ACHALASIA ACTION
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Uniting for a rare swallowing condition

Achalasia Action Panel Q & A Session 21st June 2022 5.30pm

Topic: COPING WITH ACHALASIA: Before and After Treatment

Panel:

Mr Majid Hashemi
Consultant Upper GI Surgeon
Medical Chair of Achalasia Action

Professor Daniel Sifrim
Director of Upper GI Physiology Unit
Royal London Hospital
Wingate Institute of Neuro-gastroenterology

Dr Ahmed Albusoda
Consultant Gastroenterologist
Barts and Royal London hospital
Completed a PhD on gastric pain, under Professor Qasim Aziz

(Claire Donnelly, Specialist Dietitian, Whittington Hospital, was unable to answer questions on the evening, but we aim to circulate any additional answers from her in due course)

Introduction & Preliminary Remarks

Alan introduced the meeting and welcomed the panel, saying he is delighted to have so much expertise at the meeting. Attendees should note that questions and answers are generalised, and the specialists can't give personalised advice.

Options for type of treatment

Q: I got balloon dilatation done 10 years ago and I haven't had any serious episodes of food regurgitation after that. For last two years I need bit more water to get the food down.

A: Majid: Recommend getting a timed barium swallow and if oesophageal emptying is delayed consider further treatment options

Q: I went to India couple of months back and got my endoscopy, manometry and barium swallow done, but after endoscopy my swallowing issues increased (I used to be at 70% and after endoscopy they came down to 30% not sure if anything happened during endoscopy), I

want to know what are my options like POEM or oesophageal dilatation at the moment as manometry shows LES pressure is normal but no peristalsis, I can still eat things but need lots of water to push.

A: Majid: Treatment can range from lifestyle changes to surgery, the options are:

Do Nothing (i.e. no interventions) – adjust diet, manage lifestyle to reduce anxiety, triggers etc.

Dilatation – This can have positive results in some cases once diagnosis is confirmed but it can be short-lived and the downside is that it can make any future surgery harder: It makes it more difficult to achieve a successful outcome due to breaking down/ rupturing layers in the sub-mucosa and causing scarring. In the oesophagus you have the outer muscle, the sub-mucosa and the mucosa (inside). In a dilatation the intention is to go far enough to break the muscle on the outside but not as far as the sub-mucosa as this would then be a rupture. Breaking the muscle does lead to scarring during healing.

(Dilatation is carried out via endoscope with a balloon attached to the equipment. The balloon is then inflated to stretch the lower oesophageal sphincter (LOS/LES) by breaking the exterior muscle)

Surgical (Heller's Myotomy (HM)) – A procedure that has been tried and tested worldwide over 20 years, with thousands of cases done. It involves an hour to an hour and half of surgery under general anaesthetic plus 24/48 hrs in hospital. It is performed via laparoscopic (keyhole surgery) techniques through five little holes in the abdomen. The surgery consists of cutting the muscle in the lining of the oesophagus which control the LOS in order to relax the sphincter and allow food through to the stomach. The procedure is normally combined with a fundoplication to prevent reflux – this involves wrapping the top part of the stomach (the fundus) around bottom of the oesophagus to recreate the valve effect to control reflux.

POEM (Per Oral Endoscopic Myotomy) This cuts the same muscles as in a myotomy but does not include a fundoplication. It is approached differently and performed by endoscope usually under general anaesthetic.

Majid's view (as a surgeon) is that POEM is relatively new and has raised a lot of interest in achalasia which is a good thing but that it generally offers no advantage over myotomy and may lead to reflux as no fundoplication is performed.

Ultimately if you choose to have a procedure or surgery done it needs to be tailored to you and done by someone who has either done a lot or is supported by others who have much experience in carrying out the techniques.

Timing and delay of Treatment

Q: When the oesophagus has developed diverticula (swollen/distended pouch), and gets inflamed often, with extreme pain, what is the treatment generally suggested, if there is any? And what could happen if not treated?

