Dysphagia 11:129–139 (1996)

Dysphagia



# Management of Oral-Pharyngeal Dysphagia Symptoms in Amyotrophic Lateral Sclerosis

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**Abstract.** Oral and pharyngeal dysphagia is a common symptom in patients with amyotrophic lateral sclerosis (ALS) and is the result of a progressive loss of function in bulbar and respiratory muscles. Clinicians involved in the management of ALS patients should be familiar with the common clinical findings and the usual patterns of temporal progression. The prevention of secondary complications, such as nutritional deficiency and dehydration that compound the deteriorating effects of the disease, requires careful monitoring of each patient's functional status and timely intervention with appropriate management techniques.

**Key words:** Amyotrophic lateral sclerosis (ALS) — ALS management — ALS Severity Scale — Dysphagia management — Dysphagia and respiration — Deglutition — Deglutition disorders.

Amyotrophic lateral sclerosis (ALS) is a progressive neuromuscular disease involving the degeneration of motor neurons in the cortex, brainstem, and the spinal cord. Onset of the disease is characterized by varying symptomatology, depending on the site of motor neuron involvement. The disease may affect bulbar and/or spinal systems, and symptoms may range from dysarthria and dysphagia to weakness of grasp or stumbling. The patient may exhibit paralysis and muscle atrophy, bulbar disturbances, or pyramidal tract syndromes involving exaggerated reflexes [1]. As the disease progresses, both upper and lower motor neuron signs may be evident, and symptoms may overlap. Upper motor neuron signs (UMN) include muscle weakness, increased muscle tone (spasticity,) hyperreflexia, and pseudobulbar palsy. Lower motor neuron (LMN) symptoms include decreased muscle tone (flaccidity), muscle weakness, muscle atrophy, and diminished or absent stretch reflexes.

The rate of symptom progression in ALS is extremely variable. The disease is often described as relentlessly progressive, with death occurring by 3 years of age in 50% of cases [2,3]. In some cases patients will stabilize or gradually progress over periods of more than 15 years [4,5]. Bulbar involvement, which affects speech, swallowing, and voice production is the primary initial symptom in about 25%–30% of patients [2,3,6]. (Bulbar symptoms refer to involvement of motor neurons in the brainstem which innervate the muscles of the face, tongue, pharynx, and larynx.) The rate of progression of bulbar symptoms varies, but the sequence of appearance of these symptoms is somewhat predictable. For example, the tongue is usually involved before the lips or jaw. Some research has been done in examining the bulbar affects of this disease. Investigators have examined the nature of the dysarthria accompanying ALS [7–15], as well as the changes in phonation that accompany the disease [16,17]. Although one of the most distressing bulbar symptoms is progressive dysphagia, little research has been done in examining the progression of swallowing problems for ALS patients.

As is the case in most neuromuscular diseases, the onset of dysphagia is insidious. Early symptoms may include complaints of "postnasal drip," occasional choking while drinking liquids, or even intermittent choking on saliva. As symptoms progress, weight loss and even cachexia may be found. Progressive weakness and atro-

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phy of the diaphragm and accessory muscles of respiration contribute to the patient's swallowing difficulty and account for increased susceptibility to pneumonia and respiratory failure that most often is the proximate cause of death in ALS [18–20]. In order to manage the symptoms of dysphagia in patients with ALS one must be able to predict the progression of symptoms and relate these to the clinical findings.

This paper reports data summarizing the progression of swallowing deficits associated with ALS. Two specific questions are addressed:

1. How rapidly and to what degree do dysphagia symptoms progress during disease progression in comparison to speech symptoms; in men vs. women; and in patients with initial spinal vs. bulbar vs. mixed symptoms?
2. What is the relationship of vital capacity to symptoms of dysphagia?

Clinical implications relative to the timing of steps of intervention will be discussed. This paper was motivated by the fact that data regarding symptom progression are important to clinicians' understanding of the probable course of the dysphagia, allowing them to make good, timely decisions about intervention. There is no proven therapy that affects the course of ALS, but symptomatic management of dysphagia can help to alleviate much of the misery that ALS patients experience. Problems related to malnutrition and dehydration result in secondary complications that exacerbate the effects of the disease. The data from this paper provide information important to decisions about when (and for whom) intervention should be initiated regarding dietary changes, postural and other behavioral changes, and placement of enteral feeding alternatives.

## Methods

### *Patients*

The data and the experience described in this paper were collected over an 80-month period, from January 1987 through August 1993. During that time, 140 patients (74 men and 66 women) with a confirmed diagnosis of ALS were seen in the Neuromuscular Clinic for Swallowing and Speech Disorders (NCSSD) at the University of Washington Medical Center. Patients ranged in age from 23 to 80 years with a mean age of 59.8 years and a standard deviation (SD) of 11.3 years. The mean months postdiagnosis of the first visit was 11.8 months (SD 11.4) and 49% of our patients were seen within 6 months of the diagnosis. All patients were seen as outpatients. Forty-two of the total group of patients were seen longitudinally, with time between clinic visits varying from 6 weeks to 3 months, depending on the needs of the particular individual. Each patient in this group of 42 was seen for at least three clinic visits.

