Neurol Sci (2017) 38:1059–1068
DOI 10.1007/s10072-017-2919-4

ORIGINAL ARTICLE

Edinburgh Cognitive and Behavioural ALS Screen (ECAS)-Italian version: regression based norms and equivalent scores

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Received: 30 March 2016 / Accepted: 14 March 2017 / Published online: 22 March 2017

Abstract Cognitive assessment for individuals with Amyotrophic Lateral Sclerosis (ALS) can be difficult because of frequent occurrence of difficulties with speech, writing, and drawing. The Edinburgh Cognitive and Behavioural ALS Screen (ECAS) is a recent multi-domain neuropsychological screening tool specifically devised for this purpose, and it assesses the following domains: executive functions, social cognition, verbal fluency and language (ALS-specific), but also memory and visuospatial abilities (Non-ALS specific). ECAS total score ranges from 0 (worst performance) to 136 (best performance). Moreover, a brief caregiver interview provides an assessment of behaviour changes and psychotic symptoms usually associated with ALS patients. The aim of the present study was to provide normative values for ECAS total score and subscores in a sample of Italian healthy subjects. Two hundred and seventy-seven Italian healthy subjects (151 women and 126 men; age range 30–79 years; educational level from primary school to university) underwent ECAS and Montreal Cognitive Assessment (MoCA). Multiple linear regression analysis revealed that age and education significantly influenced performance on ECAS total score and sub-scale scores. From the derived linear equation, a correction grid for raw scores was built. Inferential cut-off scores were estimated using a non-parametric technique and equivalent scores (ES) were computed. Correlation analysis showed a good significant correlation between adjusted ECAS total scores with adjusted MoCA total scores ($r_{rho} = 0.669$, $p < 0.0001$). The present study provided normative data for the ECAS in an Italian population useful for both clinical and research purposes.

Keywords Edinburgh Cognitive and Behavioural ALS Screen · ECAS · Normative values · Amyotrophic lateral sclerosis · Cognition

Introduction

Cognitive and behavioural changes in Amyotrophic Lateral Sclerosis (ALS) are now recognized as an integral feature of the disease [1]. Recent studies have underlined prominent changes in executive functions [2, 3], language [4], and social cognition [5–8] (see [1] for review). A small proportion of patients (5–15%) develop a full-blown behavioural variant of frontotemporal dementia (bvFTD) [9]. Despite the increased awareness of ALS as a multisystem disorder, the cognitive status of most ALS patients remains unknown because administration of comprehensive neuropsychological batteries or screening tests may not be feasible for patients with severe difficulties with

Electronic supplementary material The online version of this article (doi:10.1007/s10072-017-2919-4) contains supplementary material, which is available to authorized users.

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speech, writing and drawing, which are commonly implied in neuropsychological assessment [8, 10].

On the basis of the above considerations, the Edinburgh Cognitive and Behavioural ALS Screen (ECAS) have been recently developed for identifying cognitive and behavioural changes in ALS patients [8]. ECAS is a rapid screening test (15–20 min) including an ALS-specific section (100 points) that assesses executive functions and social cognition, verbal fluency and language, and a Non-ALS specific section (36 points) tapping memory and visuospatial abilities. Higher ECAS total scores indicate higher levels of cognitive functioning. Moreover, a brief caregiver interview provides an assessment of behaviour changes and psychotic symptoms usually associated with ALS [11, 12].

ECAS has been translated into several languages [8, 13, 14], but until now no normative study has been performed by means of regression-based method in Italian population. The present study was designed to provide normative data stratified by age, education and sex in a sample of healthy subjects. According to the statistical procedures adopted for most neuropsychological tests carried out on Italian population [15], a correction grid is proposed to take into account the influence of the main sociodemographic variables (gender, age and education) on raw scores and a transformation of adjusted scores into equivalent scores (ES) is proposed to compare the performance on ECAS Italian version with other tests.

