Supplementary Table 1: List of patient tissue information and experimental analyses performed.

Sample No.	Patient ID	Source	Clinical diagnosis	Age of diagnosis or sampling	Race/Gender	Region
1	90015	VABBB	CTL	66	UN/M	SC-T & OCCL
2	100036	VABBB	CTL	56	UN/M	SC-T & OCCL
3	110005	VABBB	CTL	62	UN/M	SC-T & OCCL
4	110006	VABBB	CTL	68	UN/M	SC-T
5	110023	VABBB	CTL	N/A	N/A	SC-T
6	120016	VABBB	CTL	63	UN/F	SC-T
7	90003	VABBB	fALS	73	W/M	SC-T & OCCL
8	100007	VABBB	fALS/FTD*-C9orf72 HRE	61	W/M	SC-T & OCCL
9	100038	VABBB	ALS	57	B/M	SC-T & OCCL
10	110010	VABBB	fALS-PFN1 G118V	55	W/M	SC-T & OCCL
11	120015	VABBB	ALS/FTD*	58	W/M	SC-T & OCCL
12	90005	VABBB	ALS	65	W/M	SC-T
13	90020	VABBB	fALS-SOD1 L145F	49	W/F	SC-T
14	100002	VABBB	ALS	63	W/M	SC-T
15	100039	VABBB	fALS-C9orf72 HRE	50	W/M	SC-T
16	100040	VABBB	ALS	88	W/M	SC-T
17	993	JHMI	CTL	66	W/M	IP
18	989	JHMI	CTL	59	M/M	IP
19	1058	JHMI	CTL	70	W/M	IP
20	2317	JHMI	CTL	65	B/M	IP
21	2242	JHMI	AD	62	W/M	IP
22	2260	JHMI	AD	78	W/F	IP
23	2273	JHMI	AD	62	W/F	IP
24	2277	JHMI	AD	63	W/F	IP
25	2376	JHMI	AD	61	W/M	IP
26	2417	JHMI	AD	61	W/F	IP
27	2430	JHMI	AD	62	UN/F	IP

Abbreviations: ALS (amyotrophic lateral sclerosis, fALS (familial amyotrophic lateral sclerosis), FTD* (frontotemporal dementia-like pathology), HRE (hexanucleotide repeat expansion), AD (alzheimer's disease), CTL (control), UN (unknown), W (white), M (male), F (female), B (black), N/A (not applicable), JHMI (Johns Hopkins Medical Institute), VABBB (VA biorepository brain bank), SC-T (Spinal cord-Thoracic), OCCL (occipital lobe), IP (inferior parietal cortex).

Supplementary Table 2. *C.elegans* strains

Strains	Allele	Genotype	Source
IW07	*iwIs25*	P*snb-1*::SOD1-WT	Wang lab (Wang et al., 2009)
IW5	*iwIs10*	P*snb-1*::SOD1-G85R	Wang lab (Wang et al., 2009)
IW8	*iwIs8*	P*snb-1*::SOD1-G85R-YFP	Wang lab (Wang et al., 2009)
IW377	*iwIs10;iw139*	P*snb-1*::SOD1-G85R; *lin-61(H479Y)*	Wang lab
MT12833	*n3809*	*lin-61(Q159Ochre)*	CGC (Harrison et al., 2007)
IW446	*iwIs10;n3809*	P*snb-1*::SOD1-G85R; *lin-61(Q159Ochre)*	Wang lab
IW445	*iwIs8;n3809*	P*snb-1*::SOD1-G85R-YFP; *lin-61(Q159Ochre)*	Wang lab
IW446	*iwIs25;n3809*	P*snb-1*::SOD1-WT; *lin-61(Q159Ochre)*	Wang lab
AM716	*rmIs284*	PF25B3.3::Q(67)-YFP	Richard Morimoto (Brignull et al., 2006)
IW777	*rmIs284;n3809*	PF25B3.3::Q(67)-YFP; *lin-61(Q159Ochre)*	This study
YD12	*xzEx12*	P*F25B3.3*::UbG76V-Dendra2	C. Holmberg (Hamer et al., 2010)
	xzEx12; n3809	P*F25B3.3*::UbG76V-Dendra2; *lin-61(Q159Ochre)*	This study
	xzEx12; iwIs10;n3809	P*F25B3.3*::UbG76V-Dendra2; P*snb-1*::SOD1-G85R; *lin-61(Q159Ochre)*	This study
CB4856	Hawaiian strain	Wild-type	CGC

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