear advantage for any one operation over the

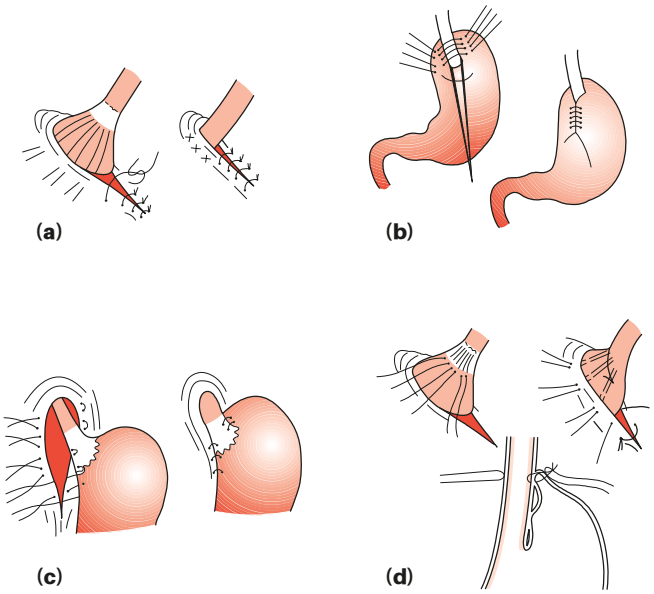


Figure 62.21 Various operations for the surgical correction of gastro-oesophageal reflux disease. **(a)** The original Allison repair of hiatus hernia (this is ineffective and is no longer done); **(b)** Nissen fundoplication; **(c)** Hill procedure; **(d)** Belsey mark IV operation.

Philip Rowland Allison, 1907–1974, Professor of Surgery, Oxford University, Oxford, UK.
Rudolph Nissen, 1896–1981, Professor of Surgery, Istanbul, Turkey, and later at Basle, Switzerland.
Lucius Davis Hill, surgeon, the Mason Clinic, Seattle, MN, USA.
Ronald Herbert Robert Belsey, d.2007, thoracic surgeon, Frenchay Hospital, Bristol, UK.

others, although one meta-analysis has come down in favour of posterior partial fundoplication over total fundoplication when performed laparoscopically.

Total fundoplication (Nissen) tends to be associated with slightly more short-term dysphagia but is the most durable repair in terms of long-term reflux control. Partial fundoplication, whether performed posteriorly (Toupet) or anteriorly (Dor, Watson), has fewer short-term side effects, although this is sometimes at the expense of a slightly higher long-term failure rate. One disadvantage of total fundoplication is the creation of an overcompetent cardia, resulting in the 'gas bloat' syndrome in which belching is impossible. The stomach fills with air, the patient feels very full after small meals and passes excessive flatus. This does not seem to occur with partial fundoplication. The problem has been largely overcome by the 'floppy' Nissen technique in which the fundoplication is loose around the oesophagus and is kept short in length. Although the other short-term side effects of fundoplication usually resolve within 3 months of surgery, this is rarely the case for gas bloat. The problem is best remedied by conversion to a partial fundoplication.

As with primary surgery, various revisional procedures have been described, usually for recurrent reflux or persistent dysphagia. For most patients, recurrent reflux relates to anatomical failure, so the solution is a revisional fundoplication. The results of surgery for recurrent reflux tend to be better than those for dysphagia, because the latter problem has many causes (too tight a wrap, slipping of the wrap, hiatal fibrosis) A very small proportion of patients may undergo more than two operations to correct recurrent reflux or unacceptable side effects. Revisional surgery carries a lower chance of success and, in some patients, local revision becomes technically impossible. The final resort is antrectomy with a Roux-en-Y reconstruction. This reduces gastric acid secretion and diverts bile and pancreatic secretions away from the stomach. Thus, the volume of potential refluxate in the stomach is reduced and, because of its changed composition, it is less damaging to the oesophagus.

For many years, the relative merits of thoracic and abdominal approaches were hotly debated. The introduction of minimal access surgery has made this debate obsolete. Most antireflux operations are now done with a laparoscopic approach.

LAPAROSCOPIC FUNDOPLICATION

Five cannulae are inserted in the upper abdomen (**Figure 62.22**). The cardia and lower oesophagus are separated from the diaphragmatic hiatus. An appropriate length of oesophagus is mobilised in the mediastinum. The fundus may be mobilised by dividing the short gastric vessels that tether the fundus to the spleen, although some surgeons feel that this is unnecessary. The hiatus is narrowed by sutures placed behind the oesophagus. In total (Nissen) fundoplication, the fundus is drawn behind the oesophagus and then sutured to itself in front of the oesophagus (**Figure 62.23a**). In partial fundoplication, the fundus is drawn either behind or in front of the oesophagus and sutured to it on each side, leaving a strip of exposed oesophagus either at the front (**Figure 62.23b**) or at the back. Robotic fundoplication has been described but comparisons with laparoscopic surgery do not indicate any clear clinical benefits.

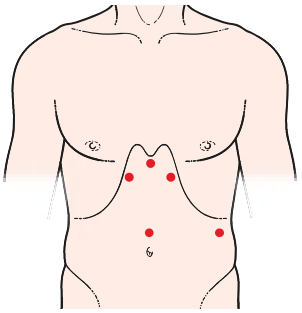


Figure 62.22 Laparoscope cannula sites for laparoscopic fundoplication.

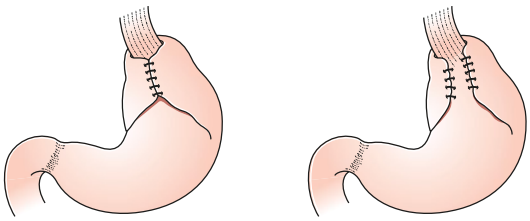


Figure 62.23 **(a)** Total (Nissen) fundoplication; **(b)** partial fundoplication (Toupet).

Complications of GORD

Stricture

Reflux-induced strictures (see **Figure 62.20**) occur mainly in late middle-aged and elderly people, but they may present in children. It is important to distinguish a benign reflux-induced stricture from a carcinoma. This is not usually difficult on the basis of location (immediately above the oesophagogastric junction), length (only about 1–2 cm) and smooth mucosa, but sometimes a cancer spreads under the oesophageal mucosa at its upper margin, producing a benign-looking stricture.

Peptic strictures generally respond well to dilatation and long-term treatment with a PPI. As most patients are elderly, antireflux surgery is not usually considered. However, it is an alternative to long-term PPI treatment, just as in uncomplicated GORD in younger and fitter patients. Most patients do not require anything other than a standard operation.

Summary box 62.7

Peptic stricture

- Day-case dilatation and PPI for peptic stricture

AM Toupet, French surgeon.
Cesar Roux, 1857–1934, Professor of Surgery and Gynaecology, Lausanne, Switzerland, described this method of forming a jejunal conduit in 1908.

Oesophageal shortening

The issue of oesophageal shortening continues to provoke debate. There can be no doubt that, in the presence of a large sliding hiatus hernia, the oesophagus is short, but this does not necessarily mean that, with mobilisation from the mediastinum, it cannot easily be restored to its normal length. The extent to which severe inflammation in the wall of the oesophagus causes fibrosis and real shortening is less clear. If a good segment of intra-abdominal oesophagus cannot be restored without tension, a Collis gastroplasty should be performed (**Figure 62.24**). This produces a neo-oesophagus around which a fundoplication can be done (Collis–Nissen operation).

Summary box 62.8

GORD

- Is due to loss of competence of the LOS and is extremely common
- May be associated with a hiatus hernia, which may be sliding or, less commonly, rolling (paraoesophageal)
- The most common symptoms are heartburn, epigastric discomfort and regurgitation, often made worse by stooping and lying
- Achalasia and GORD are diagnostically easily confused
  - Dysphagia may occur, but a neoplasm must be excluded
  - Diagnosis and treatment can be instituted on clinical grounds
- Endoscopy may be required and 24-hour pH is the 'gold standard'
  - Management is primarily medical (PPIs being the most effective), but surgery may be required; laparoscopic fundoplication is the most popular technique
  - Stricture may develop in time

Barrett's oesophagus (columnar-lined lower oesophagus)

Barrett's oesophagus is a metaplastic change in the lining mucosa of the oesophagus in response to chronic gastro-oesophageal reflux (**Figure 62.25**). Many of these patients do not have particularly severe symptoms, although they do have the most abnormal pH profiles. This adaptive response involves a mosaic of cell types, probably beginning as a simple columnar epithelium which becomes 'specialised' with time. The hallmark of 'specialised' Barrett's epithelium is the presence of mucus-secreting goblet cells (intestinal metaplasia). One of the great mysteries of GORD is why some people develop oesophagitis and others develop Barrett's oesophagus, often without significant oesophagitis. In Barrett's oesophagus, the junction between squamous oesophageal mucosa and gastric mucosa moves proximally. It may be difficult to distinguish a Barrett's oesophagus from a tubular, sliding hiatus hernia during endoscopy, as the two often coexist (**Figure 62.26**) or where the visible Barrett's segment is very short. The key

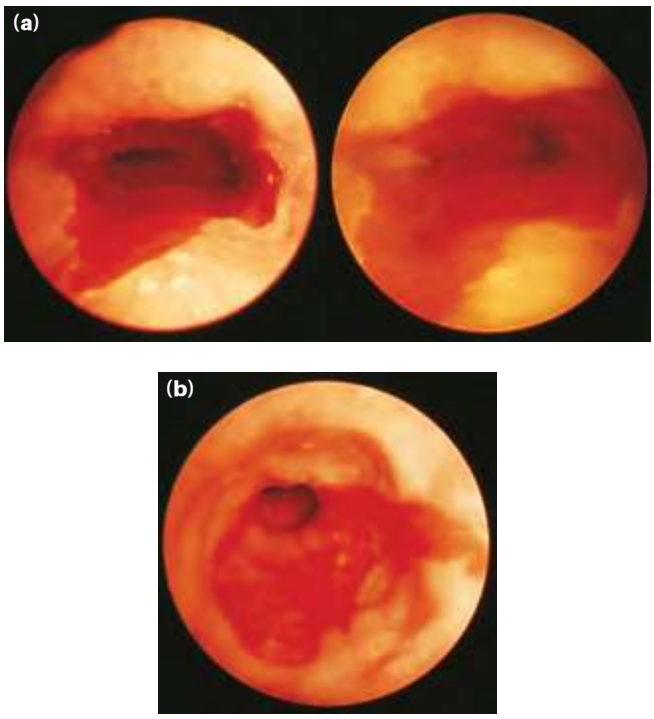


Figure 62.25 Barrett's oesophagus with **(a)** proximal migration of the squamocolumnar junction and **(b)** a view of the distal oesophagus.

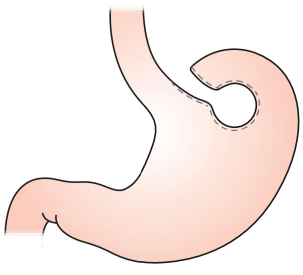


Figure 62.24 Collis gastroplasty to produce a neo-oesophagus around which a Nissen fundoplication is done. The operation may be performed by a laparoscopic as well as an open approach, using circular and linear staplers.

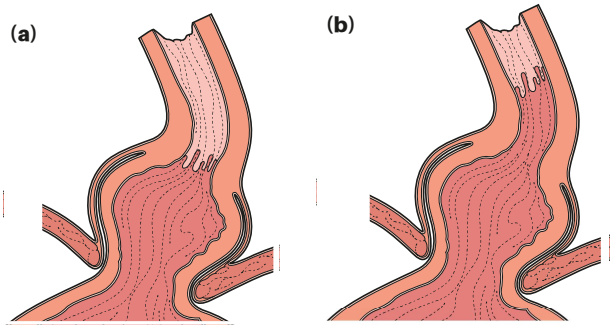


Figure 62.26 **(a)** The interrelationship of the lower oesophageal sphincter, the squamocolumnar junction and the diaphragm in sliding hiatus hernia. **(b)** Barrett's oesophagus and sliding hernia.

John Leigh Collis, 1911–2003, Professor of Thoracic Surgery, the University of Birmingham, Birmingham, UK.

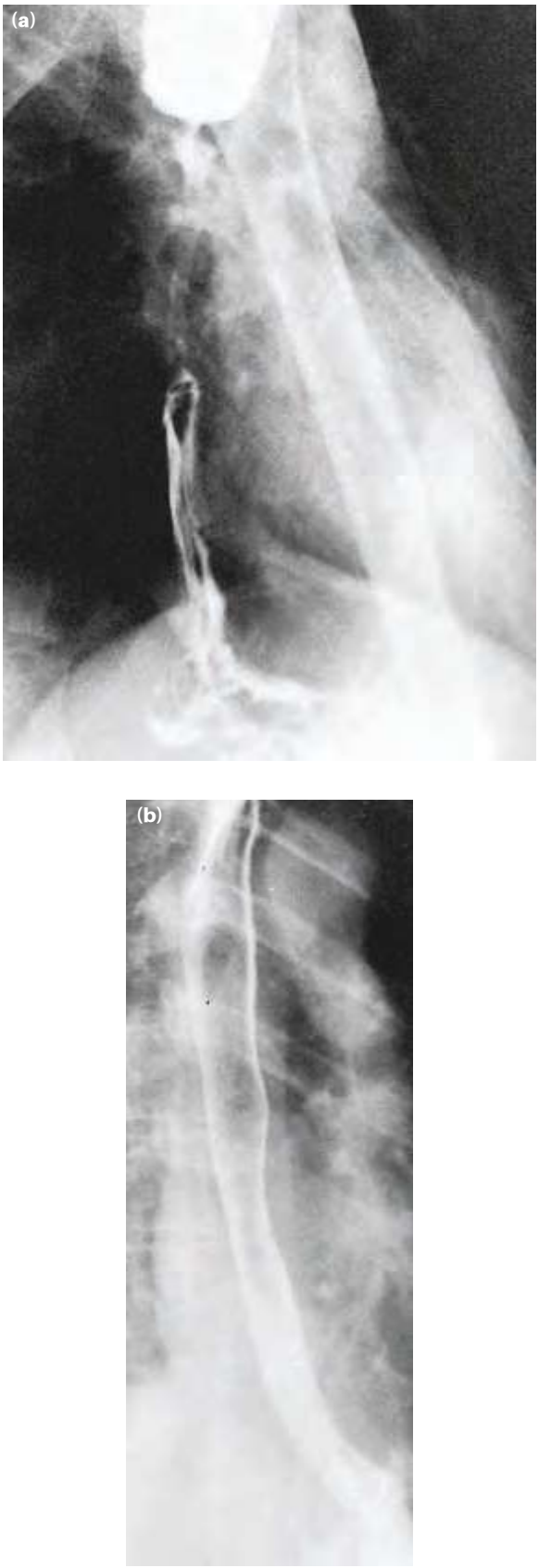


Figure 62.27 The radiological appearance of a midoesophageal stricture in **(a)**, a patient with Barrett's oesophagus and in **(b)** a normal lumen after dilatation.

is where the gastric mucosal folds end along with recognition of the mucosal vascular pattern. The mucosa in the body of the stomach has longitudinal folds; the columnar lining of Barrett's oesophagus is smooth. The lower oesophagus is characterised by palisade vessels that run longitudinally and are easily seen through the lower oesophageal mucosa. Strictures can occur in Barrett's oesophagus and nearly always appear at the new squamocolumnar junction **(Figure 62.27)**. Rarely, a stricture may occur in the columnar segment after healing of a Barrett's ulcer **(Figure 62.28)**. Although intestinal metaplasia and length of the Barrett's segment are important risk factors for the development of carcinoma, neither represents an essential feature for cancer development. The risk of transformation to cancer is probably no more than 0.5% per patient per year, which is about 25 times that of the general population **(Figures 62.29 and 62.30)**.

