

Achalasia Meetup

5-7pm, Tuesday 9 April 2019

Attendance 40 People (33 with Achalasia)

Agenda

1. Questions with Majid Hashemi
2. Overview of what Achalasia is
3. Presentation from Yan Mei Goh, Imperial College London
4. Update on development of new Achalasia charity

A note on abbreviations

LES/LOS = Lower Oesophageal Sphincter, situated at the bottom of the oesophagus and opens into the stomach

POEM = Peroral endoscopic myotomy

GERD = Gastroesophageal Reflux Disease (also known as GORD)

1. Questions with Majid Hashemi

Attendees sent questions in advance. Mr Hashemi answered some in the meeting and others by email.

Is it possible to have both achalasia and GERD?

Yes they can, it's quite a common pairing. There are two sorts of reflux or regurgitation.

- One problem is the regurgitation of food and saliva that people with untreated, or unsuccessfully treated Achalasia get. This is intra-oesophageal – the food hasn't made it into the stomach, it's been prevented by the unrelaxing Lower Oesophageal Sphincter.
- The other is reflux, where a patient may or may not have had treatment (myotomy, POEM or dilatation) and stomach contents, including stomach acids, pass up into the oesophagus. This is also what happens in people without Achalasia. This is what happens in GERD – though not all reflux is GERD
- Both of these are amongst the most worrying manifestations of Achalasia. Particularly when they impact on a patient's breathing, or cause chest infections when patients inhale regurgitated food.

What are the long term effects of taking Omeprazole? I've had conflicting information on this in the past.

- One of the most prescribed drugs. It is very safe to use in the short term and safe in the long term but there are side effects associated with long term use including osteoporosis and possible cardiac problems.
- You can of course use acid suppressants like Omeprazole, and for some people that's the only option.
- If you have a myotomy and cut through the LES, leaving it open and space for acid to travel up, it's a mechanical problem and the mechanical fix is to provide a fundoplication (wrap) that creates some sort of replacement for the LES (using the elastic stomach) It's not a perfect replacement but it work very well. This also enables you to maintain a normal stomach ph (rather than suppressing acid with medication) which can support good nutrition.
- Everything has an impact - lifestyle can make a difference. Mr Hashemi has had dozens of patients – with and without Achalasia – who have overcome reflux symptoms with diet and lifestyle.

I have noticed I have been suffering from a 'dry mouth' is this related to achalasia? I never had this before my diagnosis so would be interested to know if it is linked with this condition? Do any of your patients have this too and what do you recommend?

- It is worth considering that it may be caused by another condition, having Achalasia doesn't mean you won't develop other conditions. Do get checked by other health professionals.
- Sometimes medication can cause a dry mouth
- Think about other things. Get your blood sugar checked as dry mouth is one symptom of diabetes.
- Speak to your dentist – get your enamel and glottis checked. Regurgitation can impact on teeth and gum health and there are things your dentist can do to reduce the risk of enamel loss.

Has any research been done on lack of energy or poor energy with achalasia?

- Low energy is probably a result of a range of factors including diet and nutrition, reflux, dysphagia, managing pain. There is no reason why the condition of Achalasia would cause low energy in and of itself. Address the basics: ensure good diet (maybe vitamin supplements) and stress management

How do I find a specialist in achalasia who can advise me? The first consultant I was sent to had never treated achalasia. The second was not much better. I am losing confidence and I would like the next person I see to be able to help me!

- Specialists can be found through GP referral and via local gastroenterologists.
- Ask people at the Achalasia meetings about their experiences (although people have had different experiences with the same clinicians)

Apart from the chest pains, I am now getting 'twitches' or spasms, like muscular spasms, roughly in the centre of the chest (los?), sometimes slightly to the left (stomach?). They are short but incredibly annoying and disturbed my sleep badly by being constantly jolted awake. What can I do to help with them? Should magnesium powder help?

- See link to previous session about spasms <http://achalasia.mooc.com/showthread.php?tid=120>
- There are three sorts of pain associated with Achalasia.
 - One spasm pain we don't understand.
 - Second – pain from reflux – this usually experienced retro-sternal (centre of chest) – this can be managed with diet and / or medication – PPI like Omeprazole. If you have had a myotomy with a fundoplication (wrap) and are still experiencing reflux Mr Hashemi would consider adjusting the wrap to make it more effective.
 - Third – pain and spasm from the LES. This could be due to obstruction by food, or other reasons.
- Treating the pain in Achalasia patients is a difficulty – it may get better, or it may not with surgery. In practising since 2001 Mr Hashemi doesn't recall operating on a patient and spasms getting worse, but occasionally with other clinicians, spasms have changed in their nature or become worse, sometimes after a period of time.

A surgeon said that because I don't have 'typical' achalasia, surgery would not stop the chest pains. Is this likely to be true?

- No, as above. There isn't enough evidence about pain and surgery to make a blanket statement. Chest pain outcomes are unpredictable. In Mr Hashemi's opinion surgery is still the most effective option.

