

EMBRYOLOGY

The oesophagus develops in two parts; the upper segment which with the trachea evolves from the pharyngeal foregut and the lower part from the pre-gastric segment. The upper segment is divided by a ridge and completed by the 30th day forming the trachea and the upper segment of the oesophagus. Abnormalities in this embryological development results in the congenital defects of oesophageal atresia and trachea-oesophageal fistula.

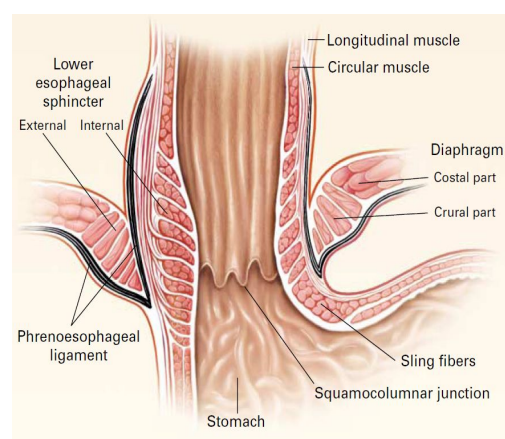
ANATOMY

The pharynx is surrounded by three constrictor muscles. The inferior constrictor divides posteriorly into the cricopharyngeus and thyropharyngeus components. There is a potential defect between the inferior constrictor and the cricopharyngeus which is important in the development of Zenker's diverticulum. The oesophagus begins at the level of C6 and is the most posterior hollow viscus in the neck and thorax. It enters the abdominal cavity through the crura of the diaphragm. The intra abdominal segment of the oesophagus is 4-5cm in length and is supported by the phreno-oesophageal ligament as it passes through the diaphragm. The oesophagus has two muscle layers; an inner circular and an outer longitudinal bundle each with a spiral component. It is striated in the upper third, smooth in the lower third, and mixed in the middle third. The mucosa of the oesophagus is stratified squamous epithelium with interspersed mucous glands. The mucosa of the junctional region between the oesophagus and the stomach consists of columnar type epithelium forming a "barrier zone" between the oesophagus and acid

secretory part of the stomach.

PHYSIOLOGY

Swallowing is accomplished through a complex series of voluntary (oral phase) and involuntary (pharyngeal and oesophageal) actions. The oral and pharyngeal phases of swallowing are controlled by the XI and XII cranial nerves. The oesophagus at rest is closed at both ends with a negative intraluminal pressure that varies with respiration. The upper sphincter (cricopharyngeus) is an anatomical sphincter exerting a pressure of 20-30 cm of water. The lower oesophageal sphincter (LOS) is approximately 4 cm in length without identifiable anatomical landmarks and functions as a physiological sphincter with a high pressure zone of 10-20 cm of water. Peristalsis in the oesophagus consists of an initial propulsive wave taking 8-12 seconds followed by secondary waves when the primary fails to propel the food bolus down the oesophagus.



FACTORS PREVENTING GASTRO-OESOPHAGEAL REFLUX

It is now generally accepted that the intra-abdominal segment of the oesophagus, (which is under the influence of the positive intra-abdominal pressure) is the most

important factor in maintaining LOS competence. Extrinsic factors such the pinch cock action of the crura of the diaphragm, phreno-oesophageal ligament and angle between the oesophagus and fundus of the stomach (angle of His) are of lesser importance. Gastro-oesophageal reflux (GOR) occurs in normal patients but only for short periods. Normal oesophageal motility is essential to ensure rapid clearance of GOR thus preventing damage to the mucosal lining of the lower oesophagus (oesophagitis).