Q: Looking for advice on when is the best timing for an HM - if the Consultant has left the decision of whether to go for a HM to the patient is it advisable to go for procedure whilst

symptoms are still manageable and being coped with or wait until they become severe and difficult to manage?

A: Majid asked Professor Daniel Sifrim for a comment on this as he has many years' experience – does he see progression of the condition over time in untreated people?
Daniel: Re the first question – in deciding which intervention to have, or whether to have one, in a patient who has been diagnosed, first ascertain whether there is an objective obstruction blocking the sphincter. Then continue with further tests and investigations before a decision about treatment.

No, he doesn't see much change over time – changes are slow – once diagnosed as type 1, 2 or 3 this rarely changes and changes in the oesophageal body are normally slow. An exception to this is that a diagnosis can change from obstruction to full/ true achalasia one or two years later. Type 2 can progress to Type 1 over time, but otherwise there are not usually any rapid changes in the oesophageal body.

Types of Achalasia

Q: Can you please explain the difference between type 2 & 3 achalasia. I understand in part, but struggle to figure out from my symptoms which type I have.

A: Majid asked Daniel to answer this:
Daniel:
Based on the manometry test, three types are identified:
Type 1 – No relaxation of the LOS, no peristalsis, and dilation of the oesophagus
Type 2 – No relaxation of LOS, less dilation, some tone in the oesophageal wall and when patient swallows there is something called oesophageal pressurisation but no contractions
Type 3 –No relaxation of LOS but very vigorous, rapid, spastic contractions in the oesophageal body

The mechanisms that lead to the three types are apparently different.
In Type 2 and Type 1 there is a loss of neurons in the oesophageal wall, particularly the type of neurons called inhibitory neurons. Whilst Type 2 can progress to Type 1, Type 3 is a totally different entity.
In Types 1 & 2 there is progressive loss of neurons in the oesophageal wall.
In Type 3 the neurons are still there but don't work well – the balance of contraction and relaxation (via excitatory and inhibitory neurons) to allow smooth progression of food through the oesophagus is lost.

The differences are important – Type 3 is a different entity to Types 1 and 2 to the point that now people working in basic sciences in these diseases suggest that Type 1 & 2 and Type 3 are two different disorders.
Symptomatically Type 3 is also different in that it is more common in younger people and more commonly associated with chest pain due to the vigorous uncoordinated or premature contractions.
Recommended management of the different types is also therefore different.

Following up patients before and after

Q : How often should patients be seen for routine follow up checks after diagnosis?

Majid welcomed Dr Ahmed Albusoda to the meeting and asked him to comment:
Ahmed:
Depends on why – as we know we can't reverse the disease then some sort of intervention may be needed to relieve symptoms. This can progress with time so he follows up patients to see how they are managing and if interventions *are* needed.
Other follow ups – regarding the theoretical increase in oesophageal cancer, may need to do some form of follow up endoscopy.
So generally, he will recommend follow ups annually plus routine gastroscopy at appropriate intervals if all going well but with contacts provided in case symptomatically needed earlier.
If pain or reflux is an issue might need more follow up tests to see if symptoms can be relieved via an intervention.

Q: How long should you be reviewed for after surgery?

A: Majid : I follow up patients annually. There has been some discussion about following up patients with periodic barium swallows to monitor any progression in dilation of the oesophagus but I don't feel this is necessary unless there are new symptoms.
Majid agrees that pain and reflux can be issues both before and after treatment and these create need for earlier follow up.

Q: How long is acceptable for a referral [by a GP?] after being discharged when problems occur again?
A: Majid: Once discharged the patient would have to go back via GP for referral to local gastroenterology dept which is the starting point. The current pressures on the NHS may well be making this more difficult at the moment, and patients may need to persist.

Achalasia and Cancer Risk
Q: How often should we be checked for oesophageal cancer and would this just be by gastroscopy?
Q: I'm interested to find out more about the statistical occurrence of oesophageal cancer 15-20 years after achalasia diagnosis.
Q: Like Barrett's Oesophagus patients who are reviewed regularly and kept on the hospital books, is there any way we can be kept on the list as having a chronic condition in case of developing problems?

