doi:10.4181/RNC.2008.16.194 original

Severity and Functional Ability Scale for Amyotrophic Lateral Sclerosis patients: preliminary results

Escala de Gravidade e Habilidade Funcional para pacientes com Esclerose Lateral Amiotrófica: Resultados Preliminares

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SUMMARY

Introduction. Amyotrophic lateral sclerosis (ALS) is a progressive and fatal illness that affects the neurons of the pyramidal tracts and the anterior horn of the spinal cord. Many evaluations methods have been proposed in order to supply better follow-up information of patients as well as improved management of secondary complications. We present, in this study, a new instrument for clinical and rehabilitation follow-up of patients with ALS. **Method.** We evaluated 96 consecutive patients with diagnosis of ALS, in the University Hospital Antonio Pedro and in the Institute of Neurology Deolindo Couto through the Severity and Functional Ability Scale. **Results.** This preliminary data allowed us to delineate a 5 domain scale that measure 1) muscle strength myotome specific, 2) functional abilities, 3) swallowing function, and 4) breathing, and 5) disease stage severity. Clinical features and functional manifestations of ALS patients were heterogeneous regarding to the most frequent clinical complications and independence levels. **Conclusion.** These preliminary results suggest that our 5 domain scale is simple, applicable, not time consuming of, as well as easily reproducible regarding clinical course and prognosis of patients with ALS. Our pilot study grants the next step of our research that includes accuracy, internal validity, reliability, factorial analysis and other needed formal methodological and statistical procedures.

Keywords: Amyotrophic Lateral Sclerosis. Scales. Neuromuscular Diseases.

Citation: Orsini M, de Freitas MRG, Nascimento OJM, Mello MP, Domingues CA, Lima JMB, Pernes M, Gress CH, Oliveira ABS, Reis CHM, Júnior W, Quintanilha G, Cavalcanti EB, Carvalho LBC. Severity and Functional Ability Scale for Amyotrophic Lateral Sclerosis patients: preliminary results.

RESUMO

Introdução. A esclerose lateral amiotrófica (ELA) é uma doença progressiva e fatal que afeta os neurônios do feixe piramidal e da ponta anterior da medula espinhal. Inúmeras avaliações têm sido propostas no sentido de fornecer um melhor acompanhamento dos indivíduos e gerenciamento das complicações secundárias. Apresentamos, no presente estudo, um novo instrumento para acompanhamento clínico e reabilitativo de pacientes com ELA. **Método.** Avaliamos 96 pacientes consecutivos com diagnóstico de ELA, no Hospital Universitário Antonio Pedro e no Instituto de Neurologia Deolindo Couto com a Escala de Gravidade e Habilidade Funcional. **Resultados.** Estes dados preliminares permitiram delinear uma escala de 5 domínios que mensuram 1) força muscular em miótomos específicos, 2) habilidades funcionais, 3) deglutição 4) respiração, and 5) estágio de gravidade da doença. As características clínicas e manifestações funcionais dos pacientes com ELA foram heterogêneas em relação as complicações mais freqüentes e ao nível de independência funcional. **Conclusão.** Estes resultados preliminares sugerem que nossa escala de 5 domínios é simples, de fácil aplicabilidade, não demorada, assim como facilmente reprodutível a respeito do curso clínico e do prognóstico dos pacientes com ELA. Nosso estudo piloto garante a etapa seguinte de nossa pesquisa que inclui a exatidão, validade interna, a confiabilidade, análise fatorial e outros procedimentos metodológicos e estatísticos formais necessários.

Unitermos: Esclerose Lateral Amiotrófica. Escalas. Doenças Neuromusculares.

Citação: Orsini M, de Freitas MRG, Nascimento OJM, Mello MP, Domingues CA, Lima JMB, Pernes M, Gress CH, Oliveira ABS, Reis CHM, Júnior W, Quintanilha G, Cavalcanti EB, Carvalho LBC. Escala de Gravidade e Habilidade Funcional para pacientes com Esclerose Lateral Amiotrófica: Resultados Preliminares.

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Recebido em: 01/08/08
Revisado em: 02/08/08 a 03/09/08
Aceito em: 04/09/08
Conflito de interesses: não

INTRODUCTION

Amyotrophic lateral sclerosis (ALS), known in UK as motor neuron disease, is a devastating illness for patients, relatives, and caregivers. It is a progressive disorder that involves degeneration of the motor system at all levels[1].

