

Supplemental Table 1: Demographic Information for iPSC Lines

iPSC Line Name	Source	Clinical Diagnosis	Age at Time of Collection	Sex	Origin
CS0002	Cedars-Sinai	Non-neurologic control	51	Male	PBMC
CS0201	Cedars-Sinai	Non-neurologic control	56	Female	PBMC
CS0202	Cedars-Sinai	Non-neurologic control	57	Male	PBMC
CS0206	Cedars-Sinai	Non-neurologic control	72	Female	PBMC
CS1ATZ	Cedars-Sinai	Non-neurologic control	60	Male	PBMC
CS2AE8	Cedars-Sinai	Non-neurologic control	50	Female	PBMC
CS8PAA	Cedars-Sinai	Non-neurologic control	58	Female	PBMC
CS9XH7	Cedars-Sinai	Non-neurologic control	53	Male	PBMC
EDi022-A	Cedars-Sinai	Non-neurologic control	79	Male	PBMC
EDi034-A	Cedars-Sinai	Non-neurologic control	79	Female	PBMC
EDi036-A	Cedars-Sinai	Non-neurologic control	79	Female	PBMC
EDi037-A	Cedars-Sinai	Non-neurologic control	79	Male	PBMC
EDi043-A	Cedars-Sinai	Non-neurologic control	80	Male	PBMC
EDi044-A	Cedars-Sinai	Non-neurologic control	80	Female	PBMC
CS0JGZ	Cedars-Sinai	sALS	56	Male	PBMC
CS1KL3	Cedars-Sinai	sALS	71	Female	PBMC
CS2EVP	Cedars-Sinai	sALS	69	Male	PBMC
CS3EPR	Cedars-Sinai	sALS	60	Female	PBMC
CS3XLK	Cedars-Sinai	sALS	55	Female	PBMC
CS4KGP	Cedars-Sinai	sALS	82	Female	PBMC
CS4PFR	Cedars-Sinai	sALS	72	Female	PBMC
CS4ZCD	Cedars-Sinai	sALS	53	Male	PBMC
CS5JPF	Cedars-Sinai	sALS	55	Female	PBMC
CS5ZHY	Cedars-Sinai	sALS	64	Male	PBMC
CS6EVH	Cedars-Sinai	sALS	56	Male	PBMC
CS6PYD	Cedars-Sinai	sALS	69	Female	PBMC
CS8JGP	Cedars-Sinai	sALS	61	Male	PBMC
CS9GXD	Cedars-Sinai	sALS	68	Male	PBMC

Supplemental Table 2: ASO Sequences

ASO Name	Sequence
Scrambled (Ionis ID: 676630)	CCTATAGGACTATCCAGGAA
CHMP7 (Ionis ID: 1508917)	TGTTACCCTCAGATACCGCC
CHMP2B ASO 1	TGTGACAAGTCCTAGTGGGA
CHMP2B ASO 2	AGGGTGAGAAAGAGTATGGG
CHMP2B ASO 3	GGGATGCAGAAATACAGAGG
CHMP2B ASO 4	AAGTGGCATCTGAAGTAGGG
CHMP2B ASO 5	TGGTAAGCGAAGATGCCAGA
CHMP2B ASO 6	ATCACTTACCACCTGCCATG
CHMP2B ASO 7	GTTGGTGGGACAGAGCAATA
CHMP2B ASO 8	CACCTAACTCTCTAGTGGCT
CHMP2B ASO 9	TCAATGGAACAGAGCCCAGA
CHMP2B ASO 10	CATTTACCCTATGACCCAGC

Supplemental Table 3: Plasmid Information

Plasmid	Backbone	Source
CHMP7 WT Flag	pcDNA3.1	Vietri et al., 2020
CHMP7 Open Flag	pcDNA3.1	Genscript, This Paper
CHMP7 Δ Helix 6 Flag	pcDNA3.1	Genscript, This Paper
CHMP7 NES1*/NES2* Flag	pcDNA3.1	Genscript, This Paper
POM121 RITE (Myc-Flag)	pLenti CMV/TO Hygro DEST	Toyoma et al., 2018

