

SUPPLEMENTAL MATERIAL

RNA analyses

Total RNA was isolated and reverse transcribed with AffinityScript QPCR cDNA Synthesis Kit (Agilent Technologies). PCR reactions were performed in triplicate in a 96-well format using the MX3005P (Stratagene). The primer sets (SYBR green) used to detect specific gene expression are as follows: mBDH1 fwd-TCTCGGACTGCCCTGCGCTAT, rev-ACCGCTGTTGCAGTAGGTTT; m36B4 fwd-TGGAAGTCCAACACTTCCTCAA m36B4 rev-ATCTGCTGCATCTGCTTGGAG; mMCT1 fwd-TGCAACGACCAGTGAAGTATC, rev-GACAACCACCAGCGATCATTA; mMCT2 fwd-ATACTTGAGGTCCCTCTCATT, rev-GGAAGAGGCAGACAACGATAA. 36B4 primer set was included in a separate well (in triplicate) and used to normalize the gene expression data.

Proteomics using Stable Isotope Labeling by Amino Acids (SILAC)

Crude mitochondrial fractions were prepared from cardiac tissue of Lys6 (¹³C6-Lysine, Silantes) labeled (heavy) and non-labeled (light) mice. Mitochondrial fractions were prepared as previously described.¹ Briefly, immediately following euthanasia, the mouse ventricles were dissected, washed and placed in ice-cold isolation buffer (220 mM mannitol, 70 mM sucrose, 5mM HEPES-KOH, pH 7.4, 1mM EGTA, 1mg/ml BSA and protease inhibitor cocktail). The tissue was then minced and homogenized using a Potter-Elvehjem glass/Teflon homogenizer. A crude mitochondrial fraction was extracted from the homogenate using differential centrifugation and resuspended in small amount of the isolation buffer (300 µl).

A 1:1 mixture of heavy and light heart mitochondrial fractions were then separated by gel electrophoresis on precast 4–12% NuPAGE gradient gels (Invitrogen) and stained with the

Colloidal Blue Staining Kit (Invitrogen). Evenly sized gel pieces were excised and processed for mass spectrometry. The gel pieces were subjected to in-gel reduction and alkylation, followed by LysC (Wako) digestion as described previously.² In brief, trypsin digested gel pieces were washed twice with 50% 50 mM NH₄HCO₃ eluent additive for LC-MS (Sigma-Aldrich) / 50% ethanol for 20 min, and dehydrated with 100% ethanol for 10 min, and then vacuum centrifuged. Gel pieces were reduced with 10 mM DTT for 45 min at 56°C and alkylated with 55 mM iodoacetamide for 30 min at RT in the dark. After two cycles of washing and dehydration, samples were dehydrated twice with 100% ethanol for 15 min and vacuum centrifuged. Gel pieces were digested overnight at 37°C in 50 µl of digestion buffer containing 12.5 ng/µl of LysC (Wako).

Released peptides were extracted once with 30% acetonitrile/ 3% trifluoracetic acid (TFA), twice with 70% acetonitrile, followed by two final extractions with 100% acetonitrile. Extracts were vacuum centrifuged to remove acetonitrile and subsequently acidified with 0.5% TFA. Peptides were desalted and concentrated with homemade "STAGE" tips (Stop and Go extraction tips) filled with C-18 (C18 Empore Disks, 3M) as described.³

Mass spectrometric experiments were performed on a nano-flow HPLC system (Agilent) connected to a LTQ-Orbitrap XL instrument (Thermo Scientific) equipped with a nanoelectrospray source (Proxeon). The mass spectrometer was operated in the data dependent mode to monitor MS and MS/MS spectra. Survey full-scan MS spectra (from m/z 300–2000) were acquired in the Orbitrap with a resolution of R=60,000 at m/z 400 after accumulation of 1,000,000 ions. The five most intense ions from the preview survey scan delivered by the Orbitrap were sequenced by collision-induced dissociation (CID) in the LTQ. Mass spectra were analyzed using MaxQuant software (Version 1.0.14.10)⁴ and all tandem mass spectra were

searched against the mouse International Protein Index protein sequence database (IPI version 3.54) and concatenated with reversed copies of all sequences. The required false positive rate was set to 1% at the protein and peptide level. Maximum allowed mass deviation was set to 7 ppm in MS mode and 0.5 Da for MS/MS peaks. The parameter settings were: LysC as digesting enzyme, a maximum of two missed cleavages, a minimum of six amino acids, carbamidomethylation at cysteine residues as fixed and oxidation at methionine residues as variable modifications.

Metabolomic analysis of organic acids and acylcarnitines

Immediately following deep anesthesia by intraperitoneal injection of pentobarbital (100 mg/kg body weight), bi-ventricle was excised and frozen. Specimens of powdered bi-ventricle tissue were diluted 20-fold (mass:volume) in 50% acetonitrile supplemented with 0.3% formate (acylcarnitines, amino acids, and organic acids). Samples were homogenized in a TissueLyser II (Qiagen). Tissue extracts were derivatized and analyzed as previously described.⁵ Levels of succinate, C4OH-carnitine, and C2-carnitine were determined using stable isotope dilution techniques. The data were acquired using a Waters AcquityTM UPLC system equipped with a TQ (triple quadrupole) detector and a data system controlled by MassLynx 4.1 operating system (Waters Corporation). Metabolites were quantified using methods described previously.⁶

Supplemental Table 1: Mitochondrial proteins identified by proteomic profiling in compensated hypertrophy (CH) and/or heart failure (HF) samples.

