

## SLC25 – Mitochondrial carrier family

Human gene name	Protein name	Aliases	Predominant substrates	Transport type / coupling ions <sup>*)</sup>	Tissue distribution and cellular / subcellular expression	Link to disease	Human gene locus	Sequence accession ID	Splice variants and their features
<a href="#">SLC25A1</a>	CIC (citrate-isocitrate carrier)	CTP, tricarboxylate carrier, SLC20A3	citrate, malate, PEP	E / citrate-H <sup>+</sup> / malate	liver, kidney, pancreas (also in brain, lung, heart) / inner mitochondrial membrane		22q11.21	<a href="#">NM_005984</a>	
<a href="#">SLC25A2</a>	ORC2 (ornithine carrier 2)	ORNT2	ornithine, citrulline, lysine, arginine, histidine	E / ornithine / citrulline + H <sup>+</sup> ; ornithine / H <sup>+</sup>	liver, testis, spleen, lung, pancreas, small intestine, brain, kidney / inner mitochondrial membrane		5q31	<a href="#">NM_031947</a>	
<a href="#">SLC25A3</a>	PHC (phosphate carrier)	PTP	phosphate	C / phosphate + H <sup>+</sup> ; E / phosphate / OH <sup>-</sup>	isoform A: heart, skeletal muscle and diaphragm; isoform B: liver, kidney, brain, thymus, lung, heart, skeletal muscle, diaphragm / inner mitochondrial membrane	mitochondrial phosphate carrier deficiency	12q23	<a href="#">NM_005888</a> isoform a <a href="#">NM_002635</a> isoform b <a href="#">NM_213611</a>	3 splice variants
<a href="#">SLC25A4</a>	ANT1 (adenine nucleotide translocase-1)	AAC1, T1, PEO2, PEO3	ADP, ATP	E / ADP / ATP	heart, skeletal muscle, much less in brain, pancreas, prostate, kidney, lung, thymus / inner mitochondrial membrane	autosomal dominant progressive external ophthalmoplegia with mitochondrial DNA deletions-2, AAC1 deficiency	4q35	<a href="#">NM_001151</a>	
<a href="#">SLC25A5</a>	ANT2 (adenine nucleotide translocase-2)	AAC2, T2	ADP, ATP	E / ADP / ATP	brain, lung, kidney, pancreas, heart, skeletal muscle, spleen / inner mitochondrial membrane		Xq24-Xq26	<a href="#">NM_001152</a>	
<a href="#">SLC25A5P1</a>	pseudogene	bK250D10, ANTP3					22q13.2	<a href="#">NG_009276</a>	
<a href="#">SLC25A6</a>	ANT3 (adenine nucleotide translocase-3)	AAC3, T3	ADP, ATP	E / ADP / ATP	brain, lung, kidney, liver, pancreas, heart, skeletal muscle, spleen, thymus / inner mitochondrial membrane		Xp22.32, Yp11.3	<a href="#">NM_001636</a>	
<a href="#">SLC25A6P1</a>	pseudogene						20p11.22	<a href="#">NG_003107</a>	
<a href="#">SLC25A7</a>	UCP1 (uncoupling protein 1)	thermogenin, proton carrier	H <sup>+</sup>	F	brown adipose tissue / inner mitochondrial membrane	(obesity)	4q28-q31	<a href="#">NM_021833</a>	
<a href="#">SLC25A8</a>	UCP2 (uncoupling protein 2)	proton carrier, UCPH	H <sup>+</sup>	F	brain, lung, kidney, spleen, heart / inner mitochondrial membrane	obesity, type 2 diabetes, congenital hyperinsulinism	11q13	<a href="#">NM_003355</a>	

