

Supplemental Table 1: Primer sequences for qRT-PCR

GENE	FORWARD	REVERSE	REF.
PGC1 α	TTGCTAGCGGTCCTCACAGA	GGCTCTTCTGCCTCCTGA	(27)
PGC1 β	CGCTCCAGGAGACTGAATCCAG	CTTGACTACTGTCTGTGAGGC	(54)
Tfam	CCAAAAAGACCTCGTTCAGC	ATGTCTCCGGATCGTTTCAC	(82)
PPAR γ	GGAAGACCACTCGCATTCTT	TCGCACTTTGGTATTCTTGGAG	(83)
Nrf2	TTCCTCTGCTGCCATTAGTCAGTC	GCTCTTCCATTTCCGAGTCACTG	(84)
NQO1	TTCTCTGGCCGATTTCAGAGT	GGCTGCTTGGAGCAAATG	(85)
ND1	CCTTCGACCTGACCTGACAGAAGGA	GATGCTCGGATCCATAGGAA	(82)
ND5	GCTCTACCTCACCATCTCTTGC	TCCAGTATGCTTACCTTGTTACG	(82)
COX I	GGTCAACCAGGTGCACTTTT	TGGGGCTCCGATTATTAGTG	(82)
COX II	TGAAGACGTCCTCCACTCATGA	GCCTGGGATGGCATCAGTT	(83)
COX III	GCAGGATTCTTCTGAGCGTTCT	GTCAGCAGCCTCCTAGATCATGT	(83)
SOD1	CAGGACCTCATTTTAATCCTCAC	CCCAGGTCTCCAACATGC	(85)
SOD2	CACAAGCACAGCCTCCAG	CGCGTTAATGTGTGGCTCC	(70)
Catalase	CAGCGACCAGATGAAGCA	CTCCGGTGGTCAGGACAT	(85)
Gpx-1	ACAGTCCACCGTGTATGCCTTC	CTCTTCATTCTTGCCATTCTCCTG	(85)
HO-1	GGTCAGGTGTCCAGAGAAGG	CTCCAGGGCCGTGTAGATA	(85)
S18	GTTGGTGGAGCGATTTGTCTGG	AGGGCAGGGACTTAATCAACGC	(82)

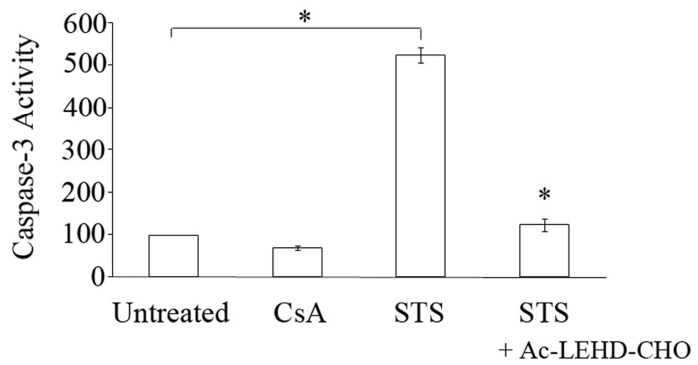
82. Noack, H., Bednarek, T., Heidler, J., Ladig, R., Holtz, J. and Szibor, M. (2006)
TFAM-dependent and independent dynamics of mtDNA levels in C2C12 myoblasts caused by redox stress. *Biochim. Biophys. Acta*, **1760**, 141-150.
83. Uldry, M., Yang, W., St-Pierre, J., Lin, J., Seale, P. and Spiegelman, B.M. (2006)
Complementary action of the PGC-1 coactivators in mitochondrial biogenesis and brown fat differentiation. *Cell Metab.*, **3**, 333-341.
84. Pehar, M., Vargas, M.R., Robinson, K.M., Cassina, P., Diaz-Amarilla, P.J., Hagen, T.M., Radi, R., Barbeito, L. and Beckman, J.S. (2007) Mitochondrial

superoxide production and nuclear factor erythroid 2-related factor 2 activation in p75 neurotrophin receptor-induced motor neuron apoptosis. *J. Neurosci.*, **27**, 7777-7785.

85. Panee, J., Liu, W., Nakamura, K. and Berry, M.J. (2007) The responses of HT22 cells to the blockade of mitochondrial complexes and potential protective effect of selenium supplementation. *Int. J. Biol. Sci.*, **3**, 335-341.

SUPPLEMENTAL FIGURE 1: Staurosporine increases cell death in MN-1 cells

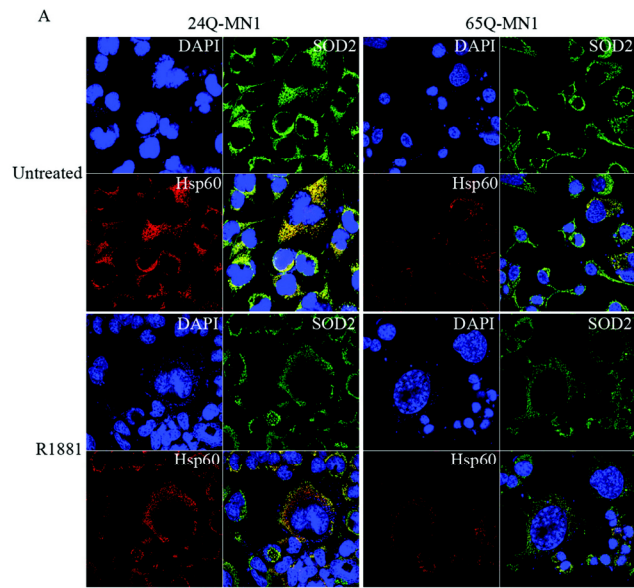
Staurosporine (STS), an apoptotic inducer was used as a positive control for caspase 3 activity in the fluorometric assay (for Fig. 1C). Parental MN-1 cells were exposed to vehicle control or STS and treated with the pan-caspase inhibitor, z-VAD. STS resulted in a 5-fold increase in caspase 3 activity that was blocked with z-VAD.



SUPPLEMENTAL FIGURE 2: Reduced SOD2 protein in MN-1 cells and mouse

tissues

AR-24Q and AR-65Q MN-1 cells (A) were either exposed to vehicle control or R1881 for 48 h. The cells were immunostained for SOD2 (FITC labeled; green), and Hsp60 (Texas-red labeled; red) with the nucleus counterstained with DAPI (blue). The images show decreased levels of SOD2 and Hsp60 in the AR-65Q MN-1 cells. (B-C) Representative immunoblots of 3-month old mouse muscle (B) and spinal cord (C) show decreased levels of SOD2. The densitometric quantitation is shown in the histograms (N = 5 for each genotype per tissue). The error bars indicate SEM; * $p \leq 0.05$.



B Muscle at manifesting stage **C** Spinal cord at manifesting stage

