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Department of surgical diseases No. 2

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DISEASES OF ESOPHAGUS

Textbook for students of 4 courses medical faculty of faculty surgery

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This teaching guide covers main issues about etiology, pathogenesis, clinical features, laboratory and instrumental diagnostics and complications of Diseases of esophagus.

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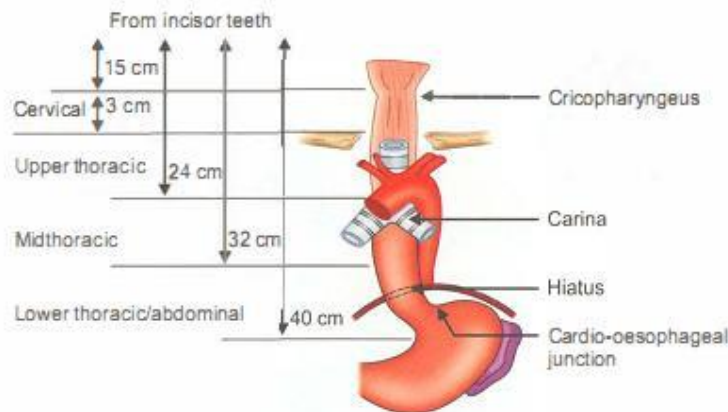
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SURGICAL ANATOMY

- Oesophagus is 25 cm in length, extending from the cricopharyngeal sphincter to the cardio-oesophageal junction (Fig. 1).



Division of oesophagus into 4 parts based on distance from incisor teeth Fig. 1

- Cardio-oesophageal junction lies to the left of T11 vertebra. It is identified at endoscopy by a Z-line.
- It is a jagged line where the oesophageal mucosa changes to gastric mucosa.
- Collar of Helvitius: It is the site at which the circular muscles of oesophagus turns to oblique muscles of the stomach at the incisura.
- It runs in the posterior mediastinum as a continuation of pharynx. 2 cm of this tube lies below the diaphragm.
- Physiological constrictions (Table 1)

Physiological constrictions			
Constrictions	Distance from incisor teeth	Diameter of oesophagus	Problems
1. Cricopharyngeal	15 cm	14 mm	Foreign body lodgement
2. Aortic and bronchial	25 cm	15–17 mm	Perforations during endoscopy
3. Diaphragmatic sphincter	40 cm	16–19 mm	Malignancy

Muscle layers

- It has an inner circular layer and an outer longitudinal layer.
- Upper 1/3rd has striated muscle fibres.
- Lower 2/3rd has smooth muscle fibres.

Mucosa

- The entire oesophagus is lined by squamous epithelium except last 3 cm which is lined by columnar cells. The columnar cells are similar to gastric mucosa but oxyntic and peptic cells are absent.
- Mucosa is the toughest coat of oesophagus.
- The smooth oesophageal mucosa gets transformed into rugal folds.

Lymphatic drainage

- Upper oesophagus drains into the left and right supra-clavicular nodes.

LYMPHATIC DRAINAGE OF OESOPHAGUS

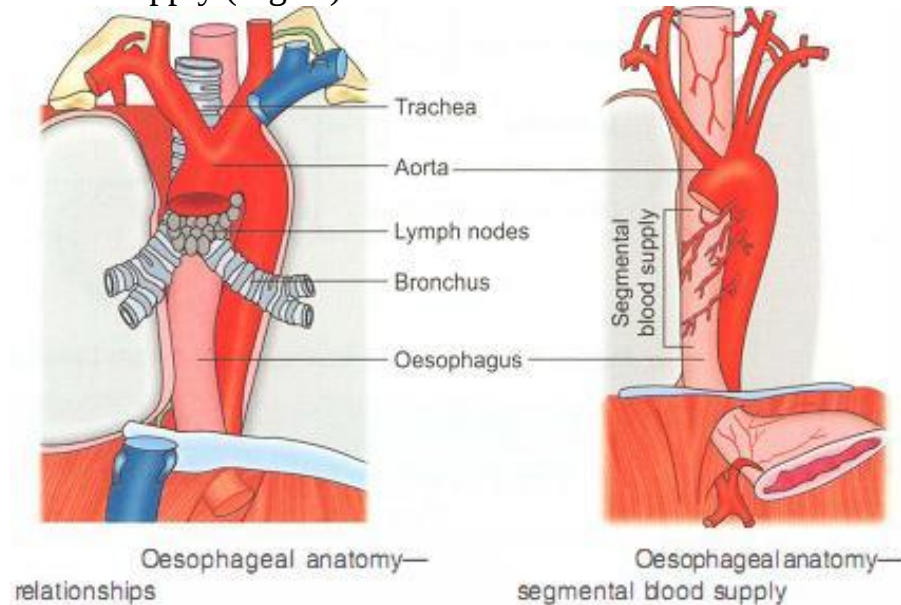
- Extensive lymphatic plexus in the submucosa which explains the easy spread
- Like in the colon they are classified as periesophageal, paraesophageal and lateral oesophageal lymph nodes
- Thus throughout the entire length of oesophagus various groups of lymph nodes are enlarged such as deep cervical, mediastinal, subcranial, hilar, diaphragmatic, para and periesophageal, gastric, coeliac, etc.
- Presence of coeliac nodes indicate inoperability
- Longitudinal lymphatics are 6 times more than transverse. Hence, distant spread occurs fast—satellite nodules occur proximal and distal to the growth. This is the reason why 10 cm margin must be given in resection of the oesophagus

- Middle oesophagus drains into the tracheobronchial nodes and paraesophageal nodes.
- Lower oesophagus drains into lymph nodes along the lesser curvature of stomach and then into coeliac nodes. Involvement of coeliac nodes indicate inoperability.

Nerve supply

The parasympathetic nerve supply is by vagus nerve through extrinsic and intrinsic plexuses. The intrinsic plexus has no Meissner's network and Auerbach's plexus is present only in the lower two-thirds.

Blood Supply (Figs 2)



Figs 2

Arterial

- Cervical oesophagus: Mainly from branches of the inferior thyroid artery.
- Upper thoracic oesophagus: Mainly from branches of the inferior thyroid artery and less consistently from anterior esophagotracheal branch from aorta.
- Mid and lower thoracic oesophagus: Supplied by bronchial arteries.
- Lower oesophagus: Small branches of the left gastric artery. Rich internal arterial anastomosis is present in the oesophagus and in the stomach.

Hence, extensive mobilisation of oesophagus can be done without compromising viability.

