

Table e-4. Demographic and clinical characteristics of *C9orf72*-negative ALS patients stratified for site of onset.

	BULBAR		SPINAL		p-values		
	Baseline	Follow-up	Baseline	Follow-up	bulbar vs spinal	bulbar	spinal
					baseline	baseline vs FU	baseline vs FU
n	74	30	192	102	-		
Follow-up interval (months)		5.07 (3.98-6.14)		5.31 (4.34-6.20)	-		
Gender, female/male (%)	26/48 (35.1/64.9)	9/21 (30.0/70.0)	64/128 (33.3/66.7)	35/67 (34.3/65.7)	0.894	0.785	0.968
Age at baseline, y	63.1 (57.2-67.4)	62.5 (59.2-64.6)	63.4 (54.1-69.0)	60.9 (51.5-67.6)	0.986	0.464	0.147
Age at onset, y	61.6 (55.9- 66.0)	61.1 (58.3-63.6)	60.8 (52.9-67.3)	58.6 (49.7-65.2)	0.729	0.445	0.121
Type, familial/sporadic (%)	3/70 (4.1/94.6) ¹	1/29 (3.3/96.7)	11/181 (5.7/94.3)	8/94 (7.8/92.2)	0.237	0.801	0.651
Spinal, arms/legs (%)	-	-	118/74 (61.5/38.5)	69/33 (67.6/32.4)	-	-	0.356
Progression rate at baseline	0.50 (0.33-0.75)	0.41 (0.25-0.64)	0.52 (0.26-0.83)	0.43 (0.19-0.64)	0.711	0.124	0.091
Disease duration at baseline, m	12.7 (9.1-19.9)	12.7 (9.6-16.8)	14.2 (9.2-21.4)	15.4 (10.2-23.0)	0.488	0.116	0.404
ALS-FRS-R at baseline	41 (37-44)	43 (40.3-45)	41 (36.5-43)	42 (38-44)	0.563	0.081	0.113

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.