

ATP-binding cassette family

Overview: ATP-binding cassette transporters are ubiquitous membrane proteins characterized by facilitated movement of a range of substrates, including ions, lipids, peptides, steroids. The functional transporter is probably dimeric, with individual subunits typically made up of two groups of 6TM-spanning domains, with two nucleotide-binding domains (NBD). The majority of eukaryotic ATP-binding cassette (ABC) transporters are 'full' transporters incorporating both TM and NBD entities. Some ABCs, notably the ABCD family, appear relatively truncated and are only functional as homo- or heterodimers. Eukaryotic ABC transporters convey substrates from the cytoplasm, either out of the cell or into intracellular organelles. Their role in the efflux of exogenous compounds, notably chemotherapeutic agents, has led to considerable interest.

Systematic name	Common abbreviation	Other names	Ensembl ID	Comments
ABCA1	ABC1, CERP	Cholesterol efflux regulatory protein	ENSG00000165029	Loss-of-function mutations are associated with Tangier disease, in which plasma HDL cholesterol levels are greatly reduced
ABCA2	ABC2	–	ENSG00000107331	–
ABCA3	ABC3, ABCC	–	ENSG00000167972	Loss-of-function mutations are associated with pulmonary surfactant deficiency
ABCA4	ABCR	Retinal-specific ATP-binding cassette transporter, RIM ABC transporter, RmP, Stargardt disease protein	ENSG00000198691	Retinal-specific transporter of <i>N</i> -retinylPE; loss-of-function mutations are associated with Stargardt disease, a juvenile onset macular degenerative disease
ABCA5	–	–	ENSG00000154265	–
ABCA6	–	–	ENSG00000154262	–
ABCA7	–	–	ENSG00000064687	–
ABCA8	–	KIAA0822	ENSG00000141338	–
ABCA9	–	–	ENSG00000154258	–
ABCA10	–	–	ENSG00000154263	–
ABCA12	–	–	ENSG00000144452	Reported to play a role in skin ceramide formation (Zuo <i>et al.</i> , 2008)
ABCA13	–	–	ENSG00000179869	–
ABCA14	–	–	ENSMUSG00000062017	No orthologue in man
ABCA15	–	–	ENSMUSG00000054746	No orthologue in man
ABCA16	–	–	ENSMUSG00000051900	No orthologue in man
ABCA17	–	–	ENSMUSG00000035435	No orthologue in man
ABCB1	MDR1, PGP1	Multi-drug resistance protein 1, P-glycoprotein 1, CD243 antigen	ENSG00000085563	Responsible for the cellular export of many therapeutic drugs
ABCB2	TAP1	Antigen peptide transporter 1, APT1, peptide transporter TAP1, peptide supply factor 1 (PSF-1), peptide transporter involved in antigen processing 1	ENSG00000168394	Endoplasmic reticulum, possibly as heterodimer with TAP2
ABCB3	TAP2	Antigen peptide transporter 2 (APT2), Peptide transporter TAP2, peptide supply factor 2 (PSF-2), peptide transporter involved in antigen processing 2	ENSG00000204267	Endoplasmic reticulum, possibly as heterodimer with TAP1
ABCB4	PGY3	Multi-drug resistance protein 3, P-glycoprotein 3	ENSG00000005471	Transports PC from intracellular to extracellular face of the hepatocyte canalicular membrane (Oude Elferink and Paulusma, 2007)
ABCB5	–	P-glycoprotein ABCB5	ENSG00000004846	–
ABCB6	MTABC3	Mitochondrial ABC transporter 3, ubiquitously expressed mammalian ABC half transporter, P-glycoprotein-related protein	ENSG00000115657	Mitochondrial
ABCB7	ABC7	–	ENSG00000131269	Mitochondrial; reportedly essential for haematopoiesis (Pondarre <i>et al.</i> , 2007)
ABCB8	MABC1	–	ENSG00000197150	Mitochondrial; suggested to play a role in chemoresistance of melanoma (Elliott and Al-Hajj, 2009)
ABCB9	TAPL	TAP-like protein, hABCB9	ENSG00000150967	Reported to be lysosomal (Kamakura <i>et al.</i> , 2008)
ABCB10	MTABC2	Mitochondrial ABC transporter 2	ENSG00000135776	Mitochondrial