Methods

Subjects

We selected for the study subjects who had no past or current history of neurologic or psychiatric diseases (including: alcohol or drug abuse, depression or major psychiatric diseases, brain injury, stroke and dementia). Subjects were also excluded from analysis if their adjusted score at the Montreal Cognitive Assessment (MoCA; [16]) was lower than or equal to 15.5. To avoid enrolment of "supernormal" subjects, we did not exclude individuals with mild hypertension and well compensated type II diabetes. Two hundred and seventy-seven Italian volunteers, distributed across age classes (age range 30–79 years), gender (151 women and 126 men) and education levels (from primary school to university) took part in this study. Mean age of the sample was 55.25 years (SD 13.15), and mean formal education was 10.88 years (SD 4.97). Adjusted MoCA score was 23.94 (SD 3.31). The distribution of the sample for age, education and gender is reported in Table 1. Informed consent was obtained from all participants included in the study.

Table 1 Distribution of the experimental sample according to age, education level, and gender

	Age, years					
	30–39	40–49	50–59	60–69	70–79	Total
Education level						
1–5 years						
Men	–	3	3	4	12	22
Women	–	4	7	8	9	28
6–8 years						
Men	–	15	14	6	4	39
Women	–	13	19	6	9	47
9–13 years						
Men	4	10	16	6	2	38
Women	6	19	13	6	2	46
>13 years						
Men	5	5	6	5	6	27
Women	6	8	5	5	6	30
Total						
Men	9	33	39	21	24	126
Women	12	44	44	25	26	151

Material and procedure

All participants were tested individually in a quiet room. All of them first underwent the Italian version of ECAS, and then completed the Italian version of MoCA [16]. ECAS consists of 15 tasks (see Supplementary Table 1) exploring the following cognitive domains: (1) executive functions and social cognition (score range 0–48), assessed by means of reverse digit span task (max score: 12 points), alternation task (12 points), sentence completion task (12 points), and social cognition task (12 points); (2) verbal fluency (score range 0–24), assessed by verbal fluency task for words beginning with the letter "S" (12 points) and verbal fluency task for 4-letter words starting with the letter "C" (12 points); (3) language (score range 0–28), assessed by means of naming (8 points), comprehension (8 points) and spelling tasks (12 points); (4) memory (score range 0–24), assessed by means of immediate recall (10 points), delayed recall (10 points) and delayed recognition tasks (4 points); (5) visuospatial abilities (score range 0–12), assessed by Dot counting (4 points), cube counting (4 points) and number location tasks (4 points). As foreseen by the original ECAS version, we administered the verbal fluency tasks either in spoken ($n = 156$) or in written version ($n = 121$), to produce a balanced normative dataset. We also computed the Verbal Fluency Index (VFI), as in the original ECAS version: after completing the fluency tasks participants are required to read (spoken version) or to copy (written version) their own responses as fast as possible, so to obtain data from a "motor control"

condition and compute VFI according to the formula [(Total time for test − Time to copy or read words)/Number of correct words generated]. The resulting VFI is then converted in a fluency score by means of the conversion table provided in Appendix Table 7 for Fluency score—letter S, and in Appendix Table 8 for Fluency score—letter C, computed as detailed below. For example, a participant who generated 5 correct words in the spoken version of the Fluency task—letter S, and needed 15 s to read the responses would be assigned a VFI of 9 [(60 − 15)/5)], corresponding to a Fluency score of 4 according to the conversion table.

Statistical analysis

Raw scores achieved by participants on the ECAS and its sub-scores were entered into several linear regression analyses to check the influence of each demographic variable. The effects of age and education level (expressed as years of schooling) were explored after several transformations (e.g., quadratic, cubic, logarithmic, reciprocal) to determine which was the most effective in reducing residual variance. By means of simultaneous regressions the effect of each predictor was weighed within the complete model by partialling out the common variance with the other variables of the model. Bonferroni correction for multiple comparisons was applied to reduce the possibility of type-I error, and variables were included in the model only when the significance level related to each of them was lower than or equal to 0.017. Based on results of the best regression model, regression equation was developed to compute the adjusted score for each subject by adding or subtracting the contribution of the concomitant variables from the original score. After correcting all the raw scores, we considered a non-parametric procedure to evaluate unidirectional tolerance limits that can classify a given score as normal or abnormal with confidence set at 95% [17]. According to the procedure described by Ackermann [18], we have computed separately the outer and inner tolerance limits, while the scores falls between them are defined "borderline scores", because inferentially controlled judgment cannot be expressed. Cut-off value was defined the score at which or below which the probability that an individual belongs to the normal population is less than 0.05 [18]. To allow adjustment of the raw scores of newly tested individuals according to demographic variables, a correction grid was built for any combination of age level (by 10-year steps) and educational level (according to the Italian schooling system). Since the use of adjusted scores is more informative when it is standardized, we have converted adjusted scores into five-point ordinal scale or Equivalent Scores (ES) divided as follows: 0 = scores equal or lower than the outer tolerance limit (5%); 4 = scores higher than the median value of the whole sample; 1, 2 and 3 were obtained by dividing into three equal parts the area of distribution between 0 and 4 [15].

