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ACADVL Rabbit pAb

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Catalog # AP58258

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P49748
Reactivity	Rat
Predicted	Pig, Dog, Human, Mouse, Rabbit, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	70390
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human ACADVL
Epitope Specificity	251-350/655
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Mitochondrion inner membrane.
DISEASE	Defects in ACADVL are the cause of acyl-CoA dehydrogenase very long chain deficiency (ACADVLD) [MIM:201475]. ACADVLD is an autosomal recessive disease which leads to impaired long-chain fatty acid beta-oxidation. It is clinically heterogeneous, with three major phenotypes: a severe childhood form, with early onset, high mortality, and high incidence of cardiomyopathy; a milder childhood form, with later onset, usually with hypoketotic hypoglycemia as the main presenting feature, low mortality, and rare cardiomyopathy; and 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	ACADVL (acyl-Coenzyme A dehydrogenase, very long chain) catalyzes the first step of the mitochondrial fatty acid beta-oxidation pathway. It is specific to esters of long-chain and very long chain fatty acids such as palmitoyl-CoA and stearoyl-CoA. Deficiencies in ACADVL are associated with reduced myocardial fatty acid beta-oxidation and cardiomyopathy.

Additional Information

Gene ID	37
Other Names	Very long-chain acyl-CoA dehydrogenase, mitochondrial, VLCAD, 1.3.8.9, Long-chain acyl-CoA dehydrogenase, mitochondrial, 1.3.8.8, ACADVL (HGNC:92)
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500

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

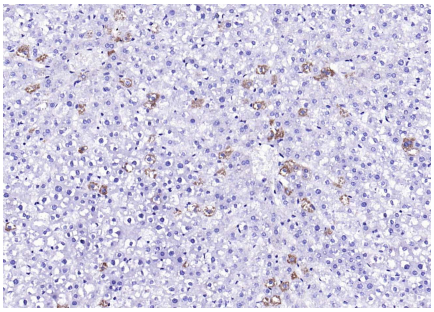
Name	ACADVL (HGNC:92)
Function	Catalyzes the first of the four reactions of the mitochondrial fatty acid beta-oxidation (FAO) pathway, which consists in the proR-proR stereospecific alpha, beta-dehydrogenation of fatty acyl-CoA thioesters using the electron transfer flavoprotein (ETF) as their physiologic electron acceptor, resulting in the formation of trans-2-enoyl-CoA ((2E)-enoyl-CoA) (PubMed:17564966, PubMed:18227065, PubMed:26474213, PubMed:7668252, PubMed:9461620, PubMed:9599005, PubMed:9839948). The mitochondrial FAO pathway is the major energy- producing process in tissues and is performed through cycles of four consecutive reactions (PubMed:26474213, PubMed:7668252). 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:26474213, PubMed:7668252). Among the different mitochondrial acyl-CoA dehydrogenases, very long-chain specific acyl-CoA dehydrogenase acts specifically on fatty acyl-CoAs with saturated 12 to 24 carbons long primary chains (PubMed:17564966, PubMed:21237683, PubMed:9839948), but can also catalyze monounsaturated fatty acids such as oleate ((9Z)-octadecenoate), (9Z)-hexadecenoate, and others (PubMed:17564966). 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). Among the different mitochondrial acyl-CoA dehydrogenases, its FAO activity overlaps with that of ACAD9 and ACADL, but plays a primary role in tissues where it is the main long-chain ACAD expressed, such as the heart and skeletal muscle (PubMed:17564966).
Cellular Location	Mitochondrion inner membrane; Peripheral membrane protein
Tissue Location	Predominantly expressed in heart and skeletal muscle (at both mRNA and protein levels) (PubMed:17564966, PubMed:8845838). Also detected in kidney and liver (at protein level) (PubMed:8845838).

Background

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Images

Paraformaldehyde-fixed, paraffin embedded (rat liver); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3%



hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (ACADVL) Polyclonal Antibody, Unconjugated (AP58258) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.

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