ID	Gene Name
IPI00387379	2,4-dienoyl CoA reductase 1, mitochondrial
IPI00379694, IPI00127625	3-hydroxy-3-methylglutaryl-Coenzyme A lyas
IPI00330754, IPI00890322, IPI00857778	3-hydroxybutyrate dehydrogenase, type 1
IPI00885424, IPI00154047	3-hydroxyisobutyryl-Coenzyme A hydrolase
IPI00132653, IPI00856651, IPI00858156	3-oxoacid CoA transferase 1
IPI00606168, IPI00856229, IPI00227445, IPI00407499	4-aminobutyrate aminotransferase
IPI00648312	5'-nucleotidase domain containing 3
IPI00648235, IPI00187512	5'-nucleotidase, cytosolic II
IPI00230589, IPI00230591, IPI00230590, IPI00894854, IPI00890007, IPI00230593, IPI00115506, IPI00230592	A kinase (PRKA) anchor protein 1
IPI00323406	ATP synthase mitochondrial F1 complex assembly factor 1
IPI00336348	ATP synthase mitochondrial F1 complex assembly factor 2; similar to ATP synthase mitochondrial F1 complex assembly factor 2
IPI00468481	ATP synthase, H ⁺ transporting mitochondrial F1 complex, beta subunit
IPI00856424, IPI00125460	ATP synthase, H ⁺ transporting, mitochondrial F0 complex, subunit F pseudogene; similar to ATP synthase coupling factor 6, mitochondrial precursor (ATPase subunit F6); ATP synthase, H ⁺ transporting, mitochondrial F0 complex, subunit F
IPI00341282	ATP synthase, H ⁺ transporting, mitochondrial F0 complex, subunit b, isoform 1; predicted gene 12231
IPI00857439, IPI00130280	ATP synthase, H ⁺ transporting, mitochondrial F1 complex, alpha subunit, isoform 1
IPI00750074, IPI00313475, IPI00775853, IPI00776084, IPI00776275, IPI00751391, IPI00776312	ATP synthase, H ⁺ transporting, mitochondrial F1 complex, gamma polypeptide 1
IPI00761646, IPI00672663, IPI00816848	ATP-binding cassette, sub-family B (MDR/TAP), member 7
IPI00856267, IPI00856425, IPI00123422, IPI00459383	ATP-binding cassette, sub-family B (MDR/TAP), member 8
IPI00135646, IPI00535376	ATP-binding cassette, sub-family D (ALD), member 3
IPI00108410	ATPase family, AAA domain containing 1
IPI00126913, IPI00464208	ATPase family, AAA domain containing 3A
IPI00127598, IPI00331463	ATPase inhibitory factor 1
IPI00880712, IPI00880589, IPI00880341, IPI00357510, IPI00357511, IPI00124900	AU RNA binding protein/enoyl-coenzyme A hydratase
IPI00606647, IPI00321499	BCL2-like 13 (apoptosis facilitator)
IPI00128346	CDGSH iron sulfur domain 1
IPI00345740, IPI00649328, IPI00649725	CDGSH iron sulfur domain 3
IPI00222203, IPI00874655, IPI00267596	COX15 homolog, cytochrome c oxidase assembly protein (yeast)
IPI00830383, IPI00830273, IPI00467840, IPI00830443	ClpB caseinolytic peptidase B homolog (E. coli)
IPI00128261	Cytochrome b
IPI00130376, IPI00355248	Cytochrome c oxidase subunit 1
IPI00116896	Cytochrome c oxidase subunit 3; ATP synthase subunit a; ATP synthase protein 8
IPI00347634, IPI00120414, IPI00279858	DnaJ (Hsp40) homolog, subfamily A, member 3
IPI00648191, IPI00226466, IPI00828649	DnaJ (Hsp40) homolog, subfamily C, member 11
IPI00624902, IPI00468992, IPI00111111, IPI00881254	DnaJ (Hsp40) homolog, subfamily C, member 19
IPI00137424	ECSIT homolog (Drosophila)
IPI00828369, IPI00474134, IPI00153350	FAD-dependent oxidoreductase domain containing 1
IPI00267299, IPI00460703	FAST kinase domains 2
IPI00881994, IPI00230283	G elongation factor, mitochondrial 1
IPI00338876, IPI00269240	G elongation factor, mitochondrial 2
IPI00622170, IPI00225294, IPI00336569	GUF1 GTPase homolog (S. cerevisiae)
IPI00117083	GrpE-like 1, mitochondrial
IPI00115598, IPI00230550	H2-K region expressed gene 6
IPI00331517	HIG1 domain family, member 1A
IPI00170051, IPI00881059	HscB iron-sulfur cluster co-chaperone homolog (E. coli)
IPI00110578, IPI00856218, IPI00856829	IscU iron-sulfur cluster scaffold homolog (E. coli); similar to nitrogen fixation cluster-like
IPI00130331	L-2-hydroxyglutarate dehydrogenase; predicted gene 7842
IPI00471097	LYR motif containing 5
IPI00117832, IPI00918067	LYR motif containing 7
IPI00122740	MACRO domain containing 1
IPI00123276	MOCO sulphurase C-terminal domain containing 2
IPI00222753	MTERF domain containing 3
IPI00116748	NADH dehydrogenase (ubiquinone) 1 alpha subcomplex 10
IPI00318645	NADH dehydrogenase (ubiquinone) 1 alpha subcomplex 11
IPI00344004	NADH dehydrogenase (ubiquinone) 1 alpha subcomplex, 12
IPI00230715	NADH dehydrogenase (ubiquinone) 1 alpha subcomplex, 13
IPI00315302	NADH dehydrogenase (ubiquinone) 1 alpha subcomplex, 2
IPI00858238, IPI00132216	NADH dehydrogenase (ubiquinone) 1 alpha subcomplex, 3
IPI00125929	NADH dehydrogenase (ubiquinone) 1 alpha subcomplex, 4
IPI00880753, IPI00331332	NADH dehydrogenase (ubiquinone) 1 alpha subcomplex, 5
IPI00133399	NADH dehydrogenase (ubiquinone) 1 alpha subcomplex, 6 (B14)
IPI00130322	NADH dehydrogenase (ubiquinone) 1 alpha subcomplex, 7 (B14.5a)

IPI00120984	NADH dehydrogenase (ubiquinone) 1 alpha subcomplex, 8
IPI00120212	NADH dehydrogenase (ubiquinone) 1 alpha subcomplex, 9
IPI00226687	NADH dehydrogenase (ubiquinone) 1 alpha subcomplex, assembly factor 1
IPI00330551	NADH dehydrogenase (ubiquinone) 1 alpha subcomplex, assembly factor 2
IPI00648625, IPI00133744	NADH dehydrogenase (ubiquinone) 1 alpha subcomplex, assembly factor 4
IPI00133403	NADH dehydrogenase (ubiquinone) 1 beta subcomplex 3; predicted gene 3192
IPI00132390	NADH dehydrogenase (ubiquinone) 1 beta subcomplex 4; predicted gene 3873; predicted gene 3244
IPI00387430	NADH dehydrogenase (ubiquinone) 1 beta subcomplex 8
IPI00880613, IPI00121288	NADH dehydrogenase (ubiquinone) 1 beta subcomplex, 10
IPI00881846, IPI00881750, IPI00882364, IPI00132531, IPI00882043	NADH dehydrogenase (ubiquinone) 1 beta subcomplex, 5
IPI00648743, IPI00341322	NADH dehydrogenase (ubiquinone) 1 beta subcomplex, 6
IPI00133215	NADH dehydrogenase (ubiquinone) 1 beta subcomplex, 7
IPI00132623	NADH dehydrogenase (ubiquinone) 1 beta subcomplex, 9
IPI00133006, IPI00918301, IPI00918079	NADH dehydrogenase (ubiquinone) 1, alpha/beta subcomplex, 1; predicted gene 445 ^c
IPI00132050, IPI00110825	NADH dehydrogenase (ubiquinone) 1, subcomplex unknown, 2; similar to NADH dehydrogenase (ubiquinone) 1, subcomplex unknown, 2
IPI00308882	NADH dehydrogenase (ubiquinone) Fe-S protein 1
IPI00830766, IPI00128023, IPI00830872	NADH dehydrogenase (ubiquinone) Fe-S protein 2
IPI00229008	NADH dehydrogenase (ubiquinone) Fe-S protein 4
IPI00649019, IPI00117300	NADH dehydrogenase (ubiquinone) Fe-S protein 5; cDNA sequence BC002163
IPI00120232	NADH dehydrogenase (ubiquinone) Fe-S protein 7
IPI00170093	NADH dehydrogenase (ubiquinone) Fe-S protein 8
IPI00130460	NADH dehydrogenase (ubiquinone) flavoprotein 1
IPI00169925, IPI00845716	NADH dehydrogenase (ubiquinone) flavoprotein 2
IPI00128285	NADH dehydrogenase (ubiquinone) flavoprotein 3
IPI00341550	NADH-ubiquinone oxidoreductase chain 1
IPI00116843	NADH-ubiquinone oxidoreductase chain 4L; NADH-ubiquinone oxidoreductase chain 4
IPI00881776	NFU1 iron-sulfur cluster scaffold homolog (S. cerevisiae); predicted gene 785 ^c
IPI00265701	NLR family member X1
IPI00407938, IPI00133608	OCIA domain containing 1
IPI00111512	OMA1 homolog, zinc metallopeptidase (S. cerevisiae)
IPI00857491, IPI00857001, IPI00457976	PET112-like (yeast)
IPI00650052, IPI00649794, IPI00895414, IPI00117264, IPI00894710, IPI00894598, IPI00649406, IPI00895518, IPI00648764, IPI00894769	Parkinson disease (autosomal recessive, early onset) 7
IPI00133706	RAB1B, member RAS oncogene family
IPI00850133, IPI00900458, IPI00762463	RIKEN cDNA 1300010F03 gene
IPI00108431, IPI00785241, IPI00918883	RIKEN cDNA 1500001M20 gene
IPI00132478	RIKEN cDNA 1700021F05 gene
IPI00113080	RIKEN cDNA 1700034H14 gene
IPI00459357	RIKEN cDNA 1700034I23 gene; predicted gene 10731
IPI00654397, IPI00133220, IPI00458085, IPI00109206, IPI00170212	RIKEN cDNA 1810020G14 gene; synaptojanin 2 binding protein; predicted gene 4116
IPI00113143	RIKEN cDNA 2010107E04 gene
IPI00110460, IPI00869373	RIKEN cDNA 2310003L22 gene
IPI00750687, IPI00222284	RIKEN cDNA 2310005E10 gene
IPI00270788	RIKEN cDNA 2310028O11 gene
IPI00153792	RIKEN cDNA 2410015M20 gene
IPI00880887, IPI00173167	RIKEN cDNA 2410091C18 gene
IPI00895415, IPI00453792	RIKEN cDNA 4930402E16 gene
IPI00221782	RIKEN cDNA 4933403F05 gene
IPI00308195, IPI00416621, IPI00856408, IPI00857151	RIKEN cDNA 9030617O03 gene
IPI00856512, IPI00267983	RIKEN cDNA 9330129D05 gene
IPI00221850	RIKEN cDNA 9430016H08 gene
IPI00228106	RIKEN cDNA A230051G13 gene
IPI00471368	RNA (guanine-9-) methyltransferase domain containing 1
IPI00761959, IPI00515530	SCO cytochrome oxidase deficient homolog 1 (yeast)
IPI00132762	TNF receptor-associated protein 1
IPI00113052, IPI00885494	Ts translation elongation factor, mitochondrial
IPI00136555	YME1-like 1 (S. cerevisiae)
IPI00132696, IPI00788370	aarF domain containing kinase 1
IPI00226414, IPI00886271, IPI00404935	abhydrolase domain containing 10
IPI00858299, IPI00170213	abhydrolase domain containing 11
IPI00154054	acetyl-Coenzyme A acetyltransferase 1
IPI00226430, IPI00653158	acetyl-Coenzyme A acyltransferase 2 (mitochondrial 3-oxoacyl-Coenzyme A thiolase
IPI00624175	acid phosphatase 6, lysophosphatidic
IPI00116074	aconitase 2, mitochondrial
IPI00122633	acyl-CoA synthetase family member 2
IPI00654091, IPI00762636	acyl-CoA synthetase family member 2

