



A Patient's Guide to Achalasia

-Produced by Achalasia Action (charity in the course of formation)

www.achalasia-action.org

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INTRODUCTION

We have written this booklet to help patients with achalasia to understand the condition better, to describe the various treatments that may be possible, and to record the various hints and tips that other patients have passed on to us. You should always be guided by your doctor, particularly a specialist, who has examined you and knows your own history and symptoms, but this book aims to be a patient's general guide. We have ensured that its contents are supported by medical knowledge and expertise.

Achalasia is rare. It affects about 6,000 people in Britain with all stages of the disease (prevalence), according to the *NHS Choices* website (www.nhs.uk/conditions/achalasia/Pages/Introduction.aspx). About 0.5 – 1 per 100,000 population are diagnosed each year (incidence); the equivalent figures in the USA and Canada are 1 and 1.6. It prevents people from swallowing properly, can have a severe effect on nutrition, and may prevent people from maintaining a healthy weight. It is often a condition that is not diagnosed readily, and can lead to considerable distress and loss of quality of life. In extreme cases it can eventually lead to the surgical removal of the oesophagus.

The condition affects both men and women. It is most common between the ages of 30 and 60 years, but first presentation to a clinic can be at any age. Some young children are diagnosed with the condition, and, rarely, it can be present from birth. There may be some genetic predisposition to the condition.

There has been little information available for achalasia patients until recently, and we hope that this booklet will help to fill that need. We hold the copyright of the text of this booklet. This is to ensure the accuracy of its contents rather than to restrict any distribution to patients and those with an interest in the condition.

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GLOSSARY

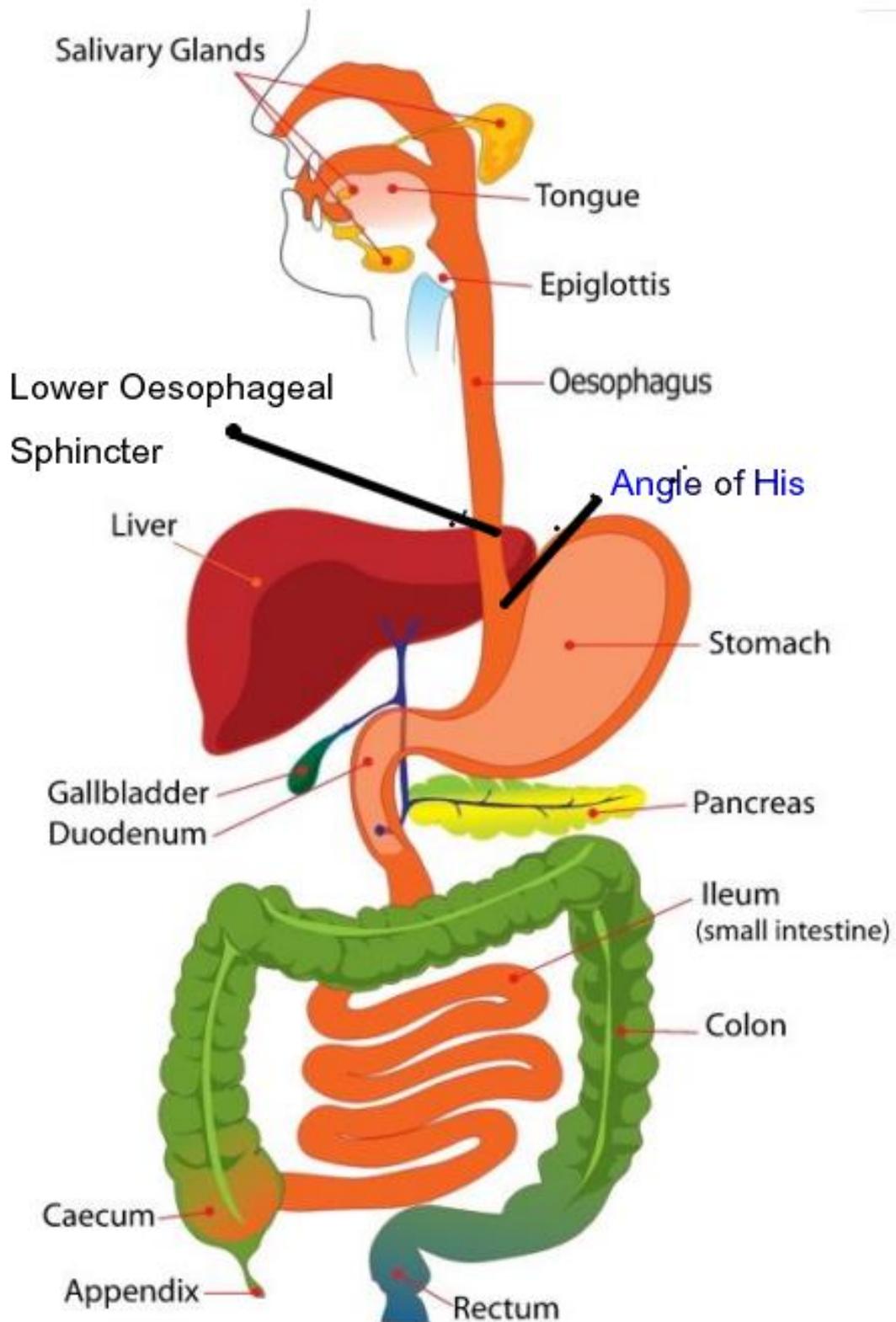
This glossary of medical terms appears here at the front of this booklet so that you will not miss it and will know where to look up if medical terms need a definition.

<i>Aperistalsis</i>	Absence of the wave of muscular actions in the oesophagus to push food towards the stomach
<i>Barium Swallow Test</i>	A test where the patient swallows a white liquid that can be tracked by a radiologist as it progresses through the oesophagus and into the stomach
<i>Barrett's Oesophagus</i>	A condition caused by prolonged exposure to stomach acid, where the lining of the oesophagus starts to change, and cells change to become columnar. Barrett's Oesophagus carries a small risk of oesophageal adenocarcinoma (cancer) which increases if dysplasia is found from a biopsy. A person aged 30 years with recently developed Barrett's oesophagus may have an 11 – 25% risk of developing adenocarcinoma before the age of 80.
<i>Bolus</i>	A lump or ball of food that has been chewed and swallowed.
<i>Candidiasis</i>	Also known as <i>Thrush</i> . A fungal infection caused by yeast that can develop in the throat and oesophagus
<i>Chagas Disease</i>	Mostly occurring in Mexico, central and southern America, Chagas disease is caused by insect bite infection and can lead to enlargement of the oesophagus.
<i>Cholinergic neurons</i>	Nerve cells that mainly use a neurotransmitter <i>acetylcholine</i> to send messages to the brain to operate part of the digestive system
<i>Chyme</i>	Partly digested food that passes through from the stomach to the intestines.
<i>Dor fundoplication</i>	Fundoplication where the wrap is around 180°-200° of the circumference of the oesophagus.
<i>Dilatation</i>	An endoscopy procedure with a 'balloon' that is inflated to stretch the lower oesophageal sphincter to allow it to pass food more easily.
<i>Dysphagia</i>	Difficulty in swallowing. Categorised 0 (none) – 4 (disabling), from Excellent – no dysphagia; Good – dysphagia once a week or less; Fair – more than once a week and requiring dietary adjustment; and Poor – dysphagia preventing ingestion of solid food.

