Master's Today Medical surgical disease

Focused Review of Surgical diseases

(A guide for exam takers)

Dr Konokbai Murasbek

Аннотаци(кырг):

Бул китеп медициналык мектептин студенттери жана улуттук аттестациялык экзамендерге даярданып жаткан адамдар үчүн атайын иштелип чыккан хирургиялык оорулар боюнча кеңири колдонмо болуп саналат. Хирургиялык оорулардын бардык негизги темаларын камтыган бул китеп ар бир теманын кыска, бирок мазмундуу түшүндүрмөлөрүн берет, бул аны тез жана натыйжалуу үйрөнүү үчүн эң сонун окуу куралына айландырат.

Материал "Schwartz`s principles of Surgery" жана "Sabiston textbook of Surgery" сыяктуу белгилүү хирургия боюнча окуу китептерин камтыган ишенимдүү булактарга негизделген. Ар бир бөлүм клиникалык аспектилерди терендирээк түшүнүүгө жардам берүүчү жана экзамендерге даярдануу жеңилдетүүчү фактылар жана мисалдар менен бекемделген. Китепте ошондой эле материалды жакшыраак өздөштүрүүгө көмөктөшүүчү иллюстрациялар жана схемалар бар.

Бул басылма башталгыч студенттер жана өз билимин жаңыртууну жана аттестациядан ишенимдүү өтүүнү көздөгөн тажрыйбалуу врачтар үчүн да баа жеткис ресурс болуп калат.

Annotation (Eng):

This book is a comprehensive guide to surgical diseases, specifically designed for medical school students and those preparing for national certification exams. Covering all the major topics of surgical diseases, the book provides concise yet substantive explanations of each topic, making it an ideal resource for quick and effective learning.

The material is based on authoritative sources, including well-known surgical textbooks such as " Schwartz`s principles of Surgery " and " Sabiston textbook of Surgery " Each chapter is supported by relevant facts and examples, allowing for a deeper understanding of clinical aspects and facilitating exam preparation. The book also includes illustrations and diagrams that help enhance material retention.

This edition will become an invaluable resource for both beginning students and experienced doctors looking to refresh their knowledge and confidently pass certification exams.

Аннотация(рус):

Эта книга представляет собой всеобъемлющее руководство по хирургическим заболеваниям, специально разработанное для студентов медицинских школ и тех, кто готовится к национальным аттестационным экзаменам. Включая в себя все основные темы хирургических болезней, книга предоставляет краткие, но содержательные объяснения каждой темы, что делает ее идеальным пособием для быстрого и эффективного обучения.

Материал основан на авторитетных источниках, включая известные учебники по хирургии, такие как " Schwartz`s principles of Surgery " и " Sabiston textbook of Surgery ". Каждая глава подкреплена актуальными фактами и примерами, что позволяет глубже понять клинические аспекты и облегчает подготовку к экзаменам. Книга также включает в себя иллюстрации и схемы, которые способствуют лучшему усвоению материала.

Это издание станет незаменимым ресурсом как для начинающих студентов, так и для опытных врачей, стремящихся обновить свои знания и уверенно пройти аттестацию.

ADAM UNIVERSITY
SCHOOL *of* MEDICINE

Foreword to "Focused Review of Surgical Diseases (A Guide for Exam Takers)

This book is an invaluable resource for medical students preparing for national exams and diploma confirmation. It is based on authoritative texts like "Schwartz's Principles of Surgery" and "Sabiston Textbook of Surgery," presenting structured and organized content. Each chapter covers essential topics from anatomy to modern clinical recommendations, simplifying complex concepts for easier understanding. Richly illustrated with diagrams and mnemonics, it aids in grasping difficult material and applying it in practice. Its user-friendly layout allows students to efficiently navigate topics and reinforce their knowledge, making it an essential tool for exam preparation and future medical careers. Ultimately, this guide represents a significant contribution to medical education, inspiring confidence in students as they embark on their professional journeys.

Top contributors:

Sirmbard Svetlana
The rector of Adam University, a national expert on the reform of higher professional education, associate Professor.

Dr najam wagan
My beloved student MD (general medicine)

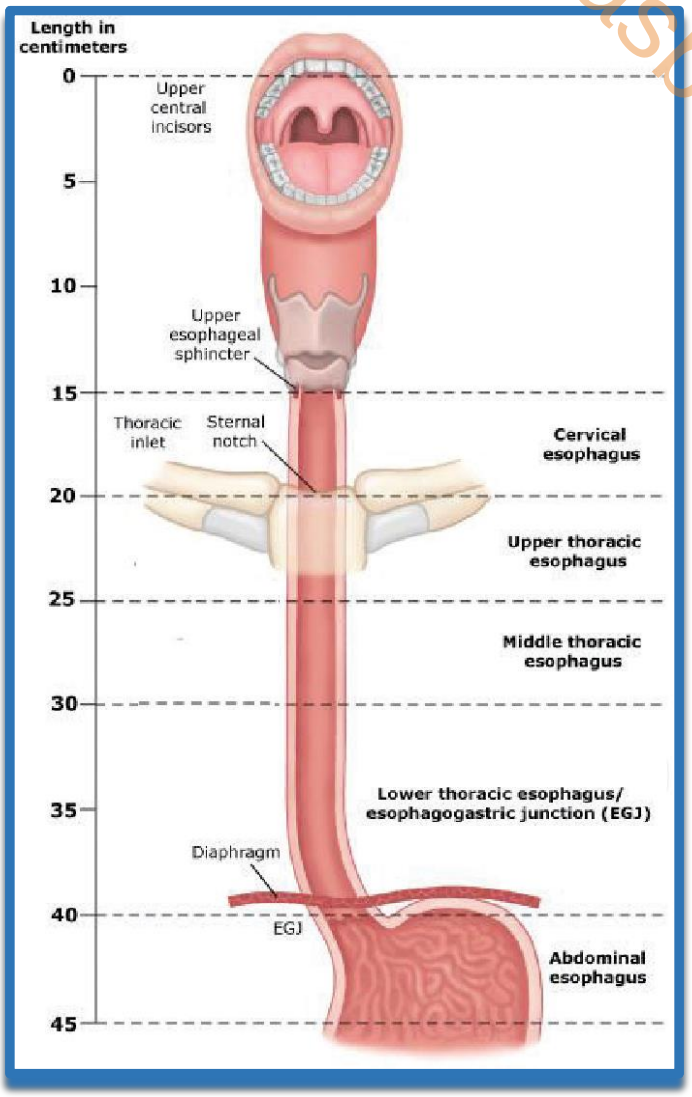
Contents

Chapter
1.

Esophagus Diseases and Surgery

- The esophagus is a muscular tube extending from the pharyngeal junction to the cardia of the stomach.

- The junction of the pharynx and esophagus is located at the level of the 6th cervical vertebra when the head is in a normal position (anteriorly corresponding to the cricoid cartilage of the larynx).



Sections of the Esophagus

- Cervical Esophagus: Approximately 5 cm long, located at the C6 to T1-2 level.

- Thoracic Esophagus: Extends from T1-2 to T10, approximately 20 cm long.

- Abdominal Esophagus: Located at the T10-T11 level, approximately 2 cm long.

Esophageal Narrowing's

- 1st Narrowing:: At the entrance of the esophagus, caused by the cricopharyngeus muscle. This is the narrowest part of the esophagus, with a diameter of about 1.5 cm.

- 2nd Narrowing: At the level of the tracheal bifurcation, with a diameter of about 1.6 cm.

- 3rd Narrowing: At the point where the esophagus passes through the diaphragm. The diameter here is 1.6-1.9 cm, caused by the lower esophageal sphincter (LES) mechanism.

Histological Layers of the Esophagus

- Mucosa: Composed of squamous epithelium.

- Muscle Layers:

    - Circular Muscle Layer: Thicker and arranged in a helical structure.

-

Longitudinal Muscle Layer: Thinner

- Serosa: The esophagus does not have a serosa.

- Cervical Esophagus: Covered by striated muscle. The circular muscle layer is thicker than the longitudinal layer.

Arterial Supply of the Esophagus

- Cervical Esophagus: Supplied by the inferior thyroid artery, which originates from the thyrocervical trunk of the subclavian artery.

- Thoracic Esophagus: Supplied by bronchial arteries, which arise directly from the aorta.

- Abdominal Esophagus: Supplied by the ascending branch of the left gastric artery and the inferior phrenic artery.

Venous Drainage of the Esophagus

- Cervical Region: Drains into the inferior thyroid veins.

- Thoracic Region: Drains into the bronchial veins, azygos, and hemiazygos veins.

- Abdominal Region: Drains into the coronary vein (left gastric vein).

Lymphatic Drainage of the Esophagus

- The lymphatics of the esophagus form a dense plexus under the submucosa. The lymphatic flow in the submucosa is primarily longitudinal, which is why esophageal cancers tend to spread longitudinally rather than transmurally. Longitudinal spread is six times more common than transmural spread.

- Cervical Esophagus: Drains into paratracheal and deep cervical lymph nodes.

- Upper Thoracic Esophagus: Drains into paratracheal lymph nodes.

- Lower Thoracic Esophagus: Drains into subcarinal lymph nodes.

- Abdominal Esophagus: Drains into superior gastric lymph nodes.

Upper and Lower Esophageal Sphincters

- Upper Esophageal Sphincter (UES): A high-pressure area formed by the cricopharyngeus muscle, which attaches to the posterior ends of the cricoid cartilage.

- Lower Esophageal Sphincter (LES): Composed of intrinsic smooth muscle and extrinsic components from the right crus of the diaphragm. The LES pressure exceeds the intra-gastric pressure by 15-25 mmHg.

- During swallowing, the LES relaxes for approximately 1 second, allowing physiological reflux

Esophageal Physiology: Swallowing

- Swallowing occurs 600-1000 times a day.

- During eating and drinking, swallowing occurs once per second, and about 70 times per hour between meals. It does not occur during deep sleep.

- The oral phase of swallowing is voluntary, while the pharyngeal and esophageal phases are involuntary.

- Peristalsis in the esophageal wall functions as a pump, while the LES allows controlled passage of food into the stomach and prevents reflux.

- The LES is also known as the gastroesophageal sphincter.

- There is no true anatomical sphincter in this region; the pressure difference prevents the reflux of stomach contents into the esophagus.

- Factors affecting LES pressure are detailed in the accompanying table.

Gastroesophageal reflux

Up to 75% of esophageal diseases is Gastroesophageal reflux. It is the condition in which stomach acid repeatedly flows back into esophagus, however definitive diagnosis is difficult, and its true incidence is unknown.

• Reflux esophagitis occurs because of abnormally prolonged contact between normal gastric content and esophageal mucosa in the distal esophagus. It is a chemical inflammation of esophageal mucosa.

•Reflux occurs in some patients, but esophagitis does not develop.

• People with esophagitis experience reflux more frequently.

• Physiological reflux is more common while standing and awake.

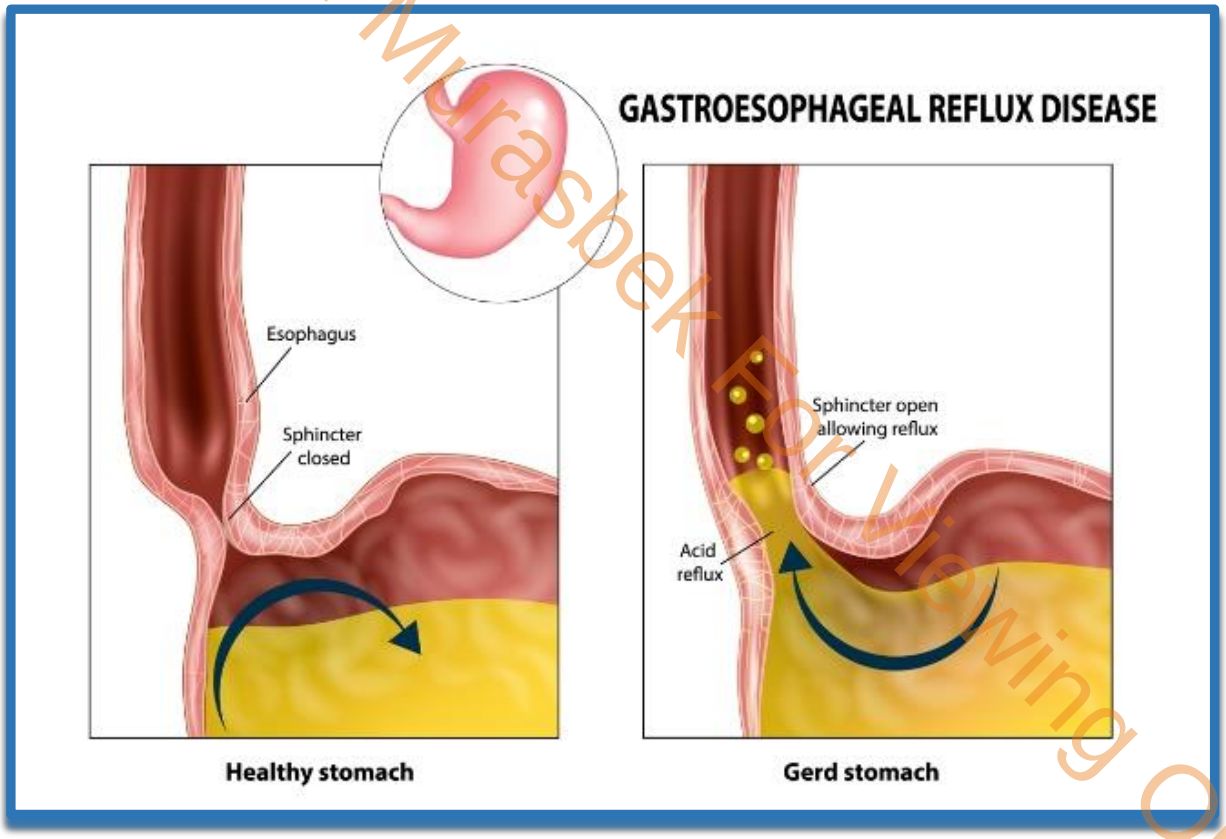
• LES pressure is significantly higher than hydrostatic pressure in the abdomen when lying down than when upright. In this lying position, it depends on the effect of pressure on the abdominal part of the sphincter.

Most important factor in the formation of reflux esophagitis is LES failure.

• Gastric fundus distension also causes a decrease in LES pressure.

•LES insufficiency criteria :

- LES pressure < 6 mmHg
- Abdominal LES < 1 cm
- Total LES length < 2 cm



Among these, the most common cause of GER is short abdominal LES. (sliding hernia)

Secondary peristaltic waves, in response to exposure of the esophagus to gastric contents, develops and the esophagus being cleaned the refluxed material thanks to these waves.

In patients with esophagitis, the process of clearing the esophagus from acid is also defective and reflux attacks last longer takes.

If normal stomach contents remain in contact with the esophagus for long time, mild esophagitis develops. Mixed contact of gastric and duodenal content causes severe esophagitis. If the combination of bile acid and acid pepsin -that causes the most severe damage to the esophageal mucosa.

Clinic

- There are no symptoms or signs specific to GER.
- Common symptoms such as burning sensation behind the sternum and regurgitation are common with many other diseases.

• Atypical symptoms due to GER may also develop~ Nausea, vomiting, fullness after meals, chest pain,

chronic cough, wheezing, feeling of suffocation, hoarse voice.

• Additionally, bronchiolitis, recurrent pneumonia, idiopathic pulmonary fibrosis and asthma are also primarily caused by GER.

Complications

• Reflux esophagitis

•Stricture

•Barret esophagitis

•Fe deficiency anemia

•Pulmonary fibrosis

•Aspiration pneumonia

•Laryngitis, asthma, paroxysmal coughing, sinusitis

Diagnosis

•GIS endoscopy (gold standard)

•24ph monitorization (Reflux is defined if the measured esophageal pH is less than 4)

•Esophageal manometry

Treatment

1) Mechanical measures and diet to reduce reflux

•It is necessary to raise the head of the bed and not eat anything until 3 hours before bedtime.

• Wearing tight clothing should be prevented, any other maneuver that can increases intra-abdominal pressure

• For overweight patients recommended losing weight.

• Intake of chocolate, alcohol, gas-forming substances and carbonated drinks that increase reflux is restricted:

• Smoking should be quit. Because it reduces lower esophageal sphincter pressure and increases reflux.

2) Reducing gastric acid or acid reflux

• Antacid may be sufficient.

• H2 receptor blockers and proton pump inhibitors reduce gastric acid secretion:

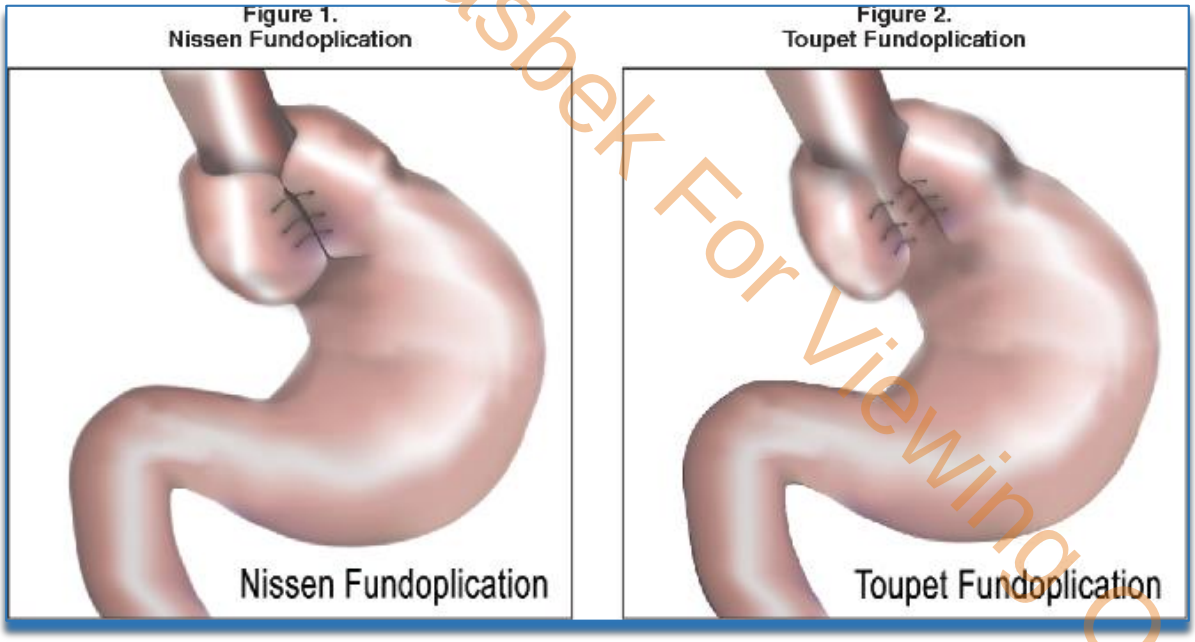
• Alginic acid forms a balloon that prevents contact between the stomach contents and the esophageal epithelium:

3)Agents that increase LES pressure

• Urocholine, metoclopramide and domperidone stimulate LES contractions:

• Cisapride stimulates esophageal contractions. However, they are not sufficient.

4) Nissen fundoplication (360-degree fundoplication)



• Ideally it should be done laparoscopically.Recurrence is < 10%.

• Too tight or long fundoplication may be cause of dysphagia.

• Other complications; Inability to belch and vomit, bloating and increased gas production.

• It is applied in the presence of Barrett's esophagus and if there is no severe dysplasia or in situ cancer. Otherwise resection is recommended.

Barrett's esophagus

It is the change of the normal squamous epithelium of the esophagus. (metaplasia)

•Barret esophagus occurs in approximately 10-15% of GER patients.

•The consequences of untreated GER disease is Barrett's esophagus

• As a result of long-term GER, first the columnar epithelium is change to the squamous cell epithelium of the esophagus. Later, in most of the patients, small intestinal epithelium develops over this columnar epithelium after metaplasia develops.

•The situation becomes clear when goblet cells appear. In order to diagnose Barrett's disease.

• Endoscopically, Barrett's esophagus may be uncomplicated or present with esophagitis, stricture,

It may be accompanied by complications such as ulceration and dysplasia it is a premalignant lesion, where the metaplastic Barrett's epithelium becomes dysplastic and progresses to adenocarcinoma.

• Antireflux surgery should be performed before dysplasia develops. If "high-grade dysplasia" is present,esophageal resection is recommended.

Hiatal hernia

1Type Sliding hernia

• It is develops as a result of weakening of the phrenoesophageal membrane

• Cardia upwards, towards the posterior mediastinum slips

• is much more common than other types.

•Median age is 48 years.

• Symptoms are usually functional due to GER It develops as a result of abnormalities.

• Retrosternal burning sensation, regurgitation and dysphagia may be observed.

• However, many people with sliding hernia most are completely asymptomatic.

•In this case, treatment is not required. Asymptomatic sliding hernia is not a pathology that needs to be corrected.

•Surgical treatment decision for sliding hernia depends on the symptoms and complications related to gastroesophageal reflux depends on the severity.

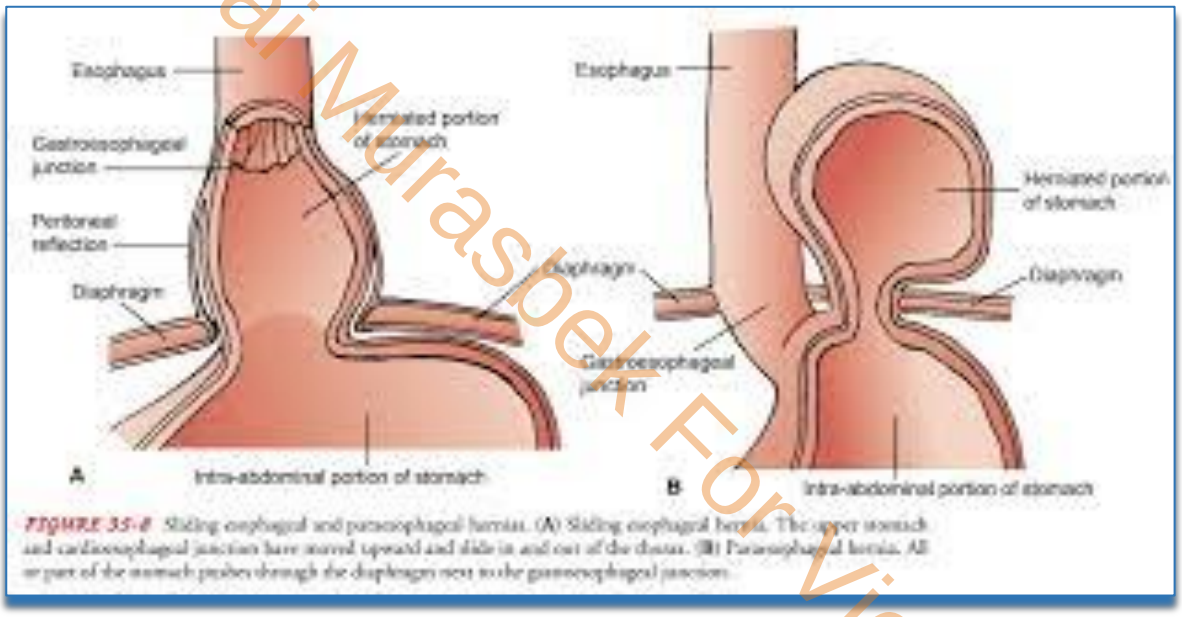


FIGURE 35-8 Sliding esophageal and paraesophageal hernias. (A) Sliding esophageal hernia. The upper stomach and cardioesophageal junction have moved upward and slide in and out of the thorax. (B) Paraesophageal hernia. All or part of the stomach pushes through the diaphragm next to the gastroesophageal junction.

2 Type Paraoesophageal hernia (PEH)

•Also called giant hiatal hernia.

•While the cardia maintains its normal position, the gastric fundus herniates above the hiatus.

• It is 4 times more common in women. Median age is 6l.

• Dysphagia and postprandial fullness are observed at a higher rate in PEH.

•Approximately one-third of patients have anemia. Due to petechial bleeding from the ischemic mucosa or mechanical gastritis and due to bleeding from mucosal erosions (Cameron's ulcer).

•Respiratory complications (dyspnea and recurrent pneumonia) Previously developed due to aspiration. Recent studies have shown that cardiac output decreases due to left atrium compression.

• As the part of the herniated stomach increases over time, the stomach rotates and foregut obstruction develops.

• However, many patients may remain asymptomatic.

• Also, PEH may cause excessive bleeding, or gastric volvulus with acute gastric obstruction or infarction.

3 Type Mixt hernia (PEH)

It is an upward displacement of both the cardia and the fundus of stomach.

• It is very rare.

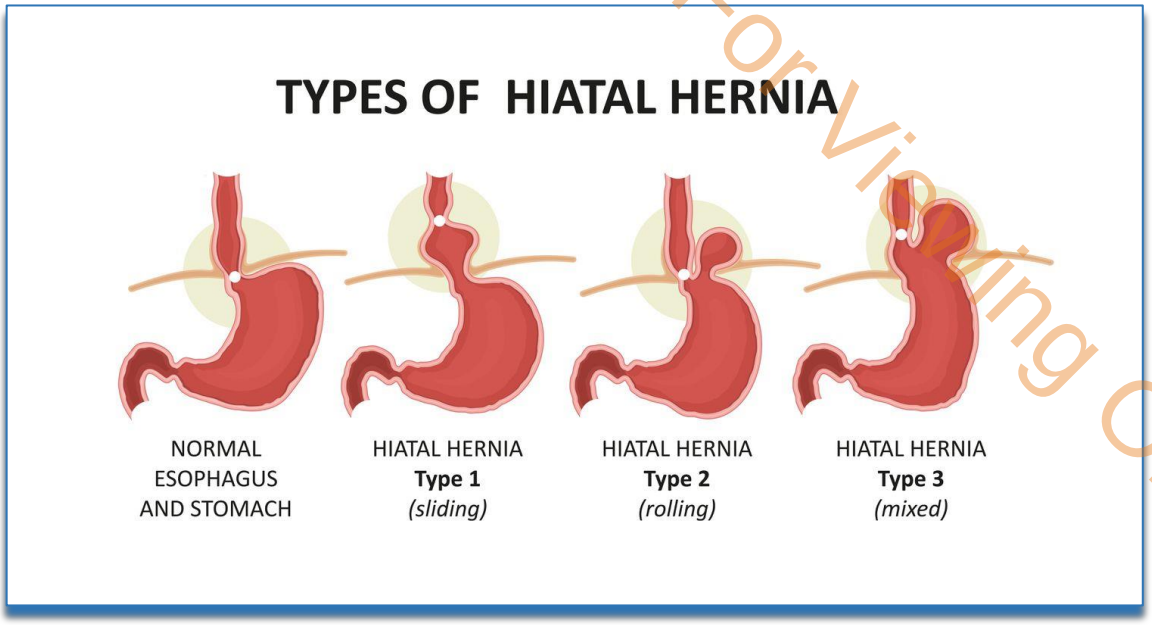
•Most cases occur when type I sliding hernia transforms into type III hernia.

4 Type Mixt hernia

The intra-abdominal organ herniate along with the stomach in these hiatal hernias.

• The most common herniation is the colon. The spleen, pancreas, and small intestines may also herniate.

• Type II and Type IV hiatus hernias are also called paraesophageal hernias.



Diagnosis:

• Observation of an air-fluid level behind the heart shadow on a chest X-ray taken while the patient is standing It is suggestive of paraesophageal hiatus hernia.

• Barium radiography for paraesophageal hernias;

• The most useful method in the diagnosis of sliding hernia is endoscopy.

Treatment

• The classical approach is always surgery, regardless of the clinical and size of paraesophageal hernia.

• Indications for surgery:

-Catastrophic complications such as severe bleeding, volvulus, strangulation and necrosis

- When these complications develop, emergency surgeries have a high mortality rate.

(mortality of elective surgery < 1%)

Achalasia

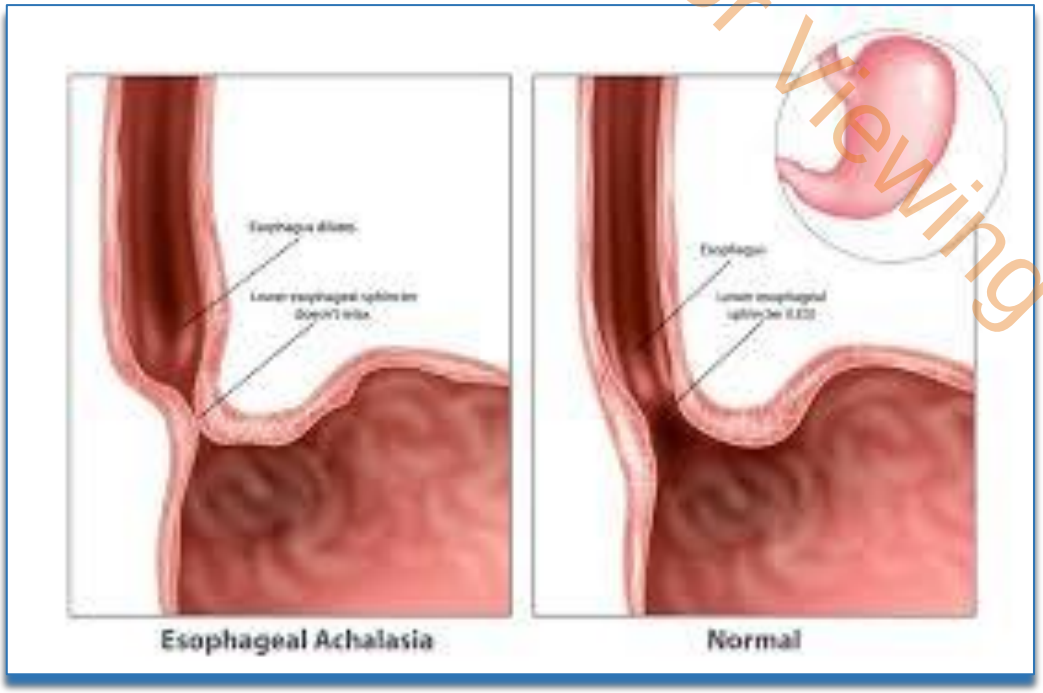
• It is the distal esophageal motility disorder, characterized by the loss of functional muscle ganglion cells in the distal esophagus and LES

• It is a primary LES disease.

• LES relaxation disorder + aperistalsis (often in the entire esophagus).

• Peristaltic contractions may occur in 5% of patients.

• It is seen equally in men and women. It can be seen at any age; It is most common between the ages of 30-50.



Pathogenesis

• Idiopathic or infection-related neurogenic degeneration.

• Degeneration of the vagus nerve or myenteric plexus ganglia of the esophagus.

• As a result of this degeneration, hypertension in the LES, lack of LES relaxation with swallowing,

increase in esophageal intraluminal pressure, dilatation in the esophagus and eventually in the esophageal body it will led to loss of peristalsis.

• Simultaneous contractions are observed in a group of achalasia patients. Complaint of severe chest pain

Clinic

Dysphagia

• May be intermittent initially; It becomes permanent over time.

•As a rule, dysphagia is more common in the beginning, especially with cold and watery foods. (paradoxical dysphagia)

• Chest pain is common.

• Pain is greater at the beginning of the disease and decreases as dilation increases. The patient may present with pneumonia.

• Classic achalasia triad > Dysphagia, regurgitation, weight loss

Diagnosis:

A “bird's beak” appearance or a columnar tortuous appearance is typical on barium radiograph.

• Although it is a chronic disease, achalasia is caused by a malignancy involving the gastroesophageal junction.It may also be the first symptom.

• Chest radiography (dilated esophagus and mediastinal dilation), esophageal radiography (aperistalsis, Lack of relaxation in LES), esophageal manometric studies (in the esophageal body and LES

increased pressure compared to normal) and endoscopy(especially normal mucosa in the narrow segment) are helpful methods in diagnosis.

• Endoscopy due to the possibility of confusion with cancer should be done. Endoscopic finding is the mucosa is normal, dilation of the proximal part of the stenosis.

•In **vigorous** achalasia, may be high-amplitude contractions. Clinically, it is accompanied by chest pain.

Definitive diagnosis is made by manometric studies.

Complications

- Aspiration pneumonia

- Carcinoma

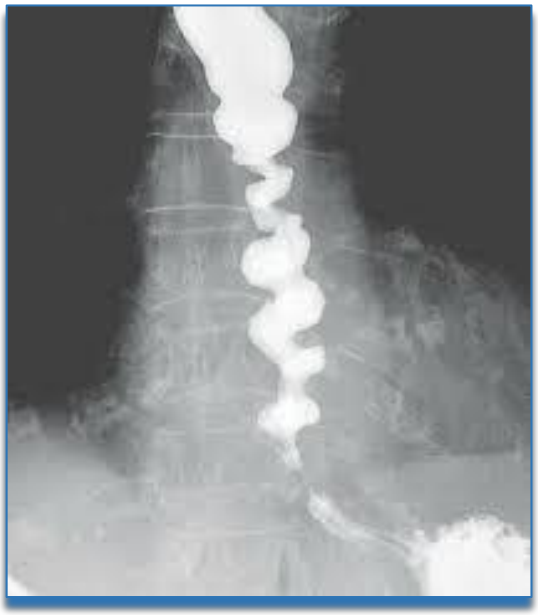
Treatment

All methods are palliative as they cannot restore esophageal motility and LES relaxation.

- There are both surgical and non-surgical treatment options.
- Non-surgical options are classified as Medical and endoscopic.
  - Sublingual nitroglycerin, nitrates or calcium channel blockers may be used symptomatically in the early stages provides relaxation.
  - Dilation with bougies is another treatment option.
  - Endoscopic botulinum toxin injections can be performed.
  - As surgical treatment; Esophagomyotomy is performed using the method described by **Heller.**

    A single 7-8 cm myotomy is made from the anterior aspect of the narrow distal segment of the esophagus.
  - Laparoscopic Heller esophagomyotomy + partial fundoplication is the most effective surgery.

DIFFUSE OR SEGMENTARY ESOPHAGUS SPASM (DES)



- Pain behind the sternum and dysphagia are the most common symptoms in DES.
- Pain is more important. Dysphagia is intermittent or absent.
- It is basically a disease of the esophageal body.
- Radiologically simple narrowing in 50% of patients, segmental spasm.
- In some cases, due to increase in pressure within the lumen of esophagus **epiphrenic** or midesophageal diverticula may develop.
- Esophageal motility studies are important in diagnosis.
- Pressure in the lumen of esophagus is high. Relaxation in LES is usually is normal; LES pressure may be high.

Treatment

• Today, primarily pharmacological or endoscopic interventions are preferred.

• Nitrate and Ca channel blockers, sedatives and anticholinergics are used.

• Bougie dilatation and botulinum toxin injection may be tried.

• If pharmacological and endoscopic interventions fail, surgery is preferred.

• Extended esophagomyotomy is performed in surgical treatment:

• Esophagomyotomy should include the LES. The upper limit of myotomy is also determined by motility studies. If sliding hernia accompanies it, antireflux surgery can also be performed in the same session.

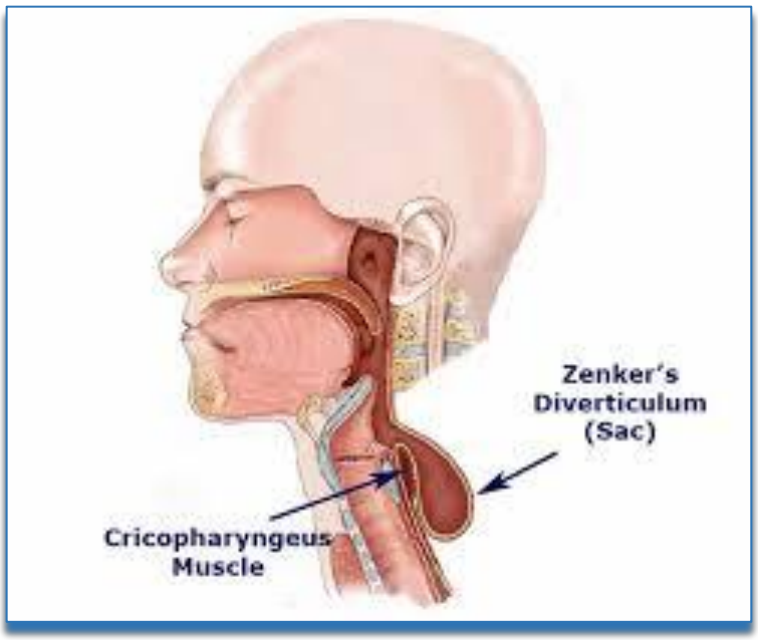
• Today, the gold standard is laparoscopic surgery.

ESOPHAGEAL DIVERTICULA

Esophageal diverticulum can be “pulsion” and “traction”. **Pulsion** (false)diverticula are those where intraluminal pressure of esophagus remains high for prolonged period due to distal obstruction. This lead to out pouch of mucosa through a weak point on its wall. **Traction**(real) diverticula are those in which there paraesophageal inflammation leading to fibrosis which exerts a pull on the wall and contains all layer of esophagus.

Pharyngeal (Zenker)diverticula

• Between the transverse fibers just above the cricopharyngeus muscle and the oblic fibers of the inferior constrictor muscles (the area called Killian's triangle) appears.



• It extends behind, usually to the left.

• It is more common in men and older people.

• Pulsion(false) diverticulum; It contains only the mucosal layer.

• Studies have shown that there is motor incoordination in the cricopharyngeus muscle and that this muscle is weak during swallowing and It is stated that it undergoes premature contraction.

• During the process, the mucosa extends outwards; It becomes a pouch.

• The most common symptom is food getting stuck in the upper esophagus (high cervical obstruction).

• As the diverticulum grows, undigested food accumulates in the pouch and causes aspiration, lung complications, halitosis, there is a sloshing sound when drinking water, swelling in the neck, and regurgitation of food into the mouth.

DIAGNOSIS

- Radiology -Barium lateral radiographs are the best diagnostic method.

TREATMENT

- Diverticulectomia and cricopharyngeal myotomia
- Cricopharyngotomy with endoscopic stapler

MIDESOPHAGEAL DIVERTICULES

• Tuberculosis etc. It is associated with mediastinal inflammatory diseases.

• A LAP attaches and pulls on the esophageal wall from the outside.

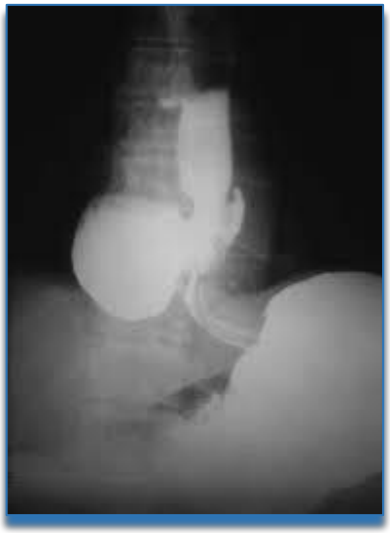
• It is a traction diverticulum. It is a true diverticulum.

• It is detected incidentally.

• It is asymptomatic.

• Treatment is directed towards the cause. Diverticula smaller than 2 cm are observed. If large or symptomatic indicated surgical treatment

EPYPHRENIC DIVERTICULES



• They are located in the last 10 cm of the esophagus, just above the lower esophageal sphincter.

• It is of the type of pulsion diverticula and causes incardination and intraluminal problems due to relaxation of the esophageal sphincter, which lead to increase in intraluminal pressure.

• The mucosa forms a pouch outside the lumen. It may be associated with hiatal hernia, diffuse spasm, and achalasia.

The most common symptoms are dysphagia and regurgitation.

Treatment is diverticulectomia and esophagomyotomia .

Esophageal perforation

40-60% occur in the cervical, 40-50% in the thoracic and 10% in the abdominal esophagus.

Etiology

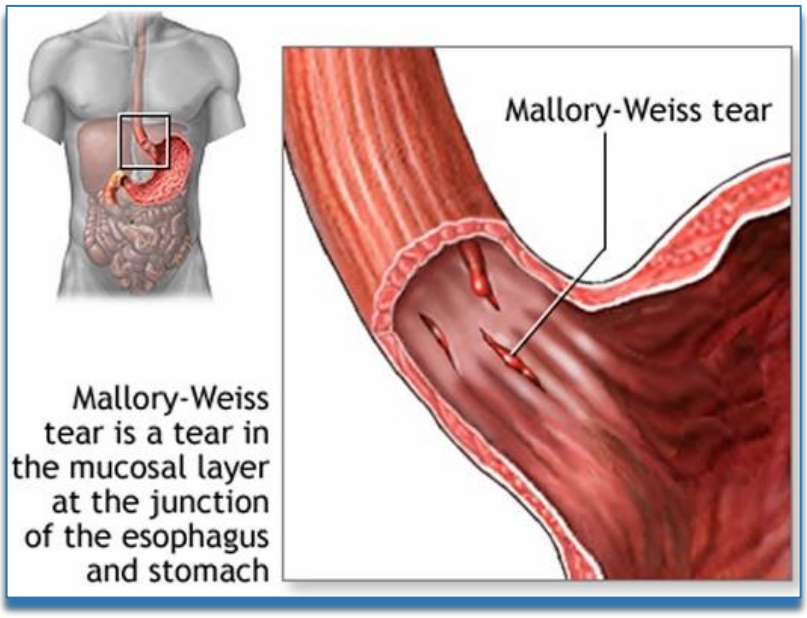
- Iatrogenic
- Spontaneous perforation
- Foreign bodies
- Trauma

Iatrogenic Perforation

- It constitutes 60-75% of esophageal perforations.
- Most of them occur during instrumental procedures such as endoscopy or dilatation.
- The cricopharyngeal muscle area is the narrowest part of the esophagus; Most of these perforations are in this area.

Spontaneous Perforation

- It corresponds to 15% of esophageal perforations.
- Most are postemetic. (Barogenic perforation) It was first described by Boerhaave.
- Spontaneous esophageal perforation is known as Boerhaave syndrome.
- In **Boerhaave** syndrome, perforation usually occurs just higher the gastroesophageal junction.It occurs proximally in the thoracic esophagus and in the left posterolateral wall of the distal esophagus and is longitudinal.



- If the tear is only in the mucosa and submucosa, it is called **Mallory-Weiss syndrome**. Mallory Weiss tears are longitudinal and occur on the lesser curvature side of the gastroesophageal junction at the level of mucosa and

submucosa. It is the cause of upper GI bleeding. Bleeding in most patients It stops automatically.

- Some of the spontaneous perforations are caused by malignant diseases or benign distal ulcers.It may occur as a result of spontaneous perforation (Non-barogenic).

Perforation Due to Foreign Objects

- is rare.
- Corresponds to 14% of esophageal perforations.
- Perforation is most common in places where foreign objects are frequently lodged, in the cervical region.

Penetrating or Blunt Trauma

- It constitutes 10% of esophageal perforations.
- Most are in the cervical esophagus.
- In blunt traumatic perforations, the mechanism is a rapid increase in esophageal intraluminal pressure.

CLINIC OF ESOPHAGEAL PERFORATIONS

• It varies depending on the level of perforation and the degree of inflammation in the surrounding tissue.

• Pain, fever, dyspnea and dysphagia are general symptoms.

• In upper esophageal perforations, pain in upper chest and neck, and lower perforations pain in the precordial and epigastric regions. It is the most important symptoms

• Dyspnea usually occurs due to pleural effusion.

• Pneumothorax may also occur with pleural effusion.

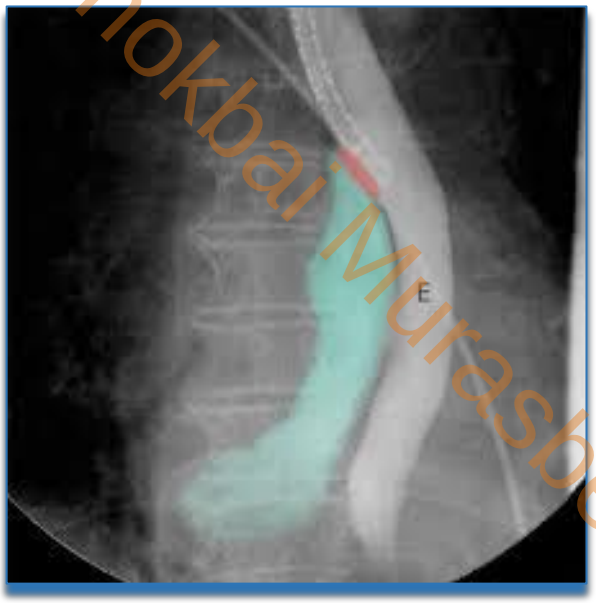
• The first sign of cervical esophageal perforation is tenderness in the cervical region.

• Crepitation due to subcutaneous emphysema in the neck is diagnostic.

• In thoracic esophageal perforations, crepitus may be seen in the neck, but there is usually no sensitivity.

• A crackling sound is heard when the heart is rested sign of Mediastinal emphysema (Hamman sign)

• In subphrenic perforations, acute abdomen with abdominal tenderness.



DIAGNOSIS

• For early diagnosis, radiological methods are used.

• Direct thorax radiography can be performed first. Expansion and air are seen in the mediastinum:

• Esophagography with a water-soluble opaque substance (gastrografin) is the standard diagnostic method (In right lateral decubitus position should be done)

TREATMENT

• The treatment of choice is early surgical intervention.

• Broad-spectrum antibiotics and fluid therapy, electrolyte therapy are administered to all cases, whether or not they undergo surgery, balance should be regulated and cardiorespiratory support should be provided.

• The most important factor determining treatment success is early repair.

• It is very important that surgery be performed within the first 24 hours.

• Basic principles of surgical treatment(Closing the perforation +Drainage of the area contaminated as a result of perforation)

• Nutrition is provided parenterally or enterally.

• Non-operative approach can be applied in selected cases.

Cameron suggested three criteria for nonoperative treatment of esophageal perforations:

1- It should be shown by drinking barium that the perforation is in the mediastinum and flows back into the esophagus.

2- Symptoms should be mild.

3- Clinical evidence of sepsis should be minimal.

ESOPHAGEAL CORROSIVE CAUSTIC INJURY

• It usually occurs in children.

• Accidental ingestion of alkaline substances is a more common problem than acidic substances because the acidic substances immediately cause severe burning in the mouth and are usually not swallowed.

• Acute and chronic damages occur in corrosive traumas.

• Acute period -Tissue injury and perforation

• Chronic period -Strictures and swallowing disorders

• Alkaline substances penetrate deeper because they cause tissues to dissolve. In acidic substances injures, they have less tissue penetration, because they cause coagulation necrosis.

• Chemical burns damage mostly the middle esophagus.

• In the early period, pain in the mouth and substernal region, hyper salivation, and dysphagia occur.

• To determine the extent of the lesion in the early period, early endoscopy (in the first 12- 24 hours) is performed as soon as the patient is stable. Radiological methods are not used because they are unreliable.

• Surgery decision is made according to the endoscopy results. While first degree burns are observed for 24-48 hours, second degree burns are observed for 24-48 hours and third degree burns are indication for surgery.

• In patients who do not require surgery, agents that will neutralize corrosive substances within the first few hours available.

• Procedures that may cause vomiting, such as the use of emetics and nasogastric is contraindicated.

• Steroid use is controversial (beneficial in experimental studies; benefit not shown in humans).

• Hypovolemia is corrected.

• Broad spectrum antibiotics are started.

• Oral feeding is started after early dysphagia subsides.

• The dilatation is controversial. There are also those who perform dilatation immediately after the injury. There are also approaches that do not apply this treatment before 6 weeks due to the risk of perforation.

ESOPHAGEAL MALIGN TUMORS

Squamous cell carcinoma and Adenocarcinoma are most common histological subtypes of esophageal carcinoma

1) SQUAMOUS CELL CARCINOMA

Squamous cell carcinoma constitutes 60-80% of esophageal tumors. It is the most common type.

It is most commonly located in the middle thoracic esophagus (32%). Other locations, in order of frequency: lower thoracic esophagus (25%), cervical (8%) and upper thoracic esophagus (3%).

ETIOLOGY

- Alcohol
- Cigarette
- Smoked or pickled foods
- Foods and drinks taken at high temperatures
- Benziprene and various nitrosamines
- Riboflavin and zinc deficiencies
- Deficiency of vitamins A, C, E
- Fanconi anemia
- Tylosis(Autosomal dominant transmission occurs. It is characterized by thickening of the skin on the soles of the hands and feet. The risk of esophageal cancer is very high (100%) in these patients)
- Corrosive strictures
- Achalasia
- Human papilloma virus
- Plummer-Vinson syndrome (sideropenic dysphagia)

2) ADENOCARCINOMA is second most common esophageal cancers and mostly developed countries in North America and Europe have higher incidence.

ETIOLOGY

- Barrett esophagus
- GERD
- High alcohol intake
- Smocking

CLINIC OF ESOPHAGEAL MALIGN TUMORS

Clinical Symptoms

• The most common symptom is dysphagia.

• Difficulty in swallowing solid foods at first, but over time, difficulty in swallowing soft and small bites, even watery ones.

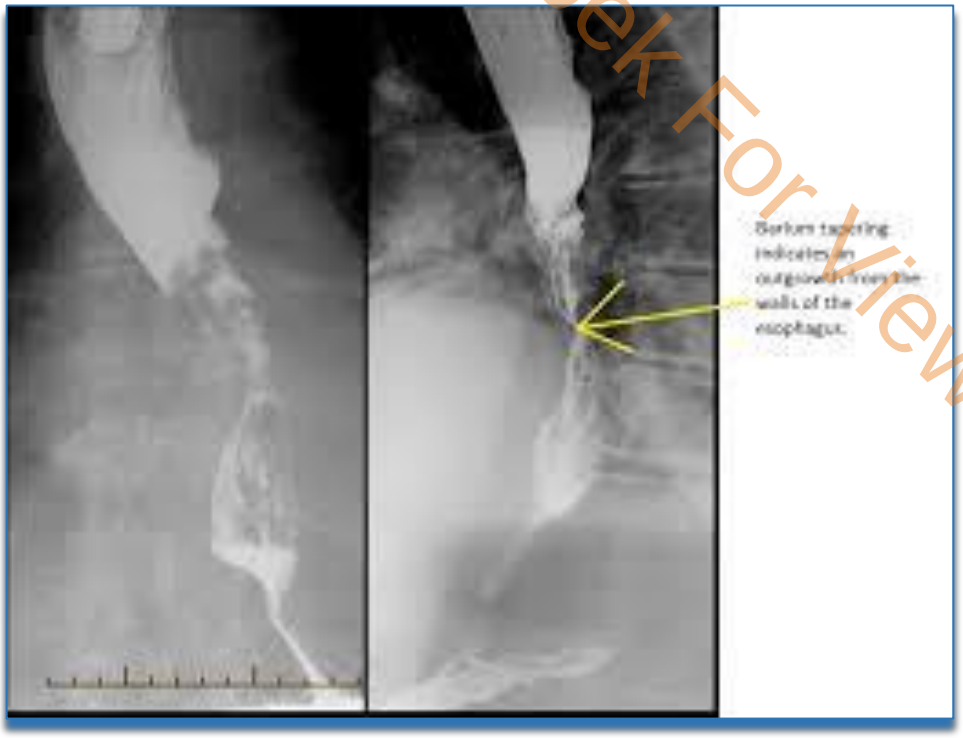
• Fistulation into the tracheobronchial structures (especially seen in the squamous type) causes cough and symptoms or pneumonia.

• Loss of appetite and weight loss occur before dysphagia.

• Other symptoms: Weight loss, fatigue, anemia. ..

• In esophageal adenocarcinoma, long-standing retrosternal pain and regurgitation may occur.

Diagnosis



- Radiological Examination

-Irregular narrowing of the esophageal lumen

- Apple-shaped appearance (if there is symmetrical narrowing all around)

- An asymmetric mass with an infiltrative appearance is mostly detected.

- Esophagoscopy

- It is a definitive diagnosis method.

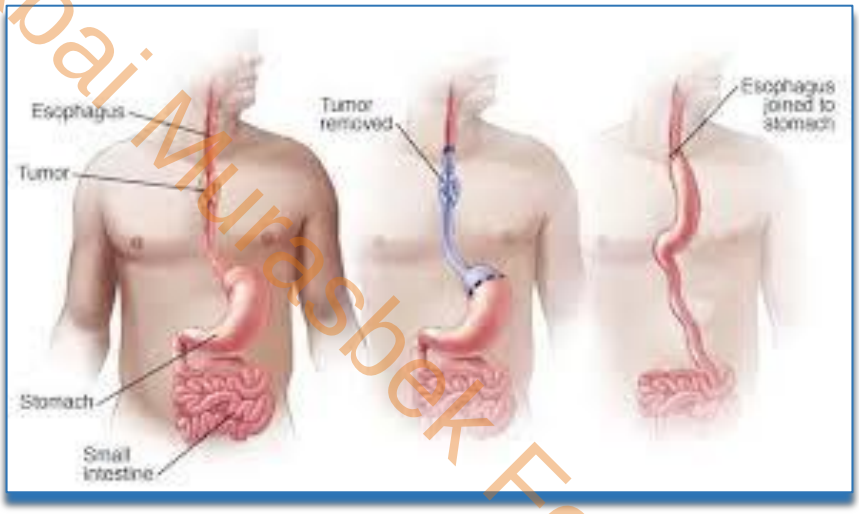
-It is used in all cases where cancer is suspected.

- Endoscopic ultrasonography (EUS)
- Positron Emission Tomography (PET)

TREATMENT

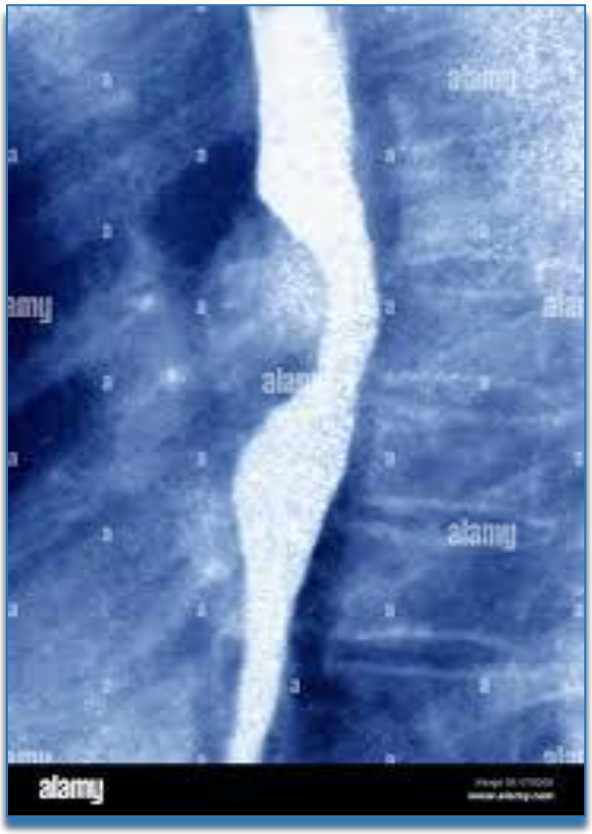
- Esophagectomy (Minimal invasive esophagectomy, Transhiatal esophagectomy,Ivor Lewis two phase procedure, Mc Keown three phase operation.
- Radiotherapy

- Chemotherapy
- Palliative treatment. The main aim of palliative treatment is to relieve the dysphagia caused by advanced esophageal tumor.



BENIGN ESOPHAGEAL TUMOR (LEIYOMYOMA)

- It corresponds to 10% of all GI leiyomyomas.
- The average age of diagnosis is 38 years.
- It is most common in men.
- Because of smooth muscle origin, 90% of it is located in lower part(2/3) of esophagus.



- Dysphagia and pain are the most common symptoms.
- Rarely, ulceration and bleeding occur.
- Barium X-ray - is the best diagnostic method. Smooth limited, half-moon shaped mass that is mobile with swallowing.
- In endoscopy, submucosal tissue growing towards the lumen
- Endoscopic biopsy is not recommended because of high risk of perforation and bleeding
- Surgical method of treatment - is enucleation.

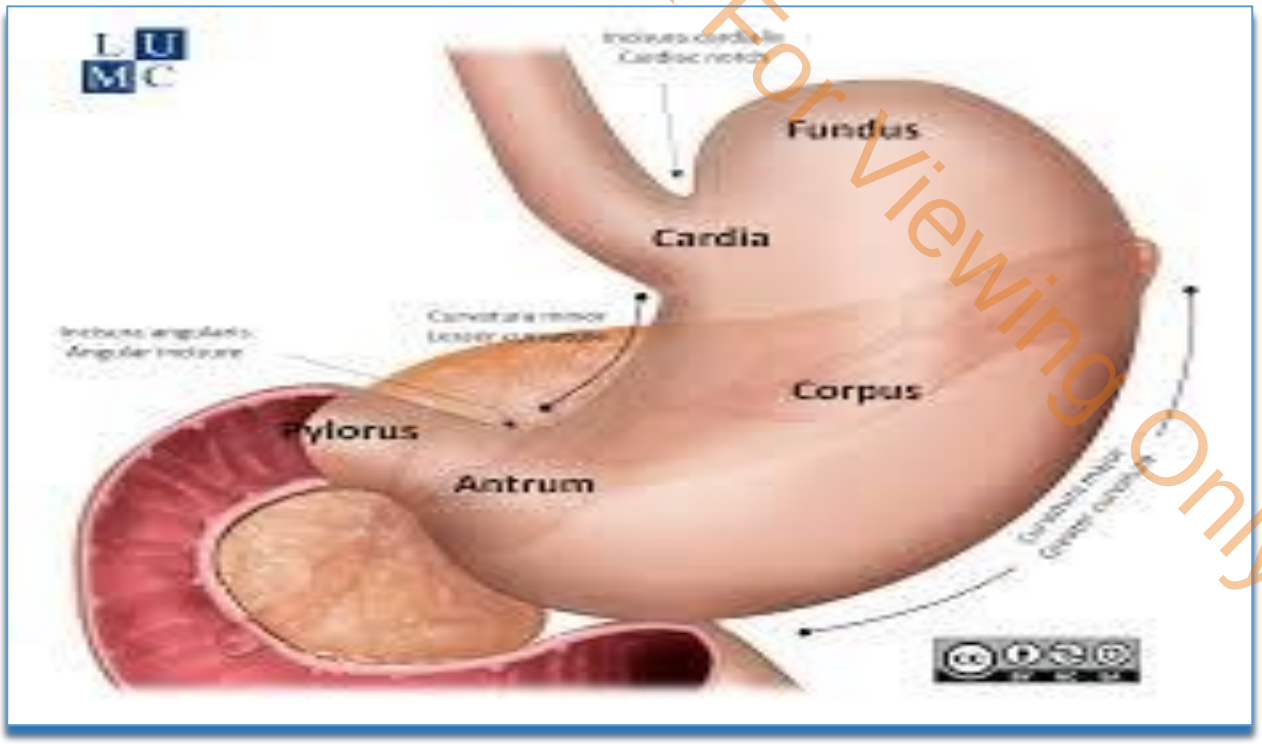
Chapter

2.

Stomach and Small Intestine Diseases and Surgery

• The stomach is a dilatation of the proximal GI tract and is used for the initial digestion and storage of food. Its capacity is approximately 1000 ml. Stomach consist of:

- Cardia - At T12 level.
- Body
- Fundus



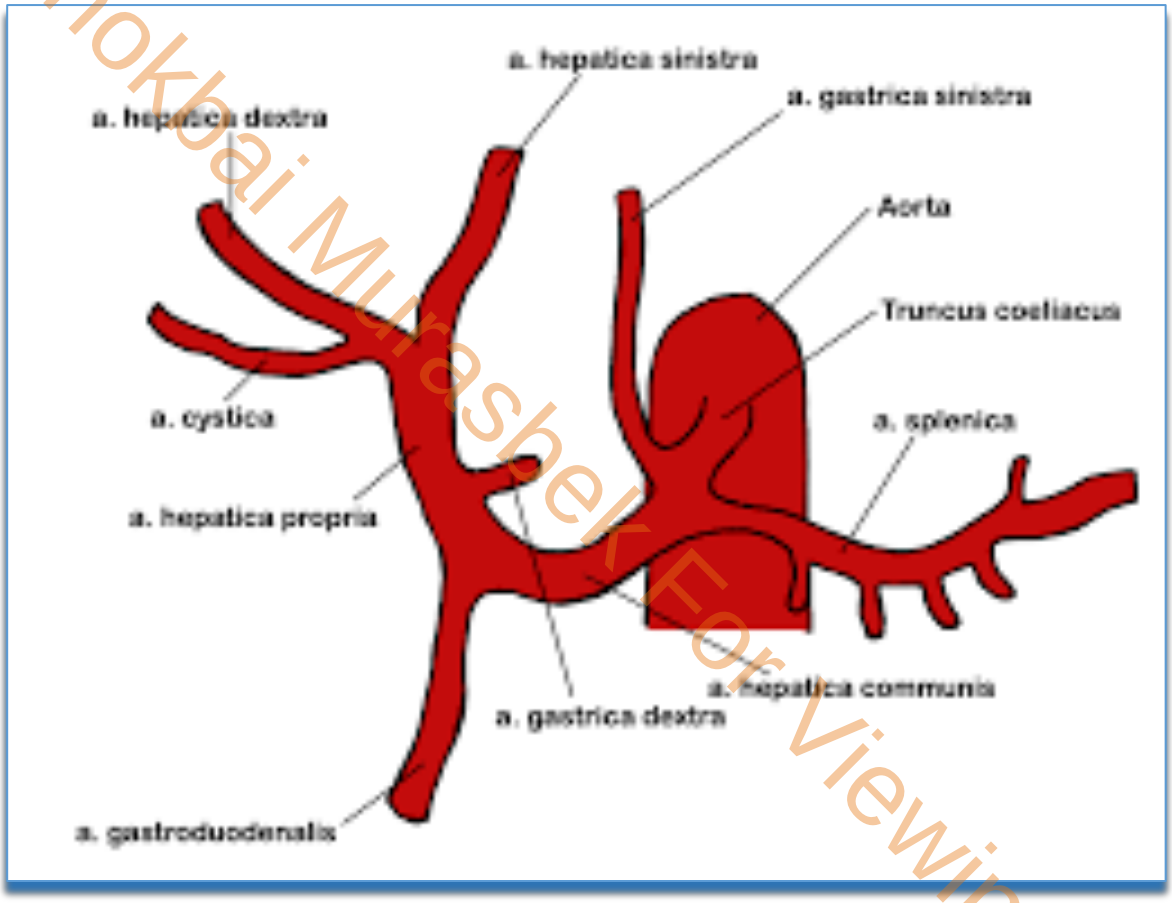
- Pylorus - It is at the level of the lower edge of L1, 1-2 cm to the right of the midline.

There is a complete sphincter in the pylorus. Circular muscle layer of the stomach. It becomes very thick in this region and forms the pyloric sphincter muscle.

- Angle of His - It is the angle between the left side of the gastroesophageal junction and fundus.

ANATOMY

Vascular supply: The arterial supply to the stomach is extremely rich:



-Left gastric artery from the truncus coeliacus

-Right gastric artery from the hepatic artery

-Right gastro-epiploic artery from a.gastroduodenale(branch of a.hepatica)

-Left gastro-epiploic artery from a.splenica

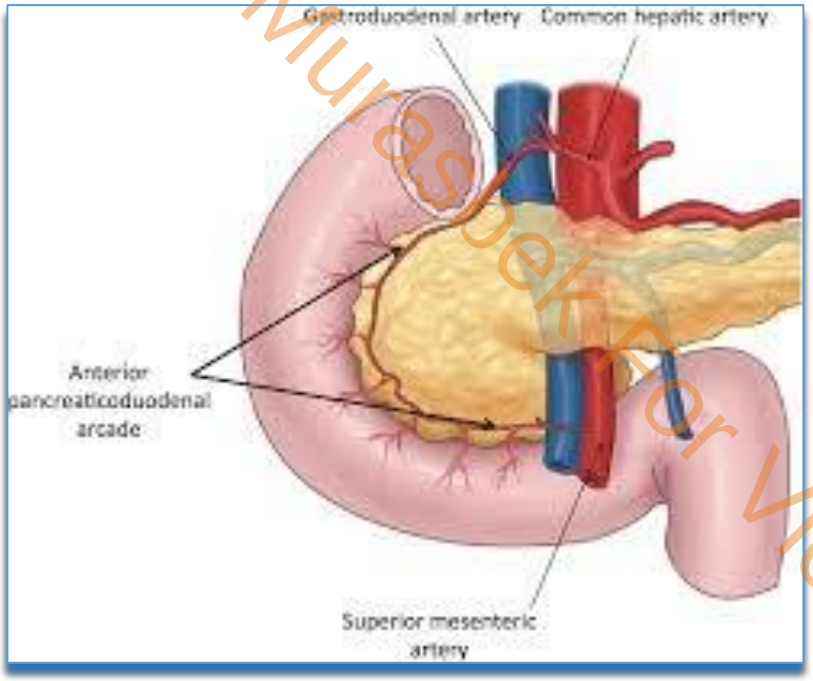
-Short gastric arteries from a.splenica

- All corresponding veins drein to v.portae

Nerves

• Stomach has parasympathetic and sympathetic nerves.
• Sympathetic nerves come from the celiac plexus around the gastric and epiploic arteries.
• Parasympathetic nerves come from the vagus.
• The left vagus is located anteriorly, the right is located posteriorly.

• The anterior vagus gives off hepatic branches under the diaphragm, and the posterior vagus gives off celiac branches.
• The anterior vagus (left) innervates the liver and gallbladder. Also the stomach (fundus and corpus) and gives branches that innervate the antrum.
• The posterior vagus (right) gives branches to the celiac plexus. It also innervates the fundus, corpus and other parts of the stomach then innervates the distal GI tract.



DUODENUM;

The duodenum, which develops from the foregut together with the stomach, resembles the small intestine in structure. Since ulcers is most commonly seen in the first part of the duodenum, it is considering among stomach diseases.
• Average length is 25 cm.
• It has no mesentery and is retroperitoneal organ.
• Duodenum consists of four parts. The first (superior) part of the duodenum is approximately 5 cm . It starts from the pylorus and ends at the neck of the gallbladder. Its anterior surface is covered with peritoneum. Other parts descending, inferior and ascending form a “C” shape which wraps around the head of the pancreas
HYSTHOLOGY
• Gastric mucosa is lined with columnar cells.
• Superficial glands in the cardia secrete mucus.
• They provide lubrication during food passage.
• In the glands called “oxyntic glands” in the fundus and corpus located mucus-secreting cells, chief (zymogenic) cells that secrete pepsinogen, **parietal (oxyntic) cells** that secrete HCl and intrinsic factor also argentaffin endocrine cells.
• There are mucus cells and **G cells** that secrete gastrin in the antral mucosa of stomach. Parietal cells no take place in this part of the stomach.
• **D cells** are somatostatin producing cells, which can find in stomach, intestine and pancreatic islets. Somatostatin acts directly on the parietal cells to reduce acid secretion. **Helicobacter pylori Increases acid secretion by disrupting the function of D cells.**
• 13% of normal gastric epithelial cells are oxyntic (parietal) cells, 44% are chief (zymogenic) cells, 40% mucus cells and 3% endocrine cells...

• The submucosa is the layer richest in blood vessels and collagen. In anastomoses in the GIS this layer determines the strength. Additionally tunica serosa (visceral peritoneum) has a role in tensile strength in GIS anastomoses.
• Mucus cells act as stem cells for other cells.

GASTRIC SECRETATION PHYSIOLOGY.

• The most important secretion of the stomach is hydrochloric acid (HCL).
• Three main stimulation for acid secretion; gastrin, acetylcholine and histamine.
• After gastrin is secreted from the antrum and proximal duodenum, it is released into the circulation system.
• Acetylcholine is secreted from cholinergic nerve endings (n.vagus).
• Histamin is secreted from enterochromaffin like cells in the lamina propria of the fundus and corpus. After as a paracrine they are effects parietal cells.
•Parietal cells have all types of receptors for these stimulators.
Acid secretion is controlled by 3 different systems:

a)cephalic phase

- It starts with the view, smell and thought of food and is basically created by the n.vagus .
- Vagal stimulation directly stimulates parietal cells and increases gastrin secretion.

b)gastric phase

- Antral distension, increasing in luminal pH value with meals containing amino acids, short peptides will lead to gastrin secretion. Gastrin secretion absolutely stops when pH <2.0

c) intestinal phase

-It begins with the entry of food into the duodenum.
Absorption of amino acid and gastrin secretion from the proximal duodenum have main role in this phase. The decrease in duodenal pH ,also affects other intestinal hormones such as secretin and neuropeptide Y.Other secretory functions include mucus and bicarbonate, intrinsic factor and pepsinogen.

STOMACH HORMONES

Gastrin

• It is released from G cells in the antrum that increases acid release from parietal cells.
• Luminal peptides and amino acids stimulate gastrin secretion.
• Luminal acid is the major factor that inhibits gastrin secretion as a “negative feedback”
Somatostatin
• It is released from D cells found in the entire gastric mucosa.

Ghrelin

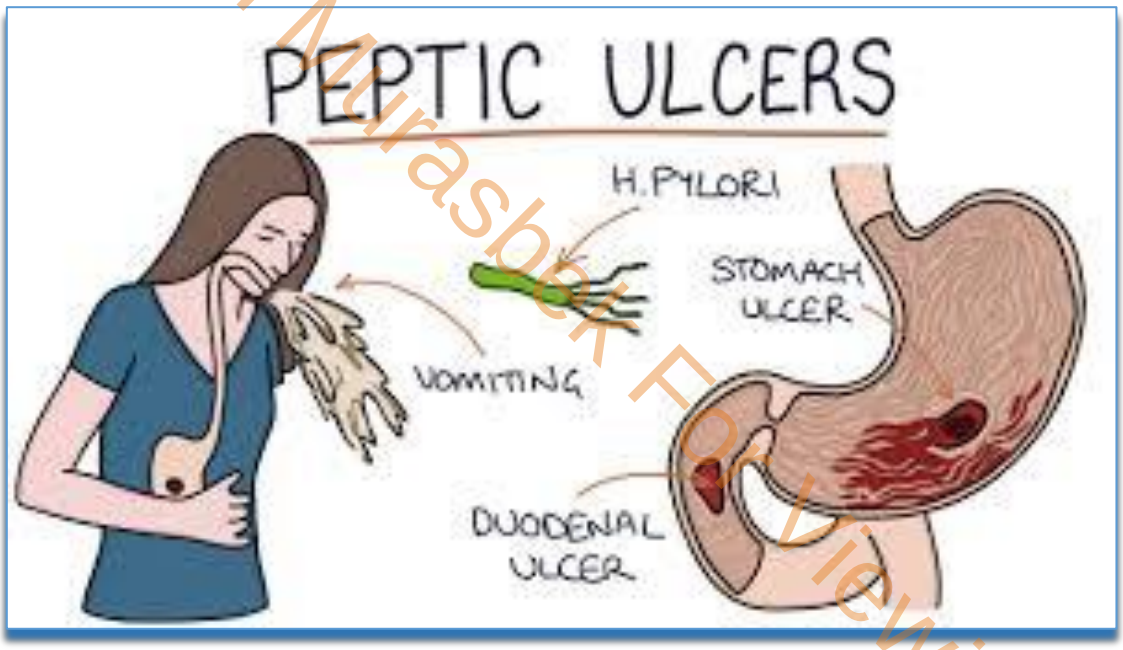
• It is an important hormone for appetite.
• When its level increases, appetite increases; when it decreases, appetite decreases.
• Stimulates growth hormone secretion from the anterior pituitary.

Leptin

• Stimulates the satiety center

PEPTIC ULCER

• Peptic ulcer is the name given to ulcers caused by stomach acid (HCL).
• Ulcer can located in esophagus, stomach, duodenum, jejunum (after gastrojejunostomy) and ileum (close to Meckel diverticulum mucosa).



• Peptic ulcer is the most common GI disorder with a lifetime risk of 10%.
• It reaches its peak around the age of 70.
• It is most commonly located in the duodenum.
• It is 2-3 times more common in men.
• Reducing gastric acid secretion leads to healing of all ulcers.
• In most patients with duodenal ulcer (DU), acid secretion is higher than normal; in patients with gastric ulcer (GU) acid secretion normal or less than normal.
• Hyperacidity-main factor in the pathophysiology of GU.
• Disruption of defense factors of mucosa- the main factors of DU.
• GU is generally more common at older ages and in women.
• The majority of peptic ulcers are caused by Helicobacter pylori or NSAID use.
• Duodenal ulcer usually occurs within the first 1-2 cm after the pylorus; It is located in the proximal duodenum.
• There is no single etiological factor. There is an interaction of many factors.
• Strengthening of aggressive factors for the mucosa and weakening of defensive factors will lead to ulcer formation.
• Important protective agents; prostaglandins, nitric oxide, intrinsic nerves and peptides (calcitonin associated peptide, gastrin-releasing peptide, gastrin and heat shock proteins

ETIOLOGY

Helicobacter pylori(disrupting of D-cells) and NSAID (reduce prostaglandin production) most common etiologic factors in Peptic Ulcer.

Helicobacter pylori (HP)

• It is a Gram (-), microaerophilic, motile and curved bacillus.
• HP Increases acid secretion; decreases the acid protection mechanisms of mucosal cells;
• Found in most patients with peptic ulcers.
• H. pylori produces urease enzyme. The enzyme urease converts urea into ammonia and bicarbonate. Bicarbonate buffers the micro acid environment in the stomach. It makes it suitable for H. pylori to live in this acidic facility. Also ammonia damages the gastric epithelium.
• HP increases acid secretion by disrupting the function of D cells.
• Helicobacter pylori is also associated with gastric lymphoma and adenocarcinoma.
• The most appropriate method for the diagnosis of H. pylori is rapid urease test.
• Microbiological culture may be preferred, when eradication therapy has failed several times (antibiotic resistance cases).
• A positive serological test in a patient who has never received treatment for H.pylori is evidence of active infection.

•Urea breath test, and stool antigen test to confirm HP eradication following appropriate

treatment is the standard method.

•HP also can be cause of carcinoma and lymphoma because of formation pangastritis with low

acidity and function disorders in stomach corpus.

Diagnostic tests for Helicobacter Pylory		
Test	Advantages	Disadvantages
Serologic	Non-invasive Specificity: 90% Sensitivity: > 80%	Active and past infection cannot distinguished
urea breath test	Simple specificity: 99% Sensitivity: 90%	If done early, may be a negative result
Histology	Specificity: > 95% Sensitivity: 80% gold standard	Invasive, requires experience and Laboratory facilities.
rapid urease test	Specificity: 95-100% Sensitivity: 80-95%	Invasive, If done early, possible negative result
Microbiologic culture	Specificity: 100% Sensitivity: 80% Antibiotic sensitivity can checked	Time consuming and Expensive

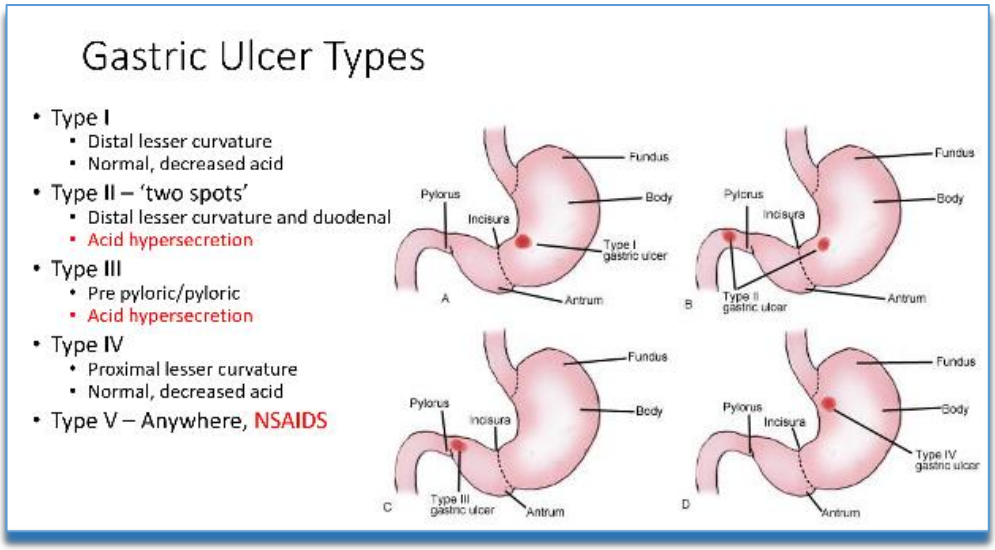
Duodenal ulcers occurs in the first part of duodenum. Gastric ulcers usually large and involve lesser curvature of the stomach.

DUODENAL ULCER (CLINIC)

- In most patients with duodenal ulcers, acid secretion is higher than normal.
- The most important symptom of duodenal ulcer is epigastric pain.
- Pain usually occurs when fasting, it is relieved with food and antacids. A few hours after eating pain develops again when food and acidic stomach contents pass into the duodenum.
- The pain does not typically radiate, but radiating toward the back may indicate that the ulcer has spread(penetrated) to the pancreas.
- Other symptoms and signs as nausea, bloating, weight loss, positive stool occult blood and anemia may be present.
- The most common complications are bleeding, perforation, obstruction and penetration.

GASTRIC ULCER (CLINIC)

- In the pathogenesis of gastric ulcer, disorders of mucosal defense is at the forefront rather than hyperacidity.
- The most common symptom is epigastric pain.
- Pain usually occurs after eating.
- The patient sometimes claim that his pain decrease by eating.
- Other symptoms :Dyspepsia, nausea, vomiting, weight loss ...
- Perforation and bleeding are less common than duodenal ulcer.
- The most common complication of stomach ulcer is perforation.



TYPES OF GASTRIC ULCERS

Type 1- Corpus and less curvature of stomach (Low/N acidity+ most common type)
Type 2 – Corpus located + Duodenal ulcer (hyperacidity)
Type 3-Prepyloric or Pyloric ulcer (hyperacidity)
Type 4- Less curvature or cardia located ulcer(Low/N acidity)
Type 5 -Ulcer caused by medications (NSAID)

COMPLICATIONS OF PEPTIC ULCER(Gastric and Duodenal ulcer)

- Bleeding
  -The most common complication of duodenal ulcer is bleeding.
  - Massive bleeding develops in 15-20% of patients with peptic ulcers.
  - Microscopic bleeding is more common.
  - Bleeding is usually caused by ulcers located on the posterior wall of the duodenum, from gastroduodenal artery.
  - It develops due to erosion of the wall.
  - 75% of patients presenting with bleeding can be treated with simple medical treatment and acid suppression.25% will continue to bleed or bleed again in the hospital.
  - The most common cause of peptic ulcer-related death is bleeding.
- Perforation and penetration
  -Perforation is the second common complication of peptic ulcer .
  -If the ulcer destroys all layers of the stomach or duodenum wall, perforation or penetration can occur.
  - The most commonly penetrated organ is the pancreas.
  - Perforation develops in 5-10% of peptic ulcer patients.
  - due to irritating the peritoneum with acidic content in perforation-sudden onset pain described as “wooden abdomen”, and paralytic ileus can develop.
  -Pneumoperitoneum due to perforation is found in 80% of patients. Free air is seen under the right diaphragm in chest radiographs (X-Ray).
  - Surgical treatment is required for perforation. However, rarely in stable patients without signs of peritonitis (spontaneously closed), non-surgical treatments can be used.
  - The prognosis of stomach perforations is worse than duodenal ulcer perforations. The reason is these patients are older and have more comorbidities.
  - Other factors affecting the prognosis in peptic ulcer perforations :
    a)24 hours between the onset of symptoms and clinical presentations.
    b) The patient is hemodynamically unstable
    c) Body mass index < 21
    d) advanced age
  - **"kissing ulcer"** – ulcer on both (anterior and posterior) walls, should be suspected in patients with acute perforation and bleeding.
- obstruction

- Lumen narrowing and obstruction because of edema (acute ulcer) and scar tissue (chronic ulcer) formation observed in 5% of cases.
- Obstruction present with nausea, vomiting and abdominal distension.
- Dehydration, hypokalemia and hypochloremic metabolic alkalosis can develop due to vomiting.
- Less indication for surgery.

INVESTIGATIONS

- Esophagogastroduodenoscopy
- Detection of HP(quick urease test, urea breath test, stool test, serologic test, histology and culture)
- Serum Gastrin level (a base line serum gastrin level is appropriate to rule out gastrinoma)
- X-Ray (only for determination free air in perforation)

Suspicious symptoms requiring esophagogastroduodenoscopy
-Weight loss
-Recurrent vomiting
-Dysphagia
-Bleeding
-Anemia
-palpable mass
-epigastric pain in patients elder 45 years (even for the first time)

TREATMENT OF PEPTIC ULCER

- Medical treatment (Gastric and Duodenal ulcer)
  - Antacids neutralize acid.
  - H2 receptor antagonists inhibit acid secretion.
  - Anticholinergics inhibit acid secretion.
  - Proton pump inhibitors inhibit acid secretion.
  - Prostaglandins increase mucosal protection.
  - Sucralfate forms a protective cover on the mucosa.
  - Colloid bismuth creates a protective cover and eradicates H. pylori.
  - Antibiotics eradicate H. pylori (various drugs containing metronidazole, amoxicillin, tetracycline).
- Surgical treatment indicated for all patients with complications as a bleeding, perforations, obstruction and that cases where 8-12 week unresponsive medical treatment exist. Also if the ulcer size more than 2 sm indication to surgery.

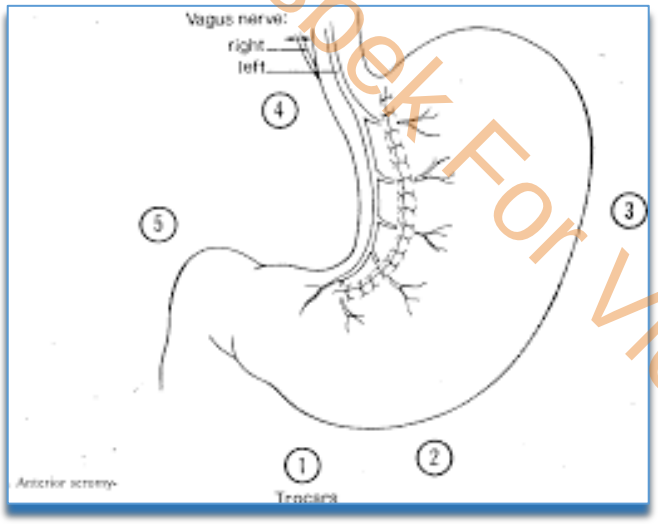
SURGICAL METHODS:

- a simple stitch for a bleeding ulcer
- For perforated ulcers (if < 1 cm) preferred simple closure. (primary repair Graham patch)
- Surgical treatment of duodenal ulcer is usually vagotomy. Because the main purpose of DU treatment is reduce acid secretion. There are two types of vagotomy: truncal and high selective vagotomy.

TRUNCAL VAGOTOMY (bilateral)

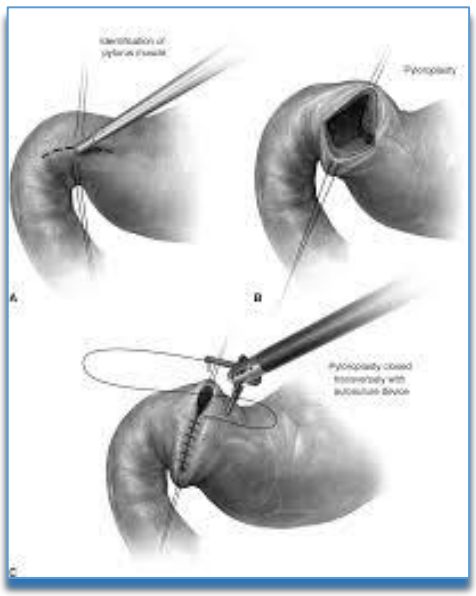
-it is the simple and most common use methods

-in the distal part of esophagus anterior and posterior n.vagus are cut.



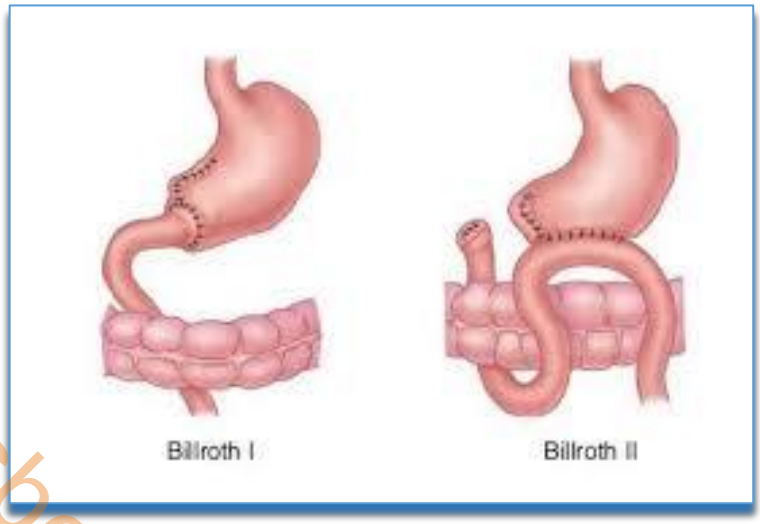
-because of this cutting, innervation of pylorus disappear and as a result gastric emptying will delayed (especially solid foods) that's why a drainage surgery definitely added (pyloroplasty or gastrojejunostomy)

HIGH SELECTIVE VAGOTOMY



-all branches of latarjet nerve which leading to corpus and fundus are cut except of "crow foot" branches that goes to antrum.

-transit time of solid food through the stomach does not delayed.



- BILROTH 1 GASTRECTOMY: Distal stomach is mobilized, resected and anastomosed with duodenum (gastroduodenostomy).
- BILROTH2 GASTRECTOMY: In this type of gastrectomy antrum and distal part of the body of the stomach is mobilized and resected, the duodenal stump is closed and distal part of stomach is anastomosed with jejunum (gastrojejunostomy).

BENIGN GASTRIC TUMORS

POLYPES - the most common benign tumors of the stomach. Most patients are asymptomatic. Some patients have epigastric pain and discomfort.

Types of gastric polyps:

- Hyperplastic polyps -the most common polyp. It develops on the basis of chronic gastritis, there is a risk of malignancy (>2 cm)
- Adenomatous polyps- 10-30% of all incidents, mostly located in antrum, high risk of malignancy, It develops more frequently on the basis of atrophic gastritis and intestinal metaplasia.
- Hamartomatous, inflammatory and beterotopic polyps – very rare and no risk of malignancy

MALIGN GASTRIC TUMORS

90-95% of stomach tumors are malignant; 95% of these are carcinoma, Lymphoma (4%), malignant GIST (1%), others carcinoid tumors of the stomach. Gastric carcinoma is major cause of cancer related mortality worldwide. This cancer is more common in Japan with incidence of the 70 cases per 100,000 populations per year.

ADENOCARCINOMA

- Costa Rica, Russia, China and Japan are the countries with the highest incidence in the world...
- Twice as common in men...
- Incidence reaches its peak at the age of 60-70.

- Environmental factors are more important than familial factors in the development of stomach cancer.
- incidence is higher in low socioeconomic areas.
- It is more common in people with blood type A.

ETHIOLOGY

- Factors that increase the risk of developing stomach cancer
  -family history
  -Diet (high nitrite, salt, fat)
  -Familial polyposis
  - Gastric adenomas
  -Hereditary non-polyposis cancer syndrome, Li-Fraumeni syndrome
  -Helicobacter pylori infection (atrophic gastritis, intestinal metaplasia, dysplasia)
  -pernicious anemia
  - Previous gastrectomy or gastrojejunostomy (at least 10 years later)
  - Smoking (alcohol is probably not a risk factor)
  -Menetrier's disease(massive gastric folds, hypertrophic gastropathy)
  -Epstein-Barr Virus infection
  - Genetic factors: P53, cox-2 (2 most common factors), APC, ras, CDHl.
  -The CDH1 gene encodes the E-cadherin protein. CDH1 mutation causes hereditary diffuse gastric cancer causes. This situation is an indication for prophylactic total gastrectomy.
  - Increased IL - I expression
- Factors that reduce the risk of developing stomach cancer:
  - Aspirin
  -Diet (intake of fresh vegetables and fruits)
  -vitamins C and E
- Premalignant conditions for stomach cancer
  -Adenomatous and hyperplastic polyp
  -Atrophic gastritis . The majority of stomach cancers develop on the basis of atrophic gastritis (95%).
  - Verrucous gastritis(chronic erosive gastritis)
  -intestinal metaplasia, dysplasia
  -Partial gastrectomy
  -Menetrier's disease(massive gastric folds, hypertrophic gastropathy)
  -Chronic superficial gastritis
  -Remnant stomach (stomach residue remaining after gastric resection)
  -40% of gastric adenocarcinomas are distal, 30% are middle and 30% are proximal tumors.

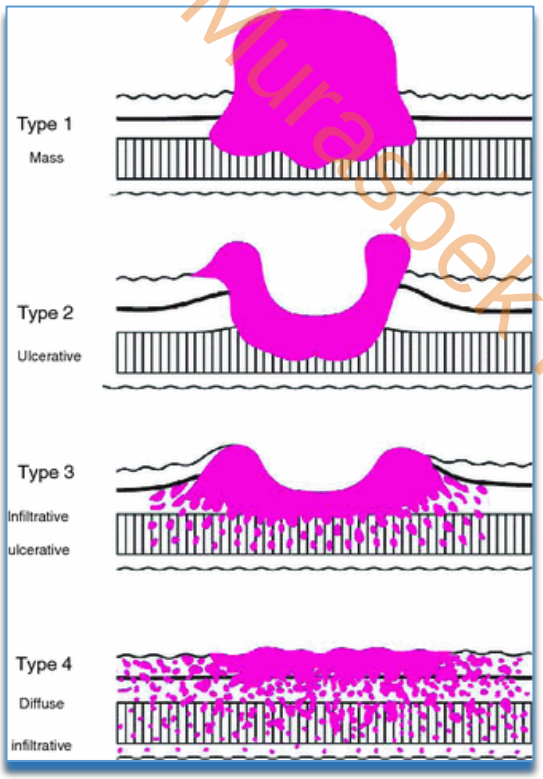
CLASSIFICATION

Gastric cancer can be divide into two types: early and advanced gastric cancer.

- Early gastric cancer involve (but not exceeding) the mucosa and submucosa.

- Metastasis may develop at any stage of early stomach cancer.
-Most commonly seen in the corpus.
-There are three types of early gastric cancer (superficial, exophytic, excavated)
- 95% cure is achieved with adequate gastric resection and adequate lymphadenectomy.

- Advanced gastric cancer is a neoplasm that has extended submucosa into muscularis propria and has classified into four types (Borman`s classification):



TYPE 1- Polyploidal well demarcated.

TYPE 2-Ulcerated with sharply demarcated margins.

TYPE 3-Ulcerative without definite margins.

TYPE 4-Diffuse infiltration without significant ulceration.

- TNM CLASSIFICATION

Tis - Carcinoma in situ

T 1 - Submucosa invasion (not exceeding the submucosa)

T2 - Muscularis propia invasion (not exceeding muscularis propria)

T3 - Invasion of subserosal connective tissue (not exceeding the serosa)

T4 - Exceeded the serosa (visceral peritoneum)

T 4a - Exceeded the serosa

T4b -Invaded neighboring structures

NO - No regional lymph node involvement

N 1 - Number of metastatic lymph nodes 1-2

N2 - Number of metastatic lymph nodes 3-6

N3 - Number of metastatic lymph nodes 7%

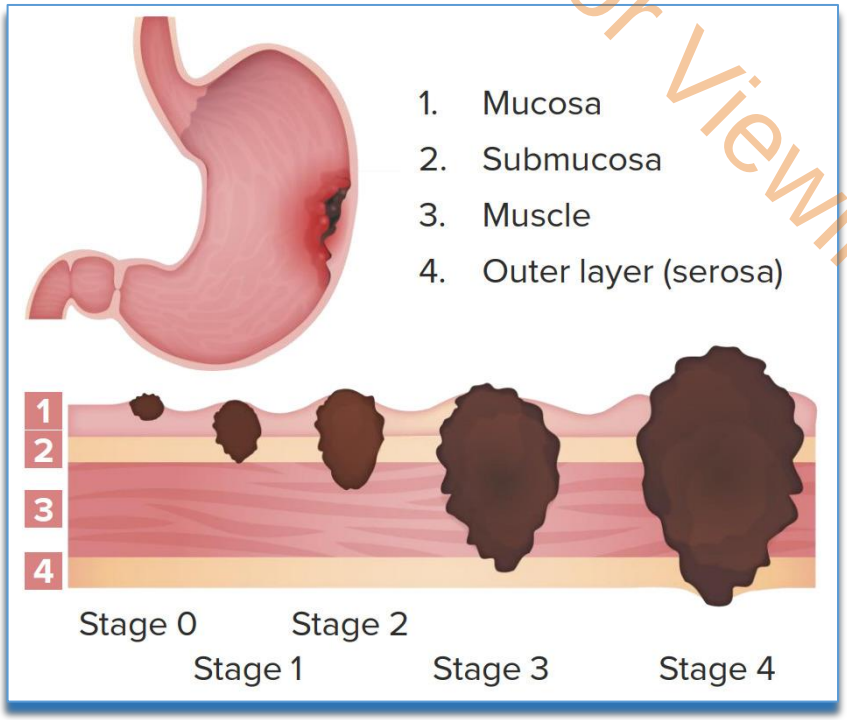
MO - No distant metastasis

M 1 - There is distant metastasis

STAGES OF GASTRIC CANCER

STAGES			
STAGE 0	Tis	N0	M0
STAGE I A	T1	N0	M0
STAGE I B	T2	N0	M0
	T1	N1	M0
STAGE IIA	T3	N0	M0
	T2	N1	M0

	T1	N2	M0
STAGE IIB	T4a	N0	M0
	T3	N1	M0
	T2	N2	M0
	T1	N3	M0
STAGE IIIA	T4a	N1	M0
	T3	N2	M0
	T2	N3	M0
STAGE IIIB	T4b	N0	M0
	T4b	N1	M0
	T4a	N2	M0
	T3	N3	M0
STAGE IIIC	T4b	N2	M0
	T4b	N3	M0
	T4a	N3	M0
STAGE IV	Any T	Any N	M1



CLINIC

- Symptoms begins quite late; The symptoms are not specific to the disease.
- Discomfort in the epigastric region is the most common initial symptom.
- In advanced stages, weight loss and abdominal pain may occur.
- Anorexia and weight loss are the most common complaints (in advanced stages).
- Weakness, dysphagia, nausea and vomiting may develop.
- Pain occurs in the late stages and it is a rare findings.
- Occult bleeding is common; In advanced stages, massive bleeding may occur.

SPREAD OF STOMACH CANCER

Direct invasion, implantation, hematogenous and lymphatic...

- Direct invasion into omentum, liver, pancreas and colon ...
- Implantation - Krukenberg tumor (ovarian), Blummer shelf (rectovesical/ rectouterine fossa) path
- Lymphatic pathway - Perigastric, iliac, splenic, pancreatic lymph nodes, ductus thoracicus pathway in advanced stages with left supraclavicular lymph node metastasis (Virchow node) . Left axillary lymph node metastasis (Irish nodule)
- Hematogenous - Most commonly to liver. Lung, bone, adrenal and skin metastases are also seen.
- Sometimes metastatic mass in the umbilicus (Sister Mary Joseph nodule).

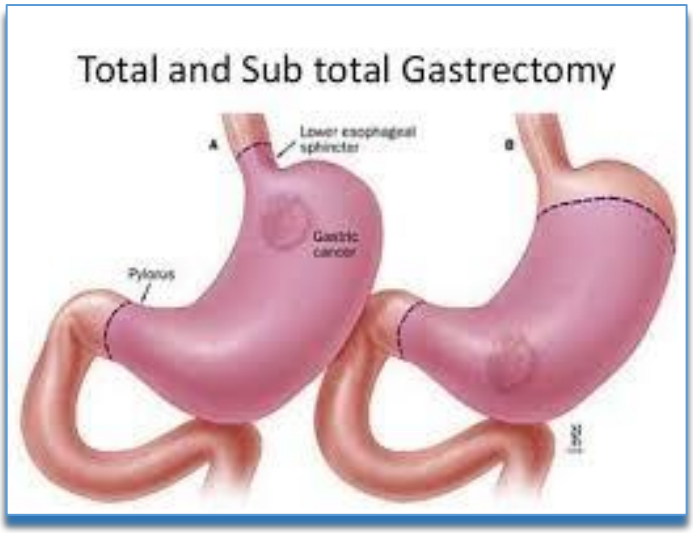
Paraneoplastic syndrome in gastric cancer:

- Trousseau syndrome: thrombophlebitis
- Acanthosis nigricans: hyperpigmentation in the axilla and inguinal region
- Peripheral neuropathy

DIAGNOSIS IN STOMACH CANCER

- Endoscopy - It is the gold standard diagnostic method.
- Endoscopic biopsy, pathological and cytological examination is performed.
- CT is used in preoperative stage and showing the extent of the disease in patients with stomach cancer.
- Endosonography and laparoscopy are helpful.
- The most useful imaging for early gastric cancer diagnosis is endosonography.

TREATMENT FOR STOMACH CANCER



- Treatment of stomach cancers is surgery.
- The most commonly applied surgical treatment for curative purposes is radical subtotal gastrectomy.
- Chemotherapy and radiotherapy are not very effective for curative treatment.
- In the most common distal 1/3 gastric carcinomas- subtotal, middle and upper 1/3 gastric locations- total gastrectomy is suitable for localized stomach cancers.

- For middle and upper 1/3 lesions, splenectomy should be added for radical surgery.
- Lymphatic dissection should be performed together with gastrectomy. Lymph node groups beyond the 3 cm border of the stomach (perigastric, left gastric, celiac,hepatic, splenic groups) is defined as 02 dissection and it is routinely applied in gastric cancer surgery.

PROGNOSIS IN STOMACH CANCER

- The most important factor determining prognosis is the stage of the disease.
- The prognosis is good in early stomach cancer.
- Cancer in mucosa-annual survival in cancers is 50-84%, in submucosal cancers it is 30-61%.
- Stages III and IV is 13-29% and 3-15% survival , respectively.
- This rate is 78.7% in early stomach cancers without lymph node metastases and 50% in those with lymph node metastases.

