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# A Patient's Guide to Achalasia



**OPA - The Oesophageal Patients Association  
and London Achalasia Meetup Group**

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# CONTENTS

	<b>Page No(s)</b>
Introduction	3
Glossary	4 – 7
The Digestive System Illustrated	8
What is Achalasia?	9 - 13
Diagnosing Achalasia	14 - 15
Treatments	16 - 21
What to expect from treatment	22
Medication	23
Hints and Tips	24 - 28
Food that causes problems	29 – 31
Frequently Asked Questions	32 - 33
Thrush (Candidiasis)	34
Sources for further help and information	35 - 36
Further Reading	37
Acknowledgements	38
Index	39

## 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](http://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

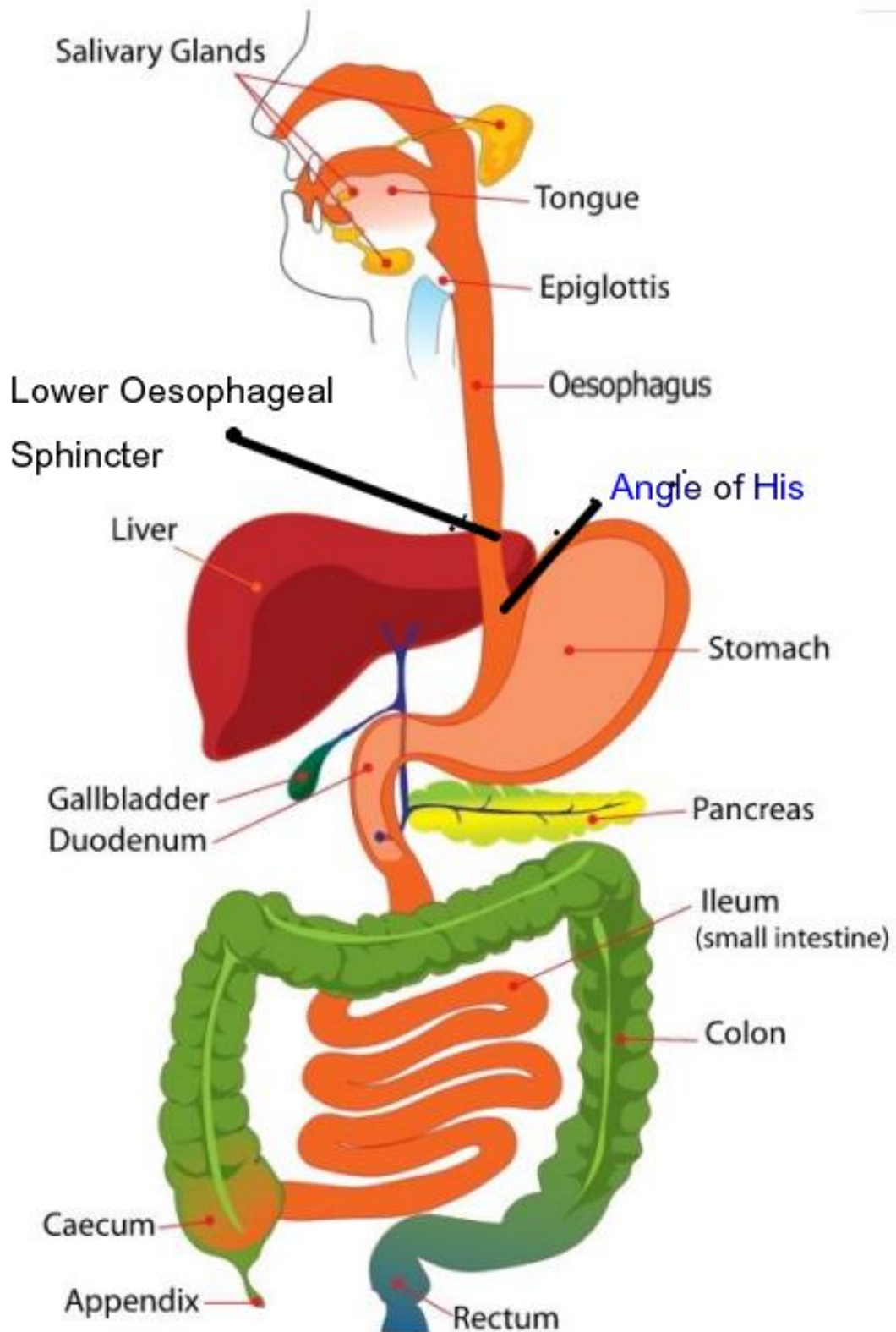
This glossary of medical terms appears here at the front of this booklet so that you will not miss it and will know where to look up if medical terms need a definition.

