abcam

Product datasheet

Recombinant Human ACADVL/VLCAD protein ab98234

1 Image

Description

Product name Recombinant Human ACADVL/VLCAD protein

Purity > 90 % SDS-PAGE.

ab98234 was purified using conventional chromatography techniques.

Expression system Escherichia coli

Accession P49748

Protein length Full length protein

Animal free No

Nature Recombinant

Species Human

Sequence MGSSHHHHHHSSGLVPRGSHMAGGAAQLALDKSDSHP

SDALTRKKPAKAE

SKSFAVGMFKGQLTTDQVFPYPSVLNEEQTQFLKELVEP

VSRFFEEVNDP

AKNDALEMVEETTWQGLKELGAFGLQVPSELGGVGLCN

TQYARLVEIVGM

HDLGVGITLGAHQSIGFKGILLFGTKAQKEKYLPKLASGET

VAAFCLTEP

SSGSDAASIRTSAVPSPCGKYYTLNGSKLWISNGGLADIFT

VFAKTPVTD

PATGAVKEKITAFVVERGFGGITHGPPEKKMGIKASNTAEV

FFDGVRVPS

ENVLGEVGSGFKVAMHILNNGRFGMAAALAGTMRGIIAKA

VDHATNRTQF

GEKIHNFGLIQEKLARMVMLQYVTESMAYMVSANMDQGA

TDFQIEAAISK

IFGSEAAWKVTDECIQIMGGMGFMKEPGVERVLRDLRIFRI

FEGTNDILR

LFVALQGCMDKGKELSGLGSALKNPFGNAGLLLGEAGKQ

LRRRAGLGSGL

SLSGLVHPELSRSGELAVRALEQFATVVEAKLIKHKKGIVN

EQFLLQRLA

DGAIDLYAMVVVLSRASRSLSEGHPTAQHEKMLCDTWCI

EAAARIREGMA

ALQSDPWQQELYRNFKSISKALVERGGVVTSNPLGF

1

Predicted molecular weight 69 kDa including tags

Amino acids 41 to 655

Tags His tag N-Terminus

Specifications

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.

Applications SDS-PAGE

Form Liquid

Additional notes Previously labelled as ACADVL.

Preparation and Storage

Stability and Storage 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.

pH: 8.00

Constituents: 0.0154% DTT, 0.316% Tris HCl, 0.0292% EDTA, 10% Glycerol (glycerin,

glycerine), 0.58% Sodium chloride

General Info

Function 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.

Pathway Lipid metabolism; mitochondrial fatty acid beta-oxidation.

Involvement in 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.

Sequence similarities Belongs to the acyl-CoA dehydrogenase family.

Cellular localization Mitochondrion inner membrane.

Images



15% SDS-PAGE analysis of 3µg ab98234

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