

Achalasia Meeting
5-7pm, Tuesday 19 September 2019

Agenda

1. Presentation on I-PASS from Prof Giovanni Zaninotto
2. Update on development of new Achalasia charity
3. Questions with Majid Hashemi

A note on abbreviations

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

POEM = Peroral endoscopic myotomy

Presentation on I-PASS from Prof Giovanni Zaninotto

Professor Giovanni Zaninotto gave an introduction to a new project, I-PASS, which stands for International - Patient reported outcome of Achalasia Symptom Score.

The presentation is available as a pdf – following are some general notes and a slide by slide description and general notes from the presentation.

Achalasia is

- Hard to diagnose
- There is no standardised treatment
- No cure, treatment is palliative and aims to improve symptoms

Prof Zaninotto is a specialist in oesophageal disease with an interest in Achalasia. Two years ago started developing guidelines for Achalasia treatment, there is currently no standardised way to establish if treatment brings improvement and how to measure the symptomatic improvement

The I-PASS project is creating a questionnaire that will enable patients to report the outcomes of treatment. It will be used by patients at before their treatment and again after treatment. The data will be used to understand, and drive improvements in, the efficacy of treatments for Achalasia. Having a standardised, internationally available Patient Reported Outcome (PRO) is important to enable future projects to develop treatments and improve care for people with Achalasia (for example, major funding organisations only support projects where there is a PRO). The PRO will enable randomised control trials of future treatments to have objective measurement of the outcome.

Stages of the I-PASS project

- 15 Achalasia experts were asked to develop a set of questions that would help demonstrate the efficacy of any treatment. This used the Delphi process (multiple rounds of questions circulated and amended based on feedback). This stage has been completed.
- A group of 15 people, from different countries, with Achalasia will read the questions and be interviewed by a psychologist to ensure that the questions are fit for purpose and to improve the questionnaire.
- Pilot study. 50 people who have not had treatment will be asked to complete the questionnaire, and then repeat it after treatment. Then the questionnaire will be refined. The project is currently funded to this stage.
- Validation study. 750 people who have not had treatment will be asked to complete the questionnaire, and then repeat it after treatment.

I-PASS would like the support of the Achalasia group in the following ways.

- 5 volunteers for cognitive interview (people in the meeting gave their details, the study will interview a cross section. A further call out will follow if people of a different demographic (eg older or younger, male or female are required)
- support for the pilot study (recommend patients to take part)
- help in asking for grants for larger study by endorsing the work (a previous similar study cost around £25k)
- help recruiting treated patients for the validation study
- There will be more information from the study after a meeting with one of the projects funders (ISDE) in November.

The meeting unanimously voted to support the project with a show of hands.

Other interesting points raised during the presentation & questions and answer session.

- Many patients have multiple treatments for Achalasia
- The first record of dilatation, most likely for achalasia was in 1650, using a whalebone with a sponge on it's tip, the first Heller's Myotomy was in 1913
- There is no difference in occurrence of Achalasia in adults globally (that we currently know of) based on race, age, sex. There is a lower incidence of achalasia in children than adults.
- There is currently an increase in cases in South America because of Chagas disease
- There are 6 trials comparing surgery and dilation that Mr Hashemi is aware of – none of them are of the best standard or comparing like with like.
- The Eckhardt score was never validated for outcomes of treatment – only to assess the initial diagnosis and severity of Achalasia. The test scores the severity of symptoms on the following factors
 - Dysphagia (difficulty swallowing)
 - Frequency of regurgitation
 - Chest pain
 - Weight loss

The new PRO should be simple, like the Eckhardt score.

Eckardt Score for achalasia severity

| Sign/symptoms | Score | | | |
|-------------------------|-------|------------|-------|---------------------|
| | 0 | 1 | 2 | 3 |
| Recent weight loss (kg) | None | <5 | 5-10 | >10 |
| Dysphagia | None | Occasional | Daily | Each meal |
| Chest pain | None | Occasional | Daily | Several times a day |
| Regurgitation | None | Occasional | Daily | Each meal |

Score: 0-12

Eckardt AJ, Eckardt VF. Treatment and surveillance strategies in achalasia: an update. Nat Rev Gastroenterol Hepatol. 2011;8(6):311-319.