<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

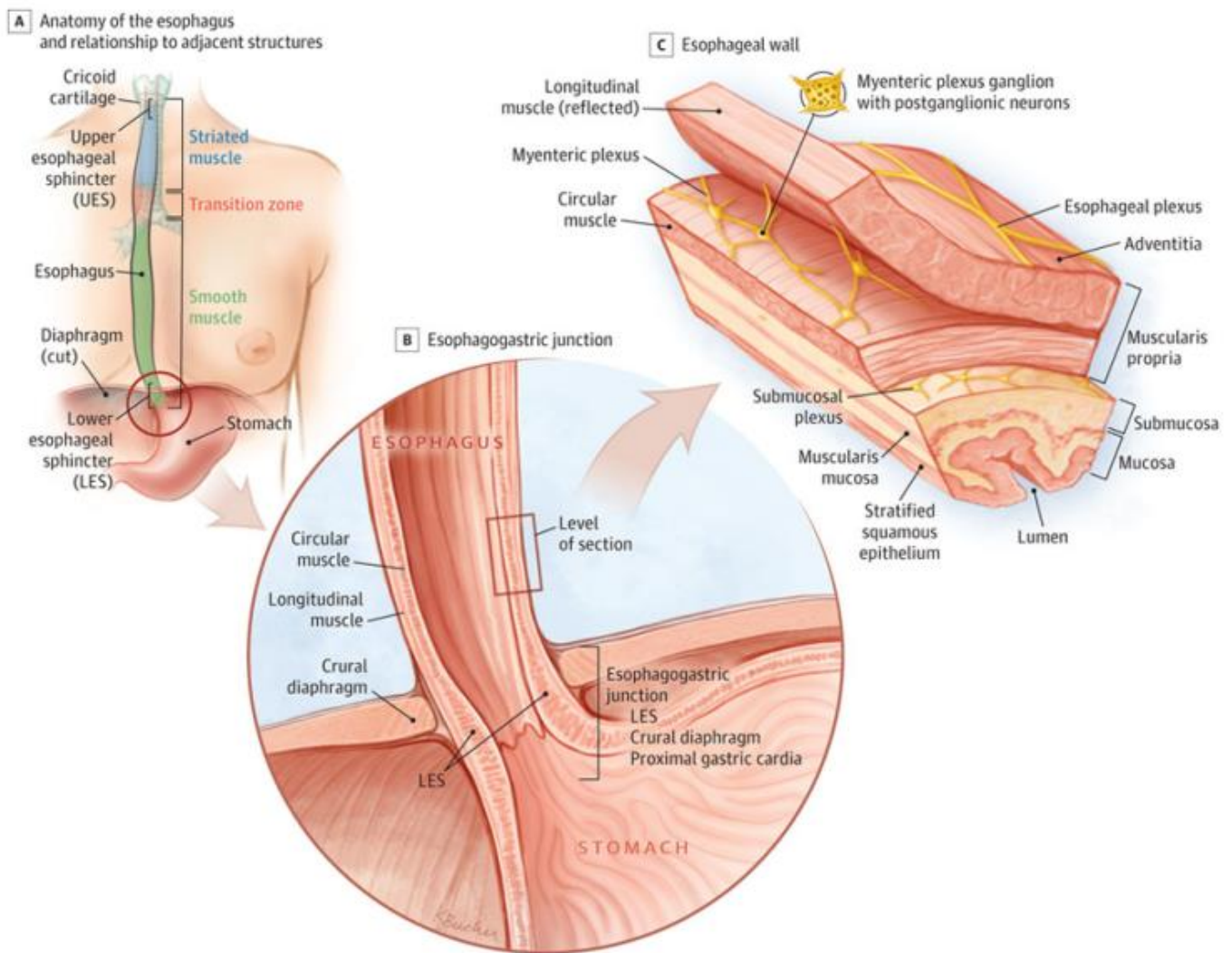
<i>Lower Oesophageal Sphincter (LOS)</i>	A sphincter valve near the abdomen between the stomach and chest that should allow food to pass downwards through to the stomach, but keeping stomach acid from rising into the oesophagus
<i>Manometry</i>	An examination involving a thin, pressure-sensitive tube 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)



