**Short communication: Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration**

**Age and education-matched cut-off-scores for the revised German/Swiss-German version of ECAS**

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**Abstract**

The Edinburgh Cognitive and Behavioral ALS Screen (ECAS) has been developed to assess cognition and behavior in patients with Amyotrophic Lateral Sclerosis (ALS). Cognitive impairments of ALS-specific and ALS-nonspecific functions can be determined using cut-off-scores based on performance of healthy subject. However, detailed analysis show that older healthy subjects performed worse than younger ones, whereas highly-educated individuals performed better than those with lower education. As a consequence this study presents new age and educated matched cut-off-scores for the revised German/Swiss-German version of ECAS based on the performance of 86 healthy subjects.

**Introduction**
The Edinburgh Cognitive and Behavioral ALS Screen (ECAS) has been developed to assess cognition and behavior in patients with Amyotrophic Lateral Sclerosis (ALS). Cognitive impairments of ALS-specific (language, verbal fluency, executive function) and ALS-nonspecific functions (memory, visuospatial) can be determined using cut-off-scores based on performance of healthy subjects to distinguish these deficits from other disorders (Abrahams et al., 2014).

The first German/Swiss-German version of ECAS, published in 2014, was revised based on experiences of the initial ECAS validation study (Lulé et al., 2015; for further information see attachment below).

Detailed analysis shows that in the initial German/Swiss-German version of the ECAS older healthy subjects performed worse than younger ones, whereas highly-educated individuals performed better than those with lower education (Lulé et al., 2015). However, age and education adjusted cut-off-scores for the revised German/Swiss-German version of ECAS as used by Lulé et al. (2015) have not yet been introduced.

Usage of the previously published cut-off-scores for the revised ECAS might possibly result in a wrong classification of pathological patterns of cognitive profile in ALS and - as a consequence - in a faulty distinction of ALS specific cognitive impairments from other disorders. We hereby provide age- and education-matched cut-off-scores for the revised German/Swiss-German version of ECAS.

**Methods**

86 healthy subjects (15 from Switzerland and 71 from Germany) completed a revised German or Swiss-German version of the ECAS, respectively. 48 subjects performed the oral and 39 the written form of the ECAS. According to Abrahams et al. (2013) all cut-off-scores were determined by subtracting two standard deviations from the mean score. Referring to Lulé et al. (2015), cut-off-scores were divided as follows: The classification of education was ≤12 for low and >12 years for high education analogous to International Standard Classification of Education – ISCED 2. Additionally, cut-off-scores were defined for <60 and ≥60 years of age, respectively, providing four different age- and education adjusted cut-off groups:

- **Group 1**: <60 years of age and ≤12 years of education (N=20, 10 female, M_{age} = 44.35)
- **Group 2**: <60 years of age and >12 years of education (N=26, 12 female, M_{age} = 47.38),
- **Group 3**: ≥60 years of age and ≤12 years of education (N=20, 11 female, M_{age} = 69.65),
- **Group 4**: ≥60 years of age and >12 years of education (N=20, 8 female, M_{age} = 67.90).
Statistics

Data distribution was tested with Kolmogorov-Smirnov. Results implicated the usage of non-parametric Spearman correlations for calculating the relationship between age and years of education with the ECAS. For the between-group-analysis non-parametric Man-Whitney-U-Tests were applied. According to Abrahams and colleagues, cut-offs were determined by subtracting twice the standard deviation from the mean score (2013). Mean and standard deviation are given in the text. A \( p \)-value of 0.05 was adopted for statistical significance.

Results

Age (\( r = -.18, p < 0.05 \)) and years of education (\( r = .4, p < 0.01 \)) correlated significantly with the ECAS-total-score as well as with the ALS-specific score (\( r = -.2, p < 0.05; r = .4, p < 0.01 \)), respectively.

Subjects with high education performed better in the ECAS total score and almost all subdomains. Moreover, older subjects performed mostly worse than younger ones in ECAS-ALS-specific and in the subdomain executive functions. For the ECAS total score and the other subdomains there was a similar pattern but differences did not reach statistical significance (table 1).

Table 1. Performance of age and education adjusted groups in the revised ECAS.