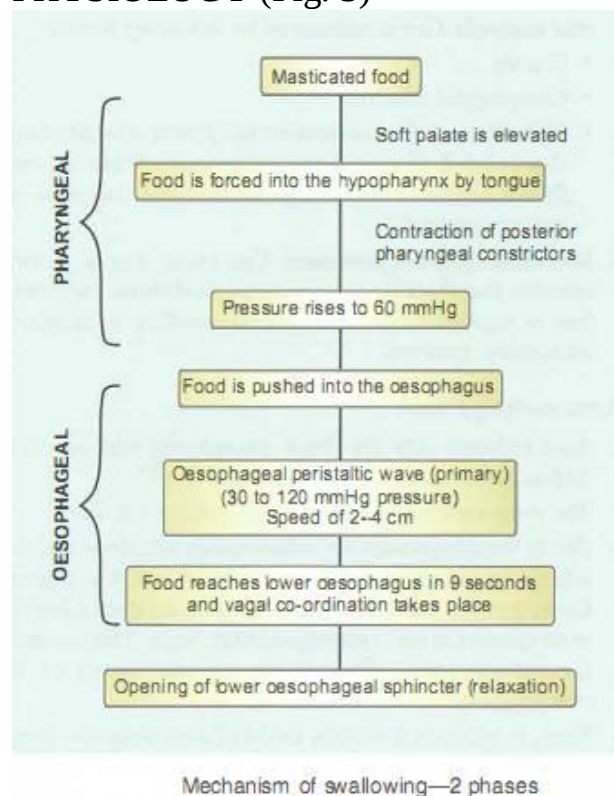
Venous drainage

Veins accompany corresponding arteries. Importantly, thoracic oesophagus drains into azygos and hemiazygos veins.

• Table 2

Surgical anatomy of the oesophagus—key points		
Surgical anatomy	Significance	Surgical points of interest
1. Upper oesophageal sphincter	Dense cricopharyngeus muscle	Zenker's diverticulum through Killian triangle
2. Lower oesophageal sphincter	High pressure zone at gastro-oesophageal junction	Weakness causes reflux oesophagitis
3. Oesophageal mucosa	Toughest coat of oesophagus	Important in oesophageal anastomosis
4. No serosa, no mesentery	Requires mobilisation	Leak rate is high, easy spread of carcinoma
5. Helicoidal muscle	Helps in peristalsis but it recoils due to elasticity	Hence, once resection takes place—specimen 'shortens' or retracts—anastomosis may become difficult especially in abdominal anastomosis
6. Segmental arterial supply	Extensive mobilisation can be done without compromising blood supply	Hence, transhiatal oesophagectomy (THE) is done with ease
7. Lower end of oesophagus—veins	Rich intercommunicating veins between portal and systemic veins	Oesophageal varices occur here
8. Azygos vein crossing oesophagus in thorax	Can get injured and bleed	Resection of midoesophageal tumours
9. Mucosal and submucosal lymphatics out number capillaries	Oesophageal tumours extend over a long distance within oesophageal wall	Need for total oesophagectomy and cervical anastomosis + lymph node clearance

PHYSIOLOGY (Fig. 3)



The main function of the oesophagus is to transfer food from the mouth to the stomach. Voluntary contraction of the oropharynx pushes food into the upper

oesophagus through a relaxed cricopharyngeal sphincter. Then, due to primary and secondary peristalsis, the food bolus is transferred to the stomach.

THE PHARYNGEAL STAGE—REFLEX

- Food stimulates mechanoreceptors
- Soft palate is pulled upwards so that food will not enter nasal cavities
- Vocal cords adduct and larynx is pulled upwards against epiglottis so that food will not enter trachea
- Cricopharyngeal sphincter opens
- Muscular wall of the pharynx then contracts from above downwards.

- Cricopharyngeal sphincter is closed at rest. It helps in preventing regurgitation of oesophageal contents into the respiratory passage.
- Lower oesophageal sphincter (LOS): It is a physiological sphincter at the gastro-oesophageal junction. It is 3-4 cm long with a resting pressure of 20-25 mmHg. Hence, it is known as high pressure zone (HPZ).
- This HPZ prevents reflux of gastric contents into the oesophagus. Cigarette smoking affects this zone and so, smokers have a high incidence of reflux oesophagitis.

GASTRO-OESOPHAGEAL REFLUX DISEASE

- Loss of competence of LOS leads to gastro-oesophageal reflux disease (GORD). The competence of LOS can be affected by obesity, smoking, excessive eating, etc. Sliding hernia is associated with GORD.
- As a result of reflux of gastric acid, extensive inflammation of the lower oesophagus occurs which results in various forms of oesophagitis.

Types

1. Acute: Following alcohol, bums, stress.
2. Chronic: It is associated with hiatus hernia or following esophagojejunostomy.

It is mainly precipitated by

1. Structurally defective LOS secondary to inflammatory injury.
2. Inadequate abdominal length (short length) of oesophagus- precipitates a reflux when gastric distension occurs.
3. Ineffective oesophageal pump affecting clearance of food into stomach. This is influenced by following actors:
 - Gravity
 - Oesophageal motility
 - Salivation-Saliva neutralises the minute amount of acid that is let following a peristaltic wave. When salivary flow is decreased (following radiotherapy), the reflux can get exaggerated.
4. Increased gastric pressure: Can occur due to pyloric stenosis and diabetic gastroparesis. Following vagotomy, loss of receptive relaxation occurs resulting in increased intragastric pressure.

Aetiopathogenesis

- Acid refluxes into the lower oesophagus and produces diffuse inflammation with multiple ulcers.
 - The symptoms are worse when the patient lies down.
 - Due to vagal hyperactivity, inflammation and ulcers develop which produce severe longitudinal muscle spasm.
- Consequently, the cardia is drawn up into the thorax, leading to an increase in the oesophago-cardiac angle. This increases the reflux. Later, fibrosis causes shortening of the oesophagus.
- Thus, it becomes a vicious circle of oesophagitis-longitudinal muscle spasm-displacement of oesophagus- increased regurgitation.

Clinical features

Most common presentation of GORD is history of heartburn and regurgitation. Heartburn is confined to epigastrium and retrosternal areas, does not radiate to the back. Clinical presentation of the GORD can be classified as follows: Atypical symptoms can be so many and can thus confuse the picture.

Oesophageal: Dysphagia, regurgitation, heartburn

Gastric: Early satiety, belching, bloating, nausea

Pulmonary: Asthma, wheezing, aspiration, cough, dyspnoea, bronchitis, hoarseness of voice due to damage to vocal cords, etc.

Ear, nose, throat: Waterbrash, globus, hoarseness

Cardiac: Chest pain

What is globus? Sensation of a substernal lump (globus)

When this occurs during fasting, it is termed 'globus hystericus'.

It is a neurotic symptom in patients with emotional instability.

- Retrosternal pain: It is burning in nature and becomes worse on lying down. The pain reduces in the sitting position. The pain is described as heartburn and can be confused for angina pectoris. It is relieved on taking antacids.

- Heartburn is otherwise called pyrosis.

Occult blood in stools and streaks of blood in the vomitus are common.

- Anaemia and weakness are uncommon features.
- Dysphagia: Transient difficulty in swallowing results from spasm due to inflammation of the lower end of oesophagus. Late dysphagia is due to stenosis or stricture of the oesophagus. Belching is not uncommon.