At present, the use and the value of standardized scales in the evaluation and follow up of patients with deficiencies/incapacities caused by neurological illnesses is consensus among medical societies and rehabilitation services. Health professionals have sought to systematically evaluate some aspects of the health of their patients in order to define specific goals of treatment and to implement more effective interventions. So, they have been introduced to the practical concept of scales, with the consequent abandonment of subjective evaluations, susceptible to individual parameters of judgment allowing contradictory results[2].

With the constantly increasing availability of research on new therapeutic possibilities for ALS, it has become evident that traditional evaluation would be insufficient to detect some alterations that future proposals of treatment (medical and rehabilitation) could bring for these patients. The great difficulty, at this moment, is the use of an instrument that would allow objective evaluation of the neurological deficit, the level of functional independence and, mainly, that could be applied to such patients in specific periods of their disease activity[3].

Many scales had been formulated, beginning with the voluntary evaluation of muscular strength and indicators of functional evaluation up to tissue samplings of affected areas of the nervous system. Besides progress has been made, no indicator proposed at this time was able to meet the criteria of biological significance, sensibility to the progression of the illness, clear relation with the general prognostic and survival, as well as easy and friendly measurement approach[4].

The main goal of the scales in neuromuscular diseases, as in ALS, is to evaluate objectively the relevant results of the illness and, at the same time, to recognize or to control the deficiencies/incapacities of the patients. The concepts of validity, efficiency, sensitivity, and specificity are essential to find an adequate instrument[5]. In this way, this study presents the partial results of the application of a new clinical and functional instrument for patients with ALS, extracted after cross-section analysis of 40 individuals.

METHOD

We evaluated a total of 96 patients with diagnosis of ALS, 50 registered at the Neurology Outcome Sector of the University Hospital Antonio Pedro (HUAP), Fluminense Federal University – UFF, and 46 at the Institute of Neurology Deolindo Couto (INDC), Federal University of Rio de Janeiro (UFRJ). The patients were seen consecutively according to the arrival in the sector for diagnosis or clinical follow up. All the participants of the study signed the Consent Form before the scale application. The Research Ethic Committee of the Fluminense Federal University approved the study (#138309).

As inclusion criteria, the patients had to have a definite diagnosis of ALS, or a probable diagnosis with or without laboratory support according to criteria of the World Federation of Neurology, revised in the year 2000 (El Escorial) and a range between 30–85 years. Fifty six patients who had presented at least one of the following items were excluded from the study: (1) electroneuromyography demonstrating either motor or sensitivity conduction block; (2) patients with emotional lability or depression that would eventually preclude follow-up; (3) patients with pain, muscular contractions or associated problems that prevented them from going through with the manual muscular test on the date of application of the scale; (4) patients with spasticity greater or equal to 2 on the Modified Ashworth Scale6; (5) patients with associated illnesses.

Assessment

One professional (MAO) applied the scale in the 40 included patients, 25 men (31 to 68 years) and 15 women (32 to 67 years), in the period from March to September 2007, and the results were scored after consensus with two other researchers.

The rooms had been protected against external noises, had systems of adjusted refrigeration and adequate length for an optimal evaluation. The evaluations took an average of 50 minutes (for each patient) and in accordance with the necessity of the individuals, pauses had been supplied.

All the participants had handbooks from HUAP and INDC with complete information related to clinical and evolutive information's about the illness, properly written, in clear and accurate form, dated and signed by each hospitals staff. The handbooks were up to date and the necessary minimum information consisted of: identification of the patient, clinical description, finding on complementary

exams, findings after application of the EL Escorial criteria[7], description of the clinical and laboratorial findings that had allowed the diagnosis of the illness, as well as reports of patients follow-up. The disease classification of the International Classification of Diseases (ICD-10) was also included in the handbooks. The purpose of the instrument was to objectively measure the functional aspects, the muscular strength, and the speech and respiratory functions of patients with ALS (Annex 1). The first stage set out to evaluate the strength in selected myotomes. For such, one key muscle for each spinal segment was established: C5 – biceps brachii; C6 – extensor carpi radialis; C7 – triceps; C8 – fingers flexors; T1 – dorsal and palmar interosseous; L2 – iliopsoas; L3 – quadriceps femoris; L4 – tibialis anterior; L5 – extensor hallucis longus; S1 – ankle plantar flexors. Such muscles had been evaluated in accordance with the criteria of the Medical Research Council[8]. As there is 10 myotomes evaluated in each half of the body and the maximum punctuation for each one of them is 5, the maximum final score possible was 100 (hundred) points and the minimum 0 (zero).

