# Impact of Expiratory Strength Training in Amyotrophic Lateral Sclerosis

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## Abstract

**Introduction**—We evaluated the feasibility and impact of Expiratory Muscle Strength Training (EMST) on respiratory and bulbar function in persons with amyotrophic lateral sclerosis (ALS).

**Methods**—25 ALS patients participated in this delayed intervention open-label clinical trial. Following a lead-in period, patients completed a 5-week EMST protocol. Outcome measures included: maximum expiratory pressure (MEP), physiologic measures of swallow and cough, and Penetration-Aspiration Scale (PAS) scores.

**Results**—Of those participants who entered the active phase of the study (n=15), EMST was well tolerated and led to significant increases in MEPs and maximum hyoid displacement during swallowing post-EMST ($P<0.05$). No significant differences were observed for PAS scores or cough spirometry measures.

**Discussion**—EMST was feasible and well tolerated in this small cohort of ALS patients and led to improvements in expiratory force-generating pressures and swallow kinematics. Further investigation is warranted to confirm these preliminary findings.

## Keywords

Amyotrophic Lateral Sclerosis; Rehabilitation; Respiratory Training; Expiratory Muscle Strength Training; Swallowing

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## Introduction

Swallowing impairment occurs in up to 85% of individuals with amyotrophic lateral sclerosis (ALS)[1, 2] and is both functionally and psychologically devastating. Malnutrition and aspiration pneumonia, both consequences of dysphagia, increase the risk of death by 7.7 times and contribute to 25.9% of patient mortality[3, 4]. Beyond these direct physical consequences, dysphagia also contributes to social isolation, fear, and decreased mental health in this patient population[5].

Dysphagia in ALS results from weakness and/or spasticity of the muscles of deglutition, including muscles of mastication, the tongue, lips, pharynx, and larynx[6], while weakness of the respiratory and ventilatory musculature further impairs airway protection by reducing the expiratory pressure generation capacities necessary to produce an effective cough[7, 8]. In a recent videofluorosopic analysis of 18 patients with ALS who aspirated, 45% demonstrated a weak and ineffective cough response, 55% demonstrated an absent cough response, and none could produce a cough response sufficient to expel tracheal aspirate[9]. Gaziano and colleagues concluded that management strategies to improve vestibule closure, expiratory force generating pressure, and cough strength might enhance airway protection during swallowing in individuals with ALS[9].

Although animal and human studies suggest that exercise may have a beneficial impact on motor function and motor neurons serving the extremities[10–19], the role of targeted exercise in ameliorating the effects of motor neuron degeneration in the bulbar and respiratory systems of ALS patients has not been studied systematically. In this study, we assessed the feasibility and impact of Expiratory Muscle Strength Training (EMST) on expiratory force generating ability, swallow kinematics, cough physiology, and airway protection in individuals with ALS.

## METHODS

### Participants

*25* individuals with a diagnosis of probable or definite ALS by the Revised El-Escorial criteria[20] were recruited. The diagnosis of ALS was confirmed in all patients by academic neuromuscular neurologists who specialize in ALS at the University of South Florida ALS Center prior to screening for enrollment. Patients were enrolled if they had: 1) reduced maximum expiratory pressure (MEP) compared with published normative data for gender and age[21]; 2) forced vital capacity greater than 60%; 3) preserved cognition as evidenced by a score of >24 points on the Mini-Mental Status Exam[22]; 4) no allergies to barium; 5) no tracheostomy or mechanical ventilation; 6) no diaphragmatic pacer; and 7) no significant concurrent respiratory disease. This study was approved by the University of South Florida Institutional Review Board (Protocol 00005321) and conducted in accordance with the Declaration of Helsinki. All included patients provided informed written consent.

### Design

A repeated measures design was used and included a 5-week lead-in period to benchmark bulbar disease progression. Patients attended 3 evaluations, each separated by 5-weeks. At

each evaluation, respiratory, swallow, and cough functions were assessed. Testing order was maintained across evaluations for each participant to minimize any potential order effect due to patient fatigue. Between baseline evaluations 1 and 2, no interventions were conducted, and participants were instructed to perform their usual daily activities and typical routine. Immediately following baseline evaluation 2, a 5-week EMST program commenced. At the completion of the EMST program, participants were evaluated for the third and final time.

