abcam

Product datasheet

Recombinant Human ACADVL protein ab98234

1 图像

概述

产品名称	重组人ACADVL蛋白
蛋白长度	Full length protein

描述

性质	Recombinant
来源	Escherichia coli

氨基酸序列

Accession	P49748
种属	Human
序列	**MGSSHHHHHHSSGLVPRGSHM**AGGAAQLALDKSDSHPSDALTRKKPAKAE SKSFAVGMFKGQLTTDQVFPYPSVLNEEQTQFLKELVEPVSRFFEEVNDP AKNDALEMVEETTWQGLKELGAFGLQVPSELGGVGLCNTQYARLVEIVGM HDLGVGITLGAHQSIGFKGILLFGTKAQKEKYLPKLASGETVAAFCLTEP SSGSDAASIRTSAVPSPCGKYYTLNGSKLWISNGGLADIFTVFAKTPVTD PATGAVKEKITAFVVERGFGGITHGPPEKKMGIKASNTAEVFFDGVRVPS ENVLGEVGSGFKVAMHILNNGRFGMAAALAGTMRGIIAKAVDHATNRTQF GEKIHNFGLIQEKLARMVMLQYVTESMAYMVSANMDQGATDFQIEAAISK IFGSEAAWKVTDECIQIMGGMGFMKEPGVERVLRDLRIFRIFEGTNDILR LFVALQGCMDKGKELSGLGSALKNPFGNAGLLLGEAGKQLRRRAGLGSGL SLSGLVHPELSRSGELAVRALEQFATVVEAKLIKHKKGIVNEQFLLQRLA DGAIDLYAMVVVLSRASRSLSEGHPTAQHEKMLCDTWCIEAAARIREGMA ALQSDPWQQELYRNFKSISKALVERGGVVTSNPLGF
分子量	69 kDa including tags
氨基酸	41 to 655
标签	His tag N-Terminus

技术指标

Our Abpromise guarantee covers the use of **ab98234** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

应用	SDS-PAGE

纯度	> 90 % SDS-PAGE. ab98234 was purified using conventional chromatography techniques.
形式	Liquid

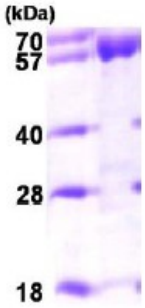
制备和贮存

稳定性和存储	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle. Preservative: None Constituents: 10% Glycerol, 0.1M Sodium chloride, 20mM Tris HCl, 1mM EDTA, 1mM DTT, pH 8.0

常规信息

功能	Active toward esters of long-chain and very long chain fatty acids such as palmitoyl-CoA, mysritoyl-CoA and stearoyl-CoA. Can accomodate substrate acyl chain lengths as long as 24 carbons, but shows little activity for substrates of less than 12 carbons.
通路	Lipid metabolism; mitochondrial fatty acid beta-oxidation.
疾病相关	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.
序列相似性	Belongs to the acyl-CoA dehydrogenase family.
细胞定位	Mitochondrion inner membrane.

图片



SDS-PAGE - ACADVL protein (ab98234)

15% SDS-PAGE analysis of 3µg ab98234

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