

ACHALASIA

Achalasia or cardiospasm is an uncommon condition which affects about 6,000 people in Britain. It results from failure of the valve (the *cardia*) between the gullet and stomach to open to allow food to pass through.

What is the cause of the illness?

Achalasia is caused by damage to the nerves in the gullet wall; the reason for which is unknown. It could be due to a viral infection earlier in life.

Symptoms

Symptoms may start at any time of life and usually come on gradually. Most people have difficulty, and sometimes pain, in swallowing their food increasing over many years. (This is called *dysphagia*.) Food may be regurgitated or vomited shortly after meals. The "vomit" may sometimes contain recognisable food eaten several meals ago, indicating that it was held up in the gullet (or *oesophagus*) for some time. Occasionally it may dribble out of the corner of the mouth to stain the pillow during the night. If it trickles down the windpipe it can give rise to repeated chest infections and even pneumonia. In some people the disease causes no symptoms and is found incidentally on a chest X-ray or other investigation. Because food is retained in the oesophagus weight loss, sometimes severe, may occur.

Investigations

Barium swallow and chest X-ray.

This involves swallowing a white liquid containing barium which allows the gullet to be seen on X-ray. In achalasia the exit at the lower end never opens properly and, together with a lack of the progressive contractions which propel food along the gullet, causes a delay in barium passing into the stomach. A chest X-ray may show a wide gullet.

Endoscopy

This is a test in which a flexible instrument called an endoscope is used to look directly at the lining of the oesophagus (gullet) and stomach. Retained food will be visible, and the tight valve at the lower end yields to allow the endoscope through. The endoscopist can then check that there is no other disorder in the stomach.

Manometry

This measures pressure waves in the gullet. A small plastic tube is passed into the gullet through either the mouth or nose. Pressure at different sites in the gullet is measured. In achalasia there are usually weak contractions of the upper gullet and sustained high pressure (spasm) of the valve at the lower end of the gullet. It is this high pressure and the failure of the valve to relax in response to a swallow that causes difficulty in swallowing and food remaining in the gullet.

Treatment

The aim of treatment is to relax the valve at the *cardia* so that food can pass easily into the stomach. The underlying disease cannot be cured, but there are various ways in which the spasm at the end of the gullet can be reduced and symptoms improved.

Drugs

The valve at the lower end of the gullet may be temporarily relaxed by drugs. Tablets such as nitrates or nifedipine will only produce a brief improvement in swallowing and are ineffective in some patients, but may be useful while a more permanent treatment is planned. These tablets may cause headache, which usually gets less with continued treatment.

Dilatation (stretching the valve)

This is done under a sedative or general anaesthetic. A small balloon (30mm; 1 ½ inches) is used to stretch and break up the muscle fibres of the valve at the lower end of the gullet. This usually improves swallowing. The dilatation may need to be repeated after one or more years.

Botulinum toxin ("Botox") injection

This substance causes relaxation of muscle fibres. It can be injected painlessly into the cardia valve through an endoscope. This is usually effective for a few months, occasionally for a few years, but then has to be repeated. It is not a permanent treatment, but is useful for patients who are unable or unwilling to have surgery.

Surgery

Under general anaesthetic the gullet can be approached through the chest wall or the abdomen. The muscle fibres that fail to relax are cut and this results in a permanent improvement in swallowing. The operation is now often performed by laparoscopy ("key-hole surgery") requiring only an overnight stay in hospital.

How to reduce symptoms after treatment

Food should be chewed well. It is wise to eat food sitting upright, accompanied by fizzy drinks to ensure that the gullet is kept clear. Using several pillows or raising the head of the bed on wooden supports so that you sleep fairly upright can also be helpful. After dilatation or surgery, acid may be able to rise from the stomach into the gullet through the weakened valve, causing heartburn.

If heartburn develops after treatment it is important to consult your doctor as medication may be needed to reduce the acid regurgitation. Any recurrence of swallowing difficulties or weight loss should be reported to your doctor. Chest pain may persist after effective treatment. This may be difficult to cure, but drinks of cold water often give relief.

Is there a familial tendency?

There is nothing to indicate that achalasia is an inherited illness. Women with achalasia can have a normal pregnancy and their children will develop normally.

Is there any risk of cancer?

If the gullet contains a large amount of food which does not pass into the stomach in the normal way, the risk of cancer of the gullet is slightly increased. It is therefore important to relieve the hold up by appropriate treatment even if symptoms are not disabling.