Supplementary Table 1. Key characteristics of commonly used mouse models of ALS (excluding C9orf72 repeat expansion models). Abbreviations: Alsin^{KO}, Alsin knockout; FUS, fused in sarcoma; hPFN1, human profilin 1; LMN, lower motor neurons; n.a., not applicable; n.s., not specified; SOD1, superoxide dismutase 1; TDP-43, TAR DNA-binding protein 43; UCHL1 null, ubiquitin carboxyterminal hydrolase 1 gene knockout; UMN, upper motor neurons; +, present; -, absent.

model	gene copy number	disease onset (months)	life-span (months)	motor phenotype (tremor, muscle weakness)	paralysis (at the humane end-stage)	UMN loss	UMN deficits	LMN loss	LMN deficits	references
SOD1^{G93A}	25	3	4	+	+	+	+	+	+	(Gurney et al., 1994; Chiu et al., 1995; Özdinler et al., 2011)
SOD1$^{G93A\text{-}low}$	8-10	4-5	8-8.5	+	+	n.s.	n.s.	+	+	(Acevedo-Arozena et al., 2011)
SOD1^{G85R}	0.2-1	8	8.5	+	+	n.s.	n.s.	+	+	(Bruijn et al., 1997)
SOD1^{G86R}	high/low	3-4	4	+	+	n.s.	n.s.	+	+	(Ripps et al., 1995)
SOD1^{G37R}	4-12	3-6	7	+	+	n.s.	n.s.	+	+	(Wong et al., 1995)
TDP-43^{Q331K}	1-1.5	3	> 24	+	+	+	+	+	+	(Arnold et al., 2013; Mitchell et al., 2015)

Prp-TDP43^{A315T}	n.a.	3-4	4.5-5.7	+	+	+	+	+	+	(Wegorzewska et al., 2009)
FUS^{P525L}	4	1	12	+	n.s.	n.s.	n.s.	+	n.s.	(Sharma et al., 2016)
FUS$^{dNLS/+}$	n.a.	10	>22	+	-	n.s.	n.s.	-	-	(Scekic-Zahirovic et al., 2016; Scekic-Zahirovic et al., 2017)
Alsin^{KO}	n.a.	12	>13	-	-	-	+	-	+	(Deng et al., 2007; Gautam et al., 2016)
UCHL1 null	n.a.	2.2	>6.6	+	-	+	+	-	+	(Jara et al., 2015) (Genç et al., 2016)
hPFN1^{G118V}	n.a.	4-4.5	5.7-7.7	+	+	+	+	+	+	(Fil et al., 2017)
Prp-PFN1^{C71G}	n.a.	4-5	6-8	+	+	-	-	+	+	(Yang et al., 2016)

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