



# ACADV Monoclonal Antibody

<b>Catalog No</b>	BYmab-07315
<b>Isotype</b>	IgG
<b>Reactivity</b>	Human;Mouse
<b>Applications</b>	WB
<b>Gene Name</b>	ACADVL VLCAD
<b>Protein Name</b>	Very long-chain specific acyl-CoA dehydrogenase, mitochondrial (VLCAD) (EC 1.3.8.9)
<b>Immunogen</b>	Synthesized peptide derived from human protein . at AA range: 471-520
<b>Specificity</b>	ACADV Monoclonal Antibody detects endogenous levels of protein.
<b>Formulation</b>	Liquid in PBS containing 50% glycerol, and 0.02% sodium azide.
<b>Source</b>	Monoclonal, Mouse,IgG
<b>Purification</b>	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
<b>Dilution</b>	WB 1:500-2000
<b>Concentration</b>	1 mg/ml
<b>Purity</b>	≥90%
<b>Storage Stability</b>	-20°C/1 year
<b>Synonyms</b>	
<b>Observed Band</b>	72kD
<b>Cell Pathway</b>	Mitochondrion inner membrane ; Peripheral membrane protein .; [Isoform 2]: Mitochondrion inner membrane ; Peripheral membrane protein .
<b>Tissue Specificity</b>	Adipose tissue,Liver,Lung,Pancreas,Peripheral blood,Placenta,
<b>Function</b>	catalytic activity:Acyl-CoA + ETF = 2,3-dehydroacyl-CoA + reduced ETF.,cofactor:FAD.,disease:Defects in ACADVL are the cause of very long chain acyl-CoA dehydrogenase deficiency (VLCAD deficiency) [MIM:201475]. VLCAD deficiency 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.,function:Active toward esters of long-chain and very long chain fatty acids

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such as palmitoyl-CoA, mysritoyl-CoA and stearoyl-

**Background**

acyl-CoA dehydrogenase, very long chain(ACADVL) Homo sapiens The protein encoded by this gene is targeted to the inner mitochondrial membrane where it catalyzes the first step of the mitochondrial fatty acid beta-oxidation pathway. This acyl-Coenzyme A dehydrogenase is specific to long-chain and very-long-chain fatty acids. A deficiency in this gene product reduces myocardial fatty acid beta-oxidation and is associated with cardiomyopathy. Alternative splicing results in multiple transcript variants encoding different isoforms. [provided by RefSeq, Jul 2008],

**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

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