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PROVISIONAL BEST PRACTICES GUIDELINES FOR THE EVALUATION OF BULBAR DYSFUNCTION IN ALS

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Abstract

Introduction: Universally established comprehensive clinical bulbar scales objectively assessing disease progression in ALS are currently lacking. The goal of this working group project is to design a best practice set of provisional bulbar ALS guidelines, available for immediate implementation within all ALS clinics.

Methods: ALS specialists across multiple related disciplines participated in a series of clinical bulbar symposia, intending to identify and summarize the currently accepted best practices for the assessment and management of bulbar dysfunction in ALS

Results: Summary group recommendations for individual speech, Augmentative and Alternative Communication (AAC) and swallowing sections were achieved, focusing on the optimal proposed level of care within each domain.

Discussion: We have identified specific clinical recommendations for each of the 3 domains of bulbar functioning, available for incorporation within all ALS clinics. Future directions will be to

establish a formal set of bulbar guidelines through a methodological and evidence-based approach.

Keywords: *Bulbar, Speech, Swallowing, Guidelines, AAC*

Introduction

Bulbar dysfunction in ALS significantly contributes to reductions in quality of life, social withdrawal, malnutrition, dehydration, aspiration pneumonia, and early mortality. A globally recognized working metric, incorporating reliable clinical assessment scales to monitor bulbar disease progression in ALS, has yet to be achieved. Previous attempts aimed at achieving this metric have fallen short of a satisfactory and comprehensive protocol [1,2,3,4,5]. The Northeast ALS (NEALS) bulbar subcommittee has recently completed a bulbar practice survey regarding current practice patterns within participating sites, which identified significant inconsistencies involving the assessment and management of bulbar dysfunction in ALS [6]. These survey results revealed an urgent need to design and incorporate a best practice set of provisional guidelines, intended to comprehensively assess and monitor bulbar dysfunction across clinical sites. In an attempt to address this need, the NEALS bulbar subcommittee recently convened to generate a

provisional working group set of best practice guidelines, as established through expert standard of care consensus. Our expected goals are to: 1) Standardize bulbar data collection across all sites, which will assist in the acquisition of patient data; 2) Develop guidelines for the timing of referrals and follow-up evaluations of speech, Augmentative and Alternative Communication (AAC), and swallowing; and 3) Propose common data elements for standardized data collection of speech, AAC, and swallowing assessments. The temporal feasibility of implementing these guidelines within the clinic setting was a critical concern, emphasizing the required time and ease of administration, given the amount of clinical testing ALS patients routinely undergo from the multiple health related disciplines during each clinic visit. A pragmatic approach was therefore underscored.

Methods

Several designated ALS bulbar symposia recently convened to establish consensus summary recommendations for the assessment of bulbar dysfunction, specifically related to the domains of speech, AAC and swallowing. In attendance were ALS specialists across multiple disciplines, including clinical neurology, biostatistics, and speech-language pathology (SLP) representing the United States, Canada, and Italy, thereby providing an expansive range of bulbar expertise from an experienced clinical perspective. This consensus group worked to establish provisional guidelines, focusing on speech and swallowing impairments. The focus of the speech group was to (1) provide guidance concerning the timing of patient referrals for a speech and AAC evaluation, and (2) propose common data elements of a clinical speech exam that are feasible to

administer in a busy ALS clinical setting. The working goal of the swallowing group was to develop a pragmatic, triage-based clinical pathway to guide screening, assessment, and management of dysphagia in patients with ALS. These symposium sessions were organized into separate speech, AAC and swallowing sections, resulting in working group summary guidelines within each section that ultimately underwent a comprehensive review by the entire committee.

Results

Speech Section: Clinical Speech Evaluation

The speech section focused on the establishment of important supplementary metrics of related systems and functions, with subsequent system and subsystem analysis: Forced Vital Capacity/Slow Vital Capacity (FVC/SVC), ALS Functional Rating Scale Revised (ALSFRS-R), Pseudobulbar Affect (PBA), dysphagia, medications, and cognition.

Guidance regarding Speech referrals: The speech section recommended SLP speech evaluations at the initial clinic visit and recommended that all clinic visits should gather routine clinical information as listed in Figure 1. Follow-up speech assessments are suggested as an integral component of each ALS clinic visit, though frequency and visit duration may vary depending on multiple factors including patient needs, SLP availability, physician recommendations, and clinic resources. All patients who present with an oral motor exam atypical for bulbar ALS (ie. asymmetrical tongue or pharyngeal weakness, oral/throat pain, raspy voice or swallow

difficulties preceding speech impairment) should undergo an otolaryngology evaluation to rule out alternative causes.