IPI00856216, IPI00112549, IPI00857822, IPI00556840,
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 IPI00169862
 IPI00655156, IPI00353887

acyl-CoA synthetase long-chain family member 1
 acyl-CoA synthetase long-chain family member 2
 acyl-CoA synthetase short-chain family member 1
 acyl-CoA thioesterase 1
 acyl-CoA thioesterase 10
 acyl-CoA thioesterase 13
 acyl-CoA thioesterase 2
 acyl-CoA thioesterase 9
 acyl-Coenzyme A dehydrogenase family, member 1α
 acyl-Coenzyme A dehydrogenase family, member 1β
 acyl-Coenzyme A dehydrogenase family, member 1γ
 acyl-Coenzyme A dehydrogenase, long-chair
 acyl-Coenzyme A dehydrogenase, medium chair
 acyl-Coenzyme A dehydrogenase, short chair
 acyl-Coenzyme A dehydrogenase, short/branched chair
 acyl-Coenzyme A dehydrogenase, very long chair
 acyl-Coenzyme A oxidase 1, palmitoyl
 adenylate kinase 1
 adenylate kinase 3
 alcohol dehydrogenase, iron containing, 1
 aldehyde dehydrogenase 1 family, member L1
 aldehyde dehydrogenase 1 family, member L2
 aldehyde dehydrogenase 2, mitochondrial
 aldehyde dehydrogenase 4 family, member A1
 aldehyde dehydrogenase 9, subfamily A1
 aldehyde dehydrogenase family 6, subfamily A1
 aldehyde dehydrogenase family 7, member A1
 aldehyde dehydrogenase family 5, subfamily A1
 aldo-keto reductase family 1, member B7
 aldolase C, fructose-bisphosphate
 alkylglycerone phosphate synthase
 alpha-methylacyl-CoA racemase
 aminolevulinic acid synthase 1
 apolipoprotein A-I binding protein
 apolipoprotein O-like
 apoptosis-inducing factor, mitochondrion-associated 1
 arginyl-tRNA synthetase 2, mitochondrial
 asparaginyl-tRNA synthetase 2 (mitochondrial)(putative)
 aspartyl-tRNA synthetase 2 (mitochondrial)
 biphenyl hydrolase-like (serine hydrolase, breast epithelial mucin-associated antigen
 brain protein 44; similar to brain protein 44; predicted gene 3982
 branched chain aminotransferase 2, mitochondrial
 branched chain ketoacid dehydrogenase E1, alpha polypeptide
 branched chain ketoacid dehydrogenase E1, beta polypeptide; similar to 3-methyl-2-oxobutanoate dehydrogenase
 cDNA sequence AK157302
 carbonyl reductase 2
 carbonyl reductase 4
 carnitine acetyltransferase

 carnitine palmitoyltransferase 1a, liver
 carnitine palmitoyltransferase 1b, muscle
 carnitine palmitoyltransferase 2

 caseinolytic peptidase X (E.coli)
 caseinolytic peptidase, ATP-dependent, proteolytic subunit homolog (E. coli)
 catalase
 cathepsin B
 cathepsin D
 caveolin 1, caveolae protein
 chaperone, ABC1 activity of bc1 complex like (S. pombe)
 chaperonin containing Tep1, subunit 7 (eta)
 citrate lyase beta like
 citrate synthase
 citrate synthase like
 clathrin, heavy polypeptide (Hc)
 coenzyme Q3 homolog, methyltransferase (yeast)
 coenzyme Q5 homolog, methyltransferase (yeast)
 coenzyme Q6 homolog (yeast)
 coenzyme Q9 homolog (yeast)
 coiled-coil domain containing 109A

IPI00137601
 IPI00153794
 IPI00854928, IPI00110180, IPI00854953
 IPI00754739, IPI00133562, IPI00757033
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 IPI00554929, IPI00229080
 IPI00117281, IPI00660262, IPI00463220, IPI00227783

coiled-coil domain containing 44
 coiled-coil domain containing 58
 coiled-coil domain containing 90A
 coiled-coil-helix-coiled-coil-helix domain containing 3; similar to coiled-coil-helix-coiled-coil-helix domain containing
 coiled-coil-helix-coiled-coil-helix domain containing 4
 complement component 1, q subcomponent binding protein
 coproporphyrinogen oxidase
 creatine kinase, mitochondrial 1, ubiquitous
 creatine kinase, mitochondrial 2
 cysteine conjugate-beta lyase 2
 cysteinyl-tRNA synthetase 2 (mitochondrial)(putative)
 cytochrome P450, family 27, subfamily a, polypeptide 1
 cytochrome b5 reductase 1
 cytochrome b5 reductase 3
 cytochrome c oxidase subunit IV isoform 1
 cytochrome c oxidase subunit VIIa polypeptide 2-like; predicted gene 6969
 cytochrome c oxidase, subunit VI a, polypeptide 2
 cytochrome c oxidase, subunit VIIa 1
 cytochrome c oxidase, subunit VIIb polypeptide 1
 cytochrome c oxidase, subunit Va
 cytochrome c oxidase, subunit XVII assembly protein homolog (yeast)
 cytochrome c, testis
 cytochrome c-1
 death associated protein 3
 dehydrogenase/reductase (SDR family) member 1
 dehydrogenase/reductase (SDR family) member 4
 demethyl-Q 7
 dihydrolipoamide S-acetyltransferase (E2 component of pyruvate dehydrogenase complex
 dihydrolipoamide S-succinyltransferase (E2 component of 2-oxo-glutarate complex
 dihydrolipoamide branched chain transacylase E2
 dihydrolipoamide dehydrogenase
 dihydroorotate dehydrogenase
 dodecenoyl-Coenzyme A delta isomerase (3,2 trans-enoyl-Coenzyme A isomerase
 electron transferring flavoprotein, dehydrogenase
 enoyl Coenzyme A hydratase domain containing 3
 enoyl Coenzyme A hydratase, short chain, 1, mitochondria
 enoyl coenzyme A hydratase 1, peroxisoma
 enoyl-Coenzyme A, hydratase/3-hydroxyacyl Coenzyme A dehydrogenas
 ethylmalonic encephalopathy 1
 family with sequence similarity 162, member A
 family with sequence similarity 82, member B
 ferredoxin reductase
 ferrochelatase
 fission 1 (mitochondrial outer membrane) homolog (yeast)
 frataxin
 fumarate hydratase 1
 fumarylacetoacetate hydrolase domain containing 1
 fumarylacetoacetate hydrolase domain containing 2A
 glioblastoma amplified sequence
 glutamate dehydrogenase 1; predicted gene 5902
 glutamate oxaloacetate transaminase 2, mitochondrial
 glutaryl-Coenzyme A dehydrogenase
 glutathione S-transferase kappa 1
 glutathione transferase zeta 1 (maleylacetoacetate isomerase)
 glycerol kinase
 glycerol kinase 2
 glycerol phosphate dehydrogenase 2, mitochondrial
 glycerol-3-phosphate dehydrogenase 1 (soluble)
 glycyl-tRNA synthetase
 glyoxalase domain containing 4
 growth factor, erv1 (*S. cerevisiae*)-like (augmenter of liver regeneration)
 growth hormone inducible transmembrane protein
 heat shock protein 1 (chaperonin 10); predicted gene, EG628438; heat shock protein 1 (chaperonin 10), related sequence 1;
 predicted gene 2903
 heat shock protein 1B; heat shock protein 1A; heat shock protein 1-like
 heat shock protein 2
 heat shock protein 9
 heat shock protein 90 alpha (cytosolic), class B member 1
 heterogeneous nuclear ribonucleoprotein L-like; glutathione peroxidase 4