ABCB11	ABC16	Bile salt export pump, BSEP, PFIC-2, PFIC2, PGY4, SPGP	ENSG00000073734	Loss-of-function mutations are associated with familial intrahepatic cholestasis (Stieger, 2009)
ABCC1	MRP1	Multidrug resistance-associated protein 1, Leukotriene C ₄ transporter	ENSG00000103222	Exhibits a broad substrate specificity (Bakos and Homolya, 2007)
ABCC2	MRP2, cMOAT	Multidrug resistance-associated protein 2, Canalicular multispecific organic anion transporter 1, Canalicular multidrug resistance protein	ENSG00000023839	Loss-of-function mutations are associated with Dubin-Johnson syndrome, in which plasma levels of conjugated bilirubin are elevated
ABCC3	MRP3	Multidrug resistance-associated protein 3, Canalicular multispecific organic anion transporter 2, Multi-specific organic anion transporter-D, MOAT-D	ENSG00000108846	Transports conjugates of glutathione, sulphate or glucuronide (see Borst <i>et al.</i> , 2007)
ABCC4	MRP4	Multidrug resistance-associated protein 4, Multi-specific organic anion transporter-B, MOAT-B	ENSG00000125257	Although reported to facilitate cellular cyclic nucleotide export, this role has been questioned (see Borst <i>et al.</i> , 2007); reported to export prostaglandins in a manner sensitive to NSAIDs (Reid <i>et al.</i> , 2003)
ABCC5	MRP5	Multidrug resistance-associated protein 5, Multi-specific organic anion transporter-C, MOAT-C, pABC11, SMRP	ENSG00000114770	Although reported to facilitate cellular cyclic nucleotide export, this role has been questioned (see Borst <i>et al.</i> , 2007)
ABCC6	MRP6	Multidrug resistance-associated protein 6, Anthracycline resistance-associated protein, Multi-specific organic anion transporter-E, MOAT-E	ENSG00000091262	–
ABCC7	CFTR	Cystic fibrosis transmembrane conductance regulator, cAMP-dependent chloride channel	ENSG00000001626	See page S131
ABCC8	SUR1	Sulfonylurea receptor 1	ENSG00000006071	See page S141
ABCC9	SUR2	Sulfonylurea receptor 2	ENSG000000069431	See page S141
ABCC10	MRP7	Multidrug resistance-associated protein 7	ENSG00000124574	–
ABCC11	MRP8	Multidrug resistance-associated protein 8	ENSG00000121270	Single nucleotide polymorphisms distinguish wet vs. dry earwax; reported to export chemotherapeutic nucleotides (Oguri <i>et al.</i> , 2007)
ABCC12	MRP9	Multidrug resistance-associated protein 9	ENSG00000140798	–
ABCC13		Putative ATP-binding cassette transporter C13	ENSG00000155288	A possible pseudogene
ABCD1	ALDP	Adrenoleukodystrophy protein	ENSG00000101986	Peroxisomal
ABCD2	ALDR	Adrenoleukodystrophy-related protein, adrenoleukodystrophy-like 1	ENSG00000173208	Peroxisomal
ABCD3	PMP70	70 kDa peroxisomal membrane protein, PXMP1	ENSG00000117528	Peroxisomal
ABCD4	PMP69	Peroxisomal membrane protein 69, peroxisomal membrane protein 1-like, PXMP1-L, P70R	ENSG00000119688	Peroxisomal
ABCE1	OABP	RNase L inhibitor, ribonuclease 4 inhibitor, RNS4I, 2'-5' oligoadenylate-binding protein	ENSG00000164163	Lacks transport function
ABCF1	ABC50	TNF- α -stimulated ABC protein	ENSG00000204574	Lacks transport function
ABCF2		Iron-inhibited ABC transporter 2	ENSG00000033050	Lacks transport function
ABCF3			ENSG00000161204	Lacks transport function
ABCG1	ABC8	White protein homolog	ENSG00000160179	–
ABCG2	ABCP	Placenta-specific ATP-binding cassette transporter, breast cancer resistance protein, BCRP, mitoxantrone resistance-associated protein, MXR, CD338 antigen, CDw338	ENSG00000118777	–
ABCG4		White2	ENSG00000172350	–
ABCG5		White3, Sterolin-1	ENSG00000138075	–
ABCG8		Sterolin-2	ENSG00000143921	–

ABCC8 and ABCC9 are unusual in that they lack transport capacity but regulate the activity of particular K⁺ channels (K_{ir}6.1-6.2), conferring nucleotide sensitivity to these channels to generate the canonical K_{ATP} channels.

Abbreviations: ABC, ATP-binding cassette; NBD, nucleotide-binding domain; *N-retinylPE*, *N*-retinylphosphatidylethanolamine; NSAID, non-steroidal anti-inflammatory drugs; PC, phosphatidylcholine

Further Reading

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