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Survey of current enteral nutrition practices in treatment of amyotrophic lateral sclerosis

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SUMMARY

Background and aims—Enteral nutrition (EN) is commonly prescribed for dysphagia and weight loss in amyotrophic lateral sclerosis (ALS), but there are currently no ALS-specific EN guidelines. We aimed to survey current practices prescribing EN to ALS patients.

Methods—An online survey was distributed using list servers administered by the Academy of Nutrition and Dietetics (AND), Muscular Dystrophy Association (MDA), and ALS Association (ALSA).

Results—A total of 148 dietitians, nurses, and physicians participated in the survey, of whom 50% were dietitians and 68% were associated with an ALS clinic. Only 47% of respondents reported their patients to be fully compliant with EN recommendations. Side effects (fullness, diarrhea, constipation, and bloating) were the most important reason for patient noncompliance, followed by dependence on caregivers. By contrast, only 3% of providers rated depression/

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hopelessness as the most important reason for noncompliance. Half of those surveyed reported that more than 25% of patients continued to lose weight after starting EN.

Conclusions—Our survey results show a high frequency of gastrointestinal side effects and weight loss in ALS patients receiving EN. These findings may be limited by sampling error and non-response bias. Prospective studies are needed to help establish EN guidelines for ALS.

Keywords

Amyotrophic lateral sclerosis; ALS; Enteral nutrition; PEG; Dysphagia; Weight loss

1. Introduction

Amyotrophic lateral sclerosis (ALS, or “Lou Gehrig’s disease”) is a neurodegenerative disease characterized by the progressive loss of upper and lower motor neurons. Patients typically survive 2 to 5 years from symptom onset until they reach respiratory paralysis and death.

Because of problems with dysphagia, muscle degeneration, and increased energy expenditure, ALS patients frequently have difficulty maintaining their body weight and nutrition status. Patients not using enteral nutrition have been shown to consume on average 10–19% less than their recommended daily calories.^{1,2} This diet deficit is correlated to their degree of weight loss and reduction in body fat percentage.^{1,3} Because weight is a prognostic factor in disease progression, weight maintenance is important both in preventing malnutrition and delaying physical decline.^{1,2,4–6}

Enteral nutrition (EN) administered via a percutaneous endoscopic gastrostomy (PEG) or radiologically inserted gastrostomy (RIG) device provides long-term nutritional support for patients suffering from dysphagia. According to the American Academy of Neurology (AAN) 2009 Practice Parameter update: “Enteral nutrition administered via PEG is probably effective in stabilizing body weight/body mass index”.⁷ The AAN guidelines reviewed two Class II studies in which EN was shown to stabilize body weight in patients receiving PEG compared to continued weight loss in controls who refused PEG.⁷ The European Federation of Neurological Societies (EFNS) guidelines from 2012 state that “PEG improves nutrition, but there is no convincing evidence that it prevents aspiration or improves quality of life or survival”.⁸ Despite the potential benefits of EN treatment in the care of ALS, there are currently no prospective randomized studies of the use of EN in ALS. While the AAN and EFNS guidelines recommend early insertion of feeding tubes before respiratory status has declined to 50% of predicted forced vital capacity, there are no guidelines regarding the best method of EN administration, target body weight, type of nutritional supplements, best method to calculate daily caloric requirements, or frequency of monitoring after EN initiation. In part as a result, EN is frequently underutilized in treatment of ALS.⁹ The aim of this study was to survey current EN practices of ALS providers in order to begin to address these important questions.

2. Methods

2.1. Participants

Partners Healthcare Institutional Review Board approval was obtained for the online survey and all recruitment letters. Participants were invited to participate in the online survey if they were active in prescribing tube feedings to patients with ALS. Invitations were sent through list servers administered by the Academy of Nutrition and Dietetics (AND), Muscular Dystrophy Association (MDA), and ALS Association (ALSA). These associations primarily serve the United States and Canada. Participants who filled out the survey were entered into a drawing for an iPad2. Participants were assured that their answers would be confidential, and that their name and contact information would only be used for the purposes of entering into the drawing. A total of 148 dietitians, nurses, and physicians participated in the survey.

We developed a 19 question survey designed to query current clinical practices and experiences in prescribing enteral nutrition to patients with ALS (see Online supplementary material). The survey was developed in conjunction with several dietitians and physicians involved in treating ALS (see author list) and pilot tested by dietitians not involved in the development of the survey. The 10 minute web-based survey was administered through LimeSurvey from the period of August through November 2011. The survey was divided into 5 parts. Part 1 consisted of questions about their work facility, experience with ALS and role in prescribing EN. Part 2 asked participants about the different EN formulas and routes of administration. Part 3 involved questions about problems with EN tolerability and compliance. Part 4 addressed how participants calculate the caloric needs and weight goals of their patients. Part 5 asked about how participants monitor their patients' weights and adjust their care to address weight loss. Participants were also invited to provide free-text answers regarding their experience with treating ALS.