As for the Fluency score, new conversion tables for the VFI using scores obtained by the 277 healthy participants have been provided. For this purpose, we adopted a non-parametric technique [17], analogous to that used for obtaining ES (see above), respecting the same intervals as suggested by ECAS guidelines (www.era.lib.ed.ac.uk/handle/1842/6592). Indeed, we assigned a converted score of 0 to VFI scores above which there should be at least 95% of population (with 95% confidence), and assigned a converted score of 12 to VFI scores lower than the median value; then, the distribution of VFI scores included between 0 and 12 was split into five regions with equal density of observations, assigning them the converted scores of 2, 4, 6, 8, and 10, respectively (as foreseen by ECAS guidelines).

Spearman's non-parametric correlation analysis was performed to investigate the association of adjusted ECAS total score and adjusted MoCA total score. Finally, we computed Spearman's correlation coefficients among the 15 tasks of the battery. Effect size for the correlation coefficient was defined by the following criteria: $r_{rho} < 0.3$ weak; $r_{rho} = 0.3$–0.5 moderate; $r_{rho} > 0.5$ strong [19].

Results

Descriptive statistics of raw ECAS total score and sub-scores are reported in Table 2 (raw scores stratified by age and education ranges, and by gender are reported in Supplementary Table 2).The distribution of ECAS total score and sub-scale scores was skewed, with a longer left tail. The individual regression analyses showed that the square root of education (in years) and the logarithmic transformation of age [log10 (100 − age)] were the most effective in reducing residual variance for all measurements. The influence of age and education was significant for all domains, while the linear effect of the gender was significant only for the visuospatial domain (Supplementary Table 3). For all tasks, except Dot counting, the influence of education level was always significant, whereas age accounted for a significant amount of the variance in reverse digit span, sentence completion, fluency—letter S, naming, comprehension, and cube counting sub-task. Moreover, the linear effect of the gender was significant for the reverse digit span, cube counting, and number location task (Supplementary Table 4).

On these bases, we provide the formulae for exact direct calculation of adjusted ECAS total score and its sub-scores, including the most suitable transformations of independent

Table 2 Descriptive statistics of Edinburgh Cognitive and Behavioural ALS Screen (ECAS) total score and its sub-scores

Task	Mean	SD	Median	Range (min–max)
ECAS total score	102.95	19.54	108	30–131
ALS-specific	76.86	15.97	81	24–97
ALS non-specific	26.09	4.93	27	6–35
Cognitive domain				
Executive	33.78	8.52	36	6–46
Reverse digit span	5.38	1.758	5	0–11
Alternation	8.74	4.26	12	0–12
Sentence completion	8.96	3.29	10	0–12
Social cognition	10.71	2.45	12	0–12
Verbal fluency	18.82	5.91	22	0–24
Fluency score—letter S	9.44	3.32	10	0–12
Fluency score—letter C	9.39	3.36	10	0–12
Language	24.25	3.8	25	11–28
Naming	6.78	1.38	7	2–8
Comprehension	7.55	0.75	8	4–8
Spelling	9.92	2.75	11	0–12
Memory	15.03	4.29	16	0–23
Immediate recall	4.97	2.06	5	0–10
Delayed recall	7.78	2.65	8	0–10
Delayed recognition	2.28	1.23	2	0–4
Visuospatial	11.06	1.36	12	5–12
Dot counting	3.84	0.48	4	0–4
Cube counting	3.39	0.943	4	0–4
Number location	3.84	0.528	4	0–4

SD standard deviation

variables (Supplementary Table 5). For a sample of 277 subjects and using a non-parametric procedure, outer and inner tolerance limits are defined by values corresponding to the 8th and 20th worst observations (for details Supplementary Table 6).