Johnson-DeMeester's scoring system

- Three important symptoms are taken into consideration Modified Savary

Miller classification of reflux oesophagitis

Grade I: Single or isolated erosion at or above GE junction

Grade II: Multiple non-circumferential erosions above GE junction

Grade III: Circumferential erosions above GE junction

Grade IV: Chronic lesion-stricture, ulceration/short oesophagus

Grade V: Columnar epithelium in continuity with Z line (Barrett's oesophagus)

Investigations (Table 3)

Investigations of GORD		
To detect structural abnormalities	To detect increased exposure to gastric juice	To detect functional abnormalities
A. Barium swallow • Detects hiatus hernia (prone position) • Detects stricture or any rings • Extrinsic compression can be detected • Can also detect small carcinoma B. Flexible endoscopy • Can detect hiatus hernia or diverticulum, etc. • Can detect oesophageal abnormalities	A. Flexible endoscopy and to biopsy the Barrett's oesophagus B. Grading of oesophagitis by endoscopy Grade I: Small, circular, nonconfluent erosions Grade II: Linear erosions lined with granulation tissue that bleeds on touch Grade III: Erosions with circumferential loss of the epithelium---cobblestone oesophagus Grade IV: Stricture	A. Oesophagus manometry • It is indicated when motor abnormality is suspected such as achalasia, diffuse spasm • Can also detect scleroderma polymyositis • Average LOS pressure < 6 mmHg, suspect GORD • High resolution manometry is more accurate

Table 3

OESOPHAGEAL MANOMETRY
• Information about LOS – Resting pressure – Length – Relaxation • Quality of oesophageal peristalsis • Manometry is essential. To place the pH probe 5 cm above upper border of LOS for ambulatory pH monitoring.
24-HOUR pH MONITORING IS THE GOLD STANDARD
• Indications: When symptoms are certain but endoscopy is normal, atypical symptoms • It involves the transnasal placement of a pH measuring electrode in the lower oesophagus. The pH electrode monitors the changes in intra-oesophageal pH over 24 h • A 24 h pH profile is thus obtained that provides information on frequency, duration and pattern of reflux • A reflux episode is defined as a pH drop to below pH 4 • Thus identification, type and duration of reflux is noted

- Barium swallow' in the Trendelenburg's position (head down position) can demonstrate the reverse flow of barium into the lower end of the oesophagus (from the stomach).
- Oesophagoscopy may reveal red, angry looking mucosa in the lower end of the oesophagus.
- Oesophageal manometry to detect motility disorders.
- 24-hour pH monitoring is the gold standard.

Medical management

Treatment of uncomplicated GOD can be discussed under following headings.

I. Lifestyle modification

- Stop smoking
- Stop alcohol
- Control obesity
- Avoid coffee, chocolate and coke
- Head up-propped-up position

- Avoid stooping
- Avoid tight garments

II. Drugs

- Antacids with alginate-antireflux floating alginate.
- Proton pump inhibitors: Pantoprazole 40 mg, esomeprazole till 20 mg may have to be given for one or two months or till symptoms are controlled. These are antisecretory drugs.
- Prokinetics: Itopride 50 mg can be given 2-3 times a day for 8 weeks on empty stomach. Prokinetics enhance motility. Cisapride and mosapride are not favoured because they can cause cardiac arrhythmias.

III. Mucosa protective agents

- Sucralfate colloidal bismuth-cytoprotective agent.
- It is a sucrose sulfate-aluminium complex which binds to the mucosa. Thus it protects mucosa of GI tract against hydrochloric acid.
- Colloidal bismuth compounds.

IV: Endotherapy

- Endoscopic plication/suturing
- Enteryx injection
- Plexiglass microspheres (PMMA): Through an endoscopic needle, microspheres suspended in gelatin are injected. Gelatin is absorbed and spheres cause tissue bulking.

V. Surgery: Indication

To summarise, quit smoking, decrease alcohol intake, avoid overweight and start walking. Do not drink coke and do not eat chocolates, take mucosal protective agents.



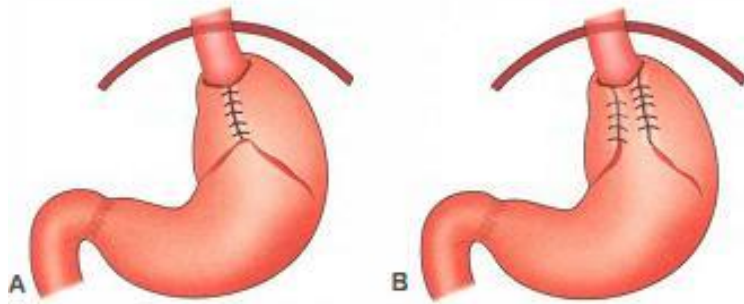
1. Intractable pain

2. Complications such as haemorrhage or stricture

- The results of antireflux surgery are good with a small mortality rate (0.1 to 0.5%). However, careful selection of patients depending upon their symptoms and lifestyle are important factors.

Types of surgery

1. Nissen's total fundoplication (Figs 4): The aim is to restore 2-4 cm of intra-abdominal oesophagus by reducing the hernia, followed by repair of the hiatus.



Nissen's fundoplication

Figs 4

- In this operation, fundus of the stomach is mobilised by dividing short gastric arteries.
- Fundus is brought behind the oesophagus and wrapped in front of oesophagus (Fig. 5). It is a loose wrap (loppy Nissen's).



Fundus brought on the right side

Fig. 5

- Diaphragmatic defect is repaired by using nonabsorbable sutures such as nylon or silk. This is the operation which serves all aims.
- Mortality and morbidity should be minimised.

Laparoscopic fundoplication

- Most popular today
- Minimal morbidity and mortality
- Early discharge, within 1-2 days
- Early recovery

All operative steps that are performed in open surgery are carried out here with a 'better vision' in laparoscopic method.

Principles of fundoplication

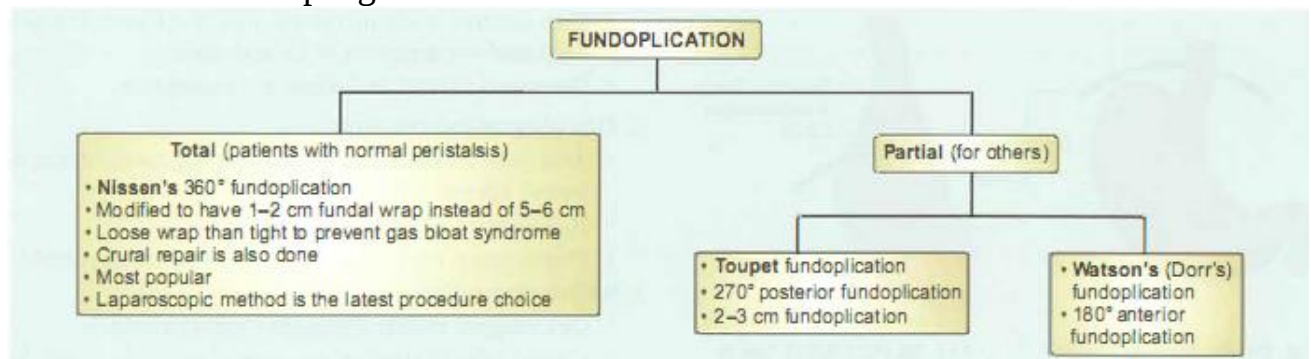
- 360° gastric fundoplication should be no longer than 2 cm.
- It should be constructed over a 60 F bougie.
- Fundoplication should be placed in the abdomen without tension.
- Only 'fundus' should be used to wrap (undus relaxes- body does not relax on swallowing).
- Vagus should not be damaged because it may result in failure of sphincter to relax.
- Lengthen the oesophagus with a Collis gastroplasty (in cases of short oesophagus).
- Patients with normal peristaltic contractions do well with 360°

Nissen's fundoplication-or others, two-thirds partial fundoplication may be the procedure of choice.