- There is the potential to create a European network , Achalasia is recognised by the EU as a rare disease, some special funds being available for research on rare diseases.
- The chronic nature of condition was raised, it changes throughout life. The new PRO will just be a snapshot. There was discussion about a longitudinal study being ideal. The PRO is a good starting point for this.
- There was a point raised about people who have not had intervention but have improved their condition over time, receiving good non-surgical advice. The questionnaire will probably include space for people to share their experiences. In the same way tips and experiences are shared at the meetings this study could share that. The international nature of this study may enable sharing of useful, different approaches from other countries.

- Manometry and barium swallow aren't pleasant tests and you don't want to repeat them without good cause. The PRO could help to create a threshold for further investigation / treatment.
- On the question of spasms, an important issue for those with achalasia, Prof Zaninotto said that chest pain was included in the study.

Overview of slides without titles

Slide 1 – Title slide

Slide 2 – Definition of Achalasia

Slide 3 – Manometry of oesophageal function in patient with Achalasia

Slide 4 -- Manometry of oesophageal function in patient with Achalasia

Slide 5 – Normal Oesophagus

Slide 6 – Manometry of oesophageal function patients with different types of Achalasia and an overview of the Chicago classification of those types.

Slide 7 – Overview of incidence (number of new patients each year) and prevalence (total number of patients)

Slide 8 – How Achalasia was managed in the UK, where there are approximately 624 new cases a year and a total of 7,487 patients (taken from NHS records, not including private hospitals)

Slide 9 -- How Achalasia was managed in the UK

Slide 10 – Definition of what a patient reported outcome is.

Slide 11 – Why a patient reported outcome is needed.

Slide 12 – Aims of the I-PASS project

Slide 13 – Stages of the project (a)

Slide 14 – Stages of the project (a)

Slide 15 – Example of the I-PASS questionnaire in current format

Slide 16 – Second example of the I-PASS questionnaire in current format

Slide 17 – Next steps for the I-PASS project

Slide 18 – What I-PASS would like from the Achalasia group

Slide 19 – Who is funding I-PASS

Slide 20 – End slide

Update on development of new charity – Achalasia Action

A working party has been setting up Achalasia Action. They have

- established objectives
- identified and drafted an appropriate constitution
- Created a website - <https://www.achalasia-action.org> (Anita also modelled an Achalasia Action t-shirt!) Please sign up to receive an occasional newsletter, the patients guide is also available to download from this website.

The charitable objectives of the new organisation are

- To provide education and raise public awareness about Achalasia and associated conditions
- To alleviate physical or mental distress of persons with Achalasia including their friends and families.
- To encourage and support research into Achalasia for the public benefit

The charity will be a Charitable Incorporated Organisation (CIO), an application is going into the Charity Commission shortly. This is posted online for comments. Once the registration process is complete the organisation can open a bank account.

The old <http://achalasia.mooc.com> forum has been closed.

There isn't currently any social media for Achalasia Action, the team hope to do this in the future.

The Meetup pages will continue to be used for the time being as they are useful for arranging the Sunday meet ups and larger meetings effectively <https://www.meetup.com/achalasia-104/>

A new Health Unlocked community has been set up – a place for discussion and sharing experiences (the Achalasia Action website is an information place, there currently isn't the resource to effectively manage a forum through this site) You can sign up through the link on the website or <https://healthunlocked.com/achalasia-action>

The meeting gave a huge thanks to the whole team who are making this happen and have given freely of their time, energy and in some cases cash.

ACTIONS

- Subscribe to the newsletter via the website
- Sign up to Health Unlocked
- Anyone who would like to get more involved in organising please email info@achalasia-action.org

Questions with Majid Hashemi

Questions had been submitted in advance of the meeting and Mr Hashemi answered a selection during the available time in the meeting. Questions are in bold, a note version of Mr Hashemi's answers are below each question.

Q. Because of the large cervical osteophyte complex at c3/c4 and great difficulty and risk of perforation getting a standard endoscopy tube past this, I am in effect limited to two options. Either leave things and wait and see, or have the myotomy.

What are the risks of just doing nothing? I've had different views including the possibility of food getting completely stuck. Could damage to the LES occur? One registrar said recovery from surgery becomes more difficult getting older (I'm 65). Also I've been trying fizzy water with some meals, but on one occasion I had severe chest pains for hours including during the night and wondered if sparkling water should be used with caution.

