



ACHALASIA ACTION

Uniting for a rare swallowing condition

Achalasia Explained

Helpline 0300 772 7795

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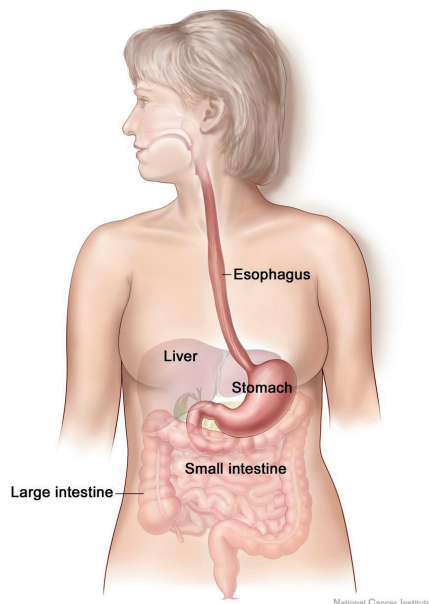
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What is Achalasia?

Achalasia is a rare swallowing condition that prevents the food you swallow from reaching your stomach properly. It affects the muscles in your oesophagus, the food pipe that runs from your throat, down through your chest and to your stomach. (In America and some other parts of the world, the spelling is esophagus). In a normal oesophagus, muscles squeeze the food down towards the stomach by tightening and relaxing in a coordinated sequence in a process called peristalsis. At the lower end of the oesophagus, a valve-like ring of muscle (the lower oesophageal sphincter or LOS) should automatically relax to let food through into your stomach. Then the LOS should close again to stop stomach acid from rising. Peristalsis normally takes two or three seconds and is so effective that some people can drink large quantities of liquid whilst upside down.



With achalasia, the muscles in the oesophagus do not work properly. They are either weak, do not tighten and relax in the right order, or do not work at all. The LOS, located just above your stomach, often fails to open. Food and drink then block back into your oesophagus and can remain there for a long time, sometimes hours or days. This food build-up can create pressure, similar to a blocked hosepipe, until

the food either trickles down into your stomach, or is brought back up through your mouth (regurgitation). This is not caused by a fault in the muscles themselves but seems to be a problem with the nerve endings that send signals to the brain to operate those muscles.

What are the symptoms of achalasia?

If you have achalasia you will not always experience the same symptoms as other people in the same way. People with achalasia can have a whole range of severity of symptoms, or sometimes a much milder version. So interpret the list of symptoms, compare them with your own experience and do not take things out of proportion. This booklet aims to be comprehensive, and whilst some of these problems are listed below, you may well not experience all of them. Although this may be difficult, you should try not to feel more anxious about eating, not least because stress and anxiety do tend to make symptoms worse. It is not inevitable that you will necessarily face the more serious problems. We try to give information about how some symptoms can be alleviated.



Food starting to 'pool' at the base of oesophagus

Stuck food

The food that remains blocked in your oesophagus can cause discomfort and sometimes pain in your chest area from the extra pressure. It can also cause anxiety and worry about food not going down into your stomach. Food can pool at the bottom of the oesophagus. Food such as rice and bread can cause a congealing problem. Food with a stringy texture and vegetable or fruit skins can also cause particular problems.

Regurgitating of food and 'drooling' of saliva

If food residue cannot go down into your stomach, the pressure on your oesophagus can only be relieved by it moving upwards and being regurgitated. People often try and help blocked food go down by carefully taking a drink of warm or cold water or a fizzy drink. In

extreme cases people have sometimes used their fingers to alleviate the problem. Sometimes saliva does not flow properly between the oesophagus and the stomach and can become trapped, leading to excess bubbles and saliva needing to be spat out, or drooling (draining outside your mouth). Regurgitation involves food that does not reach the stomach; reflux is acid and other contents from the stomach itself.

Chest pain from spasms

Spasms seem to be caused by pressure on the nerves and muscles from blocked food, but they can also be caused by reflux and other reasons. Sometimes these pains are misdiagnosed as heartburn. Spasms are not well understood. In an informal survey, people with achalasia most often associated spasm with stress, followed by reactions to certain food. As often occurs with pain, the sensation can sometimes be transferred through the nerve system and be felt in other places like the jaw or back. For some people spasms might be their first, or an early, symptom.

