

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	<a href="#">P49748</a>
种属	Human
序列	<b>MGSSHHHHHSSGLVPRGSHMAGGAAQLALDKSDSHPSDALTRKKPAKAE</b> SKSFAVGMFKGQLTTDQVFPYPSVLNNEEQTQFLKELVEPVSRFFEEVNDP AKNDALMVEETTWQGLKELGAFGLQVPSELGGVGLCNTQYARLVEIVGM HDLGVGITLGAHQSIGFKGILLFGTKAQKEKYLPKLASGETVAAFCLTEP SSGSDAASIRTSAPVSPCGKYYTLNGSKLWISNGGLADIFTVFAKTPVTD PATGAVKEKITAFVVERGFGGITHGPPEKKMGIKASNTAEVFFDGVRVPS ENVLGEVSGFKVAMHILNNGRFGMAAALAGTMRGIIAKAVDHATNRTQF GEKIHNFGLIQEKLARMVMLQYVTESMAYMVSANMDQGATDFQIEAAISK IFGSEAAWKVTDECIQIMGGMGFMKEPGVERVLRDLRIFRIFEGTNDILR LFVALQGCMDKGKELSGLSALKNPFNAGLLLGEAGKQLRRRAGLGSGL SLSGLVHPELSRSGELAVRALEQFATVVEAKLIKHKKGIVNEQFLLQRLA DGAIDLAMVVVLSRASRSLSEGHPTAQHEKMLCDTWCIEAAARIREGMA ALQSDPWQELYRNFKSISKALVERGGVVTSNPLGF
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
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**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.

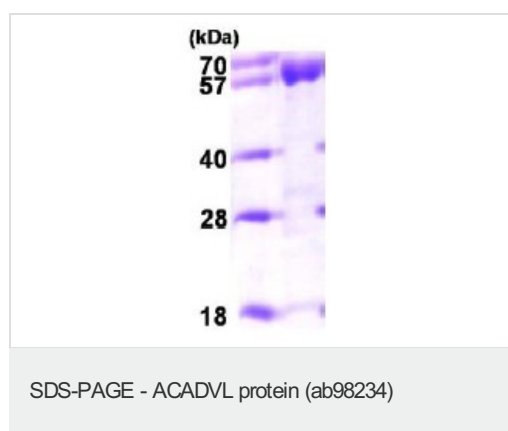
**通路** Lipid metabolism; mitochondrial fatty acid beta-oxidation.

**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



15% SDS-PAGE analysis of 3µg ab98234

**Please note:** All products are "FOR RESEARCH USE ONLY AND ARE NOT INTENDED FOR DIAGNOSTIC OR THERAPEUTIC USE"

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