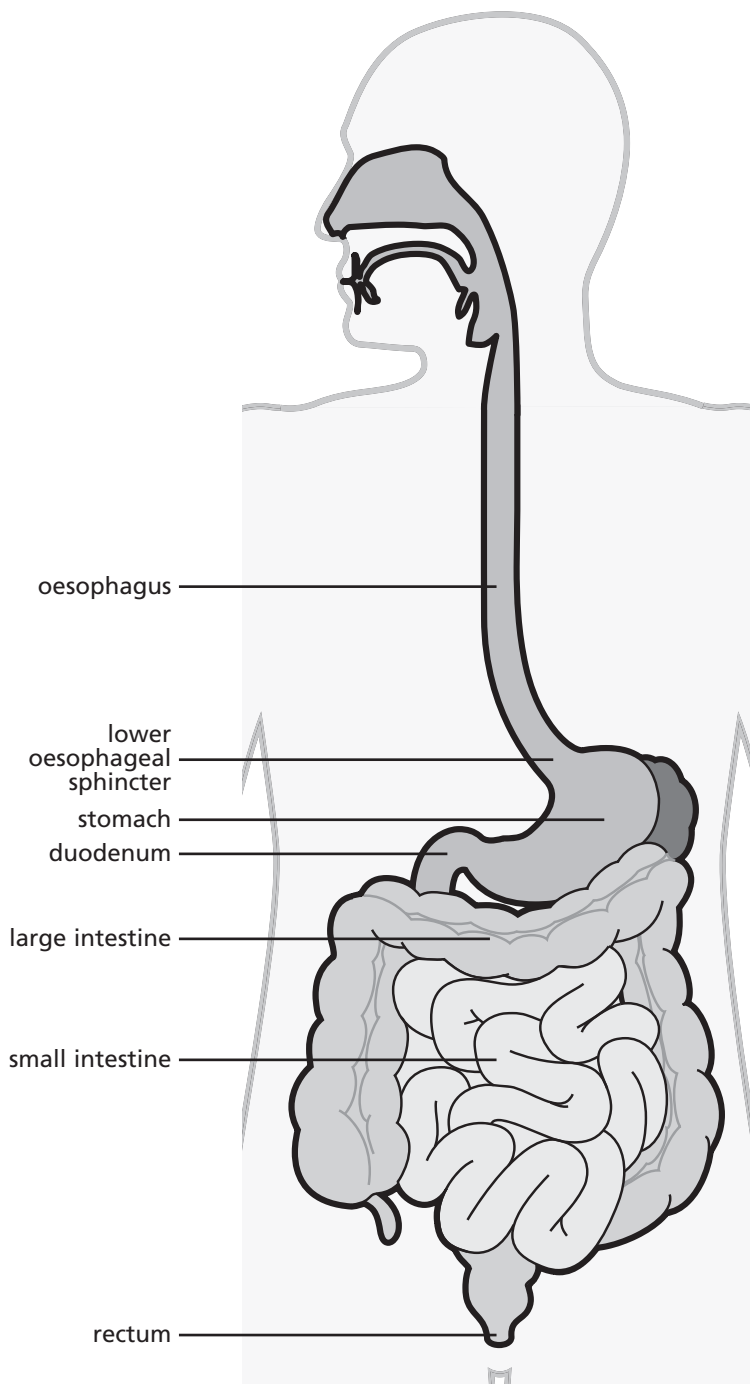




Achalasia

This leaflet explains the causes, symptoms and treatment of achalasia and where to get help from Great Ormond Street Hospital (GOSH).



The oesophagus (foodpipe) is the tube that takes food from the back of the mouth to the stomach. It contains muscles which squeeze rhythmically to push food downwards. At the end of the oesophagus is a specific area of muscle (lower oesophageal sphincter) that stops food passing back upwards from the stomach. In achalasia, the muscles and sphincter do not work properly so food cannot pass easily into the stomach to be digested.

What causes achalasia?

In achalasia, the muscles that squeeze rhythmically to push food downwards do not work properly. In addition, the sphincter at the base of the oesophagus does not relax as it should to let food pass through into the stomach.

Achalasia is most common in adults but can occur in children too. In children, achalasia affects around two in every million, with boys affected more than girls. It is most commonly diagnosed after the age of 10 years but can occur in younger children.

What are the signs and symptoms of achalasia?

The main sign of achalasia is difficulty swallowing (dysphagia). Children are more likely to have difficulty swallowing solids rather than liquids. Other symptoms include vomiting, chest pain, heartburn and reflux (where the food passes back up the oesophagus). Sometimes, stomach contents



can be breathed into the lungs, which can cause frequent chest infections. As it becomes more difficult to eat, children may not gain weight as expected.

How is achalasia diagnosed?

Imaging scans, such as chest x-rays, may show the oesophagus ballooning above the sphincter. Usually contrast studies are suggested. Your child will be asked to drink a liquid that shows up well on x-rays.

Oesophageal manometry is also used. A catheter (clear, plastic tube) is inserted into your child's digestive system through their nose and fluid is flushed through it to copy what happens when your child eats or drinks. This causes the nerves and muscles to start working, to squeeze the fluid through your child's digestive system. This activity is picked up by sensors contained in the catheter and recorded on a machine.

Most children also have an endoscopy under general anaesthetic, where thin flexible tube containing a light and a camera is passed through the mouth down into the oesophagus. This allows the doctors to see inside the oesophagus and also take tiny samples of tissue (biopsies) for testing in the laboratory.

The endoscope can also be used to inject tiny amounts of botulinum toxin into the sphincter. Botulinum toxin is produced naturally by the bacterium *Clostridium botulinum*. When purified, it can be used in tiny, controlled doses to relax excessive muscle contraction.

If your child is having manometry and endoscopy, these will usually be carried out at the same time under general anaesthetic.

How is achalasia treated?

One form of treatment is an operation called a Heller's myotomy, which aims to cut the muscle fibres in the oesophagus so that the sphincter is able to relax. This is generally done using keyhole (laparoscopic) surgery but occasionally open surgery is needed. A Heller's myotomy is usually combined with a partial fundoplication operation, where the top of the stomach is wrapped around the bottom of the oesophagus. This reduces reflux of stomach contents into the oesophagus, which can be a side effect of the Heller's myotomy.

Another form of treatment is a procedure called oesophageal dilatation, which is always carried out while your child is under a general anaesthetic. Once your child is under general anaesthetic, the doctor passes a catheter (soft plastic tube) containing a balloon down the back of your child's mouth into their foodpipe. They watch where the catheter is by using X-rays and continue to pass it down the foodpipe until it reaches the narrowed section. Once it is in place, the doctor inflates the balloon so that it stretches the narrowed section. Further X-rays are taken to check how much the balloon is inflated. At the end of the procedure, the balloon is deflated and with the catheter is brought back up the foodpipe and out of your child's mouth.

What happens next?

Children who have had achalasia will need regular follow up appointments as the condition can come back, causing similar symptoms as before. If dilatation – botulinum toxin injections are tried several times and are unsuccessful, surgery may be required to treat the condition.