GASTRIC LYMPHOMA

- Although the stomach tissue has the least lymphoid tissue in the entire GIS.The most common place for lymphoma is the stomach.
- The majority of them are non-Hodgkin lymphoma.
- Most stomach lymphomas are B-cell type and originate from mucosa associated lymphoid tissue (mucosa associated lymphoid tissue - MALT).
- Increased MALT in the stomach during the development of chronic gastritis may indicate malignant degeneration.
- Responsible factor -H. pylori
- Low grade MALT lymphoma develops on the basis of chronic gastritis associated with H. pylori
- This relatively harmless tumor may later develop into high-grade lymphoma.
- When H. pylori is eradicated and gastritis resolves, low-grade MALT lymphoma often disappears
- Low grade MALT lymphoma is not need a surgical treatment. However, close follow-up is required.
- Effective antibiotic treatment (H.pylori eradication) can lead to complete treatment of low-grade gastric lymphoma.
- Close follow-up is required (Especially patients with t 11:18 translocation).
- High-grade gastric lymphomas are very different and require aggressive treatment.
- May be stomach involvement in systemic lymphoma.
- It may also develop as a mass of tumor; however, it often occurs secondary to lymphocyte infiltration in the submucosa. It is characterized by thickening of the rugae on its wall.

CLINIC

- It is similar to stomach cancer. However, the most common symptom is epigastric pain.

- Systemic symptoms such as fever, weight loss and night sweats occur in approximately 50% of patients .
- Tumor may cause obstruction and bleeding.
- Lymphadenopathy and/or organomegaly should suggest systemic disease.
- Definitive diagnosis is made by endoscopy and biopsy.
- It shows a spread pattern similar to stomach cancers. Local invasion to surrounding organs and to regional lymph's It tends to cause node metastasis and distant metastasis.
- When gastric lymphoma is diagnosed, lung, abdominopelvic, extra gastric disease should be carefully investigated by performing tomography and bone marrow biopsy.
- Treatment is the combination of chemotherapy and radiotherapy.
- Surgical treatment is used in recurrent cases.
- Most patients with primary gastric lymphoma can be treated without gastric resection.

MALiGN GASTROiNTESTINAL STROMAL TUMOR (GiST)

- Around 1% of malignant stomach tumors.
- 2/3 of malignant GISTs are located in the stomach.
- GiST tumors originates from the interstitial cell ofCajal (inside the muscle layer; pacemaker).
- That's why it was formerly called leiomyoma and leiomyosarcoma.
- GISTs rarely include the Carney triad (gastric GIST, paraganglionoma, and pulmonary chondroma) or it may be part of tumor syndromes such as neurofibromatosis type I.
- They are slow-growing, submucosal tumors.
- Most of them are located in the body of the stomach, and are usually a single lesion.
- Small lesions are usually detected incidentally.
- Larger tumors cause weight loss, abdominal pain, fullness, early satiety, and bleeding.They can form a palpable mass in the abdomen.
- They most commonly present with epigastric pain and bleeding.
- Lymphatic spread is rare; the main spread is hematogenous to Liver and lung.
- Endoscopy and biopsy are main diagnostic methods.

TREATMENT:

- Tumor > 2 cm –Surgery: wide local excision, enucleation,sleeve gastrectomy or total gastrectomy ± en bloc resection of surrounding organs ...
- Tumor < 2 cm ~ Controversial... Surgery recommended in patient`s with high risk (ulceration, heterogeneity, irregular borders), in low-risk patients follow-up with EUS is recommended at 6-12 month intervals.
- Prognosis depends on tumor location, tumor size and level of mitosis.

- The prognosis is better than adenocarcinoma.
- Low grade tumors can be cured (5 year survival 80%), high grade tumors have a poor prognosis (5 annual survival 30%).
- **C-kit (CD 117) proto-oncogene** is usually found in GISTs. **Imatinib** is inhibits the activity of the tyresine kinase product of this gene and can use in treatment of metastatic or non-surgically resectable GISTs
- In imatinib-resistant cases, **sunitinib** is given.

HYPERTROPHIC GASTRITIS (MENETRIER DISEASE)

- It is an inflammatory disease characterized by the hypertrophic gastric folds, especially in the proximal stomach.
- Autoimmune etiology is considered.
- Mucus-secreting cells are hyperplastic, but parietal cells are reduced.
- Increased TGF-alpha is responsible for the development of the disease.
- TGF-alpha activates the epidermal growth factor receptor. This causes hyperplasia of mucus-secreting cells.
- Bleeding due to mucosal ischemia.
- Hypoproteinemia.
- Monoclonal antibody **cetuximab** against the epidermal growth factor receptor is used in the treatment.
- When surgical treatment is required, total gastrectomy is performed.

BEZOARS

It is the mass found trapped in gastrointestinal system usually in stomach. There are two types:

- Trichobezoar – is the mass of the hair balls, uncommon condition exclusively found in psychiatric patients (mostly in females) caused by ingestion of hair, which remain undigested in the stomach.
- Phytobezoar- mass of the vegetable balls found in patients with gastric stasis.
- In the treatment firstly, proteolytic enzymes and endoscopic interventions are used, and if necessary, laparotomy is performed.

WATERMELON STOMACH (GASTRIC ANTRAL VASCULAR ECTASIS)

This name comes from the parallel red lines seen at the top of the mucosal folds in the distal part of the stomach.

- Histologically, gastric antral vascular ectasia is characterized by dilated mucosal blood vessels in the lamina propria and It is characterized by the tiny clots they often contain.
- Often mucosal fibromuscular hyperplasia and hyalinization are present.

- It is similar to gastropathy due to portal hypertension(but this condition involves the proximal stomach). In the watermelon stomach, involved the distal part of stomach.
- Typical patients are women with chronic gastrointestinal bleeding that often requires transfusion.
- Most of them have accompanying autoimmune connective tissue diseases and chronic liver disease in at least 25% of cases.
- Estrogen and progesterone can be used in treatment.
- Antrectomy may be necessary to prevent bleeding.

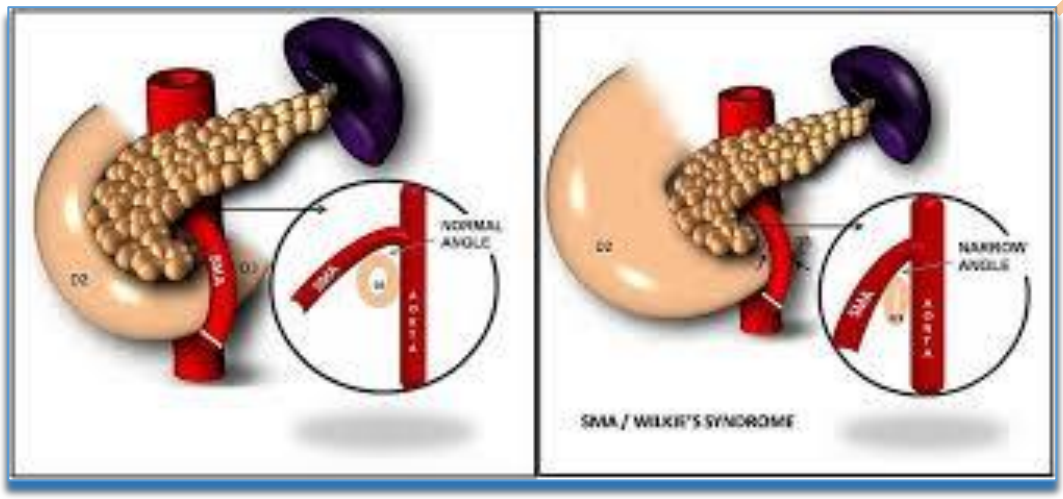
DIEULAFOY LESION

- It is a congenital arteriovenous malformation.
- There is an abnormally large submucosal artery.
- If this artery bleeds, massive, pulsatile bleeding may occur.
- Endoscopy is used in diagnosis and treatment.

PORTAL HYPERTENSIVE GASTROPATHY

- Gastric mucosa changes associated with portal hypertension is named as portal hypertensive gastropathy.
- Portal hypertensive gastropathy can cause acute or chronic bleeding.

WILKIE SYNDROME



(SUPERIOR MELENTER ARTERY SYNDROME)

- As a result of compression of the third part of the duodenum by the superior mesenteric artery(SMA), duodenal obstruction findings.
- The angle between the aorta and the SMA is less than 25°.

- Rapid weight loss (loss of retroperitoneal fat pads) and It occurs as a result of (scoliosis surgery and body casts) hyperextension of the vertebrae
- Clinical findings: feeling of fullness in the epigastrium after meals, nausea, vomiting and vomiting
- Diagnosis: Barium radiography and angiography
- Treatment

  -installation of NGT, liquid and electrolyte correlation therapy in the presence of acute symptoms. Supine position should be avoided.

  -If medical treatment is unsuccessful, duodenojejunostomy is performed.

COMPLICATIONS SEEN AFTER GASTRIC SURGERY

- They are associated with vagotomy or gastrectomy.
- Post-gastrectomy syndromes can occur due to the loss of the stomach's reservoir function, the elimination of the pyloric sphincter mechanism, due to the cutting of vagus nerve.

COMPLICATIONS RELATED TO VAGOTOMY

The complications of vagotomy are categorized into three groups: intraoperative, early postoperative, and late postoperative complications.

- Intraoperative Complications are Injuries to adjacent organs and vessels.
- Early Postoperative Complications

  - Gastric stasis.

  - Dysphagia.
- Late Postoperative Complications

  - Diarrhea. Clinically significant diarrhea develops in 5-10% of patients after truncal vagotomy. It is most common after vagotomy combined with drainage. The exact cause is unknown.

    Possible mechanisms include:

    - Intestinal motility disorders and accelerated transit.

    - Rapid passage of unconjugated bile salts from the denervated biliary system to the colon.

    - Bacterial overgrowth.

    - In prolonged cases, long-term oral **cholestyramine** is used. Medications such as **codeine** and **lomotil** are beneficial.

    - If there is no response, surgical treatment is considered (a 10 cm reversed jejunal loop is transposed 100 cm distal to Treitz ligament.

  - Cholelithiasis - Seen after truncal vagotomy.

  - Reflux esophagitis.

EARLY COMPLICATIONS RELATED TO GASTRECTOMY

- DUODENAL STUMP LEAKAGE

  - It is often seen between the 2nd and 7th postoperative days.

- It is characterized by sudden onset of pain, fever, and tachycardia, along with a deterioration in the patient's general condition.

- Emergency surgical drainage should be performed, fluid and electrolyte balance should be well monitored, and TPN should be initiated.

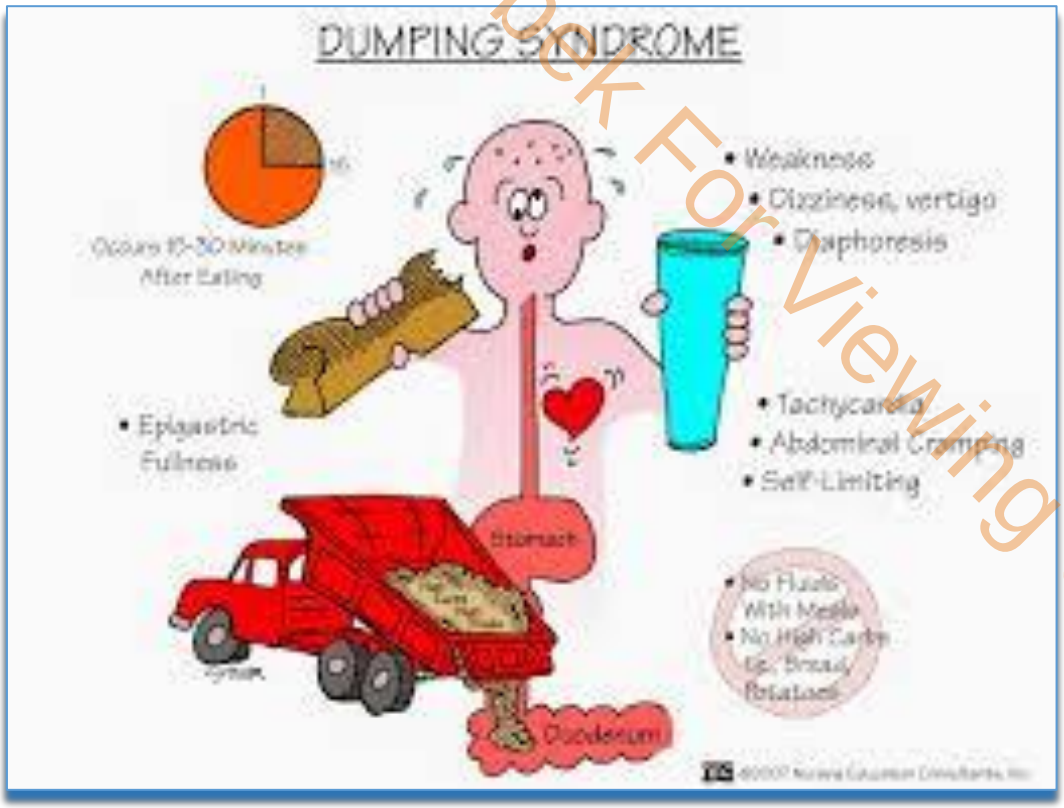
- The cause is often a technical error and the opening of the duodenal suture line.

- Despite appropriate treatment, mortality is very high.

LATE COMPLICATIONS OF GASTRECTOMY

- EARLY POSTPRANDIAL DUMPING SYNDROME

-



Symptoms begin during or immediately after a meal; they subside within 40-60 minutes.

- Vasomotor symptoms are usually predominant, overshadowing gastrointestinal symptoms.

- Vasomotor symptoms include sudden weakness, faintness, dizziness, a desire to lie down, cold sweats, and palpitations.

- Gastrointestinal symptoms include epigastric fullness and a sensation of turbulence. Vomiting is rare but may occur as explosive diarrhea and intestinal cramps.

- High-carbohydrate foods exacerbate the symptoms.

- The incidence is related to the amount of stomach removed.

- It is less common after Billroth I reconstruction.

Pathogenesis

- The fundamental initiating event is the rapid, uncontrolled emptying of hyperosmolar foods from the stomach into the jejunum.

- The critical factor is foods high in carbohydrates and hyperosmolar contents.

- The sudden passage of hyperosmolar content into the jejunum causes rapid fluid shift from the intravascular compartment to the intestinal lumen, leading to a decrease in plasma volume.

- Additionally, hyperglycemia may develop.

- Hormones released from the distended jejunal wall, such as serotonin, kinins, substance P, neurotensin, enteroglucagon, and peptide YY, also play a role.

Treatment

- A diet low in carbohydrates, eating small and frequent meals, avoiding liquids during meals, or taking them 30 minutes to an hour after eating is recommended.

- **Pectin** can be used to delay gastric emptying. **Octreotide** is beneficial.

- In patients unresponsive to treatment, surgery is performed. The aim is to slow gastric emptying.

LATE (HYPOGLYCEMIC) DUMPING SYNDROME

- It is less common. Symptoms begin 1-3 hours after eating.

- Vasomotor symptoms are the same; gastrointestinal symptoms are absent or very mild.

- The rapid emptying of carbohydrates from the stomach into the jejunum causes a rapid increase in postprandial blood glucose.

- The material passing into the jejunum leads to excessive release of enteroglucagon.

- This substance stimulates the pancreas to secrete insulin, resulting in hypoglycemia.

- Symptoms subside after glucose intake.

- The treatment is the same as for early postprandial dumping syndrome.

- After a few months of low-carbohydrate diet, enteroglucagon secretion decreases, and hypoglycemia does not occur.

- Surgical treatment is almost never necessary.

ALKALINE REFLUX GASTRITIS

- It occurs as a result of the cutting, removal, or bypassing of the pylorus (pyloroplasty, gastrectomy, gastroenterostomy).

- Uncontrolled reflux of duodenal contents into the stomach causes gastritis, leading to abdominal pain and bile-stained vomiting in patients.

- The incidence is 5-35%.

- Rarely, it can also occur in individuals who have not had surgery due to pyloric dysfunction.

- It presents with epigastric pain, bile-stained vomiting, and weight loss.

- The pain is present in 80% of cases, is burning in nature, constant, worsens with eating, and does not improve with vomiting.

- Vomit contains bile and food remnants.

- The diagnosis is made by endoscopic biopsy, which is the gold standard.

Treatment:

- Mild symptoms: Conservative treatment with mucosal protective agents (sucralfate, alginic acid).

- No response to medical treatment: Surgery.

- The aim of surgical treatment is to keep pancreatic, bile, and duodenal contents away from the stomach.

- The most common method is Roux-en-Y diversion.

AFFERENT LOOP SYNDROME

- It develops due to the obstruction of the afferent loop.

- The obstruction is usually partial.

- Characteristic symptoms include postprandial cramp-like abdominal pain, explosive bile-stained vomiting, and relief after vomiting.

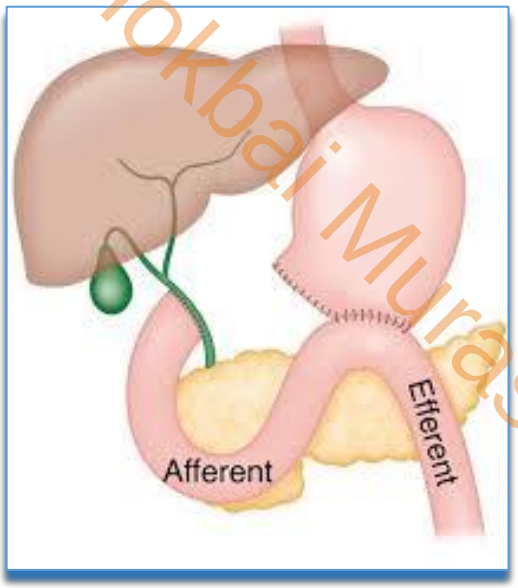
- The pain is relieved by vomiting. The vomit does not contain food remnants.

- If untreated, anemia and malabsorption develop due to vitamin B deficiency caused by bacterial colonization in the afferent loop.

Treatment:

- It is surgical.

- Billroth II can be converted to Billroth I, or a side-to-side jejunojejunostomy or Roux-en-Y diversion can be performed.



EFFERENT LOOP SYNDROME

- It is especially seen in cases with retrocolic anastomosis.

- It is usually related to a band (adhesion).

- Jejuno-gastric intussusception can also lead to efferent loop syndrome (obstruction).

- Vomiting is present (containing food remnants that have been in the stomach), but the pain does not resolve with vomiting (it does resolve in afferent loop syndrome).

- If there is improvement with conservative treatment within 7-10 days, it indicates that the obstruction is likely due to edema.

- If it does not resolve, surgery is necessary.

- **CANCER DEVELOPMENT**

- Etiological factors include hypochlorhydria and enterogastric reflux.

- The incidence of gastric remnant (stomach residue) carcinoma is 0.4-8.9%, with a latent period of 10-30 years.

- **ROUX SYNDROME**

- Difficulty in gastric emptying without mechanical obstruction following gastrectomy and Roux-en-Y gastrojejunostomy.

- Symptoms may include vomiting, pain, and weight loss.

- Roux-en-Y gastrojejunostomy should be avoided if more than half of the stomach is not removed. Otherwise, marginal ulcers and issues such as gastric stasis (Roux syndrome) may occur.

- **MARGINAL ULCER**

- Also known as recurrent ulcer, anastomotic ulcer, or stomal ulcer.

- These are ulcers seen on the jejunal side of the gastrojejunostomy.

- Parietal cell activity continues.

Predisposing Factors for Marginal Ulcer Formation:

- Gastric stasis
- Incomplete vagotomy
- Inadequate antrum resection
- Functional parathyroid adenoma
- Zollinger-Ellison syndrome
- Long afferent loop
- Helicobacter pylori colonization

Symptoms:

- Symptoms are the same as those of peptic ulcers.
- Complications such as bleeding, perforation, penetration, and obstruction can develop.

Diagnosis:

- Barium X-ray and endoscopy.
- In patients who have undergone vagotomy, the adequacy of the vagotomy can be assessed by measuring pancreatic polypeptide (PP) levels after sham feeding (e.g., chewing gum).
- An increase in PP levels > 50% at thirty minutes indicates incomplete vagotomy.

Treatment:

- If complications have not developed, treatment includes PPI and antacid therapy. Surgical treatment is planned based on the underlying cause.

- **BONE DISEASES**

- After gastric surgery, disturbances in calcium and vitamin D metabolism can sometimes occur.
- Impaired absorption of calcium and vitamin D can lead to metabolic bone diseases.
- Years after surgery, bone pain and/or fractures may develop.
- Dietary calcium and vitamin D replacement can help prevent these complications.

- **ANEMIA**

- Anemias can develop due to deficiencies in iron, vitamin B12, and folic acid.

- **Iron Deficiency Anemia**

- 30% of patients who undergo gastrectomy develop iron deficiency anemia within 5 years.

PATHOGENESIS:

- After gastrectomy, HCl decreases. Since HCl is necessary for converting ferric iron to ferrous iron and facilitating its absorption via vitamin C, iron deficiency anemia can develop.

- Gastric factor (gastroferrin), which helps with iron absorption, is lost.

- The duodenum, where a large portion of iron absorption occurs, is bypassed.

- As a preventive measure, patients are given oral iron preparations.

- **MEGALOBLASTIC ANEMIA**

- Usually due to vitamin B12 deficiency, and rarely due to folate deficiency.

- Anemia due to vitamin B12 deficiency typically develops after several years due to the body's storage of the vitamin.

- It is related to the loss of intrinsic factor.

- HCl helps facilitate the release of vitamin B12 from food. Decreased stomach acid also predisposes to vitamin B12 deficiency.

- Additionally, bacterial colonization in the afferent loop can lead to vitamin B12 deficiency.

- **GASTRIC STASIS**

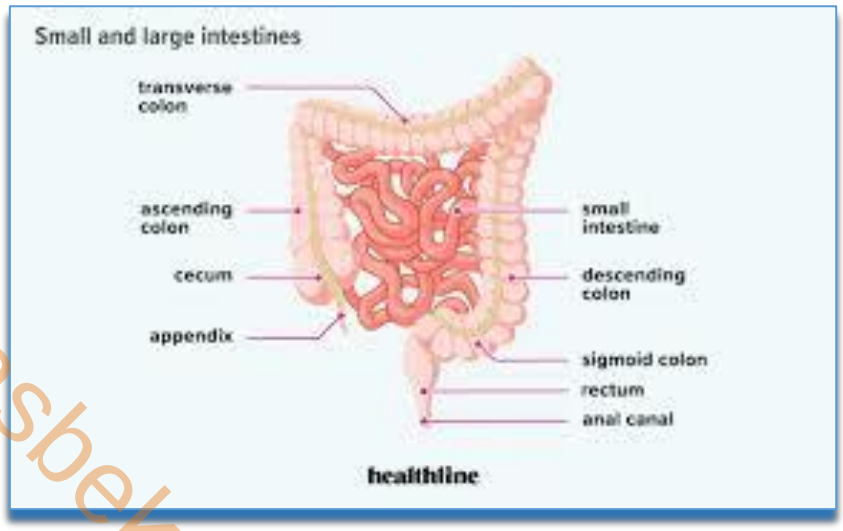
- It is due to a problem with motor function or obstruction.

- Symptoms include vomiting, bloating, weight loss, and epigastric pain.

- The best methods to measure gastric emptying are scintigraphic techniques that show the half-life of solid and liquid food emptying.

- The most commonly used prokinetic agents in medical treatment are **metoclopramide** and **erythromycin**.

Small Bowel Diseases and Surgery

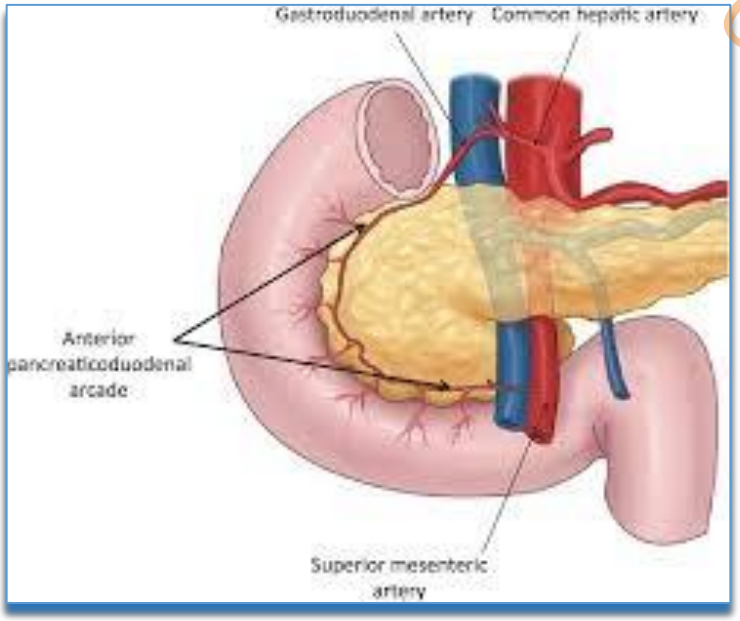


- The small intestine is the part of the digestive tract that extends from the pylorus to the large intestine.

- In adults, it is approximately 6 meters long.

- It is the largest organ of the endocrine and immune systems.

- It consists of three parts: the duodenum, jejunum, and ileum.



Duodenum

- It is approximately 20 cm long. It has no mesentery and is retroperitoneal.

- Duodenum has four parts:

- Superior Part (1st Part): 5 cm long. It starts at the pylorus and ends at the neck of the gallbladder. Its anterior surface is covered by the peritoneum.

- Descending Part (2nd Part): 8-10 cm long. It extends from the neck of the gallbladder down along the right edge of the vertebral column to the lower edge of the 3rd lumbar vertebra.

- Inferior Transverse Part (3rd Part): 10 cm long. It begins at the lower edge of the 3rd lumbar vertebra and ends in front of the abdominal aorta.

- Ascending Part (4th Part): 2-5 cm long. It forms the duodenojejunal junction. This segment is attached to the diaphragm by a fibromuscular band called the suspensory ligament of the duodenojejunal flexure (Treisz ligament).

Jejunum-Ileum

- The jejunum has an average diameter of 4 cm, while the ileum has an average diameter of 3 cm.

- The wall has four layers:

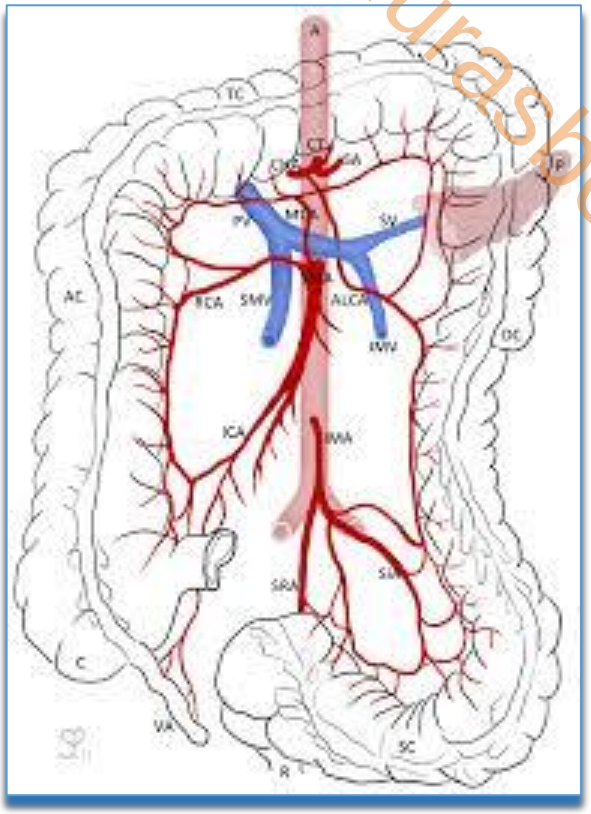
- Serosa

- Muscular Layer (longitudinal and circular)

- Submucosa. The strongest layer. It is the most important layer through which sutures must pass in anastomoses.

- Mucosa. The mucosa contains circular folds (Kerckring folds), villi, lymph follicles, and glands.

- Glutamine is a major energy source for enterocytes.



- The ileum predominantly fills the right lower quadrant, while the jejunum is more located in the left upper quadrant and is more mobile. There are also some macroscopic differences between the jejunum and ileum.

- Except for the proximal duodenum, the blood supply to the small intestine comes from the superior mesenteric artery (SMA).

- The ileum is particularly rich in lymphatic networks.

- Lymphatic flow begins in the mucosa and ends in the thoracic duct via the cisterna chyli.

- Parasympathetic innervation is provided by the vagus nerve.

- Sympathetic innervation occurs via the splanchnic nerves.

- The nerve plexuses within the small intestine wall include:

  - Myenteric Plexus (Auerbach's Plexus): Located between the two muscle layers.

  - Meissner's Plexus: Found in the submucosa.

- Proliferation in the small intestine is very rapid.

- Intestinal cell renewal in the small intestine is completed approximately every 5-7 days.

- Regeneration occurs most rapidly in the ileum (the epithelium in the ileum renews itself approximately every three days).

Small Intestine Physiology

- The small intestine is responsible for the absorption of fats, proteins, carbohydrates, fluids, and electrolytes.

- Additionally, the small intestine functions as an endocrine organ.

- The small intestine secretes several hormones including secretin, cholecystokinin, gastrin inhibitory peptide (GIP), vasoactive intestinal peptide (VIP), glucagon-like peptide (GLP-2), enteroglucagon, motilin, bombesin, somatostatin, neurotensin, and peptide YY.

- The motility of the small intestine is under neural and hormonal control.

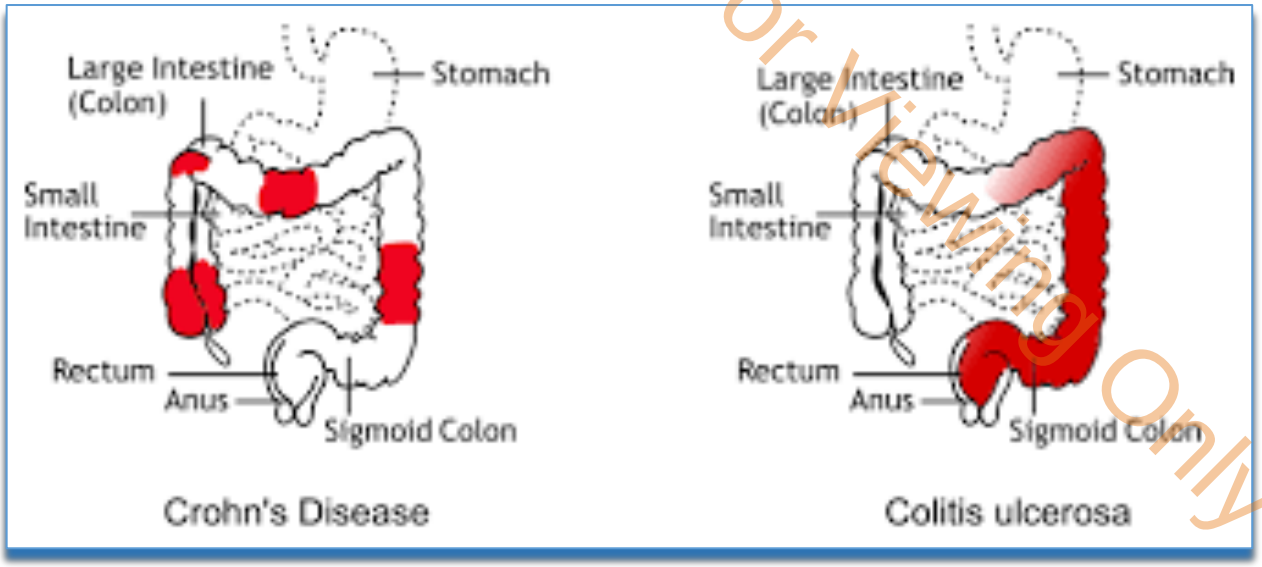
- Neural stimulation is mediated by the vagus and sympathetic nerves.

- Vagal fibers can be classified into two groups: cholinergic fibers that increase motility and peptidergic fibers that suppress motility.

- Sympathetic nerves reduce motility and regulate the vasoconstriction of intestinal blood vessels.

- Additionally, the peptides secreted by the small intestine also have effects on motility.

CROHN DISEASES



It is a transmural chronic inflammatory disease that can occur in any part of the gastrointestinal system (GIT), characterized by acute exacerbations and spontaneous remissions.

- It is more common in North America and Europe, while its frequency is significantly lower in Black and Asian populations.

- It can occur anywhere in the GIT, but it is most frequently found in the terminal ileum.

- For this reason, it is also referred to as terminal ileitis or regional enteritis.

- **Etiology**

- The etiology is not yet fully understood.

- Factors such as specific infections, immunological events, stress, genetic predisposition, and environmental influences are suspected.

- Infectious agents like Mycobacterium paratuberculosis, Mycobacterium avium, and Yersinia enterocolitica are particularly implicated.

- Although autoimmune parameters have been found to be abnormal, concrete evidence is lacking.

- The most significant risk factor identified today is genetic predisposition. A positive family history greatly increases the risk.

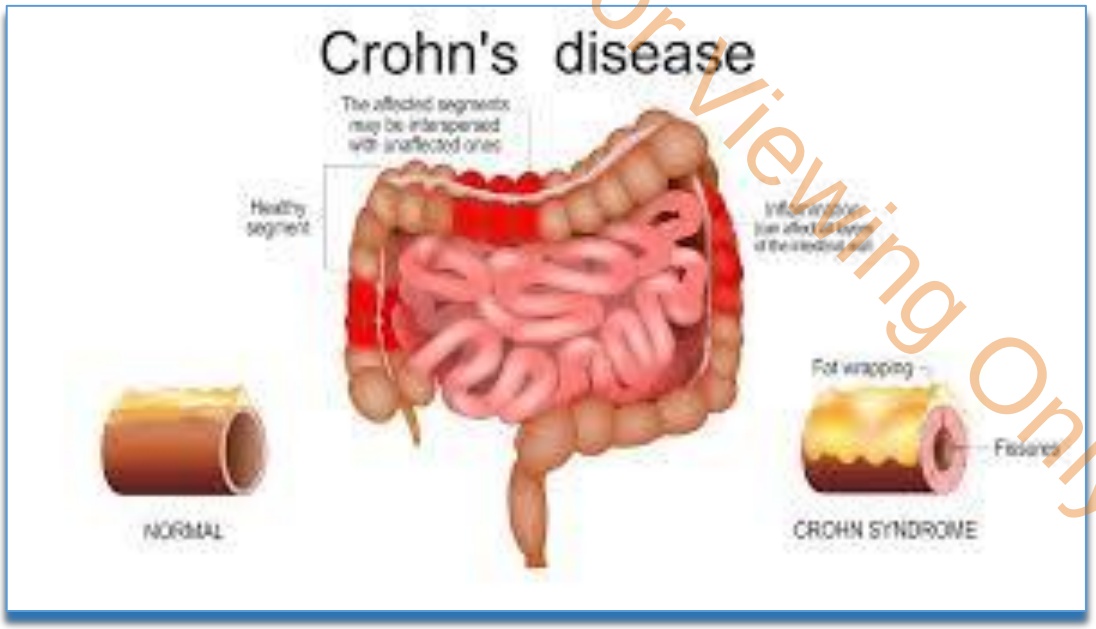
- Genes with the most frequent and strongest associations include NOD2, IL-23R, and ATG16L1.

- High socioeconomic status is a risk factor for Crohn's disease.

- Many studies have shown a protective effect of breastfeeding.

- There are publications suggesting that smoking increases the risk of developing Crohn's disease. Additionally, smoking increases the frequency of flare-ups and the need for surgical treatment in Crohn's disease.

- **Pathology**



- The earliest lesions observed in Crohn's disease are aphthous ulcers. Over time, linear ulcers develop, and in advanced stages, the entire bowel wall is affected.

- Non-caseating granulomas are highly specific for the disease.

- The most prominent pathological finding is submucosal and subserosal fibrosis, which leads to strictures:

  - Due to the stricture, dilation occurs in the proximal segments.

- Crohn's disease is a transmural disease that affects all layers of the intestine.

- There are three forms of the disease: fibrostenotic, fistulizing, and aggressive inflammatory.

- **CLINIC**

- Although it can occur at any age, the typical Crohn's patient is a young adult in their 2nd or 3rd decade of life.

- The most commonly encountered symptoms are abdominal pain, diarrhea, and weight loss.

- Pain is the most frequent symptom. It is typically intermittent and colicky, often located in the lower quadrant.

- Episodes of pain and diarrhea that alternate with periods of remission are characteristic and highlight the chronic nature of the disease.

- Diarrhea is the second most common symptom.

- Diarrhea typically follows colicky abdominal pain; it is explosive in nature and often nocturnal.

- Additionally, fever, weight loss, weakness, and fatigue may occur. Pain and other clinical findings can mimic acute appendicitis.

- The most common intestinal complications are obstruction, followed by perforation.

- Perforation can lead to widespread peritonitis, but internal fistulas and abscesses are more frequently observed.

- In 80% of cases, there is involvement of the small intestine, while 20% involve only the colon.

- Most cases with small bowel disease are located in the ileocecal region.

- Isolated small bowel involvement occurs in 15-30% of cases.

- Isolated perineal and anorectal disease occurs in about 5-10% of cases.

- Perineal disease is present in 25% of patients with small bowel involvement and in 50% of those with colonic involvement.

- The least commonly affected areas are the esophagus, stomach, and duodenum.

- **Diagnosis**

- Diagnosis is made through history, endoscopic findings, biopsies, and radiological tests.

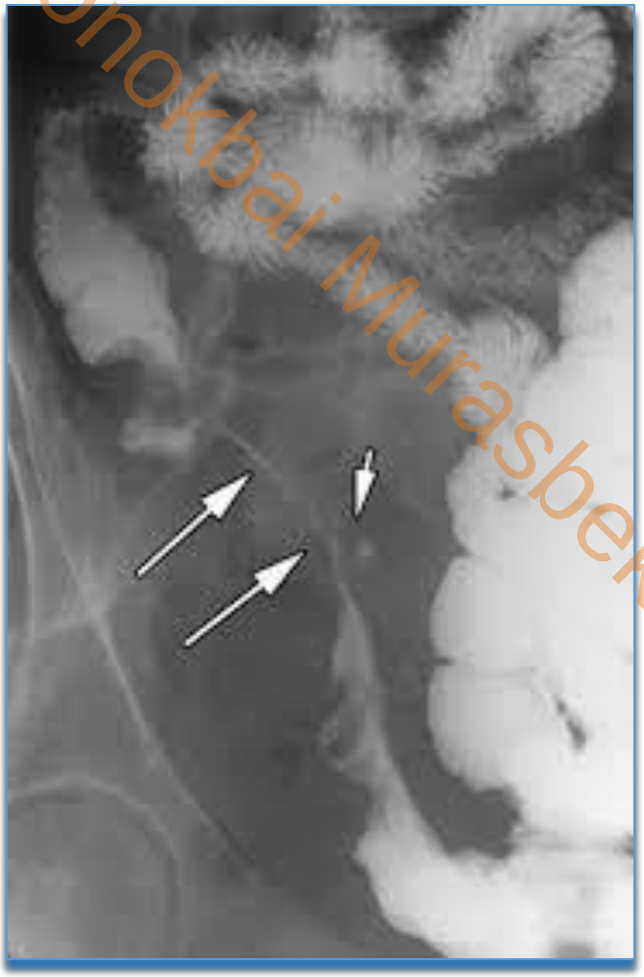
Radiological Findings:

- Nodular contour

- Linear and deep ulcers

- Fistulas and abscesses

- Cobblestone appearance



- Thickening and asymmetric involvement of the right colon wall

- Displacement of bowel loops

- Diffuse narrowing of the lumen

- Skip areas (areas without disease)

- Sinuses

- "String sign" (Terminal ileum appears very narrowed, resembling a string)

- Capsule endoscopy is useful in diagnosis and is increasingly used.

- Endoscopic advancements, such as double-balloon enteroscopy,

- CT and MRI are equally valuable in assessing disease activity and intestinal damage. However, MRI may have superior ability compared to CT in detecting intestinal strictures and wall thickening in the ileum.

- 92% of Crohn's disease patients test positive for ASCA (+) and negative for pANCA (-).

- 98% of ulcerative colitis patients test negative for ASCA (-) and positive for pANCA (+).

- **Complications**

Local Complications (Related to the gastrointestinal system):

- Obstruction- The most common complication

- Intraabdominal abscess

- Internal fistula

- Perforation

- Bleeding

- Perianal abscess and fistula

Systemic complications :

- Malnutrition

- Growth and development delays

- Anemia

- Migratory polyarthritis affecting small joints

- Extrintestinal involvement is seen in 30% of patients.

- The most common extrintestinal symptoms are skin lesions.

- Erythema nodosum, pyoderma gangrenosum

- The severity of erythema nodosum and peripheral arthritis parallels the severity of intestinal inflammation.

- **Extrintestinal Findings of Crohn's Disease**

- Dermatological: Erythema nodosum, Pyoderma gangrenosum

- Rheumatological: Peripheral arthritis, Ankylosing spondylitis, Sacroiliitis

- Ocular: Conjunctivitis, Uveitis, Iritis, Episcleritis

- Hepatobiliary: Hepatic steatosis, Cholelithiasis, Primary sclerosing cholangitis, Pericholangitis

- Urological: Nephrolithiasis, Ureteral obstruction (the most common urological complication)

- Other: Thromboembolic disease, Vasculitis, Osteoporosis, Endocarditis, Myocarditis, Pleuropericarditis, Interstitial lung disease, Amyloidosis, Pancreatitis

- **Treatment**

- There is no curative treatment.

- Initially, all patients should receive medical treatment.

- A diet that is easy to digest, low in fiber, and nutritionally supplemented is recommended.

- In addition to the diet, **sulfasalazine** is started, beginning with 500 mg twice daily, and can be increased up to 4000 mg per day.

- In severe cases, adding steroids to the treatment may be necessary. **Budesonide** is also beneficial and has fewer side effects.

- **6-mercaptopurine** and **azathioprine** can be used. **Azathioprine** is particularly useful in patients with fistulas.

- **Cyclosporine**, radiotherapy, BCG applications**, levamisole**, and immunoglobulins are other treatment methods, but their success rates are low.

- In recent years, **infliximab (Remicade**), an anti-TNF-alpha antibody, has been observed to induce remission and accelerate the closure of enterocutaneous fistulas. However, it is not used in septic conditions.

- **Adalimumab (Humira)** is a human anti-TNF antibody.

- For perineal disease, the first-line treatment is **ciprofloxacin** and **metronidazole**. If there is no improvement or if recurrence occurs after 2-4 weeks of treatment, azathioprine should be used. **Azathioprine** and **infliximab** are indicated for patients with fistulas.

- Crohn's disease is the most common surgical disease of the small intestine in Western societies.

- 60-75% of patients will require surgery at some point in their lives.

- More than 75% of patients will have symptoms and signs of obstruction to some degree.

- The cause of obstruction is often due to inflammation, fibrosis, or edema narrowing the bowel lumen, or it may result in paralytic ileus. Mechanical issues and obstructions can also arise from adhesions.

- In the presence of obstruction, resection and end-to-end anastomosis are performed.

- Resections cannot completely eliminate the disease; further surgeries may be necessary, so resections are kept limited.

- Asymptomatic enteric-enteric fistulas without signs of sepsis do not require surgical intervention.

- Resection boundaries should be determined according to the macroscopic extent of the disease.

- Recent studies have shown that stricturoplasty can correct strictures without resection.

- Strictureplasty is contraindicated in the presence of intra-abdominal abscess or fistula.

- In cases where resection cannot be performed due to extensive inflammation or in severe and high-risk patients, various bypass procedures such as ileotransversostomy may be used, though they are not the first choice.

- Abscesses are treated with percutaneous drainage and antibiotics. However, in cases of fistula development or uncontrolled sepsis, surgery is required.

- In a patient presenting with acute abdominal symptoms and undergoing surgery with a preliminary diagnosis of acute appendicitis, if Crohn's disease is found during the operation and the cecum is inflamed, recommended to do the caecum resection.

appendicitis	Crohn	Treatment tactic
+	+	Caecum resection
+	-	Appendectomy
-	-	Appendectomy
-	+	Don`t touch

TYPHOID ENTERITIS

- Typhoid is an acute, febrile illness caused by Salmonella typhi and paratyphi.

- In 10-20% of patients, serious bleeding occurs, and perforation happens in about 2% of cases. Perforation is typically observed in the last 30 cm of the ileum.

- **Fluoroquinolones** and third-generation **cephalosporins** are the first choice for antibiotics.

- In cases of perforation, primary repair is sufficient. Resection increases mortality and morbidity.

- For patients with multiple perforations (about 25%), resection with primary anastomosis or loop ostomy is preferred.

- Even in treated patients, carrier status develops in about 3% of cases, with the source being the gallbladder.

- For functional gallbladders, **ampicillin + probenecid + amoxicillin** or **trimethoprim + sulfamethoxazole** are given for up to 1 month. If drug therapy fails or the gallbladder is non-functional, cholecystectomy is performed.

BENIGN TUMORS OF SMALL INTESTINE

- The majority of benign tumors are asymptomatic.

- When symptomatic, they most commonly cause abdominal pain due to partial or intermittent obstruction, and bleeding is another frequent complication.

- They may also lead to intussusception or volvulus.

- Adenomas and stromal tumors are the most common benign tumors.

- Benign tumors are more frequently observed in the distal small intestine.

- In autopsy series, adenomas are the most frequently observed benign tumors, but stromal tumors are the most common symptomatic benign tumors of the small intestine.

Malignant Tumors of the Small Intestine

- Small intestine malignant tumors: adenocarcinoma 35-50%, carcinoid tumors 20-40%, lymphoma 10-15%, GIST 15%.

- Despite the small intestine constituting more than 90% of the mucosal surface of the gastrointestinal system, it accounts for only 1.1% to 2.4% of gastrointestinal malignancies.

- Adenocarcinomas are more frequently seen in the proximal small intestine; other malignant lesions are more common in the distal small intestine.

- The small intestine is the least common site for cancer in the gastrointestinal system.

-Reasons for the Rarity of Small Intestine Adenocarcinomas:

- The small intestine's content is low in fluid, alkaline substances, and bacteria.
- The rapid transit time of contents through the small intestine.
- The rapid turnover of small intestine cells.
- The protective apoptotic mechanisms in the small intestine wall; high levels of IgA in the wall.

- Most small intestine neoplasms are asymptomatic until they reach large sizes.

- Partial small intestine obstruction, accompanied by cramp abdominal pain, distension, nausea, and vomiting, is the most common presentation in clinical settings.

- Painless bleeding is typically the second most common symptom.

- Physical examination may reveal abdominal masses in up to 25% of cases and obstruction findings; however, there may be no detectable findings.

- Fecal occult blood tests may be positive.

Small Intestine Adenocarcinomas

- These account for approximately 50% of malignant tumors located in the small intestine.

- Adenocarcinomas, except those associated with Crohn's disease, most commonly occur in the duodenum.

- In Crohn's disease patients, they most commonly occur in the ileum.

- About half of duodenal adenocarcinomas develop from the ampulla of Vater.

- Jaundice with intermittent onset and fecal occult blood positivity are characteristic of ampullary carcinomas.

- Treatment involves wide resection of the affected segment of the intestine plus regional lymphadenectomy.

- There is no proven efficacy of chemotherapy in the adjuvant and palliative treatment of small intestine adenocarcinomas.

- The prognosis is poor.

Gastrointestinal Stromal Tumors (GIST)

- Originates from interstitial cells of Cajal.

- The defining characteristic of GISTs is the expression of the tyrosine kinase KIT receptor.

- About 60-70% of GISTs are located in the stomach.

- The small intestine is the second most common location for GISTs (25-35%).

- GISTs bleed more frequently compared to other small intestine cancers.

- Small intestine GISTs are treated with segmental resection. If the GIST diagnosis is known before resection, extensive lymphadenectomy is not meaningful, as GISTs rarely metastasize to lymph nodes.

- GISTs are resistant to classic chemotherapy agents; **Imatinib (Gleevec)** is used.

- **Imatinib** is used in adjuvant therapy for high and intermediate-risk patients after resection. Risk is determined based on tumor size, mitotic rate, and location.

- Approximately 80% of GISTs that are inoperable or metastatic benefit from **Imatinib**.

- **Sunitinib** is used for cases resistant to Imatinib.

- **Regorafenib** and **Nilotinib** are second-generation tyrosine kinase inhibitors.

- They are used for cases resistant to **Imatinib and Sunitinib**.

Carcinoid Tumors

- Develop from enterochromaffin (neuroendocrine) cells.

- Secrete various humoral factors, primarily serotonin, substance P, and histamine.

- Carcinoid tumors can be found in organs derived from the foregut, midgut, and hindgut.

- In the gastrointestinal system, 90% are found in three locations:

  - Appendix (45%) – the most common site.

  - Ileum (28%).

  - Rectum (16%).

According to the 20th edition of Sabiston:

- Appendix (38%).

- Small intestine (29%).

- Colon (13%).

- Stomach (12%).

- Rectum (8%).

- They are rarely found in the esophagus and pancreas.

- Small intestine carcinoids are often multiple in 30% of cases, while appendix carcinoids are usually solitary.

- Another feature of carcinoids is the frequent presence of a second primary neoplasm of a different histological type, often seen synchronously. Typically, this is colorectal adenocarcinoma.

- Macroscopically, carcinoids appear as slightly raised, round, firm nodules covered with normal mucosa.

- Intense desmoplastic reaction can lead to excessive fibrosis in the mesentery and bowel wall. This fibrosis can cause bowel wall obstruction.

- Abdominal pain, obstruction, diarrhea, and weight loss are the most common symptoms.

- Radiological studies often reveal numerous filling defects, some due to the tumor and others due to fibrosis.

- Mesenteric vascular angiography shows slow venous drainage in the tumor area.

- Somatostatin receptor scintigraphy is very effective in localizing carcinoid tumors.

- Chromogranin A is a non-specific neuroendocrine tumor marker.

Treatment:

- Tumor diameter < 1 cm: Segmental resection is performed.

- Tumor diameter > 1 cm, large or multiple lesions, or lymph node metastasis: Wide resection is performed.

- In the presence of metastatic disease, debulking resections are done; these provide both long-term survival benefits and reduce symptoms of the carcinoid syndrome.

Carcinoid Syndrome

- Seen in 5-10% of patients with malignant carcinoid tumors.

- For a carcinoid tumor to cause carcinoid syndrome, the serotonin it secretes must reach the systemic venous circulation without passing through the liver.

- Most patients have massive liver metastases, but not every patient with liver metastasis shows carcinoid syndrome.

- Carcinoid tumors that bypass the liver, such as those in the ovaries and retroperitoneum, can also cause carcinoid syndrome without liver metastasis.

- Carcinoid syndrome is most commonly associated with jejunoileum carcinoid tumors.

- Typically, 5-9% of malignant carcinoid tumors lead to carcinoid syndrome symptoms.

- Symptoms include episodic flushing, bronchospasm, diarrhea, and vasomotor collapse attacks.

CLINIC

- Hepatomegaly, diarrhea, and flushing: 80%

- Right heart valve diseases: 50% (most commonly pulmonary stenosis; in internal medicine sources, tricuspid insufficiency is most common)

- Asthma: 25%

- Malabsorption and pellagra (dementia, dermatitis, diarrhea) may also be observed.

- Heart valve disease results from endocardial fibrosis.

- Fibrosis, related to high levels of serotonin, is seen in the right heart endocardium, as well as in the right intestinal wall, retroperitoneum, and around mesenteric blood vessels.

- The left heart is protected because serotonin is metabolized in the lungs.

- Asthma symptoms are observed due to bronchoconstriction caused by serotonin, substance P, and bradykinin.

Diagnosis and Management of Carcinoid Syndrome:

- Basis of Diagnosis: Relies on the examination of various humoral factors.

- High levels of serotonin and substance P in circulation.

- 5-HIAA Measurement: The level of 5-HIAA in urine can be measured. It has high specificity but low sensitivity. Circulating serotonin is metabolized in the liver to 5-hydroxyindoleacetic acid (5-HIAA), which is then inactivated

- Chromogranin A: Can be used with high specificity but low sensitivity (95% specificity and 55% sensitivity).

- Using both Chromogranin A and urinary 5-HIAA together improves sensitivity.

- Studies suggest that Chromogranin A (CgA) and N-terminal pro-brain natriuretic peptide (NT-pro BNP) may be used together.

- MRI is not used for the diagnosis of carcinoid tumors but is useful for diagnosing metastatic disease, especially liver metastases.

- Treatment Options:

- Palliative Surgery: Liver resection can be performed for palliation.

- Hepatic arterial embolization, along with chemotherapy, can be applied, if Surgery is not possible.

- **Octreotide**: Effective in palliation of symptoms related to carcinoid syndrome.

Meckel's Diverticulum

- It is the most common congenital anomaly of the digestive tract, occurring in about 2% of the population.

- Development: It results from the failure of closure of the omphalomesenteric (or vitelline) duct, which normally connects the midgut to the yolk sac during intrauterine development.

- Failure of closure of this duct can lead to various anomalies, including omphalomesenteric cysts, residual fibrous bands containing enteric tissue, umbilical sinuses, or polyps.

- The most frequently observed anomaly is Meckel's diverticulum.

- It can occur alongside other congenital malformations such as esophageal atresia, anorectal atresia, and malformations of the central nervous system and cardiovascular system.

-It may contain ectopic mucosa in about 60% of cases.

Complications:

- Complications include bleeding, intestinal obstruction, inflammation (diverticulitis), perforation, Littre's hernia, and neoplasms.

- Rarely, it can also lead to malabsorption due to its blind loop effect.

- The most common symptom in children is rectal bleeding.

- In adults, intestinal obstruction, bleeding, and inflammation are more frequently observed.

- Most symptoms are seen in children under 2 years of age.

Bleeding:

- In children under 2 years of age, Meckel's diverticulum is the most common cause of severe lower gastrointestinal bleeding.

- The bleeding is usually caused by heterotopic mucosa within the diverticulum; 80-85% of the time, this is gastric mucosa. Other types of mucosa, such as pancreatic, duodenal, or colonic, may also be present.

Obstruction:

- It is the most common complication in adults.

Inflammation:

- The clinical picture is very similar to acute appendicitis.

- It can lead to localized abscesses or generalized peritonitis.

- In patients who are operated on with a diagnosis of acute appendicitis, if the appendix is found to be normal, Meckel's diverticulum should be investigated.

- If diverticulitis is identified, an appendectomy may also be appropriate.

Perforation:

- The diverticulum may perforate due to deepening of a peptic ulcer, progression of inflammation, or erosion caused by a foreign body.

Neoplasm:

- Rarely occurs (0.5-3.2%).

- The most common neoplasm is a carcinoid tumor.

Diagnosis:

- It can be incidentally discovered through a barium study.

- For uncomplicated Meckel's diverticulum, enteroclysis can be helpful.

- Technetium-99m pertechnetate scintigraphy is used to visualize ectopic gastric mucosa (reliable in children >90%).

Treatment:

- Complicated Meckel's diverticulum must be is excised.

- If the indication for surgery is bleeding or neoplasm, the diverticulum is resected together with the ileum segment from which it originates.

Small Intestine Perforations

- Iatrogenic perforations are the most common.

- The most frequent injury encountered during ERCP is duodenal injury.

- CT is the gold standard diagnostic method.

- Other causes of perforations include infections (typhoid, tuberculosis), Crohn's disease, ischemia, medications (such as NSAIDs), and radiation.

Short Bowel Syndrome

- It is a clinical condition characterized by insufficient intestinal length for adequate nutrition.

- Definition: Small intestine < 200 cm is considered short bowel syndrome.

- Digestion and absorption functions are lost.

- Electrolytes, carbohydrates, and amino acids are absorbed through active transport throughout the entire small intestine.

- Most nutrient absorption is completed in the proximal small intestine.

- Iron, folate, and calcium are especially absorbed from the proximal small intestine, while bile salts and vitamin B12 are absorbed from the terminal ileum.

- Resection of more than 50% of the small intestine is incompatible with life.

- **The most common cause is mesenteric vascular occlusions**.

- In newborns, the most common cause is necrotizing enterocolitis (NEC).

- Intestinal content is maintained isotonic in the jejunum, while it becomes increasingly concentrated in the ileum.

- In the colon, especially water and salt absorption occurs.

- After jejunal resection, the ileum and colon try to absorb increased fluids and electrolytes to minimize diarrhea.

- However, after ileal resection, the passage of isotonic content from the jejunum and unabsorbed bile salts to the colon causes severe diarrhea.

- The cause of this diarrhea is that bile salts damage the membrane structure of the colon mucosal cells, inhibiting active Na transport.

- Resection of more than 1 meter of the ileum impairs bile salt absorption.

- Fat absorption is also impaired, leading to steatorrhea.

- Along with small bowel resection, gastric acid hypersecretion occurs.

- This is likely due to the inhibition or elimination of a factor secreted from the small intestine that inhibits gastric acid secretion.

- Gastric hypersecretion is temporary. The increase in gastric secretions exacerbates diarrhea by increasing the fluid-electrolyte load of the intestines.

- In short bowel syndrome, bile stone formation increases threefold due to the disruption of the enterohepatic circulation of bile salts.

Clinical Stages:

1. Stage

- This is the period immediately after surgery.

- Fluid losses are at their highest levels.

- Fluid and electrolyte imbalance is the most significant cause of mortality.

- It lasts on average 3 weeks.

- If the remaining bowel length is < 100 cm without the colon or < 60 cm with the colon, lifelong TPN is required.

2. Stage

- Diarrhea decreases. The patient is comfortable as long as no oral fluids and food are taken.

- Diarrhea occurs with meals.

- Intestinal adaptation begins in this stage.

- It lasts on average 3-6 months.

3. Stage

- Numerous nutritional and metabolic disorders occur, proportional to the length of the bowel removed.

- Anemia is common.

- 30% of patients have osteomalacia.

- Hyperoxaluria (due to the increased absorption of oxalate in the colon after unabsorbed fatty acids bind with calcium, freeing oxalate), hypomagnesemia, gallbladder, and urinary system stones are observed.

Intestinal Adaptation

- Defined as the adaptive changes that occur in the remaining intestines after resection.

- The severity of the adaptive response is directly proportional to the length of the intestine removed.

- The ileum has a greater capacity for adaptive changes than the jejunum.

- Adaptive changes begin 48 hours after surgical intervention and can take months to complete.

- The basis of adaptation is the hyperplasia of enterocytes, which increases the absorptive surface.

- Intestinal villi lengthen without numerical increase.

- The number of cells and the rate of cell renewal increase.

- The length and diameter of the intestines increase.

Treatment

Phase 1:

- Fluid-electrolyte therapy is the most important treatment.

- TPN (Total Parenteral Nutrition) is administered.

Phase 2:

- As intestinal adaptation partially occurs, oral foods can begin.

- TPN support continues.

- Cholestyramine may be beneficial for choleretic diarrhea caused by bile salts passing into the colon.

- If the resected ileum is more than 1 meter, bile salt loss is more pronounced, and cholestyramine is not beneficial.

- Medium-chain triglycerides, which do not require micelle formation for absorption, are given.

- Elemental diets are used for patients who cannot tolerate a normal diet.

- Enteral nutrition should start with carbohydrates, followed by proteins and fats.

- Small intestine transplantation is only surgical treatment.

Phase 3:

- After 3-12 months, patients enter Phase 3.

- In addition to a normal diet, supplements of iron, vitamin B12, folate, magnesium, and vitamin D are given.

Chapter

3.

Rectum and Colon Diseases and Surgery

Anatomy of the Colon

- The average length of the colon is between 90-150 cm. It extends from the ileocecal valve to the rectum.

- The cecum is 6-7 cm, the ascending colon is 12.5-20 cm, the transverse colon is 40-50 cm, the descending colon is 25-30 cm, the sigmoid colon is 40 cm, the rectum is 12-15 cm, and the anal canal is 4 cm long.

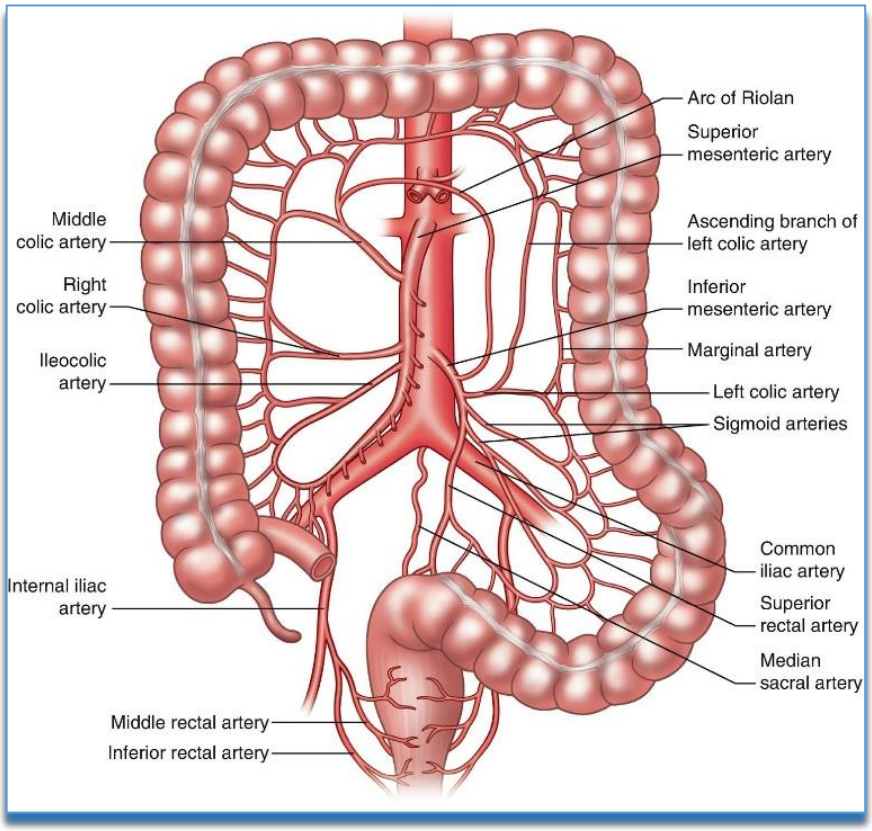
- The ascending colon, descending colon, hepatic flexure, and splenic flexure are retroperitoneal.

- The cecum, transverse colon, and sigmoid colon are intraperitoneal.

- The wall of the colon consists of the mucosa, submucosa, inner circular muscle layer, outer longitudinal muscle layer, and serosa from the inside out.

- The mucosa of the intestine shows a glandular structure.

- After the pectinate line, the mucosa transitions from a single layer of columnar cells to cuboidal, and then to stratified squamous epithelium (modified skin).

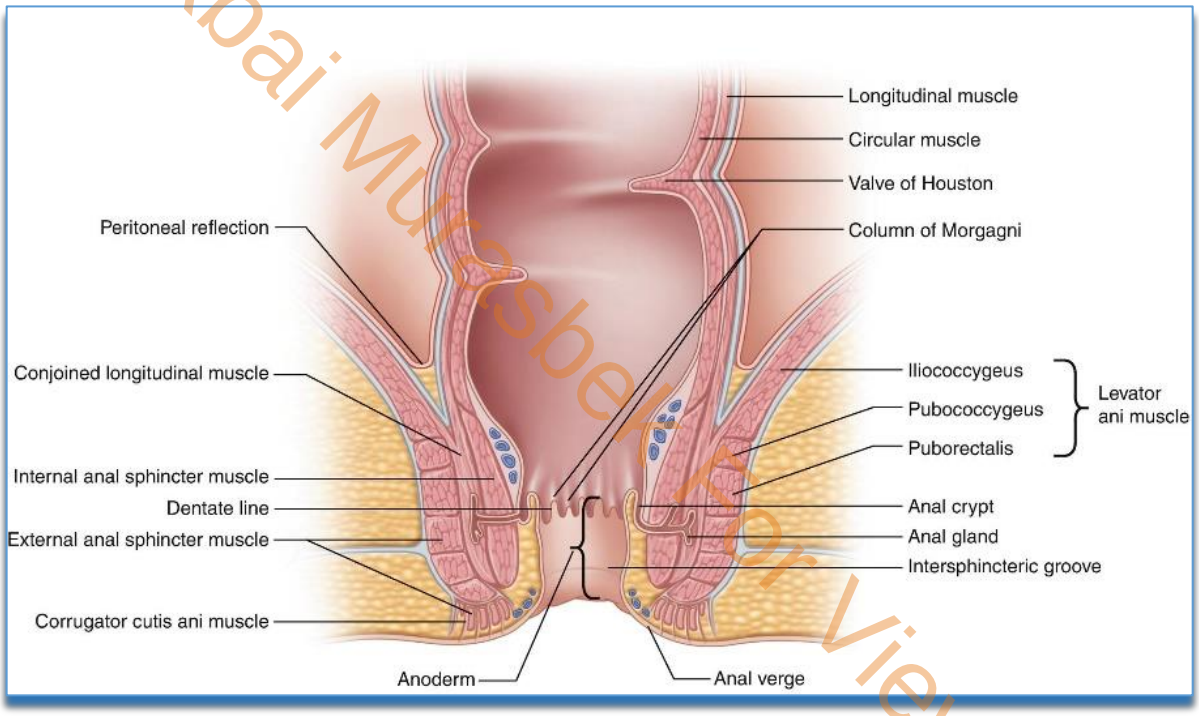


- Arterial supplying occurs with a.Mesenterica superior and a.Mesenterica inferior, all veins, except for the inferior mesenteric vein, follow the paths of the arteries.

-It has sympathetic and parasympathetic innervation.Peristalsis is inhibited by the sympathetics and stimulated by the parasympathetics. Parasympathetic stimulation up to the distal 1/3 of the transverse colon comes

from the celiac branch of the right vagus, while the distal part is innervated by the sacral plexus."

Anatomy of rectum



- It is generally considered a separate structure from the colon.

- The rectal wall consists of the mucosa, submucosa, inner circular, and outer longitudinal muscle layers. It is 12-15 cm in length.

- Posteriorly, the presacral fascia separates the rectum from the presacral venous plexus and pelvic nerves. At the level of S4, the rectosacral fascia (Waldeyer's fascia) extends forward and downward to attach to the fascia propria at the anorectal junction. Anteriorly, Denonvilliers' fascia separates the rectum from the prostate and seminal vesicles in males and from the vagina in females. Lateral ligaments support the lower rectum.

- The arteries include the superior rectal artery from the inferior mesenteric artery for the upper 1/3 of the rectum, the middle rectal artery from the internal iliac artery for the middle 1/3, and the inferior rectal artery from the internal pudendal artery for the lower 1/3.

- Venous drainage follows the paths of the arteries and occurs into both the caval and portal systems.

Normal Colon Function

- The colon absorbs water, sodium, and chloride, and secretes potassium, bicarbonate, and mucus.

- Approximately 90% of ileal fluid is absorbed by the colon.

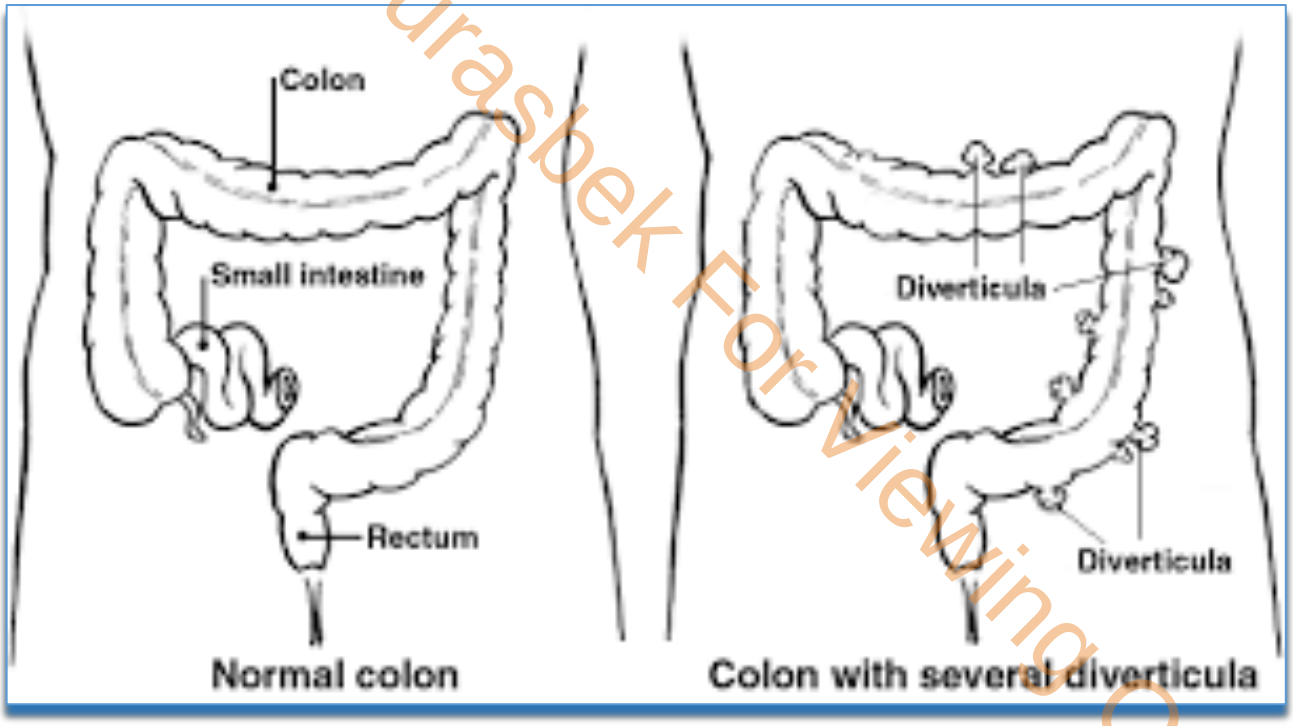
- The colon provides an environment for the formation of vitamin K by bacteria.

- Ammonia and short-chain fatty acids are absorbed from the colon.

- Short-chain fatty acids (acetate, butyrate, propionate) are an important energy source for the colon mucosa.

- The most common bacteria in the flora are B. fragilis and E. coli.

Diverticulosis



- There are two types: acquired and congenital.

- Acquired diverticulosis is usually in the left colon. It consists of multiple false diverticula.

- Congenital diverticula are single and true, located in the right colon.

- In false diverticula, the diverticular wall lacks a muscular layer and only contains mucosa and serosa.

- True diverticula include all three layers of the intestinal wall.

- Colonic diverticulosis is very common in the USA and Europe.

- It is believed that almost half of the population over the age of fifty has diverticula.

- Despite the high prevalence of colonic diverticulosis, complications are rare and patients are usually asymptomatic.

- Constipation and a low-fiber diet are implicated in its etiology. A fiber-poor diet leads to small amounts of feces, resulting in increased intraluminal pressure to move the feces forward. This leads to muscular hypertrophy of the colon wall.

- Increased intraluminal pressure causes diverticula to herniate, usually at weak points where blood vessels enter the intestinal wall, pushing the mucosa outward.

- Therefore, diverticula are located on the mesenteric side of the antimesenteric tenia.

- They most commonly develop in the sigmoid colon.

- Colonic diverticulosis is generally asymptomatic.

- The treatment for colonic diverticulosis is typically medical.

Complications:

- **Bleeding (15%)**

- It is the most common cause of massive lower gastrointestinal (GI) bleeding in adults.

- Bleeding typically occurs from the left colon.

- In cases of massive bleeding, colonoscopy may not reveal the source.

- Angiography can be helpful.

- 80% of the bleeding episodes stop spontaneously; however, if the bleeding does not stop, surgical intervention is required.

- **Diverticulitis**

- Occurs in 10-25% of patients with diverticulosis.

- Fecal impaction at the opening of a diverticulum leads to the accumulation of mucus secretions and bacterial proliferation within the diverticulum.

- If the obstruction persists, inflammation develops.

- The infection spreads to the wall of the diverticulum and surrounding areas.

- Eventually, this can result in fistulas, contained perforation, and abscess formation.

- Clinical findings: dull and persistent pain in the lower left quadrant, low-grade fever, loss of appetite, nausea, leukocytosis, diarrhea, constipation.

- The most reliable diagnostic method is CT. CT is performed initially.

- It is also useful in distinguishing between inflammation and abscess.

- During the acute phase, barium studies and colonoscopy are contraindicated.

- Preventing complications is necessary, so patients are advised to follow a high-fiber diet and use laxatives.

- Hinchey staging: Hinchey I- localized abscess(para-colonic)

Hinchey II- pelvic abscess

Hinchey III- purulent peritonitis(pus in the abdominal cavity)

Hinchey IV- feculent peritonitis

Treatment.

- is primarily medical .Most patients with uncomplicated diverticulitis are treated as outpatients with broad-spectrum oral antibiotics and a low-fiber diet.

- Approximately 10-20% of patients with more severe pain, tenderness, fever, and leukocytosis are started on IV analgesics; oral intake is discontinued; nasogastric decompression is performed; and IV fluid-electrolyte therapy is administered. Broad-spectrum parenteral antibiotics are started.

- Improvement is generally seen within 48-72 hours. If the patient's condition does not improve, surgery is performed.

- The current approach does not recommend routine surgery after recurrent uncomplicated diverticulitis attacks and uncomplicated diverticulitis attacks in young patients, once cancer is ruled out.

- In immunosuppressed patients, colectomy is still recommended after the first attack.

- Under elective conditions, sigmoid colectomy with primary anastomosis is preferred.

- Diverticulitis can also lead to complications such as abscess, obstruction, generalized peritonitis (free perforation), or fistula formation between the colon and adjacent structures.

- In emergency conditions, the most commonly performed procedure in stable patients is the Hartmann procedure (segmental resection + proximal end colostomy + closure of the rectal stump).

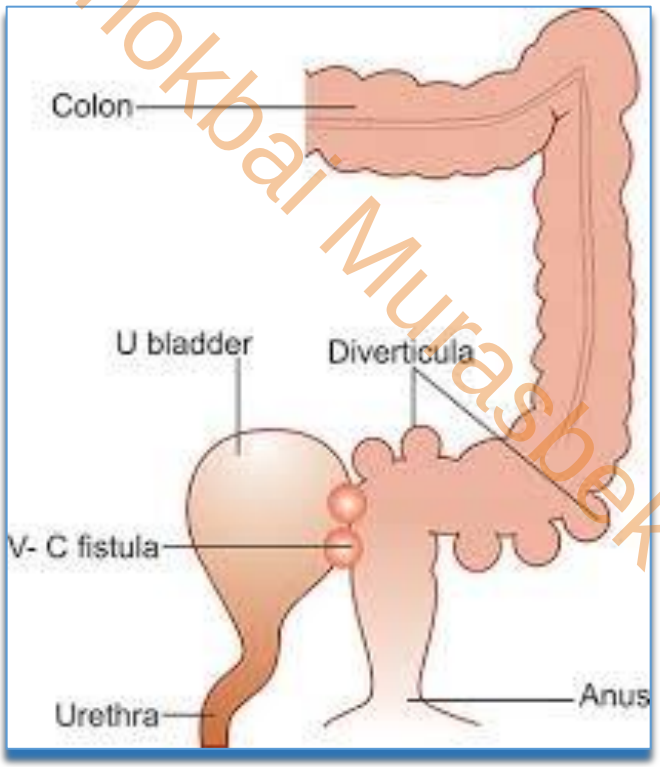
- In unstable patients or those with severe inflammation, proximal diversion and local drainage are performed.

- Additionally, obstruction and fistula can develop due to diverticulitis.

- Diverticulitis and colon cancer can be confused.

- To rule out colon cancer, a colonoscopy should be performed 4-6 weeks after the acute attack.

Colovesical Fistula



- A colovesical fistula develops between the colon and an adjacent organ in 5% of patients with complicated diverticulitis.

- It most commonly occurs between the colon and the bladder (colovesical). Less commonly, it occurs between the colon and the vagina (colovaginal) or the colon and the intestine (coloenteric).

- Other causes of colovesical fistula include sigmoid colon cancer and Crohn's disease.

- In a patient presenting with fecaluria and pneumaturia, if a tumor or Crohn's disease is not present, a diverticulitis-related fistula should be considered.

- There is often a history of previous diverticulitis attacks.

- Diagnosis is aided by barium colonography, CT, and cystoscopy. Elective surgical excision is the treatment.

Right-Sided Diverticula

- Diverticulosis of the colon is rarely seen in the cecum and ascending colon.

- More commonly, a single and true diverticulum is observed.

- It is considered to be congenital.

- It occurs more frequently in younger individuals compared to left-sided colon diverticula.

- It is more common in asymptomatic cases.

- Sometimes diverticulitis can develop.

- In young patients, it can be confused with appendicitis due to causing pain in the right lower quadrant.

- Diagnosis is usually made during surgery.

- Ileocecal resection is generally preferred over diverticulectomy.

- Bleeding is less common in right-sided diverticula.

VOLVULUS

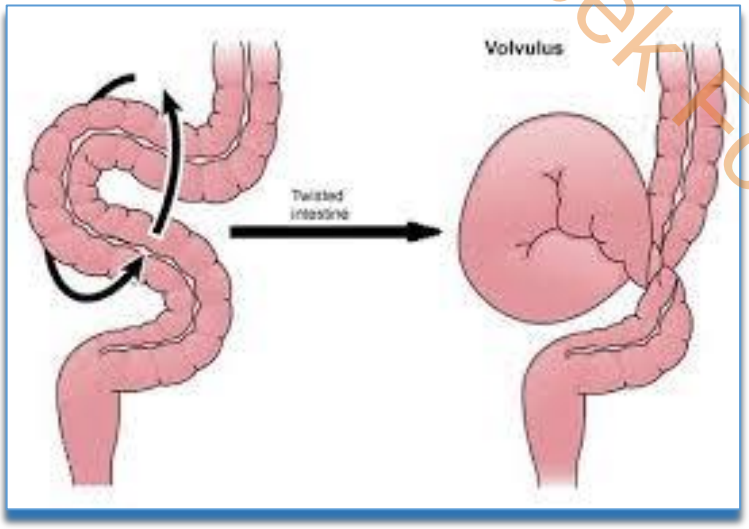
- It is the rotation of an air-filled bowel segment around its own mesentery.

- The most commonly affected areas are the sigmoid colon, cecum (less than 20%), and transverse colon.

- It can also occur in the stomach and small intestine.

- Volvulus may resolve spontaneously; however, it more commonly leads to bowel obstruction, and can progress to strangulation, gangrene, and perforation.

Sigmoid Volvulus



- Sigmoid volvulus accounts for approximately 90% of all volvulus cases.

- Patients present with crampy abdominal pain, asymmetric distension, and obstipation (inability to pass gas or stool).

- Most patients have a history of previous episodes of abdominal pain and distension.

- On abdominal X-ray, a "reversed U," omega sign, or coffee bean appearance is typical.

- In cases where diagnosis cannot be made with X-rays, barium enemas may be helpful, but should not be performed if there is a suspicion of gangrene.

- Barium studies show a progressively descending column of barium that ends at a point of obstruction. This appearance is known as "bird-beak deformity."

- If there is no strangulation, decompression with rigid sigmoidoscopy is performed following fluid resuscitation.

- If strangulation is suspected or if signs of peritoneal irritation are present, reduction should not be attempted, and the patient should proceed directly to surgery.

- Failure to achieve detorsion, presence of bloody discharge, or signs of mucosal ischemia indicate that strangulation or gangrene has developed. In such cases, sigmoidoscopy should be terminated and the patient should be taken for urgent surgery.

- Typically, a Hartmann procedure is performed.

Arteriovenous Malformations (AVMs)

- Other names: AVM, angiodysplasia, vascular dysplasia, vascular ectasia.

- AVM is the most common cause of chronic, recurrent lower gastrointestinal bleeding when the cause cannot be determined by known diagnostic methods.

- It occurs due to progressive dilation of submucosal veins in any part of the gastrointestinal tract.

- AVMs can be associated with aortic stenosis and renal failure.

- Most patients are over 50 years old, with an equal gender distribution.

- Most patients experience chronic bleeding; massive bleeding occurs in about 15% of cases.

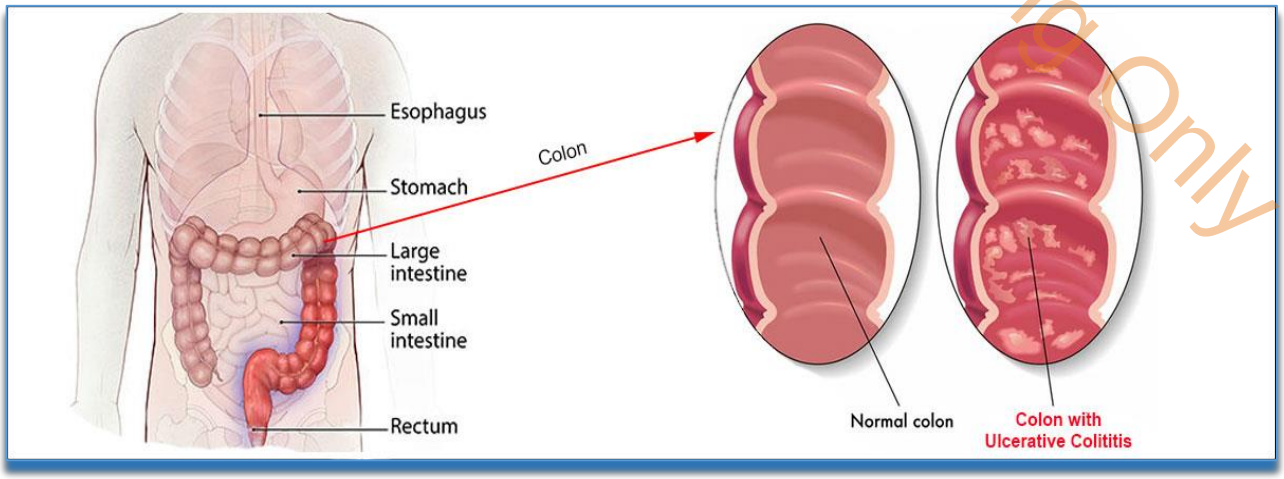
- Colonoscopy and angiography are the most useful diagnostic methods.

- The most common site is the colon, particularly the cecum.

- Diagnosis is made through colonoscopy or angiography.

Ulcerative Colitis

- An inflammatory disease affecting the colon mucosa and submucosa.



- More common in males.

- Forms of the Disease:

  - Rectal

  - Rectosigmoid

  - Left-sided colonic

  - Pancolitis

Etiology:

- Certain specific microorganisms (e.g., C. difficile, Campylobacter jejuni)

- Hypersensitivity and mucosal immune system abnormalities: In ulcerative colitis, there are particularly antibodies against certain antigens.

- Psychosomatic disorders: Emotional stress may exacerbate the disease.

- Family history: Associations with HLA and DR2 (HLA-DRB1).

- The risk of developing ulcerative colitis has been reduced in individuals who have **undergone an appendectomy**.

Pathology of Ulcerative Colitis

- Ulcerative colitis affects only the mucosa and submucosa.

- The earliest endoscopic finding is mucosal edema.

- **Typical Appearance**: Hyperemic mucosal appearance is characteristic.

- **Crypt Abscesses**: Observed in pathological examination, these are specific to the disease.

- **Ulceration**: Ulcers are present in the mucosa, covered with pus.

- **Advanced Cases**: The bowel length may be shortened due to fibrosis.

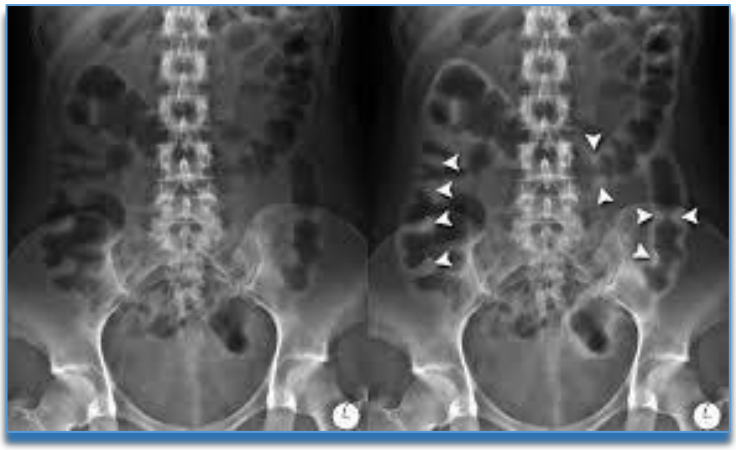
- **Stenosis**: Can also occur.

- **Sigmoid Colon**: The "S" shape curvature of the sigmoid colon is lost.

- **Disease Distribution**: The disease starts in the rectum, may remain localized to the rectum, or spread proximally.

- **Extent of Involvement**: In 30% of cases, the entire colon is affected.

- **Disease Pattern**: The disease is not segmental.



- **Terminal Ileum Involvement**: Rare (seen in 10% of cases), referred to as "backwash ileitis."

- **Pseudo-polyps**: The mucosa between ulcers can form a polypoid structure, known as pseudopolyps.

- **Barium Colonography**: Pseudopolyps give a "cobblestone" appearance on barium colonography (more pronounced in Crohn's disease).

- **Fibrosis**: Leads to the loss of haustral patterns, giving a "lead pipe" appearance on colonography.

- **Stricture Formation**: In ulcerative colitis, inflammation is confined to the mucosa, so stricture formation is very rare. If a stricture is detected, malignancy should be considered until proven otherwise.

- **Malignancy Concerns**: Strictures that raise suspicion of malignancy are typically those seen in the later stages of the disease (more than 20 years, with a 60% incidence), those located proximal to the splenic flexure, and those causing obstruction in the large intestine.

CLINIC

- Acute Fulminant

  - Seen in only 5-10% of patients.

  - Onset is sudden.

  - Severe abdominal cramps, diarrhea (30-40 times a day, always purulent and mucus-filled).

  - Fecal incontinence, tenesmus.

  - Fever and systemic toxicity are present.

  - Electrolyte loss due to profuse mucus diarrhea; hypokalemia may occur.

  - Hypoproteinemia and anemia may develop.

  - Rapid progression to toxic megacolon.

- Chronic Continuous

  - Chronic continuous forms are more common.

  - Risk of developing toxic megacolon is lower in chronic type.

- Chronic Attacking

  - Symptoms are typical; diarrhea and bloody diarrhea are the most frequent.

  - The most common finding of active disease is bloody diarrhea or hematochezia.

  - Tenesmus may be observed. The absence of tenesmus can be useful for differential diagnosis compared to Crohn's disease.

  - Paradoxically, rectal involvement is seen in 100% of patients, while anal involvement is rare.

Systemic Findings

- Liver Involvement (40-50%)

    - The liver is the most affected organ in extra-colonic manifestations.

    - Steatosis is common; cirrhosis is rare. If cirrhosis develops, it does not improve after treatment.

- Sclerosing Cholangitis

    - Ulcerative colitis is present in 40-60% of patients with primary sclerosing cholangitis.

    - Does not improve after colectomy. Cholangiocarcinoma is a rare complication seen in patients with long-standing inflammatory bowel disease.

- Arthritis

    - Commonly seen. Resolves after colectomy. Can include ankylosing spondylitis and sacroiliitis.

- Erythema Nodosum (5-15%) and Pyoderma Gangrenosum

    - Erythema nodosum is parallel to the severity of the disease.

- Ocular Lesions (10%)

    - Includes iritis, uveitis, and episcleritis. These appear during acute flare-ups of inflammatory bowel disease.

- All of these systemic findings arise due to hypersensitivity and generally resolve spontaneously after surgical treatment, with exceptions including primary sclerosing cholangitis, liver cirrhosis, and erythema nodosum.

Complications of Ulcerative Colitis

- Bleeding - Most common.

- Obstruction(strictures and stenoses)

- Perforation

- Pericolic Abscess

- Perianal Fistulas

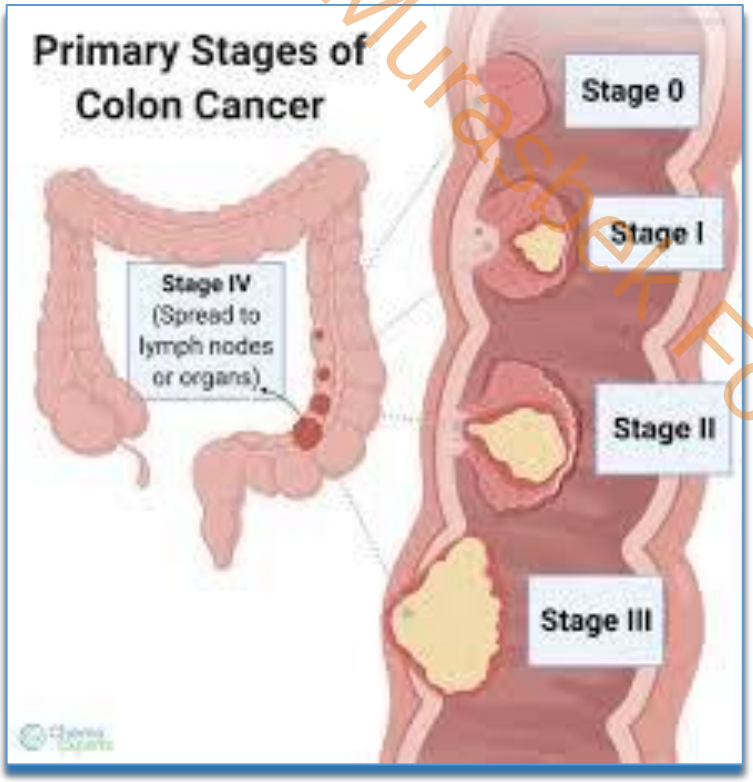
- Malabsorption

Colon Cancer

- Colon cancer is the most significant late complication of ulcerative colitis, occurring in about 3% of patients.

- It is more infiltrative and less differentiated compared to sporadic colon cancer.

- It develops from multiple foci and flat dysplastic areas.



- Early diagnosis is challenging.

- Patients with dysplasia detected in biopsy should be considered for prophylactic proctocolectomy, ileal pouch, ileoanal anastomosis, and loop ileostomy.

- The prognosis is generally worse.

- The incidence of cancer is higher in patients who develop ulcerative colitis at an early age.

- The cancer risk is approximately 2% after 10 years, 8% after 20 years, and 18% after 30 years.

- The 5-year survival rate for colon cancer secondary to ulcerative colitis is 20%.

- Perianal disease or abdominal mass and weight loss are very rare compared to Crohn's disease.

- Diagnosis is made using clinical evaluation, barium enema, sigmoidoscopy, colonoscopy, and biopsy.

- Barium enema typically shows a "cobblestone" appearance, "lead pipe" appearance, loss of haustral markings (not seen in Crohn's disease), a short and rigid colon, and abnormal mucosal patterns.

- Sigmoidoscopy, colonoscopy, and biopsy are used for definitive diagnosis.

- In differential diagnosis, antibody positivity is also utilized. The most commonly used tests are perinuclear antineutrophil cytoplasmic antibodies (pANCA) and anti-Saccharomyces cerevisiae antibodies (ASCA).

- pANCA -/ASCA+ supports a diagnosis of Crohn's disease, while pANCA+/ASCA- supports ulcerative colitis.

- Differential diagnoses to consider include amebiasis, bacillary dysentery, tuberculosis, diverticulosis, lymphogranuloma venereum, colon cancer, celiac disease, non-tropical sprue, and malabsorption syndrome.

Treatment

- Initial treatment is primarily medical, especially for acute cases. Chronic cases often do not respond well to medical treatment alone.

- Nutritional care is crucial in medical treatment. For patients with severe malnutrition or those on steroids, creating a stoma during surgery is generally safer than performing a primary anastomosis.

- Complete physical and emotional rest is essential.

- Various medications are used, including sulfasalazine, steroids, azathioprine, S-ASA, and Imuran.

- **Sulfasalazine and S-ASA** are the first-line medications in the initial stages of treatment.

- Steroid + ASA enemas are administered in acute cases.

- Steroids are started at high doses in acute cases, then tapered and discontinued; they are generally not used for maintenance.

- **Metronidazole** is beneficial in cases of active colitis and diarrhea.

- Antibiotics are not used unless there is fulminant colitis or toxic megacolon.

- **Azathioprine and 6-mercaptopurine** are useful in cases that do not respond to salicylate therapy or are steroid-dependent or resistant.

- **Tacrolimus and cyclosporine** are used as second-line immunomodulatory drugs for steroid-resistant cases.

- TNF-alpha inhibitors*such as **infliximab and adalimumab** can be used for steroid-resistant ulcerative colitis.

- **Golimumab and certolizumab** are other TNF-alpha inhibitors.

- If the disease is limited to the rectum, medical treatment can be very effective.

- In other cases, the response to medical treatment is often lower.

Surgical Treatment

Elective Surgery Indications:

- Resistance to medical treatment (most common) or development of side effects from medications (e.g., aseptic necrosis due to steroid use).

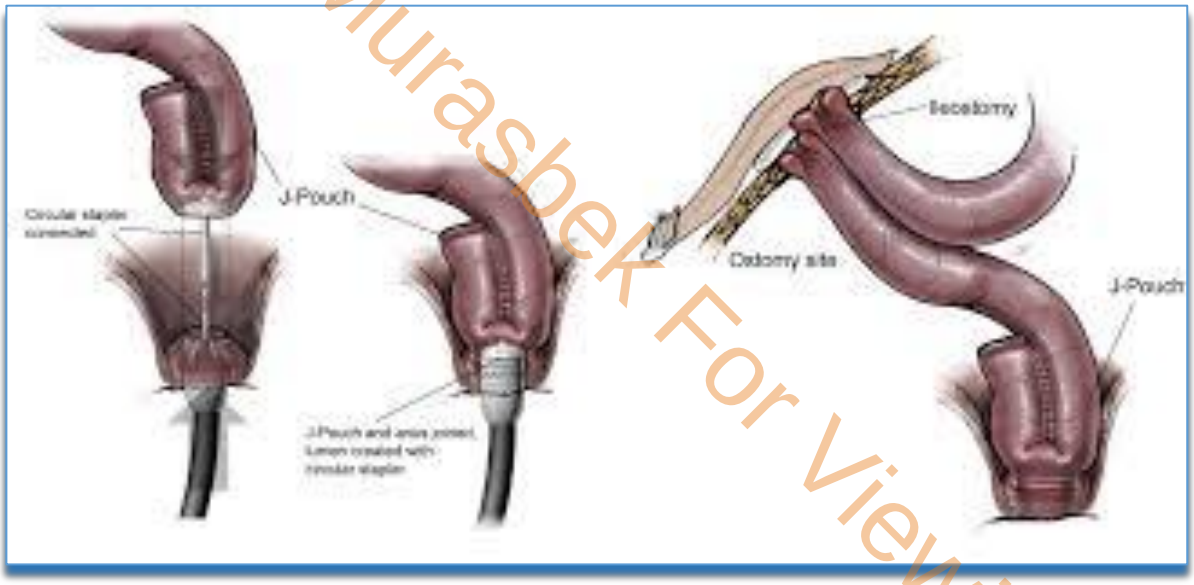
- Cancer prophylaxis.

- Severe extracolonic symptoms resistant to non-surgical measures.

Emergency Surgery Indications:

- Life-threatening massive bleeding.

- Toxic megacolon or fulminant colitis unresponsive to medication.



Surgical Treatment:

- Restorative proctocolectomy is performed. This involves total proctocolectomy + rectal mucosectomy + ileal pouch + ileal pouch-anal anastomosis. A protective loop ileostomy is typically created.

- In emergency surgeries, total colectomy + ileostomy are performed. Anastomosis is not done and is postponed to a later session.

Pseudomembranous Colitis

- **Definition:** A syndrome characterized by acute diarrhea occurring in patients who are currently receiving or have recently taken antibiotics.

- **Cause:** Antibiotics alter the bacterial population in the colon, leading to an overgrowth of Clostridium difficile.

- **Incidence:** Approximately 25% of cases develop diarrhea after the cessation of antibiotic use. Symptoms can appear up to 6 weeks after stopping antibiotics.

- Triggering Antibiotics: Can develop after the use of any antibiotic, most commonly following the use of clindamycin, ampicillin, and cephalosporins.

- **Clinical Spectrum:** Ranges from mild, self-limiting diarrhea to severe invasive colitis that can lead to megacolon and perforation.

- **Management:** Antibiotic use should be discontinued immediately if this syndrome is suspected.

- **Diagnosis**: Proctoscopy or colonoscopy can be useful for detecting pseudomembranes. Testing for toxin A in stool is the most reliable diagnostic method.

- Mild Cases: Typically self-limiting.

- Severe Symptoms: Patients with severe symptoms such as abdominal pain, fever, and leukocytosis should be hospitalized for intravenous hydration and observation.

- **Treatment:**

  - Preferred antibiotic is metronidazole (3x250-500 mg for 10 days, either IV or oral).

  - Oral vancomycin is another treatment option.

  - Cholestyramine can be given as it binds and neutralizes the toxins.

Amebic Colitis

- **Definition:** Amebiasis is an infection primarily affecting the colon but can also secondarily involve other organs, most notably the liver, caused by Entamoeba histolytica.

- **Transmission:** Spread via the fecal-oral route.

- **Pathology:** The pathological lesions of invasive amebiasis are small ulcers. While these can be found throughout the colon, they are most commonly located in the cecum.

  - The ulcers are covered with yellow exudate.

  - As the disease progresses, ulcers can grow larger than 2 cm and rarely, they may perforate, leading to fulminant peritonitis.

  - Histological examination can reveal amoebae at the ulcer margins.

- **Clinical Presentation:** Ranges from asymptomatic carriers to acute illness with bloody diarrhea resembling fulminant ulcerative colitis.

  - It is essential to differentiate amebiasis from Crohn's disease and ulcerative colitis, as steroids, used in these conditions, are contraindicated in amebiasis.

- **Diagnosis:**

  - The most reliable tests are serological tests.

- The indirect hemagglutination test is positive for E. histolytica.

- Microscopic examination of fresh stool shows trophozoites in 90% of patients.

- Appropriate stool cultures should be taken to differentiate from infectious bacterial colitis.