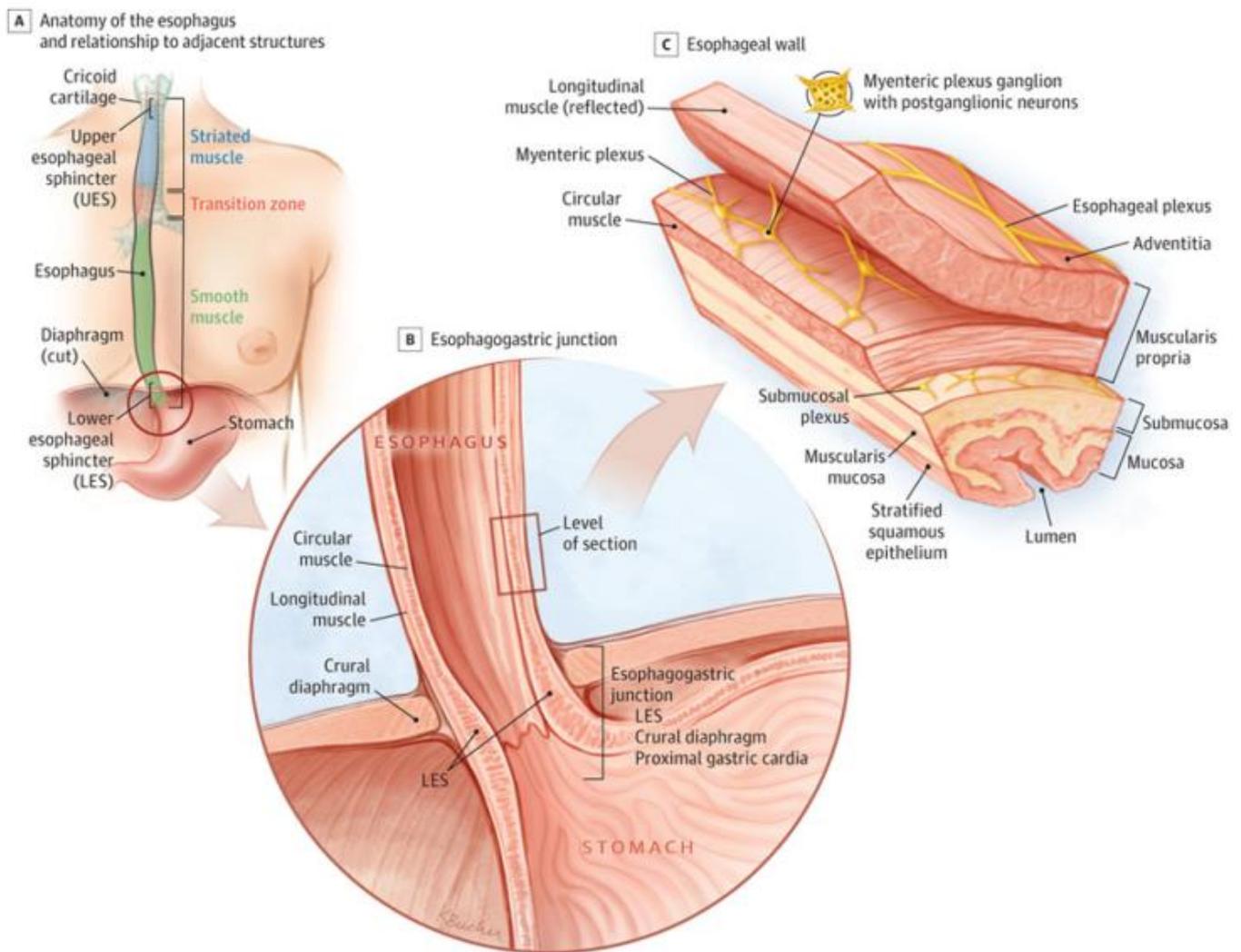
<i>Endoscope</i>	A miniature camera with a flexible tube passed through the throat (or sometimes the nose) to examine the inside of the oesophagus and stomach in a process called <i>endoscopy</i>
<i>Enteric neurons</i>	Part of the mesh-like nerve system that controls the digestive system
<i>Esophagus</i>	American spelling of Oesophagus
<i>Fundoplication</i>	A surgical procedure for wrapping the top part of the stomach around the bottom of the oesophagus to create a valve-like effect to function like a lower oesophageal sphincter and control reflux.
<i>Fundus</i>	The top part of the stomach, bulging above the junction of the oesophagus
<i>Ganglia</i>	Nerve cells.
<i>GORD</i>	(or GERD in America) Gastro-Oesophageal Reflux Disease. The complicating factors of stomach acid and other contents rising into the oesophagus causing irritation, inflammation, oesophagitis, Barrett's Oesophagus, or cancer.
<i>Globus sensation</i>	A feeling of a lump in the throat when medical examination cannot find a physical reason for it. The cause is often unknown
<i>Heller myotomy</i>	A surgical procedure to cut the muscle of the lower oesophageal sphincter to allow food to pass through more easily, first performed by surgeon Ernest Heller in 1913
<i>His</i>	The angle of His is the angle at which the oesophagus joins the stomach
<i>Hypercontractile Oesophagus</i>	Extreme form of Nutcracker Oesophagus
<i>Incidence</i>	Statistics counting how many cases are diagnosed each year, as distinct from <i>prevalence</i>
<i>Jackhammer oesophagus</i>	Alternative name for Hypercontractile Oesophagus
<i>Laparoscopic surgery</i>	Keyhole surgery, where instruments are inserted into the patient's body through small apertures rather than open surgery.
<i>Linx</i>	A recently introduced surgical device in the form of a magnetic bracelet that is wrapped around the base of the oesophagus to recreate the valve effect of the LOS. Used predominantly for anti-reflux surgery for patients for whom medication does not work

<i>Lower Oesophageal Sphincter (LOS)</i>	A sphincter valve near the abdomen between the stomach and chest that should allow food to pass downwards through to the stomach, but keeping stomach acid from rising into the oesophagus
<i>Manometry</i>	An examination involving a thin, pressure-sensitive tube that is passed into your oesophagus to monitor and record pressure at various points along the length of the oesophagus. This sometimes takes an hour; sometimes longer. 24 hour manometry is sometimes used. Testing for acid (pH) reflux can be done at the same time.
<i>Motility</i>	The muscle movements that propel food through the digestive tract
<i>Myenteric plexus</i>	Muscular coat of the oesophagus
<i>Myotomy</i>	A surgical procedure to cut muscle(s), in achalasia cases the muscles preventing the lower oesophageal sphincter from opening
<i>Nissen fundoplication</i>	Fundoplication where the wrap is around 360°, the whole circumference of the oesophagus.
<i>Nutcracker Oesophagus</i>	Exaggerated pressure from muscles and peristalsis, causing chest pain, regurgitation and difficulty in swallowing. Contractions occur in sequence rather than simultaneously.
<i>Oesophagectomy</i>	A surgical operation to remove the oesophagus. The remainder of the stomach is then re-fashioned, pulled up into the chest and joined to the stump of the oesophagus near the throat. The digestive tract thereby becomes shorter. It is a major operation, usually undertaken for cancer patients, but it is often possible, with care, to enjoy a good quality of life afterwards.
<i>Oesophagus</i>	The gullet, or food tube, around 20cms in length, that takes food from the throat to the stomach
<i>Peristalsis</i>	A series of coordinated wave-like muscle contractions and relaxations that should move food further down the digestive tract
<i>POEM</i>	A recently introduced surgical procedure, <i>Per Oral Endoscopic Myotomy</i> , that involves cutting the muscles <i>within</i> the lining of the oesophagus around the lower oesophageal sphincter (LOS) to relax the LOS and allow food to pass through. The procedure is carried out with an endoscope.

<i>Post Nasal Drip</i>	Excessive mucus inside nose or back of throat. Can be caused by a number of things that lead to regurgitation or reflux, but can be an effect of motility disorder such as achalasia.
<i>PPI</i>	Proton Pump Inhibitor medication (eg Omeprazole) switches off the production of stomach acid to reduce reflux problems
<i>Prevalence</i>	Statistic counting the number of people who have a condition within the population (sometimes many years after diagnosis)
<i>Reflux</i>	Stomach contents rising up into the oesophagus
<i>Sequelae</i>	After-effects or secondary results of a medical condition
<i>Upper GI</i>	The Upper Gastro-Intestinal Tract (oesophagus, stomach and duodenum)



The Digestive System (courtesy of Wikipedia commons, ladyofhats)



Structure of the Oesophagus (image by K Bucher)

WHAT IS ACHALASIA?

In a healthy oesophagus (about 20 cm long and around 1.5 – 2 cm wide in an adult), a bolus, or lump of chewed food, is propelled towards the stomach by a set of muscles that contract and relax in sequence. The muscles above the bolus contract, and the muscles below the bolus relax, creating a chain reaction that sends the food into the stomach after it passes through the lower oesophageal sphincter (LOS). The LOS should relax to allow food through, but then close to prevent stomach acid rising into the oesophagus, as part of this normal digestion process. There is a story of a lecturer at medical school who would demonstrate this process of *motility* by drinking a glass of milk whilst standing on his head. The milk would progress into his stomach despite the effect of gravity! In a healthy digestive system, the oesophagus is an active organ that needs to join with the stomach at a good angle (*the angle of His*). Saliva should act as a good lubricant,

and the stomach and small bowel should also empty well in a good digestive system.