Complications

- Too tight a replication may result in dysphagia or gas bloat syndrome wherein belching is prevented.

2. Partial fundoplication (Toupet) solves the above problem wherein undus is sutured around the back of oesophagus or Dorr's, where undus is sutured anterior to the oesophagus.



3. Belsey Mark IV operation

- There are 3 layers of sutures to be placed in this operation via a thoracotomy.
- First row: Four interrupted silk mattress sutures are placed between the anterior surface of the oesophagus and adjacent fundus of the stomach so as to wrap the stomach around the anterior two-thirds of the oesophagus.
- Second row: Sutures are placed between oesophago-gastric junction and under surface of the diaphragm to maintain the junction below the diaphragm.
- Third row: Posterior crural sutures are placed to tighten the opening.

4. Hill's repair: Median arcuate ligament repair.

- In this, the long intra-abdominal segment of the oesophagus is firmly fixed below the diaphragm by anchoring the gastro--oesophageal junction to the crura just above the median arcuate ligament. This is described as posterior gastropexy.

5. Recurrent GORD: Success of revision surgery is much less than primary surgery. Partial gastrectomy with Roux--en-y reconstruction is the final

surgery for GORD. It diverts bile and pancreatic juice and reduces acid secretion.

HIATUS HERNIA

Definition

Abnormal protrusion of abdominal viscus through the oesophageal hiatus into the chest.

Types

1. Sliding hernia or oesophagogastric hernia: It is the commonest type of hiatus hernia, accounting for about 80% of cases (Fig. 6). It may be associated with GORD.

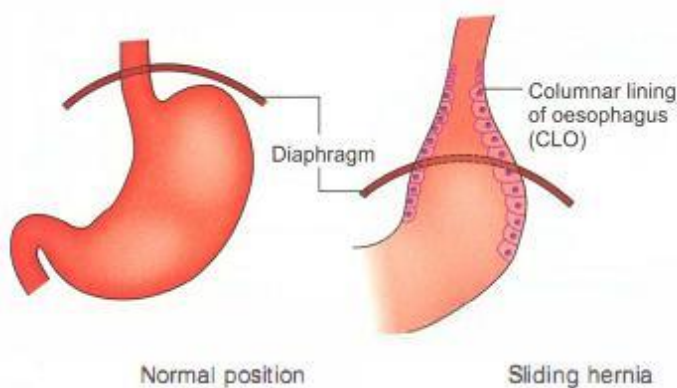


Fig. 6

2. Rolling or para-oesophageal hernia
3. Mixed hernia
4. Massive herniation

Common symptoms

1. Symptoms due to reflux: Regurgitation and heartburn are the two most common symptoms.
2. Symptoms due to complications: They are dysphagia, odynophagia, haematemesis and melaena.
3. Nonoesophageal symptoms: They are asthma and chest pain.

SLIDING HERNIA (Fig. 6)

Anatomical factors which prevent sliding hernia

1. Presence of 2 cm of intra-abdominal oesophagus.
2. The angle of His: The oesophago-cardiac angle of about 45° has valvular defect.
3. Mucosal folds at the oesophago-cardiac junction.
4. Positive intra-abdominal pressure which closes the abdominal oesophagus.

5. Lower oesophageal sphincter (LOS): It is a functional sphincter which increases the pressure during coughing, straining, etc.

Causes of sliding hernia:

1. Deposition of fatty tissue in the hiatus leads to weakening of the hiatus in obese individuals.
2. Advancing age resulting in muscular degeneration can predispose to hernia.
3. Raised intra-abdominal pressure due to lower abdominal tumours, pregnancy, etc.
4. Saint's triad: Gall stones, diverticulosis and hiatus hernia can occur together in a patient.

Clinical features

- Sliding hernia produces symptoms like reflux oesophagitis.
- More common in women, especially if obese.

Investigations

- Oesophagoscopy reveals varying degree of inflammation.
- During oesophagoscopy, when the patient is asked to strain (Valsalva's manoeuvre), the sphincter is seen to be more patulous and herniation of gastric mucosal folds can be seen. Reflux of the gastric acid is the most valuable sign.
- Barium meal demonstrates gastro-oesophageal reflux in the Trendelenburg position.

Treatment

I. Conservative treatment

In all cases of GORD, conservative treatment has to be tried first. The results of surgery are appreciated only when the symptoms are significant and conservative treatment fails.

Principles

I. Lifestyle changes

- Decrease in weight
- Diet control with increased intake of proteins and decreased consumption of fat and sugar.
- Decreased alcohol and tobacco consumption.

2. Oesophageal mucosa protection

- Antacids: Preparations containing alginates, cytoprotective agents.
- H₂ blockers: Ranitidine.
- Proton pump inhibitors: Omeprazole or esomeprazole.

3. Reflux prevention

- Oesophageal reflux: Cisapride, metoclopramide
- Gastric reflux: Domperidone, metoclopramide, cisapride.

4. Decision of surgery.

ROLLING HERNIA

In this condition, cardio-oesophageal junction is normal. Greater curvature of the stomach ascends into a preformed sac in the mediastinum (Fig. 7). Thus, there are no features of reflux oesophagitis but the sac containing stomach in the thorax causes compression of the heart and lung.

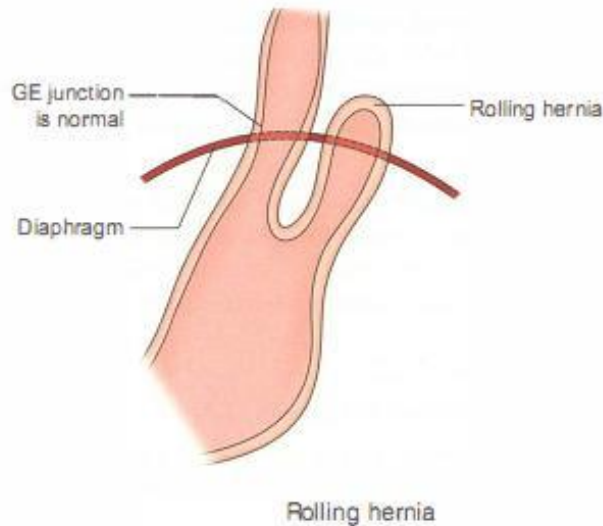


Fig. 7

Clinical features

- No retrosternal burning pain because no reflux
- Discomfort after a small meal
- Feeling of fullness after a meal or dysphagia due to large sac
- Palpitations due to compression on the heart
- Respiratory tract infection and hiccough due to irritation of phrenic nerve.

Investigation

Barium meal shows the sac in the thorax containing stomach. Sometimes, it can be upside down.

Treatment

Reduction of the sac and repair of the hiatus by using nonabsorbable suture material to approximate the right crus of the diaphragm.

MIXED HERNIA

- In this, both sliding and rolling hernia are present (Fig. 8).

