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ACADL Rabbit pAb

Catalog # AP99840

Product Information

Application	WB, E
Primary Accession	P28330
Other Accession	P28330
Reactivity	Human
Predicted	Rat, Mouse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	47656
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human ACADL
Epitope Specificity	201-300/430
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the acyl-CoA dehydrogenase family.
SUBUNIT	Homotetramer.
DISEASE	Defects in ACADL are a cause of acyl-CoA dehydrogenase very long-chain deficiency (ACADVLD) [MIM:201475]. An inborn error of mitochondrial fatty acid beta-oxidation which leads to impaired long-chain fatty acid beta-oxidation. It is clinically heterogeneous, with three major phenotypes: a severe childhood form characterized by early onset, high mortality and high incidence of cardiomyopathy; a milder childhood form with later onset, characterized by hypoketotic hypoglycemia, low mortality and rare cardiomyopathy; an adult form, with isolated skeletal muscle involvement, rhabdomyolysis and myoglobinuria, usually triggered by exercise or fasting.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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. [provided by RefSeq].

Additional Information

Gene ID	33
Other Names	ACAD4; LCAD; ACOADA; ACADL_HUMAN; ACADL; 1.3.8.8; ACADL_MOUSE; ACADL_PIG; ACADL_RAT; acyl-CoA dehydrogenase long chain; acyl-Coenzyme A dehydrogenase, long chain; long-chain acyl-CoA dehydrogenase

Dilution	WB=1:500-2000,ICC/IF=1:100-500,ELISA=1:5000-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

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).

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