Losing weight

Difficulty in swallowing can easily lead to a reduced intake of food and loss of weight. Sometimes this inability to take in food, and the understandable anxiety that arises from this, can lead to young people being wrongly diagnosed with an eating disorder like bulimia or anorexia, despite the fact people with achalasia genuinely want to eat well.

Malnutrition

Nutrition is absorbed into the body through your digestive system, so the reduced ability of food to reach your stomach will eventually affect nutrition levels. Your GP may prescribe special nutrition drinks such as Fortisip.

Effects from fermenting food, and heartburn

If blocked food in your oesophagus starts to ferment, you may feel a nasty taste in your mouth because of this fermentation of stale food. This can be confused with reflux, where acid comes up from the stomach itself.

If achalasia has not been treated, the sour taste is much less likely to be caused by stomach acid because your LOS valve is likely to be too tight to allow the stomach acid to rise in this way. If you have had a dilatation, myotomy without a fundoplication or a POEM, the LOS will inevitably be more relaxed so the cause of the sour taste is much more likely to be stomach acid.

The oesophagus is a sensitive organ. The lining will tend to be irritated by prolonged exposure to stomach acid, and to a lesser extent by fermenting food, depending on what you have eaten. This is because it is different from the lining of the stomach which is designed to withstand the effect of the strong acid that is part of a healthy digestive system.

Heartburn is a pain in the chest area, normally caused by the effect of reflux of acid from the stomach itself, but it is a feeling that can have a variety of causes. Heartburn is a common complaint, normally treated with over-the-counter indigestion remedies like Gaviscon, Rennie's or Tums. If heartburn is persistent for more than three weeks you should consult your doctor. Long term reflux can be a significant health problem.

Medication absorption

Achalasia can result in pills remaining stuck in your oesophagus rather than being dispersed into your system through your stomach. This might cause damage to the sensitive lining of your oesophagus as well as the loss of the intended benefit of the medication. A GP or pharmacist can be asked about whether medication can be prescribed in liquid form to reduce this risk.

Choking and coughing fits

Food residue can sometimes flow upwards and find its way into your lungs, especially when you are lying down. You may find it helpful to raise your head and shoulders with pillows, raising part of the mattress of your bed or even considering a bed with adjustable settings.

Chest infections and other respiratory problems

Food residue that flows up your oesophagus and into your lungs can cause infections and pneumonia.

Fatigue or tiredness

Being unable to swallow food can lead to a lack of strength and energy. It can be helpful to ask for your levels of vitamins and minerals to be tested.

Anxiety and depression

Achalasia is a long-term condition that, for some, can affect your quality of life and destroy the pleasure of eating. Some people find relaxation techniques helpful in reducing anxiety and tension.

A weak and fatigued voice

Reflux around the vocal cords can lead to your voice being affected.

How many people have achalasia?

Achalasia is rare, and statistics are not precise. The number of people diagnosed each year (incidence) is similar in most countries, and is estimated as between 0.7 to 1.6 per 100,000 inhabitants per year. The UK population of 66.8 million would therefore, in theory, generate between 467-1,069 diagnoses annually. The prevalence (number of people continuing to be affected by the condition after diagnosis) is regarded internationally as about 10 in 100,000 and is estimated by the NHS to be around 6,000 in the UK. The latest high-resolution manometry techniques may be contributing to more cases being diagnosed.

How old are those with achalasia?

The condition affects both men and women. It is most common between the ages of 30 and 60 years, but it can develop at any age. Some young children are diagnosed with achalasia, and, rarely, it can

be present from birth. A study found only 228 UK children under 16 years diagnosed with achalasia in a 10-year period until 2008 but with a rise in the annual rate within that decade¹.

What causes achalasia?

The muscles within the oesophagus that perform peristalsis are normally automatically controlled by a complex, interlinked system of nerve cells, or neurons. The cause of achalasia is related to the degeneration and malfunctioning of a specific type of nerve cell called ganglia that send signals to the brain to operate the muscles. The loss of function of these neurons impairs the normal motility (the passage of food through the digestive system) of the oesophagus and dilation of the LOS – resulting in an oesophagus that cannot channel food into the stomach properly.

The causes of the degeneration of these nerves and therefore of achalasia are not clear, and research into the causes of achalasia has so far been inconclusive. They are likely to be different for individuals.

