Achalasia

Achalasia is a disorder of the esophagus characterized by incomplete lower esophageal sphincter (LES) relaxation, increased sphincter tone and uncoordinated peristalsis. Achalasia occurs because the ganglion cells in the myenteric (Auerbach) plexus become damaged. Patients present with dysphagia to both solids and liquids. Diagnosis is made by manometry or barium swallow study. Patients with achalasia have an increased risk of esophageal squamous cell carcinoma.



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Pathophysiology

Failure to Relax Lower Esophageal Sphincter

Pinching the Lower Sphincter of a Sarcophagus Sphinx

In achalasia, there is disordered motility with inability to relax the LES. Recall the LES is located at the junction of the distal esophagus and the stomach. The distal esophageal musculature is predominately comprised of smooth muscle. Since the LES is unable to relax, there is increased tone and contraction of the LES.

Malignancies

Malignant-man

Malignancy can cause pseudoachalasia by invading the esophageal neural plexus directly or by releasing humoral factors that disrupt esophageal function as part of a paraneoplastic syndrome.

Chagas Disease

Shotgun

The ganglion cells of the myenteric (Auerbach) plexus can become damaged by Trypanosoma cruzi, a flagellated protozoan that causes Chagas disease. Chagas disease can cause megaesophagus or abnormal dilation of the esophagus often accompanied by aperistalsis. Destruction of the autonomic nervous system innervation leads to loss of smooth muscle tone and subsequent gradual dilation.

Loss of Auerbach Plexus

Hourglass

Achalasia occurs because ganglion cells in the myenteric (Auerbach) plexus become damaged. The myenteric plexus is located between the inner circular and outer longitudinal muscle layers of the muscularis propria. The ganglion cells in the myenteric plexus are important for motility and relaxing the LES.

Symptoms

Dysphagia to Solids & Liquids

Dice-fajita with Solids and Liquids

Dysfunctional peristalsis causes dysphagia or difficulty swallowing with both solids and liquids. In contrast, esophageal obstructions such as masses or esophageal webs typically cause dysphagia to solids before liquids.

Diagnosis

Barium Swallow

Swallowing Berries

Barium swallow (esophagram) is commonly used to diagnose achalasia. Barium sulfate is given to a patient in an oral suspension to swallow; once swallowed, X-ray or CT images are taken and used to assess the function and appearance of the esophagus during the swallowing process.

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Bird's Beak Appearance

Bird Beak

On barium swallow, there is an acute tapering of the LES and narrowing at the junction between the esophagus and stomach, producing an image akin to a bird’s beak.

Manometry

Man-eating-tree

Esophageal manometry, also known as an esophageal motility study, is the most accurate method to diagnose achalasia. The awake procedure involves placing a catheter into the bottom of the esophagus and obtaining pressure readings as the catheter is withdrawn and the patient swallows water or breathes deeply. These pressure readings are then correlated to esophageal activity. The diagnosis of achalasia is determined by findings that include aperistalsis in the distal 2/3 of the esophagus, incomplete LES relaxation, and elevated LES pressure.

Risk Factors

Increased Risk Squamous Cell Carcinoma

Up-arrow Risk of Square-mouse with Car-gnome

Patients with achalasia are at increased risk for developing esophageal squamous cell carcinoma. However annual surveillance for cancer following a diagnosis of achalasia is not recommended currently.

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