CLINICAL RESEARCH ARTICLE

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## Lung volume recruitment improves volitional airway clearance in amyotrophic lateral sclerosis

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## Abstract

**Introduction/Aims:** In this study we evaluated the effects of lung volume recruitment treatment (LVR), a low-tech, low-cost, manual "breath-stacking" technique used to help people cough with enough force to clear their airways, thereby reducing the risk of aspiration and choking, on five volitional airway clearance and protection behaviors used by people living with amyotrophic lateral sclerosis (PwALS).

**Methods:** Using a repeated-measures cross-over design, 29 PwALS performed five volitional airway clearance and protection behaviors in LVR treatment and in no-treatment, control conditions. Peak cough flow (PCF) was used to measure maximum expiratory rate during forced expiration, throat clearing, hawking, post-swallow coughing, and the supraglottic swallowing maneuver. Comparisons were made as a function of condition (treatment or control) and three time-points (pretreatment, and 15 and 30 minutes posttreatment).

**Results:** LVR treatment had a significant positive effect on maximum expiratory rates during all tested airway clearance and protection behaviors. Increased PCF values lasted for up to 30 minutes post-LVR for all tested behaviors in the treatment condition.

**Discussion:** We found that LVR treatment could increase control over airway clearance in PwALS, as well as provide improved airway protection for up to 30 minutes, the duration of a typical meal. This study has implications for patient care. These include offering patients control over some of the most feared symptoms of ALS, particularly choking during activities of daily living, and enhanced ALS respiratory care in low-resource settings. Findings may have implications for other neurodegenerative disorders in which dysphagia occurs with retained sensory function.

#### KEYWORDS

activities of daily living, amyotrophic lateral sclerosis, quality of life, rehabilitation, respiratory function

Abbreviations: ALS, amyotrophic lateral sclerosis; ANOVA, analysis of variance; LVR, lung volume recruitment; PCF, peak cough flow; PwALS, people living with amyotrophic lateral sclerosis; VACM, volitional airway clearance maneuver.

Portions of this work were presented at the annual convention of the American Speech-Language and Hearing Association, San Diego, California, in 2011, and the annual research forum of the ALS Society of Canada, Toronto, Ontario, Canada, in 2011.

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## 1 | INTRODUCTION

With the progressive upper and lower motor neuron degeneration that is inherent to amyotrophic lateral sclerosis (ALS), people with ALS (PwALS) have encumbered airways, compromised airway clearance capacity, and dysphagia.<sup>1,2</sup> These symptoms can have life-threatening consequences,<sup>3,4</sup> and the fear of "choking to death" is one of the most distressing features of ALS for many people living with this disease.<sup>5-8</sup> Strategies to treat and manage these challenges should be initiated early to facilitate respiratory function, prevent complications, and improve quality of life.<sup>9,10</sup>

Lung volume recruitment (LVR), also called "breath stacking," is a simple, low-cost, manual insufflation and cough augmentation technique.<sup>11,12</sup> Considered standard first-line therapy at many Canadian ALS clinics, LVR offers PwALS some degree of autonomy and control,<sup>13</sup> aspects of clinical care that are critical to patient wellbeing.<sup>14-17</sup> Although previous studies demonstrated the positive effect of breath stacking<sup>18-22</sup> and call for more timely prescription of cough adjunct devices such as LVR,<sup>23</sup> studies to date have not explored how LVR treatment may be used to augment volitional airway clearance and protection behaviors.

In previous work we investigated the effects of LVR on pulmonary function and spontaneous coughing in PwALS.<sup>18</sup> In this study we analyzed unpublished data from the same body of research and patient cohort to determine whether LVR may be used to amplify the peak cough flow (PCF) generated by five volitional airway clearance and protection maneuvers, each of which targets different regions of the throat and esophagus. The five behaviors evaluated in this investigation are clinically important, as they capitalize on sensory capacities that are generally spared in ALS-specifically those that allow PwALS to sense when and where materials are retained in their airways.<sup>24</sup> Forced expiration is used to clear the lungs, trachea, and larynx.<sup>25</sup> Throat clearing involves closure of the supraglottic folds and is effective in moving material out of the laryngeal vestibule and into the pharynx.<sup>26</sup> Hawking, a rapid exhalation followed by contact of the soft palate and the base of the tongue, is used to propel materials from the oropharynx into the anterior portion of the oral cavity.<sup>26</sup> The post-swallow cough is an airway protection behavior that guards against fluids, foods, or secretions entering the larynx and lungs. The supraglottic swallow maneuver aims to achieve volitional vocal fold closure before and during the swallow by causing the arytenoid cartilages to tilt anteriorly toward the base of the epiglottis.<sup>27</sup> This is recommended to improve two of the most significant dysphagic challenges for PwALS: laryngeal closure and airway protection.28

In this study we investigated the following questions: (1) What is the effect and effect duration of LVR on the maximal expiratory airflow for three volitional airway clearance maneuvers (VACMs) specifically, forced expiration, throat clearing, and hawking? (2) What is the effect and effect duration of LVR on maximal expiratory airflow during two compensatory swallowing techniques, the post-swallow cough and the supraglottic swallow maneuver?

