

Psychometric properties of the Russian version of the Edinburgh Cognitive and Behavioral Amyotrophic Lateral Sclerosis Screen

Kutlubaev M.A.¹, Areprintseva D.K.², Radakovic R.^{3,4,5,6}, Pervushina E.V¹.

1- Bashkir State Medical University, Ufa, Russia

2- G.G. Kuvatov Republican Clinical Hospital, Ufa, Russia

3- Faculty of Medicine and Health Sciences, University of East Anglia, Norwich, UK.

4- Euan MacDonald Centre for Motor Neuron Disease Research, University of Edinburgh, Edinburgh, UK.

5- Alzheimer Scotland Dementia Research Centre, University of Edinburgh, Edinburgh, UK

6- Cambridgeshire and Peterborough NHS Foundation Trust, Cambridge, UK.

Abstract

Background: Amyotrophic lateral sclerosis (ALS) is a neurodegenerative condition with observable cognitive and behavioral impairment. The Edinburgh Cognitive and Behavioral ALS Screen (ECAS) is a tool developed specifically for people with ALS (pwALS) and previously translated into Russian, but the psychometric properties have not yet been explored. The aim was to explore and determine the psychometric properties of the Russian-version of ECAS (ECAS-R).

Methods: 56 Russian speaking pwALS, 32 of their caregivers and 26 healthy controls were recruited for the study. They completed the ECAS-R, Patient Health Questionnaire-9 (PHQ-9) and Montreal Cognitive Assessment (MoCA). King Staging System was also utilized. Internal consistency, divergent and convergent validity, as well as culturally-derived cutoff scores of ECAS-R were determined.

Results: The internal consistency of ECAS-R was good (Cronbach's alpha=0.73). Convergent validity was observed though a strong correlation between the ECAS-R and MoCA scores. No correlation between ECAS-R and PHQ-9 were observed in terms of divergent validity. Based on culturally-derived cutoff scores, 64.2% (N=36) of pwALS displayed cognitive impairment, with the most affected cognitive domains as executive function and language. Apathy was the most common behavioral impairment for pwALS followed by a loss of sympathy/empathy.

Conclusions: The ECAS-R is valid and reliable tool for the screening for the cognitive and behavioral impairment in pwALS in Russia, with culturally-derived cutoffs presented.

Keywords: Motor neuron disease, Amyotrophic lateral sclerosis; Cognitive; Behavioral; ECAS.

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Introduction

Amyotrophic lateral sclerosis (ALS) is characterized by motor and non-motor symptoms, the latter includes cognitive and behavioral impairment [1]. The Edinburgh Cognitive and Behavioral Amyotrophic Lateral Sclerosis Screen (ECAS) is brief multidomain assessment originally designed specifically for use with patients with ALS (pwALS) [2]. It has been translated into Russian [3], but its properties have not been explored.

The aim of this study was to determine the psychometric properties of the Russian-language version of the ECAS (ECAS-R).

Material and methods

Participants

PwALS were recruited from academic hospitals in Ufa (Russia) between September 2020 and June 2021. The inclusion criteria were: 1) ALS diagnosis according to the revised El Escorial criteria, 2) ability to communicate including using assistive technologies. Exclusion criteria were: 1) advanced stages of ALS, (i.e., unable to communicate) 2) major psychiatric, medical, or other neurological diseases including dementia. The age, education-level matched controls were recruited as caregivers of the pwALS who were screened through a clinical interview to rule out any significant psychiatric/neurological diseases.

Procedure and measures

PwALS and controls completed the ECAS-R [3]. PwALS also completed Montreal Cognitive Assessment (MoCA) [4] and Patient Health Questionnaire 9 (PHQ-9) [5] as a screen for depression. The ALS Functional Rating Scale-Revised (ALSFRS-R) was employed for the assessment of the functional state for pwALS. Kings Stages were derived through clinical examination. Caregivers completed the ECAS-R behavioral interview.

This study was approved by the local ethical committee of Bashkir State Medical University. All participants provided written informed consent.

Statistical analysis

Non-parametric tests were used as the sample size was relatively small and data distribution was not normal based on inspection of histograms. Mann-Whitney test was used for comparison of continuous variables, and the Chi-square test was used for categorical variables, Cronbach's standardized alpha coefficient was used to quantified internal consistency, reliability of the ECAS-R, with $\geq .70$ classified as good. Associations between the ECAS-R and other measures were explored using Spearman's rho correlation analysis. Culturally-derived ECAS-R cutoffs were defined as two standard deviations below the control mean values. All analyses were performed using SPSS for Windows (version 25, IBM Corp., Armonk, NY, USA), with a significance level threshold set at $p < 0.05$.

Results

Fifty six pwALS, 32 of their caregivers, who completed ECAS-R behavioral interview and 26 healthy controls were recruited for the study (table 1).

