

Introduction

Reactome is open-source, open access, manually curated and peer-reviewed pathway database. Pathway annotations are authored by expert biologists, in collaboration with Reactome editorial staff and cross-referenced to many bioinformatics databases. A system of evidence tracking ensures that all assertions are backed up by the primary literature. Reactome is used by clinicians, geneticists, genomics researchers, and molecular biologists to interpret the results of high-throughput experimental studies, by bioinformaticians seeking to develop novel algorithms for mining knowledge from genomic studies, and by systems biologists building predictive models of normal and disease variant pathways.

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Literature references

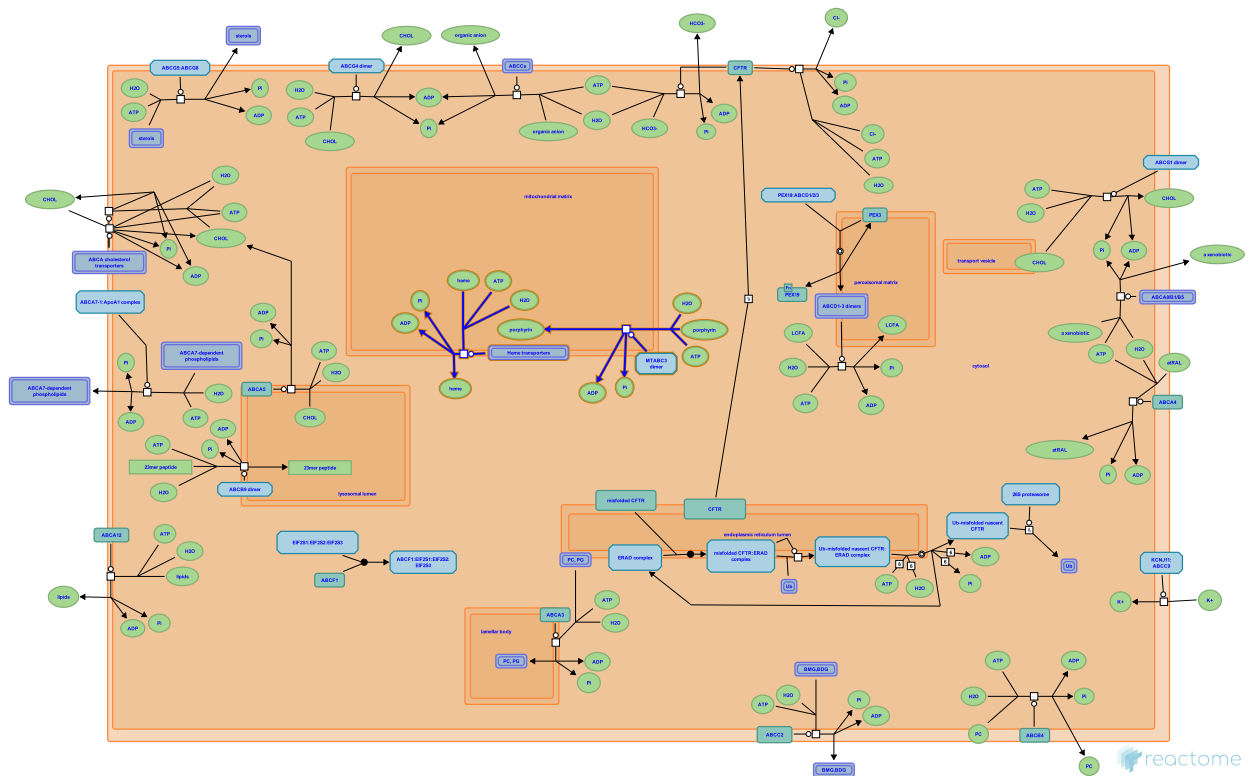
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Reactome database release: 70

This document contains 1 pathway and 2 reactions ([see Table of Contents](#))

Mitochondrial ABC transporters ↗

Stable identifier: R-HSA-1369007



Mammalian ABC transporters are usually found on the plasma membrane and on organelles such as the ER and peroxisome but a small number are also located on the mitochondria. Here they are thought to play roles in heme biosynthesis and iron-sulphur cluster synthesis (Burke & Ardehali 2007).

