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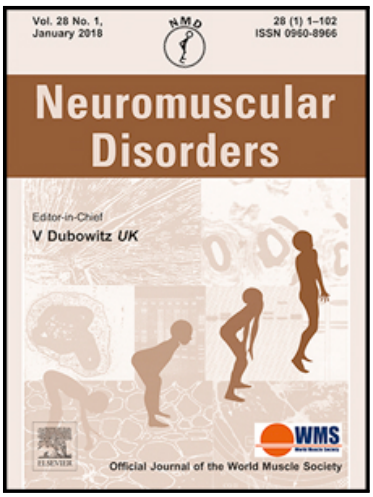

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Highlights

- First reported study investigating the use of a device to aid swallowing in IBM
- EMST is not a restorative treatment for severe dysphagia in patients with IBM
- EMST may have some benefit to patients with a shorter duration of disease

Exploring the efficacy of the Expiratory Muscle Strength Trainer to improve swallowing in Inclusion Body Myositis: a pilot study

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Abstract

Inclusion Body Myositis (IBM) is the most common acquired myopathy in older individuals with more than two thirds of patients experiencing impaired swallowing. There are currently no standardized exercise therapies to improve or sustain swallowing despite good evidence for exercise therapy in limb muscles. Reduced upper esophageal sphincter (UES) opening is a common abnormality associated with dysphagia in IBM. This pilot study recruited IBM patients with abnormal UES function and dysphagia into an exercise program. It was hypothesised that regular practice using the Expiratory Muscle Strength Trainer (EMST) device would improve hyolaryngeal movement by strengthening suprahyoid musculature and facilitate opening of the UES thereby improving swallowing and quality of life. Overall, IBM patients who used the EMST device demonstrated no improvement in swallowing function. Consistent with that result, there was also no change in measures of quality of life. However, further studies are needed to elucidate whether it has a preventative role in the development or progression of dysphagia in IBM as there is a suggestion that patients with a shorter duration of disease may have had some benefit. This research provides pilot data and recommendations that will guide future studies on exercise therapy and swallowing in this area.

Keywords: Dysphagia, Exercise therapy, Suprahyoid muscles, Pharyngeal, Upper esophageal sphincter, Inclusion Body Myositis

1.1 Introduction

Inclusion Body Myositis (IBM) is the most common acquired myopathy in patients over 50 years of age [1]. Dysphagia, defined as difficulty or discomfort in swallowing, is a common symptom of the disease and occurs in 40-80% of IBM patients [2-4]. Swallowing is an essential biological function and a major contributor to quality of life; dysphagia is linked to life-threatening medical complications such as dehydration, malnutrition and recurrent aspiration pneumonia [5]. Any intervention that improves swallowing function therefore has the capacity to significantly reduce morbidity and mortality in IBM patients and improve quality of life [6]. Currently only invasive interventions such as balloon dilation [7], cricopharyngeal myotomy [8] or injection of botulinum toxin to the Upper Esophageal Sphincter (UES) [9-12] are known to improve swallowing function in this population. There are no standardised exercise devices to improve or maintain swallowing function in IBM. Langdon et al found that dysphagia in IBM was predominantly consistent with pharyngeal and suprahyoid muscle weakness rather than failure of UES relaxation [13]. There has been a study investigating the effectiveness of the Mendelsohn Manoeuvre which improved swallowing in some IBM patients [14, 15]. This manoeuvre utilises extended hold to maintain an elevated larynx and in turn contribute to patency of UES during swallowing. Similar to the Expiratory Muscle Strength Trainer (EMST), the Mendelsohn Manoeuvre targets strengthening of the suprahyoid muscles used for hyolaryngeal movement [16]. The EMST device is however easier for patients to use. Several studies investigating the use of exercise programmes in IBM, particularly in relation to exercise of limb muscles are shown to be safe, feasible and significantly improve function in patients [17-19].