IPI00762858, IPI00283612, IPI00759938, IPI00468553, hexokinase 1
 IPI00283611
 IPI00114342
 IPI00133034
 IPI00116138
 IPI00134572, IPI00309285 histidine triad nucleotide binding protein 2
 IPI00880410, IPI00880581, IPI00115866, IPI00882305 histidyl-tRNA synthetase 2, mitochondrial (putative)
 IPI00121105 holocytchrome c synthetase
 IPI00223092 hydroxyacyl glutathione hydrolase
 hydroxyacyl-Coenzyme A dehydrogenase
 hydroxyacyl-Coenzyme A dehydrogenase/3-ketoacyl-Coenzyme A thiolase/enoyl-Coenzyme A hydratase (trifunctional protein), alpha subunit
 hydroxysteroid (17-beta) dehydrogenase 1C
 hydroxysteroid (17-beta) dehydrogenase 4
 hydroxysteroid dehydrogenase like 2
 hypothetical protein LOC675054; RIKEN cDNA 2900010M23 gene
 inner membrane protein, mitochondrial

IPI00320847, IPI00830581, IPI00626132 insulin degrading enzyme
 IPI00331628 iron-sulfur cluster assembly 1 homolog (S. cerevisiae)
 IPI00649718, IPI00117214
 IPI00133350, IPI00894954, IPI00894692 isocitrate dehydrogenase 2 (NADP+), mitochondrial
 IPI00555088, IPI00381412, IPI00228150, IPI00381413, isocitrate dehydrogenase 3 (NAD+) alpha
 IPI00554845 isocitrate dehydrogenase 3 (NAD+) beta
 IPI00119784, IPI00828796, IPI00407289 isocitrate dehydrogenase 3 (NAD+), gamma
 IPI00720014 isoleucine-tRNA synthetase 2, mitochondrial; similar to isoleucine-tRNA synthetase 2, mitochondria
 IPI00857308, IPI00318614, IPI00875110 isovaleryl coenzyme A dehydrogenase
 IPI00608078, IPI00459725 lactate dehydrogenase B; predicted gene 5514
 IPI00126635 lactation elevated 1
 IPI00109169 leucine aminopeptidase 3
 IPI00453499, IPI00830173, IPI00720234 leucine zipper-EF-hand containing transmembrane protein 1
 IPI00471246 leucine-rich PPR-motif containing
 IPI00280156, IPI00229510, IPI00776197 leucyl-tRNA synthetase, mitochondrial
 IPI00885799, IPI00885728, IPI00830170, IPI00169752 lon peptidase 1, mitochondrial
 IPI00331436, IPI00828469 lysophospholipase 1
 IPI00131177 malate dehydrogenase 1, NAD (soluble)
 IPI00875457, IPI00420706 malate dehydrogenase 2, NAD (mitochondrial)
 IPI00123138 malic enzyme 3, NADP(+)-dependent, mitochondria
 IPI00761408, IPI00319518 malonyl-CoA decarboxylase
 IPI00130018, IPI00831119, IPI00400117, IPI00831019 metaxin 2
 IPI00336324, IPI00649695 methionine sulfoxide reductase A
 IPI00323592 methionine sulfoxide reductase B2
 IPI00308976, IPI00415361 methylcrotonoyl-Coenzyme A carboxylase 1 (alpha)
 IPI00759857, IPI00114866 methylcrotonoyl-Coenzyme A carboxylase 2 (beta)
 IPI00225254, IPI00874889 methylenetetrahydrofolate dehydrogenase (NADP+ dependent), methenyltetrahydrofolate cyclohydrolase,
 IPI00475322, IPI00331442 formyltetrahydrofolate synthase
 IPI00109655 methylmalonic aciduria (cobalamin deficiency) type A
 IPI00652521, IPI00881206, IPI00320850 methylmalonic aciduria (cobalamin deficiency) type B homolog (human)
 IPI00134792, IPI00553717 methylmalonyl CoA epimerase
 IPI00122862 methylmalonyl-Coenzyme A mutase
 IPI00312720 mitochondrial ribosomal protein S7
 IPI00750681, IPI00653086, IPI00283203 mitochondrial antiviral signaling protein
 IPI00133776 mitochondrial carrier homolog 2 (C. elegans); predicted gene, 100039384; predicted gene, 100039506
 IPI00133553, IPI00850737 mitochondrial intermediate peptidase; similar to Mipep protein
 IPI00330688 mitochondrial poly(A) polymerase
 IPI00122075 mitochondrial ribosomal protein L1
 IPI00894752, IPI00132039, IPI00807902, IPI00750596 mitochondrial ribosomal protein L10
 IPI00222838, IPI00753039 mitochondrial ribosomal protein L11
 IPI00459344 mitochondrial ribosomal protein L12
 IPI00720216, IPI00118237 mitochondrial ribosomal protein L13
 IPI00132412, IPI00648692 mitochondrial ribosomal protein L15
 IPI00132470 mitochondrial ribosomal protein L16
 IPI00118963 mitochondrial ribosomal protein L17
 IPI00134011 mitochondrial ribosomal protein L18
 IPI00798614, IPI00131732 mitochondrial ribosomal protein L19
 IPI00118230 mitochondrial ribosomal protein L20
 IPI00271726, IPI00919171 mitochondrial ribosomal protein L21
 IPI00857677, IPI00118235, IPI00856509 mitochondrial ribosomal protein L22
 IPI00858168, IPI00311406 mitochondrial ribosomal protein L23
 IPI00881504, IPI00323669 mitochondrial ribosomal protein L39
 IPI00665641, IPI00607969 mitochondrial ribosomal protein L40
 IPI00132895, IPI00474272, IPI00856299 mitochondrial ribosomal protein L44
 IPI00111211 mitochondrial ribosomal protein L45
 IPI00322422 mitochondrial ribosomal protein L46
 IPI00274131, IPI00874379 mitochondrial ribosomal protein L47
 IPI00131988, IPI00271430 mitochondrial ribosomal protein L48
 IPI00132170 mitochondrial ribosomal protein L49; similar to mitochondrial ribosomal protein L49
 IPI00654010, IPI00284978 mitochondrial ribosomal protein L51
 mitochondrial ribosomal protein L9

