

Product Name: ABCD1 Rabbit Polyclonal Antibody
Catalog #: APRab06415



Summary

Production Name	ABCD1 Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	WB,ELISA
Reactivity	Human,Rat,Mouse

Performance

Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Purification	Affinity purification

Immunogen

Gene Name	ABCD1
Alternative Names	ABCD1; ALD; ATP-binding cassette sub-family D member 1; Adrenoleukodystrophy protein; ALDP
Gene ID	215.0
SwissProt ID	P33897.The antiserum was produced against synthesized peptide derived from human ABCD1. AA range:531-580

Application

Dilution Ratio	WB 1:500 - 1:2000. ELISA: 1:40000. Not yet tested in other applications.
Molecular Weight	75kD

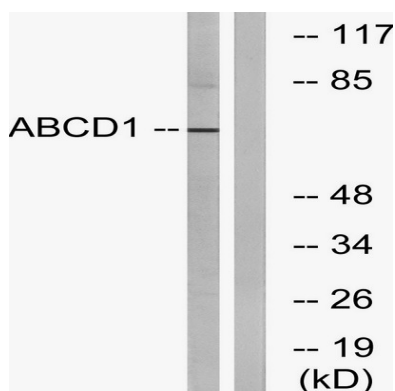
Background

The 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 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. Defects in ABCD1 are the cause of adrenoleukodystrophy X-linked (X-ALD) [MIM:300100]. X-ALD is a peroxisomal metabolic disorder characterized by progressive multifocal demyelination of the central nervous system and by peripheral adrenal insufficiency (Addison disease). It results in mental deterioration, corticospinal tract dysfunction, and cortical blindness. Different clinical manifestations exist like: cerebral childhood ALD (CALD), adult cerebral ALD (ACALD), adrenomyeloneuropathy (AMN) and "Addison disease only" (ADO) phenotype. Microdeletions in ABCD1 are involved in the contiguous ABCD1/DXS1375E deletion syndrome (CADD5) [MIM:300475]. Patients manifest profound neonatal hypotonia, subsequent failure to thrive, and cholestatic liver disease. Probable transporter. The nucleotide-binding fold acts as an ATP-binding subunit with ATPase activity. Belongs to the ABC transporter family. ALD subfamily. Contains 1 ABC transmembrane type-1 domain. Contains 1 ABC transporter domain. Can form homo- and heterodimers with ABCD2/ALDR and ABCD3/PMP70. Dimerization is necessary to form an active transporter. Interacts with PEX19.

Research Area

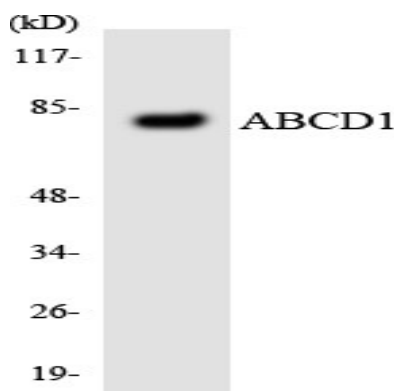
ABC transporters;

Image Data

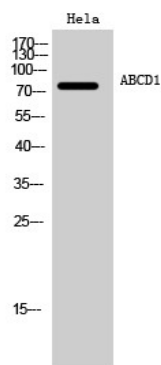


Western blot analysis of lysates from Jurkat cells, using ABCD1 Antibody. The lane on the right is blocked with the synthesized peptide.

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Western blot analysis of the lysates from HeLa cells using ABCD1 antibody.



Western Blot analysis of HeLa cells using ABCD1 Polyclonal Antibody

Note

For research use only.