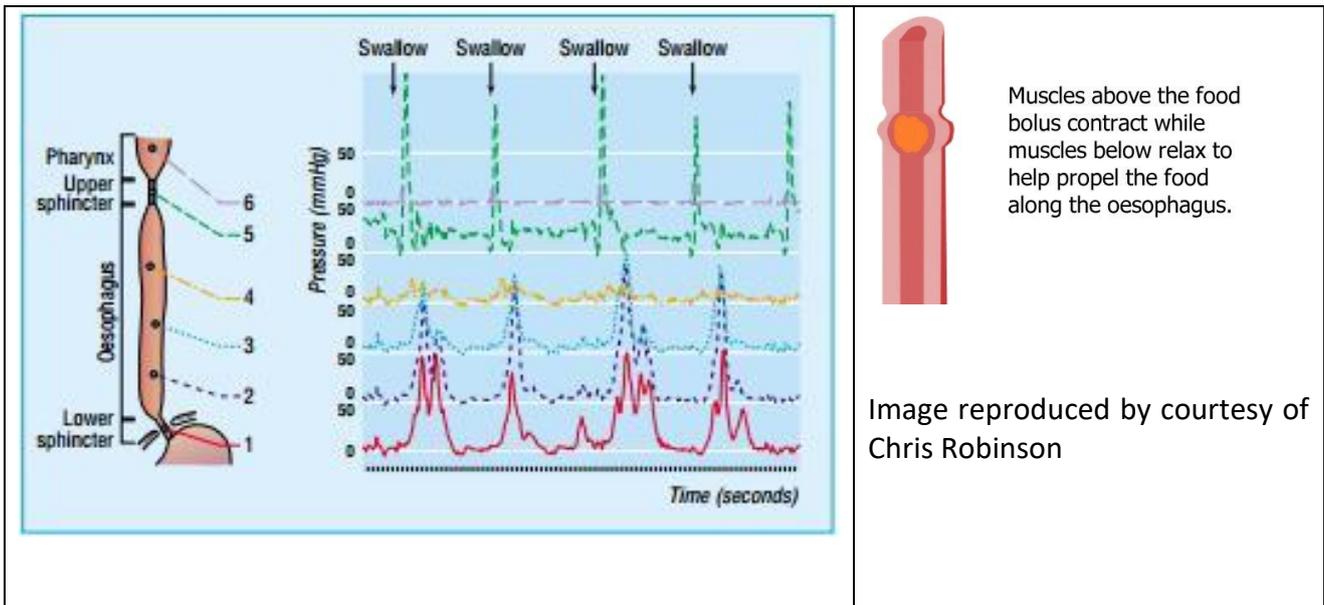
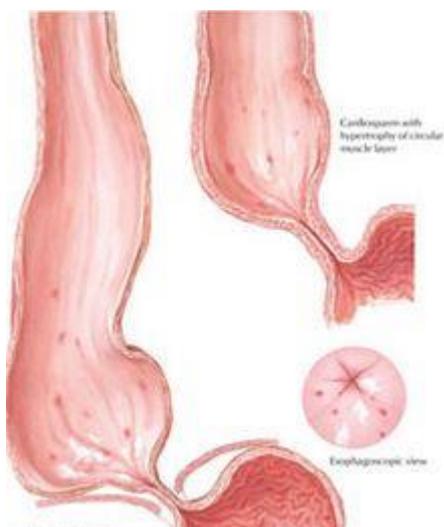


Diagram showing pressure of muscles at various points in oesophagus

The muscles that govern motility are normally automatically controlled by a complex, interlinked nerve system, and the cause of achalasia appears to be related to malfunction and loss of the inhibitory nerve cells (ganglia) controlling the muscles involved in peristalsis, rather than the muscles themselves. Loss, over time, of *cholinergic neurons* (that use a neurotransmitter *acetylcholine* to send messages to the brain to control part of the digestive system) can lead to loss of muscle tone and dilation – resulting in a big, baggy oesophagus that collects food in a pool surrounding the LOS rather than channelling food into the stomach.



Food starting to 'pool' at base of oesophagus



A 'bird's beak' image of an LOS preventing efficient passage of food

The escalating problems of not eating well, starting to miss out on a balanced diet, the effect of stagnant food on the taste buds and appetite, and the body getting 'trained out' of accepting the food and the nutrition it needs, can lead to problems that include frustration, low mood, oral thrush / oesophageal candidiasis, severe weight loss and/or malnutrition.

Achalasia is a disorder of the oesophagus where there is failure of the normal mechanisms that propel food down towards the stomach. It typically involves the muscles in the oesophageal wall and also the muscles in the lower oesophageal sphincter. The contractions in a normal oesophagus are sequential, starting from the top down, but in achalasia the contractions are uncoordinated and often occur simultaneously. When they do occur, the contractions are weak. Sometimes they are absent. The lower oesophageal sphincter (LOS) is also abnormal - this is the 'valve' between the oesophagus and the stomach – and in achalasia this fails to relax and so does not allow food to pass through into the stomach. The oesophagus thus becomes dilated and 'baggy' in achalasia. Food can therefore go down past the throat immediately after being swallowed, but it then gets blocked at the level of the lower oesophageal sphincter. This can lead to discomfort in what feels like the chest area. The food can remain there and build up, and it can start to putrefy, leading to feelings of nausea, and then regurgitation back up into the throat. The muscles can go into a spasm, leading to great pain the chest area, and this pain is sometimes very difficult to differentiate from that of a heart attack.

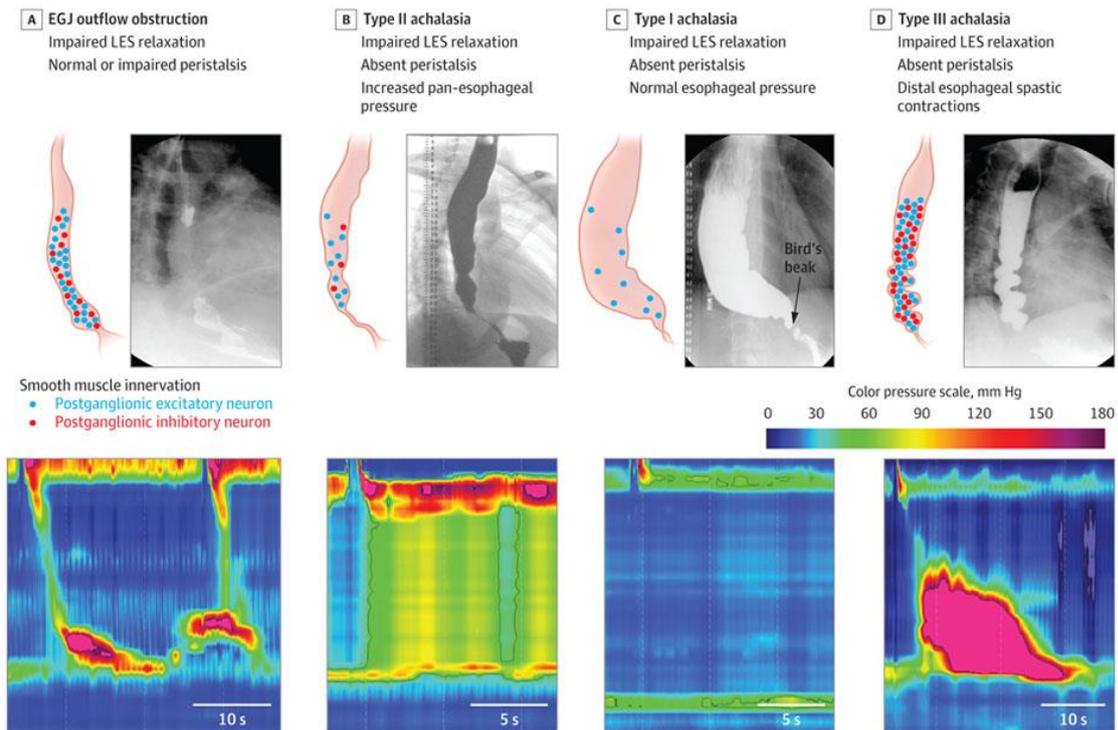
Achalasia typically involves some or all of:

- a non-relaxing LOS,
- poor muscle contractions in the oesophagus
- Simultaneous and/or badly coordinated muscles contractions in the oesophagus
- Simultaneous areas of high and low pressure in the oesophagus without relaxation of the LOS
- The oesophagus altering shape because of the weight of stuck food, leading to part of the oesophagus bulging below the LOS and letting food form a pool rather than falling into the stomach.
- Difficulty with maintaining good nutrition and body weight

- Difficulty with medication designed to be absorbed from the stomach and lower parts of the digestive system.
- Irritation and/or inflammation and/or infections of the oesophagus
- Malnutrition and its consequences
- Fatigue
- Tension and anxiety
- As well as swallowing difficulties and regurgitation, a person with achalasia might present a doctor with symptoms such as reflux, pain, respiratory problems, laryngeal problems, pneumonia caused by aspirating reflux, post nasal drip, nocturnal cough, a weak voice after an hour, and becoming tired after talking loudly.

Some medical authorities refer to different types of achalasia:

Classical, Type 1:	No peristalsis (lack of muscle contractions, and lack of LOS relaxation)
Compression, Type 2:	Problems involving pressurisation within the oesophagus (Ineffective, uncoordinated peristalsis and lack of LOS relaxation)
Vigorous, Type 3:	Vigorous, causing chest pain (high amplitude spikes, and uncoordinated vigorous contractions, with lack of LOS relaxation)



Charts showing diagrams of different patterns of achalasia, pressures, and different sorts of neurons that can become unbalanced between excitatory and inhibitory types.

