BASED IN THE UNITED KINGDOM
ACHALASIA ACTION
www.achalasia-action.org
Uniting for a rare swallowing condition
Reg Charity no 1187367

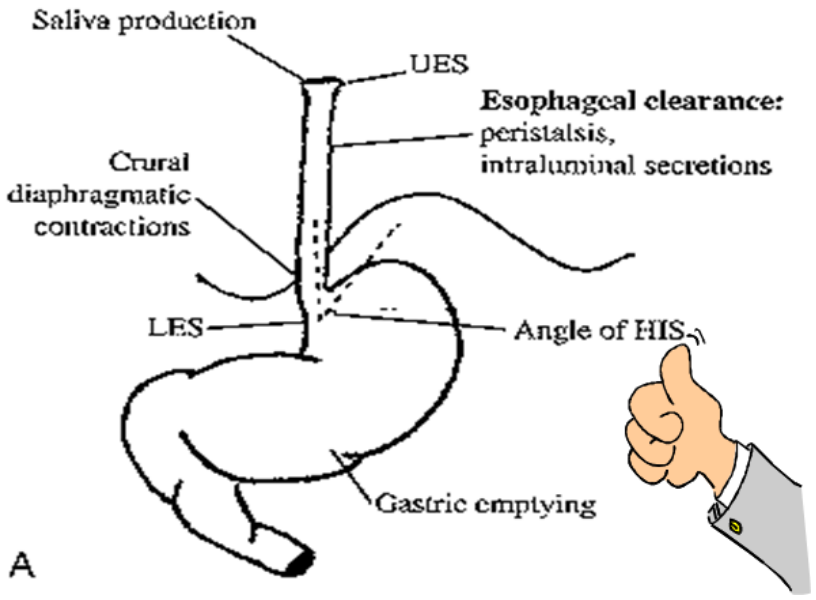
PAIN AND SPASMS - 9 November 2021

Introduction

Alan Moss (Achalasia Action Chair & Host of the meeting) welcomed everyone to the meeting and introduced **Majid Hashemi FRCS,** Consultant Upper GI Surgeon, and Medical Chair of Achalasia Action.

> **Dr Ahmed Albusoda** - Dr Albusoda is a Consultant Gastroenterologist at Barts and Royal London hospital and has completed a PHD on gastric pain, under Professor Qasim Aziz
>
> **Mr Sacheen Kumar** Mr Kumar is a Consultant Upper GI Surgeon at the Royal Marsden Hospital with wide experience of benign and cancer surgery.

Mr Hashemi commenced the session by presenting a short powerpoint explaining the basics of achalasia. (What it is, diagnosis, treatment etc).



From a medical point of view, achalasia involves a non-relaxing lower oesophageal sphincter (LES or LOS, the valve between oesophagus (or gullet) into stomach), a high resting muscular pressure in the lower oesophagus, poor contractions in the oesophagus (peristalsis), and sometimes simultaneous and badly coordinated contractions in the oesophagus. For the process of peristalsis to work properly, you need the nerve endings (ganglia) controlling the muscles to work properly both to trigger the muscles and to relax them. There is a loss of the inhibitory ganglia in the myenteric plexus of oesophagus, a non-relaxing sphincter (LES /LOS), a degeneration of inhibitory nerves in oesophagus, unopposed action of excitatory

neurotransmitters, and an issue with acetylcholine. Loss of cholinergic neurones over time leads to loss of tone, dilatation, and simultaneous contractions.

A barium swallow image will often show a 'bird's beak' where the LOS has become very narrow. People complain of difficulty in swallowing (dysphagia), regurgitation of chewed food that has not reached the stomach, reflux, pain, respiratory problems and laryngeal issues.

Diagnosis entails taking a full history, endoscopy, barium swallow test and manometry. Achalasia affects both men and women. Around 6,000 people are affected in the UK. It can first present to doctors at any age, mostly between 16 and 80s but the age range 30 - 60 is most common, and it can also affect children. There may be an element of genetic predisposition.

Treatment comprises two parallel strategies, firstly to avoid stress and exacerbating factors accompanied by medication. Then endoscopic treatments, POEM and surgery.

