

Abstract

Cognitive changes in patients with ALS often present as deficits in executive functions and changes in language and social cognition. In disease management, cognitive dysfunction may impair patient's decision-making ability.

The Edinburgh Cognitive and Behavioural ALS Screen (ECAS) has been developed to detect the specific profile of cognition and behaviour changes in ALS and to differentiate it from other disorders (1).

It is a 15-20 min screen that includes ALS-specific and nonspecific functions and a carer behaviour screen.

Results

- Data from healthy controls was used to produce abnormality cut-offs (Table 2).
- 24%** of patients scored below the cut-off in the ALS-specific domain and **27%** for the ECAS total score (Figure 2). Behavioural changes were found in **32%** of patients (Figure 3).
- Patients with **bulbar onset** of disease showed significantly more cognitive impairment than those with spinal onset.
- No correlation was found between cognitive impairment and age, education, duration, stage of disease or respiratory status.

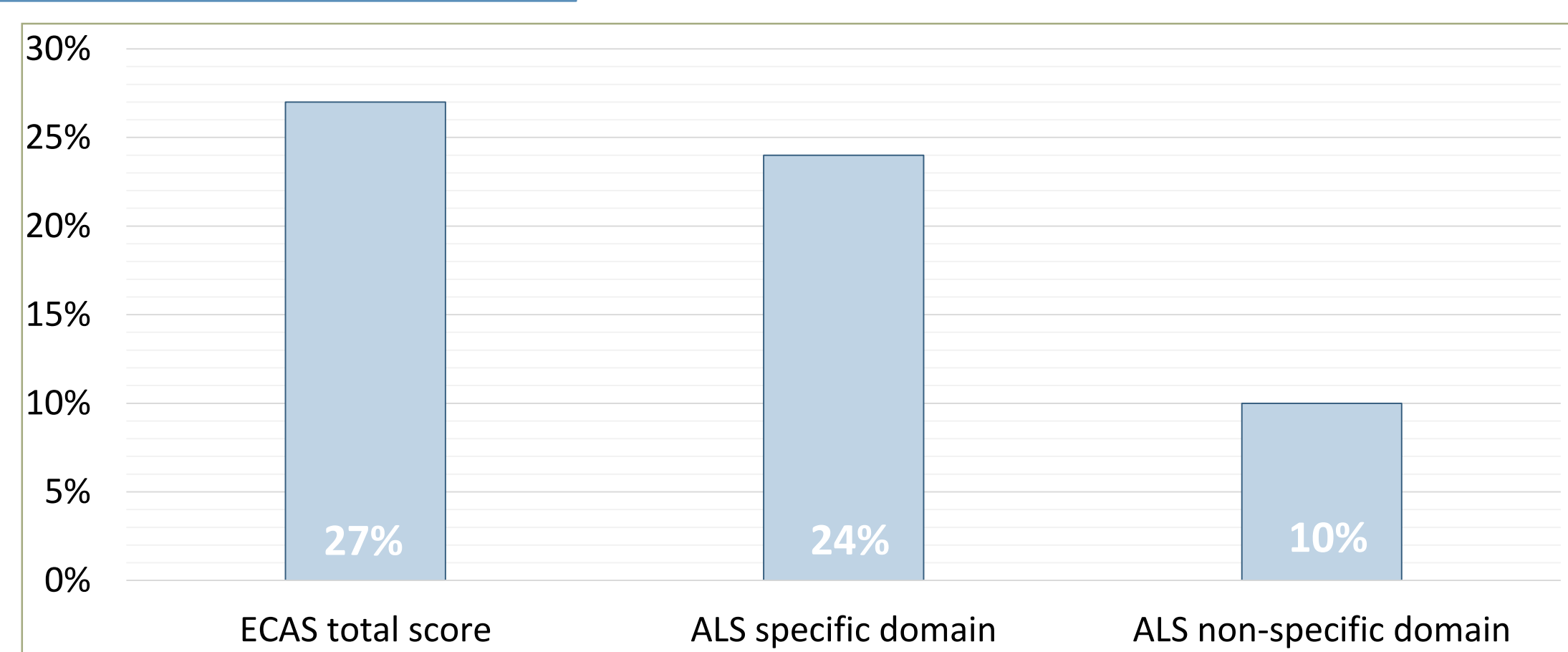


Figure 2. The percentage of patients falling below cut-off score in the ECAS total score, ALS specific domain and ALS non-specific domain.