The underlying cause of achalasia is not known for certain, but there is speculation about contributory factors being viral infections, implicated being the *varicella zoster* virus (linked with chicken pox and shingles), measles, a possible genetic predisposition in some, general association with GI motility problems, or an autoimmune process.

DIAGNOSING ACHALASIA

Swallowing problems can have a variety of different causes, but there is, regrettably, often a long delay before patients are diagnosed with achalasia. A meeting of patients in 2015 found that the period between initial problems and final diagnosis ranged from three months to twenty years. Diagnosis is often made by a gastroenterologist or surgeon at a hospital specialising in Upper GI conditions. The tests undertaken are:

- **Barium Swallow** - a white liquid is swallowed by the patient whilst being monitored in an X-ray machine that tracks the progress of the liquid
- **Endoscopy** – a miniature camera on a flexible tube is passed down the patient's throat, often after a sedative, to provide a visual examination of the inside of the oesophagus and stomach. It is important for establishing whether there are other physical causes for blockages. Endoscopy can reveal conditions like Barrett's oesophagus, ulcers or tumours. The technique can also be used for taking biopsies. An endoscopy may reveal evidence of achalasia, showing a dilated or baggy oesophagus, a tight lower oesophageal sphincter, or pooled and held up food and fluid residue with associated oesophagitis (inflammation of the lining of the oesophagus).
- **Manometry** – a flexible tube is inserted into the oesophagus that measures pressure at rest and during swallows at up to 2 cm intervals from pharynx to stomach. Although uncomfortable, the test does provide quantitative information on the contractions (or their absence) in the oesophagus at rest and also during swallowing, and it also provides a reading of the pressures in the lower oesophageal sphincter, but individuals may not necessarily require this test in all cases. The test is usually done without any sedation as an important component is measurement of the pressures during swallowing so patients need to be awake to swallow safely. Sometimes the catheter cannot be passed through the lower oesophageal sphincter (which in achalasia is closed at abnormally high pressures). Modern high resolution equipment is much more comfortable for the patient. A 24-hour monitoring process is sometimes used to test for acid (pH) reflux into the oesophagus: this involves the patient going home with the tube in place and a monitoring device strapped to the waist.
- The results of these tests are all put together and with the history and examination they should give the doctors a picture of the swallowing

process and where problems are occurring. A thorough diagnosis is important for indicating what treatment is likely to have the best, long term benefit. Do remember that sometimes the diagnosis may be a condition other than achalasia and so it is important that the diagnosis is supported by these tests. An endoscopy is essential in all cases to rule out other causes of holdup to food. The other tests are not always essential, and can be carried out in various combinations.

Stress, tension, anxiety and fatigue can contribute to the nervous system not acting effectively in operating the muscles involved in motility. Effective digestion sometimes takes time and requires the body to be relaxed, something that the modern world does not always easily allow us. If a patient has significant stress, the inability to take in nutrition may well make achalasia worse, and a downward spiral of deteriorating health can develop. Stress also accentuates the perception of one's symptoms and also their sequelae (medical consequences).

As with many other conditions, achalasia may be accompanied by one or more other problems.

Because achalasia can lead to regurgitation, weight loss, fatigue, loss of nutrition and reluctance to eat certain sorts of food, it can sometimes be confused with eating disorders such as bulimia, particularly if the patient is young.

Chagas disease can mimic achalasia. Chagas disease is a tropical disease caused by triatominae insects, mostly in Mexico, central and south America, that can cause damage to the nerves in the oesophageal wall and can lead to enlargement of the oesophagus in much the same way as in achalasia.

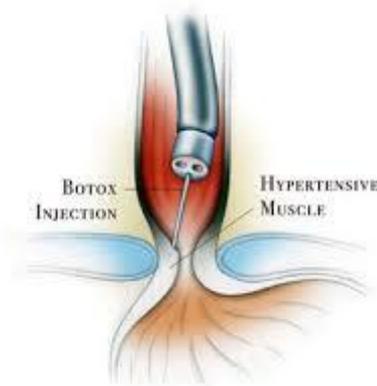
Pseudoachalasia is a term given to describe dilatation and failure of effective peristalsis as a result of obstruction at the lower end due to conditions such as cancer or a gastric band

TREATMENTS

These range from lifestyle changes to surgery, and a good doctor will treat the patient rather than being governed unduly by the test results. There are generally two parallel strategies in the treatment for achalasia. First is the medical treatment: the avoidance of stress, anxiety, remedy of nutritional deficiency, treatment of resulting chest infections or candida overgrowth and infection, and relief of symptoms as much as possible. In parallel, treatment is also directed at relieving the obstruction caused by the non-relaxing sphincter. The first course of treatment provides the best shot at obtaining sustained relief – repeated treatments each become more difficult technically, and with each treatment the chances of a good sustained longer term outcome diminish. So it is important to choose the initial treatment carefully to match the individual patient's particular circumstances – age, fitness, and disease severity.

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. Some patients have described injections that did not result in symptomatic relief.

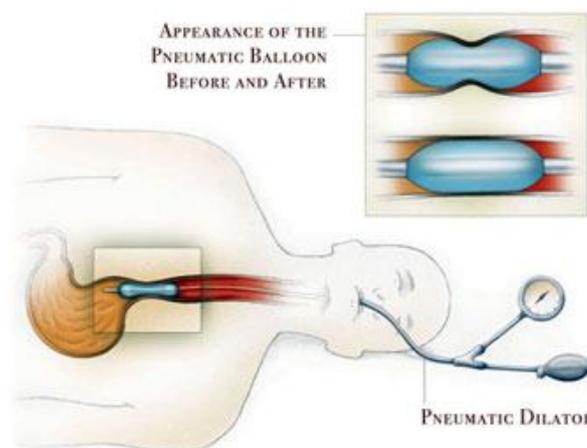


Botox injection (image courtesy of Memorial Hermann)

Dilatation

A pneumatic balloon-like device is passed through an endoscope and then inserted into the LOS and dilated to stretch the LOS. Results are variable, with most

procedures having some partial and time-limited effectiveness. Success is more likely when the oesophagus is less dilated. 15% of patients experience a certain amount of chest pain and there can be up to a 5 - 10% perforation rate, depending on the skill of the person conducting the procedure and the individual characteristics of the patient. It can be carried out with smaller 20mm balloons, often as a day surgery procedure, but a formal dilatation requires a larger balloon (40mm diameter) to provide a more sustained disruption of the lower oesophageal sphincter and this is performed under a general anaesthetic. Repeat dilatations are often required. There is usually at least some relief of symptoms but most patients require at least one repeat dilatation some weeks later, and again in the longer term. Multiple repeat dilatations at intervals of over three months are feasible, but repeat dilatations can make the LOS fibrous and this makes subsequent surgery more difficult and less effective. A dilatation can bring benefit for many years, but for others the benefits may last only weeks or months, whilst others may have little benefit at all.



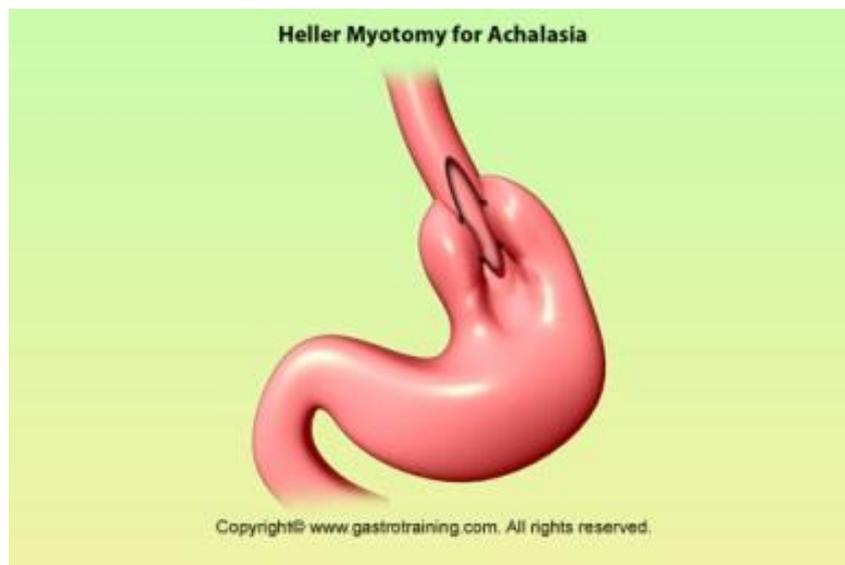
Dilatation (image courtesy of Memorial Hermann)

Heller Myotomy and Fundoplication

A myotomy procedure cuts the muscle in the LOS and the lower oesophagus and reduces the pressure that keeps the LOS closed, thus removing the obstruction to the passage of food. A normal LOS should allow food to flow through to the stomach, and also act as a competent valve to prevent stomach acid rising. Cutting the LOS leads to loss of most of this competence, so the looser the LOS, the easier food will flow, but the more likely the patient may suffer from acid

reflux from the stomach. Because of this, a myotomy on its own can result in troublesome reflux and so ideally it is always accompanied by a fundoplication. The myotomy procedure may take an hour or so, including all the necessary checks to make sure it is complete. The fundoplication takes about another twenty minutes.