Special investigations

- **Chest and abdominal X-rays**

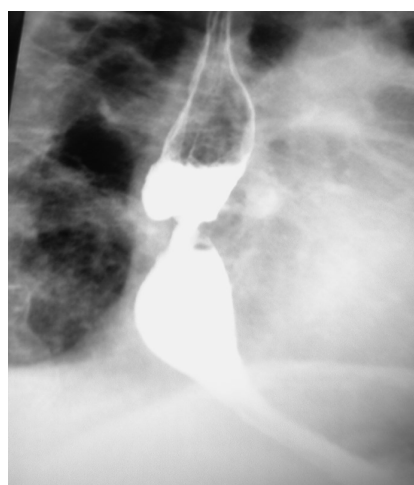
Oesophageal dilatation due to achalasia or a carcinoma of the oesophagus with lymph node metastases may show widening of the mediastinum or manifest as a fluid level on an AP or lateral chest x-ray. A large para-oesophageal hernia may also produce a fluid level (figure 2). Lung changes due to spill over from oesophageal hold-up are an important pointer to oesophageal disease and can be easily mistaken for pulmonary tuberculosis or a lung abscess. The gas bubble in the stomach is characteristically absent in patients with achalasia of the oesophagus.



Chest X-ray showing fluid level

- **Barium swallow and meal**

Contrast studies of the oesophagus, stomach and duodenum form an important part of the work-up in patients with suspected oesophageal disease, particularly if there is a history of dysphagia. Without a barium swallow, conditions such as a pharyngeal pouch may be easily missed by the endoscopists and there is a real risk of perforation. Apart from demonstrating intrinsic and extrinsic pathology of the oesophagus and cardia, barium studies will also show gross motility disorders of the oesophagus. In this respect cineradiography using barium mixed with bread or a marshmallow may help to elucidate some of these disorders. The length of a malignant stricture and axial deviation are useful markers of advanced disease (figure 3). Barium studies are also useful to distinguish between a sliding and para-oesophageal hernia. Barium is not injurious to the lung if aspirated. Gastrografin however is hygroscopic and if aspirated causes serious lung injury. If there is a chance of aspiration or a trachea-oesophageal fistula is suspected, gastrografin should not be used.



Barium meal showing a high-grade peptic stricture from long-standing GORD

- **Endoscopy**
Endoscopic examination of the oesophagus, cardia, stomach and duodenum is always indicated in patients with suspected oesophageal disease. It is superior to contrast studies in detecting early mucosal changes (eg. oesophagitis) and allows biopsy and brush cytology of suspicious lesions. Oesophageal dilatations are now preferably done with one of the fibre optic endoscopic techniques. In cases with dysphagia, many clinicians prefer to request a contrast swallow first to make an anatomical diagnosis before proceeding to flexible endoscopy.
- **CT scanning and endoscopic ultrasonography**
These imaging techniques are used mainly in patients with suspected or proven malignancies of the oesophagus to assess the extent of local and distant metastases (ie. liver). CT PET scanning may also be used to detect distant metastases.
- **Manometric studies**
This is the most reliable investigation in the assessment of patients with suspected motility disorders of the oesophagus (figure 4). Its role in the evaluation and treatment of patients with gastro-oesophageal reflux disease (GORD) is less clear and is mostly of academic interest.
- **24 hour pH monitoring**
Ambulatory systems allowing 24 hour monitoring of the pH levels in the lower oesophagus with a thin naso-oesophageal probe has facilitated the differentiation between normal and pathological gastro-oesophageal reflux. This investigation is indicated in patients to confirm GORD (in the absence of mucosal disease) and to investigate extra-oesophageal

manifestations of GORD, such as chest pain, hoarseness, chronic cough and asthma. A disposable pH probe that is attached endoscopically to the distal oesophagus has also been developed.

GASTRO-OESOPHAGEAL REFLUX DISEASE (GORD)

GORD is due to incompetence or inappropriate (premature) relaxations of the LOS. It is one of the most common gastrointestinal conditions seen in Western countries and is often associated with other conditions such as gallstones, diverticular disease and functional bowel syndrome. Many of these patients will have an associated sliding hiatus hernia with displacement of the oesophago-gastric junction into the chest. It must be stressed, however, that many patients with a hiatus hernia do not develop reflux and conversely severe gastro-oesophageal reflux may occur without a demonstrable hernia.

