

Reverse Nutech Functional Score: A Novel Scoring System to Assess Patients with Amyotrophic Lateral Sclerosis

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Abstract

Background: Amyotrophic lateral sclerosis (ALS), a progressive and fatal neuromuscular disease, is characterized by a progressive loss of the corticospinal tract, brainstem and spinal neurons that further results in muscle atrophy, weakness and respiratory failure. The Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS), a questionnaire based scale has been used to evaluate the condition of the patients. However, several essential parameters go unnoticed with this scale.

Objective: This study aimed at developing a novel scoring system that is numeric and assesses all the possible symptoms of ALS that were not being assessed in ALSFRS.

Materials and Methods: We initiated assessing the lists of recorded symptoms and prepared a single list that included all the possible symptoms associated with ALS. Reverse Nutech Functional Score (RNFS) for ALS is a 27- point positional and directional scoring system that assesses the patient's condition both before and after therapy. To facilitate the conduct of probability based studies, we have converted the scores into numeric values.

Result: Each symptom is scored as (1, 2, 3, 4, 5) that runs in GOOD →BAD direction. These five scores that are equidistant to each other and are continuous lie in a range of (0.5, 5.5).

Conclusion: RNFS appears to be a distinctive scoring system that can be adopted worldwide to assess the patients with ALS.

Keywords: Amyotrophic Lateral Sclerosis (ALS); Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS); Neuromuscular Disease; Reverse Nutech Functional Score (RNFS); Scoring System; Positional Scoring System

Abbreviations

ALS: Amyotrophic Lateral Sclerosis; ALSFRS: Amyotrophic Lateral Sclerosis Functional Rating Scale; RNFS: Reverse Nutech Functional Score

Introduction

Amyotrophic lateral sclerosis (ALS) or Lou Gehrig's disease, a progressive and fatal neuromuscular disease, is featured by a progressive loss of the corticospinal tract, brainstem and spinal neurons that further results in muscle atrophy, weakness and respiratory failure that can lead to death [1,2]. The prevalence of ALS is estimated to range between 2.16 to 3.9 cases per 100,000 persons in Europe and the U.S [3,4].

The Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS), a questionnaire based scale is used to evaluate the condition of patients with ALS. This scale measures functional changes in 10 parameters which include speech, salivation, swallowing, handwriting, cutting food and handling utensils, dressing and hygiene, turning in bed and adjusting bed clothes, walking, climbing stairs and breathing [5]. Later, this scale was revised to improve the assessment of respiratory function and it could measure 12 parameters. However, several essential parameters still go unnoticed with this scale [6].

We have developed a 27-point positional and directional scoring system, Reverse Nutech Functional Score (RNFS) that includes almost all the essential clinical parameters needed to assess the patients with ALS. In this paper, we describe RNFS and compare it with the already existing scale, ALSFRS.

Materials and Methods

The study included patients with ALS who either visited directly or were referred to our institute. The patients either had a previous diagnosis or were diagnosed at our institute with the medical procedure used routinely for ALS. All the symptoms (common or rare) that were found to be present while evaluating the patients were recorded in the diagnostic history. The institute initiated assessing the lists of recorded symptoms and prepared one single list which could be used to evaluate the patients. This list of symptoms is revised from time to time in order to maintain accuracy with literature studies. The RNFS scoring system evaluates a symptom based on five ordinal scores that run in a GOOD → BAD direction. For conducting probability based studies, the scores are converted into numeric values. All the patients who were previously assessed with ALFRS were then assessed with RNFS.

Results

We developed a 27-point scoring system that includes all the symptoms associated with ALS. Appendix 1 presents RNFS scores for all the parameters. RNFS for ALS has been classified into two groups; Group 1 that includes parameters associated with neuromuscular system and Group 2 that includes parameters associated with cognitive behavior. The term not afflicted in ailment (NA) is used to score a parameter that is found not to be associated with the ailment. Not existing (NE) is used to score a parameter for which a case is early in the diagnosis.

