

ADDITIONAL FILE 5: Tabs S1-S3

Table S1		
Reagent/Resource	Source	Identifier
Antibodies		
Mouse monoclonal anti-Strep (1:1000)	Abcam	Cat# ab184224
Rabbit polyclonal anti-HA (1:1000)	Sigma-Aldrich	Cat# SAB4300603; RRID:AB_10620829
Rat monoclonal anti-HA (3F10; used for immunofluorescence)	Roche	Cat#11867423001; RRID:AB_10094468
Mouse monoclonal anti-HA, agarose conjugate	Sigma-Aldrich	Cat#A2095 RRID: AB_257974
Mouse monoclonal anti-LDH (H-10) (1:1000)	Santa-Cruz	Cat# sc-133123; RRID:AB_2134964
Mouse monoclonal anti-PDH (1:1000)	Santa-Cruz	Cat# sc-377092
Rabbit polyclonal anti-COX17 (1:1000)	Biorbyt	Cat# orb160552
Rabbit polyclonal anti-COX6B1 (1:1000)	Sigma-Aldrich	Cat# HPA004192, RRID:AB_1847180
Rabbit polyclonal anti-NDUFB7 (1:1000)	Abcam	Cat# ab188575
Rabbit polyclonal anti-NDUFS5 (1:1000)	Abcam	Cat# ab179806
Rabbit polyclonal anti-NDUFA8 (1:1000)	Abcam	Cat# ab184952
Rabbit polyclonal anti-NDUFB10 (1:1000)	Abcam	Cat# ab196019
Rabbit polyclonal anti-CMC1 (1:1000)	Sigma-Aldrich	Cat#HPA043333; RRID:AB_10797139
Mouse monoclonal anti-HSP70 (1:1000)	Santa-Cruz	Cat# sc-66048; RRID:AB_832518
Rabbit polyclonal anti-TIMM9 (1:250)	This study	N/A
Rabbit polyclonal anti-TIMM13 (1:1000)	Sigma-Aldrich	Cat# HPA048985
Purified mouse anti-TIMM23 (1:1000)	BD Biosciences	Cat# 611223
Rabbit polyclonal anti-STARD7 (1:1000)	Proteintech	Cat# 15689-1-AP
Rabbit polyclonal anti-MIA40 (1:1000)	Fischer et al., 2013, Erdogan et al., 2018	N/A
Goat anti-Mouse - Affinity Pure, HRP Conjugate	ImmunoReagents	Cat# GtxMu-003-DHRPX
Goat anti-Rabbit - Affinity Pure, HRP Conjugate	ImmunoReagents	Cat# GtxRb-003-DHRPX
Goat anti-Rat, Alexa Fluor 488	Invitrogen	Cat# A-11006; RRID:AB_2534074
Mitotracker red	Thermo Fisher	Cat# M22425
Bacterial and Virus Strains		
One Shot™ TOP10 Chemically Competent <i>E. coli</i>	Thermo Fisher	Cat# C404010
Rosetta™ 2 (DE3) Singles™ Competent Cells	Novagen	Cat# 70954-3
Chemicals		
MG132 (Z-Leu-Leu-Leu-al)	Sigma-Aldrich	Cat# C2211; CAS: 133407-82-6
Emetine	Sigma-Aldrich	Cat# E2375; CAS: 7083-71-8
Methyl-PEG-Maleimide, mmPEG12	Thermo Fisher	Cat# 22711
Methyl-PEG-Maleimide, mmPEG24	Thermo Fisher	Cat# 22713
N-Ethylmaleimide (NEM)	Sigma-Aldrich	Ca# E3876; CAS: 128-53-0
Tris(2-carboxyethyl)phosphine hydrochloride (Tris)	Sigma-Aldrich	Cat# C4706; CAS: 51805-45-9
Doxycycline hyclate	AppliChem	Cat# A2951; CAS: 24390-14-5
4-Isopropylbenzoic acid (Cumate)	Sigma-Aldrich	Cat# 268402; CAS: 536-66-3
Puromycin	InvivoGen	Cat# ant-pr-1
Hygromycin B solution from <i>Streptomyces hygroscopicus</i>	Sigma-Aldrich	Cat# H0654; CAS: 31282-04-9
Blasticidin	InvivoGen	Cat# ant-bl-05
PrestoBlue™ Cell Viability Reagent	Invitrogen by Thermo Fisher	Cat# A13261
Digitonin	PanReac AppliChem	Cat# A1905
Benzonase® Nuclease	Sigma-Aldrich	Cat# E1014
