Inclusion body myositis and dysphagia. Presentation, intervention and outcome at a swallowing clinic

Assessment and Management of Dysphagia in IBM

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Abstract

Purpose: This study reviews a patients with inclusion body myositis (IBM) referred for assessment of dysphagia at a tertiary swallow clinic. It describes symptoms at presentation, imaging, and management strategies.

Methods: A retrospective review of electronic patient records was performed between 2016-2020.

Results: Twenty-four patients were included, with a mean age of 72. Baseline modified Sydney swallow questionnaires (m-SSQ) identified problems with hard/dry food, food sticking, and repeated swallowing. Twenty-two patients had an RSI score that could indicate significant reflux. Video swallow identified specific problems, including with tongue base retraction (96%) and residual pharyngeal pooling (92%). Seven patients (30%) had features of aspiration on imaging despite a median PAS score of 2. Four patients received balloon dilatation, and two patients underwent cricopharyngeal myotomy.

Conclusion: This study helps to profile features of dysphagia in patients with IBM. More evidence is needed to determine the most effective management pathway for these patients.

Key Words

Dysphagia

Inclusion Body Myositis

Deglutition Disorders

Neuromuscular Diseases

Introduction

Inclusion body myositis is a sporadic, progressive, inflammatory myopathy characterised by asymmetric involvement of the quadriceps and finger flexors. Its incidence varies from 1.2 to 3.2/million/year, and its prevalence is around 3-7/100,000 [1]. It is the most common myopathy after age 50 [2]. Unlike other inflammatory myopathies, and despite inflammatory histopathological findings, it is refractory to glucocorticoid treatment. Over time, it progresses to disability, which may contribute to increased mortality [3 4].

Dysphagia is reported as being a significant feature of inclusion body myositis (IBM), often being present at diagnosis [5]. Estimates of dysphagia as a symptom range from 40-80% of patients [6-8], however this may be an underestimate of the problem [8-10].

The severity of dysphagia in IBM can vary from mild to severe and is generally progressive over time. Even 'mild' problems can have an impact on quality of life due to the limitations associated with social interaction, particularly around meals. There are also potential physical, social, and psychological consequences [11]. As the dysphagia becomes more severe, it can result in a failure to maintain adequate nutrition, contributing to cachexia, and can predispose to aspiration pneumonia. These factors are considered to contribute to the mortality in patients with IBM [3, 12].

The diagnosis of dysphagia in this group and its subsequent assessment and treatment is, therefore, important for the quality of life for patients with IBM.

The aim of this review is to detail the presentation, assessment, and treatment of patients with IBM managed in the Oxford Dysphagia clinic and a review of the findings with reference to the published literature.

Materials and Methods

The aims, methods and results of this study were registered with the Ulysses Clinical Governance System (reference number 6624) and approved by Oxford University Hospitals Foundation Trust.

The medical records of all patients with IBM attending the Oxford Dysphagia clinic between 2016-2020 were interrogated retrospectively. All patients were referred from the Oxford centre for neurology. The centre has a specialist interest in IBM. All patients were referred with an established or suspected diagnosis of IBM as well as swallowing difficulties and desire to attend specialist services.

All patients attending the clinic routinely completed the self-reported modified Sydney swallow questionnaire and the Reflux Symptom Index questionnaire (see appendix).

All patients were jointly assessed by an ENT Surgeon (SW) and a speech and language therapist. Assessment included a comprehensive history and examination along with a range of clinical assessments including fiberoptic endoscopic evaluation of swallowing (FEES), with a range of textures and consistencies available.

Following clinical assessment, all patients received verbal and written advice regarding their swallowing. Additional assessments included video fluoroscopy (VFS) or, on occasions, barium swallow. All imaging was assessed by two independent speech and language therapists and scored, where possible, using imaging features mentioned previously in the literature [13].