Parameter	Description	Scores
Neuromuscular (Area affected: extremity, back, abdomen, chest, face, head)		
Paralysis of upper/lower Extremities	Not Afflicted in ALS	NA
	Not existing	NE
	More than 3 limbs involved	5
	3 limbs involved	4
	2 limbs involved	3
	1 limb involved	2
	Paralysis absent	1
Muscle weakness	Not afflicted in ALS	NA
	Not existing	NE
	More than 3 areas involved	5
	3 areas involved	4
	2 areas involved	3
	1 area involved	2
	No muscle weakness	1
Fatigue	Not afflicted in ALS	NA
	Not existing	NE
	Permanent exhausting fatigue	5
	Fatigue after daily hygiene activities	4
	Fatigue after all normal daily activities	3
	Fatigue after only gentle workout	2
	No fatigue	1

Myalgia	Not afflicted in ALS	NA
	Not existing	NE
	Constant muscle pain in the whole body all day and night despite medication	5
	Pain in limbs (U/L, L/L) and/or trunk despite medication	4
	Low pain only in the limbs or trunk as relieved by medication	3
	Slight muscle pain only after exertion	2
	Myalgia absent	1
Stiffness of muscles	Not afflicted in ALS	NA
	Not existing	NE
	More than 3 areas involved	5
	3 areas involved	4
	2 areas involved	3
	1 area involved	2
	Stiffness absent	1
Fasciculation	Not afflicted in ALS	NA
	Not existing	NE
	More than 3 areas involved	5
	3 areas involved	4
	2 areas involved	3
	1 area involved	2
	No fasciculation	1
Sitting -Balance	Not afflicted in ALS	NA
	Not existing	NE
	No sitting balance at all	5
	Requires maximum external support	4
	Requires minimum external support	3
	Sits with no external support	2
	Sitting Balance normal	1
Standing -Balance with caliper	Not afflicted in ALS	NA
	Not existing	NE
	Cannot stand at all	5
	Stand with caliper + Maximum therapist support	4
	Stand with caliper + Minimum therapist support	3
	Stand independently with caliper / with external support	2
	Stand normally without caliper	1
Walking with Aid	Not afflicted in ALS	NA
	Not existing	NE
	Walker with elbow support (without wheels)	5
	Walker alone (with wheels)	4
	Elbow crutches	3
	Cane	2
	No walking aid required	1
Walking distance	Not afflicted in ALS	NA
	Not existing	NE
	Less than five meters/ cannot walk alone	5

	Can walk up to 25 meters only	4
	Can walk from 50 meters to 100 meters only	3
	Can walk > 100 meters up to < 500 meters only	2
	Can walk normal distance	1
Chewing	Not afflicted in ALS	NA
	Not existing	NE
	Chewing absent/ Alternate feed method	5
	Severe difficulty	4
	Moderate difficulty	3
	Mild difficulty	2
	Normal chewing	1
Swallowing	Not afflicted in ALS	NA
	Not existing	NE
	On peg feeding	5
	Only liquid food by mouth	4
	Only pureed food by mouth	3
	Semi solid food by mouth	2
	Swallowing is normal	1
Speech	Not afflicted in ALS	NA
	Not existing	NE
	Dependent on alternate communication system	5
	Dysarticulated and cannot be understood	4
	Dysarticulated but can be understood	3
	Slurred but still understandable	2
	Normal speech	1
Breathing difficulties	Not afflicted in ALS	NA
	Not existing	NE
	Very severe	5
	Severe	4
	Moderate	3
	Mild	2
	Breathing difficulties absent	1
SPO ₂ (while awake at rest)	Not afflicted in ALS	NA
	Not existing	NE
	< 88 %	5
	(90 to 88) %	4
	(93 to 91) %	3
	(96 to 94) %	2
	> 96 %	1
Sensation- Deep	Not afflicted in ALS	NA
	Not existing	NE
Numbness	Not afflicted in ALS	NA
	Not existing	NE
	More than 3 areas involved	5
	3 areas involved	4
	2 areas involved	3

