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Review

Management of achalasia in the UK, do we need new guidelines?

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HIGHLIGHTS

- Questionnaire to UK Upper GI specialists on achalasia management.
- No current UK guidelines.
- NICE guidelines refer to 2011 SAGES guidelines.
- Only 15% have benign MDT to discuss complex achalasia cases.
- Disparities in management with deviations from current US guidelines.

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ABSTRACT

Aim: It is recommended that management of complex benign upper gastrointestinal pathology is discussed at multi disciplinary team (MDT) meetings. American College of Gastroenterology (ACG) guidelines further recommend that treatment delivery is provided by high volume centres, with objective post-procedural investigations, in order to improve patient outcomes. We aimed to survey the current UK practice in the management of achalasia.

Methods: 443 Upper gastrointestinal (UGI) specialist surgeons throughout the UK were sent a surveymonkey.com questionnaire about the management of achalasia.

Results: 100 responses were received. The majority of patients with achalasia are referred directly to surgeons (80%) and only 15% of units have a MDT meeting for discussing such patients. Diagnosis was mainly with oesophagogastroduodenoscopy (OGD) and contrast swallow, and only 61% of units have access to high resolution manometry (HRM). 89% of younger patients were offered surgery initially, whilst in the elderly surgery was offered as first line treatment in 55%. Partial fundoplication was carried out by 91% of responders as part of the operation, and 58% responders carry out an intraoperative OGD. The average number of operations carried out per annum is 4 per responder. Most responders (66%) did not perform routine post-intervention investigations and follow-up varied from none to lifelong.

Conclusion: Diagnosis and management of achalasia within the UK is relatively standardised, although there remains limited access to HRM. Discussion at benign MDTs however is poor and follow-up differs widely. UK guidelines may help to make these more uniform.

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1. Introduction

Achalasia is a rare oesophageal dysmotility disorder characterized by absent peristalsis of the oesophagus coupled with failure of relaxation of the lower oesophageal sphincter (LOS) [1]. The pathological changes consist of inflammation affecting the myenteric plexus with subsequent destruction and loss of ganglion cells and nerves [2]. The aetiology is thought to be autoimmune mediated, with links to certain viral infections in genetically susceptible individual [3]. The condition is rare with an incidence of 1:100,000 in the UK [1].

Dysphagia to solids and liquids is the most common presenting complaint. Other symptoms include odynophagia, regurgitation of undigested food, halitosis, chest pain and weight loss. Intra-oesophageal reflux due to food content stasis within the oesophagus can occur resulting in food bolus fermentation and consequent oesophagitis, leading to symptoms often being mistaken for gastroesophageal reflux disease [4].

The differential diagnosis includes other benign oesophageal dysmotility disorders as well as gastro-oesophageal reflux disease [5] and pseudo-achalasia.

Characteristic appearances are seen at contrast fluoroscopy and oesophagogastroduodenoscopy (OGD), and the latter investigation is mandatory to exclude alternative pathology. The gold standard investigation for diagnosis is oesophageal manometry [4].

Treatment is tailored to the patient and ranges from medical therapy to surgery. Because neuronal loss is irreversible, treatment is limited to disruption of the LOS - recurrence is a recognized problem, as is the development of mega-oesophagus in late stages [6].

Achalasia is a lifelong, debilitating condition, which significantly affects the quality of life of patients. Its low incidence renders it a condition that often requires a subspecialist interest and that is often challenging to diagnose. American College of Gastroenterology (ACG) guidelines recommend delivery of treatment in high volume centres, and multi disciplinary team (MDT) management is recommended worldwide to improve patient outcome [7]. Objective post-procedural investigations are also recommended to help identify and target recurrence early [8].

There are currently no UK based guidelines on the management of achalasia. The National Institute for Health and Care Excellence (NICE) recommends reference to Society of American Gastrointestinal and Endoscopic Surgeons (SAGES) guidelines [9].

Our aim is to characterise the current UK practice within the Upper Gastrointestinal specialist surgical community in the management of Achalasia via a web-based questionnaire, paying particular attention to the prevalence of multidisciplinary team based practice, post-procedural objective assessment and follow-up.

2. Materials and methods

The opinion of UK Upper Gastrointestinal (UGI) specialist surgeons was sought through the mailing list of the Association of Upper Gastro-Intestinal Surgeons (AUGIS) on the management of achalasia through a 21-question survey created on surveymonkey.com by the authors and reviewed independently by three Consultant UGI surgeons. AUGIS is the national UGI association in Great Britain and Ireland to which all UK UGI Specialist Surgeons are expected to hold a membership. The survey questions addressed topics spanning unit setup to management of recurrent disease and new treatment methods. The survey was left open for a period of 6 months in order to maximise response rates.

The questionnaire can be found in the [Appendix](#).

3. Results

Responses from 100 out of 443 (23%) specialist UGI surgeons on the AUGIS mailing list were collected over a period of 6 months, however all responses were received within a period of 30 days from November to December 2014.

