Supplement table 1: Characteristics of patients with ALS and/or FTD and healthy controls.

	ALS	***PLS***	***PBP***	***F A/L***	***ALS-FTD***	***FTD***	***HC***
	N=611	N=44	N=20	N=38	N=31	N=18	N=49
Age [years]	61.0 ± 12.1	59.4 ± 11.3	71.3 ± 6.1	62.2 ± 11.9	65.8 ± 9.4	62.3 ± 9.2	58.2 ± 13.0
Gender [male/female]	350 m / 261 f	25 m / 19 f	18 m / 2 f	27 m / 11 f	19 m / 12 f	15 m / 9 f	30 m / 19 f
Education [years]	13.3 ± 3.5	13.4 ± 4.5	12.5 ± 3.8	13.7 ± 3.4	11.7 ± 2.5	12.6 ± 2.5	14.6 ± 3.4
Site of onset [bulbar/spinal/bulbar&spinal]	168 b 413 s 19 b&s 11 n.a.	7 b 34 s 1 b&s 3 n.a.	5 b 11 s 5 n.a.	38 s	9 b 8 s 8 n.a.	-	-
Family history [sporadic/familial]	537 s 33 f 41 n.a.	38 s 3 f 3 n.a.	17 s 3 n.a.	38 s	29 s 1 f 1 n.a.	11 s 7 na.	-
Genetic mutations[2]	17 C9ORF72/ 5 SOD/ 1 FUS/ 189 neg	9 neg	6 neg	8 C9ORF72/ 1 SOD/ 1 neg	6 neg	1 neg	
Physical function [ALSFRS-R][1]	37.2 ± 7.9	39.7 ± 4.9	37.6 ± 5.8	38.3 ± 7.2	38.9 ± 8.3	-	-
Time since onset [months]	22.2 ± 24.5	50.7 ± 41.7	27.2 ± 22.2	30.6 ± 37.2	13.2 ± 9.4	88 ± 178.8	
	91 NIV 23 PEG	1 NIV	1 NIV	5 NIV 1 PEG	-	-	-

Mean ± SD or frequency, where appropriate; ALS=amyotrophic lateral sclerosis; PLS=primary lateral sclerosis; PBP=progressive bulbar palsy; FTD= frontotemporal dementia of the behavioural variant; HC= healthy controls; NIV=non-invasive ventilation; PEG=percutaneous endoscopic gastrostomy; n.a. not applicable. Physical function was measured with the ALS functional rating scale revised form (ALS-FRS-R).[sup1] Gene mutations (SOD1, C9orf72 and FUS) were determined according to standard protocol.[sup2]

Supplement references

1. Cedarbaum JM, Stambler N, Malta E, et al. The ALSFRS-R: a revised ALS functional rating scale that incorporates assessments of respiratory function. J Neurol Sci 1999;169:13-21.

2. Freischmidt A, Wieland T, Richter B, et al. Haploinsufficiency of TBK1 causes familial ALS and fronto-temporal dementia. Nat Neurosci. 2015;18:631-636.