Methods and Materials

- We **translated** and **adapted** the original version of ECAS (Figure 1).
- Cognitive status of **41 ALS patients** (mean age 63.7, range 41-87 years) was evaluated using ECAS (Table 1).
- 42 healthy controls** (mean age 65.4, range 41-85 years) were evaluated using ECAS (Table 1).
- 32 carers** completed the behavioural interview.

	ALS patients n = 41		Healthy controls n = 42		p
	Mean (SD)	Range	Mean (SD)	Range	
Age	63.7 (9.9)	41-87	65.4 (10.5)	41-85	0.22
Sex (male:female)	24:17	/	25:17	/	/
Years of education	10.7 (1.7)	8-16	11.1 (1.4)	7-16	0.14

Table 1. Characteristics of ALS patients and healthy controls. n: number of participants; p: p value (t-test for independent samples); SD: standard deviation.

	Max	Mean (SD)	Range	Cut-off
ECAS total score	136	103.7 (12.6)	73-121	79
ALS specific functions	100	76.9 (10.1)	51-91	57
ALS non-specific functions	36	26.8 (4.7)	9-32	17
Language	28	26.3 (1.9)	21-28	22
Verbal fluency	24	16.7 (3.4)	10-22	10
Executive functions	48	33.8 (7.8)	17-44	18
Memory	24	15.6 (4.2)	0-20	7
Visuospatial functions	12	11.3 (1.1)	9-12	9

Table 2. Normative data for the Slovenian version of ECAS. Max: maximum score; SD: standard deviation; Cut-off is based on 2 SD from the mean. A score at or below this value indicates impairment.

EDINBURŠKI KOGNITIVNI IN VEDENSKI PRESEJALNI PREIZKUS ZA ALS (EDINBURGH COGNITIVE AND BEHAVIOURAL ALS SCREEN - ECAS) Slovenska različica (2015)

Datum ocenjevanja: _____ Ime in priimek: _____
Število let izobraževanja/končana šola: _____ Datum rojstva: _____
Pokoje: _____ Ime naslov zdravstvene ustanove: _____
Rokovost (desničar/levičar): _____

JEZIK – Imenovanje z besedo

Reče: „Povejte ali napišite, kako pravilno narisano:“

1. Reče: _____ Točke: 0-5
2. Reče: _____ Točke: 0-5
3. Reče: _____ Točke: 0-5
4. Reče: _____ Točke: 0-5
5. Reče: _____ Točke: 0-5
6. Reče: _____ Točke: 0-5

JEZIK – Razumevanje

Reče: „Pokažite sliko, ki kaže:“

1. Nekaj, v čemer lahko letimo _____ Točke: 0-5
2. Nekaj, kar ima plavalno kožico _____ Točke: 0-5
3. Žival, ki pleza po drevmu _____ Točke: 0-5
4. Nekaj, kar uporabljamo za cepljenje drv _____ Točke: 0-5
5. Pnevno orodje _____ Točke: 0-5
6. Nekaj, kar se hrani z oreščki in semeni _____ Točke: 0-5

B. Koritnik, V. Štukovnik, A. Štublar, P. Prunk 1

Figure 1. First page of Slovenian version of ECAS.

Modifications

COMPREHENSION:

According to the comprehension in the respective culture, we replaced the word “sting” with “kleščce” (claws), “webbed feet” with “plavalna kožica” (the skin between the fingers, typical for water birds and ambhobia) and “chopping” with “cepljenje drv” (to split a big log into smaller logs).

MEMORY:

According to the frequency of certain geographical and individual names, we changed “Primrose Woods” to “Kočevski gozd” and “Douglas Watt” to “Janez Novak”.

FLUENCY:

We adjusted the given letters, according to the frequency of the letters in Slovenian – we used „P“ instead of „S“ and „M“ instead of „T“. Conversion tables were adjusted accordingly.

SENTENCE COMPLETION:

We used “Janez” instead of “John” and “Sabina” instead of “Sally”. We changed “local café” into “vaški lokal” (village pub).

Conclusions

The results of the Slovenian version of ECAS are comparable to other published versions (Figure 3).