### *Clinical Procedures*

Patients were seen in clinical rooms of the Otolaryngology Service at the NCSSD, *Clinic for Swallowing and Speech Disorders*. Medical history (or medical update if the patient was on a return visit), symptom description, and dietary information were collected via interview with both the patient and accompanying family or caregivers. At each visit, neurologic examinations were conducted by a neurology nurse specialist. Examination of the structure and function of the oral articulatory mechanism were completed by at least two speech pathologists. Vital capacity was measured with a hand-held Write respirometer. After ensuring good lip seal and occlusion of nares, the patient was asked to "breathe in as deeply as possible, and blow air into the respirometer until all the air is gone." Each patient was also rated on the ALS Severity Scale (ALSSS) at each visit [21]. This scale determines a functional level of severity for speech, swallowing, and upper and lower extremities (see appendix for complete description of the severity scale). A series of questions are asked about functional skill for speech, swallowing, use of arms and hands, and use of the legs. A score between 1 and 10 is assigned for each of the four categories.

Procedures for determining a rating for swallow function included (1) a clinical swallow examination, and (2) specific questions asked of the patient, and the accompanying family member or caregiver. Procedures for the swallow examination included completion of medical history and routine procedures for the clinical evaluation of swallowing [22,23].

Videofluoroscopy was completed for those patients whose subjective complaints or symptoms did not fit the routine findings for ALS patients or the predicted clinical findings of the individual. For example, individuals with complaints of solid food dysphagia greater than liquids, the sensation of obstruction, or symptoms consistent with esophageal dysfunction were studied fluoroscopically.

### *Data Analysis*

A portion of the analysis of the data for this paper was examined from the total pool of 140 patients. All data examining change over time, however, is taken from the 42 patients who have made at least three visits to the Neuromuscular Clinic for Swallowing and Speech Disorders for repeated follow-up of swallow and speech function. (In our speech and swallowing clinic, patients are seen every 1-3 months, depending on rate of progression.) Data from patient reports, clinical examinations, ALSSS scores, and vital capacity scores were entered into a database from which descriptive statistics and graphing could be completed.

## Results

The questions addressed here relate to both progression of the disease and the relationship of vital capacity to dysphagia symptoms. It is important, therefore, to identify the relative degree of severity in the ALS sample studied. There is considerable variability in the range of severity among patients with ALS, even at the time of their first visit to the Clinic. Figure 1 illustrates the speech scaled score plotted against the swallowing scaled score at the time of the patient's initial visit. Note that 76% of patients have speech and swallowing scores that fall within plus or minus one ALSS score [21]. These data indicate if a patient is exhibiting progression in speech symptoms,

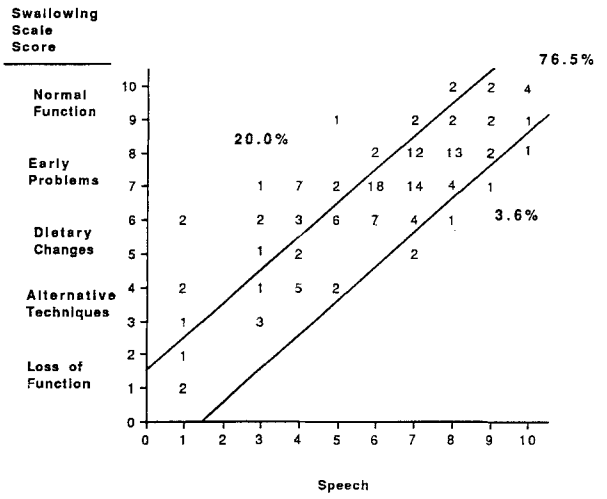


**Fig. 1.** Speech scaled scores plotted against the swallowing scaled scores for all 140 patients at the time of patients' initial visit.

there is a good chance that swallowing is also getting worse. For 20% of patient visits, the speech scaled score was 2 or more scaled scores lower than the swallowing score, and only 4% exhibited swallowing scores that were lower than speech. Not until the speech scale scores reach "6," where slow rate and reduced intelligibility are the major problems, does the functional rating for swallow begin to drop at or below the speech score.

### *Progression of Dysphagia Symptoms*

Figure 2 plots the initial and final swallowing scaled score for 42 individual ALS patients who completed at least three clinical evaluations at the speech and swallowing clinic. Figure 2A and B depict the female and male patients, respectively, who exhibited initial bulbar signs (bulbar signs refer to motor neurons in the brainstem which innervate muscles of the face, tongue, pharynx, and larynx.) Figure 2C and D depict those female and male patients with early spinal symptoms (spinal symptoms refer to those motor neurons in the spinal cord that innervate the arms, legs, and trunk.) Sixteen of 21 bulbar patients had declining swallowing scale scores by at least three levels during the time they attended our clinic. Of the spinal patients, 8 of 21 showed declining swallowing scaled scores by three or more levels. Thirteen of the 42 patients exhibited swallowing scaled scores that stayed the same or fell by only one level.

Because differences in gender have been noted with respect to ALS dysarthria [12–14], gender differences in dysphagia were examined. Although qualitative differences in swallowing performance between men and women with ALS have not been reported, differences according to gender are evident in our sample with respect to both the onset and timing of progression of dysphagia symptoms as the disease progresses. In our sample of 21 patients who exhibited initial bulbar signs, more women exhibited initial bulbar symptoms than did men (see Fig. 2A and B) and women exhibited earlier and more severe dysphagia symptoms. More men presented with initial spinal symptoms (see Fig. 2C and D), and men generally had later onset of dysphagia than women with initial spinal signs.