- Treatment:

- For invasive amebiasis, the recommended treatment is metronidazole (3x750 mg for 10 days).

- Metronidazole may not be effective for carriers and amoebae within the intestinal lumen.

- To eliminate organisms in the lumen, iodoquinol (diiodohydroxyquin) is used.

- Ameboma is a rare complication of amebic colitis. It is an inflammatory mass in the colon that can cause narrowing and may be mistaken for colon carcinoma. It is most commonly found in the cecum.Colonoscopy is helpful in diagnosis. The initial treatment is with antibiotics, surgical treatment may be necessary

Ischemic Colitis

- Definition: Ischemic colitis occurs in elderly patients with atherosclerotic stenosis or inflammatory arteriopathies.

- Prevalence: It accounts for more than 50% of all gastrointestinal ischemic episodes.

- Pathophysiology:

- Unlike ischemia of the small intestine, ischemic colitis generally does not involve major arterial or venous occlusion.

- Colonic ischemia is due to reduced blood flow and/or occlusion of small vessels.

- Risk Factors: Vascular diseases, diabetes, vasculitis, and hypotension.

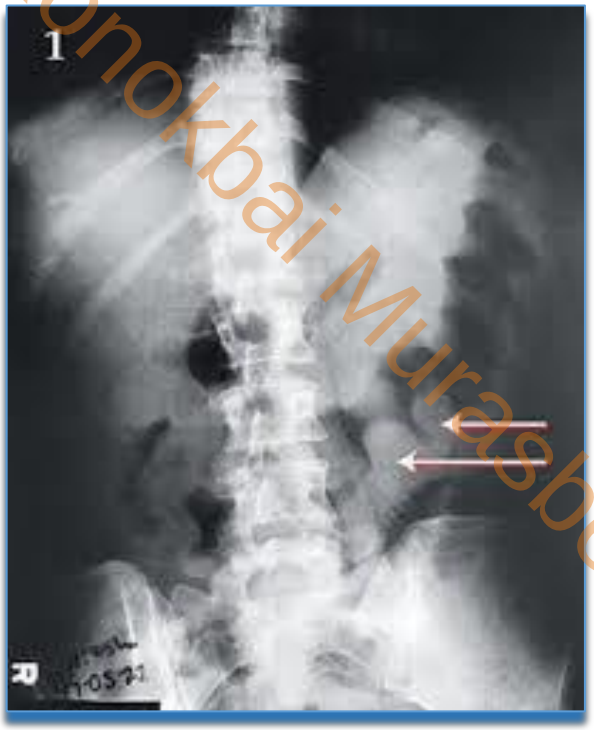
- Additionally, ligation of the inferior mesenteric artery during aortic surgery can lead to colonic ischemia.

- Common Sites: The splenic flexure is the most frequently affected area. The rectum is usually spared due to its rich collateral circulation.

- Clinical Presentation:

- Mild cases may present with painless bloody diarrhea.

- Severe ischemia may present with abdominal pain, tenderness, fever, and leukocytosis.



- Radiographic Findings:

- On plain radiographs, “thumbprinting” may be seen, indicating submucosal edema and hemorrhage. This finding can also appear on contrast studies, although contrast studies should not be performed in the acute phase.

- Sigmoidoscopy: Characteristically shows dark, hemorrhagic mucosa. However, there is a risk of perforation with this method.

- Angiography: Usually not helpful as major arterial occlusion is rare.

Treatment

- It depends on the severity of the ischemia and the symptoms.

- Unlike ischemia of the small intestine, the vast majority of ischemic colitis patients can be treated medically.

- For mild pain, minimal fever, and minimal leukocytosis, close monitoring is sufficient. The patient should improve in 2-3 days.

- If pain, fever, and leukocytosis are more pronounced, close monitoring, IV antibiotics, and bowel rest are recommended.

- The ischemic segment heals with stricture formation.

- Long-term sequelae include strictures (15-20%) and chronic segmental ischemia.

- In very few patients, the ischemia affects the entire thickness of the colon.

- In case of necrosis, resection and proximal colostomy should be performed.

Colon Neoplastic diseases (polyps and polyposis)

Polyps are epithelial protrusions seen in various organs .

- The gastrointestinal system is the most common site for polyps.

- The most frequent localization of gastrointestinal polyps is the colorectum.

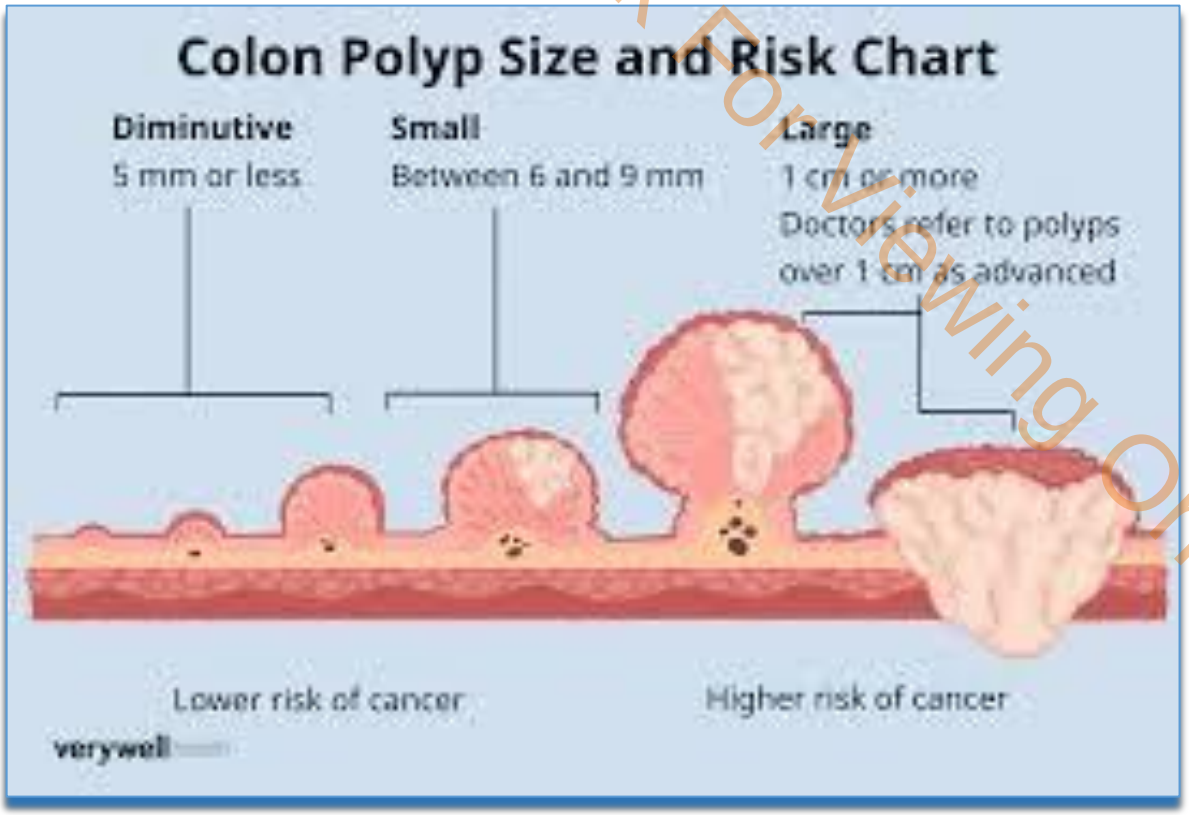
- Polyps are classified based on histological type, endoscopic appearance, and number.

- A **polyp** is a single growth or protrusion of epithelial tissue that develops on the lining of an organ, such as the colon or rectum. Polyps can vary in size and shape, and they can be benign (non-cancerous) or have the potential to become malignant (cancerous).

- **Polyposis** refers to a condition characterized by the presence of multiple polyps. It often implies a genetic predisposition or syndrome, such as familial adenomatous polyposis (FAP) or Peutz-Jeghers syndrome. Polyposis increases the risk of developing cancer due to the large number of polyps.

In summary, while a polyp is a single growth, polyposis is a condition involving multiple polyps.

Non-neoplastic Polyps



- **Hyperplastic polyps**

- The most common type of colorectal polyp. Ten times more frequent than adenomatous polyps.

- Typically small (< 5 mm).

- Composed of cells showing hyperplasia without maturation and dysplasia.

- Not considered premalignant lesions.

- However, because they cannot be distinguished from adenomatous polyps during colonoscopy, they are removed endoscopically.

- Large hyperplastic polyps (>2 cm) may have a slight risk of malignant degeneration.

- **Inflammatory polyps**

- Non-neoplastic epithelial formations.

- Most commonly associated with inflammatory bowel disease. They can also develop due to amebic colitis, ischemic colitis, and schistosomal colitis.

- Not premalignant. However, because they cannot be distinguished macroscopically from adenomatous polyps, they are removed during colonoscopy.

- **Hamartomatous polyps (Juvenile polyps)**

- Characteristic polyps of childhood, but can be seen at any age.

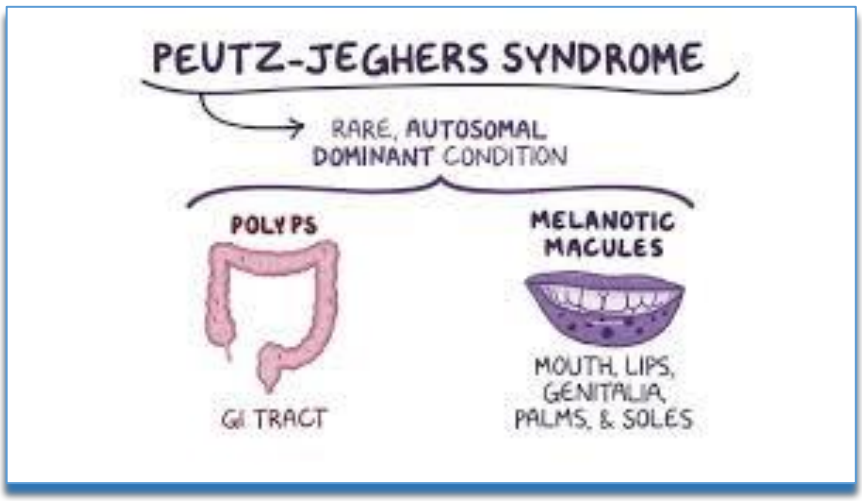
- Consist of glandular structures within a fibroblastic stroma of the lamina propria, with cystic dilations.

- Bleeding is a common symptom; intussusception and/or obstruction can also occur.

- Generally not premalignant.

- Since their macroscopic appearance is the same as adenomatous polyps, polypectomy is performed.

Hamartomatous Polyposis Syndromes



Peutz-Jeghers Syndrome

- It is inherited in an autosomal dominant manner.

- Characterized by polyposis in the small intestine and, to a lesser extent, the colon, along with melanotic spots on the oral and lip mucosa.

- Since the polyps are hamartomas, they are not considered to have a high risk of malignant degeneration. However, cancer can rarely develop.

- There is an increased risk throughout the entire gastrointestinal tract, from the stomach to the rectum.

- Polyps are most commonly found in the jejunum.

- Cancers of the small intestine, stomach, pancreas, ovary, uterus, and breast occur in 50-90% of patients.

- Patients present with abdominal pain and sometimes bleeding. Colicky abdominal pain is caused by intussusception.

- Screening begins with baseline colonoscopy and upper endoscopy at age 20, followed by annual flexible sigmoidoscopy and other system screenings.

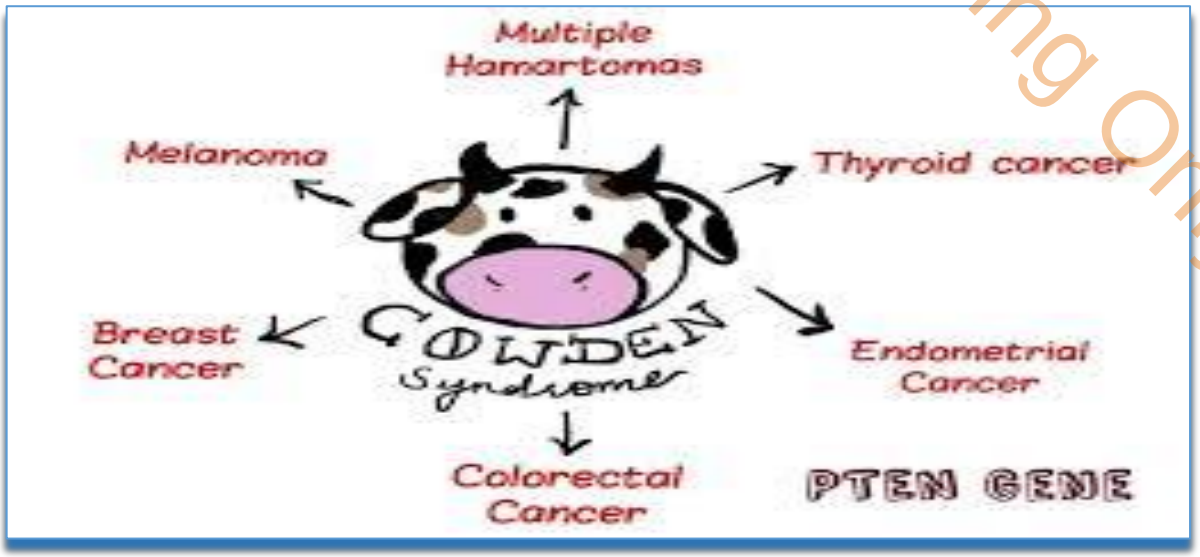
Familial Juvenile Polyposis

- Inherited in an autosomal dominant manner.

- Characterized by hundreds of polyps in the colon and rectum.

- Unlike solitary juvenile polyps, these polyps can degenerate into adenomas and eventually cancer.

- Annual screenings begin between ages 10-12. Due to the difficulty of colonoscopic follow-up, prophylactic colectomy is recommended.



Cowden Disease

- Very rare.

- Hamartomas from all three embryonic layers are observed. It is inherited in an autosomal dominant manner.

- Features include facial trichilemmomas, breast cancer, thyroid disease, and gastrointestinal hamartomatous polyps.

- Patients should be screened due to the cancer risk.

Cronkhite-Canada Syndrome

- Characterized by gastrointestinal polyps, alopecia, areas of skin pigmentation, and nail atrophy.

- It is not inherited. There is no cancer risk.

- Diarrhea is a significant symptom. Vomiting, malabsorption, and protein-losing enteropathy may develop.

Neoplastic Polyps

- **Adenomas**

- Adenomas or adenomatous polyps are benign neoplasms of the colon and rectum.

- They are divided into three types based on the dominant feature in their histological structure:

  - Tubular adenoma (65-80%)

  - Tubulovillous adenoma (10-25%)

  - Villous adenoma (5-10%)

- Adenomatous polyps are precursors to cancer.

- By definition, these lesions are dysplastic.

- Factors determining the risk of malignant degeneration include:

  - Size (>2 cm)

  - Number

  - Polyp type (cancer risk: tubular: 5%, tubulovillous: 22%, villous: 40%)

  - Presence of ulceration or bleeding

  - Epithelial atypia

- Invasive cancer in a polyp is when malignant cells have breached the muscularis mucosa layer.

- Carcinoma in situ (intramucosal cancer) is when cancer has not breached the muscularis mucosa.

Treatment

- Colonoscopic polypectomy.

- Cancers located at the head of pedunculated polyps (in situ) can also be treated with colonoscopic polypectomy.

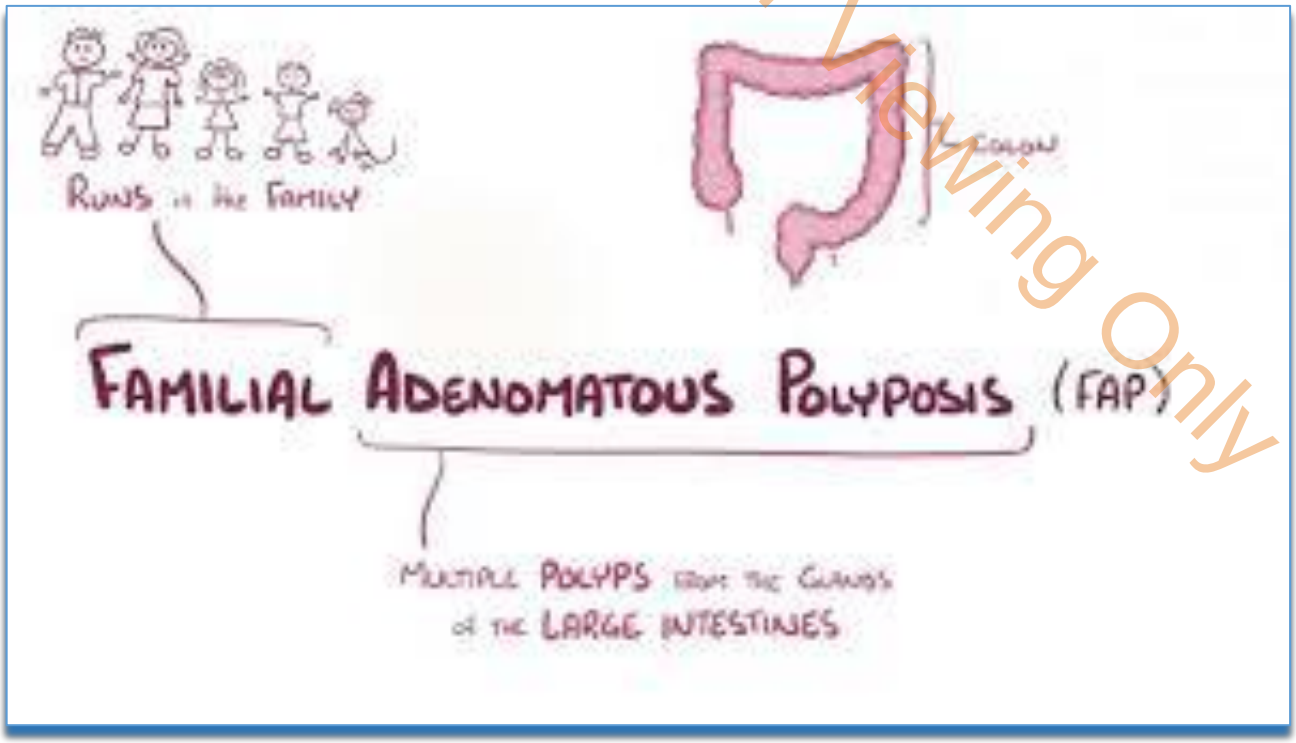
- Invasive cancer located at the head of a pedunculated polyp, if there is no involvement of the stalk, carries a low risk of metastasis (<1%) and can be treated with colonoscopic polypectomy.

- Invasive cancer developing from a sessile polyp extends to the submucosa, and the best treatment is segmental colectomy.

- Polypectomy has complications such as perforation and bleeding.

- If a small perforation occurs in a patient who has undergone complete bowel preparation, and if the patient's overall condition is stable, the patient can be managed with bowel rest, broad-spectrum antibiotics, and close monitoring. If signs of sepsis, peritonitis, or clinical deterioration appear, a laparotomy is required.

Adenomatous Polyposis



- **Familial Adenomatous Polyposis (FAP)**

- It develops as a result of a mutation in the APC gene located on chromosome 5.

- Approximately 1% of all colorectal cancers are due to FAP.

- 75% of patients have an APC mutation; 25% of cases occur without a family history.

- Hundreds to thousands of polyps appear in the colon and rectum.

- Polyps begin to appear during adolescence; if untreated, they progress to colorectal cancer.

- The likelihood of developing colon cancer by the age of 50 in FAP patients is 100%.

- FAP also includes benign extracolonic findings such as:

  - Congenital hypertrophy of the retinal pigment epithelium

  - Epidermoid cysts

  - Osteomas

  - In addition to colorectal cancer, patients with FAP have an increased risk of other cancers:

    - Upper gastrointestinal neoplasms (stomach and duodenum polyps, duodenal and periampullary cancers)

    - Hepatobiliary tumors (hepatoblastoma, pancreatic cancer, cholangiocarcinoma)

    - Thyroid cancers (with a cribriform growth pattern)

    - Desmoid tumors

    - Medulloblastomas

- The risk of developing duodenal adenomas in FAP patients is 100%. These adenomas have a risk of malignant transformation: the risk of duodenal cancer in these patients is 100 times higher than in the general population.

- Key symptoms include rectal bleeding, diarrhea, abdominal pain, and mucus discharge.

- First-degree relatives of FAP patients should be screened with flexible sigmoidoscopy starting from age 10-15.

- Congenital hypertrophy of the retinal pigment epithelium (CHRPE), visible on indirect ophthalmoscopy, should be evaluated.

- Gastric and duodenal adenomas can be seen under age 20; however, screening with upper endoscopy should begin at age 30.

- Treatment :

- The most appropriate treatment for FAP patients is the removal of the entire colon and rectal mucosa. Total proctocolectomy with ileal reservoir and ileoanal anastomosis is the most commonly recommended procedure.

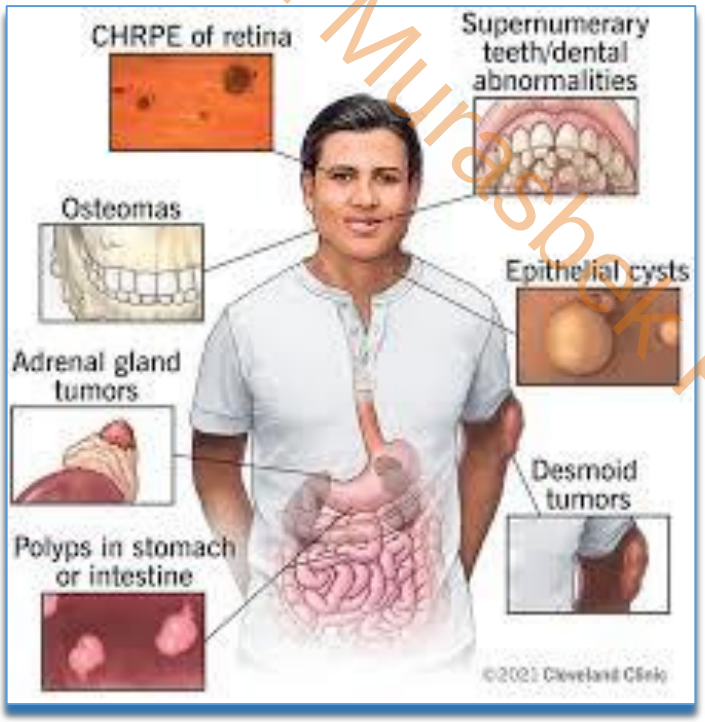
- In this approach, mucosectomy is performed up to the level of the dentate line, removing all mucosa at risk.

- COX-2 inhibitors (**sulindac, celecoxib**) can slow down or halt the development of polyps.

- In FAP patients, the potentially serious and fatal extracolonic manifestation is intraabdominal desmoid tumors.

- After surgical intervention, fibrous tissue can develop in the mesentery of the small intestine, pulling on the small intestine and leading to ischemia.

- In some patients with desmoid tumors, tamoxifen and COX-2 inhibitors have been found to be beneficial.



Gardner Syndrome

- FAP combined with osteomas, epidermoid cysts, desmoid tumors, dental anomalies, and skin lesions.

- The frequency of extracolonic adenomas is higher than in FAP.

- Consequently, the risk of duodenal and periampullary cancers is much more significant.

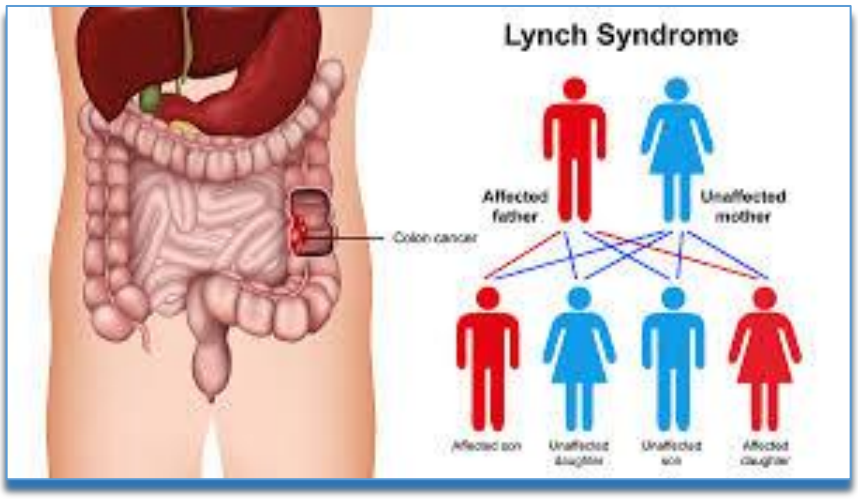
- Treatment is similar to that for FAP.

- **Turcot Syndrome**

- FAP combined with central nervous system tumors (gliomas, medulloblastomas).

Hereditary Non-Polyposis Colorectal Cancer (HNPCC)

- More common than FAP, accounting for about 3% of colorectal cancers.



- It is the most frequently observed hereditary colorectal cancer syndrome.

- Caused by mismatch repair mutations.

- About 70% of individuals affected by this syndrome will develop colon cancer.

- Colon cancers typically appear around ages 40-45.

- Although colon cancers are often mucinous adenocarcinomas, their prognosis is generally better.

- The risk of synchronous and metachronous colon cancers is 40%.

- Lynch Syndrome I and II

- Lynch I syndrome is characterized by cancers, particularly in the **proximal colon only**.

- The risk of synchronous or metachronous colorectal cancer is 40%.

- Lynch II syndrome also involves proximal colon cancer as a common cancer but includes other cancers such as endometrial cancer, as well as ovarian, pancreatic, gastric, small bowel, biliary tract, and urinary tract cancers.

- Screening with annual colonoscopy begins at ages 20-25.

- Diagnosis is based on the Amsterdam criteria:

    - At least one first-degree relative of the others, with at least three relatives having histologically confirmed colorectal cancer.

    - In at least two consecutive generations.

    - At least one of the colorectal cancers diagnosed before age 50.

Colon and Rectal Cancers

- It is the most common cancer in the gastrointestinal system.

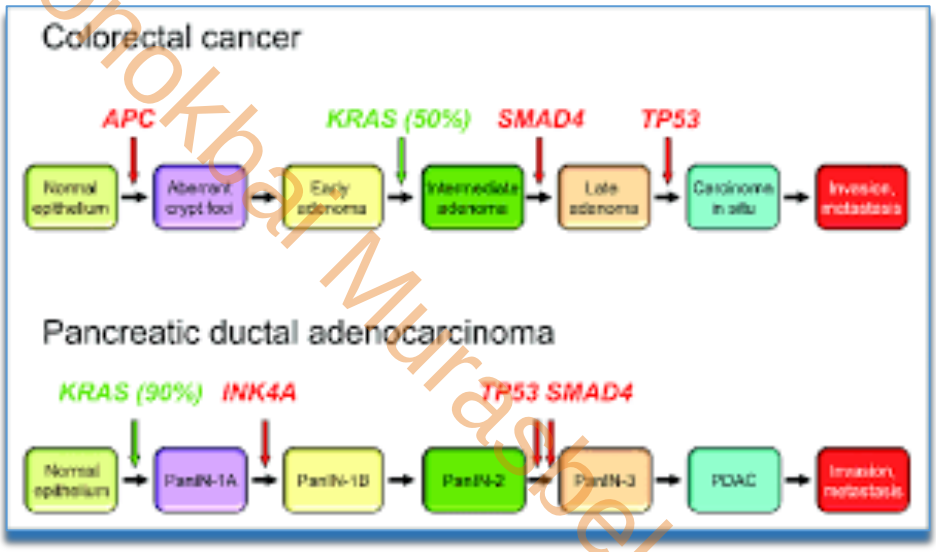
- Among visceral cancers, it is the second most common after lung cancer.

- It is responsible for 13% of cancer-related deaths.

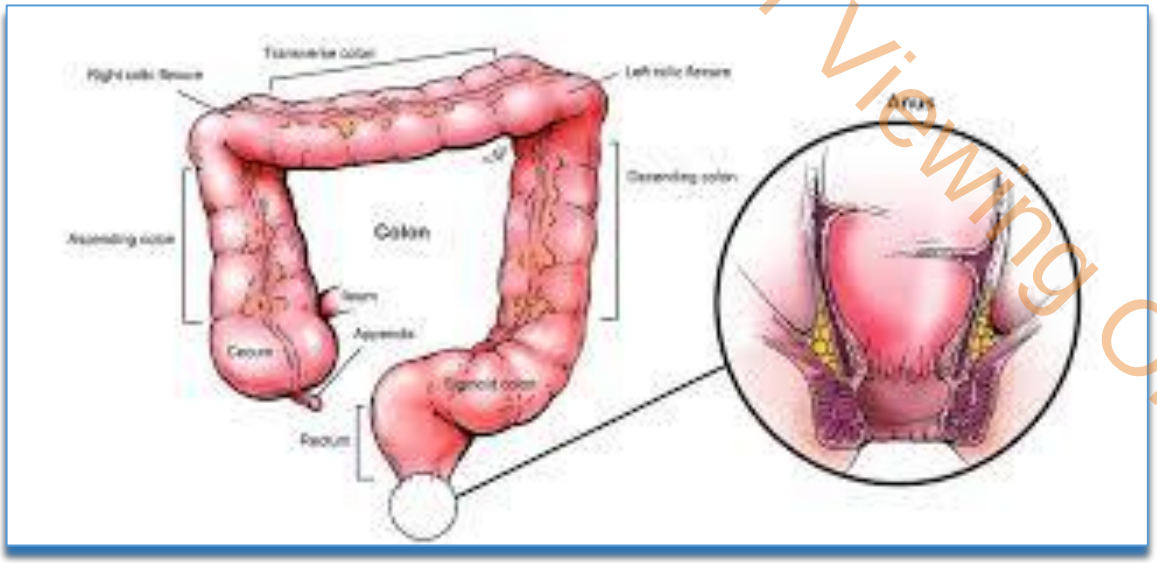
- The male-to-female ratio for colon cancer is 1.2:1, while for rectal cancer it is 1.1:1.

- The distribution of polyps and cancer is parallel, with the highest frequency in the rectosigmoid junction.

- In the last four decades, the incidence of right colon cancer has increased. About 40% of colon cancers are located in the proximal area of the flexible sigmoidoscopy view.



- Right colon cancers account for approximately 20% of all colon cancers.



- **Colorectal Cancer Development(Genes Involved in Colorectal Cancer Development)**

- APC Gene: A tumor suppressor gene located on chromosome 5.

- K-ras Gene: An oncogene that causes inactivation of G proteins. Mutations in this gene result in continuously active G proteins and increased cell proliferation.

- MYH Gene: Located on chromosome 1, responsible for the development of attenuated familial adenomatous polyposis (AFAP). It is inherited in an autosomal recessive manner.

- DCC Gene: One of the major tumor suppressor genes, thought to be related to cell differentiation.

- P53 Gene: A tumor suppressor gene. P53 mutations are found in 75% of colorectal cancers.

- **High-Risk Factors for Colorectal Cancer**

- Age over 50

- Premalignant lesions

- Ulcerative colitis lasting more than 10 years

- Crohn's disease with strictures

- Familial Adenomatous Polyposis (FAP)

- Hereditary Non-Polyposis Colorectal Cancer Syndrome (HNPCC)

- History of colon polyps

- Family history of polyps or colorectal cancer

- **Medications Thought to Reduce Colorectal Cancer Risk**

- Aspirin and NSAIDs (likely reduce colorectal cancer risk by decreasing prostaglandin synthesis)

- Oral folic acid and calcium (reduce the risk of adenomatous polyps)

- Selenium

- Antioxidant vitamins (ascorbic acid, beta-carotene, tocopherol) may reduce cancer risk

- Estrogen replacement therapy reduces colon cancer risk in women

- **Screening Tests**:

- Many colon cancers develop from polyps. Detecting and removing polyps can prevent cancer development.

- Early detection increases the chances of successful treatment.

- Screening tests are established for asymptomatic patients.

- The methods used in screening are listed in the table below.

- Fecal Occult Blood Test (FOBT)

- This is the least invasive test.

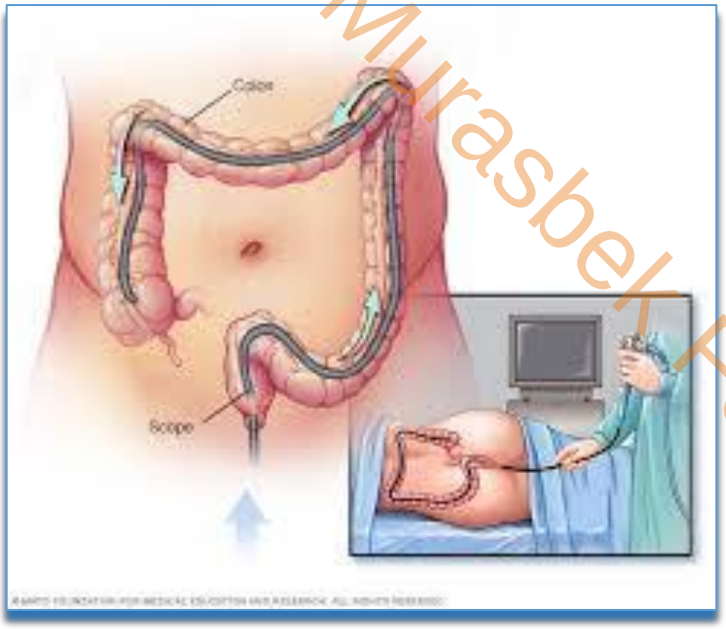
- It is known to reduce colorectal cancer mortality by 33%.

- However, FOBT has low sensitivity; it misses about 50% of cancers and a large portion of adenomas.

- Its specificity is also low; about 90% of positive results do not actually indicate colorectal cancer.

- Despite this, it remains a part of screening guidelines.

- For those over 50 years old, annual FOBT is recommended. If the result is positive, a colonoscopy is required.



- Colonoscopy

- This is the most useful but also the most invasive screening test for colorectal cancer.

- Double-Contrast Barium Enema and Sigmoidoscopy

- **Spread Pathways:**

- Direct Contiguity: Tumor cells may be present in the bowel segment observed 5 cm distal and 5 cm proximal to the lesion.

- **Lymphatic Spread:** This is the most common route of spread for colorectal cancers. Factors affecting lymph node metastasis include tumor size, poor differentiation, lymphovascular invasion, and depth of invasion. The number of metastatic lymph nodes increases the likelihood of distant metastasis and indicates a poor prognosis.

- **Hematogenous Spread:** The tumor spreads through the portal system. It most commonly metastasizes to the liver and, secondarily, to the lungs. Bone metastases can also occur. For hematogenous metastasis, the tumor must invade the venous wall.

- **Gravity-Dependent Seeding:** Cells detached from the tumor tissue due to gravity proliferate where they land. This often results in rectovesical (Blumer's shelf), ovarian (Krukenberg tumor), or peritoneal carcinomatosis.

- **Implantation During Surgery**: During surgery, manipulation and cells shed from the tumor can implant at new sites. Therefore, the tumor mass should be manipulated as little as possible during surgery.

CLINIC

- Clinical symptoms vary based on localization.

- The classic initial symptoms of colorectal cancer are changes in bowel habits and rectal bleeding, especially in distal colon cancers.

- Abdominal pain, gas, and other signs of obstruction suggest larger tumors and more advanced disease.

- The left colon has a narrower diameter and more solid contents, making obstruction more common in left colon cancers.

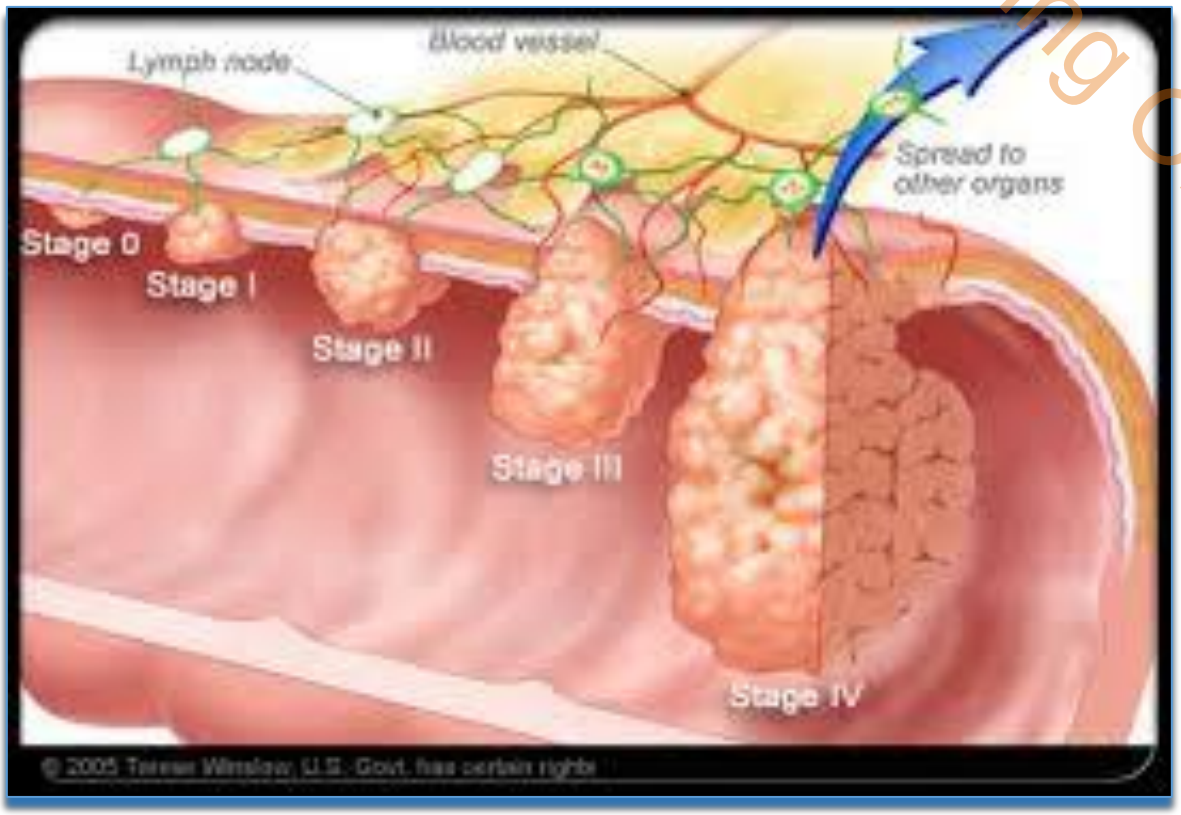
- Obstruction can lead to perforation (more commonly from the cecum) and peritonitis.

- Colon cancer patients can be asymptomatic. Non-specific symptoms such as unexplained anemia, weakness, and weight loss may be present.

- Obstruction is less common and occurs later in rectal cancer. It is usually associated with bloody mucus stools and tenesmus.

- The most common finding in rectal cancer is hematochezia (passing fresh blood through the rectum).

- **Staging**



- Colorectal cancer staging is based on tumor depth, lymph node involvement, and the presence or absence of distant metastasis.

- Older staging systems, such as the Dukes classification and its Astler-Coller modification, have been replaced by the TNM staging system.

Colon Cancer TNM Staging

- T0 - No evidence of primary tumor

- Tis- Tumor in situ (mucosa)

- T1- Tumor in submucosa

- T2- Tumor in muscularis propria

- T3-Tumor extends through muscularis propria into subserosa

- T4a-Tumor penetrates the visceral peritoneum (serosa)

- T4b- Tumor invades adjacent organs

- N0-No regional lymph node metastasis

- N1- Metastasis in 1-3 regional lymph nodes

- N1a-Metastasis in 1 lymph node

- N1b- Metastasis in 2-3 lymph nodes

- N1c-Tumor deposits in subserosa, mesentery, or peritoneum without regional lymph node metastasis

- N2- Metastasis in 4 or more regional lymph nodes

- N2a-Metastasis in 4-6 lymph nodes

- N2b- Metastasis in 7 or more lymph nodes

- N3- Metastasis in lymph nodes around major vascular structures

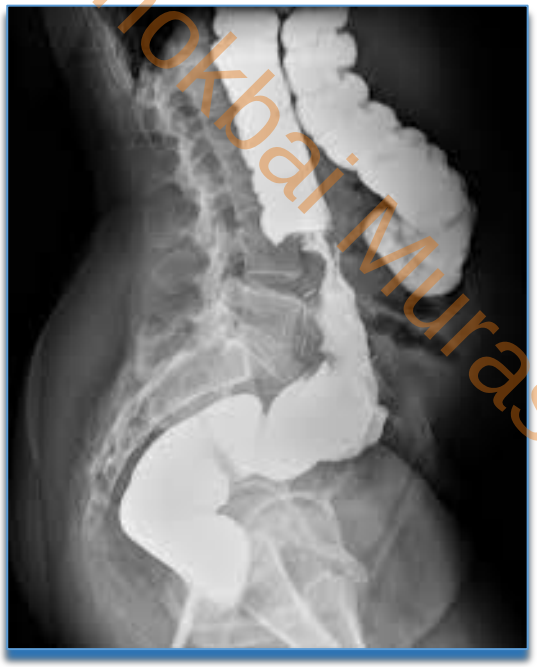
- M0- No distant metastasis

- M1- Distant metastasis present

- M1a- Metastasis to one distant organ or site

- M1b-Metastasis to more than one distant organ or site

DIAGNOSIS AND PREOPERATIVE EVALUATION



-Barium Colonography

- A filling defect or apple core appearance is typical.

-Colonoscopy

- Provides the most accurate and complete evaluation of the colon.

- Screening for synchronous tumors is performed (up to 5%).

-Other Tests

- After diagnosis, additional tests are necessary to assess the extent of the disease and to check for metastases:

- Chest X-ray and CT: Routine tests to rule out pulmonary metastases.

- Abdominal CT: Preferred method for assessing the degree of tumor invasion and intra-abdominal metastases. It is the preferred method for clinical staging.

- Abdominal Ultrasound: Has no role in the diagnosis of colorectal cancer.

- Endorectal Ultrasound and MRI: Used to determine the T and N stages (local extent) in rectal cancer. Endorectal ultrasound is the most reliable test for evaluating perirectal spread.

- If colonoscopy cannot be performed, CT colonography or barium colonography may be used.

-Carcinoembryonic Antigen (CEA)

- CEA is a glycoprotein found in embryonic and fetal tissues. It is not present in the colon mucosa of normal adults.

- In cases of early carcinoma that does not completely penetrate the bowel wall, serum CEA levels are usually normal.

- Elevated serum CEA levels are indicative of recurrence or metastasis.

- CEA levels can also be elevated in lung, breast, stomach, or pancreatic tumor

- Additionally, CEA levels may rise in smokers, patients with cirrhosis, pancreatitis, kidney failure, or ulcerative colitis. It is non-specific.

- Another tumor marker is CA 19-9.

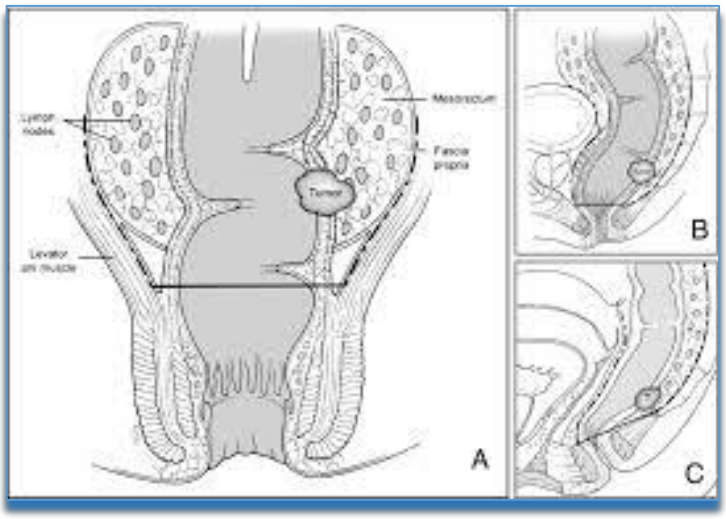
Surgical Treatment

- Surgical treatment is performed for both palliative and curative purposes.

- Curative Surgery: Involves the removal of tumor tissue along with at least 5 cm of the distal (excluding the rectum) and 5 cm of the proximal bowel segment. The lymphatic drainage and arteries of this area are also removed. Dissection of at least 12 lymph nodes is required for adequate staging. Depending on the location, procedures such as right hemicolectomy, transverse colectomy, or left hemicolectomy may be performed.

- Sigmoid Colon Tumors: For tumors in the sigmoid colon, sigmoid resection should be performed. For rectosigmoid lesions, an anterior resection is indicated.

In rectal cancers, tumors in the upper rectum are treated with anterior resection, while those close to the sphincter require abdominoperineal resection. However, with the use of staplers, it is possible to perform lower anterior resection, extending beyond these boundaries. For curative surgery, an attempt is made to adhere to a 2 cm distal surgical margin.



- Total Mesorectal Excision (TME): Nowadays, radical resection for rectal tumors is performed as "total mesorectal excision (TME)." This method involves less bleeding during surgery and has resulted in reduced local recurrence rates. Additionally, it causes less damage to the pelvic nerves and the presacral plexus.

- Complications: During TME, damage to the hypogastric plexus can result in retrograde ejaculation and bladder issues. Damage to the parasympathetic nerves during the procedure can lead to impotence and an atonic bladder.

- **Treatment of Colon Cancer by Stage:**

- Stage 1: Surgery

- Stage 2: Surgery followed by chemotherapy

- Stage 3: Surgery followed by chemotherapy

- Stage 4: Surgery if tumor-negative resection can be achieved

- **Treatment of Rectal Cancer by Stage**:

- Stage 1: Surgery

- Stage 2: Preoperative (neoadjuvant) radiation therapy and chemotherapy followed by surgery

- Stage 3: Preoperative (neoadjuvant) radiation therapy and chemotherapy followed by surgery

- Stage 4: Surgery if tumor-negative resection can be achieved

- **Indications for Emergency Surgical Treatment:**

1.Obstruction: Managed with resection, anastomosis, and/or proximal colostomy or ileostomy, or decompression.

2. Perforation

3.Bleeding

- **Follow-Up for Colon and Rectal Cancer Patients:**

- The goal is to diagnose and treat recurrences while they are still resectable.

- Most recurrences occur within the first two years, so this period should be closely monitored.

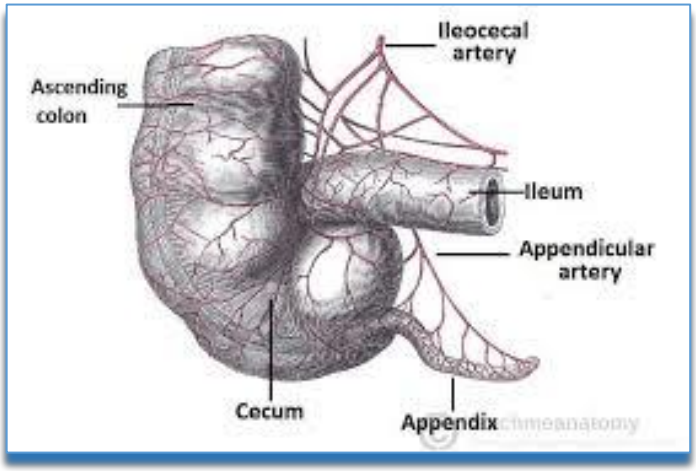
- Long-term follow-up includes regular use of CEA testing, sigmoidoscopy, colonoscopy, and CT scans.

- Elevated CEA levels require colonoscopy and/or computed tomography.

- Magnetic resonance imaging (MRI) or CT angiography is used for assessing liver metastases.

- Positron emission tomography (PET) is highly sensitive for detecting metastatic and recurrent tumors.

APPENDIX VERMIFORMIS DISEASES AND SURGERY



- The appendix vermiformis is a part of the gastrointestinal-associated lymphoid tissue (GALT) in the gastrointestinal system.

- It produces immunoglobulin A.

- There is no clear evidence showing a direct role of appendectomy or the absence of the appendix in the development of diseases in humans.

- However, a negative relationship between appendectomy and the development of ulcerative colitis has been reported. This means that appendectomy seems to be protective against the development of ulcerative colitis, but this is only valid for those who have undergone appendectomy before the age of 20.

ANATOMY OF THE APPENDIX VERMIFORMIS

- In newborns, the appendix vermiformis is an extension of the cecum and extends vertically downwards.

- Over time, the cecum grows and undergoes sacculation. The appendix also undergoes medial and internal rotation.

- The junction with the cecum (the base) does not change, but the free end can be in various locations, leading to different clinical presentations of appendicitis based on its position.

- The free end can be located in six different positions: towards the pelvis, along the iliac crest, towards the sacral promontory, under the ileum, behind the cecum, or lateral to the cecum.

- The most common location is retrocecal (but intraperitoneal).

ACUTE APPENDICITIS

- Acute inflammation of the appendix vermiformis.

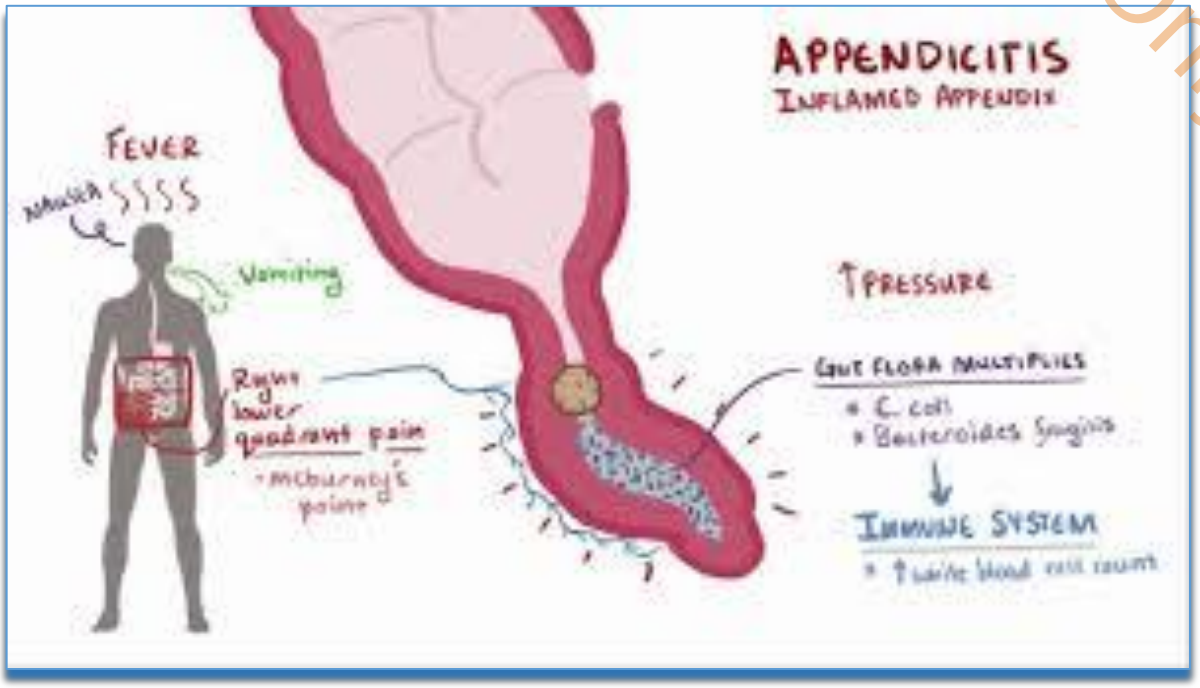
- **INCIDENCE**

- It is a common surgical disease.

- Appendectomy is the most frequently performed emergency surgery worldwide.

- Lifetime risk: 8.6% in men, 6.7% in women.

- Most commonly seen in the 2nd and 3rd decades of life (most frequent in the 2nd decade).



ETIOLOGY

-Obstruction (90%)

- The appendix vermiformis is already a narrow organ. Even the smallest particle can lead to lumen obstruction

- Fecaliths (hardened, crystallized stools) are the most common cause.

- Lymphoid hyperplasia (especially in children).

- Foreign bodies (seeds, bone fragments, barium, etc.).

- Parasites (especially Ascaris).

-Other Causes (10%)

- Generalized infections (especially respiratory tract infections).

- Fibrosis (particularly responsible for acute appendicitis in the elderly). The cause is circulatory disorder.

- In newborns, if acute appendicitis is observed, conditions such as cystic fibrosis, aganglionic megacolon, or neonatal necrotizing enterocolitis should be considered.

-Bacteriology (Multibacterial.Most commonly B. fragilis and E. coli are seen)

- **PHASES OF ACUTE APPENDICITIS**

Visceral Phase

- The issue is within the appendix vermiformis.

- Surrounding tissues and the serosa have not yet joined.

- Clinical presentation: There is a dull pain around the umbilicus.

- The pain cannot be well localized. It is visceral pain.

Somatic Phase

- The inflammation has surpassed the appendix vermiformis.

- The serous and surrounding tissues are also infected.

- With the infection of the parietal peritoneum, a well-localized, sharp pain arises.

- The pain is mostly at McBurney's point. However, the location of the pain may change depending on the position of the appendix.

- **CLINICAL SYMPTOMS**

-Abdominal Pain

- During the visceral phase, the pain is dull and around the umbilicus.

- On average, 4-6 hours (1-12 hours) later, the pain becomes localized and intensifies.

- This indicates the beginning of the somatic phase. The pain localizes to the right lower quadrant.

-Anorexia (Loss of Appetite)

- Present in 90-95% of patients, but often goes unnoticed.

- It appears before the pain. It is the first detected symptom.

-Nausea and Vomiting

- Present in 75% of patients.

- Vomiting is not severe (if vomiting is severe, other causes should be considered).

- Vomiting occurs after the onset of pain. If vomiting occurs before pain, it is usually not acute appendicitis.

- The cause of vomiting is peritoneal irritation. The stomach contents are vomited

-Fever

- Moderate fever is observed.

- Generally does not exceed 38°C.

- If the fever is higher than 39°C, a complication has likely occurred.

-Tachycardia

- Mild tachycardia is present.

- If there is severe tachycardia (120-140 beats per minute), it indicates a complication.

- **CLINICAL FINDINGS**

Muscle defanse (involuntary muscle resistance)

- The most valuable sign of peritoneal irritation.

Rovsing's Sign- When pressure is applied to the left lower quadrant and gas is pushed upwards over the abdomen, pain appears in the right lower quadrant.

Obturator Sign

- Seen in patients with a pelvic appendix.

- The right thigh is flexed and internally rotated. If pain occurs, it is positive.

Psoas Sign

- Positive in retrocecal appendicitis.

- The patient lies on their left side, and the right thigh is extended backward.

- The psoas muscle is stretched, irritating the appendix and causing pain.

Ten-Horn Sign

- Pain is felt in the right lower quadrant when the right testicle is pulled downwards.

-Dunphy's Sign

- Pain in the right lower quadrant with coughing.

Rosenstein's Sign

- Pain increases when the patient lies on their left side.

Aaron's Sign

- Pain in the epigastrium when pressure is applied to McBurney's point.

Rectal Examination

- Must be performed.

- If the appendix vermiformis is in the Douglas pouch (rectovesical fossa), pain or a mass is detected on rectal examination.

- **DIAGNOSIS**

- A good history and physical examination are essential for diagnosis.

- Laboratory findings are not very useful in diagnosis, but some routine tests should be performed.

-Complete Blood Count

- Especially the white blood cell count is important.

- In appendicitis, the white blood cell count is expected to be between 10,000-18,000/mm³.

- White blood cell count > 18,000/mm³ suggests complications.

- In 10% of cases, the white blood cell count is normal.

- Left shift in the leukocyte formula is important in neutropenic patients.

- CRP is controversial.

-**Complete Urinalysis**

- Usually normal.

- If the appendix is near the bladder or ureter, white blood cells or red blood cells may be seen in the urine, but bacteriuria is not observed.

- Complete urinalysis is done to rule out urinary tract diseases.

-Radiology

-Direct Abdominal Radiography

- Can be performed on patients presenting with suspicion of acute abdomen.

- Sensitivity and specificity for acute appendicitis are very low.

- Ultrasonography

- Clear visualization of the inflamed appendix (target sign) via USG is diagnostic.

- Sensitivity and specificity are 85-98% and 85-96%, respectively.

- Suitable for pediatric and pregnant patients.

-CT Scan

- CT is the most reliable imaging method for diagnosing acute appendicitis.

- Sensitivity and specificity are 92-97% and 85-94%, respectively.

- MRI

- Performed in pregnant patients when USG is negative.

- There is no consensus on the use and priority of imaging methods for the diagnosis of acute appendicitis.

Alvarado score	
Feature	**Score**
Migration of pain	1
Anorexia	1
Nausea	1
Tenderness in right lower quadrant	2
Rebound pain	1
Elevated temperature	1
Leucocytosis	2
Shift of white blood cell count to the left	1
Total	**10**

ALVARADO Score assessment:

- <3: Low probability of appendicitis - 4-6: Further imaging required

- ≥7: High likelihood of appendicitis

- **Differential Diagnosis**

- The most common condition that acute appendicitis is confused with in differential diagnosis is acute mesenteric adenitis.

- Other conditions it can be confused with include pelvic inflammatory disease, ruptured ectopic pregnancy, acute gastroenteritis, ruptured ovarian cyst, Meckel's diverticulitis, and Crohn's enteritis.

- **Complications**

--**Perforation**

- The most common complication.

- Frequently seen in children and the elderly (ages <5 and >65).

- In adults, surrounding tissues such as omentum, ligaments, and parietal peritoneum often confine the inflamed appendix (forming a "plastron") and prevent perforation.

- This mechanism is insufficient in children.

- In HIV-positive patients, the perforation process is faster.

- Perforation typically occurs on the anti-mesenteric side, just beyond the obstruction.

--**Generalized Peritonitis**

- Occurs when perforated contents spread throughout the abdominal cavity, leading to an adynamic ileus.

- In later stages, it can lead to mechanical obstruction due to adhesions.

--**Plastron (Phlegmon)**

- If there is a mass or fullness in the right lower quadrant of a patient suspected of acute appendicitis, a plastron or periappendiceal abscess should be considered.

- CT is the most reliable imaging method.

- A plastron without complications can be treated with observation, intravenous fluids, and antibiotics.

- An interval appendectomy is performed 6 weeks later. However, there are differing opinions on interval appendectomy.

--Abscess Formation

- Plastron formation sometimes prevents the progression of acute appendicitis and its perforation; however, if the infection is not contained, an abscess or diffuse peritonitis may develop.

- The abscess is often periappendiceal.

- The initial imaging method required for diagnosing a plastron abscess is CT.

- CT is also the most useful test for diagnosis.

- In cases of delayed abscessed acute appendicitis without diffuse peritonitis, drainage and antibiotic treatment are first performed, followed by an interval appendectomy.

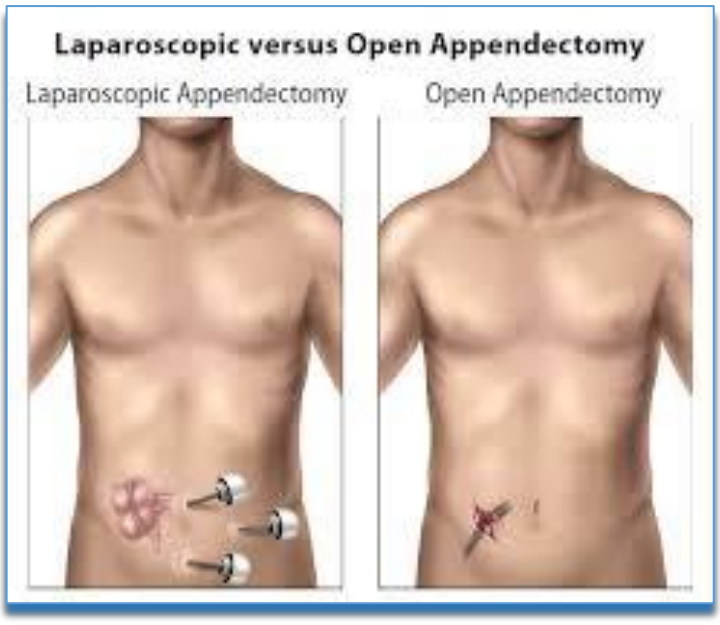
--Pylephlebitis (Pyelophlebitis)

- It is septic thrombophlebitis of the portal vein.

- A rare but very serious complication.

- Pain is located in the right upper quadrant; fever rises further, and jaundice appears.

- **Treatment**



- The treatment for uncomplicated acute appendicitis is emergency appendectomy.

- Laparoscopic appendectomy has a slight advantage over open appendectomy.

- In cases with complications like abscess or plastron (if there is no diffuse peritonitis or sepsis), the appropriate approach is intravenous fluid therapy, antibiotics, and if necessary, percutaneous drainage, followed by interval appendectomy after 6 weeks (though this is debated).

- Mortality is higher in older patients.

- The most common postoperative complication is surgical site infection.

- Laparoscopy is particularly valuable for both diagnosis and treatment in young women with suspected diagnoses and older patients with suspected malignancy. It is also considered in obese patients where appendectomy might not be feasible through a small incision.

Acute Appendicitis in Pregnancy

- Acute appendicitis is the most common non-obstetric emergency in pregnant women.

- It occurs in 1 in 766 births.

- It can occur at any stage of pregnancy; however, it is rare in the third trimester.

- After appendectomy, the fetal loss rate is about 4%, and the rate of preterm birth is about 10%.

- These rates increase if perforation occurs, making accurate preoperative diagnosis crucial in this group.

- The rate of negative appendectomy in pregnant women is approximately 25%.

- New onset of abdominal pain in a pregnant woman should raise suspicion of acute appendicitis.

- Leukocytosis is not a reliable indicator in pregnant women.

-Imaging should be routinely performed (Ultrasound ± MRI). In suspected cases, diagnostic laparoscopy may be considered. The choice between laparoscopic appendectomy and open appendectomy is debated.

- **Non-Operative Treatment**

- Days 1 and 2: Third-generation cephalosporin + Metronidazole

- Followed by 8-10 days of Fluoroquinolone + Metronidazole

Due to high failure rates (around 40%), non-operative treatment is primarily planned for acute uncomplicated appendicitis when the risk of surgery is high.

Appendix Tumors

- Tumor masses are found in approximately 1% of appendectomy specimens.

- The most common appendix tumors are carcinoids and adenomas.

- **Carcinoid**

- The appendix is the most common site for carcinoids in the gastrointestinal tract (followed by the small intestine and rectum).

- They are generally located at the tip of the appendix.

- Malignant degeneration is rare unless the tumor exceeds 2 cm.

- Carcinoid syndrome is very rare in appendiceal carcinoids.

- Treatment is based on the appendiceal carcinoid treatment algorithm.(Less then 2cm and located at the tip of the appendix-appendectomy, more then 2cm located in base of appendix)

- **Adenocarcinoma**

- Typically, one of three histological subtypes is identified:

- Mucinous adenocarcinoma

- Colonic adenocarcinoma

- Adenocarcinoid

- They usually present with symptoms of acute appendicitis.

- The treatment is a formal right hemicolectomy.

- The incidence of synchronous and metachronous cancers is increased.

- **Appendiceal mucocele**

- Refers to the dilation of the appendiceal lumen due to mucinous secretion. Mucoele can develop due to one of the following four conditions:

- Retention cyst

- Mucosal hyperplasia

- Mucinous cystadenoma

- Mucinous cystadenocarcinoma

- Clinical Presentation: Non-specific.

- Detection: Usually found incidentally during surgery for acute appendicitis.

- Management: An intact mucosal does not pose additional risk for the patient. In the presence of rupture or malignancy, more extensive resection is performed.

- Pseudomyxoma peritonei: If present, the prognosis is generally worse. Adjuvant intraperitoneal hyperthermic chemotherapy should be applied in addition to standard radical cytoreductive surgery.

- **Lymphoma**

- Appendiceal lymphoma: Extremely rare.

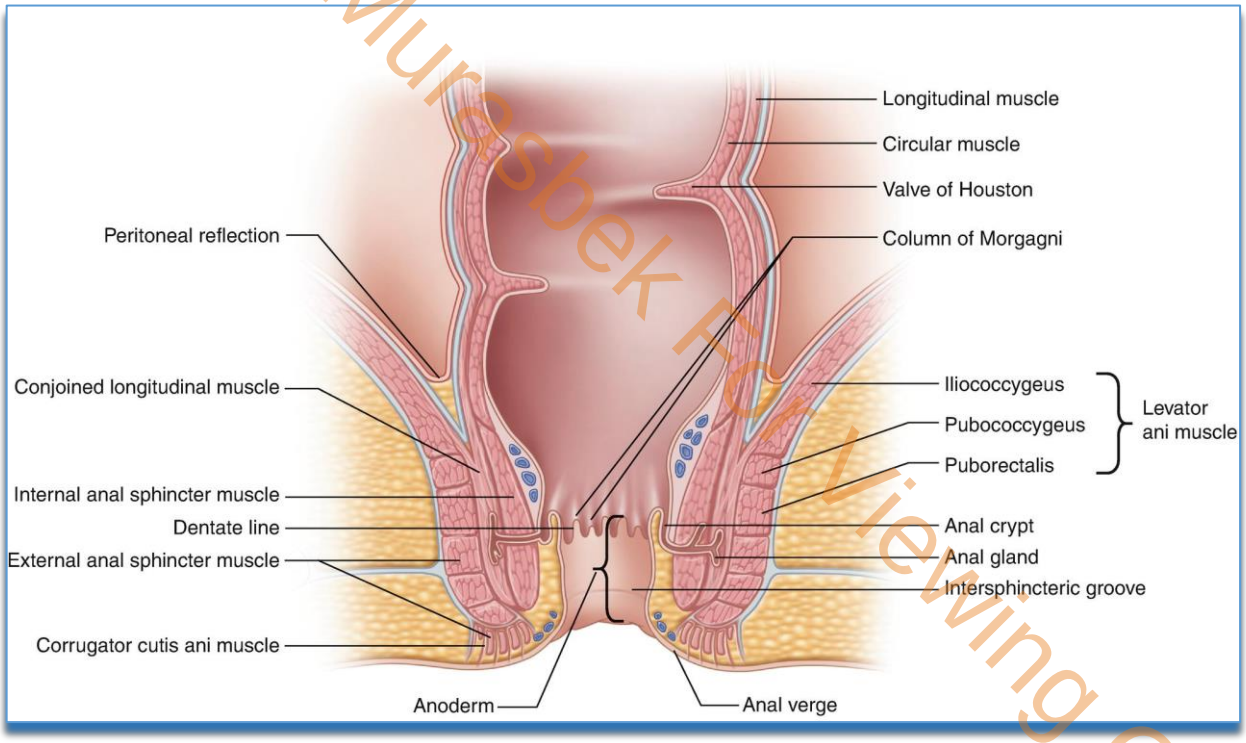
- Preoperative Imaging: If the appendiceal diameter is > 2.5 cm or if there is involvement of surrounding tissues, suspicion should arise.

- **Treatment:** If confined to the appendix, appendectomy is sufficient, and no adjuvant treatment is required. If it extends to the cecum or mesentery, right hemicolectomy is performed.

Chapter
4.

Perianal Region Diseases and Surgery

Anal Canal: The most distal part of the gastrointestinal system.



- Begins at the pelvic diaphragm and ends at the edge of the anus.
- The muscular junction between the rectum and anal canal can be felt with the finger as thickened ridge called anorectal ring.
- The epithelium of upper half of anal canal is derived from hindgut and is columnar while the epithelium of lower half of anal canal is squamous.
- The dentate line is the point where two epithelia above this line join.
- The anal canal above the dentate line is innervated by the sympathetic and parasympathetic nervous system.
- Distal to dentate line, the anal canal is innervated by the somatic nervous system.

- **INTERNAL SPHINCTER**

  - It is a continuation of the inner circular muscle of the rectum, autonomously innervated and involuntary smooth muscle.

  - It has sympathetic and parasympathetic innervation, both of which have inhibitory effects.

- **EXTERNAL SPHINCTER**

- It is a striated muscle with somatic innervation and voluntary control.

- The external sphincter is divided into three parts: subcutaneous, superficial, and deep components.

Anal Sphincter Mechanism

- The deep component of the anal sphincter begins at the symphysis pubis, encircles the rectum, and attaches back to the symphysis pubis.

-The middle component starts from the coccyx, encircles the anus, and attaches back to the coccyx.

- The lowest ring is attached to the perineal skin.

- The uppermost and lowermost rings are innervated by the internal pudendal nerve, while the middle ring is innervated by the 4th sacral nerve.

- The external sphincter is responsible for 20% of the resting pressure of the anus, while the internal sphincter accounts for 80%.

- The external sphincter is 100% responsible for the squeezing pressure.

- The external sphincter provides fine control over liquids, solids, and gases.

- The internal sphincter is responsible for the fine control of gas.

ARTERIES

- There are three main arteries supplying the anorectum:

- Superior rectal artery (superior hemorrhoidal artery): A branch of the inferior mesenteric artery.

- Middle rectal artery: Branches off the internal iliac artery on both sides. It enters the rectum at the level of the levator ani.

- Inferior rectal artery: A branch of the internal pudendal artery, which comes from the internal iliac artery. It passes through the ischiorectal fossa and enters the rectum at the level of the sphincter muscle.

- Additionally, there is the middle sacral artery, but its contribution to the blood supply of the anorectum is insignificant.

VEINS

- Superior rectal vein (superior hemorrhoidal vein)

- Middle rectal vein

- Inferior rectal vein

The superior rectal vein drains into the portal system via the inferior mesenteric vein, while the middle and inferior rectal veins drain into the caval system via the internal pudendal vein. There are anastomoses between these two systems (porto-caval) in the internal hemorrhoidal plexus, and in cases of portal hypertension, these anastomoses can open up, leading to rectal varices.

LYMPHATIC DRAINAGE

- Follows the path of arteries and veins.

- Superior rectal lymphatics primarily drain the upper and middle parts of the rectum and empty into paraaortic lymph nodes.

- Middle rectal lymphatics- drain into the internal iliac lymph nodes.

- Below the pectinate line, drainage is into paraaortic and inguinal lymph nodes.

ANORECTAL FOSSAE

- **Perianal Fossa**

- Located immediately below the transverse septum on both the right and left sides.

- These two fossae are connected by the superficial posterior anal fossa.

- Anorectal abscesses most commonly occur in the perianal fossa.

- **Intersphincteric Fossa**

- Surrounds the entire length of the anal canal and is situated between the two sphincters.

- **Ischiorectal Fossa**

- Bounded superiorly by the levator ani, laterally by the lateral pelvic walls, and medially by the external sphincter muscle.

- Inferiorly, it is limited by the transverse septum of the ischiorectal fossa.

- Located on both sides of the rectum and connected to the deep posterior anal fossa.

- Composed of fat and connective tissue, also containing the inferior rectal vein and lymphatics.

- When these two fossae combine, a horseshoe abscess can form.

- **Supralevator Fossa**

- Bounded inferiorly by the levator ani and laterally by the pelvic walls, containing fat and connective tissue.

- Posteriorly, it is connected to the retrorectal fossa.

- **Retrorectal Fossa**

- Located between the pelvic diaphragm and the levator ani.

- Bounded laterally by the pelvic wall and medially by the rectum.

- **Deep Posterior Anal Fossa**

- Connects the ischiorectal fossae on both sides.

- Bounded inferiorly by the superficial external sphincter muscle.

BENIGN DISEASES OF THE ANORECTAL REGION

- **ANAL INCONTINENCE**

Etiology:

- Mechanical Causes: Typically due to trauma, most commonly obstetric injury.

- Can also occur as a result of hemorrhoidectomy, sphincterotomy, abscess drainage, or fistulotomy.

- Neurogenic Causes: Pudendal nerve injuries can also lead to anal incontinence.

- Severe diarrhea, fecal impaction, radiation proctitis, and distal tumors can also cause incontinence.

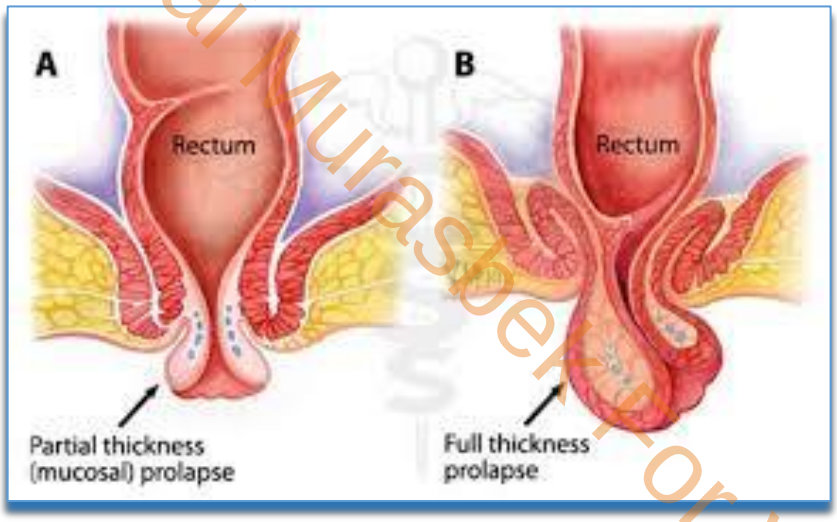
Diagnosis:

- Anal manometry and sphincter EMG are useful for diagnosis.

- Endosonography can be performed to visualize the external sphincters.

- Depending on the diagnosis, the sphincter mechanism may be surgically repaired.

- **Rectal Prolapse**



- Definition: A condition where part or all of the rectum's layers protrude through the anus.

- Common Ages: Most frequently seen in individuals under 2 years old and over 60 years old.

- Gender Prevalence: More common in women.

-Risk Factors for Rectal Prolapse:

- Chronic constipation

- Long rectosigmoid colon

- Deep rectouterine or rectovesical fossa

- Weak rectal sphincters

- Pelvic floor weakness and levator ani diastasis

- Lack of fixation of the rectum to the sacrum

- Neurological diseases (e.g., tabes dorsalis, multiple sclerosis)

- Previous anorectal surgery

- Hysterectomy

- **Patient Demographics:** 35% of patients are nulliparous (have never given birth).

- **Association:** Prolapse can also occur alongside solitary rectal ulcer syndrome.

- **Diagnosis:** May be confused with prolapsed incarcerated internal hemorrhoids. Typical prolapse shows circular mucosal folds.

- Over time and with progression, the rectum may only be reducible manually.

- **Symptoms:** Anorectal pain, bleeding, mucous discharge, and incontinence may be observed.

- Diagnosis and Classification: Performed through inspection.

- Additional Tests: Anal manometry, EMG, colon transit studies, defecography.

- **Purpose:** To assess sphincter function and choose the appropriate surgical procedure.

- **Primary Treatment:** Surgical intervention is the main treatment for prolapse.

- **Types of Surgery:**

- Classified into abdominal and perineal approaches.

- Abdominal Rectopexy: Either with or without sigmoid resection is the most durable repair.

- ## Hemorrhoidal Disease

- **Anatomy:** The upper anal canal contains vascular cushions rich in elastic fibers.

- **Function:** During defecation, these vascular structures swell with straining and prevent trauma to the anal canal epithelium.

- **Name:** These structures are called hemorrhoidal cushions.

- **Location:** There are three of these cushions located at approximately 3, 7, and 11 o'clock positions.

Pathogenesis

- **Trauma:** Trauma to the cushions is the primary mechanism.

- **Causes:** Hard stool, prolonged straining, increased abdominal pressure, and insufficient pelvic floor support.

- **Diet:** A diet low in fiber leads to constipation and hard stools.

- Straining: To pass hard stools, patients need to strain severely each time.

- **Result:** This causes excessive filling of the vascular structures.

- **Trauma**: Hard stool damages the mucosa during passage.

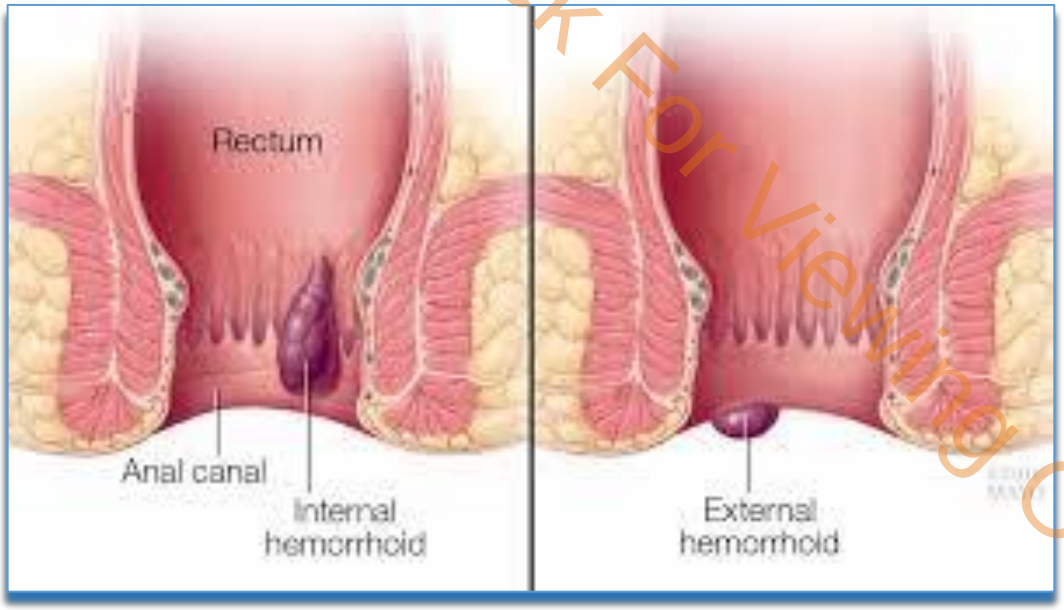
- **Bleeding:** Hemorrhage occurs in the lamina propria and may be visible on the stool.

- **Hematomas:** Hematomas may also form in the area.

- **Chronic Damage:** Repeated trauma leads to damage of the hemorrhoidal cushions, which can prolapse from the anal canal.

Internal Hemorrhoids

- **Location:** Internal hemorrhoids are located above the dentate line and are covered by



mucosa.

- **Main Symptom:** The most significant symptom is painless bleeding following defecation.

Internal Hemorrhoid Stages:

Grade 1: Painless bleeding.

Grade 2: Painless bleeding, prolapses, and spontaneously reduces.

Grade 3: Painless bleeding, prolapses, and requires manual reduction.

Grade 4: Bleeds, prolapses, and cannot be reduced; indicates a clear need for surgical intervention.

External Hemorrhoids

- **Origin:** Develop due to the dilation of inferior rectal veins.

- **Symptoms:** Usually asymptomatic.

- **Pain:** Pain becomes noticeable when thrombosed.

- **Complications:** May include ulceration and strangulation.

- Location:External hemorrhoids are located below the dentate line and are covered by anoderm.

Mixed Hemorrhoids

- **Description**: Presence of both internal and external hemorrhoids.

- **Treatment:** Typically requires hemorrhoidectomy for large, symptomatic mixed hemorrhoids.

Conservative Treatment of hemorrhoids

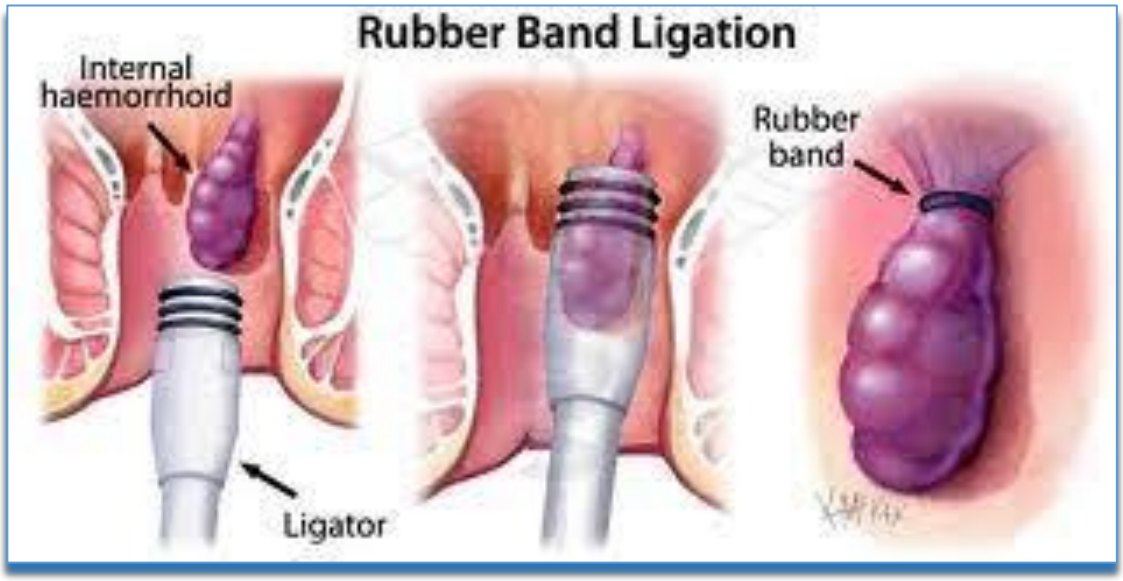
- **Diet and Lifestyle:**

  - High-fiber foods, laxatives, and warm sitz baths are recommended.

  - Spicy and hot foods should be avoided as they cause vasodilation.

- **Procedures**

  - Rubber Band Ligation: Used for persistent bleeding in Grade 1, 2, and selected Grade.3

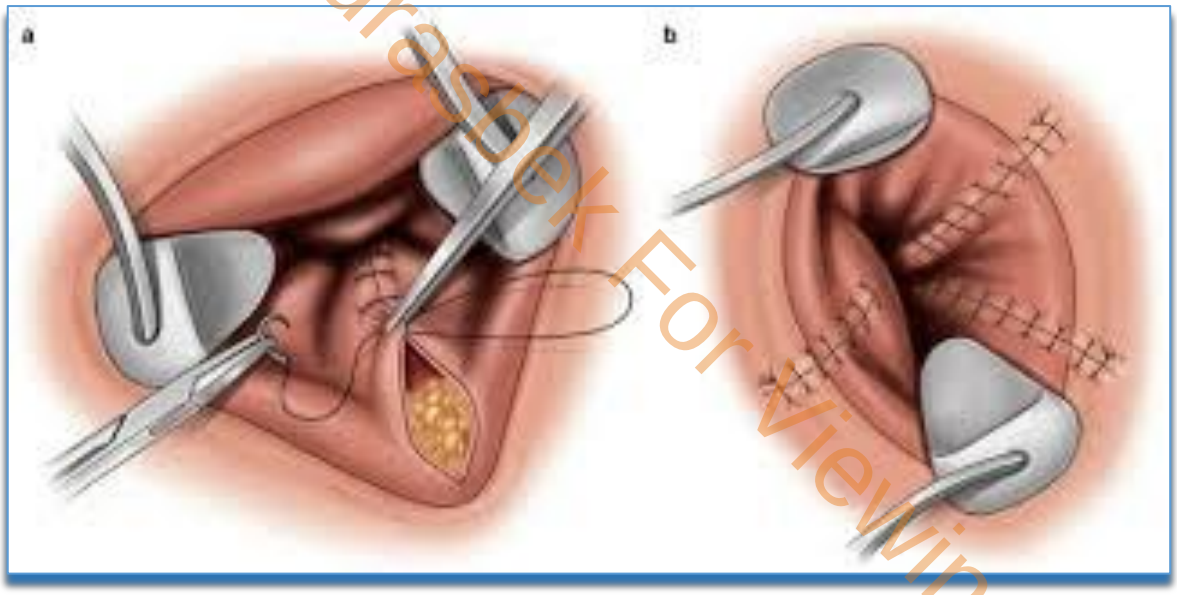


- Photocoagulation: Applied to Grade 1 and 2 small hemorrhoids.

- Sclerotherapy: Indications are similar to those for band ligation.

- Cryotherapy and Cautery: Less commonly used.

- Excisional Hemorrhoidectomy:



- Indicated for Grade 3 and 4 internal hemorrhoids and mixed hemorrhoids.

- Thrombosed External Hemorrhoids:

- Thrombectomy:

- Thrombosed external hemorrhoids cause severe pain within the first 24-72 hours.

- Thrombectomy is an effective treatment if performed during this period.

- If the patient presents late (after 72 hours), the thrombus and pain typically resolve on their own, making thrombectomy unnecessary.

- Sitz baths and analgesics are sufficient for late presentations.

- Stapled Hemorrhoidopexy:

- A procedure associated with less postoperative pain.

Hemorrhoidectomy Complications

- **Urinary Retention:** The most common complication (10-50%).

- **Fecal Incontinence**: Can occur.

- **Infection:** May present with severe pain, fever, and urinary retention as early signs.

- **Massive Bleeding**: Can occur in the early postoperative period (often in the recovery room).

- **Delayed Bleeding:** May occur 7-10 days after surgery.

- **Stricture and Ectropion:** Long-term complications.

Hemorrhoid Disease Treatment Algorithm

- **External Hemorrhoids**

    - Thrombosed:

        - < 72 hours: Thrombectomy.

        > 72 hours: Dietary adjustments, analgesic treatment, sitz baths.

    - Non-thrombosed:

        - Dietary adjustments.

        - For large, poor hygiene, painful cases: Consider excision.

- **Internal Hemorrhoids:**

    - Grade 1: Dietary adjustments, rubber band ligation.

    - Grade 2: Dietary adjustments, rubber band ligation, excision.

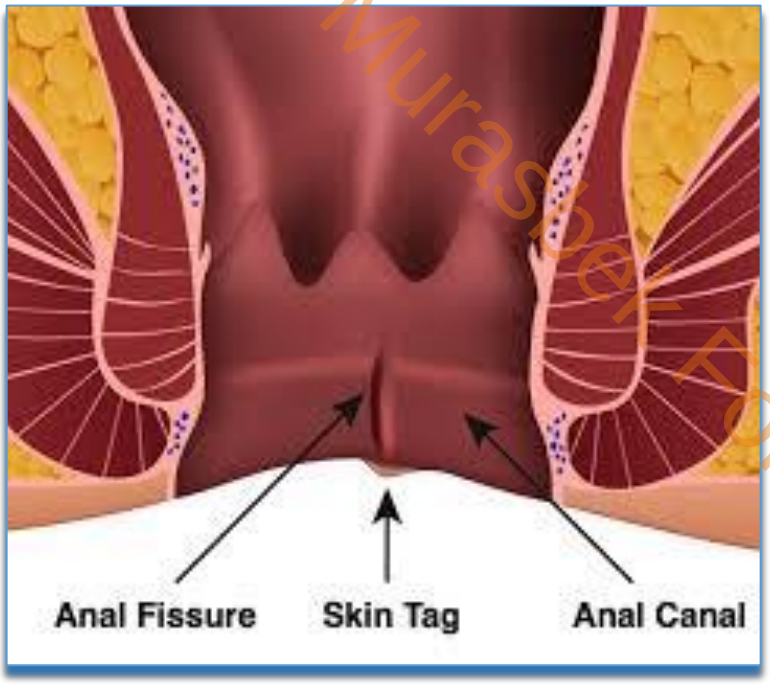
    - Grade 3: Excision, prolapse and hemorrhoid surgery, rubber band ligation.

    - Grade 4: Excision, prolapse and hemorrhoid surgery.

Anal Fissure

- **Definition:** Tears that occur in the anoderm.

- **Common Location**: Usually found in the posterior midline (at 6 o'clock position in the lithotomy position). The remaining fissures are typically located in the anterior midline.



Conditions Where Atypically Located Fissures May Be Seen
- Crohn's Disease
- Epidermoid Carcinoma
- Tuberculosis
- Syphilis
- Leukemic Infiltration
- HIV

- Contributing Factors: Low-fiber foods and constipation are responsible.

- Forms: There are acute and chronic forms.

- **Acute Form**: Characterized by a superficial tear that is very painful. There is induration and edema. The ulcer is small. Dull, dripping-like bleeding is observed after defecation. It responds well to medical treatment.

- **Chronic Anal Fissure** Components: Hypertrophied Anal Polyp,Sentinel Skin Tag (Skin Protrusion),Ulcer

- **Symptoms**:

- Pain During Defecation: This is due to spasms in the internal sphincter.

- Avoidance of Defecation: The patient avoids defecation due to pain, which leads to constipation. Hard stools cause further trauma, creating a vicious cycle that worsens the condition.

-**Clinical Diagnosis**: Easy to diagnose by visual inspection when the gluteal muscles are separated, revealing the fissure.

- Associated Conditions: Anal fissures can be seen with other systemic diseases. Therefore, attention is needed, as these conditions are significant. The most important clues are the

patient's overall clinical picture and atypical fissure location (not in the midline, and possibly multiple).

- **Treatment:**

- Initial Medical Treatment:

- Effective for acute fissures.

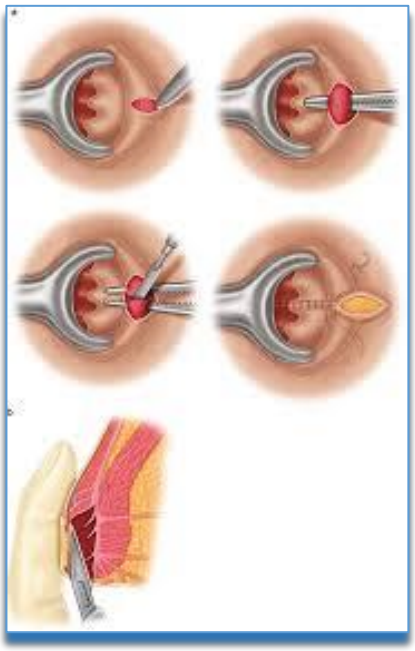
- About 50% of acute fissures heal with medical treatment.

- The main principle is to break the vicious cycle of fissure formation.

- Treatment includes topical anesthetic ointments, high-fiber foods, laxatives, and warm sitz baths.

- Topical nitroglycerin, topical betanecol, and botulinum toxin can also be used.

- Surgical Treatment:

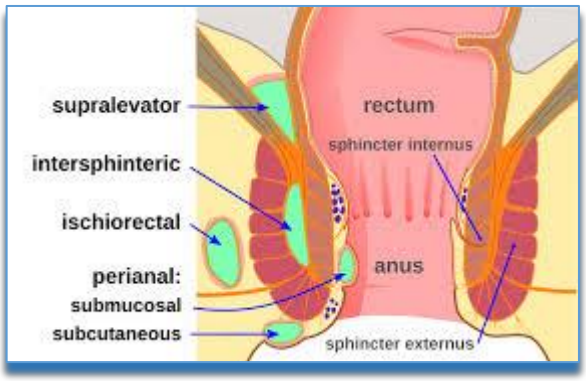


- If medical treatment fails and a chronic fissure is present, surgery is performed.

- Lateral Internal Sphincterotomy:

- Performed in the lithotomy position, cutting the internal sphincter at the 3 o'clock position. The fissure is left untouched to facilitate stool passage and eliminate traumatic effects. The fissure typically heals spontaneously within 10-15 days.

Anorectal Abscesses



- Anorectal abscesses are common.

- They are usually caused by infections in the anal glands.

- They often spread to the intersphincteric fossa.

- Most frequently located in the perianal fossa.

- Infections such as Bartholin's gland abscess, infected sebaceous cyst, pilonidal abscess, and prostatitis can also progress to anorectal spaces.

- **Symptoms**:

  - Severe anal pain is the most common symptom.

  - Fever of unknown origin may occur.

  - Redness is visible in ischiorectal and perianal abscesses.

  - A painful mass that gives fluctuation can be palpated on rectal examination.

  - Pain may increase with sneezing, coughing, and sitting.

- **Treatment:**

  - Drainage is the primary treatment. Performed via skin incision

  - Antibiotics are indicated only in cases of cellulitis, immunosuppression, diabetes, or heart disease.

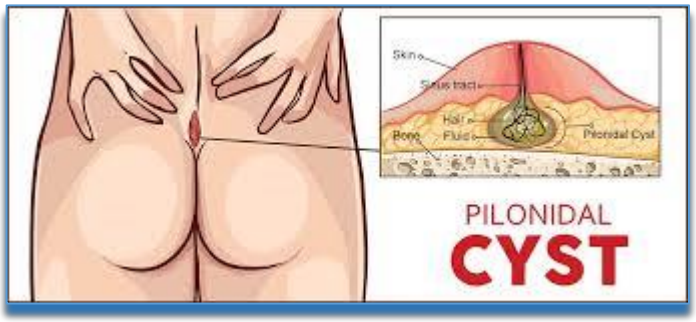
Ischiorectal Abscess:

- Drainage: Performed via skin incision.

Intersphincteric Abscess:

- Symptoms: There is dull pain around the rectum. Swelling and induration may not be detectable. A soft and tender mass can be felt on rectal examination.

- Treatment: Drained via internal sphincterotomy.



Pilonidal Abscess:

- Cause: Infection spreads from an anal gland located in the posterior midline.

- Path: Moves to the ischiorectal fossa, causing bilateral perianal symptoms on both sides.

Supralevator Abscess:

- **Diagnosis:** Difficult, hence important.

- **Symptoms:** The only finding may be fever of unknown origin.

- **Examination:** A mass with tenderness and fluctuation can be detected on vaginal and rectal examination.

- **Treatment:** If originating from an intersphincteric abscess, it should be drained from the rectum. If originating from an ischiorectal abscess, it should be drained like an ischiorectal abscess.

Perianal Fistula:

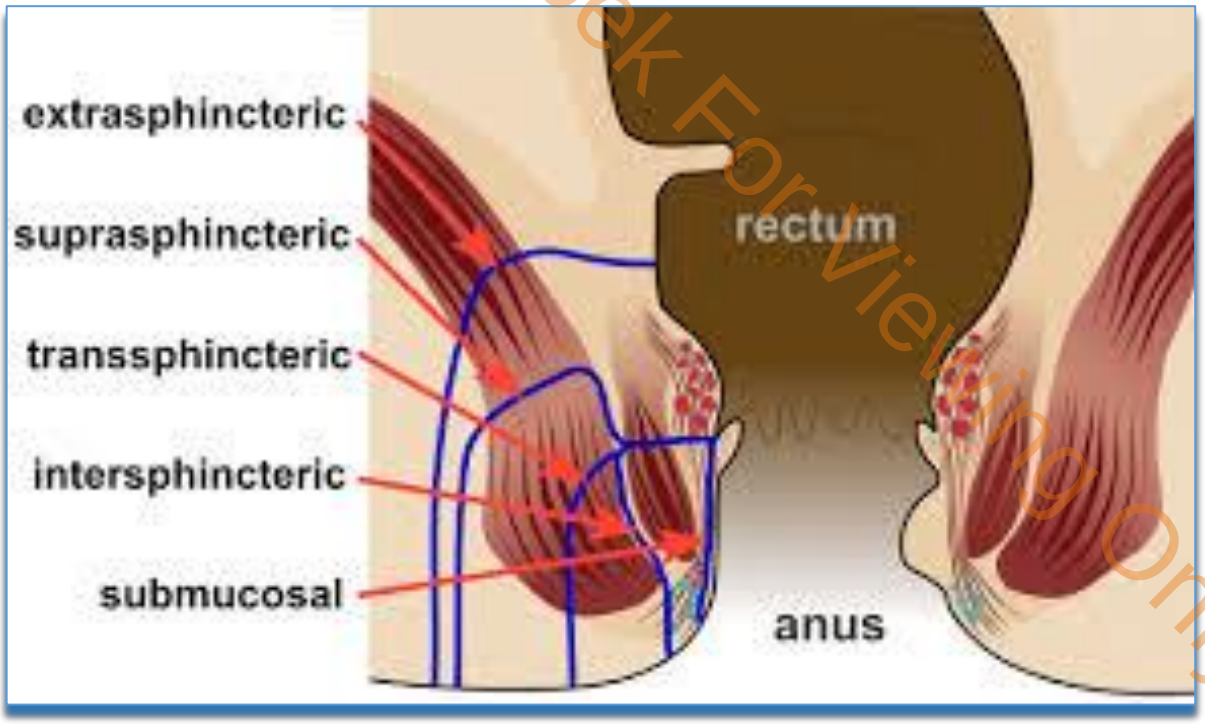
- Cause: The most common cause of perianal fistulas is anorectal abscesses.

- Incidence: About 50% of patients develop a perianal fistula after treatment of an anorectal abscess.

- Other Causes: Perianal fistulas can also result from trauma, Crohn's disease, cancer, radiation, or certain infections (e.g., tuberculosis, actinomycosis, and chlamydia).

- Formation: The fistula is a tract that extends from the anal crypt where the abscess began to the external drainage point.

- Prevention: To avoid the formation of a long fistula tract, the abscess should be drained as close to the anus as possible.



- Types: The most common type of perianal fistula is the intersphincteric fistula.

- Intersphincteric fistulas account for 45% of all anal fistulas, transsphincteric fistulas for 30%, suprasphincteric fistulas for 20%, and extrasphincteric fistulas for 5%.

- Symptoms: The fistula will have continuous purulent drainage from its internal and external openings.

- Detection: The indurated tract is usually palpable, and the internal opening can often be identified through anoscopic examination.

Goodsall's Rule helps determine the location of the internal opening of an anal fistula based on characteristics of the external opening:

- **Goodsall's Rule**: When the anus is divided by a horizontal line passing through its center:

- Fistulas with external openings anterior to this line reach the lumen directly and with a short tract.

- Fistulas with external openings posterior to this line open into the midline posteriorly through a curved tract.

- An exception to this rule is if the external opening is anterior to the line but more than 3 cm from the anus; such fistulas also open into the posterior midline through a curved tract.

- Multiple fistulas open into the posterior midline.

Treatment of Anal Fistulas:

- Principle: The main goal is to eradicate sepsis while preserving anorectal function.

- Simple Fistulas: Treated with fistulotomy, curettage or cauterization of the fistula tract, and allowing for secondary healing.

- Fistulectomy: Involves complete removal of the fistula tract, but it is less commonly used due to the increased risk of sphincter function loss, especially as the amount of removed tissue increases.

- Seton Application: Used for extrasphincteric, suprasphincteric, and high transsphincteric fistulas. In some cases, fecal diversion with a colostomy may be necessary.

Anorectal Malignant Diseases

- Anal Canal Cancers: These are rare and account for about 2% of all colorectal cancers.

