

Table e-7. Demographic and clinical characteristics of *C9orf72*-negative ALS patients stratified for behavior.

	NORMAL		ABNORMAL		p-values		
	Baseline	Follow-up	Baseline	Follow-up	normal vs abnormal	normal	abnormal
					baseline	baseline vs FU	baseline vs FU
n	98	59	51	25			
Follow-up interval (months)		4.92 (4.36-6.08)		5.38 (3.93-6.43)			
Gender, female/male (%)	40/58 (40.8/59.2)	26/33 (44.1/55.9)	13/38 (25.5/74.5)	6/19 (24.0/76.0)	0.094	0.816	1.000
Age at baseline, y	64.89 (58.62-69.50)	63.87 (53.86-69.50)	64.24 (53.17-68.63)	60.95 (52.22-65.65)	0.235	0.490	0.388
Age at onset, y	62.99 (56.04-68.33)	60.92 (52.37-68.09)	62.03 (51.68-66.72)	59.05 (50.47-63.32)	0.304	0.437	0.365
Type, familial/sporadic (%)	7/91 (7.1/92.9)	5/54 (8.5/91.5)	2/48 (3.9/94.1) ¹	2/23 (8.0/92.0)	0.286	0.761	0.467
Site of onset (%)					0.393	0.917	0.875
Bulbar	29 (29.6)	17 (28.8)	11 (21.6)	5 (20.0)			
Spinal	69 (70.4)	42 (71.2)	40 (78.4)	20 (80.0)			
Thoracic/respiratory	-	-	-	-			
Progression rate at baseline	0.43 (0.23-0.79)	0.40 (0.18-0.57)	0.49 (0.25-0.70)	0.33 (0.19-0.58)	0.858	0.187	0.250
Disease duration at baseline, m	13.07 (10.04-21.74)	16.56 (10.74-22.67)	15.51 (12.30-25.93)	15.54 (13.28-24.95)	0.343	0.412	0.774
Cognition,	80/15/3	45/13/1	39/11/1	18/7/0	0.601	0.659	0.661
normal/abnormal/unknown	(81.6/15.3/3.1)	(76.3/22.0/1.7)	(76.5/21.6/2.0)	(72.0/28.0/0.0)			
ALS-FTD-Q	5.5 (2-10)	5 (2-11)	24 (17-30)	24 (19.8-30.8)	<0.001	0.905	0.995
ECAS behavior (0/1/ \geq 2)	48/2/0	23/4/0	4/8/21	2/2/10	<0.001	0.091	0.747

Median (interquartile ranges, IQR) unless otherwise stated. Disease progression rate was calculated as follows: (48-ALSFRS-R)/disease duration (in months). ALSFRS-R=revised ALS Functional Rating Scale, m=months, y=years. ¹missing values n=1. Significant p-values <0.05 are in bold.