

### **Goat Anti-ABCD1 Antibody**

Peptide-affinity purified goat antibody Catalog # AF1011a

## **Specification**

### **Goat Anti-ABCD1 Antibody - Product Information**

Application WB
Primary Accession P33897

Other Accession NP\_000024, 215

Reactivity
Host
Clonality
Concentration
Isotype
Human
Goat
Polyclonal
100ug/200ul
IgG

Isotype IgG
Calculated MW 82937

## **Goat Anti-ABCD1 Antibody - Additional Information**

#### Gene ID 215

### **Other Names**

ATP-binding cassette sub-family D member 1, Adrenoleukodystrophy protein, ALDP, ABCD1, ALD

### **Format**

0.5 mg lgG/ml in Tris saline (20mM Tris pH7.3, 150mM NaCl), 0.02% sodium azide, with 0.5% bovine serum albumin

## **Storage**

Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

#### **Precautions**

Goat Anti-ABCD1 Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

## **Goat Anti-ABCD1 Antibody - 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:<a href="http://www.uniprot.org/citations/11248239" target="\_blank">11248239</a>, PubMed:<a href="http://www.uniprot.org/citations/15682271" target="\_blank">15682271</a>, PubMed:<a href="http://www.uniprot.org/citations/16946495" target="\_blank">16946495</a>, PubMed:<a



href="http://www.uniprot.org/citations/18757502" target=" blank">18757502</a>, PubMed:<a href="http://www.uniprot.org/citations/21145416" target="\_blank">21145416</a>, PubMed:<a href="http://www.uniprot.org/citations/23671276" target="\_blank">23671276</a>, PubMed:<a href="http://www.uniprot.org/citations/29397936" target="\_blank">29397936</a>, PubMed:<a href="http://www.uniprot.org/citations/33500543" target="blank">33500543</a>). Coupled to the ATP- dependent transporter activity has also 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: <a  $href="http://www.uniprot.org/citations/29397936" target="\_blank">29397936</a>, PubMed:<a href="http://www.uniprot.org/citations/33500543" target="\_blank">33500543</a>). Thus, plays a$ 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:<a href="http://www.uniprot.org/citations/21145416" target=" blank">21145416</a>, PubMed:<a href="http://www.uniprot.org/citations/23671276" target=" blank">23671276</a>). 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. Controls also 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

#### Goat Anti-ABCD1 Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

### Goat Anti-ABCD1 Antibody - Images



HEK293 overexpressing human ABCD1 and probed with AF1011a at 1ug/ml (mock transfection in



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first lane).

## Goat Anti-ABCD1 Antibody - 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 of the nervous system.

## Goat Anti-ABCD1 Antibody - References

Identification of novel SNPs of ABCD1, ABCD2, ABCD3, and ABCD4 genes in patients with X-linked adrenoleukodystrophy (ALD) based on comprehensive resequencing and association studies with ALD phenotypes. Matsukawa T, et al. Neurogenetics, 2010 Jul 27. PMID 20661612.

[A novel missense mutation resulting in X-linked adrenoleukodystrophy in female heterozygotes of a Chinese family] Xie HH, et al. Zhonghua Yi Xue Yi Chuan Xue Za Zhi, 2010 Apr. PMID 20376793. Spinocerebellar variant of adrenoleukodystrophy with a novel ABCD1 gene mutation. Li JY, et al. J Neurol Sci, 2010 Mar 15. PMID 20042197.

Downregulation of ABCD1 in human renal cell carcinoma. Hour TC, et al. Int J Biol Markers, 2009 Jul-Sep. PMID 19787628.

Three novel variants in X-linked adrenoleukodystrophy. Shukla P, et al. J Child Neurol, 2009 Jul. PMID 19406751.