Adjusted ECAS total score and its sub-scores lower than or equal to outer tolerance limit (or cut-off point) can be considered abnormal, values higher than inner tolerance limit indicate a normal performance while intermediate scores indicate a borderline performance, which in our study was obtained by 4.33% of the sample. We computed the correction grid for any combination of age (by 10-year steps) and educational level (according to the Italian schooling system) to allow adjustment of raw scores of newly tested individuals (Table 3 for ECAS total score and its domains and Table 4 for ECAS single tasks).

The score interval corresponding to each ES, the density of observations and the cumulative frequency of each ES are shown in Table 5 for ECAS total score and its domains and Table 6 for single ECAS tasks. As regards to the Dot counting task, we observed that the 97.5% of the sample obtained a raw score of 3 points; because of this floor effect, a raw score below 3 can be taken as the cut-off score suggesting an impaired performance.

For individuals with demographic characteristics not included in the correction grid, it is possible to use the formulae for exact direct calculation of adjusted ECAS total score and its sub-scores shown in Supplementary Table 5, but in this case adjustment factors should be treated with caution. Adjusted MoCA total scores [16] was positively correlated with adjusted ECAS total scores ($r_{rho} = 0.669$, $p < 0.0001$).

Most raw scores on single tasks showed weak to moderate correlations ($r_{rho} = 0.125$–0.477); strong correlations ($r_{rho} = 0.566$) were found between Fluency score—letter S and Fluency score—letter C (Supplementary Table 7).

Discussion

ECAS is a brief assessment designed to screen cognitive functions and behavioural disorders commonly affected in ALS patients (ALS-specific section) [1, 3, 20–22], but ECAS explores also domains not

Table 3 Correction grid for Edinburgh Cognitive and Behavioural ALS Screen (ECAS) total score and its domains, according to age, education, and gender

Education (years)	Age (years)				
	30–39	40–49	50–59	60–69	70–79
ECAS total score					
1–5	15.04*	17.06	19.5	22.54	26.62
6–8	2.2*	4.23	6.66	9.71	13.78
9–13	−6.68	−4.65	−2.22	0.82	4.89
>13	−14.99	−12.96	−10.53	−7.48	−3.41
ALS-specific					
1–5	12.45*	14.01	15.88	18.22	21.35
6–8	2.02*	3.58	5.45	7.79	10.92
9–13	−5.19	−3.64	−1.77	0.56	3.7
>13	−11.95	−10.39	−8.52	−6.18	−3.05
ALS non-specific					
1–5	2.55*	3.03	3.60	4.31	5.27
6–8	0.16*	0.63	1.2	1.92	2.87
9–13	−1.5	−1.02	−0.45	0.25	1.21
>13	−3.05	−2.57	−2	−1.29	−0.33
Executive					
1–5	5.48*	6.3	7.28	8.51	10.16
6–8	0.67*	1.49	2.47	3.7	5.35
9–13	−2.65	−1.83	−0.85	0.37	2.01
>13	−5.77	−4.95	−3.97	−2.74	−1.09
Verbal fluency					
1–5	4.88*	5.23	5.65	6.17	6.87
6–8	1.22*	1.57	1.99	2.51	3.21
9–13	−1.3	−0.95	−0.54	−0.01	0.68
>13	−3.67	−3.32	−2.91	−2.38	−1.68
Language					
1–5	2.03*	2.43	2.92	3.53	4.35
6–8	0.07*	0.48	0.97	1.58	2.4
9–13	−1.27	−0.86	−0.37	0.23	1.05
>13	−2.53	−2.13	−1.64	−1.03	−0.21
Memory					
1–5	2*	2.34	2.76	3.28	3.97
6–8	0.16*	0.51	0.93	1.44	2.14
9–13	−1.10	−0.75	−0.34	0.17	0.87
>13	−2.28	−1.94	−1.52	−1	−0.31
Visuospatial function (male)					
1–5	0.29*	0.43	0.59	0.80	1.08
6–8	−0.25*	−0.12	0.04	0.25	0.52
9–13	−0.64	−0.50	−0.33	−0.13	0.14
>13	−0.99	−0.86	−0.69	−0.48	−0.21
Visuospatial function (female)					
1–5	0.70*	0.83	1	1.21	1.48
6–8	0.14*	0.28	0.45	0.65	0.93
9–13	−0.23	−0.09	0.06	0.27	0.55
>13	−0.59	−0.45	−0.28	−0.08	0.19

