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A validity study of the Czech Edinburgh Cognitive and Behavioural ALS Screen (ECAS)

Short report

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Declaration of Competing Interest

Authors Fazio Raffaele, Nina Soósová, Thomas Bak, Evžen Růžička, and Ondrej Bezdicek do not have any disclosures.

Abstract

Background: Edinburgh Cognitive and Behavioural ALS Screen (ECAS) is a brief, standardized assessment of cognitive impairment in amyotrophic lateral sclerosis. *Objective*: We aimed at developing a normative dataset for the ECAS Czech version (ECAS-CZ) to make the assessment applicable for clinical settings. *Method*: Included were 102 healthy participants (mean age: 54.92 ± 14.55 ; education: 14.52 ± 2.44 ; 54:48 females/males) that fulfilled rigorous exclusion criteria including the control of depressive symptoms. *Results*: The internal consistency of ECAS-CZ was acceptable (Cronbach's $\alpha = 0.69$). We found medium correlations ($r \approx 0.5$) of age and education with ECAS-CZ Total score but not with gender. Cut-offs with -2 SD's threshold are presented for the differentiation of cognitive impairment. We report percentile values for ECAS-CZ Total including all subscales. *Conclusion*: We provide normative values for ECAS-CZ that are well suited for the detection of cognitive impairment in clinical settings especially for patients with ALS.

Keywords: cognition, behaviour, ECAS, normative data, screen

Background

Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease affecting upper and lower motor neurons (1). To present days, it is acknowledged that ALS goes beyond only motor problems, insofar as patients usually present behavioural and cognitive impairments overlapping with frontotemporal dementia (2,3). To accommodate the increasing physical disabilities faced by ALS patients, simple-to-use neuropsychology testing minimizing movements is preferred. The Edinburgh Cognitive and Behavioural Amyotrophic Lateral Sclerosis Screen (ECAS) is such a tool that permits an easy and quick assessment of ALS patients with motor and verbal disabilities and it has been validated demonstrating high sensitivity and specificity (4,5). Accordingly, the ECAS has been recently translated and validated in Italian and German consortia (6,7).

Objectives

To the current date, there is no validated Czech tool for the screening of cognitive impairments in ALS. In virtue of a successful pilot study (8), we aim to develop and validate a Czech version of the ECAS including preliminary normative data in healthy adults for use in clinical practice.

Subjects and methods

The Czech versions of the ECAS and the Beck Depression Inventory–Second Edition (BDI-II) were administered to 102 Czech healthy participants. All were recruited from the general community through advertisements. To be eligible for the study, all fulfilled the following inclusion criteria: free of major neurologic or psychiatric disorders, active oncologic illness, and abuse of psychoactive substances. Informed consent in compliance with the revised Declaration of Helsinki, 1987, was acquired from all participants via the Institutional Review Board of the First Faculty of Medicine. Using IBM SPSS Statistics 25 for Windows (IBM

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Corp, Armonk, NY). Continuous variables are expressed as mean, SD, and range, categorical variables as percentages. Pearson's correlation coefficients were used to describe the relationships between different continuous measures, point-biserial correlation coefficient when one variable was dichotomous (women/men). Between-group differences were analysed using the Mann-Whitney U Test.

Results

The mean age of the participants was 54.92 years (SD 14.55; range 20–90), 54 (52.90%) were females, and the mean length of education was 14.52 years (SD 2.44 years; range 9–19). None of the participants was classified as suffering from depression as based on BDI-II (mean 1.69, SD 1.50). Descriptive data on the ECAS are displayed in Table 1. Based on previous research (4) we utilized a -2 SD cut-off threshold for cognitive impairment. Pearson's correlation coefficients of age, education, and point-biserial for gender (women/men), and BDI-II with the ECAS were also calculated (see Table 2). Since age and years of education significantly correlated with the ECAS Total score and its subscales, we stratified the sample accordingly (see Table 1 in Supplementary Material). The ECAS Total score (see Table 2 in Suppl. Mat.) and the Cronbach's alpha coefficient on standardized items for internal consistency was 0.69 [95% CI: .48–.72]. Finally, normative percentile values for age and years of education are available in Table 3 (for the stratified version see Table 3 in Suppl. Mat.).

Discussion

In the current short report, we display the data obtained from the Czech version of the ECAS administered to cognitively normal individuals. Together with a successful pilot study (8), this paper presents the first normative data for the use of the ECAS in the Czech population.

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As suggested by Abrahams et al. (4), we utilized -2 SD from the mean of the ECAS Total score and its subscales to identify possible cognitive impairment. Six participants scored below the threshold of the ECAS Total score, however, five of them were above 60 years old which may influence cognitive performance in the ECAS (6).

