

## 32-2179: BCKDHA Recombinant Protein

**Alternative Name :** 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.

### Description

Source : Escherichia Coli. 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. Branched-chain  $\alpha$ -keto acid dehydrogenase E1 component  $\alpha$  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 (alpha<sub>2</sub>-beta<sub>2</sub>) 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.

### Product Info

**Amount :** 5  $\mu$ g  
**Purification :** Greater than 80.0% as determined by SDS-PAGE.  
**Content :** The BCKDHA solution (0.25 mg/ml) contains 20mM Tris-HCl buffer (pH 8.0), 5mM DTT, 30% glycerol and 0.2M NaCl.  
**Storage condition :** 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.  
**Amino Acid :** MGSSHHHHHH SGLVPRGSH MSSLDDKPQF PGASAEFIDK LEFIQPNVIS GIPIYRVM DR QGQIINPSED PHLPKKEKVLK LYKSMTLLNT MDRILYESQR QGRISFYMTN YGEEGTHVGS AAALDNTDLV FGQYREAGVL MYRDYPLELF MAQCYGNISD LGKGRQMPVH YGCKERHFVT ISSPLATQIP QAVGAAYA AK RANANRVVIC YFGEGAASEG DAHAGFNFAA TLECPIIFFC RNNGYAISTP TSEQYRGDGI AARGPGYGIM SIRVDGNDVF AVYNATKEAR RRAVAENQPF LIEAMTYRIG HHSTSDDSSA YRSVDEVNYW DKQDHPISRL RHYLLSQGWV DEEQEKAWRK QSRRKVMEAF EQAERKPKPN PNLLFSDVYQ EMPAQLRKQQ ESLARHLQTY GEHYPLDHF D K.