2.2. Data management and analysis

The survey was administered on the website LimeSurvey (<http://www.limesurvey.org/>), an open source software hosted by Partners' internal datacenter. This website was chosen over other 3rd-party survey hosting websites because of its security and Secure Sockets Layer (SSL) encryption. Analysis of results was done using LimeSurvey, as well as on Microsoft Office Excel and SAS 9.2 (SAS Institute, Cary, NC). A separate sub-analysis of dietitians treating >20 patients a year was also performed to look at concordance or deviation from all participants.

3. Results

3.1. Demographics

148 health workers responded to this survey (Table 1). Dietitians encompassed the largest proportion of participants, followed by physicians and nurses. The majority of participants worked at MDA, ALSA or other ALS clinics, followed by in-patient and out-patient hospitals. Most participants reported treating more than 30 ALS patients a year. Of those who answered the survey question, 23.2% reported being involved only in the initial

recommendations for enteral nutrition, while most reported being involved in long term follow-up care (76.8%).

3.2. Methods of administration

Bolus feeding was the most commonly used method of administering EN (53.4%), followed by gravity feeding (23.3%). Other methods such as pump feeding (6.9%) and combinations of bolus and gravity (16.4%) were used less often. Bolus feeding was associated with lower rates of patient compliance compared to gravity feeding (17.0% versus 7.4% of participants reported only 25–50% compliance rates, Fisher's Exact Test $p = 0.008$). However, gravity feeding was associated with a higher frequency of diarrhea than bolus feeding (48.2% versus 26.5%, $p = 0.03$). No other gastrointestinal side effects were reported more frequently in one method of administration over another.

3.3. Calculation of caloric requirements and ideal body weight

In order to calculate the caloric requirements of their patients, participants predominately used the kcal/kg body weight equation (58.8%) or Harris-Benedict equation (27.7%). The Mifflin St. Jeor equation (8.8%) and indirect calorimetry (5%) were rarely used as first choice options, but were sometimes used as second or third options after the first choice failed to adequately calculate the patient's caloric needs. Similar results were found in the subanalysis of dietitians (51.1% used the kcal/kg equation and 36.2% used the Harris-Benedict equation as their first choice).

The most common equations used to estimate patients' ideal body weight (IBW) were BMI (32.4%), Hamwi's equation (28.4%), and pre-illness weight (22.3%). Providers reported their nutritional goals to be for patients to maintain weight (51.6%) or to gain weight regardless of IBW (39.6%). Only 8% of providers aimed for weight maintenance when patients had lost more than 10% of their IBW.

3.4. Patient compliance

Only 47% of respondents considered 75–100% of their patients as fully compliant (Fig. 1A). Side effects were the most important reason for patient noncompliance with EN recommendations (42.1% of respondents). The most frequently reported side effects of EN included fullness, diarrhea, constipation, and bloating (Table 2). Abdominal pain, nausea, and vomiting were reported less frequently. Other common reasons for noncompliance included dependence on caregivers to administer feedings (24.0%) and the amount of time required to administer feedings (14.1%). Less important factors in noncompliance included a desire to lose weight or fear of weight gain (4.0%), and feelings of depression and hopelessness (3.3%).

3.5. Weight loss

Overall, 51% of participants reported that less than 25% of their patients lost weight after starting enteral nutrition. As shown in Fig. 1B, 36% reported observing weight loss in 25–50% of patients, 10% reported weight loss in 50–75% of patients and 3% in 75–100% of patients. Perceived rates of patient compliance were not predictive of observed rates of weight loss (linear regression $p = 0.75$).

We next examined the frequency of observed weight loss by participants based upon the formula they used most often to calculate caloric requirements. Indirect calorimetry was used by only 5 participants to calculate caloric requirements, and of these participants only 1 reported seeing weight loss in more than 25% of ALS patients. By contrast, 45% of participants who used the kcal/kg estimation and 60% of those who used the Harris-Benedict equation reported that more than 25% of their patients continued to lose weight. Comparing these equations, there was no significant difference ($p = 0.2$ by Fisher's exact test). Weight loss in more than 25% of patients was observed at a similar frequency by participants who reported that their goal was weight gain (50%) versus weight stability (45%) ($p = 0.8$).

Despite the reported frequency of weight loss, 40.5% healthcare providers reported that the predictive formula they used accurately represented their patients' caloric needs. Only 16.2% did not find the formulas accurate, while 43.2% did not provide an answer to this question.

Finally, participants were allowed to enter additional information in a free text portion of the survey. Two major themes emerge from this data including "there is no set formula or procedure that works for everyone" and "hydration is not recommended adequately".