Summary box 62.9

Barrett's oesophagus

- Intestinal metaplasia is a risk factor for the development of adenocarcinoma
- Do not confuse Barrett's ulcer with oesophagitis

Patients who are found to have Barrett's oesophagus may be submitted to regular surveillance endoscopy with multiple biopsies in the hope of finding dysplasia or in situ cancer, rather than allowing invasive cancer to develop and cause symptoms. There is no general agreement about the benefits of surveillance endoscopy, or its ideal frequency. Endoscopy at 2-year intervals is probably adequate, provided that no dysplasia has been detected. A significant problem is that the incidence of Barrett's oesophagus in the community is

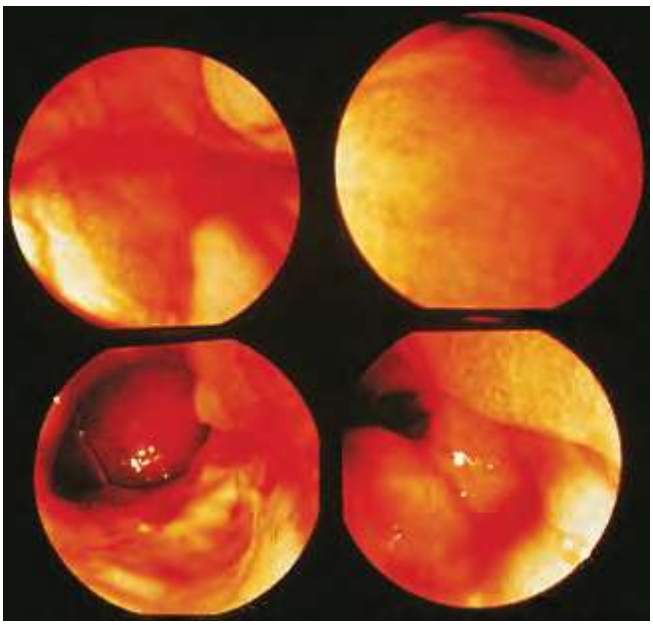


Figure 62.28 Barrett's ulcer in the columnar cell-lined oesophagus.

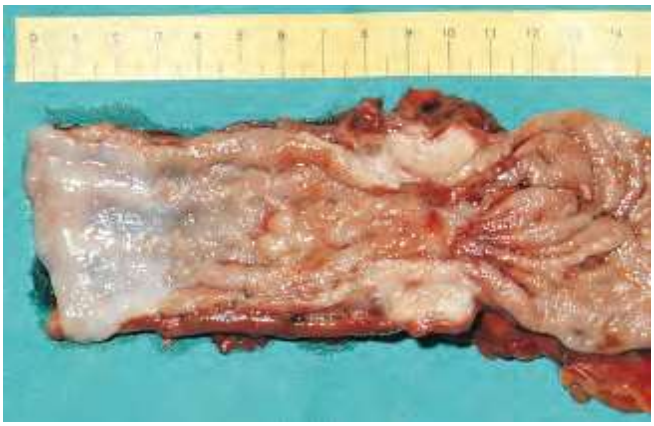


Figure 62.29 The macroscopic appearances of an adenocarcinoma in Barrett's oesophagus.

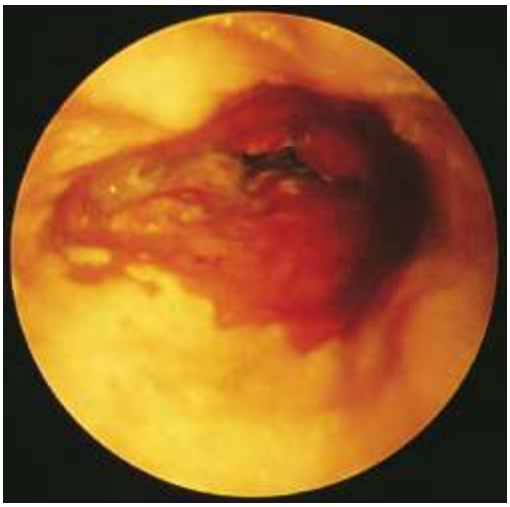


Figure 62.30 Endoscopic view of carcinoma in Barrett's oesophagus.

estimated to be at least 10 times the incidence discovered in dyspeptic patients referred for endoscopy. Thus, adenocarcinoma in Barrett's oesophagus often presents with invasive cancer without any preceding reflux symptoms.

Until recently, Barrett's oesophagus was not diagnosed until there was at least 3 cm of columnar epithelium in the distal oesophagus. With the better appreciation of the importance of intestinal metaplasia, Barrett's oesophagus may be diagnosed if there is any intestinal metaplasia in the oesophagus. The relative risk of cancer rises with increasing length of abnormal mucosa. The following terms are widely used:

- classic Barrett's (≥3 cm columnar epithelium);
- short-segment Barrett's (<3 cm of columnar epithelium);
- cardia metaplasia (intestinal metaplasia at the oesophagogastric junction without any macroscopic change at endoscopy).

When Barrett's oesophagus is discovered, the treatment is that of the underlying GORD. There has been considerable interest in recent years in endoscopic methods of treating Barrett's mucosa in the hope of eliminating the risk of cancer development. Laser, photodynamic therapy, argon-beam plasma coagulation, RFA and endoscopic mucosal resection (EMR) have all been used. Until recently, it was felt that intervention should only be offered in patients with high-grade dysplasia, but recent evidence suggests that patients with low-grade dysplasia also have a substantial risk of progressing to high-grade dysplasia of around 9% per year. For this reason endoscopic treatment is considered appropriate for patients with low-grade dysplasia when confirmed by a second independent histopathologist. EMR of dysplastic areas followed by RFA seems to be the most popular approach and, together with PPI treatment, these endoscopic methods can result in a neosquamous lining. Fears of buried glands that might give rise to cancer seem unfounded.

PARAOESOPHAGEAL ('ROLLING') HIATUS HERNIA

True paraoesophageal hernias in which the cardia remains in its normal anatomical position are rare. The vast majority of rolling hernias are mixed hernias in which the cardia is displaced into the chest and the greater curve of the stomach rolls into the mediastinum (**Figure 62.31**). Sometimes, the whole of the stomach lies in the chest (**Figure 62.32**). Colon or small intestine may sometimes lie in the hernia sac. The hernia is most common in elderly people, but may occur in young fit people. As the stomach rolls up into the chest, there is always an element of rotation (volvulus).

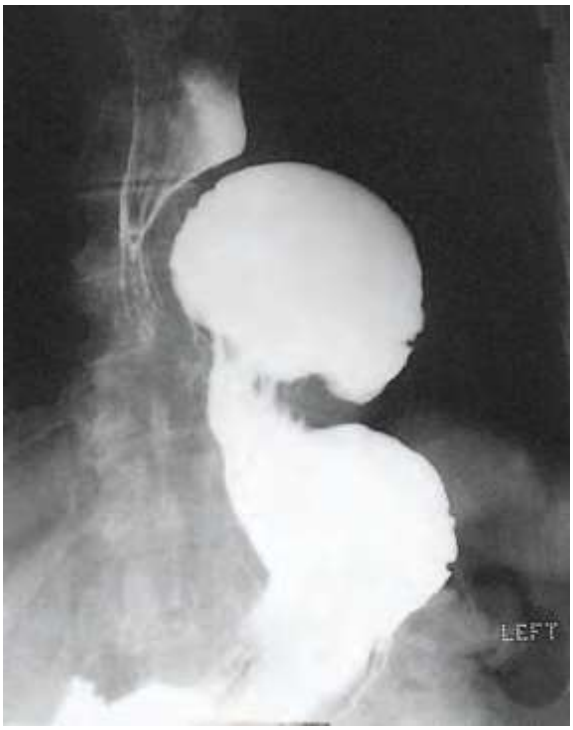


Figure 62.31 A paraoesophageal hernia showing the gastro-oesophageal junction just above the diaphragm and the fundus alongside the oesophagus, compressing the lumen.

Figure 62.32 A huge paraoesophageal hernia with an upside-down stomach and the pylorus just below the hiatus.

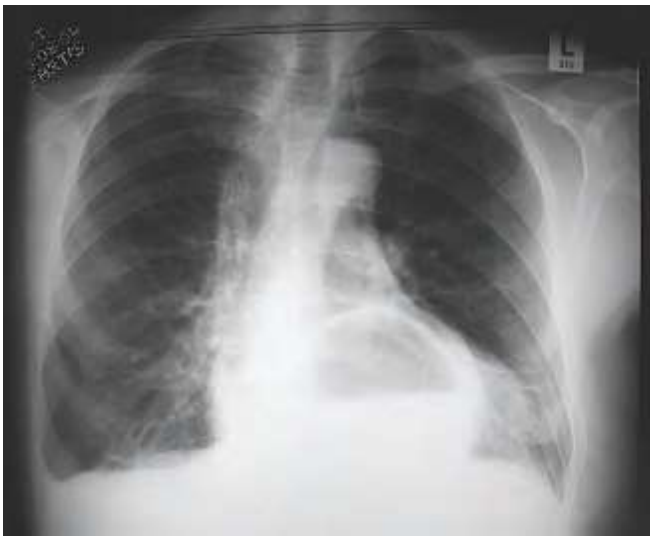


Figure 62.33 A gas bubble seen on a plain chest radiograph, showing the fundus of the stomach in the chest (courtesy of Dr Stephen Ellis, Bart's and the London NHS Trust).

Summary box 62.10

'Rolling' hiatus hernia

- Potentially dangerous, because of volvulus

The symptoms of rolling hernia are mostly due to twisting and distortion of the oesophagus and stomach. Dysphagia is common. Chest pain may occur from distension of an obstructed stomach. Classically, the pain is relieved by a loud belch. Symptoms of GORD are variable. Strangulation, gastric perforation and gangrene can occur. Emergency presentation with any of these complications carries high mortality on account of a combination of late diagnosis, generally elderly patients with comorbid diseases and the complexity of surgery involved.

The hernia may be visible on a plain radiograph of the chest as a gas bubble, often with a fluid level behind the heart (**Figure 62.33**). A CT scan with oral contrast is the best method of diagnosis, highlighting the gastric anatomy but also identifying other structures involved in the hernia. The endoscopic appearances may be confusing, especially in large hernias when it is easy to become disoriented.

Symptomatic rolling hernias nearly always require surgical repair because they are potentially dangerous. The risk of an asymptomatic patient developing a significant problem when a rolling hiatus hernia is discovered incidentally has probably been underestimated in the past. The annual risk is probably greater than the historical estimate of 1%. Patients who present as an emergency with acute chest pain may be treated initially by nasogastric tube, to relieve the distension that causes the pain, followed by surgical repair. Endoscopy is useful if nasogastric intubation is unsuccessful. If the pain is not relieved or perforation is suspected, immediate operation is mandatory.

Emergency surgery needs to be tailored to the problem encountered and the fitness of the patient. Elective surgery involves reduction of the hernia, excision of the sac, reduction of the crural defect and some form of retention of the stomach in the abdomen. Some surgeons perform a fundoplication, arguing that this is a very effective means of maintaining reduction and that it deals with the associated GORD. Others argue that fundoplication should be done only if reflux can be conclusively demonstrated beforehand. Surprisingly, both philosophies achieve good results. Laparoscopic repair has recently become popular. Full anatomical repair of a large rolling hernia can be difficult using this approach and requires considerable expertise. Secure closure of the hiatal defect can be a problem, and some surgeons advocate mesh to reinforce the repair.

NEOPLASMS OF THE OESOPHAGUS

Benign tumours

Benign tumours of the oesophagus are relatively rare. True papillomas, adenomas and hyperplastic polyps do occur, but most 'benign' tumours are not epithelial in origin and arise from other layers of the oesophageal wall (GI stromal tumour [GIST], lipoma, granular cell tumour). Most benign oesophageal tumours are small and asymptomatic, and even a large benign tumour may cause only mild symptoms (**Figure 62.34**). The most important point in their management is usually to carry out an adequate number of biopsies to prove beyond reasonable doubt that the lesion is not malignant (**Figure 62.35**).

Malignant tumours

Non-epithelial primary malignancies are also rare, as is malignant melanoma. Secondary malignancies rarely involve the oesophagus, with the exception of bronchogenic carcinoma

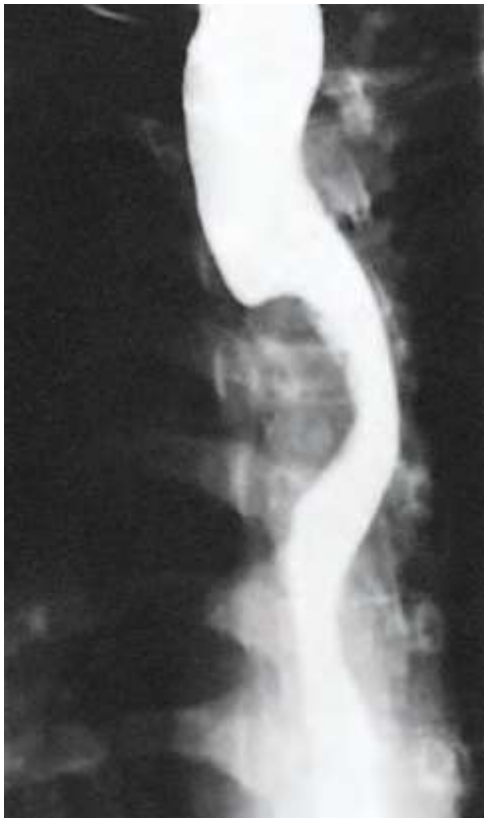


Figure 62.34 Classic appearance of a large oesophageal gastrointestinal stromal tumour on barium swallow.

by direct invasion of either the primary and/or contiguous lymph nodes.

Carcinoma of the oesophagus

Cancer of the oesophagus is the sixth most common cancer in the world. In general, it is a disease of mid to late adulthood, with a poor survival rate. Only 5–10% of those diagnosed will survive for 5 years.

Summary box 62.11

Carcinoma of the oesophagus

- Squamous cell usually affects the upper two-thirds; adenocarcinoma usually affects the lower third
- Common aetiological factors are tobacco and alcohol (squamous cell), GORD and obesity (adenocarcinoma)
- The incidence of adenocarcinoma is increasing
- Lymph node involvement is a bad prognostic factor
- Dysphagia is the most common presenting symptom, but is a late feature
- Accurate pretreatment staging is essential in patients thought to be fit to undergo 'curative' treatment

Pathology and aetiology

Squamous cell cancer (**Figures 62.36 and 62.37**) and adenocarcinoma (**Figures 62.38 and 62.39**) are the most common types. Squamous cell carcinoma generally affects the upper

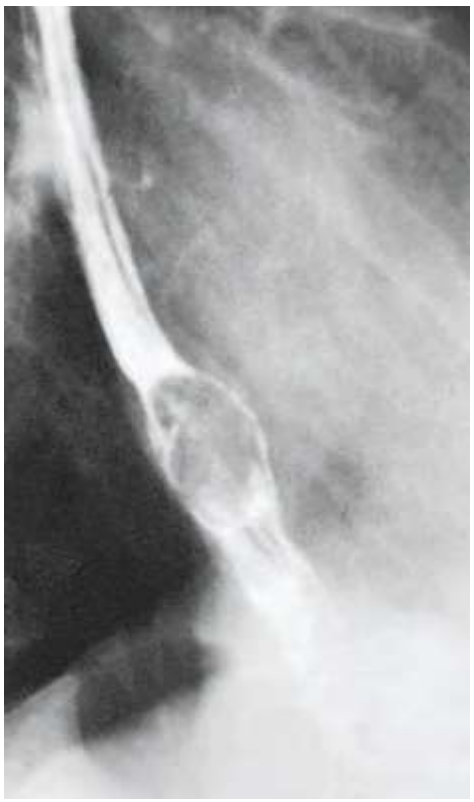


Figure 62.35 An intraluminal polyp that proved to be a leiomyosarcoma.

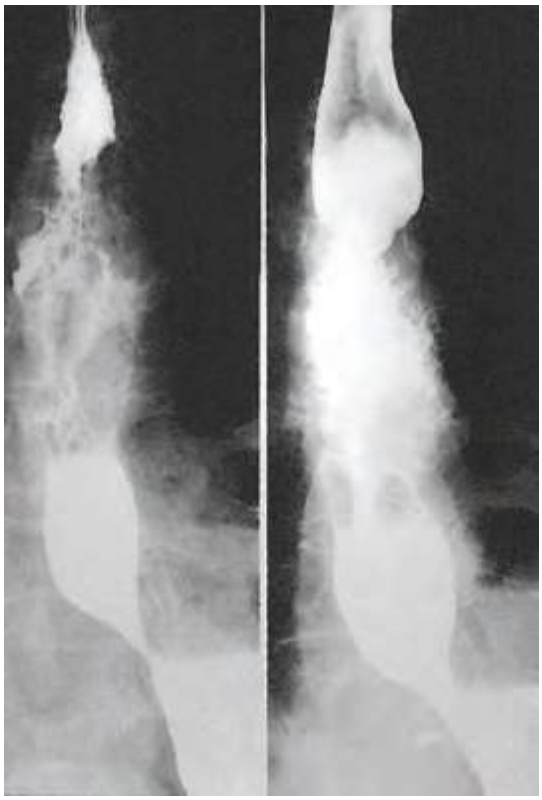


Figure 62.36 The classic appearances of a midoesophageal proliferative squamous cell carcinoma.

