DOI: 10.1590/0004-282X20160145

ARTICLE

Assessment and nutrition education in patients with amyotrophic lateral sclerosis

Avaliação e educação nutricional em pacientes com esclerose lateral amiotrófica

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ABSTRACT

Neurological patients with amyotrophic lateral sclerosis (ALS)often deteriorate to a worsening nutritional status. The aim of this study was to compare the nutritional status and food intake after nutrition education in patients with ALS. Clinical, anthropometric and functional variables were analyzed. Fifty-three patients were monitored at an early stage of the disease. The average score on the functionality scale was 33 points. Initially only 3.8% were classified as low body weight. After three months, 50% showed significant variation in anthropometric measures related to muscle mass and body fat reserves without association with clinical variables. After nutritional guidance, there was an increase in the intake of all food groups, especially the dairy group ($p < 0.05$).The change of the nutritional status occurs early in patients with amyotrophic lateral sclerosis, even in those previously eutrophic or over weight. There was an increase in food intake after nutritional guidance according to the food guide adapted to the Brazilian population.

Keywords: amyotrophic lateral sclerosis; continuing education; body composition.

RESUMO

Pacientes neurológicos com esclerose lateral amiotrófica frequentemente evoluem com piora do estado nutricional. O objetivo desse estudo foi comparar o estado nutricional e a ingestão alimentar depois da orientação nutricional em pacientes com ELA. Variáveis clínicas,antropométricas e funcionais foram analisadas. 53 pacientes foram avaliados na fase inicial da doença. A pontuação média da escala de funcionalidade foi de 33 pontos. Inicialmente apenas 3,8% foram classificados como baixo peso. Após três meses, 50% apresentaram variação significativa nas medidas antropométricas relacionadas com reservas de massa muscular e gordura corporal, sem associação com variáveis clínicas. Após orientação nutricional, houve um aumento na ingestão de alimentos de todos os grupos com relevância para o grupo de lacticínios ($p < 0,05$). A mudança do estado nutricional ocorre precocemente em pacientes com ELA, mesmo naqueles anteriormente eutróficos ou sobrepeso. Houve um aumento na ingestão de alimentos após orientação nutricional de acordo com o guia alimentar adaptado da população brasileira.

Palavras-chave: esclerose lateral amiotrófica; educação continuada, composição corporal.

Amyotrophic lateral sclerosis (ALS) is the most common form of motor neuron disease (MND), characterized by selective loss of motor neurons in the spinal cord, brainstem and motor cortex. Symptoms include atrophy, weakness and muscle fatigue, fasciculation, dysarthria, dysphagia, sialorrhea and emotional lability[1, 2].

Muscle atrophy may mask the increased metabolic demand characteristic of progressive diseases. The energy being channeled to maintain pulmonary ventilation, justifies the increase in resting energy expenditure in these patients. In recent times, the nutritional demand in ALS gained such proportions that studies sought to identify it as predictive factor, emphasizing the importance of nutritional intervention in disease treatment[3, 4].

Weight loss has multifactorial causes including oropharyngeal dysphagia, secondary loss of appetite, loss of autonomy with emotional disorders, inappropriate use of noninvasive ventilation and hypermetabolism[5]. There is a direct association between weight loss and disease severity. At the time of diagnosis, a reduction of 5% of normal weight increased the risk of death in this population by 30%. A reduction of 1 kg/m^2 in body mass index (BMI) was associated with a 20% risk of death[4].

Changes in body composition have been described since the first studies on nutrition in ALS. Kasarskis et al. [6] found

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Conflict of interest: There is no conflict of interest to declare.

Received 13 May 2016; Accepted 16 August 2016.

that men show a greater loss of skeletal muscle compared to women, suggesting different nutritional requirements. During the course of illness, decreased muscle mass is associated with lower survival, regardless of body weight[7].

Based on this evidence, nutritional intervention when applied early allows the use of this resource in the different stages of the disease. Thus, the aim of this study was to compare the nutritional status and food intake after nutrition education.

METHODS

This is a longitudinal study in patients with MND/ALS, treated at the Neurology/Neurosurgery Department, of the Federal University of São Paulo (UNIFESP), during the period of 2013–2014. Fifty-three patients with confirmed diagnosis according to the El Escorial[8], were included in the initial phase of medical treatment. Patients receiving any nutritional orientation were excluded. Patients were referred by a medical staff member.