Figure 3 represents the initial and most recent swallowing scaled score for the 42 ALS patients followed longitudinally. Median data are plotted for six groups: men and women with initial bulbar signs; men and women with initial spinal signs; and men and women with initial mixed signs. The women in our sample were generally seen earlier than the men, indicating that women may exhibit earlier swallowing problems than men. For those patients with early bulbar signs, the women also tend to exhibit progression of dysphagia symptoms more rapidly. Among those patients with early spinal signs, women again show a slightly greater slope of dysphagia progression. Because these are ordinal scores, we have expressed them in medians rather than means.

### *Relationship of Vital Capacity to Dysphagia*

In order to examine the relationship of vital capacity measures to dysphagia scaled scores, we return to the large group data. There is a corresponding progression of decreasing vital capacity and increasing dysphagia that is seen in many patients. Because disorders of swallowing and respiration may complicate one another, often one can anticipate functional swallowing difficulties by monitoring pulmonary function. Figure 4 plots the average swallowing scaled score (for spinal, bulbar, and mixed groups) with the vital capacity measure for all 140 individuals over every visit. These data indicate that it does not matter whether the initial signs were bulbar, spinal, or mixed. As respiratory capacity declines, swallowing declines. Note that at about 1½ L, the swallowing scaled scores are generally quite low. However, some patients do exhibit low vital capacity even when swallowing is not yet presenting a serious problem.

In our clinic we categorize respiratory status into three levels. Generally, we find that individuals with vital capacity over 1.5 L to be asymptomatic and therefore categorize them as having adequate respiratory function. We place individuals with vital capacities between 1 and 1.5 L into a marginal category, especially if surgical intervention is being considered. Finally, individuals with vital capacities of less than 1 L are categorized as having insufficient respiratory support, and usually consider them to be poor candidates for surgical intervention. This is primarily because, even though the procedure is done under local anesthesia, many patients with poor respira-

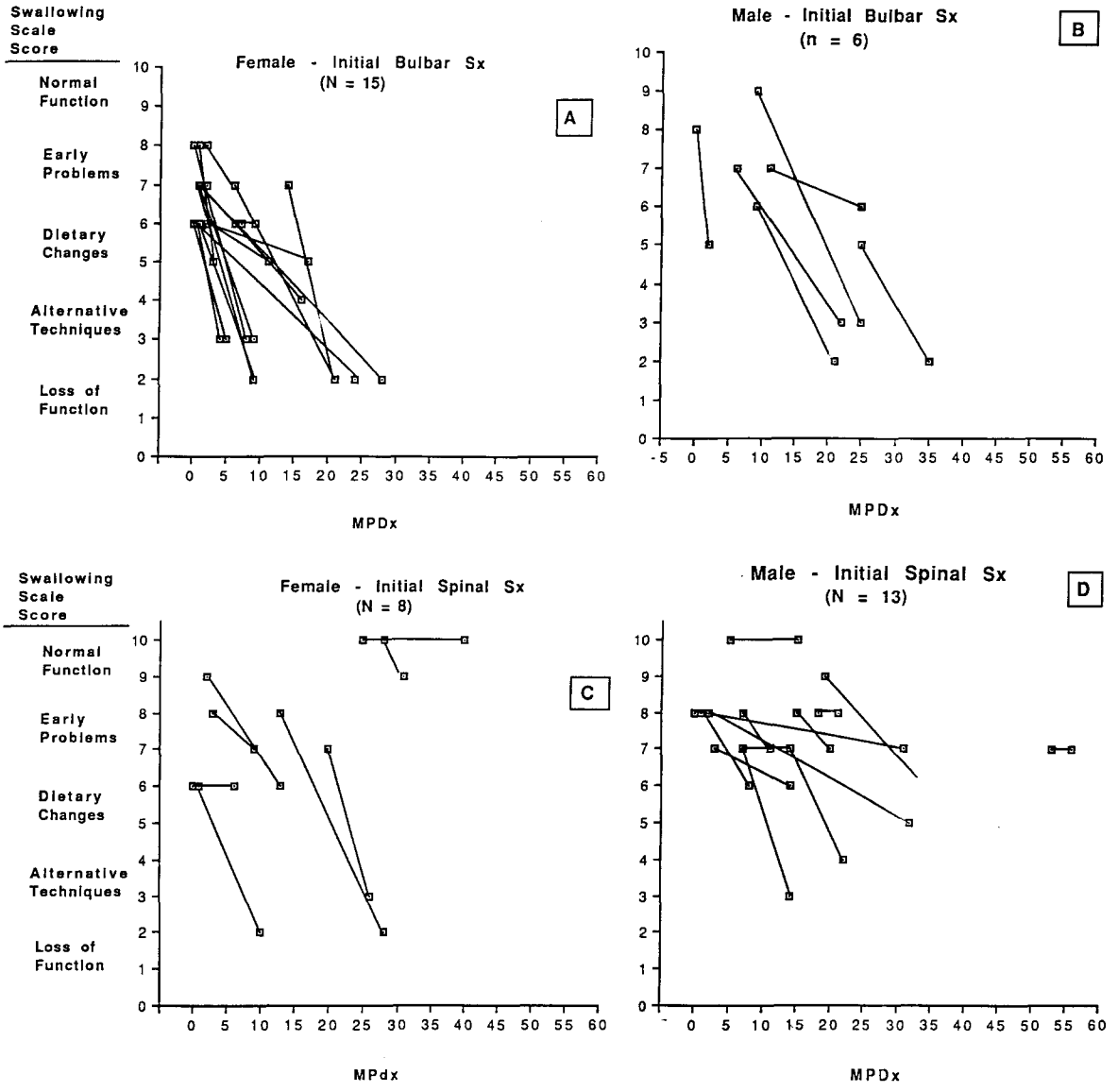


**Fig. 2.** Initial and final swallowing scaled scores for 15 women with ALS who exhibited initial bulbar symptoms **(A)**; 6 men with ALS who exhibited initial bulbar symptoms **(B)**; 8 women with ALS who exhibited initial spinal symptoms **(C)**; and 13 males with ALS who exhibited initial spinal symptoms **(D)**.

tory support have difficulty staying prone for the half-hour that the procedure requires.

