

## BCKDHA Human

**Description:** BCKDHA Human Recombinant fused with a 21 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.

**Catalog #:** ENPS-097

For research use only.

**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.

**Source:** Escherichia Coli.

**Physical Appearance:** Sterile Filtered colorless solution.

**Amino Acid Sequence:** MGSSHHHHHH SSGLVPRGSH MSSLDDKPQF PGASAEFIDK  
LEFIQPNVIS GIPIYRVMDR QGQIINPSED PHLPEKEVLK LYKSMTLLNT MDRILYESQR  
QGRISFYMTN YGEEGTHVGS AAALDNTDLV FGQYREAGVL MYRDYPLELF MAQCYGNISD  
LGKGRQMPVH YGCKERHFVT ISSPLATQIP QAVGAAYAAK RANANRVVIC YFGEAASEG  
DAHAGFNFAA TL

**Purity:** Greater than 80.0% as determined by SDS-PAGE.

### Formulation:

The BCKDHA solution (0.25 mg/ml) contains 20mM Tris-HCl buffer (pH 8.0), 5mM DTT, 30% glycerol and 0.2M NaCl.

### 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.

### Usage:

NeoBiolab's products are furnished for LABORATORY RESEARCH USE ONLY. The product may not be used as drugs, agricultural or pesticidal products, food additives or household chemicals.

### Introduction:

Branched-chain -keto acid dehydrogenase E1 component chain (BCKDHA) is a member of the BCKDHA family. The BCKD (branched-chain alpha-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 (alpha2-beta2) branched-chain alpha-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.

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