Figure 62.37 Squamous cell carcinoma of the oesophagus producing an irregular stricture with shouldered margins.

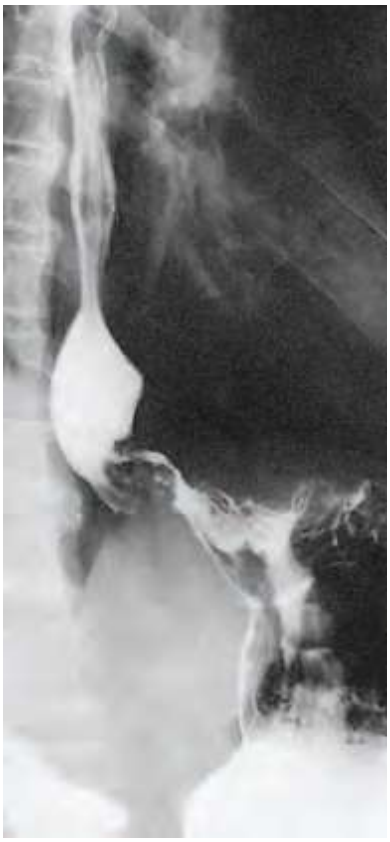


Figure 62.38 Adenocarcinoma of the lower oesophagus, spreading upwards from the cardia.

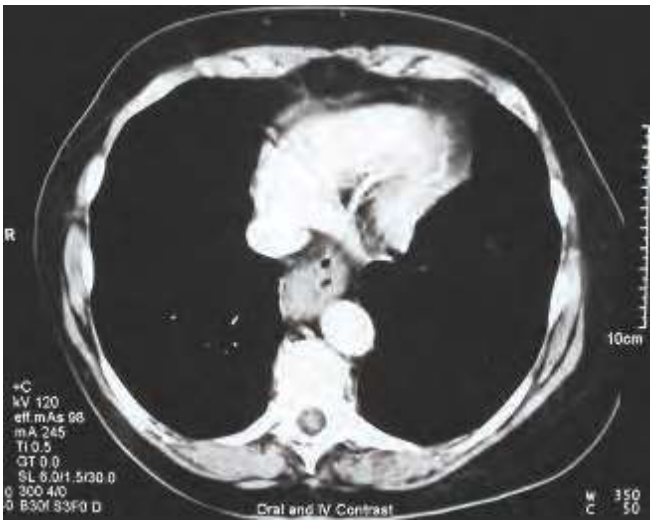


Figure 62.39 Computed tomography scan showing a primary tumour of the lower oesophagus.

two-thirds of the oesophagus and adenocarcinoma the lower third. Worldwide, squamous cell cancer is most common, but adenocarcinoma predominates in the west and is increasing in incidence.

Geographical variation in oesophageal cancer

The incidence of oesophageal cancer varies more than that of any other cancer. Squamous cell cancer is endemic in the Transkei region of South Africa and in the Asian 'cancer belt', which extends across the middle of Asia from the shores of the Caspian Sea (in northern Iran) to China. The highest incidence in the world is in Linxian in Henan province in China, where it is the most common single cause of death, with more than 100 cases per 100 000 population per annum. The cause of the disease in the endemic areas is not known. Although there is evidence of genetic susceptibility across much of central Asia, a variety of environmental factors along with nutritional deficiencies are probably involved. In Linxian, supplementation of the diet with β-carotene, vitamin E and selenium has been shown to reduce the incidence.

Away from the endemic areas, tobacco and alcohol are major factors in the occurrence of squamous cancer. Incidence rates vary from less than 5:100 000 in white people in the USA to 26.5:100 000 in some regions of France.

In many western countries, the incidence of squamous cell cancer has fallen or remained static, but the incidence of adenocarcinoma of the oesophagus has increased dramatically since the mid-1970s at a rate of 5–10% per annum.

The change is greater than that of any other neoplasm in this time. Adenocarcinoma now accounts for 60–75% of all oesophageal cancers in several countries. The reason for this change is not understood. A similar rate of increase in GORD over the same period, which mirrors an increase in obesity in the west, is likely to be an important factor, particularly through the link to Barrett's oesophagus. Obese, white men in their 60s represent the highest-risk group. There has also been an increase in the incidence of carcinoma of the cardia of the stomach, which suggests that cancer of the cardia and adenocarcinoma of the oesophagus may share common aetiological factors. With a falling incidence of cancer in the rest of the stomach, more than 60% of all upper GI cancers in the west involve the cardia or distal oesophagus.

Both adenocarcinomas and squamous cell carcinomas tend to disseminate early. Sadly, the classic presenting symptoms of dysphagia, regurgitation and weight loss are often absent until the primary tumour has become advanced, and so the tumour is often well established before the diagnosis is made. Tumours can spread in three ways: invasion directly through the oesophageal wall, via the lymphatics or in the bloodstream. Direct spread occurs both laterally, through the component layers of the oesophageal wall, and longitudinally within the oesophageal wall. Longitudinal spread is mainly via the submucosal lymphatic channels of the oesophagus. The pattern of lymphatic drainage is therefore not segmental, as in other parts of the GI tract. Consequently, the length of oesophagus involved by tumour is frequently much longer than the macroscopic length of the malignancy at the epithelial surface. Lymph node spread occurs commonly. Although the direction of spread to regional lymphatics is predominantly caudal, the involvement of lymph nodes is potentially widespread and can also occur in a cranial direction. Any regional lymph node from the superior mediastinum to the coeliac axis and lesser curve of the stomach may be involved, regardless of the location of the primary lesion within the oesophagus. Haematogenous spread may involve a variety of different organs including the liver, lungs, brain and bones. Tumours arising from the intra-abdominal portion of the oesophagus may also disseminate transperitoneally.

Clinical features

Most oesophageal neoplasms present with mechanical symptoms, principally dysphagia, but sometimes also regurgitation, vomiting, odynophagia and weight loss. Clinical findings suggestive of advanced malignancy include recurrent laryngeal nerve palsy, Horner's syndrome, chronic spinal pain and diaphragmatic paralysis. Other factors making surgical cure unlikely include weight loss of more than 20% and loss of appetite. Cutaneous tumour metastases or enlarged supraclavicular lymph nodes may be seen on clinical examination and indicate disseminated disease. Hoarseness due to recurrent laryngeal nerve palsy is a sign of advanced and incurable disease. Palpable lymphadenopathy in the neck is likewise a sign of advanced disease.

Patients with early disease may have non-specific dyspeptic symptoms or a vague feeling of 'something that is not quite right' during swallowing. Some are diagnosed during endoscopic surveillance of patients with Barrett's oesophagus and, although this does identify patients with the earliest stages of disease, such programmes have little overall impact, because most patients with Barrett's oesophagus are unknown to the medical profession and make their first presentation with a symptomatic, and therefore usually locally advanced, oesophageal cancer. The widespread use of endoscopy as a diagnostic tool does, nevertheless, provide an opportunity for early diagnosis (**Figure 62.40**). Biopsies should be taken of all lesions in the oesophagus (**Figures 62.41 and 62.42**), no matter how trivial they appear and irrespective of the indication for the examination.

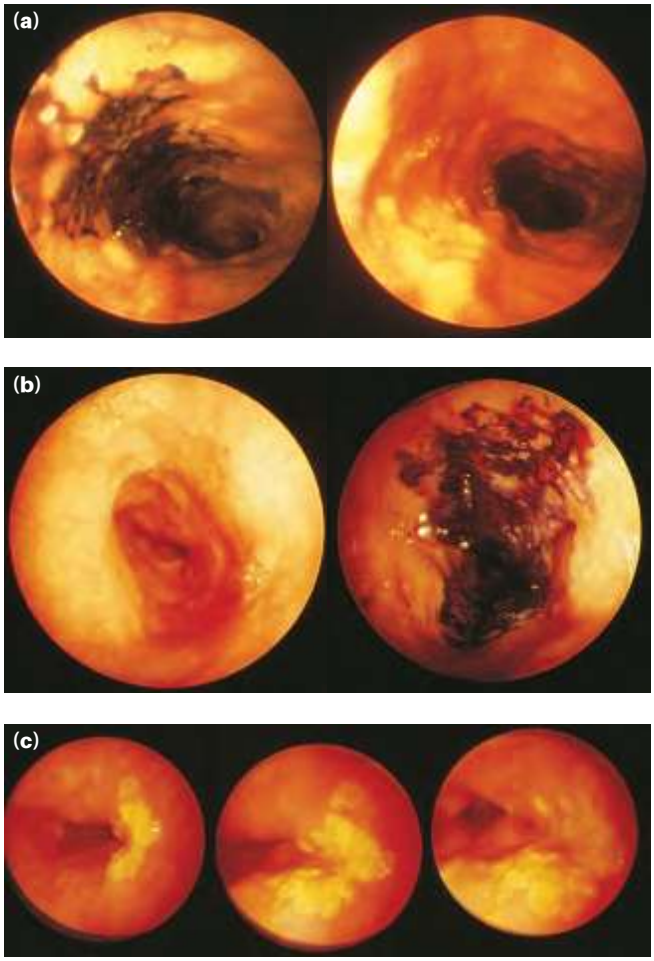


Figure 62.40 Carcinoma *in situ* showing the varied presentations: **(a)** occult form; **(b)** erythroplakia; **(c)** leukoplakia. The right-hand pictures in **(a)** and **(b)** demonstrate the use of vital staining with methylene blue.

Johann Friedrich Horner, 1831–1886, Professor of Ophthalmology, Zurich, Switzerland, described this syndrome in 1869.

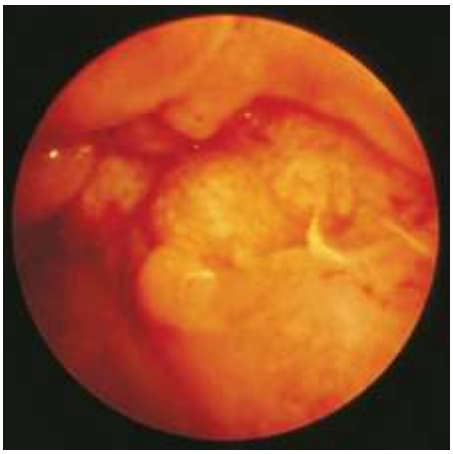


Figure 62.41 Endoscopic appearances of a midoesophageal squamous cell carcinoma.

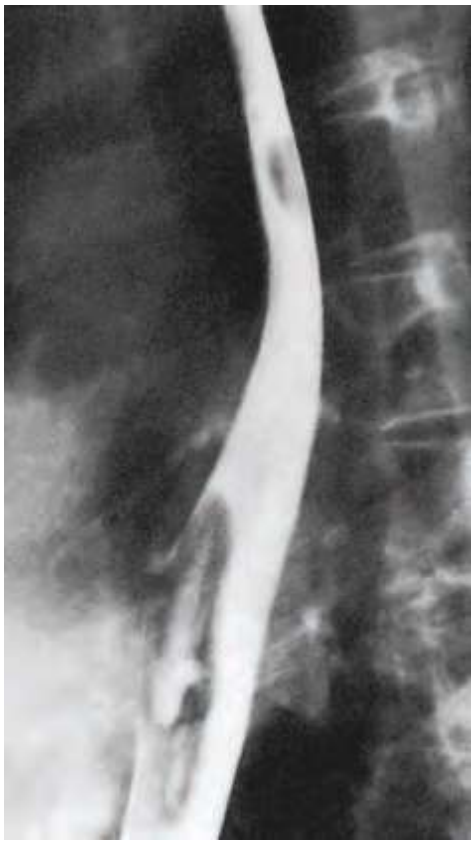


Figure 62.42 Beware the differential diagnosis of infection, for what appears to be a tumour. This midoesophageal mass was actually tuberculosis.

Investigation

Endoscopy is the first-line investigation for most patients. It provides an unrivalled direct view of the oesophageal mucosa and any lesion allowing its site and size to be documented. Cytology and/or histology specimens taken via the endoscope are crucial for accurate diagnosis. The combination of histology and cytology increases the diagnostic accuracy to more than 95%. The chief limitation of conventional endoscopy is that only the mucosal surface can be studied and biopsied. Other investigations are therefore usually required to define the extent of local or distant spread. The improved image resolution of modern endoscopes and novel techniques involving magnification and the use of dyes to enhance surface detail may lead to more early lesions being recognised.

GENERAL ASSESSMENT AND STAGING

Once the initial diagnosis of a malignant oesophageal neoplasm has been made, patients should be assessed first in terms of their general health and fitness for potential therapies. Their preferences should also be considered. Most potentially curative therapies include radical surgery, although definitive chemoradiotherapy is an alternative in squamous cell carcinoma. Patients who are unfit for, or who do not wish to contemplate, radical treatments should not be investigated further, but should be diverted to appropriate palliative therapies, depending on the symptoms and current quality of life. Only those patients suitable for potentially curative therapies should proceed to staging investigations to rule out haematogenous spread (CT scan) and then to assess locoregional stage (endoscopic ultrasonography [EUS] ± laparoscopy). This will distinguish between early (T1/T2, N0) and advanced lesions (T3/T4, N1) and indicate whether surgery alone or multimodal therapy is most appropriate. Where attempted cure is deemed possible, the aim should be to provide the best chance of cure while minimising procedural risks. In general, surgery alone should be reserved for patients with early disease, and multimodal therapy should be used in patients with locally advanced disease, in whom the chance of cure by surgery alone is small (generally <20%).

The most widely used pathological staging system is that of the World Health Organization (tumour–nodes–metastasis TNM).

Table 62.2 shows the TNM system for oesophageal cancer. Similar to all pathological systems, it relies on the nature and extent of the surgery performed, e.g. performing more

TABLE 62.2 TNM staging scheme for oesophageal cancer.

Tis	High-grade dysplasia
T1	Tumour invading lamina propria or submucosa
T2	Tumour invading muscularis propria
T3	Tumour invading beyond muscularis propria
T4a	Tumour invading adjacent structures (pleura, pericardium, diaphragm)
T4b	Tumour invading adjacent structures (trachea, bone, aorta)
N0	No lymph node metastases
N1	Lymph node metastases in 1–2 nodes
N2	Lymph nodes metastases in 3–6 nodes
N3	Lymph node metastases in 7 or more lymph nodes
M0	No distant metastases
M1	All other distant metastases
Stage	1A: T1N0M0; 1B: T2N0M0; 2A: T3N0M0; 2B: T1/2N0M0; 3A: T4aN0M0, T3N1M0, T1/2N2M0
Stage	3B: T3N2M0; 3C: T4aN1/2M0, T4bN0–3M0, T1–4N3M0; 4T:1–4N1–3M1

extensive radical surgical lymphadenectomy provides a more accurate assessment of the 'N' stage. There is evidence that many patients described as N0 in the past were probably N1, a phenomenon described as stage migration.

Staging information may be gathered before the commencement of therapy, during therapy (e.g. at open operation) or following treatment (histology or postmortem examination). The techniques commonly used to provide preoperative staging data are described in Figure 62.43, along with a suggested algorithm.

BLOOD TESTS

These are of limited value. Blood tests reveal nothing about local invasion or regional lymph node spread and, to date, no reliable tumour marker for oesophageal cancer has been isolated from peripheral blood. The presence of abnormal liver function tests (LFTs) may suggest the presence of liver metastases, but this is generally too insensitive to be diagnostic. Many patients with known liver metastases have normal LFTs. At best, abnormal LFTs only reinforce the clinical suspicion of spread to the liver, and further imaging is usually required to confirm the diagnosis.

