

ACADS Human

Acyl-Coenzyme A Dehydrogenase C-2 to C-3 Human Recombinant
ENZ0014

Product Overview

Name ACADS Human

Description

Acyl-Coenzyme A Dehydrogenase C-2 to C-3 Human Recombinant

Accession (Primary) [P16219](#)

Synonyms

Short/branched chain specific acyl-CoA dehydrogenase mitochondrial, SBCAD, 2-methyl branched chain acyl-CoA dehydrogenase, 2-MEBCAD, 2-methylbutyryl-coenzyme A dehydrogenase, 2-methylbutyryl-CoA dehydrogenase, ACADSB, ACAD7, SBCAD, 2-MEBCAD.

Introduction

Short/branched chain specific acyl-CoA dehydrogenase (ACADSB) belongs to the acyl-CoA dehydrogenase family of enzymes which 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 while having the highest affinity for (s)-2-methylbutyryl-CoA, isobutyryl-CoA and 2-methylhexanoyl-CoA as substrates. ACADSB may use valproyl-CoA as substrate. ACADSB gene defects cause the short/branched-chain acyl-CoA dehydrogenase deficiency (SBCADD), which is an autosomal recessive disorder characterized by an increase of 2-methylbutyrylglycine and 2-methylbutyrylcarnitine in blood and urine.

Source

Escherichia Coli.

Physical Appearance

Sterile Filtered clear solution.

Formulation

The ACADSB solution (0.5mg/ml) contains 20mM Tris-HCl buffer (pH 8.0), 0.1M NaCl, 10% glycerol and 1mM DTT.

Stability

Store at 4°C if entire vial will be used within 2-4 weeks. Store, frozen at -20°C for longer periods of time. For long term storage it is recommended to add a carrier protein (0.1% HSA or BSA). Avoid multiple freeze-thaw cycles.

Purity

Greater than 90.0% as determined by SDS-PAGE.

Amino acid sequence

MGSSHHHHHH SSGLVPRGSH MGS HM KSSQS EALLNITNNG IHFAPLQTFT DEEMMIKSSV KKFAQEIQIAP
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 FQGLQHVAH VATQLEAARL LTYNAAARLLE
AGKPFKEAS MAKYYASEIA GQTTSKCI EW MGGVGYTKDY PVEKYFRDAK IGTIYEGASN IQLNTIAKHI DAEY.

Precautions

ACADS Human is for research use only and not for use in diagnostic or therapeutic procedures.

Target Information: ([P16219](#))