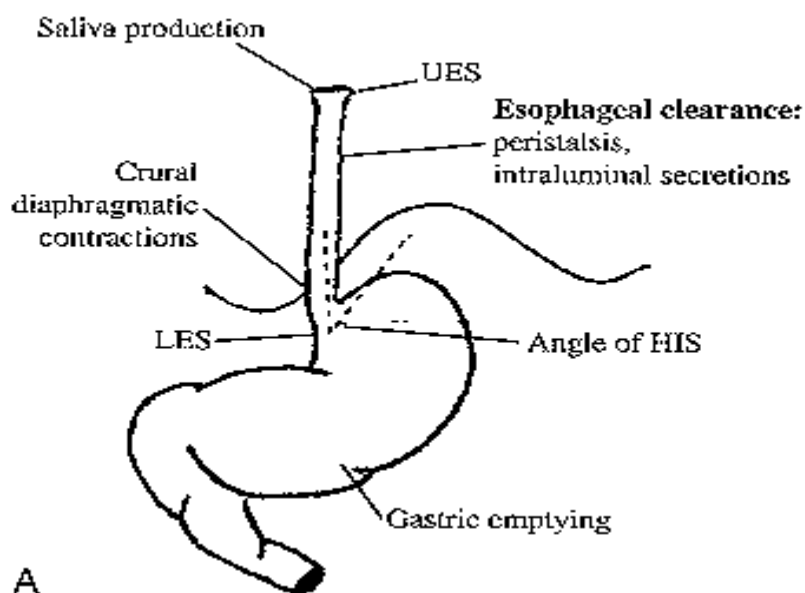


## QUESTIONS AND ANSWERS ON ACHALASIA - 21 JULY 2020 – via zoom

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- Kenneth Park, Senior Upper GI surgeon, Aberdeen and Director of Aberdeen Clinic
- Sheraz Markar, Senior Research Fellow, Imperial College with experience from USA

### General Overview



(UES = Upper oesophageal sphincter; LES/LOS = Lower Oesophageal Sphincter  
 Peristalsis = muscle contractions and relaxations that propel food through the digestive system)  
 LOS necessary to help prevent reflux

There are four manometric features common to different forms of achalasia:

- Non relaxing lower oesophageal sphincter
- High resting pressure in the lower oesophagus [making it tight shut]
- Poor or absent contractions in the body of the oesophagus [poor peristalsis]
- Simultaneous and badly-coordinated contractions in the body of the oesophagus

The nerve endings (ganglia) that control the actions of the muscles in the oesophagus do not work properly. Sometimes the cause can be a virus such as measles, chicken pox, shingles, the varicella zoster virus, or Chagas disease (mostly occurring in Mexico). There is occasionally a genetic predisposition to developing achalasia. There may be an auto-immune dimension. Nitric oxide, affecting blood supply and chemical signalling, may also be a factor.

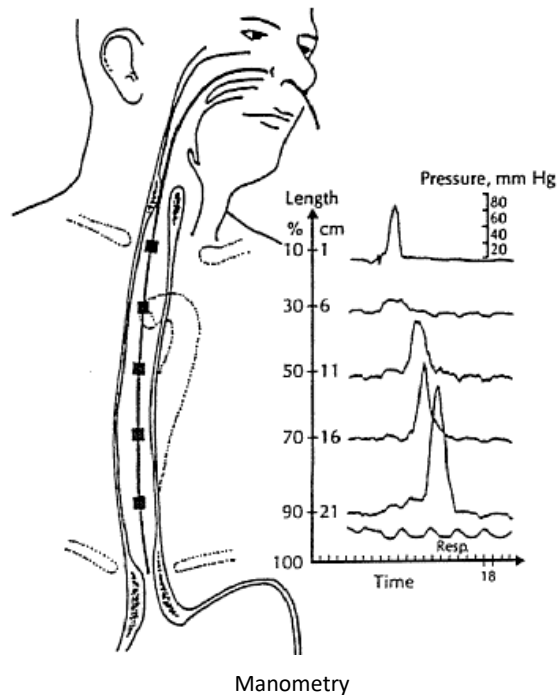
Those with achalasia complain of:

- Dysphagia [difficulty in swallowing]
- Regurgitation
- Reflux
- Pain in chest area behind breast bone or around stomach area.
- Respiratory problems [aspiration pneumonia when reflux gets into lungs]

- Laryngeal problems – hoarseness and loss of voice.

