

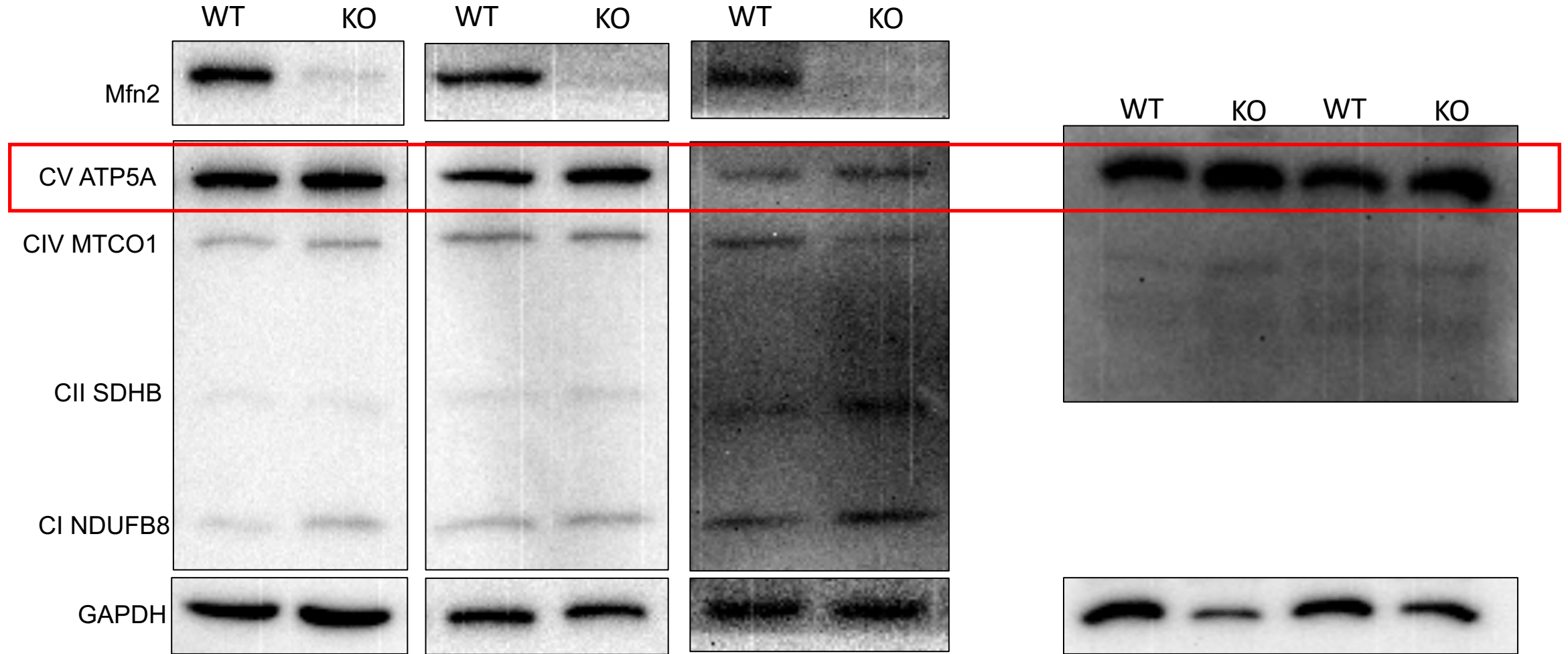
**Supplementary table 1.** Primers used for gene expression analysis and genotyping