TRANSCUTANEOUS ULTRASONOGRAPHY

It is difficult to visualise mediastinal structures with transcutaneous ultrasonography. With the relatively low-frequency sound waves used, good depth of tissue penetration is achieved at the expense of poor image resolution. In addition, the mediastinal organs are surrounded by bone and air, which renders them largely inaccessible to external ultrasound. The technique is therefore used mainly to assess spread to the liver, the whole of which can be clearly visualised by standard transcutaneous ultrasonography. Haematogenous spread can be more fully assessed by combining ultrasonography with chest radiography, although this combination is less accurate than CT scanning.

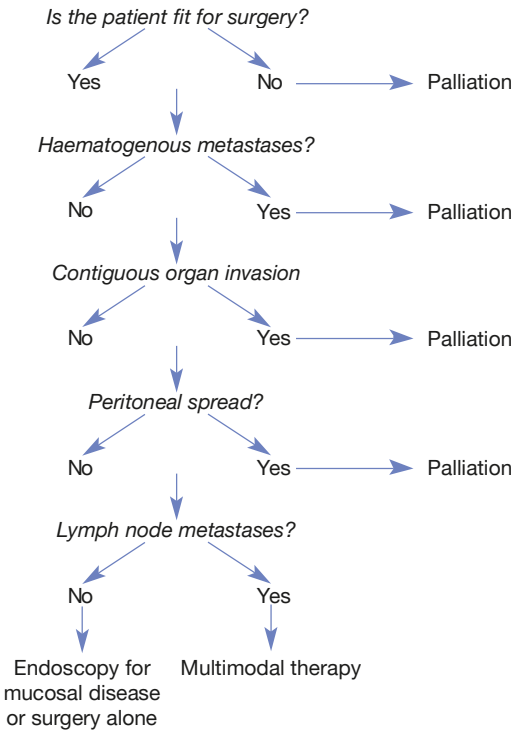


Figure 62.43 Algorithm for the management of oesophageal cancer.

BRONCHOSCOPY

Many middle- and upper-third oesophageal carcinomas (and therefore usually squamous carcinomas) are sufficiently advanced at the time of diagnosis that the trachea or bronchi are already involved (Figure 62.44). Bronchoscopy may reveal either impingement or invasion of the main airways in over 30% of new patients with cancers in the upper third of the oesophagus. In some cases, therefore, bronchoscopy alone can confirm that the tumour is locally unresectable.

LAPAROSCOPY

This is a useful technique for the diagnosis of intra-abdominal and hepatic metastases. It has the advantage of enabling tissue samples or peritoneal cytology to be obtained and is the only modality reliably able to detect peritoneal tumour seedlings (Figure 62.45). This is particularly important for tumours arising from the intra-abdominal portion of the oesophagus,

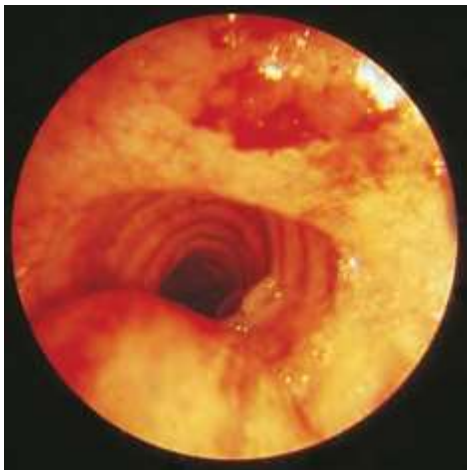


Figure 62.44 Invasion into the posterior wall of the trachea from an oesophageal carcinoma.

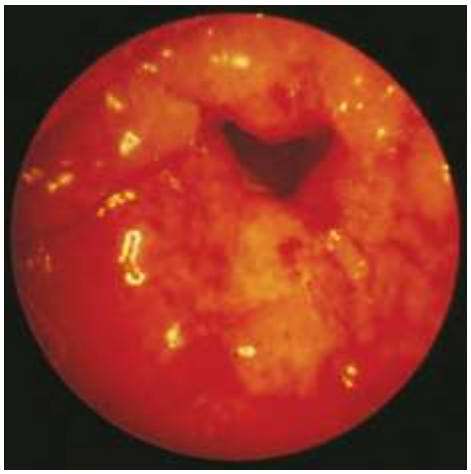


Figure 62.45 Adenocarcinoma of the cardia. Transcoelomic spread may occur with this type of lesion.

cardia and where there is a potential communication between a full-thickness tumour and the peritoneal cavity, for instance where there is a hiatus hernia.

COMPUTED TOMOGRAPHY

Computed tomography (CT) from the neck to the pelvis with intravenous contrast is the modality most used to identify haematogenous metastases (**Figure 62.46**). Distant organs are easily seen and metastases within them visualised with high accuracy (94–100%). The normal thoracic oesophagus is easily demonstrated by CT scanning. The mediastinal fat planes are usually clearly imaged in healthy individuals, and any blurring or distortion of these images is a fairly reliable indicator of abnormality. In cachectic patients with dysphagia and malnutrition, the mediastinal fat plane may be virtually absent, making local invasion difficult to assess. Thin-slice CT permits structures such as lymph nodes to be adequately imaged, down to a minimum diameter of about 5 mm. Smaller nodes cannot be reliably visualised, and it is not possible to distinguish between enlarged lymph nodes that have reactive changes only and metastatic nodes. Similarly, micrometastases within normal-sized nodes cannot be detected.

MAGNETIC RESONANCE IMAGING

Magnetic resonance imaging (MRI) does not expose the patient to ionising radiation and needs no intravascular contrast medium, although intraoesophageal air or contrast media may help to assess wall thickness. Distant metastases to organs such as the liver are usually reliably identified by MRI but, at the moment, there do not seem to be additional benefits over CT.

ENDOSCOPIC ULTRASONOGRAPHY

After haematogenous spread, the two principal prognostic factors for oesophageal cancer are the depth of tumour penetration through the oesophageal wall and regional lymph node spread. Although CT will detect distant metastasis, its limited axial resolution precludes a reliable assessment of both the depth of wall penetration and lymph node involvement. Endoscopic ultrasonography (EUS) can determine the depth of spread of a malignant tumour through the oesophageal wall (T1–3), the invasion of adjacent organs (T4) and metastasis to lymph nodes (N0 or N1) (**Figures 62.47–62.49**). It can also detect contiguous spread downward into the cardia and more distant metastases to the left lobe of the liver.

EUS visualises the oesophageal wall as a multilayered structure. The layers represent ultrasound interfaces rather than true anatomical layers, but there is close enough correlation to allow accurate assessment of the depth of invasion through the oesophageal wall. Structures smaller than 5 mm can be clearly seen, enabling very small nodes to be imaged. The EUS image morphology of such structures provides an additional means of distinguishing malignant from reactive or benign lymph nodes. For submucosal lesions, EUS can demonstrate the wall layer of origin of a lesion, suggesting the likely histological type.

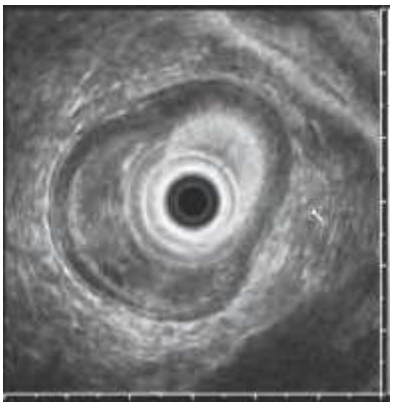


Figure 62.47 Endosonography demonstrating an 'early' tumour. Note the preservation of the outer dark wall layer that represents the muscle coat.

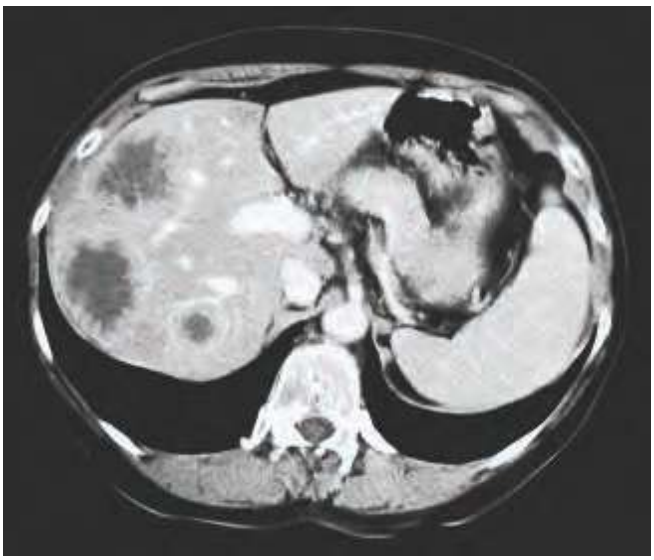


Figure 62.46 Computed tomography scan demonstrating liver metastases.

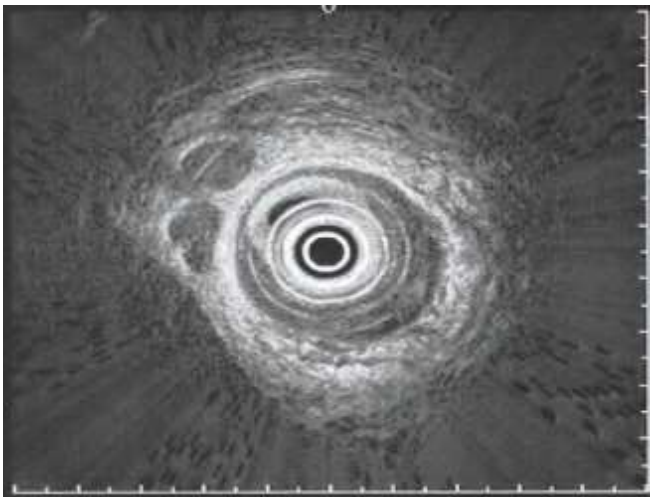


Figure 62.48 Endosonography demonstrating an 'advanced' local tumour. Note the breach of the outer white line that represents the interface between the oesophageal wall and the mediastinum.

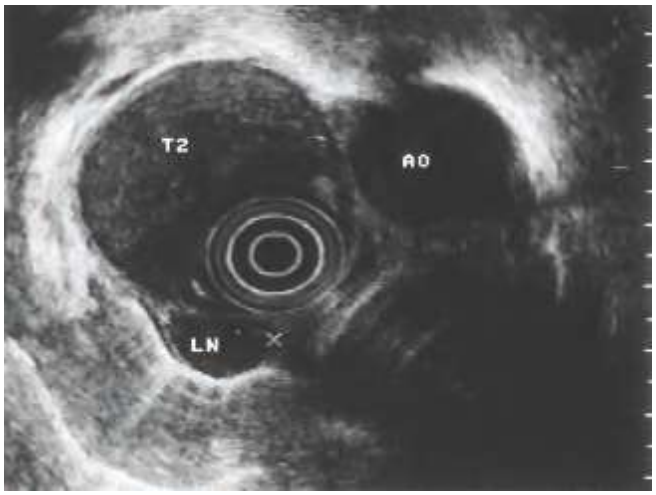


Figure 62.49 Endosonography demonstrating malignant nodes. These are usually large, hypoechoic and round compared with normal nodes.

Narrow EUS instruments are available for insertion over a guidewire to minimise the risk of technical failure, and linear array echoendoscopes can be used to biopsy lesions that might signify incurability outside the wall of the GI tract which might be considered to lie outside the proposed field of resection (usually lymph nodes outside a standard field).

Positron emission tomography with CT

Positron emission tomography (PET) with CT (PET-CT) in the context of cancer staging relies on the generally high metabolic activity (particularly in the glycolytic pathway) of tumours compared with normal tissues. The patient is given a small dose of the radiopharmaceutical agent [^{18}F]fluorodeoxyglucose (FDG). This enters cells and is phosphorylated. FDG 6-phosphate cannot be metabolised further and, as it is a highly polar molecule, it cannot easily diffuse back out of the cell. After intravenous injection of FDG, it continuously accumulates in metabolically active cells. Primary oesophageal cancers are usually sufficiently active to be easily visible, and spatial resolution of positive PET areas occurs down to about 5–8 mm. When used in isolation, there are problems with the anatomical location of these areas. This has been significantly improved by combining PET with CT (**Figure 62.50**), which covers the whole body. The main value of PET-CT is the identification of distant metastatic disease not seen on CT alone. Although there are wide variations between centres, a change in stage is frequently reported in around 15% of patients, but the timing of the scan in relation to the last date of treatment is important and probably accounts for differences in perceived effect. It has also been suggested that a reduction in PET activity after chemotherapy might be a way of predicting 'responders' to this approach, although the persistence of PET activity after a neoadjuvant treatment seems to be a poor prognostic sign.

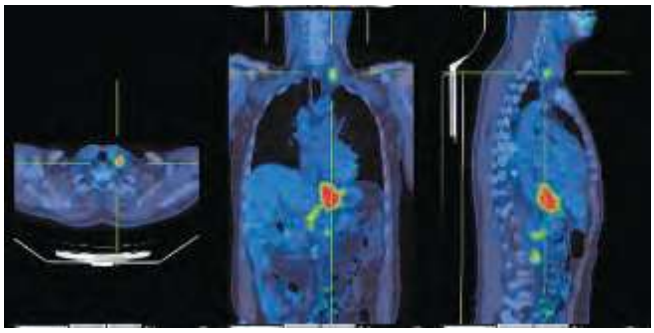


Figure 62.50 Positron emission tomography/computed tomography demonstrating a primary tumour and a distant metastatic node.

Treatment of malignant tumours

PRINCIPLES

At the time of diagnosis, around two-thirds of all patients with oesophageal cancer will already have incurable disease. The aim of palliative treatment is to overcome debilitating or distressing symptoms while maintaining the best quality of life possible for the patient. Some patients do not require specific therapeutic interventions, but do need supportive care and appropriate liaison with community nursing and hospice care services.

As dysphagia is the predominant symptom in advanced oesophageal cancer, the principal aim of palliation is to restore adequate swallowing. A variety of methods is available and, given the short life expectancy of most patients, it is important that the choice of treatment should be tailored to each individual. Tumour location and endoscopic appearance are important in this regard, as is the general condition of the patient.

Once oesophageal neoplasms reach the submucosal layer of the oesophagus, the tumour has access to the lymphatic system, meaning that, even at this early local stage, there is an incidence of nodal positivity for both squamous cell carcinoma and adenocarcinomas of about 20%. The principle of oesophagectomy is to deal adequately with the local tumour in order to minimise the risk of local recurrence and achieve an adequate lymphadenectomy to reduce the risk of staging error. Although studies in Japan would indicate that more extensive lymphadenectomy is associated with better survival, this may simply reflect more accurate staging. A number of studies support the view that the proximal extent of resection should ideally be 10 cm above the macroscopic tumour and 5 cm distal. When such a margin cannot be achieved proximally, particularly with squamous cell carcinoma, there is evidence that postoperative radiotherapy can minimise local recurrence, although this does not improve survival.

Adenocarcinoma commonly involves the gastric cardia and may therefore extend into the fundus or down the lesser curve. Some degree of gastric excision is essential in order to achieve adequate local clearance and accomplish an appropriate lymphadenectomy. Excision of contiguous structures, such as the crura, diaphragm and mediastinal pleura, needs to be considered as a method of creating negative resection margins.

The rarity of intramucosal cancer in symptomatic patients means that there are no randomised studies to compare different approaches to this type of very early disease. EMR for these apparently early (T1a) lesions has become increasingly popular, providing either a cure or at least sufficient histological information on which to base a further management strategy.

Surgery alone is best suited to patients with disease confined to the oesophagus (T1b, T2) without nodal metastasis (N0). The problem here is ensuring that patients are truly node negative. Despite careful preoperative investigation, these patients are not easily identified and so there is an argument for neoadjuvant therapy followed by surgery. A cure rate for the patient who is truly node negative having surgery alone of between 50% and 80% needs to be balanced against adverse events related to neoadjuvant therapy, the likelihood of response and the potential impact on survival. Patients with more advanced stages of disease require either multimodal approaches or entry into appropriate trials.

It is essential that oesophagectomy should be performed with a low hospital mortality and complication rate. Patient selection, volume and experience of the surgical team are all important. Preoperative risk analysis has shown that this can play a major part in reducing hospital mortality. There are really no circumstances in the western world in which surgery should be undertaken if it is not part of an overall treatment plan aimed at cure.