Has any data been gathered or research analysis carried out on people who suffer from Achalasia e.g.: age, sex, life style (smoking/drinking/drugs etc.), height/weight, fitness levels, any family history, type of jobs, hours of work, living locations (city, town, seaside, country etc.) or other past/current medical conditions.

- Yes. To some extent, there is lots' of data online (look at Google Scholar) and the new website for the Achalasia charity will aim to be a hub.
- There is no occupational association.
- There is ongoing research on the impact of stress

- There doesn't seem to be a hereditary connection, other than in the few instances where another family member has Achalasia the patient will have awareness and understanding.

Once a Heller's myotomy is carried out on the oesophagus/LOS the muscle is cut, only the mucosa is left which is wafer thin, when the scar tissue grows over how strong/weak is this area vs. the rest of the walls of the oesophagus and how thick does the scar tissue become on average.

- The mucosa is VERY thin, 2 / 3 mm but also very strong. You suture into it during surgery. During a myotomy the mucosa (or inner lining of the oesophagus) is not cut but the muscles around it are.
- The scar tissue in the muscles develops and strengthens over days/weeks. If for example you ate a piece of toast immediately after a myotomy you might perforate the mucosa which is why patients don't eat solid food for 2/4 weeks after surgery.

How often can a situation arise when a further operation is required following a Heller's myotomy, can achalasia return, e.g. type 1 lead onto types 2 or 3.

- Rare. Irrespective of type

Does the oesophagus return back to its original shape after having a bird's beak for a prolonged period of time (i.e. several months or longer)

- No. It doesn't. The point of the 'bird's beak' will be more open after treatment as the LES is more open.

The dilemma: Should I have another dilation or surgery?

- In the US 80% of patients get Myotomy as first intervention. Not the case in the UK where most patients Mr Hashemi sees have had dilatation or botox.
- When healthy the tissue of the oesophagus is very strong. After it's been botoxed, dilated or if it's inflamed it's less tough (think wet blotting paper)

2. What is Achalasia?

Achalasia is the presence of one or more of the following symptoms

- Non-relaxing LES
- Simultaneous contractions of the oesophagus
- High pressure contraction

Majid drew a time / pressure graph showing how pressure in a normal oesophagus moves in waves as food is swallowed.

When you swallow you want the sections of the oesophagus below where the bolus (ball of food) is to be relaxed, allowing the food to be propelled down.

In Achalasia the inhibitory neurons have stopped working – so they don't allow the oesophagus to relax in the correct order, or at all. The LES is tightly closed and food and liquid can't travel down to the stomach. Regurgitation can occur.

We don't know why this happens.

Some research and thinking into possible viral causes, including the chicken pox virus.

How does food get down when the LES is tight in Achalasia?

Ultimately for some patients it doesn't, it stops entirely.

Pressure works – gravity, liquid can all help food move down.

In health, the contractions of the oesophagus work together to move the food down. The swallow at the throat (pharynx sphincter) indicates to the LES that it should begin to relax to allow the food to pass into the stomach. In health, there are also occasional periods of LES relaxing, venting is going on. In Achalasia

there is no relaxation of the LES as the food passes the pharynx and down the oesophagus. As far as Mr Hashemi is aware no-one has measured any venting, periods of the LES relaxing in people with Achalasia.

The pressure of the contractions in the oesophagus can vary. In health, amplitudes can range from 30 – 180 – some people have much higher pressure than that. The pressure often diminishes with age. The impact of reflux on the oesophagus can lower pressure.

Can Achalasia progress further down the digestive system?

There are other gut motility diseases and conditions. Achalasia is limited to the oesophagus.

Mr Hashemi, anecdotally, thinks there may be a loose association with gut motility problems. A very few of his Achalasia patients have some delayed stomach emptying, some have constipation.

General trend – in a small number of patients

Achalasia is a nerve issue – it is connected with the autonomic system, the nervous system that regulates the blood pressure etc.

Mr Hashemi has had conversations about how in reflux or Achalasia patients, maybe there is something more general in the autonomic that is involved as well? He's not identified anything in his 20 years of practice, no research currently.

Mr Hashemi follows patients up and has repeated wraps where patients are experiencing reflux. One or two that he completed 10 & 15 years ago and a small number where original surgery was completed more recently, in the last 5 or 6 years. During the surgery he can calibrate the tightness of the wrap to ensure it's not too tight but is tight enough. Two ways of doing this, with a pressure reading in the sphincter during the operation or with an 'endo flip', which enables him to see the tightness. Mr Hashemi now has a low threshold for investigation – which might include manometry, barium swallow, endoscopy, or pH study. He has never regretted returning to re-do a wrap. The message is to get a regular follow up.

There was some discussion about POEM, which doesn't include a wrap. Mr Hashemi recognised that many skilled clinicians are enthusiasts and there have been good outcomes from POEM. He wonders if the outcomes of the procedure will remain as good as it gets rolled out to less skilled (because of less experience) or less enthusiastic clinicians.