It is believed that infections from the varicella zoster virus (linked with chicken pox and shingles) or a side effect from measles may be contributory factors.

Achalasia is sometimes described as an auto-immune disease, meaning that the body's immune system may have started to react differently. There may be occasional cases of achalasia that may be associated with auto-immune disease, or occur in patients who already have auto-immune diseases. As far as we understand it, this is not of the scale of some other conditions which are actually autoimmune in aetiology (how they are caused), and where, for instance, treatment would entail prescribing immune-suppressant drugs or steroids. Furthermore this does not normally mean that if you have achalasia your immunity from general infections is affected.

There can sometimes be a genetic predisposition. Children with Down syndrome are more likely to experience achalasia. Sometimes

¹ Marlaisset et al Arch Dis Child 2011;96:192–194

there is an association with other gastric disorders such as delayed gastric emptying.

One disease that can be similar to achalasia and leads to the same changes in the oesophagus is Chagas disease, caused by the triatomine bug, which can carry the *trypanosoma cruzi* parasite. The disease affects around 6-7 million people worldwide, mostly in Latin America, and can cause cardiac and sometimes digestive complications as well.

Pseudoachalasia is a term given to describe symptoms that are very similar to achalasia, but the lack of peristalsis is caused by an obstruction such as a gastric band or a tumour near the LOS.

How is achalasia diagnosed?

Swallowing problems can have a variety of different causes in addition to achalasia, and the diagnostic tests may be similar. A diagnosis depends upon tests normally given by a gastroenterologist in a specialist Upper GI (upper gastro-intestinal tract) clinic setting in a hospital.

Endoscopy

An endoscope is a miniature camera on a flexible tube that is passed through your throat and oesophagus and into your stomach to provide images that will normally reveal signs of a physical cause of swallowing or digestive problems. An endoscopy can reveal conditions like Barrett's Oesophagus, ulcers, inflammation or strictures. The device can also take biopsies for analysis.

The endoscopy might detect a tight LOS, a dilated or 'baggy' oesophagus, or undigested food residue. The test is important to confirm or disprove other causes of obstruction, but will often not be sufficient in itself to diagnose achalasia because it does not analyse the swallowing process. Endoscopy is often performed under sedation, meaning that you should have somebody to accompany you home afterwards. Some endoscopies are undertaken with a throat spray rather than under sedation.

Recently some hospitals have been using trans-nasal endoscopy where a thinner instrument and tube are used and passed through your nose. Improvements in technology have meant that these images are equivalent to conventional endoscopy, and can be performed whilst you are sitting in a chair in a non-specialist environment and without the need for sedation.

Barium swallow

This test involves your swallowing a white, chalky liquid whilst being monitored by an x-ray machine that records the progress of the liquid into your stomach. Sometimes the progress is recorded at 1, 2 and 5 minute intervals to illustrate any swallowing delay. For somebody with achalasia, the x-ray pictures will show the liquid taking a long time to reach the stomach, compared to a healthy adult, where the liquid reaches the stomach in a few seconds. The white liquid sometimes shows a 'bird's beak' profile when the liquid is blocked at the entrance of the stomach from the non-relaxing LOS.



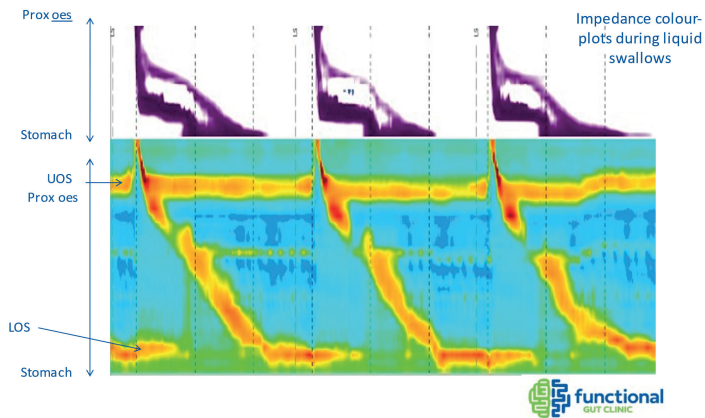
Barium swallow image showing 'bird's beak' effect of blockage at the LOS