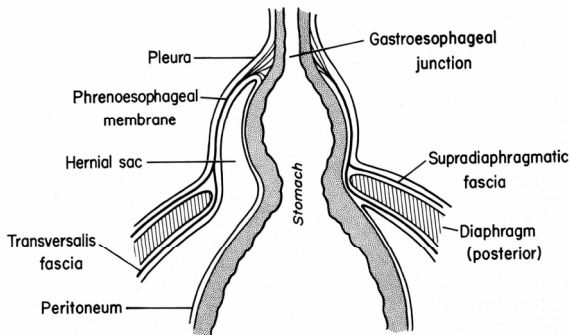
Symptoms

The symptomatology of GORD may vary from being completely asymptomatic to presenting with a complication. The most typical symptoms associated with GORD are "heartburn", described as burning sub-sternal or epigastric distress and acid regurgitation associated with meals, stooping or lying down. The latter may lead to aspiration, contribute to asthma and cause hoarseness. There is increasing awareness of the association between GORD and atypical symptoms such as chest pain, asthma, coughing and hoarseness. However, a causal relationship may be difficult to prove.

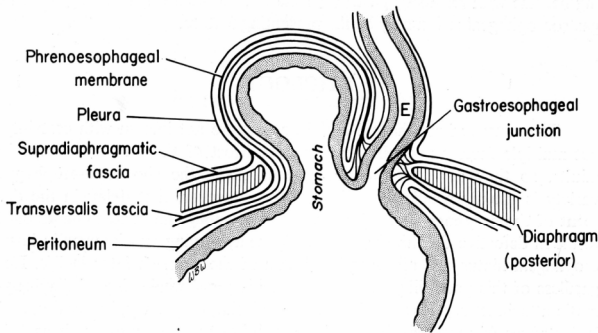
Special investigations

- **Barium meal** will show the hernia with gastric folds above the diaphragm. Reflux of barium can be demonstrated but is not a

reliable indicator of pathological GORD. The study can also help to distinguish between a sliding and para-oesophageal hernia (figure 5).



Sliding Hiatus hernia



Para-oesophageal hernia

- **Endoscopy** is important to assess oesophagitis (which is poorly demonstrated on barium meal) and to exclude other associated conditions of the stomach and duodenum. Endoscopy is mandatory to exclude malignancy in cases with an associated stricture.
- **Oesophageal motility studies** are not essential except when associated motility disorders are suspected. 24 hour ambulatory pH monitoring is indicated to confirm gastro-oesophageal reflux in cases with severe reflux symptoms without macroscopic evidence of oesophagitis on endoscopy or, when reflux symptoms do not respond to appropriate therapy.

Complications

Oesophagitis of varying severity is frequently seen in patients with GORD. In severe cases, discrete or circumferential ulceration may develop which can be complicated further by stenosis and more rarely bleeding. In cases of severe and longstanding reflux oesophagitis, metaplastic columnar epithelium replaces the squamous epithelium of the lower oesophagus to form a Barrett's oesophagus. The latter is potentially pre-malignant with a 5-10% lifetime risk of developing an adenocarcinoma in the columnar epithelium.

Treatment

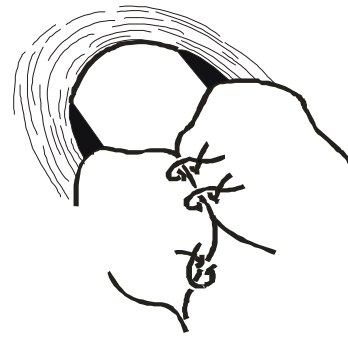
The majority of patients with GORD have mild symptoms without associated oesophagitis, commonly referred to as non-erosive reflux disease (NERD). Most of these patients are controlled with lifestyle modifications and self-medicate. In the presence of typical symptoms (heartburn and acid regurgitation) and the absence of danger signs/symptoms (loss of weight, anaemia, dysphagia) a young patient may be given empirical treatment. Others may need further investigation to confirm reflux and to exclude other upper gastrointestinal pathology. Treatment is aimed primarily at preventing pathological gastro-oesophageal reflux and its complications. Lifestyle modifications include weight reduction, avoidance of situations that will increase intra-abdominal pressure (stooping, heavy lifting) and elevation the head of the bed (not pillows). It is recommended that patients should avoid fatty foods which may decrease gastric emptying. Smoking and excessive alcohol intake should also be discouraged. Patients should be advised to take smaller meals, particularly at night and to avoid going to bed on a full stomach. In elderly patients non-steroidal anti-inflammatory drugs are often a