IPI00895548, IPI00895482, IPI00348248, IPI00895361	mitochondrial ribosomal protein S10
IPI00321858	mitochondrial ribosomal protein S15
IPI00320963, IPI00880873, IPI00556725	mitochondrial ribosomal protein S18A
IPI00223360, IPI00118193	mitochondrial ribosomal protein S18B
IPI00110918	mitochondrial ribosomal protein S22
IPI00649272, IPI00882119, IPI00882252, IPI00515295,	mitochondrial ribosomal protein S23; similar to Mrps23 protein
IPI00754363, IPI00853715, IPI00284934	
IPI00222514, IPI00462763	mitochondrial ribosomal protein S27
IPI00110672	mitochondrial ribosomal protein S28
IPI00132504	mitochondrial ribosomal protein S30
IPI00124828	mitochondrial ribosomal protein S31
IPI00894878, IPI00222538	mitochondrial ribosomal protein S35
IPI00269020	mitochondrial ribosomal protein S6
IPI00461565, IPI00855075, IPI00336292	mitochondrial ribosomal protein S9
IPI00109501	mitochondrial ribosome recycling factor; similar to mitochondrial ribosome recycling factor; predicted gene 637-
IPI00648884, IPI00121276	mitochondrial trans-2-enoyl-CoA reductase
IPI00776049, IPI00649361, IPI00649863, IPI00551118,	mitochondrial translational initiation factor 2
IPI00649625, IPI00130227	
IPI00649455, IPI00308263, IPI00649724	mitochondrial ubiquitin ligase activator of NFKB 1
IPI00653064, IPI00881102, IPI00881801, IPI00135579	mitofusin 1
IPI00226140	monoamine oxidase B
IPI00121107	myosin, light chain 10, regulatory
IPI00894940, IPI00407962, IPI00330747	nephronophthisis 3 (adolescent); acyl-Coenzyme A dehydrogenase family, member 11
IPI00314829, IPI00405682, IPI00874680	neurolysin (metallopeptidase M3 family)
IPI00115459, IPI00858044	nicotinamide nucleotide adenyllyltransferase 3
IPI00762127, IPI00874685, IPI00875011, IPI00830812	nicotinamide nucleotide transhydrogenase
IPI00119945	nitrilase family, member 2
IPI00760006, IPI00311072	nitrogen fixation gene 1 (<i>S. cerevisiae</i>); similar to Nitrogen fixation gene 1 (<i>S. cerevisiae</i>)
IPI00807996, IPI00110522	nudix (nucleoside diphosphate linked moiety X)-type motif 13
IPI00312507	optic atrophy 3 (human)
IPI00129178	ornithine aminotransferase
IPI00308001	oxidase assembly 1-like
IPI00123765, IPI00857222	oxidoreductase NAD-binding domain containing 1
IPI00460118, IPI00719841, IPI00845652, IPI00626237,	oxoglutarate dehydrogenase (lipoyamide)
IPI00420882	
IPI00126857	pentatricopeptide repeat domain 2
IPI00338458	pentatricopeptide repeat domain 3
IPI00750433, IPI00120199	peptidase (mitochondrial processing) alpha
IPI00274656	peptidase (mitochondrial processing) beta
IPI00116228	peptidylprolyl isomerase F (cyclophilin F)
IPI00649645, IPI00848536, IPI00648105, IPI00605254,	peroxiredoxin 1; predicted gene 7204
IPI00121788, IPI00648615	
IPI00116192	peroxiredoxin 3
IPI00828909	peroxiredoxin 4
IPI00475031, IPI00759999, IPI00129517	peroxiredoxin 5
IPI00322931, IPI00877214	peroxisomal delta3, delta2-enoyl-Coenzyme A isomerase
IPI00459332, IPI00720114, IPI00387505	phenylalanine-tRNA synthetase 2 (mitochondrial)
IPI00788337, IPI00788353, IPI00170126	pitrilysin metallopeptidase 1
IPI00654405, IPI00117689	polymerase I and transcript release factor
IPI00315808	predicted gene 10078; predicted gene 3544; similar to mitochondrial ribosomal protein S36; mitochondrial ribosomal protein S36; predicted gene 7258; predicted gene 4676
IPI00222419, IPI00265239	predicted gene 10108; cytochrome c, somatic; predicted gene 10053; similar to cytochrome c
IPI00133342, IPI00876323	predicted gene 10221; predicted gene 4602; predicted gene 9391; ATP synthase, H ⁺ transporting, mitochondrial FO complex, subunit G2, pseudogene; predicted gene 11477; predicted gene 9712; ATP synthase, H ⁺ transporting, mitochondrial FO complex, subunit g; predicted gene 4045
IPI00136505, IPI00623553, IPI00881799, IPI00230507	predicted gene 10250; hypothetical protein LOC676483; predicted gene 5051; ATP synthase, H ⁺ transporting, mitochondrial F0 complex, subunit d; predicted gene 4953; similar to ATP synthase, H ⁺ transporting, mitochondrial F0 complex, subunit d
IPI00116222	predicted gene 11225; 3-hydroxyisobutyrate dehydrogenase
IPI00110524, IPI00848762, IPI00659536	predicted gene 11945; predicted gene 6238; mitochondrial ribosomal protein L30
IPI00111877, IPI00408243	predicted gene 12115; single-stranded DNA binding protein 1
IPI00308885, IPI00853695, IPI00830313, IPI00845678,	predicted gene 12141; heat shock protein 1 (chaperonin)
IPI00461249	
IPI00118196, IPI00894830	predicted gene 13328; mitochondrial ribosomal protein S5
IPI00666165, IPI00125035, IPI00648501, IPI00649018,	predicted gene 13639; similar to adenylate kinase 4; adenylate kinase 3-like 1
IPI00651878	
IPI00858282, IPI00474281, IPI00115607, IPI00850941	predicted gene 13910; similar to Hydroxyacyl-Coenzyme A dehydrogenase/3-ketoacyl-Coenzyme A thiolase/enoyl-Coenzyme A hydratase (trifunctional protein), beta subunit; hydroxyacyl-Coenzyme A dehydrogenase/3-ketoacyl-Coenzyme A thiolase/enoyl-Coenzyme A hydratase (trifunctional protein), beta subunit; predicted gene 9108
IPI00115040, IPI00850545, IPI00677484	predicted gene 14506; BCL2/adenovirus E1B interacting protein 3; predicted gene 6532; similar to E1B 19K/Bcl-2-binding protein homolog

IPI00110385
IPI00116753
IPI00132347, IPI00555000
IPI00111770, IPI00875971
IPI00453724, IPI00556699, IPI00755768

IPI00133363
IPI00319731, IPI00849049, IPI00659907, IPI00874964, IPI00753028, IPI00849031, IPI00850955, IPI00850432, IPI00850377, IPI00462008, IPI00462075, IPI00850337, IPI00620663, IPI00850243, IPI00848648, IPI00848411, IPI00273646, IPI00848801, IPI00656311, IPI00850028, IPI00849532, IPI00848807, IPI00762304, IPI00850062, IPI00663957, IPI00848511, IPI00849530, IPI00135284, IPI00753102, IPI00849050, IPI00751677, IPI00752685, IPI00756025, IPI00752289, IPI00849792, IPI00849045, IPI00850760, IPI00851086, IPI00271869, IPI00622795, IPI00849046, IPI00850779, IPI00853668, IPI00850434

IPI00118986, IPI00919068
IPI00648759, IPI00115117, IPI00648577, IPI00649391
IPI00132042
IPI00131771, IPI00622837
IPI00114246
IPI00407130, IPI00845840

IPI00856384, IPI00271986
IPI00225318
IPI00128857, IPI00670735, IPI00881197

IPI00876341, IPI00122548
IPI00127417, IPI00379441

IPI00319994, IPI00751369, IPI00754398, IPI00756114
IPI00798555, IPI00113606
IPI00130589, IPI00135857
IPI00378520
IPI00625588, IPI00460132, IPI00274407
IPI00133167

IPI00124015, IPI00856157, IPI00857160
IPI00651865, IPI00880977, IPI00321718
IPI00126917, IPI00133440, IPI00649084
IPI00881014, IPI00322760, IPI00882128, IPI00475037
IPI00719901, IPI00720189, IPI00131548
IPI00918862, IPI00606510, IPI00918838, IPI00468653
IPI00330523, IPI00858257, IPI00858124
IPI00321190
IPI00312174
IPI00226766
IPI00672824, IPI00850694, IPI00857971, IPI00849866
IPI00408727, IPI00881534, IPI00881722, IPI00881568, IPI00127050
IPI00114710
IPI00337893
IPI00118594
IPI00753072, IPI00222767, IPI00752455
IPI00221407, IPI00776169
IPI00122251
IPI00881473, IPI00881737, IPI00459279
IPI00222930, IPI00307931, IPI00515349, IPI00123186
IPI00875863, IPI00474370, IPI00473646
IPI00858050, IPI00320716

predicted gene 15683; RIKEN cDNA 2310005N03 gene; similar to RIKEN cDNA 2310005N03
predicted gene 2893; electron transferring flavoprotein, alpha polypeptide
predicted gene 2962; ubiquinol-cytochrome c reductase binding protein
predicted gene 2972; ATP synthase, H⁺ transporting, mitochondrial F1F0 complex, subunit e
predicted gene 3672; similar to Glycine cleavage system H protein, mitochondrial precursor; glycine cleavage system protein H (aminomethyl carrier)
predicted gene 4237; NADH dehydrogenase (ubiquinone) 1, subcomplex unknown, 1
predicted gene 4691; predicted gene 6944; predicted gene 10290; predicted gene 10566; predicted gene 10291; predicted gene 3200; predicted gene 12070; predicted gene 7286; predicted gene 6946; predicted gene 8825; predicted gene 9081; glyceraldehyde-3-phosphate dehydrogenase; predicted gene 10359; predicted gene 10358; predicted gene 7784; predicted gene 12416; predicted gene 14148; predicted gene 13882; predicted gene 4217; predicted gene 7183; predicted gene 10313; predicted gene 2467; predicted gene 5787; predicted gene 2193; predicted gene 13292; predicted gene 380687; predicted gene 2076; predicted gene 3222; predicted gene 7507; predicted gene 7293; predicted gene 15191; predicted gene 674324; predicted gene 10284; predicted gene 8055; similar to hCG1978856; predicted gene 2574; similar to glyceraldehyde-3-phosphate dehydrogenase; predicted gene 7545; predicted gene 12033; predicted gene 3272; predicted gene 3534; predicted gene 2308; predicted gene 3839; predicted gene 9061; predicted gene 3671; predicted gene 8513; predicted gene 4654; predicted gene 12537; predicted gene 4929; predicted gene 5652; predicted gene 9568; predicted gene 8349; predicted gene 2546; predicted gene 7129; similar to Glyceraldehyde-3-phosphate dehydrogenase (GAPDH); predicted gene 8318; glyceraldehyde-3-phosphate dehydrogenase pseudogene; predicted gene 7611; predicted gene 2445; predicted gene 6283; predicted gene 11557; predicted gene 4335; predicted gene 9127; predicted gene 3695; predicted gene 5507; predicted gene 8174; predicted gene 4609; predicted gene 3809; predicted gene 9034; predicted gene 2606; predicted gene 4575; predicted gene 12286; predicted gene 11658; predicted gene 5732; predicted gene 16374; predicted gene 8100; predicted gene 11953; predicted gene, EG545741; predicted gene 2451; predicted gene 14130

