Table 1. Removal of NF-M/H tail domains protects against motor neuron degeneration in SOD1-mediated ALS in mice

Ventral (Motor) Root Axon Number (per L5 Root)

NF-(M/H)$^{wild\text{-}type}$ (control mice, at 14 months and healthy)	NF-(M/H)$^{tail\Delta}$ (control mice, at 14 months and healthy)		NF-(M/H)$^{wild\text{-}type}$ SOD1^{G37R}	NF-(M/H)$^{tail\Delta}$ SOD1^{G37R}	NF-M$^{tail\Delta}$ SOD1^{G37R}	NF-H$^{tail\Delta}$ SOD1^{G37R}
1008 ± 61 (*n*=3)	862 ± 58 (*n*=4)	presymptomatic:	1032 ± 19 (*n*=3)	862 ± 78 (*n*=3)	ND	ND
		hind limb weakness:	517 ± 29 (*n*=4)	724 ± 26 (*n*=3)	525 ± 52 (*n*=3)	580 ± 91 (*n*=2)
		end stage:	374 ± 29 (*n*=4)	613 ± 51 (*n*=3)	384 ± 33 (*n*=3)	460 ± 31 (*n*=4)

Lumbar Ventral Horn Motor Neuron Number (per L3-L6 Section)

NF-(M/H)$^{wild\text{-}type}$ (control mice, at 14 months and healthy)	NF-(M/H)$^{tail\Delta}$ (control mice, at 14 months and healthy)		NF-(M/H)$^{wild\text{-}type}$ SOD1^{G37R}	NF-(M/H)$^{tail\Delta}$ SOD1^{G37R}
27 ± 2.2 (*n*=3)[1]	23 ± 1.9 (*n*=3)[1]	hind limb weakness:	12 ± 0.1 (*n*=3)[1]	18 ± 0.4 (*n*=3)[1]
47 ± 3.5 (*n*=3)[2]	41 ± 3.1 (*n*=3)[2]		29 ± 4.0 (*n*=3)[2]	36 ± 0.2 (*n*=3)[2]
[1]large motor neurons (>25 μm diameter)		end stage:	7 ± 0.4 (*n*=3)[1]	12 ± 0.5 (*n*=2)[1]
[2]total motor neurons			17 ± 2.3 (*n*=3)[2]	28 ± 5.5 (*n*=2)[2]

Percentage of Innervated End Plates in the Gastrocnemius Muscle

NF-(M/H)$^{wild\text{-}type}$ (control mice, at 14 months and healthy)	NF-(M/H)$^{tail\Delta}$ (control mice, at 14 months and healthy)		NF-(M/H)$^{wild\text{-}type}$ SOD1^{G37R}	NF-(M/H)$^{tail\Delta}$ SOD1^{G37R}
83 ± 7.3 (*n*=3)	ND	hind limb weakness:	26 ± 2.2 (*n*=3)	60 ± 5.3 (*n*=3)
		end stage:	13 ± 4.2 (*n*=3)	25 ± 5.9 (*n*=3)

Results are mean ± SEM. Presymptomatic (at 6 months; before onset), hind limb weakness (at 10% weight loss; after onset), end stage (hind limb paralysis). Percentage of innervated end plates was determined as the fraction of bungarotoxin-positive endplates (postsynaptic) that were also positive for synaptophysin and NF (presynaptic). Per animal (*n*) 150 randomly choosen end plates were analyzed. ND, not determined.