The second part of the scale makes reference to the functional abilities. It consists of 10 items: feeding, dressing, activities that require dexterity, and fine movements, hygiene, to rise from a chair, to carry through activities (work/ social), to ramble or to touch the wheel-chair, carry out changes of position and transfer, carry out functional activities in the erect position; to go up and go down the stairs. Each item was evaluated with score: 0 (zero) – incapable of performing the activity; 5 (five) – capacity of perform the activity, with certain difficulty, being able to refer signs of muscular weakness and abnormal fatigue; 10 (ten) – capacity of carrying through the activity independently. The maximum punctuation is of 100 (one hundred) points and the minimum 0 (zero) points. Depending on the score, the individuals were classified into the following categories: complete dependence (0 points); serious dependence (5–25 points); moderate dependence (30–60 points); mild dependence (65–95 points); and independence (100 points).

The third part of the examination was destined to evaluate swallowing. Patients who possess normal alimentary habits receive the maximum score of 100. Problems of feeding associated with eventual chokings, changes in the food consistency (related to swallowing problems), and the necessity of assistance for feeding, received a score of 75, 50 and 25 points, respectively. A complete dependency requiring enteral or parenteral nutrition was equivalent to 0 (zero).

The fourth part is related to the respiratory function of the patients, being divided in 5 subitems. A score of 100 (one hundred) points signified complete ventilatory independence and therefore normal respiratory function. The score goes from 75 to 25, and the patients can present alterations in the respiratory function varying anywhere from the use of accessory muscles to the need of partial ventilatory support. In the last stage, score 0 (zero), the patient is completely dependent on ventilatory support.

Finally, the patient was further classified on a scale from 0 to 400 points, into specific categories of disease activity/intensity. Stage 5: Terminal phase of ALS (maximum score obtained in all 4 items of evaluation were equal or less than 80 of the total of 400 points); Stage 4: Serious Dependence (maximum score obtained in all 4 items of evaluation between 81 and 160 points of the total of 400); Stage 3: Moderate Dependence (maximum score obtained in all 4 items of evaluation from 161 to 240 points of the total of 400); Stage 2: Mild Dependence (maximum score gotten in the 4 items of evaluation from 241 to 320 points of the total of 400); Stage 1: Initial phase of ALS (maximum score obtained in all 4 items of evaluation higher than 321 of the total of 400). The result of the total of the four items (muscular strength, functional abilities, swallowing and breathing) defined the stage of the illness and supplied an overall view of the main functions affected by the illness.

Statistics analysis

The model of the study was cross-section. The quantitative analyses had been carried out through software SPSS, widely used in the elaboration of descriptive statistics of the data and modeling. The study sought descriptive statistics of the motor function, the functional abilities, and vital functions and the reliability of the scale in the identification of the intensity of the illness and its consequences on specific functions of daily life.

RESULTS

The patients, at the moment of the diagnosis, had age ranged from 31 to 68 years. The average time elapsed between the first manifestations of the disease and final confirmation of the clinical diagnosis of ALS was 10 months, 17.5% had received the disease

confirmation less than 5 months after the beginning of the first symptoms, 35% between 5 and 10 months, 22.5% between 10 and 15 months, and 25% more than 15 months after the first manifestations.

The majority of our patients (97%) made use of the Rilutek, some associated it with other drugs and/or vitamin supplements. Many patients also make use of vitamin E (61.76%) and natural supplements (14.71%). Other medicaments and rehabilitative therapies had been also used (29.41%).

In our study, 47.5% of the individuals related the onset of the illness in lower limbs and 42.5% in the upper limbs. The remaining pointed beginning of the illness in speech. Complaints related with not-explained stumbling, frequent falls and compromise of the dexterity in the hands were the more related by our sample. Of the total of 40 studied patients, 12 presented similar characteristics to the man-in-the-barrel syndrome. The distribution of muscular weakness in determined myotomes is presented in Table 1.

The patients presented a great number of functional damages in basic and essential daily life activities, 63% out of the patients did not carry out activities related to work or social domain; 35% needed supportive equipment (wheel-chair); 53% were unable to overcome obstacles during gait; 30% were totally dependent in changing body position and transferences (Table 2).

Of the total of our patients, 10% had presented symptoms related to the involvement of the motor neurons of the brain stem, beginning with speech changes (articulation, tone and intensity). Of the total, 57.5% presented problems related to swallowing; 27.5% presenting problems in the feeding with eventual choking, and 30.0% already needing changes in the food consistency, had the difficulties in the chewing and bronchoaspiration (Table 3).