The EMST is a therapeutic goods administration (TGA)-approved device currently used in patients suffering other neurological conditions. The use of the EMST device for dysphagia in Parkinson's Disease has been supported with class 1 evidence and a randomized control trial showed significant improvement in the swallowing of Parkinson's patients [20, 21]. Troche et al [21] suggest that their device works to improve swallowing function through strengthening of the hyolaryngeal movement, in turn improving the opening of the UES [21]. The device was also shown to be effective in improving swallowing function in neuromuscular conditions such as Motor Neuron Disease [22] and Pompe disease [23]. There have been no previous studies investigating this device in IBM.

In keeping with the Troche et al [21] and Langdon et al [13] studies, it was hypothesised that an exercise programme using the EMST device would strengthen the suprahyoid muscle group and help open the UES to facilitate and improve swallowing function and quality of life in IBM patients. The EMST device is a hand held and inexpensive home-based therapy that if found to be effective could greatly improve outcomes for these patients. It was the combination of these studies that led to our hypothesis that the EMST device would improve hyolaryngeal movement in IBM.

1.2 Aims

The aims of this pilot study were to firstly evaluate the feasibility of utilising the EMST device to drive a directed exercise- based intervention in this patient population. Secondly, we aimed to determine whether a 12-week exercise programme focussed on suprahyoid swallowing muscles using the EMST device improves objective measures of swallowing function, and quality of life for patients.

1.3 Methods

1.3.1 Participants

This pilot study was approved by the Royal Perth Hospital Human Research Ethics Committee (RGS0000000109) and conducted in accordance with the Helsinki Declaration of 1975. All included participants provided informed written consent. IBM patients living in Western Australia were invited to participate in this trial through specialised clinics at Western Australian Neurology Research Institute (WANRI) and the Institute of Immunology and Infectious Diseases (IIID) at Murdoch University by a consultant neurologist specialising in neuromuscular conditions (MN). All participants completed an abridged Dysphagia Handicap Index (DHI), SF-36 Item health survey (SF-36) [24], they underwent two instrumental swallow assessments videofluoroscopic swallow study (VFSS) and flexible endoscopic evaluation of swallowing (FEES). The Penetration-Aspiration Scale (PAS) and the Yale Pharyngeal Residue Severity Rating scale (YPRSRS) were used as validated rating tools for aspiration and residue. Participants with evidence of UES dysfunction on VFSS were invited to undergo a 12-week exercise programme using the EMST device.

1.3.2 Inclusion Criteria

Participants with a diagnosis of clinically-defined or pathologically-defined IBM as per the European Neuromuscular Centre (ENMC) criteria [25] were invited to participate. To be included in the study, participants were required have dysphagia. Dysphagia was characterised by (but not limited to) subjective reports of difficulty swallowing food. In addition, participants should have been able to complete baseline questionnaires, VFSS and FEES assessments and were required to have evidence

of UES dysfunction on VFSS. Modified Barium Swallow Impairment profile (MBSImp) rating was utilised to measure UES function identifying reduced transfer of the bolus through the sphincter and The Yale Pharyngeal Residue Severity Rating Scale (YPRSRS) was utilised to measure secretion or bolus residue specifically in the piriform sinuses.

Participants also needed to provide informed consent, agree to undergo baseline and follow up assessments and adhere to the 12-week exercise programme using the EMST device.

Participants did not receive other standard speech therapy, swallowing treatment or pharmacological treatment (e.g. steroids, intravenous immunoglobulin) during their time in the study. Further to be included in the study, participants were chosen who had not previously had botulinum toxin treatment for their dysphagia.

1.3.3 Exclusion Criteria

Participants were excluded if the dysphagia was considered due to an alternative cause than IBM (such as an esophageal web), or if there was no evidence of UES dysfunction on VFSS. Other exclusion criteria included: co-morbidities of neurological origin in addition to IBM, uncontrolled asthma, COPD, emphysema or if they were unable consent to the study, or unwilling to undertake a 12-week exercise programme using the EMST device.

