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The Evaluation of Bulbar Dysfunction in Amyotrophic Lateral Sclerosis: Survey of Clinical Practice Patterns in the United States

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Abstract

Objective: Speech and Swallowing impairments are highly prevalent in individuals with amyotrophic lateral sclerosis (ALS) and contribute to reduced quality of life, malnutrition, aspiration, pneumonia and death. Established practice parameters for bulbar dysfunction in ALS do not currently exist. The aim of this study was to identify current practice patterns for the evaluation of speech and swallowing function within participating Northeast ALS clinics in the United States.

Methods & Results: A 15-item survey was emailed to all registered NEALS centers. Thirty-eight sites completed the survey. The majority (92%) offered Speech-Language Pathology, augmentative and alternative communication (71%), and dietician (92%) health care services. The ALS functional rating scale-revised and body weight represented the only parameters routinely collected in greater then 90% of responding sites. Referral for modified barium swallow study was routinely utilized in only 27% of sites and the use of percutaneous gastrostomy tubes in ALS patient care was found to vary considerably.

Conclusions: This survey reveals significant variability and inconsistency in the management of bulbar dysfunction in ALS across NEALS sites. We conclude that a great need exists for the development of bulbar practice guidelines in ALS clinical care to accurately detect and monitor bulbar dysfunction.

Keywords

amyotrophic lateral sclerosis; bulbar; practice patterns; speech; swallow; survey

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Conflicts of Interest: None.

Introduction:

Amyotrophic lateral sclerosis (ALS) is a fatal, neurodegenerative disease resulting in progressive weakness of voluntary muscles. Motor denervation of the bulbar musculature leads to progressive speech (dysarthria) and swallowing (dysphagia) impairment in ~85% of patients [1,2]. Loss of functional oral intake contributes to social isolation and diminished quality of life [3,4] and the potential loss of speech has been rated by ALS patients as the worst aspect of the disease [5]. In addition to these psychosocial sequalae, bulbar dysfunction contributes to malnutrition, dehydration, tracheal aspiration and pneumonia [2,6–8] which together account for 26% of ALS mortality [9] and increase the risk of death 7.7 fold in this patient population [6]. These factors underscore the critical need for early detection and accurate monitoring of bulbar dysfunction to ensure the optimal nutritional intake, communication abilities, pulmonary function and quality of life in individuals with ALS [10,11].

Although limited advances have been made in the management of individuals with ALS, care at specialized multidisciplinary clinics has been shown to improve quality of life, reduce hospitalization rates and extend survival [12–15] and constitutes current practice recommendations of the American Academy of Neurology [16] and European Federation of Neurological Societies [17]. While this comprehensive multidisciplinary model allows great breadth of care (with up to eight different health care consultations), it requires that individual clinicians perform abbreviated evaluations and underscores the need for quick, efficient, and accurate screening tools to detect speech and swallowing impairments in ALS [10,11].

Internationally accepted practice guidelines for the evaluation of bulbar function in ALS are currently lacking and little is known regarding the routine, clinical utilization of assessment protocols. Furthermore, referral patterns for instrumental swallowing examination or percutaneous gastrostomy (PEG) tube placement, including who within the health care team is administrating these protocols is largely unknown. As an initial step towards the establishment of best practice guidelines, we surveyed current practice patterns throughout Northeastern ALS (NEALS) centers within the United States regarding the assessment and monitoring of speech and swallowing function in patients with ALS.

Methods:

An internet-based survey was developed and administered to members of NEALS who currently work with individuals with ALS across various clinical settings including private neurology groups, hospitals, outpatient rehabilitation centers and University clinics. The final survey was prepared on Survey Monkey's online interface (www.surveymonkey.com). Email invitations with survey links were sent using Survey Monkey's Email Invitation Collector system and remained open for a thirty-day period during June of 2015. No compensation was offered to the respondents. The complete survey is provided in the Appendix and consists of 15 questions indexing respondent's clinic patient population demographics (Q1, Q2); routinely administered tests and clinical parameters (Q3); healthcare professional services offered (Q4-Q9); and referral preferences for procedures

related to nutrition and swallowing function (Q9-Q15). Survey questions included: multiple-choice (12 items), binary yes/no (3 items) and short answer (1 item) questions. The survey was designed to be low burden (i.e., short, electronic submission) to encourage respondent participation and completion of all survey items. Descriptive summary statistics were applied and included: frequency counts, mean, mode, standard deviation and range to profile current practice patterns in responding sites.