In our study, 62.5% of the patients had respiratory disturbances. Of these 45% present dyspnea to the great and/or medium efforts with/without increase of the respiratory frequency and use of the accessory muscles. 17.5% of our sample already needs periods of non-invasive ventilation and/or oxygen support, relating dyspnea to the small efforts and even to the rest, with increase of the respiratory frequency and use of accessory muscles (Table 4).

The total score, with all parts of the scale, we classified the patients according to the stage of evolution of the ALS, showing that most of patients were in mild dependence and initial phase (Table 5).

DISCUSSION

In this regard, our data is in keeping with previous study results, which have shown a slight predominance of ALS in men[9]. More recent studies, however, show that both genders are equally affected[10]. An increase in the number of women with the illness can be related to improved identification of the at-risk population when compared to previous studies, being recently exposed to an unidentified environmental agent, and/or changes in patients lifestyle, with patients becoming more vulnerable to potential toxins (smoke, among other occupational hazards). New research has found a statistically significant association between gender and the survival of patients with ALS[11], and the results have demonstrated that men possess a more prolonged survival when compared to women.

Table 1. Distribution of the patients regarding muscular weakness in myotomes.

Key muscle for each spinal segment*	Criteria of the Medical Research Council											
	0		1		2		3		4		5	
	L	R	L	R	L	R	L	R	L	R	L	R
C5 – biceps brachii	2	2	5	4	6	5	6	6	12	12	9	11
C6 – extensor carpi radialis	4	3	6	4	8	7	9	11	10	9	3	6
C7 – triceps	5	2	2	3	6	5	8	9	8	9	11	12
C8 – fingers flexors	2	3	7	3	6	3	6	11	10	8	9	12
T1 – dorsal and palmar interosseous	10	7	6	5	6	5	6	9	5	8	7	6
L2 – iliopsoas	6	5	5	5	5	4	8	12	11	9	5	5
L3 – quadriceps femoris	4	3	4	3	2	7	6	4	14	18	10	5
L4 – tibialis anterior	8	9	9	6	2	5	5	5	9	7	7	8
L5 – extensor hallucis longus	13	13	4	6	2	1	4	2	10	12	7	6
S1 – ankle plantar flexors	8	6	4	7	3	5	5	4	3	5	17	13

* Such muscles had been evaluated in accordance with the criteria of the Medical Research Council 10. L = left; R = right.

Table 2. Distribution of the patients regarding functional abilities.

Functional Abilities	Scale		
	0	5	10
Feeding	12	17	11
Dressing	16	14	10
Activities that require dexterity and fine movements	11	20	9
Hygiene	12	13	15
To rise from a chair	15	10	15
To carry through activities (work/ social)	25	11	4
To ramble or to touch the wheel-chair	14	12	14
Carry out changes of position and transfer	12	16	12
Carry out functional activities in the erect position	17	11	12
To go up and go down the stairs	21	10	9

0 = incaple; 5 = with difficulty; 10 = independent

The peak age of ALS onset is from 55 to 75 years. Recently, a modest increase in incidence has been noted, along with a tendency for the condition to present at younger ages[12]. The patients of the present study, at the moment of the diagnosis, had age from 31 to 68 years. The factor age is identified as a strong and reliable predictor in the prognosis of patients with ALS[9]. Younger individuals (up to 40 years), at the moment of the diagnosis and/or at the moment of the first symptoms, had a more favorable prognostic when compared to the older subjects, with age from 40 to 70 years. In the youngest group, 60% survived at least 5 years, whereas only 8% of the eldest patients reached this endpoint. The mechanism underlying this phenomenon is unknown. Younger patients may compensate better for declining motor function and older patients may have fewer motor neurons to compensate. But such explanations provide little insights as to the mechanisms involved. Other studies have also found statistically significant results when associating the age to the prognostic of ALS[13].

In our study, the average time elapsed between the first manifestations of the disease and final confirmation of the clinical diagnosis of ALS was 10 months, such a factor, according to some researches, serves as an excellent prognosis marker for these patients[14]. This delay was negatively related to hazard, i.e., positively related to length of survival, the longer the delay the longer the survival[9]. These results suggest that fast progressing patients tend to seek medical care earlier, whereas those with slower disease progression are referred later on or adapt to the first symptoms for a longer time before they visit a tertiary care facility. Patients with higher economic and education level look for health services earlier than individuals of lower social classes[15].