### Primary outcome measure: Maximum expiratory pressure

MEP was the primary outcome variable. It was assessed using a handheld digital manometer (Micro Respiratory Pressure Meter, MP01, Micro Direct Inc.). To minimize labial leakage in patients who were unable to create a tight lip seal due to facial weakness, a flanged rubber mouthpiece was attached to the manometer. During MEP testing, the participant was seated with a nose clip in place to occlude the nasal cavity, and after inhaling to total lung capacity, he/she was instructed to place his/her lips around the mouthpiece and blow out as forcefully as possible. Three trials within 10% were collected, and each participant's highest MEP was used in the analysis.

### Secondary measures: Physiologic measures of swallowing

Swallow function and airway safety were evaluated radiographically using the videofluoroscopic swallowing study (VFSS). Participants were seated upright in a lateral viewing plane using a properly collimated Phillips BV Endura fluoroscopic C-arm unit (GE OEC 8800 Digital Mobile C-Arm system type 718074). A Swallowing Signals Lab unit (Pentax, Lincoln Park, NJ) digitally recorded the fluoroscopic images at 29.97 frames per second using a scan converter. A standardized bolus presentation protocol was used and consisted of 2 1-cc boluses of liquid contrast agent (barium), 1 3-cc bolus of thin liquid contrast, 1 3-cc bolus of paste, 1 20-cc bolus of liquid contrast, 1 90-cc bolus of thin liquid contrast in the lateral view, and a 20-cc bolus of liquid contrast in the anterior/posterior view. To ensure patient safety, the VFSS was terminated immediately after a second aspiration event on any of the 7 swallow trials. Images were recorded digitally onto the Pentax Swallow Workstation for subsequent analysis. Temporal and kinematic objective measures of oropharyngeal swallowing were performed by a blinded rater using the standardized and validated measures of Leonard[23]. These assessments included: 1) hypopharyngeal transit time (ms), 2) oropharyngeal transit time (ms), 3) pharyngoesophageal segment opening (mm), 4) maximum hyoid displacement (mm), 5) hyoid to larynx approximation (mm), and 6) pharyngeal constriction ratio ($cm^2$)[23]. Appendix A (supplementary material, available on-line) provides a definition for each of these measures. Airway safety was evaluated using the validated Penetration-Aspiration Scale (PAS)[24], an 8-point ordinal scale of airway safety that describes the degree of airway invasion, the participant's response, and whether the invasive material is successfully ejected from the airway (see Appendix B, supplementary material available on-line). All objective ratings were performed in a blinded fashion on the 20cc bolus challenge, or the largest bolus the participant was able to swallow.

### Secondary measures: Voluntary cough spirometry

Cough was assessed using an oral pneumotachograph (MLT 1000, ADInstruments, Inc; Colorado Springs, CO), connected to a spirometer filter (MQ 304 Spirometer Filter,

Vacumed; Ventura, CA) during voluntary cough production. The participant was seated with a respiratory facemask held in place by the examiner and instructed to complete 3 tidal breaths into the pneumotachograph, take a deep breath in and then "cough hard like something is stuck in your throat." The patient was provided with a model by the examiner that demonstrated 3 very strong coughs in succession and ensured the patient understood the requirements of this task. Airflow signal was measured and low-pass filtered at 150Hz, digitized at 1000 Hz, and displayed on a portable laptop computer using Lab Chart Version 7 (Microsoft Corp; Redmond, WA). Physiologic voluntary cough airflow measures deduced from the cough flow waveforms included: 1) inspiratory phase duration (time from onset of inspiration following tidal volume breathing to the end of inspiration prior to the compression phase of cough); 2) inspiratory peak flow (peak inspiratory flow during the inspiratory phase preceding the cough); 3) compression phase duration (time from the end of the inspiratory phase to the beginning of the expiratory phase); 4) expiratory rise time (time from the beginning of expiratory phase to the peak expiratory flow); 5) expiratory peak airflow (peak airflow during the expiratory phase of the cough); and 6) cough volume acceleration (expiratory peak flow divided by expiratory rise time). Cough spirometry measures were assessed on the first cough epoch across the 3 trials by a blinded rater.

