

SUPPLEMENTARY TABLES

Supplementary Table 1. Demographic and clinical data for iPSC-derived motor neurons.

Cell Line	Sex	Age (y)	Clinical diagnosis	Primary tissue	Mutation
CS0002iCTR	M	51	Normal	PBMC	N/A
CS83iCTR	F	21	Normal	Fibroblast	N/A
CS188iCTR	M	80	Normal	PBMC	N/A
CS14iCTR	F	35	Normal	Fibroblast	N/A
CS00iCTR	M	6	Normal	Fibroblast	N/A
FA0000011	F	49	Normal	Fibroblast	N/A
NN0003920	M	64	Normal	Fibroblast	N/A
CS0118iALS-SOD1-I114T	F	73	ALS	Fibroblast	SOD1 I113T
CS28iALS	M	47	ALS	Fibroblast	C9ORF72 (HRE ~800)
CS29iALS	M	47	ALS	Fibroblast	C9ORF72 (HRE ~800)
CS52iALS	M	49	ALS	Fibroblast	C9ORF72 (HRE ~800)
CS30iALS	F	51	ALS	Fibroblast	C9ORF72 (HRE ~70)
NN0004306	F	51	ALS	Fibroblast F10330	C9ORF72 (HRE 2.7kb)
NN0004307	M	57	ALS	Fibroblast F09152	C9ORF72 (HRE 6-8kb)

ALS, amyotrophic lateral sclerosis; F, female; HRE, hexanucleotide repeat expansion; M, male; ORF, open reading frame; PBMC, peripheral blood mononuclear cells; SOD1, superoxide dismutase 1; y, years.

Supplementary Table 2. Demographic and clinical data of tissue samples.

	Biopsy		Autopsy	
	Normal	ALS	Normal	ALS
Number	24	36	22	23
Mean age (years) ^a	52 ± 15	57 ± 13	67 ± 12	64 ± 11
Age range (years)	24 - 77	27- 86	34 - 83	40 - 81
Gender (M:F)	11:13	21:15	18:4	18:5
Duration ^b (m)		15 ± 9		51 ± 32
Diagnosis		Spinal onset (33) Bulbar onset (3)		Spinal onset (20) Bulbar onset (3)
Muscle sampled				
Biceps brachii	5	2		2
Deltoid	3	11	4	3
Vastus lateralis	15	8	3	5
Tibialis anterior	1	15		Triceps (1)

^aMean age (± SD) at time of sample collection.

^bMean duration (± SD) from onset of symptoms to sample collection. Duration was unknown for three ALS patients in the biopsy pool.

Supplementary Table 3. Demographic and clinical data of plasma samples.

	Normal	ALS
Number	23	28
Mean age (y) ^a	61 ± 9	59 ± 10
Age range (y)	45 - 84	35- 82
Gender (M:F)	12:11	18:10
Duration ^b (m)		26 ± 20
Onset		Spinal onset (22) Bulbar onset (6)

F, female; M, male; m, months; y, years.

^aMean age (± SD) at time of sample collection.

^bMean duration (± SD) from onset of symptoms to sample collection.

Supplementary Table 4. ALS study patients.

Plasma FGF21 (FC)^a	< 1.5	≥ 1.5
Number	7	9
Age (y) ^b	65 ± 9	57 ± 9 ^d
Age range (y)	58 - 82	42 – 70 ^d
Gender (M:F)	4:3	7:2
Duration ^c (m)	18 ± 10	26 ± 23 ^d
Onset	Bulbar (4) Spinal (3)	Spinal (9)

F, female; FC, fold-change; M, male; m, months; y, years.

^aFold-change over normal control group.

^bMean age (± SD) at time of sample collection.

^c Mean duration (± SD) from onset of symptoms to sample collection.

^dNo significant difference between the 2 groups.