

Mitochondrial respiratory complex IV deficiency recapitulates amyotrophic lateral sclerosis

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Table S1. Human samples information.

Samples	Disease	Tissues	Age	Gender
NBB-C1-Cer	Ctrl	Cerebellum/ Motor cortex	79	male
NBB-C2-Cer	Ctrl	Cerebellum/ Motor cortex	86	female
NBB-C3-Cer	Ctrl	Cerebellum/ Motor cortex	70	male
NBB-C4-Cer	Ctrl	Cerebellum	85	male
NBB-C5-Cer	Ctrl	Cerebellum	83	male
NBB-C7-Cer	Ctrl	Cerebellum	85	female
NBB-C8-Cer	Ctrl	Cerebellum	84	female
NBB-A1-Cer	C9orf72-ALS/FTD	Cerebellum/ Motor cortex	72	female
NBB-A2-Cer	C9orf72-ALS/FTD	Cerebellum	68	female
NBB-A4-Cer	C9orf72-ALS/FTD	Cerebellum	75	male
NBB-A5-Cer	C9orf72-ALS/FTD	Cerebellum/ Motor cortex	65	male
NBB-A6-Cer	C9orf72-ALS/FTD	Cerebellum	77	female
NBB-A7-Cer	sALS	Cerebellum/ Motor cortex	53	female
NBB-A8-Cer	sALS	Cerebellum	64	female
C1	Ctrl	Muscle	61	female
C2	Ctrl	Muscle	52	male
P1	sALS	Muscle	76	female
P2	sALS	Muscle	48	male
P3	sALS	Muscle	50	male
P4	sALS	Muscle	48	male

Table S2. The primers used for genotyping, Sanger-sequencing, Deep-sequencing, and QPCR.

Name		Sequence (5'>3')	Annealing temperature for qRT-PCR
Genotyping	ROSA-FP	GGGTGGCGAAGGTAATGTCT	
	ROSA-RP	GCTATGAACTAATGACCCCGTAATT	
	CRE-FP CRE-RP	TACTGACGGTGGGAGAATG CTGTTTCACTATCCAGGTTACG	
Sanger-sequencing	COXI-FP COXI-RP	GCTAAATACCCTACTTACTGGCTTC CAATTATTAGTGGTACAAGTCAGTTCC	
	COXII-FP: COXII-RP	ATAGGCTCATTATCTCACTTACG TTAGAACTGGGTATTAATCTCGTCTAT	
	COXIII-FP COXIII-RP	CAGTACGACTAACAGCAAACATTACAG CTGATGCTAAGAGGACTGATGTAT	
QPCR	Chat-FP Chat-RP	GAGCCTGAGCATGTCATCGT ATCCTCGTTGGACGCCATTT	58 °C
	Nefh-FP Nefh-RP	GCTGCTCGGTCAGATTCAGG CGGTCCAATCTCACTCGGAAC	60 °C
	Sv2b-FP Sv2b-RP	GGCGCCACAATCAACTTCAC ATCATCGCAGACTCGCTGTT	61 °C
	Sdk2-FP Sdk2-RP	TGGGACCTCCGAGGTGACCA TGCGTAGCATGGCTTCGCAC	63 °C
	MMP9-FP MMP9-RP	AAACCTCCAACCTCACGGAC GGCCTTTAGTGTCTCGCTGT	58 °C
	Chodl-FP Chodl-RP	GGAGGTGTCCTCCTCAGTCT GAACTGCTTCCGTCAGACCA	55 °C
	Nos1-FP Nos1-RP	TGACCCAGCCCCTCGGTCTT AGCTCCCTGCCCATGGCCTT	68 °C
	Zeb2-FP Zeb2-RP	CCCGGTGCAAGAGGCGCAA CCAGCTGTGTTCCACAACGC	68 °C
	Gpc4-FP Gpc4-RP	GGTGACCATTTGAAGATCTG CGTATGTCTTCACGAACATA	52 °C
	Slc6a1-FP	CAAGCCCCAAAACCCTTGTAG	57.5 °C

	Slc6a1- RP	AAGGAAGAGAGGAACACCCG	
Deep- sequencing	rCOX1- KO-Fwd	CTCAGCCATTTTACCTATGCTC	
	rCOX1- KO-Rev	TGTGACGATGACATTATAGATTTGG	
	rCOX2- KO-Fwd2	CCCTACAACCTGGTTTCAAGCC	
	rCOX2- KO-Rev2	GGTGTGGTCATGAAAGTTTGTAAG	
	rCOX3- KO-Fwd	ACTTGAATTTGCCGTAGCCT	
	rCOX3- KO-Rev	AGGATGTTTGTGTCAGGAGGCC	