### EMST Exercise Protocol

A 5-week exercise protocol commenced immediately following the 5-week baseline evaluation using the handheld calibrated, one-way, spring-loaded valve trainer. EMST was performed with the trainer set at 50% of a patient's individual MEP, representing a moderate load on the expiratory muscles. Expiratory force generating pressures in participants ranged between 12–122 cmH$_2$0. Therefore, 2 expiratory training devices were utilized that were capable of calibrated changes in the resistance or physiologic load to the target muscles to facilitate this patient range. A low range pressure trainer with a small range between 0–20 cm H$_2$0 was used by participants whose MEPs were <40cm H$_2$0 (Phillips Threshold PEP, Positive Expiratory Pressure Trainer, Respironics; Cedar Grove, NJ, USA). For participants whose MEPs were >40cm H$_2$0; a higher range pressure trainer capable of setting a pressure threshold between 20–150cm H$_2$0 was utilized (EMST 150; Aspire Products; Gainesville, FL, USA). A flanged rubber mouthpiece was attached to the opening of the trainer to help create a tight lip seal.

EMST therapy sessions were completed 5 days per week. During training the participants sat in a comfortable position and wore a nose clip. They took a deep breath, held their cheeks lightly (or had a caregiver do so) and blew forcefully into the training device to break the seal. A single daily training session consisted of 25 targeted forced exhalations through the trainer, performed in 5 sets of 5 repetitions. This was described to participants as the "Rule of Fives"[25]. Participants were instructed to rest between each 5-breath set, and a typical training session lasted approximately 20 minutes each day. On the first training session of each week, patients attended the university swallowing clinic where a research therapist reassessed MEP using a handheld digital manometer (MP01, Micro Direct Inc.) and recalibrated the EMST trainer to 50% of current MEP value. After this adjustment, the patient performed an EMST therapy session with the therapist. The remaining 4 therapy sessions were performed at home with the assistance of the participant's caregiver.

### Statistical Analysis

Descriptive statistics were performed to determine attrition, feasibility, and exercise tolerability in this study across time points. To assess the effect of EMST on respiratory, swallow, and cough function, a repeated measures analysis of variance (ANOVA) was performed on all outcome variables across the 3 time points with alpha set at 0.05 using SPSS (Version 22). *Post-hoc* analysis (LSD pairwise comparisons) was performed when significant main effects were observed.

## RESULTS

Table 1 summarizes demographic details of recruited patients, those who completed the study, and individuals who dropped out. Six participants withdrew following the initial baseline evaluation (during the 5-week lead in period). Of the remaining 19 patients who entered the active treatment phase, 4 individuals withdrew, 1 of whom had relocated to a different state. The overall attrition rate in this study was 40%, however of those entering the active phase, 79% completed the EMST protocol. EMST was well tolerated with no reports of pain, fatigue, discomfort, or other adverse events. The final analysis was performed on 15 ALS patients.

### Primary Outcome: Maximum Expiratory Pressure

A significant main effect was observed for the primary outcome variable, MEP, $F_{(2)}=4.26$, $P<0.03$. *Post-hoc* testing revealed a significant increase in MEPs between Baseline 2 vs. post-EMST ($P=0.01$; absolute mean difference=17.17, SE: 5.87; CI:4.57, 29.76). Mean MEP data are shown in Figure 1A.

### Secondary Outcome: Swallowing Kinematics

A significant main effect was noted for maximum hyoid displacement during swallowing, $F_{(2)}=5.48$, $P<0.02$. *Post-hoc* analysis revealed a significant increase in maximum hyoid displacement between Baseline 1 vs. Post-EMST ($P=0.01$; absolute mean difference=54mm; SE:18mm; CI:15mm, 93mm) and Baseline 2 vs. Post-EMST ($P=0.03$; absolute mean difference=57mm; SE:23mm; CI:7mm, 106mm) testing points (Figure 1B). No other significant differences were noted for objective swallowing measures or airway safety during swallowing (PAS).

