

Table R1. Definition and frequency of cognitive impairment based on normative data in the Spanish population.

Measure	Mean (SD) in Mora <i>et al.</i> , 2018	Cut-off (<1.5 SD)	n (%) of ALS* participants impaired
Executive function	35.57 (8.07)	23	8 (26)
Language	25.25 (2.50)	21	7 (23)
Fluency	17.48 (5.02)	9	10 (32)
ALS-specific ECAS	78.36 (12.91)	58	9 (29)
ALSci**	-	-	13 (42)
ALSbi***	-	-	11 (35)
ALSci** and/or ALSbi***	-	-	19 (61)

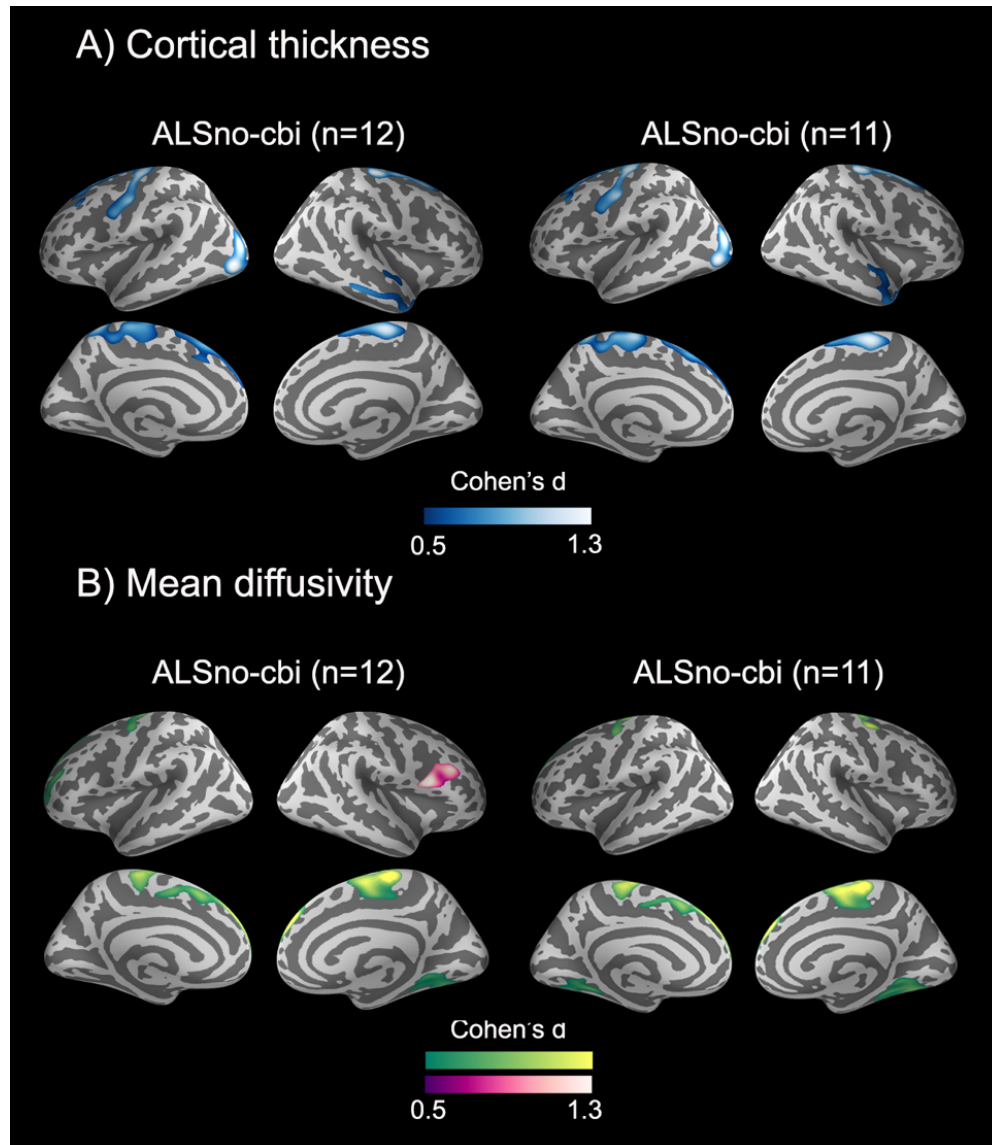
Table R1 - Footnotes: *: 31 ALS participants after excluding 3 PLS and 1 PMA cases.

** : Included ALS participants with at least one abnormal score (< 1.5 SD) on the fluency, language and executive scores of the ECAS.

*** : Included ALS participants with any behavioral impairment (at least 1 behavior feature of Rascovsky criteria)

Key: ALS=Amyotrophic lateral sclerosis; ALSno-cbi = Amyotrophic lateral sclerosis without cognitive or behavioral impairment; ALScbi = Amyotrophic lateral sclerosis with cognitive or behavioral impairment; ALS-FTD = Amyotrophic lateral sclerosis-frontotemporal dementia continuum; SD=standard deviation;

Supplementary figure 1



Supplementary Figure 1 - footnotes: Group comparisons between ALSno-cbi group (left column) and ALSno-cbi group after excluding the participant with *SOD1* mutation (right column) compared to cognitively normal controls for A) cortical thickness and B) cortical mean diffusivity. Regions in blue represent thinner cortex. Regions in green represent higher cortical mean diffusivity and regions in purple represent decreased cortical mean diffusivity. All analyses were adjusted for age, sex, education and MRI

equipment. Only clusters that survived familywise error correction p -value <0.05 are shown.

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