- **-Types of Anal Canal Tumors:**

- Tumors affecting the anal margin (distal to the dentate line).

- Tumors affecting the anal canal (proximal to the dentate line).

- Lymphatic Drainage:

- Proximal to the Dentate Line: Lymphatic flow from the anal canal drains upward through the superior rectal lymphatics to the inferior mesenteric lymph nodes and laterally through the middle and inferior rectal vessels via the ischiorectal space to the internal iliac lymph nodes.

- Distal to the Dentate Line: Lymphatic flow typically drains into the inguinal lymph nodes.

Anal Intraepithelial Neoplasia (AIN) (Bowen's Disease)

- Definition: Carcinoma in situ of the anal squamous epithelium.

- **Pathological Terms:** Carcinoma in situ and high-grade squamous intraepithelial dysplasia are synonymous terms.

- **AIN:** Used to describe these lesions, which are precursors to invasive squamous cell carcinoma (epidermoid carcinoma).

- **Etiology:** AIN is associated with human papillomavirus (HPV) infection, particularly HPV types 16 and 18.

- **Epidemiology:** Both AIN and anal epidermoid carcinoma have dramatically higher incidence in HIV-positive homosexual men.

- **Symptoms:** Patients often report perianal itching, burning, or bleeding. They may exhibit chronic irritation or an eczema-like appearance due to itching.

- **Treatment:** Treated with wide local excision.

Epidermoid Carcinoma

- **Types:** Anal epidermoid carcinomas include squamous cell carcinoma, cloacogenic carcinoma, transitional carcinoma, and basoloid carcinoma.

- **Behavior and Course:** These tumors generally have similar clinical behavior and progression.

- **Growth:** They are slow-growing tumors.

- **Presentation:** Typically present as an anal or perianal mass. Pain and bleeding may be present.

- **Diagnosis:** Chronic, non-healing ulcers in the perianal region should be biopsied to rule out squamous carcinoma.

- **Treatment**:

  - Perianal Epidermoid Carcinomas: Treated similarly to squamous cell cancers of other skin areas, as adequate surgical margins can usually be achieved without removing the anal sphincters.

  - **Anal Canal or Sphincter-Invading Epidermoid Carcinomas:** Cannot be locally excised and initially require treatment with chemotherapy and radiation therapy (e.g., 5-fluorouracil, mitomycin C, and 3000cGy external radiation).

  - **Response**: Over 80% of these tumors can be cured with this treatment.

  - **Recurrence**: If recurrence occurs, radical resection (abdominoperineal resection [APR]) is generally necessary.

- **Prognosis:** Metastasis to the inguinal lymph nodes is a poor prognostic indicator.

Verrucous Carcinoma (Giant Condyloma Acuminatum)

- **Nature:** Verrucous cancers are a locally aggressive form of condyloma acuminatum.

- **Metastasis:** They do not metastasize.

- **Treatment:** Treated with wide local excision. In very large lesions, radiation therapy may be applied.

Basal Cell Carcinoma

- **Nature:** Basal cell carcinoma of the anus is a rare lesion.

- **Growth and Metastasis:** It grows slowly and rarely metastasizes.

- **Treatment:** Wide local excision is a treatment option, but recurrence can occur in up to 30% of cases.

- Large Lesions: Radical resection and/or radiation therapy may be required for large lesions.

Adenocarcinoma

- **Nature:** Anal adenocarcinoma is very rare and typically arises from lower rectal adenocarcinomas that spread outward.

- **Origin:** Adenocarcinomas can sometimes originate from anal glands or develop in a chronic fistula.

Paget's Disease of the Anus

- **Nature:** Extramammary perianal Paget's disease is an adenocarcinoma in situ originating from the apocrine glands in the perianal region.

- **Appearance**: This lesion typically appears as a plaque, which may be indistinguishable from Bowen's disease.

- **Histology:** Characteristic Paget cells are observed histologically.

- **Associated Conditions:** This tumor is often associated with synchronous gastrointestinal cancers, so a thorough evaluation of the gastrointestinal system is necessary.

- **Treatment:** Wide local excision is generally sufficient for treatment.

Anorectal Melanoma

- Nature: Anorectal melanoma is rare, constituting less than 1% of anorectal cancers and 1-2% of all melanomas.

- Prevalence: It is the third most common site for melanoma after skin and eye melanomas.

- Symptoms: Patients typically present with pain, bleeding, and a mass.

- Prognosis: The prognosis for anorectal melanoma is poor, with 5-year survival rates of less than 10%.

- Metastasis: Approximately 40% of patients have distant metastases at diagnosis, commonly to the lungs, liver, and bones.

- Early Detection: A few patients present at a localized stage, where potentially curative resection may be possible.

- Treatment: Both radical resection (abdominoperineal resection) and wide local excision are options, with no significant difference in survival rates.

- Recommendation: Due to the morbidity associated with APR, wide local excision is recommended as the initial treatment for localized anal melanoma.

INTESTINAL OBSTRUCTIONS

Intestinal obstruction refers to the partial or complete blockage of the passage of intestinal contents through the gastrointestinal (GI) tract.

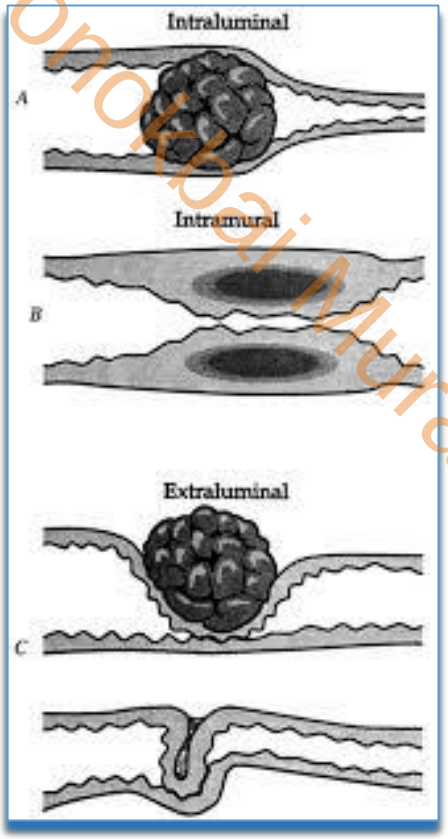
- General: Mechanical small bowel obstruction is the most common surgical disease of the small intestine.

- **-Classification**:

Intestinal obstructions can be classified based on their mechanisms, locations, and the relationship of the obstructing lesion to the intestinal wall.

Based on the relationship of the obstructing lesion to the intestinal wall:

- Intraluminal: Includes obstructions caused by foreign bodies, gallstones, or meconium.



- Intramural: Includes obstructions caused by tumors or inflammatory strictures associated with Crohn's disease.

- Extrinsic: Includes obstructions caused by adhesions, hernias, or carcinomatosis.

- **Common causes of small bowel obstructions**:

- Postoperative adhesions (75%)

- Neoplasms (20%)

- Primary small bowel neoplasms

- Secondary small bowel neoplasms (e.g., melanoma metastasis)

- Local invasion from other intra-abdominal cancers

- Carcinomatosis

- Hernias

- Crohn's disease

- Other causes include intussusception, inflammatory bowel diseases, and various other conditions.

Most Common Causes of Colon Obstructions:

- Colon cancer (60%)

- Diverticulitis (15%)

- Volvulus (15%)

- **PATHOPHYSIOLOGY**:

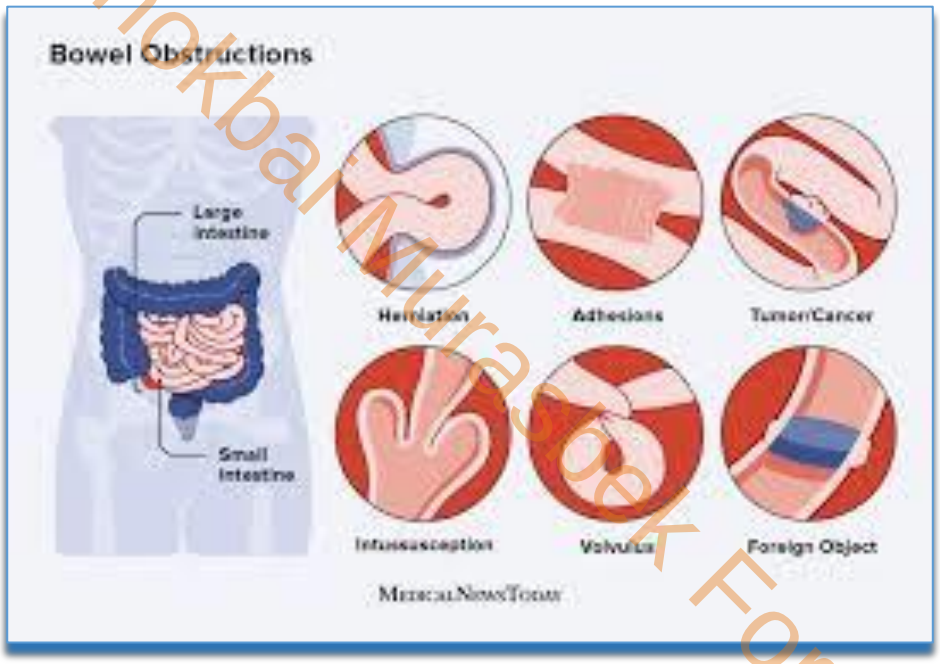
- With the development of an obstruction, gas and fluid start accumulating in the intestinal lumen proximal to the obstruction site.

- Absorption decreases, and fluid is also lost through secretion.

- Vomiting and nasogastric drainage contribute to fluid loss.

- The metabolic effects of fluid loss in intestinal obstructions vary depending on the level and duration of the obstruction.

- In proximal intestinal obstructions, vomiting is more pronounced, and distension is less.



- Gastric fluid rich in acid and potassium is lost.

- Hypochloremia, hypokalemia, and metabolic alkalosis develop.

- In distal intestinal obstructions, fluid loss into the intestinal lumen is greater.

- Changes in blood pH and electrolyte imbalances are not significant.

- However, as dehydration and hypovolemia increase, microcirculation at the cellular level is disrupted, leading to metabolic acidosis.

- Dehydration causes oliguria, azotemia, and hemoconcentration.

- If dehydration is not treated, circulatory disorders, tachycardia, decreased central venous pressure, and decreased cardiac output can occur.

- Hypotension and hypovolemic shock may develop.

- Distension occurs due to the accumulation of gas within the lumen.

- Intestinal distension increases intra-abdominal pressure.

- Venous return decreases.

- The diaphragm rises.

- Breathing may become shallow.

- In the early stages, the intestines increase peristalsis to overcome the obstruction. During this period, hyperactive bowel sounds are heard.

- However, after a while, bowel movements and sounds gradually decrease.

- A silent abdomen develops.

- The increasing edema in the bowel wall eventually disrupts the circulation in the blood vessels supplying this segment.

- This leads to necrosis (strangulation) of the bowel.

Strangulated Intestinal Obstruction

- The disruption of blood circulation to the obstructed segment of the intestine is defined as strangulated intestinal obstruction.

- It is the most serious complication of intestinal obstruction.

- In partial small bowel obstructions, only a portion of the intestinal lumen is blocked, allowing partial passage of gas and fluid distally.

- Therefore, strangulation develops more slowly and is less common compared to complete intestinal obstructions.

Colon Obstruction

- Accounts for 6% of all intestinal obstructions.

- Fluid and electrolyte balance is not significantly disturbed in colon obstructions.

- If the ileocecal valve is competent, the pressure within the colon lumen rapidly increases, compromising mucosal blood flow and integrity.

- If the ileocecal valve is not competent, partial decompression can occur through reflux into the ileum, presenting symptoms of small bowel obstruction.

- When the ileocecal valve is competent, the colon becomes a closed loop, and if untreated, the obstruction can result in perforation.

- According to Laplace's law, the pressure required to distend the wall of a hollow organ is inversely proportional to the radius of that segment; thus, under a certain pressure, the greatest distention in the colon is observed in the cecum, which has the largest diameter.

- Therefore, in colon obstructions, regardless of the level of the blockage, perforation most commonly occurs in the cecum. (Laplace's law)

- **Clinical Presentation of Intestinal Obstructions**

- The four cardinal symptoms of intestinal obstruction:

  - Colicky abdominal pain

  - Nausea and vomiting

  - Inability to pass gas or stool (obstipation)

  - Abdominal distension

- Pain is related to increased intraluminal pressure.

- The interval between colicky pain episodes can help localize the obstruction:

  - In high intestinal obstructions, the interval is 3-5 minutes.

  - In distal obstructions, the interval is 10-15 minutes.

- In complete intestinal obstructions, the passage of gas and stool completely stops.

- In partial obstructions, there may be intermittent passage of gas with small amounts of diarrhea-like stool.

- Vomiting occurs reflexively as soon as the obstruction happens.

- After a delay of a few days, continuous vomiting may begin due to regurgitation of the accumulated bowel contents.

- This vomited content, which has undergone putrefaction and has a foul smell, is referred to as fecaloid. Although dark and thick, it is not actual fecal matter.

-Frequent and bilious vomiting suggests a proximal obstruction, while infrequent and fecaloid vomiting suggests a distal obstruction.

- **DIAGNOSIS OF INTESTINAL OBSTRUCTIONS**

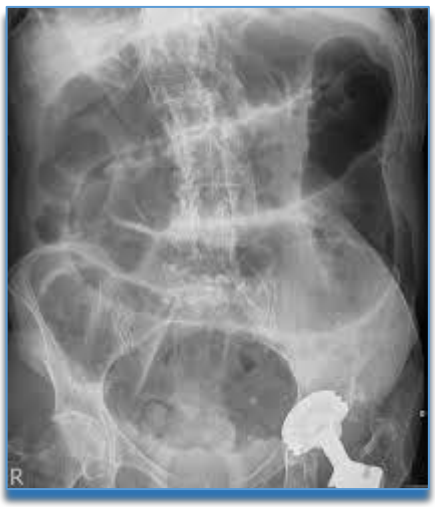
- History: It is important to inquire about previous abdominal surgeries, inflammatory bowel disease, or intra-abdominal cancers.

- Physical Examination: Investigate the possibility of hernias.

- **Laboratory Findings:** Serum amylase levels may be elevated in intestinal obstruction.

- Imaging: The diagnosis of intestinal obstruction is generally made radiologically.

Abdominal X-ray



- Initial Examination: An upright abdominal X-ray is primarily performed. The presence of air-fluid levels on X-ray is diagnostic. The sensitivity of X-ray is between 70-80%.

- Triad of Small Bowel Obstruction on X-ray:

1. Enlarged small bowel loops (> 3 cm)
2. Air-fluid levels
3. Absence of gas in the colon

- In Paralytic Ileus: There is widespread gas distension in the stomach, small bowel, and colon, with multiple air-fluid levels. However, bowel diameters are narrow.

-Additionally, gas is visible in the colon.

-However, it is not always possible to differentiate between paralytic ileus and mechanical intestinal obstruction using X-ray alone.

- Similarly, X-ray may not always distinguish whether the obstruction is in the small bowel or colon.

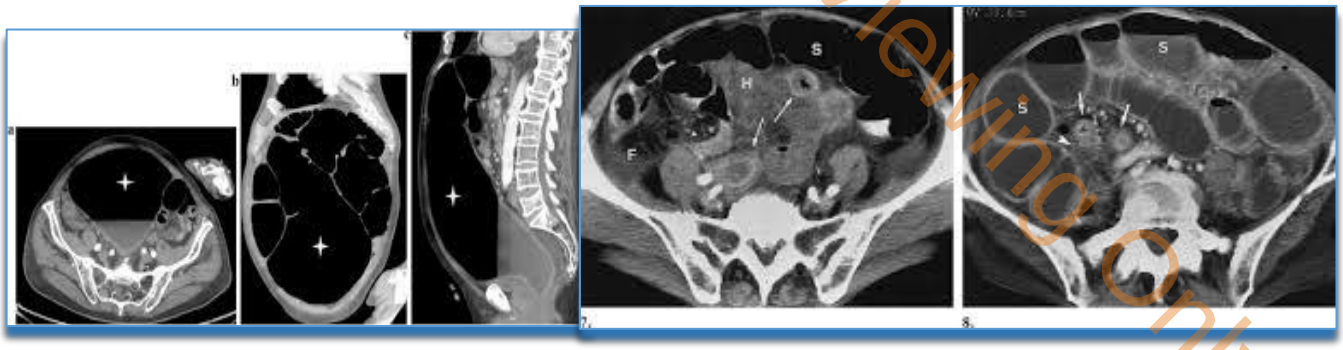
- In colon obstructions, broad-based air-fluid levels are observed.

- There is no gas distal to the obstruction.

- In sigmoid volvulus, the "coffee bean sign" or "omega sign" may be seen.

- Contrast retrograde imaging is contraindicated in the presence of perforation, peritonitis, and advanced cecal dilation.

Computed Tomography (CT)



- CT has a sensitivity of 70-80% and specificity of 70-90% for detecting small bowel obstructions.

- On CT, it can be seen that the bowel is dilated up to a certain point, after which it returns to normal, with the contrast agent not passing beyond that point.

- The presence of air in the bowel wall (pneumatosis intestinalis) and gas in the portal vein on CT may suggest strangulation.

- CT can provide a general assessment of the abdomen and help reveal the cause of the obstruction.

- In cases where CT is inconclusive, contrast-enhanced small bowel follow-through or enteroclysis may be useful.

- Barium can be used as a contrast agent, but in cases with a risk of perforation, a water-soluble contrast medium like Gastrografin is preferred.

- Small bowel follow-through studies have high sensitivity in identifying etiologies of luminal and mural obstructions, such as primary intestinal tumors.

- For enteroclysis, 200-250 ml of barium is administered to the proximal jejunum via a long nasoenteric catheter, followed by 1-2 liters of methylcellulose solution.

- The double-contrast technique allows for better evaluation of mucosal surfaces and visualization of potential small lesions in overlapping segments of the small intestine.

- CT enteroclysis has been reported to have superiority over plain contrast studies.

- **Treatment of Intestinal Obstructions**

- The initial treatment for all intestinal obstructions is fluid and electrolyte therapy.

- A nasogastric (NG) tube should be installed.

- The standard treatment for complete intestinal obstruction is surgical intervention.

- The timing of surgery should be carefully adjusted due to potential metabolic disturbances associated with the obstruction.

- During the period before surgery, it is important to investigate and clarify the etiology and assess the extent of the condition.

- The goal is to operate before necrosis develops.

- In cases of partial obstruction, medical management may be attempted.

- In cases of partial intestinal obstruction due to adhesions, it is known that the majority of patients recover completely within 24-48 hours with medical treatment.

- Therefore, for obstructions caused by adhesions, medical treatment should be administered for 24-48 hours before deciding on further intervention based on the situation.

- **Indications for Emergency Surgery**

- Strangulation

- Closed-loop obstructions

- Colonic obstructions

- Obstructions that do not improve despite medical treatment

ILEUS

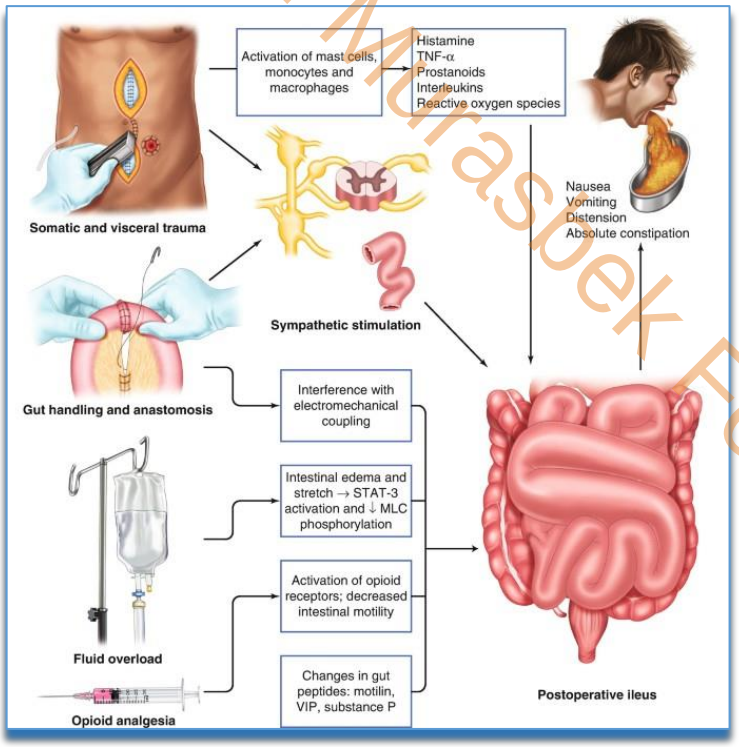
Ileus is a condition where there is impaired or absent bowel motility without mechanical obstruction.

- Most Common Type: Adynamic or paralytic ileus.

- Rare Types: Spastic ileus and ischemic ileus.

Spastic ileus is rarely seen in cases of heavy metal (lead) poisoning and in porphyria.

Adynamic ileus is most commonly observed in postoperative patients. After laparotomy, gastrointestinal motility typically returns in the following sequence: first the small intestine within ~24 hours, then the stomach, followed by the colon.



- **Postoperative ileus** is expected in the immediate postoperative period.

If ileus persists beyond the 3rd-5th postoperative day or occurs without abdominal surgery, diagnostic evaluation should be considered.

In the postoperative phase, if instead of expected improvement, there is noticeable distension, absence of gas or stool passage, nausea, and absent bowel sounds, paralytic ileus is likely.

- Radiological Findings: Numerous small and large air-fluid levels are seen on abdominal X-ray.

Treatment

-In the treatment of ileus, oral intake should be restricted, and any underlying causes of the ileus should be addressed.

-If vomiting occurs or abdominal distension persists, a nasogastric tube should be placed to decompress the stomach. Intravenous fluids and electrolyte treatment should be administered until the ileus resolves.

-If the condition persists, total parenteral nutrition (TPN) may be required.

- Some studies suggest that, if early postoperative feeding is well-tolerated, it can reduce the duration of postoperative ileus, thus shortening hospital stays.

- The use of nonsteroidal anti-inflammatory drugs (NSAIDs) like ketorolac and the concurrent reduction of opioid doses have been shown to shorten the duration of ileus.

- Similarly, thoracic epidural anesthesia/analgesia combined with local anesthesia can reduce systemic opioid doses and shorten the duration of postoperative ileus. However, it does not affect the length of hospital stay.

- Intraoperative fluid restriction can reduce postoperative ileus and shorten hospital stay.

- Many pharmaceutical agents, including prokinetic drugs, are not routinely used due to their efficacy and side effect profiles.

- The use of alvimopan, a peripheral μ-opioid receptor antagonist with limited oral absorption, shortens the duration of postoperative ileus, reduces hospital stay, and decreases readmission rates.

- Enteral feeding rich in fat can reduce postoperative ileus through CCK-dependent vagal reflexes.

- The effectiveness of prokinetic agents like metoclopramide and erythromycin is limited.

- Cisapride has been shown to palliate symptoms but is cardiotoxic.

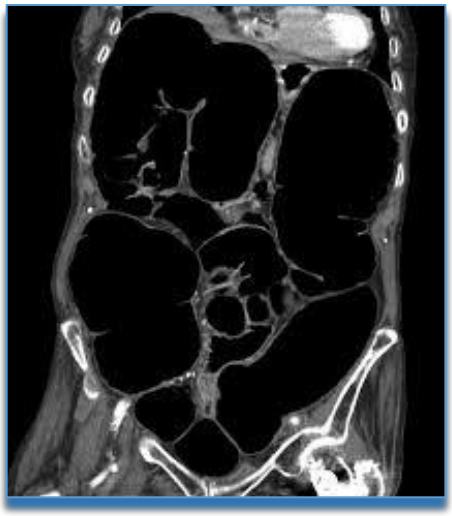
Pseudo-Obstruction

- Pseudo-obstruction is a condition where intestinal obstruction (mechanical) symptoms occur without an organic cause.

- It usually presents as small intestine and colon obstruction.

- Sometimes, motility disorders may affect the entire gastrointestinal system.

- The form observed in the colon is known as Ogilvie's syndrome.



- In Ogilvie's syndrome, there is an imbalance between parasympathetic (stimulatory) and sympathetic (inhibitory) tones.

- It can be seen in patients who are bedridden for long periods, use narcotics, or have comorbid conditions.

- Massive dilation is particularly noted in the right and transverse colon.

- Treatment is primarily medical.

- Intravenous neostigmine is effective.

- Visceral myopathies are a group of disorders characterized by degeneration and fibrosis of the intestinal muscularis propria.

- Visceral neuropathies involve degenerative diseases of the myenteric and submucosal plexuses.

- Systemic diseases such as progressive systemic sclerosis and progressive muscular dystrophy, which affect smooth muscles, or neurological conditions like Parkinson's disease, can complicate chronic intestinal pseudo-obstruction.

- Viral infections, such as cytomegalovirus and Epstein-Barr virus, may also cause pseudo-obstruction.

- The goal of treating chronic intestinal pseudo-obstruction is to palliate symptoms by managing fluids, electrolytes, and nutrition.

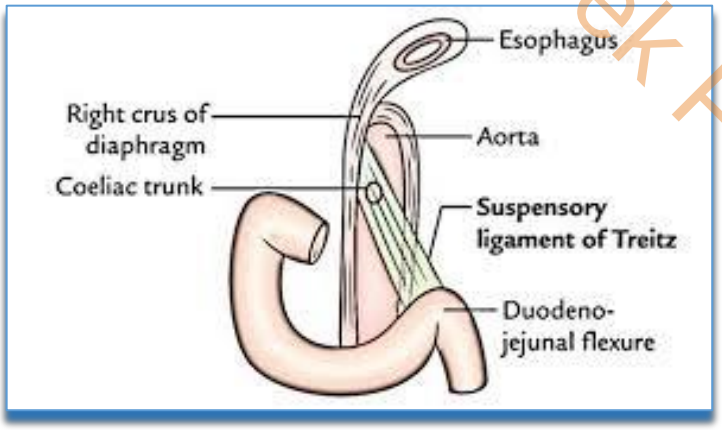
- There are no surgical methods that can prevent or delay the diseases causing intestinal pseudo-obstruction.

Chapter
5.

Gastrointestinal System Bleeding and Surgery

Definitions Related to Gastrointestinal System Bleeding

- -Treitz Ligament: Also known as the ligamentum suspensorium duodeni.



- Bleeding proximal to the Treitz ligament is referred to as upper GI bleeding, while bleeding distal to it is referred to as lower GI bleeding.

- **Hematemesis**

- Hematemesis refers to vomiting blood.

- The blood can appear fresh and bright red or resemble coffee grounds.

- The coffee ground appearance is due to the digestion of blood by gastric acid, indicating slow bleeding.

- When blood remains in the stomach, hydrochloric acid converts red hemoglobin into brown hematin.

- Hematemesis is only seen in upper GI bleeding.

- **Melena**

- Melena is the passage of dark, tarry, and foul-smelling stools.

- In cases of rapid bleeding, the transit time increases, leading to the passage of bright red stools with blood clots.

- Melena often occurs alongside hematemesis, indicating upper GI bleeding.

- Lesions distal to the pylorus can cause melena without hematemesis.

- At least 50 ml of blood loss into the GI tract is necessary for melena to occur.

- Occult blood in the stool can remain positive for up to 21 days following an episode of hematemesis and melena.

- Slow bleeding from the lower GI tract can also result in melena.

- **Hematochezia**

- Hematochezia is the passage of bright red blood in the stool.

- It typically indicates lower GI bleeding.

- In cases of rapid transit and bleeding exceeding 1000 ml, it can also be seen in upper GI bleeding.

- When melena and hematochezia are present, the first step to evaluate the bleeding location is to place an NGT (nasogastric tube) and check the aspirated stomach contents.

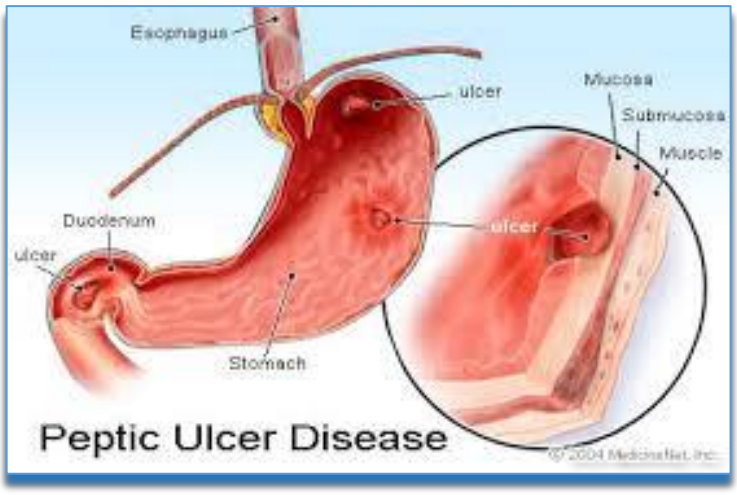
- Bloody content suggests upper GI bleeding, while bile-stained but blood-free content suggests lower GI bleeding.

UPPER GI BLEEDING

- 80% of GI bleedings are upper GI bleedings.

- The most common causes, in order, are peptic ulcer, stress gastritis, acute gastric mucosal lesions, and esophageal varices.

- PEPTIC ULCER



- Causes 40% of all GI bleedings.

- 10-15% of peptic ulcer patients will experience bleeding at some point in their lives.

- Duodenal ulcer bleeding is the most common cause of upper GI bleeding.

- Bleeding ulcers are usually chronic and located on the posterior wall.

- Duodenal ulcer bleedings are four times more common than gastric ulcer bleedings.

- The reason is the proximity to the gastroduodenal artery and the higher frequency of duodenal ulcers.

- **STRESS GASTRITIS/ULCER**

- Causes bleeding from multiple superficial erosions throughout the stomach, most commonly in the corpus (acid-secreting mucosa). Can lead to life-threatening bleeding.

Causes:

- Trauma

- Shock

- Sepsis

- Hemorrhage

- Respiratory failure

- Develop due to impaired blood flow to the gastric mucosa.

- Considered lesions caused by acid and pepsin when gastric perfusion is compromised (NSAIDs can cause similar lesions, but bleeding is not very common in NSAID-induced ulcers).

-Early Lesions: - Develop within the first 24 hours after injury.

- Typically multiple, superficial.

- Bleeding occurs if they progress to the submucosa.

- Early lesions are almost always in the gastric fundus.

- Wedge-shaped mucosal hemorrhages with coagulative necrosis of superficial mucosal cells.

-Late Lesions:

- Develop within 24-72 hours.

- Tissue reaction, inflammatory exudate, and organization around the clot.

- Late lesions resemble healing gastric ulcers.

The two most important factors increasing the risk of bleeding in stress gastritis/ulcer are:

1.Coagulopathy

2. Respiratory failure requiring mechanical ventilation support for more than 48 hours

- Sucralfate may be preferred for prophylaxis (although there is not yet enough evidence).

- Advances in the treatment of shock and sepsis, and the widespread use of acid-suppressive therapy in these patients, have led to a decrease in stress gastritis and its associated bleeding.

- Acid-suppressive therapy is generally successful in treating stress gastritis bleeding.

- If it is not successful (rarely), options include octreotide or vasopressin, endoscopic treatment, and even angiographic embolization.

- Surgical Treatment:

- Vagotomy and pyloroplasty with suturing of bleeding lesions or near-total gastrectomy.

- **Curling's Ulcer**

- Stress Erosions from Burns:

- Develops in burn patients.

- Occurs when the burn area is greater than 30%.

- **Cushing's Ulcer**

- Occurs in Patients with Severe Head Trauma:

- Due to acid hypersecretion.

- Typically solitary.

- Bleeding is rare.

- **ESOPHAGEAL AND GASTRIC VARICES**

Upper GI bleeds in patients with portal hypertension are referred to as portal hypertensive bleeding. This term includes gastroesophageal varices and hypertensive portal gastropathy. Portal hypertensive bleeding accounts for 20% of upper GI bleeds. Of these, 90% are due to gastroesophageal variceal bleeding, with the remainder due to hypertensive portal gastropathy.

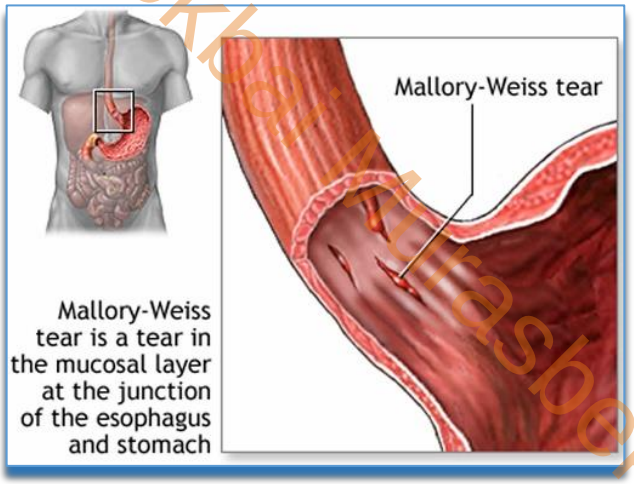
- Variceal Bleeding:

- Most variceal bleeds are massive.

- Many patients have alcoholic cirrhosis.

- It is the cause of upper GI bleeding with the highest morbidity and mortality.

- **MALLORY-WEISS SYNDROME**



- Longitudinal mucosal tears located just below the gastroesophageal junction.

- Develops due to vomiting and retching, often in those who consume excessive alcohol and in pregnant women.

- Mostly resolves spontaneously (90%).

SYMPTOMS AND SIGNS OF UPPER GI BLEEDS

Patient history is extremely important. The pattern of bleeding, pain, and medications should be inquired about. Symptoms are generally due to hypotension. Hemoglobin and hematocrit values are not reliable in the early stages. The first sign in the early stage is postural (orthostatic) hypotension. When the patient moves from a lying to a sitting position, a drop in systolic blood pressure by more than 15 mmHg and in diastolic blood pressure by more than 10 mmHg is observed. This is known as the **Tilt test**. The presence of orthostatic hypotension or a supine blood pressure below 100 mmHg indicates a bleed of more than 750 ml.Azotemia occurs in almost every bleed, especially in esophagogastric variceal bleeds. BUN returns to normal within 3 days after the bleeding stops. The cause of azotemia is bacterial flora. Therefore, azotemia may not be observed in those taking antibiotics as the intestinal flora is affected. The rise in BUN is directly proportional to the severity of the bleeding. If there is hypotension and hypovolemia, azotemia becomes more pronounced due to prerenal and renal failure.

TREATMENT:

- Large-bore catheters should be used to establish venous access, and 4-6 units of blood should be prepared.

- A urethral catheter should be placed to monitor urine output.

- A nasogastric (NGT) tube should be inserted in every patient suspected of upper GI bleeding.

- The only way to rule out upper GI bleeding is to see clear bile-stained gastric fluid from the NGT.

- After diagnosing upper GI bleeding, an endoscopy (esophagogastroduodenoscopy) should be performed first.

- It both diagnoses and offers a chance for treatment. The diagnostic success rate exceeds 90%.

- Barium studies and X-rays are absolutely contraindicated in the diagnostic approach.

- The first principle of treatment for both peptic ulcers and acute mucosal lesions is neutralizing gastric acid.

- Proton pump inhibitors are administered parenterally.

- Washing the stomach with ice-cold water is a useful method.

- Magnesium and aluminum hydroxide antacids are also beneficial.

- Selective arterial catheterization can be used to administer vasopressin or occlusive agents like "gelfoam" to the gastroduodenal artery.

- Photocoagulation can be performed using argon or Nd-YAG lasers.

- If bleeding does not stop despite these treatments, surgical intervention is required.

- Exceeding 4 units of transfusion in 24 hours and persistent bleeding is a clear surgical indication.

- Surgical intervention is based on the primary diagnosis.

Lower GI Bleeding

- These are bleedings originating from any source distal to the Treitz ligament.

- They usually lead to hematochezia or the presence of occult blood in the stool, though they can rarely cause melena as well.

- **Causes of Lower GI Bleeding**

- The most common cause is diverticular disease.

- The table below lists the causes.

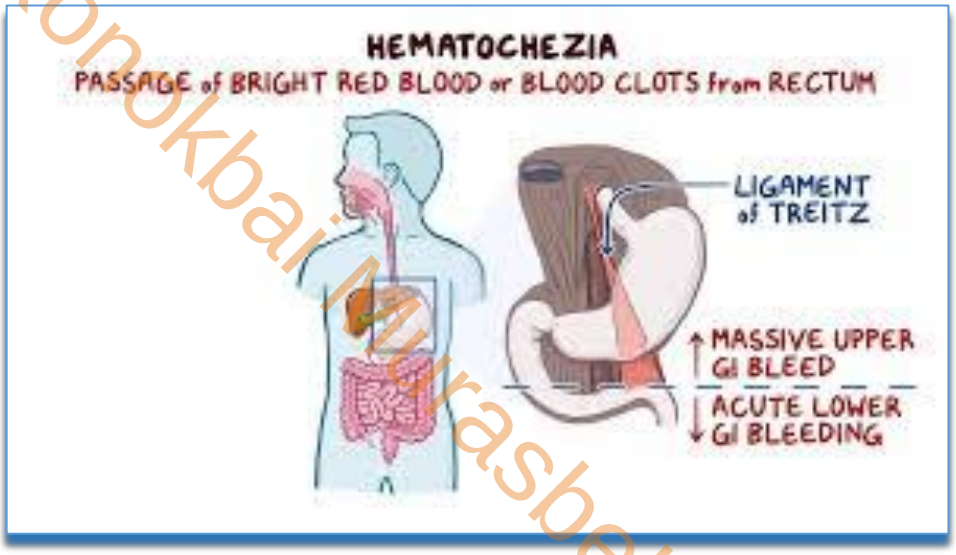
- The most frequent cause of colon-origin bleeding is diverticular disease.

- The most frequent cause of small intestine-origin bleeding is angiodysplasias.

- **Diagnosis**

-History and Physical Examination:

- The nature of the bleeding, any changes in bowel habits, and weight loss are important.



- In hemorrhoid-related bleeding, there is painless, bright red blood on the stool or toilet paper after defecation.

- In large prolapsed hemorrhoids, severe bleeding may occur from the anal canal without defecation.

- Polyps or cancers located distally can lead to bleeding mixed with bright red or slightly darker stool.

- Diverticular disease of the colon generally causes severe bleeding:

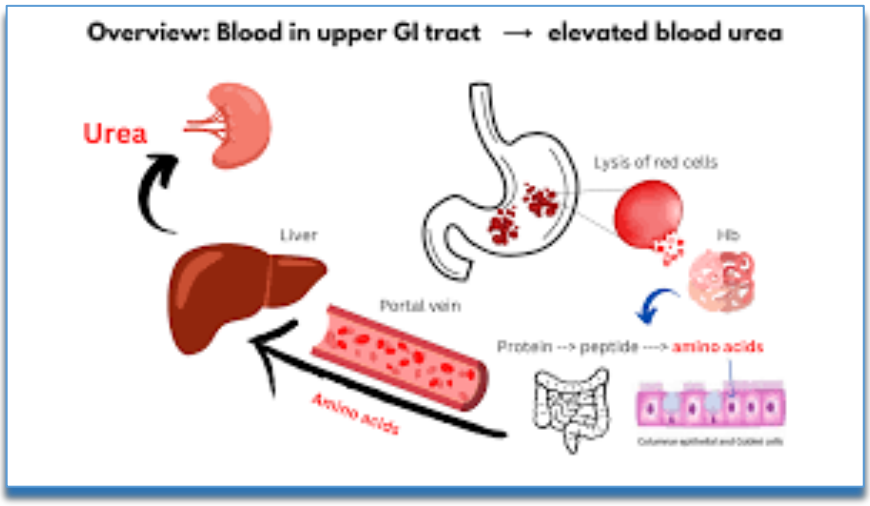
- Typically, a large amount of dark red stool is observed following a sudden urge to use the toilet.

- In ischemic colitis, bloody diarrhea is commonly observed along with abdominal pain felt on the left side.

- Diagnostic Tests

- Hemoglobin, hematocrit, and BUN levels are assessed.

- Coagulation disorders are investigated.



- If BUN > 30 mg/dL within the first 24 hours of bleeding, upper GI bleeding is more likely.

- In patients with hematochezia (bright red blood in stool), the initial step is nasogastric tube (NGT) aspiration to differentiate between upper and lower GI bleeding. (Upper GI bleeding can also present as fresh red blood in the rectum.)

- If NGT aspiration yields clear and bile-stained contents, a diagnosis of lower GI bleeding is made, and the first procedure should be rectosigmoidoscopy (or proctosigmoidoscopy).

-Proctosigmoidoscopy or Anoscopy. Must be performed: - If no bleeding source is found and the bleeding is mild enough to allow for a colonoscopy, then a colonoscopy should be done. In a young, healthy patient, if a bleeding hemorrhoid or anal fissure is found, anoscopy and sigmoidoscopy may be considered sufficient.

-Colonoscopy:- In cases of lower GI bleeding, if the bleeding is mild or has stopped, colonoscopy is indicated. Colonoscopy can also be used to intervene in bleeding lesions.

-Tagged Red Blood Cell Scintigraphy: In cases of non-massive active bleeding, red blood cell scintigraphy can be performed after colonoscopy. Scintigraphy can detect bleeding rates of 0.1 mL/min or faster. Tagged red blood cell scintigraphy with 99mTc has a sensitivity of approximately 97% and a specificity of about 85%.

-Angiography- is the first test to be performed after proctosigmoidoscopy in cases of active and massive bleeding.The bleeding rate can be at least 0.5 mL/min. Additionally, angiography can be used for therapeutic purposes, such as intra-arterial vasopressin infusion or embolization of the bleeding artery.

-Enteroclysis involves placing a nasointestinal tube into the duodenum or jejunum and directly administering contrast material into the small intestine. It is performed with barium under elective conditions.It is the best test for showing small bowel mass lesions and mucosal patterns. Enteroclysis should be performed if colonoscopy, angiography, bleeding scintigraphy, contrast colon and small bowel X-rays, and endoscopies are negative.

-Intraoperative Enterocolonoscopy.In cases of recurrent, massive bleeding where the cause remains unidentified and surgical exploration is being done, intraoperative endoscopy can be utilized.

TREATMENT

- In cases of lower GI bleeding, 75-90% of the bleeding stops spontaneously.

- If the need for blood transfusion exceeds 4 units within the first 24 hours, there is a 50% chance that surgical intervention will be required.

- If the cause of the bleeding has been identified, treatment should be directed towards that cause.

Surgical Indications for GI Bleeding

- Hemodynamic instability despite aggressive resuscitation

- Failure of endoscopic methods to stop the bleeding

- Recurrent bleeding after the bleeding has stopped (when at least two attempts at endoscopic treatment have failed)

- Shock associated with recurrent bleeding

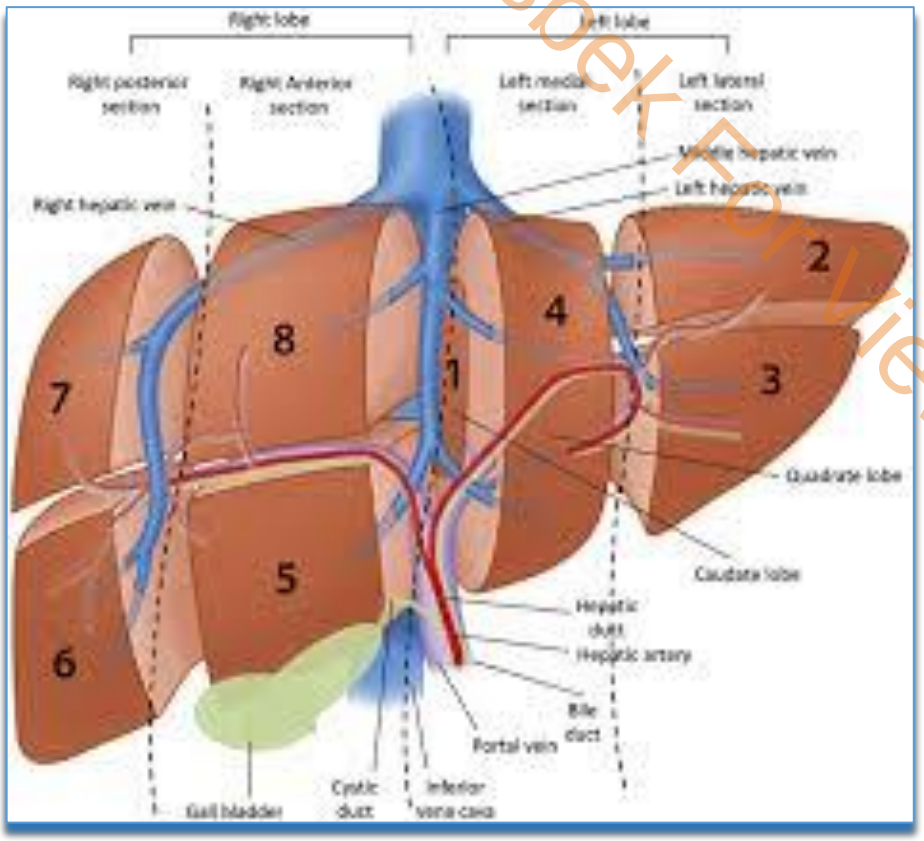
- Ongoing slow bleeding requiring more than 3 units of transfusion per day

Chapter
6.

Liver Diseases and Surgery

- **Liver Anatomy**

- The liver accounts for approximately 2% of body weight.



- It is the largest solid organ in the body.

- The liver is entirely surrounded by the peritoneum, known as Glisson's capsule.

- The right and left lobes of the liver are separated by a plane between the inferior vena cava and the gallbladder bed (Cantlie line).

- The falciform ligament separates the left lateral and left medial segments along the umbilical fissure and suspends the liver from the anterior abdominal wall.

- Based on hepatic venous drainage, the liver is divided into 8 segments.

- Segment 1 is the caudate lobe, which is independent of hepatic venous and portal branch drainage.

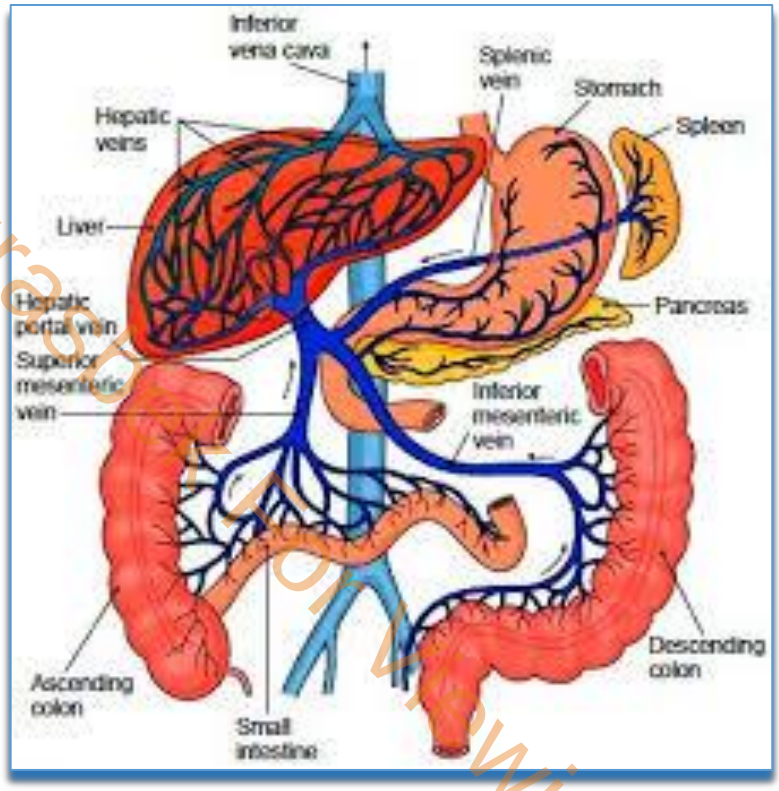
- Segments 2, 3, and 4 constitute the left lobe.

- Segment 4 is also known as the quadrate lobe.

-The smallest functional unit of the liver is the lobule. In humans, there are 50,000-100,000 lobules.

- The potential spaces between liver cells and sinusoids are called "Disse spaces."

- The liver is supplied with blood by 25% from the hepatic artery and 75% from the portal vein. The liver receives 1100 ml of blood per minute via the portal vein and 350 ml per minute via the hepatic artery, which constitutes 25-30% of cardiac output.



- The right hepatic artery branches from the celiac trunk. Exceptionally, in about 10-15% of cases, it may arise from the superior mesenteric artery (SMA) as a replaced or accessory artery.

- Many liver tumors, such as hypervascular lesions, which derive most of their blood supply from the hepatic artery, are well visualized in the arterial phase.

- Because of the liver's blood supply comes from the portal vein, normal liver parenchyma is best visualized in the portal phase.

- The hepatic venous system starts from the central veins of the hepatic lobules and drains into the inferior vena cava through three main veins: right, left, and middle hepatic veins.

- The portal vein is formed by the confluence of the superior and inferior mesenteric veins and the splenic vein, located posterior to the neck of the pancreas. It is approximately 6.5 cm long and averages 8 mm in diameter.

- The portal venous system does not have valves.

- The pressure of blood flow from the portal vein to the liver is 5-10 mmHg.

- The pressure of blood flowing from the hepatic veins into the inferior vena cava is 0 mmHg. In pathological conditions, portal vein pressure can reach 20-30 mmHg.

- The most common cause of increased hepatic vascular resistance is liver cirrhosis.

- The most frequent cause of hepatic congestion is heart failure.

- An increase in hepatic venous pressure leads to enhanced drainage due to excessive permeability of the sinusoids, causing fluid to leak from the liver capsule into the abdominal cavity. This fluid is similar to plasma and is termed ascites.

- Increased pressure in the portal system also leads to edema in the intestines.

- However, the development of collateral vessels helps reduce this pressure and decreases fluid transudation.

- **Functions of the Liver**

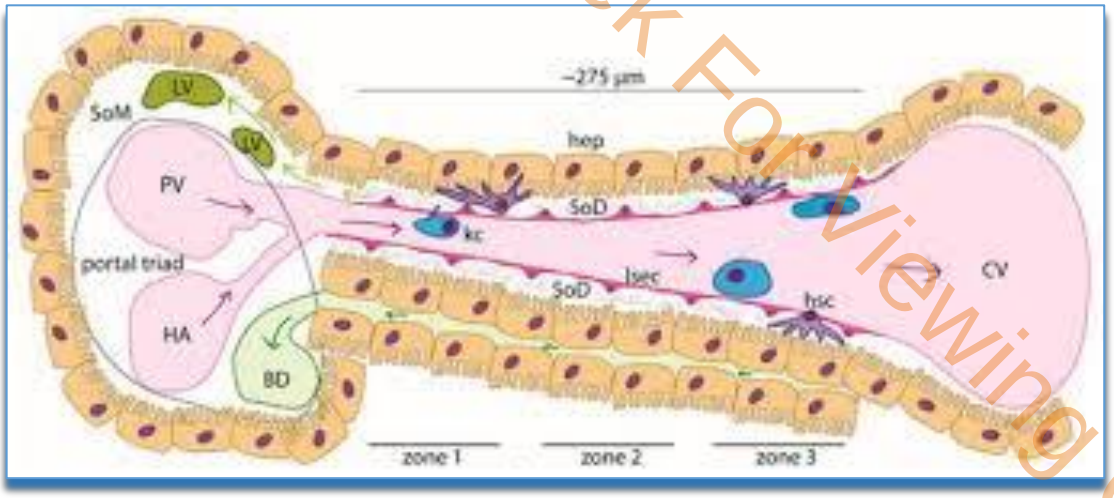
- The liver consists of four physiological anatomical units:

1- Circulatory System

2-Biliary System

3-Reticuloendothelial System: About 60% of this system is in the liver, with Kupffer cells being the most important.

4Hepatocytes: They undertake the body's metabolic load.



Liver Abscesses

- **Pyogenic Abscesses:**

- These are the most commonly observed liver abscesses in developed countries.

- Factors causing pyogenic liver abscesses can be classified by frequency as follows:

- Biliary Tract and Gallbladder Diseases (Ascending Biliary Infection): Includes choledocholithiasis, cholangitis, bile duct strictures, sclerosing cholangitis, and biliary tract, pancreatic, and ampulla of Vater carcinomas, with acute cholecystitis being the most common causes.

- **Cryptogenic Abscesses (Idiopathic):** Abscesses with an unknown cause.

- **Hematogenous Spread (via Hepatic Artery**): Infections causing bacteremia, such as endocarditis, pneumonia, otitis media, and osteomyelitis, can lead to bacterial spread to the liver via the hepatic artery, resulting in pyogenic abscess formation.

- Portal Venous Spread: Intra-abdominal infections like diverticulitis, appendicitis, omphalitis, or pancreatic abscesses can lead to pylephlebitis and subsequent pyogenic liver abscess formation.

- **Direct Spread (via Contiguity):** In subphrenic abscesses, the infection can spread from neighboring regions into the liver, leading to pyogenic abscess development.

- The most common causes are ascending biliary infections (cholangitis) and cryptogenic abscesses.

- Predisposing factors for the development of pyogenic liver abscesses include diabetes, chronic alcoholism, hematological disorders, and steroid dependency.

Abscess Characteristics:

- Pyogenic Abscesses- can be either solitary or multiple.

- Solitary Abscesses- often have a subtle progression.

- Multiple Abscesses- tend to present with an acute and toxic condition.

- Most abscesses are located in the right lobe of the liver.

- The most common pathogen in pyogenic abscesses is E. coli. However, Klebsiella or streptococci can also be involved.

- In 40% of cases, abscesses are monomicrobial; in another 40%, they are polymicrobial; and 20% of cases have negative cultures.

Clinical Signs and Symptoms

- The most frequent symptom is fever. The presentation can be complicated due to the frequent association with other infections.

- Common symptoms include abdominal pain, weakness, loss of appetite, nausea, and vomiting.

- Weight loss, cough, night sweats, and right-sided pleurisy may also occur

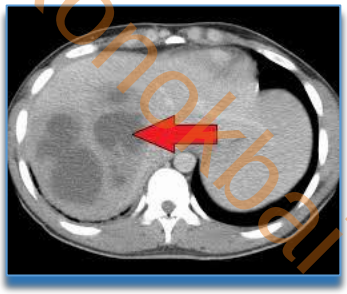
- The most common clinical signs include hepatomegaly (30-60%), liver tenderness, and jaundice if secondary to cholangitis.

DIAGNOSIS

- Laboratory findings typically include leukocytosis (18,000-20,000), anemia (50%), and hypoalbuminemia.

- Liver function tests commonly show elevated levels of ALP, and sometimes GGT.

- Chest X-ray may reveal elevation of the right diaphragm, atelectasis, and pleural effusion.



- The most reliable diagnostic methods are CT (90%), ultrasound (80%), and scintigraphy (70%).

- Ultrasound is the initial diagnostic procedure with an accuracy rate of 85-95%.

- CT and MRI can also be helpful.

- Scintigraphy can detect lesions larger than 2 cm.

- With Tc-99, cold areas are seen, while Ga-67 highlights hot areas.

Treatment

- Treat the underlying cause, administer broad-spectrum intravenous antibiotics, and consider drainage if necessary.

- Antibiotics should be effective against Gram-negative bacteria and anaerobes.

- They should be administered for at least 8 weeks.

- If percutaneous aspiration is performed, cultures can guide more accurate antibiotic selection.

- This approach is expected to be successful in 80-90% of cases.

- If unsuccessful, laparoscopic or open surgical drainage may be required.

- Percutaneous aspiration is often not very effective because the abscess content is very thick and generally cannot be drained percutaneously.

Amoebic Abscess

- Caused by Entamoeba histolytica.

- The ratio of men to women affected is approximately 10:1.

- After invading the intestinal mucosa, the parasite reaches the liver via the portal venous system.

- They typically develop as a single large abscess in the right lobe of the liver.

Clinical Signs:

- Fever and abdominal pain are the most common symptoms.

- The fever does not usually rise as high as in pyogenic abscesses.

- A history of bloody diarrhea is present in about 50% of cases.

- Jaundice is rare.

- Nausea, vomiting, weakness, and weight loss may also occur.

Diagnosis

- In the acute form, anemia is not present, but leukocytosis is pronounced. In chronic cases, anemia is significant and leukocytosis is less pronounced.

- Many patients develop normochromic normocytic or microcytic anemia.

- Leukocytosis and high sedimentation rate are present.

- Liver function tests are generally normal. The most common liver function test anomaly is prolonged prothrombin time (slightly elevated alkaline phosphatase, Schwartz's).

- Chest X-ray may show elevation of the right diaphragm and pleural effusion.

- Diagnosis is confirmed with ultrasound, CT, and MRI.

- Serological tests are used to differentiate from pyogenic abscesses.

- Indirect hemagglutination test is always positive.

- Thick content similar to anchovy paste during drainage is pathognomonic. Diagnosis is generally made after aspiration. The detection rate of amoeba is around 35%.

Complications

- Secondary Infection: The most common complication.

- Rupture: The second most common complication. Abscesses in the left lobe are more likely to rupture into the abdomen. Direct neighboring structures may lead to rupture into the pleura and lung complications. Rupture into the pericardium is the most fatal complication.

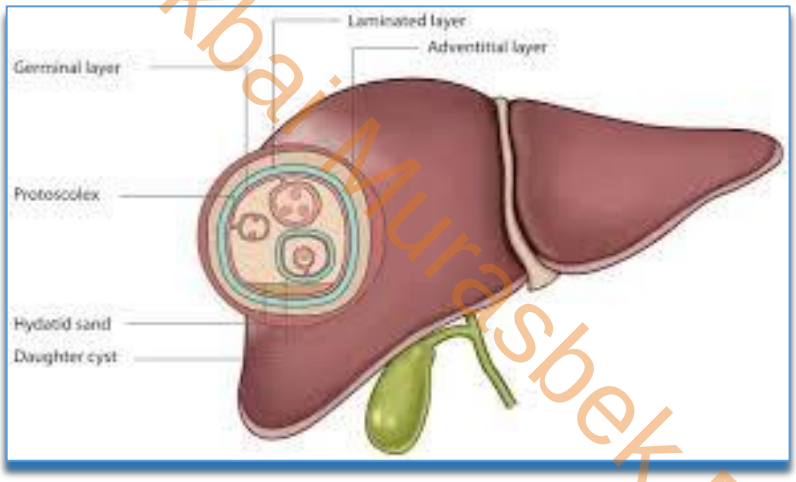
Treatment

- Metronidazole- controls both the intestinal and hepatic phases, administered at 400 mg three times a day.

- Drainage- is rarely necessary.

- Large abscesses, abscesses unresponsive to medical treatment, those with secondary infections, and abscesses located in the left lobe (which could rupture into the pericardium) may require drainage.

Hydatid Cyst



- Two different parasites are responsible for this disease.

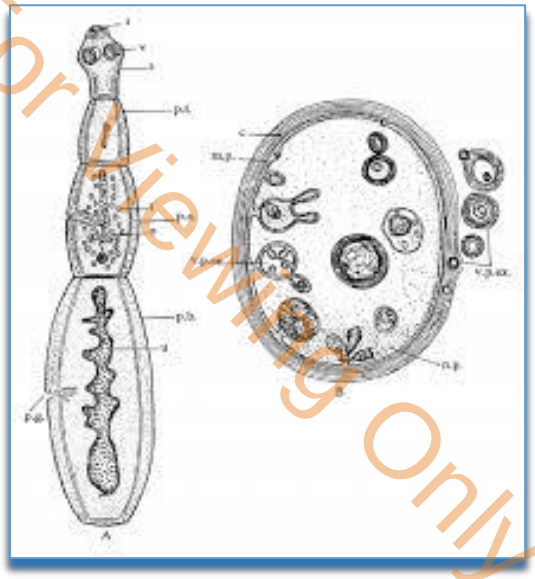
- The most common cause is Echinococcus granulosus.

- The disease caused by Echinococcus alveolaris (E. multilocularis) is hydatid cyst with an alveolar appearance and multiple compartments.

- 70% of hydatid cysts are in the liver.

- Most of these cysts are single and located in the right lobe of the liver.

- The second most affected organ is the lung (10-30%).



Parasitology

- The adult form of Echinococcus granulosus adheres to the intestinal mucosa of carnivorous animals like dogs, foxes, and wolves.

- These definitive hosts appear healthy (primary hosts), but millions of parasite eggs are dispersed through their feces.

- Herbivorous animals like sheep and contaminated vegetables and fruits consumed by humans act as intermediate hosts (secondary hosts).

- In intermediate hosts, the eggs reach the portal venous system. A small portion of the eggs that reach the liver may pass through it and go to the lungs.

- Some of those reaching the lungs may also bypass pulmonary circulation and spread to other organs.

- Inside the cyst, there is a clear, colorless, odorless fluid called "hydatid fluid," containing millions of larvae known as scolices or protoscolices.

- The structure of the cyst is consistent across all organs:

- The outermost layer is the pericyst, which is made of host tissue under pressure and protects the cyst.

- Inside this, a laminar membrane forms the second layer.

- Within this, there is a germinative membrane of a single-cell thickness. Scolices developing from the surface of this membrane fall into the cystic fluid.

- The cyst contains vesicles that can be seen as small, fluid-filled structures.

- As the cyst develops, more hydatid fluid is secreted, and the intraluminal pressure of the cyst can rise up to 100 cm H_2O.

- Hydatid cysts are well-capsulated and therefore do not usually present systemic symptoms such as fever, weakness, or weight loss.

- When the cyst is still within the parenchyma, the laminar membrane can be disrupted due to trauma or the opening of the bile duct into the cyst.

- This disruption can lead to three events:

- The antigenic cyst fluid is absorbed, leading to an allergic reaction characterized by elevated IgE levels, urticaria, and itching.

- Within the pericystic cavity, daughter vesicles are formed. Each daughter vesicle has its own laminar membrane, germinative layer, and scolices. These cysts are referred to as multivesicular cysts, with approximately 50% being multivesicular.

- Small fragments of the disrupted laminar membrane or small daughter vesicles may enter the bile ducts, causing obstructive jaundice.

Complications

- Rupture into the bile ducts: This is the most common complication.

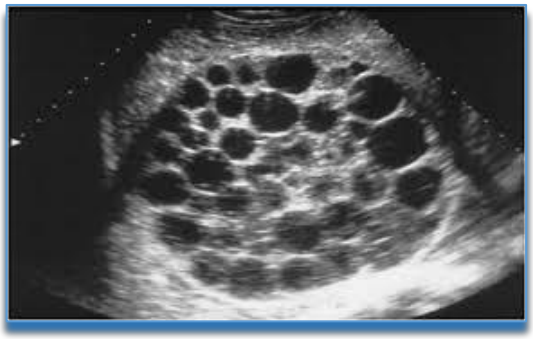
- Secondary infection: This is the second most common complication.

Immunological Methods

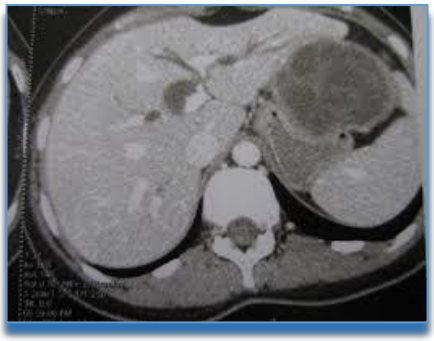
- Detection of echinococcal antigens using the ELISA method, which is positive in more than 90% of infected individuals.

- ELISA (or indirect hemagglutination test - IHA) is the most useful test for screening or epidemiological studies.

USG



CT-scan



Imaging

- Ultrasound (US) and Computed Tomography (CT): Both are equally effective, with a diagnostic accuracy of around 90%.

- Ultrasound: This is the most commonly used imaging method.

Treatment

- Surgical Intervention: The primary treatment for liver hydatid cysts is surgery.

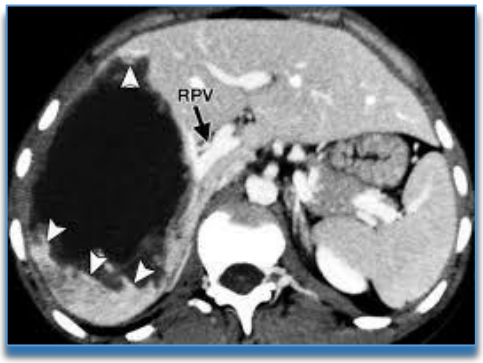
- The main principles include inactivating the parasite, removing the germinative membrane, and obliterating the remaining cavity.

- Partial cystectomy + omentopexy is the most commonly used method.

- Percutaneous Drainage: For selected cases, the PAIR (Puncture, Aspiration, Injection, Reaspiration) method can be applied. After percutaneous aspiration of the cyst, alcohol is injected into the cyst and then reaspirated. Its effectiveness is greater than 75%.

- Preoperative or Tolerable Cases: Albendazole or mebendazole can be used prophylactically or when surgery is not tolerated.

- Small Asymptomatic Cysts: Albendazole can be curative.



Alveolar Hydatid Cyst

- Echinococcus multilocularis causes a more severe form of hydatid cyst.

- No Capsule: The cyst does not have a distinct capsule.

- Invasive: It is invasive and continually progresses.

- Treatment: In complex cases, lobectomy may be performed if limited to a single lobe. In cases of widespread involvement, liver transplantation is considered as a last resort.

Benign Liver Tumors

- Prevalence: Benign liver tumors are found in about 10-20% of people and are much more common than malignant tumors.

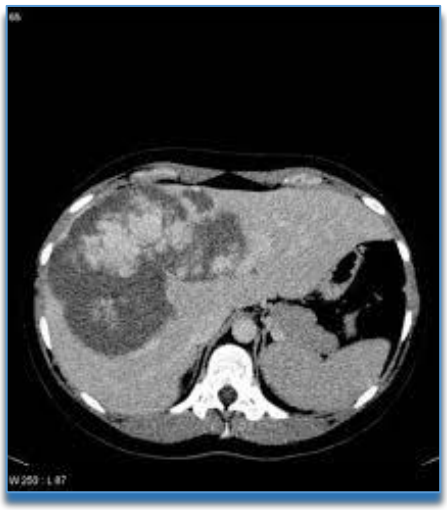
- Most Common: The most frequently observed benign liver tumor is a -hemangioma.

- Other Benign Tumors: Following hemangiomas, the next most common benign liver tumors are -focal nodular hyperplasia (FNH), hepatic adenomas, and mesenchymal hamartomas.

Liver Tumors Classification

1. Benign Tumors
   - Cavernous Hemangiomas: The most common benign liver tumors.
2. Primary Malignant Tumors
   - Primary Epithelial Carcinomas
     - Hepatocellular Carcinoma (HCC): The most common primary malignant liver tumor.
     - Cholangiocarcinoma
     - Hepatoblastoma
   - Mesenchymal Tumors
     - Mixed Tumors: Includes -rhabdomyosarcoma and angiosarcoma.
3. Metastatic (Secondary) Malignant Tumors
   - Colorectal Cancer Metastases: The most common source of secondary liver malignancies.

Hemangioma (benign tumor)



- **Prevalence:** Hemangiomas are the most common solid benign tumors of the liver.
- **Organ:** The liver is the organ where hemangiomas are most frequently observed.
- **Age:** Typically found in adults around 30-40 years old.
- **Gender:** More common in women than in men, with a ratio of about 5:1.
- **Types:** Can be either cavernous or capillary.
- **Malignant Transformation:** There is no risk of malignant transformation.
- **Symptoms:** Most are small and asymptomatic.
- **Effects of Growth:** As they grow, they can cause pressure on surrounding organs and pain.
- **Kasabach-Merritt Syndrome**: Large hemangiomas can consume blood elements(anemia), leading to this syndrome.
- **Platelet Count:** Among benign liver tumors, hemangiomas have the highest likelihood of causing thrombocytopenia.
- **Pregnancy:** Rarely, hemangiomas may grow during pregnancy.

- **Rupture Risk:** The risk of spontaneous rupture is very low.

- **Diagnosis:** Diagnosis can be made using ultrasound (USG) and computed tomography (CT). Magnetic resonance imaging (MRI) with T2 weighting and scintigraphy are also reliable diagnostic methods.

- **Percutaneous Biopsy:** Not recommended.

Treatment

- **Asymptomatic Cases:** Surgery is not required due to the very low risk of bleeding; follow-up is recommended.

- **Indications for Surgery:** The primary surgical indication is pain.

Hepatic Adenoma (benign tumor)

- **Prevalence:** More common in women who use oral contraceptives.

- **Risks with Oral Contraceptives:** Women using oral contraceptives have a higher risk of larger tumors, and the risk of intratumoral or intraperitoneal bleeding is increased.

- **Risk of Hepatocellular Carcinoma:** There is a risk of developing hepatocellular carcinoma in patients with adenomatous conditions.

- **Clinical Presentation:** Hepatic adenomas can present in various ways.

- **Symptoms:** About 75% of adenomas are symptomatic at diagnosis, with pain being the most common symptom.

- **Rupture:** Rupture into the abdominal cavity can lead to intraabdominal bleeding.

- **Percutaneous Biopsy:** Contraindicated.

- **Surgical Planning:** Due to the risk of malignant transformation and rupture, surgical resection is usually planned at the time of diagnosis.

- **Emergency Surgery:** Bleeding constitutes an emergency surgical indication.

- **Transplantation:** In the presence of adenomatous conditions, liver transplantation may be necessary.

Focal Nodular Hyperplasia (FNH)

- **Prevalence:** Mainly seen in young women.

- **Frequency:** It is the second most common benign liver tumor.

- **Characteristics:** Typically presents as a small (less than 5 cm) nodular mass, developing equally in the right and left lobes.

- **Symptoms and Risks:** Unlike adenomas, FNH is typically asymptomatic and has no risk of rupture or malignant degeneration.

- **Relation to Oral Contraceptives:** There is no clear association with oral contraceptive use. It is considered a hyperplastic response rather than a neoplastic event.

- **Detection:** Nearly all patients are asymptomatic. FNH is often discovered incidentally or diagnosed during laparotomy.

- **Imaging:** Diagnosis is typically made using CT or MRI imaging techniques.

- **Biopsy:** Deep biopsy is sufficient once the mass is detected.

- **Histology:** Macroscopically and microscopically, FNH features a star-shaped central scar.

- **Differential Diagnosis:** This characteristic may cause confusion with fibrolamellar hepatocellular carcinoma, a subtype of hepatocellular carcinoma.

- **Treatment:** Conservative management is preferred.

- **Complications:** Resection is performed in the presence of complications.

- **Hormonal Therapy:** Discontinuation of oral contraceptives and estrogen use is recommended.

Liver Malignant Tumors

- **Prevalence:** The most common malignant liver tumors are metastatic tumors.

- Primary Malignant Tumors: Among primary malignant tumors, 80-90% are hepatocellular carcinoma (HCC).

Hepatocellular Carcinoma (HCC)

- **Overview:** Hepatocellular carcinoma (HCC) is the most common primary malignant liver tumor.

- **Gender Disparity:** HCC is 2-8 times more common in men than in women.

Etiology

- Associated Factors: The development of HCC is linked with hepatic viral infections, cirrhosis, genetic metabolic disorders, oral contraceptive use, alcohol consumption, and smoking.

- Hepatic Viral Infections: 75-80% of HCC cases are associated with hepatic viral infections (HBV 50-55%, HCV 25-30%).

- Cirrhosis and Chronic Hepatic Inflammation: 60-90% of HCC cases have cirrhosis and chronic hepatic inflammation, though cirrhosis is not always a prerequisite for HCC development.

- HCV Infection: In patients with chronic HCV infection, cirrhosis is always present before HCC develops. However, HCC may occur without cirrhosis in cases of hepatitis B virus infection.

- Chemical Agents:

- Aflatoxins: Potent hepatotoxins produced by Aspergillus.
- Vinyl Chloride
- Thorium Dioxide (Thorotrast)
- Nitrites
- Hydrocarbons

- **Genetic Metabolic Disorders**

- Hereditary Hemochromatosis
- Alpha-1 Antitrypsin Deficiency
- Wilson's Disease

- Other Factors:

- Androgenic Steroids
- Oral Contraceptives
- Hepatic Adenoma

Clinical Presentation

-Symptoms: Patients with HCC commonly present with right upper quadrant pain, weight loss, and a palpable abdominal mass.

- Acute Presentation: They may also present with sudden onset of bleeding and hypovolemic shock due to intra-abdominal hemorrhage from tumor rupture.

- Additional Signs: Obstructive jaundice, hemobilia, and fever may also be observed.

DIAGNOSIS

- Imaging:

- Ultrasound (USG): Important for screening and early diagnosis.
- CT and MRI: Used for definitive diagnosis and treatment planning. They also help evaluate peritoneal metastases, lymph node involvement, and vascular and biliary invasions.

Tumor Markers:

- Alpha-Fetoprotein (AFP): Useful in diagnosis.

- About 75% of patients with HCC have AFP levels >20 ng/ml.

- A hypervascular mass consistent with HCC and AFP >400 ng/ml is diagnostic.

- AFP is also useful for monitoring treatment and detecting recurrences.

- **Treatment**

- Staging: After diagnosing HCC, the disease stage is assessed to plan appropriate treatment.

- Metastases: HCC commonly metastasizes to the lungs, bones, and peritoneum.

- Survival: The prognosis is influenced by both the tumor and associated cirrhosis.

- Lesion Development: Most HCCs start as a single tumor but can eventually develop multiple satellite lesions due to portal vein invasion and metastases.

- *Surgical Treatment*: The best outcomes are achieved with partial hepatectomy or total hepatectomy combined with liver transplantation to remove the tumor. However, only 10-20% of patients have resectable disease.

- Patient Selection: Choosing the right patient for surgery is crucial. The extent of disease and the condition of the liver both play important roles in determining the outcome.

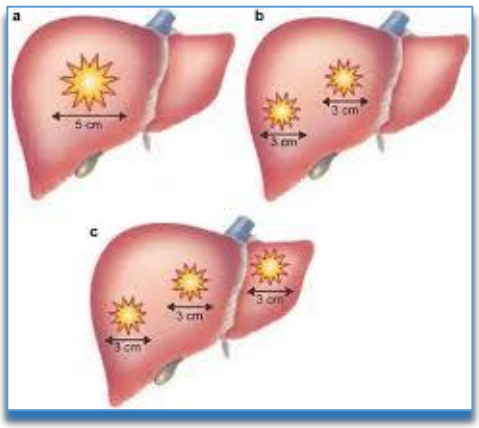
- Child-Pugh Classification:

- Child A: Patients generally tolerate liver resection well.

- Child B, C, and those with portal hypertension: These patients are usually not suitable for resection.

Milan Criteria for Liver transplantation

- For Liver Transplantation: Milan criteria are used to assess eligibility for liver transplantation in patients with HCC , candidates for resection with:



- Single nodule ≤ 5 cm

- Up to 3 nodules, each ≤ 3 cm

- No vascular invasion

- For patients who are not candidates for resection, a range of non-surgical local ablation treatments can be applied.

- Other treatment options include radiofrequency ablation, ethanol ablation, cryosurgery, microwave ablation, chemoembolization, hepatic artery infusion chemotherapy (TACE), Yttrium-90 microspheres, stereotactic radiotherapy, and intensity-modulated radiotherapy.

- Sorafenib, a multi-kinase inhibitor, has been found to have some effect in the treatment of HCC.

Fibrolamellar Hepatocellular Carcinoma

- This variant of HCC is found in younger patients without a history of cirrhosis.

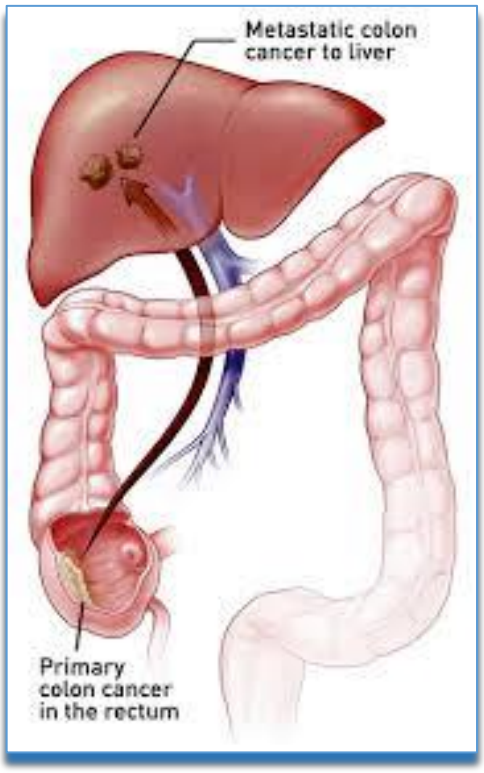
- It is well-defined and encapsulated, with a central fibrotic area. This central scar makes it challenging to differentiate fibrolamellar HCC from FNH.

- In 75% of cases, it presents as a single, large lesion in the left lobe.

- It does not produce AFP but has elevated levels of neurotensin.

- Overall, it has a better prognosis compared to HCC.

Metastatic (Secondary) Malignant Tumors



- These are the most common malignant tumors of the liver.

- Due to portal venous spread, the liver is the organ most frequently metastasized by gastrointestinal tumors.

- Colorectal cancer metastases are the most common type of liver metastasis that surgeons deal with and treat.

Cancers that commonly metastasize to the liver:

- Colorectal cancers

- Upper gastrointestinal tract cancers (stomach, pancreas, bile ducts)

- Genitourinary system (kidney, prostate)

- Neuroendocrine tumors

- Breast cancer

- Skin cancers (melanoma)

- Soft tissue sarcomas (retroperitoneal sarcomas)

- Gynecological cancers (ovary, endometrium, cervix)

Metastatic Spread to the Liver:

- Portal Venous Spread:Colorectal carcinomas, stomach, and pancreatic cancers.
- Lymphatic Spread: Breast and lung carcinomas.
- Hepatic Artery: Lung cancer and melanoma.
- Direct Spread: Gallbladder and bile duct carcinomas, stomach, and colon cancers.

- When cancer metastasizes to a distant site, it is considered a systemic disease, and regional treatments (such as surgery and radiation therapy) are often not effective.**

- Some metastatic liver tumors, particularly colorectal cancer and neuroendocrine tumor metastases, are exceptions to this rule.

- Limited metastatic colorectal cancers to the liver can be resected because they offer the potential for cure and long-term survival.

- CT scans are generally sufficient for imaging liver metastases.

- Intraoperative ultrasound also provides good results.

- Elevated CEA levels may indicate metastatic colorectal cancer.

- While normal liver parenchyma receives 75% of its blood flow from the portal system, primary and metastatic liver tumors primarily receive their blood supply from hepatic artery branches.

- The only current absolute contraindication for metastasectomy is the presence of unresectable disease in the liver.

Poor prognosis in patients with Liver Metastases:

- Presence of extrahepatic metastases.

- Lymph node involvement in the primary colorectal tumor.

- Presence of synchronous tumors.

- Short disease-free interval (<12 months).

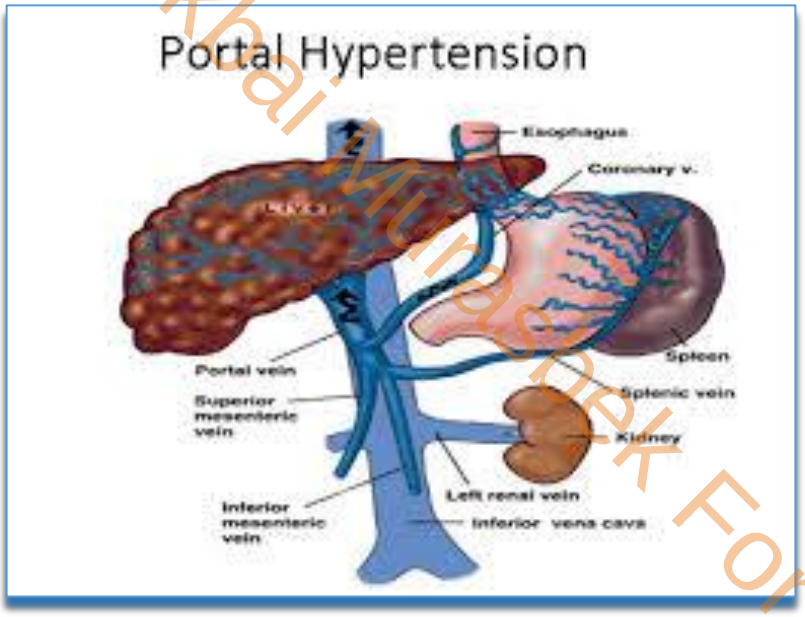
- High number of metastases.

- Bilobar metastases.

- CEA levels >200 ng/ml.

- Largest metastatic tumor diameter in the liver >5 cm.

Portal Hypertension



- The portal system starts with the venous capillary network of the digestive system and spleen, continuing with the portal vein formed by the convergence of the splenic and mesenteric veins.

- It ends in the liver's capillary system known as the sinusoids.

- The portal system supplies 75% of the liver's blood and 72% of its oxygen.

- Normal portal vein pressure is 5-10 mmHg.

- Portal hypertension is defined as a portal vein pressure exceeding 5 mmHg.

- When the pressure exceeds 12 mmHg, esophageal varices may be observed.

- Portal blood is in contact with liver cells in the sinusoids.

- Under normal conditions, not all sinusoids are open; some are collapsed.

- The number of open sinusoids increases proportionally with the amount of blood flowing into them. Portal venous blood partially mixes with arterial blood in the sinusoids, then flows to the central veins, hepatic veins, and eventually to the inferior vena cava.

- There are no valves in the portal system, pressure is equal at all points within the system.

- Increased pressure in the portal venous system is associated with liver disease or anatomical anomalies in the extrahepatic vascular system.

- Pressure increase and congestion in collateral pathways can lead to esophagogastric varices, ascites, hypersplenism, or encephalopathy.

Classification of Etiological Factors in Portal Hypertension:

- Prehepatic:

- Portal vein thrombosis

- Splenic vein thrombosis

- Hepatic arterial-portal venous fistula

- Splenic arterial-venous fistula
- Schistosomiasis

- Intrahepatic:

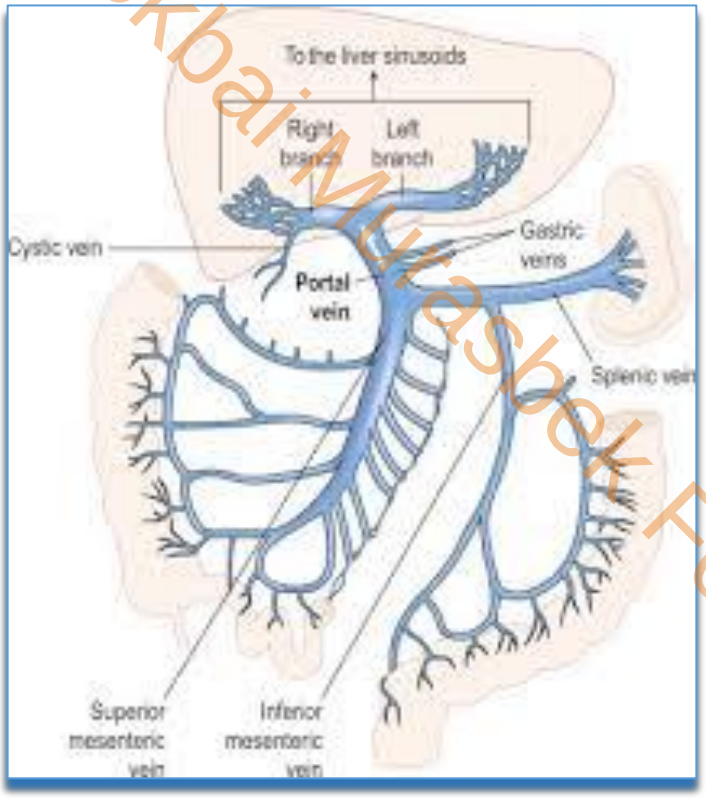
- Pre-sinusoidal:
  - Congenital hepatic fibrosis
  - Idiopathic portal fibrosis
  - Infiltrative lesions (e.g., sarcoidosis, Gaucher's disease)
- Sinusoidal:
  - Chronic active hepatitis
  - Nodular regenerative hyperplasia
  - Graft-versus-host disease
- Post-sinusoidal:
  - Cirrhosis
  - Alcoholic hepatitis
  - Hemochromatosis
  - Wilson's disease
  - Veno-occlusive disease

- Post hepatic:

- Cirrhosis
- Budd-Chiari syndrome
- Constrictive pericarditis
- Congestive heart failure

Collateral Circulation in Portal Hypertension

- **Hepatopedal Collaterals:**



- In the presence of prehepatic obstruction, these collaterals come into play and transport a limited amount of portal venous blood to the liver. Accessory veins of Sappey, Deep cystic veins,Epiploic veins, Hepatocolic and hepatorenal veins

- *Porto-portal anastomoses*: These are connections that attempt to bypass obstructions in the branches or root of the portal vein. Three important anastomoses are:

1. Connection between the splenic vein and the portal vein via the posterior gastric and left gastric veins

2. Splenic-superior mesenteric anastomosis through the gastroepiploic vein (Barkow's arc)

3. Splenic-inferior mesenteric vein anastomoses with pancreatic veins

- **Hepatofugal Collaterals:**

- In the presence of portal hypertension, these collaterals frequently provide flow from the liver to the caval system.

- Also known as **portocaval anastomoses.**

Esophageal Collateral Anastomoses

- Coronary and Esophageal Veins: Anastomoses with the azygos and hemiazygos veins, which then connect to the vena cava.Esophageal variceal bleeding originates from these anastomoses.

Hemorrhoidal Anastomoses

- Superior Hemorrhoidal Vein: Connects with the middle and inferior hemorrhoidal veins, which then connect to the inferior vena cava.

Umbilical Anastomoses

- Umbilical and Paraumbilical Veins: Connect with the superficial abdominal veins, superior and inferior epigastric veins, and then the caval system. Dilation of the paraumbilical veins leads to the "Medusa head" appearance in the abdomen.

- In "**Curvethier-Baumgarten Syndrome**", there is liver atrophy or agenesis. This condition results from the umbilical vein remaining open after birth, leading to reduced blood flow to the liver due to the formation of porto-systemic circulation. Clinically, the expansion of the umbilical vein is observed, and a thrill can be felt upon auscultation.

Retroperitoneal Anastomoses

- Anastomoses between lumbar veins draining into the inferior vena cava and retroperitoneal veins draining into the superior mesenteric vein.These are also known as "Retzius veins".

Splenorenal Anastomoses

- Short Gastric Veins (Vasa Brevia): Connect with the fundal veins of the stomach, esophageal veins, and diaphragmatic veins, and eventually drain into the superior vena cava.

- Spontaneous splenorenal anastomoses can also transport portal blood to the inferior vena cava.

Diagnosis of Portal Hypertension

- **Pressure Measurements:** Differentiates pre- and post-sinusoidal diseases by measuring portal system pressure and hepatic venous pressure (HVPG).

- Radiology: X-rays of the esophagus, stomach, and duodenum, as well as endoscopy, are used to detect varices.

- Splanchnic Arteriography: Used to investigate the etiology of portal hypertension without intra- or extrahepatic obstruction.

- **Doppler Ultrasound:** The simplest initial test is abdominal ultrasound. Doppler ultrasound helps determine the anatomy of the portal vein by detecting thrombosis and assessing the direction of portal venous blood flow.

CLINIC

- Complications include bleeding, ascites, hypersplenism, and encephalopathy.

- Bleeding and hypersplenism occur in all types.

- Ascites is most commonly seen in post-sinusoidal portal hypertension.

- Ascites and hepatic failure are rarely observed in pre-sinusoidal portal hypertension.

- The most significant factor contributing to mortality in portal hypertension is bleeding.

- In post-sinusoidal portal hypertension, ascites can also cause serious problems.

- In children, acute abdominal pain is generally the first sign of portal hypertension.

Complications and Treatment of Portal Hypertension

Bleeding

- The most important cause of upper gastrointestinal (GI) bleeding in portal hypertension is gastroesophageal variceal bleeding.

- Dilation of veins leads to the dilation of the submucosal plexus and thinning of the mucosa.

- Varices can also develop in the stomach's cardia and lesser curvature.

- High pressure in dilated veins and ulceration due to esophagitis are blamed for bleeding.

- About 30% of cirrhotic patients with varices experience variceal bleeding, usually within the first two years after varices are detected.

- Prognosis is worse for variceal bleeding due to cirrhosis, with 70% of patients dying within one year following the bleed.

- Gastroesophageal variceal bleeding accounts for about 15-20% of upper GI bleeds.

- In cirrhotic patients, esophageal varices are not the only cause of bleeding; 20% of patients may have bleeding due to gastritis or duodenal ulcers. Gastric varices can also cause bleeding.

- Due to the specific treatment required, a rapid diagnosis of variceal bleeding is crucial.

- Peripheral signs can be detected through physical examination.

- Esophagoscopy is the most reliable method for diagnosing variceal bleeding.

TREATMENT

Direct Treatment for Bleeding

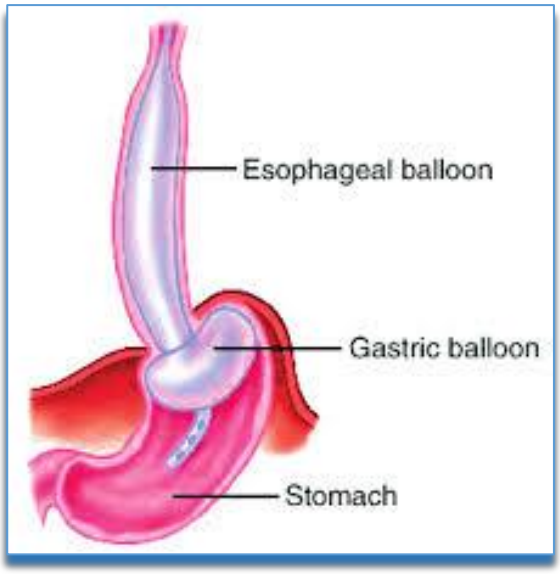
- Control of the bleeding site:

    - Tamponade (with Sengstaken-Blakemore tube):

    - Complications: esophageal rupture, asphyxia, aspiration pneumonia.

- Esophagoscopic sclerotherapy

- Esophagoscopic band ligation: (Also used for bleeding prophylaxis).



Indirect Treatment - Reducing Portal Pressure

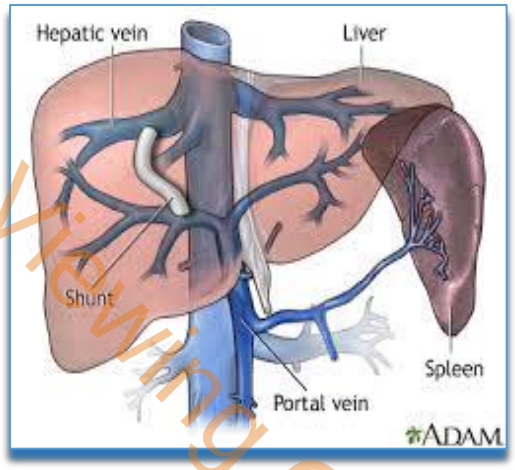
- Vasopressin / Terlipressin: The most effective vasoconstrictor, but its ischemic vasoconstrictive effects can cause hypertension, myocardial ischemia, arrhythmias, ischemic abdominal pain, and gangrene in the limbs, which limits its use.

- Propranolol:Used in acute bleeding and for bleeding prophylaxis.

- Somatostatin / Octreotide: Currently preferred as the first-line pharmacological agent in acute variceal bleeding.

- Short-term prophylactic antibiotic use (e.g., ceftriaxone 1g/day intravenously):Shown to reduce bacterial infection rates and increase survival.

TIPS (Transjugular Intrahepatic Portosystemic Shunt)



- Application as a bridge to liver transplantation in patients with poor general condition.

- Procedure:

- Catheterization of the hepatic vein is performed via the jugular vein.

- A rigid needle is then used to create a path to a large branch of the portal vein.

- A guide wire is inserted, followed by an 8-10 mm angioplasty balloon catheter to dilate the path and create a portosystemic fistula.

- A metal stent is left in the fistula.

- Stenosis occurs in two-thirds of patients post-TIPS.

- Indications for TIPS:

- Bleeding resistant to endoscopic and medical treatment.

- Refractory ascites.

- Budd-Chiari syndrome.

- Hepatopulmonary syndrome.

- Absolute contraindications for TIPS: - Right heart failure.

- Polycystic liver disease.

Surgical Treatment

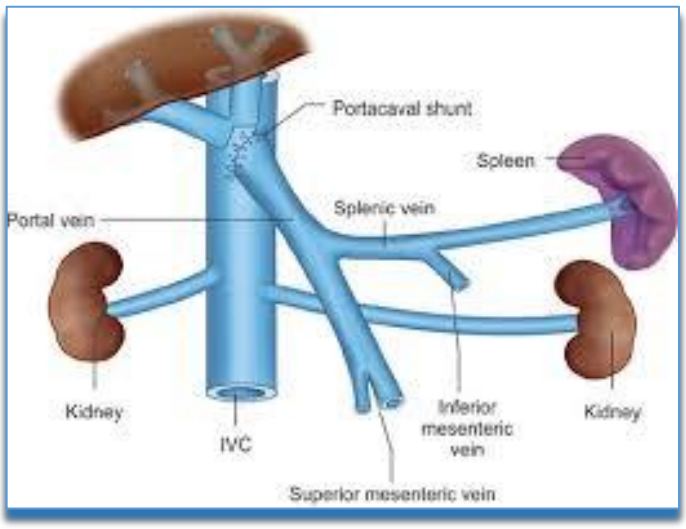
Direct Methods for Controlling Bleeding:

- Transesophageal ligation
- Esophageal transection (using a stapler)
- Devaskularization (Sugiura procedure)
- Gastroesophageal resection: Interposition with colon or jejunum

Indirect Methods for Reducing Portal Pressure:

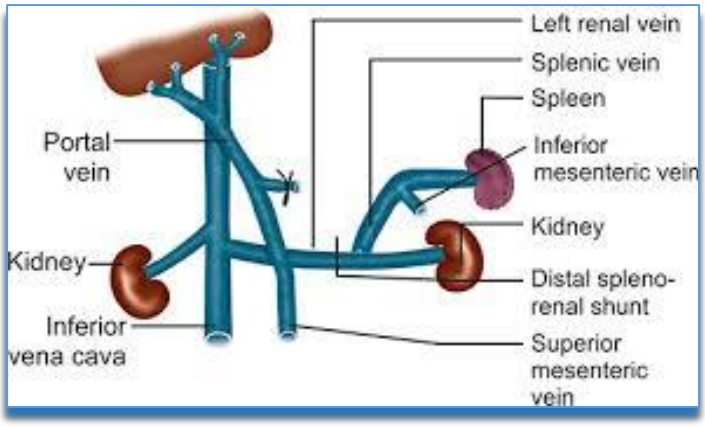
- Splenectomy: Particularly effective in treating portal hypertension due to splenic vein thrombosis.

- Portosystemic Shunts:Aim to reduce portal pressure. An ideal shunt should stop esophageal variceal bleeding, prevent recurrence, not increase the tendency for encephalopathy, and have low surgical mortality. Procedures that completely alter the direction of portal flow have a high risk of encephalopathy, while those that partially change the flow have less reduction in portal pressure.



- End-to-side portocaval shunt: Rarely used due to the high risk of encephalopathy.

- *Side-to-side portocaval shunt*: Best at controlling ascites, preferred more in postsinusoidal portal hypertension.



- *Distal splenorenal shunt (Warren shunt):* - the lowest risk of encephalopathy.Involves anastomosing the distal splenic vein to the left renal vein.

ASCITES

- Ascites is a common complication of portal hypertension and cirrhosis.

- Portal hypertension is a minor factor, and there is no relationship between the degree of portal hypertension and the prevalence of ascites.

- Impaired hepatic venous flow results in increased liver congestion, leading to the dilation of lymphatic vessels and an increase in lymphatic fluid.

- Hypoalbuminemia also contributes by decreasing serum osmotic pressure.

- These pathophysiological events result in intravascular volume loss.

- To compensate for this volume loss, aldosterone secretion increases.

- This causes sodium and water retention, creating a vicious cycle in the formation of ascites.

- Medical treatment is effective in 90% of cirrhotic patients. A high-calorie diet rich in carbohydrates and proteins, along with vitamin supplementation, supports hepatic functions.

- Restriction of sodium intake (2 grams/day) and diuretic therapy are essential.

- Diuretic therapy usually begins with spironolactone since most patients have secondary hyperaldosteronism.

- If symptomatic ascites causing abdominal distension is present, paracentesis may be necessary.

- For ascites resistant to medical treatment, options include paracentesis, TIPS (Transjugular Intrahepatic Portosystemic Shunt), peritoneovenous shunt, or liver transplantation.

- There is no difference in survival between paracentesis and TIPS in patients with advanced liver disease.

- Patients with relatively better liver function may benefit significantly from TIPS.

Hypersplenism

- Hypersplenism is a condition in which the spleen is overactive and removes too many blood cells.

- One of the most common causes of hypersplenism is congestive splenomegaly due to portal hypertension.

- If surgery is needed for hypersplenism in these patients, the first choice is a distal splenorenal shunt.

Encephalopathy

-Encephalopathy is condition that cause brain disfunction.

- Encephalopathy related to portosystemic shunts is characterized by neuropsychiatric symptoms and signs such as changes in consciousness, impairment of intellectual functions, personality changes, and "flapping tremor."

- As neurological disorders progress, coma can also develop.

- Different types of portosystemic shunt surgeries can lead to varying rates of encephalopathy.

- Portocaval shunts have a higher rate of encephalopathy, while distal splenorenal shunts have a lower rate.

-Many of these factors increase **blood ammonia** levels.

- Dietary protein is the primary source of intestinal nitrogen. Additionally, when bleeding occurs, the accumulated blood in the intestine becomes a significant source of intestinal nitrogen.

- EEG is a sensitive indicator of portosystemic encephalopathy.

- In patients with hepatic coma, blood ammonia levels are above 125 ng/dl.

- Treatment aims at correcting facilitating factors and reducing the ammonia and bacterial load in the intestine.

- Daily protein intake (~50 grams) is restricted.

- Glucose in the diet inhibits bacterial ammonia production.

