

ECAS-based neuropsychological phenotyping in Amyotrophic Lateral Sclerosis: a retrospective study comparing different algorithms

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Supplementary Table 1. Patients' background and clinical measures

	Total
<i>N</i>	901
Age (years)	63.5 ± 10.99 (20-88)
Sex (male/female)	56%/44%
Education (years)	11.39 ± 4.27 (3-25)
Disease duration (months)^a	19.25 ± 19.27 (1-264)
ALSFRS-R^b	38.77 ± 5.85 (12-47)
ΔFS^c	.83 ± .89 (.03-7)
King's (Stage 1/2/3/4 %)^b	38%/33%/25%/4%
MiToS (Stage 0/1/2/3 %)^b	78%/20%/2%/0%
Genetics (N)^d	
<i>C9orf72</i>	56
<i>SOD1</i>	25
<i>TARDBP</i>	21
<i>FUS</i>	3
<i>OPTN</i>	4
<i>SQSTM1</i>	3
Other	17
ECAS [impaired %]	
Total	98.86 ± 19.05 (24-133) [33%]
ALS-specific	72.41 ± 15.69 (13-98) [33%]
ALS-nonspecific	26.45 ± 4.95 (6-36) [19%]
Language	23.55 ± 3.95 (1-28) [20%]
Naming	6.73 ± 1.4 (0-8) [15%]
Comprehension	7.63 ± 0.74 (0-8) [9%]
Spelling	9.19 ± 2.87 (0-12) [16%]
Fluency	16.49 ± 5.74 (0-24) [20%]
Verbal fluency - S	8.48 ± 3.01 (0-12) [20%]
Verba fluency - C	8.02 ± 3.45 (0-12) [19%]
Executive	32.36 ± 8.5 (6-48) [31%]
Backward digit span	5.26 ± 1.89 (0-12) [1%]
Alternation	8.63 ± 4.28 (0-12) [32%]
Sentence completion	8.9 ± 2.63 (0-12) [25%]
Social cognition	9.57 ± 3.23 (0-12) [11%]
Memory	15.17 ± 4.41 (0-24) [16%]
Immediate recall	5.12 ± 1.98 (0-10) [7%]
Retention score	7.83 ± 2.66 (0-10) [20%]
Recognition	2.22 ± 1.23 (0-4) [12%]
Visuospatial	11.28 ± 1.2 (4-12) [9%]
Number position	3.82 ± 0.57 (0-4) [12%]
Dot-counting	3.87 ± 0.39 (1-4) [1%]
Cube-counting	3.59 ± 0.8 (0-4) [5%]
ECAS-CI	
Total	0.96 ± 1.48 (0-10)
Disinhibition	7%
Socially inappropriate behaviour	3%
Loss of manners/decorum	3%
Impulsiveness	3%
Loss of sympathy/empathy	19%
Diminished response to others' needs/feelings	9%
Diminished social interest	15%
Apathy	33%
Perseveration	11%
Simple repetitive movements	9%
Complex/compulsive/ritualistic behaviour	9%
Altered eating behaviour	7%
Altered food preferences	5%
Binge eating/hyperorality	5%
Psychosis	2%
Suspicion	1%
Delusions	<1%
Hallucinations	<1%

Notes. ALSFRS-R=Amyotrophic Lateral Sclerosis Functional Rating Scale-Revised; ΔFS=progression rate; ECAS=Edinburgh Cognitive and Behavioural ALS Screen; MiToS=Milano-Torino staging system; ECAS-CI=ECAS Carer Interview; ALS=amyotrophic lateral sclerosis; cbn=cognitively and behaviourally normal; ci=cognitively impaired; bi=behaviourally impaired; cbi=cognitively and behaviourally impaired. ^adata available for *N*=854 patients; ^bdata available for *N*=827 patients; ^cdata available for *N*=788 patient; ^ddata available for *N*=720 patients.