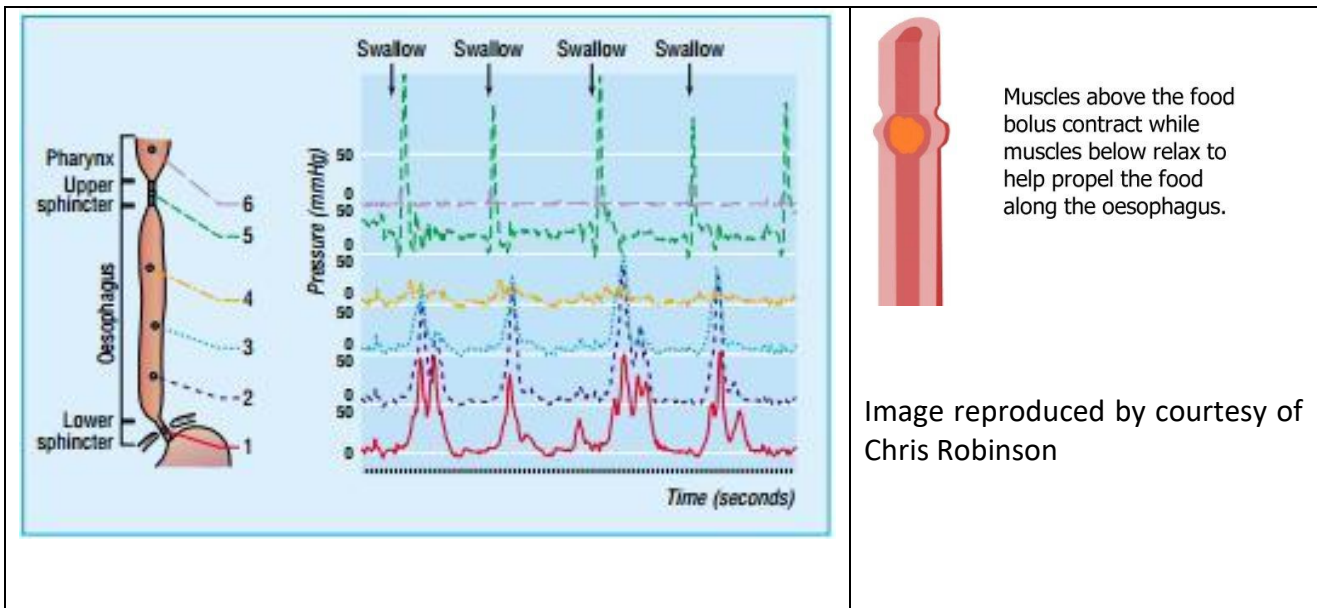


Structure of the Oesophagus (image by K Bucher)

## WHAT IS ACHALASIA?

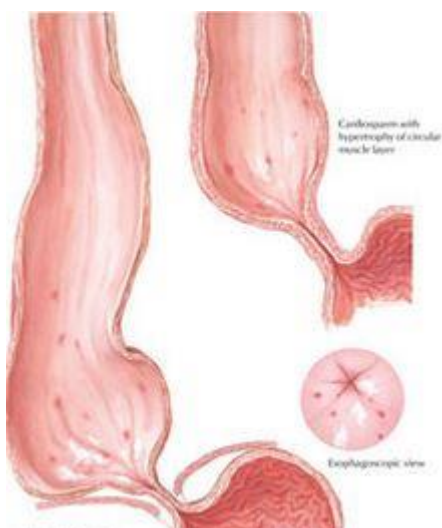
In a healthy oesophagus (about 20 cm long and around 1.5 – 2 cm wide in an adult), a bolus, or lump of chewed food, is propelled towards the stomach by a set of muscles that contract and relax in sequence. The muscles above the bolus contract, and the muscles below the bolus relax, creating a chain reaction 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.

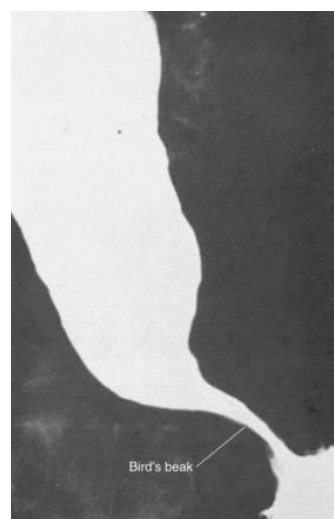


**Diagram showing pressure of muscles at various points in oesophagus**

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.