3.1. Unit set up

Achalasia is referred to an UGI Surgical Specialist in 80% of units. Physicians are the primary point of contact in the remaining 20%. Access to a MDT forum to discuss management of achalasia is available in 15% of units.

3.2. Diagnosis

To diagnose achalasia and exclude alternative pathology, 99% perform OGD, 89% also use contrast swallow and 61% use HRM. Botulinum toxin is used by 12% of responders to aid diagnosis in challenging cases.

3.3. Management

For patients under the age of 60, 89% of responders (71 out of 80) would offer surgery as first line treatment, 10% (n = 8) would opt for Pneumatic Dilatation (PD) first whilst 1% (n = 1) would opt for Botulinum toxin A injections into the LOS.

For patients over the age of 60, 55% of responders (43 out of 78) reported they would offer surgery as first line treatment, 37% (n = 29) would offer PD and 7.7% (n = 6) would offer Botulinum toxin A injections.

3.4. HRM subtypes

When asked to specify if HRM types would result in a change in first line management, 11 of 32 (34%) responders favoured conservative management over surgery for Type 3 achalasia.

3.5. Pneumatic Dilatation

PD is offered by 55 of 74 respondents (74%), with 27 (49%) offering it for patients who are unfit for surgery, 11 for recurrent disease (20%), 13 for patient choice (24%) and 3 first line treatment (5%). Fluoroscopic control is used routinely by 42 of the 55 respondents (76%) who offer PD, with a starting minimum balloon diameter ranging from 10 to 30 mm (mean 25 mm, median 25 mm) and a starting maximum balloon diameter ranging from 15 to 40 mm (mean 30 mm, median 30 mm). Of those who offer PD without fluoroscopic guidance, the minimum starting balloon diameter ranges from 12 to 30 mm (mean 24 mm, median 25 mm) and the maximum starting balloon diameter ranges from 15 to 40 mm (mean 32 mm, median 30 mm).

3.6. Surgery

Of 100 responders, 66 answered the questions on specific surgical options for achalasia. Of these, 97% ($n = 64$) would offer a Laparoscopic Heller Myotomy (LHM), 91% ($n = 58$) routinely perform a wrap (85% anterior wrap, 3% Nissen and 12% Toupet) and 58% ($n = 37$) will do a check intra-operative OGD. The average length of myotomy is 3 cm on the stomach and 6 cm on the oesophagus. On average, responders performed 4 myotomies a year each between 2011 and 2013 (range 1–15).

Of 61 responders, 24 (39%) would consider Per Oral Endoscopic Myotomy (POEM).

3.7. Objective post-operative assessment

There were 64 responders to the questions on post-operative assessment. A contrast swallow is performed by 33% ($n = 21$), while 16% ($n = 10$) perform manometry (including HRM), 19% ($n = 12$) perform an OGD and 66% ($n = 42$) do not offer objective post-procedure assessments. Of the 64, 90% ($n = 57$) do not use a scoring system such as the Eckardt score and only 33% ($n = 21$) offer long-term OGD surveillance.

The question pertaining to management of recurrent disease was answered by 62 responders, of these, 53% ($n = 33$) recommend PD, 39% ($n = 24$) would re-operate and 7% ($n = 4$) would use botulinum toxin.

Follow-up varies from 3 month to lifelong. 23 of 63 responders (36.5%) recommend lifelong follow up.

4. Discussion

Despite widespread recognition that decision making in the management of achalasia can be challenging, only 15% of responders have access to a MDT forum to discuss the management of achalasia. There is a paucity of evidence in the literature to support the implementation of a formal timetabled MDT for benign oesophago-gastric disease, and it would clearly be challenging logistically for many units to do so. Nevertheless, patients with achalasia clearly need a multidisciplinary approach to their management, and we would suggest that this is best done in a formal MDT, with a suggested core team of UGI surgeons, gastroenterologists and radiologists.

Only 61% use HRM despite good evidence that results from treatment vary considerably with achalasia subtype – of these, 34%

would change their first line treatment strategy to conservative if faced with a diagnosis of HRM type III achalasia. Achalasia can be subdivided into 3 subtypes categorized by HRM results: in Type I (classic achalasia) there is absence of peristalsis, in Type II (pan-oesophageal pressurization) there are some simultaneous contractions with amplitudes <40 mmHg and in type III (spastic or vigorous achalasia) there are simultaneous contractions >40 mmHg. Numerous publications report different prognoses for the 3 achalasia types. Type II has the best prognosis following serial dilatations or myotomy, whereas type I has a slightly worse prognosis and type III has the worst prognosis [10–12].

The original oesophagocardiomyotomy was first described by Ernst Heller in a publication in 1914 [8]. With the advent of minimally invasive surgery, the LHM has become the favoured first line treatment for achalasia, recommended by SAGES guidelines [9]. Our results show that LHM is the mainstay of treatment for achalasia in the United Kingdom, with 89% of responders saying that it is first line treatment in their unit. This number falls to 55% in patients over the age of 60. This is likely due to both patient and surgeon choice, as well as a reflection on the presence of comorbidities in this population age group, although a study in 2010 suggests LHM can achieve good results in patients over the age of 60 [13].