Human gene name	Protein name	Aliases	Predominant substrates	Transport type / coupling ions <sup>3)</sup>	Tissue distribution and cellular / subcellular expression	Link to disease	Human gene locus	Sequence accession ID	Splice variants and their features
<a href="#">SLC25A9</a>	UCP3 (uncoupling protein 3)	proton carrier	H <sup>+</sup>	F	skeletal muscle, lung / inner mitochondrial membrane	(obesity, type II diabetes)	11q13	<a href="#">NM_003356</a> <a href="#">NM_022803</a>	2 splice variants
<a href="#">SLC25A10</a>	DIC (dicarboxylate carrier)		malate, phosphate, succinate, sulphate, thiosulphate	E / malate / phosphate	liver, kidney, heart, brain, lung, pancreas / inner mitochondrial membrane		17q25.3	<a href="#">NM_012140</a>	
<a href="#">SLC25A11</a>	OGC (oxoglutarate carrier)	oxoglutarate / malate carrier, SLC20A4	2-oxoglutarate, malate	E / oxoglutarate / malate	heart, skeletal muscle, liver, kidney, brain, pancreas / inner mitochondrial membrane		17p13.3	<a href="#">NM_003562</a>	
<a href="#">SLC25A12</a>	AGC1 (aspartate / glutamate carrier 1)	aralar1	aspartate, glutamate	E / aspartate / glutamate + H <sup>+</sup>	brain, heart, skeletal muscle, lung, pancreas, kidney, but not in liver / inner mitochondrial membrane	AGC1 deficiency, (autism)	2q24	<a href="#">NM_003705</a>	
<a href="#">SLC25A13</a>	AGC2 (aspartate / glutamate carrier 2)	citrin, CTLN2, aralar2	aspartate, glutamate	E / aspartate / glutamate + H <sup>+</sup>	liver, kidney, pancreas, heart, skeletal muscle, brain / inner mitochondrial membrane	citrullinemia type II (CTLN2), neonatal intrahepatic cholestasis (NICCD)	7q21.3	<a href="#">NM_014251</a>	
<a href="#">SLC25A14</a>	UCP5 (uncoupling protein 5)	BMCP1, brain mitochondrial carrier protein	O		widely expressed, with highest levels in brain and testis		Xq24	<a href="#">NM_003951</a> <a href="#">NM_022810</a>	2 splice variants
<a href="#">SLC25A15</a>	ORC1 (ornithine carrier 1)	ORNT1, HHH	ornithine, citrulline, lysine, arginine	E / ornithine / citrulline + H <sup>+</sup> ; ornithine / H <sup>+</sup>	liver, pancreas, lung, testis, small intestine, spleen, kidney, brain, heart / inner mitochondrial membrane	hyperornithinemia-hyperammonemia-homocitrullinuria (HHH) syndrome	13q14.11	<a href="#">NM_014252</a>	
<a href="#">SLC25A15P</a>	pseudogene						Yq11.223	<a href="#">NG_002817</a>	
<a href="#">SLC25A16</a>	GDC (Graves' disease carrier)	GDA, ML7, HGT.1	O		liver, kidney, thyroid, lung, heart, skeletal muscle, brain / inner mitochondrial membrane		10q21.3	<a href="#">NM_152707</a>	
<a href="#">SLC25A17</a>	PMP34	ANC1, ANT1	ATP	E / ATP / AMP	brain, kidney, lung, heart, liver, pancreas / peroxisomal membrane		22q13.2	<a href="#">NM_006358</a>	
<a href="#">SLC25A18</a>	GC2 (glutamate carrier 2)		glutamate	C / glutamate + H <sup>+</sup> ; E / glutamate / OH <sup>-</sup>	brain, testis, heart, pancreas, kidney, lung / inner mitochondrial membrane		22q11.2	<a href="#">NM_031481</a>	