All patients were treated with an advice-based approach initially and offered a follow-up to assess their progress. Where appropriate, community-based speech and swallowing therapy

was initiated with advice regarding the specifics of IBM to support the community-based therapist.

Subsequent treatment was formulated through a discussion between the patient, speech and language therapist and surgeon, utilising information from the self-reported questionnaires and investigations. This could involve further therapy and exercises, including advice about strategies to optimise safe swallow, pleasure whilst eating, and adequate nutrition status. Treatment also could comprise balloon dilatation of the upper oesophageal sphincter, percutaneous feeding tube insertion, cricopharyngeal myotomy, or botulinum toxin injection.

Where visualised data followed a normal distribution, mean and standard deviation were calculated, otherwise median and range were used. Correlation between questionnaires (ordinal data) was measured using Spearman's rank correlation coefficient.

Results

Between 2016 and 2020, twenty-six patients with IBM were referred to the clinic; one was later found to be incorrectly diagnosed, and there was no available data for one; both were removed from analysis. Of the twenty-four patients included in this study, there were 13 males and 11 female patients. The mean age at first visit to the dysphagia clinic was 72 years (range 54 – 84 years). For four patients (17%), the diagnosis of IBM was made whilst investigating their dysphagia symptoms. This included one patient who was investigated for dysphagia for eight years before eventually receiving a diagnosis of IBM and being referred to the clinic.

At their first visit to the clinic, seventeen patients (71%) completed baseline modified SSQ and RSI questionnaires, whilst the remaining six completed these soon after.

The mean and standard deviation of the total scores of the modified SSQ are 79 and 31, range 3-130 (maximum possible score is 180). The profile of responses to the modified SSQ demonstrated a broad range (figure 1). The symptoms that scored highest were problems with hard foods, food sticking, choking and repeated swallow. The symptoms that were least reported were odynophagia and nasal regurgitation.

The results from the Reflux Symptom Index (RSI) of all patients on their first presentation to the clinic demonstrated a mean score of 21 (range 10 – 36, standard deviation 7). Twenty-two of the 23 patients (96%) for which we have results had a score above 13, suggesting that the reflux-related symptom burden appears to be quite high in this cohort. All patients received written self-management advice, along with reflux medication where appropriate.

All patients underwent a contrast swallow assessment: seventeen underwent video fluoroscopy and seven underwent barium swallow for their first assessment. Barium swallows were initially performed at the inception of the clinic; they have been superseded by video fluoroscopy due to the higher image resolution and detail. All video swallows were performed with a speech and language therapist in attendance.

The baseline imaging features of the cohort upon presentation to the clinic are displayed in table I. The most prevalent features were impairment of tongue base retraction, residual pharyngeal pooling, and pharyngeal constrictor impairment. Cricopharyngeal hypertrophy was found in eighteen patients (75%). Seven patients (30%) had objective features of aspiration, with a median Penetration Aspiration Score for the cohort of 2, range 1 – 8 (figure 2).

To see whether certain scales from the modified SSQ could ‘predict’ Penetration-Aspiration Scale (PAS, [14]) scores calculated on imaging, correlation coefficients were calculated. All questions had coefficients between -0.5 and +0.5 (data not presented).

All patients received targeted speech and swallowing advice and were offered a review appointment, either remotely or face-to-face. Sixteen patients (67%) returned to clinic for review. All patients had the option of being followed up by the community speech and language teams.

Sixteen patients (67%) in the cohort received swallowing advice but no surgical intervention. We followed them up over time. Three patients (13%) completed repeat modified-SSQs after a few months, after being given the swallowing advice; their serial scores were largely quite similar, despite reporting their dysphagia to be improved overall.
Four patients (17%) in this group had serial video swallows over the course of several years, which allows us to see the natural progression of dysphagia in IBM. These limited data show increasing PAS scores and impairment over time (data not shown).