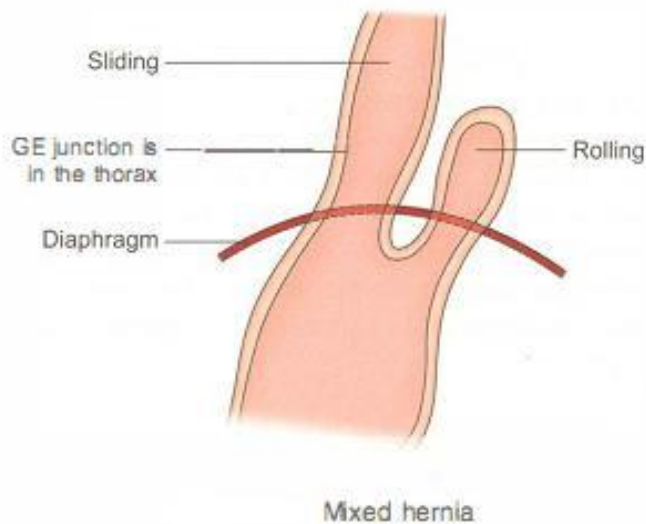


Fig. 8

- Symptoms are mixed.
- Treatment is also mixed and is done for both sliding and rolling hernia.

Complications of GORD

1. Stricture oesophagus: It is seen in middle-aged and elderly patients. Due to repeated reflux, ulcers, fibrosis and stricture develop in the lower end of the oesophagus. Early diagnosis by endoscopy followed by frequent dilatation and proton pump inhibitors will help the situation.

- Peptic strictures are difficult to manage surgically.
- Surgery is indicated in refractory cases of dilatation, in the form of gastroplasty.

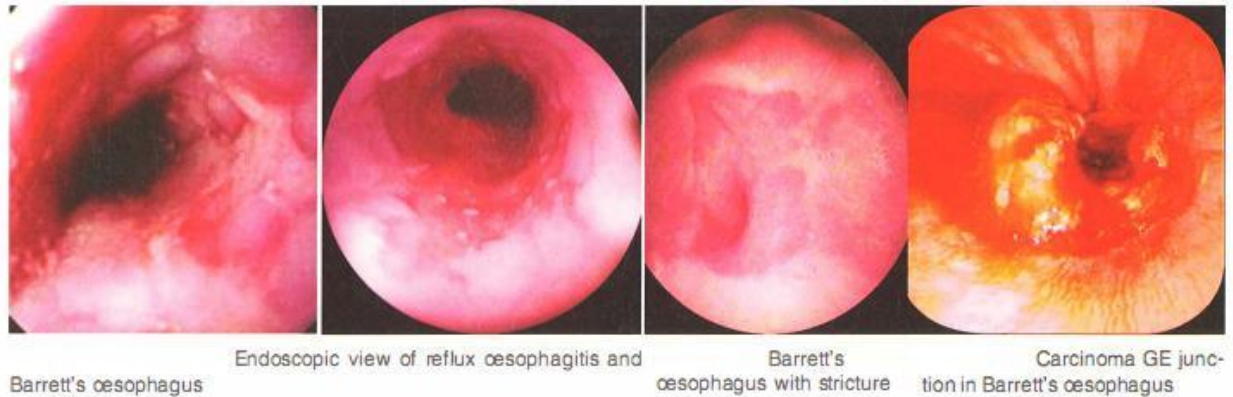
2. Oesophageal shortening is also treated by Collis gastroplasty by using stomach (short oesophagus).

3. Barrett's oesophagus: Also known as columnar-lined oesophagus (CLO).

BARRETT'S OESOPHAGUS

Definition

When columnar mucosa extends at least 3 cm into oesophagus or when it shows intestinal metaplasia, it is described as Barrett's oesophagus (Figs 9).



Figs 9

- Ulcer in the Barrett's CLO is called Barrett's ulcer.

Pathogenesis

Repeated reflux results in shifting of the oesophago-gastric junction upwards, which further increases the reflux resulting in intestinal metaplasia of middle and lower oesophagus.

Pathological types

1. Gastric type with chief and parietal cells.
2. Intestinal type with goblet cells is a marker of intestinal metaplasia. This mucosa is smooth (unlike gastric folds).
3. Junctional type: It has mucus glands and resembles gastric cardia.

Clinical types

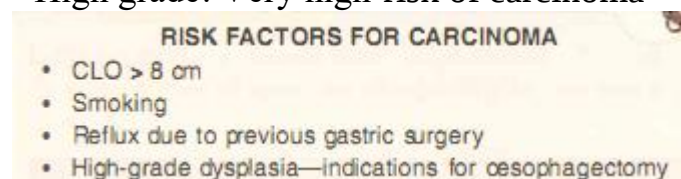
1. Long segment: Metaplastic changes involving more than 3 cm.
2. Short segment: Metaplastic changes involving less than 3 cm.

Incidence of malignancy

40 times more prone to carcinoma of the lower and middle oesophagus as compared to the general population.

Types of dysplasia

- Low grade: Negligible risk of carcinoma
- High grade: Very high-risk of carcinoma



Screening programme

- It is important to screen these patients regularly with repeat endoscopies and multiple biopsies to find dysplasia.

Treatment

- High dose proton pump inhibitors for 8 weeks.
- Laser photodynamic therapy.
- Argon beam plasma coagulation
- Asymptomatic, symptomatic Barrett's oesophagus responds well to laparoscopic antireflux surgery. Antireflux surgery also prevents progression to high-grade dysplasia or adenocarcinoma.
- Endoscopic mucosal resection or Barrett's oesophagus with low-grade dysplasia.
- Oesophagectomy in cases of high-grade dysplasia.

Complications of Barrett's oesophagus

1. Oesophageal ulcers: Barrett's ulcer-pain, bleeding and perforation.
2. Oesophageal stricture: Usually located in the middle or upper oesophagus.
 - Peptic stricture occurs in the distal oesophagus.
3. Dysplasia and adenocarcinoma.

Non-reflux Oesophagitis

This is also a condition which occurs due to several factors such as corrosives, drugs, chemoradiation, AIDS, etc. They are summarised in Table. Basic treatment is similar to that of reflux oesophagitis and avoid the causative agents.

Types of non-reflux oesophagitis		
	Agents causing	Clinical features
1. Corrosive:	Lye, acid, alkali	Burns, stricture
2. Infective:	Candida, Herpesvirus	Chronic illness, AIDS
3. Radiation:	20-40 Gy units to mediastinum	Ulceration and stricture
4. Chemotherapy	Adriamycin	—
5. Drug induced:	NSAID, doxycycline	↑ reflux

ACHALASIA CARDIA

- It is a primary oesophageal motility disorder.
- It is also called cardiospasm because of severe spasm of the circular muscle fibres of the lower end of the oesophagus. The contracted segment does not relax during the act of swallowing (achalasia = failure of relaxation). As a result of this, there is dilatation, tortuosity and hypertrophy of the oesophagus above.
- Incidence: 6 in 1,00,000 population/year.

Aetiopathogenesis

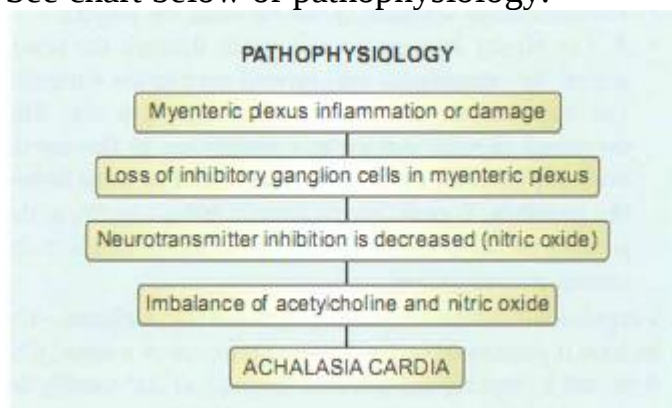
1. Idiopathic: This occurs due to absence or degeneration of Auerbach's plexus throughout the body of the oesophagus leading to improper integration of the parasympathetic impulse. This is called primary achalasia.