Literature references

Burke, MA., Ardehali, H. (2007). Mitochondrial ATP-binding cassette proteins. *Transl Res*, 150, 73-80. ↗

Editions

2011-07-04	Authored, Edited	Jassal, B.
2011-08-23	Reviewed	D'Eustachio, P.

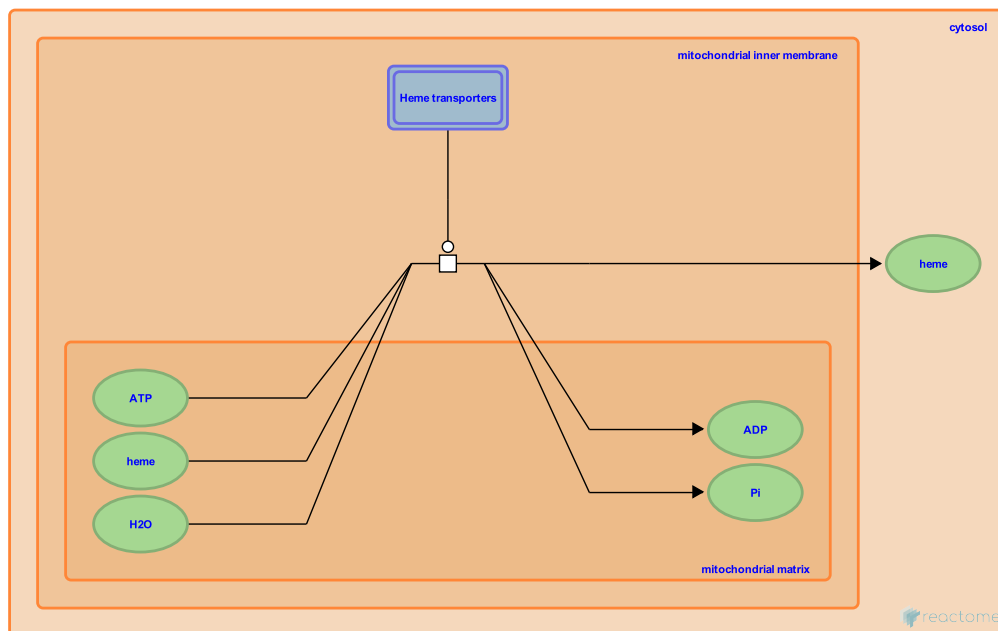
ABC7, mABC1 and mABC2 mediate heme transport ↗

Location: Mitochondrial ABC transporters

Stable identifier: R-HSA-382560

Type: transition

Compartments: mitochondrial inner membrane, cytosol, mitochondrial matrix



Mitochondrial ABC transporters are thought to play a key role in iron metabolism and heme biosynthesis. All mitochondrial ABC transporters described to date are of the half-transporter type and would probably function as dimers (Ramjeesingh et al. 2003) but their dimerization partners have not yet been identified. ABC7 is the functional human orthologue of yeast *Atm1p* (Csere et al. 1998), is predicted to dimerize in the same way as *Atm1p* (Chloupková et al. 2004) and is probably involved in iron homeostasis. Defects in ABCB7 are the cause of X-linked sideroblastic anemia with ataxia (ASAT) [MIM:301310] (Allikmets et al. 1999). Human genes ABCB8 and ABCB10 encode mABC1 and mABC2 respectively (Hogue et al. 1999, Zhang et al 2000 respectively). They would be expected to dimerize, as demonstrated for mABC2 (Graf et al. 2004). Both are believed to have similar functionality to ABC7 although this has not been demonstrated yet.