Six patients (25%) received active surgical intervention. Three received more than one different procedure: oesophageal dilatation (performed in four patients), radiologically inserted percutaneous gastrostomy (RIG) (one patient), cricopharyngeal myotomy (two patients) and Botox injection to the cricopharyngeus muscle (one patient).

The effect of the different procedures was measured by serial modified SSQs. Given the low numbers, data has not been presented nor statistically analysed, but briefly described here. Balloon dilatation was offered to patients with cricopharyngeal hypertrophy on contrast swallows and symptomatic obstruction. Of the four patients in this group, only one derived lasting benefit from a single dilatation. The other three either derived no benefit or experienced a recurrence in their symptoms: one went on to have two repeat dilatations (the last with botulinum toxin injection to the cricopharyngeus muscle) without benefit; one patient improved after gastrostomy (RIG) insertion; and one patient only benefited from a cricopharyngeal myotomy. Cricopharyngeal myotomy was offered to two patients based on

imaging features and severity of dysphagia; both reported subjective improvement in their swallow (supplementary figure 1).

Discussion

Our patient cohort were referred to the clinic due to concerns over dysphagia. They therefore represent a subset of IBM patients encountered in clinical practice. Subtle features of dysphagia in IBM may be present without spontaneous reporting: in a study by Cox [8], 37 of 57 patients had symptoms of dysphagia picked up by a questionnaire, but only 17 spontaneously reported dysphagia during clinical assessment.

The patients in our cohort were almost evenly split between the sexes: 13 males and 11 females. This is an interesting finding given that other studies have found the majority of patients with IBM are male [13, 15]. The mean age at presentation is comparable to other studies [8, 13]. One patient in our study died due to aspiration pneumonia, though our patients were only followed up over the four-year study duration.

Four patients (17%) had dysphagia as their presenting symptom of IBM – this is unusual since dysphagia is thought to usually present later in the disease [8, 13, 16, 17]. One patient in our cohort was investigated for dysphagia for eight years before their diagnosis of IBM, similar to a previous case report [18].

Twenty-three patients presenting to our clinic completed a Reflux Symptom Index. The median score was 21. Normative data suggests that an RSI greater than 13 may be indicative of significant reflux. Twenty-two (96%) had scores over 13, suggesting this could be a common component of the symptom burden. When reflux was identified, this was addressed with discussion, written advice, and provision of reflux medication. However, the RSI has not been correlated with reflux in an IBM population. It is therefore possible that the high score reflects the underlying dysphagia associated with IBM. Additional studies would be needed to evaluate the correlation of the RSI in this population.

Results from the cohort's modified-SSQs suggests particular difficulties with hard and dry food, food sticking, and repeated swallowing. These results resemble those found previously in the literature [7, 8, 13], suggesting these are common features of the dysphagia in IBM. These questions could provide a good screening for dysphagia in these patients.

All patients underwent contrast swallow assessment (table I). The most common features identified were impairment in tongue base retraction (96%), pharyngeal constriction (82%), and residual pharyngeal pooling (92%). Cricopharyngeal dysfunction and hypertrophy were identified in 75% of patients, a figure comparable to previous studies [13]. Aspiration was identified in 30% of patients at baseline. Median penetration aspiration score was 2, similar to a previous study [9], though the distribution of scores was uneven (figure 2). However, it should be noted that we did not have a standardised protocol to ensure inter-rater reliability, nor any software to measure dynamics of upper oesophageal sphincter opening. Moreover, barium swallows have a lower frame per second rate and quality compared to videofluoroscopy. Therefore, we recommend that future studies use videofluoroscopy to better define swallowing abnormalities, and that there is a validated protocol and analysis tool (e.g. MBSiMP). Given the reported difficulties with solids, incorporating both fluid and solid boluses within the VFS protocol could be informative for this population.

All patients received support by our speech and language team. This support depended on several factors: the swallow mechanism, risks associated with oral intake, imaging results, patient goals, and effect on life. The amount and timing of therapy was tailored to individual need.