- Bowel emptying is achieved through cathartics and enemas, with lactulose often preferred as a cathartic. Lactulose reduces ammonia absorption from the colon mucosa by lowering the pH of the intestine.

- Non-absorbable antibiotics (such as neomycin) are given to reduce the number of bacteria in the intestine.

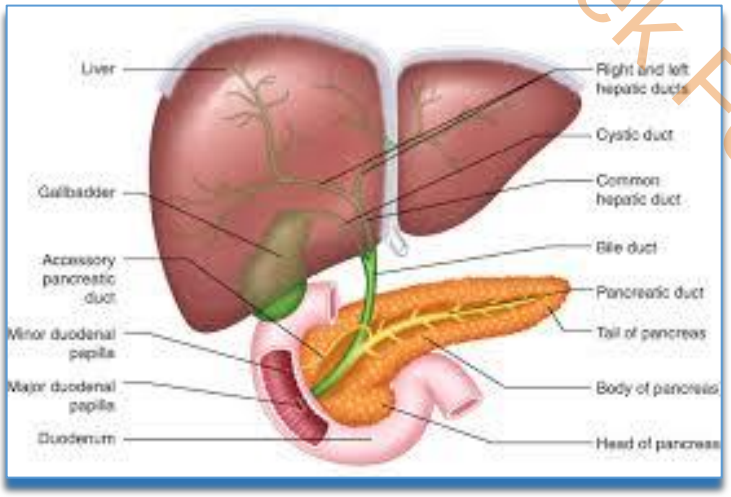
Chapter

7.

GALLBLADDER AND BILE DUCTS DISEASES AND SURGERY

ANATOMY OF THE GALLBLADDER AND BILE DUCTS

- The gallbladder is located beneath the liver, measuring 5-7 cm in width and 7-10 cm in length, with a pear-shaped appearance.



- Its volume is 30-60 cm^3, but it can reach up to 300 mL in case of obstruction.

- The gallbladder consists of four parts: fundus, corpus, neck, and infundibulum.

- The infundibulum of the gallbladder is also known as Hartmann's pouch, where stones are most commonly found.

- The gallbladder mucosa is made up of columnar epithelium. Around the mucosa, there are the muscularis mucosa and serosa. There are mucus-secreting glands in the infundibulum.

- The gallbladder is positioned adjacent to the liver without a mesentery. Luschka ducts are found in this area; these ducts can directly transport a small amount of bile from the liver to the gallbladder in some individuals.

- The bile exchange between the liver and the gallbladder and common bile duct occurs via the cystic duct, which has an internal diameter of 2 mm. An upper limit of 3 mm is considered normal. If larger, it is considered dilated, indicating a stone might have passed through.

- The cystic duct contains spiral mucosal valves known as Heister valves.

- The gallbladder is adjacent to liver segments 4b and 5.

ARTERY AND VEIN OF THE GALLBLADDER

- The cystic artery typically arises from the right hepatic artery in 90% of cases. However, it can also originate from the left hepatic, common hepatic, gastroduodenal, or superior mesenteric arteries.

- In 25% of individuals, there may be more than one artery.

- The cystic artery is located within the hepatocystic or Calot's triangle. The boundaries of this triangle are the cystic duct, the common hepatic duct, and the liver margin.

- The veins usually consist of small veins that directly enter the liver. There is typically one larger vein that drains into the portal vein.

LYMPHATICS

- The lymphatic drainage is directly to the liver, the ganglia around the common bile duct, the ganglia around the pancreas, and the retroperitoneal system.

NERVES

- The nerves come from the celiac ganglion.

- These include sympathetic nerves that travel around the arteries and motor nerves from the vagus.

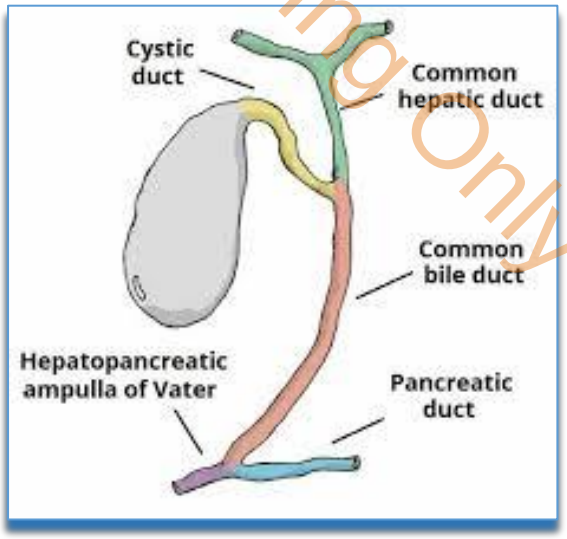
- Sensory nerves are associated with the right side, traveling via the thoracic 8-9 posterior roots through the sympathetic nerves.

- As a result, pain from the gallbladder is felt in the 8th-9th intercostal spaces on the right side and under the right shoulder blade.

BILE DUCTS

Intrahepatic Ducts

- Capillary branches merge and grow, eventually opening into the extrahepatic ducts.



Extrahepatic Ducts

- They start with the right and left hepatic ducts.

- The right hepatic duct is 1 cm long, while the left hepatic duct is 3 cm long.

- The left hepatic duct is longer than the right and expands more in cases of distal obstructions.

- These two ducts join to form the common hepatic duct, which is 1-4 cm in length and approximately 4 mm in diameter.

- The common hepatic duct merges with the cystic duct to form the common bile duct (choledochus).

- The common bile duct is 7-11 cm long and has a diameter of 5-10 mm.

- The common bile duct passes behind the first part of the duodenum and opens into the second part of the duodenum.

- Here, the pancreatic duct also has its opening. Sometimes both ducts merge first and then open into the duodenum, while other times they open separately into the duodenum.

- The common bile duct is divided into three parts: suprapancreatic, infrapancreatic, and intrapancreatic. It can also be extrapancreatic.

PHYSIOLOGY OF THE GALLBLADDER AND BILE DUCTS

- The daily volume of bile is 500-1000 ml.

- The primary function of the gallbladder is to store bile from the liver, concentrate it, and release the concentrated bile into the duodenum in response to food.

- The mucosa of the gallbladder has the highest absorption capacity in the body.

- The filling of the gallbladder occurs due to the tonic contractions of the sphincter of Oddi, which creates a pressure gradient between the gallbladder and the bile ducts.

- Vagal stimulation stimulates gallbladder contraction, while sympathetic stimulation inhibits motor activity.

- Cholecystokinin (CCK) and secretin increase bile flow.

- Cholecystokinin is released into the bloodstream when there is acid, fat, or amino acids in the duodenum, stimulating gallbladder contraction. It also relaxes the sphincter of Oddi and the duodenum.

- In patients who have undergone vagotomy, the response to cholecystokinin stimulation decreases, and gallbladder volume increases.

- VIP and somatostatin relax the gallbladder.

- Bile consists of water, electrolytes, bile salts, bile pigments, fat, and protein.

- Cholesterol and phospholipids constitute the essential components of fats.

- 90% of the phospholipids are lecithin.

- The bile in the gallbladder contains 10 times more bile pigments and cholesterol than hepatic bile.

- Primary bile acids, chenodeoxycholic and cholic acid, form bile salts when conjugated with taurine and glycine.

- Bile salts are separated in the intestines, and primary bile acids are converted into secondary acids, deoxycholic and lithocholic acids, through dehydroxylation. These secondary acids are then reabsorbed from the intestines by passive diffusion (enterohepatic circulation).

Diagnostic Methods for Gallbladder and Bile Duct Diseases

- **Blood Biochemistry**

- When a disease related to the gallbladder or bile ducts is suspected, a complete blood count and liver function tests should be ordered.

- In obstructive bile duct diseases, blood bilirubin levels and alkaline phosphatase(ALP) levels increase.

- Urinary urobilinogen decreases.

- Leukocytosis is observed in inflammatory diseases.

- **Direct Radiography**

- Approximately 15% of gallstones are radiopaque and can be visualized on direct radiography.

- Air can be seen in the bile ducts in biliary-enteric fistulas.

- Sometimes a porcelain gallbladder can be seen on direct radiography.

- **Ultrasound (USG)**

- Today, USG is the first imaging method used for bile duct diseases.

- It is the most commonly used diagnostic method for bile duct diseases.

- It is the most reliable method for diagnosing gallstones (cholelithiasis).

- It shows gallstones with over 95% accuracy.

- It can differentiate between stones and polyps.

- It is useful in diagnosing acute and chronic cholecystitis.

- It can show edema, wall thickening, pericholecystic fluid, hydrops, porcelain gallbladder, polyps, and cancers.

- It is generally the preferred method in the presence of jaundice.

- It can show the gallbladder, liver, bile ducts, and pancreas.

- Biliary dilation, wall thickening, and pericholecystic fluid can be detected.

- It can be used in cases of jaundice and pregnancy.

Situations where the value of ultrasound decreases:

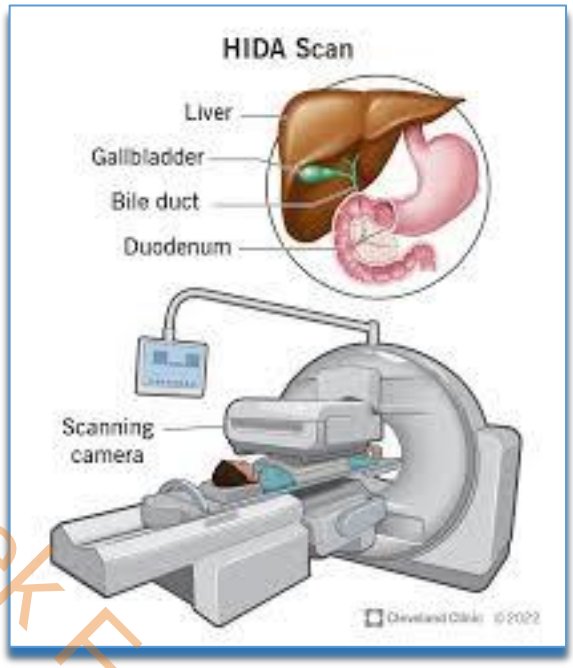
- Excessive intestinal gas

- Obesity

- Presence of ascites

- Recent barium film

- **Biliary Scintigraphy (HIDA / PIPIDA)**



- It is 95% reliable in diagnosing acute cholecystitis.

- It is a scintigraphic imaging method using dimethyl iminodiacetic acid or para-isopropyl iminodiacetic acid (HIDA/PIPIDA) labeled with technetium 99M.

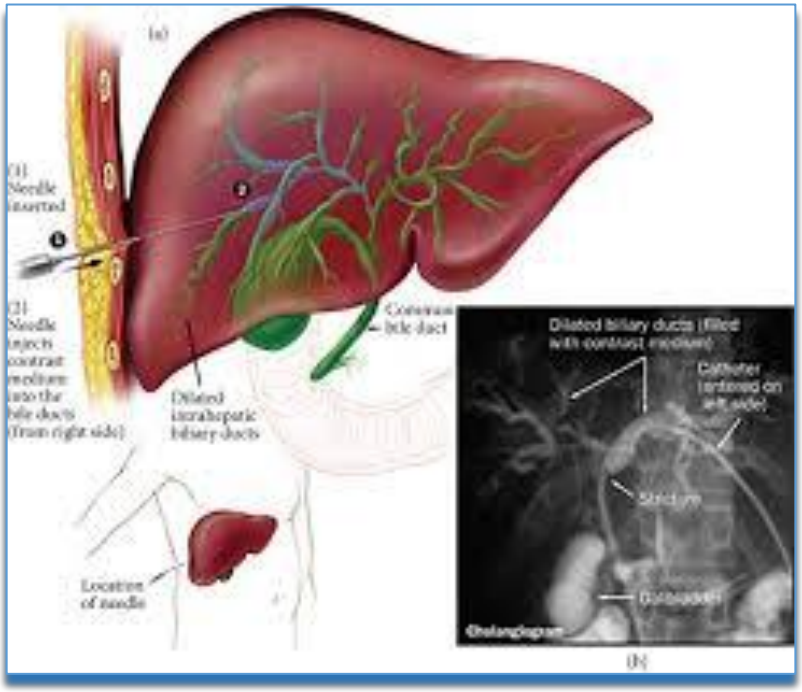
- It is used for diagnosing acute cholecystitis and cholecystopathy.

- **Tomography (CT)**

- It is less successful than USG in showing gallstones.

- It is superior in the differential diagnosis of jaundice and mass lesions.

- **Percutaneous Transhepatic Cholangiography (PTC)**



- It differentiates between mechanical biliary duct obstruction and intrahepatic cholestasis.

- It is performed with a fine needle (CHIBA).

- A dye is injected into one of the intrahepatic ducts.

- Since intrahepatic ducts are dilated in mechanical biliary obstructions, the entry is easy.

- PTC is not performed if the bile ducts are normal.

- It is also not performed in patients with bleeding diathesis.

- The ducts proximal to the obstruction appear dilated.

- Complications such as bleeding, cholangitis, and bile leakage may occur.

- **Magnetic Resonance Cholangiopancreatography (MRCP)**

- It is the only non-invasive method used for direct imaging of the pancreatic duct and bile ducts.

- **Endoscopic Retrograde Cholangiopancreatography (ERCP)**

- It is used to demonstrate bile duct pathologies in patients with jaundice.

- The papilla of Vater is cannulated, and a dye is injected into the pancreatic duct and common bile duct.

- Pathologies related to the bile ducts and lumen can be visualized, stents can be placed, and cytology and biopsy can be performed.

- This method is particularly suitable for lesions in the head of the pancreas or ampulla of Vater.

- Additionally, it is possible to take biopsies or perform cytology studies from these areas using this method.

- ERCP provides both diagnostic and therapeutic capabilities.

- Complications include pancreatitis, cholecystitis, cholangitis, bleeding, and duodenal perforation, with a complication rate of about 10%.

- The most common complication is pancreatitis.

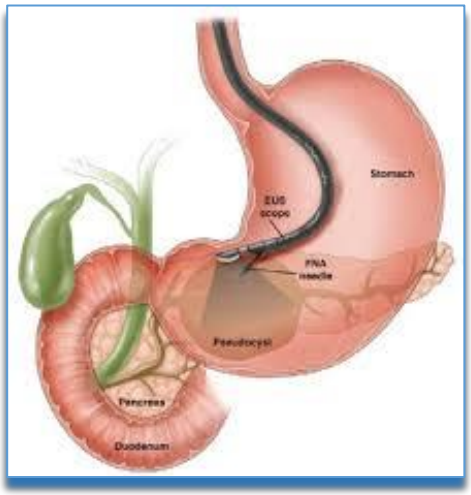
- Mortality is generally 1% or less.

- It is not contraindicated in acute pancreatitis, but the risk of complications is high.

- It cannot be performed on patients who have undergone Billroth II reconstruction due to technical reasons.

- In recent years, endoscopic choledochoscopy or wirsungoscopy can also be performed with small cameras.

- **Endoscopic Ultrasound (EUS)**



- Performed using a special endoscopic probe equipped with ultrasound.

- It is crucial for assessing tumors and resectability.

- It also includes a biopsy probe.

- **Operative Cholangiography**

- Can be performed by accessing the cystic duct during surgery or directly entering the common bile duct or gallbladder.

- Aims to prevent unnecessary exploration of the common bile duct.

Indications for Operative Cholangiography

- Preoperative elevation of AST, ALT, ALP, and bilirubin.

- Preoperative presence of an enlarged common bile duct.

- Jaundice.

- Enlarged cystic duct with small stones.

- Enlarged common bile duct with small stones.

- Preoperative failure of ERCP.

- History of pancreatitis.

- Presence of a single faceted stone within the gallbladder.

- Anatomy that cannot be visualized during laparoscopic dissection.

- Suspected injury to the bile duct during surgery.

- **Operative Choledochoscopy**

- This technique is used in addition to operative cholangiography and exploration of the common bile duct.

- It reduces the incidence of leaving stones behind to about 2.5%.

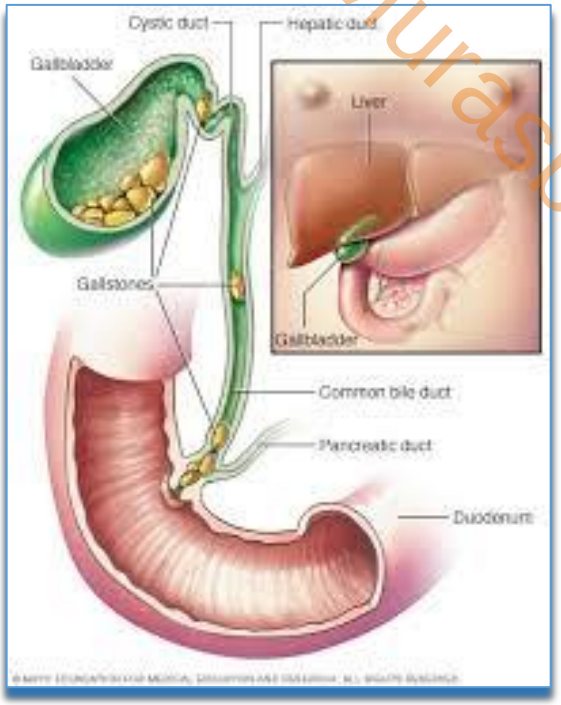
- It is useful for evaluating the functional anatomy of the Oddi sphincter and allows for biopsy collection.

- **Operative Ultrasound**

- Publications suggest that it is more sensitive than operative cholangiography in identifying common bile duct stones.

- It has lower morbidity compared to operative cholangiography as it is non-invasive.

Gallstones



- Gallstone disease is one of the most common conditions affecting the digestive system.

- According to autopsy reports, the prevalence ranges from 11% to 36%. This prevalence depends on various factors, including age, sex, and ethnic group.

- One in four women over the age of forty has gallstones.

- Predisposing factors include obesity, pregnancy, dietary factors, Crohn's disease, terminal ileum resection, gastric surgery, hereditary spherocytosis, sickle cell anemia, and thalassemia.

- Individuals with a family history of cholelithiasis have twice the risk of developing gallstones.

- Factors associated with the development of gallstones are shown in the table.

- Types of Gallstones Based on Formation

- **Cholesterol Stones**: All gallstones can contain cholesterol, but to be classified as cholesterol stones, they must contain more than 70% cholesterol. Their colors can range from white-yellow to green or black. They are typically large and have a smooth surface. Most gallstones are cholesterol stones. Even mixed stones are 50% cholesterol. Therefore, the majority of gallstones are radiolucent.

- However, 10-15% of stones are visible on plain films, and these are primarily calcium bilirubinate stones

- **Pure Bilirubin (Pigment) Stones**: These stones are dark green or black with a smooth surface. They are common in hemolytic diseases such as sickle cell anemia and spherocytosis.

- **Calcium Bilirubinate Stones**: These stones form in the presence of infection or inflammation in the bile ducts and an increase in unconjugated bilirubin.

- **Mechanisms Involved in Gallstone Pathogenesis**

- Conditions such as liver disease, diabetes, obesity, pregnancy, and hypercholesterolemia disrupt the balanced ratio between bile salts, phospholipids, and cholesterol, leading to the formation of cholesterol stones.

- In hemolytic conditions, the increased bilirubin level results in an elevated concentration of bilirubin in bile, causing excess bilirubin to precipitate and form stones.

- High cholesterol levels in bile also contribute to the formation of cholesterol stones.

- Obesity, consumption of high-cholesterol foods, and rapid weight loss are associated with gallstone formation.

- Medications such as clofibrate, which reduce high blood lipid levels, can increase bile cholesterol levels and lead to stone formation.

- Resection of the terminal ileum and terminal ileum pathologies result in the malabsorption of bile salts, disrupting the balance of bile contents and predisposing to stone formation.

- **Clinical Aspects of Gallbladder Stones**

- Most patients are asymptomatic. (Detected incidentally through various imaging methods. About two-thirds of patients with asymptomatic gallstones remain asymptomatic over a 20-year period.Therefore, prophylactic cholecystectomy is rarely necessary for these patients.)

- Mild symptoms may include dyspepsia, gas, and discomfort in the right upper quadrant.

- If stones block the cystic duct, severe pain can occur.

- Various complications may develop within 5-10 years.

- **General Symptoms of Gallbladder Diseases**

- Pain

- Jaundice (icterus)

- Indigestion, abdominal bloating, gas, nausea, and constipation

- Itching

- **Biliary Colic:**

- The most common symptom of cholelithiasis.

- Results from temporary obstruction of the gallbladder (cystic duct), causing severe, cramp-like pain.

- Typically occurs after evening meals.

- Pain is localized to the epigastrium and right upper quadrant and may radiate to the right scapula.

- Nausea, vomiting, and belching may also occur.

- Symptoms resolve if the stone falls back into the gallbladder or moves to the common bile duct.

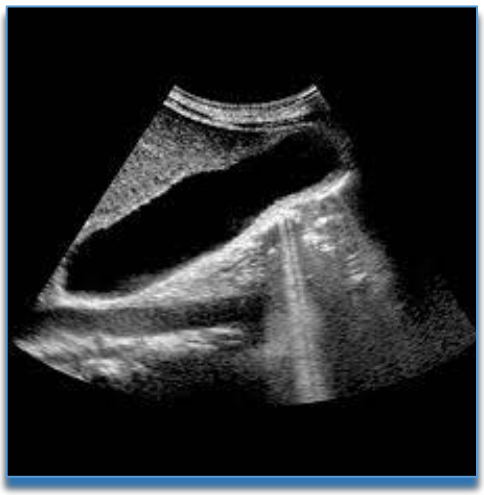
- Recurrent biliary colics are common.

- There is no fever or leukocytosis.

- Liver function tests are usually normal.

- Elective cholecystectomy is performed in such cases.

- **Gallbladder Hydrops**



- Gallbladder hydrops occurs when the gallbladder fills with mucus due to obstruction, provided there is no infection. The bile within the gallbladder is absorbed, and the gallbladder fills with mucus.

- If an infection develops, the gallbladder fills with purulent material, a condition known as empyema.

- Following this, subhepatic abscesses, intraperitoneal abscesses, and subphrenic abscesses may develop.

- The treatment for gallbladder hydrops is an urgent cholecystectomy.

- **Complications of Cholelithiasis**

- Acute cholecystitis

- Chronic cholecystitis

- Choledocholithiasis

- Cholangitis

- Internal biliary fistula

- Pancreatitis

- Gallbladder cancer

Acute Cholecystitis

- **Etiology**

- Obstruction due to gallstones is the most common cause.

- Less common causes include systemic diseases (such as typhoid fever), collagen vascular diseases, and cystic artery thrombosis.

- Inflammation develops as a result of obstruction of the cystic duct by a stone.

- Substances thought to be responsible for the inflammation include lysolecithin, PAF (Platelet Activating Factor), and bile acids.

- **Clinical Presentation**

- Women are more commonly affected, and it is usually seen after the age of 40.

- Acute attacks often occur in individuals with a history of cholelithiasis or chronic cholecystitis.

- Acute symptoms often arise after a heavy and fatty meal.

- Pain in the right upper quadrant can radiate to the back and right shoulder, and while it is typically biliary colic pain, it does not resolve.

- The patient may have fever and severe vomiting, and loss of appetite .

- On physical examination, the right upper quadrant is tender, and **Murphy's sign** may be positive.

- **Murphy's sign** is described as the sudden cessation of breathing when palpating deeply in the right upper quadrant and is characteristic of acute cholecystitis.

- A hydroptic gallbladder may be palpable, and jaundice may be present.

- In differential diagnosis, peptic ulcer perforation or penetration, pancreatitis, hepatitis, appendicitis, right basal pneumonia, myocardial infarction (MI), and herpes zoster should be considered.

- Leukocytosis is present, and there may be an increase in bilirubin and alkaline phosphatase (ALP).

- Amylase levels may also be elevated.

- Elevation in bilirubin levels is mild and does not exceed 4 mg/dl.

- More severe hyperbilirubinemia should prompt consideration of choledocholithiasis or Mirizzi syndrome.

- Electrocardiogram (ECG) and chest X-ray are necessary for differential diagnosis.

- Ultrasound (USG) is the primary diagnostic tool and is the most effective method for diagnosing acute cholecystitis.

- **Treatment**

- For patients suspected of having acute cholecystitis, intravenous fluid replacement, antibiotics, and analgesics are administered.

- Antibiotics should cover gram-negative and anaerobic bacteria. A common combination is a third-generation cephalosporin + metronidazole.

- The definitive treatment for acute cholecystitis is cholecystectomy.

- Laparoscopic cholecystectomy is the preferred method.

- In patients who cannot undergo general anesthesia, cholecystostomy may be performed under local anesthesia.

- **Complications of Acute Cholecystitis**

- **Pericholecystic Abscess:** The most common complication.

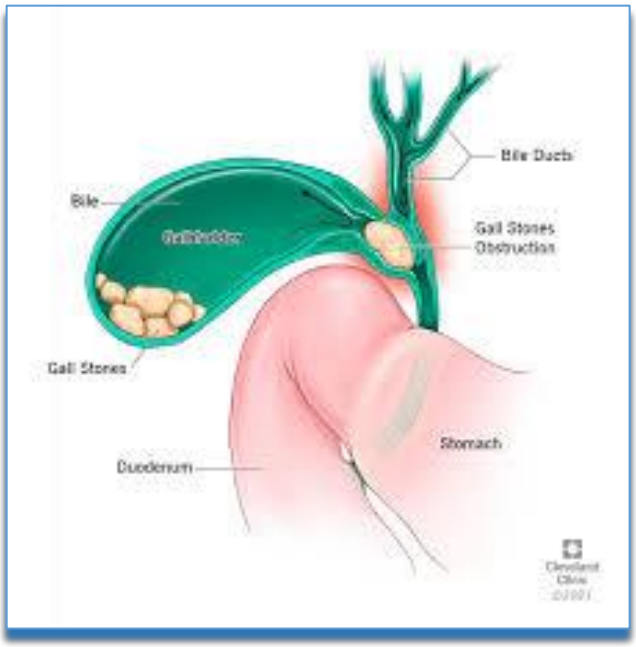
- **Cholecystic Empyema**: Infection of the bile within the gallbladder.

- **Gallbladder Gangrene:** Impaired circulation of the gallbladder wall.

- Gallbladder Perforation.

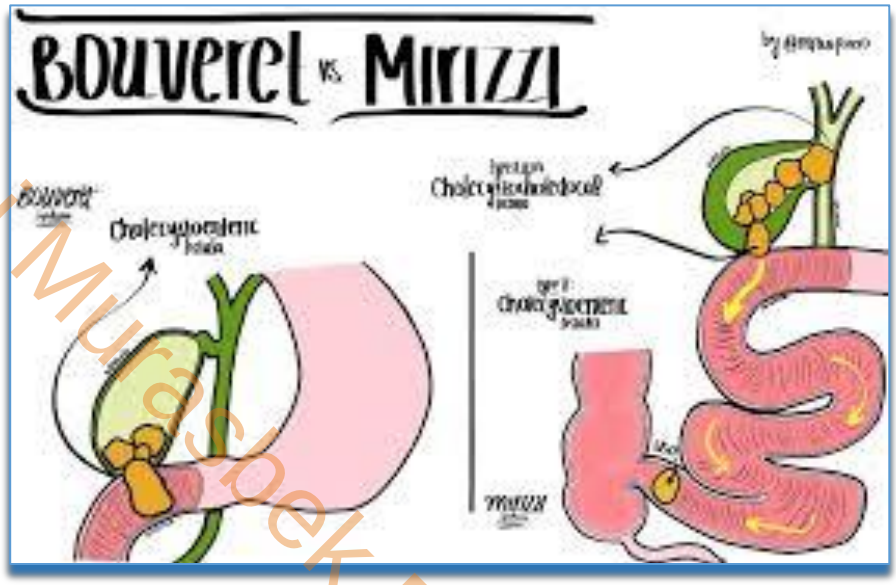
- Internal Biliary Fistula.

• Mirizzi Syndrome



Sometimes, stones within the gallbladder can cause obstruction of ductus cysticus, by applying pressure to common bile duct,without falling into the common bile duct or bile ducts. This condition, characterized by jaundice due to the obstructive effect of the stones, is known as Mirizzi syndrome.

- **Bouveret Syndrome**



This is a condition where a gallstone, passing into the stomach through a cholecystogastric fistula, causes obstruction at the gastric outlet.

- Bilioenteric Fistula and Gallstone Ileus

-Gallstones in the gallbladder can sometimes erode through the gallbladder wall due to inflammation and enter the bile ducts or intestines.

-The most common location for a bilioenteric fistula is the duodenum, with colonic fistulas occurring second most frequently at a rate of about 15%.

-Fistulas often result as a complication of serious inflammation, such as acute cholecystitis or empyema.

-Gallstones most frequently cause obstruction in the terminal ileum.

-History is important for diagnosis; 50-75% of cases have a history of cholelithiasis, while acute cholecystitis or jaundice is seen in 25% of cases. Symptoms may include vomiting, distension, and cramping.

-In abdominal X-rays (ABG), the presence of air in the bile ducts and air-fluid levels in the abdomen should suggest the diagnosis.

-Treatment involves cholecystectomy and closure of the fistula.

Acalculous Cholecystitis

Acalculus cholecystitis is inflammation of the gallbladder without the presence of stones.

- Acute acalculus cholecystitis is often associated with severe systemic illness.

- It commonly arises in conditions such as burns, sepsis, trauma, collagen vascular diseases, multiple organ failure, or in debilitated patients who cannot receive oral nutrition.

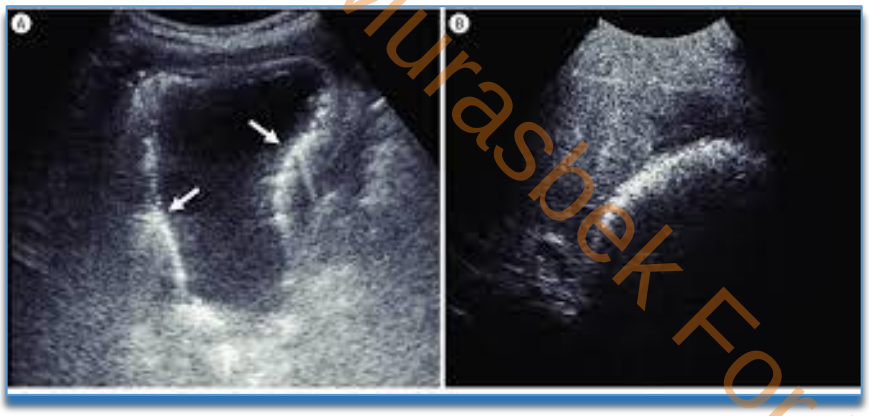
- The chronic form is a type of biliary dyskinesia.

- Ultrasound (USG) shows wall thickening.

- Cholecystectomy is the preferred treatment. If the patient's general condition does not allow for surgery, cholecystostomy is performed.

- In children, cholecystostomy alone is usually sufficient.

Emphysematous Cholecystitis



- Emphysematous cholecystitis is a rare type of acute cholecystitis, often leading to gangrene.

- It is more common in men.

- It involves the formation of submucosal gas pockets within the gallbladder and its wall.

- The clinical presentation is similar to that of acute cholecystitis.

- About half of the patients have cholelithiasis, and many are diabetic.

- Due to the high risk of perforation (40-60%), emergency surgery is required. Antibiotics targeting Clostridia and coliforms are administered.

Chronic Cholecystitis

- Characterized by recurrent attacks on the background of cholelithiasis.

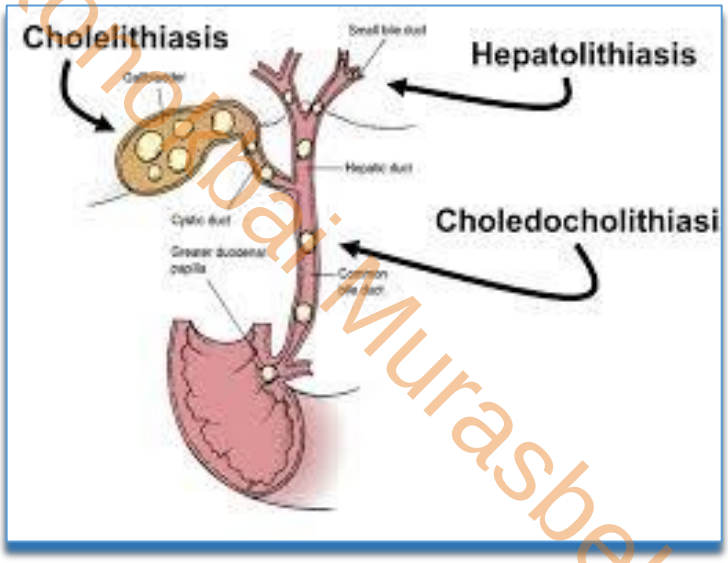
- Features cellular infiltration and fibrosis in the wall.

- The gallbladder is contracted and its wall is thickened.

- Aschoff-Rokitansky sinuses may be present in the mucosa; these result from the extension of the mucosa into deeper tissue layers.

- Cholesterolosis: Results from the aggregation of cholesterol in macrophages within the gallbladder mucosa, forming polyps. The classic macroscopic appearance is described as "strawberry gallbladder."

- Adenomyomatosis or Glandular Proliferative Cholecystitis: Microscopic findings include hypertrophy of smooth muscle bundles and the growth of mucosal glands into the muscle layer (epithelial sinus formation).



Clinic

- The most significant and frequent symptom in patients is pain (biliary colic).

- The pain is variable, moderate in intensity, located in the right upper quadrant or epigastrium; it may radiate to the right shoulder and back.

- The main difference from acute cholecystitis pain is that it is shorter in duration (hours) and less severe.

- Pain lasting more than 24 hours with fever suggests acute cholecystitis.

- History of intolerance to fatty foods and gas is common.

- On physical examination, tenderness is noted in the right upper quadrant during deep palpation.

- **Diagnosis**

- Ultrasound (USG) is the most reliable diagnostic method.

- **Treatment**

- The treatment of choice is laparoscopic cholecystectomy.

- In diabetics, earlier surgery is planned.

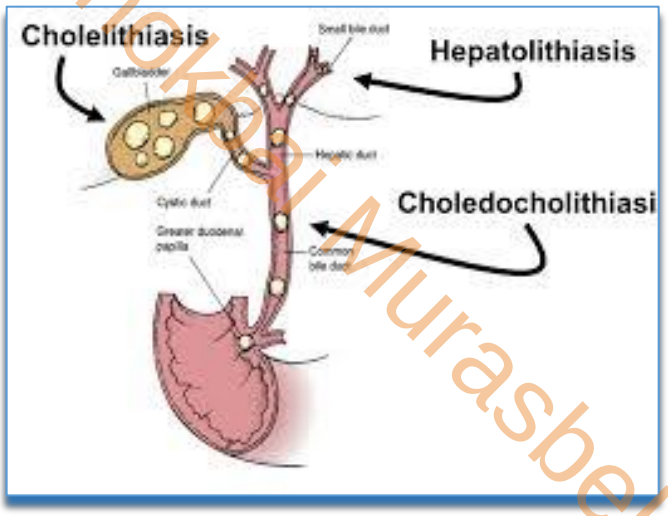
- In pregnant women, laparoscopic cholecystectomy can be performed during the second trimester.

Choledocholithiasis

Choledocholithiasis refers to stones located in the common bile duct, which can be either primary or secondary (originating from the gallbladder). Secondary stones are seen in approximately 90% of cases.

- Primary stones are typically parallel to the axis of the duct and have an elliptical shape.

- They are often soft because they are primarily composed of bilirubin.



Clinic

- Choledocholithiasis can be asymptomatic and is often discovered incidentally.

- It can cause either complete or incomplete obstruction and may lead to cholangitis or biliary pancreatitis.

- Pain caused by choledocholithiasis is similar to biliary colic.

- Signs of obstruction include jaundice, pale stools, and dark-colored urine.

- Nausea and vomiting may occur.

- Physical examination may reveal tenderness in the epigastric or right subcostal areas, in addition to jaundice.

- Prolonged obstruction can lead to dilation of the common bile duct.

- Increased intraluminal pressure results in bile stasis. If an infection occurs, cholangitis can develop, presenting with right upper quadrant pain, fever, and jaundice, **known as Charcot's triad**.

- In cholangitis, bacteria usually ascend from the gastrointestinal tract, but they can also reach the bile duct via hematogenous routes.

- In acute suppurative cholangitis, **Reynolds' pentad is present**, which includes Charcot's triad plus shock and neurological signs.

- Choledochal cysts, tumors, and various interventions can all lead to cholangitis, but the most common cause of cholangitis is choledocholithiasis.

- The risk of developing cholangitis with choledocholithiasis is higher compared to other obstructive lesions.

- The most frequently encountered pathogen secondary to bile stasis is *E. coli*. Broad-spectrum antibiotics should be used.

- Recurrent obstruction and infections can lead to long-term biliary cirrhosis.

- The initial diagnostic approach should be ultrasound (USG).

- In patients with choledocholithiasis, the presence of jaundice and biliary colic attacks along with a common bile duct diameter > 8 mm on USG suggests choledocholithiasis.

- Magnetic Resonance Cholangiopancreatography (MRCP) is also very helpful in diagnosis.

- However, the most reliable diagnostic method is Endoscopic Retrograde Cholangiopancreatography (ERCP).

- **Treatment**

For patients diagnosed with or suspected of choledocholithiasis, the next step is Endoscopic Retrograde Cholangiopancreatography (ERCP).

- Stones can be removed by performing sphincterotomy.

- The initial treatment for cholangitis includes intravenous antibiotics and fluid resuscitation.

- Intensive care monitoring and vasopressor support may be needed. Most patients respond well to these treatments.

- Once the obstructed bile duct is stable, it should be drained.

- Approximately 15% of patients do not respond to antibiotics and fluid resuscitation; urgent biliary decompression may be required.

- Biliary decompression can be performed endoscopically, percutaneously (via percutaneous transhepatic approach), or surgically.

- The choice of method depends on the level and type of biliary obstruction.

-For obstructions caused by choledocholithiasis or periampullary tumors, endoscopic approaches are most effective. Sphincterotomy and placement of an endoscopic biliary stent are performed.

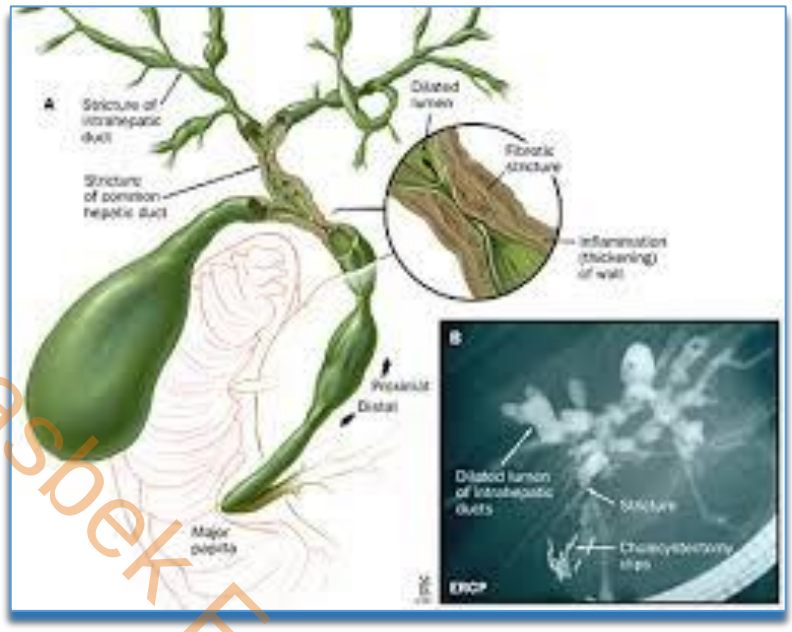
- If the obstruction is proximal or perihilar, or if there is suspected stricture in a biliary-enteric anastomosis, or if endoscopic intervention fails, percutaneous transhepatic drainage is applied.

-If neither ERCP nor percutaneous intervention is possible, emergency surgery with T-tube drainage of the common bile duct may be life-saving.

-Definitive surgery is considered once the cholangitis has resolved.

- The mortality rate for acute cholangitis is around 5%.

Sclerosing Cholangitis



- **Associated Conditions:**

- Ulcerative colitis

- Crohn's disease

- Riedel's thyroiditis

- Retroperitoneal fibrosis

- Porphyria

- **Etiology:**

- The cause is unknown, but atypical immune responses are suspected.

- **Characteristics:**

- Thickening of the bile duct walls leads to narrowing.

- Characterized by strictures and dilatations in the bile ducts.

- **Complications:**

- Increased risk of developing cholangiocarcinoma.

- **Clinical Presentation:**

- Generally occurs between ages 30-45 and is twice as common in men.

- Patients present with recurrent right upper quadrant pain and jaundice.

- Can progress to biliary cirrhosis and complications, potentially leading to portal hypertension.

- In patients with ulcerative colitis and impaired liver function tests, sclerosing cholangitis should be considered.

- **Diagnosis:**

- Imaging and ERCP (endoscopic retrograde cholangiopancreatography) with sphincterotomy and drainage are performed.

- Common bile duct strictures are observed, often described as a "string of beads" appearance.

- Definitive diagnosis is made through liver biopsy, which shows normal mucosa but intense fibrosis in the submucosa.

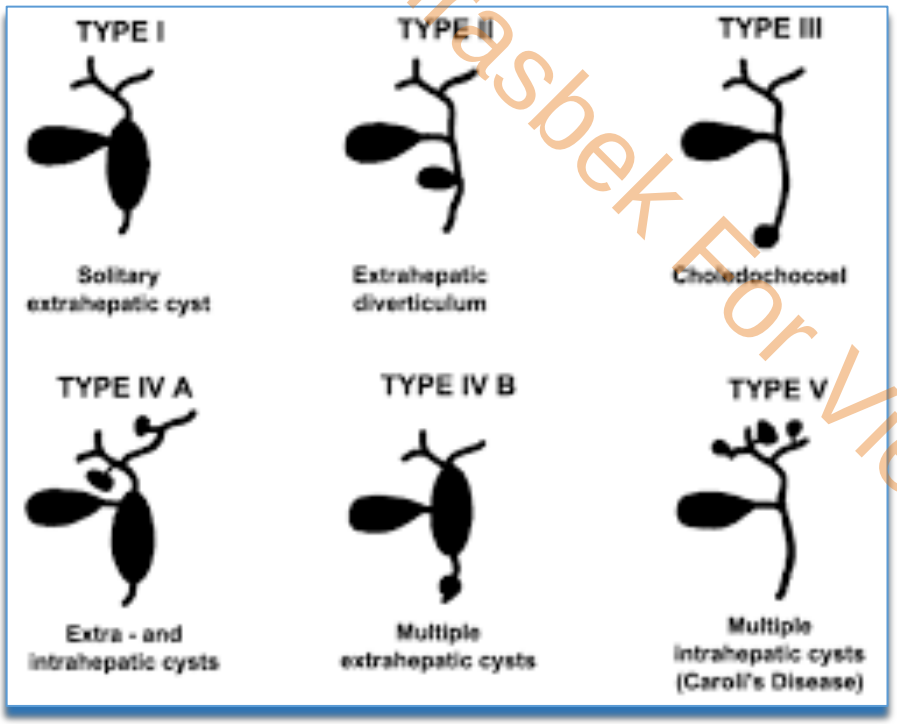
- **Treatment:**

- Patients without complications are monitored with steroid therapy.

- Biliary diversion surgeries are performed for stenosis and obstruction.

- In cases of widespread involvement, liver transplantation is considered as a last chance.

Choledochal Cysts:



- Type I: Fusiform dilatation of the common bile duct (the most common type, 80-90%)

- Type II: Diverticulum of the bile duct

- Type III: Choledochocele (cystic dilation in the duodenum)

- Type IVa: Widespread cystic dilatation in both intrahepatic and extrahepatic ducts

- Type IVb: Widespread cystic dilatation only in the extrahepatic ducts

- Type V: Widespread intrahepatic ductal dilatation (Caroli's Disease), which is very rare. Intrahepatic ducts show congenital segmental saccular dilatations.

- **Associated Conditions:**

- Often associated with cystic renal diseases.

- **Clinical presentation**

- Choledochal cysts are four times more common in women.

- The classic triad includes abdominal pain, a palpable mass, and jaundice. However, this triad is present in less than half of the patients.

- More common presentations include intermittent abdominal pain occurring over months or years, and often unnoticed mild jaundice.

- If not diagnosed, complications such as cholangitis and pancreatitis can develop.

- **Diagnosis:**

- Cholangitis can progress to cirrhosis and portal hypertension.

- Choledochal cysts are a risk factor for cholangiocellular carcinoma.

- Diagnosis is typically made through ultrasound (USG), endoscopic retrograde cholangiopancreatography (ERCP), and percutaneous transhepatic cholangiography (PTC).

- **Treatment:**

- Due to the risk of cancer, cyst excision and biliary reconstruction are necessary.

- For Type IVa cysts and Caroli's disease, liver transplantation may be required.

- Initial treatment typically involves ERCP (endoscopic retrograde cholangiopancreatography) and percutaneous transhepatic cholangiography (PTC) for drainage.

- If the cyst is limited to a single lobe, resection may be attempted.

- Liver transplantation may be necessary in some cases.

Postcholecystectomy Syndrome

- The presence of symptoms that develop or persist after cholecystectomy.

- Can be due to biliary or extra-biliary causes.

- Differential diagnosis should be made.

- **Causes of Biliary Postcholecystectomy Syndrome:**

- Retained stones in the common bile duct

- Long residual cystic duct stump

- Stones in the cystic duct stump

- Stenosis of the Oddi sphincter

- Biliary stricture

- **Extra-biliary causes of Postcholecystectomy Syndrome:**

- Hiatal hernia

- Pancreatitis

- Peptic ulcer

- Irritable bowel syndrome

- Food intolerance

Gallbladder Cancer

- The most common carcinoma of the biliary system.

- Occurs three times more frequently in women.

- The average age of diagnosis is around 70 years.

- Found in 1% of patients who have undergone biliary system surgery.

- 80-90% of cases are adenocarcinomas; rarely encountered are adenosquamous, squamous, small cell, or anaplastic carcinomas.

- **Pathological Types:**

- Papillary

- Nodular

- Tubular

- The papillary type has the best prognosis.

- The tumor primarily metastasizes via lymphatic routes, but transmural spread and liver metastasis are more common findings.

- Liver metastasis most often occurs in segments 4 and 5.

- **Etiology and Risk Factors for Gallbladder Cancer:**

- *Cholelithiasis*

  - The most significant risk factor.

  - 80-95% of patients have stones.

  - The risk of cancer is 10 times higher with large stones (>3 cm).

  - Symptomatic cholelithiasis patients are more likely to develop cancer compared to asymptomatic patients.

- *Polypoid Lesions*

  - Particularly those > 10 mm.

- *Porcelain Gallbladder*

  - Cancer risk is 10-20%.

- *Choledochal Cysts*

  - *Pancreatic and Biliary Tract Junction Anomalies*

- *Sclerosing Cholangitis*

- *Carcinogen Exposure* (e.g., azotoluene, nitrosamines)

- **Clinical Presentation**:

- The most common symptom is pain in the right upper quadrant.

- Nausea and vomiting may accompany, and sometimes jaundice.

- Symptoms may be confused with cholecystitis.

- In advanced stages, a mass may be palpable.

- Diagnosis is usually made after surgery.

- Laboratory tests are not very helpful; ultrasound (USG) and computed tomography (CT) are useful.

- **Treatment**

- In early stages, a cholecystectomy alone may be sufficient.

- If the serosa is breached, additional procedures such as liver wedge resection and lymphadenectomy are required.

- For cases that have breached the serosa, the 5-year survival rate is low.

- Incidental diagnoses generally have a better prognosis.

- For patients diagnosed with the tumor after surgery, the 5-year survival rate is 5%.

Biliary Tract Cancers (Cholangiocarcinoma)

- They are rare.

- More common in men (Male to Female ratio: 1.3:1).

- Two-thirds are located proximal to the bifurcation, most frequently at the junction of the right and left hepatic ducts (Klatskin tumor).

- Sporadic cases typically occur at the bifurcation.

- Those associated with primary sclerosing cholangitis are often multifocal; the chance for resection is very low.

- Overall, the chance for effective treatment is generally low.

- **Etiology and Risk Factors**

- Primary Sclerosing Cholangitis

- Choledochal Cysts

- Ulcerative Colitis

- Hepatolithiasis

- Bilioenteric Anastomoses

- Recurrent Pyogenic Cholangitis

- Chronic Typhoid Carrier Status

- Liver Parasites (e.g., Clonorchis sinensis, Opisthorchis viverrini)

- Exposure to Thorotrast, Asbestos, and Dioxins

- Smoking

- Oral Contraceptives

- Foods Containing Nitrosamines

- In many patients, no defined risk factor is identified.

- **Pathology**

- Most cases (>95%) are adenocarcinomas.

- Four types:

- Nodular (most common)(Scirrhous, Diffuse Infiltrative.Papillary)

- **Clinical presentation :**

- Painless jaundice is the most common symptom.

- Anorexia, weight loss, pruritus and right upper quadrant pain may also be present.

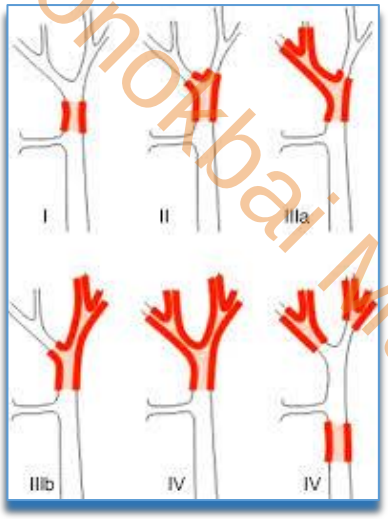
- Courvoisier's sign may be observed.

- **Diagnosis**

- USG, PTK, and ERCP are used for diagnosis.

- CA 19-9 is the tumor marker and is valuable in diagnosis.

- The tumor typically presents as a mass affecting a segment of the bile ducts, showing symptoms according to the segment involved.



- It is classified as adenocarcinoma and referred to as cholangiocarcinoma.

- The tumor can involve the distal choledochus, common hepatic duct, cystic duct, or right and left hepatic ducts separately.

- When located at the hepatic duct confluence, it is called a "**Klatskin tumor**".

- Primarily, they metastasize via lymphatic spread, grow slowly, and show both direct and metastatic spread to the liver.

- **Treatment:**

- Surgical intervention is the only treatment option.

- At the time of diagnosis, the resectability rate is approximately 10-20%.

- Distal tumors have a higher chance of treatment success.

- Polypoid tumors generally have a better prognosis.

- Pancreaticoduodenectomy may be performed for distal choledochal tumors.

- For more proximal tumors, resection and biliary reconstruction (hepaticojejunostomy) are applied.

- In cases where curative resection is not possible, palliative procedures such as intrahepatic hepaticojejunostomy, choledochojejunostomy, or surgical or radiological stenting can be considered.

- Without any intervention, survival generally does not exceed 6 months.

- With stenting, survival can be extended to 1-1.5 years, and with resection, the average survival is 23 months.

- The cause of death is often not directly related to the tumor but is usually due to progressive biliary cirrhosis, recurrent cholangitis, abscess formation, or sepsis.

Obstructive Jaundice

- When bilirubin levels exceed approximately 2.5 mg/dl, a yellowish discoloration of the sclerae develops.

- Jaundice is a similar discoloration of the skin that occurs when serum bilirubin levels rise above 5 mg/dl.

- Obstructive jaundice is relevant to surgery.

- Etiology includes: choledocholithiasis, periampullary region cancers, choledochal strictures, hydatid cysts, external compressions, and other causes.

- Choledocholithiasis

- Biliary tract obstruction.

- Periampullary Region Tumors (Pancreaticoduodenal Region Tumors)

- Pancreatic head cancer is the most common, accounting for 60-75% of cases.Pancreatic cancers have the worst prognosis and are all adenocarcinomas.

- Other tumors in this region may show occult blood in the stool.

- Cholangitis and sepsis may occur.

- Jaundice rarely resolves except in cases of ampullary tumors.

- Cirrhosis and liver abscesses are rare in these tumors because patients often do not survive long enough.

- Extrahepatic Bile Duct Tumors
- Ampulla of Vater Tumors

- These constitute 10% of periampullary cancers.

- They develop from the duodenal mucosa.

- Early stages may present with intermittent jaundice.

- Stool may test positive for occult blood.

- Most tumors have a chance of resection.

- The five-year survival rate is 35-40%.

- Among periampullary tumors, those with the best prognosis and the highest chance of resection are ampullary tumors.

- **Choledochal Strictures**

- ***Postoperative***(Typically develop postoperatively, often following cholecystectomy. They are iatrogenic).

Iatrogenic strictures are the most common cause of benign biliary strictures.

- ***Inflammatory***

- Fibrosis resulting from inflammation can lead to strictures. Causes include: chronic pancreatitis, choledocholithiasis, acute cholangitis, biliary obstruction due to cholelithiasis,

sclerosing cholangitis, and cholangiopathies. T-tube irritation can also lead to fibrosis and strictures.

- Choledochal Cysts also cause of mechanic jaundice
- Parasites- Particularly Ascaris. Hydatid cysts.
- Lymphadenopathy and Metastatic Tumors

Diagnosis and Differential Diagnosis of Obstructive Jaundice

- Diagnosis can be made with laboratory methods.Clinical examination does not differentiate between intrahepatic and extrahepatic cholestasis.

- **Ultrasound (USG).** The initial imaging method for obstructive jaundice.

  - 90% reliability overall; decreases to 60% as it approaches the lower end of the common bile duct.

  - Dilated intrahepatic bile ducts observed on USG indicate extrahepatic cholestasis.

  - Can sometimes reveal the cause of the obstruction.

  - Can identify choledocholithiasis or head of the pancreas tumors.

-Intravenous Cholangiography

  - Not useful if bilirubin is above 3 mg/dl.

-Percutaneous Transhepatic Cholangiography (PTC)

- Used to identify the cause of extrahepatic cholestasis when intrahepatic bile ducts are dilated.

- Particularly successful in diagnosing proximal choledochal and hepatic ductal lesions.

- Also allows for biliary stenting.

-Endoscopic Retrograde Cholangiopancreatography (ERCP)

- Can reveal the location and cause of the obstruction.

- More effective for distal obstructions.

- The most common complication of ERCP is the development of post-procedural pancreatitis.

-Computed Tomography (CT)

- The most successful method for identifying mass lesions.

-**Kinetic Scintigraphy**

- Performed with radioactively labeled substances that are filtered with bile.

- More effective than static scintigraphic methods and has a lower margin of error.

Treatment

1. Eliminate Obstruction

- Remove stones from the bile duct and perform cholecystectomy if necessary.

2. Address Oddi Sphincter Fibrosis

- If fibrosis of the Oddi sphincter is present, perform sphincteroplasty, choledoco-duodenostomy, or choledoco-jejunostomy.

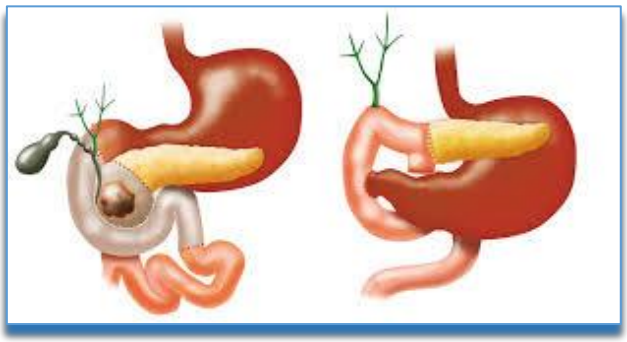
3. Manage Bile Duct Strictures

- If strictures are present in the bile duct, perform reconstruction or bypass.

4. Resect Tumors

- Remove any tumors that are present.

5. Pancreaticoduodenal Region Tumors



- Whipple's procedure is commonly used for tumors in the pancreaticoduodenal region.

- After surgery, perform gastro-jejunostomy, choledoco-jejunostomy, and pancreaticojejunostomy as required.

6. Mortality and Survival Rates

- There is an approximate 10% mortality rate associated with the surgery.

- Postoperative 5-year survival rates: 15-20% for pancreatic head cancers and 25-30% for ampullary cancers.

7. Postoperative Diabetes Mellitus

- Post-surgery, diabetes mellitus may develop, and managing it can be challenging.

8. Bypass Procedures

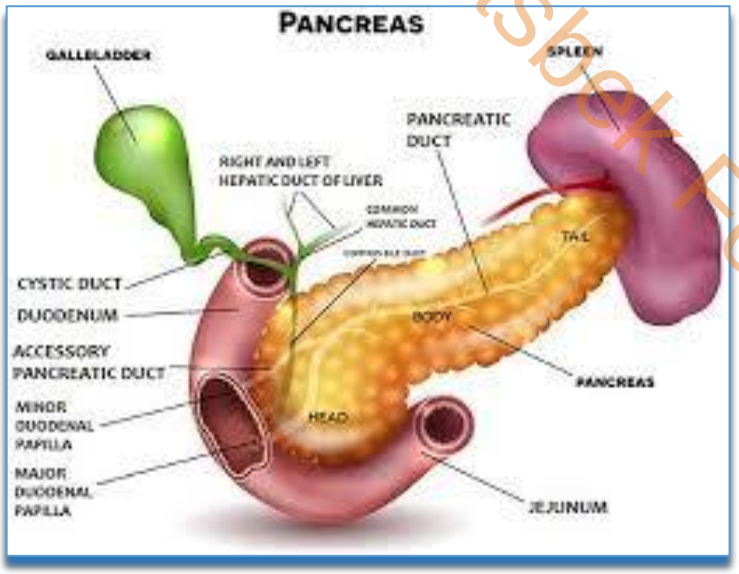
- If the bile duct is obstructed and the patient is not curable, consider bypass surgeries to ensure bile flow.

Chapter

8.

PANCREATIC DISEASES AND SURGERY

PANCREAS ANATOMY



The pancreas is an organ that extends obliquely between the "C"-shaped segment of the duodenum and the hilum of the spleen, situated retroperitoneally.

- Surgically, it is divided into four parts: the head, neck, body, and tail.

- The main pancreatic duct (Wirsung) starts from the tail of the pancreas and enters the duodenum distal to the common bile duct.

- The common bile duct and the duct of Wirsung usually run side by side for a few millimeters before merging to form a single channel.

- This single pancreaticobiliary channel opens into the lumen of the duodenum at the papilla of Vater, which is located in the second part of the duodenum, posteromedially.

- The ampulla of Vater is the dilation within the papilla where the common channel is located. In 90% of cases, there is an ampulla, which is usually 5 mm or shorter. In 10% of cases, there is no ampulla, and both ducts open separately into the duodenum.

- The accessory duct (Santorini) usually drains the anterior and superior portion of the head of the pancreas.

- In 60% of cases, the duct of Santorini enters the duodenum at the minor papilla, which is located 2 cm cranial and anterior to the papilla of Vater.

-The blood supply to the head of the pancreas primarily comes from the superior pancreaticoduodenal artery, originating from the gastroduodenal artery, and the inferior pancreaticoduodenal artery, originating from the superior mesenteric artery.

-The body and tail of the pancreas receive blood supply from branches of the splenic artery and the inferior pancreatic artery.

- Venous drainage largely parallels the arteries.

- The lymphatics of the pancreas are quite rich and generally follow the venous drainage in all directions.

- The pancreas receives sympathetic innervation from the splanchnic ganglia and parasympathetic innervation from the vagus nerve.

- Pain sensation is typically carried by visceral afferent fibers passing through the celiac plexus and ganglia.

- For palliative pain relief in cases of cancer and chronic pancreatitis, celiac ganglion destruction can be performed.

PANCREAS PHYSIOLOGY

The physiology of the pancreas can be divided into the exocrine and endocrine pancreas.

Exocrine Pancreas:

- It constitutes about 85% of the pancreatic mass, while the endocrine pancreas makes up only about 2%.

- With the replacement of insulin and digestive enzymes, a human can survive without a pancreas.

- If about 20% of the pancreatic parenchyma remains and the remaining tissue is normal, pancreatic insufficiency can be prevented.

- Produces 500-800 ml of clear, odorless, alkaline (pH: 8.0-8.3) pancreatic fluid daily.

- This fluid contains more than 20 digestive enzymes and is the joint product of acinar and ductal cells.

- The fluid stimulated by secretin, rich in electrolytes and isoosmolar with plasma, mainly contains sodium and potassium as cations, and bicarbonate and chloride as anions.

- The alkaline pancreatic fluid helps neutralize stomach acid and provides the optimal pH necessary for pancreatic enzyme activity.

- Pancreatic enzymes are divided into three groups: proteolytic (trypsin, chymotrypsin, carboxypeptidase, ribonuclease, deoxyribonuclease, elastase), lipolytic (lipase, colipase, phospholipase A2), and amylolytic (amylase), responsible for digesting proteins, fats, and carbohydrates, respectively.

- Acinar cells have the capacity to synthesize all types of enzymes and can secrete them in response to stimulation by cholecystokinin and vagal cholinergic stimulation.

- Amylase and lipase are secreted in their active forms, while proteolytic enzymes and phospholipase A2 are secreted as inactive zymogens that require activation.

- Trypsinogen, secreted into the duodenum, encounters the duodenal enzyme enterokinase, which converts it to its active form, trypsin. Trypsin then activates other zymogens into their active forms.

- The activation of trypsinogen within the pancreas is prevented by inhibitors also secreted by acinar cells.

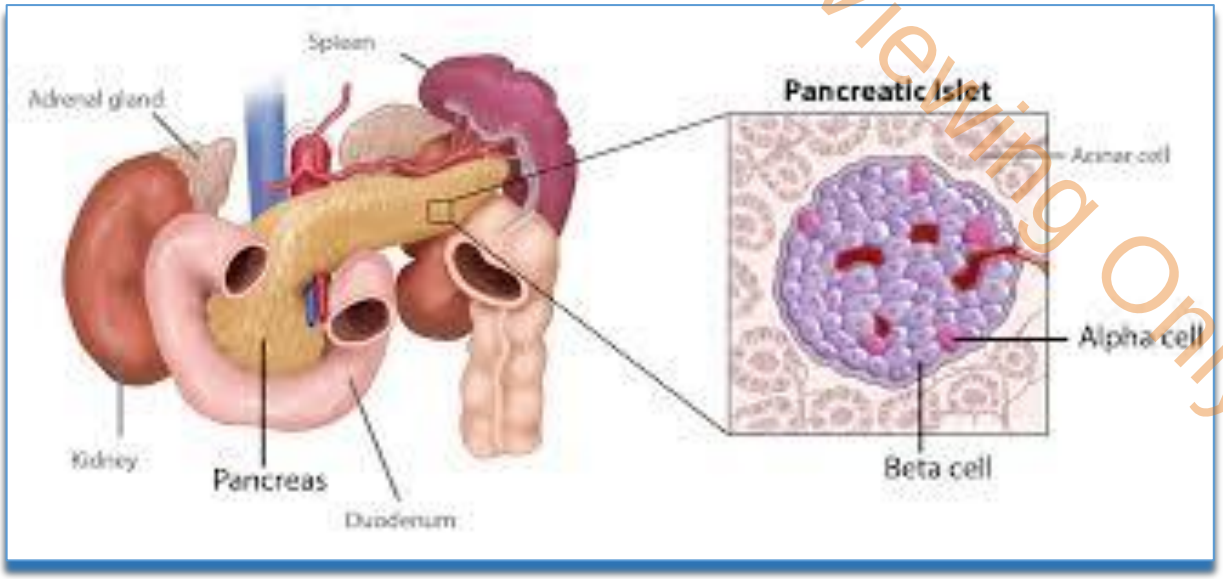
- A deficiency in the production of normal trypsinogen inhibitors, known as pancreatic secretory trypsin inhibitor (PSTI) or SPINK1, can cause familial pancreatitis.

- These inhibitors ensure that trypsinogen remains inactive while inside the pancreas and only becomes active in the duodenum. Without this regulation, pancreatic cells would be damaged, leading to pancreatitis.

- Mutations in the cationic trypsinogen gene (PRSS1) can cause premature activation of trypsinogen within the pancreas.

- This is responsible for two-thirds of hereditary pancreatitis cases.

ENDOCRINE PANCREAS



The islets of Langerhans constitute 1-2% of the pancreatic cell mass.

These islets are composed of 3000-4000 cells that secrete four main hormones.

These include beta cells that secrete insulin, alpha cells that secrete glucagon, delta cells that secrete somatostatin, and PP cells that secrete pancreatic polypeptide.

- Insulin - Beta cells (Decreased gluconeogenesis, glycogenolysis, fatty acid breakdown, and ketogenesis

Increased glycogenesis, protein synthesis, glucose uptake)

- **Glucagon** - Alpha cells- (Opposite of insulin, increased hepatic glycogenolysis, and gluconeogenesis)

- **Somatostatin** - Delta cells (Inhibits GI secretions, functions of all GI endocrine peptides, and cell growth)

- **Pancreatic polypeptide** - PP cells(Inhibits pancreatic exocrine secretion and insulin release.Facilitates the hepatic effects of insulin,

- **Amylin** - Beta cells (antagonizes insulin release and function)

- **Pancreastatin** - Beta cells (Reduces the release of insulin and somatostatin. Increases glucagon release.Reduces pancreatic exocrine secretion)

- **Ghrelin** - Epsilon cells (Reduces insulin release and its functions)

PANCREATITIS

- Pancreatitis is the inflammation of the pancreas, which can range from mild edema to hemorrhagic necrosis, and can present with various pathological and clinical pictures, potentially resulting in fibrosis and permanent exocrine and endocrine dysfunction.

- It is primarily classified into two groups: acute and chronic.

- In acute pancreatitis, morphological changes return to normal with the treatment of the primary cause, whereas in chronic pancreatitis, repeated attacks and inflammation lead to permanent and progressive tissue damage.

- Despite repeated attacks, acute pancreatitis does not tend to evolve into chronic pancreatitis.

- Fibrosis is very rare.

ACUTE PANCREATITIS

ETIOLOGY OF ACUTE PANCREATITIS

- Acute pancreatitis (AP) has many causes.

- Biliary and alcoholic pancreatitis account for 75-80% of all pancreatitis cases.

Gallstones

- The most common cause.

- 40% of acute pancreatitis cases are biliary pancreatitis.

-

Typically occurs in women aged 50-70.

- Alcohol

- Excessive alcohol consumption is the second most common cause.

- Accounts for 30% of cases.

- Typically occurs in men aged 30-45.

- About 5-10% of heavy alcohol users develop acute pancreatitis.

- Factors increasing the risk of alcohol-related pancreatitis: Excessive alcohol consumption (> 100 g/day, for at least 5 years), Smoking and Genetic predisposition

- Triggers pro-inflammatory pathways such as nuclear factor kappa B (NF-KB).

- NF-KB stimulates the production of TNF-α and IL-1.

- Increases the expression and activity of caspases.

- Caspases are proteases that regulate apoptosis.

- Reduces pancreatic perfusion.

- Leads to Oddi's sphincter spasm.

- Causes protein precipitation in pancreatic ducts, leading to ductal obstruction.

- **Pathophysiology of Severe Acute Pancreatitis:**

- Local injury triggers the secretion of TNF-α and IL-1.

- Both cytokines exacerbate pancreatic injury.

- They also stimulate the release of other inflammatory mediators, amplifying the inflammatory response.

- This can lead to damage in distant organs.

- This mechanism is responsible for early deaths observed in severe acute pancreatitis.

DIAGNOSIS OF ACUTE PANCREATITIS

- Abdominal pain consistent with acute pancreatitis (sudden onset, severe epigastric pain radiating to the back) + elevated amylase/lipase (>3X) = Acute pancreatitis.

- CT scan is not necessary for the diagnosis of acute pancreatitis unless the above diagnostic criteria are not met.

- The half-life of amylase is shorter than that of lipase. Therefore, in patients presenting 24-48 hours after symptom onset, lipase is more sensitive.

- Amylase can also be elevated in other conditions such as peptic ulcer, salpingitis, mesenteric ischemia, and macroamylasemia.

- AP patients are typically hyperglycemic.

- Leukocytosis may be present, and liver enzymes may be elevated.

- Elevated alanine aminotransferase (ALT) in a patient diagnosed with AP with high amylase/lipase levels indicates acute biliary pancreatitis with 95% confidence.

- Other upper abdominal diseases that can be confused with AP include peptic ulcer perforation, gangrenous small bowel obstruction, and acute cholecystitis.

CLINICAL FINDINGS IN ACUTE PANCREATITIS

- **Pain**

- The most common finding is epigastric pain, which is severe and constant, often radiating to the back and can be belt-shaped.

- It often occurs following a heavy meal or alcohol intake and is accompanied by nausea and persistent vomiting.

- The severity of the pain is proportional to the severity of the pancreatitis.

- **Fever**

- Mildly elevated. If fever exceeds 38°C (100.4°F), infectious complications should be considered.

- **Dehydration Findings**

- Hypotension, tachycardia, and tachypnea. Hemoconcentration.

Cullen's Sign- Ecchymosis in the periumbilical area due to retroperitoneal bleeding.

Grey-Turner's Sign- Ecchymosis in the flank due to retroperitoneal bleeding. Both Cullen's and Grey-Turner's signs are indicative of hemorrhagic pancreatitis; they are present in approximately 1% of patients.

DIAGNOSIS. Laboratory and instrumental Findings in Acute Pancreatitis

Serum Amylase Level

- Elevated amylase without abdominal pain is not meaningful.

- Serum amylase can also be elevated in conditions such as acute cholecystitis, ulcer perforation, and intestinal obstruction.

- Amylase is generally elevated in pancreatitis.

- Levels rise with the onset of the disease and remain high for 3-5 days.

- There is no direct correlation between amylase elevation and the severity of pancreatitis.

- Mild forms of pancreatitis can also present with elevated amylase levels.

- In acute pancreatitis due to hyperlipidemia, serum amylase levels are usually normal due to interference between lipids and amylase.

- Persistent amylase elevation lasting more than a week may suggest ongoing inflammation or complications such as pseudocyst, abscess, or pancreatic necrosis.

- Levels of pancreatic lipase, trypsin, and elastase may be elevated.

- Among pancreatic exocrine enzymes, lipase has the highest specificity for diagnosing pancreatitis.

Direct Radiographs(X-ray)

- Direct radiographs are not useful for diagnosing acute pancreatitis (AP).

- They are helpful for ruling out conditions that can be confused with AP, such as ulcer perforation.

- In direct abdominal radiographs, a segment of the small intestine secondary to adynamic ileus in the left upper abdomen may be detected, including sentinel loops and sentinel loops.

- Inflammation of the pancreas can cause spasm in the neighboring colon, resulting in an interruption in the colon gas pattern (colon cut-off sign).

CT Scan

- CT is the most reliable and preferred imaging method for diagnosing acute pancreatitis (AP).

- A CT scan is essential for all patients who do not improve within 2-3 days after the diagnosis of AP to investigate potential complications.

- On CT, the pancreas may appear normal, edematous, or show signs of phlegmon, peripancreatic fluid collection, necrosis, abscess, and spread of inflammation.

- The presence of necrosis indicates severe pancreatitis.

- Contrast-enhanced CT provides better visualization of pancreatic perfusion and necrosis.

- It is the most reliable imaging method anatomically for the pancreas.

- It is the gold standard for diagnosing necrosis, fluid collections, and pseudocysts.

- The presence of air bubbles supports the presence of infection (such as infected pancreatic necrosis or pancreatic abscess).

Ultrasound (USG)

- After diagnosing acute pancreatitis (AP), ultrasound is used to understand the etiology of the condition.

- It helps in diagnosing biliary pancreatitis by showing gallstones and dilation of the bile ducts.

- It can reveal edematous pancreas, peripancreatic fluid collections and ascites, pseudocysts, and dilation of the Wirsung duct.

- Samples can be taken from pancreatic collections.

- Determining the Severity of Acute Pancreatitis

- Knowing whether the disease is mild or severe is important for determining the treatment plan.

- Parameters commonly used to determine the severity of the disease include clinical findings, early prognostic indicators, serum markers, and CT scans.

- The presence of necrosis on CT scan indicates severe pancreatitis.

- Recent multicenter studies have defined four categories of severity in acute pancreatitis: mild, moderate, severe, and critical.

-Ranson's criteria are used to assess the severity of acute pancreatitis and predict its prognosis. Based on the Swartz textbook of surgery, Ranson's criteria include both the initial and the 48-hour criteria. Here's a summary:

Initial Criteria (upon admission):

1. Age: Over 55 years old.

2. White Blood Cell Count (WBC): Greater than 16,000 cells/µL.

3. Blood Glucose Level: Greater than 200 mg/dL.

4. Lactate Dehydrogenase (LDH): Greater than 350 IU/L.

5. Aspartate Aminotransferase (AST):Greater than 250 IU/L.

48-Hour Criteria (measured after 48 hours):

1. Hematocrit Drop: Greater than a 10% decrease.

2. BUN Increase: Greater than 5 mg/dL increase.

3. Serum Calcium: Less than 8 mg/dL.

4. Arterial PaO2: Less than 60 mm Hg.

5. Base Deficit: Greater than 4 mEq/L.

6. Fluid Sequestration: Greater than 6 liters.

Scoring:

- Each criterion met scores 1 point.

- The total score ranges from 0 to 11.

- A higher score correlates with a more severe prognosis and increased mortality risk.

In the Swartz "Textbook of Surgery", Ranson's criteria for acute pancreatitis are used to estimate the severity of the disease and predict mortality risk. According to this textbook, the scoring system and associated mortality risk are as follows:

- 0-2 points: Mortality rate of approximately 1-5%

- 3-4 points: Mortality rate of approximately 10-20%

- 5-6 points: Mortality rate of approximately 30-50%

- 7 or more points: Mortality rate of approximately 50% or higher

TREATMENT

The treatment of acute pancreatitis involves several key components. Here's an overview based on the textbook:

1. Initial Management:

- Supportive Care:

- Fluid Resuscitation: Start with aggressive intravenous fluid therapy to manage hypovolemia and prevent shock. This is crucial for maintaining blood pressure and ensuring adequate perfusion of vital organs.

- Electrolyte Management: Monitor and correct electrolyte imbalances, including hypocalcemia and hypokalemia.

- Pain Control: Administer analgesics, typically opioids, to manage severe abdominal pain. Avoid to use the morphine

- Nutritional Support: Early Enteral Feeding: Once the patient's condition stabilizes, enteral nutrition via a nasojejunal tube is preferred over parenteral nutrition as it helps maintain gut integrity and reduces complications.

- Diet: Begin with a clear liquid diet and gradually advance as tolerated.

2. Treatment of Underlying Causes:

- Biliary Pancreatitis:

- ERCP (Endoscopic Retrograde Cholangiopancreatography): If the pancreatitis is due to gallstones, perform ERCP to remove the stones and relieve bile duct obstruction. This may be done early or delayed based on the patient's condition.

- Alcohol-Induced Pancreatitis: Encourage and support alcohol abstinence. Address any alcohol use disorder with appropriate interventions.

3. Surgical Interventions:

- Necrotizing Pancreatitis:

- Surgical Debridement: If there is evidence of pancreatic necrosis or infection, debridement may be necessary. This can be done surgically or via percutaneous techniques.

- Timing of Surgery: Delay surgery until the patient's condition stabilizes and necrotic tissue becomes demarcated. Early surgical intervention is generally avoided.

4. Management of Complications:

- Infection. Antibiotics: Administer broad-spectrum antibiotics if there is evidence of infection, especially in cases with pancreatic necrosis or pseudocysts.

- Organ Failure.Supportive Care: Monitor and manage any organ failure, such as renal or respiratory failure, with appropriate supportive measures including renal replacement therapy or mechanical ventilation if needed.

5. Long-Term Management:

- Follow-Up: Regular follow-up is necessary to monitor recovery and manage any ongoing issues such as diabetes or digestive problems.

- Lifestyle Modifications: Educate patients about lifestyle changes, including diet and avoidance of alcohol, to prevent recurrence of pancreatitis.

These treatment strategies are designed to address the underlying cause of acute pancreatitis, manage symptoms, and prevent complications. The management approach may vary based on the severity of the condition and the presence of complications.

Complications in Acute Pancreatitis

1.**Systemic Complications:**

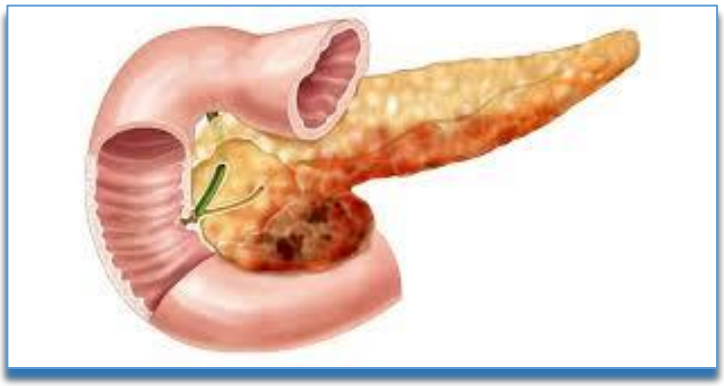
- ARDS (Acute Respiratory Distress Syndrome) and Respiratory Failure

- Renal Failure

- Myocardial Depression

2.Local Complications:

-**Peripankreatic Fluid Collections** (Sterile and Infected): Fluid collections are observed in 30-57% of patients with acute pancreatitis.These collections do not have a fibrotic capsule around them. Typically managed with supportive care and monitored over time. Most collections are absorbed spontaneously.



-Necrosis:

- Infected Necrosis:Presence of devitalized pancreatic tissue or peripancreatic fat tissue.

- Diagnostic Method: CT (Computed Tomography) is the best diagnostic tool.

- Prevalence: Necrosis occurs in up to 20% of acute pancreatitis patients.

- Autopsy Findings: Necrosis is seen in 80% of patients who die from acute pancreatitis.

- Main Complication: Infection is the primary complication of necrosis.

- Infection Risk: Directly related to the amount of necrosis.

- Indicators of Infection: Persistent fever, elevated white blood cell count, and clinical deterioration suggest infected necrosis.

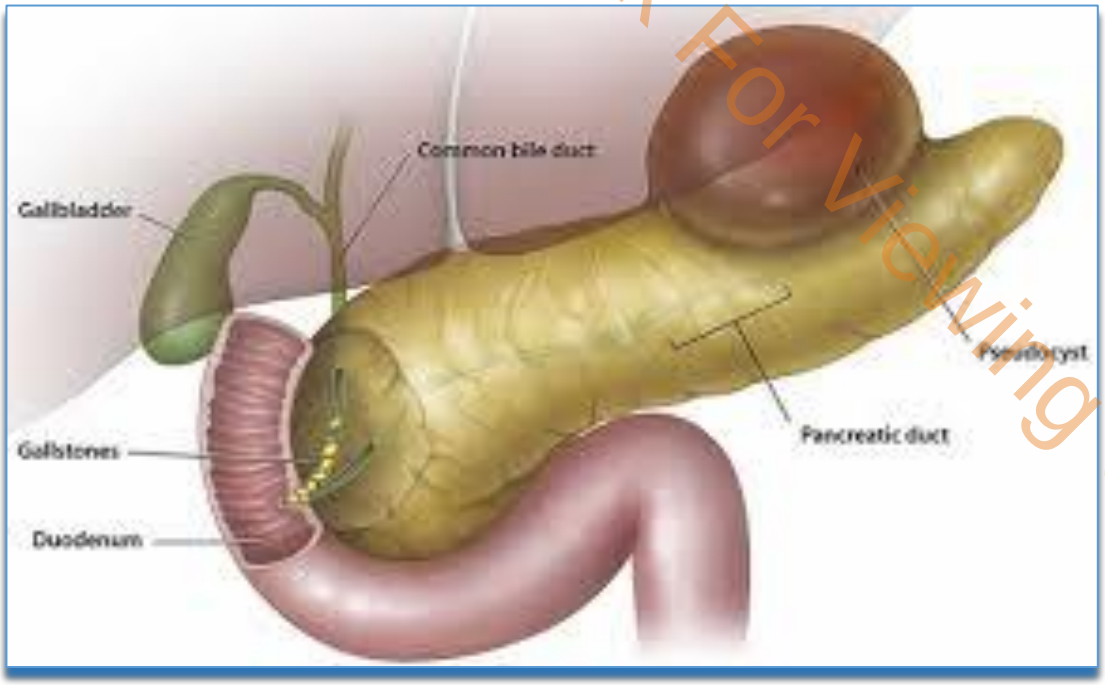
- CT Findings: Presence of air in the necrotic tissue indicates infection.

- Management:If infection is suspected, a percutaneous drainage catheter should be placed.

- Treatment: IV antibiotics should be started, with carbapenems being the first choice.

-Pseudocyst:

- Occurrence: Pseudocysts develop in about 5% of patients with fluid collections during the acute phase of pancreatitis.



-

Description: A pseudocyst is a fluid collection containing pancreatic enzymes.

- Management in Acute Phase: If a pseudocyst is detected during the acute phase, it should be monitored for about 6 weeks.

- Resolution:There is a possibility that the pseudocyst may resolve within this period (up to 70% of cases).

- Treatment Options:

- Small Cysts:Endoscopic transgastric or transduodenal drainage is performed.

- Cysts Associated with the Main Pancreatic Duct: Transpapillary drainage may be used.

- Surgical Intervention: If endoscopic methods are unsuccessful, surgery is required, most commonly cystogastrostomy.

-Pancreatic Ascites:

- **Definition:** Occurs when pancreatic secretion leaks freely into the peritoneal cavity instead of being confined like in a pseudocyst.

- **Diagnosis:** Paracentesis is helpful in diagnosing pancreatic ascites.

- **Treatment:** Drainage is performed to manage the condition.

CHRONIC PANCREATITE

Etiology of Chronic Pancreatitis:

1.Chronic Alcoholism:

- The most common cause of chronic pancreatitis.

- Excessive alcohol consumption is the most frequent cause, accounting for 70-80% of cases (according to Sabiston, 20th edition).

- According to Schwartz, 10th edition, idiopathic causes are the most common, followed by genetic factors, with alcohol being the third most common cause.

- 3-7% of heavy alcohol users develop chronic pancreatitis, suggesting that alcohol acts more as a cofactor.

- The toxic effects of alcohol on the pancreas were discussed in the acute pancreatitis section.

- Important Discovery in the Late 1990s:

    - The identification of pancreatic stellate cells (PSCs).

    - PSCs are differentiated, dormant fibroblasts located at the base of pancreatic acinar cells.

    - When activated, PSCs transform into active myofibroblasts that synthesize proteins forming the extracellular matrix.

    - Examples of these synthesized proteins include collagen I and III, fibronectin, laminin, and matrix metalloproteases.

    - Chronic necrosis and inflammation activate PSCs.

    - The release of these substances leads to collagen deposition and pancreatic fibrosis.

    - Ethanol-induced necroinflammation activates PSCs, leading to fibrosis.

    - Ethanol and some of its metabolites (such as acetaldehyde) have been shown to activate PSCs.

- Current preclinical treatments for chronic pancreatitis target PSC activation.

- Antioxidants, ACE inhibitors, peroxisome proliferator-activated receptor gamma ligands, and vitamin A inhibit PSC activity.

2.Smoking:

- Smoking increases the risk of developing chronic pancreatitis caused by alcohol.

- Active smokers develop chronic pancreatitis at a younger age compared to non-smokers.

- Additionally, smokers have an increased risk of pancreatic calcification and diabetes.

3.Genetic Mutations:

- Mutations in the cationic trypsinogen gene, also known as protease serine 1 gene (PRSS1), are common in hereditary chronic pancreatitis patients, but not frequent in other types.

- Mutations in the **PRSS1** gene lead to intra-acinar trypsinogen activation.

- **SPINK-1** is a gene released by acinar cells that controls the premature activation of trypsinogen.

- SPINK-1 mutations are present in 1-2% of healthy individuals. However, chronic pancreatitis is much rarer.

- SPINK-1 mutation alone is likely insufficient to cause chronic pancreatitis but facilitates the disease and leads to a more severe course.

- SPINK-1 mutations are more pronounced in alcoholic, hereditary, and idiopathic pancreatitis.

- **CFTR Gene**:

- The secretion of bicarbonate and chloride in pancreatic and respiratory secretions is controlled by the CFTR gene.

- In CFTR mutation, normal bicarbonate secretion is affected, reducing the volume of pancreatic fluid and increasing enzyme concentrations within the pancreatic duct.

- Homozygous CFTR mutations cause cystic fibrosis. Mild heterozygous mutations increase susceptibility to pancreatic exocrine insufficiency and chronic pancreatitis.

- The prevalence of CFTR mutations is higher in patients with alcoholic, idiopathic, and hereditary pancreatitis compared to the general population.

- Genetic Defect Linked to Alcoholic Chronic Pancreatitis (2012 Study):

- A genetic defect explaining susceptibility to alcohol-related chronic pancreatitis was discovered in men in 2012.

- 26% of men without pancreatitis and 50% of men with alcoholic pancreatitis were found to have a DNA variant located on the X chromosome.

- This variant, CLDN2, codes for a tight junction protein in pancreatic ductal cells.

- In chronic pancreatitis patients, CLDN2 protein is abnormally expressed, altering enzyme secretion dynamics.

- This anomaly alone does not cause pancreatitis, but if pancreatitis develops in these individuals, it is likely to become chronic, especially in alcohol users.

Chronic Pancreatitis Pathology

- **Fibrosis**:

- In the early stages, nodular scars and fibrosis in the lobular area are observed.

- It has been shown in vitro that vitamin A can prevent this fibrosis.

- **Stone Formation**:

- Normal pancreatic fluid contains proteins, known as lithostatin, that prevent calcium crystallization.

- Alcohol consumption inhibits lithostatin secretion, leading to the formation of plugs.

- If alcohol consumption is stopped early, this process is reversible.

- **Duct Distortion**:

- In the early stages, protein-containing plugs are seen in the ducts, which later progress to fibrosis and calcifications.

- In more advanced cases, ductal dilation resembling a "chain of lakes" may be seen, along with biliary and duodenal obstructions due to fibrosis.

Clinical Findings in Chronic Pancreatitis

- Abdominal pain is the most common symptom:

- The pain is very severe, located in the epigastrium, deep, radiates to the back, or wraps around the waist in a belt-like fashion.

- It is not colicky. As the disease progresses, the pain tends to disappear.

- Loss of appetite:

- The second most common symptom after pain.

- Weight loss and malabsorption.

- Steatorrhea: Caused by exocrine pancreatic insufficiency. It appears first as lipase levels decrease.

- Glucose intolerance or actual diabetes mellitus (DM).

- Pseudocyst formation.

- Biliary obstruction and jaundice.

- Portal hypertension.

- Pyloric obstruction.

Laboratory and Radiological Findings:

- Laboratory tests:Usually performed when imaging is insufficient, but typically not necessary.

- Blood pancreatic products:

- Pancreatic enzyme levels: Not very helpful.

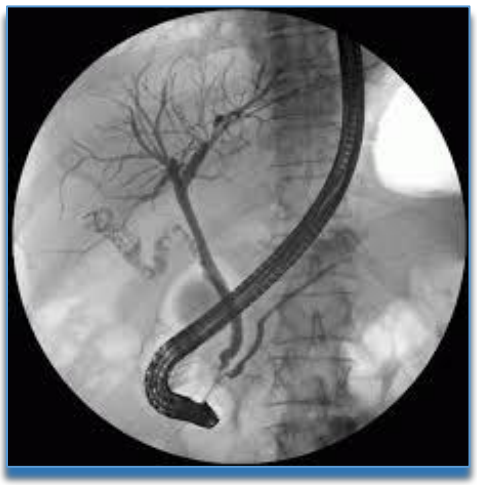
- Pancreatic polypeptide levels: Levels do not increase despite stimulation, reflecting the severity of chronic pancreatitis.

- Ultrasonography (USG)

- Used for initial evaluation and monitoring.

- Computed Tomography (CT) Detects biliary obstruction, dilatation, pseudocysts, dilated Wirsung's duct, and mass lesions.

- **Endoscopic Retrograde Cholangiopancreatography (ERCP)**



- Assesses ductal conditions, dilatation, and allows for biopsy.

- It is the most sensitive imaging method for diagnosing chronic pancreatitis.

Endoscopic Ultrasonography (EUS)

- Increasingly used for evaluating patients with chronic pancreatitis.

- Comparable to ERCP in detecting changes associated with chronic pancreatitis, and recent studies suggest it may be more sensitive than ERCP in early or mild disease.

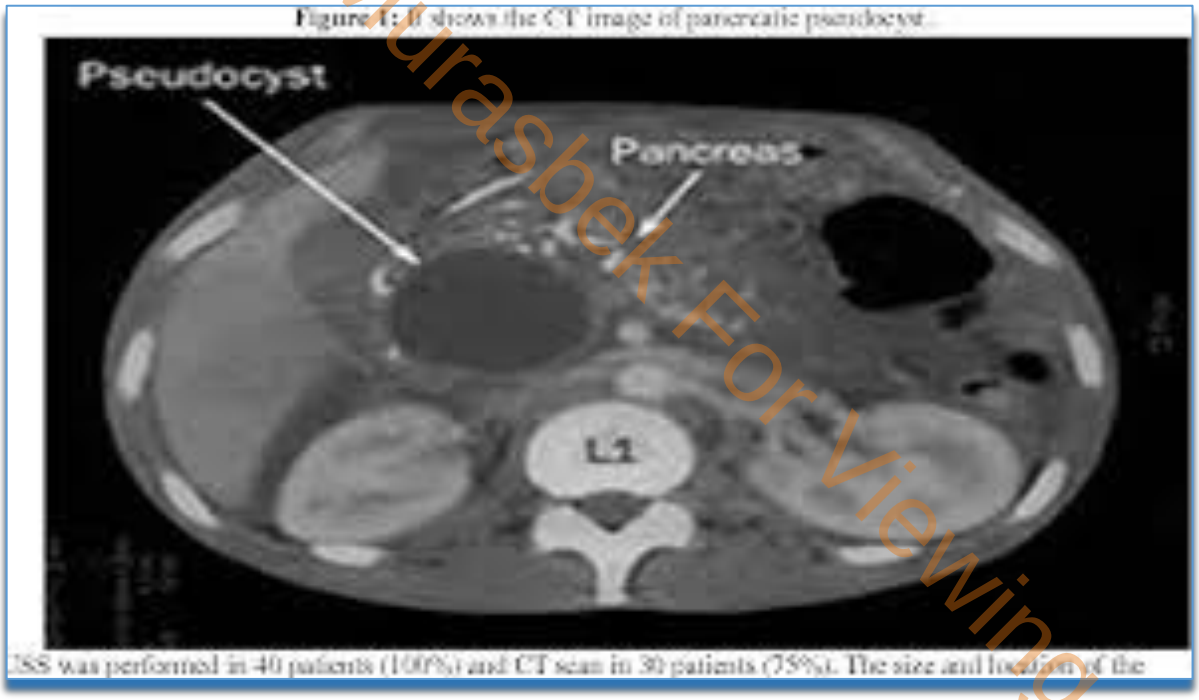
- Highly reliable in ruling out pancreatic carcinoma when findings are normal.

- Differentiates periampullary malignancies.

- Sometimes, surgery may be required for differential diagnosis.

Complications in Chronic Pancreatitis

- **Pseudocyst**



- Most Common Complication: It is the most frequently seen complication of chronic pancreatitis.

- Most Common Pancreatic Cystic Lesion: Pseudocysts are the most common cystic lesions of the pancreas.

- Definition: A pseudocyst is a chronic collection of pancreatic fluids within a fibrous and granulation tissue wall, which lacks an epithelial lining.

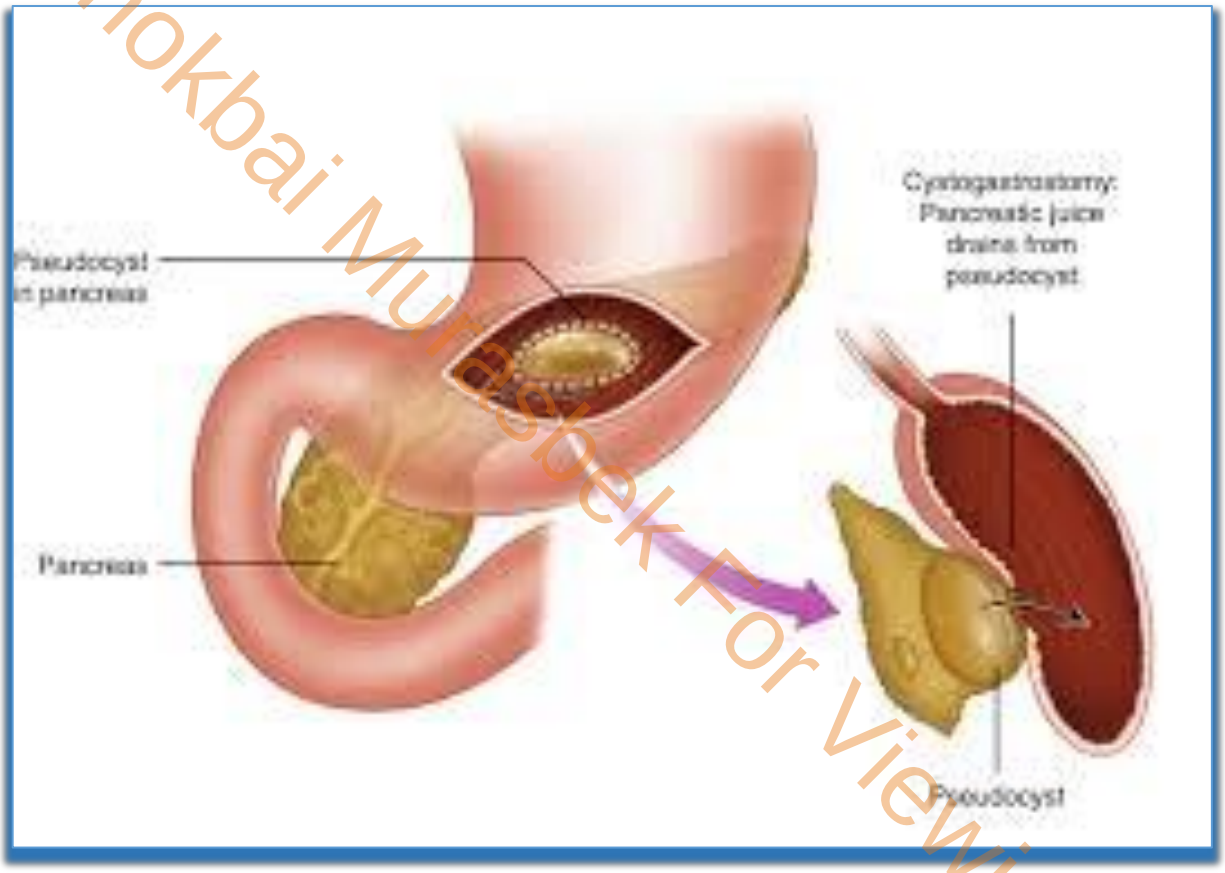
- Early Stages: In the early stages, there is no wall around the fluid collection.

- Wall Maturation: Therefore, the cyst wall's maturation is awaited.

- Diagnostic Tools: Ultrasonography (USG) and computed tomography (CT) can be used for diagnosis. However, CT is the most reliable method.

- Classification: Pseudocysts are classified into two types: those related to the pancreatic duct and those unrelated to it.

Diagnosis and Management of Pseudocysts in Chronic Pancreatitis



- Best Diagnostic Tool: Endoscopic Retrograde Cholangiopancreatography (ERCP) is the best test to determine if the pseudocyst is related to the pancreatic duct. Therefore, treatment is determined based on the ERCP results.

- Management of Pseudocysts Unrelated to Pancreatic Duct: These pseudocysts can be managed through observation or fine-needle aspiration.

- **Management of Pseudocysts Related to Pancreatic Duct:**

  - Initially, ERCP is performed to place a stent in the pancreatic duct.

  - If ERCP fails, surgical treatment is considered after waiting at least 4-6 weeks.

  - Surgical options include cystogastrostomy, cystoduodenostomy, or cystojejunostomy.

- **Major Complications of Pseudocysts:**

    - Bleeding

    - Rupture

    - Infection

    - Splenic vein or superior mesenteric vein thrombosis

    - Gastrointestinal (GI) or bile duct obstruction

- **Incidental Pancreatic Cystic Lesions:** When a pancreatic cystic lesion is incidentally discovered, endoscopic ultrasonography should be performed to distinguish between neoplasm and pseudocyst, followed by aspiration if necessary.

- **Chronic Pseudocyst:** Defined as a pseudocyst that persists for more than six weeks.

- **Jaundice**

- Jaundice frequently occurs due to bile duct obstruction, which is often caused by fibrosis in the head of the pancreas.

- **Pancreatic Duct Obstruction**

- Can occur in chronic pancreatitis.

- **Splenic and Portal Vein Thrombosis**

- Vascular complications of chronic pancreatitis are rare.

- In 4-5% of cases, chronic pancreatitis is associated with portal venous obstruction and splenic vein thrombosis.

- Obstruction of the portal or splenic veins can lead to the development of varices.

- **Duodenal or Colonic Obstruction**

- Can occur when a mass effect is present. This is very rare.

- **Pancreatic Enteric Fistula**

- Results from erosion of the walls of adjacent luminal organs by a pseudocyst, leading to a pancreatic enteric fistula.

- The most common locations for fistula development are the transverse colon or the splenic flexure.

- **Pancreatic Head Mass**

- Approximately 30% of patients with advanced chronic pancreatitis develop an inflammatory mass in the head of the pancreas.

- This mass can cause severe pain, obstruction of bile ducts and the duodenum, compression of the portal vein, and narrowing of the proximal main pancreatic duct.

- Treatment is typically achieved through pancreatic head resection while preserving the duodenum in most cases.

- **Pancreatic Acid**

- Definition: Pancreatic acid occurs when pancreatic fluid leaks from a damaged pancreatic duct and is not confined to a pseudocyst, but instead flows freely into the peritoneal cavity.

- Thoracic Leakage: Occasionally, pancreatic fluid can leak upwards into the thoracic cavity, leading to a pancreatic pleural effusion.

- Occurrence: These complications are more commonly seen in chronic pancreatitis than in acute pancreatitis.