Common data elements of clinical speech examination: The common elements of the speech examination are listed in Figure 1. The Center for Neurologic Study-Bulbar Function Scale (CNS-BFS), a recently validated patient reported outcome measure[7] that has demonstrated clinical utility[8], may also include a family member or caregiver for guidance to more completely assess the communication status of the patient. The speech assessment should include a spontaneous speech sample and the reading of a short paragraph, such as the Bamboo[9] or Rainbow Passage[10] (only for fluent English speakers and readers; in non-English speakers/readers another short paragraph should be selected based upon the patient's native language). These samples will allow clinician-based ratings of dysarthria severity (0=normal, 4=severe), speaking rate (words per minute), and involvement of one or more of the four speech subsystems (respiratory, phonatory, articulatory, and resonatory). Clinician based ratings of speech should be supplemented with objective testing of the involved individual speech subsystems (Figure 2). The estimated time necessary to perform the entire speech assessment is 8 to10 minutes.

AAC Section: Clinical Communication Evaluation

The AAC section defined the scope of augmentative communication options, identifying the optimal timing of an evaluation and stressed a proactive approach. It also focused upon

minimizing the patient energy expenditure while maximizing their collaborative engagement. Guidance on when to refer for speech and AAC evaluations, and frequency of follow-up, was established. Understanding that speech can rapidly deteriorate, patient education with AAC exposure and training is critical during the early stages of disease progression, even prior to the onset of overt bulbar symptoms. The AAC evaluation should therefore be recommended at the time of diagnosis, regardless of whether speech impairment exists. The initial screening conducted in the clinic can introduce the AAC concepts to patients and families early in the disease course, with the implementation of technology options at a later date, as appropriate (Figure 3). Patient and family counseling regarding the broader definition of augmentative communication should also be initiated early, emphasizing the focus upon broad support of communication through speaking, writing, texting and/or computer interfacing in the event of ineffective speech.

Guidance regarding AAC referrals: The AAC section agreed that the initial clinic visit was identified as the time to arrange referral to an SLP for an AAC evaluation, which is intended to be an ongoing, dynamic collaboration between the patient, physician, and SLP, and not limited to specific technology selection. The purpose of each evaluation is to identify and implement strategies to preserve communication, and to compensate for reduced ability to communicate effectively.

Swallowing Section: Clinical Swallow Evaluation

The group initially identified existing barriers or challenges within ALS centers, including: 1) timing of referral to speech-language pathology services, 2) inconsistencies in the assessment and management practices for dysphagia, and 3) specific timing of subsequent swallow interventions. It was recommended that the SLP clinical swallowing screen should include objective testing in each of the following five domains: 1) Patient-reported outcomes; 2) Dietary intake; 3) Pulmonary function and airway defense physiologic capacity; 4) Bulbar function; and 5) A dysphagia/aspiration screen.

Guidance regarding Swallowing referrals: SLP clinical swallow referrals should be established early in the disease course. Table 1 provides an overview of the importance in gathering clinical information across each of the above domains, as well as the supporting evidence within the ALS population for specific assessment tools. Objective measures allow for documentation of disease progression on swallowing, including dysphagia-related symptoms, impact on quality-of-life and dietary intake. The estimated time and cost of each of these proposed assessments is as follows: the EAT-10 is free of charge and can be administered to the patient while in the waiting room, the IOPI takes four minutes to perform, the CPF takes roughly one minute with a device cost of approximately $45, and the Yale Swallow Protocol is free of charge, requiring one minute to complete. Objective measures of voluntary cough airflow have been noted to demonstrate significant discriminant ability to detect ALS patients at risk for penetration/aspiration[11], and

have been suggested as a useful index of airway defense physiologic capacity for inclusion in the clinical swallowing evaluation by the SLP [11,14].

Common data elements of clinical swallowing exam:

The distinction between a swallowing screen and a swallowing assessment was emphasized. The screen should incorporate a pragmatic approach in managing a triage flow of care within a busy ALS clinical setting. Specifically, it was recommended that all ALS patients undergo a swallowing (dysphagia/aspiration) screen, and those patients presenting with markers of dysfunction (e.g., failed screening measure) be routed for a comprehensive swallowing evaluation, which may also include an instrumental assessment of swallowing function (e.g., videofluoroscopic swallow study – VFSS). Although the panel of experts in this section felt that an instrumental swallowing exam was important for patients demonstrating a high risk for aspiration and dysphagia, recent survey data reveal that only 27% of sites are routinely administering a VFSS[6]. Given the established high rate of silent aspiration in ALS (i.e., no attempt to expectorate aspirated material)[13], the panel agreed that direct visualization of swallowing function (safety and efficiency) and evaluation of the effectiveness of trialed swallowing maneuvers or strategies will most reliably be assessed through the use of instrumental techniques in this patient population. An open dialogue concerning the relative role, timing, and utility of the VFSS in the evaluation and monitoring of swallowing function in individuals with ALS should continue to evolve, with consideration given to the availability of

specialized and highly trained SLPs in the ALS multidisciplinary clinic setting, in whose absence the VFSS may become an essential tool to accurately determine swallowing risk.