1.3.4 EMST Exercise Protocol

The EMST 150 is an expiratory muscle strength training device made of a plexiglass tube with a one-way valve which can be calibrated between 25cmH_20 and 150cmH_20. A mouthpiece was used to allow better fit and more comfortable use.

Participants were instructed to hold the EMST 150 mouthpiece between their teeth, breathe in, tighten their lips around the mouthpiece and exhale forcefully and quickly. Airflow is blocked until the valve receives enough pressure from the exhalation to open and produces a short and sharp whistling sound.

The pressure-threshold load on the EMST device was increased daily throughout the first week of training by the clockwise rotation of the pressure dial. The participants maximum threshold was when the valve failed to release with exhalation. The dial was then rotated one quarter turn counter-clockwise to reduce the load, equating to 75% of the maximum threshold. After each week of training, this process was repeated by the speech pathologists to determine the new threshold load.

Participants were trained by an experienced speech pathologist for 30 minutes a day for 5 consecutive days either face to face or by telephone in the use and titration of the EMST device similar to a prior study [21]. Participants were then given a therapy diary to prompt them to record their daily therapy of 5 sets of 5 breaths through the EMST device each day for 5 days per week. Following the training, participants were seen by the speech pathologist at least once per week for the first 5 weeks to ensure the exercises were performed correctly, and to titrate the pressures on the device. They were then seen fortnightly thereafter either in person or by video teleconference, (Figure 1). After 12 weeks of exercises, the swallowing assessments including surveys Abridged Dysphagia Handicap Index (DHI), The SF-36 Health Survey (SF-36)[24], Videofluoroscopic swallow study (VFSS) and Flexible Endoscopic Evaluation of Swallowing (FEES) were performed.

1.3.5 Videofluoroscopic Swallow Study

All participants underwent oropharyngeal VFSS swallowing examination. Participants sat upright and their swallowing function was recorded in the lateral and AP viewing plane with nine boluses of varying consistency and volume as per the Martin-Harris study [26]. All VFSS assessments were rated by group consensus with experienced speech pathologists. The severity of airway invasion was scored using the 8-point ordinal Penetration Aspiration Scale (PAS), where 1-2 is normal, 3-5 represents penetration, and 6-8 aspiration[27]. Swallowing was specifically assessed through all 17 components of the MBSImp as per the protocol outlined in the Martin-Harris study [26]. Each score was determined by a distinguishable observation and group consensus. Scales of each component ranged from 0 to 3 or points. The higher the score, the worst the swallowing function.

1.3.6 Flexible Endoscopic Evaluation of Swallowing (FEES)

Initial transnasal FEES was performed utilising Kay Pentax EPK15000 unit including Kaypentax camera, video and endoscope model 9213 HD, fibreoptic light source Model LH- 150P and Olympus Medical Systems Corp ENT scope Model ENFGP2. During the study FEES equipment was upgraded to Xion endoportable Model CFT-004 with videonasendoscope EN-VC CE0482. Patients were in a sitting position and video-recorded with a temporal resolution of 25 frames per second (FPS). Data was collected for six boluses varying in consistency and volume in accordance with the study protocol (thin fluids 10ml, thin fluids sequential, Level 3 moderately thick (L400) 10ml, Level 2 mildly thick (L150m) 10ml, Puree 10ml, regular diet). The YPRSRS [28] was used to quantify the severity of the residue in the pyriform sinus and vallecula observed during FEES. If the residue could not be viewed in any trial due to an

inability to visualise the vallecula or pyriform sinus it was scored as N/A. In addition, severity of airway invasion was scored using the 8-point Penetration-Aspiration (PAS) Scale.