predicted gene 5436; similar to ATP synthase, H⁺ transporting, mitochondrial F1 complex, O subunit; ATP synthase, H⁺ transporting, mitochondrial F1 complex, O subunit
predicted gene 5626; stomatin (Epb7.2)-like 2
predicted gene 6123; pyruvate dehydrogenase (lipoamide) beta
predicted gene 6265; similar to cytochrome c oxidase, subunit VIc; cytochrome c oxidase, subunit VI
predicted gene 6444; NADH dehydrogenase (ubiquinone) 1 beta subcomplex, 11
predicted gene 6560; predicted gene 2124; predicted gene 6992; pyruvate kinase, muscle; similar to M2-type pyruvate kinase
predicted gene 6835; ATP synthase, H⁺ transporting, mitochondrial F0 complex, subunit f, isoform 2; predicted gene 6581

predicted gene 6997; mitochondrial ribosomal protein L22
predicted gene 7049; similar to NADP-dependent malic enzyme (NADP-ME) (Malic enzyme 1); malic enzyme 1, NADP(+)-dependent, cytosolic
predicted gene 7591; voltage-dependent anion channel 3
predicted gene 7730; non-metastatic cells 2, protein (NM23B) expressed in; predicted gene 5566; predicted gene 5425; similar to Nucleoside diphosphate kinase B (NDK B) (NDP kinase B) (P18)
predicted gene 7997; lactate dehydrogenase A; predicted gene 5452
predicted gene 8546; acylglycerol kinase
predicted gene 8566; superoxide dismutase 1, soluble; similar to Superoxide dismutase
predicted gene 9481; mitochondrial ribosomal protein L41
predicted gene 9755; Tu translation elongation factor, mitochondrial
predicted gene 9803; mitochondria-associated protein involved in granulocyte-macrophage colony-stimulating factor signal transduction
presenilin associated, rhomboid-like
prohibitin 2
prohibitin; predicted gene 4773; RIKEN cDNA 1700071K01 gene
proline dehydrogenase
proline synthetase co-transcribed
propionyl Coenzyme A carboxylase, beta polypeptide
propionyl-Coenzyme A carboxylase, alpha polypeptide
prosaposin
prostaglandin E synthase 2
protein phosphatase 1K (PP2C domain containing)
protein phosphatase 2C, magnesium dependent, catalytic subunit
pyrophosphatase (inorganic) 2

pyruvate carboxylase
pyruvate dehydrogenase E1 alpha 1
pyruvate dehydrogenase E1 alpha 2
pyruvate dehydrogenase complex, component X; similar to pyruvate dehydrogenase complex, component X
pyruvate dehydrogenase kinase, isoenzyme 1
pyruvate dehydrogenase kinase, isoenzyme 2
quinoid dihydropteridine reductase; similar to Quinoid dihydropteridine reductase
ras homolog gene family, member T1
ras homolog gene family, member T2
reticulon 4 interacting protein 1

IPI00895515, IPI00229040, IPI00857145	retinol dehydrogenase 13 (all-trans and 9-cis)
IPI00129164	sepiapterin reductase
IPI00318006	serine hydrolase-like
IPI00454008	serine hydroxymethyltransferase 2 (mitochondrial)
IPI00109354	seryl-aminocyl-tRNA synthetase 2
IPI00170357, IPI00850215	similar to AFG3(ATPase family gene 3)-like 2 (yeast); AFG3(ATPase family gene 3)-like 2 (yeast)
IPI00225288, IPI00471416	similar to Coiled-coil domain containing 90B; coiled-coil domain containing 90B
IPI00848492, IPI00121440, IPI00757916	similar to Electron transferring flavoprotein, beta polypeptide; electron transferring flavoprotein, beta polypeptide
IPI00121309, IPI00475158	similar to NADH dehydrogenase (ubiquinone) Fe-S protein 3; NADH dehydrogenase (ubiquinone) Fe-S protein 3; predicted gene 12251
IPI00673568, IPI00886222, IPI00128345, IPI00674389	similar to NADH dehydrogenase (ubiquinone) Fe-S protein 6; NADH dehydrogenase (ubiquinone) Fe-S protein 6; predicted gene 6415
IPI00653905, IPI00757305, IPI00830434, IPI00850402, IPI00153266	similar to Saccharopine dehydrogenase (putative); saccharopine dehydrogenase (putative)
IPI00850430, IPI00124771	similar to Solute carrier family 25 (mitochondrial carrier, phosphate carrier), member 3; solute carrier family 25 (mitochondrial carrier, phosphate carrier), member 3
IPI00553262, IPI00480233	similar to Unknown (protein for IMAGE:4910858); predicted gene 4076
IPI00753287, IPI00122547, IPI00750490	similar to Voltage-dependent anion-selective channel protein 2 (VDAC-2) (mVDAC2) (mVDAC6) (Outer mitochondrial membrane protein porin 2); predicted gene 7832; voltage-dependent anion channel 2
IPI00269076, IPI00648318	similar to adenylate kinase 2; adenylate kinase 2
IPI00848729, IPI00918701, IPI00124292	similar to brain protein 44-like protein; brain protein 44-like; predicted gene 3452; predicted gene 8219
IPI00315794	similar to cytochrome b5 outer mitochondrial membrane precursor; cytochrome b5 type E
IPI00111255, IPI00850715	similar to cytochrome c oxidase, subunit VIIc; predicted gene 3386; cytochrome c oxidase, subunit VIIc
IPI00785410, IPI00116154	similar to cytochrome c oxidase, subunit Vb; predicted gene 11273; cytochrome c oxidase, subunit Vt
IPI00133284	similar to es1 protein; DNA segment, Chr 10, Johns Hopkins University 81 expressed
IPI00109293, IPI00890317	similar to lactamase, beta; lactamase, beta
IPI00469103	similar to lysyl-tRNA synthetase; lysyl-tRNA synthetase
IPI00342938	similar to mitochondrial malonyltransferase isoform b precursor; malonyl CoA:ACP acyltransferase (mitochondrial)
IPI00894705, IPI00857674, IPI00312244, IPI00649816, IPI00415517, IPI00816883, IPI00894813	similar to mitofusin 2; mitofusin 2
IPI00855011, IPI00117657, IPI00918906, IPI00918980, IPI00918878, IPI00403336, IPI00919056	similar to optic atrophy 1 (autosomal dominant); optic atrophy 1 homolog (human)
IPI00321923, IPI00556889	similar to polynucleotide phosphorylase-like protein; polyribonucleotide nucleotidyltransferase
IPI00169883	sirtuin 5 (silent mating type information regulation 2 homolog) 5 (S. cerevisiae)
IPI00137194	solute carrier family 16 (monocarboxylic acid transporters), member 1
IPI00131584	solute carrier family 25 (mitochondrial carnitine/acylcarnitine translocase), member 2
IPI00230754, IPI00648357, IPI00648119	solute carrier family 25 (mitochondrial carrier oxoglutarate carrier), member 11
IPI00308162	solute carrier family 25 (mitochondrial carrier, Aralar), member 12
IPI00135651	solute carrier family 25 (mitochondrial carrier, adenine nucleotide translocator), member 12
IPI00115564	solute carrier family 25 (mitochondrial carrier, adenine nucleotide translocator), member 4
IPI00127841, IPI00658303	solute carrier family 25 (mitochondrial carrier, ADP/ATP translocase 2 (Adenine nucleotide translocator 2) (ANT 2) (ADP,ATP carrier protein 2) (Solute carrier family 25 member 5); predicted gene 5529; predicted gene 8429; predicted gene 5256
IPI00885254, IPI00276926	solute carrier family 25 (mitochondrial carrier, citrate transporter), member 1
IPI00317074	solute carrier family 25 (mitochondrial carrier, dicarboxylate transporter), member 10
IPI00756073, IPI00754831, IPI00109275, IPI00856458, IPI00754329	solute carrier family 25 (mitochondrial carrier, glutamate), member 22
IPI00849619, IPI00226854	solute carrier family 25, member 4C
IPI00320503	solute carrier family 25, member 42
IPI00387392, IPI00652424	solute carrier family 25, member 46
IPI00221608	sorting and assembly machinery component 50 homolog (S. cerevisiae)
IPI00830372, IPI00830269, IPI00170128, IPI00830563	spastic paraplegia 7 homolog (human)
IPI00123129	staphylococcal nuclease and tudor domain containing 1
IPI00648476, IPI00648299, IPI00134131, IPI00648007	sterol carrier protein 2, liver
IPI00230351	succinate dehydrogenase complex, subunit A, flavoprotein (Fp)
IPI00338536	succinate dehydrogenase complex, subunit B, iron sulfur (Ip); similar to succinate dehydrogenase Ip subunit
IPI00831418, IPI00319111	succinate dehydrogenase complex, subunit C, integral membrane protein
IPI00406442	succinate-CoA ligase, GDP-forming, alpha subunit
IPI00261627	succinate-Coenzyme A ligase, ADP-forming, beta subunit
IPI00665996, IPI00459487	succinate-Coenzyme A ligase, GDP-forming, beta subunit
IPI00750524, IPI00313998	sulfide quinone reductase-like (yeast)
IPI00153144	sulfite oxidase
IPI00109109	superoxide dismutase 2, mitochondrial
IPI00314919	suppressor of var1, 3-like 1 (S. cerevisiae)
IPI00119022	tRNA 5-methylaminomethyl-2-thiouridylate methyltransferase
IPI00187402, IPI00651932	tRNA nucleotidyl transferase, CCA-adding, 1
IPI00125652, IPI00828588	thioredoxin 2
IPI00471266, IPI00350590, IPI00124699, IPI00271280	thioredoxin reductase 2; similar to thioredoxin reductase 2
IPI00223216, IPI00762128	thiosulfate sulfurtransferase, mitochondrial
IPI00895166, IPI00112822, IPI00895392, IPI00230512	transcription factor A, mitochondrial