## 2 | METHODS

# 2.1 | Standard protocol approval, registration, and patient consent

This study was approved by the health research ethics board at the University of Alberta, Canada (#B-031106). All study participants provided informed consent.

## 2.2 | Participants

Participants were recruited through a multidisciplinary, hospital-based ALS clinic and met the following inclusion criteria: (1) diagnosis with possible, probable, laboratory-supported probable, or definite ALS by the revised El Escorial criteria<sup>29</sup>; (2) fluent in English and able to provide informed consent; (3) able to consume some food and liquid by mouth; and (4) prescribed LVR therapy, received instruction in its use from a respiratory therapist, and used LVR successfully for at least 2 weeks before the study.

## 2.3 | Research design

We used a repeated-measures cross-over research design to examine the effects of LVR on pulmonary function and spontaneous coughing (as reported by Cleary et al<sup>18</sup>), and on three VACMs and two compensatory swallowing techniques (current investigation). Participants completed a treatment session and a no-treatment control session within the same week, separated by at least 24 hours. Half of the participants received the LVR treatment session followed by the control session, and the other half participated in the reverse order. Participants were asked to refrain from LVR for 24 hours before each research session. Participants performed the volitional and protective behaviors in the following order: forced expiration, throat clearing, hawking, post-swallow cough, and supraglottic swallow maneuver. Participants swallowed a 10-mL calibrated cup sip of cold water for the compensatory swallow techniques. The investigator provided standard instructions and modeling for each behavior in both treatment and nontreatment sessions. After each set of instructions, participants performed the target behavior. For each behavior, baseline measures were taken (time 1), followed by LVR with posttreatment peak cough flow (PCF) measurement at 15 minutes (time 2) and 30 minutes (time 3). LVR was performed according to accepted protocols (see Supplementary File S1) and using a manual resuscitation bag (Mercury Medical, Clearwater, Florida<sup>30</sup>) equipped with a one-way flow valve and mouthpiece. Assistance with squeezing the resuscitator bag was provided as needed, and participants who were accustomed to using a facemask instead of a mouthpiece were permitted to do so. The protocol was the same for control sessions, except LVR was not administered.

We used PCF, the maximum expiratory airflow produced as the vocal folds open during a cough or air expiration, to measure the effects of the five target behaviors with and without LVR at times 1, 2, and 3. We measured PCF using a flow meter (Health Scan ASSESS Peak Cough Flow Meter; Phillips Respironics, Amsterdam, The Netherlands).<sup>31-33</sup> Outcomes were compared with the following standard measures: at least 160 L/min PCF is required to move mucus from the lungs into the upper airway;<sup>32,34-36</sup> at least 80 L/min is effective for airway protection<sup>36</sup>; and individuals with PCF less than 270 L/min are considered at risk for airway encumbrance and increased respiratory morbidity.<sup>35,37</sup>

## 2.5 | Data analyses

Methods used to analyze the effect and effect duration of LVR on VACMs were described previously<sup>18</sup> and are summarized here. We conducted separate two-way repeated-measures analysis of variance (ANOVA) tests for each dependent variable. The within-subjects' factors were Condition (LVR treatment vs control) and Time (times 1, 2, and 3). The Condition main effect, Time main effect, and Condition × Time interaction effects were tested using the multivariate criterion of Wilks'  $\Lambda$ . Tests for significant ANOVA effects were conducted using paired sample *t* tests. The Bonferroni adjustment procedure was used to control for increased risk of type I error across the *t* tests. Alpha was set at  $P \leq .001$  for all post-hoc tests.

Participants' best scores out of three test trials at each time-point in each condition were used in the analyses, as per standards of the American Thoracic Society and European Thoracic Society.<sup>38</sup> If all test trials for the target behavior in each condition could not be completed, we excluded participant data for that specific behavior from analysis.