The nutritional instrument was applied at both phases of the study, and consisted of a structured interview based on clinical evaluation, nutritional assessment, analysis of food intake and functional evaluation, according to the Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS)[9]. The ALSFRS-R is a questionnaire-based scale for activities of daily living. This scale contains 12 items grouped into three domains that encompass appendicular function (gross motor tasks), bulbar and respiratory function. Each item has a 5-point scale (0 for unable; 4 for normal) and scores ranging from 0 to 48. Low scores denote a serious disease status.

The assessment of nutritional status was performed using anthropometric measurements and analyzed according to population scores[10]. The measures were: weight, height, arm circumference, triceps skinfold, biceps skinfold, suprailiac skinfold, subscapular skinfold, arm muscle circumference, arm muscle area, arm fat area and calf circumference for individuals over 60 years of age. For the anthropometric index, the BMI was used[10]. The adequacy percentile[10] was calculated using the software Decision Support System in Nutrition version 2. 5 - UNIFESP / Escola Paulista de Medicina.

For compartmented analysis of body composition, the arm muscle area measures for muscle mass and triceps skinfold for body fat were selected. For analysis of food intake, the food consumption frequency questionnaire was applied and analyzed according to the food guide for the Brazilian population[11].

The study occurred in two phases: an initial evaluation at the time of referral (T0) and after three months (T1). In the first evaluation, patients were divided randomly into two groups: intervention group (IG) and control group (CG). The criterion for the division of groups was the type of nutritional guidance provided. For the IG patients the food pyramid tool, adapted to the Brazilian population, was used[11]. For the CG, general guidelines were provided, including changes in food consistency and dividing up the components of meals.

The project was approved by the Ethics Committee in Research of the Federal University of São Paulo under number 492 239 CEP.

Statistical analysis

Categorical variables were described in absolute value and relative frequency. Continuous variables, after being submitted to the Kolmogorov-Smirnovnormality test, were described using central tendency and dispersion measures. We considered a decline in the anthropometric measures to be the distribution of values below the 25th percentile of the variation in both phases of assessment (T0 and T1). The association between predictor factors and the decline of selected anthropometric measurements was evaluated according to Pearson's chi-square test or Verisimilitude ratios. Results with type I error probability of less than 5% were considered statistically significant.

RESULTS

Among the characteristics of the studied population, there was a predominance of males, aged 57 years. For the type of manifestation of the disease, an appendicular presentation was predominant (79. 2%). The factors that characterized the early stages of the disease were the time between symptom onset and diagnosis (360 days) and the scores found on the ALSFRS functionality scale (33 points). As for nutritional status, only 3. 8% of patients presented with low body weight, with similar proportions among those considered normal weight or overweight (Table 1).

Table 1.General characteristics of the study population.

Variable	n	%
Gender		
Male	31	58,5
Female	22	41.5
Age (years)		
Average (min – max)	57.0	(33 – 76)
< 60 years	34	64,2
≥ 60 years	19	35.8
Manifestation of thedisease		
Appendicular	42	79.2
Bulbar	11	20.8
Symptom onset (days)		
Average (min – max)	360	90 – 3270
ALSFRS score		
Average (min – max)	33	13 – 44
Nutritional status according to BMI		
Lowweight (< 18,9)	2	3,8
Eutrophy (19–24,9)	26	49.1
Overweight (≥ 25)	25	47.2
Nutritionalcounseling		
Foodguide	35	66
Standard orientation	18	34

ALSFRS:amyotrophic lateral sclerosis functional rating scale;BMI: body mass index.

Table 2 shows the variation in anthropometric measurements between the two assessments: at baseline (T0) and after three months of follow-up (T1). Despite the biceps skinfold measures, suprailiac skinfold, subscapular skinfoldand arm muscle circumference, at least half of the subjects showed declines in measures, with a significant difference for BMI, arm circumference, biceps skinfold, triceps skinfold, and subscapular skinfold, compared with the initial phase.

The results of variables related to the decline of body weight are shown in Table 3. We observed a decline ≥ 0. 53 kg/m^2 in BMI between the period of evaluations. The gender, age and time of onset of symptoms showed very similar proportions among categories, without significant difference. We observed that individuals with bulbar involvement, and those who were overweight showed the largest decline in BMI ($p < 0.4$ and $p < 0.1$ respectively).

For the changes of arm muscle area and triceps skinfold, shown in Tables 4 and 5, we found no significant difference between the study variables and reducing measurement. It is worth mentioning that the age group and functionality were the variables that were most related to the decline of the measurement.