Results.

Survey Response Demographics:

Thirty-eight surveys were submitted electronically between June 1 and June 22 of 2015. Respondents included neurologists (63.1%, n=24), nurse practitioners (13.2%, n=5), and speech-language pathologists (SLP's) (10.5%, n=4). Five respondents (13.2%) reported anonymously on behalf of their ALS clinic. All respondents currently work with ALS patients across a variety of clinical settings, with most (66%) working in a university medical setting (see Table 1 for full clinical setting data).

ALS Patient Characteristics:

Table 2 presents data for the number and reported degree of bulbar involvement in ALS patients seen at clinical sites completing this survey. Twenty sites (52.6%) report seeing greater than 21 ALS patients per month (new and returning patients), with the most common response item being "11-20 patients" (32.2% of responders). Seventy-four percent (n=28) of respondents indicated that bulbar symptoms are present in 15-34% of their patients.

Routinely Collected Clinical Parameters:

Question 3 prompted clinicians to select clinical parameters routinely reported on ALS patients during their clinical visits (permitted to select as many clinical parameters from the ten available options, see Appendix, Question 3). The average number of clinical parameters reported by respondents was 5.5 (SD: 1.7, range: 1 – 8). Figure 1 summarizes the relative percentage of sites routinely reporting each of the ten bulbar-related clinical parameters surveyed. No single clinical parameter was routinely reported across all 38 sites. The ALS Functional Rating Scale Revised (ALSFRS-R) [18] represented the most commonly reported parameter with 97.4% (n=37) of clinical sites reporting its routine use. Other widely utilized clinical outcomes included body weight (94.7%, n=36), forced vital capacity (FVC; 89.5%, n=34), and body mass index (BMI; 63.2%, n=24). The least utilized clinical indices of the ten available options included: speaking rate (18.4%, n=7), the Sentence Intelligibility Test (SIT; 18.4%, n=7), maximum expiratory pressure (MEP; 28.9%, n=11), and a calorie count (36.8%, n=14).

Health Care Services Offered (Questions 4-8):

92.1% (n=35) of sites surveyed reported the provision of an SLP in their ALS clinic. Of the three sites who did not offer SLP services, two represented university clinics and one was an anonymous respondent. SLP's were reported to see all patients in 63.4% (n=23) of sites; per physician referral in 30.6% (n=11) of sites; and only in ALS patients with bulbar symptoms in 5.6% (n=2) of clinics. An Augmentative and Alternative Communication (AAC) expert

was present in 74.4% (n=28) of responding clinics, who provided AAC evaluations (71.0% of sites), communication training (22.6% of sites), and AAC therapy (6.5% of sites). A dietician was part of the healthcare team in 92.1% (n=35) of ALS clinics surveyed.

Referral Patterns for Instrumental Swallowing Evaluation (Questions 9-11):

Figure 2 summarizes response data for Question 9, "What percentage of your patients do you refer for a modified barium swallow study", and denotes the most common response as 'in less than 15% of patients' (51.4%, n=19). Overall, 73% (27/37) of clinicians reported utilizing the MBS examination in less than half (50%) of their patients (one missing data point). Reported criterion for MBS referral in those responding to Question 10 (n=32) included: in all patients with suspected dysphagia and/or weight loss (47%, n=15), in all ALS patients (6%, n=2), and in bulbar onset patients (3%, n=1) and 44% (n=14) of responding sites reported that they "do not refer for MBS in any ALS patients" in question 10. Question 11 represented a follow-up question, "What is the reason you do not perform an MBS evaluation?", yielding a total of 18 responses that are summarized below in Table 3.

