

Table S1. Summary information about genes analyzed in the study. We have selected twenty mitochondria- and ATPase-related genes from GeneChip Rat Gene 1.0 ST Arrays (Affymetrix, California, USA). The selection included genes encoded by both mtDNA and nuclear DNA; ATPase and OXPHOS subunits, ATPase assembly factors and other transcription factors involved in mitochondrial metabolism regulation. Marked genes (†) were selected for analysis by qPCR to validate more precisely expression profile revealed by microarray analysis.

Encoded in	Gene	Protein	Cell localization	Function	Notes
<i>mtDNA</i>	<i>mt-Nd2</i>	NADH dehydrogenase subunit 2	mitochondria, complex I	core subunit	
	<i>mt-Nd5</i>	NADH dehydrogenase subunit 5	mitochondria, complex I	core subunit	
	<i>mt-Co1</i>	Cytochrome c oxidase subunit I	mitochondria, complex IV	core subunit	
	<i>mt-Co2</i>	Cytochrome c oxidase subunit II	mitochondria, complex IV	core subunit	
	<i>mt-Atp6</i>	ATP synthase subunit a	mitochondria, F ₀ -ATP synthase	proton translocation	†
	<i>mt-Atp8</i>	ATP synthase subunit A6L	mitochondria, F ₀ -ATP synthase	unknown	
<i>nuclear DNA</i>	<i>Atp5a1</i>	ATP synthase subunit α	mitochondria, F ₁ -ATP synthase	ATP synthesis, binding IF1	†
	<i>Atp5g2</i>	ATP synthase subunit c	mitochondria, F ₀ -ATP synthase	rotor, proton translocation, “rate-limiting” subunit	†
	<i>Atp5d</i>	ATP synthase subunit δ	mitochondria, F ₁ -ATP synthase	rotor, connects F ₁ with F ₀	
	<i>Atp5o</i>	ATP synthase subunit OSCP	mitochondria, F ₀ -ATP synthase	connects stator with F ₁ , estrogen binding	†
	<i>Atpaf1</i>	ATP synthase mitochondrial F ₁ complex assembly factor 1	mitochondria	ATP synthase assembly, incorporation of subunit β (Wang <i>et al.</i> 2001)	
	<i>Atpaf2</i>	ATP synthase mitochondrial F ₁ complex assembly factor 2	mitochondria	ATP synthase assembly, incorporation of subunit α (De Meirleir <i>et al.</i> 2004)	
	<i>Tmem70</i>	Transmembrane protein 70	mitochondria, inner membrane	ATP synthase assembly, incorporation of subunit a and A6L (Torraco <i>et al.</i> 2012)	†
	<i>Ppargc1a</i>	Peroxisome proliferator-activated receptor gamma, coactivator 1 α	cytosol, nucleus	chromatin binding, coactivator, hypoxia and fatty acid response	
	<i>Ppargc1b</i>	Peroxisome proliferator-activated receptor γ , coactivator 1 β	nucleus, mitochondria	estrogen receptor and AF-2 domain binding, cAMP and glucocorticoid response	
	<i>Pprc1</i>	Peroxisome proliferator-activated receptor γ , coactivator-related 1	nucleus	poly(A) RNA binding	
	<i>Esrra</i>	Estrogen related receptor α	nucleus	DNA binding, positive transcription regulation	
	<i>Hnf4a</i>	Hepatocyte nuclear factor 4 α	cytoplasm, nucleus	DNA and fatty acid binding, cell differentiation	
	<i>Nrf1</i>	Nuclear respiratory factor 1	cytoplasm, nucleus, exosomes	DNA binding, interaction with estradiol, noradrenalin and others	
	<i>Gabpa</i>	Nuclear respiratory factor 2 α	nucleus	chromatin binding (RNAPol II promoter), interaction with noradrenaline, carbon monoxide, copper	
	<i>Tfam</i>	Transcription factor A, mitochondrial	mitochondria, nucleus	DNA binding (mitochondrial promoters)	

† – analyzed by qPCR to validate microarray profile; F₁ – peripheral domain of F₀F₁-ATP synthase; F₀ – domain of F₀F₁-ATP synthase, which is integrated in the inner mitochondrial membrane; IF1 – ATPase inhibitory factor 1 (Atpif1); AF-2 – activation function 2

Supplement references and other sources:

<http://www.ensembl.org> (Flicek *et al.* 2014)

<http://rgd.mcw.edu/> (Shimoyama *et al.* 2015)

- DE MEIRLEIR L, SENECA S, LISSENS W, DE CLERCQ I, EYSKENS F, GERLO E, SMET J, VAN COSTER R: Respiratory chain complex V deficiency due to a mutation in the assembly gene ATP12. *J Med Genet* **41**: 120-124, 2004.
- FLICEK P, AMODE MR, BARRELL D, BEAL K, BILLIS K, BRENT S, CARVALHO-SILVA D, CLAPHAM P, COATES G, FITZGERALD S, GIL L, GIRÓN CG, GORDON L, HOURLIER T, HUNT S, JOHNSON N, JUETTEMANN T, KÄHÄRI AK, KEENAN S, KULESHA E, MARTIN FJ, MAUREL T, McLAREN WM, MURPHY DN, NAG R, OVERDUIN B, PIGNATELLI M, PRITCHARD B, PRITCHARD E, RIAT HS, RUFFIER M, SHEPPARD D, TAYLOR K, THORMANN A, TREVANION SJ, VULLO A, WILDER SP, WILSON M, ZADISSA A, AKEN BL, BIRNEY E, CUNNINGHAM F, HARROW J, HERRERO J, HUBBARD TJ, KINSELLA R, MUFFATO M, PARKER A, SPUDICH G, YATES A, ZERBINO DR, SEARLE SM: Ensembl 2014. *Nucleic Acids Res* **42**: D749-D755, 2014.
- SHIMOYAMA M, DE PONS J, HAYMAN GT, LAULEDERKIND SJ, LIU W, NIGAM R, PETRI V, SMITH JR, TUTAJ M, WANG SJ, WORTHEY E, DWINELL M, JACOB H: The Rat Genome Database 2015: genomic, phenotypic and environmental variations and disease. *Nucleic Acids Res* **43**: D743-D750, 2015.
- TORRACO A, VERRIGNI D, RIZZA T, MESCHINI MC, VAZQUEZ-MEMIJE ME, MARTINELLI D, BIANCHI M, PIEMONTE F, DIONISI-VICI C, SANTORELLI FM, BERTINI E, CARROZZO R: TMEM70: a mutational hot spot in nuclear ATP synthase deficiency with a pivotal role in complex V biogenesis. *Neurogenetics* **13**: 375-386, 2012.
- WANG ZG, WHITE PS, ACKERMAN SH: Atp11p and Atp12p are assembly factors for the F(1)-ATPase in human mitochondria. *J Biol Chem* **276**: 30773-30778, 2001.