Study variables according to the type of nutritional guidance are applied in Tables 6 and 7. The age group and the time of onset of symptoms were similar between the IG and CG. We found a higher frequency of female patients and with bulbar manifestation in the IG. Similar averages paired were found between anthropometric measures highlighting averages of BMI, arm circumference and skinfolds, but with no significant difference. The same was not observed for the functionality ($p = 0.03$). The IG patients had lower scores on the ALSFRS scale in both phases (T0 and T3), as shown in Figure.

Table 2. Distribution of variation (T0 – T1) of anthropometric measurements according to the percentiles of the study population.

Variable	Min.	P25	Average	P75	Max.	p**
BMI (kg/m^2)	-3.63	-0.53	-0.09	0.0	+1.47	0.02
AC (cm)	-3.0	-1.0	-0.75	0.0	+3.0	0.04
TSF (mm)	-5.0	-1.0	-0.90	0.0	+4.0	0.008
BSF (mm)	-6.0	-1.0	0.0	0.0	+2.0	0.001
SISF (mm)*	-8.0	-1.0	0.0	0.0	+3.0	0.003
SSF (mm)	-3.0	-1.0	0.0	0.0	+3.0	0.005
AMC (cm)	-3.0	-1.0	0.0	2.0	+4.0	0.69
AMA (cm^2)	-8.0	-3.0	-2.0	0.0	+16.0	0.94

T0: time of initial evaluation; T1: time after three months; P25: values below the 25th percentile; P75: values below the 75th percentile; BMI: body mass index; AC: arm circumference; TSF: triceps skinfold; BSF: biceps skinfold; SISF: suprailiac skinfold; SSF: subscapular skinfold; AMC: arm muscle circumference; AMA: arm muscle area; *missing values, n = 1 (1.9%); **T-test paired.

Table 3. Body mass index decline category according to clinical, demographic and nutritional variables.

Variable	BMI decline		p
	< 0.53 Kg/m^2	≥ 0.53 Kg/m^2	
Gender			
Male	23 (76.7%)	7 (23.3%)	0.74
Female	16 (72.7%)	6 (27.3%)	
Age group			
< 60 years	25 (75.8%)	8 (24.2%)	0.86
≥ 60 years	14 (73.7%)	5 (26.3%)	
Disease form			
Appendicular	32 (78%)	9 (22%)	0.32
Bulbar	7 (63.6%)	4 (36.4%)	
Symptom onset			
≤ 360 days	24 (72.7%)	9 (27.3%)	0.61
> 360 days	15 (78.9%)	4 (21.1%)	
ALSFRS			
≥ 33 points	23 (79.3%)	6 (20.7%)	0.42
< 33 points	16 (69.6%)	7 (30.4%)	
Prior nutritional status			
Eutrophy/lowweight	22 (84.6%)	4 (15.4%)	0.08
Overweight	16 (64%)	9 (36%)	
Nutritionalcounseling			
FoodGuide	25 (71.4%)	10 (28.6%)	0.39
Standard orientation	14 (82.4%)	3 (17.6%)	

BMI: body mass index; ALSFRS:amyotrophic lateral sclerosis functional rating scale.

Table 4. Arm muscle area decline categories according to clinical, demographic and nutritional variables.

Variable	Arm muscle area decline		p
	< 3.0 cm^2	≥ 3.0 cm^2	
Gender			
Male	20 (64.5%)	11 (35.5%)	0.31
Female	17 (77.3%)	5 (31.3%)	
Age group			
< 60 years	22 (64.7%)	12 (35.3%)	0.27
≥ 60 years	15 (78.9%)	4 (21.1%)	
Disease form			
Appendicular	29 (69%)	13 (31.%)	0.45
Bulbar	8 (72.7%)	3 (27.3%)	
Symptom onset			
≤ 360 days	24 (72.7%)	9 (27.3%)	0.55
> 360 days	13 (65%)	7 (35%)	
ALSFRS			
≥ 33 points	19 (65.5%)	10 (34.5%)	0.45
< 33 points	18 (75%)	6 (25%)	
Prior nutritional status			
Eutrophy/lowweight	19 (70.4%)	8 (29.6%)	0.85
Overweight	17 (68%)	8 (32%)	
Nutritionalcounseling			
FoodGuide	24 (68.6%)	11 (31.4%)	0.78
Standard orientation	13 (72.2%)	5 (27.8%)	

ALSFRS:amyotrophic lateral sclerosis functional rating scale.