ECAS is an effective and useful clinical tool that can improve the quality of ALS patient care.

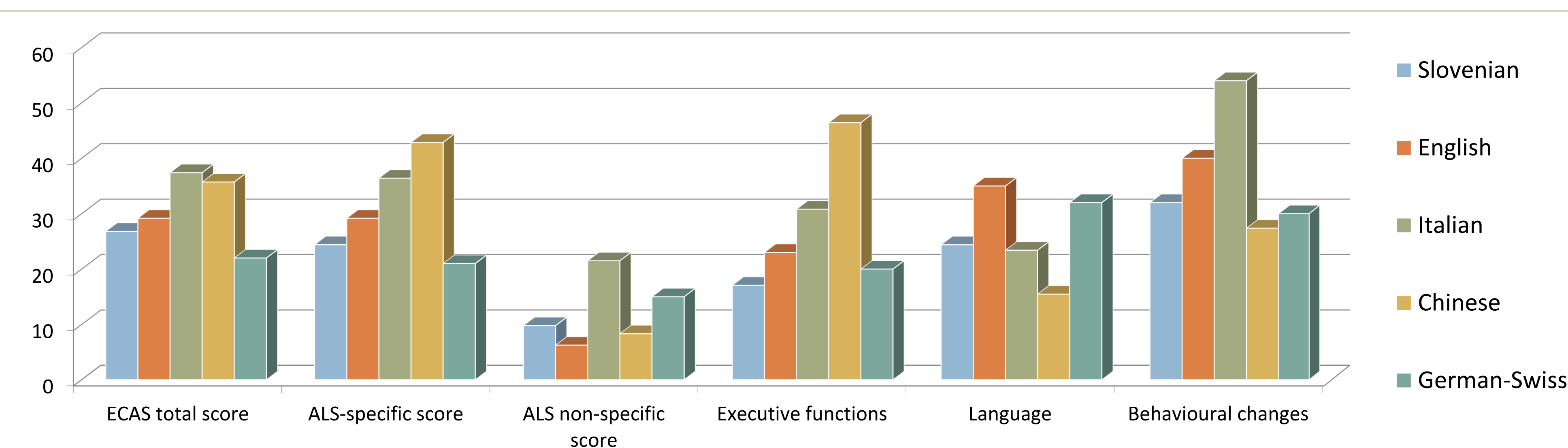


Figure 3. The comparison between percentage of patients with cognitive or behavioural impairments according to ECAS cut-off scores in different language versions (1, 2, 3, 4, 5).

Cognitive changes are, like in other similar studies, most often found in ALS-specific domains. In our study, the most prominent changes have been found in the **Language sub-domain** (24% of patients) that is comparable to the results found by Abrahams et al (35% of patients) and Lule et al (32% of patients).

Contact

Petra Prunk
Ljubljana ALS Centre
Email: petraprunk@gmail.com

Vita Štukovnik
Unit for Neurorehabilitation
Email: vita.stukovnik@kclj.si

Ana Štublar
Ljubljana ALS Centre
Email: bogomila.stublar@gmail.com

Blaž Koritnik
Ljubljana ALS Centre
Email: blaz.koritnik@kclj.si

References

- Abrahams S et al. Screening for cognition and behavioural changes in ALS. *Amyotroph Lateral Scler Frontotemporal Degener* 2014; 15: 9–14.
- Lule D et al. The Edinburgh Cognitive and Behavioural Amyotrophic Lateral Sclerosis Screen: a cross-sectional comparison of established screening tools in a German-Swiss population. *Amyotroph Lateral Scler Frontotemporal Degener*. 2015; 16 (1-2): 16-23.
- Poletti B et al. The validation of the Italian Edinburgh Cognitive and Behavioural ALS Screen (ECAS). *Amyotroph Lateral Scler Frontotemporal Degener*. 2016; 24: 1-10.
- Ye S et al. The Edinburgh Cognitive and Behavioural ALS Screen in a Chinese Amyotrophic Lateral Sclerosis Population. *PLoS One*. 2016; 11(5): 1-11.
- Loose M et al. Age and education-matched cut-off scores for the revised German/Swiss-German version of ECAS. *Amyotroph Lateral Scler Frontotemporal Degener*. 2016; 17: 374-6.