Critical Commercial Assays		
EasyTag EXPRESS 35S Protein Labeling Mix	Perkin-Elmer	Cat# NEG772
TnT Quick Coupled Transcription/Translation System	Promega	Cat# L1170
GSTrap 4B column	GE Healthcare	Cat# 28401745
Superdex 16/600 75 pg High Load column	GE Healthcare	Cat# 28989333
FuGENE® HD Transfection Reagent	Promega	Cat# E2311
Experimental Models: Cell Lines		
Human: Flp-In T-REx-293-cell lines – see Table S2	Invitrogen	Cat# R78007
Human: HEK293T-MIA40 CRISPR #2 MIA40 – see Table S2	Habich et al., 2019	Habich et al., 2019
Human: Flp-In T-REx-293 CRISPR ko YME1L1 – see Table S2	Langer lab	

Oligonucleotides		
See Table S3		
Recombinant DNA		
pcDNA5/FRT/TO Mammalian Expression Vector	Invitrogen	Fischer et al., 2013
pOG44	Invitrogen	Fischer et al., 2013
PB-CuO-MCS-IRES-GFP-EF1α-CymR-Puro cDNA Inducible Cloning and Expression Vector	System Biosciences	Cat# PBQM812A-1
Super PiggyBac Transposase Expression Vector	System Biosciences	Cat# PB210PA-1
pGEX-6p-1	GE Healthcare	Cat# 28954648
Software and Algorithms		
Fiji (<i>Fiji Is Just ImageJ</i> 1.51n)	Schindelin et al., 2012 Schneider et al., 2012	imagej.net/Fiji
Jalview 2.11.0	Waterhouse et al., 2009	jalview.org
PyMOL 2.3.4	The PyMOL Molecular Graphics System, Version 1.2r3pr, Schrödinger, LLC	pymol.org
Image Lab 5.2	Biorad Laboratories	
ImageQuantTL 8.1	GE Healthcare Life Sciences	
Clustal Omega	Sievers and Higgins, 2018 Madeira et al., 2019	
RCSB PDB	Berman et al., 2000 Berman et al., 2003	rcsb.org , wwpdb.org
JPred4	Drozdetskiy et al., 2015	www.compbio.dundee.ac.uk/jpred/

Table S2					
Cell lines	Plasmid	Gene	TAG	Reference	
Flp-In T-REx-293-Mock	pcDNA5/FRT/TO	-	-	Fischer et al., 2013	
Flp-In T-REx-293-MIA40 WT	pcDNA5/FRT/TO	ORF <i>MIA40</i>	Strep (C-terminal)	Fischer et al., 2013	
Flp-In T-REx-293-MIA40 Δ131	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ131-142	Strep (C-terminal)	this study	
Flp-In T-REx-293-MIA40 Δ122	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ122-142	Strep (C-terminal)	this study	
Flp-In T-REx-293-MIA40 Δ108	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ108-142	Strep (C-terminal)	this study	
Flp-In T-REx-293-MIA40 WT	pcDNA5/FRT/TO	ORF <i>MIA40</i>	Strep (N-terminal)	this study	
Flp-In T-REx-293-MIA40 Δ108	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ108-142	Strep (N-terminal)	this study	
Flp-In T-REx-293-MIA40 WT	pcDNA5/FRT/TO	ORF <i>MIA40</i>	-	this study	
Flp-In T-REx-293-MIA40 Δ131	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ131-142	-	this study	
Flp-In T-REx-293-MIA40 Δ122	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ122-142	-	this study	
Flp-In T-REx-293-MIA40 Δ108	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ108-142	-	this study	