predisposing factor to oesophagitis and should be stopped if possible. PPIs are the most widely prescribed agents for the medical management of GORD. H2 Receptor antagonists (H2-RA) are still used for mild symptomatic disease but are not effective in healing erosive oesophagitis. Prokinetic drugs have been advocated but there is increasing evidence that these are not effective. It is difficult to provide clear guidelines for medical treatment, particularly with regard to which patients should stay on maintenance treatment. As a general rule, maintenance PPIs are required in those patients with oesophagitis as the relapse rate following cessation of therapy is up to 80% within 12 months of stopping therapy. For the rest, "on demand" PPI therapy to control symptoms will suffice.

Antireflux Surgery

With the advent of laparoscopic surgery, more patients are now referred for surgery. (Figure 6). The success rate is about 80% but it should be noted that side effects such as dysphagia, gas related symptoms and diarrhoea may be incapacitating and difficult to treat. Thus, patients with GORD should not be referred for surgery before they have had adequate conservative treatment which includes life-style adjustments and PPI therapy. Severe and persistent oesophagitis despite adequate medical treatment is a clear indication for surgery but this is uncommon today with modern medical treatment. Young patients who require maintenance treatment with PPIs to control symptoms and oesophagitis are also strong candidates for surgery. Some patients with troublesome regurgitation due to poor lower oesophageal sphincter pressure do not respond well to PPIs are also potential candidates for surgery. Complications such as stenosis are no longer a strong indication for surgery. Most of these patients are elderly and will respond to repeated dilatations

and continuous PPI therapy.



Nissen Fundoplication

PARA-OESOPHAGEAL HERNIA

This is less common than sliding hiatus hernia. Unlike the sliding hiatus hernia, the fundus of the stomach herniates alongside the oesophagus while the oesophago-gastric junctions, remains below the diaphragm. Most of these hernias are asymptomatic, even when the entire stomach is displaced into the chest. It does not predispose to gastro-oesophageal reflux except when there is an associated sliding component. These hernias have the potential to develop serious complications such as a gastric volvulus with obstruction, incarceration, strangulation and pulmonary complications. However, these complications are rare and in asymptomatic patients surgery is therefore not recommended.

MOTILITY DISORDERS

Any disturbance of the complex swallowing mechanism and or coordinated peristalsis of the oesophagus may lead to serious impairment of swallowing and propulsion of food down the oesophagus into the stomach.

1. Pharyngeal dysphagia

Psychiatric disorders of swallowing must be distinguished from the motor disorders of the cricopharyngeus sphincter. These functional disorders

are seen in psychiatric patients who have a fear of choking. They become "rabbit-chewers" and take a long time before propelling the food bolus into the hypo pharynx. This may ultimately lead to malnutrition, vitamin and iron deficiencies and may result in the Plummer-Vinson Syndrome (Fe-deficiency anaemia, glossitis, koilonychias, and cervical web). This disorder must be distinguished from "globus hystericus", where patients complain of a "lump in the throat" sensation and which is relieved by continuous swallowing.

2. Motor disorders of the upper sphincter of the oesophagus

There are many disorders that may cause disturbance of pharyngo-oesophageal function. These include bulbar poliomyelitis, cerebrovascular accidents, myasthenia gravis and radical oropharyngeal surgery. Premature contraction of the cricopharyngeus muscle after relaxation during swallowing is another important cause of dysphagia and may be the principle underlying cause of pharyngo-oesophageal diverticular. Inadequate relaxation of the cricopharyngeus muscle (erroneously called cricopharyngeal spasm) on swallowing causes upper dysphagia and can result in formation of a false (Zenker's) diverticulum.