- Incidence: In about 14% of patients, both ascites and pleural effusion are present; in 18% of patients, only pleural effusion is observed.

- Symptoms: Patients typically present with progressive abdominal swelling and weight loss. Pain and vomiting are rare.

- Imaging: CT may show ascites, chronic pancreatitis, and a partially collapsed pseudocyst.

- Fluid Analysis:

- Paracentesis or thoracentesis may reveal non-infected fluid.

- Fluid has high amylase levels and a protein level > 25 g/L.

- Serum amylase may also be elevated.

- Serum albumin may be low, and accompanying liver disease may be present, which can confuse the diagnosis with hepatic ascites. Paracentesis helps differentiate.

- ERCP: This is the most useful method for detecting pancreatic leakage.

- Initial Treatment: Medical treatment includes antisecretory agents (such as somatostatin and its analogs), bowel rest, and Total Parenteral Nutrition (TPN).

- Approximately half of the patients respond to medical treatment.

- Surgical Intervention: If medical treatment is ineffective, surgical intervention may be necessary.

Chronic Pancreatitis Treatment

Medical Management:

- Pain Control: The primary approach is managing pain.

- Alcohol Abstinence: Alcohol consumption should be discontinued.

- Somatostatin Analogs: These may be administered.

- Narcotic Analgesics: Often required for pain relief.

- Exocrine Pancreatic Enzymes: These can be given to aid digestion.

- Diabetes Management:

- Ketoacidosis is rare in diabetes associated with chronic pancreatitis.

- Insulin is preferred for diabetes; oral antidiabetic medications are generally ineffective.

- ERCP: Can also be useful in treatment.

Surgical Management:

Indications for Surgery:

- Pain: Persistent pain or pain unresponsive to medical treatment is the most common indication.

- Biliary Obstruction: Due to obstruction of the bile duct.

- Duodenal Obstruction: Blockage of the duodenum.

- Colonic Obstruction: Blockage of the colon.

- Suspicion of Pancreatic Cancer: To rule out malignancy.

- Splenic or Portal Vein Obstruction: Associated with portal hypertension.

- Pseudocyst: Complications related to pseudocysts.

Surgical Procedures:

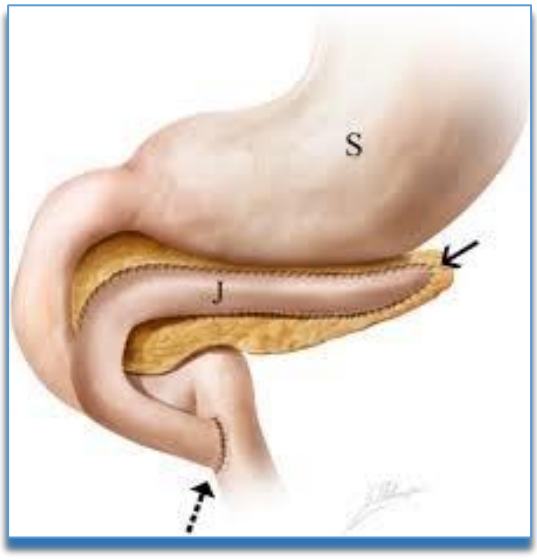
Drainage Surgeries:

- Indication: Often performed for pain palliation.

- Criteria: Typically done when the diameter of the Wirsung duct is greater than 7 mm; provides pain relief in about 80% of cases.

- Procedure:

- Lateral Pancreaticojejunostomy (Puestow Procedure):A common surgical method for pain relief in chronic pancreatitis.



Resections:

- Distal Pancreatectomy: Removal of the distal part of the pancreas.

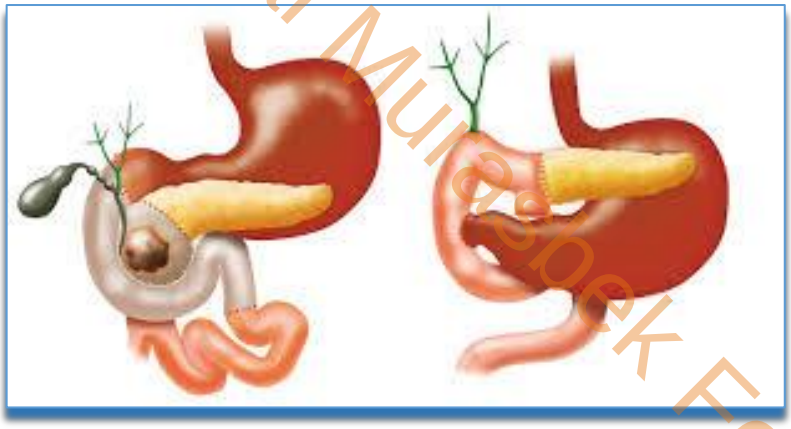
- Subtotal Pancreatectomy: Partial removal of the pancreas, which can be:

- 50% Subtotal: Removal of 50% of the pancreas.

- 80-85% Subtotal: Removal of 80-85% of the pancreas.

- 95% Subtotal:Removal of 95% of the pancreas.

- Total Pancreatectomy: Complete removal of the pancreas.



- Pancreaticoduodenectomy (Whipple Procedure): Removal of the pancreas head, part of the duodenum, gallbladder, and sometimes part of the stomach.

Transplantation:

- Pancreas Transplantation: Very rarely used for chronic pancreatitis.

EXOCRINE PANCREAS TUMOR(CANCER)

RISK FACTORS

- Smoking is a significant risk factor for pancreatic cancer.

- Type II diabetes can increase the risk of developing pancreatic cancer.

- The sudden increase in insulin requirements in a newly diagnosed diabetic patient or a diabetic patient should raise suspicion of pancreatic cancer.

- It has been shown that the risk of developing pancreatic cancer is increased up to 20 times in patients with chronic pancreatitis. This risk increase is independent of the type of chronic pancreatitis, although the mechanism of carcinogenesis is not known.

- As with other gastrointestinal cancers, a diet high in fat and low in vegetables and fruits may be associated with an increased risk of pancreatic cancer.

- The risk of developing pancreatic cancer is increased in individuals with a family history, especially if two or more first-degree relatives have pancreatic cancer.

- Coffee and alcohol consumption have also been investigated as potential risk factors, but no definitive relationship has been established.

GENETIC FEATURES AND MOLECULAR BIOLOGY

- Pancreatic carcinogenesis is associated with the accumulation of numerous mutations in oncogenes and tumor suppressor genes, as well as errors in the production of growth factors and their receptors.

- Most pancreatic cancers have three or more mutations.

- The K-ras oncogene is found in approximately 90% of tumors.

- The K-ras oncogene is the most frequently mutated gene in pancreatic cancers.

- The HER2/neu oncogene may also be overexpressed in pancreatic cancers.

- Additionally, deletions and/or mutations can occur in tumor suppressor genes such as p53, p16, and DPC4.

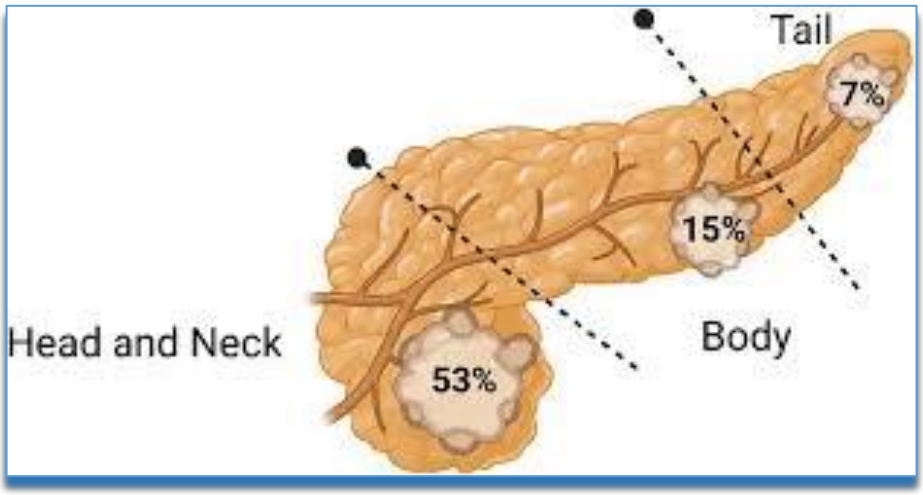
- It is estimated that approximately 10% of pancreatic cancers are associated with genetic predisposition.

PATHOLOGY

The precursor lesion of pancreatic cancer is termed pancreatic intraepithelial neoplasia (PanIN).

- Three stages of PanIN have been identified. In the pancreatic cancer progression model, the histological progression from normal ductal epithelium to low-grade PanIN and high-grade PanIN is associated with the accumulation of specific genetic changes.

- Early changes involve HER2/neu and K-ras mutations, while changes in the intermediate stage include p16 mutations. Changes associated with in situ or early invasive cancer are related to mutations in p53, BRCA2, and DPC4.



Pancreatic Adenocarcinomas

- Roughly two-thirds (about 70%) of pancreatic adenocarcinomas originate from the pancreatic head or uncinate process, 15% from the body, and 10% from the tail, with the remaining cases showing diffuse involvement in the gland.

- Pancreatic adenocarcinomas are typically larger than 3 cm at the time of diagnosis and often present with both nodal and distant metastases.

- Tumors located in the body and tail are generally larger at diagnosis and thus have a lower chance of resection.

- Approximately 70-80% of all pancreatic neoplasms are ductal adenocarcinomas and their variants.

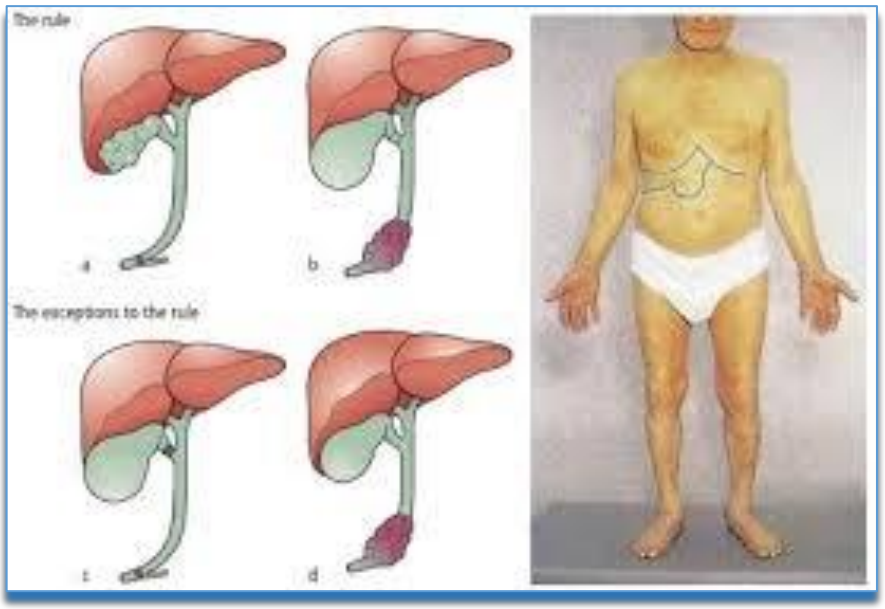
Symptoms and Clinical Findings

- Due to the deep location of the pancreas within the abdomen, the early symptoms of pancreatic cancer are often vague, leading to a prolonged asymptomatic period before typical symptoms develop and the disease progresses significantly.

- Most patients present with weight loss, pain, and jaundice.

- The most common symptom and finding is weight loss.

- On physical examination, weight loss is prominent, and scleral icterus (jaundice) may be noted.



-Courvoisier's Sign- This refers to the palpation of a non-tender, distended gallbladder in a patient with jaundice. Its presence primarily raises suspicion of pancreatic head cancer.

- In obstructive jaundice, ultrasound may reveal an enlarged common bile duct, a distended gallbladder, and no stones, suggesting pancreatic head cancer.

- Pain is a common symptom associated with pancreatic cancer and is often the first symptom.

- Therefore, in elderly patients with newly onset diabetes, especially if accompanied by vague abdominal pain, pancreatic cancer should be investigated.

- Recent onset diabetes may also be related to pancreatic cancer.

- Unexplained migratory thrombophlebitis (Trousseau's syndrome) can be associated with pancreatic and other cancers.

- There is no sufficiently sensitive and specific serum marker to aid early diagnosis of pancreatic cancer.

- In tumors located in the pancreatic head, direct hyperbilirubinemia and elevated alkaline phosphatase can indicate biliary obstruction.

- Prolonged prothrombin time due to vitamin K deficiency may be seen in those with long-standing obstructive jaundice.

- The two most commonly used serum markers for pancreatic cancer are CEA and CA19-9.

- Both are often elevated in advanced disease but are typically normal in the early stages where curative treatment may be possible. Therefore, they are not very useful in screening or early diagnosis of patients with vague symptoms or in high-risk groups.

INSTRUMENTHAL METHODS

For patients with jaundice, the initial and appropriate diagnostic imaging method is abdominal ultrasound.

- The detection of stones in the gallbladder along with dilation of the bile ducts suggests that choledocholithiasis may be the cause of the jaundice.

- If no gallstones are detected, malignant obstruction of the bile duct should be considered, and a CT scan should be performed next.

- For patients without jaundice but with suspected pancreatic cancer, ultrasound is not suitable, and the first diagnostic method should be CT.

-The current preferred imaging method for diagnosing and staging pancreatic cancer is dynamic contrast-enhanced spiral CT.

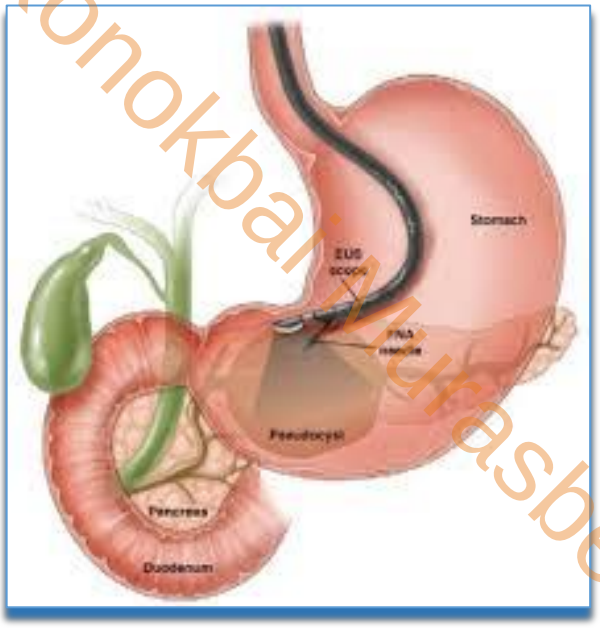
- On CT, findings such as invasion of the hepatic artery or superior mesenteric artery, enlarged lymph nodes beyond resection margins, distant organ invasion, or distant metastasis can prevent resection.

- The success rate of CT in identifying unresectable disease is between 90-95%.

- MRI has the same sensitivity and specificity as CT, but CT is more commonly preferred.

- PET is increasingly used and can be helpful in distinguishing pancreatic cancer from chronic pancreatitis.

Endoscopic ultrasound (EUS) can be used to detect small pancreatic masses not visible on CT.



- EUS is also a sensitive method for identifying portal and superior mesenteric vein invasion.

- For evaluating patients with obstructive jaundice where a mass is not detected on CT, ERCP can be useful.

- The detection of "double duct sign," indicating strictures in both the bile and pancreatic ducts on ERCP, strongly suggests pancreatic cancer.

TREATMENT

- In pancreatic cancer patients considered operable by all imaging methods, at least 20% may have small peritoneal or liver metastases that are not detected by imaging and are found during surgery.

-To avoid unnecessary laparotomies, laparoscopy is recommended at the start of the operation.

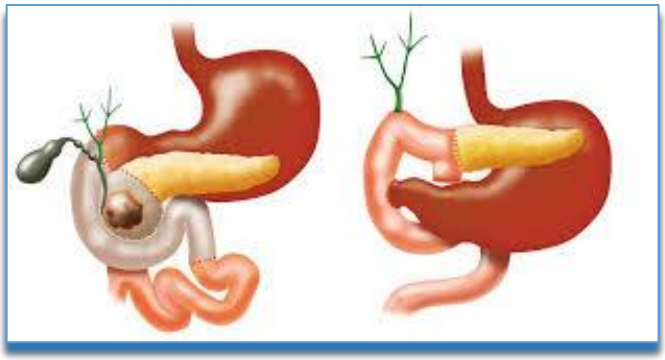
-For patients suspected of pancreatic cancer through clinical and imaging methods, tissue diagnosis is not required before pancreaticoduodenectomy.

- However, for patients not considered for resection, biopsy is important for tissue diagnosis as these patients may receive palliative chemotherapy and radiotherapy.

-Surgical treatment is the only option for patients who respond poorly to radiotherapy and chemotherapy.

Curative Surgeries

- Pancreaticoduodenectomy (Whipple procedure), which can be performed as a pylorus-preserving type.



- Total pancreatectomy.

- Extended Whipple resection.

- Distal pancreatectomy can be applied for lesions in the tail.

Palliative Surgeries

- In patients with pancreatic head cancer, palliative treatment may involve addressing three conditions that require palliation: jaundice, gastric outlet obstruction, and pain.

Findings Contraindicating Resection:

- Liver metastases
- Celiac lymph node involvement
- Peritoneal implants
- Transverse mesocolon invasion
- Involvement of lymph nodes at the liver hilum

Findings Not Contraindicating Resection:

- Invasion of the duodenum or distal stomach
- Involvement of peripancreatic lymph nodes
- Lymph node involvement along the porta hepatis that can be removed with the specimen

The most common causes of death are sepsis, bleeding, and cardiovascular events.Postoperative complications such as delayed gastric emptying, pancreatic fistula, and bleeding are still quite frequent.

- Delayed gastric emptying is commonly seen after pancreaticoduodenectomy and is treated conservatively unless complete gastric outlet obstruction is identified through contrast studies.

- Octreotide, a synthetic analogue of somatostatin with a longer half-life, is a pharmacological treatment used to reduce pancreatic enzyme secretion and therefore reduce pancreatic fistula after pancreatic resection.

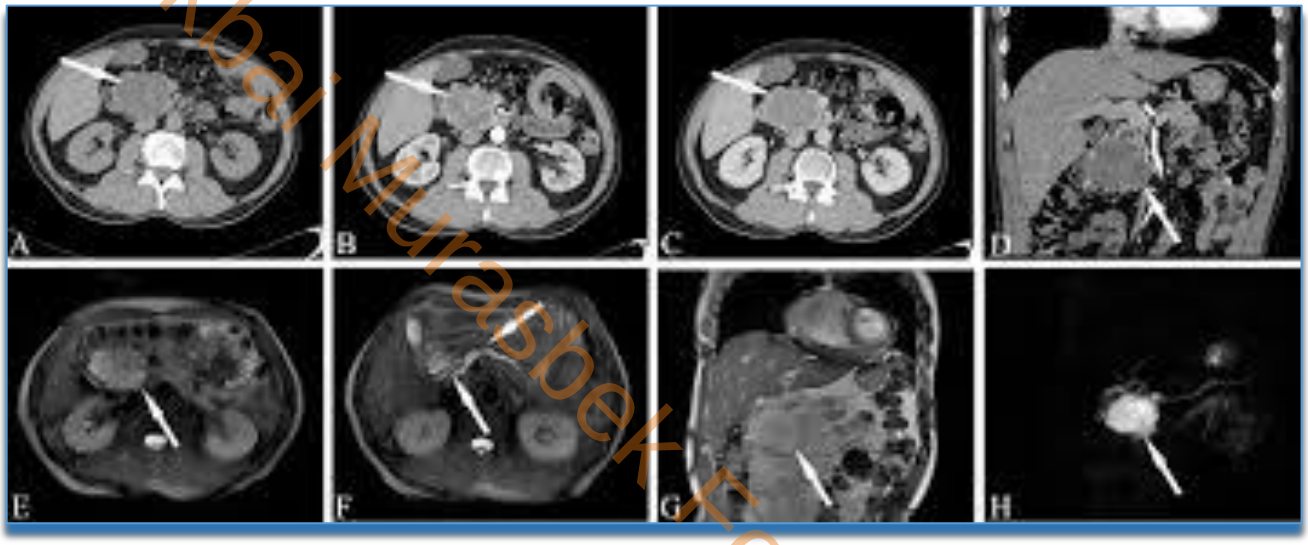
- Postoperative bleeding may result from inadequate vessel ligation during surgery.

- Bleeding can also be related to erosion of a retroperitoneal vessel due to a biliopancreatic leak.

The “median survival” after pancreaticoduodenectomy is 22 months.

- Even those with long-term (5-year) survival often die due to late recurrence of pancreatic cancer.

Cystic Tumors of the Exocrine Pancreas



Most benign exocrine pancreatic tumors are cystic, but not all cystic tumors, which constitute 10-15% of pancreatic tumors, are benign. When a patient presents with a cyst in the pancreatic localization, the possibility of a pancreatic cystic neoplasm should be ruled out. The presence of a solid component within the cyst, the presence of septations within the cyst, and the absence of a history of pancreatitis should be warning signs that the lesion may be a cystic neoplasm.

Pancreatic cystic neoplasms include benign serous cystadenoma, benign and malignant mucinous cystic neoplasms, and benign and malignant intraductal papillary-mucinous neoplasms.

- **Mucinous Tumors**

Approximately 45% of cystic tumors are mucinous. Mucinous cystadenoma is the most common benign tumor of the exocrine pancreas. Even though it is a benign tumor, it is considered to have malignant potential. These cysts are lined by columnar, mucin-producing, and sometimes papillary epithelium. They should be surgically removed before invasive malignancy develops. Survival rates are better even after malignant changes and invasion develop compared to ductal adenocarcinoma.

- **Serous Cystadenoma**

Approximately 20% of cystic pancreatic tumors are serous cystadenomas. These tumors are lined by epithelium containing glycogen in their cytoplasm and do not stain with mucin. Most are composed of multiple microscopic cysts, giving them a spongy appearance. Radiologically, they appear as well-circumscribed cystic masses with small septations, sometimes showing a central calcified scar. They are generally benign tumors with no malignant potential. They are mostly found in the body and tail of the pancreas and are asymptomatic. When localized in the

head of the pancreas, they can be symptomatic. If there is diagnostic suspicion and if they are symptomatic, surgical resection is indicated.

- **Intraductal Papillary Mucinous Neoplasm (IPMN)**

Intraductal papillary mucinous neoplasm (IPMN) is a newly identified type of cystic tumor of the pancreas. It can involve the major duct, smaller ducts, or both. Although it can be found throughout the pancreas, it is most commonly seen in the head of the pancreas. It is characterized by an enlarged pancreatic duct due to mucin production from the lesion. The diagnosis of IPMN can be made with high confidence if mucin is seen coming from a wide, fish-eye-shaped papilla during endoscopy. The ducts involved by IPMN are usually lined by columnar, mucin-producing cells that form papillary projections. It is believed that IPMNs can transform into carcinoma and are categorized as PanIN-1 lesions in the pancreatic cancer development model. Resection performed before the development of invasive malignancy is generally curative. However, the cure rate is relatively lower when resection is performed after the development of invasive malignancy.

ENDOCRINE PANCREAS NEOPLASMS

INSULINOMA

- The most common of the pancreatic islet cell tumors.

- Associated with 10% of MEN Type 1 cases, which involves widespread involvement.

- 80-90% are benign solitary adenomas; 10% are malignant.

Diagnosis

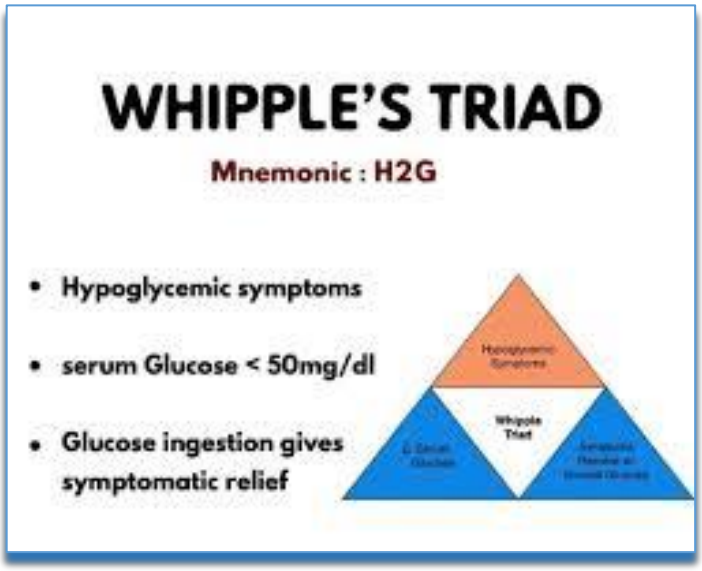
- Fasting hypoglycemia is the most significant clinical symptom.

- Whipple's triad is observed:

1. Symptoms of hypoglycemia occurring during fasting.

2. Blood glucose <50 mg/dl during symptoms.

3. Relief of symptoms with intravenous glucose administration.



- A plasma insulin/glucose ratio >0.4 is diagnostic.

- Elevated plasma C-peptide levels are present.

- If proinsulin level >50%, malignant insulinoma should be considered.

- Preoperative localization studies are crucial for all islet cell tumors.

  - CT scan is primarily used.

  - Endoscopic ultrasound is the most useful test for localization.

  - If necessary, selective angiography or selective venous sampling may be performed.

- Insulinomas are distributed equally among the head, body, and tail of the pancreas.

Treatment

- Tumor enucleation is performed to remove the tumor.

- Intraoperative ultrasound can be used.

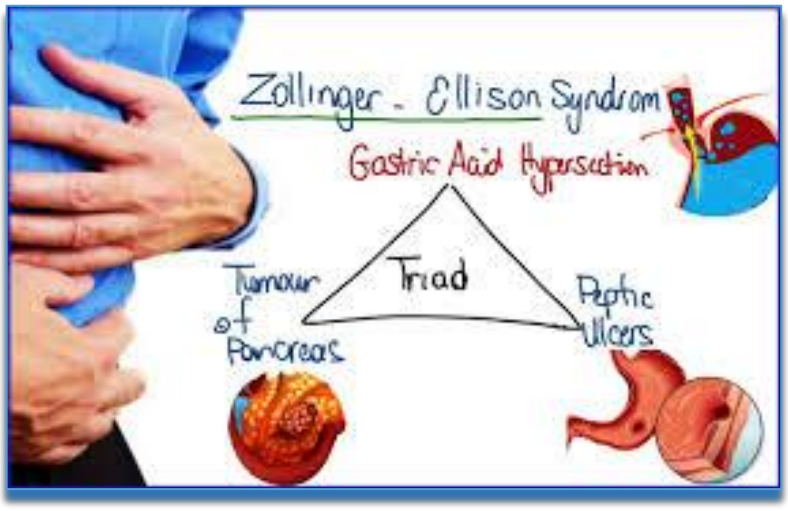
- In extensive cases, debulking surgery (tumor reduction) may be done.

- Medical treatment:

  - Diazoxide is the primary choice.

  - Streptozotocin is used for malignant cases with no chance of cure.

GASTRINOMA (ZOLLINGER-ELLISON SYNDROME)



- Characterized by excessive gastrin secretion leading to peptic ulceration.

- Most commonly found in the duodenum.

- 80% are sporadic and 20% are genetically inherited.

- Among genetic tumors, MEN1 syndrome is the most frequently associated syndrome.

- In MEN1 patients, gastrinoma is the most common pancreatic tumor.

- Sporadic gastrinomas are usually solitary and suitable for surgical treatment. In genetically inherited tumors, the tumor focus is multiple and resistant to surgical treatment.

- 50-60% are malignant.

- With metastatic disease, the 5-year survival rate is 40%.

- Patients with sporadic and completely resectable gastrinoma tumors have a greater than 90% chance of a cure.

Clinical Features

- Pain due to peptic ulcers.

- Diarrhea, gastric hypersecretion, and steatorrhea due to increased intestinal motility directly related to gastrin.

- Gastrointestinal bleeding, ulcer perforation, or pyloric obstruction.

- Dehydration and malnutrition.

Diagnosis

- Zollinger-Ellison syndrome should be ruled out in the presence of persistent peptic ulcer disease, severe esophagitis, and persistent secretory diarrhea.

- Diagnosis is made with high gastrin levels associated with high acid secretion.

- Fasting gastrin levels above 100 pg/ml are considered high; gastrin levels above 1000 pg/ml are almost diagnostic.

- High serum gastrin levels and a gastric juice pH below 2 confirm the diagnosis of gastrinoma.

- Low gastric pH is important for diagnosis because very high gastrin levels can also be seen in pernicious anemia or PPI users, where gastric acid production in the mucosa is not present.

If Zollinger-Ellison syndrome is suspected or gastrin levels are between 100-1000 pg/ml, a “secretin stimulation test” is performed to differentiate Zollinger-Ellison syndrome from other conditions with elevated gastrin levels.An increase of 200 pg/ml in serum gastrin levels after intravenous administration of secretin (2 U/kg) supports the diagnosis of gastrinoma.

- In Zollinger-Ellison syndrome, G-cell hyperplasia and gastric outlet obstruction lead to increased gastrin secretion.

- Gastrinomas are found within the gastrinoma triangle in 70-90% of patients.

- The corners of the gastrinoma triangle are the cystic duct-common bile duct junction, the second and third portions of the duodenum, and the pancreatic head.

- Initial localization studies typically involve a CT scan.

- The most useful test for localization is somatostatin (octreotide) receptor scintigraphy.

- Endoscopic ultrasound can also be performed.

- Approximately 50% of gastrinomas have metastases to surrounding lymph nodes and/or the liver.

- The presence of lymph node metastases does not affect prognosis.

Treatment

- A localized tumor must be surgically removed.

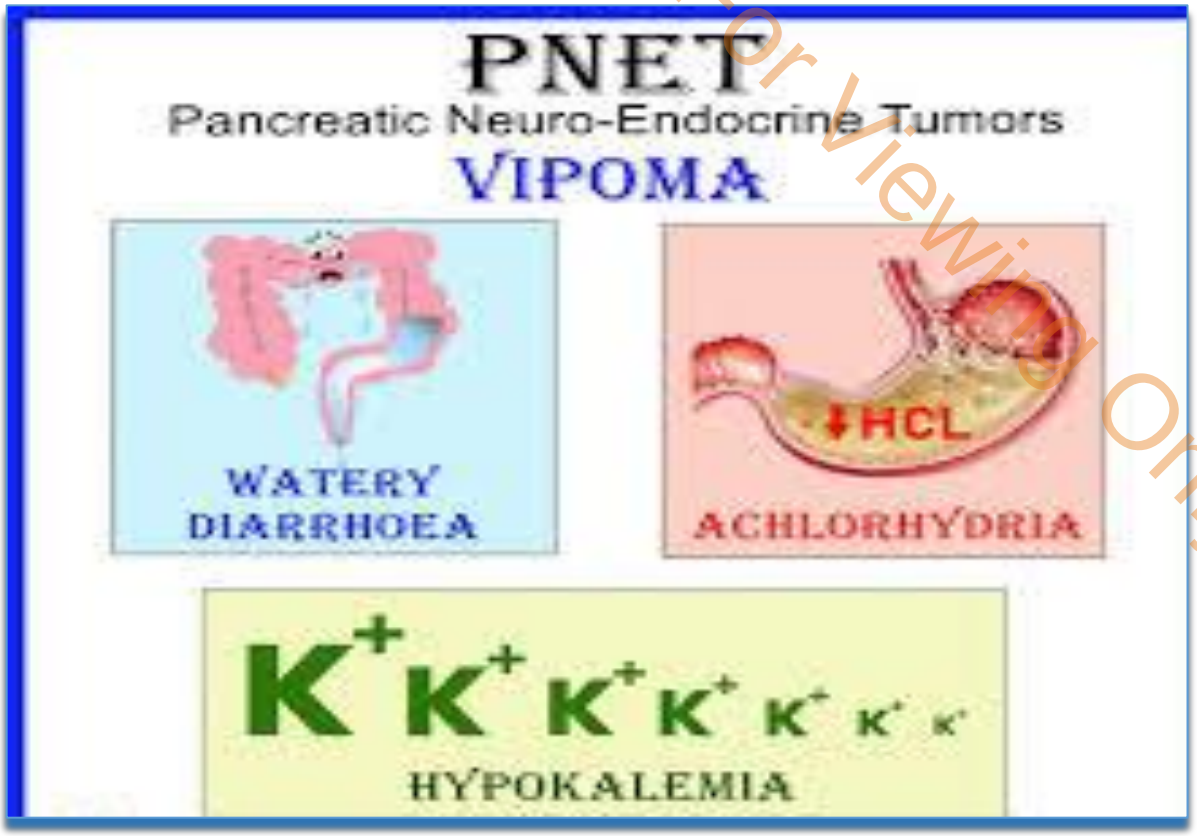
- If resection is not possible, PGV (palliative gastric vagotomy) may be considered during surgery.

- For widespread tumors, especially in MEN1 syndrome, medical treatment is applied.

- The first-line medication in medical treatment is omeprazole.

- Acid-reducing surgeries and medications have made total gastrectomy unnecessary.

VIPOMA (Pancreatic Cholera, Verner-Morrison Syndrome, WDHA Syndrome)



- Typically, the tumor is solitary in 80% of cases, often located in the body or tail of the pancreas.

- The likelihood of malignancy is 50%.

- The presence of" tea-colored diarrhea" suggests VIPoma.

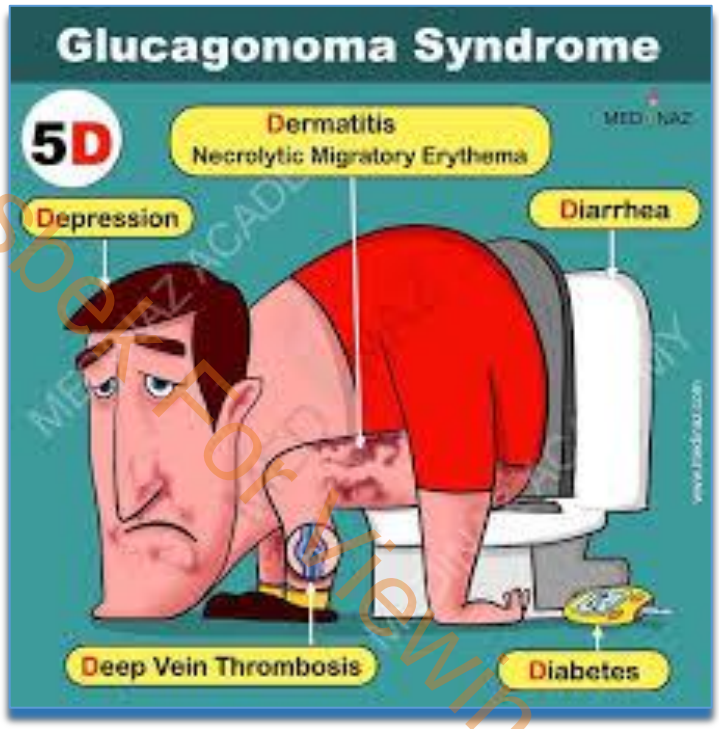
- Hypokalemia is typical despite metabolic acidosis.

- Endoscopic ultrasound is the most useful test for localization studies.

- Surgical excision is preferred if possible.

- If surgery is not successful, “debulking” can help improve diarrhea.

GLUCAGONOMA



- Originates from alpha cells of the pancreas.

- Rare occurrence.

- Patients present with severe diabetes, weight loss, dermatitis, anemia, stomatitis, and glossitis.

- Characteristic “necrotizing migratory erythema” (erythema with central healing and peripheral spreading) is observed.

- Deep vein thrombosis in the lower extremities is common.

- 65-70% are malignant.

- Tumors are generally large (average diameter at diagnosis is 4 cm).

- Treatment includes surgical resection or debulking.

- Adjuvant therapy is effective.

- For non-resectable tumors, streptozocin and somatostatin are administered.

SOMATOSTATINOMA

- Somatostatin-producing pancreatic tumors are often characterized by a combination of mild diabetes mellitus, malabsorption, diarrhea, and gallbladder dilation, often accompanied by gallstones.

- Severe weight loss may occur.

- Tumors are often located in the head of the pancreas and are typically malignant.

- Metastases are commonly present at the time of diagnosis.

- Localized tumors can be treated with surgery.

- For “metastatic disease”, medical treatments such as streptozocin and dacarbazine can be used.

NON-FUNCTIONAL PANCREATIC ISLET CELL TUMORS

- These are pancreatic islet cell tumors that do not produce any hormones.

- The most common non-functional islet cell tumor, after insulinoma, is the non-functional islet cell tumor.

- Since they do not secrete any hormones, they generally present as malignant forms.

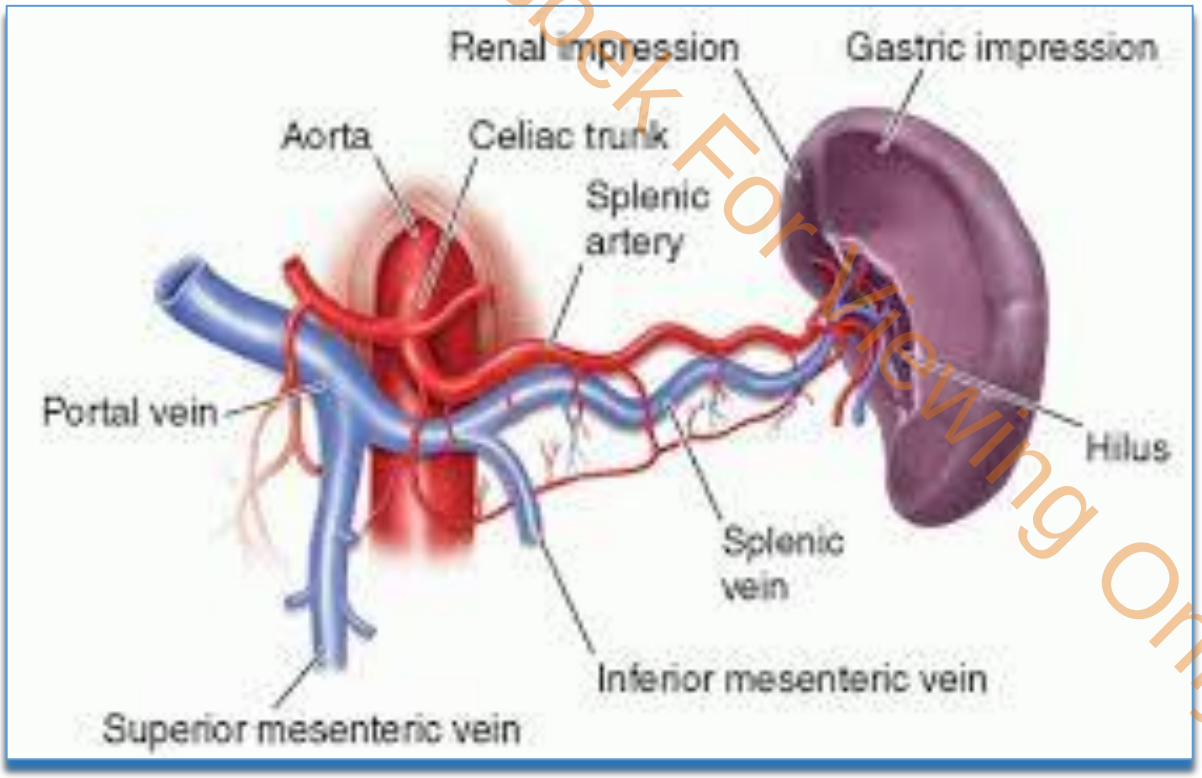
- Some tumors have been shown to secrete pancreatic polypeptide.

Chapter

9.

SPLEEN DISEASES AND SURGREY

Anatomy of the Spleen



- It is the largest reticuloendothelial organ in the body.

- Weight: 80-300 grams.

- Its blood supply is provided by the splenic artery, which arises from the celiac trunk. The splenic artery is 70% distributed (short main trunk with numerous long branches at the hilum) and 30% magistral (long main trunk with numerous short terminal branches at the hilum).

- There are 4 ligaments that hold the spleen in place:

  - Splenocolic (lienocolic) ligament

  - Gastrosplenic ligament (contains short gastric vessels)

  - Phrenosplenic ligament

  - Splenorenal (lienorenal) ligament

- The most common embryological anomaly is an accessory spleen (14-30%).

- Most commonly located at the splenic hilum (80%).

- Technetium-99m sulfur colloid radionuclide scan reveals splenic localization and size.

Functions of the Spleen

1. Filtration

The filtration function of the spleen is one of its primary roles in maintaining blood health and immune function. The spleen acts as a blood filter, removing abnormal, old, or damaged cells and foreign particles from the bloodstream. Here's a detailed overview of spleen's filtration function:

- Erythrocyte membrane abnormalities: Spherocytes (hereditary spherocytosis)

- Erythrocyte surface pits and craters: Sickle cells

- Howell-Jolly bodies (nuclear remnants)

- Hemoglobin C crystals

- Heinz bodies (denatured hemoglobin)

- Antibody-coated erythrocytes

- Pappenheimer bodies (iron granules)

- Antibody-coated platelets

- Acanthocytes (spur cells)

- Antibody-coated white blood cells

- Aged erythrocytes

- Antigen particles

2. Immunological Functions

- Opsonization: The spleen plays a critical role in the opsonization process, especially against encapsulated microorganisms.

- Antibody Synthesis: The spleen is crucial for the synthesis of antibodies, particularly IgM.

- Protection Against Infections: It provides protection against infections, notably those caused by Streptococcus pneumoniae (pneumococcus), Haemophilus influenzae, and Neisseria meningitidis (meningococcus).

- Postsplenectomy Infections: The most significant complication after splenectomy is infections, which have high mortality rates. These infections are most frequently seen within the first two

years after splenectomy, but the risk remains high for a lifetime. The risk of infections after splenectomy is about 3-3.5%.

- Mortality Risk: Among patients who develop postsplenectomy infections, the mortality risk is between 40-50%. The highest mortality risk is observed in patients with thalassemia major and sickle cell anemia.

- Immunoglobulin Levels: After splenectomy, levels of immunoglobulin M (IgM) and functional immunoglobulin G (IgG) decrease.

- Properdin and Tuftsin Levels: The levels of properdin and tuftsin, which function as opsonins, also decrease after splenectomy.

3. Storage Function

- Platelet Storage: The spleen stores about one-third of the body's total platelets.

Hypersplenism

- Not every case of splenomegaly or hypersplenism requires surgical intervention.

- Splenomegaly refers to the condition where the spleen is larger than normal; it is purely an anatomical term.

- When the spleen size exceeds 15 cm, it is termed splenomegaly; if it exceeds 20 cm, it is termed massive splenomegaly.

- Hypersplenism is a general term that describes various pathological conditions arising from the spleen's overactivity.

Indications for Splenectomy

- The most common indication for splenectomy is spleen trauma.

- Elective splenectomy is most commonly performed for Immune Thrombocytopenic Purpura (ITP).

- Purposes of splenectomy:

  - To control the disease

  - Due to chronic and severe hypersplenism

  - Additionally, splenectomy can be performed for staging in Hodgkin's lymphoma.

1. Indications for Splenectomy (For Control Disease):

- Hereditary Spherocytosis

- Elliptocytosis

- Autoimmune Anemia

- Thrombotic Thrombocytopenic Purpura

- Spleen Rupture (spontaneous or massive trauma)

- Immune Thrombocytopenic Purpura (ITP)

- Primary Cysts or Tumors (spleen abscess, echinococcal cyst)

- Sickle Cell Anemia

- Hb SC Disease

- AIDS

2.Indications for splenectomy Due to Chronic and Severe Hypersplenism:

- Hairy Cell Leukemia

- Lymphoproliferative Diseases (Chronic Lymphocytic Leukemia (CLL), Non-Hodgkin Lymphoma)

- Felty Syndrome

- Agnogenic Myeloid Metaplasia

- Thalassemia Major

- Gaucher Disease

- Hemodialysis-associated Splenomegaly

- Splenic Vein Thrombosis (bleeding esophagogastric varices)

Splenectomy

- Can be performed either openly or laparoscopically.

- If the spleen is longer than 22 cm or weighs more than 1600 grams, hand-assisted laparoscopic or open splenectomy is appropriate.

- Laparoscopic splenectomy has similar hematological outcomes compared to open surgery.

- Compared to open splenectomy, laparoscopic splenectomy results in less intraoperative blood loss, shorter hospital stays, and lower morbidity rates.

- All patients must receive vaccines for Haemophilus influenzae type b, meningococcus, and polyvalent pneumococcus before surgery.

- Ideally, these vaccinations should be administered 15 days before the surgery.

- In emergency splenectomies, due to the transient immunosuppression observed postoperatively, vaccinations should be given at the earliest 2 weeks after the surgery.

Complications of Splenectomy:

- Atelectasis in the Left Lung Base: This is the most common complication.

- Injury to Adjacent Organs: The most frequently injured are the greater curvature of the stomach and the tail of the pancreas.

- Postoperative Bleeding: Usually due to inadequate hemostasis.

- Subdiaphragmatic Abscess: The use of drains particularly increases the risk.

- Thrombocytosis: Sometimes thrombocytosis can exceed 1,000,000, requiring anticoagulant therapy.

Postsplenectomy Sepsis

- Higher Risk in Children: The risk of sepsis is higher in children post-splenectomy.

- Most Common Pathogen: Streptococcus pneumoniae (80-90% of cases).

- Risk Factors for Sepsis: The most important factor determining the risk of sepsis after splenectomy is the underlying reason for the splenectomy. The risk is particularly high in malignant diseases, hematologic diseases, and reticuloendothelial system diseases.

- Increased Risk: Particularly if splenectomy is performed for reticuloendothelial system disease.

- Treatment: High-dose antibiotics are administered.

- Occurrence: Some cases can develop within the first 2 years postoperatively.

Vaccination Guidelines

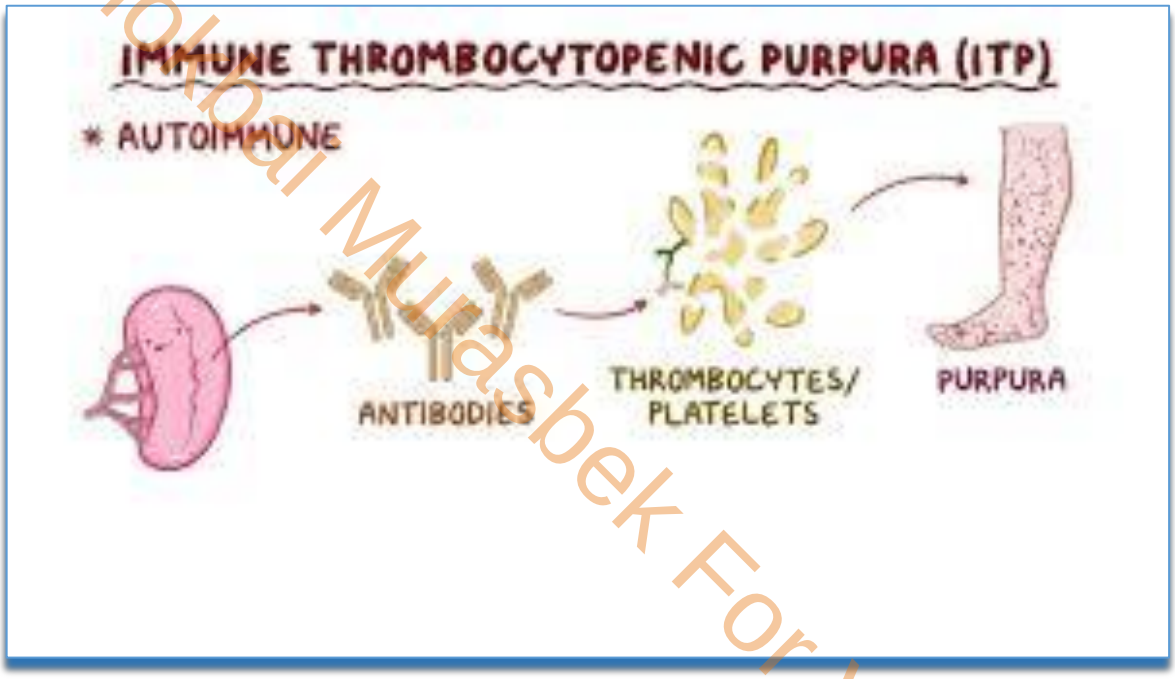
- Elective Cases: Vaccination should be done 2 weeks before the surgery.

- Emergency Splenectomy: Vaccinations for Haemophilus influenzae B and meningococcus should be given 2 weeks after the surgery.

- High-Risk Patients: A pneumococcal vaccine booster should be administered after 5 years.

- Booster Efficacy: There is no evidence that boosters reduce the incidence of infections.

Immune Thrombocytopenic Purpura (ITP)



Immune Thrombocytopenic Purpura (ITP): A condition characterized by a low platelet count (thrombocytopenia) due to the immune system mistakenly attacking and destroying platelets. It typically presents with easy bruising, petechiae, and mucosal bleeding. The diagnosis is made by excluding other causes of thrombocytopenia and confirming the presence of isolated low platelet counts without other abnormalities in the blood. Treatment may include corticosteroids 1mg/kg, immunoglobulins, and in chronic cases, splenectomy or other immunomodulatory therapies.

Platelet Count Monitoring:

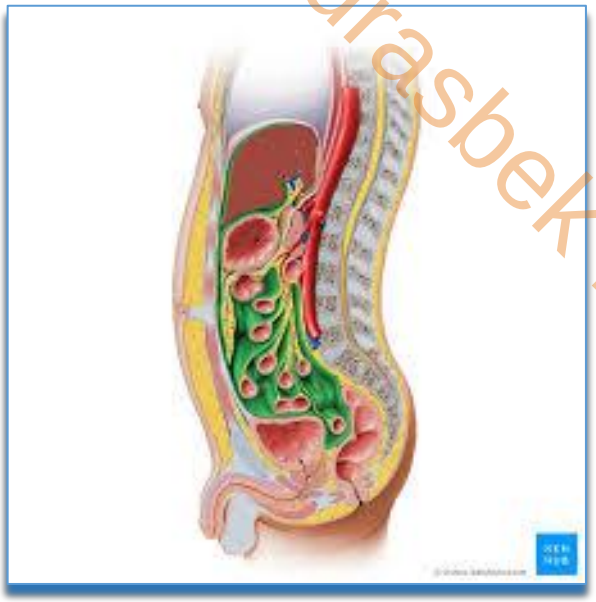
-Platelet counts above 50,000 can be monitored.

-Counts between 30,000 and 50,000 require close monitoring.

-Counts below 30,000 necessitate treatment.

Chapter
10.

Peritonitis and Surgery



- **Definition:** Inflammation of the peritoneum, either in its entirety or in part.

- **Causes:**

- Primary Peritonitis:Caused by bacterial, chlamydial, fungal, or mycobacterial infections without gastrointestinal perforation.

- Secondary Peritonitis: Results from gastrointestinal perforation, involving purulent exudate from enteric sources within the abdomen.

- Common Causes of Secondary Peritonitis: Peptic ulcers, acute appendicitis, colonic diverticulitis, and pelvic inflammatory disease.

"**Blumberg's Sign"** is a clinical sign used in physical examination to assess for peritoneal irritation, which may indicate acute abdominal conditions such as appendicitis. It is often described as follows: During the physical examination, the examiner presses down gently and slowly on the abdomen, and then quickly releases the pressure. If the patient experiences a sharp pain upon the release of pressure, this is considered a positive Blumberg's sign.

Primary Peritonitis

- Spontaneous Bacterial Peritonitis:

- A peritoneal infection secondary to a non-peritoneal source, often hematogenous.

- Defined as a bacterial infection of ascitic fluid when no intra-abdominal surgical infection source is present.

- Most commonly occurs in adults with liver cirrhosis, and less frequently in lupus patients.

- In adults, common pathogens include aerobic enteric bacteria such as Escherichia coli and Klebsiella pneumoniae.

- In children, it is most common in those with nephrotic syndrome, with Streptococcus pneumoniae being the primary pathogen. Other possible pathogens include streptococci and S. aureus.

- Primary peritonitis is usually a monomicrobial infection.

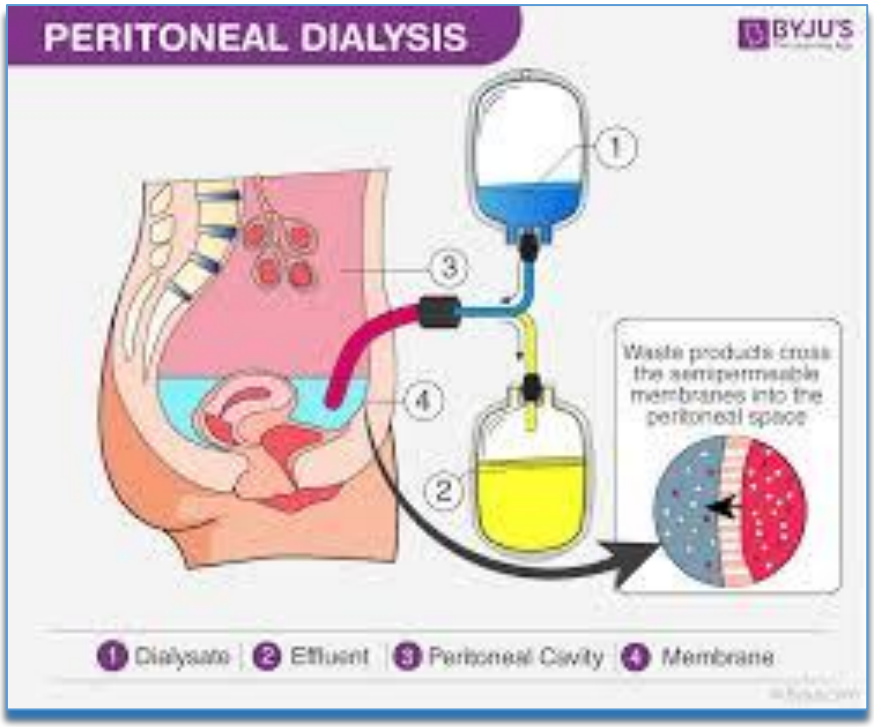
- Diagnosis: In a patient with low protein ascites, the presence of supporting symptoms (abdominal pain, fever, and leukocytosis) and an increased neutrophil count in ascitic fluid (Sabiston >250 neutrophils/mm^3; Schwartz >100 neutrophils/mm^3) confirms the diagnosis.

- Ascitic fluid culture should be monomicrobial; polymicrobial growth, especially with Gram-negative enterics, suggests secondary peritonitis.

- Treatment: Broad-spectrum antibiotics, such as third-generation cephalosporins.

Peritoneal Dialysis-Associated Peritonitis

- Overview:One of the most common complications of Continuous Ambulatory Peritoneal Dialysis (CAPD).



- Incidence: At least one peritonitis episode occurs every 1-3 years.

- Major Cause of CAPD Failure: Refractory or recurrent peritonitis.

- Diagnosis: Characterized by abdominal pain, fever, cloudy dialysis fluid, and elevated leukocytes >100/mm^3 with >50% neutrophils.

- Pathogens: >75% Gram-positive bacteria, with S. epidermidis being the most frequent (30-50%).

- Treatment: Intraperitoneal antibiotics are administered first. Infections are typically resolved with antibiotic treatment guided by culture results.

- Infections caused by S. aureus, Gram-negative bacteria, and fungi have lower success rates.

- For refractory or recurrent peritonitis, the catheter should be removed.

Tuberculous Peritonitis

- **Pathogenesis:** The tuberculosis bacillus may reach the peritoneum via hematogenous spread from infected organs or through transmural spread from affected intestinal walls, tuberculous salpingitis, or nephritis.

- Clinical Presentation:

- Fever, ascites, abdominal pain, and weakness, which is more common (80% of cases).
- Ascites is absent.

- **Diagnosis for tuberculous peritonitis:**

  - Ascitic fluid is predominantly lymphocytic.

  - High protein concentration in ascitic fluid.

  - Serum-ascitic albumin gradient (SAAG) is low (<1.1).

  - Ascitic fluid glucose is <30 mg/dL.

  - **Definitive Diagnosis:** Made through laparoscopic peritoneal biopsy.

- Treatment: Antituberculous agents are used. Surgical interventions are reserved for cases where needle biopsy is insufficient or to address complications such as fecal fistulas.

Intraabdominal Abscesses

- Definition: Intraabdominal abscesses are localized intra-abdominal infections with a significantly better prognosis compared to diffuse peritonitis. The formation of an abscess indicates a successful peritoneal defense mechanism against infectious agents, which helps contain bacteria and prevent systemic infection or sepsis.

- **Left Subphrenic Abscesses**

- Characteristics: These are the most common type of upper abdominal abscesses. They can develop following splenectomy (especially if the splenic fossa was drained) or due to pancreatitis.

- Symptoms: May present with left costal tenderness, left shoulder pain (Kehr's sign), left pleural effusion, and restricted movement of the left diaphragm.

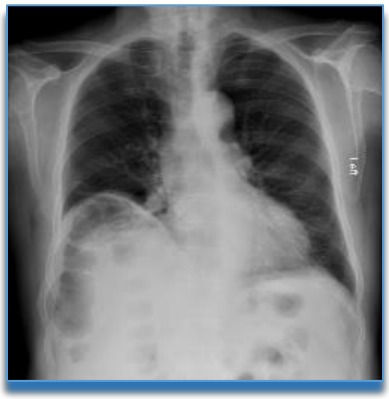
- **Lesser Sac (Omental) Abscesses**

- Characteristics: Often arise secondary to pancreatic abscesses or infections of pancreatic

pseudocysts. They can also develop due to perforation of gastric ulcers, duodenal ulcers, or gastric cancer.

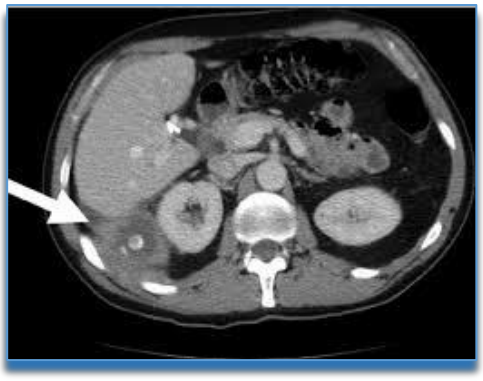
- Prognosis: These abscesses have the worst prognosis and are the rarest.

- **Right Subphrenic Abscesses**



- Characteristics: Commonly occur secondary to rupture of hepatic abscesses.

Right Subhepatic Abscesses



- Characteristics: Often caused by complications from gastric ulcer surgeries. They can also develop after biliary tract surgeries, and less frequently after colon surgery or appendicitis.

- **Interloop (Midabdominal) Abscesses**

- Characteristics: Located between segments of the small intestine, mesentery, abdominal wall,

and omentum, these are multiple abscesses.

- **Diagnosis:** They are the most difficult type of intra-abdominal abscess to diagnose.

- **Pelvic Abscesses**

- **Causes:** The most common cause is perforation of colon diverticula. They can also develop following pelvic inflammatory disease, perforated appendicitis, or generalized peritonitis.

Treatment of Intraabdominal Abscesses

- **Antibiotic Therapy:** Treating intraabdominal abscesses with antibiotics alone is usually insufficient.

- **Drainage:** Once localized purulence occurs, it must be drained to achieve effective treatment.

- **Treatment Approach:** The treatment of intraabdominal abscesses involves appropriate antibiotic therapy and adequate drainage.

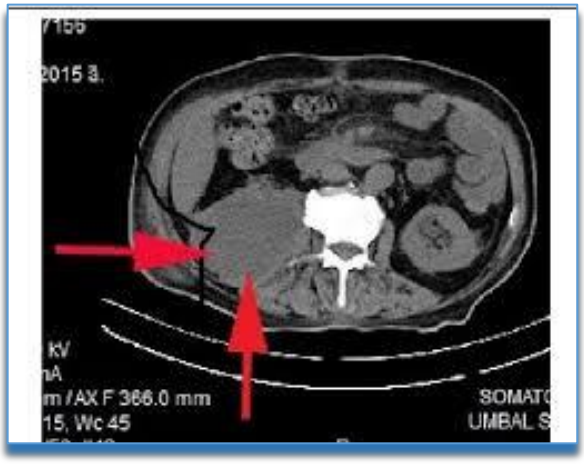
- **Drainage Methods**: Percutaneous drainage guided by ultrasound (USG) or computed tomography (CT) is generally preferred.

- **Surgical Drainage: If** percutaneous drainage is not feasible or does not provide adequate drainage, open surgical drainage is performed.

Retroperitoneal Abscesses

- **Causes:** Retroperitoneal abscesses most commonly develop due to kidney diseases. They can also arise from gastrointestinal conditions such as appendicitis, diverticulitis, pancreatitis, and Crohn's disease.

- **Symptoms:** The most common symptoms include abdominal or lower back pain, fever, weakness, and weight loss.



- **Diagnosis:** The diagnosis is made with computed tomography (CT), which shows a low-density mass in the retroperitoneum along with surrounding inflammation; gas may be present in one-third of cases.

- **Treatment:** Management involves appropriate antibiotics and adequate drainage.

Chapter
11.

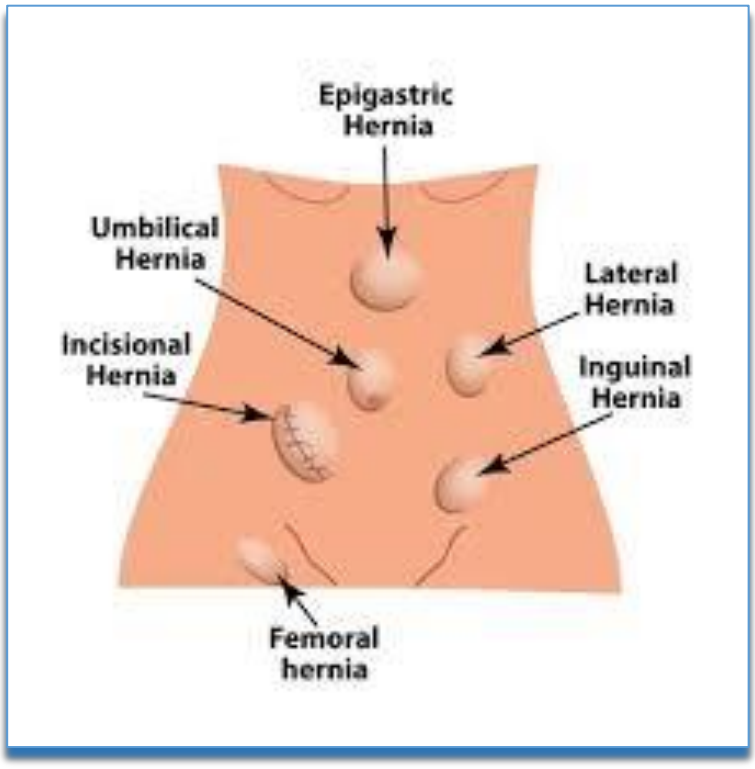
Abdominal Wall Hernias and Surgery

Hernias occur when abdominal organs, along with the peritoneum, protrude through weak points in the abdominal wall into the subcutaneous tissue.

- **Hernia Sac:** The peritoneum surrounding the herniated organ is called the hernia sac.

- **Common Contents:** Intraperitoneal organs that move freely in the abdomen can easily enter the hernia sac. Retroperitoneal organs, except for the pancreas, can also enter the sac. The omentum and small intestines are most commonly found within the hernia sac. Gender: 75% of hernias occur in men, and 25% in women. The prevalence of hernias increases with age.

- Types of Hernias:



- **Inguinal Hernias:** The most common type, particularly the indirect inguinal hernia, which is prevalent among men, women, and children.

- **Incisional Hernias:** The second most common type.

- **Umbilical Hernias:** These are the third most common type.

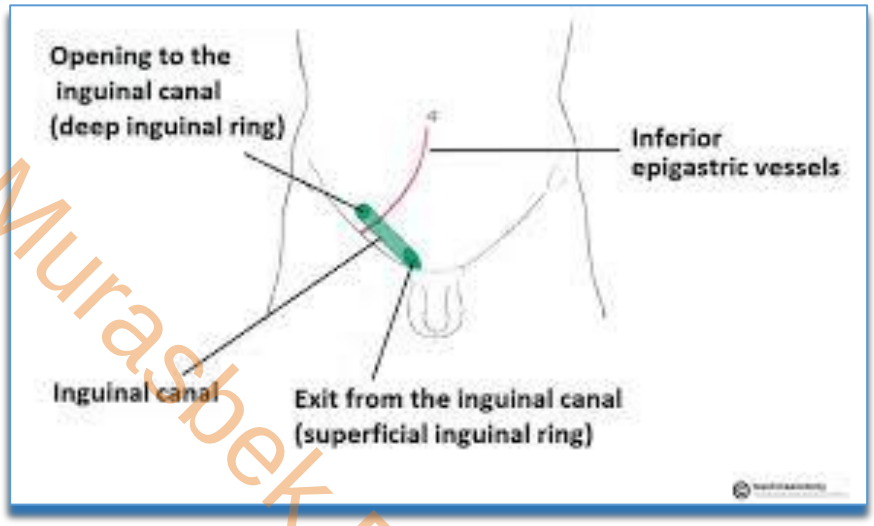
- **Reducible Hernia:** A hernia whose contents can be pushed back into the abdomen.

- **Incarcerated (Irreducible) Hernia:** A hernia whose contents are trapped and cannot be pushed back into the abdomen.

- **Strangulated Hernia:** A condition where the blood supply to the tissue trapped in the hernia sac is compromised, leading to ischemia and possible necrosis.

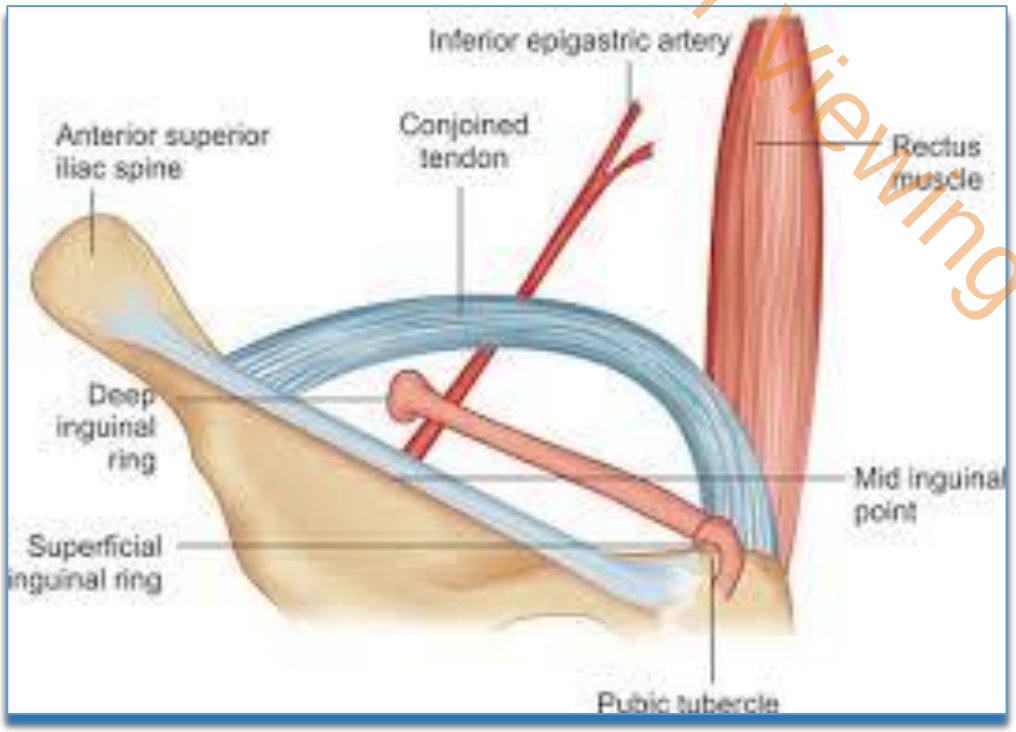
Anatomy of the Inguinal Region

- Deep Inguinal Ring (annulus inguinalis profundus):



- Located in the transversalis fascia, lateral to the inferior epigastric vessels.

- The anterior part is formed by the internal oblique muscle, and the posterior part by the transversalis fascia.



- **Superficial Inguinal Ring (annulus inguinalis superficialis):**

  - An opening in the aponeurosis of the external oblique muscle.

- **Inguinal Ligament (Poupart's ligament):**

  - The lower edge of the aponeurosis of the external oblique.

  - Extends from the anterior superior iliac spine to the pubic tubercle.

- **Lacunar Ligament (Gimbernat's ligament):**

  - Formed by the insertion of the inguinal ligament into the pubis.

  - Forms the medial boundary of the femoral opening

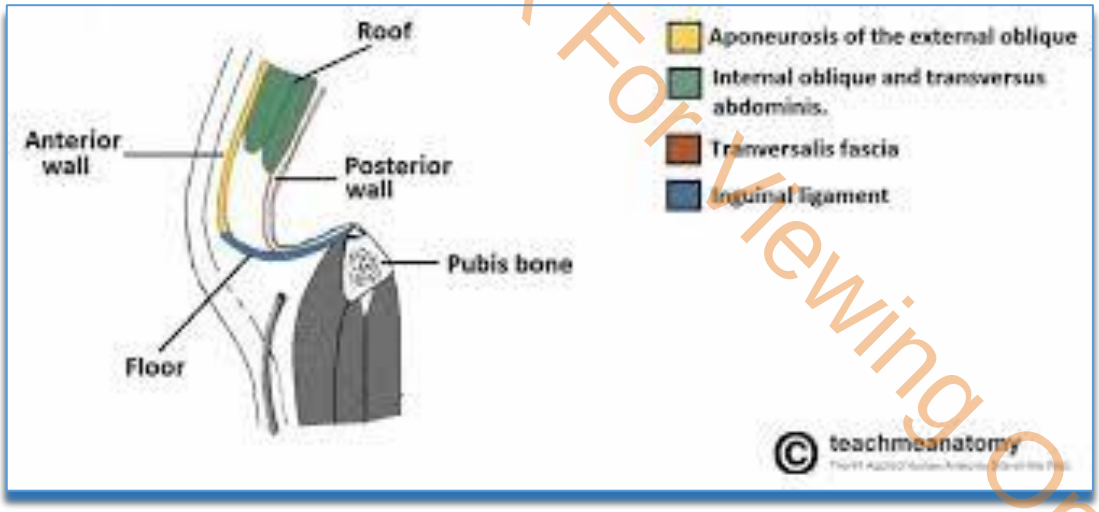
- **Inguinal Canal:**

  - A passage between the deep and superficial inguinal rings.

  - Length: 4-6 cm.

  - Contains the spermatic cord in males and the round ligament of the uterus in females.

Boundaries of inguinal canal:



- **Anterior wall:** Aponeurosis of the external oblique muscle.

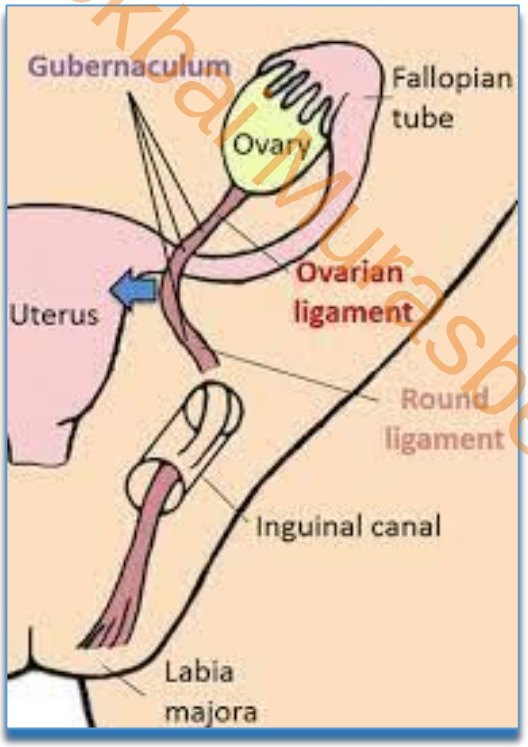
- **Inferior wall:** Inguinal ligament (Poupart's ligament) and its reflection.

- **Superior wall (roof):** Internal oblique and transversus abdominis muscles and their aponeuroses (conjoined tendon).

- **Posterior wall:** Transversalis fascia and aponeurosis of the transversus abdominis muscle.

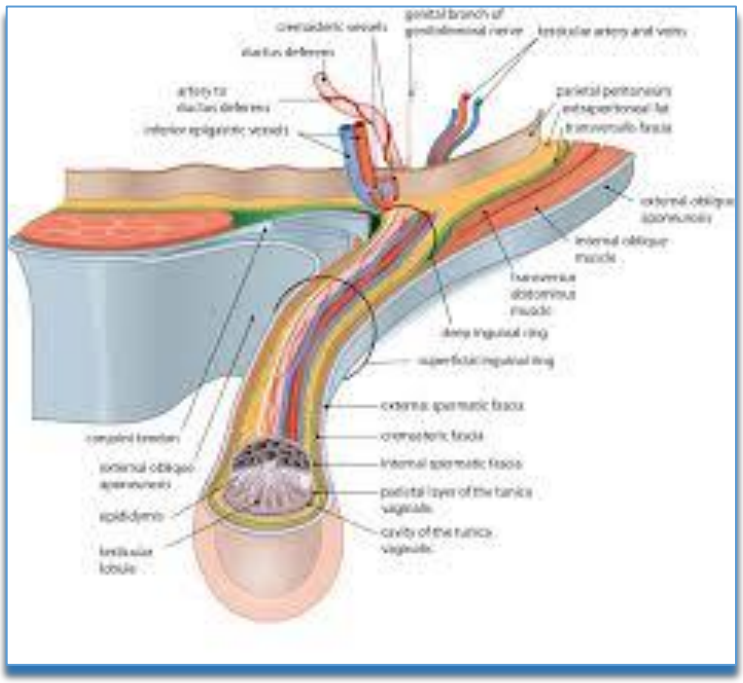
Contents of the Inguinal Canal:

- In males: vas deferens, testicular and cremasteric arteries, pampiniform plexus veins, nerves (ilioinguinal, genital branch of the genitofemoral nerve, and sympathetic nerves), and the processus vaginalis.



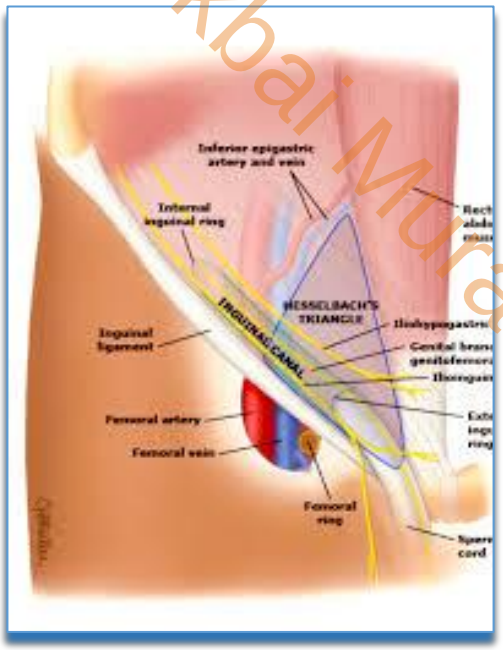
- In females: round ligament of the uterus.

- The genital branch of the genitofemoral nerve and ilioinguinal nerves pass over, not within, the spermatic cord



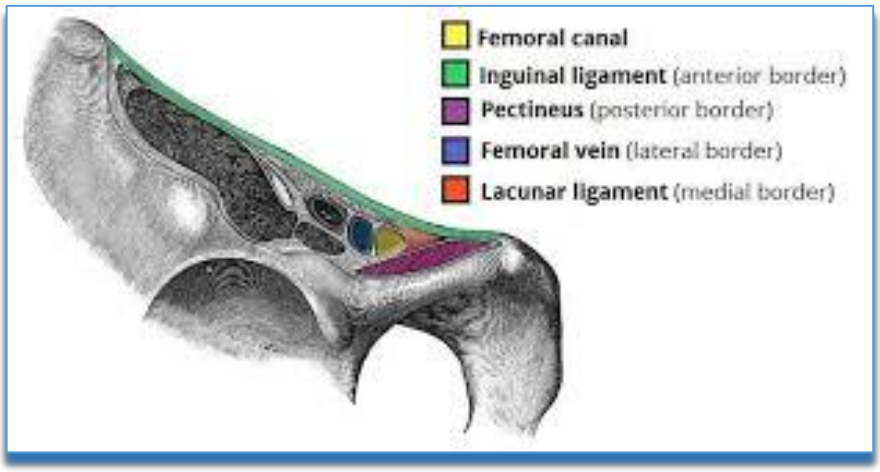
Hesselbach's Triangle:

- Located in the posterior wall.



- Bordered laterally by the inferior epigastric vessels, inferiorly by the inguinal ligament, and superomedially by the lateral edge of the rectus sheath.

- **Femoral Canal:**



- Boundaries:

- Anterior: Iliopectineal tract.

- Posterior: Cooper's ligament (pectineal ligament).

- Lateral: Femoral vein.

- Medial: Lacunar ligament.

- **Internal Oblique Muscle and Aponeurosis**:

- Forms the upper boundary of the inguinal canal.

- Medial fibers of the internal oblique aponeurosis combine with the fibers of the transversus abdominis aponeurosis to form the conjoined tendon, which is present in only 5-10% of people.

- **Iliopectineal Tract:**

  - A thickened inferior edge of the transversalis fascia and transversus abdominis aponeurosis.

  - Acts as the deep part of the inguinal ligament and forms the lower boundary of the deep inguinal ring.

- **Cooper's Ligament (pectineal ligament):**

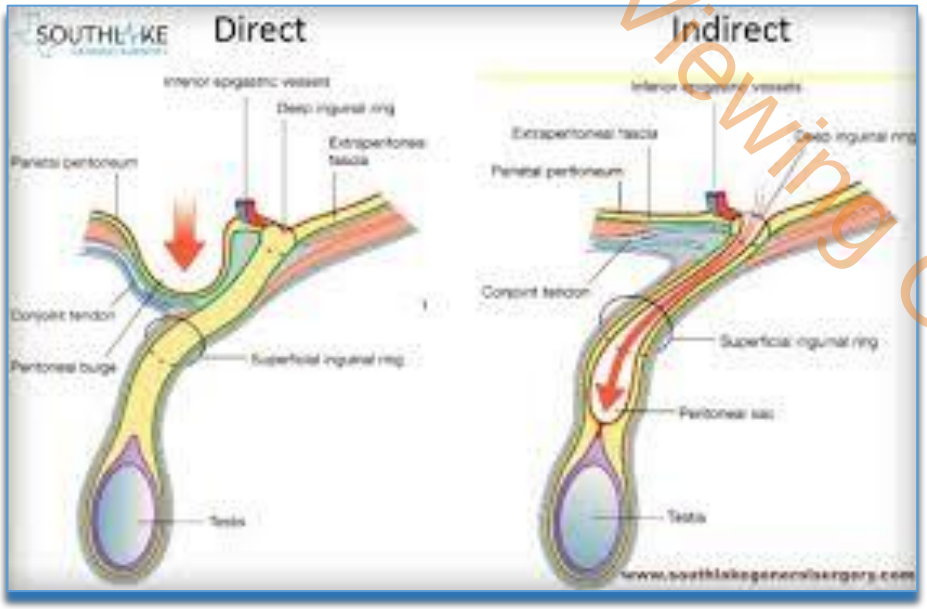
  - Extends along the superior ramus of the pubis.

  - Forms the posterior boundary of the femoral canal.

- **Transversalis Fascia:**

  - A connective tissue layer located under the abdominal wall muscles.

Indirect Inguinal Hernia



  - The most common type of hernia in both men and women.

  - Male to female ratio is approximately 5-10:1.

  - Indirect hernias are five times more common than direct hernias.

  - Location: The hernia sac is located lateral to the inferior epigastric vessels.

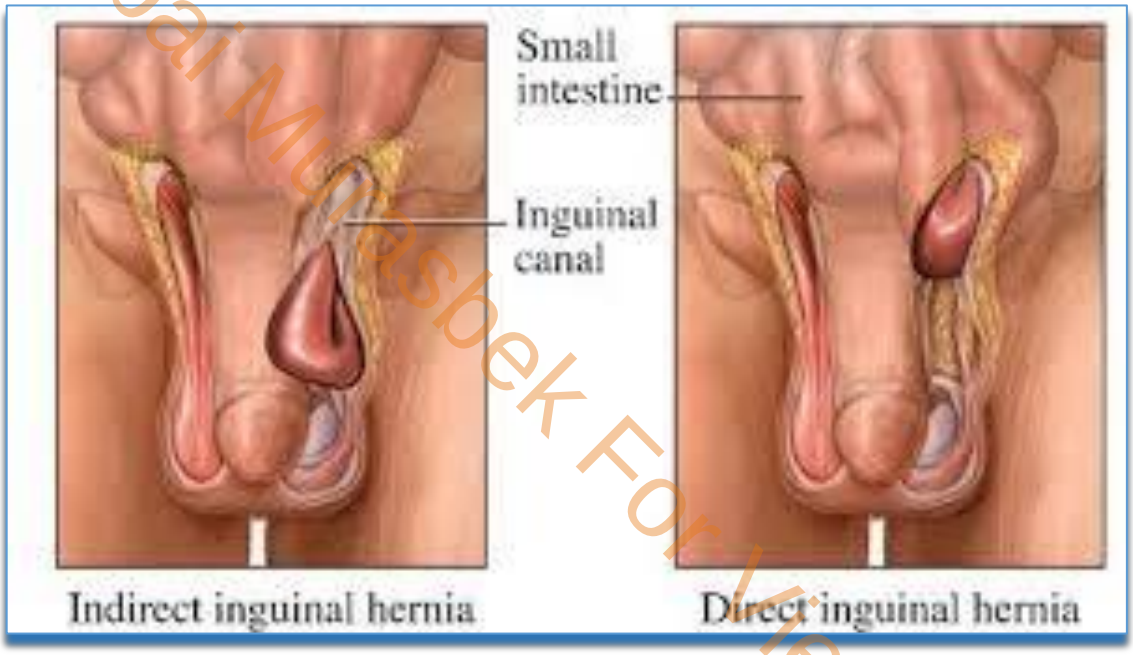
- Pediatric Cases: Nearly all pediatric inguinal hernias are indirect.

  - High risk of incarceration.

- 75% of indirect inguinal hernias are on the right side.

- In children, there is a high risk of bilateral occurrence.

- About 10% of indirect hernias have a bilateral open processus vaginalis.



Direct Inguinal Hernia

Occurs due to weakness in the transversalis fascia, emerging through the Hesselbach triangle.

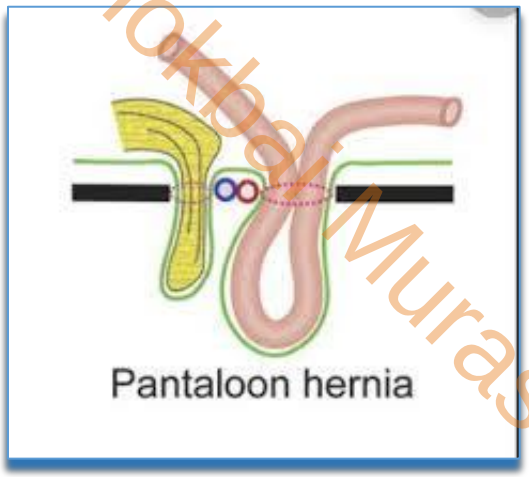
- Location: The hernia sac is located medial to the inferior epigastric vessels.

- Characteristics:

- The defect typically has a wide base, resulting in a lower risk of incarceration and strangulation.

- Frequency increases with age and is associated with physical activity.

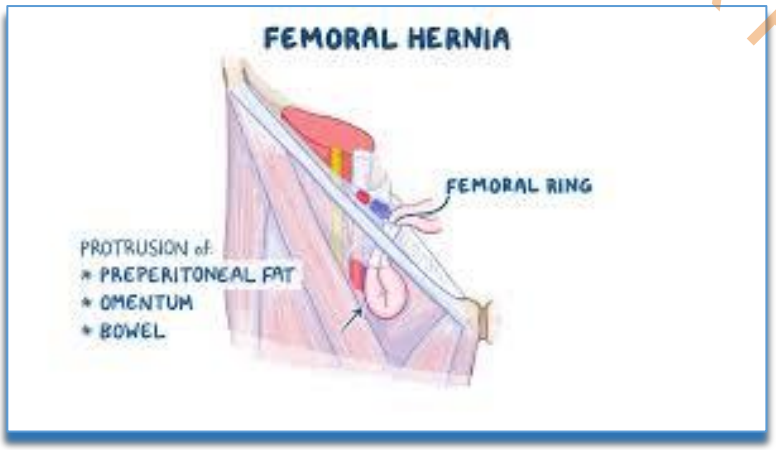
Pantaloon Hernia



Presence of direct and indirect hernias on the same side.

- Herniation occurs both medial and lateral to the inferior epigastric vessels.

Femoral Hernia



- Herniation occurs inferior and posterior to the inguinal ligament, extending into the femoral canal.

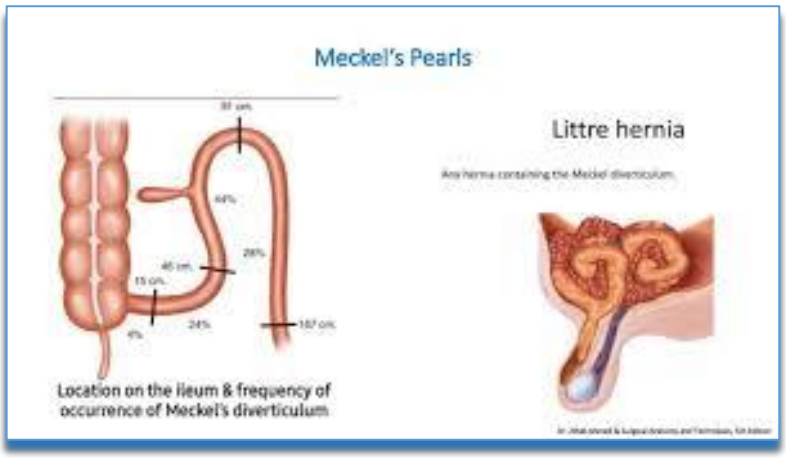
- Located medial to the femoral vessels.

- Linked to physical exercise and pregnancy.

- More common in women.

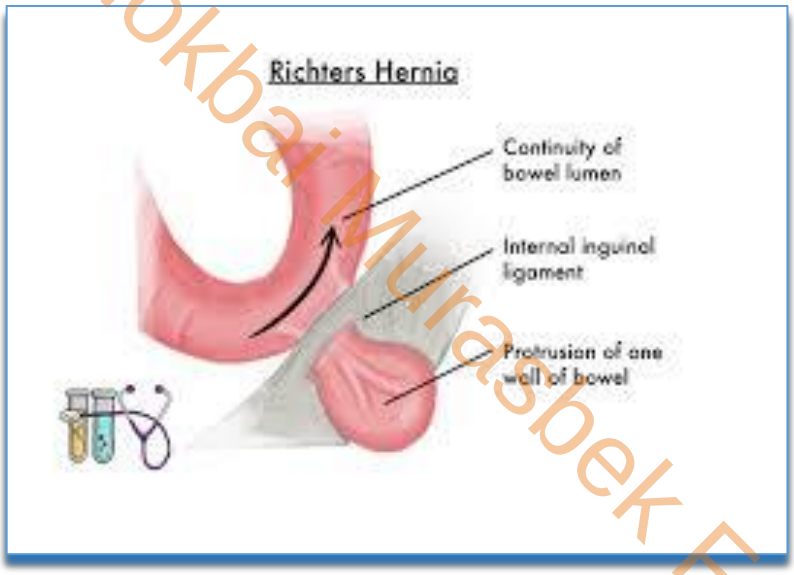
- Carries the highest risk of incarceration and strangulation among hernia types (30-40%).

Littre's Hernia:



- A hernia that contains a Meckel's diverticulum within the hernia sac.

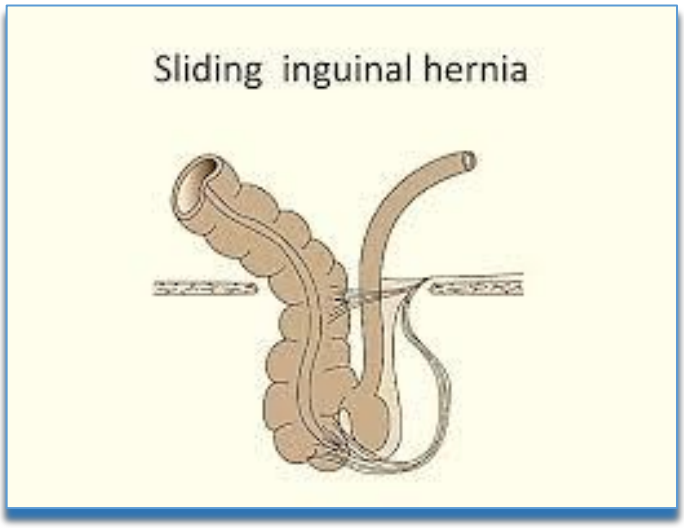
Richter's Hernia (Lateral Pinch):



- A hernia that contains only a part of the bowel wall within the sac.

- Strangulation can occur without causing obstruction.

Sliding Hernia:

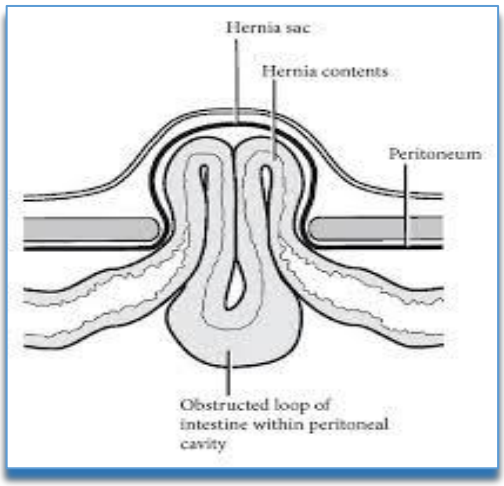


- In a sliding hernia, the hernia sac wall is not formed entirely by parietal peritoneum but is partially made up of intra-abdominal organs like the colon or bladder, which slide and accompany the hernia sac.

- On the right side, the cecum most commonly accompanies the hernia sac, while on the left side, the sigmoid colon is the most common.

- Sliding hernias are most frequently seen in indirect inguinal hernias.

Maydle Hernia:



- These hernias occur when the intestine enters the hernia sac in a "W" shape.

Amyant Hernia:

- A hernia in which the appendix is found within the hernia sac.

Etiology of Hernias

- Inguinal Hernias:

- Can be congenital or acquired. Both types have a strong family history, indicating a genetic predisposition for most inguinal hernias.

- All indirect inguinal hernias are congenital, resulting from the incomplete closure of the processus vaginalis.

- In adults, about 20% have an open processus vaginalis. An open processus vaginalis is a potential factor for the development of an indirect hernia, but it doesn't necessarily mean a hernia will develop; other contributing factors are also needed.

- Muscle Insufficiency (Impairment of the Valve Mechanism):

- Congenital or acquired (due to pelvic bone fractures or poorly performed appendectomy incisions) internal oblique muscle insufficiency leaves the internal ring exposed to intra-abdominal pressure.

- Congenital structural malformations in the transversalis fascia and transversus abdominis aponeurosis can lead to indirect hernias.

Possible Causes of Inguinal Hernias:

- Coughing

- Chronic Obstructive Pulmonary Disease (COPD)

- Obesity

- Straining

- Constipation

- Prostatism

- Pregnancy

- Low birth weight (<1500 g)

- Family history of hernias

- Valsalva maneuver

- Ascites

- Standing for prolonged periods

- Congenital connective tissue diseases

- Defective collagen synthesis

- Previous lower right quadrant incisions

- Arterial aneurysms

- Smoking

- Lifting heavy weights

- Physical exertion

Complications:

- Intestinal Obstruction

- Intestinal Strangulation: The most serious complication.

  - It is most commonly seen in indirect inguinal hernias.

  - The risk of occurrence is highest in femoral hernias.

- Perforation, Gangrene, Peritonitis

Conditions Associated with Inguinal Hernias (Differential diagnosis):

- Hydrocele

- Varicocele

- Spermatic cord lipoma

- Lymphadenitis

- Undescended testis

- Abscess

- Metastatic neoplasm

- Sebaceous cyst

Treatment of Inguinal Hernia

General Principles:

- Reduction: Returning the hernia contents to the peritoneal cavity.

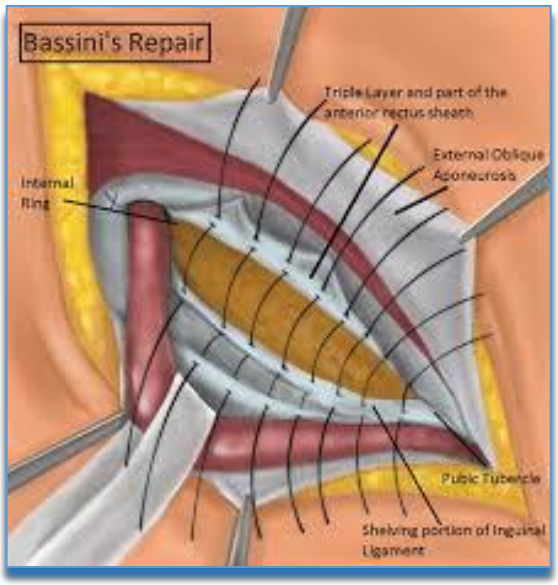
- High Ligation: Tying off the base of the hernia sac.

- Repair of the Abdominal Wall Defect: To prevent recurrence.

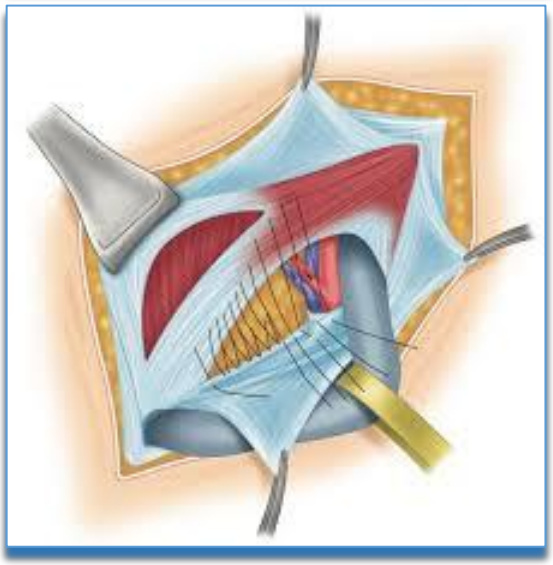
In Children:

- High Ligation Alone: Usually sufficient, as there is no abdominal wall defect requiring repair.

Techniques for Abdominal Wall Defect Repair:



- **Bassini Repair**: The "conjoined tendon" is sutured to the transversalis fascia and inguinal ligament. Only direct and indirect hernia repair are performed.



- **Cooper Ligament Repair (McVay Repair):**

- Covers direct, indirect, and femoral hernia areas.

- The conjoined tendon and transversalis fascia are sutured to the pubic ramus periosteum (Cooper's ligament).

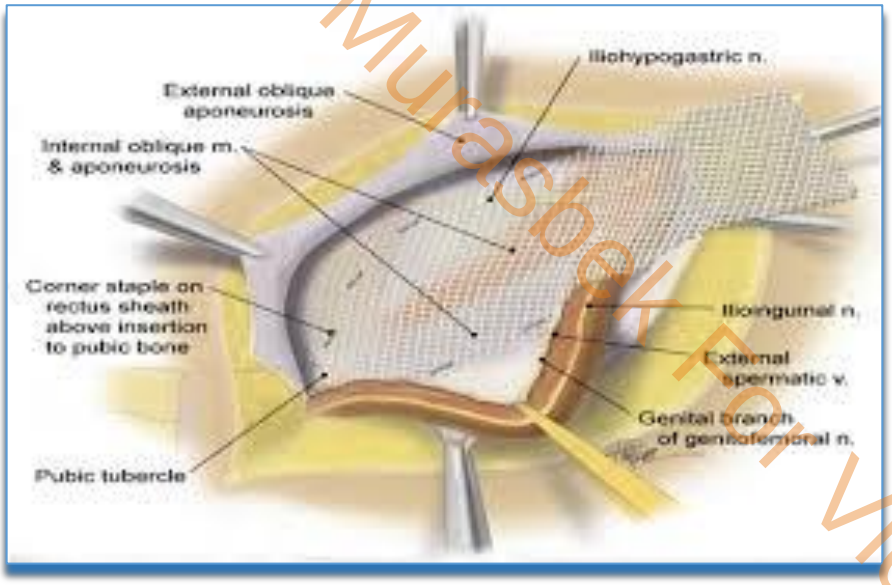
- Indicated for patients where prosthetic materials cannot be used and who have femoral hernias.

- Repairs direct, indirect, and femoral hernias.

- **Shouldice Repair**:

- A modification of the Bassini operation.

- The transversalis fascia is incised and imbricated. Direct and indirect hernia repair are performed.



- **Lichtenstein Hernioplasty (Tension-Free Mesh Repair)**:

- The most commonly used hernia repair technique today.

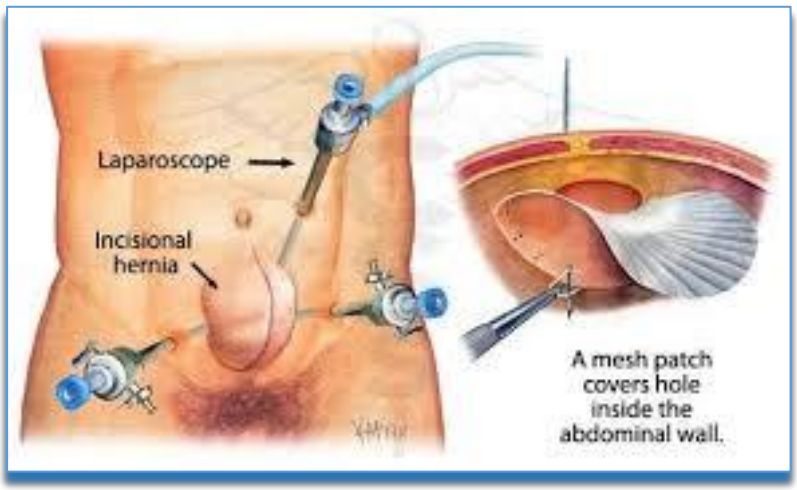
- The defective posterior wall is repaired with a mesh (prosthetic material or graft).