Diagnosis involves:

- History
- Endoscopy
- Barium swallow
- Manometry



Patients used to describe symptoms experienced for 5-7 years, but a recent trend towards two years or less may indicate that awareness of achalasia, and referrals, are improving. *Endoscopy* is essential to check on whether other conditions are present in the oesophagus. A Schatzki ring, for instance, can also lead to swallowing difficulties because of narrowing of the oesophagus but would be treated with dilatation. Endoscopy would also check on gastric emptying, but in some cases there is no need to repeat an endoscopy as a matter of routine. *Barium swallow* is helpful for showing a picture of *before and after*. High resolution *manometry* is very useful but can be limited by difficulty of interpretation when the tube cannot pass through the LOS and is a temporary snapshot of muscle pressures.

Who does it affect?

- Men and women
- 1/100,000 annually incidence [ie those diagnosed]
- 10/100,000 prevalence [ie all stages of the condition - about 6,000 in the UK]
- Any age 16 – 80s first presentation
- 30 – 60 years
- Some genetic predisposition

Achalasia can tend to get worse over time and with age.

## Different types of achalasia and treatments

Achalasia is not a simple or consistent problem, sometimes it is an issue about muscle propulsion; sometimes predominantly about failure of the LOS to relax; sometimes a combination. High resolution manometry has helped to distinguish the details of the condition, and whereabouts the problems are occurring through the length of the oesophagus, ie around the LOS, or higher up towards the throat. John Pandolfino developed the Chicago classification into types of achalasia that can sometimes guide what kind of treatment is likely to be best:

- *Type 1* is predominantly a failure of the lower end of the oesophagus and the LOS to relax. There is little or no muscle pressure in the main body of the oesophagus. This type is reported as being more likely to be amenable to treatment by dilatation, botox or Heller's myotomy. It can be like a hosepipe with a knot in it where pressure can build up, and treatment will often involve measures to relieve this obstruction.
- *Type 2* involves some pressure in the main body of the oesophagus, sometimes intermittent or compartmentalised. This type is less likely to be successfully treated by dilatation and surgical intervention around the LOS, and more likely to respond to medical therapy.
- *Type 3* involves premature / spastic / uncoordinated contractions in the body of the oesophagus, and is sometimes regarded as a condition that can only be helped by medication rather than surgical procedures.

These labels are not always helpful in practice. It is possible, for instance, to alleviate problems with Type 3 by addressing removal of the cause of the food obstruction and to contemplate making worthwhile improvements in a patient's condition, with realistic and limited expectations. The treatment tends to be best when concentrating on relieving the dominant symptom, and the patient history can be really important in deciding on treatment.

### Treatment:

- 2 parallel strategies – aiming to get the patient into the best situation before any intervention.
- Avoiding stress, and other exacerbating factors
- Medical treatments
- Endoscopic treatments (botox, POEM, dilatation)
- Surgery eg myotomy and fundoplication.
- *First intervention shot is the best chance of getting it right.* Subsequent interventions become more difficult. This can be relevant if too many dilatations are carried out, notwithstanding that sometimes a dilatation can be the best option if surgery is ruled out for other reasons.

*Myotomy* involves cutting about 10cm length of muscle in oesophageal lining. *Fundoplication* involves wrapping the fundus, or top part of the stomach, around the base of the oesophagus to create a valve-like effect to prevent reflux from rising.

### Problems after myotomy:

- Obstruction not relieved
- Spasm not relieved
- Reflux
- Recurrence

## **Follow Up**

Mr Hashemi stated that he himself routinely invited patients for follow-up appointments after treatment for achalasia because it is a long term condition, and that it was important for monitoring reflux and other issues. There was not a consistent approach nationally, however, and he stated that patients should approach their GP for a referral if their condition was changing or if there were situations that created concern. Sometimes people might feel that their fundoplication wrap had loosened or was losing effectiveness.

## **Spasms**

Q: Why do we get them? The disease would be easier to live with without them. What can I do for the extreme chest pains that shoot to the back, especially at night. Can I ask about nortriptyline low dosage that is an anti-depressant but works with spasms?

A: Spasms are difficult to explain, difficult to treat and difficult to live with. They may be more common with Type 3 achalasia where medical therapies more likely to be more effective. Nortriptyline and amitriptyline sometimes used, but each patient tends to be different.

There is a need to define the pain and establish whether or not it is heartburn-type pain around the sternum (breast bone), or is it spasms. Important also to undertake cardiac assessment for chest pain. Sometimes relieving the obstruction (eg tight LOS) relieves the spasms. If spasms persist, calcium channel blockers can be prescribed, and if these fail, amitriptyline 10mg three times daily. Calcium channel blockers and nitrates can relax the LOS. Stress can exacerbate the problem. Buscopan (available over the counter) 10mg 6-hourly can be an option. Spasms suffered before surgery may persist afterwards. In a study (Patti et al, 2008) involving 167 myotomies, 55% had pain beforehand. 95% improved afterwards.

