
Product Data Sheet

Product Name: ACADSB Human
 Cat. No.: GP21332
 Batch No.: 1

Product Data

Purity	>98%	Source	Escherichia Coli.
Physical Appearance	solid	Shipping Condition	Shipped with Ice Packs.
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.		
Amino Acid Sequence	<p>MGSSHHHHH SSGLVPRGSH MGSHMKSSQS EALLNITNNG IHFAPLQFTT 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 FDYTIPIYIKE RIQFGKRLFD FQGLQHQVAH VATQLEAARL LTYNAARLLE AGKPFIKEAS MAKYYASEIA GQTTSKCI EW MGGVGYTKDY PVEKYFRDAK IGTIYEGASN IQLNTIAKHI DAEY.</p>		
Formulation	The ACADSB solution (0.5mg/ml) contains 20mM Tris-HCl buffer (pH 8.0), 0.1M NaCl, 10% glycerol and 1mM DTT.		

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-

Caution: Product has not been fully validated for medical applications. For research use only.

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methylbutyrylcarnitine in blood and urine.

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.

Background

ACADSB Human Recombinant produced in E.coli is a single, non-glycosylated polypeptide chain containing 424 amino acids (34-432) and having a molecular mass of 46.4kDa. ACADSB is fused to a 25 amino acid His-tag at N-terminus & purified by proprietary chromatographic techniques.

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