Many causal and pathogenic hypotheses for ALS have been proposed over the years, ranging from heavy metal toxic effects to environmental and occupational exposures. More recent studies focus largely on excitotoxicity and oxidant stress. The excitotoxic hypothesis has led to the identification of Riluzole, a glutamate-release inhibitor, as the first licensed disease-modifying treatment for ALS[16]. The majority of our patients (97%) made use of the Rilutek®, some associated it with other drugs and/or vitamin supplements. In individuals with ALS rehabilitation is mainly designed to prevent fatigue and contracture, to improve independence and activities for as long as possible, to optimize ability to live with the handicap, and finally to maximize quality of life. The functional impairment must be defined and physical therapy techniques have to be adapted to each patient and reevaluated frequently during the course of the disease[17]. Strengthening or endurance exercises are controversial as exercise may injure remaining muscle fibers and motor neurons. Isometric exercise, short of fatigue, of unaffected muscles is recommended.

Range of motion exercise is critically important for preventing contraction. Assistive and adaptative equipments are essential for maintaining the patient's activities of daily living and home equipment preserves independence. Several orthoses for hand, arm, foot or cervical weakness are available. A wheelchair is an important adaptative device when walking becomes too fatiguing or impossible. The choice for special options and features may require attention. Pulmonary complications are prevented with adapted techniques for bronchic obstruction[18].

Some patients of the present study presented with clinical acute, reactive depression after the diagnosis had made necessary psychological accompaniment. Clinically significant depression should be sought and treated regardless of the stage. Selective serotonin reuptake inhibitors are most frequently

Table 3. Distribution of the patients regarding swallowing evaluation.

Criteria	N	Score
Normal alimentary habits	16	100
Problems of feeding, eventual chokings	12	75
Changes in the food consistency (provoked for difficulty in swallowing)	11	50
Necessity of sounding lead for feeding	0	25
Necessity of enteral or parenteral nutrition – complete dependence	1	0

100 = independence; 75 = choking; 50 = difficulty of swallowing; 25 = help for feeding; 0 = complete dependence

Table 4. Distribution of the patients regarding breathing.

Criteria	N	Score
Patient without necessity of ventilatory support. Normal respitatory function	15	100
Patient presenting dyspnea to the great and/or medium efforts with/without increase of the respiratory frequency and use of the accessory muscles. Not needing support of oxygen and/or non invasive ventilation	18	75
Patient needing periods of not invasive ventilation and/or support of oxygen. Already relates dyspnea to the small efforts or the rest, increase of the respiratory frequency and use of the accessory muscles.	7	50
Patient partially dependent on mechanical ventilation (support mode). Receives a pressure from support to assist its ventilation.	0	25
Patient dependent on mechanical ventilation (Assst/ Control or controlled mode)	0	0

100 = independence; 25–75 = respiratory disorders; 0 = dependent on ventilatory support.

used; however, amitriptyline has its advantages in ALS because this drug may exert favorable effects on other symptoms such as drooling, emotional lability, and sleep disturbances[19].

The first part – The strength in selected myotomes

Motor unit enlargement by sprouting is an important compensatory mechanism for loss of functional motor units during neuromuscular diseases. Perisynaptic Schwann cells at neuromuscular junctions extend branchings that bridge between denervated and reinnervated endplates, and guide axonal sprouts to reinnervate the denervated endplates. The progression is accelerated in motoneuron disease, progressing more rapidly in the post-polio syndrome after prolonged denervation and extremely fast in ALS[20].

In most cases, the patients perceived the onset as weakness in a distal part of one limb. The majority of the patients relate that the illness began with slips, inexplicable falls or problems in activities that demanded dexterity. The disease begins with equal frequency in upper and lower limbs (30 to 40% of cases each) and the clinical picture depends on the area of the nervous system that is damaged[21]. In our study, 47.5% of the individuals related the onset of the illness in lower limbs and 42.5% in the upper limbs. The muscles of the upper arm and shoulder girdles were typically involved later. When an arm is the first limb affected, all this occurs while the thigh and leg muscles seem relatively normal, and there may come a time in some cases when the patient walks about with useless, dangling arms and mimicking the man-in-the-barrel syndrome[22].

Of the total of 40 studied patients, 12 presented similar characteristics to the Man-in-the-barrel Syndrome. Later the atrophic weakness spreads to the neck, tongue, pharyngeal, and laryngeal muscles, and eventually those in the trunk and lower extremities yield to the onslaught of the disease.

