

ACHALASIA MEET UP - 12 DECEMBER 2013

Brampton House, Hospital of St John and St Elizabeth,
60 Grove End Road, St John's Wood, London NW8 9NH

Speakers: Mr Majid Hashemi – Consultant Upper GI Surgeon and Dr Rehan Haidry Consultant Gastroenterologist

Attendance: 44 total. 30+ achalasia patients. 11 had had more than two dilatations (five more than twice); 9 had had myotomies (two twice); Fundoplication 8 (one twice); 1 POEM procedure.

What is Achalasia?

When you swallow food, you need a wave of active muscle contractions working in a coordinated series down your oesophagus to move the bolus of food progressively from your mouth to your stomach. The lower oesophageal sphincter (LOS) valve (by your diaphragm, stopping stomach acid rising into your oesophagus) is at the bottom of your oesophagus and this should also relax in its turn in order to let the food pass through to the stomach. You also need the pyloric sphincter (at the bottom of your stomach) to be working properly so that your stomach can empty, and your bowel to empty normally. The *Angle of His*, the angle formed where the oesophagus joins the stomach, needs to be properly positioned. The upper oesophagus needs to prevent food rising upwards.

With achalasia, the nerve system does not drive the muscle contractions effectively. Several things can be failing to work properly: a non-relaxing LOS; high resting pressure in the lower oesophagus; poor contractions in the oesophagus; simultaneous and badly co-ordinated contractions in the oesophagus.

The pathology indicates:

- Loss of inhibitory ganglia (nerve cells) in *myenteric plexus* (the muscular coat) of the oesophagus.
- Non-relaxing sphincter
- Degeneration of inhibitory nerves in oesophagus – unopposed action of excitatory neurotransmitter *acetylcholine*
- Loss of *cholinergic neurons* over time leads to loss of tone, dilation, simultaneous contractions (ie big baggy oesophagus)
- Nerves gone but muscles work

(Sometimes other, physical, causes can affect the process, for instance the presence of a gastric band can lead to achalasia-type symptoms – triggering the oesophagus to stop working)

Suggestions for underlying cause:

- Viral? Measles? *Varicella zoster virus* (linked with chicken pox and shingles? *Chagas* disease (a tropical parasitic disease)
- (the stomach itself can also fail)
- Genetic? (possible genetic predisposition)
- Nitric oxide? – poor manufacture in body
- Autoimmune? – weak link

Achalasia is 'easy' in medical terms to diagnose, but the causes are not known. 5 – 7 years usually before diagnosis. Some people have it at 16 years of age, another at 12. It can be thought they were anorexic.

Half of achalasia patients have an oesophagus not much more than 3 cm in diameter, which is normal.

Symptoms:

- Dysphagia (ie difficulty in swallowing)
- Regurgitation
- Reflux
- Pain
- Respiratory problems
- Laryngeal problems

Diagnosis and investigation:

- History
- Endoscopy
- Barium swallow
- Manometry

Endoscopic Therapy for Achalasia (Dr Rehan Haidry)

Achalasia is one of the benign oesophageal diseases

- 0.5–1 per 100,000 cases diagnosed per year
- No clear age predilection
- Characterised by loss of enteric neurons
 - *Aperistalsis* (absence of the wave of muscular contractions) + impaired LOS relaxation
 - *A1* response (medical term for oesophageal reaction) triggered by viral infection
- Approach is first to rule out anatomical lesions
 - Endoscopy +/- radiology
 - Endoscopy may show as normal in early stages of disease
 - *OGD* (oesophago-gastro-duodenoscopy) diagnostics in 1/3 and radiology in 2/3
 - Endoscopy + history + barium meal + manometry

High resolution manometry:

Not comfortable!

- A series of pressure sensors, less than 2 cm apart, at intervals from pharynx to stomach
- Recordings analysed and displayed as:
 - Line plot
 - Spatio-temporal plot
- Sometimes the LOS pressure is so intense that doctors cannot get tube past the sphincter

Achalasia Types

(but the classifications are not always very clear cut)

- Classical Type I
- Compression Type II
- Vigorous Type III

Classical

Lack of peristalsis (muscle contractions).

Failure of relaxation of LOS

Compression achalasia

Ineffective, asynchronous (uncoordinated) peristalsis
Failure of LOS relaxation.

Vigorous achalasia

High amplitude peristaltic spikes
Asynchronous / vigorous contractions in oesophagus)
Failure of LOS relaxation

Treatment options:

- Lifestyle/diet/drugs
- Botulinum toxin (ie 'botox')
- Pneumatic balloon dilatation
- Surgery
- POEM (per oral endoscopic myotomy)

Pharmacological

- Calcium channel blocks
- Inhibitors ?
- Nitrates

Initial improvement 50 – 90%

- Diminishing improvement
- 30% side effects
- used as a bridge

Botox injection

Injection of botox into the LOS.

- influence in LOS tone
- improved symptoms
- decreased LOS pressure
- improved oesophageal emptying
- 3 or 4 minutes for a botox injection
(3 in the room reported injections that did not work)
- No sustained benefits
- 50% relapse within 3 months
- Used for those who have limited surgery options
- 1 in 10 have a perforation after dilatation

Pneumatic Dilatation for Achalasia

Best outcome:

- greater than 40%
- women
- normal oesophagus
- 3-5% perforation rate
- 15% get chest pain

Does it work?

- 201 patients in a study – 2 groups – split up for pneumatic dilatation or myotomy
- Outcomes monitored
- Follow up over 4 years – no difference between 2 groups

POEM:

For oesophageal achalasia with no skin incision. A Japanese expert has visited London recently and supervised training in the technique, which involves using an endoscope carefully to tunnel through the wall of the oesophagus and cut muscles within the wall itself.