Values marked by the asterisk (*) should be taken cautiously because they were obtained by extrapolation from the formulas reported in Supplementary Table 5

usually affected in these patients (ALS Non-specific functions) to differentiate cognitive change characteristic of ALS from other disorders (e.g., Alzheimer disease) [8]. The collection of normative data is the first step to introduce ECAS in clinical practice. For this purpose we took care to enrol participants with a normal score on the Italian version of the MoCA [16] and we excluded subjects without any formal education to avoid the huge effect of illiteracy on neuropsychological measures [22].

The mean of raw ECAS total score of 102.95 (SD 19.54) observed in the present study is lower than that achieved by healthy controls in the original paper (118.48 ± 6.64) [8]. The samples enrolled in the two studies had similar age but differed for educational level, lower in our study (10.88 ± 4.97 years; score range 1–25) than in the original paper (12.28 ± 2.52 years; score range 9–20), reflecting the mean educational level of the general population in our geographic area. It is worth mentioning that the cut-off scores proposed in our study refer to age and education-adjusted scores and not to raw scores, in line with the procedures adopted by Italian normative studies [15]. The present cut-off values are lower than those reported in a recent Italian validation study [14], where a parametric procedure (2 SD below the mean of healthy subjects) has been used to compute cut-off on the basis of raw scores. The discrepancy between the two Italian normative studies can be explained by differences in the features of the samples and by the different statistic procedures to estimate cut-off values.

Nonetheless, by computing age- and education-adjusted scores our study allows clinicians to assess cognitive functioning independently from individual demographic features. Therefore, the adjusted cut-off value for the ECAS total score (67.06), albeit being lower than that proposed in the previous studies [8], takes into account the specific sociodemographic variables in the Italian population.

Beyond ECAS total score, we evaluated the possible effects of sociodemographic variables on ALS-specific functions score, Non-ALS specific functions score, the five

Table 4 Correction grid for tasks including in the Edinburgh Cognitive and Behavioural ALS Screen (ECAS), according to age, education, and gender