Percutaneous Gastrostomy Tube Placement (Questions 12-15):

Responses to Questions 12, 14 and 15 are provided in Figure 3. Responses to question 12 ('what percentage of your patients ultimately undergo PEG placement') indicated that most respondents report '50-69% of patients,' (36.8%, n=14) with the '<15% of patients' representing the least chosen item selection (7.9%, n=3). An equal number of responses for each of the remaining options (15-34%, 35-49% and >70%) was noted (n=7 in each, Figure 3A) indicating widespread responses for this particular question. When making feeding tube recommendations; 89.5% of clinicians surveyed reported that FVC was a factor in this clinical decision, with 47% recommending PEG placement in patients whose FVC is between 30-50% predicted; 44% recommending PEG placement when FVC is >50% predicted; and only 9% making this recommendation when FVC falls below 30% predicted (see Figure 3B). PEG tube procedures were most often performed by gastroenterologists (63.9%, n=23), followed by general surgeons (16.7%, n=6). Four clinicians (11.1%) reported that they refer to an outside facility for PEG placement while 3 clinicians selected the 'unknown' response option (8.3%).

Discussion:

This survey includes a broad, demographic representation of clinical ALS research centers throughout the United States, including both university and non-university based centers. With 118 registered U.S NEALS sites, our survey included 38 centers with one designated responder allowed per site. These responders included neurologists, nurse practitioners and speech language pathologists. The questions posed were limited in both scope and content for this initial study, focusing on practice patterns for the evaluation of bulbar function in ALS, thereby attempting to avoid excessive time demands for completion and enhancing overall survey participation.

Most of these sites offered SLP, AAC, and dietary professional health care services. Routinely collected bulbar clinical parameters varied in number and type, with the ALSFRS-

R and body weight representing the only two parameters routinely collected in >90% sites. Referral for instrumental swallowing examination (MBS) was noted to be significantly underutilized, with only 27% of sites routinely utilizing this swallowing examination and 44% of clinical sites not utilizing MBS in their practice. The relative percentage or number of ALS patients undergoing a PEG tube placement varied widely across sites. Although 90% of clinicians were in consensus that forced vital capacity (FVC) was a key consideration when making PEG placement recommendations, the specific FVC thresholds used in making this decision was not consistent across sites.

Clinical Parameters Routinely Used in ALS Clinics:

Survey results highlighted consistent discrepancies involving routinely obtained clinical parameters across sites (Question 3, Figure 1). The selection of utilized clinical parameters from the available item list ranged from 1 to 8, from which no single outcome or test was routinely implemented across all (100%) of the clinical sites. The ALSFRS-R [18] represented the most commonly used outcome in 37/38 of the respondents. This finding was not surprising given that the ALSFRS-R represents one of the few validated scales for disease progression in ALS. This scale contains a ‘bulbar subscale’ with one question for each bulbar domain of speech, swallowing, and salivation. Realizing that this is a patient self-report scale with recognized limitations including known floor and ceiling effects, the ALSFRS-R may represent an insensitive tool for the identification and tracking of bulbar function over time [19]. Further, it has been noted that ALS patients tend to under-report swallowing impairment during the *early stages* of the disease due to progressive adaptation or compensation to subtle changes in swallowing function [20]. Body weight (36/38) and FVC (34/38) were two other outcome measures collected by most sites, consistent with their recognized importance as markers of disease progression. BMI was routinely tracked in only 63% of sites (n=24) and a clinical swallowing evaluation (“clinical bedside”) was reported in only 55% of sites (n=21).

Recognizing that dysphagia is reported to occur in up to 85% of ALS patients throughout the disease course, the finding that 45% of sites surveyed do not perform a basic clinical swallowing test raises the question of how dysphagia is currently being evaluated. Also of interest, objective speech testing represented the least utilized clinical parameter reported, with speaking rate and the sentence intelligibility test (SIT) routinely performed at only 18% (n=7) of sites. This may be clinically relevant given that recently published data identified speaking rate as a very sensitive marker and predictor of subsequent speech intelligibility decline [19, 21–24]. These tests have been designed to represent an easy to acquire and clinically useful outcome measure to longitudinally track ALS patients for optimal timing of augmentative communication devices before a significant reduction in communication abilities ultimately occurs. Evidence-based recommendations currently state that when speaking rate falls below 120 words per minute, referral for AAC services should be pursued, as rapid speech intelligibility decline occurs once this critical threshold has been reached [25–27].

Health Care Professionals and Services Offered:

The majority of clinical sites utilized SLP’s, dieticians and AAC experts within their clinics.