	1 area involved	2
	Numbness absent	1
Cognitive and others		
Short-term memory	Not afflicted in ALS	NA
	Not existing	NE
	No short-term memory at all	5
	Severe short- term memory loss	4
	Moderate short- term memory loss	3
	Mild short-term memory loss	2
	Memory becomes normal	1
Orientation (Time, Place, Person, Situation)	Not afflicted in ALS	NA
	Not existing	NE
	No orientation for all 4 categories	5
	3 of the 4 categories affected	4
	2 of the 4 categories affected	3
	1 of the 4 categories (mostly time) affected	2
	Orientation assumed normalcy	1
Depression	Not afflicted in ALS	NA
	Not existing	NE
	Severe depression with suicidal tendencies	5
	Severe depression without suicidal tendencies	4
	Moderate depression	3
	Mild depression	2
	No depression	1
Irritability	Not afflicted in ALS	NA
	Not existing	NE
	Most of waking hours	5
	Only if in company with people	4
	Only if in company with certain people	3
	Only Off and On	2
	Irritability absent	1
Sleep Disorder – Hypersomnia	Not afflicted in ALS	NA
	Not existing	NE
	> 13 hours sleep per day	5
	11 to 13 hours sleep per day	4
	9 to 11 hours sleep per day	3
	8 to 9 hours sleep per day	2
	Hypersomnia absent	1
Sleep Disorder – Hyposomnia	Not afflicted in ALS	NA
	Not existing	NE
	No sleep despite sleeping medicines	5
	< 4 hours sleep with medicines	4
	4 to 6 hours sleep with medicines	3
	6 to 8 hours sleep with medicine	2
	Hyposomnia absent, <i>i.e.</i> normal sleep with no medicines	1

Hand functions - Gross	Not afflicted in ALS	NA
	Not existing	NE
	Unable to do	5
	Need maximum assistance	4
	Need moderate assistance	3
	Need minimum assistance	2
	Total independence	1
Hand functions - Fine	Not afflicted in ALS	NA
	Not existing	NE
	Unable to do	5
	Need maximum assistance	4
	Need moderate assistance	3
	Need minimum assistance	2
	Total independence	1
Calipers for standing	Not afflicted in ALS	NA
	Not existing	NE
	Trunk-hip-knee-ankle-foot orthosis (THKAFO)	5
	Hip-knee-ankle-foot orthosis (HKAFO)	4
	Knee-ankle-foot-orthosis (KAFO) + / Shanon Brace (brace around the waist with handles on the side)	3
	Ankle-foot-orthosis (AFO) + / Knee Brace / Shanon Brace	2
	Without AFO / Knee Brace / Shanon Brace	1

Appendix 1: Reverse Nutech Functional Score for ALS.

In RNFS, the five ordinal scores (1, 2, 3, 4, 5) run in the direction 1 → 5, i.e., GOOD →BAD. These five scores that are equidistant to each other and are continuous lie in a range of (0.5, 5.5). To conduct probability based studies, which requires a range of (-1, 1) or (0, 1), we have converted the scores into numeric values. This configuration can be used universally for one symptom. We have used the polynomial smoothing and graphical methods to derive an equation for converting categorical scores into numeric scores as: $Y_n = 0.096 \times (Y_c + 0.5) - 0.166$, where Y_n = numeric score and Y_c = categorical score.

Table 1 shows the conversion of five/three categorical scores (0.5- 5.5) in to five/three numeric scores in the range (0, 1).

No. of Scores	Numeric (Y_n)	Categorical Scores (Y_c)				
		5	4	3	2	1
5	Score	0.122	0.310	0.500	0.690	0.89
	Range	0 - 0.241	0.241 - 0.379	0.379 - 0.621	0.621 - 0.759	0.759 - 1.00
3	Score	0.167	0.500	0.833	-	-
	Range	0 - 0.333	0.333 - 0.667	0.667 - 1.00	-	-

Table 1: Conversion Table from Categorical Scores to Numeric Range for RNFS.

Discussion

The symptoms presented by ALS in its early stages are similar to several other neuronal disorders. Thus, diagnosing the possibility of the condition has to be done carefully [1]. Several tests are available that are used to diagnose the presence of ALS, including electromyography (EMG) and nerve conduction study (NCS) [7,8]. Blood and urine tests are also available but some infectious diseases, like human T-cell leukemia virus (HTLV), human immunodeficiency virus (HIV), polio, Lyme disease and West Nile virus may also manifest symptoms similar to ALS. It has been established that no single test can be relied upon to confirm the diagnosis of ALS. Thus, the physicians rule out the diagnosis of ALS based largely upon the symptoms and signs and a series of diagnostic tests [1].