- Repairs direct and indirect hernias.

- The most important cause of recurrence in hernia surgeries is tension at the repair site.

- The least recurrence occurs with tension-free, mesh-based repairs.

- **Laparoscopic Hernia Repair**:



- Laparoscopic hernia surgeries also involve tension-free repairs using mesh.

- The primary advantage is that it allows tension-free preperitoneal closure of direct, indirect inguinal, and femoral hernia areas (myopectineal orifice) with a single large mesh.

- Ideal for bilateral and recurrent hernia repairs.

- If anterior approaches to hernia repair recur, laparoscopic repair is ideal.

- Recurrence rates are low, similar to Lichtenstein repair.

Post-Hernia Surgery Complications

- Bleeding

- Disruption of Testicular Blood Flow

- Cutting of the Vas Deferens

- Bowel Injury

- Strangulation

- Bladder Injury

- Compression of the Femoral Vein

- Urinary Retention

- Scrotal Bruising

- Testicular Swelling and Atrophy

- Hydrocele

- Wound Infection

- Neuroma and Nerve Injury

- Missed Hernia

- Recurrence

- Chronic Groin Pain

- Chronic Residual Neuralgia: Can develop due to manipulation of sensory nerves during surgery, compression from scar tissue, or inflammatory granulomas.

- Risks of injuring following nerves: Ili oinguinal,Iliohypogastric, Genitofemoral,Lateral Femoral Cutaneous Nerves

- **Anterior Repair Techniques:** Higher risk of entrapment of the ilioinguinal and iliohypogastric nerves.

- **Laparoscopic Repair:** Higher risk of involvement with the genitofemoral and lateral femoral cutaneous nerves (Lateral Femoral Cutaneous Nerve Injury: May cause tingling and burning pain on the outer thigh, known as meralgia paresthetica.)

Ventral Hernias

- Ventral Hernias: These are anterior wall hernias occurring outside the inguinal region. The most common type is the incisional hernia.

Incisional Hernia. Develops from a problem with wound healing at the incision site.

- Factors Increasing Risk of Incisional Hernia Development:

- Postoperative wound infection

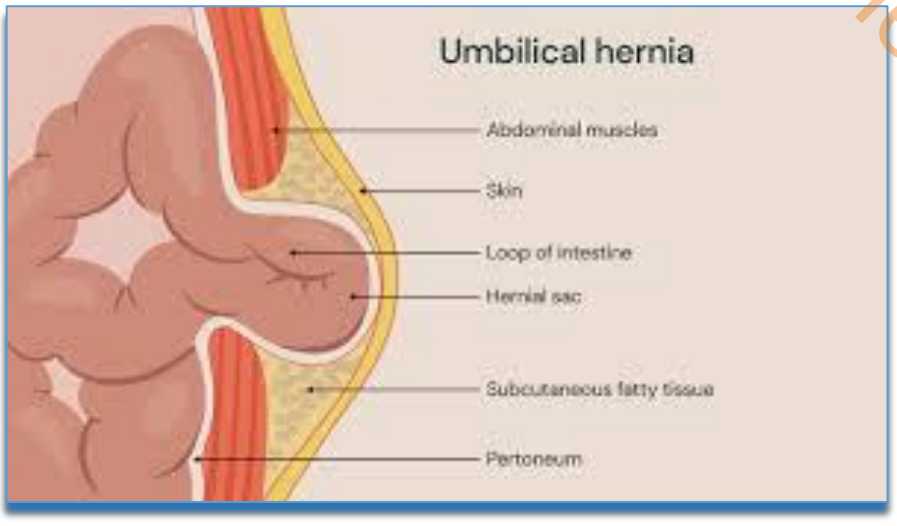
- Malnutrition

- Obesity

- Immunosuppression

- Increased intra-abdominal pressure

Umbilical Hernia



- Associated with ascites and pregnancy.

- Ten times more common in women.

- Congenital umbilical hernias may close spontaneously within the first year. If closure does not occur, elective surgery is recommended.

- In adults, small, asymptomatic umbilical hernias can be monitored.

- Surgical treatment involves primary repair or, for larger defects (>2 cm), repair with prosthetic mesh.

Epigastric Hernia

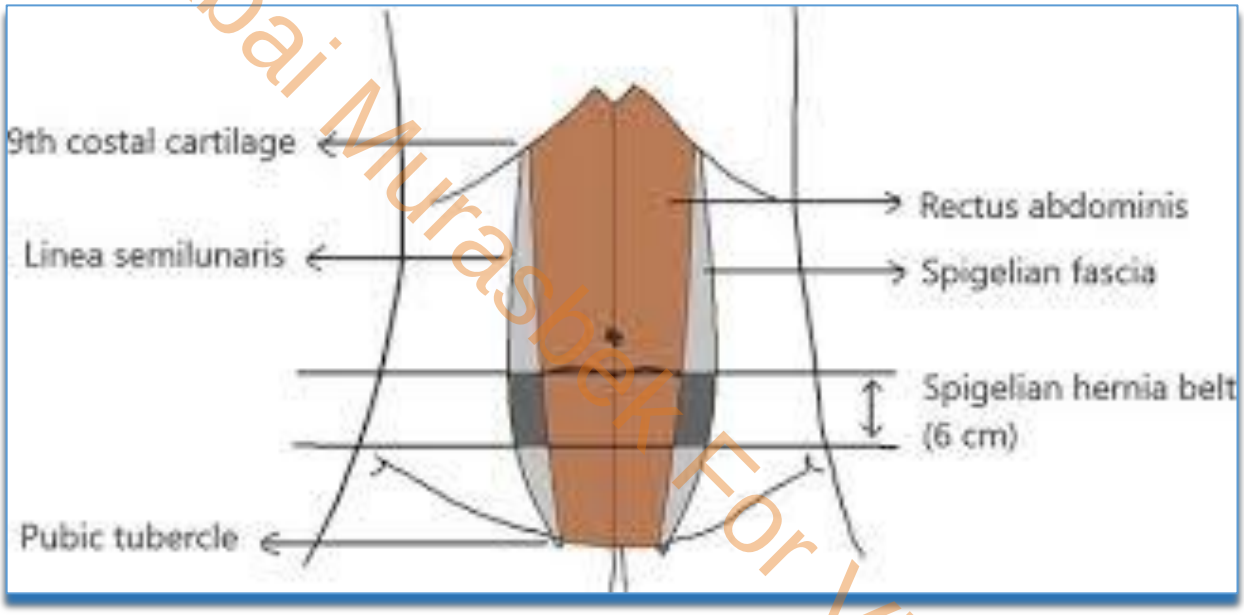
Hernias that occur through a defect in the linea alba.

- Three times more common in men.

- Can be very painful.

Spigelian Hernia

- Occurs at the lateral edge of the rectus sheath along the semilunar line.



- Classified as an interparietal hernia because it protrudes between the layers of the abdominal wall.

- Most commonly develops above and below the arcuate line.

- The hernia sac is usually not palpable because it is small and almost always remains beneath the external oblique aponeurosis.

- The most common site is where the semilunar line intersects the arcuate line.

- Patients present with pain in the hernia area. Diagnosis is made using ultrasound (USG) or computed tomography (CT).

Parastomal Hernia

- Develops near the site of a colostomy or ileostomy.

Lumber Hernia

- Can be congenital or acquired.

- Superior Lumbar Triangle (Grynfeltt) Hernias: More common; the boundaries of the superior lumbar triangle are the 12th rib, paraspinal muscles, and internal oblique muscle.

- Inferior Lumbar Triangle (Petit) Hernias: The boundaries are the iliac crest, latissimus dorsi muscle, and external oblique muscle.

- These hernias develop due to weakness in the dorsolumbar fascia.

- Small lumbar hernias can be asymptomatic.

- Incarceration does not occur in lumbar hernias.

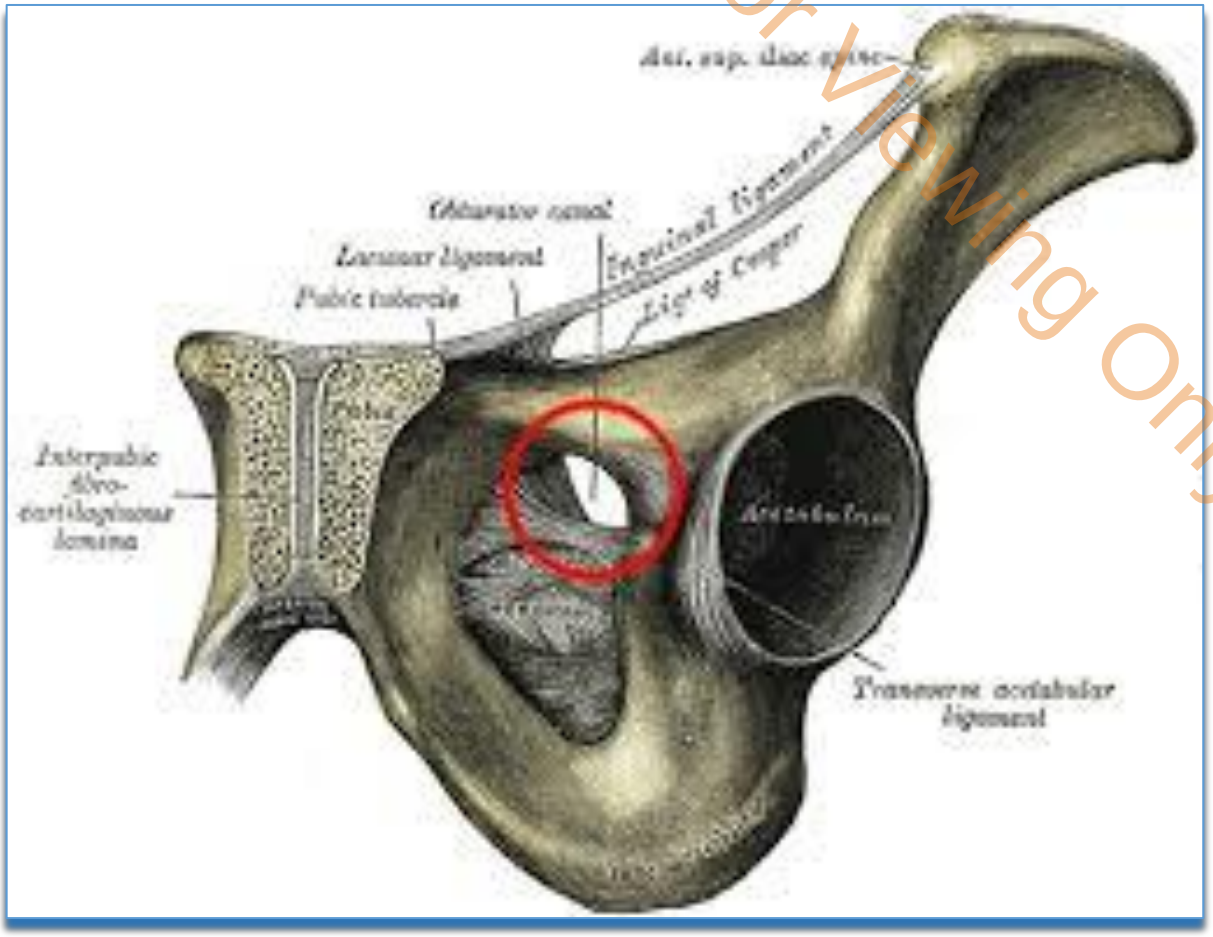
- Diagnosis is made with CT.

- Repair is performed with mesh.

Pelvic Hernias

Obturator Hernia

- Diagnosis is difficult; it is never visible externally. Rarely, a mass may be palpable in the upper-inner thigh during a pelvic or rectal examination.



- The most common pelvic hernia.

- More common on the right side.

- More frequent in women.

- Compression of the obturator nerve causes pain in the hip, knee, and inner thigh, which is relieved by flexion. This characteristic pain, seen in half of the patients, is known as the Howship-Romberg sign.

- Additionally, due to compression of the obturator nerve, the patellar reflex may be preserved while the adductor reflex may be lost. This finding is called the Hannington-Kiff sign.

- 50% of patients present with intestinal obstruction.

- Diagnosis is made with abdominal CT.

- Treatment involves surgical closure of the obturator foramen with a patch (prosthetic mesh).

Sciatic Hernia

- Involves abdominal organs exiting through the large sciatic foramen.

- Extremely rare hernias.

- Diagnosis is challenging.

- Typically asymptomatic unless intestinal obstruction develops.

- The most common symptom is a painful mass in the gluteal region.

- High risk of strangulation.

- Usually treated via a transabdominal approach.

Perineal Hernia

- Can be congenital or acquired defects. Rarely seen.

- May develop after abdominoperineal resection or perineal prostatectomy.

- Primary perineal hernias are rare and usually seen in older, multiparous women.

- Symptoms include a protruding mass from the defect, with pain increasing while sitting or standing.

Internal Hernias

- Occur within the abdominal cavity through natural or abnormal openings.

- Types include paraduodenal, transmesenteric, omental, and retroanastomotic hernias.

In the context of diaphragmatic hernias, Swartz's textbook on general surgery provides detailed classifications and management guidelines. Here's a summary based on Swartz's descriptions:

Diaphragmatic Hernias

1. Types of Diaphragmatic Hernias

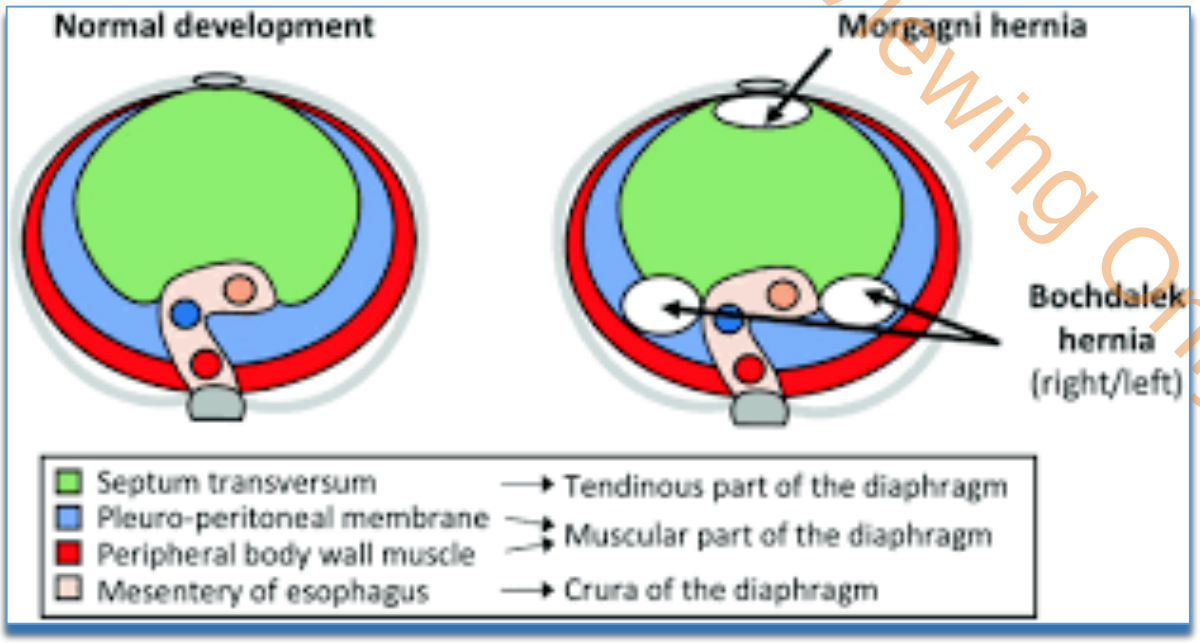
Congenital Hernias:

- **Posterolateral (Bochdalek) Hernias:**

- Most common congenital diaphragmatic hernia.

- Typically occur on the left side.

- Result from a failure of the fusion of the pleuroperitoneal membrane during embryonic development.



- Can cause significant respiratory distress in neonates due to herniation of abdominal contents into the thoracic cavity, leading to compromised lung development.

- **Anteromedial (Morgagni) Hernias:**

- Less common than Bochdalek hernias.

- Occur through the foramen of Morgagni, located in the anterior diaphragm.

- Often asymptomatic and discovered incidentally.

- **Central (Hiatal) Hernias:**

- Occur through the esophageal hiatus of the diaphragm.

- Can be classified into sliding or paraesophageal hernias.

- **Sliding Hernias:** The most common type, where the stomach and possibly other abdominal organs slide into the thoracic cavity.

- **Paraesophageal Hernias:** The stomach herniates beside the esophagus but the esophagus remains in its normal position.

2. Diagnosis and Management

- **Clinical Examination:** Symptoms may include difficulty breathing, decreased breath sounds on one side, and signs of gastrointestinal distress.

- **Chest X-ray:** Can reveal displacement of abdominal organs into the thoracic cavity.

- **CT scan:** Provides a more detailed view of the hernia and associated structures, often used to confirm the diagnosis.

***Management*:**

- **Surgical Repair:** The primary treatment for symptomatic diaphragmatic hernias.

- **Congenital Hernias:** Often repaired early in life, especially in symptomatic cases to prevent complications such as pulmonary hypoplasia.

- **Acquired Hernias:** Requires surgical intervention to repair the defect and address any associated injuries or complications.

- **Supportive Care:** May include respiratory support, nutritional support, and management of associated conditions.

Acute Abdomen

- Acute abdomen is a condition that requires immediate medical intervention.

- It begins suddenly and progresses rapidly.

- A prompt decision needs to be made in acute cases—whether surgery or medical treatment is required. This decision takes precedence over making a definitive diagnosis.

History in Acute Abdomen

- Taking a detailed history is crucial in cases of acute abdomen.

- It guides further investigations.

Presenting Symptoms

- Pain

  - It is the most important symptom.

  - The presence of pain alone is not sufficient; its characteristics must be thoroughly investigated.

Localization of Pain

- Initially, the pain is diffuse and does not provide clear information (visceral phase).

- Visceral pain is transmitted by the dorsal 6-12 spinal nerves and is not localized; it is widespread.

- Pelvic pain is transmitted by spinal nerves below L1.

- Visceral pain cannot be localized and is felt when there is stretching or ischemia.

- Somatic phase pain can be localized.

- This pain originates from the parietal peritoneum or mesentery.

- Organ pathology can be generally predicted based on pain localization:

    - Epigastrium: Stomach and pancreas pathologies.

    - Right upper quadrant: Gallbladder pathologies.

    - Umbilical region: Small intestine pathologies.

    - Suprapubic region: Colon, rectum, and genitourinary pathologies.

Onset and Nature of Pain

- Sudden onset pain suggests perforation, volvulus, or circulatory disturbances.

- Gradual onset pain suggests inflammatory conditions.

- Sharp, stabbing pain suggests vascular occlusion or perforation.

- Burning pain suggests an ulcer.

- Dull pain is diffuse and often associated with inflammatory diseases.

- Colicky pain occurs when the lumen of an organ with smooth muscle (e.g., urinary, biliary, or intestinal colic) is obstructed.

- Cramping is a more frequent version of colicky pain.

Pain Radiation

- Radiation of pain provides important clues.

- Right upper quadrant pain may radiate between the shoulders or to the right shoulder.

- This occurs via diaphragmatic proximity or through the sympathetic nervous system.

- Left upper quadrant and spleen pathologies may radiate to the left shoulder (**Kehr's sign**).

- Urinary system pathologies may cause pain that radiates along the ureter's course to the groin.

- Pancreatic pathologies may cause a band-like pain in the lower back and central back.

Referred Pain Locations

- Right shoulder: Liver, gallbladder, right hemidiaphragm.

- Left shoulder: Heart, pancreas tail, spleen, left hemidiaphragm.

- Scrotum and testicles: Ureter.

- Back pain: Pancreatic pathologies.

Pain in Relation to Other Symptoms

- In acute appendicitis, pain typically precedes nausea and vomiting (reflex nausea and vomiting).

- If nausea and vomiting precede the pain, this makes acute appendicitis less likely.

- Activities that exacerbate or relieve the pain are also important.

- Oral intake often exacerbates pain associated with intestinal obstruction, biliary colic, pancreatitis, diverticulitis, or bowel perforation.

- In non-perforated peptic ulcer and gastritis, oral intake may relieve or eliminate the pain.

Fever and Leukocytosis

- Sudden onset or long-standing fever that occurs in episodes can provide clues.

- A high fever and leukocytosis over 16,000 may indicate a complication of any disease.

Nausea and Vomiting

- The nature of the vomit is important.

- In obstructions, initial vomiting is reflexive and consists of stomach contents, followed by different characteristics depending on the level of obstruction, ranging up to fecal matter.

- Additionally, attention should be paid to whether the vomit contains blood or bile.

Bowel Motility

- The ability or inability to pass stool, and the nature of the stool, can be extremely important.

- Passing gas is more important in the context of obstruction.

- Whether symptoms start after meals or not is also significant.

- Decreased or absent bowel sounds are a sign in favor of surgical acute abdomen.

Anorexia

- Weight loss is important in considering malignancies.

Hiccups

- Hiccups may develop due to irritation of the phrenic or vagus nerve.

- Conditions like gastric dilation, alcohol and tobacco use, thoracic and abdominal surgical trauma, hiatus hernias, subphrenic abscesses, etc., can cause hiccups along with pain.

Urinary Symptoms

- Stones and infection should be ruled out.

- Changes in urinary frequency (polyuria, oliguria), and whether there is burning or pain, should be investigated.

Obstetric and Gynecological History in Women

- Menstrual periods, last menstrual period, pregnancy status, history of criminal abortion attempts, etc., should be investigated.

Past Medical History and Surgeries

Hematological Diseases

- Sickle Cell Anemia: Particularly important as it can cause acute abdominal pain crises.

Metabolic Disorders

- Diabetes, porphyria, etc.: Can mimic acute abdomen.

Lead Poisoning

- Can cause abdominal pain.

Alcohol Consumption

- Should be considered in the history.

Allergies

- Important to note any known allergies.

Family History

- Familial Mediterranean Fever (FMF): Can provide information about hereditary metabolic diseases.

System Review

- Useful for differential diagnosis: For example, a good cardiovascular history might lead to considering myocardial infarction as a cause of acute epigastric pain.

Physical Examination in Acute Abdomen

Inspection

- If peritonitis is present, the patient may have difficulty moving, with changes in facial expression and overall behavior.

- Patients with stone diseases may appear restless.

- Look for distension, previous surgical scars, and assess for the presence of petechiae or ecchymosis.

Auscultation

- Decreased or absent bowel sounds suggest a serious intra-abdominal pathology.

- Early in intestinal obstructions, bowel sounds may be increased, with tympanic or metallic sounds noted.

Percussion

- Can help differentiate between peritoneal fluid and masses.

- Closure of Traube's space suggests splenomegaly.

Palpation

- Evaluate for masses and hepatomegaly.

- Physical examination findings supporting acute abdomen include tenderness, rebound tenderness, and guarding.

- The most important sign of acute abdomen is involuntary guarding (rigidity).

- This indicates peritoneal irritation.

- "Board-like abdomen" refers to the entire abdomen being involuntarily tense.

- Voluntary guarding occurs when the patient consciously tenses their abdomen.

- **Rebound test**: The doctor presses firmly on the patient's abdomen and then suddenly releases it.

  - If the pain increases in the same quadrant when the hand is released, it is called direct rebound.

  - If the pain increases in the opposite quadrant when pressure is released, it is called indirect rebound.

- Peripheral arterial pulses: Important for assessing cardiovascular disease and mesenteric artery occlusions.

- Edema: Important in the context of cardiovascular health.

- Hernias: Must be carefully evaluated.

- Rectal and, in women, vaginal exams should be performed in patients suspected of having an acute abdomen.

Laboratory Tests in Acute Abdomen

- Should be kept to a minimum to avoid unnecessary delays.

- First, request rapid, simple, and reliable tests.

- Routine tests include complete blood count and urinalysis.

- If pancreatitis is suspected, request amylase and lipase levels.

- Blood glucose and bilirubin can be requested if needed.

- Beta-hCG should be tested in female patients.

Imaging in Acute Abdomen

- Abdominal X-rays (upright and supine) and chest X-rays are generally routine.

- Free air under the right diaphragm suggests perforation of a hollow organ (chest X-rays can be done for the same purpose).

- Ultrasound (US), CT, IVP, IV cholangiography, and angiography can be performed if necessary.

- CT is the best test for detecting free air in the abdomen.

- Abdominal MRI is not used for diagnosing acute abdomen.

- Barium studies are contraindicated in patients with suspected acute abdomen due to the risk of barium peritonitis and interference with US, CT, and angiography.

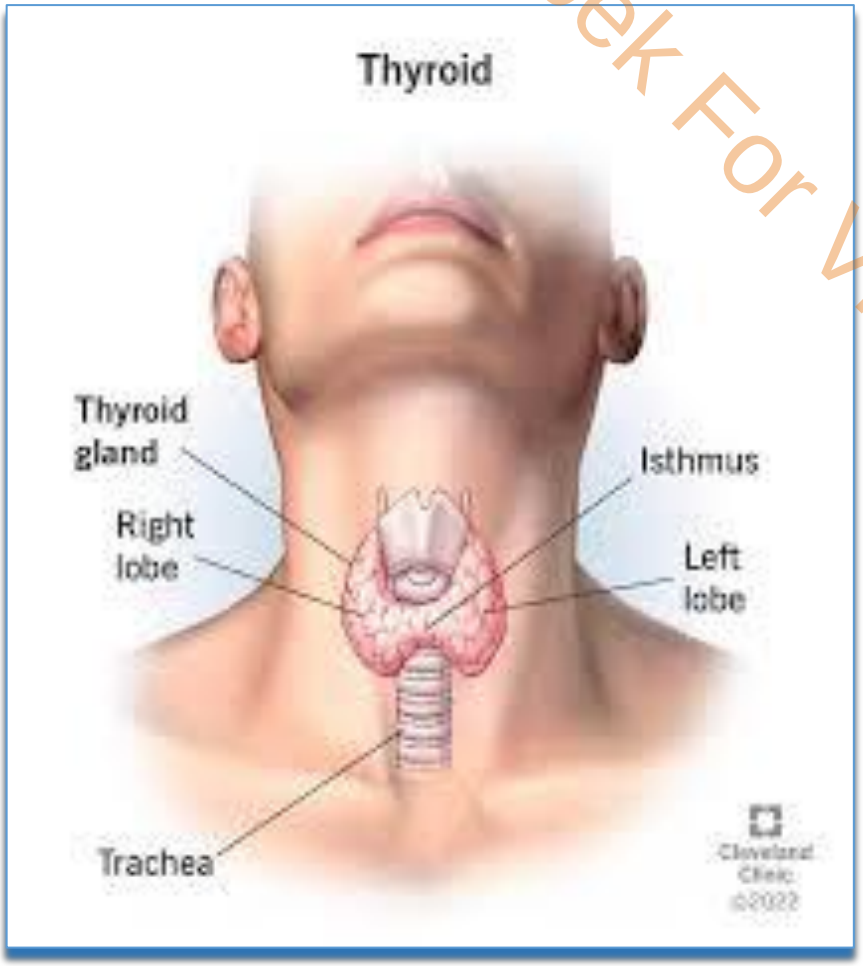
- Barium studies are only done under elective conditions.

Chapter
12.

Thyroid Diseases and Surgery

Anatomy of the Thyroid Gland

-Innervation: The thyroid gland receives sympathetic innervation from the cervical ganglia (vasomotor) and parasympathetic innervation from the vagus nerve.



- Nerves:

- Inferior laryngeal nerve (N. recurrens, RLS) and superior laryngeal nerve are branches of the vagus nerve.

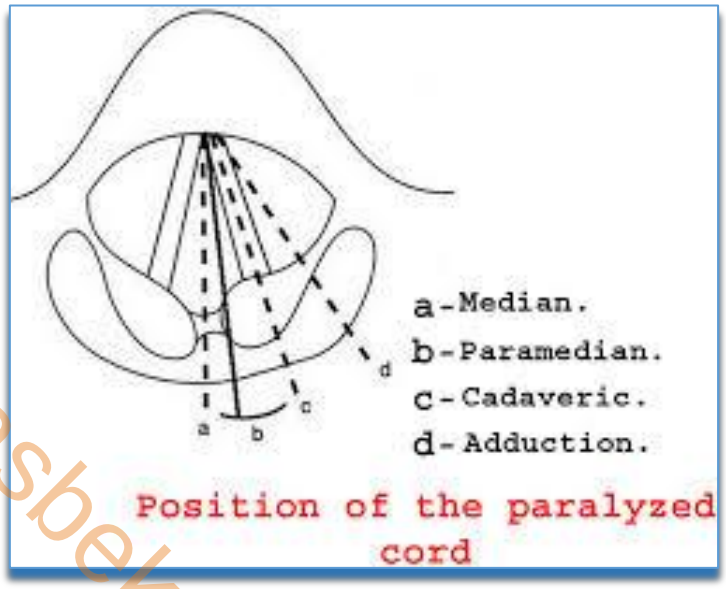
- On the left side, they loop under the ligamentum arteriosum, and on the right side, under the subclavian artery, to reach the larynx.

- In 0.5-1% of cases, the nerve may be non-recurrent, often associated with a vascular anomaly.

- The recurrent laryngeal nerve (RLS) innervates all intrinsic muscles of the larynx.

- The cricothyroid muscle is innervated by the superior laryngeal nerve.

- **Consequences of Nerve Injury:**



- Unilateral Injury of the Recurrent Laryngeal Nerve:

- Leads to paralysis of the vocal cord on the affected side.

- The vocal cord may move to a medial position, resulting in a paramedian or "abducted" position.

- In the paramedian position, the voice is normal but weak.

- In the abducted position, hoarseness, coarseness, and inadequate coughing are observed.

- **Bilateral Injury:**

- Both vocal cords move medially, causing the rima glottis to narrow, leading to loss of voice and necessitating an emergency tracheostomy.

- Superior Laryngeal Nerves:

- These also branch off from the vagus nerve.

- The internal branch carries sensory information from the supraglottic larynx.

- Injury during thyroid surgery is rare, but if it occurs, it may lead to aspiration.

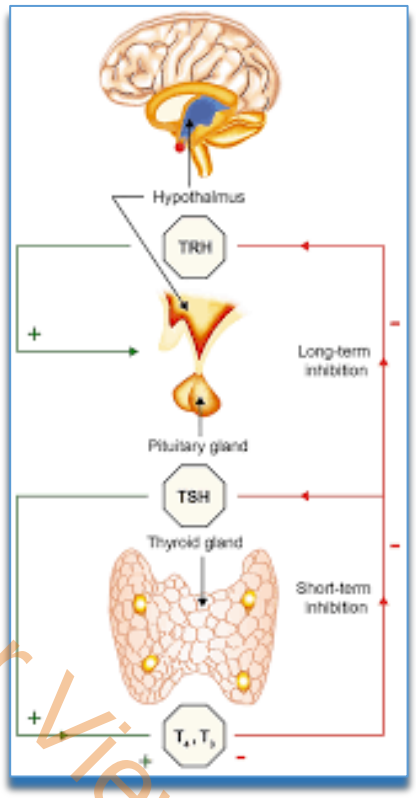
- The external branch travels to the cricothyroid muscle and is in close proximity to the superior thyroid vessels.

- The risk of injury increases if these vessels are ligated away from the thyroid tissue.

- Also known as the Amelita Galli-Curci nerve or the "high note" nerve.

- Injury to this nerve prevents the vocal cord from tensing, resulting in the inability to produce high-pitched sounds and may cause vocal fatigue during prolonged speech.

Thyroid Function Tests and Radiologic methods for diagnosis



- **Serum TSH Measurement**

- Normal Range: 0.5-5.0 µIU/mL.

- The normal range for TSH, measured using conventional methods, varies between 0.5-5.0 µIU/mL.

- For patients with thyroid nodules who appear clinically euthyroid, TSH is often the only necessary test.

- Ultra-sensitive TSH measurements have become the most sensitive and specific tests for diagnosing hyperthyroidism and hypothyroidism, as well as for optimizing T4 replacement therapy and suppressive therapy.

- **Total T4 and T3**

- The levels of total T4 and T3 reflect the amount bound to TBG (thyroxine-binding globulin), and thus, they are affected by changes in serum protein levels.

- **Free T4 and T3**

- These tests provide accurate and sensitive measurements of biologically active thyroid hormones.

- Measurement of free T3 is particularly useful in confirming the diagnosis of early hyperthyroidism.

- **TRH Test**

- This test is used to diagnose and differentiate secondary and tertiary hypothyroidism.

- **Thyroglobulin**

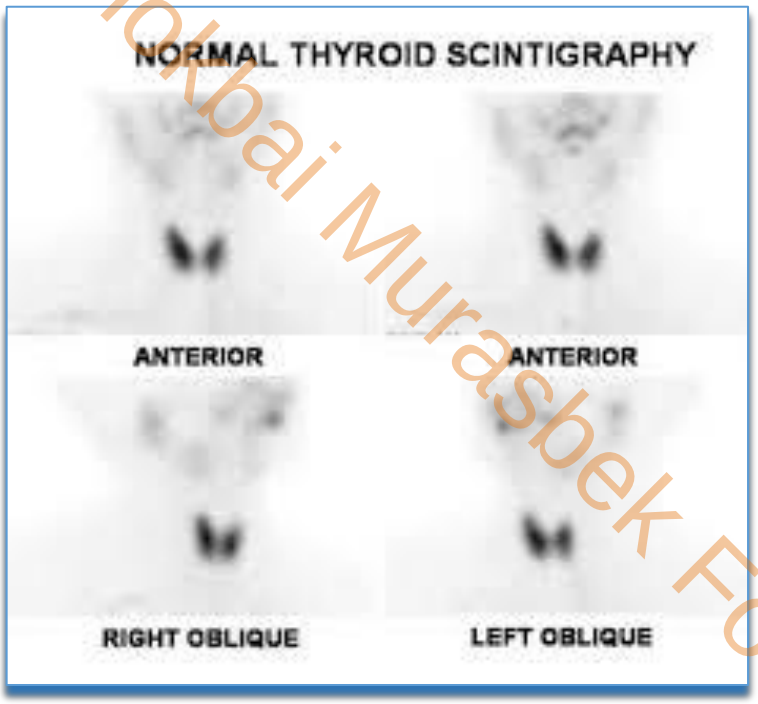
- The primary use of thyroglobulin measurement is in the follow-up to detect recurrences of differentiated thyroid cancers.

- **Thyroid Antibodies**

- Tests can be conducted for antibodies such as antithyroglobulin (anti-Tg), antimicrosomal or antithyroid peroxidase (anti-TPO), and thyroid-stimulating antibodies (TSI).

- Anti-Tg and anti-TPO antibodies typically indicate underlying conditions such as autoimmune thyroiditis, rather than directly reflecting thyroid function.

- **Radionuclide Imaging (Scintigraphy)**



- Iodine-123 (I-123) and Iodine-131 (I-131) are used in thyroid imaging.

- Iodine-123:

- Half-life: 12-14 hours.

- Emits low-dose radiation.

- Used for imaging lingual thyroids and goiters.

- Iodine-131:

- Half-life: 8-10 days.

- Emits high-dose radiation.

- Used for screening and **treatment of differentiated thyroid cancers.**

- Scintigraphy can also assess nodule function:

- Hypoactive (cold) or Hyperactive (hot) nodules.

- Technetium-99m Pertechnetate:

- Very short half-life.

- Minimal radiation.

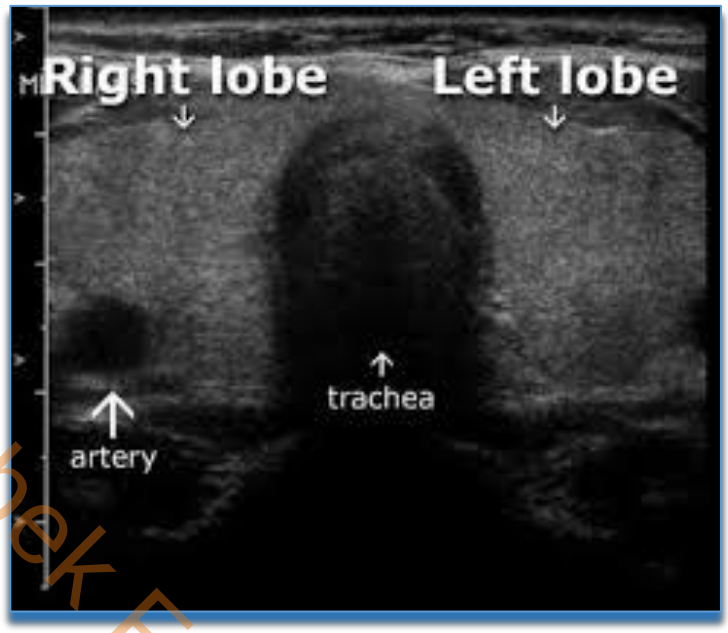
- Particularly sensitive for detecting lymph node metastases.

- **PET (Positron Emission Tomography)**

- 18-Fluorodeoxyglucose (FDG) PET-CT:

- Used for detecting thyroid cancer metastases, especially when other imaging methods yield negative results.

- **Ultrasonography**



- Useful for distinguishing between cystic and solid nodules.

- Provides information on nodule size and multicentricity.

- Shows nodule shape, borders, vascularity, and presence of calcifications.

- Valuable in assessing cervical lymph nodes.

- Guides aspiration needle placement.

- Suspicious findings for malignancy:

- Microcalcifications, hypoechogenicity, irregular borders or absence of a halo, solid nodules, intranodular blood flow, and greater length than width.

- **CT/MRI**

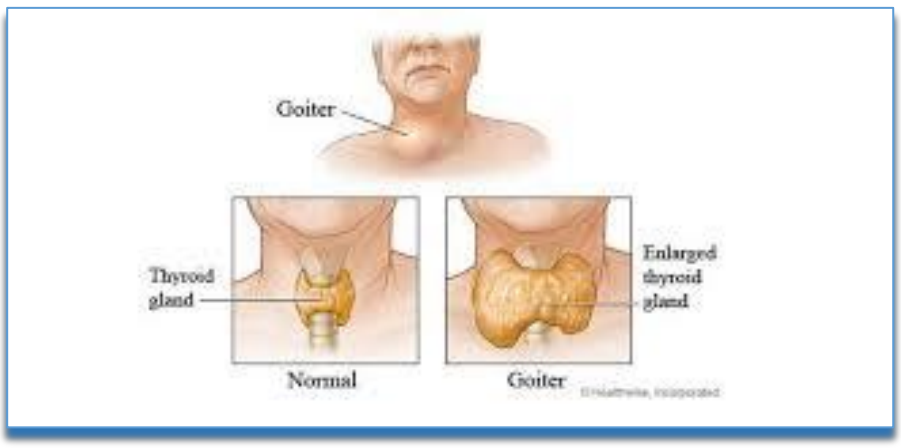
- Useful for evaluating large, fixed substernal goiters, particularly when assessing the relationship to the airway and vascular structures (where USG is insufficient).

Goiter

- Definition: Enlargement of the thyroid gland for any reason.

- Common Cause:

- Often due to insufficient thyroid hormone synthesis and increased TSH levels resulting from other paracrine growth factors.



- The thyroid enlarges to maintain the patient in a euthyroid state.

- Types:

- Goiters can be diffuse or nodular (single or multiple nodules).

- Even if initially diffuse, asymmetric nodularity may develop later.

Causes of Non-Toxic Goiter

1. Familial Goiter:

- Caused by congenital deficiencies in the enzymes required for thyroid hormone synthesis.

2. Endemic Goiter:

- The most significant factor is iodine deficiency.

- Other factors include the consumption of goitrogenic foods (like turnips and cabbage) and genetic factors.

- Excessive iodine administration in endemic goiter patients may lead to thyrotoxicosis, known as **Jod-Basedow syndrome.**

- The suppression of thyroid function due to excess iodine is known as the **Wolff-Chaikoff effect**.

3. Other Causes of Goiter:

- Thyroiditis-induced goiter.

- Drug-induced goiter: Medications like iodide, amiodarone, and lithium can cause goiter.

- Tumor-related goiter.

- Goiter due to thyroid hormone resistance.

- Sporadic goiter:

  - This term refers to thyroid enlargements not linked to a specific cause.

  - Most sporadic goiters do not have an identifiable cause.

Pathology

- The thyroid gland may be diffusely enlarged with a smooth surface or nodular.

- In the early stages, the thyroid gland is hyperplastic, which may be reversible with iodine supplementation.

- Non-toxic nodular goiters can contain nodules of varying sizes.

- Degeneration of nodules may lead to cyst formation, bleeding within the nodule, and calcifications.

Clinical Presentation

- Symptoms:

- Most patients are asymptomatic.

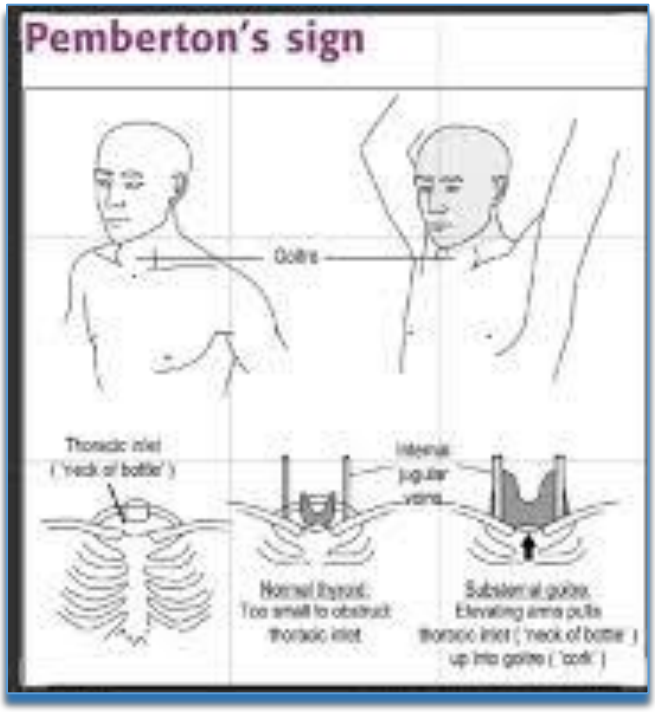
- The most common symptom is swelling and pain in the neck area.

- If the goiter becomes very large, compressive symptoms like dyspnea and dysphagia may occur.

- Hoarseness is rare but may result from recurrent nerve paralysis due to tension caused by the enlarging gland.

- Compression is more pronounced in goiters that grow into the thorax.

- Pemberton's Sign:



- When the patient raises their arms, the substernal goiter may cause increased compression symptoms, facial flushing, and venous distension in the neck. This is known as Pemberton's sign.

- Sudden Enlargement:

- Acute pain and sudden increase in gland size may indicate bleeding into a nodule or cyst.

Diagnosis

- Thyroid Function Tests:

- Typically, thyroid function tests are normal.

- Fine-Needle Aspiration Biopsy (FNAB):

- Recommended in cases of dominant nodules, painful or enlarging nodules, since 5-10% of multinodular goiters have been reported to be carcinoma.

Treatment

- Conservative Management:

- Most patients with small, diffuse, euthyroid goiters do not require treatment.

- Many cases respond well to thyroxine treatment that suppresses TSH.

- In endemic goiters, particularly those without nodules, iodine and thyroxine replacement can significantly reduce the gland size.

- Indications for Surgical Treatment:

- Compressive symptoms.

- Continued growth despite thyroxine suppression.

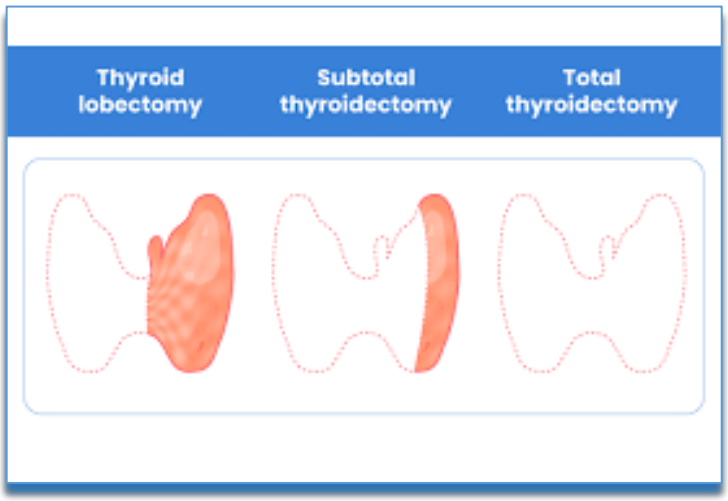
- Retrosternal extension.

- Suspicion of malignancy.

- Confirmation of malignancy via FNAB.

- Cosmetic concerns.

- Surgical Approach:



- Typically involves total or near-total thyroidectomy.

- Patients should receive lifelong thyroxine (T4) therapy post-surgery.

Thyrotoxicosis

- Definition: Clinical signs associated with increased thyroid hormones in circulation.

- Hyperthyroidism: Refers to clinical signs related to increased thyroid hormones in the blood due to an overactive thyroid.

Causes of Thyrotoxicosis

1. Increased Thyroid Hormone Secretion:

- Graves' disease

- Toxic nodular goiter

- Toxic adenoma

- Jod-Basedow syndrome

2. Thyroid Hormone Secretion Not Increased:

- Subacute thyroiditis

- Functional metastatic thyroid cancer

- Struma ovarii

- Iatrogenic (over-treatment of hypothyroidism)

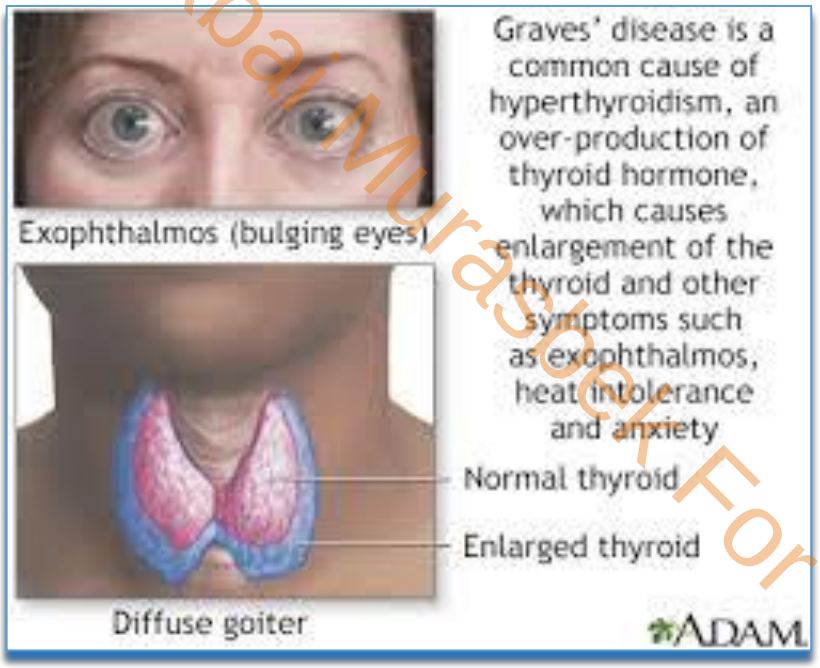
- Radioactive Iodine Uptake (RAIU):

- Diseases associated with increased thyroid hormone synthesis lead to increased RAIU, while others are characterized by decreased RAIU.

- Struma ovarii is the presence of thyroid tissue within an ovarian teratoma.

Graves' Disease (Toxic Diffuse Goiter)

- Prevalence: Responsible for 60-80% of all cases.



- Most Common Cause of Hyperthyroidism.

- Etiology: An autoimmune disease with a strong familial predisposition; exact cause is unknown.

- Demographics:

  - More common in women (5:1 ratio).

  - Most frequently affects women aged 20-40 years.

Features of Graves' Disease

- Thyrotoxicosis

- Diffuse Goiter

- Extra-Thyroidal Manifestations: Ophthalmopathy, dermopathy (pretibial myxedema), thyroid acropachy, gynecomastia, and vitiligo.

- Pathophysiology:

- The thyroid gland enlarges diffusely with increased vascularity.

Clinical Presentation

- Thyrotoxicosis: Heat intolerance, increased sweating, thirst, weight loss, palpitations, nervousness, fatigue, hyperkinesis, and tremors.

- Most common gastrointestinal symptom: increased bowel motility and diarrhea.

- Women may experience amenorrhea and increased miscarriage rates.

- Elderly may present with atrial fibrillation and congestive heart failure.

- Skin is warm and moist, with possible darkening.

- Fine tremor, muscle wasting, and increased tendon reflexes are common.

- Ophthalmopathy occurs in approximately 50% of patients.

Eye Manifestations:

- Upper Eyelid: Spasm and retraction (Dalrymple sign)

- Lid Lag: Von Graefe sign

- External Ophthalmoplegia

- Proptosis and Exophthalmos

- Conjunctival Edema and Congestion (Chemosis)

- Cause: Retro-orbital tissue edema and mucopolysaccharide accumulation, not directly related to thyrotoxicosis, and does not resolve with treatment (euthyroid state).

Diagnosis

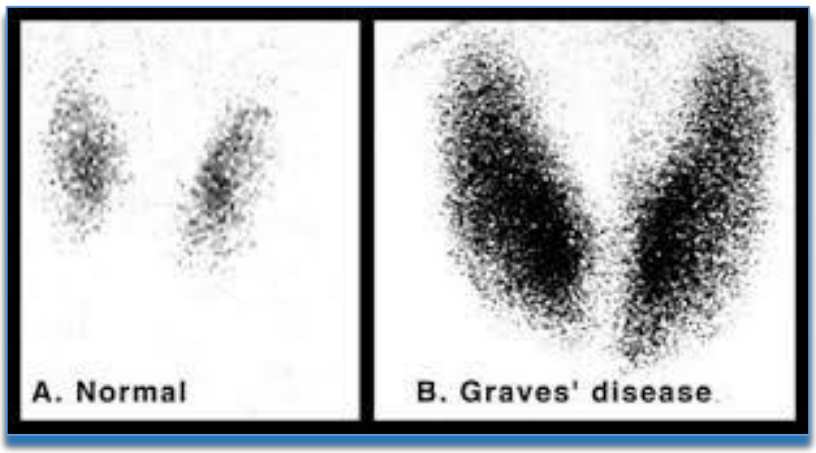
- Clinical Diagnosis: Based on clinical presentation.

- Laboratory Findings:

- Elevated T3, T4 levels, suppressed TSH.

- Presence of eye signs is diagnostic.

- In the absence of eye signs, an iodine-123 (I-123) scan is recommended.



- Thyroid Scintigraphy: Shows diffuse gland enlargement.

- Antibodies:

- Anti-Tg and anti-TPO elevated in 75% of cases, but are not specific.

- Diagnostic: Detection of thyroid-stimulating antibodies (TsAb) and TSH receptor antibodies.

- Ophthalmopathy Evaluation: Orbital MRI may be indicated.

- RAIU Values: Elevated (45-90%).

Treatment

- Graves' Disease Treatment:

- Classic methods include antithyroid drugs, radioactive iodine (I-131) ablation therapy, and thyroidectomy.

Criteria for Treatment Selection:

- Patient's age

- Severity of the disease

- Gland size

- Associated pathology

- Severity of ophthalmopathy

- Patient preference

- Desire for pregnancy

- Surgery was the most common treatment in the past.

- Non-Surgical Options are now preferred.

Antithyroid Drugs

- Typically used in preparation for radioactive iodine or surgery.

- **Propylthiouracil (PTU):**

    - Initial dose: 100-200 mg every 8 hours; Maximum dose: 1600 mg.

    - Major side effect: Agranulocytosis.

- Methimazole:

    - Initial dose: 10-30 mg every 8 hours.

- Both drugs inhibit all stages of thyroid hormone synthesis.

- PTU also inhibits the peripheral conversion of T4 to T3.

- Both drugs cross the placenta and can suppress fetal thyroid function; they also pass into breast milk.

- PTU is preferred in pregnant and breastfeeding women.

Side Effects:

- Fever

- Skin rashes

- Granulocytopenia

- Peripheral neuritis and polyarteritis

- Vasculitis

- Agranulocytosis and aplastic anemia (rare).

- Serious Complications: Agranulocytosis, aplastic anemia, and toxic hepatitis are the most serious complications of antithyroid drugs.

- Combination Therapy: Severe thyrotoxicosis may require antithyroid drugs alongside a beta-blocker like propranolol, especially effective in treating tachycardia.

- Clinical Improvement: Symptoms improve within the first two weeks of treatment, with most patients achieving a euthyroid state within six weeks.

- Monitoring and Dose Adjustment: Based on weight, pulse rate, TSH, and T4 levels.

- Treatment Duration:

  - Controversial. While some Graves' patients achieve long-term remission, thyrotoxicosis recurs in most, necessitating definitive treatment with radioactive iodine or surgery.

Radioactive Iodine Therapy

- Mainstay in Some Practices: A key treatment for Graves' disease.

- Advantages: Avoids surgical complications.

- Outcome: Six months post-treatment, only about 50% of patients achieve a euthyroid state; others remain hyperthyroid or develop hypothyroidism.

- Ophthalmopathy: Risk of worsening or progression after radioactive iodine therapy.

- Indications:

  - Small to moderate-sized goiters in elderly patients.

  - Recurrence after medical or surgical treatment.

- Contraindications to antithyroid drugs or surgery.

- Absolute Contraindications: Pregnancy and lactation.

- Relative Contraindications:

  - Young patients.

  - Patients with thyroid nodules.

  - Patients with ophthalmopathy.

Surgical Treatment

- Indications:

- Patients where radioactive iodine is contraindicated.

- Suspected or confirmed thyroid cancer.

- Young patients.

- Women planning to conceive shortly after treatment.

- Severe reactions to antithyroid drugs.

- Large, compressive thyroid gland (>80 grams).

- Patients who do not want radioactive iodine.

- Relative Indications:

- Smokers with moderate to severe ophthalmopathy.

- Patients seeking rapid control of thyrotoxicosis.

- Non-compliance with antithyroid medications.

- Pregnancy: Surgery is relatively contraindicated but may be necessary if rapid control of hyperthyroidism is required and antithyroid drugs cannot be used. The optimal timing for surgery is the second trimester.

- Surgical Advantages: Rapid and effective treatment of hyperthyroidism.

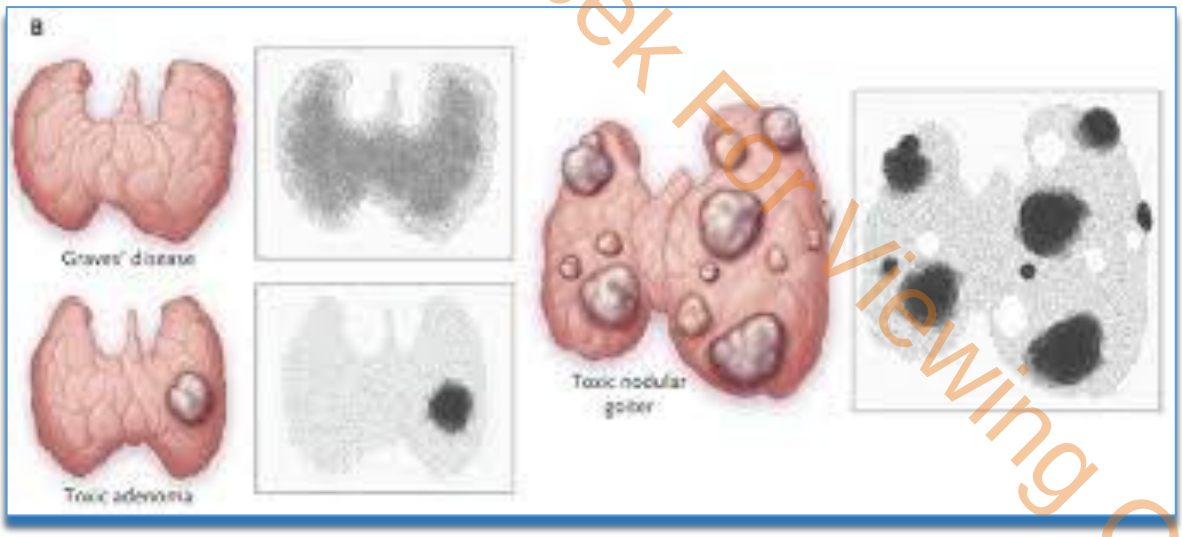
- Ophthalmopathy: May stabilize or improve following total thyroidectomy, likely due to the removal of the antigenic stimulus.

- **Surgical Recommendation:** Total or near-total thyroidectomy is recommended for Basedow Graves disease according to ATA and AACE guidelines.

- **Recurrent Thyrotoxicosis:** Typically managed with radioactive iodine therapy.

Toxic Multinodular Goiter

- **Development:** Typically develops from a non-toxic multinodular goiter.



- **Pathophysiology:** Some nodules may become autonomous and start producing excess hormones.

- **Demographics:** Patients are usually over 50 years old.

- **Onset:** The onset of hyperthyroidism is generally insidious.

- **T3 Thyrotoxicosis:** Some patients may have T3 thyrotoxicosis, where free T4 is normal but free T3 is elevated.

- **Trigger:** In euthyroid multinodular goiter patients, thyrotoxicosis can be triggered by the administration of iodine-containing medications (Jod-Basedow phenomenon).

- **Clinical Presentation:** The clinical picture of hyperthyroidism is similar to Graves' disease but milder, with no extra-thyroidal manifestations.

- **Laboratory Findings:**

  - Elevated T3 and T4 levels, suppressed TSH.

- Increased Radioactive Iodine Uptake (RAIU).

- Imaging: Scintigraphy reveals hyperactive ("hot") nodules with suppression of the remaining thyroid tissue.

Treatment

- **Initial Management:** Hyperthyroidism should first be controlled.

- **Definitive Treatment:** Both radioactive iodine (RAI) therapy and surgery are options.

- **Surgery:** The surgical approach is total or near-total thyroidectomy.

- **Radioactive Iodine Therapy:** This is a suitable option for very elderly patients who have a high surgical risk.

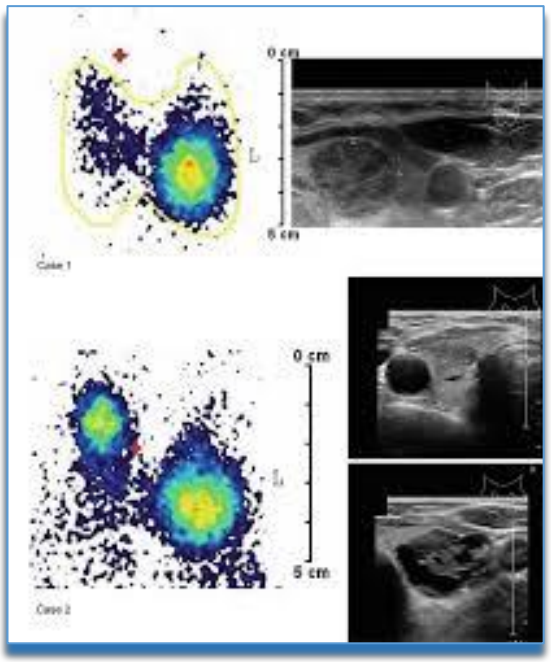
Toxic Adenoma (Plummer's Disease)

- **Characteristics:** There is a single hyperfunctional nodule.

- **Demographics:** Typically seen in younger individuals.

- **Pathology:** A solitary toxic adenoma is usually a follicular neoplasm.

- **Genetics:** Most toxic adenomas are characterized by somatic mutations in the TSH receptor gene.



- **Size:** Generally, the nodule is larger than 3 cm.

- **Physical Examination:** A single thyroid nodule can be palpated.

- Imaging: Scintigraphy shows a single "hot" nodule with suppression of the remaining thyroid tissue.

- Laboratory Findings: T3 and T4 levels are elevated; TSH is suppressed.

- Malignancy Risk: It is rare for a hyperfunctional solitary thyroid nodule to be malignant.

- Treatment:

- Small Nodules: After controlling hyperthyroidism with antithyroid medications, treatment is typically with radioactive iodine.

- Young Patients and Large Nodules: Lobectomy and isthmusectomy are preferred.

Causes of Hypothyroidism

- Primary (Increased TSH levels):

- Hashimoto's thyroiditis

- Radioactive iodine therapy for Graves' disease

- Post-thyroidectomy

- Excessive iodine intake

- Subacute thyroiditis

- Medications: Antithyroid drugs, lithium

- Rare causes: Iodine deficiency, dyshormonogenesis

- Secondary (Decreased TSH levels):

- Pituitary tumor

- Pituitary resection or ablation

- Tertiary (Decreased TSH levels):

- Hypothalamic insufficiency

Refetoff Syndrome (Thyroid Hormone Resistance Syndrome)

- **Pathophysiology:** There is a reduced response to the metabolic effects of thyroid hormones in the pituitary and peripheral tissues.

- **Clinical Features:** Characterized by growth retardation, goiter, and low IQ.

- **Laboratory Findings:** T3, T4, and TSH levels are normal.

- **Mechanism**: Due to tissue resistance, circulating levels of T3 and T4 are insufficient to meet the metabolic needs of cells, leading to increased TSH release from the pituitary. This stimulates further thyroid hormone production until T3 and T4 levels rise enough to inhibit TSH release through negative feedback.

Inflammatory Diseases of the Thyroid Gland (Thyroiditis)

Thyroiditis refers to inflammatory diseases of the thyroid gland, which can be classified as acute, subacute, or chronic.

- **Acute Suppurative Thyroiditis**

- Incidence: It is the rarest form of thyroiditis in adults but more common in children.(after bacterial infection)

- **Subacute Thyroiditis (De Quervain's, Granulomatous, Giant Cell Thyroiditis)**

- Presentation: Subacute thyroiditis can manifest in painful or painless forms.

Painful Subacute Thyroiditis

- **Demographics:** More common in women.

- **Etiology:** The exact cause is unknown. The painful form is often viral in origin and may be associated with a post-viral inflammatory response (linked to HLA-B35 tissue type).

- **Common Age Group:** Frequently seen in women aged 30-40 years.

- **Clinical History:** Often preceded by a history of upper respiratory tract infection.

- **Symptoms:** Characterized by a sudden or gradually increasing neck pain. The thyroid gland is enlarged, tender, and firm.

- **Phases of the Disease:**

1. **Thyrotoxicosis:** Due to the release of thyroid hormones from damaged follicles.

2. Euthyroid Phase: Following the thyrotoxic phase.

3. Hypothyroidism: Occurs in 20-30% of patients.

4. Return to Euthyroid State: More than 90% of patients eventually return to a normal thyroid state.

- Laboratory Findings: Early in the disease, T3 and T4 levels are high, and TSH is suppressed.

- Erythrocyte Sedimentation Rate (ESR): > 100 mm/hour.

- Radioactive Iodine Uptake (RAIU): Low, despite thyrotoxicosis.

- Treatment: The condition is self-limiting. Symptomatic treatment includes aspirin, nonsteroidal anti-inflammatory drugs (NSAIDs), steroids, and beta-blockers.

Painless Subacute Thyroiditis

- Etiology: Believed to be of autoimmune origin.

- Incidence: Occurs sporadically or in the postpartum period (typically about 6 weeks after delivery, particularly in women with high anti-TPO antibodies in early pregnancy).

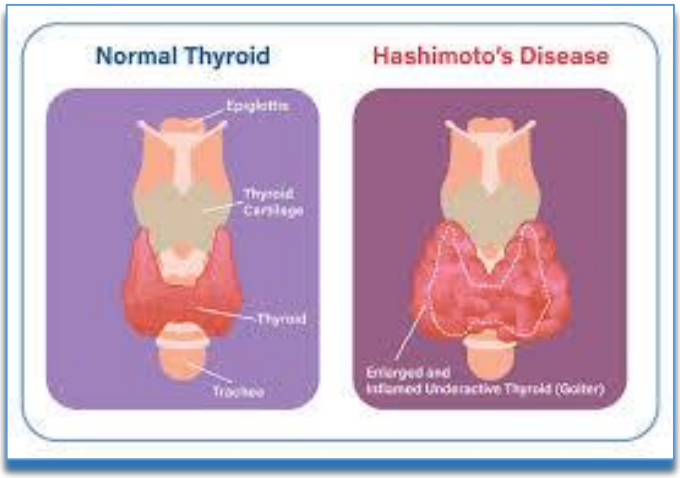
- Demographics: More common in women, usually between the ages of 30-60 years.

- Laboratory and Imaging: Similar to painful thyroiditis, except for a normal ESR.

- Symptoms: Similar clinical presentation to painful thyroiditis.

- Treatment: Symptomatic patients are treated with beta-blockers and thyroid hormone replacement.

Hashimoto's Thyroiditis (Lymphocytic Thyroiditis)



- Alternative Names: Chronic lymphocytic or autoimmune thyroiditis.

- Prevalence: It is the most common type of thyroiditis.

- Common Cause: The most frequent cause of hypothyroidism.

- Pathophysiology: An autoimmune disease initiated by the activation of CD4+ (helper) T cells specific to thyroid antigens, leading to hypothyroidism.

- Causes of Hypothyroidism:

1. Destruction of thyrocytes by cytotoxic T cells.

2. Blockage of TSH receptors by autoantibodies.

- Antibodies: Antibodies are formed against three main antigens:

- Thyroglobulin (Tg) (60%)

- Thyroid peroxidase (TPO) (95%)

- TSH receptor (TSH-R) (60%)

- Associated Factors: Increased iodine intake, interferon-alpha, lithium, and amiodarone use.

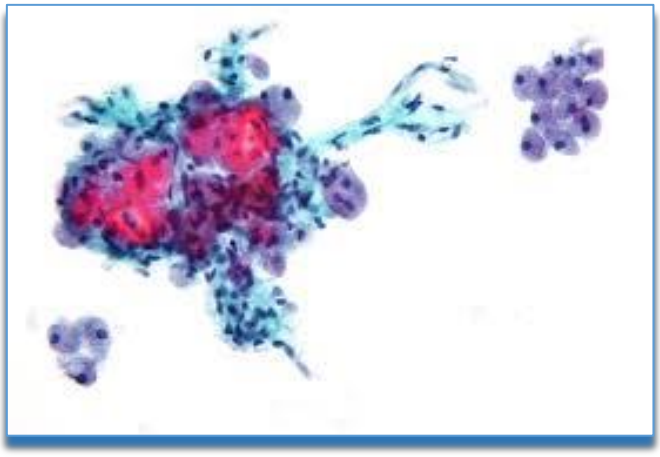
- **Genetic Predisposition:** There is evidence supporting genetic susceptibility.

- Demographics: Women are affected 10 times more often than men. Typically occurs between the ages of 30-50.

- Physical Findings: The thyroid is often symmetrically enlarged, pale, and semi-firm.

- Pathology:

- Diffuse infiltration of lymphocytes and plasma cells in the thyroid.



- Reduced colloid content and smaller thyroid follicles.

- Increased fibrous tissue between cells.

- Diagnostic presence of **Hürthle/Askanazy** cells, characterized by abundant eosinophilic, granular cytoplasm.

Clinical Features:

- Common Presentation: Most often presents as a painless neck mass, with physical examination revealing a mildly to moderately enlarged, firm thyroid gland.

- Thyroid Function: In early stages, hyperthyroidism (Hashitoxicosis) can sometimes occur, but most patients are euthyroid or hypothyroid at the time of diagnosis.

- Nodular Growth: Nodular enlargement is seen in 20% of cases.

- Lab Findings: Low T4 and T3 levels with elevated TSH. Positive thyroid autoantibodies.

- Complications: A solitary nodule or rapidly enlarging goiter in the setting of Hashimoto's thyroiditis may suggest thyroid lymphoma, requiring fine-needle aspiration biopsy (FNAB).

Treatment:

- **Hypothyroidism:** Treated with thyroid hormone replacement therapy.

- **Surgical Indications (Rare):**

  - Growth despite treatment, especially asymmetric growth.

  - Suspicion of malignancy.

  - Compression symptoms.

  - Cosmetic concerns.

Riedel's Thyroiditis (Riedel's Struma)

- **Alternative Names:** Invasive fibrous thyroiditis.

- **Incidence:** Rare condition with a debated etiology.

- **Association:** Can be found alongside autoimmune diseases like pernicious anemia or Graves' disease, and with primary sclerosing pathologies like retroperitoneal, mediastinal, periorbital, and retro-orbital fibrosis, and sclerosing cholangitis.

- **Physical Findings:** The thyroid is rock-hard due to fibrosis.

- **Demographics:** Most commonly seen in women aged 30-60.

- **Symptoms:** Characteristically presents as a painless, hard thyroid gland.

- **Compression Symptoms:** Dyspnea, dysphagia, cough, and hoarseness.

- **Complications:** Hypothyroidism and hypoparathyroidism may develop due to fibrous tissue infiltration.

- **Physical Examination:** The thyroid gland is palpated as "wood-like" in hardness, fixed to surrounding tissues, and bilateral involvement is typical.

- **Differential Diagnosis:** Clinically resembles anaplastic carcinoma, and can only be distinguished by open biopsy, as FNAB is insufficient.

Treatment:

- **Primary Treatment:** Surgery, specifically isthmusectomy (wedge resection of the isthmus).

- **Additional Therapies:** For patients with persistent symptoms, corticosteroids and tamoxifen may be beneficial.

Solitary or Dominant Thyroid Nodules

- **Overview:** Thyroid nodules are common lesions, most of which are benign. These include cysts, colloid nodules, or adenomas.

- **Prevalence:** The incidence of thyroid nodules increases with age. Solitary nodules are four times more common in women than in men.

- **Management:** The choice between conservative and surgical treatment depends on history, clinical findings, physical examination, imaging, and diagnostic information.

Risk Factors for Thyroid Cancer:

- Radiation Exposure: The most significant risk factor is a history of radiation to the head and neck area. Patients exposed to low-dose radiation to the thyroid gland have an increased risk of both benign and malignant thyroid diseases. Cancer risk increases linearly between 6.5 cGy and 2000 cGy.

- Age and Gender: Nodules that appear at a young age or late in life are more likely to be malignant. The highest risk is in men over 60 years old.

- Nodule Characteristics: Rapid growth, changes in size, pain, dyspnea, dysphagia, and hoarseness increase the likelihood of malignancy.

- Family History: A family history of thyroid cancer, both medullary and non-medullary, is a risk factor.

Physical Examination

- Malignancy Indicators: Solitary, hard nodules with irregular surfaces and fixation to surrounding tissues are more likely to be cancerous.

- Multiple nodules or diffuse nodularity is more commonly associated with benign conditions.

- Enlarged lymph nodes in the neck or thyroid isthmus (Delphian node) may suggest metastasis.

Imaging Methods

- Ultrasound:

    - Detects non-palpable thyroid nodules.

    - Determines the size of the nodule.

    - Differentiates cystic vs. solid nodules.

    - Identifies adjacent lymphadenopathy.

    - Guides fine-needle aspiration biopsy (FNAB).

- CT and MRI: Generally not required.

- Thyroid Scintigraphy (I-131 and Tc-99m):

    - Its use has significantly decreased.

    - Recommended for: - Autonomous thyroid nodules and Graves' disease patients.

        - Cases with follicular lesions found on FNAB.

        - Patients with suppressed TSH levels.

Laboratory Evaluation

- Thyroid Function Tests: Routine use is debated. Most patients with thyroid nodules are euthyroid, but measuring TSH levels can be beneficial.

- Cancer Risk: - If hyperthyroidism is found in a patient with a thyroid nodule, cancer risk is about 1%.

- Serum thyroglobulin levels are not useful in distinguishing between benign and malignant nodules but very high levels may suggest metastatic thyroid cancer.

Fine-Needle Aspiration Biopsy (FNAB)

- Importance: The most crucial test for evaluating thyroid nodules.

- Guidance: Can be performed under ultrasound guidance or directly.

- **Diagnostic Categories:**

- Non-diagnostic / Inadequate (2%-20%): Due to cyst fluid or clot. Cancer risk in this group is 1%-4%. Repeating the aspiration is recommended.

- Benign (60%-70%): Common lesions include follicular nodules (adenomatous nodules, colloid nodules, and follicular adenomas), lymphocytic (Hashimoto) thyroiditis, and granulomatous thyroiditis. False-negative results occur in about 3%, requiring follow-up.

- Atypia of Undetermined Significance (AUS) / Follicular Lesion of Undetermined Significance (FLUS) (3%-6%): Cancer risk for FLUS is about 10%-35%, and for AUS, 60%-75%. Sabiston reports a cancer risk of 15%. Repeat FNAB is appropriate for these cases. If clinical and ultrasound findings are concerning, surgery (lobectomy + isthmusectomy) is performed.

- Follicular Neoplasm Group: Includes cases that may be follicular carcinoma. Lobectomy + isthmusectomy is performed. Cancer rate in this group is 15%-35%. Hürthle cell neoplasms are also included in this group.

- Suspicious for Cancer: Most papillary and other carcinomas are identifiable via FNAB. Sometimes FNAB results are unclear (e.g., in the follicular variant of papillary carcinoma). Lesions with uncertain diagnosis are classified as "suspicious for cancer." Surgery (lobectomy or near-total thyroidectomy) is performed, with malignancy rates around 60%.

- Malignant: Cancer risk is 97%-99%. Near-total or total thyroidectomy is indicated.

Treatment

- Cystic Nodules: - Aspiration: Often cures 75% of the time. Some cysts may require 2-3 aspirations.

- If the cyst persists after 3 aspirations, lobectomy is appropriate.

- For cysts larger than 4 cm or those with a solid component, direct lobectomy is preferred.

Malignant Tumors of the Thyroid Gland

- Papillary, Follicular, and Hürthle Cell Carcinomas: These account for 90-95% of thyroid cancers and are well-differentiated.

- **Hürthle Cell Carcinomas:** Considered a subtype of follicular cancers.

- **Medullary Carcinomas:** Represent about 5% of thyroid cancers.

- **Anaplastic Carcinomas:** Occur in approximately 1% of cases.

- **Other Types:** Lymphomas and metastatic cancers can also be observed.

Thyroid Oncogenesis

- Role of Genetic Mutations:

- Both mutated proto-oncogenes and changes in tumor suppressor genes play a role in thyroid neoplasms.

- **Common Mutation:** The RET mutation is the most frequently encountered.

- **Endemic Goiter Regions:** Anaplastic and follicular cancers primarily develop in areas with iodine deficiency (endemic goiter regions).

Here is a list of thyroid-related genes:

- **RET:** Proto-oncogene involved in several thyroid cancers, including medullary thyroid carcinoma.

- **MET:** Proto-oncogene associated with various cancers, including thyroid cancer, affecting cell growth and differentiation.

- **TRK1:** A receptor tyrosine kinase involved in neurotrophin signaling, which can be implicated in thyroid cancer.

- **TSH-R:** Thyroid-stimulating hormone receptor, which, when mutated, can contribute to thyroid disorders.

- **Gsa (gsp):** Mutant form of the G protein subunit (Gsa) involved in stimulating thyroid hormone production.

- **PAX8/PPARγ1:** A fusion gene associated with follicular thyroid carcinoma.

- **BRAF:** A gene encoding a protein involved in cell signaling; mutations are commonly found in papillary thyroid carcinoma.

- CTHNB1: Likely referring to CTNNB1 (Beta-catenin), a protein involved in the Wnt signaling pathway; mutations can lead to thyroid cancer.

- p53: A tumor suppressor gene that helps regulate the cell cycle; mutations can contribute to thyroid cancer.

- p16: A tumor suppressor protein involved in regulating the cell cycle; its alteration can be associated with various cancers, including thyroid cancer.

- PTEN: A tumor suppressor gene that regulates cell growth and division; mutations can be linked to thyroid cancer and other malignancies.

Papillary Thyroid Cancer

- The most common type of thyroid cancer.

- Accounts for 80% of all thyroid cancers.

- Most common thyroid cancer in children and those with a history of radiation exposure.

- Typically seen around the ages of 30-40.

- Has the best prognosis among thyroid cancers.

- Can be more aggressive in older age.

- Twice as common in women.

- Clinical Features:

- Most patients are euthyroid.

- Presence of a slowly growing, painless thyroid mass.

- Local invasive disease may present with hoarseness, dysphagia, and dyspnea.

- Cervical lymph node metastases are common, especially in children and young adults.

- Distant metastases most frequently occur in the lungs, followed by bones, liver, and brain.

- Suspicion of thyroid cancer typically starts with palpation of a single, hard nodule during physical examination and assessment of the patient's history.

- Diagnosis is confirmed by fine-needle aspiration biopsy (FNAB) of the thyroid nodule or lymph node.

- Pathology:

- Papillary thyroid cancer originates from follicular cells that produce thyroid hormone.

- Histologically, the defining feature of papillary cancer is the presence of papillae.

- Characteristic cellular features in FNAB for papillary thyroid cancer include (Orphan Annie nuclei):

- Presence of **psammoma** bodies in the stroma.

- Subtypes of papillary cancer include: - Classic type variants: Long cell (tall cell). Insula .Columnar . Diffuse sclerosing . Clear cell . Trabecular . Poorly differentiated type

- These subtypes represent less than 1% of papillary cancers, are more common in older age, and have a worse prognosis.

- Due to intra-thyroidal lymphatic spread, the rate of multifocality is very high (85%).

- Occult or microcarcinomas: Tumors < 1 cm. No local invasion or angioinvasion. No lymph node metastases.

- Lymph node metastases do not significantly affect mortality.

- Prognosis:

- Papillary cancers generally have a slow progression and a good prognosis.

- Ten-year survival rate exceeds 95%.

- Prognostic scoring systems have been developed to assess factors affecting prognosis:

- Age at diagnosis is a crucial clinical prognostic factor; age < 40 is favorable.

- Absence of distant metastases at initial treatment and tumor size < 4 cm are also indicators of a good prognosis.

- Lymph node metastases do not affect long-term survival in patients whose papillary cancer has not extended beyond the thyroid capsule.

Treatment:

- Total or near-total thyroidectomy is performed.

- For incidental findings of < 1 cm, low-risk, unifocal, intrathyroidal papillary cancers with no history of head and neck radiation or cervical lymph node involvement, a thyroid lobectomy alone may be sufficient.

- If there is cervical lymph node involvement, a modified radical neck dissection is performed on the affected side.

Follicular Thyroid Cancers

- Account for approximately 10% of thyroid cancers.

- Typically seen in individuals over 50 years of age.

- The female-to-male ratio is 3:1.

- The incidence of follicular cancer is higher in areas with iodine deficiency (endemic goiter).

- A subtype of follicular cancer, known as Hürthle cell cancer, originates from oxyphilic cells and tends to occur in older individuals (60-75 years).

Pathology:

- Macroscopically, it is a tumor with a capsule.

- Histologically, it is composed of follicles, but the lumen may not contain colloid.

- The diagnosis of cancer is confirmed by the presence of capsular and vascular invasion.

- In the absence of capsular and vascular invasion and if composed of well-differentiated follicular cells, it is classified as a follicular adenoma (benign tumor).

- Capsular invasion alone has a better prognosis compared to vascular invasion.

- Multifocality is much less common compared to papillary cancers.

- Cervical lymphadenopathy is rare (5%).

- Less than 1% of cases may present as a hyperfunctional nodule.

- Metastatic spread occurs via hematogenous routes; common sites include bone, lung, and liver.

Clinical Features:

- Follicular cancers typically present as a painless, solitary thyroid nodule.

- Hoarseness and a hard, fixed mass suggest advanced disease.

- Cervical lymphadenopathy is rare; distant metastases, especially bone metastases, may be present.

- Follicular cancer rarely shows hyperfunction (<1%).

- Most are euthyroid.

- Most cases are solitary.

- FNAB cannot differentiate between follicular cancer and follicular adenoma because the diagnosis requires evidence of capsular and/or vascular invasion, which FNAB cannot provide. Hence, FNAB results are reported as follicular lesions (neoplasms).

- Thus, it is challenging to diagnose follicular cancer pre-operatively unless distant metastases are present.

- However, in older men, follicular tumors larger than 4 cm have a higher likelihood of malignancy.

- Treatment:

- The primary treatment for follicular cancer is surgery.

- For patients with FNAB results showing follicular lesions, a thyroid lobectomy including the isthmus and pyramidal lobe (on the affected side) should be performed.

- Most follicular lesions are adenomas, and if pathological examination confirms this, lobectomy plus isthmusectomy will be sufficient.

- "Frozen section" examination during surgery is generally not helpful for diagnosis. However, it should be performed if significant capsular or vascular invasion or lymphadenopathy is observed.

- If cancer is confirmed after lobectomy and isthmusectomy, a total thyroidectomy is performed.

- For patients with lymph node metastases, therapeutic neck dissection is also added.

- Follicular cancers are more aggressive than papillary cancers and have a worse prognosis.

- Factors Affecting Prognosis:

- Age at diagnosis > 50 years

- Tumor size > 4 cm

- High grade

- Significant vascular invasion

- Extrathyroidal invasion

- Distant metastases at diagnosis

- **Prognosis**:

- The prognosis after treatment depends on age.

- The best prognosis is in patients under 40 years old, with a 5 and 10-year survival rate of 95%.

Postoperative Surveillance and Treatment of Differentiated Thyroid Cancers

- Thyroid Hormone Therapy:

- Reasons for Administering Thyroxine:

  - Thyroid hormone replacement.

  - TSH suppression.

- TSH suppression reduces tumor recurrence rates.

- Radioactive Iodine Therapy:

- Reasons for Administering Radioactive Iodine:

  - To ablate residual thyroid tissue after surgery.

  - To treat any remaining primary tumor or metastases to surrounding tissues.

  - To treat distant metastases.

- **Current ATA Guideline for RAI (Radioactive Iodine) Indications**

  - Definite Indications:

    - Distant metastases after total thyroidectomy.

    - Patients with gross extrathyroidal extension.

    - Tumors larger than 4 cm.

  - For tumors between 1-4 cm, RAI may be recommended based on high-risk factors:

    - Certain histological subtypes (tall cell, columnar, insular, solid variants, and poorly differentiated thyroid cancer).

    - Intrathyroidal vascular invasion.

- Gross or microscopic multifocal disease.

- No indication for RAI if tumor size is < 1 cm or if multifocal tumors sum < 1 cm.

- Patient Follow-Up:

- Recurrence of differentiated thyroid cancers can occur years later, so long-term follow-up is necessary to reduce morbidity and mortality related to recurrence.

- Recurrence tends to occur more quickly in follicular cancers and later in papillary cancers.

- Measurement of thyroglobulin levels is the most sensitive test for detecting recurrence and metastases, with an accuracy of 85%-95%.

- Thyroglobulin and anti-thyroglobulin levels should be monitored every 6 months for the first 2 years and then annually.

- If thyroglobulin levels are high:

  - Full body iodine scan (131I) should be performed.

  - If iodine scan is negative, other investigations (chest X-ray, neck ultrasound, CT/MRI of the mediastinum, bone scintigraphy, PET scan) should be conducted.

- If thyroglobulin levels are normal:

  - Full body iodine scan (131I) may not be required. It is recommended to perform a total body iodine scan once yearly for the first 2 years, and then every 3-5 years if initial tests are negative.

Medullary Thyroid Cancer

- Represents about 5% of thyroid cancers.

- Originates from C cells, which are derived from neural crest cells and are part of the APUD system.

- C cells are predominantly located in the upper thyroid lobes and produce calcitonin.

- Approximately 25% of medullary cancers are familial, while most develop sporadically.

- Familial medullary cancers can be associated with or without multiple endocrine neoplasia (MEN) syndromes.

- All familial medullary cancers have mutations in the RET proto-oncogene.

- Familial Medullary Cancer Syndromes:

- Familial Medullary Thyroid Cancers:

- Part of MEN-2 (Sipple syndrome) or can occur without other endocrine pathologies.

- MEN-2 is divided into MEN-2A and MEN-2B.

- MEN-2A:

- The most common form of MEN.

- Associated with medullary cancer, pheochromocytoma or adrenal medullary hyperplasia, and hyperparathyroidism.

- Some patients may also have Hirschsprung disease and cutaneous amyloidosis.

- MEN-2B:

- Characterized by medullary cancer and pheochromocytoma.

- Mucosal neuromas on the tongue, lips, and conjunctiva, ganglioneuromas in the intestines, and a marfanoid appearance are observed.

- Pathology:

- Sporadic medullary cancers are typically unilateral, while familial cases are usually multicentric.

- In familial cases, C cell hyperplasia, considered a premalignant lesion, is frequently observed.

- The presence of amyloid deposits in the tumor stroma is diagnostic.

- Tumors are also positive for CEA (Carcinoembryonic Antigen) and calcitonin gene-related peptide.

- Clinical Features:

- Usually occurs between ages 50-60; familial cases are seen at younger ages.

- Familial forms are inherited in an autosomal dominant manner.

- Slightly more common in women (female-to-male ratio of 1.5:1).

- Typically presents as a neck mass, with cervical lymph nodes involved in 15-20% of cases.

- Local pain is more common in these patients.

- Tumors with local invasion may cause dysphagia, dyspnea, or hoarseness.

- Medullary thyroid cancer can secrete various peptides such as calcitonin, CEA, histaminase, prostaglandin E2 and F2-alpha, serotonin, ACTH, VIP, substance P, somatostatin, bombesin, and beta-endorphin, leading to uncommon symptoms.

- Disease-specific symptoms (paraneoplastic features) may include:

  - Diarrhea (most common)

  - Episodic flushing

  - Cushing's syndrome (due to ectopic ACTH secretion in about 2-4% of patients)

- Tumor primarily spreads to cervical lymph nodes, then to the upper mediastinum.

- Hematogenous spread can occur to the liver, bones, and lungs.

- Diagnosis:

- Diagnosis is based on history, physical examination, elevated serum calcitonin or CEA levels, and fine-needle aspiration biopsy (FNAB) of the thyroid mass.

- Elevated calcitonin levels (>300 pg/ml, normal range 100-200 pg/ml) with a thyroid mass are diagnostic of medullary thyroid cancer.

- Since sporadic and familial forms cannot be distinguished initially, all patients diagnosed with medullary thyroid cancer should be evaluated for RET point mutations, pheochromocytoma, and hyperparathyroidism.

- Treatment:

- Standard Initial Surgery:

  - Total thyroidectomy with central neck dissection.

  - Modified radical neck dissection is added if palpable or imaging-detected cervical lymph nodes are present.

- Prophylactic Modified Radical (Lateral) Neck Dissection:

  - Controversial; recommended if tumor >1.5 cm or central lymph nodes are involved.

- Pheochromocytoma: If present, it should be treated first with bilateral total adrenalectomy.

- Hypercalcemia: Parathyroidectomy if necessary.

- RET Mutation Carriers:Total thyroidectomy is indicated when RET mutations are detected, ideally before age 5 for MEN-2A and before age 1 for MEN-2B.

- **Prognosis:**

- Related to disease stage.

- From best to worst prognosis: Familial, non-MEN medullary thyroid cancer--- MEN-2A--- Sporadic medullary thyroid cancer--- MEN-2B

- Prognostic Significance of Calcitonin and CEA: Calcitonin is a more sensitive tumor marker. CEA provides better prognostic information.

Anaplastic Thyroid Cancer

- Represents about 1% of thyroid cancers.

- The most aggressive type of thyroid cancer.

- Common in endemic goiter regions.

- More frequent in women, with most patients in their 70s and 80s.

- Often develops from other differentiated thyroid cancers.

- Characterized by a hard, fixed, rapidly progressing tumor that invades surrounding structures.

- Common symptoms include hoarseness, dyspnea, dysphagia, and cervical tenderness.

- Lymph nodes in the neck are often enlarged at the time of diagnosis, and distant metastases may be present.

- Total thyroidectomy, if possible, is a treatment option that can slightly increase survival.

- Adjuvant combined radiotherapy and chemotherapy can extend survival in patients who can undergo surgery.

- Radioactive iodine is ineffective.

Thyroidectomy Complications

- General Complications:

- Complication rates are higher in cases of thyroid cancer, thyroiditis, and hyperthyroidism.

- The risk of complications is higher in recurrent cases compared to the initial surgery.

- Hypoparathyroidism:

- The most common complication of thyroidectomy.

- Temporary hypocalcemia occurs in about 50% of cases.

- Most cases resolve within approximately one year.

- Permanent hypoparathyroidism is much rarer (<2%).

- More common in situations such as:

  - Reoperation for recurrence

  - Central and lateral neck dissection

  - Surgery for Graves' disease

- Recurrent Laryngeal Nerve Injury:

- Innervates all intrinsic muscles of the larynx except the cricothyroid muscle.

- Rarely enters the larynx directly from the vagus nerve (non-recurrent).

- Frequently traumatized in the last 2-3 centimeters of its course in the neck (Berry's ligament).

- During thyroidectomy, the nerve should be identified and preserved due to its proximity to the thyroid.

- If recurrent nerve injury is detected during surgery, suturing the nerve ends together is recommended.

- Early postoperative hoarseness, especially if bilateral, may cause respiratory distress.

- Non-recurrent nerve presence (usually on the right) can increase the risk of injury.

- Superior Laryngeal Nerve Injury:

- The external branch innervates the cricothyroid muscle.

- The internal branch provides sensory innervation to the larynx.

- The external branch may be injured in about 20% of patients during ligation of the upper thyroid vessels.

- Injury to this nerve leads to significant voice quality deterioration and difficulty producing high-pitched sounds.

- Injury to the internal branch is much rarer but can cause aspiration of liquids postoperatively.

- Bleeding and Hematoma:

- The thyroid is a highly vascular organ.

- Postoperative bleeding can lead to neck swelling and pressure symptoms (e.g., respiratory distress).

- Emergency drainage is required if there is severe respiratory distress.

- Bleeding and hematoma occur in less than 1% of cases with meticulous hemostasis.

- Thyroid Storm:

- A life-threatening complication characterized by extreme symptoms and signs of hyperthyroidism.

- Very rare if proper preoperative preparation is done.

- Cervical Sympathetic Chain Injury:

- Risk is increased in invasive thyroid cancers and retroesophageal goiters.

- May lead to Horner's syndrome.

- Spinal Accessory Nerve Injury:

- Can occur due to neck dissection.

- Results in inability to lift the shoulder and weakness in shoulder movements due to denervation of the trapezius muscle.

Chapter
13.

Parathyroid Gland Diseases and Surgery

Embryology and Anatomy of the Parathyroid Glands

- The parathyroid glands originate from the branchial pouches.

- The superior parathyroid glands develop from the 4th branchial pouch, while the inferior parathyroid glands arise from the 3rd branchial pouch.

- Number of Glands:

- Typically, there are 4 parathyroid glands (about 80% of cases).

- In about 13% of cases, there are more than 4 glands; in 5% of cases, there are 3 or fewer glands; and in another 5% of cases, there are 6 glands.

- Location:

- The location of the superior parathyroid glands is more consistent. In 80% of cases, they are found near the cricoid cartilage level, typically located on the posterior aspect of the thyroid gland, in the middle to upper region.

- The location of the inferior parathyroid glands is less consistent. They are generally found within a 2 cm diameter circle centered at the junction of the inferior laryngeal nerve and the inferior thyroid artery. However, they can vary in location and may be found in places such as the thymus, upper mediastinum, pericardium, or paraesophageal groove. Approximately 2% are intrathyroidal.

- Cellular Composition:

- The parathyroid glands consist of several types of cells, including:

  - **Chief Cells:** These cells secrete parathyroid hormone (PTH).

  - **Oxyphil Cells:** Acidophilic cells that are more abundant in older individuals.

  - **Clear Cells:** Cells that contain a large amount of glycogen, giving them a clear appearance.

Hyperparathyroidism

- Definition: Hyperparathyroidism is the condition where the parathyroid glands are overactive.

- Classification: It can be classified as primary, secondary, or tertiary.

Primary Hyperparathyroidism

- **Pathophysiology:** In primary hyperparathyroidism, there is an increased secretion of parathyroid hormone (PTH) from pathological parathyroid glands.

- Clinical Presentation:

- Characterized by elevated PTH and calcium levels with low phosphate levels.

- It is the most common cause of hypercalcemia in the general population.

- Among hospitalized patients, it is the second most common cause of hypercalcemia after cancer.

- Commonly seen in 0.1% to 0.3% of the population.

- It is four times more common in women and its incidence increases with age, particularly in postmenopausal women.

- Most cases of primary hyperparathyroidism are sporadic.

- It can also occur in association with hereditary syndromes such as MEN1 (Multiple Endocrine Neoplasia type 1), MEN2A, isolated familial hyperparathyroidism, and jaw tumor syndrome.

MEN 1 (Wermer Syndrome, PPP)

- Parathyroid: Hyperparathyroidism is the earliest and most common manifestation of MEN 1, often presenting before the age of 40.

- **Pituitary Adenoma:** Frequently presents as prolactinoma.

- **Pancreatic Islet Cell Tumor:** Gastrinoma is the most common.

- **Etiology of Primary Hyperparathyroidism**

- Exact Cause: The exact cause is often unknown.

- Low-dose Radiation Exposure: There is an association with low-dose radiation exposure, typically manifesting 30-40 years after exposure.

- **Familial Predisposition:** Genetic factors may play a role in some cases.

- **Diet:** Certain diets may be associated with the condition.

- **Intermittent Sunlight Exposure:** May be related to the development of primary hyperparathyroidism.

- **Aging and Kidney Function:** Decreased renal function and calcium leakage with advancing age may contribute.

- **Lithium Therapy:** Lithium treatment is known to be associated with increased PTH levels and mild hypercalcemia.

- **Pathogenesis**

- **Single Gland Disease:** About 80% of cases are due to a single parathyroid adenoma.

- **Multiple Gland Disease:** In 15-20% of cases, multiple gland involvement (hyperplasia or multiple adenomas) occurs.

- **Parathyroid Cancer:** About 1% of cases are related to parathyroid carcinoma.

- **Clinical Manifestations**

- **Symptomatology:** Most patients are minimally symptomatic or asymptomatic. However, detailed questioning often reveals non-specific symptoms such as fatigue, polyuria, polydipsia, nocturia, bone and joint pain, constipation, anorexia, nausea, itching, depression, and memory loss.

- **Renal Complications:**

  - Renal symptoms or some degree of kidney dysfunction are present in most patients.

  - The most severe clinical consequences of primary hyperparathyroidism are often kidney-related.

  - Kidney stones occur in approximately 25-30% of cases.

  - Other renal complaints include polyuria, polydipsia, and nocturia.

- **Hypertension:**

  - Occurs frequently, in about 50% of cases, and is more common in elderly patients.

  - It is related to the degree of renal dysfunction and is the symptom least likely to improve after parathyroidectomy.

- **Bone Disease:**

  - Overt bone disease is rare (5-15% of cases).

- Bone pain, tenderness, and pathological fractures are uncommon, whereas osteopenia and osteoporosis are more frequent.

- Osteitis fibrosa cystica is very rare.

- Brown tumors and bone cysts may be present.

- The extent of bone damage correlates with alkaline phosphatase levels.

- Neurological, Psychiatric, and Neuromuscular Symptoms:

- Patients may exhibit a range of neurological or psychiatric disorders, from depression and anxiety to psychosis or coma.

- Symptoms such as muscle fatigue, weakness, and exhaustion, especially in the proximal muscles, may also develop.

- Gastrointestinal Symptoms:

- Increased incidence of peptic ulcers, pancreatitis, and gallstones.

- Pancreatitis is generally seen only in patients with severe hypercalcemia (calcium levels above 12.5 mg/dL).

- **Diagnosis**

- Laboratory Findings:

- Diagnosis is made by elevated calcium and serum intact PTH (iPTH) levels.

- About half of the patients have low serum phosphate levels.

- Approximately 60% have increased 24-hour urinary calcium excretion (>200 mg/day).

- In benign familial hypocalciuric hypercalcemia, 24-hour urinary calcium excretion is distinctly low (<100 mg/day).

- Chloride levels are elevated, and mild hyperchloremic metabolic acidosis may be present in 80% of cases.

- A chloride/phosphate ratio >33 is diagnostic.

- Alkaline phosphatase is elevated in 10-40% of patients.

- Normocalcemic Primary Hyperparathyroidism:

- Conditions that may cause normocalcemia in primary hyperparathyroidism include:

- Vitamin D deficiency

- Hypoalbuminemia

- Excessive hydration

- High dietary phosphate intake

- Imaging:

- Routine radiological investigations often do not detect bone lesions in most patients.

- Imaging is recommended only for patients with elevated alkaline phosphatase levels.

- Advanced cases with severe hypercalcemia may show characteristic findings of osteitis fibrosa cystica on X-rays.

- Skull X-rays may reveal a "pepper pot" appearance due to cortical bone erosion.

- Hand X-rays may show subperiosteal resorption, particularly in the middle and distal phalanges.

- Bone mineral densitometry studies are increasingly used to assess the impact of primary hyperparathyroidism on bone and have become the standard method for diagnosing osteoporosis in these patients.

> **Physical Examination:** A palpable neck mass in a patient with primary hyperparathyroidism suggests either parathyroid cancer or a thyroid nodule, as parathyroid tumors are generally not palpable.

- **Treatment**

- Surgery is recommended if the patient is symptomatic or younger than 50 years old.

- Asymptomatic Patients:

- Asymptomatic primary hyperparathyroidism is defined by the absence of common symptoms and findings associated with bone, kidney, gastrointestinal, or neuromuscular issues.

- Asymptomatic patients with primary hyperparathyroidism can be managed with observation rather than immediate surgery.

- However, some of these patients may eventually require surgical treatment.

Indications for Surgery in Asymptomatic Primary Hyperparathyroidism

- Significant Hypercalcemia: Calcium levels greater than 1 mg/dL above the normal range.

- Decreased Glomerular Filtration Rate (GFR): GFR less than 60 ml/min.

- Reduced Bone Mineral Density: A decrease in bone mineral density (measured in the lumbar spine, hip, or distal radius) by more than 2.5 standard deviations from normal bone mass (T-score < -2.5).

- Inability to Ensure Close Monitoring: When close follow-up is not feasible.

- Age Below 50 Years.

Surgical Treatment

- Single Adenoma: The treatment is solitary parathyroidectomy.