Education (years)	Age (years)				
	30–39	40–49	50–59	60–69	70–79
Reverse digit span (male)					
1–5	0.79*	0.94	1.12	1.35	1.65
6–8	−0.13*	0.01	0.2	0.42	0.73
9–13	−0.77	−0.62	−0.44	−0.21	0.08
>13	−1.37	−1.22	−1.04	−0.81	−0.51
Reverse digit span (female)					
1–5	1.3*	1.46	1.64	1.86	2.17
6–8	0.38*	0.53	0.71	0.94	1.24
9–13	−0.26	−0.11	0.07	0.29	0.6
>13	−0.86	−0.71	−0.52	−0.3	0
Alternation					
1–5	3.1*	3.1	3.1	3.1	3.1
6–8	1.14*	1.14	1.14	1.14	1.14
9–13	−0.21	−0.21	−0.21	−0.21	−0.21
>13	−1.48	−1.48	−1.48	−1.48	−1.48
Sentence completion					
1–5	1.2*	1.5	1.85	●	●
6–8	−0.04*	0.25	0.60	1.05	1.65
9–13	−0.91	−0.61	−0.25	0.19	0.78
>13	−1.72	−1.42	−1.06	−0.61	−0.02
Social cognition					
1–5	1.62*	1.62	1.62	1.62	1.62
6–8	0.59*	0.59	0.59	0.59	0.59
9–13	−0.11	−0.11	−0.11	−0.11	−0.11
>13	−0.77	−0.77	−0.77	−0.77	−0.77
Fluency score—letter S					
1–5	2.21*	2.49	●	●	●
6–8	0.35*	0.63	0.96	1.38	1.94
9–13	−0.92	−0.64	−0.31	0.1	0.66
>13	−2.12	−1.85	−1.51	−1.09	−0.53
Fluency score—letter C					
1–5	1.62	1.62	1.62	1.62	1.62
6–8	0.59	0.59	0.59	0.59	0.59
9–13	−0.11	−0.11	−0.11	−0.11	−0.11
>13	−0.77	−0.77	−0.77	−0.77	−0.77
Naming					
1–5	0.25*	0.40	0.58	0.81	1.12
6–8	−0.15*	0	0.17	0.40	0.71
9–13	−0.44	−0.28	−0.10	0.12	0.42
>13	−0.70	−0.55	−0.37	−0.14	0.16
Comprehension					
1–5	0.27*	0.34	0.43	0.54	0.68
6–8	−0.01*	0.05	0.14	0.24	0.39
9–13	−0.22	−0.14	−0.06	0.04	0.19
>13	−0.40	−0.33	−0.24	−0.14	0
Spelling					
1–5	2.26*	2.26	2.26	2.26	2.26
6–8	0.83*	0.83	0.83	0.83	0.83
9–13	−0.15	−0.15	−0.15	−0.15	−0.15
>13	−1.08	−1.08	−1.08	−1.08	−1.08
Immediate recall					
1–5	●	●	●	●	●
6–8	0.62*	0.62	0.62	0.62	0.62
9–13	−0.11	−0.11	−0.11	−0.11	−0.11
>13	−0.81	−0.81	−0.81	−0.81	−0.81
Delayed recall					
1–5	1.12*	1.12	1.12	1.12	1.12
6–8	0.41*	0.41	0.41	0.41	0.41
9–13	−0.07	−0.07	−0.07	−0.07	−0.07
>13	−0.53	-0.53	−0.53	−0.53	−0.53
Delayed recognition					
1–5	0.65*	0.65	0.65	0.65	0.65
6–8	0.24*	0.24	0.24	0.24	0.24
9–13	−0.04	−0.04	−0.04	−0.04	−0.04
>13	−0.31	−0.31	−0.31	−0.31	−0.31
Cube counting (male)					
1–5	0.10*	0.18	0.28	0.40	0.56
6–8	−0.19*	−0.11	−0.01	0.10	0.26
9–13	−0.40	−0.32	−0.22	−0.10	0.06
>13	−0.59	−0.51	−0.41	−0.29	−0.13
Cube counting (female)					
1–5	0.39*	0.47	0.57	0.69	0.85
6–8	0.09*	0.17	0.27	0.39	0.55
9–13	−0.11	−0.02	0.06	0.18	0.35
>13	−0.30	−0.22	−0.12	0	0.15
Number location (male)					
1–5	0.23*	0.23	0.23	0.23	0.23
6–8	0.04*	0.04	0.04	0.04	0.04
9–13	−0.09	−0.09	−0.09	−0.09	−0.09
>13	−0.22	−0.22	−0.22	−0.22	−0.22
Number location (female)					
1–5	0.37*	0.37	0.37	0.37	0.37
6–8	0.17*	0.17	0.17	0.17	0.17
9–13	0.04	0.04	0.04	0.04	0.04
>13	−0.08	−0.08	−0.08	−0.08	−0.08

Values marked by the asterisk (*) should be taken cautiously because they were obtained by extrapolation from the formulas reported in Supplementary Table 5; (●) this test is not applied to these groups

ECAS domains, and the fifteen tasks, and provided normative data not yet available for the Italian version. In this respect, it is worth considering that the Italian version of ECAS standardized by Poletti et al. [14] includes the oral

Table 5 Equivalent scores (ES) for adjusted values on Edinburgh Cognitive and Behavioural ALS Screen (ECAS) total score and its domains