The present analysis revealed that ECAS scores were sensitive to the effects of age and education of the participants. Since this finding has been repeatedly observed in the literature concerning the ECAS (5–7). Therefore, we stratified the sample according to these two variables to minimize their influence on differential reasoning. Furthermore, the internal consistency could be regarded as acceptable (< 0.7) considering the heterogeneity of ECAS subscales, however, it is lower than in other validity studies (6,7). Hence, we do not know whether they also included the ALS-Specific and ALS Non-specific subscales. If this would be the case, our study would display a satisfactory Cronbach's alpha coefficient of 0.84. The present study has multiple limitations. The sample size can be considered relatively small and future studies should address this issue by recruiting more healthy people. Additionally, it would be important for a future study to assess the ECAS screening specificity to ALS in comparison with other neurodegenerative diseases in the Czech population to disclose the discriminative validity and classifications accuracy.

In conclusion, we believe that the introduction of the ECAS as a validated screening tool for ALS patients in the Czech Republic is of utmost importance because of its quick and easy assessment properties, even when motor impairments are present. To this, the result of our study is to be considered satisfactory to use the Czech version of the ECAS in a clinical population.

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Table 1. Descriptive statistics of the ECAS with cut-off scores (below -2 SD threshold).

| | Mean (SD) | Range | Cut-off |
|-------------------------------------|----------------|--------|---------|
| Language functions (max 28) | 25.29 (3.21) | 15–28 | 19 |
| Executive functions (max 48) | 37.96 (5.79) | 21–46 | 26 |
| Fluency (max 24) | 17.89 (4.32) | 8–24 | 9 |
| ALS-Specific functions (max 100) | 81.12 (9.87) | 56–96 | 61 |
| Memory functions (max 24) | 16.73 (2.56) | 8–21 | 12 |
| Visuospatial functions (max 12) | 11.85 (0.53) | 9–12 | 11 |
| ALS Non-specific functions (max 36) | 28.42 (2.81) | 20–33 | 23 |
| ECAS Total score (max 136) | 109.42 (11.81) | 76–127 | 86 |

SD: standard deviation; ECAS: Edinburgh Cognitive and Behavioural ALS Screen.

| | Age | Education | Gender | BDI-II |
|----------------------------|--------------|-------------|------------------------|--------------|
| Function | r | r | r _{pb} | r |
| Language | -0.40^{**} | 0.37** | 0.06 | -0.22^{*} |
| Executive | -0.45^{**} | 0.38^{**} | -0.10 | 0.01 |
| Fluency | -0.21^{*} | 0.36** | 0.09 | -0.18 |
| ALS-Specific | -0.49^{**} | 0.50^{**} | -0.00 | -0.14 |
| Memory | -0.25^{*} | 0.32^{**} | -0.03 | -0.07 |
| Visuospatial | -0.31** | 0.19 | 0.00 | -0.27^{**} |
| ALS Non-specific functions | -0.27^{**} | 0.30^{**} | 0.04 | -0.17 |
| ECAS Total score | -0.48^{**} | 0.50^{**} | 0.02 | -0.15 |

Note. BDI-II: Beck Depression Inventory, Second Edition; ECAS: Edinburgh Cognitive and Behavioural ALS Screen; r = Pearson correlation coefficient; $r_{pb} =$ Point-biserial correlation coefficient; *p < 0.05; **p < 0.01.

| | Percentiles | | | | | | |
|----------------------------|-------------|-----|-----|-----|-----|--|--|
| Function | 10 | 25 | 50 | 75 | 90 | | |
| Language | 21 | 25 | 26 | 27 | 28 | | |
| Executive | 30 | 36 | 39 | 42 | 44 | | |
| Fluency | 10 | 16 | 20 | 20 | 28 | | |
| ALS-Specific | 66 | 75 | 84 | 89 | 91 | | |
| Memory | 14 | 15 | 17 | 19 | 20 | | |
| Visuospatial | 12 | 12 | 12 | 12 | 12 | | |
| ALS Non-specific functions | 24 | 27 | 29 | 30 | 32 | | |
| ECAS Total score | 88 | 102 | 113 | 118 | 121 | | |

Table 3. Percentile values on the ECAS.

ECAS: Edinburgh Cognitive and Behavioural ALS Screen.