The main theme of the question and answer session was Pain and Spasms and the session was commenced by Dr Albusoda giving an overview of his research work on pain:

Dr Albusoda (Ahmed):

We do not know everything about achalasia. Doctors concentrate on the movement of food through the oesophagus (gullet), and pain itself is not always their priority; the issue of pain can be elusive. It is not always clear what causes pain and it can vary in many ways between individuals, including how, where and whether we feel it.

Pain felt in the gullet (oesophagus) is mostly linked to irritation e.g. by hot or acidic foods, or by food getting stuck etc. If, for instance, you burn yourself, the area surrounding that burn will become extra sensitive, and you should not touch that area in order to give the flesh time to heal. The same principle applies to pain in the gullet and how sensitivity spreads from the immediate location of the problem causing the pain. Pain occurs because the gullet is trying to protect itself and is telling us to stop whatever the cause of irritation was.

Secondary pain can then occur, ie an increase in sensitivity may follow a couple of days of problems. Pain signals are passed through the nerves to the spinal cord and up to the brain which then interprets what is happening in the body. If the pain signals are repeated constantly, the nerves in the spinal cord themselves then become more sensitive and this creates ever stronger pain signals that are passed to the brain.

With achalasia, food remains in the gullet rather than being cleared through into the stomach and this causes irritation. Saliva normally washes this food residue through to the stomach, and when saliva also gets stuck, this process does not work properly and irritation gets worse. Sometimes the Upper Oesophageal Sphincter (UES) does not relax well at the top of the oesophagus, and this can trap gas within the gullet. Normally this will be burped out, but trapped gas can also be a cause of pain. Muscles can develop spasms, and these can also be painful. Fermentation of food in the gullet can also be a contributory factor to irritation.

Pain is not just an on/off switch. When the pain signals reach the brain they integrate with emotions, how you are feeling, anxieties and memories of previous pain (eg previous experience of choking).

Mr Hashemi (Majid):

In looking to relieve pain as a surgeon, I would first check for any obstruction and whether an intervention such as myotomy might be needed to relieve this.
Secondly - check for and rule out reflux, which can become very painful, and fermentation.
Thirdly, the role of external stress is frequently raised by patients and has been the topic of Achalasia Action's collaboration with the University of Reading and Melika Kalantari on coping mechanisms.

Mr Hashemi then led the **question and answer session**, with help from guest speakers:

Question and Answer Session

Questions had been submitted by attendees prior to the meeting.

Stress

Q: *I feel that achalasia is at least partly stress related, at least for me. I find that warm water stops a spasm progressing. I also find that a hot water bottle held across the chest can help especially at night when drinking water is not a good idea*

A: Majid - Yes he is 100% sure that stress affects pain experienced by achalasia patients.
Ahmed - Stress is linked to our coping mechanism. If, say, you were in an isolated location, you might hear the same noise and react differently according to whether you were stressed or not. There are three systems: *Rest and Digest* for when you are calm; *Fight or Flight* that will make you react if something bad happens; and thirdly, the *Enteric Nervous System* including in the digestive tract. These are very old reactions that were useful if you were, say, at risk of getting into a fight. They can be helpful in anticipating problems, but stress can make the muscles more tense and make pain and spasms worse, as part of a vicious cycle.

The enteric / autonomic nervous system operates below any level at which we can control our reactions. It can accelerate the digestive system either to empty the bowels, or sometimes to delay such movement until the danger is over (eg the person has escaped from a tiger). Stress acts as an accelerator on the digestive system and makes things more chaotic.

Acupuncture

Q: I was diagnosed with achalasia in 2017 but have been suffering with it for over 10 years. My father had it and eventually died with it. He went through all the conventional medical treatments. I am taking the Chinese medicine route, and want to know if anyone else has had success with acupuncture and the Chinese herbal medicine treatments? And if so , What specific Medicines did you use?

A: Ahmed stated that he did not refer patients for acupuncture as he was not aware of clinical trials that would give good enough evidence, but theoretically pain can be diverted through the vagus nerve system that affects the digestive tract. He knew of one trial where pain was reduced in the oesophagus when the person's foot was placed in cold water, for instance.

Physiology and Aspiration

Q: *What is the physiology of the oesophagus and windpipe? How do they work in relation to each other and what happens to them/that relationship when Achalasia occurs?*

A: This was covered in the introductory powerpoint. The windpipe relies on valves (Upper and Lower Oesophageal Sphincters) in the oesophagus to work properly to stop food going into the windpipe (trachea) and lungs. If food builds up in the oesophagus the pressure can increase until it suddenly releases upwards and can then enter the lungs or affect the vocal cords. This can lead to aspiration pneumonia which is a major trigger for medical intervention.