Manometry

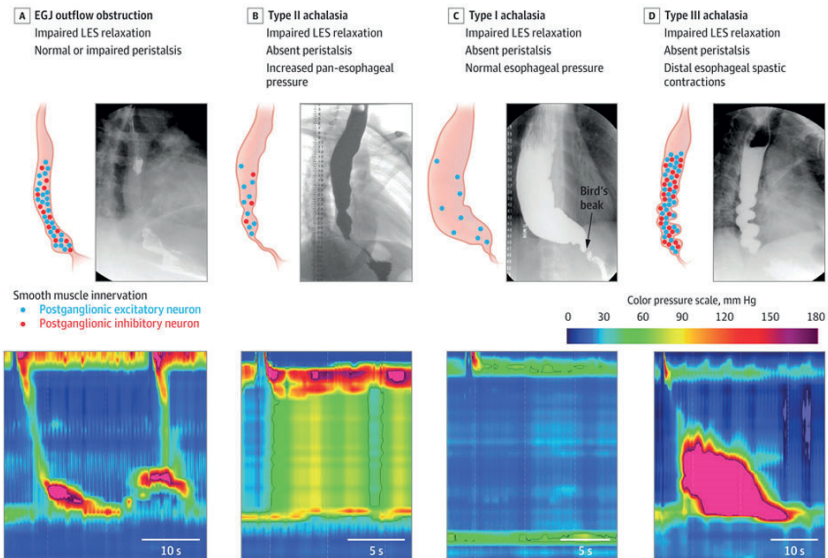
With manometry, a thin and flexible tube that contains pressure sensors is passed through your nose and normally into your stomach. Your throat and/or nose will be numbed with a spray or gel. Once the tube is in your oesophagus, you will then be asked to drink small sips of water and/or eat some food. This then measures the pressure at set intervals as the bolus (a portion of undigested food) travels down towards your stomach.

Modern equipment is likely to provide high resolution manometry that gives much more information to help achieve a detailed diagnosis of where and how the muscles are working.

High Resolution Oesophageal Manometry and Impedance – Normal Swallow and Bolus Clearance



A manometry chart shows, in different colours, the pressures at different points from the throat (at the top of the chart) to the stomach (at the bottom) on the vertical axis; the timing of the process runs from left to right. In a normally-operating healthy oesophagus the line of greater pressure will be seen at the top of the chart running diagonally down to the bottom.



It is possible to measure the effect when different 'challenges' are undertaken. Examples of these challenges include trying to drink water rapidly, or eating food of varying consistency. This can be helpful for identifying problems that occur only in particular circumstances.

In recent years, gastroenterologists have been developing criteria (known as the Chicago classification), partly based on manometry results, for classifying achalasia into different types that can help to determine what treatment is likely to be most effective in relieving the symptoms. It is not always very helpful to be over-concerned about what type of achalasia you have, because each case is liable to be different. You may not fit easily into one specific category rather than another, and the classifications can be confusing. Sometimes the condition develops from one type into another according to the stage of the disease.

- **Type 1 Achalasia**

Type 1 achalasia means that there is no peristalsis at all and the LOS does not relax. This type often occurs when achalasia is more established and can sometimes develop from types 2 and 3.

- **Type 2 Achalasia**

Type 2 achalasia also involves the LOS not relaxing. The peristaltic muscles do react but in an ineffective or uncoordinated manner.

- **Type 3 Achalasia**

Type 3 involves the LOS not relaxing, but the muscles create chest pain with high amplitude spikes and uncoordinated, premature, vigorous contractions.

24-Hour pH monitoring

Sometimes it can be helpful to conduct a 24-hour acid monitoring test. A thin tube is inserted through your nose at the hospital. This process takes about an hour. The tube is attached to a monitor that you wear whilst you continue all your normal daily activities, keeping a diary of what you eat and drink. You then return to

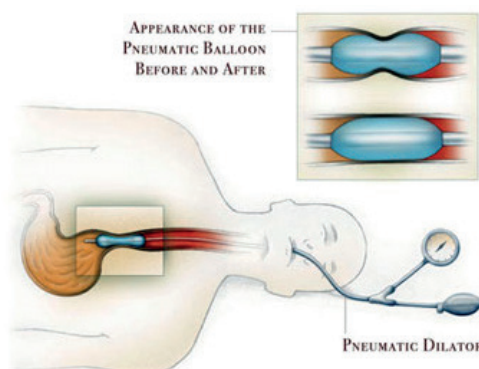
the hospital 24 hours later for removal of the tube and for the data in the monitoring device to be downloaded for analysis.