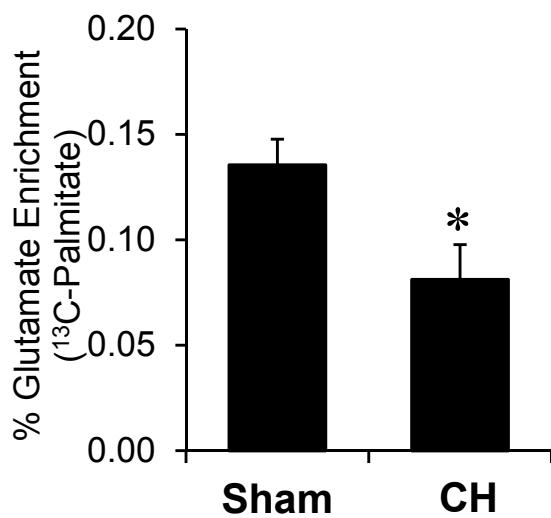
IPI00649594, IPI00848390, IPI00453815, IPI00828226, IPI00881541, IPI00648278	transforming growth factor beta regulated gene 4; similar to Transforming growth factor beta regulated gene 4
IPI00126861	transglutaminase 2, C polypeptide
IPI00165902	translocase of inner mitochondrial membrane 10 homolog (yeast)
IPI00134484	translocase of inner mitochondrial membrane 13 homolog (yeast)
IPI00885780, IPI00315325, IPI00515156	translocase of inner mitochondrial membrane 22 homolog (yeast)
IPI00135068, IPI00889924	translocase of inner mitochondrial membrane 44
IPI00111045	translocase of inner mitochondrial membrane 50 homolog (yeast)
IPI00125513	translocase of inner mitochondrial membrane 9 homolog (yeast)
IPI00474157, IPI00875326	translocase of outer mitochondrial membrane 40 homolog (yeast)
IPI00751137, IPI00377728	translocase of outer mitochondrial membrane 70 homolog A (yeast)
IPI00122499, IPI00885696, IPI00830213	transmembrane protein 143
IPI00130661	tripeptidyl peptidase I
IPI00460008, IPI00111613, IPI00307938	tryptophanyl tRNA synthetase 2 (mitochondrial)
IPI00118384, IPI00828266, IPI00885651	tyrosine 3-monooxygenase/tryptophan 5-monooxygenase activation protein, epsilon polypeptide
IPI00119138	ubiquinol cytochrome c reductase core protein 2
IPI00857580, IPI00109603, IPI00857713, IPI00919103, IPI00858246, IPI00858043, IPI00857602, IPI00230305, IPI00749564	ubiquinol-cytochrome c reductase complex chaperone, CBP3 homolog (yeast)
IPI00653598, IPI00111885	ubiquinol-cytochrome c reductase core protein 1
IPI00133240	ubiquinol-cytochrome c reductase, Rieske iron-sulfur polypeptide 1
IPI00648640, IPI00224210	ubiquinol-cytochrome c reductase, complex III subunit VII
IPI00114153	uncoupling protein 3 (mitochondrial, proton carrier)
IPI00624653	upregulated during skeletal muscle growth 5
IPI00222180	valyl-tRNA synthetase 2, mitochondrial (putative); similar to valyl-tRNA synthetase 2-like
IPI00230540, IPI00122549, IPI00857067	voltage-dependent anion channel 1
IPI00221569	zinc binding alcohol dehydrogenase, domain containing 2

Mitochondrial proteomic profiling in compensated hypertrophy (CH) and heart failure (HF). Quantitative proteomic analysis was done using the Stable Isotope Labeling by Amino ACids (SILAC) method in mitochondria isolated from the ventricles of sham-operated, CH, and HF animals. Mitochondrial proteins identified in all samples are listed. GOCC annotation in DAVID and the Mitocarta were used to define proteins as mitochondrial.

Supplemental Table 2: Proteins regulated in compensated hypertrophy (CH) and heart failure (HF) samples.