The inclusion of patient and caregiver education was especially stressed to address: 1) the role of feeding tubes for supplemental nutrition and hydration, 2) the importance of proper oral hygiene, emphasizing the association between poor oral hygiene and aspiration pneumonia in dysphagia, 3) compensatory swallow strategies and maneuvers, 4) dietary modifications including food texture and consistency, and 5) pulmonary hygiene with airway clearance and basic life saving techniques.

To summarize, if the patient fails a swallow screen or presents with other signs or symptoms of bulbar dysfunction (e.g., patient reported difficulties or reduced pulmonary clearance), a referral for a thorough swallowing assessment is warranted.

If an SLP is not present in the clinic or does not routinely see all scheduled clinic patients, the neurologist should initially examine each patient, focusing on the following items: the presence of swallowing difficulties, how food is being prepared, assistance with feeding, unintentional weight loss, coughing or choking with meals, length of time needed to eat, saliva management, speaking rate and slurring of speech. Should the clinical exam identify bulbar dysfunction, i.e., findings reveal tongue weakness, dysarthria with weak cough (peak cough flow <240L/min) and/or compromised respiratory support (FVC<60% predicted), then an SLP referral is warranted for further comprehensive assessment. The addition of a set of minimally invasive, quick and sensitive (high yield) objective swallowing metrics were recommended, as presented

in Table 1. This highlights the NEALS Bulbar recommendations for consideration of five important domains related to swallowing function, and the current suggestions for objective, validated tests with the inclusion of associated outcomes.

Discussion

The primary intent of this project was to develop and establish the currently recognized and optimal bulbar ALS practice parameters, representing an expert-based standard of care for immediate implementation within the ALS clinical setting. Ultimately, through the application of methodological techniques incorporating available systematic online reviews of each of the summary recommendations, a formal set of bulbar ALS guidelines will be established. These formal guidelines would subsequently require the inclusion of patient and caregiver participation, and undergo future modifications as new evidence-based, clinical bulbar assessment strategies and applications are identified.

A recognized limitation of this project is the lack of existing evidence-based support for the currently proposed bulbar measures—a critical direction for future research. The inclusion of cognitive and behavioral assessment scales are also omitted due to the time constraints within a busy ALS clinic, and the lack of a universally accepted and validated cognitive scale which adequately assesses patients with bulbar dysfunction. The time required to complete these proposed bulbar assessments may also represent a limitation, yet the estimated combined time for completion of speech and swallowing assessments should be less than 20 minutes. The

limited existing and validated objective swallow measures identified to date highlights the critical need to pursue ongoing clinical research, focusing on the assessment of bulbar dysfunction as it relates to swallowing dynamics and its impact on clinical management.

In conclusion, the purpose of this working group project is to create a best practice set of bulbar assessment guidelines, designed for clinical implementation throughout the broader ALS community. This goal should most effectively be accomplished through the establishment of a productive collaboration between international ALS researchers, clinicians, patients, and caregivers, specifically focusing on motor speech, swallow, and communication assessment. This proposed ongoing 'bulbar dialogue' should hasten the creation of an evidence-based and validated set of formal bulbar ALS guidelines, attainable within the foreseeable future.

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Declaration of Interest

The authors report no financial declarations of interest with this paper.

Abbreviations

AAC, Alternative and Augmentative Communication; ALS, amyotrophic lateral sclerosis; ALS-FRS-R, ALS functional rating scale-revised; CNS-BFS, Center for Neurologic Study Bulbar Function Scale; FTD, Frontotemporal dementia; FVC, Forced vital capacity; NEALS, Northeast ALS consortium; PBA, Pseudobulbar affect; SLP, Speech-language pathologist; SVC, Slow vital capacity; VFSS, Videofluoroscopic swallow study.

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Tables and Figures

Figure 1: Speech Evaluation—The common data elements suggested for inclusion in all clinical evaluations.

ALSFRS-R—ALS Functional Rating Scale-Revised

CNS-BFS—Center for Neurological Study-Bulbar Function Scale

Figure 2: Bulbar Case History Information—Speech Evaluation.

CNS-BFS—Center for Neurological Study-Bulbar Function Scale

Figure 3: Referral Goals for AAC Evaluation. This triage tool should provide guidance to the speech language pathologist in the ALS clinic as to the immediate needs to be addressed in an AAC assessment for a person with ALS. It may serve as the basis for defining the referral content.