Instrumental Swallowing Examination Referral Patterns:

Seventy-three percent of sites do not routinely refer for the MBS (≤ 50% patients) and the most common response option for Question 9 indicated that 19 sites (51% of sites) recommended the instrumental swallowing evaluation in less than 15% of their patients. Furthermore, in the follow up question, 14 of 32 sites (44%) responded that they "do not refer for MBS". While the provided rationales for this clinical decision varied, several consistent themes emerged which included: 1) the belief that their clinical assessment alone was sufficient; 2) the decision to make management recommendations based on patient report of symptoms; 3) the decision to immediately refer for PEG if choking with weight loss occurs; 4) the belief that MBS does not provide useful information, treatment recommendations or patient education opportunities; 5) physicians trained to believe that the MBS is not necessary since dysphagia is expected, and the results would not influence management; 6) risk of barium aspiration during the MBS; and 7) no access to MBS testing on-site. Given these survey results, we feel that further clarification on the utility of MBS in patients with ALS is warranted.

Clinical education in this area is important as it would highlight the role of instrumental assessment versus the potential limitations of less objective forms of clinical bulbar assessments to adequately and accurately determine dysphagia status. This may also help to address the following clinical concerns facing our bulbar patients, which include: 1) swallowing efficiency and safety impairments which cannot be directly visualized on the CSE; 2) the relative high reports of 'silent' aspiration in ALS [29–30]; 3) to determine the impact of specific swallowing treatments, compensations and dietary manipulations on swallowing function; and 4) to facilitate education to patients and caregivers of the impact of recommended treatments to help ensure compliance and maintenance of safe swallowing for the patient.

Percutaneous Gastrostomy Tube Placement:

No consistent patterns were revealed for question 12, "what percentage of ALS patients ultimately undergo PEG placement", with even responses (n=7) across options of 15-34%, 35-49% and >70%. The most commonly chosen option for this question indicated that 37% of sites (n=14) report that 50-69% of patients undergo PEG placement. These data may relate to different physician and patient preferences that are currently unclear. Gastroenterologists most often performed PEG placement, and although there was great variability regarding the number of PEG's placed, FVC was a consistently important consideration when referring for PEG. This may be directly related to the practice parameter update report by Miller and colleagues (2009) noting an increased risk of adverse events when FVC drops below 50%. It is therefore not surprising that only 9% responded they would recommend a PEG if FVC dropped below 30%, but is noteworthy that 47% of respondents would recommend a PEG if FVC fell between 30-50%.

Of significant clinical interest is the potential application of these survey results with practice patterns throughout the wider international ALS community. With regard to PEG placement, our survey questioned the percentage of patients undergoing this procedure, the methods of placement and impact of FVC on this decision. Recent UK studies have focused

on the variability of gastrostomy practice patterns [32] and the large ProGas study, which assessed not only on the comparative methods of gastrostomy placement, but provided valuable clinical insight on the optimal timing of this procedure [33]. Our findings compared similarly with ProGas results in terms of GI placement of the PEG, yet the total number of patients undergoing PEG in our study differed significantly across responding sites. The primary intent of this survey was to provide initial screening information on bulbar practice patterns, without specifically focusing on the overall management of bulbar disease. Future clinical studies incorporating international collaboration to identify universally accepted evaluation and management guidelines for our bulbar ALS patients are essential, and should be actively pursued.

This reported survey of current clinical bulbar practice patterns is limited by a relatively small number of sites responding (n=38) and the brief number of questions employed. Nevertheless, these findings have helped to establish initial bulbar practice pattern findings in ALS clinics geographically located throughout the US. This survey has also assisted in recognizing several critical areas of concern, highlighting the need to establish an accepted consensus on bulbar assessment guidelines in order to advance the management and care of bulbar dysfunction in ALS.

A summary of the major points of concern raised from this survey would include: 1) a high degree of inconsistencies in clinical bulbar parameters routinely assessed in ALS clinics; 2) a preference for use of non-specific clinical tests and parameters to accurately track and monitor speech and swallowing function over time; 3) the underutilization of both the clinical swallow evaluation and referral for instrumental swallowing examination; 4) physician education on the role of the MBS in both the assessment and treatment of dysphagia in ALS; and 5) inclusion of speaking rate measurement as an aide to guide optimal timing of AAC interventions; and finally 6) the development of a validated, minimally invasive and efficient clinical assessment battery, sensitive enough to accurately track and monitor progressive bulbar decline, which could be routinely employed throughout all ALS clinics.