## 3 | RESULTS

### 3.1 | Sample

The study sample consisted of 13 women and 16 men (n = 29). Participants' average age was 65.4 (standard deviation [SD],

11.5) years. Twenty-two participants (75.9%) had limb-onset ALS, six (20.7%) had bulbar onset, and one had respiratory onset. Time since diagnosis was, on average, 21.68 (SD, 19.86) months. Most participants lived in urban settings and in their own homes (n = 26). Two resided in long-term care facilities. One was in hospital during data collection. At the time of the study, participants had been doing LVR treatment for an average of 4.8 (SD, 7.01) months. Nine participants required assistance with squeezing of the resuscitator bag and six participants used a facemask instead of the mouthpiece. A summary table of participants' characteristics at study onset is available in Supplementary File S2.

# 3.2 | Effect of LVR on maximal expiratory airflow during forced expiration, throat clearing, and hawking

Twenty-four participants completed all forced expiration and throat-clearing tasks; 21 completed all hawking tasks. Dropouts occurred because of fatigue or inability to consistently perform the required behavior. LVR treatments, when compared with notreatment (control) conditions, produced statistically significant therapeutic effects on PCF results with all VACMs over time. This is demonstrated by the interaction effect of Condition × Time for forced expiration [ $\Lambda = .56$ , *F* (2, 22) = 8.82, *P* = .002, eta<sup>2</sup> = 0.445], throat clearing [ $\Lambda = 0.67$ , *F*(2, 22) = 5.49, *P* = .012, eta<sup>2</sup> = 0.333], and hawking [ $\Lambda = 0.40$ , *F*(2, 19) = 14.26, *P* < .001, eta<sup>2</sup> = 0.600] (Table 1).

## 3.2.1 | Forced expiration

Thirty minutes after LVR treatment (time 3), participants had significantly higher PCF results than they did in the control condition at the same time-point [t(26) = 3.79, P < .001]. Within the LVR treatment condition, significant differences were evident between times 1 and 3 [t(26) = -4.81, P < .001]. This indicates that the LVR treatment effect, when used in conjunction with forced expiration, lasted for 30 minutes (Table 1).

Condition	Time 1 (L/min)	Time 2 (L/min)	Time 3 (L/min)
Forced expiration (n = 24)			
LVR treatment	217.08 (115.4)	249.38 (110.8)	251.04 (112.1)
Control	223.54 (113.2)	214.79 (115.4)	218.13 (113.8)
Throat clearing (n = 24)			
LVR treatment	160.42 (65.6)	204.58 (77.8)	190.84 (62.9)
Control	167.71 (72.5)	165.42 (71.0)	172.29 (113.9)
Hawking (n $=$ 21)			
LVR treatment	116.19 (58.4)	156.67 (54.5)	148.33 (56.3)
Control	134.29 (64.1)	123.09 (55.8)	120.00 (52.5)

 TABLE 1
 PCF during VACMs by

 forced expiration, throat clearing, and

 hawking

Note: Data expressed as mean (standard deviation).

Abbreviations: LVR, lung volume recruitment; PCF, peak cough flow; VACM, volitional airway clearance maneuver.

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 TABLE 2
 PCF during compensatory

 swallowing techniques with post-swallow
 cough and supraglottic swallow

 maneuver
 maneuver

Condition	Time 1 (L/min)	Time 2 (L/min)	Time 3 (L/min)	
Post-swallow cough (n $=$ 27)				
LVR treatment	244.31 (101.18)	299.64 (141.82)	292.24 (119.11)	
Control	255.17 (119.91)	251.61 (115.24)	254.48 (117.43)	
Supraglottic swallow maneuver (n $=$ 19)				
LVR treatment	265.00 (147.4)	310.26 (151.9)	304.47 (139.5)	
Control	267.64 (134.0)	261.58 (134.3)	269.47 (138.9)	

Note: Data expressed as mean (standard deviation).

Abbreviations: LVR, lung volume recruitment; PCF, peak cough flow.

## $\label{eq:table_transform} \begin{array}{ll} \textbf{TABLE 3} & \text{Numbers of participants above and below PCF flow} \\ \text{threshold of 180 L/min (n = 29)} \end{array}$

Condition	Baseline time 1	15 mins post-treatment time 2	30 mins post-treatment time 3
LVR treatment			
<180 L/min	9	5	5
≥180 L/min	20	23	24
Control			
<180 L/min	8	8	8
≥180 L/min	21	20	21

Abbreviation: LVR, lung volume recruitment.