Figure 5 represents the percentage of the 140 patients over all visits, who exhibit adequate respiratory function, marginal respiratory function, and insufficient respiratory function at each level of swallowing function. In general, patients have increased difficulty with swallowing as respiration declines. Note, however, that there are a number of people who exhibit marginal vital capacity before they exhibit a swallowing scaled score of 4. For some patients, respiration becomes problematic and may impede swallowing before functional swallowing capability precludes oral feeding.

## Discussion

The data reported in this paper address progression of dysphagia symptoms. We discuss specific clinical issues related directly to the data then focus on clinical implications, especially those related to timing of intervention.

### *Progression of Dysphagia Symptoms*

In our sample of 140 ALS patients, 76% exhibited speech and swallowing scores that were within one scaled score of each other. It appears, then, that progression of swallowing symptoms follow a rate similar to that of speech

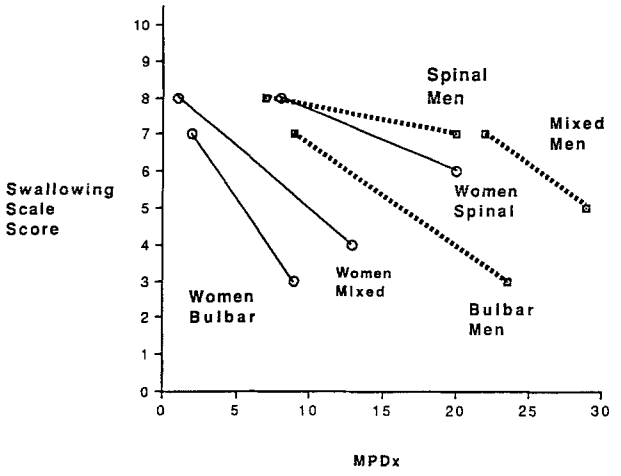


**Fig. 3.** Median swallowing scaled score for the initial and most recent clinic visit for 42 ALS patients who completed at least three clinical evaluations. The data are plotted for six groups; men and women with initial bulbar signs; men and women with initial spinal signs; and men and women with initial mixed signs.

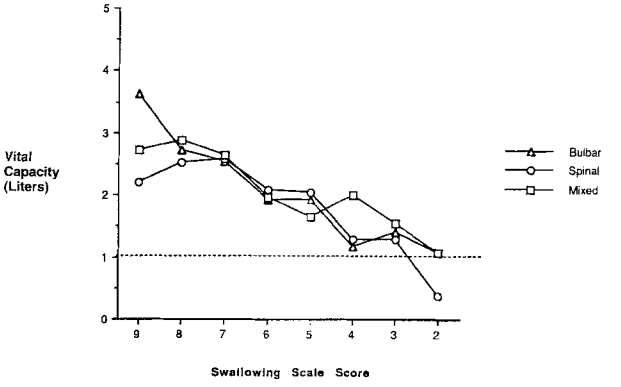


**Fig. 4.** Average swallowing scaled score for spinal, bulbar, and mixed groups plotted against mean vital capacity measures for all 140 individuals over every visit.

and that functional disability resulting from bulbar deficits will affect speech and swallowing to a similar degree. This is important because some patients will report little difficulty with swallowing, even though speech is obviously quite impaired. These data indicate that when intelligibility begins to be compromised, swallowing deficits are likely to follow and may progress rapidly. It is important to note that one cannot rely on interview data alone when assigning the swallowing scaled score, as patients may often not report phenomenon that may be dangerous (i.e., frequency of choking episodes) or they may feel there is less problem than there really is. Family report is helpful here, as is careful repeated measures of weight. Though some weight loss is inevitable due to muscle atrophy, consistent or dramatic loss is indicative of probable chewing and/or swallowing deficits.

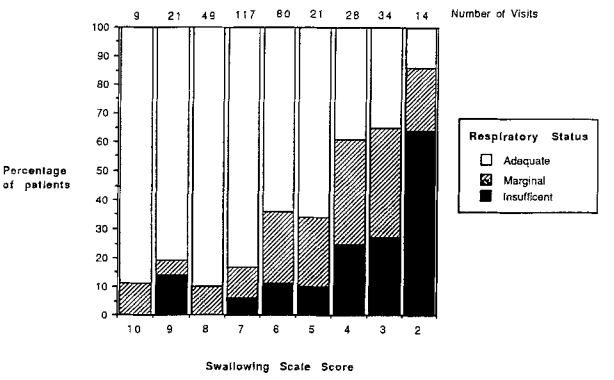


**Fig. 5.** Percentage of patients who exhibit adequate, marginal, and insufficient respiratory function at each level of swallowing function.

Much intersubject variability in rate and overall degree of progression in dysphagia was evident in our data. For example, 13% of the 42 patients followed over time exhibited functional swallowing ability that declined very little over the course of their disease; conversely, 24% declined three or more levels. This variability among patients points out that clinicians may need to provide dysphagia education and intervention to different patients at different points in time during the course of their disease.