Q: *How do we best keep the windpipe clear/healthy?*

A: A number of basic things like elevating the head of the bed at night, and not eating late in the evening.

Pain and Spasms, and attending A&E

Q: What should I do if aspiration occurs? Should I go to A&E (ie Accident and Emergency hospital department)? At what point and how long do we wait before going to A&E which we would like to avoid doing? A few hours? A day? What about severe pain occurring at night with an empty oesophagus and whilst sleeping without any known stress.

A: Not really as A&E may not be able to help much. It is best to get referred to a specialist GI (gastrointestinal) clinic for investigation. If a diagnosis of achalasia has already been given and the person has had treatment and advice from the clinician about what to do, then it is likely that people will be able to cope without going to A&E. But if people are not sure, there is always the risk that it might be related to a heart problem. Somebody with achalasia is not excluded from also having cardiac problems so you should seek help if you are in severe/unusual pain. Patients should get their heart checked out if they are regularly getting chest pain, which might be angina. Majid has referred patients to cardiac colleagues for this when the pain was not reflux-related but was an urgent heart problem.

Even if the oesophagus is apparently empty there can be small amounts of reflux occurring or small amounts of undigested food fermenting. 24-hour manometry might provide an answer. Some medication can be given through suppositories or with a spray when people cannot swallow. There is an element of experimentation when specialists prescribe these things however.

Q: *Can antidepressants help?*

A: Antidepressants can come in many different forms and are unlikely to work directly on the oesophagus or spasms. Some might even make the muscles more tense, but they can have a beneficial effect on pain perception in the brain. Ahmed would expect perhaps one in four patients to find pain relief from antidepressants. Sometimes it is more beneficial to concentrate on other issues such as reasons for lack of sleep and anxiety. There can be side effects, such as constipation with tricyclic medication, and this would then be counter-productive in slowing down digestive motility.

Fatigue

Q: *I often have a pain which goes from my chest through to my shoulder blade. Sometimes it's dull and in the background and sometimes it's sharp and quite intense. It can be quite exhausting and tiring to be in pain so much of the time. When it is intense or comes on suddenly I will speak to the paramedics, when it is there and present a lot of the time i wonder what causes it? Is this normal? And is there anything that I could do to settle it?*

A: Chronic pain does cause exhaustion, but check other things eg. vitamin and mineral deficiencies (eg lack of iron and vitamin D). Aspiration could affect sleep, and ability to sleep can be a factor. Make sure to address all these issues which may help.

Balloon dilatation

Q: I've had a Heller myotomy (HM) and a year later balloon dilation. I suffer with intensely painful spasms. I have been offered another balloon dilation as a solution but no one will explain why. How could this help spasms?

A: It does beg the question of why any dilatation is required after a HM. This is probably due to an area still left tight after the myotomy, the fundoplication wrap, perhaps because the myotomy was not long enough, or scar tissue? If so, it is reasonable to try the balloon. It may be that this case should end up with further full investigation, potential further myotomy surgery, and perhaps having an endoscopy during the operation itself to get things right. It would need a surgeon experienced in revision surgery. One should not be contemplating revision surgery solely because of spasms; the surgery should have a clear aim for dealing with a myotomy or fundoplication, and revision surgery is generally something to avoid if possible.

It does need a barium swallow test to establish whether the food is flowing through well. One would need to check that the myotomy and fundoplication were working well. If everything is working well but there is still pain it would need to be further investigated on a medication or pain relief basis.

Balloon dilatation does have a clear role as a primary treatment, and can be as good as surgery for some patients depending on the type of achalasia experienced, but not for young people with many decades of life ahead. The dilatation stretches and breaks the muscle at the bottom of the oesophagus which then allows food to flow through, and therefore solves part of the problem; the trouble is that reflux can then flow upwards, irritating the gullet and then this can create spasms.

Sneezes

Q: Usually cold water helps my spasms but why are mine often brought on by sneezing? Not necessarily sneezing through a cold but just general dust etc. I wonder if anyone else has this and lives in fear every time they sneeze that they will end up having that well known pain we all dread?