Table 5. Triceps skinfold decline categories according to clinical, demographic and nutritional variables.

Variable	TSF decline		p
	<1.0 mm	≥ 1.0 mm	
Gender			
Male	15 (50%)	15 (50%)	1.0
Female	11 (50%)	11 (50%)	
Age group			
< 60 years	18 (54.5%)	15 (45.5%)	0.38
≥ 60 years	8 (42.1%)	11 (57.9%)	
Disease form			
Appendicular	21 (51.2%)	20 (48.8%)	0.73
Bulbar	5 (45.5%)	6 (54.5%)	
Symptom onset			
≤ 360 days	18 (54.5%)	15 (45.5%)	0.38
> 360 days	8 (42.1%)	11 (57.9%)	
ALSFRS			
≥ 33 points	17 (58.6%)	12 (41.4%)	0.16
< 33 points	9 (39.1%)	14 (60.9%)	
Prior nutritional status			
Eutrophy/lowweight	14 (53.8%)	12 (46.2%)	0.48
Overweight	11 (44%)	14 (56%)	
Nutritionalcounseling			
FoodGuide	14 (40%)	21 (60%)	0.39
Standard orientation	12 (70.6%)	5 (29.4%)	

TSF:triceps skinfold; ALSFRS:amyotrophic lateral sclerosis functional rating scale

Table 6. General and demographic data of the study population according to the nutritional guidance.

Variable	Control (n = 18)	Intervention (n = 35)	p
Age			
Average ± DP	54 ±10.5	56.8 ±10.5	0.35
Gender			
Male	13 (72.2%)	18 (51.4%)	0.14
Female	5 (27.8%)	17 (48.6%)	
Disease form			
Appendicular	16 (88.9%)	26 (74.3%)	0.21
Bulbar	2 (11.1%)	9 (25.7%)	
Symptom onset			
Average	315	360	0.62
Min – Max	150 - 1560	90 - 3270	
ALSFRS			
Average	37	33	0.23
Min – Max	18 - 44	13 - 45	

ALSFRS: amyotrophic lateral sclerosis functional rating scale.

Table 7. Frequency of anthropometric and functional outcomes according to nutritional guidance groups

Variable	Control (n = 35)	Intervention (n = 18)	p
BMI*	%	%	
Delta + orstable	9 (52.9)	17 (48.6)	0.76
Delta -	8 (47.1)	18 (51.4)	
AMA			
Delta < -10%	14 (77.8)	25 (71.4)	0.62
Delta ≥ -10%	4 (22.2)	10 (28.6)	
AMC			
Delta < -10%	17 (94.4)	34 (66.0)	0.62
Delta ≥ -10%	1 (5.6)	1 (2.9)	
TSF			
Delta <10%	13 (76.5)	24 (68.6)	0.55
Delta ≥10%	4 (23.5)	11 (31.4)	
AC*			
Delta <10%	17 (100)	34 (97.1)	
Delta ≥10%	0 (--)	1 (2.9)	
ALSFRS			
Delta + orstable	10 (55.6)	9 (25.0)	0.03
Delta -	8 (44.4)	26 (74.3)	

BMI: body mass index; AMA: arm muscle area; AMC: arm muscle circumference; TSF: triceps skinfold; AC: arm circumference; ALSFRS: amyotrophic lateral sclerosis functional rating scale; *missing values. n=1 (1.9%).

For both groups, a low frequency in food intake of leguminous groups, cereals and tubers, vegetables and fruits was found, especially the cereal groups, tubers and vegetables. We found a frequency greater than 50% in the consumption of dairy groups, meat and eggs, oils and fats in the IG, as shown in Figure. The qualitative nutritional guidance of the diet seems to be more effective for the consumption of dairy products and fruit groups, particularly the dairy group, which showed a significant increase in the recommended consumption frequency ($p < 0.05$).

DISCUSSION

The population studied portayedthe epidemiological characteristics of ALS described in the literature with a predominance of males compared to females, prevalence in the sporadic form and initial appendicular involvement of the bulbarregion[12]. The basic characteristics between the studied groups were shown to be compatible with the purpose of the study and randomization allowed comparison.

The median age group was 57 years old. In studies of the Brazilian population, the average is 52 years, whereas other countries range from 59-65 years of age. The population studied was older than that described in the Brazilian literature[13].

