



ABCD1 Monoclonal Antibody

Catalog No	YP-mAb-00655
Isotype	IgG
Reactivity	Human;Rat;Mouse;
Applications	WB
Gene Name	ABCD1
Protein Name	ATP-binding cassette sub-family D member 1
Immunogen	The antiserum was produced against synthesized peptide derived from human ABCD1. AA range:531-580
Specificity	ABCD1 Monoclonal Antibody detects endogenous levels of ABCD1 protein.
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-1:2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	ABCD1; ALD; ATP-binding cassette sub-family D member 1; Adrenoleukodystrophy protein; ALDP
Observed Band	75kD
Cell Pathway	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.
Tissue Specificity	Brain,Pancreas,
Function	disease: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.,disease: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.,function:Probable transporter. The nucleotide-binding fold acts as an ATP-binding subunit with ATPase activity.,similarity:Be



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

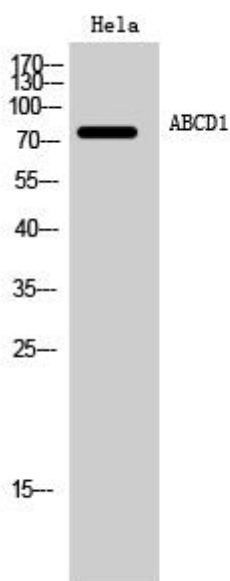
matters needing attention

Avoid repeated freezing and thawing!

Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western Blot analysis of various cells using ABCD1 Monoclonal Antibody