

<b>HGNC gene code</b>	<b>Pathway</b>	<b>Protein</b>	<b>Disease</b>	<b>OMIM (gene)</b>	<b>OMIM (disease)</b>
<i>ACADM</i>	FAO	Medium-chain Acyl-CoA dehydrogenase (MCAD)	MCAD deficiency	607008	201450
<i>ACADVL</i>	FAO	Very long-chain Acyl-CoA dehydrogenase (VLCAD)	VLCAD deficiency	609575	201475
<i>ACAT1</i>	AA	Acetoacetyl-CoA thiolase	$\beta$ -ketothiolase deficiency	607809	203750
<i>ALDH2</i>	MISC	Aldehyde dehydrogenase 2	Alcohol sensitivity, handover	100650	610251
<i>ALDH6A1</i>	AA	Methylmalonate semialdehyde dehydrogenase (MMSD)	MMSD deficiency	603178	603178
<i>BAX</i>	APOP	Bcl2 associated protein x	Cancer susceptibility	600040	600040
<i>CPT1A</i>	FAO	Hepatic CPT1	Hepatic CPT1 deficiency	600528	255120
<i>DARS2</i>	TRANS	Aspartyl t-RNA synthetase	Leukoencephalopathy	610956	611105
<i>DECR1</i>	MISC	2,4-dienoyl-CoA reductase (DECR) Dihydrolipoamid dehydrogenase E3 subunit	DECR deficiency	222745	222745
<i>DLD</i>	TCA		$\alpha$ -keto glutarate dehydrogenase deficiency	238331	238331
<i>ETFA</i>	FAO	Electron transfer flavoprotein (ETF), $\alpha$ subunit	Multiple acyl-CoA dehydrogenase deficiency (MADD), Glutaric aciduria, type II (GAII)	608053	231680
<i>ETFB</i>	FAO	Electron transfer flavoprotein (ETF), $\beta$ subunit	Multiple acyl-CoA dehydrogenase deficiency (MADD), Glutaric aciduria, type II (GAII)	130410	231680
<i>ETFDH</i>	FAO	Electron transfer flavoprotein ubiquinone oxidoreductase, ETFQO	Multiple acyl-CoA dehydrogenase deficiency (MADD), Glutaric aciduria, type II (GAII)	231675	231680
<i>ETHE1</i>	MISC	ETHE1 protein	Ethylmalonic encephalopathy	608451	602473
<i>FH</i>	TCA	Fumarase	Fumarase deficiency	136850	606812
<i>GLUD1</i>	AA	Glutamate dehydrogenase 1	Hyperinsulinism-hyperammonemia syndrome	138130	606762
<i>GPD2</i>	MISC	Glycerol 3-phosphate dehydrogenase 2	Noninsulin dependent diabetes mellitus	138430	125853
<i>HADH</i>	FAO	3-hydroxyacyl-CoA dehydrogenase, HADH (or M/SCHAD)	HADH (or M/SCHAD) deficiency	601609	231530
<i>HADHA</i>	FAO	Mitochondrial trifunctional protein (MTP), $\alpha$ subunit	LCHAD deficiency; MTP deficiency	600890	609016
<i>HADHB</i>	FAO	Mitochondrial trifunctional protein (MTP), $\beta$ subunit	MTP deficiency	143450	609015
<i>HSD17B4</i>	MISC	17- $\beta$ hydroxy steroid dehydrogenase IV, D-bifunctional protein	D-bifunctional protein deficiency	601860	261515
<i>HSPD1</i>	PQC	Heat shock 60-kd protein (HSP60)	Spastic paraplegia 13	118190	605280
<i>ME2</i>	MISC	Malic enzyme 2	Susceptibility to idiopathic generalized epilepsy	154270	600669
<i>NDUFA13</i>	RESP	NADH-ubiquinone oxidoreductase 1 $\alpha$	Papillary thyroid carcinoma	609435	607464
<sup>o</sup> <i>NDUFS1</i>	RESP	NADH-ubiquinone oxidoreductase Fe-S 1	Mitochondrial complex I deficiency	157655	252010
<i>NDUFS2</i>	RESP	NADH-ubiquinone oxidoreductase Fe-S 2	Mitochondrial complex I deficiency	602985	252010
<sup>o</sup> <i>NDUFS3</i>	RESP	NADH-ubiquinone oxidoreductase Fe-S 3	Mitochondrial complex I deficiency	603846	256000
<i>NDUFV2</i>	RESP	NADH-ubiquinone oxidoreductase flavoprotein 2	Mitochondrial complex I deficiency	600532	168600
<i>OGDH</i>	TCA	$\alpha$ -keto glutarate dehydrogenase (E1k)	$\alpha$ -keto glutarate dehydrogenase deficiency	203740	-
<i>OPA1</i>	APOP	Optic atrophy 1 protein	Mitochondrial dysfunction	605290	165500
<i>PCK2</i>	MISC	Phosphoenolpyruvate carboxykinase	PEPCK deficiency, mitochondrial	261650	-
<i>PDHA1</i>	TCA	Pyruvate dehydrogenase complex E1 $\alpha$	Pyruvate dehydrogenase deficiency	312170	308930
<i>PDHB</i>	TCA	Pyruvate dehydrogenase complex E1 $\beta$	Pyruvate dehydrogenase deficiency	312170	179060
<i>PHB</i>	PQC	Prohibitin	Breast cancer, sporadic	176705	-
<i>SDHB</i>	TCA / RESP	Succinate dehydrogenase subunit B	Paraganglioma and gastric stromal sarcoma	185470	606864
<i>SUCLA2</i>	TCA	Succinate-CoA ligase subunit $\beta$	Mitochondrial DNA depletion syndrome,	603921	612073

<sup>a</sup> Gene products with statistically significant altered level in the galactose cultivations.