The mechanism by which the progression of the disease is faster in older individuals is still unknown, however it seems to be associated with the natural loss of motor neurons. This does not occur for the adults in balancing the decline of motor function[12].

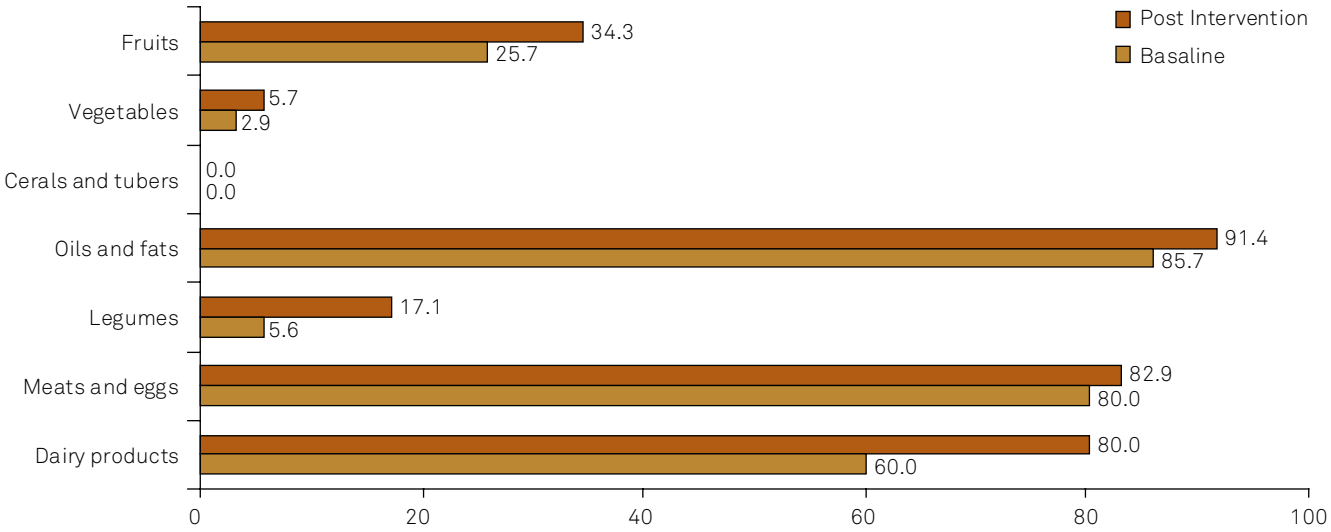

Figure. Food frequency of patients in the initial phase, and after nutritional guidance.

The time between the first symptoms and diagnosis was 360 days, similar to that described in the literature pointing to an average of 12 months, when 50% of the motor neurons have been lost; although the symptoms and severity of the disease occur differently between individuals[14, 15].

As far as functionality goes, the patients presented with a slightly impaired function with independence for activities of daily living. So far, a prognostic score for functionality has not been described. It is known that scores greater than or equal to 30 points may correspond with longer survival[14]. The same applies to nutritional matters. The ALSFRS has been associated with objective measures of muscle strength and lung function, however, few studies were found that assess the scale items with nutritional parameters[16, 17, 18].

Unlike studies that observed low body weight as a symptom in the early stages of the disease, the same was not found in this study. The change in body weight for values above the recommended, according to the age group and stature of the Brazilian population, has been growing in recent decades. When degenerative diseases affect individuals older than 45 years of age, being overweight and obese are commonly-observed symptoms[16, 19]. Another reason could be the inclusion of patients in the early stages of the disease where the respiratory functions were not affected; a fact that would help the preservation of body fat reserves. The importance of multidisciplinary assessment in the literature is well described, including early nutritional counseling as a positive factor for maintaining the nutritional status with a preventive approach in the treatment of ALS[14, 15, 16, 19].

Although there was a variation in BMI, it was not enough to change the classification of nutritional status, stressing that as an isolated measure, the BMI is not sensitive enough to detect body alteration in the course of the disease[16]. Muscle atrophy is itself the triggering factor of changes, justifying the periodic anthropometric evaluation, with a recommendation for the anthropometry of the arm[20].

There was a significant decline of the measures from the third month on, characterizing early changes in nutritional status, and these findings are in agreement with other studies to reduce anthropometric measurements and their maintenance after the completion of education on the habitual diet, with calorie and protein supplements[4, 20, 21].