Literature references

- Csere, P., Lill, R., Kispal, G. (1998). Identification of a human mitochondrial ABC transporter, the functional orthologue of yeast *Atm1p*. *FEBS Lett*, 441, 266-70. ↗
- Chloupková, M., Reaves, SK., LeBard, LM., Koeller, DM. (2004). The mitochondrial ABC transporter *Atm1p* functions as a homodimer. *FEBS Lett*, 569, 65-9. ↗
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- Ramjeesingh, M., Ugwu, F., Li, C., Dhani, S., Huan, LJ., Wang, Y. et al. (2003). Stable dimeric assembly of the second membrane-spanning domain of CFTR (cystic fibrosis transmembrane conductance regulator) reconstitutes a chloride-selective pore. *Biochem J*, 375, 633-41. ↗
- Hogue, DL., Liu, L., Ling, V. (1999). Identification and characterization of a mammalian mitochondrial ATP-binding cassette membrane protein. *J Mol Biol*, 285, 379-89. ↗

Editions

2008-11-23	Authored, Edited	Gopinathrao, G.
2008-12-02	Reviewed	Matthews, L.
2011-07-04	Revised	Jassal, B.
2011-08-23	Reviewed	D'Eustachio, P.

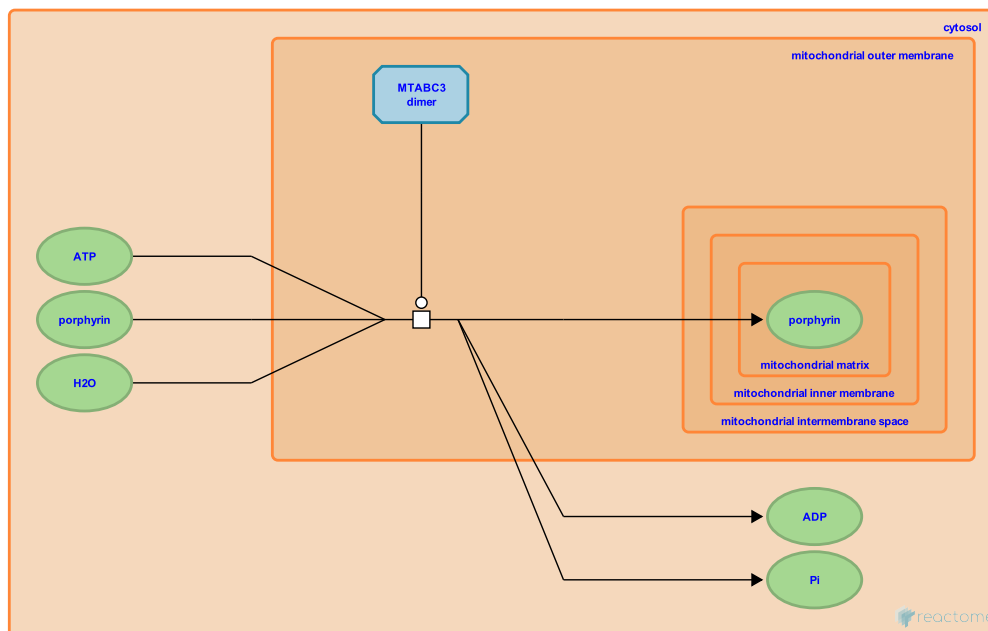
ABCB6 transports porphyrin from cytosol to mitochondrial matrix ↗

Location: [Mitochondrial ABC transporters](#)

Stable identifier: R-HSA-1369065

Type: transition

Compartments: mitochondrial outer membrane, cytosol, mitochondrial matrix



The human gene ABCB6 encodes a mitochondrial half-type ATP-binding cassette (ABC) protein MTABC3 which is uniquely located on the outer mitochondrial membrane and is functional as a homodimer (Krishnamurthy et al. 2006). It plays a crucial role in heme synthesis by mediating porphyrin uptake into mitochondria (Mitsuhashi et al. 2000, Krishnamurthy et al. 2006).

Literature references

Krishnamurthy, PC., Du, G., Fukuda, Y., Sun, D., Sampath, J., Mercer, KE. et al. (2006). Identification of a mammalian mitochondrial porphyrin transporter. *Nature*, 443, 586-9. ↗

Mitsuhashi, N., Miki, T., Senbongi, H., Yokoi, N., Yano, H., Miyazaki, M. et al. (2000). MTABC3, a novel mitochondrial ATP-binding cassette protein involved in iron homeostasis. *J Biol Chem*, 275, 17536-40. ↗

Editions

2011-07-04	Authored, Edited	Jassal, B.
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