The myotomy involves a longitudinal incision starting over the LOS to disrupt both longitudinal and circular muscle fibres of the oesophagus, extending proximally 5–7 cm and distally 2–3 cm to include the gastric sling fibres [8]. Wang et al. showed an association between larger volume of surgery and improved perioperative outcome [14] due to a steep learning curve for LHM [4,7,8]. Long-term efficacy of LHM is reported to range between 88% and 95% [6,15–19]. PD, when applied in a graded fashion, has been shown to be non-inferior in efficacy to LHM in the short term [15,16]. A recent meta-analysis suggests that long-term results however are in favour of surgery [17].

A partial fundoplication following the myotomy is carried out by 91% of responders despite the lack of good evidence supporting the routine use of a fundoplication.

Initially described in 4 patients in 2008, POEM involves an endoscopic, mucosotomy, and myotomy via a submucosal tunnel. Current evidence suggests POEM boasts equivalent short term efficacy and safety to LHM, but long term efficacy remains unclear [18]. Of 100 responders, 39 would consider POEM.

We found significant variation in use and type of post-procedural investigations to assess the results of treatment. Eckardt et al. used contrast swallows to assess treatment impact and recommend the use of objective investigations such as contrast swallow and manometry or questionnaires post-procedurally [8]. Vaezi et al. have shown that patients' symptoms or physician impression of treatment success may not be a reliable predictor of outcome as symptom resolution may occur without a significant improvement in oesophageal emptying [20].

Manometry is routinely used to monitor treatment success in patients with achalasia at many European centres. In a prospective study investigating the long term outcome of patients with achalasia treated with PD, a post-interventional LOS pressure of <10 mmHg was shown to predict a favourable long-term treatment response [21].

Gockel et al. compared the value of different severity scoring systems as an adjunct in managing achalasia. The Eckardt score was found to be the most useful for clinical practice [22]. Severity scoring systems are not currently included within the SAGES recommendations for post-procedural assessment [9]. In the current survey, 6 responders use the Eckardt score and 22 use manometry and/or imaging to assess treatment success.

Recurrence can occur years after initial treatment, presenting with its own set of challenges: diagnostic dilemmas, the

development of mega-oesophagus, the difficulty of revisional surgery, and fibrotic stricturing secondary to previous treatment modalities [23]. Up to 20% of patients develop symptoms that might need further treatment within 5 years [24–26]. Progression to mega-oesophagus or end-stage disease is seen in a further 6–20% [27]. Subspecialist input is likely to be required long term. Vaezi et al. recommend that treatment of achalasia should be centralised in high volume centres [4], and SAGES guidelines recommend reoperation by experienced surgeons over PD in the treatment of recurrent disease following surgery. 62 responders answered the question on management of recurrent disease; 53% recommend PD, and 39% would carry out revisional surgery.

There are currently no UK guidelines for the medical and surgical management of achalasia. A section of the 2004 British Society of Gastroenterology (BSG) guidelines on oesophageal dilatation is dedicated to PD in achalasia. NICE guidelines currently recommend reference to the SAGES guidelines of 2009. American guidelines, and the current literature, recommend the use of an MDT to discuss management, post-procedural follow up with objective measures of treatment success, and surgery as first line therapy. SAGES guidelines do not recommend PD as salvage therapy after myotomy.

The current survey demonstrates the need for the establishment of national guidelines to address the UK-specific issues in the management of achalasia in the setting of the current National Health Service.

Based on our survey, the current review of the literature and American guidelines, we propose the following recommendations:

In line with the current AUGIS Provision of Services document [28], we recommend that primary LHM are performed in UGI units with at least 2 consultant surgeons with an interest in such operations, with the expectation that each surgeon will perform at least 5 cases per year.

All achalasia cases should be discussed in the setting of a benign UGI MDT. The decision on the core team composition should be left for each unit to address, but we suggest that the MDT should include an UGI radiologist, UGI surgeon and luminal gastroenterologist.

All patients with possible achalasia should be investigated with high resolution manometry.

Patients with recurrent achalasia should be referred to a high volume tertiary unit.

Surgery as first line therapy should not be restricted to those less than 60 years, and should be considered for all medically fit patients.

All patients should have some form of post-procedural objective assessment of success by repeat manometry or contrast radiology.

All units should use a scoring system such as the Eckardt score.

Ethical approval

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Author contribution

All authors have contributed equally to all aspects of this study (design, data collection, analysis and writing).

Conflicts of interest

None.

Guarantor

Jihene El Kafsi (corresponding author).

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There are no conflict of interests.

Appendix A. Supplementary data

Supplementary data related to this article can be found at <http://dx.doi.org/10.1016/j.amsu.2016.10.009>.

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