Sometimes the acid readings can be the result of reflux that comes from the stomach; sometimes they reflect the fermentation of food residue that remains in the oesophagus.

How is achalasia treated?

Achalasia is a long-term condition that cannot be cured at present, but there are treatments that can improve the symptoms. Treatments can range from lifestyle changes to surgery. Doctors should look at the patient as a whole, and take into account their overall physical health rather than being solely governed by test results.

Pneumatic dilatation



Dilatation (image courtesy of Memorial Hermann)

Dilatation involves passing an endoscope through your throat with a balloon device attached to the equipment. The balloon is then inflated so that the lower oesophageal sphincter (LOS) is stretched. This is sometimes best done in two sessions about three or more weeks apart. Dilatation often results in at least some relief from symptoms and can bring significant improvement, but sometimes the benefits can be only short-term. The effect of a dilatation can make the tissue fibrous and less amenable to future procedures or surgery, so repeated dilatations sometimes have to be balanced against a potentially more definitive long-term treatment.

Heller myotomy and fundoplication

This procedure was first performed in 1913 by surgeon Ernst Heller, using open surgery rather than modern laparoscopic (keyhole surgery) techniques. A laparoscopic Heller myotomy (LHM) consists of cutting the muscles in the lining of the oesophagus and stomach that control the LOS. Cutting the muscles reduces the compression of the LOS and allows it to remain permanently relaxed, thereby allowing food to move down into the stomach by gravity.

The length of the myotomy is an important issue for success of the procedure, and, in an adult, is typically about 10 cms in length, 3 cms of which would be in the stomach wall. A number of small incisions are made to allow access for the surgical instruments, but there is no conventional operation scar that would have been part of open surgery.

Normally, the LOS acts as a valve to stop stomach acid refluxing up into the oesophagus. Prolonged exposure to stomach acid can cause heartburn and can also damage the lining of the oesophagus. To prevent this from happening after the myotomy, a fundoplication is carried out.

The fundus is the top part of the stomach, and the procedure wraps the top part of the stomach round the bottom of the oesophagus to recreate the valve effect to control reflux. There are various techniques for this, all of which depend on the circumstances of the individual patient. Sometimes the wrap is made around the back of the oesophagus; sometimes from the front. A Nissen fundoplication covers the full 360° circumference, Toupet 270°, Dor 180°-200°, and Watson 120°. If the wrap is too tight it can cause too much restriction, and can affect your ability to vomit or belch. If there is a problem with the fundoplication wrap loosening over time, a surgeon can sometimes revise the procedure.

This surgery has been well established and often provides sustained benefits for the longer term. A trial conducted by Patti and others in the period 1991–1998 in California involved 168 patients, 29 of whom had an oesophagus with a diameter greater than 6 cms (2-3 cms is normal)², 90% of the patients had good or excellent results from the surgery.

² Ann Surg. 1999 Oct; 230(4): 587

If reflux occurs after the surgery this can be controlled by PPI (proton pump inhibitor) medication like Omeprazole that is designed to reduce stomach acid. This medication is safe. There have been concerns expressed about the side effects of this medication when it is taken over a long period, such as potential issues relating to osteoporosis and the effects on gut bacteria, but these would be unlikely to be factors in the short term compared to the benefit of relief from the reflux symptoms. It is generally prudent to ask your GP for a review of medication that you have been taking on a long-term basis so that any alternatives can be considered.