Six of our patients (25%) underwent surgical procedures – a lower proportion than in previous studies [13]. Three of the four who underwent balloon dilatation received no lasting

benefit and required further procedures. These findings are similar to Oh (2008) who found that the majority of dilatations resulted in no benefit [13].

Only one patient received Botulinum toxin injection to alleviate their dysphagia; symptoms improved for just one month before recurrence. By contrast, some studies have shown longer lasting benefits [10, 19] – whereas others have suggested limited efficacy [13].

Cricopharyngeal myotomy was only offered to three patients in our cohort and was performed in two patients; it was effective at improving dysphagia in both without documented complications. Cricopharyngeal myotomy was reported to be an effective procedure in several previous studies when used appropriately, using either an endoscopic or transcervical approach [5, 6, 13, 20, 21].

There are several limitations to this study. There is a small sample size, but IBM is a rare condition. The study is retrospective and there was limited follow-up for a few patients who were most recently referred. However, despite these limitations, we recommend that all patients with IBM and dysphagia have serial modified SSQs and videofluoroscopy in order to fully assess their dysphagia and to quantify the effect of the different interventions. Moreover, the high reporting of reflux symptoms suggests that actively screening and treating reflux at an early stage should be considered. Future research should include the use of validated swallowing scales such as the Modified Swallow Impairment Profile (MBSImP [22]) to allow for standardised analysis of swallow features, alongside the use of additional outcome measures such as the Penetration Aspiration Scale (PAS [23]) and the Dynamic Imaging Grade of Swallowing Toxicity (DIGEST [24]) to detail residue, penetration and aspiration.

Conclusion

This study helps to profile features of dysphagia in patients with IBM, both subjective (via questionnaires) and objective (contrast imaging). We describe progression of dysphagia in IBM in a cohort of our patients and describe our practice. We outline suggestions on how to improve the quality of research in this area. More work is needed on these patients in order to better evaluate common symptoms, imaging features, and management strategies, and perform relevant statistical analysis.

Declarations

Funding: The authors declare no sources of funding for this work There was no financial nor material support. The work analysed existing practice.

Conflicts of interest: The authors declare no conflicts of interest for this work.

Availability of data and material: all data are available on request.

Ethics approval: The aims, methods and results of this study were registered with the Ulysses Clinical Governance System (reference number 6624) and approved by Oxford University Hospitals Foundation Trust. There is consent for publication. Retrospective, anonymised information was used. As this study audited existing service provision, additional consent to participate was not required.

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Summary

- This study describes a UK cohort of patients with inclusion body myositis, presenting with dysphagia to a tertiary centre.
- The reflux symptom index suggests an issue with significant reflux in these patients, something not previously realised.
- Swallow questionnaires and imaging revealed common problems with swallowing amongst these patients.
- Whilst the majority of these patients can be managed by a community speech and language team, they should be assessed by a specialised service for consideration of operative intervention.

Figures and Tables

All figures can be provided as separate files for higher quality.