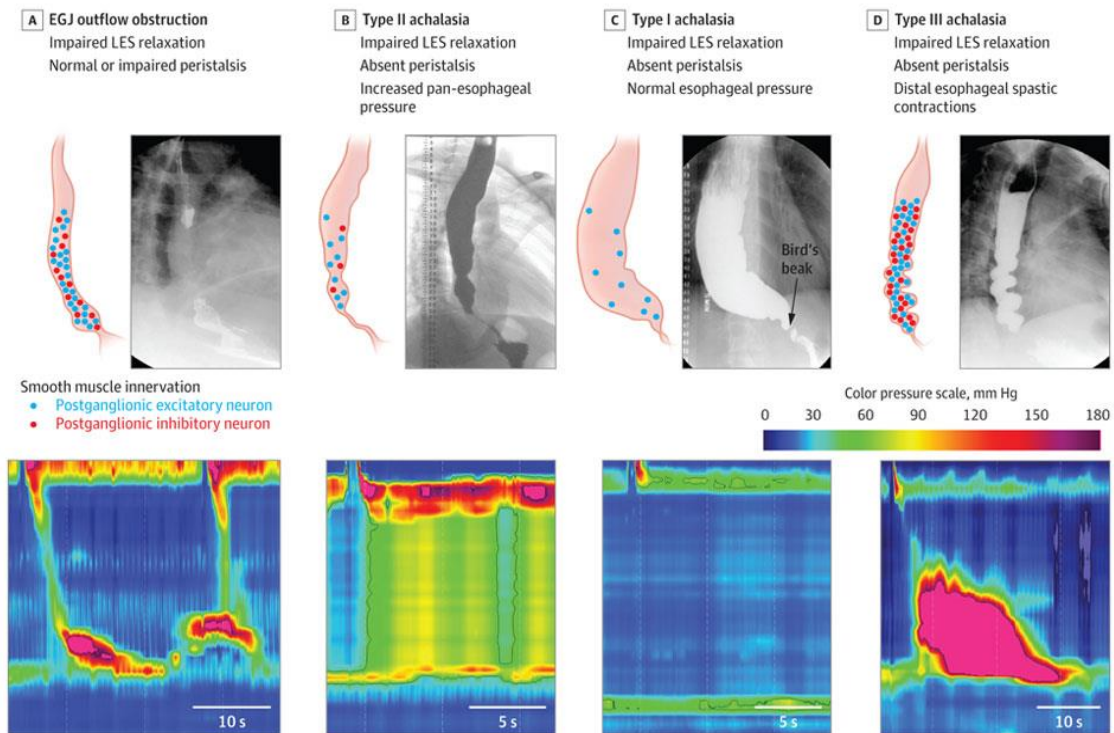
Achalasia typically involves some or all of:

- 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:

<b>Classical, Type 1:</b>	No peristalsis (lack of muscle contractions, and lack of LOS relaxation)
<b>Compression, Type 2:</b>	Problems involving pressurisation within the oesophagus (Ineffective, uncoordinated peristalsis and lack of LOS relaxation)
<b>Vigorous, Type 3:</b>	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

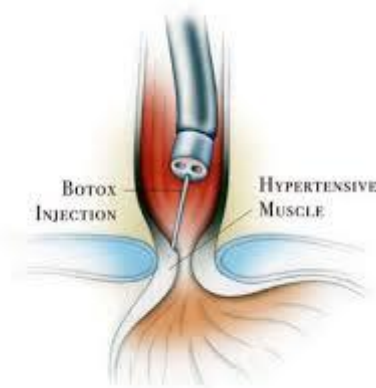


## TREATMENTS

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.



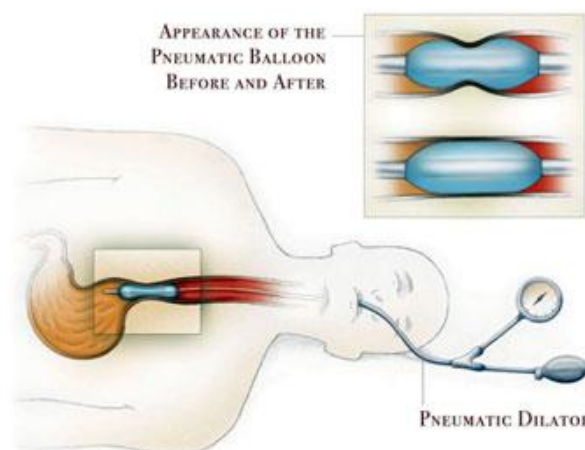
**Botox injection (image courtesy of Memorial Hermann)**

### **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

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