If spasms are associated with meal times, the cause might relate to food. If at night, they perhaps are more likely related to reflux? In one case spasms had improved after surgery, stopping of PPI medication and a change of diet, but could continuing, milder spasms at night be caused by reflux? Gaviscon Advance is recommended.

Some have found that drinking warm water helps to relieve the spasms, and at night, when drinking is not wise, placing a hot water bottle over the chest.

## **Medication**

Some people have problems of regurgitating medication, or retaining it too long in the oesophagus. Pharmacists can be consulted to provide medication in suspension / liquid form. Different hospitals vary. Calcium channel blockers and nitrates can be in suspension form. Sometimes medication can be used in the form of suppositories or can be absorbed from under the tongue, subject to pharmacy advice.

## **Reflux:**

Q: Can reflux contribute to achalasia? A: Reflux is caused by stomach contents rising into the oesophagus, but there is also the effect of a corrosive effect of unswallowed food fermenting in the oesophagus, so the cause of the problem needs to be established. Gastro-oesophageal reflux disease (GORD/GERD) occurs when the LOS does not prevent stomach acid from rising into the oesophagus and the acid then damages the lining, which is not designed to withstand strong acid like the stomach.

There can be damage to the lining of the oesophagus from fermenting food that is not passed through into the stomach. Gastric emptying is also important because the problem can be made worse if the stomach is not emptying food to propel it further down into the digestive system. Reflux can tend to occur after POEM, a myotomy or dilatation because the LOS is widened and tends to act as an open valve. Proton pump inhibitor (PPI) medication like Esomeprazole switches off acid production in the stomach to combat reflux.

Medical literature quotes 60% of *POEM* cases not requiring PPI medication afterwards, but it is not certain whether this relates only to short term reflux rather than long term. Patients can be inappropriately persuaded that they do not need PPI medication. Fundoplication is the surgical answer to providing a valve effect against reflux. Long term exposure to reflux is a health issue. Normally it is treated with medication (eg PPI such as Omeprazole). Conventional anti-reflux surgery includes fundoplication which routinely occurs at the same time as a myotomy is performed. Sometimes the fundoplication wrap needs to be adjusted. There is a device called *Linx* that consists of a magnetic bracelet around the base of the oesophagus but this is rarely suitable for achalasia patients. Reflux is not known to cause achalasia as such, but might provoke spasms. It often creates heartburn pain because of the reaction of the lining of the oesophagus against the assault of the acid reflux, and this can in turn lead to oesophagitis or Barrett's Oesophagus. A regular review is prudent.

## Age

Q: Is 72 too old for a myotomy? A: Botox can be a short-term answer, but can also make later surgery more difficult. Age of 72 no reason in itself not to have a myotomy.

## Speech and language therapists

Some have found this useful for when losing their voice because of reflux.

## Barrett's Oesophagus

Q: Is radio frequency ablation for Barrett's oesophagus advisable with a myotomy? A: Radio frequency ablation is now considered a safe procedure. It removes layers of the lining to deal with Barrett's Oesophagus with dysplasia. High grade dysplasia magnifies cancer risk, low grade dysplasia less so, but some Barrett's Oesophagus does not involve dysplasia at all. Length of Barrett's Oesophagus cells can relate to increased risk of cancer. If this is *before* a myotomy, the ablation is likely to thicken the oesophageal lining and make surgery more difficult. It would be difficult to find a practitioner who would be willing to do ablation *after* a myotomy. Achalasia treatments need to be considered for implications throughout a patient's life. It would depend on whether the dominant problem was achalasia or the Barrett's Oesophagus.

## Cancer Risk of Achalasia

Achalasia is an unusual condition. Sheraz Markar had co-written a paper: *Incidence and risk factors for esophageal cancer following achalasia treatment: national population-based case-control study. (Diseases of the Esophagus (2019) 32, 1–7 DOI: 10.1093/dote/doy106)*. It involved a group of 7,487 patients diagnosed in England between 2002-2012. When they had had several unsuccessful treatments for achalasia in a short period there was a slightly elevated risk of cancer, but this may be because there were other problems with the oesophagus rather than achalasia itself, and cancer was associated with age. Therefore some patients do need regular follow up. There is a case for following up patients at, say, 3-year intervals, especially over the age of 60 years. GPs can be requested to refer patients for

follow up. Also it is reasonable to ask the surgeon for regular follow up to monitor reflux issues. Reflux after achalasia surgery can be an issue, and it is possible to revise the fundoplication wrap.

### **Peristalsis**

A: Complete loss of peristalsis is quite common in achalasia

### **Sleeping position in hospital**

Q: Will NHS nurses be aware of importance of sleeping position to avoid reflux and choking?