<b>Primer</b>	<b>Sequence (5'→3')</b>	<b>location</b>	<b>PCR production length (bp)</b>	<b>Application</b>
Mfn1-F	CCTACTGCTCCTTCTAACCCA	exon 13	86bp	qRT-PCR
Mfn1-R	AGGGACGCCAATCCTGTGA			
Mfn2-F	CTGGGGACCGGATCTTCTTC	exon9 to exon 10	143bp	qRT-PCR
Mfn2-R	CTGCCTCTCGAAATTCTGAAACT			
β-actin-F	GTCCCTCACCCCTCCCAAAG	mRNA1805 to 2071bp	266bp	qRT-PCR
β-actin-R	GCTGCCTCAACACCTCAACCC			
mtDNA-F	CCTATCACCCCTTGCCATCAT	DNA 3920 to 4094bp	194bp	qRT-PCR
mtDNA-R	GAGGCTGTTGCTTGTGTGAC			
neuDNA-F	ATGGAAAGCCTGCCATCATG	DNA106699862 to 106699646bp	236bp	qRT-PCR
neuDNA-R	TCCTTGTTGTTTCAGCATCAC			
Mfn2-F	TTTGGAAGTAGGCAGTCTCCA	exon 6	Mutant 210bp, WT 193bp	Genotyping
Mfn2-R	CAGGCAGCACTGAAAAGAGA			
MyoDCre-F-Mutant	GCGGATCCGAATTCGAAGTTCC	exon 1	Mutant 149bp, WT 343bp	Genotyping
MyoDCre-F-Wildtype	CGGCTACCCAAGGTGGAGAT			
MyoDCre-R-Common	TGGGTCTCCAAAGCGACTCC			

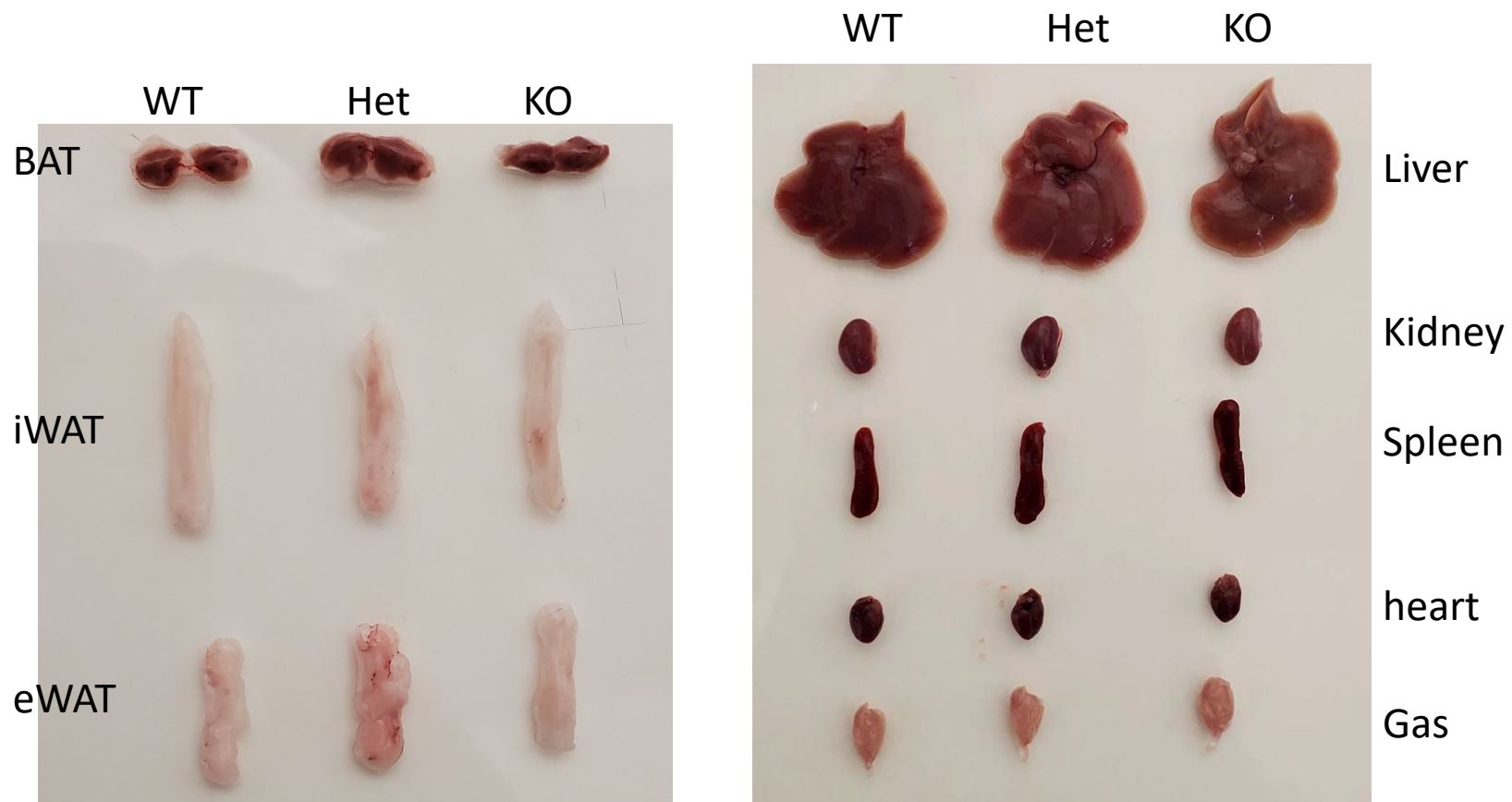
**Supplementary table 2.** Antibody used for Immunohistochemistry (IHC) or immunofluorescence (IF) and western blot (WB)