A: Ahmed:
The increased risk is not huge so the recall for tests via endoscopy could be up to ten years after initial diagnosis, then 5-yearly
A: Majid – doesn't endoscope if no new symptoms and not too much reflux or other problems unless patients are very anxious
No current recommendation in the guidelines re intervals for follow up or retesting
A: Alan Gave some further information/ statistics:

10% of people with recurrent, significant heartburn over a number of years develop Barrett's Oesophagus and of these 10% develop a type of cancer called adenocarcinoma- i.e. 1% of those with persistent heartburn. The key to this is whether there are changes in the cells – called dysplasia – identified by biopsies taken with endoscopy. A new development called Cytosponge (a pill on a string swallowed and then drawn back out to collect samples) will definitely detect Barrett's Oesophagus and probably some other conditions like cancer also.
However, people with achalasia are not at particular risk for this type of cancer - squamous cell carcinoma is more likely with achalasia patients and this is not really related to reflux. It might be no more than a raised risk because of general stress and strain within the oesophagus? If concerned ask for an endoscopy and get biopsies taken.
Overall the increased risk is very low.

Heller's Myotomy & Fundoplication (wrap)

Q: With the Heller's procedure with wrap, what do you do if you need to vomit and are unable to post procedure?
A: Majid: If done properly the wrap should be only a partial wrap, a gentle 90° or 120° so that you can still vomit, belch etc. The problems are with a tight 360° wrap which should not be necessary in achalasia.

Q: Is it common to experience frequent nausea after a Heller Myotomy + wrap like I do? As I can't physically vomit any more and can only dry retch so it's very unpleasant because I can't get rid of whatever it is that's making me sick. I've experimented recently with taking Omeprazole and it seems to be helping. What could be causing the nausea? Any kind of food seems to trigger it.
A: Majid & Daniel: Could be reflux if responds to Omeprazole but would recommend further tests before starting any treatment for reflux like PPIs (proton pump inhibitor medication like Omeprazole) – only treat if sure it's reflux. Don't self-treat. It might sometimes be the effect of undigested food fermenting in the oesophagus.

Nausea and Reflux

Q: I had my HM and partial wrap in January 2022 following diagnosis.
My main struggle is constantly being woken by discomfort/pain in the chest area most nights. Why is this happening and are there any solutions?

Q: I had the stomach wrap surgery some years ago. I'm interested in finding out how to reduce acid reflux episodes without medication like Omeprazole? I'm relying on Greek yoghurt at present to neutralise acid reflux when it occurs.

A: Majid asked the others for their views on reflux:

Daniel gave this summary:
In *non-treated* patients diagnosed with achalasia, reflux is very unlikely as the sphincter (LOS) isn't opening – so reflux is likely to be a misdiagnosis - patients can also be diagnosed and treated for reflux prior to getting a full achalasia diagnosis which isn't helpful. Some achalasia patients' first symptom is heartburn so it's important that they are investigated for achalasia.

If they have achalasia and a dilated oesophagus the heartburn symptoms are more likely to be food fermenting in the oesophagus and producing acid – not reflux from the stomach.
In *treated* patients it may be reflux as the sphincter has been opened up. This can be problematic as even a small amount of acid can sit for long periods in the oesophagus due to the lack of peristalsis and can cause damage resulting in Barrett's Oesophagus. Symptoms could also be fermentation if the food is still not clearing properly.
If treated patients whose initial symptoms now (post operation) were only dysphagia (difficulty in swallowing) have heartburn, they should be referred for a pH test to actively document if the patient has acid reflux. If confirmed reflux must be treated.
Majid – He follows up patients and if they are experiencing reflux will re-do the wrap rather than just prescribing PPIs as is common practice. Majid thinks this should be adopted more widely. Has never performed a wrap post POEM and would be very cautious about doing one.
Majid asked Ahmed for his views of long-term use of PPIs and possible side effects:

Ahmed : All medication has some side effects if it is effective. It is appropriate to prescribe PPIs if the patient has long-standing reflux and has, or is at is at risk of developing, strictures. But the important thing is initially try to avoid reflux i.e., do a HM with wrap.
There are new acid suppressing medications being introduced, currently used in Japan. Japanese are also working on an endoscopic procedure which incorporates an anti-reflux solution. This may become standard in few years' time.
Majid – Durability of an endoscopic procedure is key and he's not convinced so far.