1.3.7 Statistical Analyses

The data collected represented a sample of convenience and results were descriptive in nature. Data was analysed through SPSS ver. 25 (SPSS, Inc., Chicago, IL, USA). The design of the study was single arm pre-test post-test study. Comparison measures between VFSS, FEES and quality of life measures (DHI and SF-36) were undertaken using paired T tests. In order to take a representative of swallowing function, the Level 0 thin sequential, regular diet and Level 3 moderately thick bolus (L400) conditions were selected for both VFSS and FEES. The relationship between the PAS for VFSS and FEES was analysed through non-parametric correlation statistics. Following advice from the speech pathologist, the FEES data and the Yale Pharyngeal Residue Severity Rating Scale were simplified into a binary format such that scores 1, 2 and 3 of the scale (none, trace, mild) were coded 0 and scores 4 and 5 (moderate and severe) were coded as 1 to reflect the clinical utility of the scale. Significance was set at $p \leq 0.05$ for all comparisons.

1.4 Results

1.4.1 Demographics

Fifteen participants were screened for the study. Two participants could not tolerate the FEES assessment so were excluded from the study. One participant died during the course of the trial following a severe stroke. A total of twelve participants completed the pilot study and were very highly compliant with the intervention. There

were seven females (mean age =65) and five males (mean age=74 years). The mean age of participants was 68.8 years. The mean duration of disease was 9.5 years. Table 1 depicts the demographics of the participants during this study.

1.4.2 Swallowing Function

In all IBM patients, there was no difference in the cumulative videofluroscopic pharyngeal rating in the Level 0 thin sequential (p= 0.309) or regular bolus conditions (p=0.139) pre-and post-training illustrated in Figure 2(a) and 3(b). There was however a statistical difference in the pharyngeal rating in the Level 3 moderately thick (L400) bolus condition following training illustrated Figure 2 (c) (p=0.013). FEES analysis showed no improvement in the level of post swallow residue in the vallecular and pyriform sinus following training with the EMST device.

1.4.3 Penetration and Aspiration Score

There was no difference in the PAS scores in both VFSS and FEES imaging modalities pre-and post the EMST treatment. Given that PAS was measured in both the VFSS and FEES imaging modalities, the strength of association was measured between the two scores from each modality in two different contexts using non-parametric correlation statistics. The relationship between the PAS scores was poor for: level 0 thin bolus condition pre (r= 0.098) and post training (r=0.188) and level 3 moderately thick condition pre (r=0.101) and post training (r=0.661).

1.4.4 EMST Device in early IBM

Four participants were classified as having early IBM (duration of disease <5 years). There was no difference in cumulative pharyngeal videofluroscopic swallow study measures (level 0 thin, p=0.18; regular, p=0.09; level 3 moderately thick, p=0.06) or

quality of life in all four participants. Two participants with a more recent diagnosis (had significant reduction of residue in the vallecula after the use of the EMST training device (ID 2, p=0.001 & ID 9, p=0.01). There was also significant reduction of residue in the piriform sinus in one of the participants (ID 9) with recently diagnosed IBM (p=0.04). Unfortunately, the data obtained from the other two participants with early IBM could not be analysed to verify a trend due to inability to visualise the vallecula or pyriform sinus in the FEES assessment either pre-or post the use of the device in these participants (ID 8 and 12).

1.4.5 Quality of life

There was no change in either the abridged Dysphagia Handicap Index or the SF-36 quality of life measures after EMST exercise regime as illustrated in Tables 2 and 3. However, the results of the abridged Dysphagia Handicap Index illustrate that participants scored higher and therefore generally report more distress with the functional and emotional aspects of their dysphagia compared to the physical (Table 2). Similar to this trend, where 100 denotes the highest value of each subset of the SF-36 scoring system, Table 3 shows that participants scored higher in the Emotional Well-being, Pain and Social Functioning questions compared to the Physical Functioning questions.

1.5 Discussion

This exploratory study was designed to establish whether the EMST device would improve suprahyoid strength and in turn hyolaryngeal complex movement and therefore its' associated role in the swallowing of participants diagnosed with IBM. The overall results of this pilot study did not support the proposed prediction that 12

weeks of EMST training will result in improved swallowing function and better quality of life for participants. However, the severity of dysphagia and duration of disease were not taken into consideration, as there was a trend towards improvement in patients with a shorter disease duration. This study was inadequately powered to draw firm conclusions regarding disease duration and utility of the EMST intervention. In addition, this was a short-term pilot study designed as a short-term intervention to detect a positive difference, which may inform future longer prospective studies where it may play a more significant role in prevention of dysphagia.