- 65 is not old (hooray!) Numerical age needs to be in perspective, if a patient is fit and active that is significant. Quality of life is no less important because someone is older.
- Sometimes teams focus on less invasive options for older people. There may be medical reasons why, but just because you are older is not a reason not to have a myotomy. Mr Hashemi has operated on patients in their 70s and 80s where safe and appropriate.
- An experienced surgeon can provide a safe myotomy.
- The risks of doing nothing are loss of quality of life – restriction of diet, regurgitation. More damaging weight loss, and chest infections (from aspirating food).
- No evidence either way on drinking sparkling water – different people have different experiences. Some find it very helpful; others experience discomfort. Learning about how you respond is the key.

Q. I would like to know Mr Hashemi views on long term use of PPIs. I have googled and talked to my doctor, but would appreciate his take on this.

- PPIs are Proton Pump Inhibitors – medication that suppresses the production of stomach acid.

- I don't think they are a great choice for the long term. There are side effects that increase with longevity of use – including potential difficulty absorbing some nutrients, cardiac issues and osteoporosis.
- I prescribe on a case by case basis – many of my patients are on PPIs but I'm cautious
- I recommend trying Gaviscon Advance as required – lots of people can get good results
- When taking PPIs everyone will potentially have some degree of SIBO (Small Intestine Bacterial Overgrowth). Some people may experience symptoms, others may not. PPIs will alter your gut flora – this is what I don't like as an Upper GI specialist. PPIs turn healthy acidic stomach juice to alkali. The reflux changes from acidic to alkali – it's not fixing the problem and it's changing the ideal, naturally occurring state.
- For the short term - months / couple of years – FINE
- I would really investigate if you need them.
- A possibility is H2 antagonists – the last generation of treatment are weaker and taken as required not all the time eg ranitidine
- Diet and lifestyle changes can help.
- The overall message is be sure that you are taking PPIs for a good reason.

Q. (i) I have been told that dilatation, dilatation, dilatation is the only way (lots of dilatation!) - completely loosening up the oesophagus so that gravity is the stronger force opening up the constricted oesophagus.

- There are studies comparing Myotomy and Dilatation where Dilatation results are favourable. But are they actually? It's worth looking more at all the factors involved – expertise of the clinicians, complexity of condition of the patients.
- Mr Hashemi would go for a Heller's Myotomy if done well on a fresh oesophagus (no prior treatments)
- Has done dilatation on patients (not for people with Achalasia)
- Doing a HM after multiple dilatations is tricky as there is more scar tissue. In 18 years has had less than 10 'fresh' patients – in the US most people go straight to HM.
- Risk of perforation is much less, recovery is quicker when HM is first treatment.
- Each patient needs to be assessed on a case by case basis

Q. (ii) Can someone who has had a Heller's Myotomy + fundoplication have a POEM procedure?

- Mr Hashemi would want to know why? Has the Heller's Myotomy not achieved the desired result?
- The suggestion is the surgeons can cut higher up the oesophagus with the POEM than with the myotomy.
- If the myotomy hasn't worked – what is going on? If the surgery has been shown to anatomically work then why do more? It's basic plumbing! Remember that the treatment is only effective for the swallowing symptoms of Achalasia.

Q. I had a Heller's Myotomy 3 years ago which has been successful in restoring my life. Recently I have been experiencing changes which have been diagnosed as dumping syndrome. Could Mr Hashemi please explain this and what his management would be. I have been informed it can occur post gastro surgery.

- Alan gave an overview of Dumping Syndrome, which is when the food goes down more quickly than it should – and tricks your body into releasing more insulin. When the insulin: sugar is out of balance you get unpleasant symptoms including sweating, palpitations, nausea and you can faint. Manage dumping syndrome with diet. Balance – what you eat / amount of sugar e.g. sugary food like cornflakes could trigger dumping. A low GI diet helps the problem – body processes food slower.
- Dumping syndrome is not common after myotomy. Maybe they have cut the vagus nerve? It's worth getting an investigation

- If your food is not going down / going down slowly – dumping is not a problem (this is more relevant for people with Achalasia)
- If your LES is loose you may get reflux – and food may travel more quickly.
- Looking for food with the right texture is the right priority for those with achalasia, then keeping nutritional balance is important.