There is a good body of evidence for the effectiveness of myotomy. POEM can create a longer and higher cut in the oesophagus but myotomy is also able to cut quite high. The majority of myotomy cuts are no longer than 10cm. Myotomy shouldn't necessitate cutting through the patient's diaphragm. With laparoscopic (keyhole surgery), there are five cuts in the abdomen to get access to the inside but then nothing other than the muscles of the oesophagus should be cut. In POEM you have no incisions on the stomach as you go down the throat but you have to cut the mucosa, which you don't in a myotomy. No surgery is 100%.

Mr Hashemi has become more sure that he would recommend myotomy and dissuade a member of his own family from POEM (a stronger position than he has ever taken before!)

The final decision has to be between an informed patient and the clinician treating them.

He referred to people with Achalasia at the meeting who had managed their reflux through diet and lifestyle changes and one person present offered their experience. They have had the POEM procedure which was effective for them. They control reflux through diet – limiting acidic foods, following an almost vegan diet, drinking lavender oil in water and camomile tea. They don't eat 3 hours before bed and try not being too active after eating. They avoid lemon in water.

3. Presentation from Yan Mei Goh, Imperial College London

Please see attached presentation, some key notes below.

Oesophageal cancer is the ninth most common cause of death from cancer in the UK and the team are trying to develop less invasive, early detection options using breath testing.

There is an old study from early 1990s that suggests a **very slight** (2.8- 3%) increased risk of oesophageal cancer in people with untreated Achalasia (lower in those with treated Achalasia). This was based on a study of squamous cell carcinomas which are now less common (having been overtaken by Adenocarcinoma cancers which are now more common). Mr Hashemi has only seen one Achalasia patient who has developed cancer in his practice, who had avoided surgery, having seen hundreds of patients since 2001. He would like to reassure everyone that the additional risk is low, though you should of course always speak to your clinician if symptoms change or get worse.

There are now more than 250 Volatile Organic Compounds (VOC) we can identify in breath and breath testing for cancer works, for lower GI cancers and others at the moment.

Imperial College are inviting Achalasia patients to contribute to a study to help develop a breath test for oesophageal cancers. Achalasia patients would be in the positive control group – we have symptoms similar to oesophageal cancers, but do not have cancer.

There was a brief discussion about SIBO (Small Intestine Bacterial Overgrowth) which is also detected with a breath test. Mr Hashemi noted that anyone on long term PPIs will probably have some level of SIBO, in fact most patients with GI issues have some level of SIBO. He no longer breath tests before prescribing a probiotic, only breath testing if he suspects the SIBO is of a significant enough problem to warrant a stronger treatment (high dose antibiotics)

This is similar to his experience with the PEP test, which tests for Pepsin in saliva, an indication of lots of reflux / GERD. However, people with reflux can test negative for Pepsin and people without reflux can test positive.

With both of these things Mr Hashemi prefers to be guided by the symptoms that patient is actually experiencing.

What happens if you participate in the study?

There are two tests, which will take place at the hospital.

- A breath test, breathe into a device.
- A saliva test, place a small plastic device on the tongue for 3 minutes. They want to collect two samples so a total of 6 minutes.
- Also need to complete a questionnaire about medical history
- Will need to fast for 6 hours before the test
- Transport costs covered
- The commitment is to a one off study
- Although, they are keen to invite us back to develop into a longitudinal study. Taking part in the first visit doesn't commit you to this.

Thanks was given to Yan Mei for her presentation.

Sign up was available at the meeting, but anyone not present who would like to participate can email Amanda Ladell. amandaladell@yahoo.co.uk

4. Update on development of new Achalasia charity

- At the last meeting (22nd January 2019), it was generally agreed that setting up an Achalasia charity was a good idea. A working party has been set up and are exploring how to establish an appropriate structure, eg. charity working within a company limited by guarantee.
- The proposed name is Achalasia Action (ACAC)
- Broadly it will exist to
 - Raise awareness
 - Support people who suffer
 - Encourage research
 - Raise funds
- One group member (Neil), is designing a website – it will contain case studies, information (A Patient’s Guide to Achalasia created by the group is being refreshed). For a while, Meetup will run as well.
- There was some conversation about whether the website should contain a directory of where you can find treatment, what centres are available. Some questions and concerns about liability (legal and moral). At the moment, case studies and supporting people with Achalasia to become expert patients actively managing their own condition was favoured. Different people can have very different experiences with the same clinician. The website will probably include a mailing list sign up for events and an occasional newsletter.
- There was a question about membership, would people be asked to pay a subscription? Not in the first instance, something for future discussion.
- Suggested future t-shirt slogan – ‘You can have your cake and eat it’
- There was some hope to extend the reach, possibly with more regional meetings (one member present is moving to Scotland)– creating a sense of community across the UK and further afield. Amanda has had people attend the Sunday meetups from all over the world.
- The working party are looking for people to fulfil more roles. Amanda has offered to be treasurer, as she has experience in a similar role, but is happy for anyone else to take that role if interested. Get in touch with Amanda if you are interested in getting involved and how you think you will be able to help.
- Those present asked for a more detailed outline of the options for the structure of the charity and clearer questions about what the working party would like guidance around. This could be circulated in a paper prior to a meeting, or shared via Meetup?

Thanks given to Amanda, Alan and Mr Hashemi for their ongoing support in making the meetings happen.