With attention to the studied population, we did not find any characteristics strongly associated with the decline. Elderly patients, with time from onset greater than 12 months and a score less than 33 points on the ALSFRS scale were the ones with the greatest loss of body fat. In the case of a diagnostic study and nutrition education, data on a prognosis would not fit in this scope of work, but it is worth recalling that the reduction of body fat has been described as a negative prognostic factor[4, 21, 22].

The initial nutritional status of individuals was probably the mitigating factor for the changes in BMI. We found a high frequency of eutrophic (within ideal weight range) individuals (49%) and overweight (47%) patients. After the three months, patients with excess weight had minor variations of BMI when compared to eutrophic individuals, however without statistically significant difference, suggesting that losses in fat reserves can precede the decrease in body weight.

As for the frequency of anthropometric outcomes between the groups, although few significant differences have been found, both showed a decrease of anthropometric measurements between the study periods (T0 and T1). In 50% of patients there was a reduction of the measures, in particular the biceps skinfold, subscapular skinfoldand arm muscle circumference, with statistically significant difference for BMI, arm circumference and all skin folds (biceps, triceps, and subscapular). The same was observed by Stanich et al. [20] who found their analysis of body composition was effective for introducing nutritional supplementation, and also by Slowie et al. [23]who analyzed body composition in patients with ALS.

Salvioni et al. [18]also analyzed body composition using anthropometric measures and correlated them with the time between diagnosis and nutritional assessment of patients with ALS. The study showed that the delay in nutritional intervention negatively influences the loss of muscle mass, demonstrating the importance of early intervention.

Initially, the dietary pattern presented characterized the food inadequacy commonly observed in the Brazilian population, with respect to the components and composition of meals. Other studies, in particular, have demonstrated a high frequency (70%) of subjects with ALS who had a lower food intake, especially of calories and proteins[24, 25, 26].

The importance of nutrition in ALS is so relevant that even without a socio-demographic study analyzing factors associated with the onset of the disease, it was found that a low intake of fruit and vegetables would be a risk factor[23]. In the frequency of food consumption questionnaire applied, apart from the inadequacies, the lack of knowledge about the importance of food groups and how to incorporate them into the daily habit was identified.

Determining the components of the meals was not an instrument of analysis of this study, however we observed an inadequacy related to the number of meals per day, with consumption of just three meals (breakfast, lunch and dinner), a fact that can combine with the non-intake of other groups recommended by the food guide. We observed that food from the dairy group, meats, eggs, oils and fats were found to have a higher intake frequency, explaining in part the low intake of other food groups, since the former kinds of food are often consumed during the main meals. After nutritional counseling for both IG and CG patients, an increase in the frequency of intake of all food groups, especially dairy products, was observed. Anhypothesis for the increase in the intake of milk and dairy products would be their accessibility and for the taste and the eating habits of the Brazilian population. Another possibility would be the ease of dairy as a vehicle for thickening of fluids, a common solution for patients with oropharyngeal dysphagia[27].

Nutrition education, using the food guide for the Brazilian population[11], has been used as a tool in an intervention strategy, as it was noted that its use facilitated learning, thereby orienting the basic principles for a healthy diet. In general, all food groups that make up the object of intervention showed an increase in the proportion of consumers who met the recognized recommendations.

The intervention purpose of this study was nutritional counseling in the early stages of the disease, in order to minimize the reduction and/or inadequate diet during treatment. The guidelines were provided to the patient and caregivers during the quarterly outpatient monitoring. Periodic verification of anthropometric measurements and food intake provided the patients with their own monitoring of nutritional status. Thus, there was greater involvement with the proposed nutritional treatment. It can be inferred that, possibly, nutritional counseling has been an opportunity for appreciation of nutritional monitoring by patients and caregivers.

Another factor that may have positively influenced the compliance of the proposed therapy, was the multidisciplinary treatment offered by specialists in referral centers for patients with ALS. This service differs by providing preventive guidance to patients and caregivers, preparing them for the implications that will arise with disease progression. The multidisciplinary team aims for the ultimate goal of improving the quality of life of these individuals[27, 28]. The results of this study reinforce the importance of early nutrition counseling as part of the treatment of patients with ALS.

Under the conditions of this study, it can be concluded that: patients present with early reduction in body weight, in lean mass and body fat measurements; the change in the nutritional status occurs regardless of nutritional adequacy characterizing the hypermetabolic state of the disease; the food guide adapted for the Brazilian population, as a nutrition education tool, has been effective in increasing the frequency of consumption of food groups.

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