Table 1. Demographic and clinical characteristics of pwALS and healthy controls.

	pwALS (n=56)	Controls (n=26)	P
Age (median, interquartile range (IQR))	62.5 (58-68)	61.5 (55-68.5)	0,214
Sex, males/females (N, percent)	35/21 (62.5%/37.5%)	16/10 (61.5%/38.5%)	0,576
Disease onset (bulbar/spinal onset) (N, percent)	13(23%)/43(77%)		
King Staging System (N, percent)			
I	4 (7.1%)		
II	36 (64.3%)		
III	13 (23.2%)		
IV	3 (5.4%)		
ALSFRS-R score (median, IQR)	40 (36-42)		
MoCA score (median, IQR)	24 (21-25)		
PHQ-9 (median, IQR)	8.5 (5-12)		

Validity and reliability

The ECAS-R Cronbach's standardized alpha in pwALS was 0.73. In terms of convergent validity, a positive correlation was found between the MoCA score and the total ECAS-R ($r=0.72$, $p=0.0001$), ALS-specific ($r=0.67$, $p=0.0001$) and ALS-non-specific ($r=0.55$, $p=0.0001$) scores. In terms of divergent validity, no significant correlation between PHQ-9 scores and the total ECAS-R ($r=-0.161$, $p=0.236$), scores of the ALS-specific ($r=-0.152$, $p=0.263$) and ALS-non-specific ($r=-0.172$, $p=0.204$) domains was observed.

Further, statistical analysis revealed a moderate significant correlation between age and total ECAS-R score ($r=0.33$; $p=0.011$), ALS-specific score ($r=0.37$; $p=0.005$), and no correlation between age and ALS non-specific score ($r=0.03$; $p=0.829$). There was also no correlation between ALSFRS-R score and total ECAS-R score ($r=-0.023$; $p=0.865$), ALS-specific score ($r=-0.038$; $p=0.752$), ALS non-specific score ($r=-0.103$; $p=0.451$). Comparative analysis of the severity of cognitive impairment in groups divided by sex, form and stage of ALS did not show any significant differences.

Normative cognitive data and behavioral impairment

Calculated cutoffs and number of pwALS scoring below cutoff on the ECAS-R are shown in table 2.

Table 2. Russian language normative data and proposed cutoffs on the ECAS-R and subdomains (N=26)

	Mean±standard deviation	Cut-off score	Number of pwALS with cognitive impairment
ALS-specific score	85.4±6.2	73	40 (71.4%)
<i>Language</i>	27.0±1.2	25	28 (50.0%)
<i>Verbal fluency</i>	17.8±3.7	11	12 (21.4%)
<i>Executive functions</i>	40.5±2.9	35	47 (83.9%)
ALS non-specific score	26.3±4.7	17	13 (23.0%)

<i>Memory</i>	14.3±3.6	8	15 (26.7%)
<i>Visuospatial</i>	11.9±1.6	9	6 (10.7%)
Total score	111.7±8.9	94	36 (64.2%)

The ECAS-R behavioral interview showed apathy as the most common impairment in 16 pwALS (50%), followed by a loss of sympathy/empathy (N=13, 41%). Behavioral disinhibition was observed in 7 pwALS (22%) followed by hyperorality and altered food preferences (N=4, 13%) and then by perseverative/stereotyped/compulsive/ritualistic behaviors (N=2, 6.5%). No pwALS showed symptoms of psychosis.

Discussion

The ECAS-R demonstrated good internal consistency reliability, and strong convergent validity when compared to the MoCA. Additionally, around two thirds of pwALS were below cutoff for the ECAS-R, calculated based on the control group. The proportion of pwALS with impairment in ALS-specific domains was three times higher than in ALS-nonspecific domains. Executive functions were impaired most often, affecting almost 84% of pwALS. These findings are reflective of the wider cognitive impairment profile in ALS [6].

Frequency of cognitive impairment in our cohort was higher than in previous research, which reported cognitive decline in 42-43% of pwALS [7,8]. This partially could be explained by relatively small sample size and non-normal data distribution. Our results were more closely aligned to the results of the Arabic-language ECAS, which showed that 63% of pwALS were cognitively impaired [9]. In terms of behavioral impairment, apathy and loss of sympathy/empathy were most commonly observed in this study, in keeping with the results of previous research [2,10].

There are some limitations to this study. Due to the control sample size, it may be beneficial for future studies to improve the robustness of culturally-derived cutoff scores for the ECAS-R, using larger sample sizes and more comprehensive methodologies (i.e. ROC analysis).

In conclusion, ECAS-R is valid and reliable tool to screen for cognitive and behavioral impairment in Russian speaking pwALS and their caregivers.

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