Figure 1

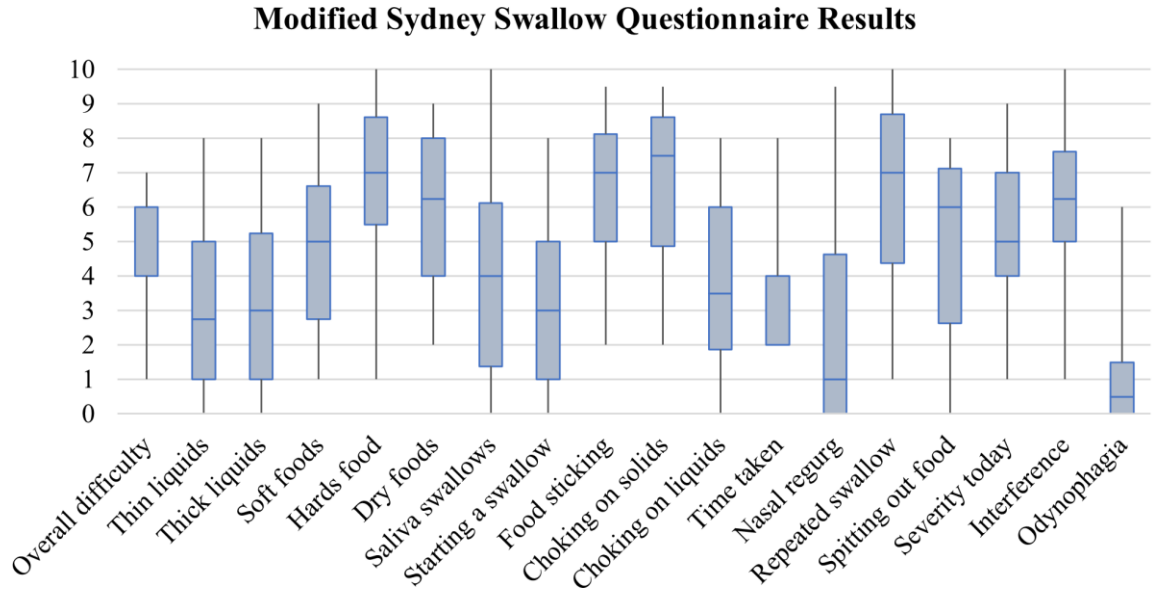


Fig 1 shows results of modified Sydney Swallow Questionnaires filled out in the initial presentation to the dysphagia clinic. The questions are displayed on the x axis, with severity scored out of 10.

Figure 2

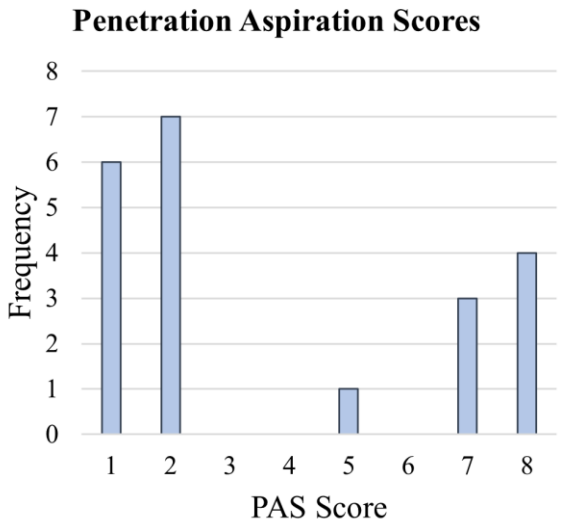


Fig 2 displays penetration aspiration scores for the cohort. The worst score across all consistencies trialled was recorded. Median score was 2, range 1-8.