The myotomy is carried out under a general anaesthetic and by means of a laparoscope through five small cuts measuring 5mm to 11mm. In experienced hands it is a very safe procedure and is all under direct vision with clear views of the lower oesophagus and stomach and with full control of the depth and extent of cutting throughout. Patients are encouraged to drink soon after recovery from the procedure. They require one or two nights' stay in hospital to ensure that they can tolerate fluids and a soft diet adequately.

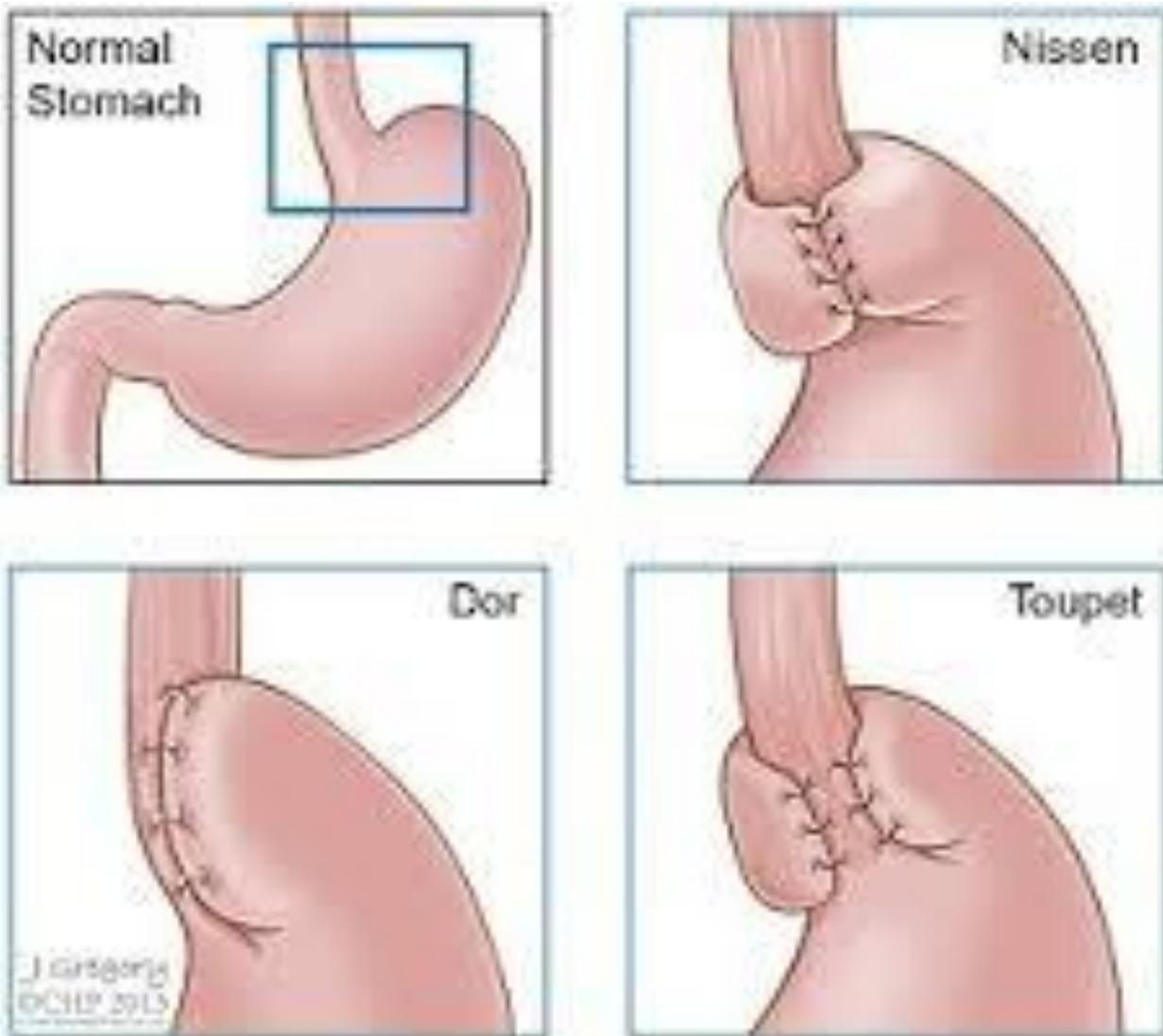


Heller Myotomy with a 'posterior' fundoplication (© Gastrotraining)

The fundoplication procedure wraps part of the top of the stomach (the fundus) around the base of the oesophagus with an aim to replicate nature's intended valve-like effect to reduce reflux. The fundoplication is usually partial, as opposed to a full 360° (that is the whole circumference of the oesophagus). Surgeons have gradually developed variations on how best to manage the extent of fundoplication, each depending upon the condition of the individual patient (Toupet 270°, Dor 180° - 200°, and Watson 120°). A fundoplication can be tightened later if it loosens over time. Too tight a wrap can cause too much restriction.

PPI medication (eg Omeprazole) can be taken to reduce acid reflux. There are side effects on the bacteria in the gut for this medication, and potential issues linked to osteoporosis, but these would be unlikely to be a factor in the short term, compared to the benefits of relief of reflux symptoms.

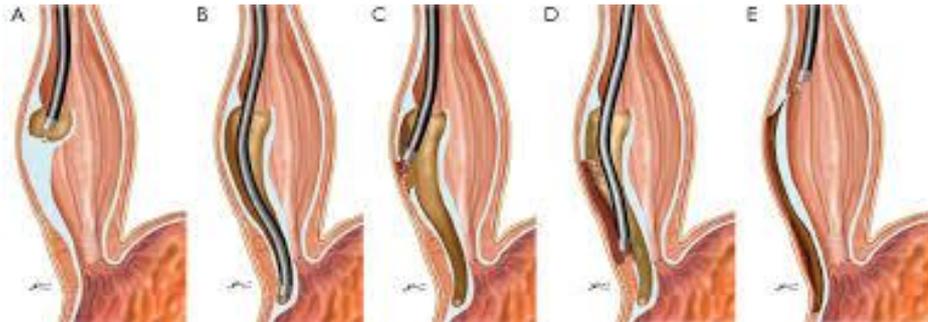
This myotomy surgery is well established and often provides sustained benefits for the longer term. A trial conducted by Pelligrini and others in California involved 168 patients, 29% of whom had an oesophagus with a diameter greater than 6cm (2-3cm is normal). 90% of the patients had good or excellent results from the surgery. Other studies have shown that this is sustained over the longer term.



Fundoplication methods (© J Gregory, Mount Sinai Health System)

POEM

POEM (Per Oral Endoscopic Myotomy) is performed through an endoscope that takes several hours and requires a general anaesthetic. The muscles of the oesophagus and the LOS are cut. It is a new procedure that has not been performed in large numbers and the mid to long term results are not known. It does not allow formation of a fundoplication and so the ensuing reflux requires PPI acid suppressing medication.



Endoscope entering the lining of oesophagus (POEM)

(©Inoue H. Showa University Northern Yokohama, 2010).

The best path is to aim for a conclusive diagnosis as early as possible and then to choose the procedure that will give greatest benefits for the long term.

Medication

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 can be used as a temporary bridge pending other treatment.

Oesophagectomy

Very much at the extreme end of experience and a last resort for achalasia sufferers, 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 operation is most often carried out in relation to cancer sufferers. It 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, but patients do achieve a reasonably good quality of life after this surgery.