Supplementary Material

Table 1. Descriptive statistics of the ECAS stratified for education and age with cut-off scores (below -2 SD threshold).

| | | | Lower | education (< 14 | 4 years) | | Higher education (\geq 14 years) | | | | | | | |
|----------------------------|------------------------|--------|---------|-------------------|----------|---------|-------------------------------------|--------------|------------------|---------|-------------------|--------|---------|------------------|
| | Age $\leq 60 \ n = 24$ | | | Age > $60 n = 25$ | | | | $Age \leq$ | 60 <i>n</i> = 41 | | Age > $60 n = 12$ | | | |
| | Mean (SD) | Range | Cut-off | Mean (SD) | Range | Cut-off | <i>p</i> -value* | Mean (SD) | Range | Cut-off | Mean (SD) | Range | Cut-off | <i>p</i> -value* |
| Language functions | 26.18 (1.86) | 21-28 | 22 | 22.16 (4.55) | 15-28 | 13 | < 0.001 | 26.39 (1.69) | 21-28 | 23 | 26.33 (1.44) | 23-28 | 23 | 0.718 |
| Executive functions | 38.17 (5.44) | 21-45 | 27 | 33.40 (5.94) | 21-44 | 22 | 0.003 | 40.71 (3.93) | 31–46 | 33 | 37.67 (6.08) | 22-46 | 26 | 0.074 |
| Fluency | 17.96 (4.48) | 8-24 | 9 | 14.80 (4.44) | 8-22 | 6 | 0.010 | 19.61 (3.01) | 10-24 | 14 | 18.33 (4.58) | 8-22 | 9 | 0.659 |
| ALS-Specific functions | 82.33 (8.63) | 56–95 | 65 | 70.36 (8.51) | 56–90 | 53 | < 0.001 | 86.61 (5.40) | 66–93 | 76 | 82.33 (9.73) | 56–96 | 63 | 0.042 |
| Memory functions | 17.21 (1.96) | 14–21 | 13 | 14.92 (1.73) | 12-18 | 11 | < 0.001 | 17.71 (2.53) | 8-21 | 13 | 16.17 (3.24) | 8-20 | 10 | 0.113 |
| Visuospatial functions | 11.96 (0.20) | 11-12 | 12 | 11.52 (0.96) | 9–12 | 10 | 0.045 | 11.95 (0.22) | 11-12 | 12 | 12.00 (0.00) | 12 | 12 | 0.459 |
| ALS Non-specific functions | 28.75 (2.42) | 22-33 | 24 | 26.44 (2.08) | 22-30 | 22 | < 0.001 | 29.51 (2.70) | 20-33 | 24 | 28.17 (3.24) | 20-32 | 22 | 0.178 |
| ECAS Total score | 111.08 (9.36) | 84–127 | 92 | 96.32 (9.86) | 83–119 | 77 | < 0.001 | 116.12(6.91) | 86–123 | 102 | 110.50(12.08) | 76–126 | 86 | 0.022 |

* Mann-Whitney U test. Bold numbers indicate statistical significance with p < 0.05.

| | ECAS Total |
|----------------------------|------------|
| Subscale | r |
| Language functions | 0.59* |
| Executive functions | 0.84^* |
| Fluency | 0.67^* |
| ALS-Specific functions | 0.98^* |
| Memory functions | 0.65^{*} |
| Visuospatial functions | 0.37^{*} |
| ALS Non-specific functions | 0.68^* |
| | |

Table 2. Pearson correlation of ECAS Total score with ECAS subscales.

ECAS: Edinburgh Cognitive and Behavioural ALS Screen; *p < 0.01.

| | | Lo | wer educa | tion (< 14 | years) | | Higher education (≥ 14 years) | | | | | | |
|----------------------------|------------|-----------------------|-----------|-------------------|--------|-----|-------------------------------------|-------------------------|-----|--------------------|-----|-----|--|
| | | $Age \le 60$ $n = 24$ | | Age > 60 $n = 25$ | | | | Age ≤ 60 n = 41 | | Age > 60 n = 12 | | | |
| | Percentile | | | Percentile | | | Percentile | | | Percentile | | | |
| | 25 | 50 | 75 | 25 | 50 | 75 | 25 | 50 | 75 | 25 | 50 | 75 | |
| Language functions | 26 | 27 | 27 | 18 | 23 | 26 | 26 | 27 | 28 | 26 | 27 | 27 | |
| Executive functions | 37 | 40 | 42 | 28 | 34 | 38 | 39 | 42 | 44 | 36 | 38 | 42 | |
| Fluency | 18 | 19 | 21 | 12 | 16 | 18 | 18 | 20 | 22 | 18 | 20 | 21 | |
| ALS-Specific functions | 80 | 85 | 87 | 64 | 70 | 76 | 84 | 88 | 90 | 81 | 83 | 88 | |
| Memory functions | 16 | 17 | 18 | 14 | 15 | 16 | 16 | 18 | 20 | 16 | 17 | 18 | |
| Visuospatial functions | 12 | 12 | 12 | 12 | 12 | 12 | 12 | 12 | 12 | 12 | 12 | 12 | |
| ALS Non-specific functions | 28 | 29 | 30 | 24 | 27 | 28 | 28 | 29 | 32 | 28 | 29 | 30 | |
| ECAS Total score | 108 | 114 | 116 | 88 | 96 | 103 | 114 | 118 | 120 | 110 | 113 | 115 | |

Table 3. Percentile values on the ECAS stratified for education and age.