3. Motor disorders of the body of the oesophagus and lower oesophageal sphincter

A number of motor disorders may affect the body of the oesophagus which, in broad terms, can be divided into those affecting bolus transit and those who don't (Table 1). The disorders can also be divided into "Primary" (only the oesophagus involved) and "Secondary" (oesophagus involved secondary to a generalised systemic disorder or due to reflux disease). It is often difficult to distinguish between these disorders, particularly during the

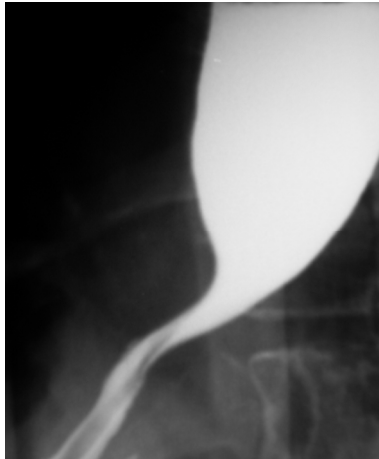
early stages of the disease. GORD may induce motility disorders such as diffuse oesophageal spasm and must be excluded before a diagnosis primary motility disorder is made.

ACHALASIA

Hypo-functional disorders

In this condition there is failure of the motor function of the body of the oesophagus and absence of relaxation of the LOS. The aetiology remains obscure but a viral cause has been implicated in the destruction of Auerbach's nerve plexus seen in these patients. Chagas' disease, which is seen in South America and caused by a trypanosome, manifests in a similar way. The oesophagus gradually dilates over a variable period (often years) until it becomes grossly dilated taking on a sigmoid configuration. It affects any age but is more commonly seen in the 20-40 year age group. There is no gender preponderance. The onset of achalasia is usually insidious. Solids and, characteristically, fluids "stick" in the lower oesophagus, particular when taken cold. Pain is usually absent, but may be present during the early stage of the disease. Regurgitation becomes a problem once the oesophagus starts to dilate. This occurs commonly at night with soiling of the pillow, and may be complicated by aspiration pneumonia, bronchiectasis or lung abscesses. Malnutrition and loss of weight may be seen in the advanced stage of the disease. The diagnosis is often missed during the early stage of the disease when the oesophagus is normal in size on barium meal. Oesophageal stasis may be misinterpreted as heartburn (possibly due to fermentation of food) leading to the erroneous diagnosis (GORD). The diagnosis is more readily made once the oesophagus starts to dilate. Barium meal study will reveal absence of peristalsis of the body of the oesophagus, a dilated oesophagus with a fluid level and a "bird's beak"

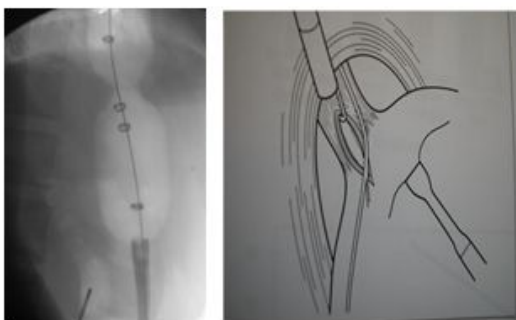
appearance at the level of the LOS. (Figure 7) Endoscopy is important to exclude a malignant lesion at the oesophago-gastric junction mimicking achalasia. The diagnosis is confirmed on manometric studies showing absence of peristalsis in the body of the oesophagus and failure of adequate relaxation of the LOS.



Barium meal showing the typical distal "bird's beak" tapering.

Treatment

Medical treatment with drugs such as prokinetics, antispasmodics, nitrites, or even calcium channel blockers are invariably unsuccessful. Intra-sphincteric injection with botulinum toxin (Botox) usually provides short-term relief but is expensive. Forceful pneumatic balloon dilatation (Figure 8) gives good results in 70-80% of cases but there is a 50% long-term failure rate and it is less satisfactory in younger patients. The procedure carries a 2-4% risk of perforation. Oesophago myotomy (Heller's operation), which is now performed laparoscopically, provides better long-term results and is now increasingly recommended as first line treatment (Figure 9). The operation consists of dividing the muscles of the lower



oesophagus and is extended onto the stomach for 1 cm. A partial anti-reflux procedure is usually added to prevent GORD.

Scleroderma is an important secondary cause of oesophageal motility failure but unlike achalasia, the LOS becomes incompetent with resultant (GORD).

