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ABCD1 Antibody (Center)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP10454c

Product Information

Application	WB, IHC-P, FC, E
Primary Accession	P33897
Other Accession	P48410, NP_000024.2
Reactivity	Human, Mouse
Predicted	Mouse
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB18846
Calculated MW	82937
Antigen Region	257-285

Additional Information

Gene ID	215
Other Names	ATP-binding cassette sub-family D member 1, Adrenoleukodystrophy protein, ALDP, ABCD1, ALD
Target/Specificity	This ABCD1 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 257-285 amino acids from the Central region of human ABCD1.
Dilution	WB~~1:1000 IHC-P~~1:100~500 FC~~1:10~50 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	ABCD1 Antibody (Center) is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	ABCD1 (HGNC:61)
Synonyms	ALD

Function	ATP-dependent transporter of the ATP-binding cassette (ABC) family involved in the transport of very long chain fatty acid (VLCFA)- CoA from the cytosol to the peroxisome lumen (PubMed:11248239, PubMed:15682271, PubMed:16946495, PubMed:18757502, PubMed:21145416, PubMed:23671276, PubMed:29397936, PubMed:33500543). Coupled to the ATP- dependent transporter activity also has a fatty acyl-CoA thioesterase activity (ACOT) and hydrolyzes VLCFA-CoA into VLCFA prior their ATP-dependent transport into peroxisomes, the ACOT activity is essential during this transport process (PubMed:29397936, PubMed:33500543). Thus, plays a role in regulation of VLCFAs and energy metabolism namely, in the degradation and biosynthesis of fatty acids by beta-oxidation, mitochondrial function and microsomal fatty acid elongation (PubMed:21145416, PubMed:23671276). Involved in several processes; namely, controls the active myelination phase by negatively regulating the microsomal fatty acid elongation activity and may also play a role in axon and myelin maintenance. Also controls the cellular response to oxidative stress by regulating mitochondrial functions such as mitochondrial oxidative phosphorylation and depolarization. And finally controls the inflammatory response by positively regulating peroxisomal beta-oxidation of VLCFAs (By similarity).
Cellular Location	Peroxisome membrane; Multi-pass membrane protein. Mitochondrion membrane; Multi-pass membrane protein. Lysosome membrane; Multi-pass membrane protein Endoplasmic reticulum membrane; Multi- pass membrane protein

Background

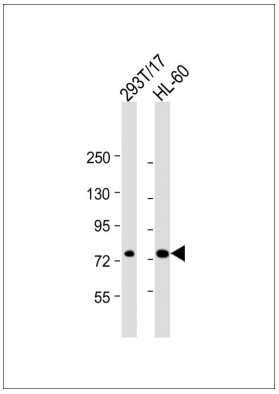
ABCD1 is a member of the superfamily of ATP-binding cassette (ABC) transporters. ABC proteins transport various molecules across extra- and intra-cellular membranes. ABC genes are divided into seven distinct subfamilies (ABC1, MDR/TAP, MRP, ALD, OABP, GCN20, White). This protein is a member of the ALD subfamily, which is involved in peroxisomal import of fatty acids and/or fatty acyl-CoAs in the organelle. All known peroxisomal ABC transporters are half transporters which require a partner half transporter molecule to form a functional homodimeric or heterodimeric transporter. This peroxisomal membrane protein is likely involved in the peroxisomal transport or catabolism of very long chain fatty acids. Defects in this gene have been identified as the underlying cause of adrenoleukodystrophy, an X-chromosome recessively inherited demyelinating disorder of the nervous system.

References

Matsukawa, T., et al. Neurogenetics (2010) In press :
Xie, H.H., et al. Zhonghua Yi Xue Yi Chuan Xue Za Zhi 27(2):144-148(2010)
Li, J.Y., et al. J. Neurol. Sci. 290 (1-2), 163-165 (2010) :
Hour, T.C., et al. Int. J. Biol. Markers 24(3):171-178(2009)
Shukla, P., et al. J. Child Neurol. 24(7):857-860(2009)

Images

All lanes : Anti-ABCD1 Antibody (Center) at 1:2000 dilution Lane 1: 293T/17 whole cell lysate Lane 2: HL-60 whole cell lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/10000 dilution. Predicted band size : 83 kDa Blocking/Dilution buffer: 5% NFDM/TBST.



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