2. Acquired variety is seen in South American countries caused by *Trypanosoma cruzi* (sleeping sickness)--

Chagas disease. This organism destroys the ganglion cells of the Auerbach's plexus.

3. Stress and emotional factors and vitamin deficiencies are also associated with this disease.

See chart below on pathophysiology.



- Classical achalasia: Loss of ganglion cells and neural fibrosis occurs.
- Vigorous achalasia: No loss of ganglion cells but ganglionitis occurs.

Clinical features

- Women around 30-40 years of age are commonly affected.

The ratio of affected females to males is 3:2.

- Dysphagia develops slowly and it is progressive.

- Solids, by forming a bolus and aided by gravity, as they touch the contracted segment, may partially open up the sphincter. Thus, there is no dysphagia for solids. Dysphagia for liquids is an important feature and it results in regurgitation (oesophageal pseudo vomiting). The regurgitant material contains foul-smelling oesophageal contents. Malnourishment, ill health and weight loss follow soon. Dysphagia, regurgitation and weight loss form the triad of achalasia cardia.

- Recurrent respiratory tract infection due to spillage of liquids can also occur.
- Features of anaemia-glossitis, stomatitis, pallor, bald tongue.
- Retrosternal discomfort and radiation of pain to the interscapular region may be present.
- Pseudoachalasia: Tumours of cardia mimicking achalasia. Often patients present with features of recent achalasia (dysphagia). During endoscopy, some difficulties are encountered and once the scope enters the GE junction,

growth will be seen.

Investigations

1. Barium swallow

- Uniformly dilated oesophagus above, with a smooth tapering segment below-cucumber oesophagus.
- In chronic cases, it may be sigmoid-shaped (Figs 10).



Barium swallow
showing dilated oesophagus

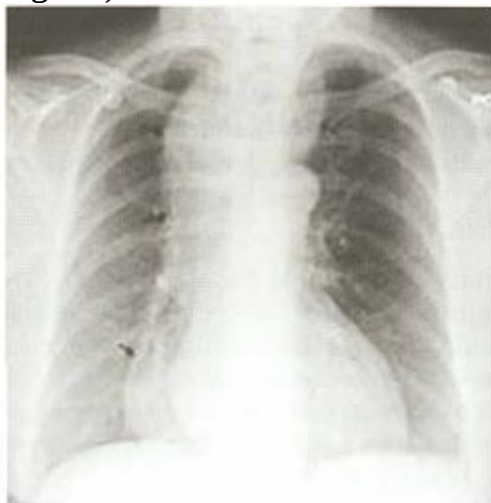


Barium swallow
showing sigmoid oesophagus

Figs 10

2. X-ray chest

- Mediastinal mass (pseudotumor) produced by dilated oesophagus can be seen (Fig. 11).



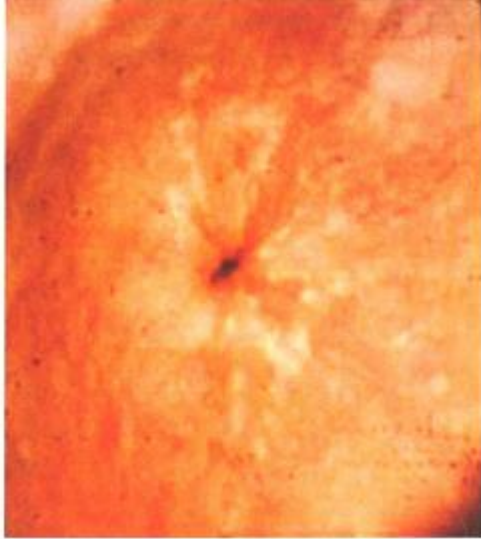
Achalasia—chest X-ray

Fig. 11

- Retrocardiac air-fluid level is seen in the lateral view.
 - Aspiration pneumonitis can be diagnosed.
3. Plain X-ray abdomen erect: Fundic air bubble is absent because of the stasis of fluid in the oesophagus.

4. Oesophagoscopy: It reveals a dilated sac containing stagnant blood and fluid due to stasis which splashes out with each heartbeat and with each respiratory movement.

LES: Lower esophageal sphincter is closed, with air insulation. It has a 'rosette' appearance.



Endoscopy showing rosette appearance

- Oesophagoscopy is also done to rule out proximal malignancy.
 - Also done to evaluate oesophagitis, stricture or a tumour at cardia.
5. Oesophageal manometry: Following features are characteristic of achalasia cardia:
- Hypertensive lower oesophageal sphincter (LOS): It does not relax on swallowing.
 - Aperistalsis in the body of oesophagus
 - Increased resting pressure in the oesophagus
6. Ultrasound: It may detect subepithelial tumour infiltration in secondary achalasia due to a distal carcinoma.

Treatment (Table 4)

Treatment of achalasia cardia			
Pharmacological therapy	Botulinum toxin injection	Pneumatic dilatation	Surgery
• Sublingually used • Nitrates and calcium channel blockers • Nitrate 5–20 mg before meal and Ca channel blockers 10–30 mg before meal • They relax smooth muscle • Nitrates have more intense effect and more side-effect vice versa with Ca channel blocker • Headache is the side-effect	• BoTx A 80–100 units are injected in one sitting • 20–25 IU are injected into each quadrant • Use 5 mm sclerotherapy needle • It acts by blocking acetylcholine release • Repeat injections are necessary as effect is temporary • Pregnancy, hypersensitivity to albumin are contraindications	• Most effective nonsurgical option • It disrupts the circular muscle fibres of LES • Polyethylene balloon is used • Dilators are 3, 3.5 and 4 cm • 70% permanent relief • Side-effect is perforation, gastro-oesophageal reflux, haemorrhage	• Modified lap. Heller's cardiomyotomy is the gold standard • 6 cm of lower oesophagus and 1 to 2 cm of proximal stomach myotomy is done • 90% success • 3% reflux oesophagitis

Table 4

- Aim of the treatment is to cure the disease.
 - More importantly, the obstruction at the lower end of oesophagus must be relieved.
 - Modified laparoscopic Heller's cardiomyotomy is the choice of surgery now.
1. Heller's cardiomyotomy: The aim is to reduce outflow resistance at the lower oesophageal sphincter.
 - With a left thoracoabdominal incision, the oesophagus and the stomach are completely mobilised.
 - The contracted segment is felt between the fingers.
 - A 7 to 10 cm long incision is made through the lower end of the oesophagus and carried over to the stomach. The muscles are cut till the mucosa bulges out. The myotomy should extend proximally up to the aortic arch and distally up to the stomach to 1 to 2 cm below the junction. Success rate is around 90%. 3 to 5% of the patients develop reflux oesophagitis which needs to be treated conservatively.
 2. Forceful dilatation: By using Pneumatic balloon-the balloon is positioned under fluoroscopic control within LOS. It should be rapidly inflated to a pressure of 300 mmHg for 15 seconds. Success rate is around 70%. 20% chances of reflux are present. Oesophageal perforation can also occur. Recurrences are common (Figs 12 and 13).