### *Relationship of Vital Capacity to Dysphagia*

Impaired respiratory function is a progressive feature throughout the course of ALS. It is a finding that is present in bulbar, spinal, and mixed forms of the disease, and is directly related to timing of some aspects of dysphagia management, such as gastrostomy or enteral feeding. Our data indicate that functional swallowing abilities (as measured by the ALSSS) decline as respiratory capacity declines, regardless of whether the initial signs were bulbar, spinal, or mixed. By the time swallowing problems are noted in patients who had initial spinal symptoms, it is probable that the patient will have respiratory problems. The swallowing problems for these spinal ALS patients may have more to do with respiratory muscle deficits than oral agility or pharyngeal contraction problems.

Monitoring of respiratory capacity is important to intervention for dysphagia even for those patients who are not yet exhibiting swallowing problems, because it is necessary when attempting to stage surgical intervention for alternative means of feeding. Our data indicate that a number of patients (over 30% as reported in this study) exhibit marginal or insufficient respiratory capacity before they exhibit a swallowing scaled score of 4 (needing supplemental tube feeding). Individuals with poor respiratory support are not good surgical candidates, even for procedures that can be carried out under local

anesthetics such as percutaneous endoscopic gastrostomies (PEG), because of their difficulty in remaining prone for the procedure. This has important implications for decisions about when to begin education on gastrostomies and when to schedule them. Early intervention and education about enteral feeding, therefore, is sometimes based on respiration vs. swallowing function.

### *Clinical Implications and Timing of Intervention*

The data presented in this paper are directed toward helping the clinician anticipate points of functional decline before they become a problem. Timing of dysphagia intervention, as well as providing the appropriate intervention, is important in maintaining optimum quality of life. Dysphagia symptoms can be categorized according to a continuum of five stages which correspond to the ALSSS scores: (1) normal eating habits (SS of 10, 9); (2) early eating problems (SS of 8, 7); (3) dietary consistency changes (SS of 6, 5); (4) needs tube feeding (SS of 4, 3); and (5) NPO (SS of 2, 1). Because the rate of dysphagia symptom progression varies from individual to individual in ALS, timing of most intervention is not suggested according to any time interval postonset, or according to first, second, or third clinic visit, but is based on the particular complex of symptoms, as well as the psychologic and attitudinal status of the patient.

### *Normal Eating Habits (SS 10-9)*

Our data indicate that speech and swallowing scale scores tend to parallel each other throughout disease progression. Dysphagia education, however, is usually begun before initial speech or swallowing symptoms appear. Education regarding the importance of hydration and nutrition is usually provided at the first clinic visit. Patients are encouraged at this time to make sure they are getting at least 2 quarts of liquid a day. They are discouraged from using caffeinated beverages because the diuretic effect can impede the advantage of increased liquid intake. The benefits of avoiding dehydration, which can cause fatigue and thickening of saliva, are discussed. Early education about fluid requirements is especially helpful for some patients with spinal ALS who will intentionally limit fluid intake because of the difficulty in transferring to a toilet for urination.

A diet history is taken during the first visit, so that the nutritionist can predict from habitual patterns what problems may arise later if dysphagia becomes a problem. Patients with either spinal forms of ALS, or early bulbar ALS, generally describe normal chewing, swallowing, and rate of ingestion. Some patients, however, upon receiving the diagnosis of ALS and gaining some knowledge regarding the progression of symptoms, will experience sensations of obstruction or "gagging" for which no physical basis has been found on clinical or radiologic examination. Such patients should be reassured regarding the relationship between swallowing dysfunction and physical findings. Patients with nominal abnormalities will often report food lodging in the gingivobuccal sulci or food sticking in the throat. Counseling regarding positioning, food consistency, and use of liquids is given at this time. Counseling on caloric intake and how to increase calories is begun when more than a few pounds weight loss is noticed, or when the patient reports that it is taking more effort to eat the same quantity of food.

### *Early Eating Problems (SS 8-7) and Dietary Consistency Changes (SS 6-5)*

Suggestions for changing consistency of preferred foods, the avoidance of thin liquids, and how to choose foods that will be safer and easier to eat are delayed until the patient begins to report difficulty, or the family or caregiver reports coughing incidents associated with meals, or unusually long meal times. This usually corresponds with a swallowing scaled score of 8. Therefore, during the second and third stages of swallowing difficulty, intervention is focused on dietary modification. Tongue, facial muscle, and masticatory muscle weakness, and prolonged meal time, chewing fatigue, and oral transport problems lead to difficulty with solid foods. Foods that are dry and crumbly, or that require longer mastication and oral preparation such as steak and leafy salads are usually the earliest textures to present difficulty. When tongue involvement, particularly spasticity (too much muscle tone), is combined with pharyngeal weakness, coughing with the ingestion of thin liquids occurs early in disease progression. The first complaint is most frequently with the ingestion of water or another thin liquid. In many instances, patients will reduce their fluid intake because of difficulty in swallowing, and they present with a mild chronic dehydration. This confounding condition produces changes in the salivary consistency that the patient describes as a thickening of secretions, or even dryness of the mouth. Some with tenacious secretions describe it as "too much saliva" because they cannot get rid of it. Additionally, the patient may suffer from excessive fatigue and malaise that is associated with a chronic state of dehydration. At this point, patients may be advised of the need to modify the diet to very soft food that is cohesive as a single bolus, rather than blenderize all foods to a thin liquid consistency. Fluids may need to be thickened to allow the patient to control the oral transport. Fiber may need to be added to the diet of those patients

with abdominal weakness or glottic impairment who experience problems with constipation. Additionally, almost all patients are advised to increase fluid intake to avoid the complications associated with chronic, insidious dehydration. Caffeine, because of its diuretic effect, should be eliminated from the diet. Hypertensive and cardiac patients should be reevaluated to see if their new medical priorities obviate the need for diuretic therapy.