A: It is not the same for everybody, but sneezing and sudden movement can create a shock through the chest and could cause a spasm.

Post Surgery

Q: What causes spasms after surgery when there is no evidence of food being held up?

A: First thoughts would be to investigate for reflux.

Medication

Q: *What medication is used for somebody who is suffering pain and spasms when the prime causes like obstruction, food hold-up and reflux have been ruled out, and the patient is well nourished and relaxed? Is there any type of painkiller to help stop spasms that does not increase the chance of heartburn? Paracetamol is not strong enough. What analgesics can be used, especially when there is difficulty in swallowing?*

A: Success of medication varies greatly between patients but the following can be helpful:

- Start with simple things like peppermint oil
- Buscopan can be helpful for spasms and is readily available, and not overly strong
- Calcium channel blockers/ nitrates can work for some - use of a GTN spray can be useful if patients cannot swallow
- Sudenafil (Viagra) is not licensed for this purpose and very few people prescribe it, but it is a powerful smooth muscle relaxant that can be used as a last resort.
- Diazepam can be used rectally in extreme cases
- Generally agreed that there is no benefit in liquid opiates and if pain is that bad, the patient needs to be seen for intervention
- Medicinal cannabis (CBD) has been spoken of but current evidence not convincing - it might be a development in future
- Ahmed does not particularly use Diplofenac
- Some of these prescriptions may need to come from a specialist initially but most can then be continued by a GP.

Q: *Over time my spasms have lessened both in severity and in frequency. Could this be just due to learning how to manage them, by avoiding triggers and using learned techniques to soften the pain before it sets in? Or could it also be due to the nerves dying off?*

A: Could be both. It is difficult to analyse what is happening with nerves. Patients learn coping strategies, and the functionality of nerves can be altered by surgery.

Q: *Can/do you test for sensitivity of oesophagus? Or work with a pain team?*

A: Sensitivity testing is not conducted in clinical practice, only for research. Ahmed works occasionally with a pain team to help manage pain but has to ensure they understand the condition as some types of medication make things worse eg opiates which can create addiction problems and constipation. He would normally use anti-depressants rather than opiate medication.

Q: *I am currently waiting to have the POEM procedure. Does this procedure alleviate pain/spasm? And will any effects/benefits from the POEM be long lasting for pain/spasms?*

A: Patients can get spasms before or after (or without) any procedure and unfortunately you cannot predict what the effect on an individual would be. There will need to have been

a discussion with specialists about whether POEM is the right course as POEM does not in itself involve a fundoplication to combat reflux.

Q: *Why do I get pain and spasms in the middle of the night, sometimes regularly at a similar time?*

A: First thing would be to check for reflux and whether the sleeping position can be changed to stop it flowing towards the throat from the stomach. Gaviscon can help at night. Pain generally is often worse at night and might depend on whether you can sleep or not. There is a diurnal variation of a stress hormone Cortisol that might affect some people.

Oesophageal Diverticula (pouch)

Q: *I am suffering from episodes of regurgitation like never before, and I am choking saliva. Although my passage in the lowest part of my esophagus is pretty tight at the moment (less than 18mm), I can still manage with small bites of semi-solid food. But strangely I noticed that I start to regurgitate food after two days and there is no gastric acid taste in it, just a sickening smell of ammonia.*

Can it be a case of Diverticula ? At my last check up 6 months ago there was no sign of Hiatal hernia.

This is the list of my symptoms:

- *Dysphagia (difficulty swallowing) Odynophagia (pain with swallowing) Chest pain. Continual cough. A feeling of food stuck in the throat. Continuous Heartburn. Regurgitation (the return of partially digested food to the mouth), especially when bending over or lying down. Bad breath. Feeling of fainting and cold creep when the food reaches the Cardia area, cold sweat. If it is Diverticula what chance is there to heal from it?*

A: Sach - This is a long list of symptoms which may not all be related. Need an endoscopy to establish whether or not there is a diverticulum in the oesophagus, or perhaps a pharyngeal pouch. Then cross-sectional contrast imaging if suspected diverticula. Start at the beginning with investigations. True diverticula are difficult to treat and many people simply live with them without problems, but surgery is possible once there is a clear diagnosis of the problem. The smell might be a sign of fermentation of food and the stasis of stuck food might be creating some of the other problems.