1. Submucosal skin incision tunneling
2. Submucosal tunneling beyond GE junction
3. Dividing circular muscular bundles
4. Complete division of minor circular muscular bundles (myotomy from inside)
5. Closure of mucosal entry
6. Procedure takes 2 ¼ - 2 ½ hours
7. No stomach wrap-around
8. POEM here to stay?

*

GENERAL:

- Early diagnosis essential
- Important to treat the patient, not the test results
- 5 – 10% risk of perforation
- Wide age group: 16 – 80s age group with first presentation

Treatment:

Two parallel strategies

- Avoiding stress, and exacerbating factors (lifestyle and eating habits can affect swallowing and digestion issues)
 - Medical
 - Endoscopic treatments
 - Surgery

Spasm

- Calcium channel blocks
- Dietary
- Stress
- *Buscopan* (medication that helps relieve spasms)

Reflux

- 'Real' reflux
- Fermentation of food inside the digestive system
- Anti-secretory drugs (ie medication that prevents secretion of eg stomach acid like omeprazole)
- Pain treatment to soothe effects of reflux – Sucralfate
- Relief of obstruction
- Improvement in gastric emptying

Questions

1. Is Achalasia stress-related? – *Yes, there seems to be a link.* Warm water stops spasm progressing. Suggestions from patients to try water bottle/warm wheat bag held across the chest can help especially. Discussion re effect of constituent parts of cannabis helping muscular spasms like, say, for MS patients, subject to legal constraints. Fizzy drinks can help – champagne?!
2. Is it usual for someone who has had a number of dilatations to have blood at the back of the throat for a few days? *Yes, because muscles are sometimes ruptured, but blood should only be apparent for a few days at most.*

3. Why do some people with achalasia get bile reflux? *The sphincter valve(s) can remain open and allow bile from the gallbladder to rise in the digestive system. A hospital can test whether reflux is bile (an alkali) or stomach acid.*
4. Guidelines for achalasia management? – *Very scattered practice. Diagnosis and pathway standard – treatment variable*
5. Does achalasia get better? *Yes, from time to time. It shouldn't get worse. Mix of vigorous and no contractions - varied in group. Need lifestyle changes and removing the stressors.*
6. When would be the best time to have the operation? *It is always best to try for the conditions to maximize chances of success first time round, as there is rarely a good second chance. Dilatation occurs in about 40% of cases in centres of excellence, but repeated dilatations make it harder to achieve success further down the line, eg with Hellers Myotomy.*
7. How can we avoid spasms of pain that occur randomly? – *Buscopan, nitrates, calcium blockers. Often gets better over time. Relieve obstruction downstream.*
8. Is the cause of the retrosternal pain known (the one that feels like a heart attack, not reflux)? *It is hard to treat but will come to an end and is not in itself harmful. Buscopan, eating bananas, drinking Actimel, taking Manuka Honey, coconut milk, fizzy water, all can help. But chest pain for other reasons also has to be excluded!*
9. Having had a *Dor Fundoplication* wrap (a variation of surgery to wrap part of the stomach round the base of the oesophagus) – is there any reason why a POEM procedure could not be carried out? *Covers the front of the stomach. Cannot have a POEM after Dor wrap.*
10. Heading towards 3rd myotomy (50/50 results – trying to find out why it didn't work) so would like to know of alternative procedures and what the future is going to be like with this condition? – *Need to get it right first time but there is hope. Sometimes it may be that the myotomy is not at the appropriate depth, but each case is individual.*

Dysphagia: Categorising its severity:

0 = none, 4 = disabling

- Excellent – no dysphagia
- Good – dysphagia once a week or less
- Fair – more than once a week requiring dietary adjustment
- Poor – dysphagia preventing ingestion of solid food

Summary of a medical trial:

(Pelligrini and others)

- 168 patients aged 11 – 96, 29% had oesophagus diameter of more than 6.7 cm
- 49% had previous dilatations (average 2)
- 20% had botox
- 90% good or excellent results

Reasons for poorer results:

Dysphagia

- If there is transmucosal fibrosis (damage to muscles) – no remedy by myotomy
- Technical reasons in the rest
 - 10/43 1st 3 years 23%
 - 4/124 next 4 years 3%

Persistent dysphagia or recurrent dysphagia:

- In 5 because of transmucosal stricture
- In 11 persistent because of incomplete myotomy
- 4 mal-positioned fundoplication

General:

- gassy drinks help with pain
- Muscle can grow back again
- If it is treated well at the outset, results are generally going to be good.

- If you are OK after first year, you are likely to be fine
- Surgeon sees 4 or 5 achalasia patients every week, but has seen only one cancer patient as a result of achalasia over a period of 10 years.
- On the issue of fears about exposure to acid reflux and Barrett's Oesophagus, it was not worth doing an endoscopy every year. All who had a myotomy without a wrap had reflux. Do NOT need regular testing after achalasia surgery but always the situation is individual.
- No association between ulcers and achalasia, but ulcers are common at any rate in the population. Peptic ulcers are common in high risk people. Heliobacter pylori is being given higher importance as 90% of ulcers are associated with this.
- One patient in the room reported having a Laparoscopic Heller Myotomy with fundoplication procedure in July 2003 and hasn't had the symptoms since.
- Mr Hashemi had done well over 100 myotomies and only a handful needed revision.
- For a first time patient – dilatation or myotomy is the definitive treatment.