There was a brief discussion around feeling anxious about eating when you are worried about regurgitation and swallowing. Other Achalasia patients have experienced this, especially around eating in social situations. One patient reported having had a positive experience with therapy to address this issue. Achalasia is something we have to learn to live with.

Q. 1 – Could anyone use Ayurvedic approach through NHS and does it work ?

- Ayurvedic (traditional Indian medicine) not available on the NHS as far as Mr Hashemi is aware
- Anything that reduces stress - promotes / smooth muscle relaxation is good for management of Achalasia

Q. 2- How do we decide what's best to do first if manometry failed ? (eg manometry device curls up rather than passing through into stomach)

- The next step is to go to a barium swallow

Q. 3- Is a barium swallow necessary if manometry fails?

- Barium swallow can be very helpful – helps to identify exactly where the issues are
- 50% of manometry don't work, especially in patients with Achalasia – the catheter curls around
- Ideally needs to be done in a relaxed environment, which is difficult in hospital. It's an unpleasant study for the patient.

Q.4- I had a combined endoscopy and manometry at the same time, literally together while you awake - has it ever been performed on anyone before at UCL? It has created lot of side effects for me - lots of tummy upset after the horrible test.

- Mr Hashemi has not done both together himself – as a research fellow was aware that it can be done if required.
- You ideally have your manometry in a good relaxed state – endoscopy is more uncomfortable.
- It's safe to do both of these together though not at all pleasant. Talk to your clinician and ask why they are taking / have taken this approach.

Q. After having a POEM procedure in January 2019 my condition has improved but not completely resolved. On the 30 July 2019 my Eckhardt score was 4 principally for the symptoms of dysphagia and very occasional regurgitation. My manometry demonstrates a failure of relaxation of the lower oesophageal sphincter, as well as lack of peristalsis. At my last appointment a clinical research fellow on their team suggested a repeat POEM or pneumatic dilatation with risks involved with both procedures. What would your medical opinion be on my symptoms?. It was also suggested to me to stop taking 20mg omeprazole daily which I have done since the beginning of August 2019 but noticed stomach cramps/pain, IBS symptoms. I changed my diet to gluten free as much as possible which has lessened the symptoms. Shall I continue not taking omeprazole as I am worried about the symptoms I have experienced and if they are causing more damage to the oesophagus. Are my symptoms due to coeliac disease ? I would appreciate your medical opinion on any or all of the questions that I have put to you and thank you in advance.

- Be very cautious of further treatment - have a full set of investigations again – endoscopy, manometry and barium swallow.
- Check the investigations – has all the improvement that can be made, been made? Check what sort of improvement could be made with further treatment.
- Get a 2nd and even 3rd opinion.
- Sadly, recurrent Achalasia is very difficult to manage.

- Revision surgery is always tricky with increasing risks and a longer recovery.

Q. I went for a routine pre-assessment appointment last week and the nurse asked me what post-op arrangements I'd made. I asked her what she meant and she said she'd noted I lived alone and without anyone to support me. She reckoned I'd need assistance for up to four weeks for things like shopping, domestic cleaning, lifting items, generally keeping an eye on me. In years gone by we had convalescent homes but not now. None of this has been raised until now. I've spoken with someone else who has had the operation who said I certainly would need help and suggested private nursing home for a while. I wondered if you had a view, or even if Mr Hashemi has recommendations for the post-operative period.

- Mr Hashemi does not think a convalescent home is necessary not unless you have other issues
- Surgery should be keyhole and you should be able go home after 2/3 days after uncomplicated surgery
- Don't do any lifting for a month or so – this is where help could be required.
- Asking other patients about their experiences and what they have found helpful post operatively.

Q. Does a baggy oesophagus matter? (commented on by a radiographer after a chest x-ray unrelated to Achalasia)

- The oesophagus can get baggy, technically called a mega- oesophagus, this isn't the state the oesophagus should be in.
- Always worth a follow up after treatment to ensure that the all improvement that can be made has been.
- Treat the blockage first (the unrelaxing LES) if not, the options are to get round the symptoms by managing diet.