## 3.2.2 | Throat clearing

Participants had significantly higher PCF results at time 3 in the LVR treatment condition as compared with the control condition [t(23) = 4.23, P < .001]. After LVR treatment, significant differences were also found between times 1 and 2 [t(26) = -4.55, P < .001], and between times 1 and 3 [t(26) = -4.60, P < .001]. This indicates that LVR had a significant positive effect on PCF during throat clearing that lasted for 30 minutes.

## 3.2.3 | Hawking

PCF measures were significantly higher in the treatment condition as compared with the control condition for hawking at times 2 [t(21) = 4.84, P < .001] and 3 [t(23) = 4.23, P < .001]. After LVR treatment, we also found significant differences between baseline and 15 minutes [t(22) = -4.67, P < .001], and between baseline and 30 minutes [t(22) = -4.85, P < .001] posttreatment. This indicates that, when used in conjunction with hawking, LVR had a significant positive effect on PCF that lasted for 30 minutes.

# 3.3 | Effect of LVR on post-swallow coughing and supraglottic swallow maneuver

### 3.3.1 | Post-swallow coughing

Twenty-seven participants successfully completed all post-swallow coughing trials. Dropouts occurred because of fatigue or inability

to consistently perform the required behavior. We found no significant differences in PCF rates between the treatment and control conditions at baseline; however, PCF values were significantly higher 15 and 30 minutes after LVR treatment when compared with the same time-points in the control condition, time 2 [t(26) = 5.329, P < .001], and time 3 [t(28) = 4.650, P < .001]. At these time-points, and with LVR treatment, PCF values were greater than 270 L/min. In the treatment condition, there was a significant difference in PCF results between times 1 and 2 [t(27) = -5.848, P < .001] and between times 1 and 3 [t(28) = -8.115, P < .001]. There were no significant differences across time in the control condition (Table 2).

Individual-level data were analyzed in relation to the minimum PCF threshold of 180 L/min for airway protection. In the treatment condition, nine participants (31%) were below this threshold at baseline. Five (17%) remained below this threshold at times 2 and 3. No individual with baseline PCFs greater than 180 L/min had flow rates less than 180 L/min at subsequent time-points. In the control sessions, eight participants (28%) produced below-threshold PCFs at all three time-points (Table 3).

### 3.3.2 | Supraglottic swallow maneuver

Nineteen (65.5%) participants completed all six supraglottic swallowing trials. As with the previous behaviors, dropouts occurred because of fatigue or inconsistent performance of the target behavior. Among the ten participants unable to complete all supraglottic swallowing trials, nine completed all but one or two trials and one was unable to perform the maneuver in any trial. We found significant differences between treatment and control conditions at times 2 [t(21) = 4.24, P < .001] and 3 [t(23)= 4.41, P < .001]. No significant differences were found between the baseline flow rates as a function of condition. Within the treatment condition, we found significant differences between times 1 and 2 [t(22) = -5.78, P < .001] and times 1 and 3 [t(25) =-4.98, P < .001]. This indicates that LVR had a significant, positive effect on PCF when used in conjunction with the supraglottic swallow maneuver, and that the treatment effect was maintained for 30 minutes after treatment. Further, in the treatment condition at times 2 and 3, PCF values exceeded 270 L/min (Table 2).

## 4 | DISCUSSION

Our findings suggest that, when used in conjunction with targeted airway clearance and protection behaviors, LVR holds promise for improving independence, function, and safety during activities of daily living-critical goals for ALS management.<sup>39,40</sup> The simple, portable nature of the device, for example, allows users to take it with them on community outings or when traveling. This may instill a sense of confidence in handling symptoms of respiratory insufficiency when away from home. Increased posttreatment PCF for the three VACMs, each of which targets different areas in the upper and lower airways, may facilitate more effective management of airway secretions when LVR is used before eating, conversation, or other daily activities. Posttreatment measurements for forced expiration and throat clearing exceeded the 180-L/min PCF threshold that is considered effective for airway protection. Although posttreatment PCF for hawking remained below the minimum threshold required to move mucus from the lungs into the upper airway, the significant difference between treatment and control conditions at times 2 and 3 is clinically relevant, as hawking targets oropharynx clearance rather than clearance of the lower respiratory system. A minimum PCF threshold for oropharynx clearance is not known. Critically, PwALS usually retain pharyngeal sensation. This provides initiation cues for specific VACMs, as well as capacity to judge clearance effectiveness. In addition, spared mucociliary escalator function aids the movement of substances through the pharynx. In keeping with a strength-based treatment approach that seeks to capitalize on relatively spared functions to support more impaired functions, retained physiological abilities may aid purposive selection and implementation of VACMs.