In addition to thickening liquids, successful swallowing can be enhanced by emphasizing taste, texture, and temperature. Because water may be the most difficult liquid to swallow, juices and sodas can be substituted to provide taste and add calories. The facilitory effects of sensation achieved by temperature should be enhanced by cooling the liquids. Heating does not provide the same advantage since patients tend to take small sips and allow it to cool to almost body temperature before initiating the swallow. We have also found that carbonation provides some "texture" enhancement that is often beneficial. Popsicles, Jell-O, ice, and fresh fruit can also be used as a last resort to maintain free water intake by mouth.

Intervention for patients experiencing dysphagia at this stage also includes addressing safety techniques and precautions to avoid aspiration. These include postural adjustments, avoiding distraction, and teaching the Heimlich maneuver.

### *Needs Tube Feeding (SS 5-4)*

Often the loss of enjoyment of meals and the effort required to complete a meal will cause the patient to dread mealtime. It is at this point that most enter the fourth stage of dysphagia, "need tube feeding." This includes all patients who use tubes for primary feeding, who require tube feedings to supplement oral intake, and who, in the examiner's judgment (judgments based on weight loss, dehydration, dread of eating, etc.), should be using a tube. Choking episodes, food spillage, the need for special meal preparation, prolonged meal times, and respiratory or chewing fatigue all contribute to an aversion to eating. Tube feedings may be used because of swallowing incompetence or as a means of relieving the burdens associated with eating.

Several factors are extremely important at this point of intervention. Patients may be very resistant to feeding tubes, as they want to avoid all life-sustaining measures. We advise patients to separate decisions regarding life support (respirators) from decisions regarding feeding tubes. We provide information about the ways feeding tubes can improve quality of life by allowing the patient to stay nourished and hydrated while avoiding the fatigue and effort required by eating. We emphasize that the patient can continue to eat and drink for pleasure (to the degree that it is safe for them), and that the tube is for their convenience.

Another important factor at this point of intervention relates to the data regarding respiratory function. When vital capacity is less than 1.5 L, patients generally complain of respiratory fatigue. Vital capacities of less than 1.0 L usually leave patients symptomatic and without sufficient respiratory reserve. Problems of secretion management, ineffective cough, and reduction in speech loudness are evident. In patients with respiratory insufficiency, shortness of breath while eating may be evident. Because swallowing requires a momentary pause in respiration, and the duration of the swallow reflex is prolonged in ALS patients, it is an ominous sign when the patient's respiratory reserve is inadequate to compensate for the required pause in breathing.

The role of monitoring respiratory function is especially important to decisions regarding surgical alternatives such as placement of a PEG. Patients who exhibit vital capacity measures at or below 1 L do not tolerate the procedure for placement of the PEG. Monitoring vital capacity allows the clinician to know when to begin education regarding feeding tubes and their role in nutrition and hydration, and therefore quality of life. For some patients who exhibit adequate swallowing skills but decreasing vital capacity, early placement of a PEG may be advised.

Two cases will illustrate the role of respiration in making decisions about the timing of intervention for dysphagia. Figures 6 A and B and 7 A and B illustrate longitudinal data for 2 patients with ALS. Although their speech, swallowing, and upper extremity scale scores were not markedly different at the time of intervention, these cases were managed differently because of differences in respiratory status. For patient SH, a PEG was recommended for placement at 19 months postdiagnosis. At that time, he received a swallowing scale score of "6," indicating only dietary modifications such as a soft diet would be the usual recommendation. This early surgical intervention was undertaken because of his marginal respiratory status. On the other hand, for patient SB, only a dietary change was recommended in response to a steady weight loss because his adequate vital capacity suggested that a delay would not be harmful. In both cases, the goal of halting the weight loss was achieved, although by different means.

### *NPO (SS 2, 1)*

The final stage of dysphagia is reached when the patient cannot manage any oral intake. At this point many patients require assistance with the management of secretions in the form of an aspirator and/or medications used