ES	Interval	Cumulative frequency	Density
ECAS total score			
0	≤67.06	8	8
1	67.07–82.4	28	20
2	82.41–95.22	73	45
3	95.23–105.4	139	66
4	>105.4	277	138
ALS-specific			
0	≤48.81	8	8
1	48.82–59.66	28	20
2	59.67–70.85	73	45
3	70.86–78.2	139	66
4	>78.2	277	138
ALS non-specific			
0	≤15.6	8	8
1	15.61–20.97	28	20
2	20.98–24.16	73	45
3	24.17–26.67	139	66
4	>26.67	277	138
Executive			
0	≤18.29	8	8
1	18.30–23.88	28	20
2	23.89–29.76	73	45
3	29.77–34.8	139	66
4	>34.8	277	138
Verbal fluency			
0	≤7.26	8	8
1	7.27–11.44	28	20
2	11.45–16.19	73	45
3	16.20–19.92	139	66
4	>19.92	277	138
Language			
0	≤15.46	8	8
1	15.47–19.95	28	20
2	19.96–22.79	73	45
3	22.80–24.91	139	66
4	>24.91	277	138
Memory			
0	≤4.83	8	8
1	4.84–10.07	28	20
2	10.08–13.36	73	45
3	13.37–15.52	139	66
4	>15.52	277	138
Visuospatial function			
0	≤7.81	8	8
1	7.82–9.42	28	20
2	9.43–10.57	73	45
3	10.58–11.38	139	66
4	>11.38	277	138

spelling task, as in the original paper [8] and in the German–Swiss validation [13]. Since Italian is a strongly transparent language with very regular grapheme-to-phoneme correspondence, the spelling task is used rarely and might fail to detect language deficits, differently from what happens in opaque languages. For this reason future studies on the Italian version of ECAS might consider replacing spelling with other tasks more suitable for exploring the language domain in a transparent language (e.g., a grammaticality judgement task; [23]).

As reported in the original paper [8] and in the German–Swiss and Italian validation studies [13, 14], we observed that age and education were associated with performance on all ECAS sub-scores. Gender was found to influence visuospatial domain only, with females achieving significantly lower scores than males. The finding of a male advantage in this domain appears to be consistent with Italian normative data for the Battery for Visuospatial Abilities [24], and with other studies reporting a male advantage in spatial skills [25, 26]. Moreover, we observed that age, education level, and gender influenced performance on ECAS single tasks to different extent. Taken together, our findings strongly confirmed that the sociodemographic variable must be always taken into account when evaluating raw scores, particularly those obtained by old or less educated individuals [27]. The significant correlation between adjusted ECAS total score and adjusted MoCA total score is consistent with the idea that the two screening tools assess closely related performance reflecting global cognitive status, but the ECAS, unlike MoCA and other cognitive screening tools such as the Addenbrooke's Cognitive Examination Revised ([28, 29] for Italian norms), takes into account motor impairments and is able to distinguish ALS-specific from Non-ALS specific cognitive deficits [13].

In conclusion, the present study provided Italian clinicians with normative data for ECAS total score and sub-scores, allowing to fully exploiting this tool. Since age and education influence performance on ECAS total score and on sub-scores, specific reference values must be considered in research and clinical contexts.

Acknowledgements The authors thank Dr. Barbara Poletti for her contribution in providing the Italian version of Edinburgh Cognitive and Behavioural ALS Screen (ECAS). Moreover, the authors thank Dr. Marianna Vicidomini, Dr. Dalila Biondi, and Dr. Carla Caterino for their contribution in collecting data.

Compliance with ethical standards

Conflict of interest The authors have no conflict of interest to disclose.

Table 6 Equivalent scores (ES) for adjusted values on tasks included in the Edinburgh Cognitive and Behavioural ALS Screen (ECAS)