Summary box 62.12

Treatment of carcinoma of the oesophagus

- Radical oesophagectomy is the most important aspect of curative treatment
- Neoadjuvant treatments before surgery may improve survival in a proportion of patients
- Chemoradiotherapy alone may cure selected patients, particularly those with squamous cell cancers
- Useful palliation may be achieved by chemo-/radiotherapy or endoscopic treatments

Treatments with curative intent

SURGERY

Histological tumour type, location and the extent of the proposed lymphadenectomy all influence the surgical approach. This is largely an issue of surgical preference, although it should be recognised that a left thoracoabdominal approach is limited proximally by the aortic arch and should be avoided when the primary tumour is at or above this level. Similarly, transhiatal oesophagectomy is unsuitable for most patients with squamous cell carcinoma because a complete mediastinal lymphadenectomy is not easily achieved by this approach. The most widely practised approach in the west is the two-phase Ivor Lewis (sometimes called Lewis–Tanner) operation (Figure 62.51), with an initial laparotomy and construction of a gastric tube, followed by a right thoracotomy to excise the tumour and create an oesophagogastric anastomosis. The closer this is placed to the apex of the thoracic cavity, the fewer problems there are with reflux disease. Three-phase oesophagectomy (McKeown) may be more appropriate for more proximal tumours in order to achieve better longitudinal clearance, although the additional distance gained is less than many surgeons believe. A third cervical incision also permits lymphadenectomy in this region.

The extent of lymphadenectomy is highly controversial. For squamous cell carcinoma, because a higher proportion of patients will have middle- and upper-third tumours in the thoracic oesophagus, the rationale behind a three-phase operation with three-field lymphadenectomy is more understandable, even though this approach has not been widely adopted in the west. For adenocarcinoma, the incidence of metastases in the neck is relatively low in the context of patients who would otherwise be curable. For this reason, two-phase operations with two-field lymphadenectomy seem the most logical operations. Although two-field lymphadenectomy does not substantially increase surgical morbidity or mortality, the same cannot be said for more extended operations.

Minimal access techniques, pioneered in Australia by Gotley and Smithers and in North America by Luketich, have enjoyed increasing popularity, often combined with enhanced recovery programmes after surgery (ERAS). Hybrid (where at least one phase is performed as open surgery), total minimally invasive and robotic oesophagectomy all have their advocates, although there are no high-quality comparative studies and little evidence to indicate clear superiority of one approach

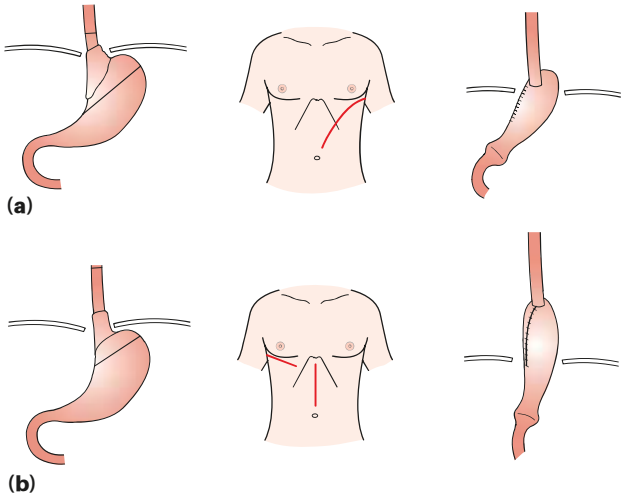


Figure 62.51 The two usual approaches for surgery of the oesophagus are (a) the thoracoabdominal, which opens the abdominal and thoracic cavities together, and (b) the two-stage Ivor Lewis approach, in which the abdomen is opened first, closed and then the thoracotomy is performed. In the McKeown operation, a third incision in the neck is made to complete the cervical anastomosis.

Ivor Lewis, 1895–1982, surgeon, the North Middlesex Hospital, London, UK, and later at Rhyl, North Wales, UK.
Norman Cecil Tanner, 1906–1982, surgeon, Charing Cross Hospital, London, UK.
Kenneth Charles McKeown, 1912–1995, surgeon, Darlington Memorial Hospital, Darlington, County Durham, UK.

over the others. In experienced hands, the open operation can be reproduced by less invasive approaches, without significant compromise. Many groups have reported similar lymph node yields and rates of resection margin positivity with open and minimal access approaches. As yet, benefits seem to be confined to reduced wound pain and the absence of specific complications associated with long incisions.

Although many centres have reduced hospital mortality to low single figures after oesophagectomy, the complication rate remains high. At least a third of all patients will develop some significant complication after surgery. The most common of these is respiratory, followed by anastomotic leakage, chylothorax and injury to the recurrent laryngeal nerves. The most common late problem is benign anastomotic stricture, which seems to be higher with cervical rather than with intrathoracic anastomoses, although the problem is usually easily dealt with by endoscopic dilatation.

Lesions of the cardia that do not involve the oesophagus to any significant extent may be dealt with by extended total gastrectomy to include the distal oesophagus, or by proximal gastrectomy and distal oesophagectomy.

Summary box 62.13

Oesophagogastric surgery

- Beware of satellite nodules proximal to the primary lesion
- Carefully preserve the blood supply of the stomach, both venous and arterial
- Right thoracic approach gives easy access to the oesophagus

TWO-PHASE OESOPHAGECTOMY (ABDOMEN AND RIGHT CHEST, IVOR LEWIS)

Mobilisation of the stomach must be done with care because it is essential to have a tension-free, well-vascularised stomach for transposition. The left gastric, short gastric and left gastroepiploic arteries are all divided. The viability of the transposed stomach mainly depends on the right gastroepiploic and, to a lesser extent, the right gastric vessels. It should be noted that venous drainage is as important as arterial supply, and it is essential to perform an accurate anatomical dissection that preserves the right gastroepiploic vein as well as the artery. The stomach is divided to remove the cardia and the upper part of the lesser curve, including the whole of the left gastric artery and its associated lymph nodes.

The approach to the oesophagus through the right chest is straightforward, providing excellent access to the mediastinum, the thoracic inlet and the hiatus. The azygos vein is divided, and the whole of the intrathoracic oesophagus can be mobilised along with the thoracic duct (which is ligated by most surgeons) and the mediastinal lymph nodes. The oesophagus is divided just below the thoracic inlet. As most lesions are in the lower or middle third, this usually gives adequate proximal clearance of at least 5 cm. Carcinomas of the upper thoracic oesophagus are almost always incurable at the time of diagnosis, and invasion of the trachea is common. If one of these lesions is resectable, it is essential to use an incision in the neck (McKeown or three-phase operation) and to resect more of the oesophagus than is customary in the operation of subtotal oesophagectomy.

Oesophagogastric anastomosis may be performed equally well by hand or stapler. Both methods require attention to detail. In experienced hands, clinical anastomotic leakage should be less than 10%. Most surgeons still prefer to keep patients nil by mouth for 5–7 days. Most centres have abandoned the use of routine contrast swallows in patients who are clinically well. Conversely, aggressive investigation of a suspected leak is mandatory for any unexplained fever or clinical event. This should involve early endoscopy, which has been shown to be safe and is the most reliable method for identifying necrosis in any part of the replacement conduit and/or CT scan to resolve the situation adequately.

Postoperative nutritional support remains controversial. There is general agreement that parenteral feeding is associated with more nosocomial infection, including pneumonia, than enteral feeding. It is also expensive. The simplicity of placing a modern feeding jejunostomy device at the time of resection means that this is in routine use in many centres.

TRANSHIATAL OESOPHAGECTOMY (WITHOUT THORACOTOMY)

This approach was popularised for cancer by Orringer, adapting a technique developed in Brazil by Pinotti for the removal of chagasic megaoesophagus (see the section on achalasia, p. 1095). The stomach is mobilised through a midline abdominal incision or by laparoscopy, and the cervical oesophagus is mobilised through an incision in the neck. The diaphragm is then opened from the abdomen, and the posterior mediastinum is entered. The lower oesophagus and the tumour are mobilised under direct vision, and the upper oesophagus is mobilised by blunt dissection. This approach can provide an adequate removal of the tumour and lymph nodes in the lower mediastinum, but it is not possible to remove the nodes in the middle or upper mediastinum. It may be a useful procedure for lesions of the lower oesophagus, but is hazardous for a middle third lesion that may be adherent to the bronchus or to the azygos vein.

NEOADJUVANT TREATMENTS WITH SURGERY

Apart from the earliest stages of disease, surgery alone produces relatively few cures in either squamous cell carcinoma or adenocarcinoma patients. This led to a number of trials throughout the 1980s and 1990s to investigate the value of chemotherapy and surgery or chemoradiotherapy and surgery compared with surgery alone. Some studies relate only to squamous cell cancer, and many are open to criticism on the grounds of trial design or patient numbers. Nevertheless,

Mark Burton Orringer, surgeon, Ann Arbor, MI, USA.
Walter Pinotti, Professor of Surgery, Sao Paulo, Brazil.
Carlos Justiniano Ribeiro Chagas, 1879–1934, Director of the Oswaldo Cruz Institute and Professor of Tropical Medicine, the University of Rio de Janeiro, Brazil.

positive results in favour of neoadjuvant therapy for adenocarcinoma in two large studies as well as a limited meta-analysis indicated that it was no longer appropriate to consider surgery alone as the 'gold standard' treatment for most patients who are surgical candidates with adenocarcinoma. The Dutch trial (CROSS) that compared chemoradiotherapy and surgery versus surgery alone has provided the most convincing evidence so far of survival benefit for squamous cell carcinoma. The same study was initially unable to show a survival benefit for adenocarcinomas, although subsequent analyses again suggested survival benefit of similar magnitude to that seen with chemotherapy and surgery.

GASTRO-OESOPHAGEAL REFLUX FOLLOWING OESOPHAGOGASTRIC RESECTION

Gastro-oesophageal reflux may be a major problem after any operation that involves resecting the cardia. Reflux may present with the typical symptoms of GORD or with a peptic stricture at the site of the anastomosis. However, the presentation may be different with a miserable patient who fails to thrive after the operation and who is then suspected of having recurrent cancer. This atypical presentation is particularly common after total gastrectomy with an inadequate reconstruction that allows bile reflux.

Summary box 62.14

Postoesophagectomy

- Reflux may be a problem after resection
- Symptoms may be atypical
- Reflux may be limited or avoided by subtotal oesophagectomy and gastric transposition high in the chest

Non-surgical treatments

Radiotherapy alone was widely used as a single-modality treatment for squamous cell carcinoma of the oesophagus until the late 1970s. The 5-year survival overall rate was 6%. As a result, multimodal approaches were adopted throughout the 1980s, initial trials indicating that similar long-term survival rates could be obtained with surgery. Subsequent randomised studies, essentially confined to patients with squamous cell carcinoma, have indicated significant survival advantages with chemoradiotherapy over radiotherapy alone. Although it is clear that chemoradiotherapy does offer a prospect of cure for patients who may not be fit for surgery, particularly in squamous cell carcinoma, the high rate of locoregional failure has meant that surgery remains the mainstay of attempted curative treatments for both adenocarcinoma and squamous cell carcinoma in patients who have potentially resectable disease and are fit for oesophagectomy. In most western series, this represents about a third of patients with adenocarcinoma and a slightly lower percentage of patients with squamous cell carcinoma. There has been no high-quality randomised comparison of the results of definitive radiotherapy versus chemoradiotherapy and surgical resection, and it is therefore impossible to make dogmatic statements about the relative merits of each form of treatment.

Summary box 62.15

Alternative therapeutic approaches

- Chemoradiotherapy may be a useful alternative to surgery, especially in unfit patients

Palliative treatment

Surgical resection and external beam radiotherapy may be used for palliation, but are not suitable when the expected survival is short, because most of the remainder of life will be spent recovering from the 'treatment'. Surgical bypass is likewise too major a procedure for use in a patient with limited life expectancy. A variety of relatively simple methods of palliation is now available that will produce worthwhile relief of dysphagia with minimal disturbance to the patient.

Summary box 62.16

Palliation

- Palliation should be simple and effective

Intubation has been used for many years after the invention of the Souttar tube, which was made of coiled silver wire. A variety of rigid plastic or rubber tubes had been developed for placement under endoscopic and/or radiological control. The technology of intubation has now moved on with the development of various types of expanding metal stent (**Figure 62.52**). These are also inserted under radiographic or endoscopic control. The stent is collapsed during insertion and released when it is in the correct position. Expanding stents produce a wider lumen for swallowing than rigid tubes. More importantly, it is not necessary to dilate the oesophagus to beyond 8 mm to insert the unexpanded stent through the tumour, so there is a lower risk of injury to the oesophagus.

Endoscopic laser treatment may be used to core a channel through the tumour. It is based on thermal tumour destruction. It produces a worthwhile improvement in swallowing, but has the disadvantage that it has to be repeated every few weeks. Lasers may also be used to unblock a stent that has become occluded by tumour overgrowth. Other endoscopic

Figure 62.52 Expanding metal stents, covered and uncovered.

Sir Henry Sessions Souttar, 1875–1964, surgeon, the London Hospital, London, UK.

methods include bipolar diathermy, argon-beam plasma coagulation and alcohol injection.

Brachytherapy is a method of delivering intraluminal radiation with a short penetration distance (hence the prefix 'brachy') to a tumour. An introduction system is inserted through the tumour, and the treatment is then delivered in a single session lasting approximately 20 minutes. The equipment is expensive to purchase, but running costs are low.

Although the above methods are suitable for patients with very advanced disease, elderly people and those with significant comorbidities that would make more aggressive strategies inappropriate, an increasing proportion of patients (particularly with adenocarcinoma) are being treated with platinum-based chemotherapy. In general, this leads to only a modest prolongation of survival but a better quality of life than in those receiving an endoscopic treatment alone.

Malignant tracheo-oesophageal fistula

Malignant tracheo-oesophageal fistula is a sign of incurable disease. Some have advocated surgical bypass and oesophageal exclusion, but this is a major procedure. An expanding metal stent is probably the best treatment.

Post-cricoid carcinoma

Post-cricoid carcinoma is considered in Chapter 47.

MOTILITY DISORDERS AND DIVERTICULA

Oesophageal motility disorders

A motility disorder can be readily understood when a patient has dysphagia in the absence of a stricture, and a barium-impregnated food bolus is seen to stick in the oesophagus. If this can be correlated with a specific abnormality on oesophageal manometry, accepting that this is the cause of the patient's symptoms may be straightforward. Unfortunately, this is often not the case. Pain, with or without a swallowing problem, is frequently the dominant symptom, and patients often undergo extensive hospital investigation before the oesophagus is considered as a source of symptoms. Symptoms are often intermittent, and the correlation between symptoms and test 'abnormalities' is poor. Confirmation of a specific motility disturbance is made by high-resolution manometry. Much harm may be done by inappropriate enthusiastic surgery for ill-defined conditions. It should also be remembered that oesophageal dysmotility may be only a feature of a general disturbance in GI function.

Summary box 62.17

Oesophageal motility disorders

- May be part of a more diffuse GI motility problem
- May be associated with GORD

Oesophageal motility disorders are currently best classified by incorporation of the Chicago classification developed for use with high-resolution manometry as shown in *Tables 62.3 and 62.4*.

Functional pain and the oesophagus

Pain that is assumed to arise from dysfunction of the GI tract may reflect abnormal motor activity, abnormal perception or a combination of the two. There is evidence that all three exist. Very high-pressure uncoordinated contractions ('spasm') have been shown to correlate with pain. Distension of a balloon in the oesophagus indicates that some patients have a low threshold for the sensation of pain (visceral hypersensitivity), and this itself may reflect local or central neuronal dysfunction. In practice, the difficulty is in understanding the relative contributions of these elements, so that a logical treatment might follow.

Achalasia

Pathology and aetiology

Achalasia (Greek 'failure to relax') is uncommon, but merits its prominence because it is reasonably understood and responds to treatment. It is due to loss of the ganglion cells in the myenteric (Auerbach's) plexus, the cause of which is unknown. In South America, chronic infection with the parasite *Trypanosoma cruzi* causes Chagas' disease, which has marked clinical similarities to achalasia. Achalasia differs

TABLE 62.3 General classification of oesophageal motility disorders.

