Pathology

● Sheet ○ Slide

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Esophageal Diseases

The Esophagus is a muscular tube that starts at the pharynx and ends at the stomach. It is about 25 cm long. It functions as a passage of food from the mouth to the stomach by peristalsis.

Peristalsis: waves of contraction and relaxation to deliver food from the mouth to the stomach.
It is important for food to move in one direction, so there has to be mechanisms, which are the sphincters, to prevent reflux of food from the stomach up to the esophagus, because acidity of the stomach will cause inflammation to the squamous epithelium of the esophagus.

There are two sphincters:

1-Upper esophageal sphincter (UES): between the pharynx and the esophagus, prevents regurgitation of food from the esophagus to the mouth.
2-Lower esophageal sphincter (LES): between the esophagus and the stomach, prevents regurgitation of the food from the stomach to the esophagus.
Histologically, the esophagus is composed of several layers:

1-Epithelial layer (stratified squamous)
2- Submucosa (supportive connective tissue)
3- Muscular layer (3 muscular layers, important for the control of the peristaltic movement)
Types of diseases that affect the esophagus:

*Narrowing and obstruction
*Inflammation
*Vascular diseases
*Tumors

Esophageal Obstruction:

Esophageal obstruction can be mechanical or functional.

Mechanical obstruction is due to developmental abnormality

(atresia, fistula, duplication → all of them are explained later), it can be caused by a tumor, large bolus of food. They are either congenital or acquired.

Functional obstruction is due to a muscle or neural problem, paralysis, or dystrophy (any condition that affects the muscle's motility).

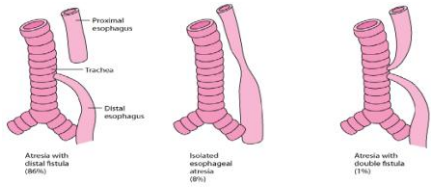
Mechanical Congenital Diseases:

1. Agenesis: no formation of esophagus. This is a very rare condition.
2. Atresia: (انسداد، رتق)

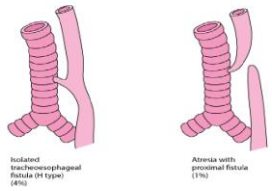
Absence or closure of a natural passage or channel in the body.
There are two types of atresia:

a. Either there is a thin non-canalized cord that replaces a segment of the esophagus (presence of stenosis in the esophagus).
b. The esophagus is formed as two <u>completely separated sacs</u>, called upper and lower pouches.

The most common site of atresia is at or near the tracheal bifurcation, no matter what the type of atresia it is.



In both types of atresia there could be a connection between the esophagus and the trachea. This connection is called a **fistula**. This fistula could be in the atretic esophagus, in the lower, or upper pouches (or in both. Any combination can occur)



0-13 min*

The most common type of atresia: complete separation of the esophagus with the lower pouch connected to the trachea by a fistula (86%).

So if a baby was born with atresia it will be detected shortly after birth, because every time the baby feeds there will be regurgitation, because the milk can't reach the stomach. The only treatment for these cases is surgical correction, the two pouches are connected, and if there was a fistula it will be removed.

- Complications of atresia:

1. Starvation
2. Severe electrolyte and fluids imbalance
3. Aspiration and suffocation
4. Pneumonia: because of the foreign bodies and fluids entering the lungs, macrophages will start reactive conditions to get rid of it, causing inflammation.

So this disease is dangerous and not compatible with life if not treated surgically.

Mechanical Acquired Diseases:

Stenosis (تضيق) is an acquired condition which leads to narrowing of the esophagus. It is caused by proliferation, sclerosis, or fibrosis in the submucosa (scar formation due to repair due to severe inflammation). it could also be associated with atrophy of muscularis propria and secondary epithelial damage.

(Inflammation → repair and fibrosis → stenosis)

*Tumors are not considered a cause of stenosis, because it is like there is something pushing the esophagus to close, while stenosis is concerned with problems that are changes within the walls of the esophagus.

Reflux is one of the major causes of inflammation leading to stenosis, in addition to irritation and caustic injury.

*Caustic injury is also called corrosive injury, it is a chemical injury to muco-cutaneous surfaces, with tissue destruction due to contact with a strong base or acid.

These patients have dysphagia, which is progressive. Difficulty in eating solids occurs long before problems with liquids.

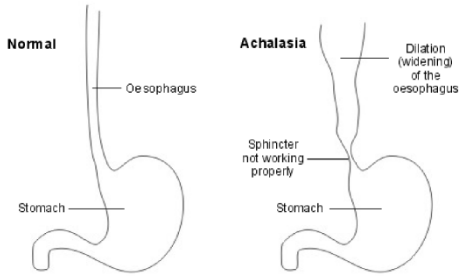
Functional Acquired Diseases:

In these diseases the passage is clear but there is a problem with the coordination of the peristalsis of muscles that helps the food descend to the stomach.

*Esophageal dysmotility interferes with the peristaltic contractions.
*Achalasia is the most important cause of functional obstruction.

- **Achalasia**

It is an acquired condition that causes aperistalsis (loss of peristalsis, thus the esophagus becomes dilated due to the presence of food that can't pass to the stomach due to LES contraction)



a→ means no, chalasia→ means relaxation.

(No relaxation in the LES).

The muscle tone increases, so there is no relaxation in the lower esophageal sphincter, and it becomes 'over-contracted'.

*13-21 min

It is caused by a problem in the neurons (ganglion cells to be exact). There will be no innervation of the lower esophageal sphincter muscles (which are normally contracted to prevent reflux of food, and is relaxed when the bolus of food reaches it). If there was no impulses it will stay contracted and there will be no relaxation (failure in the distal esophageal inhibitory neurons).