Flp-In T-REx-293-MIA40 WT	pcDNA5/FRT/TO	ORF <i>MIA40</i>	HA (C-terminal)	this study	
Flp-In T-REx-293-MIA40 Δ131	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ131-142	HA (C-terminal)	this study	
Flp-In T-REx-293-MIA40 Δ122	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ122-142	HA (C-terminal)	this study	
Flp-In T-REx-293-MIA40 Δ108	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ108-142	HA (C-terminal)	this study	
Flp-In T-REx-293-MIA40 WT	pcDNA5/FRT/TO	ORF <i>MIA40</i>	HA (N-terminal)	this study	
Flp-In T-REx-293-MIA40 Δ108	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ108-142	HA (N-terminal)	this study	
Flp-In T-REx-293-MIA40 Δ108	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ1-40	HA (C-terminal)	this study	
Flp-In T-REx-293-MIA40 Δ108 + 14 positively charged aa	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ108-142 + 14 positively charged aa	-	this study	
Flp-In T-REx-293-MIA40 Δ108 + 14 neutral charged aa	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ108-142 + 14 positively charged aa	-	this study	
Flp-In T-REx-293-MIA40 Δ108 + GSS	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ108-142 + GSS	-	this study	
Flp-In T-REx-293-MIA40 Δ131 + 12 positively charged aa	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ131-142 + 12 neutral charged aa	-	this study	
Flp-In T-REx-293-MIA40 Δ131 + 12 neutral charged aa	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ131-142 + 12 neutral charged aa	-	this study	
Flp-In T-REx-293-MIA40 Δ131 + GSS	pcDNA5/FRT/TO	ORF <i>MIA40</i> Δ131-142 + GSS	-	this study	
Flp-In T-REx-293-COX19	pcDNA5/FRT/TO	ORF <i>COX19</i>	HA (C-terminal)	Fischer et al., 2013	
Flp-In T-REx-293-COX19-C-MIA	pcDNA5/FRT/TO	ORF <i>COX19</i> + aa108-142 <i>MIA40</i>	HA (C-terminal)	this study	
Flp-In T-REx-293-COX19-C-neutral	pcDNA5/FRT/TO	ORF <i>COX19</i> + 35 neutral charged aa	HA (C-terminal)	this study	
Flp-In T-REx-293-COX19 4CS	pcDNA5/FRT/TO	ORF <i>COX19</i> (G89,119,152,182►C)	HA (C-terminal)	Habich et al., 2019	
Flp-In T-REx-293-COX19 4CS-C-MIA	pcDNA5/FRT/TO	ORF <i>COX19</i> (G89,119,152,182►C) + aa108-142 <i>MIA40</i>	HA (C-terminal)	this study	
Flp-In T-REx-293-COX19 4CS-C-neutral	pcDNA5/FRT/TO	ORF <i>COX19</i> (G89,119,152,182►C) + 35 neutral charged aa	HA (C-terminal)	this study	

Flp-In T-REx-293-AIF^{MTS}-MIA40 WT	pcDNA5/FRT/TO	ORF <i>AIF^{MTS}-MIA40</i>	-	this study
Flp-In T-REx-293-AIF^{MTS}-MIA40 Δ108	pcDNA5/FRT/TO	ORF <i>AIF^{MTS}-MIA40 Δ108-142</i>	-	this study
Flp-In T-REx-293-SOD2	pcDNA5/FRT/TO	ORF <i>SOD2</i>	HA (C-terminal)	this study
HEK293T	-	-	-	Habich et al., 2019
HEK293T MIA40 knockout #2-Mock	PB-CuO-MCS- IRES-GFP-EF1α- CymR-Puro	-	-	Habich et al., 2019
HEK293T MIA40 knockout #2-MIA40 WT	PB-CuO-MCS- IRES-GFP-EF1α- CymR-Puro	ORF <i>MIA40</i>	-	Habich et al., 2019