Disorders of the pharyngo-oesophageal junction
Neurological – stroke, motor neuron disease, multiple sclerosis, Parkinson's disease
Myogenic – myasthenia, muscular dystrophy
Pharyngo-oesophageal (Zenker's) diverticulum
Disorders of the body of the oesophagus
Diffuse oesophageal spasm
Nutcracker oesophagus
Autoimmune disorders – especially systemic sclerosis (CREST)
Reflux associated
Idiopathic
Allergic
Eosinophilic oesophagitis
Non-specific oesophageal dysmotility
Disorders of the lower oesophageal sphincter
Achalasia
Incompetent lower sphincter (i.e. GORD)

CREST, calcinosis, Raynaud's syndrome, (o)esophageal motility disorders, sclerodactyly and telangiectasia.
GORD, gastro-oesophageal reflux disease.

TABLE 62.4 The Chicago Classification of oesophageal motility v3.0.

Achalasia and EGJ outflow obstruction	Criteria
Type I achalasia (classic achalasia)	Elevated median IRP (>15 mmHg), 100% failed peristalsis (DCI <100 mmHg·s·cm) *Premature contractions with DCI values* <450 mmHg·s·cm *satisfy criteria for failed peristalsis*
Type II achalasia (with oesophageal compression)	Elevated median IRP (>15 mmHg), 100% failed peristalsis, panoesophageal pressurisation with ≥20% of swallows *Contractions may be masked by oesophageal pressurisation and DCI should not be calculated*
Type III achalasia (spastic achalasia)	Elevated median IRP (>15 mmHg), no normal peristalsis, premature (spastic) contractions with DCI >450 mmHg·s·cm with ≥20% of swallows *May be mixed with panoesophageal pressurisation*
EGJ outflow obstruction	Elevated median IRP (>15 mmHg), sufficient evidence of peristalsis such that criteria for types I–III achalasia are not met
Major disorders of peristalsis	(Not encountered in normal individuals)
Absent contractility	Normal median IRP, 100% failed peristalsis *Achalasia should be considered when IRP values are borderline and when there is evidence of oesophageal pressurisation* *Premature contractions with DCI values* <450 mmHg·s·cm *meet criteria for failed peristalsis*
Distal oesophageal spasm	Normal median IRP, ≥20% premature contractions with DCI >450 mmHg·s·cm. Some normal peristalsis may be present
Hypercontractile oesophagus (jackhammer)	At least two swallows with DCI >8000 mmHg·s·cm *Hypercontractility may involve, or even be localised to, the LES*
Minor disorders of peristalsis	(Characterised by contractile vigour and contraction pattern)
Ineffective oesophageal motility (IEM)	≥50% ineffective swallows *Ineffective swallows can be failed or weak (DCI* <450 *mmHg·s·cm)* *Multiple repetitive swallow assessment may be helpful in determining peristaltic reserve*
Fragmented peristalsis	≥50% fragmented contractions with DCI >450 mmHg·s·cm
Normal oesophageal motility	Not fulfilling any of the above classifications

DCI, distal contractile integral (mmHg·s·cm); EGJ, o(e)sophaogastric junction; IRP, integrated relaxation pressure (mmHg); LES, lower o(e)sophageal sphincter. (From Kahrilas PJ *et al*. The Chicago Classification of esophageal motility disorders v3.0. *Neurogastroenterology and Motility* 2015; **27**(2): 160–74, with kind permission, John Wiley & Sons.)

from Hirschsprung's disease of the colon because the dilated oesophagus usually contains few ganglion cells, whereas the dilated colon contains normal ganglion cells proximal to a constricted, aganglionic segment. Histology of muscle specimens generally shows a reduction in the number of ganglion cells (and mainly inhibitory neurons) with a variable degree of chronic inflammation. When powerful non-peristaltic contractions are still present, perhaps representing an early stage of the disease, inflammation and neural fibrosis may be seen with normal numbers of ganglion cells.

Summary box 62.18

Achalasia

- Is uncommon
- Is due to selective loss of inhibitory neurons in the lower oesophagus
- The causes dysphagia and carcinoma must be excluded
- Treatment is by either endoscopic dilatation, or endoscopic or surgical myotomy

The classic physiological abnormalities are a non-relaxing LOS and absent peristalsis in the body of the oesophagus. The Chicago classification identifies three variants. Although most patients have almost no recognisable contractions (type I), some patients continue to exhibit pressurisation throughout the oesophagus (type 2), whereas in others the oesophagus is of normal calibre and exhibits high-pressure contractile (although non-peristaltic) activity (spastic oesophagus, vigorous achalasia). In some patients, these uncoordinated contractions result in pain as much as a sense of food sticking. High-resolution manometry recognises these contraction patterns, which may be important in predicting the outcome of treatment. With time, the oesophagus dilates and contractions disappear, so that the oesophagus empties mainly by the hydrostatic pressure of its contents. This is nearly always incomplete, leaving residual food and fluid. The gas bubble in the stomach is frequently absent, as no bolus with its accompanying normal gas passes through the sphincter. The 'megaoesophagus' becomes tortuous with a persistent retention oesophagitis due to fermentation of food residues (**Figure 62.53**), and this may account for the increased incidence of carcinoma of the oesophagus

Harald Hirschsprung, 1831–1916, physician, the Queen Louise Hospital for Children, and Professor of Paediatrics, Copenhagen, Denmark, described congenital megacolon in 1888.

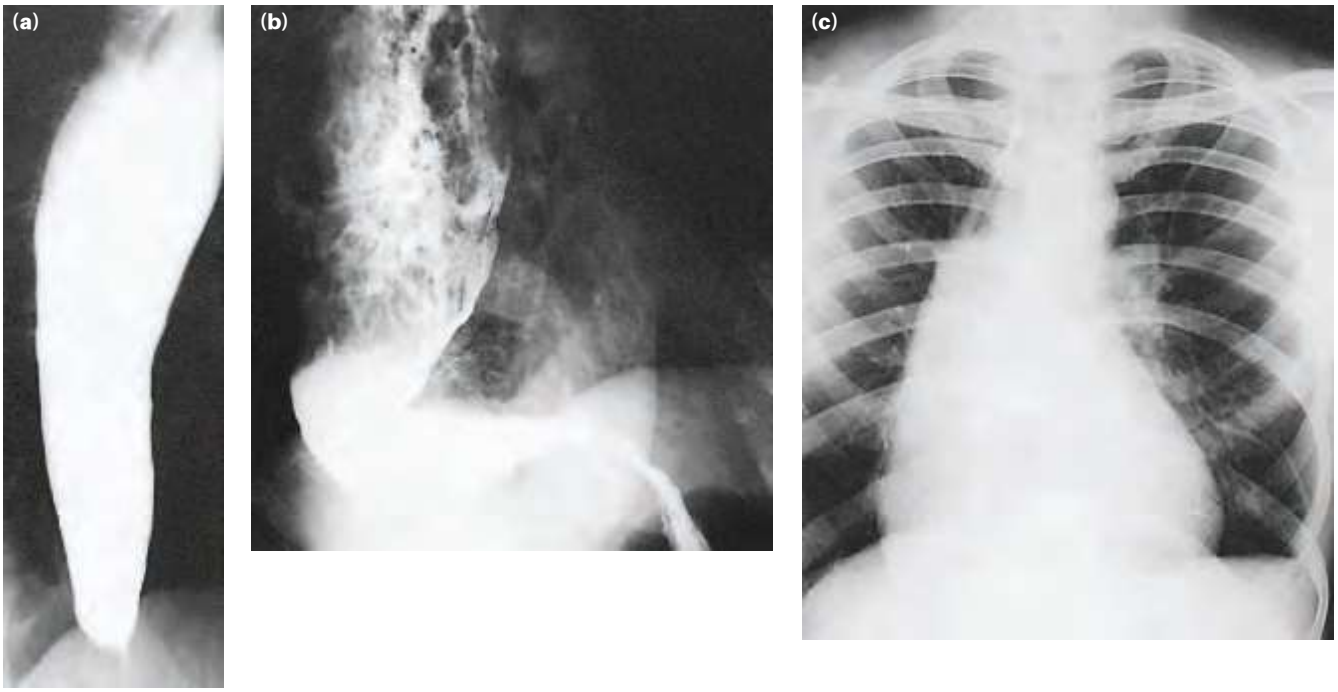


Figure 62.53 Achalasia of the oesophagus. **(a)** Barium swallow showing the smooth outline of the stricture, which narrows to a point at its lower end. **(b)** Tortuosity and sigmoid appearance of the lower oesophagus. **(c)** Mediastinal shadow due to a large, fluid-filled oesophagus.

Summary box 62.19

Lower oesophageal stricture

- Beware pseudoachalasia; look for tumour

Pseudoachalasia is an achalasia-like disorder that is usually produced by adenocarcinoma of the cardia (Figure 62.54), but has also been described in relation to benign tumours at this level. It has been presumed that the inability of the sphincter to relax is linked to the loss of body peristalsis, although other cancers outside the oesophagus (bronchus, pancreas) have also been associated with pseudoachalasia.

Clinical features

The disease is most common in middle life, but can occur at any age. It typically presents with dysphagia, although pain (often mistaken for reflux) is common in the early stages. Patients often present late and, having had relatively mild symptoms, remain untreated for many years. Regurgitation is frequent, and there may be overspill into the trachea, especially at night.

Diagnosis

Achalasia may be suspected at endoscopy by finding a tight cardia and food residue in the oesophagus. Barium radiology may show hold-up in the distal oesophagus, abnormal contractions in the oesophageal body and a tapering stricture in the distal oesophagus, often described as a 'bird's beak' (see

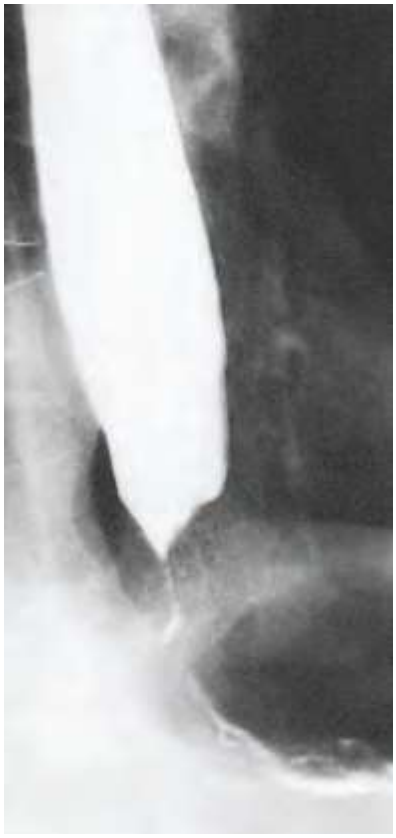


Figure 62.54 Almost achalasia, but note the irregularity of the taper, which indicates carcinoma of the cardia.

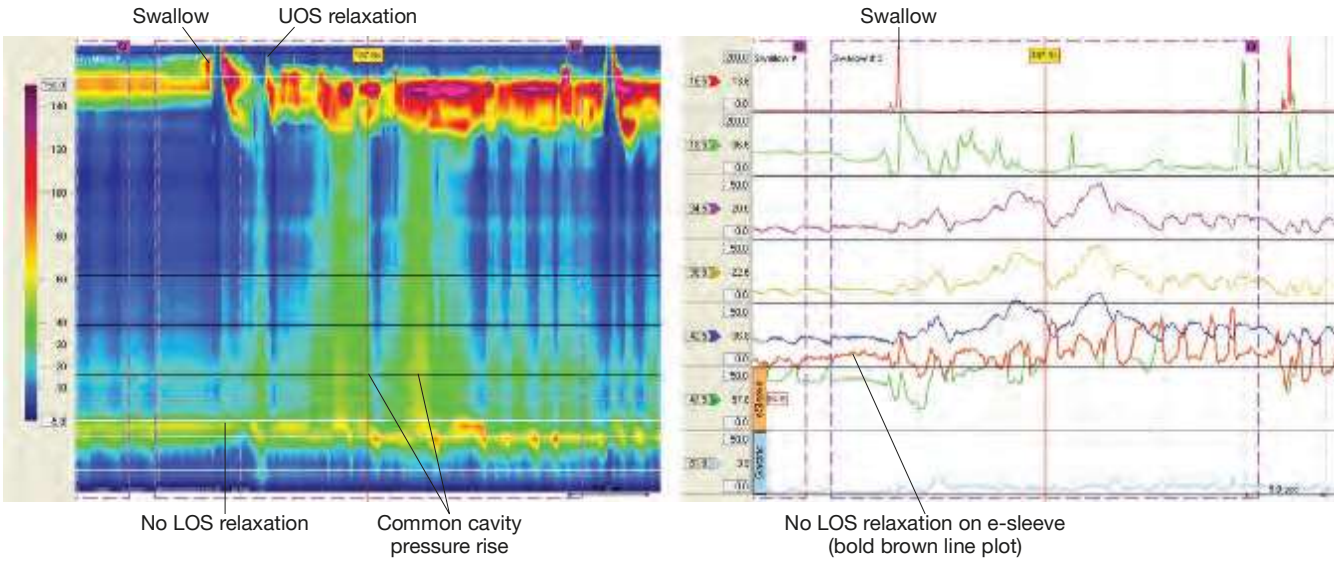


Figure 62.55 High-resolution oesophageal manometry from a patient presenting with dysphagia and regurgitation. The swallow is followed by a common cavity rise in oesophageal pressure indicating filling. Lower oesophageal sphincter (LOS) relaxation is absent and there is a positive oesophagogastric pressure gradient. Upper oesophageal sphincter (UOS) relaxation shortly after the swallow was related to regurgitation of oesophageal contents.

Figure 62.53). The gastric gas bubble is usually absent. These typical features of well-developed achalasia are often absent, and endoscopy and radiology can be normal. A firm diagnosis is established by high-resolution oesophageal manometry. Classically, the LOS does not relax completely on swallowing, there is no peristalsis and there is a raised resting pressure in the oesophagus (Figure 62.55). The LOS pressure may be elevated, but is often normal.

Treatment

Alone among motility disorders, achalasia responds well to treatment. The two main methods are forceful dilatation of the cardia and laparoscopic cardiomyotomy (conventionally performed together with a partial fundoplication), although in recent years there has been growing interest in the use of peroral endoscopic myotomy (POEM). Comparative studies between pneumatic dilatation and surgical myotomy suggest equivalence in terms of safety, effectiveness and cost when considered over a number of years.

PNEUMATIC DILATATION

This involves stretching the cardia with a balloon to disrupt the muscle and render it less competent. The treatment was first described by Plummer. Many varieties of balloon have been used but, nowadays, plastic balloons with a precisely controlled external diameter are used. If the pressure in the balloon is too high, the balloon is designed to split along its length rather than expanding further. Balloons of 30–40 mm in diameter are available and are inserted over a guidewire (Figure 62.56). Perforation is the major complication. With a 30-mm balloon, the incidence of perforation should be less than 0.5%. The risk of perforation increases with bigger balloons, and they should be used cautiously for progressive dilatation over a period of weeks. Forceful dilatation is curative in 75–85% of cases. The results are best in patients aged over 45 years.

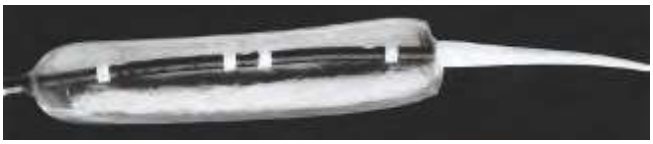


Figure 62.56 Balloon dilator for the treatment of achalasia by forceful dilatation.

Summary box 62.20

Achalasia

- Beware perforation due to dilatation of achalasia
- Beware postoperative reflux

HELLER'S MYOTOMY

This involves cutting the muscle of the lower oesophagus and cardia (Figure 62.57). The major complication is

Henry Stanley Plummer, 1874–1937, physician, the Mayo Clinic, Rochester, MN, USA.
Ernst Heller, 1877–1964, surgeon, St George's Krankenhaus, Leipzig, Germany.