A: Probably best to remind them as it might depend on the particular hospital.

### **Coronavirus risk at work**

Q: What risk is there in a large school with many special needs pupils and narrow corridors?

A: Nobody knows for sure whether or not there is a particularly higher risk of contracting COVID19 for those with achalasia, but if there have been aspiration problems, the lungs may have been weakened and less able to resist the virus if the person does become infected. COVID19 has affected interventions because of the risk of aerosol-borne infection, so that endoscopies and surgical treatments are currently performed on an emergency basis only. Surgery is currently dependent upon a 14-day quarantine and a negative virus test before admission. We are not in a position to recommend shielding just on the basis of achalasia alone. Some people with achalasia are in good health, but each individual has to assess their situation with their GP according to their own circumstances.

### **Excess drinking a potential cause?**

A: No [but not wise for other reasons!]

### **Any links with mental health?**

A: Stress can be connected in so far as stress can make the perception of symptoms, or the symptoms themselves, worse, but it would not be true to say that mental health causes achalasia.

### **Impact on the bowels?**

A: If you are not drinking enough, this can cause constipation.

It is possible to have motility problems throughout the digestive tract. Sometimes PPI medication can cause constipation. If the digestive tract lower down is working well, it helps the oesophagus and stomach to work better higher up the system.

### **Dumping Syndrome**

Q: After a Heller myotomy as an 8-year-old and now suffering from irritable bowel syndrome, the person suffers from urgent diarrhoea 30 minutes or 1.5 hours after eating. A: For those who have had part of their oesophagus or stomach removed, there can be an issue with insulin spikes caused by semi-digested food reaching the intestines and creating palpitations, clamminess etc (see factsheet from GUTS UK <https://gutscharity.org.uk/wp-content/uploads/2018/08/Guts-UK-Dumping-Syndrome-leaflet.pdf#:~:text=org.uk.%20This%20factsheet%20is%20about%20dumping%20syndrome.%20Our,is%20not%20adequately%20prepared%20to%20allow%20efficient%20absorption.>)

It is then worth considering concentrating on food with a low glycemic index to reduce initial sugar load. Small Intestine Bacterial Overgrowth (SIBO) can be diagnosed with a hydrogen breath test. It occurs when bacteria change in the intestines, especially after surgery or with strong medication, and can be treated with antibiotics. Also worth checking on diabetes issues, and whether there might be an IBS flare up? Sometimes fatigue can be associated with digestion issues as liquids naturally transfer in and out of the intestines but it would be prudent to seek medical advice if this is causing concern.

### **Fear and severe dysphagia**

Q: How long should one leave it if food remains stuck in the oesophagus for three days? A: Sometimes resilience and denial can be a good thing, but three days is too long and one should seek medical help before this. An endoscopist may have methods for dislodging or clearing the food. If you already have been having treatment, consult the specialist direct.

### **Genetic issues**

A: There is a proportion of people for whom achalasia seems to have a genetic connection and therefore more rapid referral for treatment is justified.

### **Research comparison between long and short POEM**

A: There has been a comparison study published in the New England Journal of Medicine about POEM compared to Heller's myotomy (HM), where the length of the myotomy was found to be crucial for success in HM, and therefore it is likely to be true for POEM also, notwithstanding that the panel had not heard of any results from the specific research project referred to by Giacomo Frisoni. Where revision surgery takes place, a relatively frequent cause of the first surgery not being successful is the original myotomy not being long (or deep) enough. It is possible to have POEM after a Heller's myotomy, but it needs to be done by a large-scale centre with great experience. Sometimes there is a case for POEM where there is need for the myotomy to be extended in the mid-oesophagus.

### **Hiatus Hernia**

A: Hiatus hernias are common and involve part of the stomach rising above the diaphragm, whereas achalasia is rare. Hiatus hernias can be repaired and the surgery for achalasia can be completed at the same time. It is possible for a hiatus hernia to develop after a myotomy. Hiatus hernias are likely to cause reflux problems. Occasionally a very large hiatus hernia can create pressure on the oesophagus and create symptoms similar to achalasia but it is not achalasia as such. Repairing the hiatus hernia then solves the problem.

### **Food**

Individuals can vary an enormous amount, but in general, beware of food with a texture that might create a blockage (eg tomato skins, stringiness), congeal (eg white rice, white bread), or cause irritation if it remains in the oesophagus for too long (eg spicy food). The results of a small informal survey in 2015 of nineteen people with achalasia are contained in *A Patient's Guide to Achalasia* that can be downloaded from [www.achalasia-action.org](http://www.achalasia-action.org)