<table>
<thead>
<tr>
<th>Effect of education</th>
<th>Effect of age</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gr. 1 (Vs Gr. 2)</td>
<td>Gr. 3 (Vs Gr. 4)</td>
</tr>
<tr>
<td></td>
<td>Gr. 1 (Vs Gr. 3)</td>
</tr>
<tr>
<td></td>
<td>Gr. 2 (Vs Gr. 4)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Groups</th>
<th>1 (&lt;60 ≤12)</th>
<th>2 (&gt;60 &gt;12)</th>
<th>3 (&gt;60 ≤12)</th>
<th>4 (&gt;60 &gt;12)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean</td>
<td>SD</td>
<td>Cut-Off</td>
<td>Mean</td>
<td>SD</td>
</tr>
<tr>
<td>--------</td>
<td>-------------</td>
<td>-------------</td>
<td>-------------</td>
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</tr>
<tr>
<td>ECAS total</td>
<td>109.1  7.8  93.5</td>
<td>116.9  8.6  99.7</td>
<td>104.9  13.0  78.8</td>
<td>114.8  8.3  98.2</td>
</tr>
<tr>
<td>ECAS ALSspec</td>
<td>79.1  11.4  56.3</td>
<td>86.9  1.2  84.5</td>
<td>77.4  10.5  56.4</td>
<td>85.6  5.8  73.9</td>
</tr>
<tr>
<td>ECAS ALSnon-spec</td>
<td>27.4  2.5  22.4</td>
<td>30.0  3.7  22.6</td>
<td>27.6  3.7  20.1</td>
<td>29.2  3.3  22.6</td>
</tr>
<tr>
<td>Language</td>
<td>26.4  1.7  23.1</td>
<td>27.7  0.7  26.3</td>
<td>24.9  3.2  18.6</td>
<td>27.4  1.1  25.3</td>
</tr>
<tr>
<td>Fluency</td>
<td>18.1  3.3  11.4</td>
<td>19.4  4.7  9.9</td>
<td>18.7  2.5  13.6</td>
<td>20.1  1.5  17.1</td>
</tr>
<tr>
<td>Executive</td>
<td>39.0  3.2  32.5</td>
<td>41.7  3.4  34.9</td>
<td>34.9  5.0  24.8</td>
<td>38.8  4.4  30.0</td>
</tr>
<tr>
<td>Memory</td>
<td>15.8  2.4  11.0</td>
<td>18.1  3.7  10.8</td>
<td>16.0  3.6  8.8</td>
<td>17.3  3.4  10.6</td>
</tr>
<tr>
<td>Visuospatial</td>
<td>11.6  0.7  10.2</td>
<td>11.9  0.3  11.4</td>
<td>11.6  0.6  10.4</td>
<td>12.0  0.2  11.5</td>
</tr>
</tbody>
</table>

Means, SDs, Cut-Offs are given. Cut-offs were determined by subtracting twice the standard deviation from the mean score (Abrahams et al. (2013); Mann-Whitney-U-test was used for differences between age and education adjusted groups. * \( p<0.05 \); ** \( p<0.01 \); *** \( p<0.001 \); \( p \)-values are reported one-tailed.

Discussion

The present data shows significant effects of age and education on different domains of the German/Swiss-German version of ECAS. Subjects with high educational level usually show significantly better results than those with lower educational level. Regarding the relationship between age and performance in ECAS, lower scores for older subjects were prominent only in some domains (executive and ALS non-specific functions). Thus, the significant association of age and education with ECAS previously shown by Lulé et al. (2015) is further supported.
Conclusion

The results implicate that the ECAS cut-off-scores should take age and education into account and adjusted cut-off scores are suggested, at least for the German/Swiss-German sample but in other samples, age and education might also have an effect.

Acknowledgments

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Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

References


Supplementary Material available online

Revised Version of ECAS:
http://ecas.network/download.html

Information about changes in the revised German/Swiss-German version of ECAS:
http://ecas.network/Germanversion.html

German version of ECAS

The German version of ECAS was established in 2014. It has adopted the English tasks and has been adjusted to the cultural and linguistic specificities. Respectively, a German and a Swiss-German version with marginal distinctions were developed. The German/German-Swiss version of the ECAS was validated in a sample of N=139 ALS patients and N=160 healthy controls. Due to the experiences of the validation study, the ECAS was adjusted (all adjustments of the ECAS are mentioned below) and another validation in a sample of N=80 healthy controls was performed. The standard values for the revised German version of the ECAS can be found further below.

Most important modifications in the German translation of ECAS

- Naming: According to the frequency of objects in the respective culture the images were replaced (igloo instead of bow, hedgehog instead of fox).
- Comprehension: specialization of the formulation Giftstachel (poison sting) instead of Stachel (sting)
- Memory: German version: Fahrrad (bike) – Swiss-German version: Velo (bike); Dieter Keller instead of Douglas Watt
- Language/Spelling: German version: Plätzchen (cookie)/Holz hacken (to chuck wood) - Swiss-German version: Kuchen (cake) /Spalten (to split)
- Fluency: Adjustment of the conversion table - letter G instead of T and increase in time from 60 to 120 and 90 to 180 seconds. Conversion tables were adjusted accordingly.
- Sentence completion: Sarah instead of Sally; Joseph instead of John; restaurant instead of Café
- Recognition: photographer instead of local council

The translation into the different languages should always be performed according to the guidelines.