The use of the manual muscle testing (MMT) when compared to the maximal voluntary isometric contraction (MVIC) is the recommended option in patients with ALS. Reproducibility between MVIC and MMT are equivalent. However, sensitivity to detect progressive weakness over time favored MMT, an effect largely accounted for by the muscles sampled. MMT was found to be the preferred measure of global strength because of its better Pearson correlation coefficients, essentially equivalent reproducibility, and more favorable coefficient of variation[23]. We opted for the utilization of the scale established by the Medical Research Council for the justifications pointed above.

Over the last decade, motor unit number stimulation (MUNE) methods have been applied with increasing frequency to the study of ALS. MUNE is the ideal tool for the assessment of diseases in which the primary defect is motor unit loss, as it enables quantization and tracking of motor unit numbers while simultaneously gauging countervailing collateral reinervation. These properties make it particularity useful for assessing the effects of both neuroprotective therapies and therapies designed to enhance collateral reinervation, not only in animal models but also in the living patient[24]. More recent efforts have incorporated MUNE into ongoing, multi-center clinical trials as a putative early biological marker, with encouraging results[25].

Table 5. Classification of the patient into specific categories of disease.

Stage	Score (points)	N
Stage 5 – Terminal phase of ALS	0–80	1
Stage 4 – Level of Serious Dependence	81–160	5
Stage 3 – Level of Moderate Dependence	161–240	7
Stage 2 – Level of Mild Dependence	241–320	16
Stage 1 – Initial phase of ALS	321–400	11

The second part – The functional abilities

Knowing the presentation of the patients with ALS in terms of functional dependence allows the rehabilitation services to be structured as to take meet the demands of this population in a more efficient manner. Changes in home environment also are necessary[26]. The patients of this study presented countless functional damages in basic and essential daily life activities. 63% out of the patients did not carry out activities related to work or social activities; 35% needed supportive equipment (wheel-chair); 53% were unable to overcome obstacles during gait; 30% were totally dependent in changing decubitus and transfers. We must presume that such motor disabilities are directly related to corresponding myotomes affected and, therefore points to a massive destruction of the cells of the anterior horn of the spinal cord.

The third part – The swallowing function

Bulbar symptoms are the initial manifestations in 19% to 25% percent of ALS cases[21]. Of the total of our patients, 10% had presented symptoms related to the involvement of the motor neurons of the brain stem, beginning with speech changes (articulation, tone, and intensity). Swallowing and speech disorders are the dramatic consequences of bulbar and pseudo-bulbar syndrome in ALS. Control of dysphagia requires an adjustment in diet consistency. Specific swallowing techniques can help to prevent aspiration[26]. When oral food intake becomes intolerable because of choking, percutaneous endoscopic gastrostomy should be undertaken[27]. Dysarthria can lead to complete loss of oral communication. Speech therapy is helpful initially if progression is slow. Modern computer technology can enable even quadriplegic patients to communicate effectively[28].

Discussion of percutaneous endoscopic gastrostomy tube placement should be initiated early; placement should be presented as a positive option rather than a sign of failure. Many patients and families need time to adjust to this strategy and may be helped by information from patient groups/associations. Modern PEG techniques have low morbidity, and tubes may be placed while the patient can still swallow, to enhance nutrition and avoid exhaustion. Nutritionists and home health nurses can provide essential support to families in managing PEG tubes and achieving adequate nutrition[29]. Unfortunately, the majority of the patients of the present study did not present the financial conditions necessary to meeting the cost of acquiring such equipment and/or services. Counseling was supplied to patients/familiar members.

Most previous studies have reported that disease onset in the limbs rather than in the bulbar muscles were predictive of longer survival time[13]. Bulbar-onset patients may have shortened survival from earlier involvement of respiratory muscles, a higher rate of respiratory complications, malnutrition, and dehydration. Some patients of the present study had presented the Dropped Head Syndrome, disabling them for a more formal communicative process and compromising the spatial orientation. Pathological crying and laughter was also shown to affect outcome. This does not seem to be due to some primary mood disorder but rather to an abnormal manifestation of affect, which can be very disturbing in certain social situations. The symptom responds well to several drugs[30].

The fourth part – The respiratory function

Many patients have a great fear of being unable to breath, particularly at night. Symptoms usually begin with nocturnal dyspnea and orthopnea, and signs of poor nocturnal oxygenation, such as morning headache, frequent walking, nightmares, and daytime sleepiness. Although mucolytics, expectorants, theophylline, antibiotics and oxygen can contribute to respiratory management, ventilatory support should be anticipated and the options explored before clinical respiratory failure develops. Pulmonary consultants and respiratory therapists can help patients and families learn about the many approaches, and the indications for and implications of each alternative. As with gastrostomy, patients need time to consider various options and require objective information. Patient associations and organizations can be very helpful. It is believed that fewer than 5% of patients eventually use long-term ventilatory support[31].