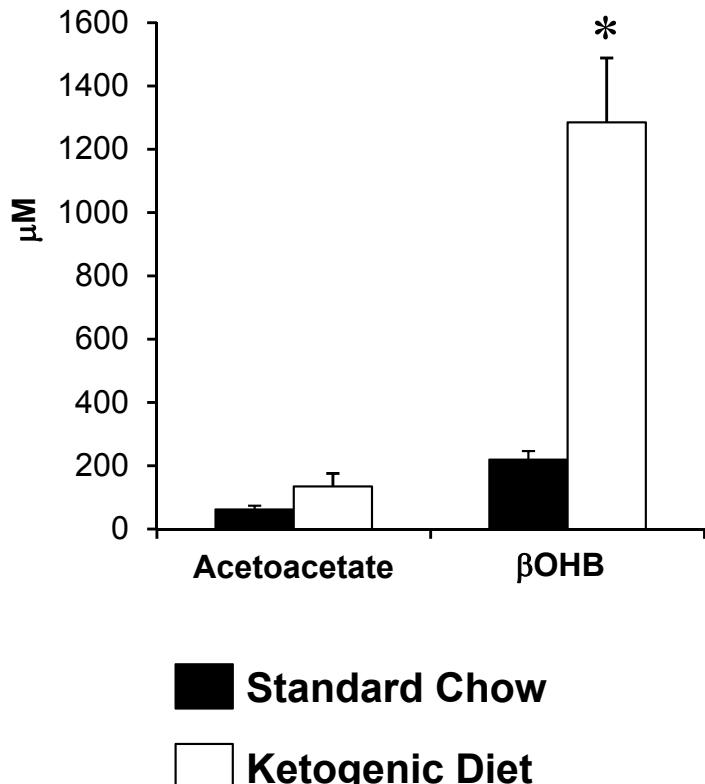
Gene Symbol	Protein Names	CH/CTRL	HF/CTRL
Abhd11	Abhydrolase domain-containing protein 11	1.28	3.45
Nfu1	Putative uncharacterized protein	1.19	2.71
Fahd2a	Fumarylacetoacetate hydrolase domain-containing protein 2A	-1.07	2.02
Abat	4-aminobutyrate aminotransferase	1.84	1.95
Bdh1	3-hydroxybutyrate dehydrogenase	2.74	1.89
Hagh	Hydroxyacylglutathione hydrolase	1.73	1.80
Cltc	Clathrin heavy chain	1.38	1.78
Nln	Neurolysin	1.63	1.65
Slc25a10	Mitochondrial dicarboxylate carrier	1.17	1.55
Rars2	Probable arginyl-tRNA synthetase	1.43	1.52
Cbr2	Carbonyl reductase [NADPH] 2	1.68	1.49
Ccdc58	Coiled-coil domain-containing protein 58	1.51	1.44
Hk1	Hexokinase 1	1.65	1.44
Ctsb	Cathepsin B;Cathepsin B1;Cathepsin B light chain;Cathepsin B heavy chain	1.53	1.43
Slc25a1	Solute carrier family 25	1.75	1.43
Cyb5r3	Cytochrome b5 reductase 3	1.84	1.38
Nudt13	Nucleoside diphosphate-linked moiety X motif 13	1.62	1.37
Acot9	Acyl-coenzyme A thioesterase 9	1.67	1.37
Cyb5r1	NADH-cytochrome b5 reductase 1	1.76	1.37
Cpox	Coproporphyrinogen-III oxidase	1.68	1.33
Hk2	Hexokinase-2	1.69	1.32
Ghitm	Growth hormone-inducible transmembrane protein	1.53	1.31
Gyk	Glycerol kinase	1.54	1.31
Tomm70a	Translocase of outer membrane 70 kDa subunit	1.65	1.28
Gfer	FAD-linked sulfhydryl oxidase ALR	1.69	1.20
Acsf2	Acyl-CoA synthetase family member 2	1.59	1.14
Tmem126a	Transmembrane protein 126A	3.00	1.13
Mrps30	28S ribosomal protein S30	1.54	1.10
Timm22	Mitochondrial import inner membrane translocase subunit	-1.27	1.02
Slc25a42	Solute carrier family 25 member 42	-1.44	-1.18
Acss1	Acetyl-coenzyme A synthetase 2-like	-1.27	-1.19
Agps	Alkylglycerone phosphate synthase	-1.73	-1.19
Cat	Catalase	-1.37	-1.21
Gstk1	Glutathione S-transferase kappa 1	-1.37	-1.25
Opa3	Optic atrophy 3 protein homolog	1.13	-1.26
Tmem143	Transmembrane protein 143	1.06	-1.26
Cisd1	CDGSH iron sulfur domain-containing protein 1	1.27	-1.26
Acaa2	Acetyl-Coenzyme A acyltransferase 2	-1.32	-1.27
9030617O03Rik	Probable 10-formyltetrahydrofolate dehydrogenase	-1.55	-1.29
Slc25a22	Mitochondrial glutamate carrier 1	-1.39	-1.32
Cox6a2	Cytochrome c oxidase subunit 6A2	-1.38	-1.35
Sep2	Non-specific lipid-transfer protein	-1.33	-1.37
Ak1	Adenylate kinase isoenzyme 1	-1.29	-1.38
Hsd17b4	17-beta-hydroxysteroid dehydrogenase 4	-1.46	-1.38
Ech1	Delta(3,5)-Delta(2,4)-dienoyl-CoA isomerase	-1.47	-1.39
Gpd1	Glycerol-3-phosphate dehydrogenase [NAD ⁺]	-1.35	-1.39
Abcd3	ATP-binding cassette, sub-family D (ALD), member 3	-1.64	-1.41
Acad11	Acyl-CoA dehydrogenase family member 11	-1.66	-1.42
Ppm1k	Protein phosphatase 1K	-1.61	-1.42
Acox1	Acyl-Coenzyme A oxidase 1	-1.71	-1.54
Mdh1	Malate dehydrogenase	-1.51	-1.54
Ehhadh	Enoyl-CoA hydratase/3,2-trans-enoyl-CoA isomerase	-1.36	-1.60
Ldhb	L-lactate dehydrogenase B chain	-1.75	-1.83
Ucp3	Mitochondrial uncoupling protein 3	-1.56	-1.84
Pkm2	Pyruvate kinase isozymes M1/M2;Pyruvate kinase muscle isozyme	-1.10	-1.91

Quantitative proteomic analysis was done using the Stable Isotope Labeling by Amino ACids (SILAC) method in mitochondria isolated from the ventricles of sham-operated, CH, and HF animals. Proteins that exhibited a significant difference in the CH and/or HF models vs. corresponding sham-operated controls as determined using mass spectrometry-based quantitative proteomic analysis (MS/MS processing; Thermo Scientific LTQ Orbitrap Velos) are listed. A cut-off of < -1.25 or > 1.5 FC was used to identify regulated proteins.

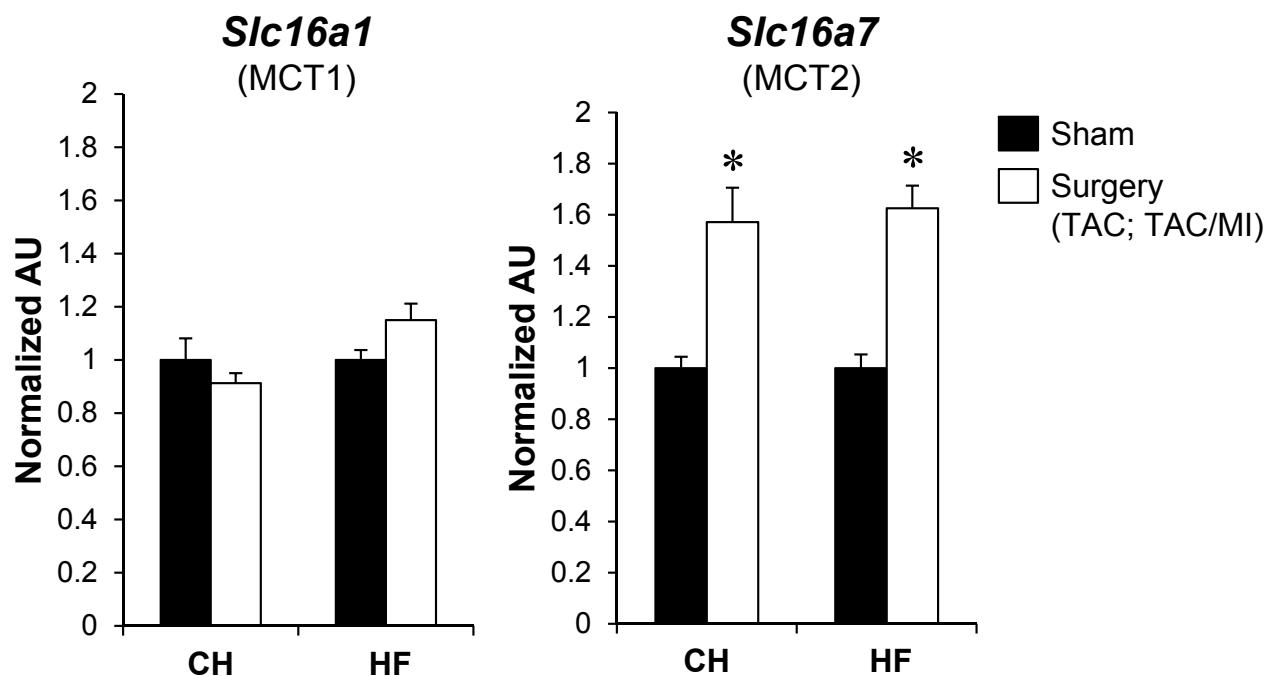


Supplemental Figure 1. Reduced palmitate oxidation in the hypertrophied heart. The relative contribution of palmitate to TCA carbon cycle flux is shown for CH and sham-operated controls. Data are shown as mean \pm SEM (n=6, Sham and 7, CH).

Plasma Ketones



Supplemental Figure 2. Ketogenic diet results in an increase in circulating ketone bodies. Plasma ketone levels measured in wild-type C57BL/6J mice fed a ketogenic diet for 4 weeks compared to control diet fed mice. Bars represent mean \pm SEM values (n=6-7 per group) * $p < 0.05$.



Supplemental Figure 3. *Slc16a1* expression is induced in the hypertrophied and failing mouse heart. *Slc16a1* (MCT1) and *Slc16a7* (MCT2) mRNA levels in cardiac ventricular tissue from mice 4 weeks after sham, TAC (CH), or TAC/MI (HF) surgeries. Samples were taken 24 h (Fasted) after feeding. Expression is normalized to *Rplp0* (36B4). Bars represent mean \pm SEM values (n=6-21 per group) * $p < 0.05$.

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