Esophagectomy

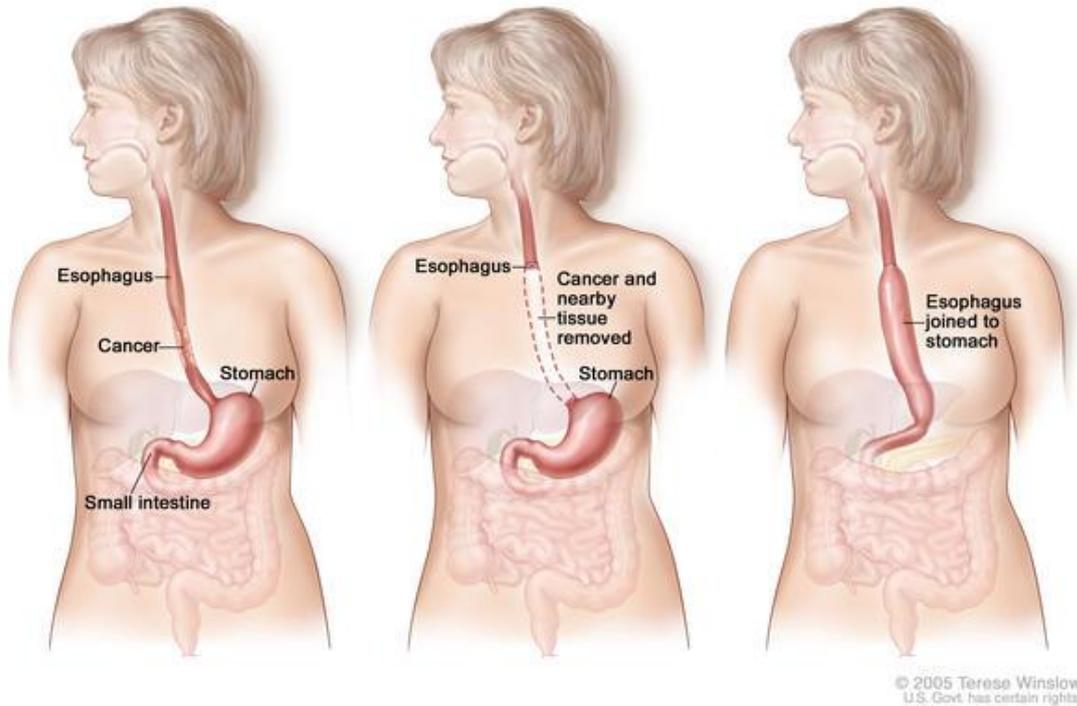


Diagram of an oesophagectomy (© Terese Winslow; St Francis Care)

WHAT TO EXPECT FROM TREATMENT

Many people with achalasia have a poor quality of life around food and will also be suffering the sequelae of obstruction and regurgitation. Despite an understandable reluctance to undergo surgery, the risks and the benefits should always be balanced against each other - temporary discomfort following a 1 - 2 hour operation and several days of recovery against the 80% plus likelihood of significant improvement in symptoms. To some extent it is easier to try the simpler procedures (eg botox, dilatation) first but these treatments usually do not provide sustained relief and additional treatments are then required later. Surgery frequently improves matters, and often makes a dramatic difference, but the primary problems causing the achalasia will not have disappeared. Better digestion will be achieved largely because the digestive tract has been surgically adapted and the obstruction relieved so that gravity will cause food to make progress into the stomach. It will probably still be necessary to be careful about eating, to avoid rushing meals, to keep an upright posture, to avoid some foods, and to avoid eating late in the evening. Trying to remove stress, tension and anxiety will always have a beneficial effect: the surgery will not allow you to completely ignore that aspect of a busy lifestyle. So you will probably not be able to 'rush' eating again as you once might have done before you had achalasia!

After surgery you will be given instructions about having 'sloppy' food such as soup for a period, and you should then be able to progress to more solid food. Your nutrition and energy levels will probably improve significantly and you should be able to resume many pleasurable aspects of life that have been denied to you by achalasia.

MEDICATION

This is a list of medication that you may come across. It is intended to help you, but is not a recommendation that you should try anything without being properly prescribed by a doctor or pharmacist.

Buscopan	Used to combat spasms in the gastro-intestinal tract
Omeprazole	A proton pump inhibitor (PPI) that switches off acid production in the stomach
Sucralfate	Used to treat reflux, and works by creating a protective coating in the lower oesophagus against acid, pepsin and bile
Gaviscon	Available over the counter. An alginate that creates a protective raft against all types of reflux
Nitrates	Nitrates improve blood flow and can relieve chest pain

HINTS AND TIPS

These quotes / snippets of advice have been gleaned from a large number of UK members of our achalasia support group between 2013 and 2016, and are shown for your information. But do be aware that we are all different, and what works for one may not work for all. Some of the points are expressed in people's own words.

General eating habits:

- Eat smaller meals more frequently – 'little and often'
- Eat slowly and chew well
- Beware of eating more when the first mouthfuls have not passed through into the stomach; it will simply pile up as further blockage in the oesophagus
- Don't eat too late in the evening e.g. after 8.30pm (when you go to bed, gravity will not be there to help your digestion)
- Keep a good posture with your back vertical.
- If going to a restaurant in the evening (for those who can), do not eat solids all day beforehand
- Beware of eating when feeling stressed
- Eat fairly moist food, and try drinking sips of water with food.
- Lifting the chest and taking a deep breath also helps
- Don't eat too many nibbles such as crisps and nuts, or drink too many glasses of wine before a meal in the evening (I know one should only have one or two glasses of wine but . . . !)
- Salads are best eaten with lots of dressing and in small quantities
- Be aware of the types of food you personally need to avoid, and what can be digested easily
- Basically it seems it is the consistency of the food which has more influence than anything
- Try mixing up textures of food as this makes them less likely to stick together.
- A little bit of spice can liven up a bland diet, according to each individual's tolerance.
- Grow your own vegetables or otherwise concentrate on young, tender items

Drinking:

- Don't have drinks too cold

- Don't drink before bedtime
- Always have a drink with meals: sparkling water can be beneficial, and gulps between every few mouthfuls help
- The most important factor in managing eating has been drinking hot water (temperature is important - half cold and half boiling). Mastering a technique which worked for me took time, but is worth it because I am now comfortable eating out and the only downside is the copious amounts of hot water I might need to drink to get the food down. The technique involves judging how much food I can eat before I have to gulp down some water.
- Actimel / milk-based drinks can be helpful (but probably this is because it is a soothing method of taking in nutrition rather than necessarily because they are milk-based)
- Coca cola, despite its unhealthy nature generally, can unblock the system (as happens for lemonade for people who are fitted with oesophageal stents), but fizzy drinks can also cause problems.

Food worth trying:

- A good start to the day is a fruit smoothie using a wide variety of fruit liquidised with a live yoghurt and probiotic (actimel) and a good teaspoon of manuka honey.
- With a good variety of nutrients early in the day, I don't have to worry about having to eat much else. Ironically, that then makes it easier to eat.
- Multigrain toast with Somerset brie is also a favourite, helped down with hot water.
- Soups are a good way of eating a variety of nutrients as they can be liquidised. I include all vegetables and pulses and experiment to get something I really like, sometimes topped with cheddar cheese.
- Fish - salmon or battered cod seem good
- Salads
- Stir-fry food is usually fine
- Funnily enough, quiche or similar is usually not a problem
- Cheese with crackers
- For dessert, ice cream is best
- Best foods were Weetabix, Readybrek, custard, sponge puddings and mashed potato. Not the most healthy range but I was advised by the dietician at the hospital that it was more important to keep my calorie intake up than eat healthily
- Manuka honey, fruit smoothies, soup and warm water all helpful
- Banana (but not for all), coconut milk and honey all good
- Warm cocoa or drinking chocolate good

- Adding gravy and sauces to help lubrication of digestion system
- Buttermint sweets can help
- Pineapple juice can help

Food worth avoiding (and see the chapter on Food that Causes Problems):

- Skins on fruit and veg, but do churn them up in smoothies and soups
- I avoid fatty meat and eat mostly chicken, fish or vegetarian dishes.
- I can even manage pizza if I drink enough hot water with it.
- I also avoid spicy food and drinking alcohol with food is very difficult
- Large lumps of meat. The only meat I consider eating is mince, usually in the form of a cottage or shepherd's pie
- Dry chicken can be a problem
- Meat in a sauce or casserole is usually better than anything else
- Pasta of any sort
- Too much bread. I can eat crackers better than bread.
- Don't eat too much bread in one sitting and eat good quality bread rather than soft white bread which is particularly bad for blocking the oesophagus
- Naan bread , buns or cakes that do not break up well . Water will not wash it through.
- Potatoes can be a problem if boiled but thin french fries are not too bad
- Rice. My first bad experience of achalasia was with a dish of paella. Fried rice is better than fluffy stuff.
- Spotted dick or similar dry sponge puddings are best avoided
- Avoid very dry food like falafels, raw cauliflower, raw carrot
- Peas (but mushy peas without skins might be OK?)

Keeping up nutrition:

- I was prescribed Fortisip milkshakes which were a lifesaver as they are full of vitamins and nutrients.
- Echinacea and Manuka honey to boost immune system
- Try suckable vitamins
- Monitor your vitamin and mineral levels as the effect of not eating well may have led to a serious shortfall in some areas that normal multi-vitamin supplements may not be able to cope with.

Medication:

- Absorption of medication may be a problem or even harmful if strong pills get stuck in the oesophagus rather than passing normally into the stomach. Pills are often designed to be absorbed from the stomach, with coatings that last for the normal duration period for reaching the stomach before the coating dissolves and releases the medication. Ask for advice from your pharmacy. Medication will often be available in liquid form. Some pills can be absorbed by placing them under your tongue.