Survey Questions:

1. How many new and return ALS patients are seen in clinic each month?

   <10%

   11-20

   21-30

   31-40

   >40

2. What percentage of patients seen in the ALS clinic present with bulbar symptoms?

   ≤15%

15-24

25-34

35-44

45

3. Which of the following clinical parameters are routinely collected at each clinic visit (*check all that apply*).

ALSFRS-R

Weight

BMI

FVC

MIP

MEP

Speaking Rate

Sentence Intelligibility

Clinical Bedside Swallow Evaluation

Calorie Count

4. Are there Speech-Language Pathology (SLP) services provided in your ALS clinic?

Yes/No

5. How are SLP's utilized in the clinic?

See all ALS patients

Only ALS patients with bulbar symptoms

Per referral from the M.D. or other staff

Per patient/family request

6. Is there a specific Augmentative and Alternative Communication (AAC) expert in your clinic?

Yes/No

7. If yes, what services does this person provide?

AAC Evaluation

Communication Training

AAC Therapy

8. Is there a dietician routinely utilized in the ALS clinic?

Yes/No

9. What percentage of your patients do you refer for MBS?

<15%

15-34

35-49

50-69

≥70

10. What is the criterion for referral for an MBS?

Every patient

All patients with suspected dysphagia and weight loss

All those with bulbar onset

We do not refer for MBS

11. If you selected the last option above, what is the reason why you do not perform MBS evaluation?

Open Ended Answers. See Table 1 below.

12. What percentage of ALS patients ultimately undergo PEG placement?

<15%

15-34

35-49

50-69

≥70

13. By whom are PEG tubes routinely placed at your institution?

Gastroenterologist

Surgery

Outside Facility

Unknown

14. Is FVC a factor in PEG placement?

Yes/No

15. If yes, do you recommend PEG for ALS patients whose FVC:

Is >50%

Between 30-50%

Is <30%

Biographical Notes:

Emily K. Plowman: Dr. Plowman is an Associate Professor in the Departments of Speech-Language and Hearing Sciences, Neurology and Physical Therpay and Co-director of the Swallowing Systems Core Laboratory at the University of Florida. She is also the clinical director of the Center for Respiratory Research and Rehabilitation.

Lauren C. Tabor: Ms. Tabor is a Speech-Language Pathologist and current doctoral student in the University of Florida Rehabilitation Science Program. Her clinical and research interests focus on identifying efficacious interventions in individuals with ALS.

James Wymer: Dr. Wymer is a neurologist and professor practicing at The University of Florida where he specializes in the diagnosis and management of neuromuscular diseases including ALS, muscles diseases and peripheral neuropathies. He is also a Fellow of the American Association of Neuromuscular & Electrodiagnostic Medicine and an active member of the American Academy of Neurology.

Gary Pattee: Dr. Pattee is the Co-medical director at University of Nebraska MDA clinic and an active member of the Northeastern ALS consortium where he serves on the bulbar subcommittee. He is also a Fellow of the American Association of Neuromuscular & Electrodiagnostic Medicine and an active member of the American Academy of Neurology.

