

产品名称: ABCD1 Rabbit Polyclonal Antibody
产品货号: APRab06415



产品概述 (Summary)

产品名称 (Production Name)	ABCD1 Rabbit Polyclonal Antibody
描述 (Description)	Rabbit polyclonal Antibody
宿主 (Host)	Rabbit
应用 (Application)	WB,ELISA
种属反应性 (Reactivity)	Human,Rat,Mouse

产品性能 (Performance)

偶联物 (Conjugation)	Unconjugated
修饰 (Modification)	Unmodified
同种型 (Isotype)	IgG
克隆 (Clonality)	Polyclonal
形式 (Form)	Liquid
存放说明 (Storage)	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
储存溶液 (Buffer)	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
纯化方式 (Purification)	Affinity purification

免疫原信息 (Immunogen)

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

产品应用 (Application)

稀释比 (Dilution Ratio)	WB 1:500-1:2000,ELISA 1:20000-1:40000
蛋白分子量 (Molecular Weight)	75kDa

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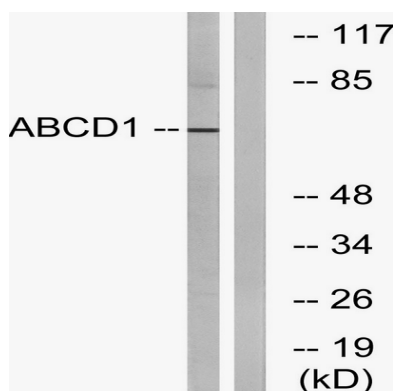
研究背景 (Background)

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.

研究领域 (Research Area)

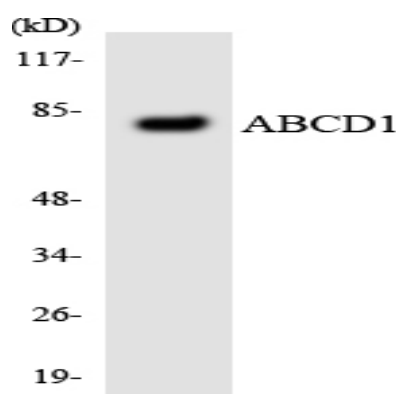
ABC transporters;

图片 (Image Data)

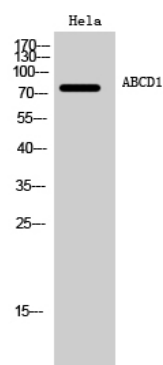


Western blot analysis of lysates from Jurkat cells, using ABCD1 Antibody. The lane on the right is blocked with the synthesized peptide.

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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

注意事项 (Note)

For research use only .