4. Discussion

This is the first large effort to survey current practices for prescribing enteral nutrition to ALS patients, including commonly used methods for feeding, formulas for calculating caloric requirements, and weight goals. Although other surveys have been performed of ALS healthcare providers, they have focused more on the reasons for initiating EN rather than the clinical outcomes of EN.^{10,11} Our survey is consistent with the results of Rio et al. in revealing that methods currently used for assessing nutritional status and daily energy requirements for EN in ALS patients are nonspecific to the disease process and vary between institutions.¹¹

The primary results of our survey include: (1) gastrointestinal side effects of EN are common in ALS; (2) side effects are the primary reason for patient non-compliance with tube feed recommendations; (3) weight loss is frequently seen after starting EN, and does not appear to correlate with the methods used to calculate caloric needs or ideal body weight. The frequency of gastrointestinal side effects, specifically bloating, constipation, diarrhea and fullness are higher than have been reported in persons with normal gastrointestinal function given enteral nutrition, including patients with neurological diseases such as stroke, intracranial hemorrhage and trauma.^{12,13} Whether this is an effect of malnutrition on the gastrointestinal tract or whether this is a symptom of gastrointestinal dysfunction due to ALS itself is unclear, and would be an important avenue for investigation.¹⁴⁻¹⁶ The frequency of weight loss after institution of EN has not been reported in prior case series and clinical trials involving feeding tube placement in ALS.⁷ This may suggest that our survey data are less reliable than case series and clinical trial data. Or it may be due to the fact that we surveyed a large number of dietitians and clinicians whose real world experiences and patients may differ from clinical trials.¹⁷ A prospective observational study to look at the incidence of weight loss and gastrointestinal side effects

after initiation of EN would be extremely important clinically, as malnutrition should be avoided in ALS.²

The AAN practice parameter recommendations for future nutritional research identified several important questions including: “What is the effect of enteral nutrition administered via percutaneous endoscopic gastrostomy (PEG) on weight stability?”, “What is the efficacy of nutritional support via PEG in prolonging survival?” and “What is the effect of enteral nutrition delivered via PEG on quality of life (QOL)?”.⁷ Our survey results identify several additional questions for future prospective nutrition studies, including:

1. How frequently do ALS patients continue to lose weight after starting EN?
2. If weight loss continues, is this due to inadequate calorie recommendations or due to intolerance of gastrointestinal side effects?
3. What is the best method to estimate the caloric goals for ALS patients?
4. How can we reduce gastrointestinal side effects of enteral nutrition for ALS patients in order to improve compliance?
5. How can we improve patient nutrition in the absence of caregiver support?
6. What disease variables may explain weight loss in patients who start EN?

Our survey data and methods were subject to several limitations. Because there is no database of all ALS providers who prescribe EN, we do not know the percent who responded to the survey. We also could not contact non-responders, therefore participants may not be a representative sample of all ALS providers of EN. With these limitations in mind, this study is still relevant for collecting the largest number of survey respondents in this field.

Despite these limitations, the findings in this study are significant to the growing literature about the indications of EN in different neurological causes of dysphagia. Several multicenter prospective randomized trials have found non-significant trends toward both reductions in death and poor outcome in dysphagic stroke patients fed with early enteral nutrition.^{18,19} While more specific EN treatment guidelines exist for these illnesses, further research is necessary to establish guidelines for the administration of EN in treating ALS. Laying down these guidelines is crucial for giving providers more structure in their care and addressing the frequent underutilization of EN as a tool in ALS treatment.⁹

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Abbreviations

AND	Academy of Nutrition and Dietetics
ALS	amyotrophic lateral sclerosis
ALSA	Amyotrophic Lateral Sclerosis Association
BMI	body mass index
EN	enteral nutrition
MDA	Muscular Dystrophy Association
PEG	percutaneous endoscopic gastrostomy
RIG	radiologically inserted gastrostomy

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Fig. 1.

A. Proportion of patients reported to be fully compliant with tube feed recommendations. Participants were asked to estimate the proportion of their patients who were fully compliant with their prescribed enteral nutrition. B. Proportion of patients reported to experience weight loss. Participants were asked to estimate the proportion of their patients who experienced weight loss after starting enteral nutrition.

Table 1

Participant demographics.

Demographics	N (%)
Profession	
Dietitian	74 (50)
Physician	19 (12.8)
Nurse	35 (23.7)
Missing	20 (13.5)
Workplace (multiple answers allowed)	
MDA/ALSA clinic	100 (67.6)
In-patient hospital	42 (28.4)
Out-patient hospital	19 (12.8)
Homecare Company	5 (3.4)
Other	16 (10.8)
ALS patients treated annually	
1–10	16 (10.8)
11–20	14 (9.46)
21–30	16 (10.8)
>30	89 (60.1)

Participants in the nutritional survey by licensure, type of practice, and number of ALS patients treated annually. Under type of practice, participants were allowed to choose more than one answer. ALS: amyotrophic lateral sclerosis; ALSA: Amyotrophic Lateral Sclerosis Association; MDA: Muscular Dystrophy Association.

Table 2

Common side effects of EN.

Common side effects	Most often (%)	Frequently (%)	Sometimes (%)	Rarely (%)	Never (%)	No answer (%)
Fullness	21.5	27.3	31.4	5.8	0	14.1
Diarrhea	15.7	11.6	36.4	19.8	0	16.5
Constipation	10.7	14.1	33.9	21.5	1.7	18.2
Bloating	9.9	21.5	40.5	10.7	0	17.4
Abdominal pain	7.4	1.7	35.5	36.4	2.5	16.5
Nausea	5.8	2.5	40.5	33.9	2.5	14.9
Vomiting	1.7	0	15.7	55.4	10.7	16.5

Frequency of gastrointestinal side effects reported by all participants expressed as a percent.