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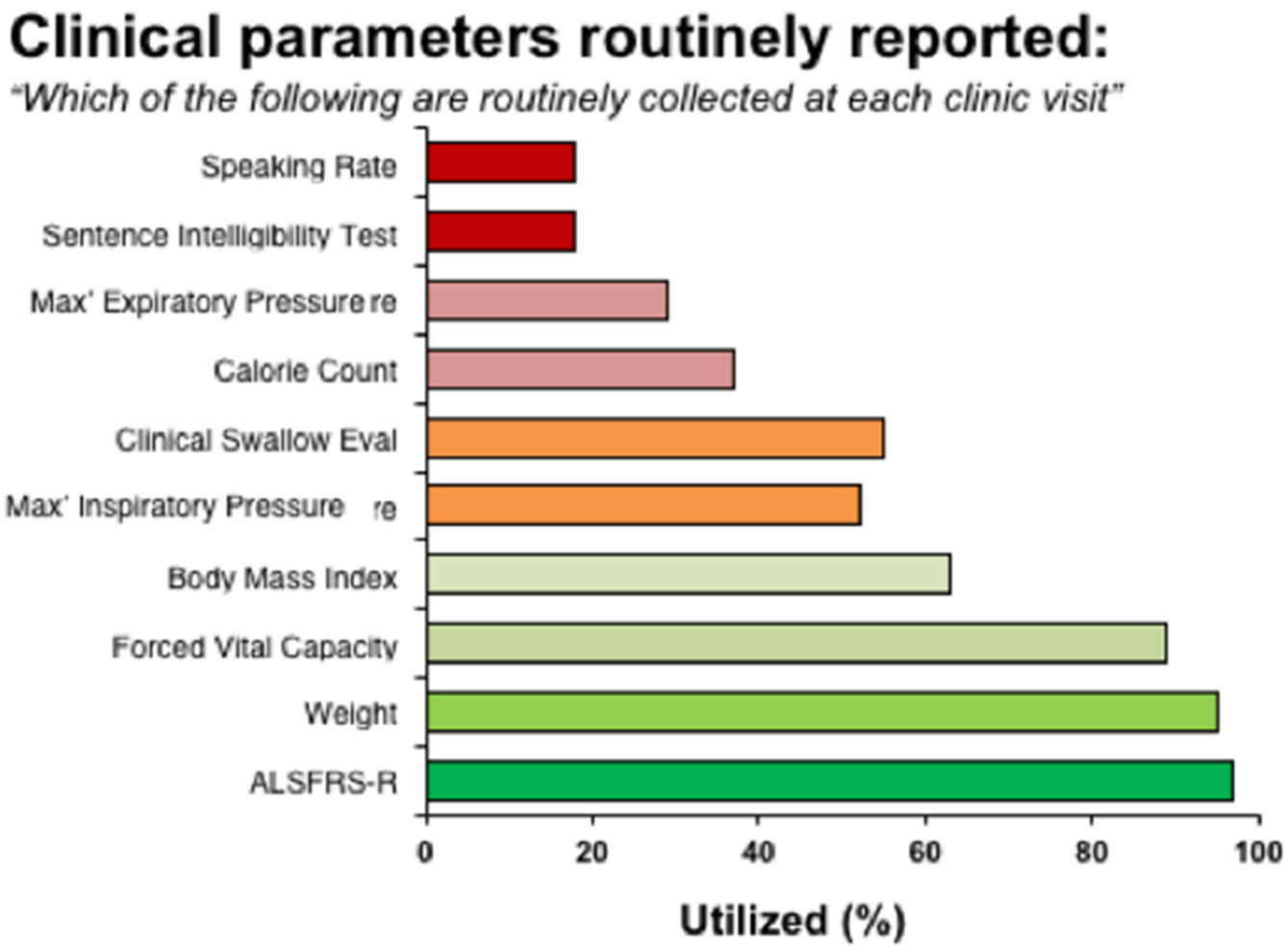

Figure 1.

Question 3 group data indexing clinical parameters routinely collected in ALS clinics. Data represent percentage of respondents who report using a specific parameter.