Figure 3

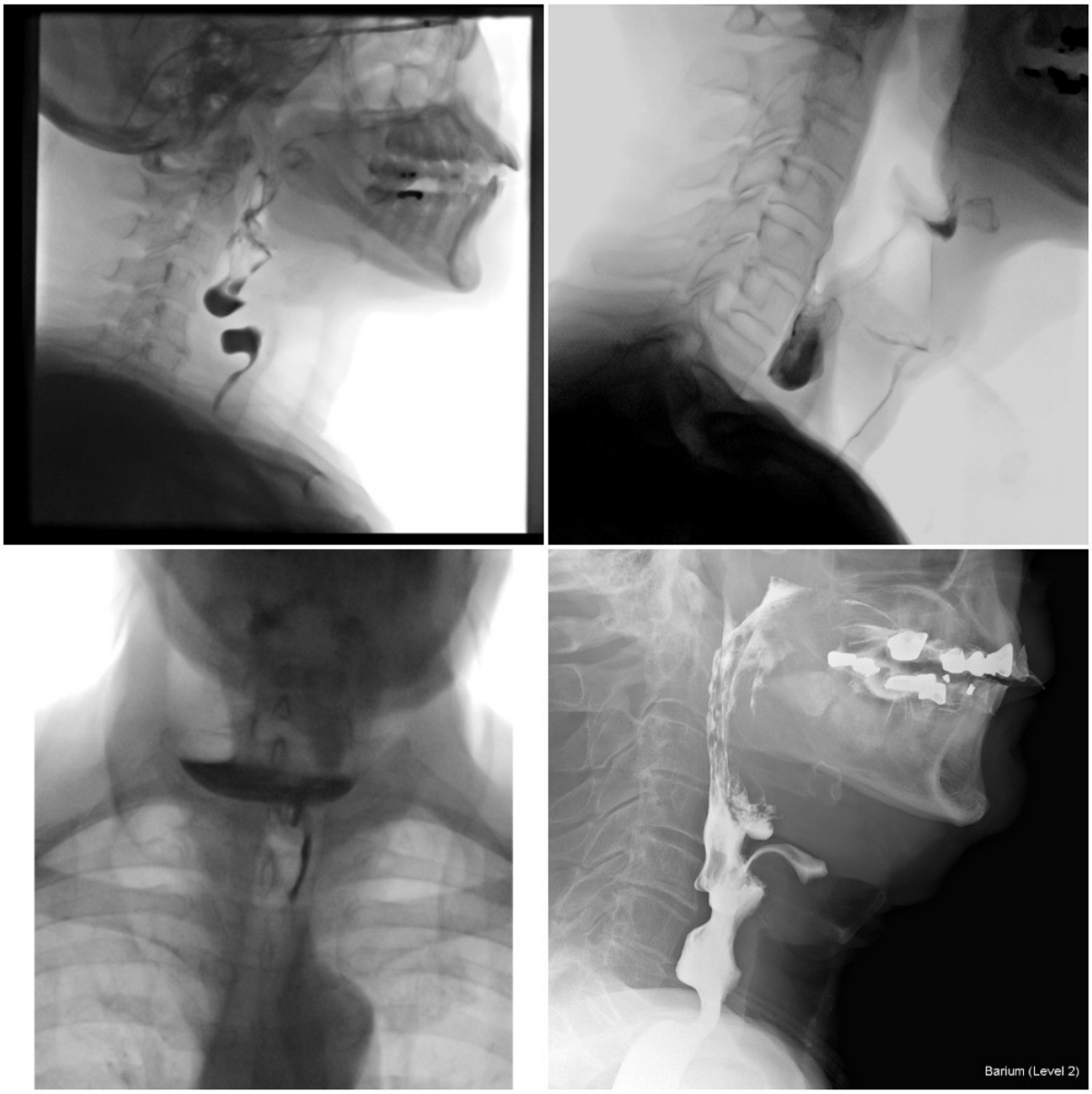
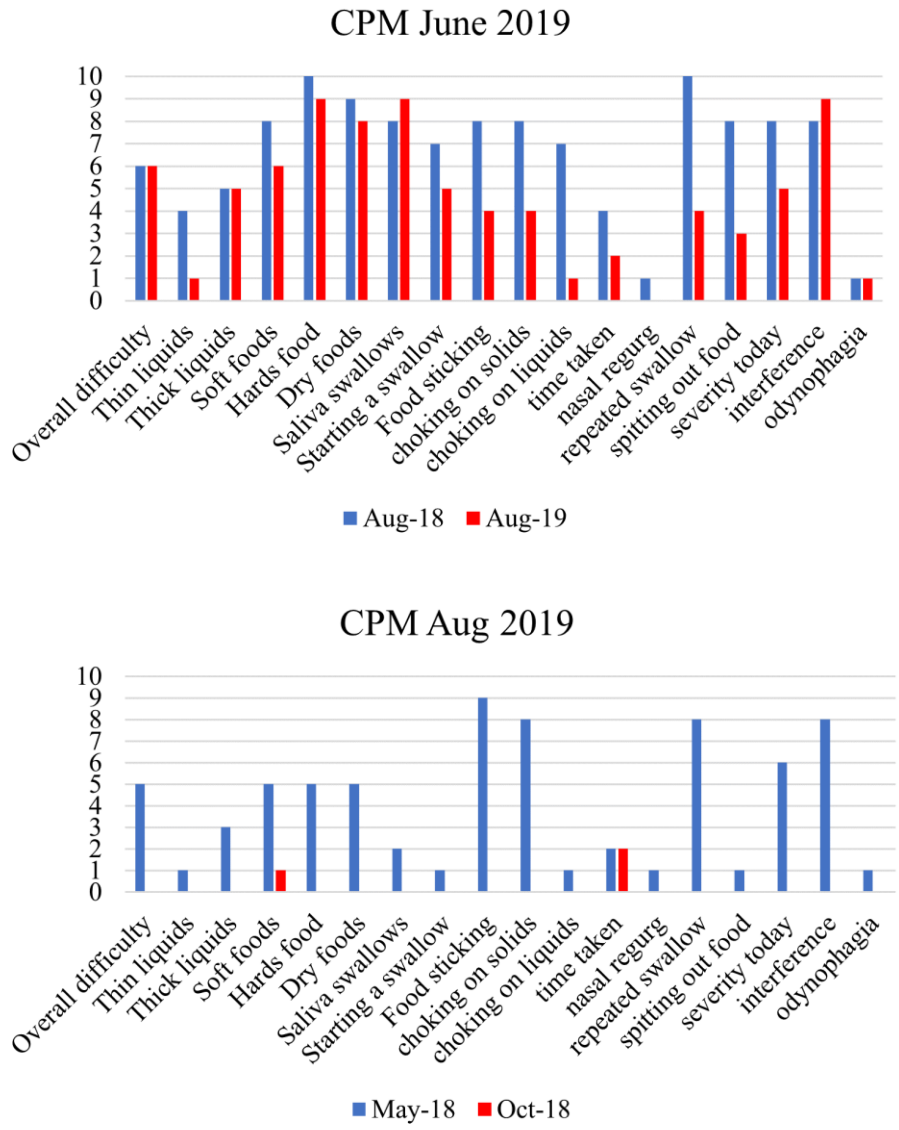


