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ACADL Antibody (N-term)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP13134a

Product Information

Application	WB, E
Primary Accession	P28330
Other Accession	NP_001599.1
Reactivity	Human, Rat, Mouse
Predicted	Bovine, Canine, Rabbit
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB32459
Calculated MW	47656
Antigen Region	14-43

Additional Information

Gene ID	33
Other Names	Long-chain specific acyl-CoA dehydrogenase, mitochondrial, LCAD, ACADL
Target/Specificity	This ACADL antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 14-43 amino acids from the N-terminal region of human ACADL.
Dilution	WB~~1:1000 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	ACADL Antibody (N-term) is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	ACADL (HGNC:88)
Function	Catalyzes the first of the four reactions of the mitochondrial fatty acid beta-oxidation (FAO) pathway, which consists in the proR-proR stereospecific alpha, beta-dehydrogenation of fatty acyl-CoA thioesters using the electron

transfer flavoprotein (ETF) as their physiologic electron acceptor, resulting in the formation of trans-2-enoyl-CoA ((2E)-enoyl-CoA) (PubMed:17564966, PubMed:21237683, PubMed:24591516, PubMed:32389575). The mitochondrial FAO pathway is the major energy-producing process in tissues and is performed through cycles of four consecutive reactions (PubMed:17564966, PubMed:26474213). Each FAO cycle shortens the fatty acyl-CoA by two carbons, yielding one acetyl-CoA (for the citric acid cycle), one FADH(2), and one NADH (which donate electrons to the respiratory chain for ATP production) (PubMed:17564966, PubMed:26474213). Among the different mitochondrial acyl-CoA dehydrogenases, long-chain specific acyl-CoA dehydrogenase activity overlaps with that of ACADV and ACAD9, acting on saturated and unsaturated acyl-CoAs with 6 to 24 carbons with a preference for 8 to 18 carbons long primary chains (PubMed:17564966, PubMed:21237683, PubMed:32389575, PubMed:8823175). Can use (4Z,7Z,10Z,13Z,16Z,19Z)-docosahexaenoate as substrate in vitro (which is not primarily used for energy but mainly beta-oxidized in the peroxisomes) (PubMed:17564966, PubMed:26474213). In addition, based on its established catalytic mechanism, and combined genetic interaction or mutant phenotype evidence, it is predicted to act also on substrates that have not been tested experimentally but are metabolized by mitochondrial FAO, including long-chain unsaturated fatty acids such as linoleate (9Z,12Z-octadecadienoate), linolenate (9Z,12Z,15Z- octadecatrienoate), and others (PubMed:26474213). Plays a primary role in FAO in tissues where it is the main long-chain ACAD expressed, such as the lung, specifically in type 2 alveolar cells (responsible for surfactant production) (PubMed:17564966, PubMed:24591516). Probably responsible for beta-oxidation of bulky substrates including branched chain fatty acyl-CoAs and sterol derivatives thanks to its enlarged substrate-binding cavity (PubMed:38839792).

Cellular Location

Mitochondrion matrix {ECO:0000250|UniProtKB:P15650}

Tissue Location

Expressed at mRNA and protein levels in lungs, where it localizes specifically in alveolar epithelial cells (alveolar type II pneumocytes) (PubMed:17564966, PubMed:24591516). Also expressed at mRNA levels in prostate, thyroid gland, kidney, heart and muscle (PubMed:17564966).

Background

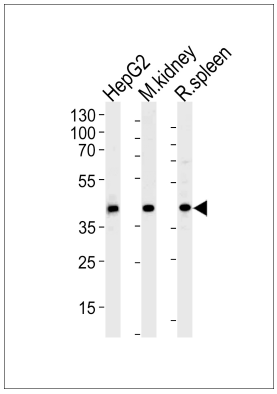
The protein encoded by this gene belongs to the acyl-CoA dehydrogenase family, which is a family of mitochondrial flavoenzymes involved in fatty acid and branched chain amino-acid metabolism. This protein is one of the four enzymes that catalyze the initial step of mitochondrial beta-oxidation of straight-chain fatty acid. Defects in this gene are the cause of long-chain acyl-CoA dehydrogenase (LCAD) deficiency, leading to nonketotic hypoglycemia.

References

Bailey, S.D., et al. Diabetes Care 33(10):2250-2253(2010)
Maher, A.C., et al. Mol. Genet. Metab. 100(2):163-167(2010)
Illig, T., et al. Nat. Genet. 42(2):137-141(2010)
Talmud, P.J., et al. Am. J. Hum. Genet. 85(5):628-642(2009)
Lu, Y., et al. J. Lipid Res. 49(12):2582-2589(2008)

Images

Western blot analysis of lysates from HepG2 cell line and mouse kidney, rat spleen tissue lysates(from left to right),



using ACADL Antibody (N-term)(Cat. #AP13134a). AP13134a was diluted at 1:1000 at each lane. A goat anti-rabbit IgG H&L(HRP) at 1:5000 dilution was used as the secondary antibody. Lysates at 35ug per lane.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.