This study contributes to the literature regarding the lack of correlation between two different instrumental swallowing modalities (VFSS and FEES). Prior studies have also reported a similar discrepancy between the measurements of PAS in VFSS and FEES [29, 30]. The accurate assessment of penetration and aspiration is important and the lack of reliability between the two different imaging modalities is concerning. This discrepancy between the measurement of penetration or aspiration between FEES and VFSS was also found in this study in IBM patients. This has potential critical implications for aspiration and penetration risk in IBM patients. The VFSS results are subjected to more stringent measurements. Similarly, factors affecting the internal reliability of the FEES include the fact that some patients find it difficult to tolerate.

In this pilot study, there was improvement of swallowing function noted in the VFSS of the level 3 moderately thick bolus condition (L400) compared with thin and regular fluid consistencies after EMST training. This was not reproducible in the FEES assessment. The literature shows that 82% of people with dysphagia associated with IBM have most difficulty swallowing solids rather than liquids [15, 31, 32]. Cook et. al,

and Dantas et. al, demonstrated that with a larger bolus, pharyngeal function began earlier so as to prepare the deglutitive chamber for a larger and subsequently higher velocity bolus [33, 34]. Specifically, according to the Dantas et al study the major effect of increased bolus volume was earlier onset of anterior tongue base movement, superior palatal movement, anterior laryngeal movement and UES opening [34]. Earlier UES opening was documented to be associated with an increased duration of sphincter opening [33]. It is plausible, therefore, that the difference in the large bolus condition may have been functional improvement due to the EMST device. However, as this result was not evident in the FEES assessment, and whether the significant result of the VFSS moderately thick bolus condition is indicative of a true result or measurement error needs to be further investigated. Perhaps, the effectiveness of the EMST device would have been better assessed through manometry which has the potential to evaluate pressures generated by the pharyngeal muscles pre-and post the device, however this is not a standard test in our clinic thus not selected for this pilot. In addition, the use of different outcome measures that are more specific for the physiologic measures of the swallow mechanism such as the SWAL-QoL, SSQ or Anderson may have been a better measure for quality of life than the DHI and SF-36 [35].

This pilot study had many limitations. Firstly, and importantly for an exercise intervention, we did not control for disease duration or severity of dysphagia which would have limited sample size at this single centre. We would anticipate that this intervention would be more effective in earlier disease stages than when the muscle involvement is already too severe to respond to an exercise intervention. While there was no improvement observed on VFSS assessment, our results suggest that there was improvement in swallowing in the FEES assessment in this early group where it

could be measured however given this trend was observed in only two patients whether it is representative of a true result warrants further investigation. The level of statistical significance obtained in the two patients is however very large suggesting that patients in earlier disease stages are most likely to benefit.

There are several issues that future studies investigating dysphagia in IBM could consider. Firstly, future studies should attempt to control IBM disease duration and dysphagia severity. The segregation of early verses late-stage IBM may reveal the role of EMST as a preventative intervention for dysphagia rather than restorative once the muscle weakness is already too severe. A correlation analysis between swallowing function and skeletal muscle weakness or cN1A may be fruitful. Secondly the variable outcomes detected between VFSS and FEES leave open the best way to diagnose and monitor dysphagia in IBM patients. Perhaps manometry or MRI may be more specific and more sensitive [36]. Although currently they are not readily available in a clinical environment. Lastly, the small cohort size and the lack of a placebo arm serves as a limitation to this study albeit difficult to facilitate.

Given these limitations, this pilot study does not exclude exercise therapy has an effect on maintaining or perhaps even improving swallowing function in IBM. The effect of exercise therapies in swallowing should be investigated in IBM patients through different modalities.

