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Rabbit Anti-ABCB7 antibody

SL12331R

Product Name:	ABCB7
Chinese Name:	ATPBinding protein家族7抗体
Alias:	ABC transporter 7 protein; ABC7; Abcb7; ABCB7_HUMAN; ASAT; Atm1p; ATP binding cassette 7; ATP binding cassette sub family B (MDR/TAP) member 7; ATP binding cassette sub family B member 7; ATP binding cassette sub family B member 7 mitochondrial; ATP binding cassette transporter 7; ATP-binding cassette sub-family B member 7; ATP-binding cassette transporter 7; EST140535; MDR7; mitochondrial; Multidrug resistance protein 7; P-glycoprotein 7; PGP7.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Rabbit,Sheep,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	83kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from Human ABCB7:201-300/752
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	The peroxisomal membrane contains several ATP-binding cassette (ABC) transporters, ABCD1–4 that are known to be present in the human peroxisome membrane (1). All

four proteins are ABC half-transporters, which dimerize to form an active transporter (1). A mutation in the ABCD1 causes X-linked adrenoleukodystrophy (X-ALD), a peroxisomal disorder which affects lipid storage (2,3). ABCD2 in mouse, is expressed at high levels in the brain and adrenal organs, which are adversely affected in X-ALD (4). The peroxisomal membrane comprises 2 quantitatively major proteins, PMP22 and ABCD3 (5). ABCD3 is associated with irregularly shaped vesicles which may be defective peroxisomes or peroxisome precursors (5). ABCD4 localizes to peroxisomes (1). The genes which encode ABCD1–4 map to human chromosome Xq28, 12q11-q12, 1p22-p21 and 14q24.3, respectively (3,6–8). ABCB7 is a half-transporter involved in the transport of heme from the mitochondria to the cytosol and maps to human chromosome Xq13.1-q13.3 (9).

Function:
Could be involved in the transport of heme from the mitochondria to the cytosol. Plays a central role in the maturation of cytosolic iron-sulfur (Fe/S) cluster-containing proteins.

Subunit:
Homodimer or heterodimer (Potential).

Subcellular Location:
Mitochondrion inner membrane.

DISEASE:
Defects in ABCB7 are the cause of X-linked sideroblastic anemia with ataxia (ASAT) [MIM:301310]. ASAT is a recessive disorder characterized by an infantile to early childhood onset of nonprogressive cerebellar ataxia and mild anemia with hypochromia and microcytosis.

Similarity:
Belongs to the ABC transporter superfamily.
ABCB family.
Heavy Metal importer (TC 3.A.1.210) subfamily.
Contains 1 ABC transmembrane type-1 domain.
Contains 1 ABC transporter domain.

SWISS:
O75027

Gene ID:
22

Database links:

Entrez Gene: 22Human

	Entrez Gene: 11306Mouse Entrez Gene: 302395Rat Omim: 300135Human SwissProt: O75027Human SwissProt: Q61102Mouse SwissProt: Q704E8Rat Unigene: 370480Human Unigene: 426128Mouse Unigene: 20068Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.