Classification of the disease stages

Many evaluations are proposed for patient follow-up in order to analyze the state of motor function and their consequences on activities of everyday life. Few recommendations can be formulated. Scales must be validaded and be relatively simple to use and generate results for statistical analysis. The choice of which scale to use depends on the clinical objective. Global scales can be used to evaluate progression of the disability. Some of these scales are strongly correlated with patient survival. Other scales are used to classify patients by homogeneous state of severity. The clinician should be aware of these different

Figure 1. Severity and Functional Abilities Scale.

SEVERITY AND FUNCTIONAL ABILITY SCALE (GFSA)
AMYOTROPHIC LATERAL SCLEROSIS

IDENTIFICATION
Patient Name:
Handbook Number: Gender: M ( ) F ( ) Occupation: Diagnostic Date:
Age of the time of the diagnosis: ( ) years; Present moment ( ) years
Begin of the first symptoms: ( ) Upper Limbs ( ) Lower Limbs ( ) Speech ( ) Swallowing
Received diagnosis from the illness how much time of beginning of the first symptoms ( ) months
Carries through rehabilitative treatment: ( ) Y ( ) N How much time: ( ) months
Medicaments treatment for the illness in question: ( ) Y ( ) N
Which: ( ) Rilutek ( ) Vitamin E ( ) Natural supplements ( ) Others

PART 1 – MOTOR ASPECT

Level	Key muscle	Strength Grade	
		L	R
C5	Biceps Brachii		
C6	Extensor Carpi Radialis		
C7	Triceps		
C8	Fingers Flexors		
T1	Dorsal And Palmar Interosseous		
L2	Iliopsoas		
L3	Quadriceps Femoris		
L4	Tibialis Anterior		
L5	Extensor Hallucis Longus		
S1	Ankle Plantar Flexors		

Legend for motor evaluation
0 – Paralysis (movement absent); 1 – Paresis (contraction visible or palpable); 2 – Active movement, does not win the severity; 3 – Active movement, against the severity; 4 – Active movement against some resistance (outside the severity); 5 – Active movement against the ultimate strength of the examiner.

Total Score of Muscle Strength: ( )/100 points
Left Upper Limb ( ): total 25 points; Right Upper Limb ( ): total 25 points;
Left Lower Limb ( ): total 25 points; Right Lower Limb ( ): total 25 points.

PART 2 – FUNCIONAL ABILITIES

Functional Ability	Points
Feeding	
Dressing (upper and lower limbs)	
Activities that require dexterity and hold	
Hygiene	
To get up of a chair	
To carry through activities (work/ social) in the community	
To ramble or to touch the wheel-chair	
Carry out changes of position and transferences	
Carry out functional activities in the foot position	
To go up and go down the stairs	

Total Score of Functional Ability: ( ) / 100 points
Dependence grade: Complete: ( ) Modified Independence ( ) Functional Independence ( )
Grade: Complete Dependence: 0 points; Serious Dependence: 5–25 points; Mild Dependence: 30–60 points; Light Dependence : 60–95 points; Independence: 100 points.

Score of Functional Ability
0 – performance total compromised (incapable you play the activity)
5 – Performance partially compromised (executes the activity with certain difficulty), being able to refer signs of muscular weakness and abnormal fatigue.
10 – Independence (executes the activity normally) with/without signals of fatigue, and muscular weakness only in the end of the activity.
* With regard to the item "To carry through activities (work/social) in the community", a maximum punctuation, 10 points, is supplied when the patient is capable to play it with complete independence in the community. In case that the patient one only carry through such activities in periods of reduced time or with aid of assistants, it receives punctuation 5. Case not to execute more activities (work/social) and to remain confined in domiciliary environment it is graduated with punctuation 0.

Figure 1. Severity and Functional Abilities Scale (cont.)

