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ABCD4 Rabbit pAb

Catalog # AP97110

Product Information

Application	WB
Primary Accession	O14678
Other Accession	O14678
Reactivity	Human
Predicted	Rat, Mouse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	68597
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human ABCD4
Epitope Specificity	351-450/606
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the ABC transporter superfamily. ABCD family. Peroxisomal fatty acyl CoA transporter (TC 3.A.1.203) subfamily. Contains 1 ABC transmembrane type-1 domain. Contains 1 ABC transporter domain.
SUBUNIT	Homodimer or heterodimer
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The peroxisomal membrane contains several ATP-binding cassette (ABC) transporters, ABCD1-4 that are known to be present in the human peroxisome membrane. All four proteins are ABC half-transporters, which dimerize to form an active transporter. A mutation in the ABCD1 gene causes X-linked adreno-leukodystrophy (X-ALD), a peroxisomal disorder which affects lipid storage. ABCD2 in mouse is expressed at high levels in the brain and adrenal organs, which are adversely affected in X-ALD. The peroxisomal membrane comprises two quantitatively major proteins, PMP22 and ABCD3. ABCD3 is associated with irregularly shaped vesicles which may be defective peroxisomes or peroxisome precursors. ABCD1 localizes to peroxisomes. ABCB7 is a half-transporter involved in the transport of heme from the mitochondria to the cytosol.

Additional Information

Gene ID	5826
Other Names	CK820_G0002881; ABC41; EST352188; MAHCJ; P70R; P79R; PMP69; PXMP1L; P69r; ABCD4_HUMAN; ABCD4; ATP-binding cassette sub-family D member 4; PMP70-related protein (P70R); Peroxisomal membrane protein 1-like (PXMP1-L); Peroxisomal membrane protein 69 (PMP69); 7.6.2.8; ABCD4_MOUSE; ATP binding cassette subfamily D member 4; ATP-binding cassette, sub-family D (ALD), member 4; Lysosomal cobalamin transporter

	ABCD4
Dilution	WB=1:500-2000
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	ABCD4 (HGNC:68)
Synonyms	PXMP1L
Function	Lysosomal membrane protein that transports cobalamin (Vitamin B12) from the lysosomal lumen to the cytosol in an ATP-dependent manner (PubMed:22922874, PubMed:28572511, PubMed:31467407, PubMed:33845046). Targeted by LMBRD1 lysosomal chaperone from the endoplasmic reticulum to the lysosomal membrane (PubMed:27456980). Then forms a complex with lysosomal chaperone LMBRD1 and cytosolic MMACHC to transport cobalamin across the lysosomal membrane (PubMed:25535791).
Cellular Location	Endoplasmic reticulum membrane; Multi-pass membrane protein. Lysosome membrane; Multi-pass membrane protein. Note=Targeted by LMBRD1 lysosomal chaperone to the lysosomal membrane.
Tissue Location	Ubiquitous.

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