- Causes of achalasia:
  1. In most cases the cause of achalasia is idiopathic (primary) (unknown cause, but the suggested cause is autoimmune destruction)
  2. The second most common cause is *Trypanosoma cruzi* infections (secondary), which causes Chagas disease. *Trypanosoma cruzi* causes destruction of myenteric plexus neurons.
  3. Any damage to the neurons that supply the muscles of the esophagus, like: diabetic autonomic neuropathy, polio virus infection, and surgical ablation.
  4. Any disease that infiltrates through the wall of the esophagus and destroys it, such as: sarcoidosis, amyloidosis and malignancies.
  - Sarcoidosis: systemic disease, in this disease there are granulomas in several areas in the body, which are inflammatory processes in which there are accumulations of macrophages. If it affected the esophagus it could destroy the surrounding ganglion cells
  - Amyloidosis: infiltrative disease that is characterized by accumulation of abnormally folded proteins that will result in the destruction of the tissue.

Symptoms Of achalasia: regurgitation, dilation, difficulty in swallowing, sometimes chest pain, and increased risk of aspiration because of regurgitation.

*All esophageal diseases may cause chest pain if it had the element of muscle spasm. And it's chest pain can mimic heart diseases if it was really severe.

Treatment of achalasia: achalasia can't be treated by fixing the cause since neurons cannot be repaired, and in Chagas disease the parasite has already destroyed the neural plexus.

Achalasia is treated by making the muscles of the lower esophageal sphincter relax permanently, like putting a balloon-like object in the sphincter to keep it opened, the **Botulinum toxin** (Botox), which paralyzes the muscle, and Cutting the muscle (**myotomy**) are also

used to treat achalasia.

*These treatment methods will increase the risk of regurgitation, because the sphincter is dilated all the time.

*A student suggested that if they placed a device to give impulses to the muscle cells in order to relax (like they do with the pacemaker in the heart), and the doctor said that it is not used but it is a great idea to develop.

- Myotomy is the best option→ 90% success rate
- Botulinum toxin should be injected every 3 months because its effect isn't permanent.

*21-30 min

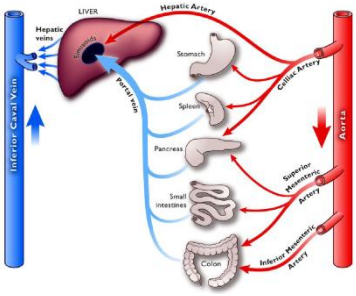
Diseases of Blood Vessels:

The most important disease is esophageal varices. Varicose veins can affect the esophagus, because of accumulation of blood inside veins.

- Blood can accumulate in the esophagus due to problems in the liver
- Valves have a secondary effect in esophageal varices.

So what does the liver have to do with the venous drainage of the esophagus?

Normal blood flow to the GI tract:



All of the blood in the GI tract is drained to the portal vein to the liver and doesn't go directly to the inferior vena cava. Why? Because the blood needs to be detoxified and cleaned from drugs and toxins and other materials that are absorbed in the GI tract to the blood before it reaches the systemic circulation.

There is a circulation for the GI tract called splanchnic circulation. all the venous drainage goes to the portal vein and then from the portal vein, following detoxification, to the inferior vena cava then to the heart.

So if there was any obstruction in the portal vein or any branch of the

portal vein inside the liver, portal hypertension will develop. If any of the portal vein branches in the liver was obstructed by a liver disease, blood will accumulate before it goes to the vena cava, and one of the organs that will be affected is the esophagus, because both the splanchnic and systemic circulations communicate with it (the esophageal veins make a communication between the splanchnic and the systemic circulations).

That is why when the portal hypertension increases, collateral vascular channels develop in the esophageal veins to allow blood to shunt from portal to caval system (inferior vena cava). These collateral veins (varices) enlarge and can rupture.

[This is the mechanism of esophageal varices]

Causes of esophageal varices: (the liver diseases that will result in portal hypertension and cause the varices)

1. The most common cause worldwide is liver cirrhosis, especially alcoholic liver disease.
   The normal Liver has a smooth surface. Excessive fibrosis will cause nodules to appear on the liver, so this fibrosis leads to liver cirrhosis, which affects the veins leading to esophageal varices.
2. The second most common cause is infection of the liver (hepatic schistosomiasis infection)

Morphology of the esophageal varices:

They appear as tortuous dilated veins within the submucosa of distal esophagus and proximal stomach. Varices are usually asymptomatic, but these dilated veins can rupture and cause bleeding.

- 50% of the cases where there is bleeding from varicose veins result in death, the other 50% develop a second bleeding because the dilated veins are still there.
- The most common cause of death of liver cirrhosis is varicose vein rupture.

Esophageal Inflammation (Esophagitis):

One type of esophageal inflammation is lacerations. Lacerations mean tears in the esophagus (this tear is called Mallory Weis tear), this tear can be caused by mechanical stretch (because of severe vomiting or with acute alcohol intoxication).

Normally the vomit is an anti-peristaltic wave, so the lower esophageal sphincter senses that there is an anti-peristaltic contractile wave, and as a reflex it dilates to allow the food to exit the stomach without causing damage to the esophagus, but if there was persistent vomiting this reflex fails and the sphincter stays closed. So when the food comes out of the stomach it will cause a tear in the esophagus and the patient is present with hematemesis (bloody vomiting).

These tears are longitudinal and they cross the gastroesophageal junction and is usually superficial and don't need any treatment. they heal by themselves, with no surgical intervention needed.

The End