Oesophageal balloon dilators



Oesophageal narrowing of lower end

Figs 12 and 13

3. Injection treatment: Inj. Botulinum toxin is injected into the lower oesophageal sphincter (LOS) endoscopically. Injection acts by interfering with cholinergic excitatory neural activity at LOS and blocking acetylcholine release from nerve terminals. It is temporary and repeat injections are necessary.

4. Drugs: Sublingual nifedipine can produce short-term relief.

5. Endoscopic myotomy-popularly called POEM: In this, through endoscope, a submucosal injection of a mixture of indigo rouge, epinephrine, and saline is done. A 2 cm long incision is made and submucosal tunnel is created. Then circular muscle bundle is cut slowly, beginning from 7-8 cm above GE junction to 2-3 cm below GE junction.

Complication of achalasia cardia

Due to prolonged stasis and chronic Irritation, it can predispose to carcinoma of the mid and lower oesophagus (due to metaplasia). Hence, it is a precancerous condition.

- Squamous cell carcinoma is the most common type identified in a patient with achalasia.

OESOPHAGEAL PERFORATIONS

- Oesophageal perforation is a serious, acute emergency carrying a very high mortality rate. Fortunately it is not common. There are many causes and are totally different from gastric or duodenal perforations.
- Instrumentation perforations are the most common cause of perforation.
- Cervical oesophagus is the most likely site of perforation.
- Cricopharyngeal region is the most common site.

Causes

1. Instrumentation: Endoscopy, dilatation of strictures, injection sclerotherapy and laser treatment are the common causes.

2. Operative: Thyroidectomy, vagotomy, spine surgery, mediastinoscopy.
3. Traumatic: Caustic injury, sharp and blunt trauma
4. Oesophageal diseases: Carcinoma, Barrett's oesophagus
5. Spontaneous rupture: Boerhaave's syndrome.

Clinical features

- Severe pain in the neck, chest or abdomen, stiffness of the neck depending upon the site of perforation.
- Haematemesis, dysphagia, dyspnoea, hypotension, tachycardia, shock and pleural effusion are the other features.
- Mackler's triad: Chest pain, vomiting and subcutaneous emphysema. It is present only in 10 to 15% of patients.
- 'Hamman's sign' escape of air into the mediastinum resulting in "mediastinal crunch" which is produced by the heart beating against air-filled tissues.

Investigations

- The investigation of choice is plain chest X-ray which can demonstrate pneumomediastinum, subcutaneous emphysema and mediastinal air fluid levels. When in doubt, CT chest is diagnostic.
- Contrast swallow can also be done.

Treatment principles

- Infection is polymicrobial: Staphylococcus, Streptococcus, Pseudomonas and Bacteroides.
- Early surgery should be done within 24 hours. Closure of perforation and external drainage is the treatment.
- Perforation older than 24 hours-treated by temporary cervical oesophagostomy, ligation of oesophagus proximal to GE junction and feeding jejunostomy. After 6- 8 weeks, oesophagectomy and gastric or colonic pull up is done.
- Conservative treatment has risk of continuing leak, sepsis and mediastinitis.

DIVERTICULUM OF OESOPHAGUS

Types of diverticulum (Table 5)

Types of diverticulum		
Name	Situation	Aetiology
1. Zenker's diverticulum (cervical or pulsion)	Proximal to the upper oesophageal sphincter	Protrusion of oesophageal mucosa between cricopharyngeus muscle inferiorly and thyropharyngeus muscle superiorly
2. Epinephric diverticulum	Proximal to the lower oesophageal sphincter	It is due to some kind of motility disturbance, wherein protrusion occurs in the lower end
3. Parabronchial diverticulum	Midoesophagus	Tuberculosis, by causing enlargement of mediastinal nodes, fibrosis and adhesions produce traction on the oesophagus, which results in the diverticulum

Table 5

- Cervical diverticulum (Fig. 14) is the commonest type of diverticulum which can present with regurgitation of meals, aspiration of blood contents into the lungs or recurrent respiratory tract infections. It is treated by excision of the sac and repair of the defect along with cricopharyngeal myotomy-posterior midline.

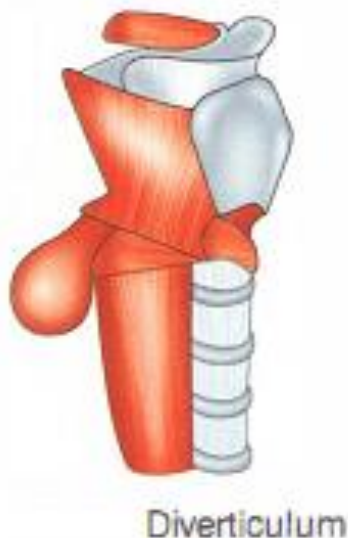


Fig. 14

DIFFERENTIAL DIAGNOSIS OF DYSPHAGIA (Fig. 15)

I. Causes from outside (extraluminal) oesophagus

- Thyroid swellings
- Cardiomegaly
- Aortic aneurysm
- Mediastinal nodes: Tuberculosis, lymphoma or secondaries
- Rolling hiatus hernia