### Secondary Outcome: Voluntary Cough

No significant differences were noted for voluntary cough spirometry measures. For the patients who completed EMST, mean (SD) bulbar subscale ALSFRS-R scores were 9.0 (2.0), 8.5 (2.1), and 7.92 (2.2) for baseline 1, baseline 2 and post-EMST time points respectively. Mean (SD) ALSFRS-R respiratory subscale scores were 9.2 (3.1), 9.1 (2.8), and 8.6 (3.3) for baseline 1, baseline 2, and post-EMST time points respectively.

## DISCUSSION

These findings indicate that for participants who entered the active phase of the trial, EMST was feasible, safe, and well tolerated in this small cohort of ALS patients and led to

immediate gains in expiratory force and greater hyoid displacement during swallowing. The dropout of more severely affected ALS patients could be due to generalized fatigue, increased transfer and travel problems, or other unidentified factors, but it could also be that EMST is best tolerated by ALS patients in the early stages of the disease. Future studies could target mildly to moderately affected ALS subjects in early stages of the disease and could also incorporate weekly home therapy visits to minimize patient travel.

In this study, a 5-week moderate load EMST program led to increases in maximum expiratory force-generating capacities and hyoid displacement during swallowing. The noted impact of EMST in this cohort of ALS patients is similar to those reported in the sedentary elderly[26], patients with Parkinson disease[25, 27, 28], and multiple sclerosis[29]. Maintenance of subglottic air pressure-generating abilities is of high relevance in ALS patients who have severely diminished capacities to generate adequate expiratory pressures to produce an effective cough and to protect their airways[7,8,9].

The increase we observed in hyoid excursion during swallowing following EMST has also been documented in Parkinson disease patients who underwent a similar EMST program[25]. This improvement in hyolaryngeal kinematics is likely attributed to supramaximal engagement and activation of the submental and suprahyoid musculature that have been observed with expiratory resistance threshold tasks[30]. Therefore, EMST likely engaged this muscle complex that is primarily responsible for the anterior and superior excursion of the hyoid during swallowing. Hyoid displacement is an important kinematic event during swallowing, as the anterior and superior movement passively aids relaxation and opening of the upper esophageal sphincter to allow effective bolus transit from the pharynx into the esophagus. Improvement in hyoid displacement has relevance for individuals with ALS who may experience upper esophageal sphincter dysfunction[31] which can lead to significant residue in the pyriform sinuses and further increases the risk of aspiration.

Although no statistical differences were observed in cough spirometry measures, inspiratory phase duration (IPD) was, on average, 22% shorter following EMST ($P$=0.06). This likely represents a clinically meaningful improvement, given the fact that the IPDs in this group of ALS patients were not within the expected normal reference ranges and were approximately 2.5 times longer than the pre-intervention IPDs noted in a comparable intervention study in individuals with Parkinson disease[32]. The reduction noted in IPD may have led to improvements in the ability of the ALS patient to efficiently inflate the lungs as well as improve the time and efficiency to glottic closure. During the treatment exercises, patients are instructed to take in a deep breath to fill their lungs prior to the forceful expiration maneuver. This may be important to reach efficiency in building up sub-glottic pressure to produce an effective cough.

This pilot study was performed in a relatively small cohort of patients, and although a lead-in period was utilized to serve as a within-subject control, we did not test a comparison control group. Given the nature of this pilot study, a lead-in design to assess for baseline vs. treatment phase differences in the same subjects over time was chosen to afford the ability to control for potentially substantial differences in native progression rates by comparing subjects over their treatment periods against themselves during a defined period prior to

treatment (an important feature for pilot studies not large enough to wash out large progression rate differences between randomized placebo and treatment groups). Further, although an attempt was made to screen cognitive function using the MMSE, this test is not sensitive in detecting the cognitive and behavioral impairment in ALS, and future work should incorporate a more sensitive instrument to screen for cognitive dysfunction. Additionally, patients were not followed over time to examine the long-term impact or any detraining effects following the exercise intervention. These limitations form the basis for our current 8-week randomized sham control trial being conducting in a larger cohort of mild to moderately impaired ALS patients. In addition, this trial utilizes a home therapist to minimize patient demands and includes follow-up evaluations to determine the impact of EMST on global disease progression and survival.