Supplemental Table 4: Antibody Information

Primary Antibodies			
Antibody	Source	Catalog Number	Application and Concentration
Rabbit Anti-CHMP7	ProteinTech	16424-1-AP	IF: 1/250
Mouse Anti-CHMP7	ProteinTech	68406-1-Ig	IF: 1/250
Rabbit Anti-POM121	ProteinTech	15645-1-AP	IF: 1/250
Rabbit Anti-CHMP2B	Cell Signaling	76173S	Western: 1/1000 IF: 1/250
Mouse Anti-CHMP2B	Invitrogen	MA5-21591	IF: 1/250
Rabbit Anti-CHMP2B	ProteinTech	12527-1-AP	Western: 1/1000 IF: 1/250
Mouse Anti-CHMP2B	R&D Systems	MAB7509	Western: 1/1000
Guinea Pig Anti-Map2	Synaptic Systems	188004	IF: 1/1000
Rat Anti-Flag	Thermo Fisher Scientific	MA1-142	IF: 1/2500
Mouse Anti-Myc	Cell Signaling	2276S	IF: 1/500
Mouse Anti-GAPDH	Life Technologies	AM4300	Western: 1/10,000
Secondary Antibodies			
Goat Anti-Mouse Alexa 488	Invitrogen	A11029	IF: 1/1000
Goat Anti-Rabbit Alexa 488	Invitrogen	A11034	IF: 1/1000
Goat Anti-Rat Alexa 488	Invitrogen	A11006	IF: 1/1000
Goat Anti-Mouse Alexa 568	Invitrogen	A11031	IF: 1/1000
Goat Anti-Rabbit Alexa 568	Invitrogen	A11036	IF: 1/1000
Goat Anti-Rat Alexa 568	Invitrogen	A11077	IF: 1/1000
Goat Anti-Guinea Pig Alexa 568	Invitrogen	A11075	IF: 1/1000
Goat Anti-Mouse Alexa 647	Invitrogen	A21236	IF: 1/1000
Goat Anti-Rabbit Alexa 647	Invitrogen	A21245	IF: 1/1000
Goat Anti-Rabbit IgG HRP	Cell Signaling	7074S	Western: 1/5000
Horse Anti-Mouse IgG HRP	Cell Signaling	7076S	Western: 1/5000

Supplemental Table 5: Demographic Information for Postmortem Human Tissue

	Clinical Diagnosis	Age of Death	Sex
Control-1	Non-neurologic control	71	Female
Control-2	Non-neurologic control	52	Male
Control-3	Non-neurologic control	74	Female
Control-4	Non-neurologic control	92	Female
Control-5	Non-neurologic control	72	Male
Control-6	Non-neurologic control	50	Male
Control-7	Non-neurologic control	62	Male
Control-8	Non-neurologic control	67	Male
Control-9	Non-neurologic control	63	Female
Control-10	Non-neurologic control	82	Female
sALS-1	sALS	67	Female
sALS-2	sALS	59	Male
sALS-3	sALS	68	Female
sALS-4	sALS	69	Male
sALS-5	sALS	65	Male
sALS-6	sALS	78	Male
sALS-7	sALS	68	Female
sALS-8	sALS	57	Female
sALS-9	sALS	61	Female
sALS-10	sALS	51	Male
sALS-11	sALS	80	Male
sALS-12	sALS	72	Female
sALS-13	sALS	70	Female
sALS-14	sALS	68	Male
sALS-15	sALS	69	Male

Supplemental Table 6: Primer Sequences and TaqMan Probe IDs for qRT-PCR

Primer Sequences		
Target	Sequence	
STMN2	Forward	AGCTGTCCATGCTGTCACTG
	Reverse	GGTGGCTTCAAGATCAGCTC
Truncated STMN2	Forward	GGA CT CGGCAGAAGACCTTC
	Reverse	GCAGGCTGTCTGTCTCTCTC
GAPDH	Forward	GAAGGTGAAGGTCGGAGTC
	Reverse	GAAGATGGTGATGGGATTTC
ACTL6B CE	Forward	ATCCTGGATCACACCTACAGC
	Reverse	AGGAGGATTGCTTGAACCC
ARHGAP32 CE	Forward	CACCTTCTAAATTCTGGTTTTGAAG
	Reverse	CAGATACAGACGAAAAAGCTGAGTT
CAMK2B CE	Forward	GAGTGCAGAGACTTCCCCC
	Reverse	CTGCTCCGTGGTCTTAATGAT
CDK7 CE	Forward	GCAGTGTGGACATGACTGATA
	Reverse	GTCCACACCTACACCATACATC
DNM1 CE	Forward	TGACCCTTTCGGCCCT
	Reverse	CACGAAATCAACATGGCAGT
HDGFL2 CE	Forward	TCACACCTGAGAAGAAAGCAG
	Reverse	TCCTCTCTTCTGTGTCCCTCT
MYO18A CE	Forward	AAGTCCAGGGATGAGATTGTG
	Reverse	GCAGAGTTTTGTCCTCCTCTTTA
SYT7 CE	Forward	GCAGTGAGAAGAAGGCTATCAA
	Reverse	CGGCAGACTGGAGCCT
TaqMan Probe IDs		
Target		TaqMan ID
ELAVL3		Hs00154959_m1
PFKP		Hs00737347_m1
RCAN1		Hs01120954_m1
SELPLG		Hs05033974_s1
POM121		Hs00208021_m1

Actin	Hs03023943_g1
GAPDH	Hs02786624_g1