General:

- Getting up and walking around during meals, or even jumping, can be helpful to encourage gravity helping digestion.
- Standing on toes and dropping on to heels sometimes jerks food through the system
- Massaging the chest area

Bedtime:

- Sleep propped up with lots of pillows (before surgery) to help stop food and drink coming back up at night with reflux
- A bed wedge is a useful alternative to lots of pillows to keep you propped up at night
- I always finish the day with a good teaspoon of manuka honey and lemon juice in hot water. This is after I have ensured as much as possible that all food has been washed down. That way I am left with manuka honey in my oesophagus overnight. I haven't had a cold for the last couple of years so I think it might have given me some protection
- Lying on right side found to be helpful for one individual (possibly because the oesophagus 'bags' in one direction rather than another?) – but most people find lying on left side better – less pressure on organs and better for reflux.

Spasms:

- Medications worth trying – nitrates, calcium channel blockers, Buscopan or Sucralfate
- Gently press downwards in the middle area of the chest
- Do deep breathing exercises
- Stretch upwards, including elongating your neck

- Relaxation exercises
- Try drinking warm water
- Try holding a hot water bottle on the chest to give pain relief
- Try eating a piece of banana
- Try dropping on to heels vigorously if food is blocking the LOS
- Keep a small bottle of water handy in case one's oesophageal muscles go into spasm when out and about
- Relaxation helps to avoid spasms and pain with the sphincter.
- I find yoga helps as does drinking hot water to relieve the pain.
- Pain from the sphincter can be avoided by warming up cold food and drink in the mouth first before allowing it to go down. Avoid letting the chest get cold. A cold breeze can set up pain: I wear a scarf even if I don't feel cold
- Pain may be related to fermentation and reflux rather than simultaneous contraction and spasms of muscles
- Controversially, some, in parts of the world where cannabis is allowed for medical purposes, report relief from pain, similar to those with some other medical conditions such as multiple sclerosis.

Avoiding stress and anxiety:

- The digestive system needs to be relaxed in order to work properly, but the inevitable anxiety about swallowing can make us tense, and make achalasia worse. Relaxing, and trying to eat in a calm environment can help matters.
- Avoid stress!

Sharing experiences:

- Communicate with other achalasia sufferers. It helps so much to know you're not alone!!

FOOD THAT CAUSES PROBLEMS

This is the result of an informal survey amongst nineteen achalasia sufferers at a meeting in July 2015, who placed plastic counters into beakers with labels according to whether that food / drink caused them problems or not. This gives a rough indication of food and drink that is likely to be troublesome (including spasms rather than motility issues), but bear in mind that not all people will have tried each item, and that some were at different stages and severity of achalasia than others. Some food might cause bloating or other digestive problems, and some people may have digestion issues in addition to achalasia. Some things may be difficult because they create a shock to the system (eg iced water); others because of how they react with the oesophagus because they are hanging around so long (eg spicy food).

The answers are consistent with a need to avoid :

- a) texture, stringiness or skins that will stick in the oesophagus;
- b) food that creates a heavy bolus or lump if it sits in the oesophagus and congeals (eg rice, bread and pasta)
- c) spicy food that causes extra irritation because it remains in the oesophagus to irritate the lining rather than passing through to the stomach where the acid contents can cope with the spiciness; and
- d) food that *might* contribute to spasms. A grey area, but cold items might do this for some.

Food	Score (max 19)	Comments (which may not be completely representative)
Bread	15	Especially cheap white bread. Multi-seeded better texture? Toast not so much of a problem. Sometimes can be shifted with lots of water.
Red Meat	15	Needs to be chewed well. Take tiny morsels. If stringy can clog the lower oesophageal sphincter. Avoid gristle and fat. Even mince can be a problem for some. Go for very lean meat, and/or slice very thin
Apple	15	The skin seems to cause the problem, but also the flesh inside. Sharp edges cause pain in throat. Stewed apple OK. Try grated apple with cinnamon?

Rice	12	The variety of rice can make a difference
Potato Chips	12	
Cakes	10	Cakes, sponge and scones can cause pains the following day (fermentation / colic??)
Spicy curry	9	Yoghurt can be added to reduce spiciness
Pastry	8	Needs to be chewed really thoroughly, and sometimes washed down with lots of water.
Pineapple	8	Pineapple juice could be used instead
Pasta	7	It sticks, like some other carbohydrates, causing a ball in oesophagus. Thin noodles in a stock go down well, but thick pasta sits more heavily. More sauce might help.
Salad	7	But be careful about ingredients
Lettuce	7	Sticks in oesophagus. Crisp lettuce might be preferable. Sometimes heavier items like meat take the green salad through with it.
Boiled potatoes	7	Mashed with butter or cheese can be good, and softened with milk / water
Risotto	6	Can depend on what is in the risotto with the rice. Too 'gloopy' and tends to bounce back. Vegetables like carrots, mushrooms etc can improve the overall texture
Falafel	6	
Chilli	7	'Insides on fire for days'
Toast	6	Sometimes better than soft white bread
Tomatoes	5	Skins are a problem. Can cause coughing. Removing skins can improve things. Small pieces of uncooked tomato can cause blockage.
Raw cauliflower	5	Score might have been higher if more people had tried to eat this!
White meat	5	Tends not to be so stringy as red meat
Banana	4	
Onion	4	Can stick. Causes reflux
Breakfast cereal	3	Weetabix found by some not to have enough 'weight' to go down. Muesli can be easy to swallow but still cause difficulty further down the system
Runner beans	3	But beware potential stringiness
Carrot	3	
Fish	2	Beware bones

Yoghurt	2	
Nut roast with gravy	2	Can be too soft and stick in a ball in oesophagus
Soup	0	But some cannot cope even with soup when feeling stressed.
Drink		
Wine	9	Starting meal with wine can lead to the oesophagus closing a little and making eating more difficult. Starting with water is better, leaving wine until later.
Fizzy drinks	8	All right some of the time. Some people have never been able to drink them. Beer can have same effect. Can lead to bloating. Swallowing not the problem so much as after-effects / reflux. <i>San Pellegrino</i> carbonated water seems not to be so gassy as others and can help digestion.
Iced Water	7	Warm water better than very cold or hot water. Icy water can cause oesophageal spasms.
Orange Juice	6	Can cause pain from wind / colic?
Coffee	4	Sometimes this just won't go down, for no obvious reason.
Warm water	0	

FREQUENTLY ASKED QUESTIONS

(Based on meetings held in December 2013 and March 2016
with Mr Majid Hashemi)

Is achalasia stress-related? – Yes, there seems to be a link.

Is it usual for someone who has had a number of dilatations to have blood at the back of the throat for a few days? Yes, because muscles are sometimes ruptured, but blood should only be apparent for a few days at most.

Why do some people with achalasia get bile reflux? The sphincter valve(s) can remain open and allow bile from the gallbladder to rise in the digestive system. A hospital can test whether reflux is bile (an alkali) or stomach acid.

Are there standard guidelines for achalasia management? – Very scattered practice. Diagnosis and pathway is standard – but treatment practices are variable

Does achalasia get better? Yes, from time to time. It shouldn't get worse if the oesophagus and other factors (eg stress levels) remain the same

When would be the best time to have the operation? It is always best to try for the conditions to maximize chances of success first time round, as there is rarely a good second chance. Dilatation occurs in about 40% of cases in centres of excellence, but repeated dilatations make it harder to achieve success further down the line, eg with Heller's Myotomy.

How can we avoid spasms of pain that occur randomly?– Buscopan, nitrates, calcium blockers. Often gets better over time. Relieve obstruction downstream (ie avoid constipation)

What is the cause of the retrosternal pain (the one that feels like a heart attack, not reflux)? Sometimes the muscular contractions work in an uncoordinated spasm, causing the pain. It is hard to treat but will come to an end, and is not in itself harmful. Buscopan, eating a piece of banana, drinking Actimel, taking manuka honey, coconut milk, fizzy water, all can help. But chest pain for other reasons also has to be excluded!

Having had a *Dor Fundoplication* wrap (a variation of surgery to wrap part of the stomach round the base of the oesophagus) – is there any reason why a POEM procedure could not be carried out? You cannot have a POEM after Dor wrap.

Heading towards 3rd myotomy (50/50 results – trying to find out why it didn't work) are there alternative procedures - and what is the future going to be like with this condition? – Need to get it right first time, but there is hope. Sometimes it may be that the myotomy is not at the appropriate depth, but each case is individual.

Does dilatation sometimes make the pain/swallowing worse? No!

How likely is it that subsequent dilatations will be needed? Very likely.

Oesophageal candidiasis (inflammation of the oesophagus with candidiasis) How can we avoid it? What is the best way of curing it and stopping it from recurring? Need to get it tested so that it can be treated. See also Tips for Oral thrush as they may help. You don't necessarily get oral thrush at the same time as oesophageal candidiasis. Medication such as antibiotics affects bacteria in the gut.

