International Foundation for
Gastrointestinal Disorders

IFFGD
3015 Dunes West Blvd.
Suite 512
Mt. Pleasant, SC 29466
www.iffgd.org

(543)

Distal Esophageal Spasm (DES)

Formerly Known as Diffuse Esophageal Spasm

By: R. Matthew Gideon, M.D. and Philip O. Katz, M.D., Head, Esophageal Testing Laboratory, Albert Einstein Medical Center, Philadelphia, PA

At a Glance

DES is a rare disorder.

Usual symptoms are chest pain and trouble swallowing.

The chest pain can feel like a heart attack.

Tests are needed to diagnose DES.

DES does not lead to other serious illnesses.

Most patients can be treated successfully.

Distal esophageal spasm (DES) is a motility (movement) disorder of the lower (distal) two thirds of the esophagus. When the esophagus functions normally, it produces a stripping wave of muscular contractions (peristalsis). In this way swallowed solid and liquid food moves from the top of the esophagus into the stomach in an orderly fashion. In contrast, in DES intermittent simultaneous contractions are mixed with normal motility. Patients with this disorder experience interruption of the normal sequential contractions with long segments of the esophagus contracting at the same time (simultaneous contractions). The disorder is rare with no known cause. It occurs equally in both men and women. We do not fully understand DES in part because it is typically treated medically. Surgical intervention is rare and therefore little tissue has been made available for study to better understand the disorder.

Symptoms of distal esophageal spasm

The most common symptoms of DES are chest pain and difficulty swallowing. Some patients may experience unusual symptoms like an unexplained cough. Symptoms are usually irregular with varying intensity. Chest pain is usually felt beneath the breast bone, but often radiates to the back. It can be brought on by eating and emotional stress, but is rarely brought on by exertion. It may awaken patients from sleep. Feeling like the pain of angina pectoris (heart pains), it results in many patients going to hospital emergency rooms believing they are having a heart attack. Because simultaneous contractions occur at irregular intervals, difficulty swallowing is usually intermittent for both solids and liquids. Weight loss from fear of eating, and food impaction in the esophagus are rare but have been reported.

Tests for distal esophageal spasm

The diagnosis may be suggested by a barium swallow (an x-ray in which a contrast material is swallowed that coats and outlines the esophagus) but the findings are not specific to this disorder. Many specialists will perform a test that views the inside of the esophagus through a thin scope (endoscopy) to be certain there is no structural cause for the chest pain or trouble swallowing. An esophageal motility test should be performed to confirm the diagnosis of distal (diffuse) spasm. The esophageal motility test measures both muscular strength and coordination and is performed by passing a small tube containing pressure sensitive transducers through the nose and into the stomach.

The tube is withdrawn until the transducers, which are hooked to a computer, are in the esophagus. Water (5cc) is given every 20 seconds to evaluate the peristaltic contractions. Abnormalities of the lower esophageal sphincter (LES) (the valve where the esophagus joins the stomach) may also be seen. While the sphincter is often normal it may also become hypertensive (squeezing harder than normal) or it may not relax or open normally during swallowing.

Management of distal esophageal spasm

There are many different treatments for DES. Some patients will respond quickly with little intervention while some will require multiple treatments. Each patient must be treated individually and may require several different approaches to find the right one for them.

It must be emphasized that DES rarely progresses to more serious diseases like achalasia and is *not* a risk for cancer or true obstruction. It is important to understand that breathing will continue during an attack.

Some patients can relieve the pain of spasm simply with a glass of warm water, relaxed breathing or a single sublingual (under the tongue) dose of hyoscyamine (Levsin, NuLev/SL). Calcium channel blockers, or nitrates are the most common agents used to treat DES. Nitrates may be given as needed (sublingual nitroglycerin) either as prevention (if symptoms have a well defined onset) or for rapid relief if symptoms are infrequent. Long acting nitrates or calcium channel blockers taken once to four times per day may be effective for those with more regular symptoms. These agents work by relaxing smooth muscle (the lower or distal portion of the esophagus) but are not without side effects. Calcium channel blockers can cause patients to become constipated, and nitrates may cause headaches. Both medications may cause lightheadedness.

Some reports have shown botulinum toxin to be effective when injected into the smooth muscle of the LES or distal esophagus. This is believed to work by interfering with the transmission of nerve impulses to the muscle, facilitating relaxation of the muscles. Some patients may experience chest pain beneath the breastbone after injection, which may last for up to 30 minutes but otherwise appears to have few side effects. This treatment works best with the initial injection, however the effect often does not last, may require retreatment, and lose effectiveness after 2 or 3 treatments.

Other treatments may be effective including medications used to treat anxiety or panic attacks (like Xanax) or pain from functional bowel diseases (such as tricyclic agents, or anticholinergic agents like dicyclomine, for example)). Peppermint may relieve painful spasms in some, while biofeedback or hypnosis may be helpful in others. Severe cases of DES, though rare, may require surgical intervention to perform a long incision (myotomy) in the LES and esophageal muscle to relieve spasms and improve swallowing.

Summary

Distal (diffuse) esophageal spasm is a rare but well-defined problem characterized by intermittent chest pain and or trouble swallowing. The cause is unknown. Diagnosis should be confirmed by an esophageal manometric study. Major problems are extremely rare and most patients respond successfully to careful, individualized treatment.