POEM

Esophagectomy

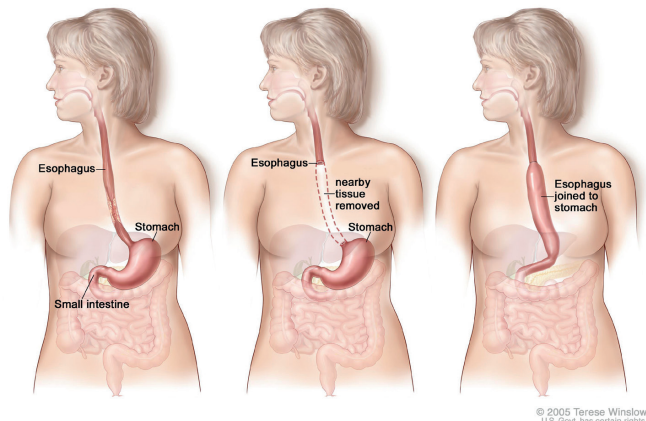
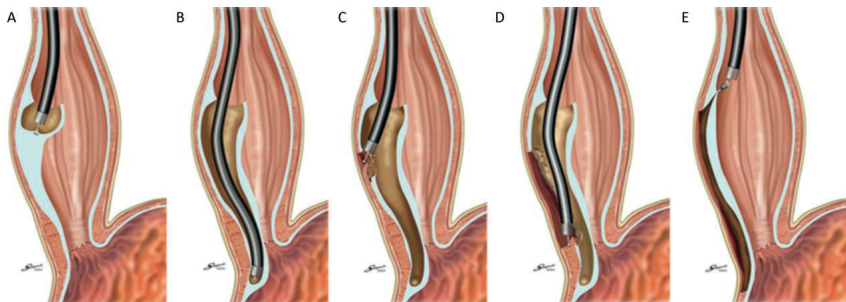


Diagram of an oesophagectomy
(© Terese Winslow; St Francis Care)

POEM (per oral endoscopic myotomy) is a relatively new procedure that is performed with an endoscope passed down into the oesophagus through your mouth under sedation or general anaesthetic. This procedure does not require incisions to be made in your belly for the laparoscopic instruments used for a Heller myotomy. The procedure may be performed by either gastroenterologists or surgeons. The endoscope is used to make a cut in the lining of the oesophagus from the inside, and the instrument is then passed through the interior wall of the oesophagus and the stomach to sever the same muscles as would have been cut in a Heller myotomy.

This procedure does not involve a fundoplication wrap to recreate the LOS valve effect against reflux. If acid reflux occurs afterwards, PPI medication may be required. There may be some specific situations where POEM may be a preferable technique compared to a Heller myotomy. Previous botox or dilatations may make the tissue more fibrous and may sometimes prevent the endoscope from reaching the full distance desirable for a full POEM procedure. It is sometimes possible for a POEM to be repeated using a different part of the oesophageal lining. As with many medical procedures, the experience of the medical practitioner involved is an important factor in success rates.

Medication



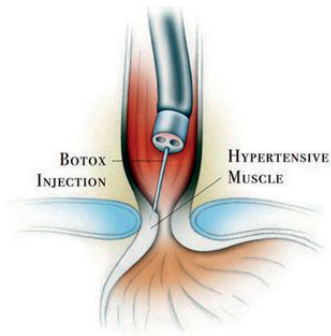
Endoscope entering the lining of oesophagus (POEM)
(©Inoue H. Showa University Northern Yokohama, 2010).

Although not regarded as standard treatment as such, calcium channel blockers can be used to reduce hypertension and reduce blood pressure. Nitrates can relax muscles in the LOS within a few minutes.

Generally there tends to be an initial improvement in 50%-90% of cases, but 30% of patients have some form of side-effects. This approach may sometimes be used as a temporary bridge pending other treatment.

Botox

Botulin toxin injections (Botox) into the LOS take three or four minutes and can reduce the muscle tone in the LOS, decrease LOS pressure and improve the flow of food into the stomach, but the relapse rate within three months is about 50%, so there are reduced chances of this having sustained benefits. It can be a useful approach for when surgical options are limited, but this option is regarded as suitable only for those unable to tolerate more definitive therapies.



Botox injection
(image courtesy of
Memorial Hermann)

Oesophagectomy

Very much at the extreme end of experience and a last resort for people with achalasia, an oesophagectomy involves removal of the oesophagus. The top of the stomach is then drawn up into the chest and joined to the remainder of the oesophagus below the throat. This surgery is a major operation with consequences for the digestive system like having to eat little and often, and having to avoid sugary food that might create insulin spikes. Patients can achieve a reasonably good quality of life after this surgery. Those facing the prospect of this surgery would normally discuss with a specialist surgeon a number of factors. These may include the prospect of relieving the severe loss of quality of life that end-stage achalasia can bring, whether they have other medical conditions, the risk of undigested food being aspirated into their lungs, potential general malnutrition, and their level of fitness to cope with a significant period of recuperation after the surgery.

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