The lasting LVR treatment effect may be clinically important. At 15 and 30 minutes after treatment, group means for both compensatory swallow maneuvers exceeded 270 L/min PCF. These flow rates are sufficient to deal with secretion encumbrance and are associated with more favorable clinical outcomes for PwALS.<sup>35</sup> Moreover, these flow rates were achieved during trial sessions in which participants may have experienced fatigue from sequentially performing the five airway tasks. In everyday life, during which individuals select a single or limited number of airway clearance or protection behaviors, LVR treatment may have an even greater positive effect on flow rates. Therefore, LVR administered before eating and used in conjunction with compensatory swallowing techniques may provide airway protection over the duration of a typical meal, thus reducing aspiration and its concomitant risks.

A central experience for PwALS is a loss of control. A small body of literature indicates that, in response, people seek and appear to benefit from control of their health care.<sup>14,16,17,41</sup> By teaching PwALS to effectively leverage LVR in conjunction with innate airway clearance behaviors and therapeutic swallowing strategies, more effective and sustained management of airway secretions and encumbrances may be achieved. This may offer PwALS a sense of control over their own care<sup>14</sup> and symptoms,<sup>13</sup> an increased sense of self-efficacy with respect to unpredictable choking, and improved adaptation to disease-related losses.<sup>14,16</sup> Although over half of those with ALS will exhibit some degree of cognitive impairment with disease progression,<sup>42</sup> LVR is simple to perform, it does not require calibration or setting changes, and it has been successfully used by PwALS who are physically capable and by caregivers. We did not formally assess cognitive function and are therefore unable to determine whether, or the degree to which, patients with cognitive dysfunction would have difficulty performing LVR. However, when individuals have PCF of less than or equal to 270 L/min, guidelines recommend training in low-tech approaches, such as LVR, to prevent acute respiratory distress and to "prepare a patient" for potential mechanical ventilation.<sup>1</sup> As a low-tech, low-cost intervention, LVR also holds potential for enhancing ALS respiratory care in low-resource contexts.

There are methodological limitations to this study. First, participants were recruited from a single multidisciplinary clinic. Although this may limit generalizability, our findings provide an empirical basis for further study of LVR across settings and for exploration of its use as a first-line therapy in low-resource areas. Second, due in part to resource limitations, each participant was seen by a single assessor, with research visits occurring two times within a 7-day period at approximately the same time of day. As a result, assessors were not blinded to the treatment vs control conditions. Finally, duration of LVR treatment before the study may have had a confounding influence on the data. All participants had received instruction in the use of LVR from a respiratory therapist and, at the time of the study, from the investigator. However, it is possible that variability in the duration of participants' LVR use and performance influenced the study results.

Further research is needed to better substantiate the treatment effect of LVR used in conjunction with volitional airway clearance and protection behaviors for PwALS, including the use of a comparison condition in which related maneuvers are used. In addition, researchers should compare PCF data with biomechanical data on swallowing function derived from more traditional methods such as videofluoroscopy and nasoendoscopy. Assessment of different but interrelated respiratory and swallowing function, and qualitative data on outcomes in everyday life situations, will build the evidence base for LVR. Finally, these findings may have implications for other neurodegenerative disorders, such as Parkinson disease and related disorders, where dysphagia occurs with retained sensory function. Research is needed to explore LVR, used in conjunction with volitional airway clearance behaviors, in other appropriate populations.

In conclusion, our findings support LVR treatment as a simple, lowcost, first-line therapy for PwALS. LVR had a positive effect on five volitional airway clearance and protection behaviors. The duration of the treatment effect may confer protective benefits for those with compromised airways and dysphagia, as well as offering some degree of autonomy and control over symptoms. Encouraging PwALS to use LVR before eating may be beneficial for those with dysphagia. With maximal expiratory airflow during volitional airway clearance and protection behaviors remaining elevated for up to 30 minutes after LVR treatment, individuals may be better able to protect their airway for at least that period, thus avoiding potentially life-threatening consequences and improving their quality of life by enhancing capacity to participate in activities of daily living.

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### CONFLICTS OF INTEREST

The authors declare no potential conflict of interest.

### ETHICAL PUBLICATION STATEMENT

We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

### DATA ACCESSIBILITY

The data that support study findings are available from the first author (S.C.) upon request.

## DATA AVAILABILITY STATEMENT

Data available on request from the authors

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#### SUPPORTING INFORMATION

Additional supporting information may be found in the online version of the article at the publisher's website.

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