Several functional rating scales based on symptoms have been developed to evaluate the patient with ALS including the Norris scale, the ALS severity scale, the Appel ALS rating scale and the ALFRS [9]. ALFRS-revised (ALFRS-R) scale measures 12 activities, each rated as 0 to 4 in the direction of bad to good. This revised ALFRS-R scale also measures dyspnea, orthopnea, and the need for ventilatory support in addition to the other measured activities. It has been demonstrated to possess strong construct validity [6]. However, this scale does not measure several parameters that seem to be crucial in assessing the condition of patients with ALS; including numbness, memory, orientation, depression, sensation, sleep - hypersomnia, sleep - hyposomnia, irritability, stiffness of muscles, myalgia, fatigue and paralysis of upper/lower extremities. RNFS includes almost all the parameters that should be assessed to evaluate the patients with ALS.

RNFS scoring system scores each parameter based on 5 scores such as paralysis of upper/lower extremities is scored as 5; if more than 3 areas are involved, 4; if 3 areas are involved, 3; if 2 areas are involved, 2; if 1 area is involved, there is no paralysis. Description and scores of all other parameters are presented in appendix 1. In RNFS, a patient who is normal for all the parameters will have a total score of 27 and the patient in worst condition will have a total score of 135. Whereas, in ALFRS, the patient is considered normal if he scores total 48 and has a worst condition if the score is 0.

Let’s take a hypothetical example of a patient with ALS who is scored with RNFS and ALSFRS both before and after therapy. Table 2 presents the scores of a patient as per RNFS and ALSFRS. Before therapy, the RNFS and ALSFRS scores of this patient are 82 and 12, respectively. After the therapy, an improvement is observed in the patient and the patient scores 39 and 24, with RNFS and ALFRS, respectively. By scoring a patient with RNFS, we are able to assess his condition for several parameters that are not included in ALSFRS. These include myalgia, stiffness of muscles, fasciculation, sitting balance, standing balance, walking aid, walking distance, sensation- deep, numbness, memory, orientation, depression, irritability, sleep disorder - hypersomnia, sleep disorder - hyposomnia, hand functions -gross and hand functions - fine and need of calipers for standing. It is known from previous studies that these neuromuscular and cognitive parameters that are left out in ALSFRS have a significant impact on patient with ALS [5]. Thus, assessing these parameters gives more information about the overall status of the patient. This in turn helps the physician to decide the treatment strategy and note even minor improvement in the parameters. Since, RNFS is a numeric scoring system; the scores can be subtracted or added which makes it more users friendly.

Symptoms	NFS Scores		Symptoms	ALFRS Scores	
	Before Therapy	After Therapy		Before Therapy	After Therapy
Paralysis of upper/lower Extremities	NA	NA	Speech	3	4
Muscle weakness	5	2	Salivation	2	4
Fatigue	NA	NA	Swallowing	0	2
Myalgia	4	3	Handwriting	1	1
Stiffness of muscles	2	2	Cutting food and handling utensils	0	0
Fasciculation	4	1	Dressing and hygiene	0	0
Sitting balance	4	2	Turning in bed and adjusting bed clothes	1	3
Standing balance with caliper	5	4	Walking	0	0
Walking with aid	5	4	Climbing stairs	0	0
Walking distance	5	4	Dyspnea	0	2
Chewing	NA	NA	Orthopnea	2	4
Swallowing	NA	NA	Respiratory support	3	4
Speech	4	3			
Breathing difficulties	NA	NA			
SPO ₂ (while awake at rest)	NA	NA			
Sensation- Deep	4	1			
Numbness	3	1			
Short-term memory loss	5	2			
Orientation	5	2			
Depression	4	1			
Irritability	4	1			
Sleep - Hypersomnia	3	1			
Sleep - Hyposomnia	4	1			
Hand functions -Gross	5	2			
Hand functions - Fine	4	1			
Calipers for standing	3	1			
Total	82	39		12	24

Table 2: A hypothetical example showing RNFS scores and ALFRS scores of a patient before and after therapy.

Our scoring systems seems to be better than ALSFRS in that it scores all the possible symptoms associated with ALS, whereas ALSFRS only measures the activities essential in daily life.

RNFS has only been used at our facility to assess patients. Extended use of this scoring system by other healthcare professionals will help in obtaining the evidences to support its use. We recommend the use of these numeric scores universally as they are pure numbers that lie within the range of (0, 1) and are thus compatible to all variables that distribute likewise including the probability distributions.

Conclusion

RNFS can be considered as a distinctive tool to assess the improvement in patients with ALS. Future studies are needed from other centers to help this scale to be adopted universally.

Disclosure

The author has no conflict of interest.

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