Human gene name	Protein name	Aliases	Predominant substrates	Transport type / coupling ions <sup>1)</sup>	Tissue distribution and cellular / subcellular expression	Link to disease	Human gene locus	Sequence accession ID	Splice variants and their features
<a href="#">SLC25A19</a>	DNC (deoxynucleotide carrier)	MUP1, TPC, MCPHA	thiamine pyrophosphate, thiamine monophosphate, (deoxy)nucleotides	E / thiamine pyrophosphate / thiamine monophosphate, thiamine pyrophosphate / (deoxy)nucleotide	brain, testis, lung, kidney, liver, spleen, skeletal muscle, heart / inner mitochondrial membrane	Amish microcephaly (MCPHA), neuropathy and bilateral striatal necrosis	17q25.3	<a href="#">NM_021734</a> <a href="#">NM_001126122</a> <a href="#">NM_001126121</a>	3 splice variants
<a href="#">SLC25A20</a>	CAC (carnitine / acylcarnitine carrier)	CACT, carnitine carrier	carnitine, acylcarnitine	E / carnitine / acylcarnitines; F (at slow rate)	heart, skeletal muscle, liver (also in lung, kidney, brain, pancreas, lung, placenta) / inner mitochondrial membrane	carnitine-acylcarnitine translocase (CACT) deficiency	3p21.31	<a href="#">NM_000387</a>	
<a href="#">SLC25A20P</a>	pseudogene						6p12	<a href="#">NG_001087</a>	
<a href="#">SLC25A21</a>	ODC (oxoadipate carrier)		oxoadipate, oxoglutarate	E / oxoadipate / oxoglutarate	kidney, gall bladder, colon, liver, placenta, testis, lung, spleen, skeletal muscle, brain, heart / inner mitochondrial membrane		14q11.2	<a href="#">NM_030631</a>	
<a href="#">SLC25A22</a>	GC1 (glutamate carrier 1)		glutamate	C / glutamate + H <sup>+</sup> ; E / glutamate / OH <sup>-</sup>	pancreas, brain, liver, testis, spleen, kidney, heart, lung, small intestine, pancreatic $\beta$ cells / inner mitochondrial membrane	early infantile epileptic encephalopathy-3 (EIEE3)	11p15.5	<a href="#">NM_024698</a>	
<a href="#">SLC25A23</a>	APC2	ScaMC-3, MCSC2	ATP-Mg <sup>2+</sup> , ATP, ADP, AMP and Pi	E / ATP-Mg / Pi	kidney, lung, small intestine, pancreas, brain, liver, skeletal muscle, heart, inner mitochondrial membrane		19p13.3	<a href="#">NM_024103</a>	
<a href="#">SLC25A24</a>	APC1	ScaMC-1	ATP-Mg <sup>2+</sup> , ATP, ADP, AMP and Pi	E / ATP-Mg / Pi	testis, inner mitochondrial membrane		1p13.3	<a href="#">NM_013386</a> <a href="#">NM_213651</a>	2 splice variants
<a href="#">SLC25A25</a>	APC3	ScaMC-2, MCSC, PCSCL	O		brain, heart, skeletal muscle, liver, small intestine, lung, pancreas, testis, inner mitochondrial membrane		9q34.11	<a href="#">NM_052901</a> <a href="#">NM_001006641</a> <a href="#">NM_001006642</a> <a href="#">NM_001006643</a>	4 splice variants
<a href="#">SLC25A26</a>	SAMC		S-adenosyl-methionine, S-adenosyl-homocysteine	E / S-adenosyl-methionine / S-adenosyl-homocysteine	ubiquitous (testis), inner mitochondrial membrane		3p14.1	<a href="#">NM_173471</a>	

