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Recombinant human ACADSB protein

Catalog Number: ATGP1628

PRODUCT INFORMATION

Expression system

E.coli

Domain

34-432aa

UniProt No.

P45954

NCBI Accession No.

NP 001600

Alternative Names

Short/branched chain specific acyl-CoA dehydrogenase mitochondrial, Short/branched chain specific acyl-CoA dehydrogenase, mitochondrial, 2-MEBCAD, ACAD7, SBCAD

PRODUCT SPECIFICATION

Molecular Weight

46.4 kDa (424aa)

Concentration

0.5mg/ml (determined by Bradford assay)

Formulation

Liquid in. 20mM Tris-HCl buffer (pH 8.0) containing 0.1M NaCl, 10% glycerol,1mM DTT

Purity

> 90% by SDS-PAGE

Tag

His-Tag

Application

SDS-PAGE

Storage Condition

Can be stored at +2C to +8C for 1 week. For long term storage, aliquot and store at -20C to -80C. Avoid repeated freezing and thawing cycles.

BACKGROUND

Description

Short/branched chain specific acyl-CoA dehydrogenase (ACADSB), is a member of the acyl-CoA dehydrogenase family of enzymes that catalyze the dehydrogenation of acyl-CoA derivatives in the metabolism of fatty acids or branch chained amino acids. ACADSB catalyzes the degradation of L-isoleucine and has the highest affinity for (s) -2-methylbutyryl-CoA, isobutyryl-CoA and 2-methylhexanoyl-CoA as substrates. Defects in ACADSB are the cause of short/branched-chain acyl-CoA dehydrogenase deficiency (SBCADD), an autosomal recessive disorder



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characterized by an increase of 2-methylbutyrylglycine and 2-methylbutyrylcarnitine in blood and urine. Recombinant human ACADSB protein, fused to His-tag at N-terminus, was expressed in E. coli and purified by using conventional chromatography techniques.

Amino acid Sequence

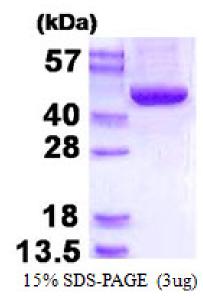
MGSSHHHHHH SSGLVPRGSH MGSHMKSSQS EALLNITNNG IHFAPLQTFT DEEMMIKSSV KKFAQEQIAP LVSTMDENSK MEKSVIQGLF QQGLMGIEVD PEYGGTGASF LSTVLVIEEL AKVDASVAVF CEIQNTLINT LIRKHGTEEQ KATYLPQLTT EKVGSFCLSE AGAGSDSFAL KTRADKEGDY YVLNGSKMWI SSAEHAGLFL VMANVDPTIG YKGITSFLVD RDTPGLHIGK PENKLGLRAS STCPLTFENV KVPEANILGQ IGHGYKYAIG SLNEGRIGIA AQMLGLAQGC FDYTIPYIKE RIQFGKRLFD FQGLQHQVAH VATQLEAARL LTYNAARLLE AGKPFIKEAS MAKYYASEIA GQTTSKCIEW MGGVGYTKDY PVEKYFRDAK IGTIYEGASN IQLNTIAKHI DAEY

General References

Arden K C., et al. (1995) Genomics. 25: 743-745. Korman S H., et al. (2005) Clin Chem. 51: 610-617.

DATA

SDS-PAGE



3ug by SDS-PAGE under reducing condition and visualized by coomassie blue stain.