ES	Interval	Cumulative frequency	Density
Reverse digit span			
0	≤2.71	8	8
1	2.72–3.53	28	20
2	3.54–4.46	73	45
3	4.47–5.34	139	66
4	>5.34	277	138
Alternation			
0	≤0.76	8	8
1	0.77–2.76	35	23
2	2.77–6.97	79	44
3	6.98–9.88	139	60
4	>9.88	277	138
Sentence completion			
0	≤2.15	8	8
1	2.16–4.51	28	20
2	4.52–7.37	73	45
3	7.38–9.76	139	66
4	>9.76	277	138
Social cognition			
0	≤3.57	8	8
1	3.58–7.28	28	20
2	7.29–10.53	73	45
3	10.54–11.57	139	66
4	>11.57	277	138
Fluency score—letter S			
0	≤2.64	8	8
1	2.65–5.51	28	20
2	5.52–7.85	73	45
3	7.86–10.04	139	66
4	>10.04	277	138
Fluency score—letter C			
0	≤1.87	8	8
1	1.88–4.23	28	20
2	4.24–8.32	73	45
3	8.33–9.99	139	66
4	>9.99	277	138
Naming			
0	≤3.52	8	8
1	3.53–4.94	28	20
2	4.95–6.17	73	45
3	6.18–7.04	139	66
4	>7.04	277	138
Comprehension			
0	≤5.72	8	8
1	5.73–6.67	28	20
2	6.68–7.25	73	45
3	7.26–7.77	139	66
4	>7.77	277	138
Spelling			
0	≤3.44	8	8
1	3.45–6.4	28	20
2	6.41–8.61	73	45
3	8.62–10.46	139	66
4	>10.46	277	138
Immediate recall			
0	≤1.55	9	9
1	1.56–2.42	30	21
2	2.43–3.87	75	45
3	3.88–4.84	139	64
4	>4.84	277	138
Delayed recall			
0	≤0.56	8	8
1	0.57–4.24	28	20
2	4.25–6.89	73	45
3	6.90–8.28	139	66
4	>8.28	277	138
Delayed recognition			
0	≤0.16	11	11
1	0.17–0.77	34	23
2	0.78–1.41	77	43
3	1.42–2.36	139	62
4	>2.36	277	138
Cube counting			
0	≤1.08	8	8
1	1.09–2.05	28	20
2	2.06–3.05	73	45
3	3.06–3.66	139	66
4	>3.66	277	138
Number location			
0	≤2.98	9	9
1	2.99–3.18	30	21
2	3.19–3.82	75	45
3	3.83–3.98	139	64
4	>3.98	277	138

Equivalent scores for the Dot Counting task are not provided since 97% of the sample obtained a raw score of 3. For this task a score below 3 has to be considered as pathologic

Appendix

See Tables 7 and 8.

Table 7 Conversion table to convert Verbal Fluency Index (VFI) to a fluency score—letter S

Verbal Fluency Index (VFI) calculation	VFI conversion to score table		
	Spoken (VFI)	Written (VFI)	Score
If spoken	≥14	≥17	0
$VFI = \frac{60\ s - \text{no. of seconds to read aloud words}}{\text{No. of correct words generated}}$	$9.5 \leq VFI < 14$	$13.71 \leq VFI < 17$	2
	$7 \leq VFI < 9.5$	$8.09 \leq VFI < 13.71$	4
If written	$6.11 \leq VFI < 7$	$6.5 \leq VFI < 8.09$	6
$VFI = \frac{120\ s - \text{no. of seconds to copy words}}{\text{No. of correct words generated}}$	$4.42 \leq VFI < 6.11$	$4.5 \leq VFI < 6.5$	8
	$3.6 \leq VFI < 4.42$	$3.41 \leq VFI < 4.5$	10
	<3.6	<3.41	12

Table 8 Conversion table to convert Verbal Fluency Index (VFI) to a fluency score—letter C

Verbal Fluency Index (VFI) calculation	VFI conversion to score table		
	Spoken (VFI)	Written (VFI)	Score
If spoken	≥50	≥104	0
$VFI = \frac{60\ s - \text{no. of seconds to read aloud words}}{\text{No. of correct words generated}}$	$29.5 \leq VFI < 50$	$53 \leq VFI < 104$	2
	$28 \leq VFI < 29.5$	$36.67 \leq VFI < 53$	4
If written	$19.33 \leq VFI < 28$	$27 \leq VFI < 36.67$	6
$VFI = \frac{120s - \text{no. of seconds to copy words}}{\text{No. of correct words generated}}$	$11.6 \leq VFI < 19.33$	$18 \leq VFI < 27$	8
	$9.67 \leq VFI < 11.6$	$14 \leq VFI < 18$	10
	<9.67	<14	12

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