1.6 Conclusion

This pilot study investigated the use of the EMST device in twelve participants with dysphagia secondary to IBM. This is the first reported study that investigated the use of an exercise device to aid swallowing function in IBM patients and showed it was feasible for IBM patients. This pilot study provides some useful lessons for future

studies on exercise therapies and dysphagia in IBM. A study on a larger, less severe cohort over a longer period and using different measures is warranted to clarify the role if any, of the EMST device in IBM patients.

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Disclosure

The author reports no conflicts of interest relevant to this work

References

[1] Needham M, Corbett A, Day T, Christiansen F, Fabian V, Mastaglia FL. Prevalence of sporadic inclusion body myositis and factors contributing to delayed diagnosis. J Clin Neurosci 2008;15:1350-3.

[2] Murata KY, Kouda K, Tajima F, Kondo T. A dysphagia study in patients with sporadic inclusion body myositis (s-IBM). Neurol Sci 2012;33:765-70.

[3] Cox FM, Verschuuren JJ, Verbist BM, Niks EH, Wintzen AR, Badrising UA. Detecting dysphagia in inclusion body myositis. J Neurol 2009;256:2009-13.

[4] Olthoff A, Carstens P-O, Zhang S, et al. Evaluation of dysphagia by novel real-time MRI. Neurology 2016;87:2132.

[5] Houser SM, Calabrese LH, Strome M. Dysphagia in patients with inclusion body myositis. Laryngoscope 1998;108:1001-5.

[6] Price MA, Barghout V, Benveniste O, et al. Mortality and Causes of Death in Patients with Sporadic Inclusion Body Myositis: Survey Study Based on the Clinical Experience of Specialists in Australia, Europe and the USA. J Neuromuscul Dis 2016;3:67-75.

[7] Murata KY, Kouda K, Tajima F, Kondo T. Balloon dilation in sporadic inclusion body myositis patients with Dysphagia. Clin Med Insights Case Rep 2013;6:1-7.