Specific punctuation for gait or use chair of wheel-chair 0 – Performance total compromised (it does not touch the chair) on account of the degree of muscular weakness and fatigue. 5 – Walk with caregiver aid or equipment of assistance or touches the wheel chair with certain difficulty due to the fatigue and muscular weakness. It does not carry through the task in a long time and for great distances. 10 – Walk of independent form for 50 meters, with/without signals of abnormal fatigue.		
PART 3 – SWALLOWING		
Swallowing		Points
Normal alimentary habits		100
Problems of feeding, eventual choking		75
Changes in the food consistency (provoked for the difficulty in the deglutition)		50
Necessity of sounding lead for feeding		25
Necessity of enteral or parenteral nutrition –complete dependence.		0
Total Score for Swallowing: ( )/100 points Dependence grade: Complete: ( ); Partial ( ); Functional Independence ( ) Grade: Complete: 0 points; Partial: 25–75 points; Independence: 100 points.		
PART 4 – BREATHING		
Breathing		Points
Patient without necessity of ventilatory support. Normal respiratory standard.		100
Patient presenting dyspnea to the great and/or medium efforts with/without increase of the respiratory frequency and use of the accessory muscles. Not needing support of oxygen and/or non-invasive ventilation.		75
Patient needing periods of not invasive ventilation and/or support of oxygen. Already relates dyspnea to the small efforts or the rest, increase of the respiratory frequency and use of the accessory muscles.		50
Patient partially dependent of mechanical ventilation (support mode). He receives a pressure from support to assist its ventilation. PSV (pressure support ventilation) + PEEP (positive end-expiratory pressure).		25
Dependent patient of mechanical ventilation (Assist/Control or controlled mode).		0
Total Score for Breathing: ( )/100 points Dependence grade: Complete: ( ); Partial ( ) ; Functional Independence ( ) Grade: Complete: 0 points; Partial: 25–75 points; Independence: 100 points.		
FINAL SCORE OF SEVERITY AND FUNCTIONAL ABILITY SCALE		
Stage 5 – Maximum score gotten in the 4 items of evaluation are equal or lower than 80 points of the total of 400. Strength ( ) Functional Ability ( ) Breath ( ) Deglutition ( ) – **Terminal phase of ALS**.		
Stage 4 – Maximum score gotten in the 4 items of evaluation between 81 and 160 points of the total of 400. Strength ( ) Functional Ability ( ) Breath ( ) Deglutition ( ) – **Phase of Serious Dependence**.		
Stage 3 – Maximum score gotten in the 4 items of evaluation between 161 and 240 points of the total of 400. Strength ( ) Functional Ability ( ) Breath ( ) Deglutition ( ) – **Phase of Moderate Dependence**.		
Stage 2 – Maximum score gotten in the 4 items of evaluation between 241 and 320 points of the total of 400. Strength ( ) Functional Ability ( ) Breath ( ) Deglutition ( ) – **Phase of Mild Dependence**.		
Stage 1 – Maximum score gotten in the 4 items of evaluation higher than 321 of the total of 400. Strength ( ) Functional Ability ( ) Breath ( ) Deglutition ( ) – **Initial phase of ALS**.		

tools and their relative utility. Knowledge of these scales, their validity, their sensitivity to modification, and their specificity and interpretation pitfalls are prerequisite to good evaluation in daily practice and clinical research[14].

Although the present study supplies only preliminary results related to a new instrument of evaluation of patients with ALS, we believe that such a scale can be useful for professionals involved in the clinical, laboratorial, or rehabilitative care of these individuals. Moreover, our findings may be important in designing new treatment trials that use disease progression as primary endpoint.

The appropriate stratification of eligible patients could result in recruitment of patients at an early stage of the disease, at a time when they might be more likely to respond to mediation, physical therapy as well as to rehabilitative measure in general and more likely to complete the trial.

Other symptoms and secondary complications presented by our patients included: cramps, muscular pain, joint pain, spasticity, pressure ulcers, sialorrhoea, muscular contractures, and other joint abnormalities. The use of specific medications in addition to the physical therapy/nursing services is strategies used for a better management of these problems[32].

CONCLUSION

These preliminary results suggest that our 5 domain scale is simple, applicable, not time consuming of, as well as easily reproducible regarding clinical course and prognosis of patients with ALS.

The Severity and Functional Ability Scale (GFSA) is an option of easy utilization, fast application, and can be used for sequential ambulatory evaluation, allowing the to individualization of the speed of functional compromise. The classification by severity stages allows anticipating conducts. The results supplied by this scale can facilitate to take decision of the multidisciplinary team for the most adequate treatment.

At present, new possibilities of treatment for patients with neuromuscular diseases are related with the principles of the scientific process, time, and efforts of countless researchers regarding therapeutic decision-making based on statistically significant results and outcomes. Considering the great number of clinical studies in the recent past, there are relatively few papers that supply results justifying new therapeutic modalities. Thus, some professionals make use of not-friction therapies and monitor the progress of the patients with empirical data.

Our pilot study grants the next step of our research that includes accuracy, internal validity, reliability, factorial analysis and other needed formal methodological and statistical procedures.

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