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ABCB6 Rabbit pAb

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Catalog # AP54796

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q9NP58
Reactivity	Rat
Predicted	Pig, Dog, Human, Mouse, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	93886
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human ABCB6
Epitope Specificity	401-520/842
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Cell membrane. Mitochondrion outer membrane; Multi-pass membrane protein. Endoplasmic reticulum. Golgi apparatus.
DISEASE	Microphthalmia, isolated, with coloboma, 7 (MCOPCB7) [MIM:614497]: A disorder of eye formation, ranging from small size of a single eye to complete bilateral absence of ocular tissues. Ocular abnormalities like opacities of the cornea and lens, scaring of the retina and choroid, cataract and other abnormalities like cataract may also be present. Ocular colobomas are a set of malformations resulting from abnormal morphogenesis of the optic cup and stalk, and the fusion of the fetal fissure (optic fissure). Note=The disease is caused by mutations affecting the gene represented in this entry.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The membrane-associated protein encoded by this gene is a member of the superfamily of ATP-binding cassette (ABC) transporters. ABC proteins transport various molecules across extra- and intra-cellular membranes. ABC genes are divided into seven distinct subfamilies (ABC1, MDR/TAP, MRP, ALD, OABP, GCN20, White). This protein is a member of the MDR/TAP subfamily. Members of the MDR/TAP subfamily are involved in multidrug resistance as well as antigen presentation. This half-transporter likely plays a role in mitochondrial function. Localized to 2q26, this gene is considered a candidate gene for lethal neonatal metabolic syndrome, a disorder of mitochondrial function.

Additional Information

Gene ID	10058
Other Names	ATP-binding cassette sub-family B member 6, ABC-type heme transporter

	ABCB6, 7.6.2.5, Mitochondrial ABC transporter 3, Mt-ABC transporter 3, P-glycoprotein-related protein, Ubiquitously-expressed mammalian ABC half transporter, ABCB6 (HGNC:47)
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	ABCB6 (HGNC:47)
Function	ATP-dependent transporter that catalyzes the transport of a broad-spectrum of porphyrins from the cytoplasm to the extracellular space through the plasma membrane or into the vesicle lumen (PubMed:17661442, PubMed:23792964, PubMed:27507172, PubMed:33007128). May also function as an ATP-dependent importer of porphyrins from the cytoplasm into the mitochondria, in turn may participate in the de novo heme biosynthesis regulation and in the coordination of heme and iron homeostasis during phenylhydrazine stress (PubMed:10837493, PubMed:17006453, PubMed:23792964, PubMed:33007128). May also play a key role in the early steps of melanogenesis producing PMEL amyloid fibrils (PubMed:29940187). In vitro, it confers to cells a resistance to toxic metal such as arsenic and cadmium and against chemotherapeutics agent such as 5-fluorouracil, SN-38 and vincristin (PubMed:21266531, PubMed:25202056, PubMed:31053883). In addition may play a role in the transition metal homeostasis (By similarity).
Cellular Location	Cell membrane; Multi-pass membrane protein. Mitochondrion outer membrane; Multi-pass membrane protein. Endoplasmic reticulum membrane; Multi-pass membrane protein. Golgi apparatus membrane; Multi-pass membrane protein. Endosome membrane; Multi-pass membrane protein. Lysosome membrane. Late endosome membrane {ECO:0000250|UniProtKB:O70595}. Early endosome membrane {ECO:0000250|UniProtKB:O70595}. Secreted, extracellular exosome. Mitochondrion. Endosome, multivesicular body membrane. Melanosome membrane. Note=Present in the membrane of mature erythrocytes and in exosomes released from reticulocytes during the final steps of erythroid maturation (PubMed:22655043). Traffics from endoplasmic reticulum to Golgi during its glycans's maturation, therefrom is first targeted to the plasma membrane, and is rapidly internalized through endocytosis to be distributed to the limiting membrane of multivesicular bodies and lysosomes (PubMed:18279659, PubMed:21199866, PubMed:25627919). Localized on the limiting membrane of early melanosomes of pigment cells (PubMed:29940187). Targeted to the endolysosomal compartment (By similarity) {ECO:0000250|UniProtKB:O70595, ECO:0000269|PubMed:18279659, ECO:0000269|PubMed:21199866, ECO:0000269|PubMed:22655043, ECO:0000269|PubMed:25627919, ECO:0000269|PubMed:29940187}
Tissue Location	Widely expressed. High expression is detected in the retinal epithelium (PubMed:10837493, PubMed:22226084). Expressed in mature erythrocytes (PubMed:22655043).

Background

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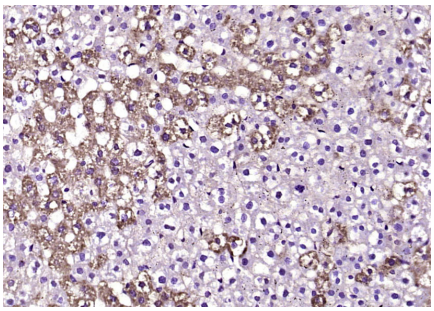
Images



Paraformaldehyde-fixed, paraffin embedded (rat liver); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (ABCB6) Polyclonal Antibody, Unconjugated (AP54796) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.