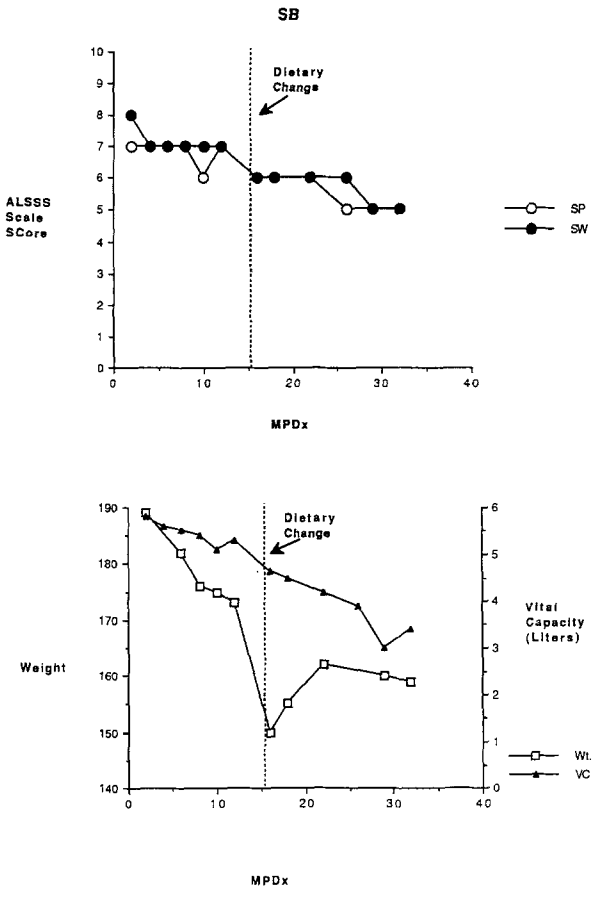


**Fig. 6.** Longitudinal data [speech (SP) and swallowing (SW) scaled scores, weight, and vital capacity] for patient SB, showing PEG intervention at 19 months postdiagnosis.

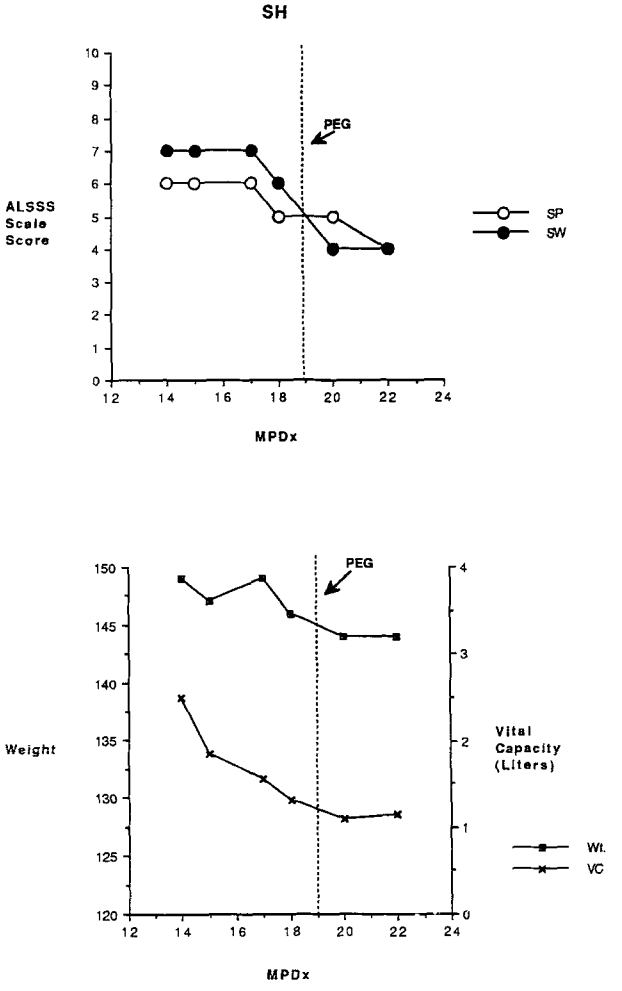


**Fig. 7.** Longitudinal data (speech and swallowing scaled scores, weight, and vital capacity) for patient SH, showing dietary change intervention at 15 months postdiagnosis.

to decrease salivary flow. Although initially a swallow reflex can still be elicited, the frequency with which the patient spontaneously swallows secretions diminishes. The most advanced patients are those who can no longer manage their own secretions. Unlike patients with a sudden onset of neuromuscular dysphagia, such as from stroke, aspiration pneumonia is a surprisingly rare complication in our ALS population.

## Conclusions

Dysphagia is a common symptom accompanying ALS. The onset of swallowing problems are often insidious and the progression relentless. Although to date there is no proven treatment to effectively reverse or arrest the progression of ALS, patients can be provided with intervention directed toward compensatory function, environmental modifications, and alternatives to oral feeding. This intervention commonly takes the form of education regarding the importance of hydration and nutrition; information regarding dietary and postural modifications; behavioral changes to avoid distraction, reflux, and other common problems; information regarding safety techniques including the Heimlich maneuver; and early education about enteral feeding options. Intervention also includes monitoring respiration and advising the patient about how respiratory deficits can impinge on swallowing function. By monitoring the patient's status in the areas of swallowing and respiration and anticipating problems based on patterns of progression illustrated in our data, the prevention of secondary complications such as dehydration and nutritional deficiency is possible. An understanding of the progression of dysphagia symptoms and the relationship of swallowing problems to speech and respiration is essential to the timing of intervention.

*Acknowledgment.* This research was supported in part by NIDCD grant 1K08 DC00043-01A1 and by NIDCD P60 DC00976.

## Appendix

### Amyotrophic Lateral Sclerosis Severity Scale

*Swallowing Scale*

Rating

NORMAL EATING HABITS

10 Normal Swallowing: Person denies any difficulty chewing or swallowing. Examination demonstrates no abnormality.

9 Nominal Abnormality: Only the individual with ALS notices slight indicators such as food lodging in the recesses of the mouth or sticking in the throat.

EARLY EATING PROBLEMS

8 Minor Swallowing Problems: Complains of some swallowing difficulties. Maintains essentially a regular diet. Isolated choking episodes.

7 Prolonged Time or Small Bite Size: Mealtime has significantly increased and smaller bit sizes are necessary. Must concentrate on swallowing liquids.