Relieving food obstruction at home

Q: If food gets stuck how should we handle/be sick?

A: There are some tried and tested techniques which include:

- Raising arms/ extending neck/ stretching/ standing up & walking around
- Fizzy drinks – but only tiny sip of coke
- Warm water

(See the Achalasia Action website and "Living with Achalasia" booklet for more tips)

Research for a Cure?

Q: Chatting with president of ALMA, one of two Italian Achalasia groups, I understand that a group of researchers in Verona (Italy), guided by Prof. Salvador and Prof. Guy Boeckxstaens from Belgium, are close to revealing the origin of Achalasia.
Do you think we are really so close? And if this will be real shortly, do you think we can hope for a real cure now?

A: To understand possible future treatments or the possibility of a cure you need to understand the causes of achalasia:
Daniel gave this explanation:
in the development of achalasia there are 3 factors:
1. Genetic background

2. Initial triggering of an immunological reaction which causes antibodies to act against antigens in the oesophagus
3. Disappearance of neurons in the oesophageal wall that are critical for the organisation of contractions and opening the LOS.
So research would be looking at three factors:

1. Genetics – identifying the series of genes present in achalasia
2. Immunological reaction – antibodies identified triggered by previous viral infections
3. Neurons that are disappearing and do not regenerate unlike other cells.

Current research is mainly around 2 - whether it's possible to block the immunological response in 2 at an early stage to avoid 3 (destruction of the neurons) by identifying what antibodies are working and if it's possible to block this reaction. In the future it may be possible to use new genetic knowledge, to understand the sequencing of the DNA and immunological actors in the oesophageal wall and to stop the reaction.
Concerning 3, the neurons which disappear, there is some very early-stage work looking at replacing the lost neurons. It may be possible further in the future – ten/ twenty years - to transplant neurons to modify the motility of the oesophagus. This sounds very sci- fi but it is based on research in the field – i.e. research into stem cell treatment
Majid – we have Connor McCann doing some work on exactly the last point – stem cell treatment in mice. But a long way to go before translating this to humans.

What makes a good centre for treatment?

Q: 'Struggling to find a centre of excellence. Is there somewhere a register of experts (both gastroenterologists, surgeons and nutritionists) that I could refer to find local support?

Q: Aged 64. Diagnosed with Type II achalasia after many years of being advised it was reflux disease. Trying to decide what is best choice between Heller's Myotomy or Dilatation. It seems that the recommended path forward is largely dictated by what sort of expert I speak to – e.g., a surgeon will recommend a surgical solution. How within the NHS can I get a multidisciplinary recommendation so that I can decide what is best for me?

Q: How do I find a solution within the NHS where I will be monitored into the long term so that I can try to keep it under control as best I can?

Q: After two dilatations and being diagnosed in 2020 but with symptoms for roughly 5 years prior to this, I know that this is a progressive disease and I would like advice from an achalasia specialist about what they feel would be the best plan of action to take. I live in Norfolk, so where should I go to get that advice?"

A: Majid/ Ahmed: Look for volume – i.e. surgeons who have performed many operations.
Ahmed would always refer patients to someone who specialises in the field and has done many procedures, not a general surgeon. Manometry 90% of time will be clear but 10% of time symptoms don't correlate with the indications of the test, but if looked at by experienced people they will recognise achalasia. Which is why you need experienced people doing/ interpreting the test, at centres where a lot of tests are carried out.