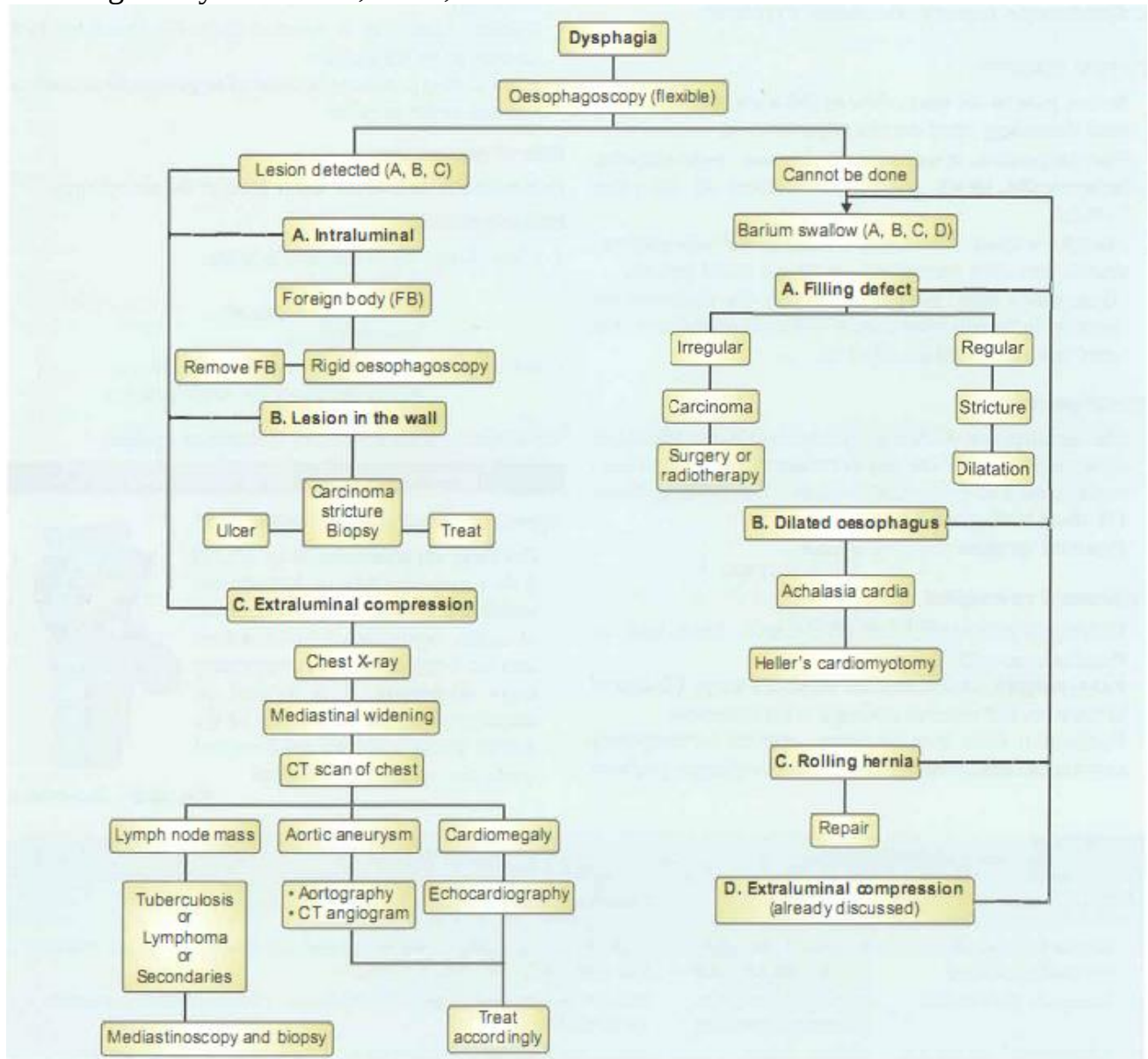
II. Causes in the wall of oesophagus (luminal)

1. Oesophageal stricture
 - Corrosive acid poisoning
 - Tuberculous stricture
2. Carcinoma oesophagus
3. Oesophageal diverticulum
4. Muscular spasm: Plummer-Vinson syndrome and achalasia cardia.

5. Tetanus

III. Causes in the lumen of oesophagus

1. Foreign body: Dentures, coins, etc.



Investigations of a case of dysphagia

Fig. 15

History

1. Acute dysphagia: Common in children. Foreign bodies are common causes. Acute dysphagia with pain suggests tonsillitis or pharyngitis.
2. Chronic dysphagia: Stricture, achalasia, Plummer-Vinson syndrome and carcinomas produce chronic dysphagia. The increasing difficulty to swallow, first to solids and later to liquids is typical of carcinoma oesophagus (in achalasia cardia, it is the reverse).
3. Age and sex: Achalasia and Plummer-Vinson's syndrome is common in females, carcinoma in men and foreign body in children.

4. Change in voice or even hoarseness with dysphagia suggests advanced laryngeal carcinoma.

1. Following are disorders of the pharyngo-oesophageal junction except:
 - A. Stroke
 - B. Myasthenia
 - C. Cricopharyngeal achalasia
 - D. Nutcracker oesophagus

2. Following are true for oesophageal lymphatics except:
 - A. Extensive lymphatic plexus in the submucosa
 - B. They are classified as perioesophageal, paraoeso-phageal and lateral oesophageal lymph nodes
 - C. Presence of coeliac nodes indicate inoperability
 - D. Longitudinal lymphatics are 6 times less than transverse
3. Which one of the following is highest percentage of premalignant condition or carcinoma oesophagus?
 - A. Tylosis B. Lye strictures
 - C. Achalasia D. Oesophagus
4. Factors or adenocarcinoma oesophagus are following except:
 - A. Barrett's oesophagus
 - B. Obesity
 - C. Reflux esophagitis
 - D. Oesophageal web
5. Factors for development of reflux disease of oesophagus include following except:
 - A. Obesity
 - B. Absence of intra-abdominal length of oesophagus
 - C. Helicobacter pylori infection
 - D. Defective angle of His
6. Clinical features of reflux disease of oesophagus include following xcept:
 - A. Heartburn
 - B. Vomiting
 - C. Epigastric pain
 - D. Regurgitation
7. Gold standard or the treatment of reflux disease is:
 - A. Barium meal
 - B. Esophagoscopy
 - C. 24-hour pH monitoring
 - D. Endosonography
8. The most effective drugs or reflux disease are:
 - A. Alginates
 - B. Antacids
 - C. H₂ receptor antagonists
 - D. Proton pump inhibitors
9. The most effective curative treatment for carcinoma oesophagus is:
 - A. Radiotherapy
 - B. Oesophagectomy
 - C. Photodynamic therapy
 - D. Chemotherapy
10. Following are features of Barrett's oesophagus except:
 - A. 3 cm or more of columnar epithelium

- B. Cardiac metaplasia
- C. Development of squamous cell carcinoma
- D. Presence of mucus secreting goblet cells (intestinal metaplasia)

ANSWERS:

1-D 2-D 3-A 4-D 5-C 6-B 7-B 8-D 9-B 10-C

Literature:

1. K Rajgopal Shenoy, Anitha Shenoy. Manipal Manual of Surgery. CBS Publishers & Distributors. Forth Edition. 429-460.
2. Harold Ellis, Sir Roy Calne, Christopher Watson. General Surgery Lecture Notes. Wiley BlackWell. 2016. 155-163
3. Dellon ES, Gonsalves N, Hirano I, et al. ACG clinical guideline: evidenced based approach to the diagnosis and management of

- esophageal eosinophilia and eosinophilic esophagitis (EoE) Am J Gastroenterol. 2013;108:679–92
4. Wolf WA, Jerath MR, Sperry SL, et al. Dietary elimination therapy is an effective option for adults with eosinophilic esophagitis. Clin Gastroenterol Hepatol. 2014.
 5. Katzka DA, Ravi A, Geno DM, et al. Cytosponge evaluation of eosinophilic esophagitis in comparison to endoscopy: accuracy, safety and tolerability. Gastroenterology. 2014;146.
 6. American Gastroenterological Association American Gastroenterological Association medical position statement on the management of Barrett's esophagus. Gastroenterology. 2011;140:1084–1091.
 7. Sikkema M, De Jonge PJ, Steyerberg EW, Kuipers EJ. Risk of esophageal adenocarcinoma and mortality in patients with Barrett's esophagus: a systematic review and meta-analysis. Clin Gastroenterol Hepatol. 2010;8:235–244
 8. Inoue H, Onimaru M, Ikeda H, et al. Clinical results of 500 POEM cases for esophageal achalasia and related diseases in a single institute. Gastrointest Endosc. 2014.
 9. Holzheimer RG, Mannick JA, editors. Munich: Zuckschwerdt; 2001. Surgical Treatment: Evidence-Based and Problem-Oriented.
 10. Redman JE, Newton R, Malhotra S, et al. Biomarkers for early detection of esophageal squamous cell dysplasia and carcinoma. Gastroenterology. 2014;146.

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