
Product Data Sheet

Product Name: BCKDHA Human
 Cat. No.: GP21435
 Batch No.: 1

Product Data

Purity	>98%	Source	Escherichia Coli.
Physical Appearance	solid	Shipping Condition	Shipped with Ice Packs.
Synonyms	2-oxoisovalerate dehydrogenase subunit alpha mitochondrial; Branched-chain alpha-keto acid dehydrogenase E1 component alpha chain; BCKDE1A; BCKDH E1-alpha; BCKDHA; MSU; MSUD1; OVD1A; FLJ45695.		
Amino Acid Sequence	<p>MGSSHHHHHH SGLVPRGSH MSSLDDKPQF PGASAEFIDK LEFIQPNVIS GIPIYRVMDR QGQIINPSED PHLPKKVLK LYKSMTLLNT MDRILYESQR QGRISFYMTN YGEEGTHVGS AAALDNTDLV FGQYREAGVL MYRDYPLELF MAQCYGNISD LGKGRQMPVH YGCKERHFVT ISSPLATQIP QAVGAAYAANK RANANRVVIC YFGEGAASEG DAHAGFNFAA TLECPIIFFC RNNGYAISTP TSEQYRGDGI AARGPGYGIM SIRVDGNDVF AVYNATKEAR RRAVAENQPF LIEAMTYRIG HHSTSDDSSA YRSVDEVNYW DKQDHPISRL RHYLLSQGWW DEEQEKAWRK QSRKRVMEAF EQAERKPKPN PNLFSVDVYQ EMPAQLRKQQ ESLARHLQTY GEHYPLDHF D K.</p>		
Formulation	The BCKDHA solution (0.25 mg/ml) contains 20mM Tris-HCl buffer (pH 8.0), 5mM DTT, 30% glycerol and 0.2M NaCl.		

Introduction

Branched-chain α -keto acid dehydrogenase E1 component α chain (BCKDHA) is a member of the BCKDHA family. The BCKD (branched-chain α -keto acid dehydrogenase) complex is an inner mitochondrial enzyme complex which catalyzes the second major step in the catabolism of the branched-chain amino acids leucine, isoleucine, and valine. This complex consists of 3 catalytic components: a heterotetrameric (α_2 - β_2) branched-chain α -keto acid decarboxylase (E1), a dihydrolipoyl transacylase (E2), and a dihydrolipoamide dehydrogenase (E3). Mutations in the BCKDHA gene result in maple syrup urine disease, type IA.

Stability

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

Tel: (909) 407-4943 Fax: (626) 353-8530 E-mail: tech@glpbio.com

Address: 10292 Central Ave. #205, Montclair, CA, USA

Product Data Sheet

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

BCKDHA Human Recombinant fused with a21 amino acid His tag at N-terminus produced in E.Coli is a single, non-glycosylated, polypeptide chain containing 421 amino acids (46-445 a.a.) and having a molecular mass of 47.8kDa. The BCKDHA is purified by proprietary chromatographic techniques.

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

Tel: (909) 407-4943 Fax: (626) 353-8530 E-mail: tech@glpbio.com

Address: 10292 Central Ave. #205, Montclair, CA, USA