A good centre would also be where gastroenterologists and surgeons are working together and where treatment is tailored for the individual. Look for a clinical background where practitioners are seeing patients in clinic.
Majid: Essential to get a proper diagnosis via manometry prior to any treatment – it gives precise figures and diagnosis. In the past before high resolution manometry was widely available, surgeons were diagnosing and accepting patients for surgery based only on endoscopy and a barium swallow test.

We don't keep a register of centres and surgeons as expertise and personnel can be fluid, and may change frequently. Get a diagnosis then take time to decide where to go for treatment - get a recommendation based on the above.

Dealing with a swollen / distended oesophagus

Q: A GI Dr said there are tablets to make your oesophagus contract. Is this true, can they be taken long term & do they have any long-term effects?

A: Dr Albusoda:
These cannot be used for achalasia as the requirement is for contractions to take place in a coordinated fashion and the tablets can't do this.
It is more likely that tablets are prescribed that make the oesophagus *relax* but these are only really useful for pain relief
Daniel – no pill can make the oesophagus contract if there are zero contractions as a starting point (Type 1). The pills can't correct the uncoordinated Type 3 contractions.
The pills described are used for a different disorder of the oesophagus, not achalasia and should not be taken for achalasia

Delayed gastric emptying

Majid asked question to Daniel : How often is delayed gastric emptying seen in achalasia and how often is a pan gut dysmotility/ slow transit colon problem seen?
Daniel – Not often they but don't look for it. Delayed gastric emptying has been mentioned in the literature but not with pan dysmotility unless a geriatric patient with a genetic disorder
Ahmed – Don't often hear patients complaining of generalised motility problems, probably because they are more focussed on the big problems they have with achalasia.
Majid – It's interesting sharing the panel with Ahmed and Daniel today as we all see the patients at slightly different stages. Majid finds that constipation is one of the biggest problems he has to deal with post operation, as well as pain (if they have pain beforehand, they may also have it afterwards). Finds generally that people who are very constipated or have confirmed slow gut transit have more problems with their response to myotomy and with swallowing and that when constipation is treated, they get better overall.

Diet

Q: I have decided not to join my son and husband on holiday. The stress can be too much: travelling and finding things to eat and worrying if I get food stuck

or pain. However I want to go. Any ideas when travelling? I take powder shakes normally to mix up but not ideal for a week.

A: Majid: It's important to relax so you should go on holiday, take the shakes and take your medication history. Avoid risky emergency treatment e.g., urgent endoscopies out of hours where there's a risk of perforation. Stress does tend to make achalasia worse

Q: What is the advice for diet after a Heller's Myotomy, and for how long?

A: Majid: This depends how surgery goes but as a general rule 24 hrs only liquids then a few days of soft diet and then solids introduced gradually.
It is important to avoid vomiting or retching in early days as this may cause damage or aspiration and heaving may occur due to pain meds/ anaesthesia so need to avoid adding to nausea by introducing food too early. But he would not want return to the old days of weeks on liquids.

After dilatation recommendation is usually one day liquid, one day soft

Medication

Q: With Achalasia I am fine one minute and feel NORMAL dancing or walking, then within an hour I can have pain or food sticking so my mood goes down and pain kicks in. Medication such as Amytripline is long term. Is there any good ideal pain relief to ask for liquid form or dissolve etc which could see us through a few hours or days Eg Buscopan for spasm does this come in liquid form we can ask for?

A: Amytripline is available in liquid form, we don't think Buscopan is but check with the pharmacy about any medication that might be in liquid / syrup form.

There were some questions in the Chat which were answered by the panel:

Q: Partial wrap but can't vomit

Majid – would need to see barium swallow result and how partial the wrap is. Usually no problems with a gentle / partial wrap. Get checked out

Q: Had friend who had wrap that became partially undone which is pushing him towards the POEM.

Majid – with any surgery or intervention you need to ensure you get best first shot. There always needs to be a clear surgical target. Go with something tried and tested. You can have a repeat Heller's but if done properly shouldn't be needed
POEM can help in some circumstances, if, say, if a Heller's hasn't worked as POEM can go a few cms higher.