DIETARY CONSISTENCY CHANGES:

6 Soft Diet: Diet is limited primarily to soft foods. Requires some special meal preparation.

5 Liquified Diet: Oral intake adequate. Nutrition limited primarily to liquified diet. Adequate thin liquid intake usually a problem. May force self to eat.

NEEDS TUBE FEEDING

4 Supplemental Tube Feedings: Oral intake alone is no longer adequate. Person uses or needs a tube to supplement intake. Person continues to take significant nutrition (greater than 50%) by mouth.

3 Tube Feeding with Occasional Oral Nutrition: Primary nutrition and hydration accomplished by tube. Receives less than 59% of nutrition by mouth.

NOTHING BY MOUTH

2 Secretions Managed with Aspirator/Medication: Cannot safely manage any oral intake. Secretions managed by aspirator and/or medications. Swallows reflexively.

1 Aspiration of Secretions: Secretions cannot be managed noninvasively. Rarely swallows.

*Speech Scale*

Rating

NORMAL SPEECH PROCESSES

10 Normal Speech: Individual denies any difficulty speaking. Examination demonstrates no abnormality.

9 Nominal Speech Abnormality: Only the individual with ALS or spouse notices that speech has changed. Maintains normal rate and volume.

DETECTABLE SPEECH DISTURBANCE

8 Perceived Speech Changes: Speech changes are noted by others, especially during fatigue or stress. Rate of speech remains essentially normal.

7 Obvious Speech Abnormalities: Speech is consistently impaired. Affected are rate, articulation, and resonance. Remains easily understood.

BEHAVIORAL MODIFICATIONS

6 Repeats Messages on Occasion: Rate is much slower. Repeats specific words in adverse listening situations. Does not limit complexity or length of message.

5 Frequent Repeating Required: Speech is slow and labored. Extensive repetition or a "translator" is commonly needed. Person probably limits the complexity or length of messages.

USE OF AUGMENTATIVE COMMUNICATION

4 Speech Plus Augmentative Communication: Speech is used in response to questions. Intelligibility problems need to be resolved by writing or a spokesperson.

3 Limits Speech to One-Word Response: Vocalizes one-word response beyond yes/no; otherwise writes or uses a spokesperson. Initiates communication nonvocally.

LOSS OF USEFUL SPEECH

2 Vocalizes for Emotional Expression: Uses vocal inflection to express emotion, affirmation, and negation.

1 Nonvocal: Vocalization is effortful, limited in duration, and rarely attempted. May vocalize for crying or pain.

X Tracheostomy

*Upper Extremities Scale Rating*

NORMAL FUNCTION

10 Normal Function: Person denies any weakness of unusual fatigue of upper extremities. Examination demonstrates no abnormality.

9 Suspected Fatigue: Person suspects fatigue in upper extremities during exertion. Cannot sustain work for as long as normal. Atrophy not evident on examination.

INDEPENDENT AND COMPLETE SELF-CARE

8 Slow Self-Care Performance: Dressing and hygiene performed more slowly than usual.

7 Effortful Self-Care Performance: Requires significantly more time (usually double or more) and effort to accomplish self-care. Weakness is apparent on examination.

INTERMITTENT ASSISTANCE

6 Mostly Independent: Handles most aspects of dressing and hygiene alone. Adapts by resting, modifying (electric razor), or avoiding some tasks (e.g., buttons, tie)

5 Partial Independence: Handles some aspects of dressing and hygiene alone. However, routinely requires assistance for many tasks such as makeup, combing, shaving, etc.

NEEDS ATTENDANT FOR SELF-CARE

4 Attendant Assists Person: Attendant must be present for dressing and hygiene. Person performs the majority of each task with the assistance of the attendant.

3 Person Assists Attendant: The attendant assists the person with ALS for almost all tasks. The person moves in a purposeful manner to assist the attendant. Does not initiate self-care tasks.

TOTAL DEPENDENCE

2 Minimal Movement: Minimal movement of one or both arms. Cannot reposition arms.

1 Paralysis: Flaccid paralysis. Unable to move upper extremities.

*Lower Extremities Scale*

Rating

NORMAL

10 Normal Ambulation: Person denies any weakness or fatigue. Examination detects no abnormality.

9 Fatigue Suspected: Person suspects weakness or fatigue in lower extremities during exertion.

EARLY AMBULATION PROBLEM

8 Difficulty with Uneven Terrain: Difficulty and fatigue when walking long distances, climbing stairs, and walking over uneven ground (even thick carpet).

7 Observed Changes in Gait: Noticeable change in gait. Pulls on railing when climbing stairs. May use leg brace.

WALKS WITH ASSISTANCE

6 Walks with Mechanical Device: Needs or uses canes, walker, or assistant to walk. Probably uses wheelchair away from home.

5 Walks with Mechanical Device and Attendant: Does not attempt to walk without an attendant. Ambulation limited to less than 50 feet. Avoids stairs.

FUNCTIONAL MOVEMENT ONLY

4 Able to Support Weight: At best can shuffle a few steps with the help of an attendant for transfers.

3 Purposeful Leg Movements: Unable to take steps but can position legs to assist an attendant in transfers. Moves legs purposefully to maintain mobility in bed.

NO PURPOSEFUL LEG MOVEMENT

2 Minimal Movement: Minimal movement of one or both legs. Cannot reposition legs independently.

1 Paralysis: Flaccid paralysis. Cannot move lower extremities.

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