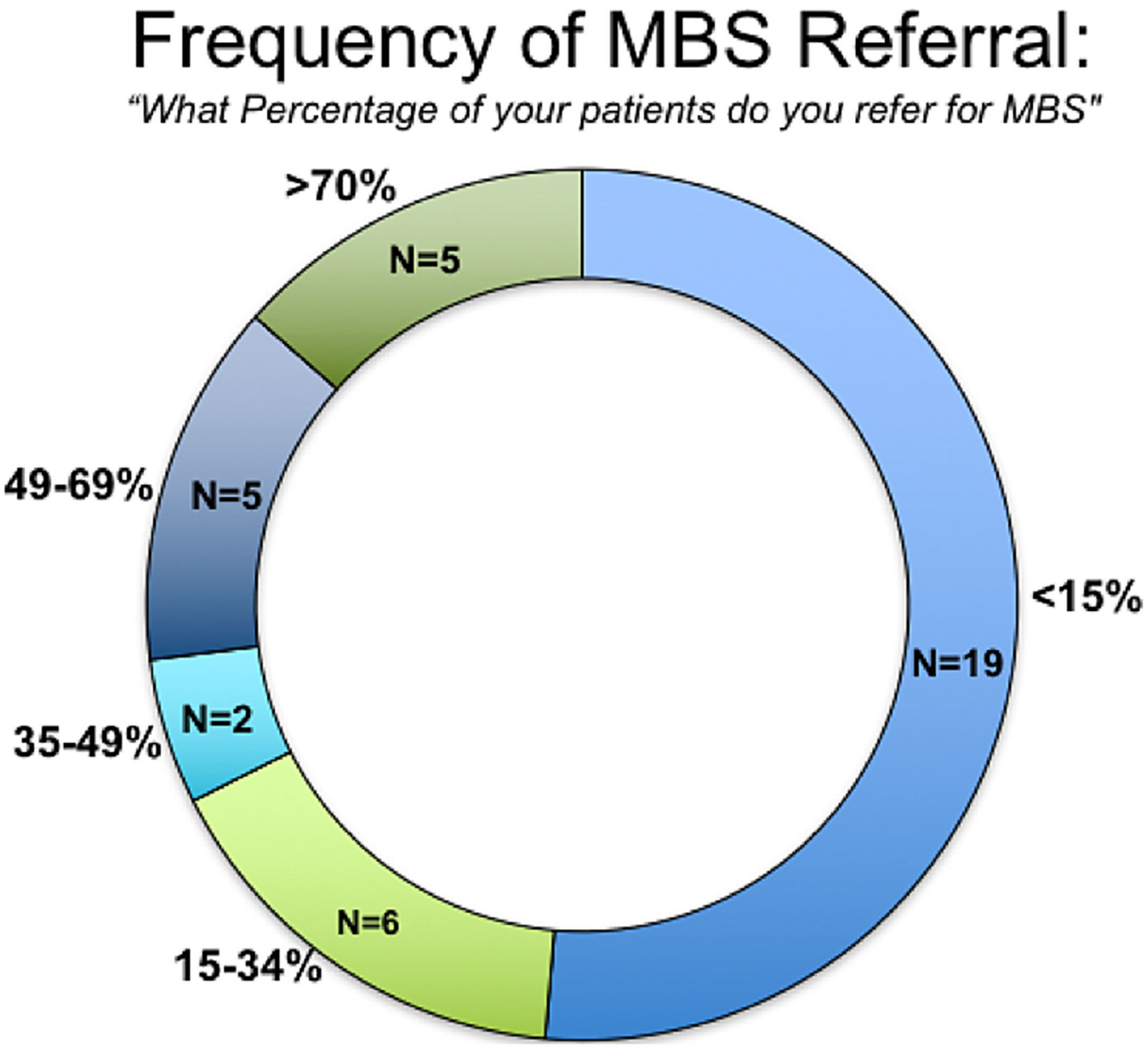

Figure 2.

Question 9 group data fot the reported percentage of ALS patients who are referred for a modified barium swallow (MBS) exam *(numbers indicate the number of responses for each choice option).*