## CONCLUSION

This pilot trial suggests that EMST improves maximum expiratory pressure in ALS patients. Additional investigation is needed to validate these preliminary findings in a larger randomized, controlled trial. Future studies will investigate the long-term impact of EMST on breathing, cough, and airway protection and on global disease progression and survival.

## Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

## Acknowledgments

## Abbreviations

| | |
|---|---|
| **(ALS)** | Amyotrophic Lateral Sclerosis |
| **(EMST)** | Expiratory Muscle Strength Training |
| **(MEP)** | Maximum Expiratory Pressure |
| **(ANOVA)** | Analysis of Variance |
| **(PAS)** | Penetration aspiration scale |
| **(ALSFRS-R)** | Amyotrophic Lateral Sclerosis Rating Scale – Revised |

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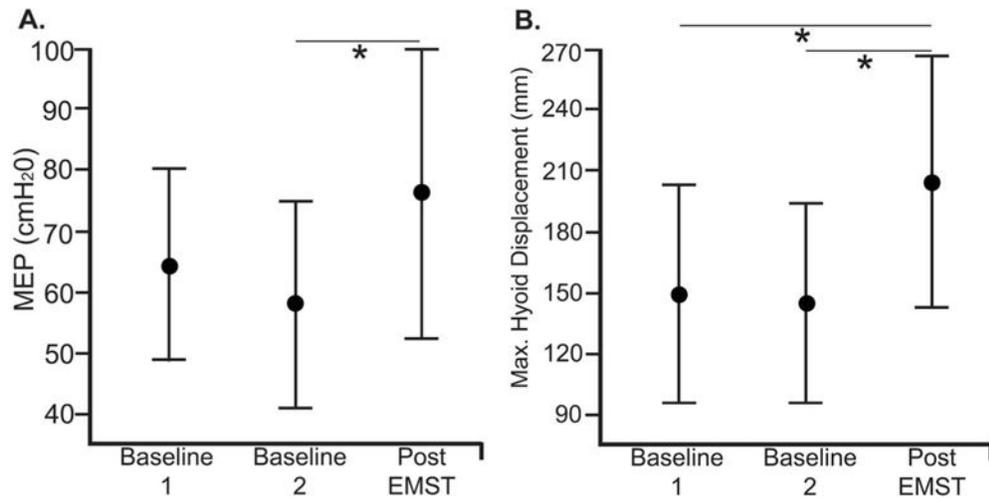Author Manuscript

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**Figure 1.**
**A**. Mean Maximum Expiratory Pressure (MEP) across testing sessions. MEPs were significantly higher following the EMST intervention compared to Baseline 2 ($P<0.02$). **B**. Mean maximum hyoid displacement (mm) across testing sessions. Hyoid displacement during swallowing was greater post-EMST intervention compared to both baseline tests ($P<0.05$). Error bars are 95% C.I. and * denotes a significant difference between evaluation time points.

**Table 1**

Summary of participant demographics.

| | Entire Cohort (N=25) | Completed (N=15) | Dropped Out (N=10) |
|---|---|---|---|
| Gender (M/W) | 14/11 | 7/8 | 7/3 |
| Age (years) | 62.2 (10.5) | 62.1 (13.2) | 62.3 (4.4) |
| ALSFRS-R Total | 32.0 (8.5) | 33.0 (7.7) | 30.4 (9.9) |
| Disease Duration (months) | 14.5 (11.7) | 16.3 (13.5) | 11.8 (8.5) |
| Onset Type (Spinal/Bulbar) | 15/10 | 10/5 | 5/5 |
| Swallowing Status: (Safe/Penetrator/Aspirator) | 15/4/6 | 10/1/4 | 5/3/2 |

Summary data are provided as mean (standard deviation) for age, ALSFRS-R, and disease duration. Raw counts are provided for gender, disease onset type, and swallowing status.