Achalasia

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FIGHTING DIGESTIVE DISEASES
Registered charity 1137029

This factsheet is about Achalasia

Achalasia is a condition that affects the nerves and muscles of the oesophagus (food pipe), mainly at the lower end where it meets the stomach. This hinders food passing down the oesophagus and entering the stomach resulting in difficulty swallowing food and liquids. It is uncommon and in most cases the causes is unknown.

Causes of Achalasia

Achalasia occurs when the nerves and muscles of the lower part of the oesophagus waste away. There is failure of relaxation of the lower oesophageal sphincter. This is a band of muscle at the between the oesophagus and the stomach that acts like a valve. Because it cannot relax, food is prevented from entering the stomach. In addition, the muscles of the oesophagus lose their ability to contract in a coordinated way, further impairing the passage of food down the oesophagus. Over time, the oesophagus can become enlarged (dilated). In most cases the cause of achalasia is unknown. However, in rare cases it can be caused by specific infections.

What are the usual symptoms?

The most common symptom of achalasia is difficulty swallowing (dysphagia) that occurs with both food and liquids. This is because the oesophagus is not functioning properly and passage of food and liquid down the oesophagus and into the stomach is hindered. Food may feel like it is stuck in the oesophagus after eating, causing chest discomfort. Heartburn is common. Regurgitation of food (bringing food back up) may occur as food is retained in the oesophagus and has nowhere to go except upwards. This can cause choking or coughing and can lead to chest infections if the food goes back down the wrong way and into the lungs. As a result of food not passing into the stomach and difficulty swallowing, less food is ingested and it is common to lose weight. Sometimes, no symptoms are experienced, and it is found incidentally during tests for a different condition.

How is Achalasia diagnosed?

Achalasia may be suspected by the doctor based on symptoms the patient has but tests are needed to confirm the diagnosis. The non-specific nature of the symptoms and the relative rarity of the condition mean that there can be a delay in diagnosis. There are a number of tests:

- Oesophageal manometry – this measures pressure waves in the oesophagus and is the main investigation for diagnosis of achalasia. This involves placing a small plastic tube into the oesophagus via the nose. The tube is lined with pressure sensors and three abnormalities are normally detected – high pressure in the sphincter at rest, failure of the sphincter to relax/open after swallowing and absence of useful muscle contractions in the lower part of the oesophagus.
- Barium swallow and chest X-ray –this involves drinking a white liquid called barium that highlights the shape/outline of the oesophagus when a chest X-ray is taken. The chest X-ray may show the characteristic enlargement/dilation of the oesophagus, then a narrowed region at the sphincter where the oesophagus meets the stomach as it is unable to relax.
- Endoscopy - This is where a small tube (the width of a small finger) with a camera on the end is inserted into the oesophagus and stomach via the mouth. Retained food may be seen in the oesophagus and the tight sphincter on entrance to the stomach may be noted.

How can Achalasia impact a person?

Achalasia can impact a person in many ways. These include complications of the condition and its overall impact on general wellbeing due to the symptoms.

Weight loss and poor nutrition can be a problem. If the patient is unable to maintain a healthy weight and becomes malnourished, other feeding methods may be needed such as nasogastric feeding (feed through a tube which is placed into the stomach via a tube). Recurrent chest infections from regurgitation of food can be also problematic.
A complication of achalasia is mega-oesophagus. This is where over a long period of time the oesophagus continues to dilate until it becomes severely dilated and the muscles cannot stretch anymore, sometimes causing the oesophagus to tear or burst. This is an emergency. Symptoms can include sudden onset abdominal/chest pain that can be tearing, bringing up blood and the patient can become very unwell very quickly.

Written by Samantha Morgan and Mark Samaan. Published in 2016. Next review in 2018. Full range at www.corecharity.org.uk.
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Achalasia can - very rarely - be associated with developing cancer of the oesophagus. Although highly unusual it is one of the reasons you should remain under follow up with a specialist.

What treatment is available for Achalasia?
There are a number of treatment options for achalasia, including medications, endoscopy procedures or surgery. Unfortunately, treatments can't cure the underlying process causing achalasia, but they can significantly help with symptoms by aiming to relax the sphincter so food can pass into the stomach. There is controversy about the "best" treatment, and so the right approach is to discuss with one's specialist which is the optimal management for one's own situation - taking into account factors like age, previous medical history, time to diagnosis, personal choice.

- Medications – The aim of medications are to relax the sphincter at the lower end of the oesophagus, and reduced spasm in the oesophagus. Medications include two classes of drugs called nitrates or calcium channel blockers (e.g. nifedipine). These may not be effective in all patients and often become less effective over time. Side effects include headache and low blood pressure.
- Dilatation – this is where the sphincter is stretched open using a small balloon passed via an endoscope under sedation or general anaesthetic. Once in position, the balloon is inflated which stretches and breaks the fibres in the sphincter. It relives symptoms in about 60% of patients, but sometimes more than one procedure is needed. There are some risks which the doctor will explain prior to the procedure.
- Botox – A substance called Botulinum toxin is injected into the sphincter, guided by endoscopy. This relaxes the muscle fibres in the sphincter by temporarily paralysing nerves that signal the sphincter to contract. It can help with symptoms for up to a few months to one year. It is not a permanent treatment, and may be used if patient is unfit for other treatments such as surgery.
- Endoscopy – new endoscopy therapies (POEM – per-oral endoscopic myotomy) are being developed which may offer hope of an alternative non-surgical permanent treatment.
- Surgery – A procedure called a Heller myotomy is used to cut muscle fibres of the sphincter, aiming to permanently improve swallowing. The procedure is performed using key hole surgery (laparoscopy).

Unfortunately these treatments can lead to side effects such a heartburn or reflux and persistent chest pain. To reduce symptoms of heartburn after treatment ensure food is chewed well, eat sitting upright, sleep with a few pillows so not completely flat and avoid eating within three hours of bed time. If heartburn or pain is experienced, see the doctor who can prescribe some medications to help with this.

Does Achalasia need to be monitored and, if so, how?
Monitoring of symptoms with regular follow-up visits is important, as mentioned above. A dietician may be needed if the patient is struggling with intake.

How does Achalasia behave over time?
Unfortunately there is no cure for achalasia. However, some patients will achieve symptom control with treatment. Unfortunately, a small number of patients may have ongoing symptoms despite adequate treatment.

What to ask your doctor when you see them?
May I be referred to a dietician to see if there are any changes to my diet that may help with my symptoms?
How often do I need follow-up? Which is the best treatment for me in the long term?

What more research needs to be done on Achalasia?
Further research into the cause of achalasia is needed, as currently the cause of most cases is unknown.
For more information about research in this area please contact Core.

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