Human gene name	Protein name	Aliases	Predominant substrates	Transport type / coupling ions <sup>*)</sup>	Tissue distribution and cellular / subcellular expression	Link to disease	Human gene locus	Sequence accession ID	Splice variants and their features
<a href="#">SLC25A27</a>	UCP4 (uncoupling protein 4)		O		brain / inner mitochondrial membrane		6p11.2-q12	<a href="#">NM_004277</a>	
<a href="#">SLC25A28</a>	Mitoferrin 2 (Mfrn2)	MRS3/4, MRS4L	Fe <sup>2+</sup>		ubiquitous (heart, liver, kidney)		10q23-q24	<a href="#">NM_031212</a>	
<a href="#">SLC25A29</a>	ORNT3	CACL	ornithine, acylcarnitine		heart, skeletal muscle, liver, brain (inner mitochondrial membrane)		14q32.2	<a href="#">NM_001039355</a>	
<a href="#">SLC25A30</a>		KMCP1, UCP6	O		kidney		13q14.12	<a href="#">NM_001010875</a>	
<a href="#">SLC25A31</a>	AAC4, ANT4 (adenine nucleotide carrier-4)	SFEC	ADP, ATP	E / ADP / ATP	testis	(spermatogenesis)	4q28.1	<a href="#">NM_031291</a>	
<a href="#">SLC25A32</a>	MFT	MFTC	folate				8q22.3	<a href="#">NM_030780</a>	
<a href="#">SLC25A33</a>	PNC1 (pyrimidine nucleotide carrier 1)	BMSC-MCP	UTP		ubiquitous		1p36.22	<a href="#">NM_032315</a>	
<a href="#">SLC25A34</a>			O				1p36.21	<a href="#">NM_207348</a>	
<a href="#">SLC25A35</a>			O				17p13.1	<a href="#">NM_201520</a>	
<a href="#">SLC25A36</a>			O				3q23	<a href="#">NM_018155</a> <a href="#">NM_001104647</a>	2 splice variants
<a href="#">SLC25A37</a>	Mitoferrin 1 (Mfrn1)	HT015, MSC, MSCP	Fe <sup>2+</sup>		spleen, placenta		8p21.2	<a href="#">NM_016612</a>	
<a href="#">SLC25A38</a>			glycine?		erythroid?	sideroblastic anemia	3p22.1	<a href="#">NM_017875</a>	
<a href="#">SLC25A39</a>		CGI-69	O		ubiquitous		17q12	<a href="#">NM_016016</a> <a href="#">NM_001143780</a>	2 splice variants
<a href="#">SLC25A40</a>		MCFP	O				7q21.12	<a href="#">NM_018843</a>	
<a href="#">SLC25A41</a>	SCaMC-3L	APC4	ATP-Mg / Pi	E	liver, testis, brain		19p13.3	<a href="#">NM_173637</a>	
<a href="#">SLC25A42</a>			CoA, ADP, ATP, adenosine 3',5'-diphosphate	E	ubiquitous (adipose tissue), inner mitochondrial membrane		19p13.11	<a href="#">NM_178526</a>	
<a href="#">SLC25A43</a>			O		adrenal gland, skeletal muscle		Xq24	<a href="#">NM_145305</a>	
<a href="#">SLC25A44</a>			O				1q22	<a href="#">NM_014655</a> <a href="#">NM_001135672</a>	2 splice variants
<a href="#">SLC25A45</a>			O		skeletal muscle, intestine		11q13.1	<a href="#">NM_182556</a> <a href="#">NM_001077241</a>	2 splice variants
<a href="#">SLC25A46</a>			O				5q22.1	<a href="#">NM_138773</a>	
<a href="#">SLC25A47</a>		C14orf68, HDMCP					14q32.2	<a href="#">NM_207117.2</a>	
<a href="#">SLC25A48</a>		FLJ44862					5q31.1	<a href="#">NM_145282.2</a>	

\*) C: Cotransporter; E: Exchanger; F: Facilitated transporter; O: Orphan transporter

**References:**

*Original version of the SLC table:*

[Palmieri F.](#) The mitochondrial transporter family (SLC25): physiological and pathological implications. *Pflugers Arch.* 2004 Feb;447(5):689-709

*Recent reviews:*

[Palmieri F, Pierri CL.](#) Mitochondrial metabolite transport. *Essays Biochem.* 2010;47:37-52.

[Palmieri F, Pierri CL.](#) Structure and function of mitochondrial carriers - role of the transmembrane helix P and G residues in the gating and transport mechanism. *FEBS Lett.* 2010 May 3;584(9):1931-9

[Palmieri F.](#) Diseases caused by defects of mitochondrial carriers: a review. *Biochim Biophys Acta.* 2008 Jul-Aug;1777(7-8):564-78

Questions & Comments