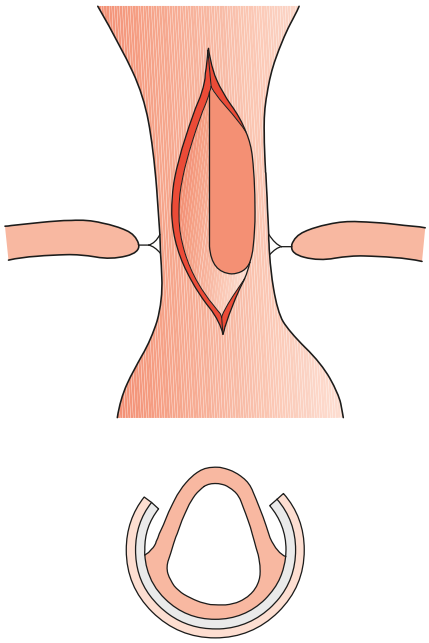


Figure 62.57 Heller's myotomy: the incision should not go too far on to the stomach. The lateral extent must enable the mucosa to pout out, to prevent the edges healing together.

gastro-oesophageal reflux, and most surgeons therefore add a partial anterior fundoplication (Heller–Dor operation). The procedure is ideally suited to a minimal access laparoscopic approach, and most surgeons use intraoperative endoscopy to judge the extent of the myotomy and to ensure that the narrow segment is abolished.

It is successful in more than 90% of cases and may be used after failed dilatation.

ENDOSCOPIC MYOTOMY

This was introduced in Japan in 2009. It involves the creation of a submucosal tunnel in the midoesophagus that can be developed distally, allowing a long myotomy to be performed. The oesophageal mucosal defect is clipped shut at the end. Many cohort studies indicate that the procedure can be performed safely and early clinical results imply equivalence to other approaches.

BOTULINUM TOXIN

This is done by endoscopic injection into the LOS. It acts by interfering with cholinergic excitatory neural activity at the LOS. The effect is not permanent, and the injection usually has to be repeated after a few months. For this reason, its use is restricted to elderly patients with other comorbidities.

DRUGS

Drugs such as calcium channel antagonists have been used but are ineffective for long-term use. However, sublingual nifedipine may be useful for transient relief of symptoms if definitive treatment is postponed.

Other oesophageal motility disorders

Disorders of the pharyngo-oesophageal junction

With the exception of Zenker's diverticulum (see below), most patients with oropharyngeal dysphagia have generalised neurological or muscular disorders with pharyngeal involvement. A small number of patients who have sustained a cerebrovascular accident benefit from myotomy of cricopharyngeus to alleviate pooling of saliva and nocturnal aspiration, but they should have good deglutition and phonation before this is performed. The operation is also effective in patients with oculopharyngeal muscular dystrophy.

Disorders of the body of the oesophagus

The older terms, 'diffuse oesophageal spasm' and 'jackhammer (nutcracker) oesophagus', have been replaced in the Chicago classification. There are, nevertheless, patients with incoordinate contractions of the oesophagus who experience dysphagia and/or chest pain. The condition may be dramatic, with spastic pressures on manometry of 400–500 mmHg, marked hypertrophy of the circular muscle and a corkscrew oesophagus on barium swallow (**Figure 62.58**). These abnormal contractions are more common in the distal two-thirds of the oesophageal body (**Figure 62.59**) and this may have some relevance to treatment. Making the diagnosis when chest pain is the only symptom may be difficult. Prolonged oesophageal manometry that correlates episodes of chest pain with manometric abnormalities may establish the diagnosis.

There is no proven pharmacological or endoscopic treatment. Calcium channel antagonists, vasodilators and endoscopic dilatation have only transient effects. Although the severity and frequency of symptoms may be tolerated by most

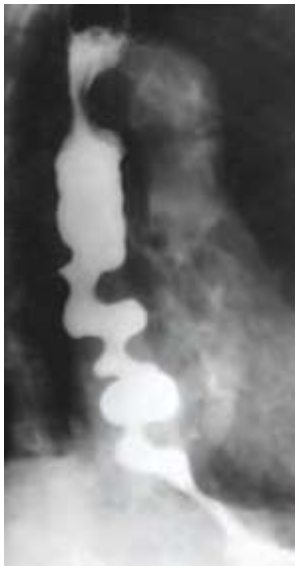


Figure 62.58 Corkscrew oesophagus in diffuse oesophageal spasm.

Friedrich Albert Zenker, 1825–1898, Professor of Pathology, Dresden, Germany.

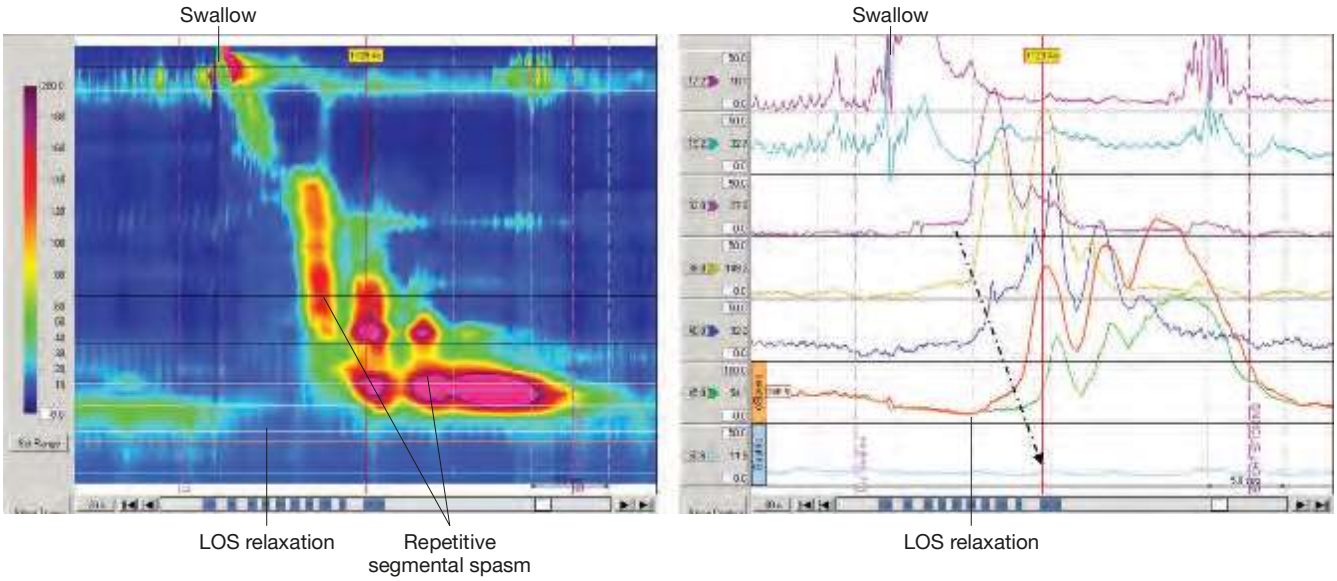


Figure 62.59 High-resolution oesophageal manometry from a patient presenting with dysphagia and chest pain. The swallow is followed by simultaneous, repetitive contractions in the mid-distal smooth muscle oesophagus. Lower oesophageal sphincter (LOS) relaxation is preserved. Note the sequential simultaneous contractions first in the mid- and distal segments of the oesophagus and then in the LOS make it appear as if there is progressive peristalsis on the conventional line plots (dotted arrow). Repetitive contractions are seen clearly on both.

patients, sometimes the combination of chest pain and dysphagia is sufficiently severe that malnutrition begins. In these patients, extended oesophageal myotomy up to the aortic arch may be required. Surgical treatment is more successful in improving dysphagia than chest pain, and caution should be exercised in patients in whom chest pain is the only symptom.

Jackhammer or nutcracker oesophagus is a condition with characteristic high-pressure manometric features (see *Table 62.4*). The correlation of manometric abnormalities with symptoms remains poor.

OESOPHAGEAL INVOLVEMENT IN AUTOIMMUNE DISEASE

Oesophageal involvement is mainly seen in systemic sclerosis, but may be a feature of polymyositis, dermatomyositis, systemic lupus erythematosus, polyarteritis nodosa and rheumatoid disease. Although most involve weak peristalsis, swallowing difficulties may be compounded by pharyngeal problems in the disorders that primarily affect skeletal muscle (e.g. polymyositis) or extraoesophageal problems such as involvement of the cricoarytenoid joint in rheumatoid disease or dry mouth in Sjögren's syndrome. In systemic sclerosis, smooth muscle atrophy causes hypoperistalsis (**Figure 62.60**). The LOS is involved, leading to a loss of the antireflux barrier. A wide range of symptoms can follow from mild to severe dysphagia accompanied by regurgitation and aspiration. Reflux can be severe and is exacerbated by weak acid clearance so that strictures can occur. There are no drugs that specifically correct the motor disorder, and medical treatment is mainly directed at minimising reflux-induced damage with PPIs. A small number of patients may require anti-reflux surgery.

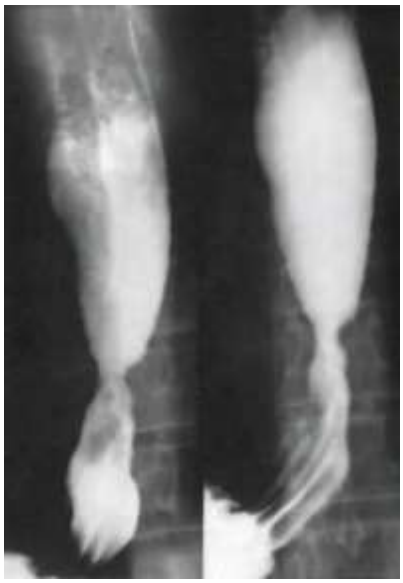


Figure 62.60 Advanced scleroderma of the oesophagus. The oesophagus dilates, and the lower oesophageal sphincter is widely incompetent.

Eosinophilic oesophagitis is a disorder that occurs in children and adults either alone or as a manifestation of

Henrik Samuel Conrad Sjögren, 1899–1986, Professor of Ophthalmology, Gothenburg, Sweden.

eosinophilic gastroenteritis. It is characterised by eosinophilic infiltration of the oesophageal wall, presumably of allergic or idiopathic origin. The most common presenting symptom is dysphagia, and more than half have some history of atopy. The oesophagus often seems narrow and friable on endoscopy and may include mucosal rings. The most important feature is the development of deep ulcers, leading to stricture development, especially in the proximal oesophagus. The diagnosis is established by endoscopic biopsy.

Elimination diets, and topical and systemic steroids all seem to be helpful in the short term, but there is scant information on the long-term impact of any particular approach. Immunotherapy directed against interleukin (IL)-5, which has a major role in eosinophil recruitment, seems to be a promising innovative approach. Although endoscopic dilatation has been recommended, this can create deep ulcers and further scarring, so should be used with caution and only when the above therapies fail.

Pharyngeal and oesophageal diverticula

Most oesophageal diverticula are **pulsion** diverticula that develop at a site of weakness as a result of chronic pressure against an obstruction. Symptoms are mostly caused by the underlying disorder unless the diverticulum is particularly large. Traction diverticula (**Figure 62.61**) are much less common. They are mostly a consequence of chronic granulomatous disease affecting the tracheobronchial lymph nodes due to tuberculosis, atypical mycobacteria or histoplasmosis. Fibrotic healing of the lymph nodes exerts traction on the oesophageal wall and produces a focal outpouching which is usually small and has a conical shape. There may be associated broncholithiasis, and additional complications may occur, such as aerodigestive fistulation (**Figure 62.62**) and bleeding.

Zenker's diverticulum (pharyngeal pouch) is not really an oesophageal diverticulum as it protrudes posteriorly above the cricopharyngeal sphincter through the natural weak point (the dehiscence of Killian) between the oblique and horizontal (cricopharyngeus) fibres of the inferior pharyngeal constrictor (**Figures 62.63 and 62.64**). The exact mechanism that leads to its formation is unknown, but it involves loss of the coordination between pharyngeal contraction and opening of the upper sphincter. When the diverticulum is small, symptoms largely reflect this incoordination with predominantly pharyngeal dysphagia. As the pouch enlarges, it tends to fill with food on eating, and the fundus descends into the mediastinum. This leads to halitosis and oesophageal dysphagia. Treatment can be undertaken endoscopically with a linear cutting stapler to divide the septum between the diverticulum and the upper oesophagus, producing a

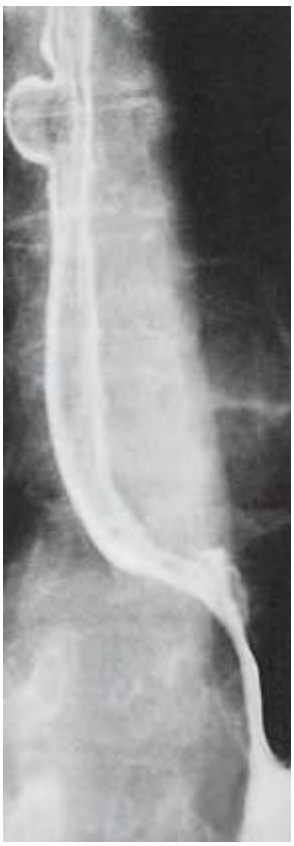


Figure 62.61 Midoesophageal traction diverticulum with the mouth facing downwards.

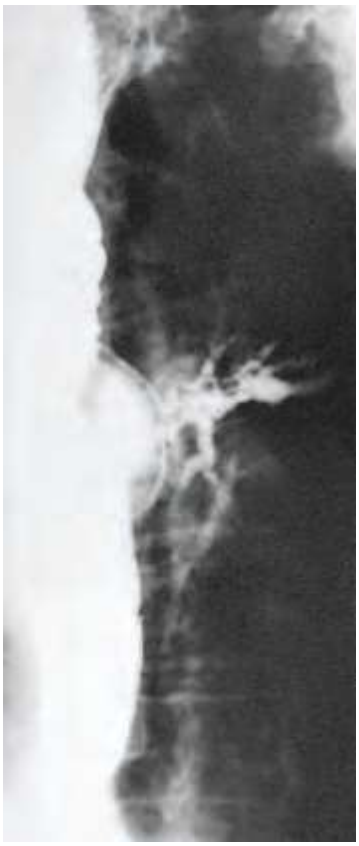


Figure 62.62 Midoesophageal diverticulum with a trachea-oesophageal fistula.

Gustav Killian, 1860–1921, Professor of Laryngology at Freiburg, and later at Berlin, Germany.

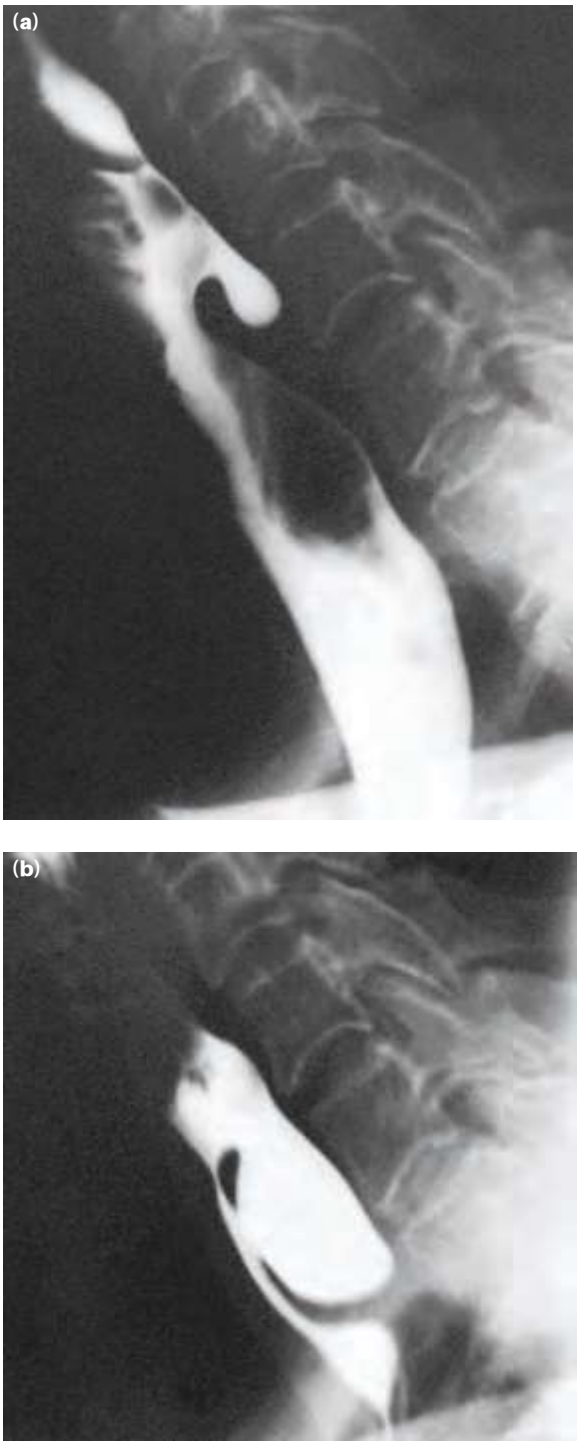


Figure 62.63 The typical appearances of: **(a)** a small pharyngeal pouch with a prominent cricopharyngeal impression and 'streaming' of barium, indicating partial obstruction; and **(b)** a large pouch extending behind the oesophagus towards the thoracic inlet.

diverticulo-oesophagostomy, or can be done by open surgery involving pouch excision, pouch suspension (diverticulopexy) and/or myotomy of cricopharyngeus. All techniques have good results.