Fig 3 compiles images of VFS showing common features identified. Top left to bottom right: a) cricopharyngeal hypertrophy evident at the height of swallow (maximal displacement of the hyoid), b) silent aspiration of post-swallow pharyngeal residue, c) spontaneous, passive opening of the CP to allow eventual bolus transit into the oesophagus, d) cricopharyngeal hypertrophy and laryngeal penetration evidence at the height of swallow (maximal displacement of the hyoid).

Supplementary Figure 1



Supplementary Figure 1: pre-procedure and post-procedure modified Sydney Swallow Questionnaires scores (questions on x-axis, scores on the y-axis) presented for two patients who underwent cricopharyngeal myotomy (CPM). Both show an improvement (reduction in m-SSQ scores).

Table I

Imaging feature	Patients	%
Tongue control impairment	2 / 21	10
Bolus control impairment	2 / 20	10
Tongue base retraction impairment	22 / 23	96
Laryngeal elevation impairment	11/ 22	50
Pharyngeal constrictor impairment	18 / 22	82
Residual pharyngeal pooling	22 / 24	92
Cricopharyngeal dysfunction	18 / 24	75
Cricopharyngeal hypertrophy	18 / 24	75
Penetration	14 / 23	61
Aspiration	7 / 23	30

Table I displays features found on baseline video fluoroscopy or barium swallow on initial presentation to the clinic. Not all features could be scored from the imaging due to the frame rate of barium swallow, hence there are variable denominators. All features were scored as binary presence or absence and were scored on fluid boluses only.