HEK293T MIA40 knockout #2-C53S	PB-CuO-MCS- IRES-GFP-EF1α- CymR-Puro	ORF <i>MIA40</i> (T157►G)	-	Habich et al., 2019
HEK293T MIA40 knockout #2-MIA40 Δ131	PB-CuO-MCS- IRES-GFP-EF1α- CymR-Puro	ORF <i>MIA40 Δ131-142</i>	-	This study
HEK293T MIA40 knockout #2-MIA40 Δ122	PB-CuO-MCS- IRES-GFP-EF1α- CymR-Puro	ORF <i>MIA40 Δ131-122</i>	-	This study
HEK293T MIA40 knockout #2-MIA40 Δ108	PB-CuO-MCS- IRES-GFP-EF1α- CymR-Puro	ORF <i>MIA40 Δ131-108</i>	-	This study
Flp-In T-REx-293-YME1L1 ko	-	-	-	Hartmann et al, 2016
Flp-In T-REx-293-YME1L1 ko-Mock	PB-CuO-MCS- IRES-GFP-EF1α- CymR-Puro	-	-	This study
Flp-In T-REx-293-YME1L1 ko-MIA40 WT	PB-CuO-MCS- IRES-GFP-EF1α- CymR-Puro MIA40 WR	ORF <i>MIA40 WT</i>	-	This study
Flp-In T-REx-293-YME1L1 ko-MIA40 Δ108	PB-CuO-MCS- IRES-GFP-EF1α- CymR-Puro MIA40 Δ108	ORF <i>MIA40 Δ108-142</i>	-	This study
Flp-In T-REx-293-YME1L1 ko + YME1L1	-	-	-	Hartmann et al, 2016
Flp-In T-REx-293-YME1L1 ko + YME1L1	PB-CuO-MCS- IRES-GFP-EF1α- CymR-Puro	-	-	This study
Flp-In T-REx-293-YME1L1 ko + YME1L1-MIA40	PB-CuO-MCS- IRES-GFP-EF1α- CymR-Puro MIA40 WR	ORF <i>MIA40 WT</i>	-	This study
Flp-In T-REx-293-YME1L1 ko + YME1L1-MIA40 Δ108	PB-CuO-MCS- IRES-GFP-EF1α- CymR-Puro MIA40 Δ108	ORF <i>MIA40 Δ108-142</i>	-	This study
Flp-In T-REx-293	-	-	-	Thermo Fisher Scientific
Flp-In T-REx-293 -Mock	PB-CuO-MCS- IRES-GFP-EF1α- CymR-Puro	-	-	This study
Flp-In T-REx-293 -MIA40 WT	PB-CuO-MCS- IRES-GFP-EF1α- CymR-Puro MIA40 WR	ORF <i>MIA40 WT</i>	-	This study
Flp-In T-REx-293 -MIA40 Δ108	PB-CuO-MCS- IRES-GFP-EF1α- CymR-Puro MIA40 Δ108	ORF <i>MIA40 Δ108-142</i>	-	This study
Rosetta2 (DE3)-MIA40 WT	pGEX-6p-1	ORF <i>MIA40</i>	GST (N-terminal)	Friederich et al., 2017
Rosetta2 (DE3)-PreScission Protease	pGEX-6p-1	ORF PreScission Protease	GST (N-terminal)	Friederich et al., 2017
Rosetta2 (DE3)-MIA40 Δ131	pGEX-6p-1	ORF <i>MIA40 Δ131-142</i>	GST (N-terminal)	this study
Rosetta2 (DE3)-MIA40 Δ122	pGEX-6p-1	ORF <i>MIA40 Δ122-142</i>	GST (N-terminal)	this study
Rosetta2 (DE3)-MIA40 Δ108	pGEX-6p-1	ORF <i>MIA40 Δ108-142</i>	GST (N-terminal)	this study
Rosetta2 (DE3)-MIA40 WT	pRSET-A	ORF <i>MIA40</i>	6xHis (N-terminal)	this study
Rosetta2 (DE3)-MIA40 Δ108	pRSET-A	ORF <i>MIA40 Δ108-142</i>	6xHis (N-terminal)	this study

Table S3		
Oligonucleotides	Sequence (5'-3')	Restriction site
Fw for pcDNA5/FRT/TO MIA40 WT, MIA40 Δ131, MIA40 Δ122, MIA40 Δ108, MIA40 Δ131-Strep, MIA40 Δ122-Strep, MIA40 Δ108-Strep, MIA40-HA, MIA40 Δ131-HA, MIA40 Δ122-HA, MIA40 Δ108-HA, MIA40 108+14 pos. aa, MIA40 108+14 neut. aa, MIA40 108+GSS, MIA40 131+12 pos. aa, MIA40 131+12 neut. aa, MIA40 131+GSS	GGCGGATCCGCAGCCATGTCCTATTGCCG	BamHI
Rev for pcDNA5/FRT/TO MIA40 Δ131-Strep	CTACTCGAGTTATTTCTCAAATTTGTGGATGACTCCATC CTCCAGCAATGGGAGCTGTTTCTTCTG	XhoI