[8] Danon MJ, Friedman M. Inclusion body myositis associated with progressive dysphagia: treatment with cricopharyngeal myotomy. Can J Neurol Sci 1989;16:436-8.
[9] Di Pede C, Masiero S, Bonsangue V, Ragona RM, Del Felice A. Botulinum toxin and rehabilitation treatment in inclusion body myositis for severe oropharyngeal dysphagia. Neurol Sci 2016;37:1743-5.
[10] Liu LW, Tarnopolsky M, Armstrong D. Injection of botulinum toxin A to the upper esophageal sphincter for oropharyngeal dysphagia in two patients with inclusion body myositis. Can J Gastroenterol 2004;18:397-9.
[11] Schneider I, Thumfart WF, Pototschnig C, Eckel HE. Treatment of dysfunction of the cricopharyngeal muscle with botulinum A toxin: introduction of a new, noninvasive method. Ann Otol Rhinol Laryngol 1994;103:31-5.
[12] Schrey A, Airas L, Jokela M, Pulkkinen J. Botulinum toxin alleviates dysphagia of patients with inclusion body myositis. J Neurol Sci 2017;380:142-7.
[13] Langdon PC, Mulcahy K, Shepherd KL, Low VH, Mastaglia FL. Pharyngeal dysphagia in inflammatory muscle diseases resulting from impaired suprahyoid musculature. Dysphagia 2012;27:408-17.
[14] Maugars YM, Berthelot JM, Abbas AA, Mussini JM, Nguyen JM, Prost AM. Long-term prognosis of 69 patients with dermatomyositis or polymyositis. Clin Exp Rheumatol 1996;14:263-74.
[15] Oh TH, Brumfield KA, Hoskin TL, Kasperbauer JL, Basford JR. Dysphagia in inclusion body myositis: clinical features, management, and clinical outcome. Am J Phys Med Rehabil 2008;87:883-9.
[16] Brooks M, McLaughlin E, Shields N. Expiratory muscle strength training improves swallowing and respiratory outcomes in people with dysphagia: A systematic review. Int J Speech Lang Pathol 2019;21:89-100.
[17] Arnardottir S, Alexanderson H, Lundberg IE, Borg K. Sporadic inclusion body myositis: pilot study on the effects of a home exercise program on muscle function, histopathology and inflammatory reaction. J Rehabil Med 2003;35:31-5.
[18] Heikkillä S, Viitanen JV, Kautiainen H, Rajamäki T, Mäntyvuo P, Harju T. Rehabilitation in Myositis: Preliminary study. Physiotherapy 2001;87:301-9.
[19] Spector SA, Lemmer JT, Koffman BM, et al. Safety and efficacy of strength training in patients with sporadic inclusion body myositis. Muscle & nerve 1997;20:1242-8.
[20] Pitts T, Bolser D, Rosenbek J, Troche M, Okun MS, Sapienza C. Impact of expiratory muscle strength training on voluntary cough and swallow function in Parkinson disease. Chest 2009;135:1301-8.
[21] Troche MS, Okun MS, Rosenbek JC, et al. Aspiration and swallowing in Parkinson disease and rehabilitation with EMST: a randomized trial. Neurology 2010;75:1912-9.
[22] Plowman EK, Watts SA, Tabor L, et al. Impact of expiratory strength training in amyotrophic lateral sclerosis. Muscle Nerve 2016;54:48-53.
[23] Jones HN, Moss T, Edwards L, Kishnani PS. Increased inspiratory and expiratory muscle strength following respiratory muscle strength training (RMST) in two patients with late-onset Pompe disease. Mol Genet Metab 2011;104:417-20.
[24] Ware J, Snoww K, Ma K, Bg G, SF36 Health Survey: Manual and Interpretation Guide. Vol. 30. 1993.
[25] Rose MR. 188th ENMC International Workshop: Inclusion Body Myositis, 2-4 December 2011, Naarden, The Netherlands. Neuromuscul Disord 2013;23:1044-55.
[26] Martin-Harris B, Brodsky MB, Michel Y, et al. MBS measurement tool for swallow impairment--MBSImp: establishing a standard. Dysphagia 2008;23:392-405.
[27] Balou M, Herzberg EG, Kamelhar D, Molfenter SM. An intensive swallowing exercise protocol for improving swallowing physiology in older adults with radiographically confirmed dysphagia. Clin Interv Aging 2019;14:283-8.
[28] Neubauer PD, Rademaker AW, Leder SB. The Yale Pharyngeal Residue Severity Rating Scale: An Anatomically Defined and Image-Based Tool. Dysphagia 2015;30:521-8.
[29] Kelly AM, Drinnan MJ, Leslie P. Assessing penetration and aspiration: how do videofluoroscopy and fiberoptic endoscopic evaluation of swallowing compare? Laryngoscope 2007;117:1723-7.
[30] Butler SG, Markley L, Sanders B, Stuart A. Reliability of the penetration aspiration scale with flexible endoscopic evaluation of swallowing. Ann Otol Rhinol Laryngol 2015;124:480-3.
[31] Mulcahy KP, Langdon PC, Mastaglia F. Dysphagia in inflammatory myopathy: self-report, incidence, and prevalence. Dysphagia 2012;27:64-9.
[32] Oh TH, Brumfield KA, Hoskin TL, Stolp KA, Murray JA, Bassford JR. Dysphagia in inflammatory myopathy: clinical characteristics, treatment strategies, and outcome in 62 patients. Mayo Clin Proc 2007;82:441-7.

[33] Cook IJ, Dodds WJ, Dantas RO, et al. Timing of videofluoroscopic, manometric events, and bolus transit during the oral and pharyngeal phases of swallowing. Dysphagia 1989;4:8-15.
[34] Dantas RO, Kern MK, Massey BT, et al. Effect of swallowed bolus variables on oral and pharyngeal phases of swallowing. Am J Physiol 1990;258:G675-81.
[35] McHorney CA, Robbins J, Lomax K, et al. The SWAL-QOL and SWAL-CARE outcomes tool for oropharyngeal dysphagia in adults: III. Documentation of reliability and validity. Dysphagia 2002;17:97-114.
[36] Carstens PO, Schmidt J. Diagnosis, pathogenesis and treatment of myositis: recent advances. Clin Exp Immunol 2014;175:349-58.