- Multiple Adenomas: If two or three parathyroid glands are enlarged, the enlarged glands are removed. Therefore, it is essential to visualize and evaluate all four parathyroid glands during standard surgical treatment.

- Multiglandular Hyperplasia: If all glands are enlarged, subtotal parathyroidectomy (removal of 3.5 glands) or total parathyroidectomy with autotransplantation is performed.

Secondary Hyperparathyroidism

- In secondary hyperparathyroidism, PTH levels are elevated, but this is due to chronic stimulation of the parathyroid glands caused by decreased serum calcium levels.

- The primary cause of the low serum calcium is usually kidney disease. However, it can also develop due to insufficient calcium or vitamin D intake or malabsorption.

- The parathyroid glands undergo chief cell hyperplasia.

- Patients are generally hypocalcemic or normocalcemic.

- Accompanying hyperphosphatemia further increases PTH secretion. Intact PTH (iPTH) levels, which normally range from 10-65 pg/mL, often rise to 500-1500 pg/mL.

- In patients with chronic kidney failure, significant alkaline phosphatase elevation and itching are warning signs.

- The skeletal changes in secondary hyperparathyroidism are similar to those in primary hyperparathyroidism, with the key difference being that ectopic calcifications are much more common in secondary hyperparathyroidism.

- The initial treatment for secondary hyperparathyroidism is medical.

- Dietary phosphate intake is restricted, and oral phosphate binders are given to treat hyperphosphatemia.

- Calcium and vitamin D replacement therapy is administered.

- Calcimimetic drugs may be used to reduce plasma PTH levels.

Surgical Indications in Secondary Hyperparathyroidism:

- Persistent bone pain and itching despite maximal medical therapy.

- Calcium-phosphate product > 70.

- Calcium > 11 mg/dL with very high PTH levels.

- Calciphylaxis:

  - A severe complication of secondary hyperparathyroidism that can affect extremities and be life-threatening.

  - Characterized by painful, purplish extremities.

  - Frequently becomes necrotic, leading to non-healing ulcers, gangrene, sepsis, and death.

- Progressive renal osteodystrophy:

  - Patients with soft tissue calcification and tumoral calcinosis.

- Localization studies are not necessary.

- Since all glands are hyperplastic, bilateral neck exploration is required.

- Subtotal parathyroidectomy or total parathyroidectomy with forearm autotransplantation is performed.

Tertiary Hyperparathyroidism:

- Tertiary hyperparathyroidism develops due to the autonomy of the chief cell hyperplasia seen in secondary hyperparathyroidism.

- Surgical intervention is indicated if symptomatic disease is present or if autonomous PTH secretion persists for more than a year following a successful kidney transplant.

Complications of Parathyroidectomy:

- Vocal cord paralysis due to recurrent laryngeal nerve injury.

- Hypoparathyroidism:

  - Both complications are considered permanent if they persist for more than 6 months.

  - Hypoparathyroidism is usually temporary, resulting from impaired glandular blood supply, and permanent cases are rare.

Hypoparathyroidism:

- Most common cause: Thyroid surgery, especially total thyroidectomy.

- Hypoparathyroidism can also develop after parathyroid surgery, such as the removal of a parathyroid adenoma or after subtotal parathyroidectomy or total parathyroidectomy with forearm autotransplantation in patients with hyperplasia.

- Distinguishing hypoparathyroidism from "hungry bone syndrome":

  - Hungry bone syndrome: Occurs after parathyroidectomy when calcium shifts back into the bones, leading to hypocalcemia.

  - Commonly seen in patients with kidney disease and severe bone disease, where calcium is deposited in the bones after surgery.

- **Laboratory findings in hypoparathyroidism:**

  - Low serum calcium levels.

  - High phosphorus levels.

  - Low PTH levels.

- **Pseudohypoparathyroidism:** Characterized by resistance to PTH in target organs.

  -Lab findings: Low calcium.High phosphorus.High PTH levels.

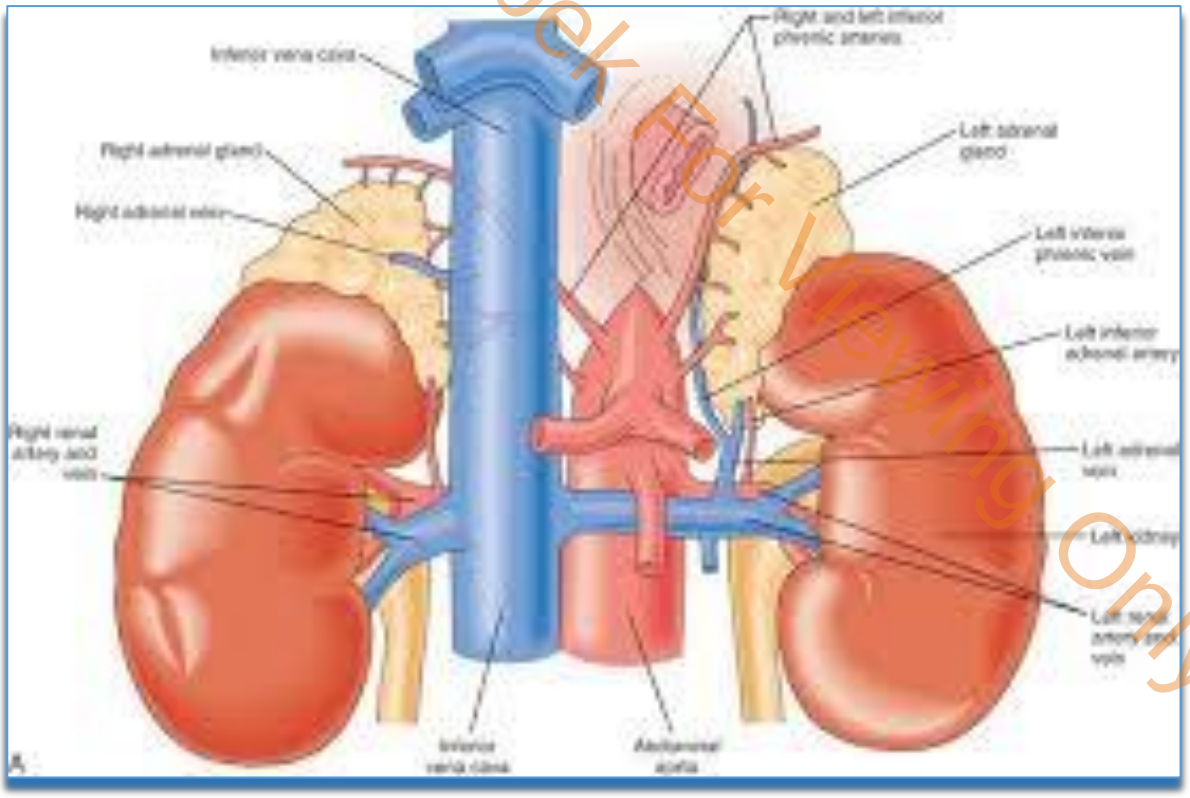
  - **Hungry bone syndrome:** Both calcium and phosphorus levels are low, but PTH levels are normal.

- DiGeorge syndrome: A congenital condition where the parathyroid glands may be absent.

Chapter
14.

ADRENAL GLAND DISORDERS AND SURGERY

ANATOMY OF THE ADRENAL GLANDS



- Adrenal Cortex:

- Blood Supply:

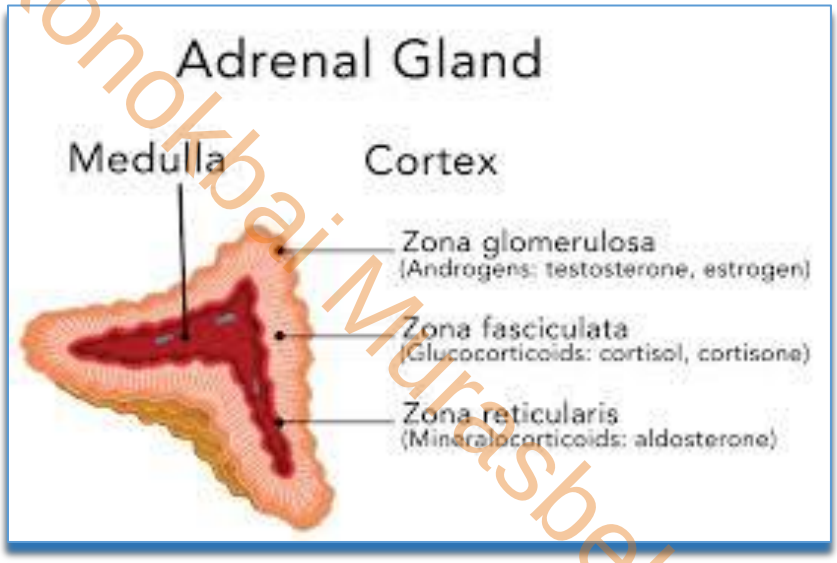
- Right and left inferior phrenic arteries.

- Inferior vena cava.

- Aorta.

- Structure:

- The adrenal cortex is divided into three layers:



- Zona Glomerulosa: The outermost layer, responsible for aldosterone secretion.

- Zona Fasciculata: The middle layer, responsible for cortisol secretion.

- Zona Reticularis: The innermost layer, responsible for androgen secretion.

- The zona fasciculata makes up 75% of the cortex, while the zona glomerulosa accounts for 15%.

- The medulla, located at the center of the adrenal gland, is composed of cells that secrete catecholamines.

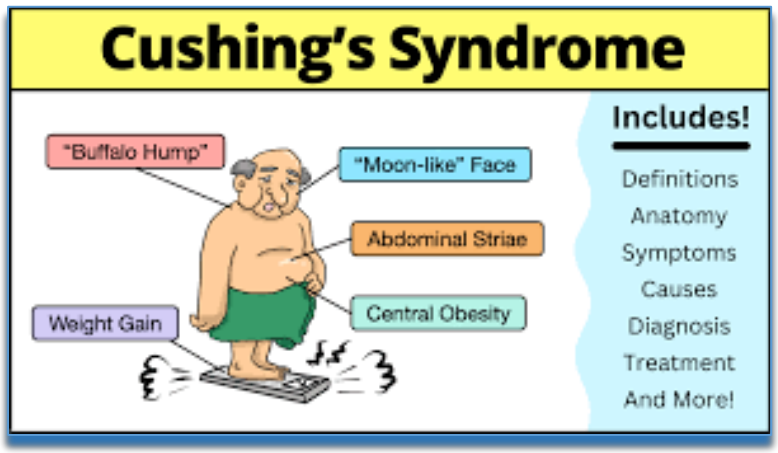
- The medullary cells are chromaffin-positive.

ADRENAL CORTEX DISORDERS

CUSHING SYNDROME

- **Etiology of Cushing Syndrome:**

- <u>ACTH-Dependent Causes:</u>



- Pituitary adenoma or Cushing's disease (70%).

- Ectopic ACTH production (10%).

- Ectopic CRH production (<1%).

- <u>ACTH-Independent Causes</u>:

- Adrenal adenoma (10-15%).

- Adrenal carcinoma (5-10%).

- Primary adrenal hyperplasia - pigmented micronodular cortical hyperplasia or gastric inhibitory peptide-sensitive macronodular hyperplasia (5%).

- Other Causes:

- Pseudo-Cushing's syndrome

- Due to iatrogenic exogenous steroid administration.

- **Clinical Features:**

- Cushing syndrome affects multiple systems.

- The most common symptom is progressive truncal obesity.

- Cushing Syndrome Signs and Symptoms by System:

- General:

- Truncal obesity (centripetal obesity, 95%).

- Buffalo hump.

- Supraclavicular fat deposition.

- Opportunistic infections.

- Skin:

- Hirsutism.

- Plethora, purple striae.

- Acne.

- Ecchymoses.

- Neuropsychiatric:

- Emotional instability.

- Psychosis.

- Depression.

- Cardio vascular system:

- hypertension

- **Metabolic:**

  - Diabetes, glucose intolerance.

  - Hyperlipidemia.

- **Renal:**

  - Polyuria.

  - Kidney stones.

- **Gonadal:**

  - Impotence.

  - Decreased libido.

  - Menstrual irregularities.

- **Diagnosis:DIAGNOSTIC STEPS**

- Confirm Hypercortisolism and identify the source:

- Screening Tests:

1. 24-Hour Urine Free Cortisol Measurement: The best screening test for diagnosing hypercortisolism (sensitivity 95-100%; specificity 98%).

2. Overnight Dexamethasone Suppression Test (DST): 1 mg dexamethasone is administered at 11:00 PM. Cortisol is measured at 8:00 AM. Normally, cortisol is suppressed (adults < 3 µg/dL). In hypercortisolism, suppression does not occur.

3. Late-Night Salivary Cortisol: An alternative test.

4. ACTH Levels: Helps in determining the cause of hypercortisolism.

- Primary adrenal tumor: Very low ACTH.

-Pituitary tumor: Moderately elevated ACTH.

- Ectopic ACTH-producing tumor: Very high ACTH.

-Imaging:

- Adrenal CT and MRI: Detect adrenal tumors with 95% sensitivity and help differentiate between adenomas and carcinomas.

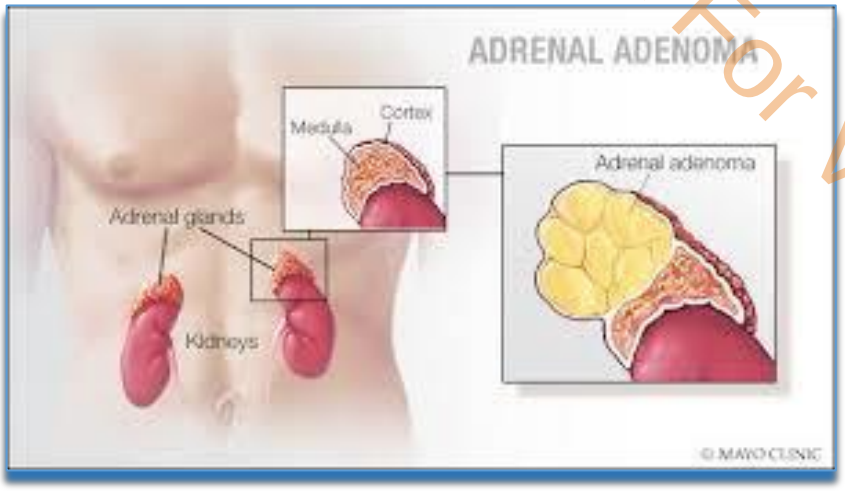
- **NP-59 Scintigraphy:** Useful for distinguishing between adrenal adenoma, hyperplasia, and carcinoma, and is the best method for diagnosing primary pigmented micronodular hyperplasia.

- **Pituitary Imaging:** For Cushing disease, the most definitive method to detect and localize a pituitary tumor is petrosal sinus sampling.

- Helps differentiate between an ectopic ACTH-producing tumor and a pituitary tumor.

- **CRH Stimulation**: If the petrosal sinus ACTH to peripheral plasma ACTH ratio is > 3 after CRH stimulation, it indicates Cushing disease with 100% sensitivity.

ADRENAL CORTEX ADENOMA



- **Definition:** A benign neoplasm of adrenal cortex cells.

- **Diagnosis:** Distinguishing adrenal adenomas from carcinomas based on cellular morphology alone can be challenging.

- **Effects**: While adenomas can cause syndromes associated with hypercortisolism and hyperaldosteronism, they rarely lead to adrenogenital syndrome.

- **Malignancy**: Malignant adrenal cortex tumors are typically large (>6 cm) and can cause adrenogenital syndrome.

ADRENAL CORTEX CARCINOMA

- **Definition:** A malignant neoplasm of adrenal cortex cells.

- **Associated Syndromes:** May be related to Li-Fraumeni syndrome, MEN 1 syndrome, Beckwith-Wiedemann syndrome, and Carney complex.

- **Functionality:** About 50% of adrenocortical cancers are hormonally inactive.

- **Hormone Secretion:** Among active tumors, cortisol is the most commonly secreted hormone (30%).

- **Clinical Presentation:** Functional tumors often present with rapidly onset Cushing syndrome and virilization signs.

- Diagnosis:

- Size: Tumor size is a key indicator of malignancy.

- Typically, lesions are larger than 6 cm.

- May contain areas of necrosis and hemorrhage.

- Invasion and metastasis are possible.

Treatment:

- Surgical Options:

- For Carcinoma or Adenoma: Unilateral total adrenalectomy.

- Preferred Approach for Adrenal Adenoma: Laparoscopic adrenalectomy.

- Open Adrenalectomy: Performed for larger tumors (~6 cm) or if there is suspicion of cancer.

- Additional Treatments:

- Mitotane: May be used for adrenocortical cancer as it is toxic to the adrenal cortex.

- Nelson Syndrome: Can develop in 20% of total adrenalectomy cases, characterized by a pituitary chromophobe adenoma.

ADRENAL HYPERPLASIA

- Definition: Increased number of cells in the adrenal glands, which can lead to hyperfunction.

- Congenital Adrenal Hyperplasia: The most common enzyme deficiency is 21-hydroxylase deficiency.

- Types of Hyperplasia:

- **Primary Adrenal Hyperplasia:** Can be micronodular, macronodular, or massive macronodular.

- **ACTH-Stimulated Hyperplasia:** Typically macronodular.

- Primary Pigmented Micronodular Adrenal Hyperplasia: Hyperplastic disease not associated with excessive ACTH secretion from the adrenal glands.

- **Treatment:** Patients with congenital adrenal hyperplasia are generally treated medically.

ECTOPIC ACTH SYNDROME

- Caused by tumors that produce ACTH outside the pituitary gland.

- **Common Tumors:**

  - Small cell lung cancer

  - Bronchial carcinoma

  - Thymic carcinoma

  - Pancreatic islet cell tumors

  - Medullary thyroid carcinoma

  - Pheochromocytoma

  - Gastrointestinal carcinoids

  - Ovarian adenocarcinomas

  - Pancreatic cystadenoma

  - Unknown origin adenocarcinomas

- **Treatment:**

  - Resection of the primary tumor if possible.

  - **For Inoperable Tumors:** Palliative medical adrenalectomy using metyrapone, aminoglutethimide, or mitotane, or surgical bilateral adrenalectomy.

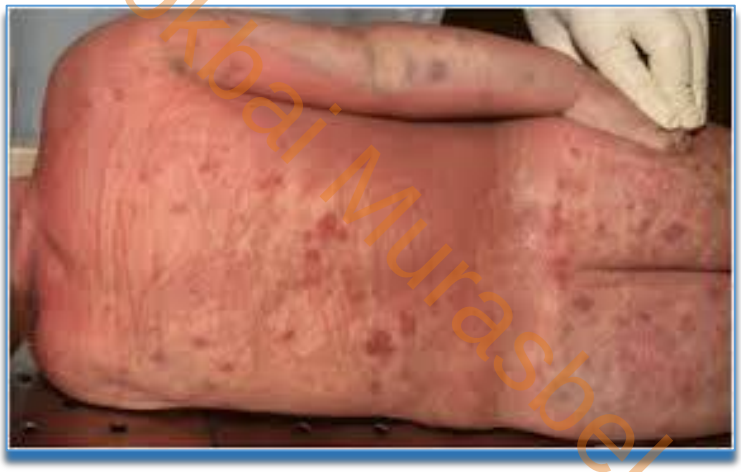
ADRENAL INSUFFICIENCY (ADDISON'S DISEASE)

- **Primary Adrenal Insufficiency:**

- **Cause:** Degeneration of the adrenal cortex, usually without involvement of the adrenal medulla.

- **Autoimmune Addison's Disease:** Can be associated with other endocrine autoimmune diseases (e.g., Schmidt's syndrome).

- **Secondary Adrenal Hemorrhage:** May occur due to sepsis, anticoagulant therapy, or coagulopathy.



- **Waterhouse-Friderichsen Syndrome**: Acute adrenal hemorrhage secondary to severe sepsis, commonly following meningococcal sepsis.

- **Secondary Adrenal Insufficiency:**

- **Cause:** Results from abnormalities in the hypothalamus or pituitary.

- **Common Cause:** Iatrogenic, due to prolonged glucocorticoid use.

- **Perioperative Risk:** If the hypothalamic-pituitary-adrenal axis does not respond appropriately to stress, it can lead to an adrenal crisis during the perioperative period.

Causes of Adrenal Insufficiency

Primary Adrenal Insufficiency:

- Autoimmune adrenalitis (most commonly, autoimmune polyglandular syndrome type 1 and 2)

- Tuberculosis

- Adrenomyeloneuropathy

- Fungal infections

- AIDS

- Metastatic carcinoma

- Familial isolated glucocorticoid deficiency

- Adrenal surgery

- Empty sella syndrome

- Hypothalamic tumors

Secondary Adrenal Insufficiency:

- Exogenous steroid use (most common)

- Pituitary disorders

- Pituitary tumor

- Craniopharyngioma

- Pituitary surgery

- Pituitary radiation therapy

- Sarcoidosis

- Histiocytosis

Clinical Presentation

- **Acute Adrenal Insufficiency:** Can mimic conditions like sepsis, myocardial infarction, or pulmonary embolism. Always be cautious in high-risk patients.

- **Symptoms:** Severe hypotension, fever, weakness, contusions, nausea, vomiting.

- **Pigmentation:** In primary adrenal insufficiency, increased CRH and ACTH secretion can lead to hyperpigmentation due to cross-reactivity with melanocyte-stimulating hormone.

Diagnosis

- **Laboratory Findings:** Characteristic findings include hyponatremia, hyperkalemia, hypoglycemia, eosinophilia, and mild azotemia.

- **Most Reliable Test:** The corticotropin (ACTH) stimulation test.

- **Procedure:** Administer 250 µg ACTH (cosyntropin) intravenously or intramuscularly before 10 AM. Measure plasma cortisol levels before administration, and at 30 and 60 minutes post-administration.

- **Normal Function:** If the basal plasma level is at least 20 µg/dL and shows an increase of more than 7 µg/dL at the 60-minute mark, adrenal function is considered normal.

Treatment

- **Acute Adrenal Crisis:** If suspected, administer intravenous hydrocortisone immediately.

- **Diagnostic Confirmation:** Check blood cortisol, glucose, sodium, potassium, blood urea nitrogen (BUN), and creatinine levels, and perform a complete blood count.

- **Initial Treatment:** Administer 200 mg of intravenous hydrocortisone while awaiting lab results.

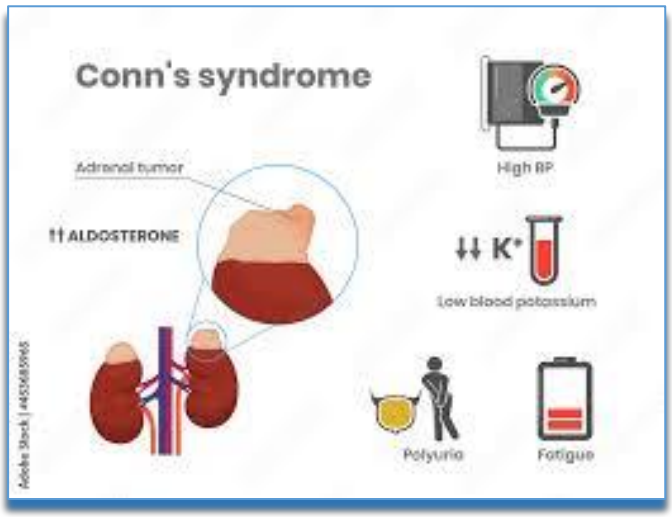
- **Correction of Hypoglycemia and Dehydration:** Provide dextrose and balanced salt solutions.

- **Dosage Adjustment:** Start with 100 mg hydrocortisone three times daily for two days, then taper off over the next few days with high-dose steroids.

- **Mineralocorticoid Addition**: For primary adrenal insufficiency, initiate mineralocorticoid therapy (e.g., fludrocortisone 0.1-0.2 mg/day) when the hydrocortisone maintenance dose is reached.

- **Surgical Considerations:** For patients with primary or secondary adrenal insufficiency undergoing surgery, glucocorticoid treatment should be administered based on the size of the surgery.

Primary Hyperaldosteronism (Conn's Syndrome)



- **Aldosteronoma:** The most common cause of primary hyperaldosteronism; a benign tumor that secretes aldosterone.

- **Idiopathic Adrenal Cortical Hyperplasia (IACH):** The second most common cause.

- **Adrenal Cortex Carcinoma:** Less common.

- Characteristics:

- Primary Hyperaldosteronism: Elevated plasma aldosterone levels with suppressed renin.

- Secondary Hyperaldosteronism: Elevated levels of both plasma renin and aldosterone, often due to conditions like renal artery stenosis, cirrhosis, or decreased renal perfusion.

Clinical Presentation

- **Hypertension:** Systolic hypertension may not always be present, but diastolic hypertension is common.

- **Symptoms:** Muscle weakness, muscle cramps, polyuria, polydipsia.

- **Edema:** Typically absent in primary hyperaldosteronism.

Diagnosis

- Laboratory Findings:

- **Hypokalemia:** Serum potassium < 3.9 mEq/L.

- **Renin:** Low in primary hyperaldosteronism.

- **Plasma Aldosterone to Renin Ratio:** Ratio > 25-30.

- **Urinary Potassium Excretion:** Elevated > 25-30 mEq/day.

- Imaging and Other Tests:

- **CT scan** ;(0.5 mm slices): Detects aldosteronoma with 90% sensitivity.

- **Tumor Characteristics:** A tumor 0.5 - 2 cm on one side with normal findings on the other side, combined with supportive biochemical results, is pathognomonic for aldosteronoma.

- **MRI:** Less sensitive but has higher specificity.

- **Venous Sampling for Aldosterone:** Sensitivity of 95%, specificity of 90, but is invasive.

- 131I-6-beta-Iodometyl-19-Nor-Iodocortisol Scintigraphy (NP-59 Scintigraphy): Detects aldosteronoma with very high accuracy but is not commonly used.

- Scintigraphy Findings:

- Aldosteronoma: Appears as a "hot" nodule.

- Bilateral Hyperplasia: Shows bilateral symmetric uptake.

Treatment for Primary Hyperaldosteronism (Conn's Syndrome)

- **Medical Management:** The best treatment involves medications such as spironolactone, nifedipine, and/or amiloride. Additional antihypertensive drugs may be used as needed.

- **Surgical Management:** Most aldosteronomas are small and benign, making them suitable for laparoscopic adrenalectomy.

Adrenal Incidentalomas

- Adrenal incidentalomas are found in 0.4% to 4% of abdominal CT scans performed for unrelated indication

- Most incidentalomas (36% to 94%) are benign, non-functioning adrenocortical adenomas.

- **Clinical Symptoms**: Investigate if there are symptoms such as hypertension, signs of Cushing's syndrome, virilization, feminization, or menstrual cycle changes.

- **Subclinical Cushing's Disease:** Perform a low-dose (1 mg) overnight dexamethasone suppression test and check for 17-ketosteroids if there is suspicion of excess sex steroids.

- **Pheochromocytoma:** Measure catecholamines, metanephrines, and vanillylmandelic acid (VMA) in 24-hour urine or plasma metanephrine levels.

- **Aldosteronoma:** For hypertensive patients, assess serum electrolytes, plasma aldosterone, and plasma renin levels.

- **Subclinical Cushing Suspected:** If there is strong suspicion of subclinical Cushing's (e.g., hypertension, obesity, diabetes), perform the following tests: Dexamethasone suppression test, salivary cortisol test and 24-hour urinary free cortisol

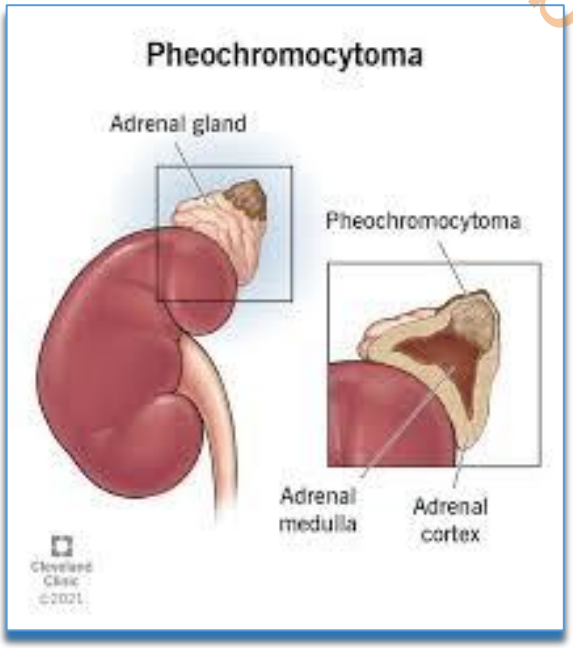
- **Androgens and Estrogens:** Screening for androgen and estrogen levels is only necessary if there are symptoms suggesting these disorders.

- **Fine Needle Aspiration Biopsy:** This can be used for evaluating adrenal masses in patients with a history of cancer and when biochemical tests have ruled out pheochromocytoma.

Chapter
15.

Adrenal Medulla Diseases and Surgery

Pheochromocytoma



- Pheochromocytoma originates from chromaffin cells.

- Chromaffin cells are widespread in the body during fetal life and are associated with sympathetic ganglia.

- After birth, most chromaffin cells degenerate, but those in the adrenal medulla persist. Therefore, 85-90% of pheochromocytomas arise from the adrenal medulla.

- However, they can occur anywhere there are sympathetic ganglia.

- Extra-adrenal pheochromocytomas can be located in the carotid body, heart, along the aorta, and in the bladder.

- The most common extra-adrenal location is the Zuckerkandl organ, located to the left of the aortic bifurcation where the inferior mesenteric artery branches from the aorta.

- Bilateral pheochromocytomas are often associated with familial syndromes such as MEN type IIA and IIB.

- Other hereditary syndromes associated with pheochromocytomas include Von Hippel-Lindau disease, neuroectodermal disorders (Sturge-Weber and tuberous sclerosis), and Carney syndrome.

- The 10% rule generally applies: 10% of pheochromocytomas are malignant, 10%bilateral, 10%extra-adrenal, or 10%occur in children.

Clinical

- Severe hypertension is seen due to excessive catecholamine secretion from the pheochromocytoma.

- The pheochromocytoma triad includes headaches, palpitations, and sweating (diaphoresis).

- Chronic hypovolemia and excessive alpha-adrenergic stimulation and vasoconstriction may cause lactic acidosis.

- Weight loss is common.

- Diabetes can develop due to excessive catecholamine secretion.

- Increased body temperature may be observed.

- Cardiovascular complications such as myocardial infarction or cerebrovascular events can occur.

- Sudden death can occur if a pheochromocytoma is operated on without being identified.

Diagnosis

- The diagnosis of pheochromocytoma is made by measuring catecholamines and their metabolites in 24-hour urine samples and, if necessary, determining plasma metanephrine levels.

- Urinary metanephrine levels are 98% sensitive and 98% specific. They are the most reliable and cornerstone tests for pheochromocytoma.

- Measuring plasma epinephrine and norepinephrine levels has lower sensitivity, generally less than 85%.

- Plasma chromogranin A levels can be checked but are not very sensitive.

- Plasma metanephrine levels have very high sensitivity (around 100%) but lower specificity (85%). It may not be suitable for every case.

Preoperative Preparation

- The foundation of preoperative preparation is alpha-adrenergic blockade.

- Patients are given 10 mg of phenoxybenzamine orally 2-3 times daily.

- If tachycardia is present, beta-adrenergic blockade (propranolol) is added.

- Beta-blockers should not be started before adequate alpha-adrenergic blockade and hydration, as they may worsen hypertension by causing vasoconstriction.

- Other alpha-blockers like prazosin, terazosin, and doxazosin provide selective alpha-blockade. Phenoxybenzamine may be preferred for patients needing long-term pharmacological treatment due to its lower side effect profile.

- Nicardipine is the most commonly used calcium channel blocker. When used as primary treatment, it is as effective as preoperative alpha- and beta-blockade.

- Alpha-methyltyrosine is another drug that can be used in preoperative preparation. It is a competitive inhibitor of tyrosine hydroxylase, the rate-limiting step in catecholamine synthesis.

Intraoperative Approach

- Despite preoperative preparation, blood pressure may rise when the tumor is manipulated.

- Hypertensive attacks can be controlled with alpha-adrenergic blockers like **phentolamine** (Regitine) or smooth muscle relaxants like **sodium nitroprusside**.

- Surgical treatment options include open or laparoscopic adrenalectomy.

Surgical Approach

- Laparoscopic adrenalectomy can be performed.

- Laparoscopic approach is preferred for tumors smaller than 6 cm and benign.

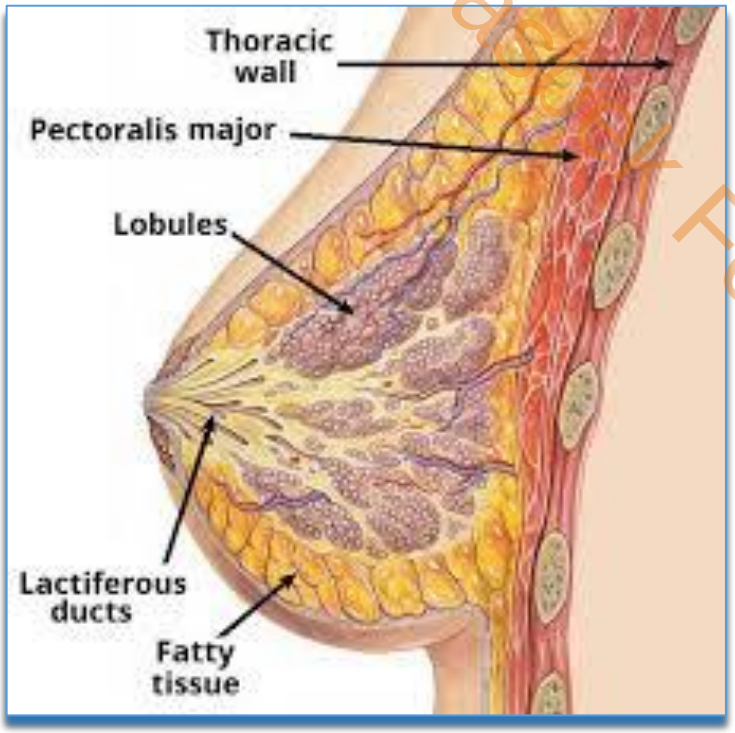
- For pheochromocytomas smaller than 5 cm, laparoscopic adrenalectomy can be safely performed.

- For malignant or large pheochromocytomas (>6 cm), open adrenalectomy is more appropriate.

Chapter
16.

BREAST DISEASES AND SURGERY

BREAST ANATOMY



- The breast is a tubuloalveolar gland composed of 15-20 lobes (segments) arranged in a radial manner starting from the nipple.

- Each breast lobe consists of 20-40 lobules, and each lobule contains 10 to 100 acini.

- Cooper's ligaments (fibrous bands) provide structural support to the breast by attaching perpendicularly to the dermis.

BLOOD SUPPLY

- The central and inner parts of the breast are supplied by perforating branches of the internal mammary artery, while the upper outer part is supplied by the lateral thoracic artery.

- The veins accompany the arteries, and venous flow is directed toward the axilla.

- There are three main groups of veins that carry venous blood from the breast and chest wall:

1. Perforating branches of the internal thoracic vein.

2. Branches draining into the axillary vein.

3. Perforating branches of the posterior intercostal veins.

- The vertebrae are one of the most common sites of metastasis for breast cancer.

- There are anastomoses between the perivertebral venous plexus (Batson's plexus), which extends from the base of the skull to the sacrum, and the posterior 3rd, 4th, and 5th intercostal veins. There are no valves within these channels, allowing breast carcinoma to metastasize to the vertebrae, skull, pelvic bones, and central nervous system without passing through the lungs.

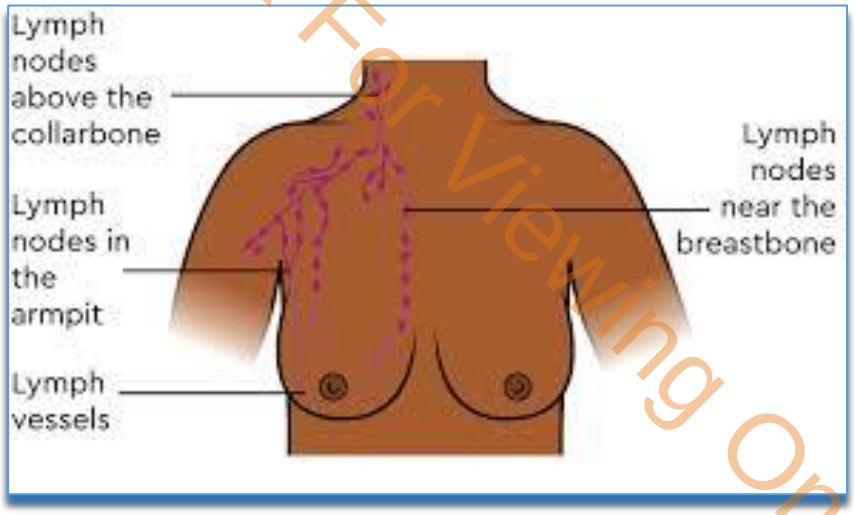
LYMPHATICS

- 75% of the breast's lymphatic drainage is directed to the axilla, while 25% drains into the internal mammary lymph nodes.

- Axillary lymph nodes constitute the most important drainage area for lymphatic flow from the breast.

- These lymph nodes are divided into 6 groups:

1. External mammary
2. Scapular
3. Axillary vein
4. Central
5. Interpectoral (Rotter)
6. Subclavicular



- To determine the extent of metastatic spread, axillary lymph nodes are divided into three levels based on their location relative to the pectoralis minor muscle:

- **Level I:** Those lateral to or below the lower edge of the pectoralis minor.

- **Level II:** Those behind the pectoralis minor.

- **Level III:** Those medial to the upper edge of the pectoralis minor.

- The internal mammary lymph nodes (parasternal) are located in front of the pleura, around the internal thoracic vessels.

MEDICAL INVESTIGATION OF BREAST DISEASES

HISTORY

- Complaint

- Menstrual cycles, menopausal status

- History of lactation and pregnancy

- Hormonal therapy

- Family history of breast disease

PHYSICAL EXAMINATION

- The most appropriate time for a breast examination is on the 5th to 7th day after menstruation.

IMAGING

- Mammography and ultrasonography (USG) are generally performed routinely (mammography for those over 40 years old).

- Magnetic resonance imaging (MRI) and positron emission tomography (PET) are performed with special indications.

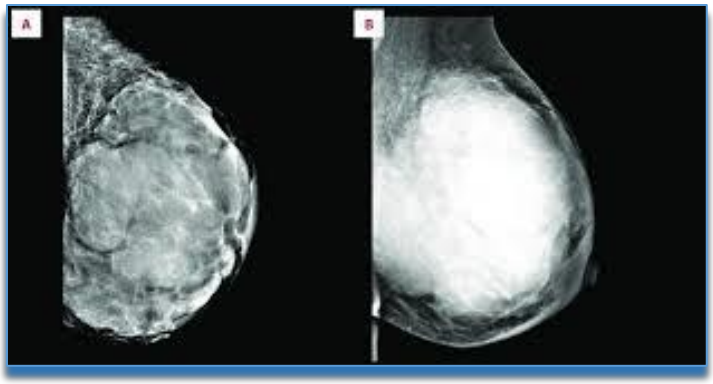
- Ultrasonography is important for distinguishing whether lesions are cystic or solid.

MAMMOGRAPHY

- Although the sensitivity of mammography is high, its specificity is low.

- The American Cancer Society recommends annual mammography screening starting at age 40.

Cancer Findings on Mammography



- Lesion with spiculated extensions

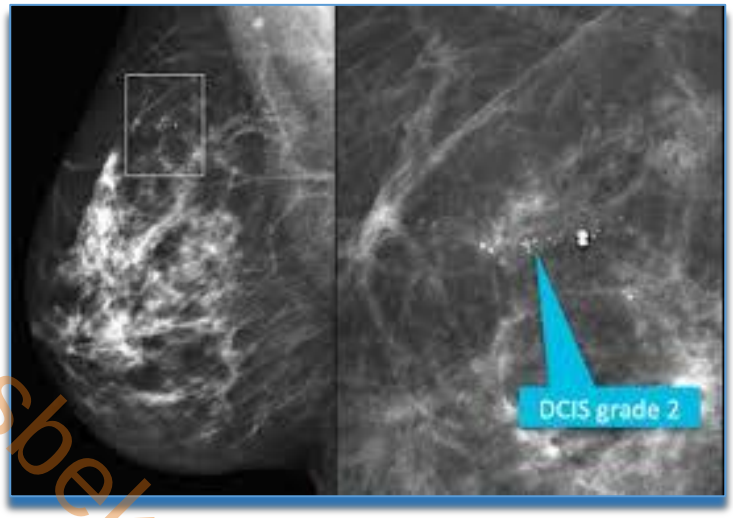
- Scattered microcalcifications (especially if the number is around 5 and pleomorphic)

- Asymmetric density

- Ductal asymmetry

- Distortion of normal breast structure

Microcalcifications Associated with Cancer



- Varying shapes and densities

- < 0.5-1 mm

- Thin and linear

Breast Imaging Reporting and Data System (BI-RADS)

0. Incomplete assessment

1. Negative (routine mammography)

2. Benign findings (routine mammography)

3. Probably benign: Short-interval follow-up

4. Suspicious lesion for malignancy, biopsy needed

5. Lesion is most likely malignant

6. Pathologically proven malignancy

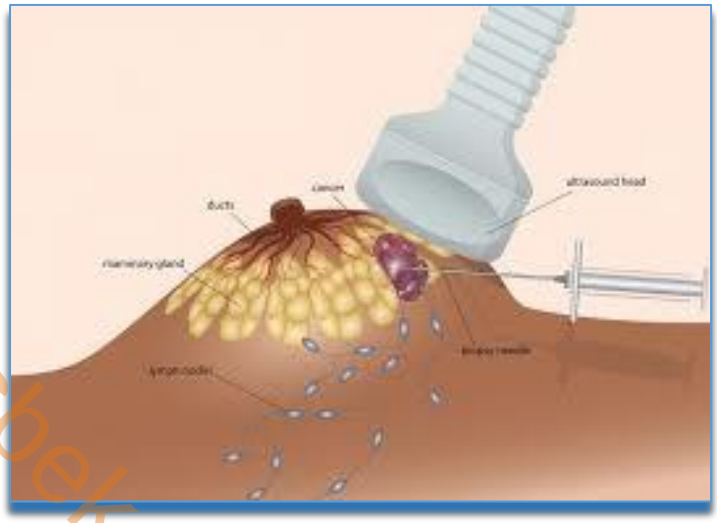
MAGNETIC RESONANCE IMAGING (MRI)

- Its sensitivity is extremely high.

- Specificity is moderate.

- It can lead to unnecessary biopsies.

BREAST BIOPSIES



- Fine needle biopsy

- Core needle (core biopsy, Tru-Cut) biopsy

- Today, core needle biopsy is considered the ideal biopsy method for breast lesions because it is less invasive than incisional and excisional biopsies and allows tissue examination.

Nonpalpable Lesions

- Biopsy is performed under imaging guidance.

- If there is a mass, it is done under USG guidance.

- If there is no mass, stereotactic mammography (for microcalcification clusters or structural distortion) is more appropriate.

- Wire localization under imaging (mammography) + excisional biopsy

- ROLL (radioguided occult lesion localization + excisional biopsy)

- Core needle biopsy under imaging (mammography) guidance (core needle biopsy)

BREAST DISEASES

Patients with breast diseases typically present with three main complaints:

a) Breast pain

b) Nipple discharge

c) Breast lump

MASTALGIA (MASTODYNIA)

- Breast pain is often cyclic.

- Pain rarely occurs with a lump.

- Cyclic mastodynia is usually most severe just before the menstrual period.

- Nonsteroidal anti-inflammatory drugs are generally sufficient.

NIPPLE DISCHARGE

- The likelihood of nipple discharge being due to breast cancer is low.

- The most common cause of spontaneous nipple discharge is intraductal papilloma.

- The most common cause of bloody nipple discharge is intraductal papilloma.

- Discharge from an intraductal papilloma can be bloody or clear like water.

Characteristics of Nipple Discharge That May Be Related to Cancer:

- Spontaneous

- Unilateral

- Localized to a single duct

- Occurs in women around 40 years old

- Bloody

- Accompanied by a lump

- In such cases, mammography and USG are indicated.

- Ductography can also be helpful.

BREAST LUMP

- Breast lumps can be classified into two categories: benign and malignant lumps.

- The most common benign pathologies include fibroadenoma, fibrocystic disease, sclerosing adenosis, radial scar, fat necrosis, galactocele, ductal ectasia, and intraductal papilloma.

BENIGN BREAST DISEASES

Classification of Benign Breast Diseases

1.Non-proliferative breast lesions (70%)

- Cysts and apocrine metaplasia

- Ductal ectasia

- Mild ductal epithelial hyperplasia

- Calcifications

- Fibroadenoma and related lesions

2.Proliferative breast lesions without atypia

- Sclerosing adenosis

- Radial scar and complex sclerosing lesions

- Ductal epithelial hyperplasia

- Intraductal papillomas

3.Atypical proliferative lesions

- Atypical lobular hyperplasia

- Atypical ductal hyperplasia

70% of benign breast lesions are non-proliferative types and do not carry a risk for the development of breast cancer.

FIBROADENOMA

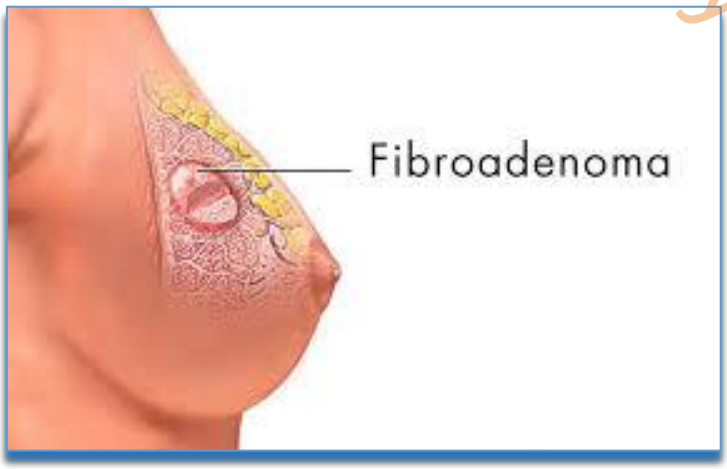
- It is the most common breast tumor in adolescents and young women.

- It is the most common benign tumor of the breast.

- Contains both epithelial and stromal elements.

- Fibroadenomas usually grow up to 1 or 2 cm in diameter and then stabilize.

- They are well-circumscribed, mobile, painless masses.



- They do not have a characteristic mammographic appearance.

- The diagnosis can be more clearly made with ultrasonography.

- Excision is not necessary.

- In young women (< 25 years old) with a characteristic fibroadenoma on USG, a needle biopsy is not required; follow-up is sufficient.

- If the fibroadenoma is > 3 cm, excision is appropriate.

- Other treatment options: cryoablation, vacuum-assisted biopsy.

FIBROCYSTIC DISEASE (FIBROCYSTIC CHANGES)

- It is a nonspecific term.

- It is often used as a diagnostic term to describe associated symptoms, rationalize the need for breast biopsy, or explain biopsy results.

- Other terms synonymous with fibrocystic changes include cystic mastopathy, chronic cystic disease, chronic cystic mastitis, Schimmelbusch disease, mazoplasia, Cooper disease, Reclus disease, and fibroadenomatosis.

- Pathologically, macro or microcysts, fibrosis, adenosis, and lymphocytic infiltration are observed.

- Generally, treatment is not required.

- If there is only a macrocyst, it can be aspirated or excised.

- There is no increased risk of cancer development.

CYSTS

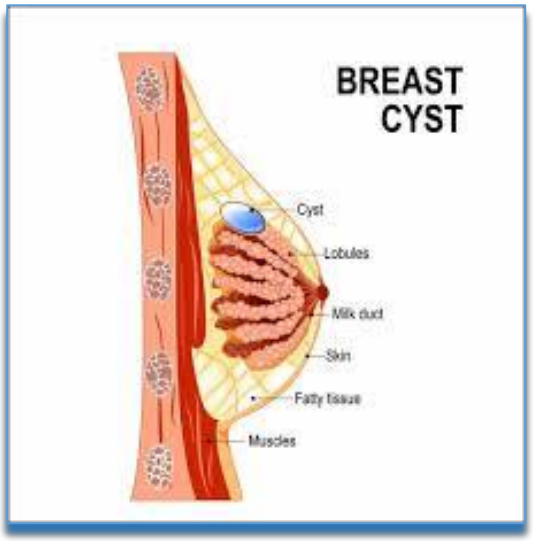
- Occur in 1 in 4 women.

- 50% of breast cysts are multiple or recurrent.

- Breast cysts are usually found in women over 35 years old.

- The incidence increases until menopause.

- It drops sharply after menopause.



- Ovarian hormones affect cysts, leading to cyclic mastalgia.

- Intracystic carcinoma is extremely rare (0.1%).

- If a palpable mass is suspected to be a cyst, aspiration and/or USG is performed.

- If the cyst disappears after aspiration and the cyst fluid is not bloody, cytology is not required.

- If a cyst recurs more than twice, pneumocystography (to determine whether there is a solid component) is recommended.

- If there is a solid component, a core needle biopsy is performed.

- Excision of the breast cyst is generally not necessary.

- If it recurs multiple times or atypia is found in needle biopsy, excision is planned.

FAT NECROSIS

- Its mammographic appearance is similar to breast cancer.

- It can develop after trauma, breast surgery, or radiotherapy.

- Calcifications are a characteristic finding of fat necrosis and may also be seen on USG.

- Pathologically, macrophages containing fat and large foreign body cells are characteristic.

- It has no malignant potential.

DUCTAL ECTASIA

- Presents with palpable dilated subareolar ducts and thick nipple discharge.

- Stagnation of secretions and local inflammation play a role in the pathogenesis of the disease.

- Nipple inversion may develop.

- Curative treatment is subareolar duct excision.

GALACTOCELE

- It is a cyst filled with milk.

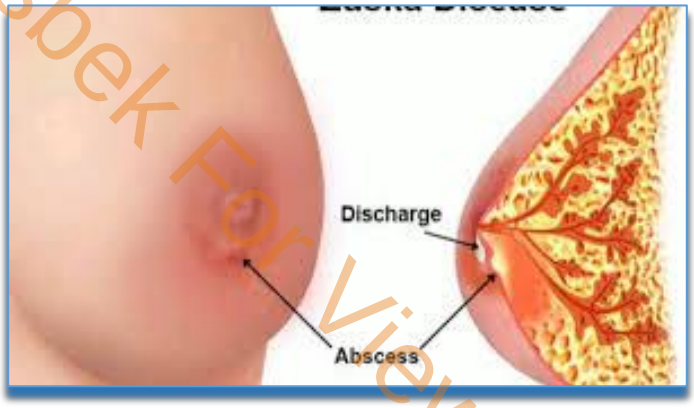
- It is more commonly seen in mothers who are not breastfeeding.

- Mammography reveals fat-fluid levels.

- Aspiration is sufficient for treatment.

ZUSKA DISEASE

- Also called recurrent periductal mastitis.



- Characterized by recurrent retroareolar infections and abscesses.

- Treatment:

  - Antibiotics, incision, and drainage if necessary.

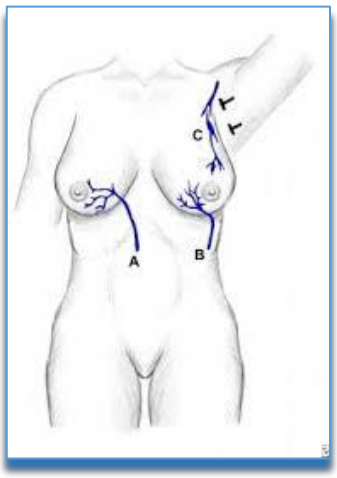
  - In rare cases, excision of chronically infected areas along with the nipple and areola may be required.

- Smoking is an important risk factor for Zuska disease.

- Another risk factor is diabetes.

MONDOR'S DISEASE

- Thrombophlebitis of the veins in the breast and thoracic wall.



- It often affects the lateral thoracic vein, thoracoepigastric vein, and less commonly the superficial epigastric vein.

- It is a benign, self-limiting disease.

- It is not neoplastic.

- The most important symptom is acute onset pain, typically located in the outer half of the breast or anterior chest wall.

- Treatment:

- Anti-inflammatory drugs

- Warm compresses

- Resolves in 4-6 weeks

- If it does not improve, the painful vein is excised.

INTRADUCTAL PAPILLOMA

- The most common cause of bloody nipple discharge.

- True polyps originating from the epithelium of the breast ducts.

- Mostly located near the areola and smaller than 1 cm.

- It does not increase the risk of breast cancer.

- Diagnosis: Ultrasonography, ductography, or MR lactography.

- Treatment: Subareolar duct excision.

SCLEROSING ADENOSIS

- Adenosis refers to an increase in the number of small terminal ductules and acini. It is usually associated with stromal tissue proliferation.

- When these two conditions coexist, the lesion is called sclerosing adenosis.

- Clinically, radiologically, and macroscopically, it can be confused with breast cancer.

- It presents as a poorly defined, rubbery nodularity.

- It is more common in women of childbearing age and those in the perimenopausal period.

- It has no malignant potential.

- In many series, sclerosing adenosis is the most common pathological diagnosis found under clusters of microcalcifications.

RADIAL SCAR AND COMPLEX SCLEROSING LESIONS

- Radial scar and complex sclerosing lesions are characterized by central sclerosis, epithelial proliferation, apocrine metaplasia, and papilloma formation.

- If the lesion is < 1 cm, it is called a radial scar.

- If the lesion is > 1 cm, it is referred to as a complex sclerosing lesion.

- These lesions can be confused with breast carcinoma.

  - They present as firm masses.

  - Show central sclerosis and radiating calcifications.

  - May cause dimpling of the breast skin.

- They are differentiated by core needle biopsy.

- Sometimes excision is required.

PHYLLODES TUMORS (CYSTOSARCOMA PHYLLODES)

- Classified as benign, borderline, or malignant.

- Benign phyllodes tumors are histologically difficult to distinguish from large fibroadenomas.

- Malignant phyllodes tumors are also difficult to differentiate histologically. Mammographic calcifications and macroscopic necrosis are not distinguishing features.

- Clinically, they present as well-circumscribed, mobile, firm, smooth-surfaced masses.

- They can be of any size but typically reach 4-5 cm and are more commonly seen in slightly older women than those with fibroadenomas.

- Benign phyllodes tumors are treated with local excision, similar to fibroadenomas.

- Borderline tumors (with suspicious malignancy features) are treated by re-excision with a 1 cm clear margin around the cavity.

- For clearly malignant phyllodes tumors, if a clear surgical margin cannot be achieved, a total (simple) mastectomy is performed.

- Mastectomy is also the appropriate treatment for large phyllodes tumors.

- Axillary metastasis is rare (around 0.9%), so axillary dissection is generally not necessary.

- Indications for radiotherapy after excision of malignant phyllodes:

  - Close surgical margin

  - Fascia or chest wall invasion

  - Tumor > 5 cm

INFLAMMATORY DISEASES

- Nonspecific breast infections mostly occur during lactation.

- Lactational mastitis, when it forms an abscess, is called a lactational abscess.

- The most common causative agent is *S. aureus*.

- Treatment includes antibiotics and, if there is an abscess, drainage as well.

- Breastfeeding is recommended even with a mastitic breast.

GRANULOMATOUS MASTITIS

- Granulomatous mastitis is a rare chronic inflammatory disease of the breast.

- Clinically and radiologically, it can mimic breast cancer.

- Specific etiological causes include tuberculosis, Corynebacterium, sarcoidosis, various fungal and parasitic infections, but the disease is often idiopathic.

- Since breast cancer is the primary consideration in the preoperative differential diagnosis, biopsy is usually the first approach.

- In patients who are found not to have cancer after biopsy, no additional treatment is often given, leading to frequent recurrences.

- There is no universally accepted treatment modality for idiopathic granulomatous mastitis.

- The role of steroid therapy in preventing recurrences is debated, but successful outcomes have been reported. In patients who cannot be medically controlled or present with severe clinical symptoms from the outset (such as recurrent abscesses or fistulas), surgery is unavoidable.

MALIGNANT BREAST DISEASES

RISK FACTORS FOR BREAST CANCER

Epidemiological Risk Factors

- Age: Being over 30 years old.

- Gender: Male to female ratio = 1:130.

- **Suspected Genetic Carrier:** Being a suspected carrier of a gene associated with breast cancer.

- **Strong Family History:** Two or more first-degree relatives with breast cancer (bilateral or premenopausal).

- Atypical Ductal or Lobular Hyperplasia or Lobular Carcinoma in Situ.

- Ductal Carcinoma in Situ (DCIS).

- Family History of Breast Cancer.

- **Menstrual History:** Menarche before age 12.Menopause after age 55.

- **Pregnancy History:** Never having been pregnant.First pregnancy after age 30.

- History of Breast Cancer in the Other Breast.

- Other Cancers that Increase Breast Cancer Risk: Ovarian, colon, and endometrial cancers.

- Diet: High-fat and high-calorie diets.

- Hormone Replacement Therapy (HRT): It is believed that 5 years of HRT with estrogen-only medications does not increase risk.

- Increased Risk with Family History: The risk of breast cancer increases in individuals with first-degree relatives who have breast cancer, particularly if the breast cancer in these relatives appeared premenopausally and bilaterally.

- **Risk of Cancer in the Other Breast:** A woman with cancer in one breast has a 1% annual risk of developing cancer in the other breast.

- Hereditary Breast Cancer Syndrome:

- For those with a family history, the risk of breast cancer increases 2-3 times.

- Women with first-degree relatives who have premenopausal and bilateral breast cancer have a 50% chance of developing breast cancer themselves.

Breast Cancer Classification

1.Non-invasive Epithelial Cancers

- Lobular Carcinoma in Situ (LCIS)

- Ductal Carcinoma in Situ (DCIS) or Intraductal Carcinoma (including Comedo, Non-comedo, and Mixed types)

2.Invasive Epithelial Cancers

- Paget's Disease

- Invasive Ductal Carcinoma (simplex, NST, or scirrhous, about 80%)

- Invasive Lobular Carcinoma (about 10%)

- Medullary Carcinoma (about 4%)

- Mucinous (Colloid) Carcinoma (about 2%)

- Tubular Carcinoma (about 2%)

- Papillary Carcinoma (about 2%)

- Rare Cancers (including Adenoid Cystic, Squamous Cell, Apocrine)

3.Mixed Connective (Stromal) and Epithelial Tumors

- Phyllodes Tumor (Benign, Malignant)

- Carcinosarcoma

- Angiosarcoma

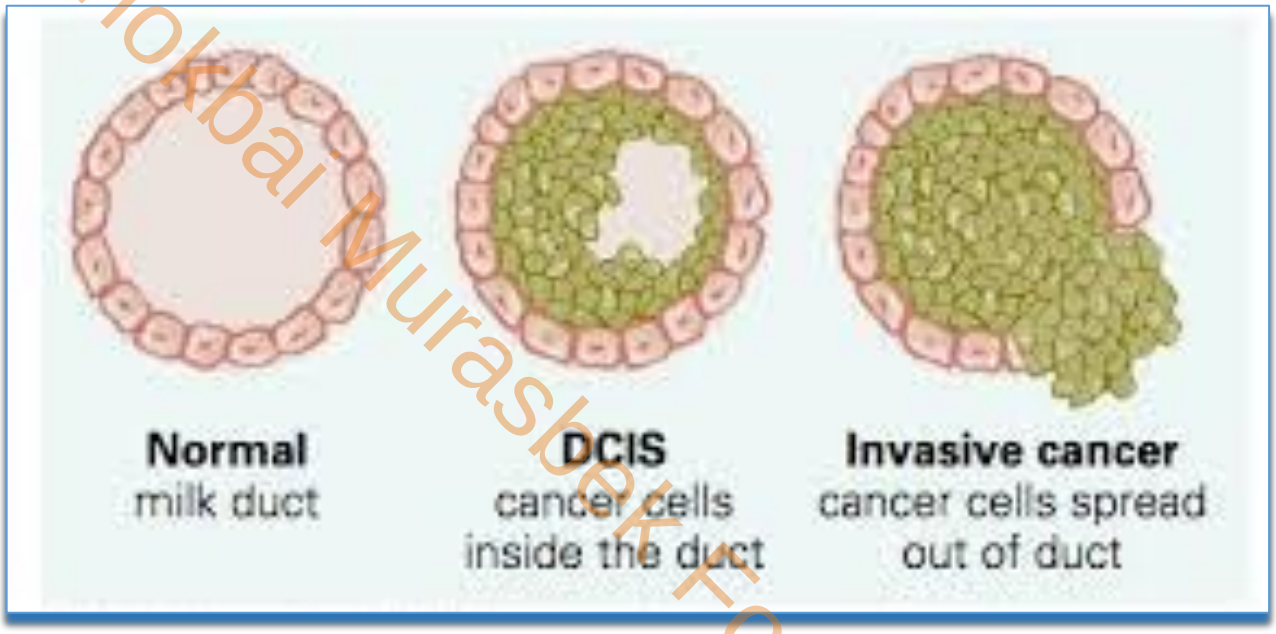
1.Non-invasive Breast Cancers

- These cancers do not penetrate the basal membrane.

- They do not metastasize because they cannot enter lymphatic and vascular structures.

- They can originate from ductal or lobular tissues.

Ductal Carcinoma in Situ (DCIS)



- Also referred to as intraductal carcinoma.

- The presence of clustered microcalcifications on mammography can be indicative of DCIS.

- DCIS constitutes about 60% of non-palpable breast cancers.

- The main pathological feature of DCIS is that cancer cells remain within the ducts and do not invade beyond the basal membrane.

- Subtypes include:

- Comedo

- High nuclear grade

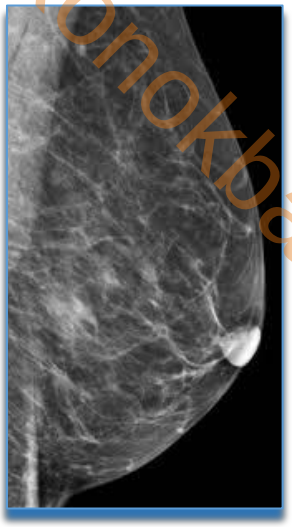
- Multicentricity

- Microinvasion

- Aggressive biological behavior

- **Non-comedo (including cribriform, solid, and papillary types)**

- If multicentric or if widespread microcalcifications are seen on mammography, mastectomy is performed.



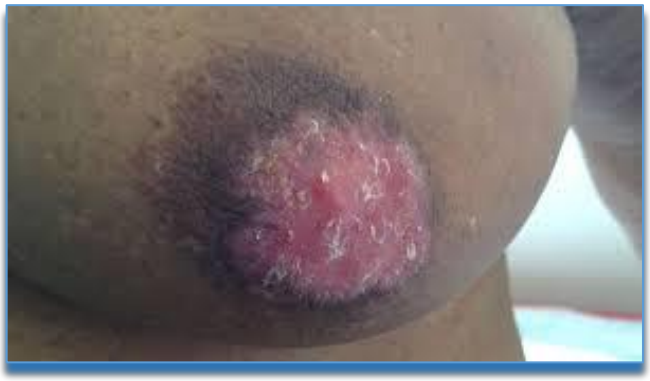
Lobular Carcinoma in Situ (LCIS)

- Occurs only in women.

- Average age of diagnosis is between 44-47 years.

- Usually found incidentally.

Paget's Disease



- An eczema-like lesion of the nipple.

- Often associated with edema and inflammation.

- Biopsy may reveal malignant cells in the milk ducts.

- Nipple and areola show large cytoplasmic, pale vacuolated cells (Paget cells) between epithelial layers.

- May be confused with superficially spreading melanoma.

- Typically associated with central DCIS or invasive ductal carcinoma.

- If invasive carcinoma is present, modified radical mastectomy is performed.

- Alternative options include excision of the nipple-areola complex, axillary staging, and radiotherapy.

2. Invasive Breast Cancers

- These cancers have penetrated the basal membrane.

Infiltrative Ductal Carcinoma (scirrhous, simplex, non-special type: NST)

- Accounts for 80% of invasive breast cancers.

- The most common type of breast cancer.

- Most likely to spread to axillary lymph nodes and has the worst prognosis among ductal cancers.

Invasive Lobular Carcinoma

- Accounts for 10% of invasive breast cancers.

- Histopathology:

- Small cells with round nuclei.

- Characteristic features include inconspicuous nucleoli.

- Sparse cytoplasm typical of lobular carcinoma cells.

- Mucin (signet-ring cells) present in the cytoplasm.

- Clinical presentation is variable; can present as non-palpable tumors or as large masses filling the breast.

- Microcalcifications are not seen in lobular carcinomas, making radiological diagnosis difficult.

- High likelihood of being multicentric and bilateral.

- Moderately poor prognosis.

- Estrogen Receptor (ER) positivity rate > 90%.

Medullary Carcinoma

- Accounts for 4% of invasive breast cancers.

- Highest rate of hormone receptor negativity among breast cancer types.

- A common phenotype of BRCA1 hereditary breast cancer.

- Grossly presents as soft, hemorrhagic masses.

- Typically large and deep-seated upon examination.

- Bilaterality occurs in approximately 20% of cases.

- Can rapidly increase in size due to necrosis and hemorrhage.

- Microscopic Features:

- Dense lymphoreticular infiltration.

- Large pleomorphic nuclei.

- Poorly differentiated.

- Active mitosis.

- Better prognosis compared to NST and lobular types.

- High rate of triple-negative status (according to some pathologists).

Mucinous Carcinoma (Colloid Carcinoma)

- Accounts for 2% of invasive breast cancers.

- More common in older individuals.

- Pathologically characterized by extracellular mucin surrounding tumor cells.

- Axillary lymph node metastasis rate is about 33%.

Papillary Carcinoma

- Accounts for less than 2% of invasive breast cancers.

- Typically seen in the 7th decade of life.

- Usually small lesions (< 2-3 cm).

- Low rate of axillary lymph node metastasis.

- A slow-progressing disease with survival rates similar to tubular carcinoma.

Tubular Carcinoma

- Accounts for 2% of invasive breast cancers.

- Axillary lymph node metastasis rate is about 10%.

- Typically detected in the perimenopausal and early menopausal period.

- Has the highest survival rate among invasive breast cancers.

Clinical Features of Breast Cancer

- The first clinical sign of breast cancer is usually a mass.

- Most commonly develops in the upper outer quadrant of the breast.

- Subsequently, it may occur in the central region, upper inner quadrant, and lower quadrants.

- Pain is rare and may develop later in the disease.

- Other findings suggestive of breast cancer include skin edema, dimpling, crusting of the nipple, nipple retraction, and nipple discharge.

Spread of Breast Cancer

- Cancer cells primarily spread to axillary lymph nodes through lymphatic channels.

- Metastasis to regional lymph nodes or blockage of lymphatic vessels can obstruct lymphatic drainage from the breast.

- Accumulation of lymphatic fluid in the breast skin can lead to a peau d'orange (orange peel) appearance.

Permeation Spread:

- Refers to cancer cells adhering to the wall of lymphatic vessels and moving against the flow.

- Results in satellite skin nodules and inflammatory breast cancer.

Bone Metastases:

- Breast cancer frequently metastasizes to bones. Common sites include:

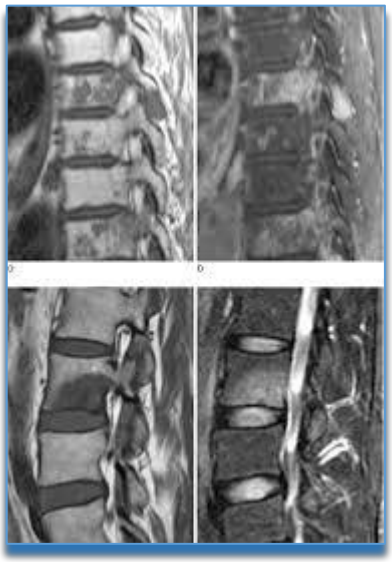
    - Vertebrae: especially lumbar vertebrae

    - Pelvic bones

    - Upper femur

    - Ribs

    - Skull bones



Explanation for Frequent Vertebral Metastases:

- The reason breast cancer often metastasizes to the vertebrae is due to the anastomoses between posterior intercostal veins (3rd, 4th, and 5th) and the Batson plexus (perivertebral venous plexus), which drains part of the breast's venous blood. This potential connection allows metastases to spread to the vertebrae, pelvis, skull bones, and central nervous system before entering the systemic circulation or causing pulmonary metastases.

Breast Cancer Pathological Staging (TNM)

- TX: Tumor cannot be assessed.

- T0: No evidence of a primary tumor.

- Tis: Carcinoma in situ or Paget's disease without a palpable tumor.

- T1: Tumor ≤ 2 cm

- T2: Tumor > 2 cm and ≤ 5 cm

- T3: Tumor > 5 cm

- T4: Tumor directly extending to the skin or chest wall

- NX: Regional lymph nodes cannot be assessed

- N0: No regional lymph node metastasis

- N1: Metastasis to 1-3 ipsilateral lymph nodes and/or internal mammary lymph nodes (with biopsy)

  - N1a: 1-3 axillary lymph nodes

  - N1b: Internal mammary lymph nodes positive on sentinel biopsy

  - N1c: N1a + N1b

- N2: Metastasis to 4-9 ipsilateral axillary lymph nodes or clinically positive internal mammary lymph nodes, with no axillary lymph node involvement

  - N2a: 4-9 axillary lymph nodes

  - N2b: Clinically positive internal mammary lymph nodes, with no axillary lymph node involvement

- N3: Metastasis to approximately 10 axillary lymph nodes or axillary lymph nodes plus internal mammary lymph nodes

- N3a: Approximately 10 axillary lymph nodes or infraclavicular lymph nodes

- N3b: Positive internal mammary lymph nodes plus approximately 1 axillary lymph node or > 3 axillary lymph nodes plus internal mammary lymph nodes positive on biopsy

- N3c: Supraclavicular lymph nodes on the same side

- M0: No distant metastasis

- M1: Distant metastasis (including contralateral metastasis, supraclavicular lymph node metastasis)

Breast Cancer Treatment

- Surgical Treatment: The goal of surgical treatment is to achieve local control of the tumor and to determine the stage.

- For early-stage breast cancer (Stage I-II), the current treatment options are either mastectomy or breast-conserving surgery, along with determining the status of the axillary lymph nodes.

- Both treatment methods are considered equivalent for Stage I-II breast cancer patients.

- Clinical detection of axillary lymphadenopathy or metastasis in the sentinel axillary lymph node requires axillary lymph node dissection.

Breast-Conserving Surgery (BCS)

- Can be applied to all Stage I and II breast cancers.

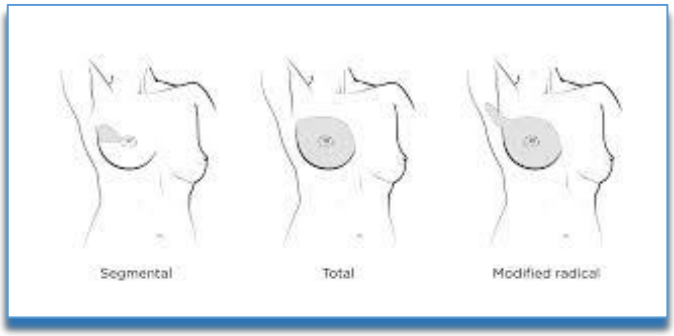
- Components of BCS include lumpectomy + determining the status of the axillary lymph nodes + radiotherapy.

- Radiotherapy is mandatory to prevent local recurrence.

Simple Mastectomy

- Involves the removal of only the breast tissue.

- Not frequently used for breast cancer.



Modified Radical Mastectomy (MRM)

- Involves the removal of the entire glandular tissue of the breast, most of the breast skin (including the nipple-areola complex), the pectoralis major fascia, and the axillary lymph nodes en bloc (as a single piece).

Radical Mastectomy

- Involves the removal of the entire glandular tissue of the breast, all of the breast skin, the pectoralis major muscle, and the axillary lymph nodes (Levels I, II, and III).

Mastectomy Complications

Early Complications:

- Seroma: The most common complication.

- Pneumothorax

- Infection

- Necrosis of skin flaps

Late Complications:

- Lymphedema: Develops in approximately 20% of patients after modified radical mastectomy; with added radiotherapy, the rate can reach 50-60%.

- Postmastectomy Pain Syndrome

Adjuvant Hormone Therapy and Chemotherapy

- Surgical Treatment Goal: To achieve local and regional control.

- **Hormone Therapy and Chemotherapy Goal:** To control occult metastases, reduce recurrence, and increase survival.

Hormone Therapy

- Tumors that are estrogen receptor (ER) positive respond better to hormone therapy.

- Tamoxifen: A selective estrogen receptor modulator. A weak estrogen agonist. The first choice for ER+ and progesterone receptor (PR) positive premenopausal and postmenopausal breast cancer.

- New selective aromatase inhibitors (letrozole, anastrozole, exemestane) can be used in the adjuvant treatment of postmenopausal breast cancer.

- LHRH agonists (goserelin or leuprolide) can also be added to hormonal therapy to suppress ovarian function and reduce estrogen levels in premenopausal patients.

Chemotherapy

- Post-surgical chemotherapy aims to eliminate clinically undetectable distant deposits of the disease using cytotoxic drugs.

- In HER-2-positive breast cancer, trastuzumab is added to chemotherapy combinations.

Post-Surgical Radiotherapy

- Radiotherapy should be added to breast-conserving surgery for all patients.

- Post-mastectomy radiotherapy is indicated for:

  - Metastatic involvement of approximately 4 lymph nodes

  - Premenopausal patients with metastasis in 1 or more lymph nodes, T3-T4 tumors

  - Extracapsular spread of lymph nodes

Breast Cancer in Pregnancy and Lactation

- Breast cancer occurs in 1 in 3,000 pregnant women.

- 75% of these patients have axillary lymph node metastasis.

- The average age is 34.

- Less than 25% of breast masses that arise during pregnancy and lactation are malignant.

- Diagnosis is made with ultrasound and needle biopsy.

- Mammography is not indicated due to increased breast density, but can be performed if necessary with fetal shielding.

- Modified radical mastectomy can be performed in the first and second trimesters, although anesthesia in the first trimester may increase the risk of miscarriage.

- In the third trimester, if radiotherapy can be deferred until after delivery, lumpectomy and axillary dissection can be performed.

- Lactation is suppressed.

- Chemotherapy is not administered in the first trimester. There is no evidence suggesting teratogenic effects of chemotherapy in the second and third trimesters.

- Therefore, many clinicians prefer to administer neoadjuvant chemotherapy during the second and third trimesters and postpone local surgical treatment until after delivery.

- Breast cancer in pregnancy is often diagnosed at an advanced stage, leading to a poorer prognosis. However, when stage-matched, the prognosis is similar to that of non-pregnant patients.

Gynecomastia

- Refers to the enlargement of breast tissue in men.

- Physiological gynecomastia most commonly occurs at three stages of life:

1. Neonatal Period: Caused by placental estrogen effects.

2. Adolescent Period (usually unilateral): Resulting from a relative excess of estrogen compared to testosterone.

3. Later Life (usually bilateral): Due to a decline in plasma testosterone levels with a relative increase in estrogen.

- Estrogen increase can result from testicular or non-testicular tumors.

- Endocrine disorders such as hyperthyroidism or hypothyroidism, as well as liver diseases, can lead to excess estrogen. Estrogen excess may also be related to nutritional changes such as protein and fat deficiencies.

- Conditions that cause androgen decline, such as aging, primary or secondary testicular insufficiency, and kidney failure, can lead to gynecomastia.

- Medications including digitalis, estrogens, anabolic steroids, marijuana, and other estrogenic drugs or drugs affecting estrogen activity can cause gynecomastia. Drugs like cimetidine, ketoconazole, phenytoin, spironolactone, antineoplastic agents, diazepam, as well as reserpine, theophylline, verapamil, tricyclic antidepressants, and furosemide can cause gynecomastia through various mechanisms including inhibition of testosterone synthesis or effects, increased estrogen synthesis by hCG, or through unknown mechanisms.

- In drug-induced gynecomastia, discontinuation of the medication is preferred if possible. In cases of gynecomastia due to androgen deficiency, testosterone therapy may be given. Danazol and tamoxifen can also be used as medical treatments. Subcutaneous mastectomy is the most effective treatment option for cases unresponsive to medical treatment.

- Gynecomastia does not increase the risk of breast cancer.

- However, primary testicular insufficiency in Klinefelter Syndrome increases the risk of breast cancer.

Pathophysiological Mechanisms of Gynecomastia

1. Conditions with Increased Estrogen

- Gonadal Origin: True hermaphroditism.

- Testicular gonadal stromal neoplasms: - Leydig cell tumors - Sertoli cell tumors - Granulosa-theca cell tumors

- Germ cell tumors: Choriocarcinoma.Seminoma, teratoma.Embryonal carcinoma

- Non-Testicular Tumors: Skin nevi. Adrenal cortical neoplasms. Lung cancers.Hepatocellular carcinoma

- Endocrine Disorders

- Liver Diseases (Cirrhosis)

- Conditions Related to Nutritional Changes

2. Conditions with Decreased Androgen

- Age-Related (Senile)

- Hypogonadism:

- Primary Testicular Insufficiency: Klinefelter Syndrome (XXY),Reifenstein Syndrome (XY),Rosewater, Gwinup, Hamwi familial gynecomastia (XY), Kallmann Syndrome, Kennedy Disease related to gynecomastia, Congenital anorchia, Congenital defects in androgen biosynthesis,ACTH deficiency

- Secondary Testicular Insufficiency: Trauma. Orchitis. Radiation. Hydrocele. Varicocele.Spermatoceles

- Kidney Failure

3. Drug-Related Gynecomastia

4. Systemic Diseases:Non-neoplastic lung diseases. Trauma to the chest wall.Central nervous system-related causes such as anxiety and stress. AIDS

Male Breast Cancer

- Less than 1% of all breast cancer cases occur in men.

- Male breast cancers are often diagnosed at more advanced stages compared to women.

- Risk factors for male breast cancer include Klinefelter syndrome, testicular feminization syndrome, estrogen therapy, radiation, cirrhosis, and obesity.

- The risk is increased in men with BRCA-2 gene mutations.

- Since men do not have lobular structures in their breasts, male breast cancers are usually of ductal origin.

- Infiltrative lobular carcinoma is rarely encountered.

- About 80% of male breast cancers are estrogen receptor positive.

- The prognosis is similar to that of female breast cancers at the same stage.

Chapter
17.

Burn Management

- Burns are the most severe form of trauma.

- Nowadays, the treatment of burn patients is a specialized area that combines multiple disciplines, primarily general surgery, and extends to rehabilitation teams during the recovery phase. It involves a team approach that includes various specialties.

The emergency evaluation of a burn patient includes four critical points:

1- Airway management (ensuring it remains open)

2- Identification of additional injuries

3- Calculation of the burn area

4- Confirmation of carbon monoxide and cyanide poisoning

Airway Management

- The first and most crucial step is maintaining an open airway.

- Signs suggesting inhalation injury include:

- Burns around the mouth and nasal hair

- Hoarse voice, wheezing, stridor, or patient-reported respiratory distress

(There is a high likelihood of developing respiratory failure, requiring elective intubation.)

- Direct thermal damage to the airways due to smoke inhalation can lead to fatal airway edema (emergency intubation is required).

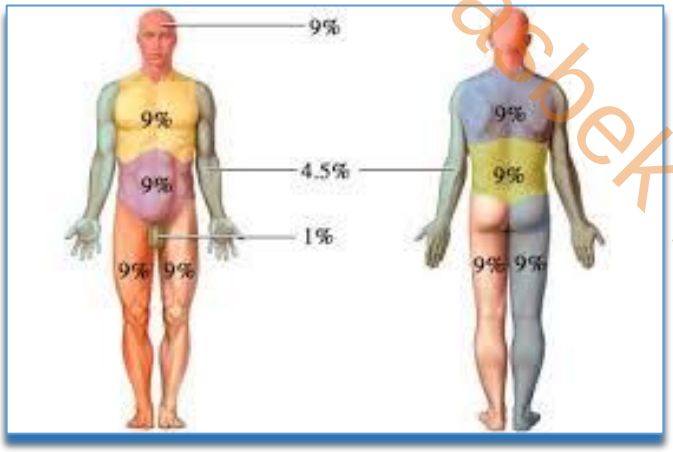
- Simultaneously with the initial examination, a thick-lumen peripheral intravenous catheter is placed, and fluid resuscitation is initiated.

- If the burn area is >40%: Two thick-lumen peripheral intravenous catheters are ideal.

- Prophylactic antibiotics are never used in burn patients.

- Local wound care is performed. If an infection develops, therapeutic antibiotics are administered.

- Tetanus prophylaxis should be administered in the emergency room.



Calculating the Burn Surface Area

- When calculating the total burn surface area, superficial (first-degree) burns are not included.

- In patients with electrical burns, the situation is different due to the development of myoglobinuria.

- Free myoglobin is toxic to kidney tubules, leading to acute tubular necrosis and anuria.

- To facilitate the rapid elimination of this toxic pigment, intravenous hydration is increased to achieve a urine output of 100-150 ml/hour.

Carbon Monoxide and Cyanide Poisoning Diagnosis

- The most important factor increasing mortality in the presence of smoke inhalation is associated carbon monoxide intoxication.

- The most reliable method for diagnosing carbon monoxide intoxication is carboxyhemoglobin levels.

- The gold standard treatment is 100% oxygen. This approach reduces the half-life of carbon monoxide fivefold.

- Hyperbaric oxygen therapy is also beneficial as a supportive treatment.

Cyanide poisoning

- Cyanide poisoning may also accompany smoke inhalation injuries.

- The presence of resistant lactic acidosis or ST elevation on ECG supports the diagnosis.

- Treatment includes sodium thiosulfate, hydroxocobalamin, and 100% oxygen.

Indications for Transfer to a Burn Center

- Second-degree burns covering more than 10% of the total body surface area

- Third-degree burns in any age group

- Burns involving critical areas such as the face, hands, joints, or genital organs (regardless of extent)

- Electrical burns, including lightning strikes

- Inhalation injuries

- Chemical burns

- Burns in patients with preexisting medical conditions that complicate treatment, delay healing, or increase morbidity/mortality

- Burn patients with accompanying trauma (e.g., fractures). If the trauma presents a more urgent situation, the patient should be stabilized before transfer to a burn center.

- Children with burns in hospitals that lack adequate staff and equipment for pediatric care

- Burns requiring special social, psychological, and/or long-term rehabilitation support

TYPES OF BURNS

- Burns are categorized into three main groups: thermal, electrical, and chemical burns.

1.Thermal Burns

- Flame

- Contact

- Scalding (from hot liquids)

Flame Burns

- These are the most common type of burns.

- They have the highest mortality rate.

2.Chemical Burns

- These are rare but potentially severe burns.

- Initial treatment involves removing the toxic substance and washing the area for at least 30 minutes.

- An exception to this is strong alkaline substances in powder form (such as sodium hydroxide). When washed with water, it can activate aluminum hydroxide. Therefore, it should be removed by wiping.

Hydrofluoric Acid Burns

- Due to its widespread use in industry, hydrofluoric acid burns are a common type of chemical burn.

- They cause hypocalcemia.

- Treatment, depending on severity, involves topical or intravenous calcium gluconate.

- The effectiveness of treatment in hydrofluoric acid burns is indicated by a reduction in pain.

Formic Acid Burns

- These are rare.

- Hemolysis and hemoglobinuria develop.

- Metabolic acidosis, kidney failure, intravascular hemolysis, and ARDS (Acute Respiratory Distress Syndrome) can occur.

- Treatment involves sodium bicarbonate.

- If a large amount of formic acid is absorbed, hemodialysis may be required.

- Diuresis can be induced with mannitol.

- Formic acid wounds typically appear greenish and are often deeper than expected.

- The best treatment is surgical excision.

BURN DEPTH

- The depth of a burn is the fundamental determinant of the long-term physical and functional outcomes of the burn patient.

- Burns are dynamic events within the first 48-72 hours.

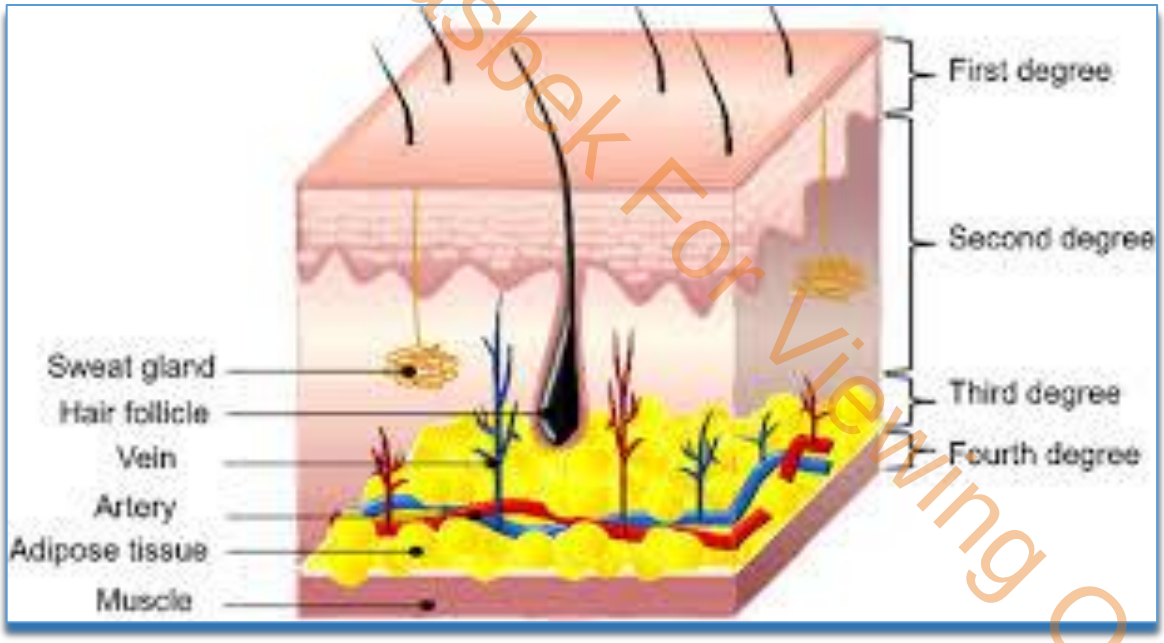
- A burn that initially appears superficial may become deep by the third day.

- Despite all new diagnostic methods, the methods used to determine burn depth cannot replace serial examinations by experienced burn surgeons.

- The depth of the burn depends on the amount of heat applied, the duration of contact, and the thickness of the skin.

BURN DEPTH CLASSIFICATION

Burns are classified into four degrees based on their depth:



1. First-Degree Burns (Superficial)

- Involve only the epidermis.

- They are reversible.

- There is hyperemia (increased blood flow).

2. Second-Degree Burns (Partial Thickness)

- Involve the epidermis and part of the dermis.

Superficial Second-Degree Dermal Burns

- Involve the epidermis and the upper layers of the dermis.

- Blisters filled with plasma and vitamin A-rich fluid form between the epidermis and dermis.

- Blisters may appear immediately after the burn or 12-24 hours later.

- Re-epithelialization occurs in 1-2 weeks (if no infection develops).

- There is no functional loss.

- Minor color changes may occur. Hypertrophic scars are rare.

Deep Second-Degree Dermal Burns

- Extend into the reticular dermis.

- They appear paler and mottled.

- They do not blanch when pressed.

- Blisters may develop.

- Re-epithelialization occurs in 2-5 weeks (thanks to keratinocytes from hair follicles and sweat glands).

- Significant scarring usually occurs.

3. Third-Degree Burns (Full Thickness)

- The skin is completely burned through.

- The elasticity of the skin is lost.

- They are painless.

- The skin takes on a brown or white color, and the blood vessels are thrombosed.

- These burns typically develop eschar.

- **Eschar** is completely burned skin that has lost its elasticity and vitality but retains its structural integrity.

- Escharotomy may be required.

- Healing is attempted through epithelialization and contraction at the wound edges.

- However, full healing is often not achieved. Contractures (permanent tightening of muscles, tendons, skin, and nearby tissues) may develop.

- Skin grafts are often necessary.

4. Fourth-Degree Burns

- Involve complete burning of the skin and underlying structures.

- These burns are usually seen in electrical burns, contact burns, or in patients who lost consciousness during the burn event.

TREATMENT

Resuscitation in Burn Patients

- Various formulas are used for fluid management in burn patients.

- The most commonly used formula is the Parkland or Baxter formula.

- According to the Parkland formula, the amount of fluid required in the first 24 hours is:

- **Fluid to be given = 4ml x Body weight x Burn percentage** (in children: 6 x burn percentage x body weight)

- Half of the amount calculated by this formula is administered in the first eight hours, and the second half is given over the next sixteen hours.

- The fluid typically used in the first 24 hours is -**Ringer's lactate**.

- To reduce the amount of resuscitation fluid required in burn patients, colloid solutions, albumin, or hypertonic solutions have been tried.

- It has been shown that the use of albumin may not be beneficial and could even be harmful.

- Colloids may be beneficial in the later stages after capillary permeability has decreased.

- High doses of Vitamin C reduce the need for fluid replacement and respiratory distress.

- Additionally, plasmapheresis can reduce fluid requirements by filtering out inflammatory mediators.

- The most useful parameters for monitoring the adequacy of resuscitation are mean arterial pressure (MAP) and urine output.

- The desired urine output is 30 ml/hour in adults and 1-1.5 ml/kg/hour in children.

- The target mean arterial pressure (MAP) is 60 mmHg.

- The use of blood transfusions in burn patients has been reevaluated based on current findings.

- Blood transfusions are considered to be immunosuppressive.

- It has been observed that as the amount of transfusion increases, so do infection rates and mortality.

- In the absence of a specific indication, maintaining a hemoglobin level above 7 g/dl in burned children is sufficient for resuscitation and does not lead to negative outcomes.

- Recombinant human erythropoietin does not prevent anemia or reduce the number of transfusions required.

Inhalation Injury and Ventilation

- Approximately one-third of burn patients may have accompanying inhalation injuries.

- The presence of inhalation injury increases the risk of developing Acute Respiratory Distress Syndrome (ARDS) and mortality.

- In patients with burns covering more than 60% of their body surface area, who also have inhalation injury and ARDS, mortality is 100%.

- The most common physiological disruption caused by inhalation injury is an increased need for fluids during resuscitation.

- Initial treatment is primarily supportive, including:

  - Aggressive pulmonary hygiene

  - Nebulized medications such as: Bronchodilators, N-acetylcysteine,

  - Heparin

  - Bronchodilators like Albuterol

  - Hypertonic saline

  - Epinephrine

Indications for intubation in patients with inhalation injury include:

1. $PaO_2 < 60$ mmHg
2. $PaCO_2 > 50$ mmHg
3. $PaO_2 / FiO_2 < 200$
4. Respiratory and ventilatory dysfunction
5. Severe edema of the upper airways

TOPICAL ANTIMICROBIAL THERAPY

1.Silver nitrate

2.Silver Sulfadiazine(Low systemic absorption with minimal metabolic effects. Used for infection prophylaxis.Not used on newly grafted areas as it can damage skin grafts. It may also delay epithelial migration in partial-thickness burns).

3.Mafenide Acetate(Has limited use due to its painful application.

4.Bacitracin, neomycin, or polymyxin B can be used.

5.MRSA (Methicillin-resistant Staphylococcus aureus) infections are treated with **mupirocin ointments.

Surgery and Burn Wound Care

- Blisters in outpatients are usually kept intact.

- In hospitalized patients, blisters are punctured and debrided.

- Escharotomy is rarely necessary within the first 8 hours; it should not be performed until there is an indication due to the risk of poor aesthetic outcomes.

- Infection is the most important cause of mortality and morbidity in burn patients.

- Therefore, topical antimicrobial agents for wound care and wound closure can reduce the risk of infection.

- The choice of dressing varies according to the burn's severity:

  - First-degree burns do not require dressing.

  - Topical creams and systemic anti-inflammatory agents are used to reduce pain and keep the skin moist.

  - Superficial second-degree burns are treated with daily dressings using topical antibiotics, elastic gauze, and elastic bandages.

  - Alternatively, wounds can be covered with biological and synthetic dressings.

  - Deep second-degree and third-degree burns require excision and grafting.

- Currently, burns that will take longer than 3 weeks to heal should undergo early tangential excision and grafting.

- Early grafting offers many advantages:

  - It not only reduces mortality but also decreases the need for reconstructive surgery, hospital stay, and costs.

Burn Complications

- Pneumonia in critically ill patients and those on mechanical ventilation is a significant cause of mortality. Early tracheostomy may be necessary.

- Abdominal compartment syndrome

- Deep vein thrombosis:

  - Occurs in up to 25% of cases.

  - Pulmonary embolism can develop.

  - Heparin prophylaxis is recommended.

- Catheter infections

- Curling ulcers:

  - Stress ulcers found in the duodenum and stomach.

  - Etiology: Ischemia of the gastric mucosa is the most likely cause.

  - Can be prevented with oral feeding and neutralization of stomach acid.

  - Upper gastrointestinal bleeding occurs in 20% of patients.

- Cellular and humoral immunity is suppressed.

- Infection complications:

  - Invasive burn wound sepsis

  - Pneumonia: The respiratory system is often the infection site in burn patients.

- Marjolin ulcer:

  - Skin cancers that develop on old burn scars.

  - The most common is squamous cell carcinoma.

  - Basal cell carcinoma, fibrous histiocytoma, sarcoma, and neurotropic malignant melanoma can also occur.

  - These cancers tend to be aggressive.

  - Approximately 30% are found in the head and neck area.

Electrical Burns

- Although the skin manifestation of electrical burns may seem limited, there can be significant deep tissue damage.

- In electrical burns, skin damage is the tip of the iceberg.

- Electrical burns carry risks of cardiac arrhythmia, compartment syndrome, and concurrent rhabdomyolysis.

- A baseline ECG is recommended for all patients. Hospitalization may not be necessary for patients with a normal ECG and low-voltage injuries because compartment syndrome and rhabdomyolysis often occur in high-voltage injuries.

- Electrical currents travel primarily along more conductive tissues such as muscles, blood vessels, and nerves, leading to more severe damage in blood vessel-rich tissues.

- The risk of kidney failure due to myoglobin released from muscles in electrical burns is high. Therefore, the amount of fluid administered should be increased, and urine output should be maintained above 100-150 ml per hour.

- High-voltage electrical burns may lead to cardiopulmonary arrest or arrhythmias.

- Patients who experience cardiac arrest should be monitored for at least 48 hours.

- The nervous system is particularly sensitive to electricity, and permanent neurological symptoms may result in chronic pain syndromes known as post-traumatic stress disorder. Another complication of electrical burns is the development of cataracts, observed in 5-7% of cases. Cataracts typically develop bilaterally within 1 or 2 years after the burn.

- Because these complications can arise even in burns with very small surface areas, hospitalization is indicated regardless of the burn percentage.

- In electrical burn patients, there are two situations where early surgical treatment is indicated:

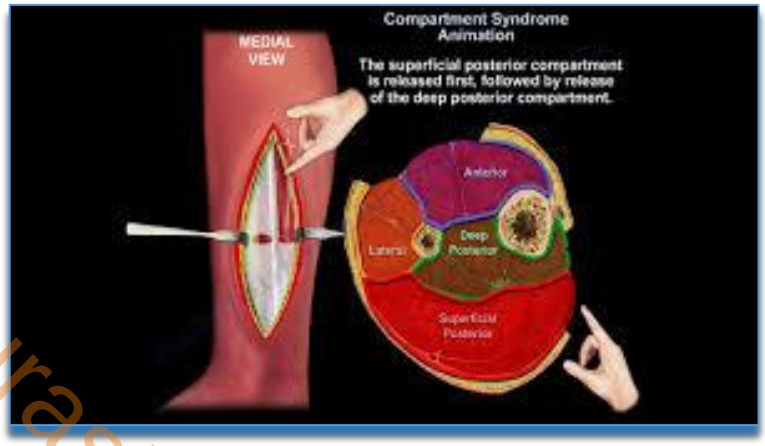
  - Massive deep tissue necrosis resulting in acidosis or myoglobinuria unresponsive to fluid therapy may require urgent major debridement and/or amputation.

  - Escharotomy and fasciotomy to prevent compartment syndrome, which is more commonly seen.

Signs of Compartment Syndrome:

- Increased pain

- Pallor



- Absence of pulse

- Temperature difference (proximal warm, distal cold; poikilothermia)

- Paresthesia

- Paralysis

- If emergency decompression or debridement is not necessary, surgical treatment can be performed between the 3rd and 5th days, before bacterial contamination occurs and the boundaries of necrosis become clear.

- Abdominal compartment pressure is monitored with a Foley catheter placed in the bladder.

- If the pressure exceeds 30 mmHg, a complete abdominal escharotomy is performed.

- If pressure remains high despite escharotomy, a decompression laparotomy should be performed.

Frostbite Burns

- Treatment for frostbite burns includes:

  - Rapid rewarming

  - Close monitoring

  - Elevation and splinting

  - Daily hydrotherapy

  - Sequential "limited" debridement as needed (even if necrosis is present)

  - Rewarming is done by wrapping the patient in warm blankets in a stable, warm environment or by placing the patient in a tub filled with water heated to 40-42°C.

- Movement of frostbitten extremities can increase damage, so it is beneficial to splint or wrap the extremities.

- Rubbing, massaging, or similar actions to rewarm are very harmful.

Master's Today Medical surgical disease

Focused Review of Surgical diseases (A guide for exam takers)

This essential study guide and workbook is designed for students aiming to enhance their understanding of surgical diseases. Perfect for aspiring medical professionals, it serves as an invaluable resource to deepen knowledge and effectively prepare for examinations in the field.

With this book, you will be able to:

- Grasp essential concepts in surgical diseases
- Learn and understand the fundamentals
- Strengthen your understanding through focused reviews.

ABOUT AUTHOR

Dr Konokbai Murasbek
Graduated from KSMA 1998
National Surgical Center -2000
Head of surgical department ADAM university