I have suffered achalasia and had Heller's myotomy– but it hasn't stopped the symptoms? Have a simple reinvestigation with manometry, endoscopy and barium swallow. Quite often if it hasn't worked, it might be that the myotomy is not long enough or the fundoplication is too tight or too loose.

How is the type of treatment that is likely to work best for someone decided? In consultation with that patient, taking into account the test results and clinical judgement.

Could doing nothing except being careful with what food is consumed, and how, be the best solution for some individuals? Can be fine, but needs monitoring.

What should one do if food gets stuck and you can't throw it up? Suggestions – massaging chest can help food go down. Lots of warm water to drink can help.

How to cope on holiday? The OPA have printed out cards in various languages explaining that we have a medical problem with eating and that we can only manage small portions. OPA tel no. 0121 704 9860

THRUSH / CANDIDIASIS

Tips on how I have coped with Oral Thrush

Please note these are from an achalasia sufferer who had candida in her oesophagus long term before surgery. Since surgery, there has been no recurrence of the candida.

- For a month at a time, I cut out all dairy foods, alcohol, sugar (particularly important), mushrooms, melon, grapes (the last three tend to harbour mould very easily).
- Best to follow a vegan diet but make sure any nuts you eat are reasonably fresh. I store nuts for the muesli I make, in the freezer, as nuts can get an invisible mould.
- Watch invisible mould in left-overs, particularly rice (don't keep cooked rice longer than 24 hours). Nothing more than 48 hours – bin it!
- No yeast-based products, such as bread, beer, wine.
- Vodka is OK as a spirit.
- Eat organic where possible.
- Good quality French cheese is good for candida if it has bacteria as these are not fungal and are actually helpful for candida.
- Can have yogurt.
- Citric Acid can help – very concentrated and tastes foul. X drops in water and gargle with it. You can swallow a little bit to help kill off the fungal infection.
- Cortisol mouth wash can help if it is painful.
- Vitamin B supplement (not yeast based).
- Get a decent candida cookbook for recipes for candida (see *Further Reading*)
- Anti-fungal *Oreganol* can also help with oral thrush – taken at breakfast and evening meal
- No coffee or chocolate.
- Take good probiotic breakfast, lunch and evening meal (preferably purchased from Metabolics) to build up immune system
- Also take good quality Zinc – breakfast, lunch and evening meal also obtainable from Metabolics.
- Ionic silver (2 teaspoons) taken before breakfast
- Watch too many chemical cleaning products. I changed from treating swimming pool with chlorine to non-chlorine based product.

NB I took all these supplements only when the Oral Thrush was at its height. Normally, I just take one zinc and one probiotic last thing. I have been clear of candida for years now but I DO have a very small chronic ulcer near the valve with the stomach.

Ann Elms / 18.12.2014

[NB longer term use of probiotics beyond two months is generally not advised]

SOURCES FOR FURTHER HELP AND INFORMATION

Oesophageal Patients Association - OPA

Formed 1985. 90% of members have had oesophagectomy and gastrectomy, most often from cancer, but other conditions also included. They produce information booklets and run support group meetings.

22 Vulcan House

Vulcan Road

Solihull

West Midlands

B91 2JY

Helpline 0121 704 9860 Email: Amanda Ladell amandaladell@yahoo.co.uk Alan

Moss alanmoss32@gmail.com or Majid Hashemi

majid.hashemi@btopenworld.com

www.opa.org.uk Achalasia page: www.opa.org.uk/pages/achalasia.html

The Achalasia group meets once or twice a year at the Hospital of St Elizabeth and St John, 60 Grove End Road, St John's Wood, London NW8 9NH. People come from London and sometimes considerable distances from other parts of the UK.

London Achalasia Meetup Group

<http://www.meetup.com/achalasia-104/>

Sign up with Meetup to get notifications about the big OPA Achalasia meetings as above and the more informal monthly meetings in central London held typically on a Sunday at 2pm at the Marriott Hotel, 140 Park Lane, London W1K 7AA.

Their internet forum is at <http://achalasia.mooo.com/index.php> with threads on Events, Food Issues, Symptoms, Treatments and General Discussions.

Contact Amanda Ladell if you have any questions about the Achalasia Meetup Group or Forum. amandaladell@yahoo.co.uk

Oesophageal Achalasia and Spasms Forum on www.patient.info

<http://www.patient.info/forums/discuss/browse/oesophageal-achalasia-and-spasm-1655>

Forum specifically for achalasia sufferers, but the threads can be long and varied.

HealthUnlocked (Internet forum for questions and blogs):

<https://healthunlocked.com/oesophagus>

(300 members, mostly after oesophagectomy, occasional medical staff, some from abroad) with ability to search on words eg *achalasia*)

In Germany (and elsewhere): Achalasie-Selbsthilfe (Achalasia Self Help)

www.achalasia-selbsthilfe.de (Some content is written in English) 386 members suffering from achalasia are in this organisation as at July 2015. There are also regional contacts. Email: achalasia.web.de

German language forum <http://www.achalasia.com/forum-neu>

They have contact points for Netherlands, Austria, Switzerland, Belgium, Spain and USA (available through their website or from alanmoss32@gmail.com)

Achalasia Awareness Organization

Worldwide organisation founded in USA by Nancy Lazarus for raising awareness of achalasia, with 259 UK members as at May 2015.

Email info@achalasiaawareness.org

<http://achalasiaawareness.org> <https://facebook.com/achalasiaawarenessorg>

<https://twitter.com/achalasiaaware> www.pinterest.com/AAORG

FURTHER READING

Reading medical journals can be a daunting prospect for the non-qualified, and one needs to be careful about placing too much importance on findings where only a small number of patients are included in research. They are written for people who are medically qualified and able to make discerning judgements. So whilst some examples are shown below, there will almost certainly be important articles elsewhere. The best course in cases of doubt is to seek advice from the doctor who is dealing with your condition who can put things in proper context for your own situation.

Minimally Invasive Surgery for Achalasia: An 8-year experience with 168 Patients
Marco G Patti, Carlos A Pelligrini and others *Ann Surg.* 1999 Oct; 230(4): 587
<http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1420907/>

Achalasia: A Systematic Review John E Pandolfino and Andrew Gawron *JAMA* 2015; 313(18):1841-1852 doi:10.1001/jama.2015.2996
www.ncbi.nlm.nih.gov/pubmed/25965233

Is it an Eating Disorder, or Achalasia, or Both? A Literature Review and Diagnostic Challenges Deborah L Reas and others.
Published online 22 July 2014 in Wiley Online Library DOI: 10.1002/erv.2307
<http://www.ncbi.nlm.nih.gov/pubmed/25053457>

Diagnosis and Management of Achalasia Michael F Vaezi and Joel E Richter
American Journal of Gastroenterology Vol 94 No 12 1999.
<http://s3.gi.org/physicians/guidelines/DiagnosisofAchalasia.pdf>

Candida Cookbooks

The following books have been found to be helpful to some extent:

Cooking for Candida - Recipes & Diet Plans, Using foods you CAN eat
Jo Hampton ISBN 0 9521544 1 2 (1996)

The Candida Diet Book Karen Brody
ISBN-13: 0-85969-761-3 & ISBN-10: 0-85969-761-4 (1997)

How to cope successfully with Candida the Drug-free Way Jo Dunbar-Lane
ISBN 1-903784-11-5 (2003)

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Much of this booklet has been drawn from talks given by the Achalasia Group Chair, consultant oesophageal surgeon Mr Majid Hashemi, from University College Hospital, London, who has given enormous support for our patient support group. His colleague, Dr Rehan Haidry, consultant gastroenterologist, has also been extremely helpful.

Amanda Ladell has composed many of the practical hints and tips from her own experience and from talking to countless achalasia sufferers. Ann Elms has also helped with this process, not least by summarising the meetings, and the section on Oral Thrush is a straight reproduction of the advice that she has written.

Alison Gardiner and Alice Champion, specialist Upper GI dieticians from St Thomas' hospital have also passed on useful advice.

We have attempted to contact the source and copyright holders of the images used in this book and believe that they have been available and attributed to the best of our knowledge and belief for the purpose of this booklet. We would be pleased to amend or correct any errors that have appeared.

In exploring the activities of other achalasia groups, we have been helped by Antje Krieger-Wehnsen of Achalasia-Selbsthilfe who travelled to London and gave us a copy of their German guidebook.

But this booklet is very much a collation of individual experiences from 'achalasia' who have come together to support and help each other through this unusual and distressing condition. This booklet will inevitably grow to accommodate future additions and amendments, and we hope that it will reach many others who suffer from this condition in the future.

We have made every effort to ensure that the information contained in this booklet is medically correct, but we take full responsibility for any mistakes that have crept in, and would be pleased to hear from any reader who notices material that should be changed, amended or expanded.

Alan Moss

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August 2016

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