Q: *In the USA I was only offered a POEM?*

A: The US healthcare system is different from the UK. In the NHS there is generally much more collaboration between gastroenterologists, surgeons and other clinicians over whether procedures by gastroenterologists or surgery is better for an individual. POEM procedures can be undertaken by both gastroenterologists and surgeons.

Q: *Drugs such as Omeprazole are prescribed to people with Achalasia to control reflux, heartburn and related stomach pain. These are often prescribed over long periods of time and for post POEM can be life long. What is the view of the panel on taking these drugs for long periods and are there any effective alternatives with fewer side effects that can be recommended?*

A: Every medication, even drinking large amounts of water, has side effects. PPI medication is relatively safe, and provided that there is a clear reason for it, can properly be prescribed long term. They can affect sleep, the acidity in the stomach that controls bacteria and absorption of calcium, however, so regular reviews are always prudent for any long term medication.

Q: *Is age 76 an exclusion for treatment?*

A: No, not in itself. Majid and colleagues have performed surgery on patients in their 80s and older, subject to their general health and fitness, the extent of any other health conditions for the individual, and their understanding of risks of anaesthetic. Extra care is taken because of the trend for the oesophagus to become a bit 'lazy' with age (presbyoesophagus).

Q: *My 12yr old son has very recently undergone a Heller Myotomy, (11/10/21)… he is still experiencing lots of chest pain during the night and early mornings. We understand that this pain is acid reflux. It has lasted up to 2hrs on occasion and he has been in agony. We give him cold water which helps and also chewing gum and gaviscon but he still wakes during the night in pain. We have recently got him an electric bed to change his sleeping position however we have concerns about this pain long term and the impact on his oesophagus. It is happening 3-4 nights per week so far. He is not eating after 6pm*

A: None of the medical experts attending the meeting are specialists in paediatrics, however the advice given above regarding adult symptoms and relief apply generally to children, but prescription doses might be different. We have had a session with Dr Osvaldo Borrelli from Great Ormond Street in the past. It would be prudent to check that there is nothing by way of post-surgery after-effects that can be remedied.

Mega Oesophagus

Q: Diagnosed with Type 1 in 2019. I haven't had surgery yet. I started to experience pain and dizziness in left breast area last year: results showed left bundle branch block and double right border in heart, which 'might be due to your Achalasia cardia'.

Question 1: could I have a mega oesophagus that is applying pressure on my heart?

Question 2: I used to experience spasms of pain in the middle of my back prior to being diagnosed (especially at night). This pain would radiate into my jaw. After the Achalasia symptoms started, I didn't experience pain, until recently. What is going on? It often results in a belch. My dad had angina and experienced similar pain

A: Upper GI Surgery can quite frequently be close to the heart, and it seems unlikely that the heart would normally be adversely affected. It is possible to have an enlarged oesophagus without being aware of that fact. A mega oesophagus is probably not a reason in itself to have surgery, but the condition might be associated with aspiration.

Q: *I have Achalasia type 1 and recently lost weight rapidly. I am being led to believe by my Gastro consultants it's due to inflammation in the small intestine.*

I have Ulcerative Colitis & have been on Remission since 2003. They now suspect Crohn's. So

starting today on Reducing steroids, endoscopy and CT scan of small intestine to be done very soon. I had thought weight loss was due to a new obstruction in the esophagus.

So the question is: Can inflammation of the small intestine create symptoms of Achalasia?

A: The short answer is No. One would need to investigate with an endoscopy to make sure there is no stricture or blockage in the oesophagus. It may be just an unfortunate coincidence that one has two separate diseases. Motility through the whole digestive system will tend to improve the symptoms of achalasia.

Q: Covid booster jab, sharp stabbing pain in my left shoulder blade, just ten minutes after receiving the vaccine. Severe vomiting followed. So I suppose my question is that I have clearly found that severe muscle pain can bring on an equally severe achalasia attack, currently lasting about four weeks! What can one do about it?

A: COVID is a new thing and it takes the medical world a long time to understand the full effects of new diseases. It could be an inflammatory response, or it could have triggered additional stress to make achalasia worse, but there is no clear correlation as far as is known at the moment. COVID can cause problems for those with achalasia because of infection in the lungs.