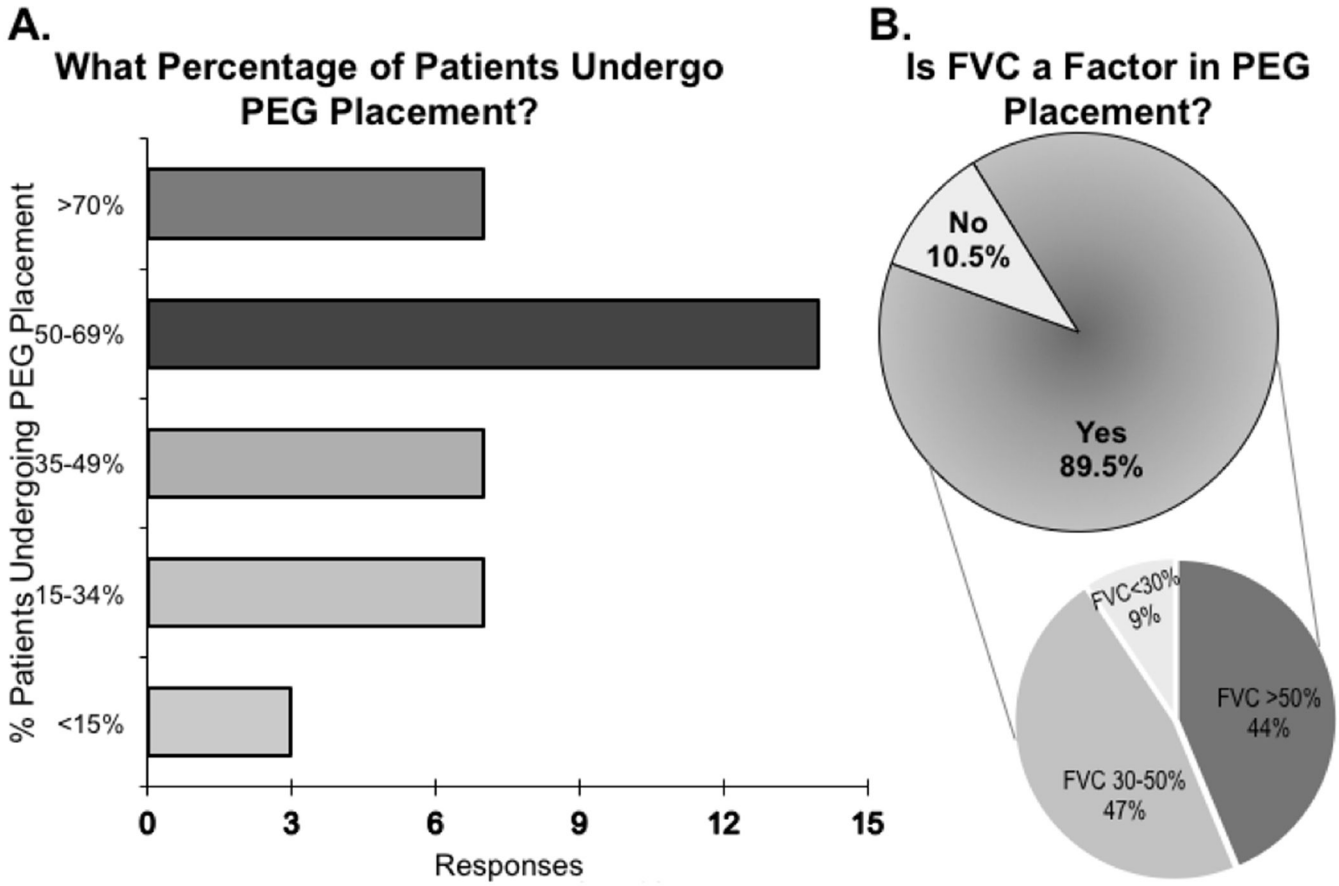

Figure 3.

Percutaneous gastrostomy (PEG) tube practice patterns regarding the frequency (expressed as % of patients seen) of placements (3A) and the influence of a patients forced vital capacity (FVC) on clinical decision making (3B)

Table 1.

Demographic information of clinical sites completing the survey.

Clinical Setting:	**% (n)**
University ALS Clinic	65.8% (25)
Private Neurology Group	7.9% (3)
Hospital	5.3% (2)
Rehabilitation Center	2.6% (1)
Veterans Affair	2.6% (1)
Unknown	2.6% (1)

Table 2.

ALS patient characteristics of responding clinical sites.

1. How many new and return ALS patients are seen each month?				
<10	**11-20**	**21-30**	**31-40**	**>40**
13.2% (n=5)	34.2% (n=13)	15.8% (n=6)	18.4% (n=7)	18.2% (n=7)
2. What percentage of patients present with bulbar symptoms?				
<15%	**15-24%**	**25-34%**	**31-40%**	**>40%**
7.9% (n=3)	44.7% (n=13)	28.9% (n=11)	10.3% (n=4)	7.9% (n=3)

Table 3.

Summary of Question 10 short answer responses (n=18).

Rationale for not performing a Modified Barium Swallowing Study
Physician preference only.
SLP and physician clinical assessments are sufficient.
MBS poses the risk for barium aspiration.
The disease progresses too quickly for MBS to be useful.
It does not add useful information.
Clients often seen for an MBS during differential diagnosis prior to being referred to our clinic.
We refer only a small number of patients who want to remain eating (and defer PEG tube placement) to enhance and educate on safe swallowing techniques.
No access to MBS on-site.
MBS does not provide education and treatment recommendations to make it worthwhile.
The SLP (seen outside of our clinic) will usually send for this.
We go by patient report of symptoms.
Taught in fellowship MBS not necessary since dysphagia is expected and results don’t change management.
If they report choking episodes associated with weight loss, we refer for a feeding tube.