Figure 1 EMST study timeline

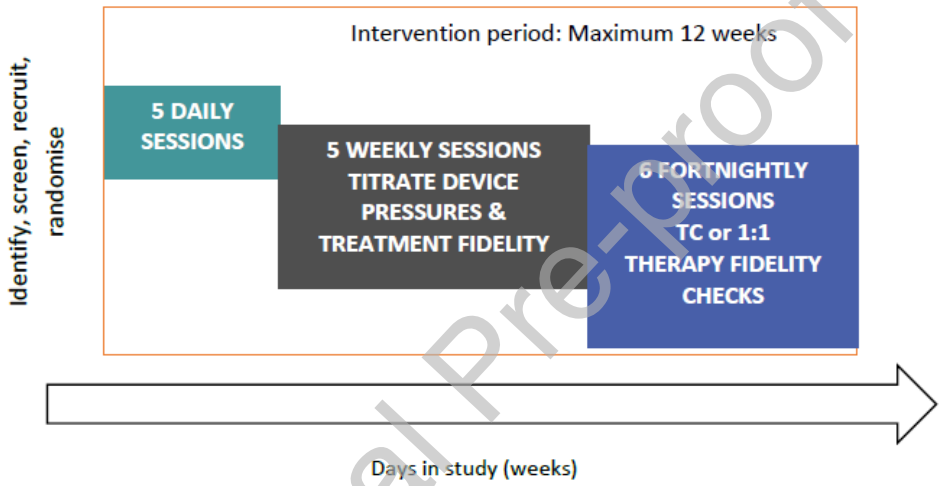

Figure 2a and 2b Cumulative pharyngeal MBSImp score pre-and post the EMST device in the Level 0 thin and regular sequential bolus condition

Figure 2c Cumulative pharyngeal MBSImp score pre-and post the EMST device in the Level 3 Moderately thick bolus condition (L400)

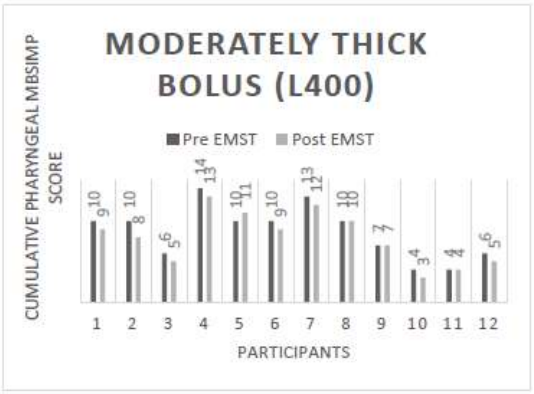

Table 1 Demographics of the participants recruited in this pilot study

Participant ID	Sex	Age (years)	Duration of disease (years)
1	F	70	17
2	F	59	3
3	M	76	13
4	M	77	18
5	F	69	6
6	M	77	10
7	F	72	12
8	F	56	1
9	M	67	4
10	F	61	10
11	F	68	16
12	M	73	4

Table 2 The abridged Dysphagia Handicap Index results

	Pre EMST	***Post EMST***	***Mean Difference***	***p value***
Physical	10.08	9.83	0.25	0.760
Functional	18.25	19.17	-0.917	0.511
Emotional	13.42	13.82	-0.415	0.724

Table 3 The SF-36 Health Survey results

	Pre EMST	***Post EMST***	***Mean Difference***	***p value***
Physical Functioning	24.17	24.92	-0.75	0.804
Emotional Well-being	75.33	74	1.33	0.762
Pain	66.46	66.67	-0.21	0.970
Social Functioning	62.5	62.5	0	1.0