Rev for pcDNA5/FRT/TO MIA40 Δ122-Strep	CTACTCGAGTTATTTCTCAAATTTGTGGATGACTCCATCCTCCAG CTGCTGGCTTCTTCTCTTTCC	XhoI
Rev for pcDNA5/FRT/TO MIA40 Δ108-Strep	CAACTCGAGTTATTTCTCAAATTTGTGGATGACTCCATCCTCCAG CTTGGGGATAGAGGTCTGGGTATTCTGC	XhoI
Fw for pcDNA5/FRT/TO Strep-MIA40 WT, Strep-MIA40 Δ108	ATTGGATCCGCAGCCATGTGGAGTCATCCACAATTTGAGAAAG GAGCTTCTATTGCCGGCAGG	BamHI
Rev for pcDNA5/FRT/TO MIA40 WT, Strep-MIA40 WT, HA-MIA40 WT, AIF^{MTS}-MIA40 WT	CGGCTCGAGTTAACTTGATCCCTCTCTTTGGTTGCAGTGG	XhoI
Rev for pcDNA5/FRT/TO MIA40 Δ108, Strep-MIA40 Δ108, HA-MIA40 Δ108, AIF^{MTS}-MIA40 Δ108	CGGCTCGAGTTATTGGGGATAGAGGTCTGGGTATTCTGCATG CATTC	XhoI
Rev for pcDNA5/FRT/TO MIA40 Δ131	CGGCTCGAGTTAAATGGGAGCTGTTTCTTCTGCTTGTCTGC	XhoI
Rev for pcDNA5/FRT/TO MIA40 Δ122	CGGCTCGAGTTATGCTGGCTTCTTCTCTTTCTCTTCC	XhoI
Rev for pcDNA5/FRT/TO MIA40 WT-HA, MIA40 Δ40-HA	GTACTCGAGTTAAGCATAATCTGGAACATCATATGGATATCCA GCACTTGATCCCTCCTCTTC	XhoI
Fw for pcDNA5/FRT/TO MIA40 Δ40-HA	GATGGATCCGCAGCCATGGGATTGATACTGCCAAATGG	BamHI
Rev for pcDNA5/FRT/TO MIA40 Δ131-HA	GTACTCGAGTTAAGCATAATCTGGAACATCATATGGATATCCA GCAATGGGAGCTGTTTCTTCTGC	XhoI
Rev for pcDNA5/FRT/TO MIA40 Δ122-HA	GTACTCGAGTTAAGCATAATCTGGAACATCATATGGATATCCA GCTGCTGGCTTCTTCTCTTTCC	XhoI
Rev for pcDNA5/FRT/TO MIA40 Δ108-HA	GTACTCGAGTTAAGCATAATCTGGAACATCATATGGATATCCA GCTTGGGGATAGAGGTCTGG	XhoI
Fw pcDNA5/FRT/TO HA-MIA40 WT, HA-MIA40 Δ108	ATTGGATCCGCAGCCATGTATCCATATGATGTTCCAGATTATG CTGGAGCTTCTATTGCCGGCAGGAAGG	BamHI
Rev for pcDNA5/FRT/TO MIA40 Δ108+14 pos. aa	CTACTCGAGTTATGCTGGCTTCTTCTTCTCTTCTCTCTTCTCTT TCTTCTCTTTGGGGATAGAGGTCTGGG	XhoI
Rev for pcDNA5/FRT/TO MIA40 Δ108+14 neut. aa	ATACTCGAGTTATGCTGGTGAACCTGCAGCCCCTGAACCAGAA GCTCCAGAAGCTTGGGGATAGAGGTCTGGG	XhoI
Rev for pcDNA5/FRT/TO MIA40 Δ108+GSS	CGACTCGAGTTAACTTGATCCTTGGGGATAGAGGTCTGGGTA	XhoI
Rev for pcDNA5/FRT/TO MIA40 Δ131+12 pos. aa	GTACTCGAGTTAACTTGATCCACGCTTACGCTTAGTAGCTGTTG CCC GAATGGGAGCTGTTTCTTCTG	XhoI
Rev for pcDNA5/FRT/TO MIA40 Δ131+12 neut. aa	GTACTCGAGTTAACTTGATCCTGACGACGAGTAGTAGCTGTT GCAGCAATGGGAGCTGTTTCTTCTG	XhoI
Rev for pcDNA5/FRT/TO MIA40 Δ131+GSS	GTACTCGAGTTAACTTGATCCAAATGGGAGCTGTTTCTTCTGC	XhoI
Fw (1) for pcDNA5/FRT/TO COX19-C-MIA, pcDNA5/FRT/TO COX19-4SC-C-MIA, pcDNA5/FRT/TO COX19-C-neutral, pcDNA5/FRT/TO COX19-4SC-C-neutral	TATGGATCCGCCAACATGTCGACCGCCATGAATTC	BamHI
Fw (2) for pcDNA5/FRT/TO COX19-C-MIA, pcDNA5/FRT/TO COX19-4SC-C-MIA	GGAAAATCAGAGGCCAAAAAAGAGGATGAGGATGAGG	-
Rev (1) for pcDNA5/FRT/TO COX19-C-MIA, pcDNA5/FRT/TO COX19-4SC-C-MIA	CCTCATCTCATCTCTTTTTTGGCTCTGATTTCC	-
Rev (2) for pcDNA5/FRT/TO COX19-C-MIA, pcDNA5/FRT/TO COX19-4SC-C-MIA	GTACTCGAGTTAAGCATAATCTGGAACATCATATGGATATCCA GCACTTGATCCCTCCTCTTC	XhoI
Fw (2) for pcDNA5/FRT/TO COX19-C-neutral,	GGAAAATCAGAGGCCAAAAAAGCTTCTGGAGCTTCTGGTTCT	-