HYPERFUNCTIONAL DISORDERS

1. Diffuse esophageal spasm

In this condition oesophageal contractions are simultaneous, repetitive and may be of greater amplitude. The pain is often intense but there is usually no hold-up of food. The pain may simulate angina pectoris with radiation into the neck and jaw. The cause is unknown in most instances. Treatment is mainly conservative.

2. Nutcracker oesophagus

Another condition which may be associated with gastro-oesophageal reflux disease is the so-called nutcracker oesophagus. This condition is characterised clinically by chest pain and manometrically by high amplitude co-ordinated peristaltic waves.

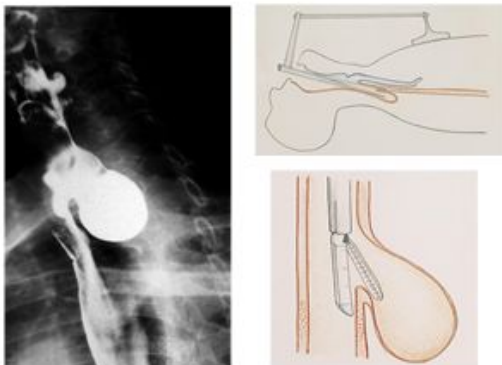
The medical treatment of these hypermotility disorders is usually unsatisfactory unless an underlying cause (such as GORD) can be identified and treated. Surgery is usually not indicated.

There is a group of conditions that cannot be categorised under the broad groups of hyper- or hypo-motility. These include tertiary contractions seen commonly in older patients and a large group of "non specific motility disorders" which have non-specific symptomatology.

DIVERTICULA

Pharyngo-oesophageal (Zenker's)

This is a pulsion diverticulum emerging posteriorly between the inferior constrictor muscle of the pharynx and the cricopharyngeus muscle (Figure 10). The pathogenesis remains uncertain but uncoordinated contraction of the cricopharyngeus has been implicated. The diverticulum usually emerges on the left side, enlarges gradually and may extend into the mediastinum. The diverticulum usually occurs in elderly patients. Symptoms are progressive as it enlarges and consist of cervical dysphagia, foul breath, noisy deglutition and regurgitation of food particles. The latter may be complicated by aspiration pneumonia. Weight loss and malnutrition are common in elderly and neglected cases. It is also a common cause of inadvertent perforation of the cervical portion of the oesophagus during endoscopy. The conventional treatment of surgical excision and cricopharyngeal myotomy is now superseded by endoscopic staple-assisted oesophago-diverticulostomy (Figure 11) or by septotomy using a flexible endoscope and energy source by Argon beam or electro-coagulation.



Zenker's diverticulum treated by endoscopic diverticulostomy

Traction diverticula are usually found in the mid-oesophagus and results from contiguous pathology such as tuberculosis.

Epiphrenic Diverticula are rare and is mostly the result of functional or mechanical oesophageal obstruction. These are situated in the lower 10cm of the supra-diaphragmatic portion of the oesophagus. In most cases it is an incidental finding on barium meal and the symptomatology relates in the main to the underlying cause rather than to the diverticulum per se. The treatment is directed at the underlying cause and diverticulectomy is only indicated when it is large and associated with entrapment of food.

OESOPHAGEAL PERFORATION

There are a variety of causes of oesophageal perforation which can be divided into two broad groups; instrumental and non-instrumental.

Instrumental

- Endoscopy
- Oesophageal dilatation
- Intubations of stents for malignant obstruction
- Injection sclerotherapy for varices

Non-instrumental

- Post emetic
- This is caused by excessive Valsalva or vomiting. It presents in three forms:
- Mallory-Weiss
 - Submucosal haematoma
 - Free rupture (Boerhaave's syndrome)
 - Foreign body
 - Penetrating injuries
 - Anastomotic leak

This is potentially a lethal condition particularly when the thoracic segment of the oesophagus is involved as these results in mediastinitis. Any delay in diagnosis and treatment increases morbidity and mortality.

Symptoms and signs

The severity of symptoms depends to a large extent on the site of the injury and whether it is a localized or free perforation (ie. into the pleural cavity). Most patients will present with some

degree of pain, respiratory distress, dysphagia and fever.