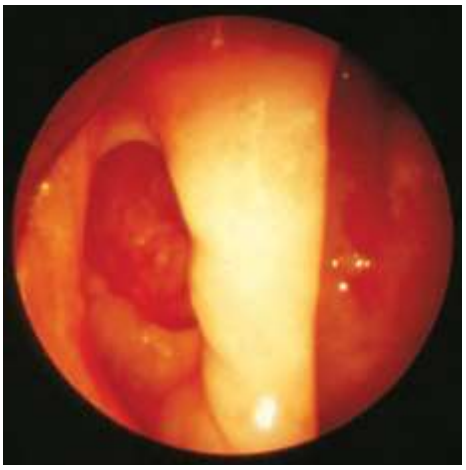


Figure 62.64 The endoscopic appearance of the mouth of a pharyngeal pouch posterior to the normal opening (left) of the oesophagus.

Midoesophageal diverticula are usually small pulsion diverticula of no particular consequence. The underlying motility disorder does not usually require treatment. Some pulsion diverticula may fistulate into the trachea (see **Figure 62.62**), but this is more common with traction diverticula in granulomatous disease.

Epiphrenic diverticula are pulsion diverticula situated in the lower oesophagus above the diaphragm (**Figure 62.65**). They may be quite large, but cause surprisingly few symptoms. They again probably reflect some loss of coordination between an incoming pressure wave and appropriate relaxation of the LOS. This needs to be acknowledged in the surgical management of the patient. The diverticulum, in isolation, should not be assumed to account for a patient's illness just because it looks dramatic on a radiograph. Large diverticula may be excised, and this should be combined with a myotomy from the site of the diverticulum down to the cardia to relieve functional obstruction.

Summary box 62.21

Oesophageal diverticula

- Diverticula are indicators of a motor disorder and not necessarily the cause of symptoms

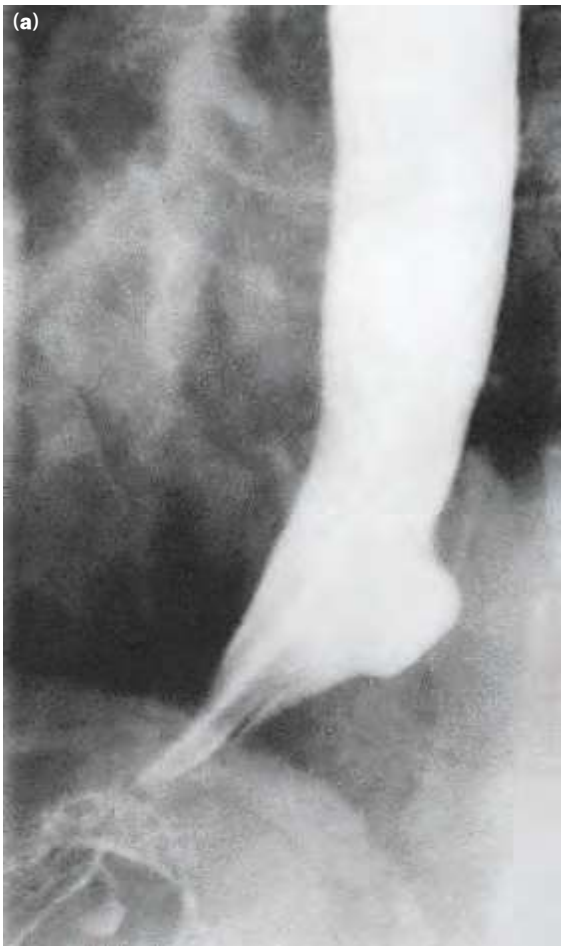
Diffuse intramural pseudodiverticulosis is a rare condition in which there are multiple tiny outpouchings from the lumen of the oesophagus. The pseudodiverticula are dilated excretory ducts of oesophageal sebaceous glands. It is questionable whether the condition produces any symptoms in its own right.

OTHER NON-NEOPLASTIC CONDITIONS

Schatzki's ring

Schatzki's ring is a circular ring in the distal oesophagus (**Figure 62.66**), usually at the squamocolumnar junction. The

Richard Schatzki, 1901–1992, American radiologist.



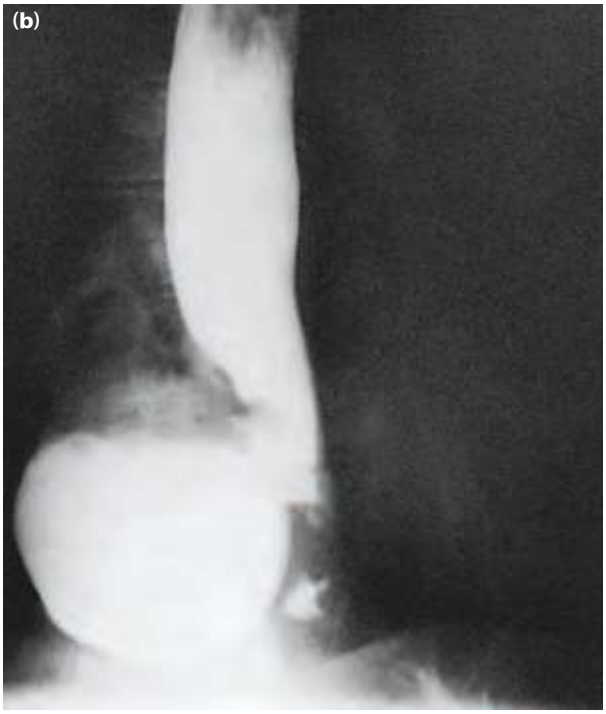


Figure 62.65 Epiphrenic diverticulum proximal to the gastro-oesophageal sphincter. **(a)** Small and asymptomatic; **(b)** large, symptomatic and appearing as a gas-filled bubble on the chest radiograph.

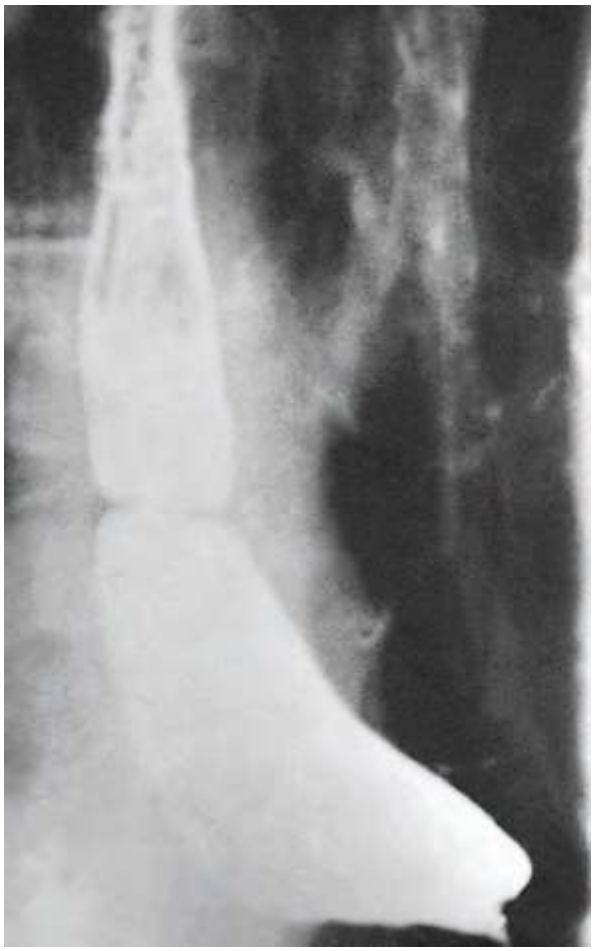


Figure 62.66 Schatzki's ring, a thin submucosal web completely encircling the whole of the lumen, usually situated at the squamo-columnar junction.

cause is obscure, but there is a strong association with reflux disease. The core of the ring consists of variable amounts of fibrous tissue and cellular infiltrate. Most rings are incidental findings. Some are associated with dysphagia and respond to dilatation in conjunction with medical antireflux therapy.

Oesophageal infections

Bacterial infection of the oesophagus is rare, but fungal and viral infections do occur. They are particularly important in immunocompromised patients.

Oesophagitis due to *Candida albicans* is relatively common in patients taking steroids (especially transplant recipients) or those undergoing cancer chemotherapy. It may present with dysphagia or odynophagia. There may be visible thrush in the throat. Endoscopy shows numerous white plaques that cannot be moved, unlike food residues (**Figure 62.67**). Biopsies are diagnostic. In severe cases, a barium swallow may show dramatic mucosal ulceration and irregularity that is surprisingly similar to the appearance of oesophageal varices (**Figures 62.68 and 62.69**). Treatment is with a topical antifungal agent.

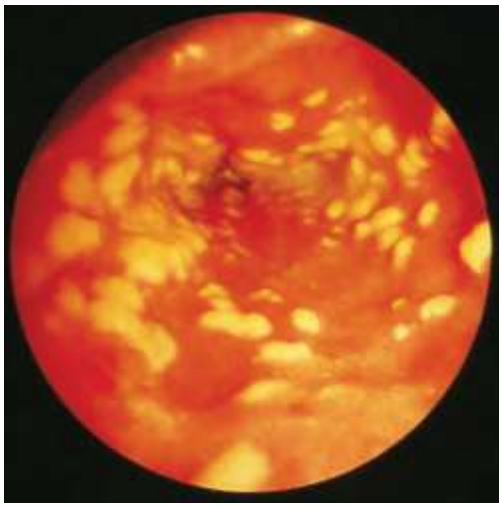


Figure 62.67 Endoscopic appearance of oesophageal candidiasis.

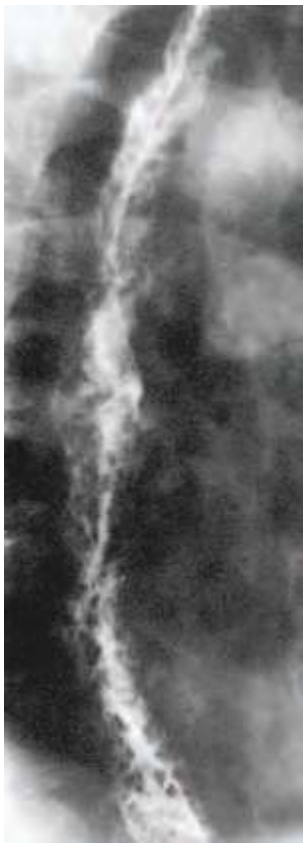


Figure 62.68 Oesophageal candidiasis with shaggy appearance of mucosal defects.

Dysphagia and odynophagia can also be caused by herpes simplex virus and cytomegalovirus (CMV). With the former, there may be a history of a herpetic lesion on the lip some days earlier, and endoscopy may reveal vesicles or small ulcers with raised margins, usually in the upper half of the oesophagus. CMV infection may be apparent in graft-versus-host disease following bone marrow transplantation. It has a characteristic endoscopic appearance with a geographical, serpiginous border. In both cases, endoscopic biopsy is diagnostic.

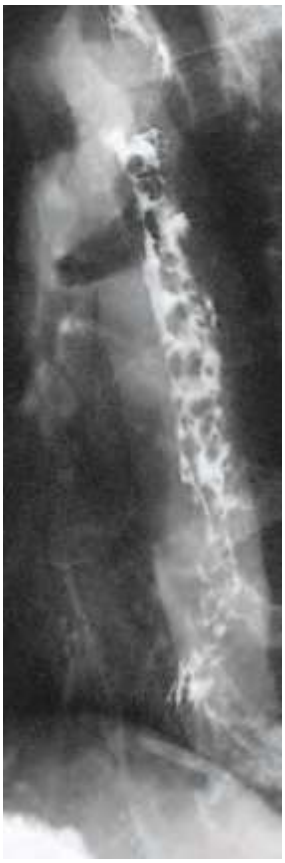


Figure 62.69 Oesophageal varices with smooth outline of the filling defects.

Chagas' disease

This condition is confined to South American countries, but is of interest because oesophageal symptoms occur that are similar to severe achalasia. It is caused by a protozoan, Trypanosoma cruzi, transmitted by an insect vector. Parasites reach the bloodstream and, after a long latent period, there is damage particularly to cardiac and smooth muscle. Destruction of both Auerbach's and Meissner's plexus leads to acquired megaoesophagus.

Crohn's disease

The oesophagus is not commonly affected by symptomatic Crohn's disease. However, pathological studies indicate that it may be present in 20% of patients without symptoms. Symptoms are often severe, and a diagnosis of reflux oesophagitis is usually made on the basis of retrosternal pain and dysphagia.

Burrill Bernard Crohn, 1884–1983, gastroenterologist, Mount Sinai Hospital, New York, NY, USA, described regional ileitis in 1932.

Endoscopy shows extensive oesophagitis that extends much further proximally than reflux oesophagitis. Biopsies may be diagnostic, but may show only non-specific inflammation. In severe cases, deep sinuses occur, and fistulation has been described. Crohn's oesophagitis is said to respond poorly to medical treatment and, although balloon dilatation of strictures and surgical resection for multiple internal fistulae have both been described, these interventions should be used with great caution.

Plummer–Vinson syndrome

This is also called the Paterson–Kelly (or Paterson–Brown Kelly) syndrome or sideropenic dysphagia. The original descriptions are vague and poorly supported by evidence of a coherent syndrome. Dysphagia is said to occur because of the presence of a postcricoid web that is associated with iron deficiency anaemia, glossitis and koilonychia. The classic syndrome is rarely complete. Some patients may have oropharyngeal leukoplakia, and this may account for an alleged increased risk of developing hypopharyngeal cancer.

Webs certainly occur in the upper and middle oesophagus, usually without any kind of associated syndrome. They are nearly always thin diaphanous membranes identified coincidentally by contrast radiology. Even symptomatic webs that cause a degree of obstruction may be inadvertently ruptured at endoscopy. Few require formal endoscopic dilatation.

Vascular abnormalities affecting the oesophagus

Several congenital vascular anomalies may produce dysphagia by compression of the oesophagus. Classically, this results from an aberrant right subclavian artery (arteria lusoria). However, the oesophagus is more commonly compressed by vascular rings, such as a double aortic arch. Dysphagia occurs in only a minority of cases and usually presents early in childhood, although it can occur in the late teens. Treatment is usually by division of the non-dominant component of the ring.

In adults, acquired causes include aneurysm of the aorta, diffuse cardiac enlargement and pressure from the left common carotid or vertebral arteries. It is rare that symptom severity justifies surgical intervention.

Mediastinal fibrosis

This rare condition can occur alone or together with retroperitoneal fibrosis. The cause is unknown and, although the major consequences are usually cardiovascular as a result of caval compression, dysphagia can occur. The existence of irreparable cardiovascular problems usually precludes surgical intervention on the oesophagus.

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Henry Stanley Plummer, 1874–1936, American internist and endocrinologist.
Porter Paisley Vinson, 1890–1959, Physician, the Mayo Clinic, Rochester, MN, who later practised in Richmond, VA, USA.
Adam Brown Kelly, 1865–1941, surgeon, Ear, Nose and Throat Department, the Royal Victoria Infirmary, Glasgow, UK.
Donald Rose Paterson, 1863–1939, surgeon, Ear, Nose and Throat Department, the Royal Infirmary, Cardiff, UK.
Vinson, Kelly and Paterson all described this syndrome independently in 1919.