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Rabbit Anti-ACADL antibody

SL10150R

Product Name:	ACADL
Chinese Name:	酰基辅酶A脱氢酶长链抗体
Alias:	mitochondrial; ACAD4; ACADL; ACADL_HUMAN; Acyl Coenzyme A dehydrogenase long chain; FLJ94052; LCAD; Long chain acyl CoA dehydrogenase; Long-chain specific acyl-CoA dehydrogenase.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	44kDa
Cellular localization:	cytoplasmicMitochondrion
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human ACADL:31-130/430
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	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].

Subunit:
Homotetramer.

Subcellular Location:
Mitochondrion matrix.

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.

Similarity:
Belongs to the acyl-CoA dehydrogenase family.

SWISS:
P28330

Gene ID:
33

Database links:

Entrez Gene: 33Human

Entrez Gene: 614508 Cow

Entrez Gene: 11363 Mouse

Entrez Gene: 396931 Pig

Entrez Gene: 25287 Rat

Omim: 609576 Human

SwissProt: P28330 Human

SwissProt: P51174 Mouse

SwissProt: P79274 Pig

	SwissProt: P15650 Rat Unigene: 471277 Human Unigene: 2445 Mouse Unigene: 174 Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.