<b>Antibody</b>	<b>Source or referenc</b>	<b>Identifiers</b>	<b>Dilution rate</b>
Mfn2	Cell Signaling Technology	Cat# 9482	IHC/IF (1:500), WB (1:1000)
Mito-Tracker	Cell Signaling Technology	Cat# 9082	IF (1:200)
Pax7	Developmental Studies Hybridoma Bank	Cat# pax7	IHC/IF (1:15), WB (1:100)
MyoG	Developmental Studies Hybridoma Bank	Cat# PCR- MYOG1C5	IF (1:1000), WB (1:1000)
MF20	Developmental Studies Hybridoma Bank	Cat# MF 20	IF (1:500), WB (1:1000)

MyHC I	Developmental Studies Hybridoma Bank	Cat# BA-D5	IF (1:200)
MyHC 2A	Developmental Studies Hybridoma Bank	Cat# SC-71	IF (1:200)
MyHC 2B	Developmental Studies Hybridoma Bank	Cat# BF-F3	IF (1:200)
MyHC	Developmental Studies Hybridoma Bank	Cat# MF 20	IF (1:500), WB (1:1000)
MyoD	Santa Cruz Biotechnology	Cat# sc-377460	WB (1:1000)
GAPDH	Santa Cruz Biotechnology	Cat# sc-47724	WB (1:1000)
Dystrophin	Abcam	Cat# ab15277	IHC (1:1000)
OxPhos	Thermo Fisher Scientific	Cat# 45-8099	WB (1:1000)
CellROX	Thermo Fisher Scientific	Cat# 10422	IF (1:1000)
H2DCFDA (H2-DCF, DCF)	Thermo Fisher Scientific	Cat# D399	IF (1:1000)
488 goat polyclonal antimouse IgG1	Thermo Fisher Scientific	Cat# A-21121	IF (1:1000)
488 goat polyclonal antirabbit IgG	Thermo Fisher Scientific	Cat# A-11034	IF (1:1000)
568 goat polyclonal antimouse IgG1	Thermo Fisher Scientific	Cat# A-21124	IF (1:1000)
647 goat polyclonal antimouse IgG2b	Thermo Fisher Scientific	Cat# A-21242	IF (1:1000)
DAPI	Thermo Fisher Scientific	Cat# sc-24941	IF (1:1000)
Hoechst 33342	Thermo Fisher Scientific	Cat# H3570	IF (1:1000)
HRP-conjugated goat polyclonal anti-rabbit IgG	Jackson Immuno Research Labs	Cat# 111-035003	WB (1:10000)
HRP-conjugated goat polyclonal anti-mouse IgG	Jackson Immuno Research Labs	Cat# 115-035003	WB (1:10000)



**Supplemental Figure S1.**  $Mfn2^{MKO}$  (KO) myoblasts have higher levels of complex V protein ATP5A (shown in the red box) than wildtype (WT) myoblasts.



**Supplemental Figure S2:** *Mfn2*<sup>MKO</sup> (KO) does not affect development of non-skeletal muscle tissues. Mice were analyzed at 2 months of age. Het: Heterozygous. WT: wildtype. BAT: Brown adipose tissue. iWAT: inguinal white adipose tissue. eWAT: epididymal white adipose tissue. Gas: Gastrocnemius muscle.