The diagnosis is often delayed in the non-instrumental group, either due to late presentation such as in the post-emetic group when patients are frequently intoxicated, or when the leak is small and contained. On the other hand, the presentation is dramatic when there is free perforation into the pleural cavity, particularly after instrumentation. This gives rise to severe respiratory distress and pleuritic pain, crepitus in the neck from air escaping into the mediastinum and shock, due to septicaemia. Low oesophageal perforations may simulate other chest and intra-abdominal catastrophes.

The negative intra-thoracic pressure is responsible for rapid and widespread contamination of the mediastinum with saliva and gastric content, giving rise to a virulent necrotising mediastinitis and pleural contamination.

Special investigations

X-rays of the chest are useful for the early diagnosis. Free air in the retro-oesophageal space in the neck and posterior mediastinum are diagnostic. Pleural effusion is common and is seen mostly on the left with distal perforations.

Gastrografin swallow is indicated to locate the site of the leak but it is important to note that there is an appreciably high false negative rate. Endoscopy is usually contra- indicated unless it is done to remove a foreign body or if an experienced endoscopist intends to close the perforation with endoscopic clips or with a covered expandable stent.

Treatment

Treatment is often complex and depends on many factors: ie. cause of, site and extent of the perforation, presence or absence of associated oesophageal pathology and duration

of perforation.

The general principles of treatment include supportive treatment with intravenous fluids, broad spectrum antibiotics, maintaining an adequate airway and meticulous debridement and drainage of the contaminated space. Most patients will require long-term nutritional support.

Cervical oesophagus

Perforations at this level usually respond to conservative treatment. Patients are kept nil per mouth or their intake is restricted to clear fluids and broad spectrum antibiotics are administered. The formation of an abscess or signs of spread into the mediastinum requires urgent drainage.

Thoracic oesophagus

Localised perforations without mediastinitis can also be treated conservatively. The passage of a fine bore feeding tube into the stomach or proximal jejunum is often advised for nutritional support.

Transmural perforation of the thoracic oesophagus requires urgent thoracotomy and exploration of the mediastinum. Repair of the perforation soon after the event may be possible but breakdown with fistula formation is frequent.

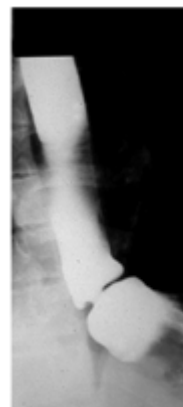
Late perforation in desperately ill patients poses a major management problem. The principles of drainage, antibiotics and nutritional support also apply here, but some would advocate a more radical approach such as oesophageal exclusion with or without an oesophageal resection and the creation of a cervical oesophagostomy and gastrostomy. The oesophagus is then replaced at a later date with colon or the stomach. The mortality remains high despite all these innovative methods.

DYSPHAGIA

Dysphagia is a very important presenting symptom in diseases of the pharynx and oesophagus and requires careful evaluation to determine the underlying cause. The term odynophagia is used when this is associated with severe pain. Tables 2 and 3 lists the causes respectively with some examples in Figures 12 and 13.

Table 2 Causes of dysphagia	
Benign	Malignant
Peptic(GORD)	squamous carcinoma
Caustic	adenocarcinoma
Webs	metastases
Schatzki ring	
Motility disorders	
Drug induced	
Post Nissen fundoplication	
Eosinophilic oesophagitis	

Table 3 Causes of odynophagia
Hypermotility disorders
Candidiasis
Herpes simplex
Drugs
- antibiotics (eg. tetracyclins)
- NSAIDs
- Ascorbic acids
- Emepromium bromide



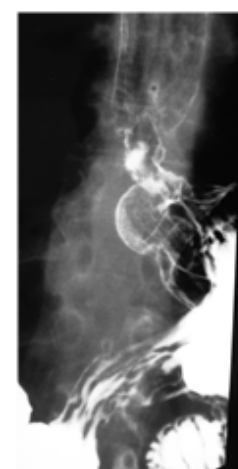
Schatzki ring



bolus obstruction



Caustic stricture



OG junction carcinoma



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