## 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 E1alpha,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 47.8 kDa . The BCKDHA is purified by proprietary chromatographic techniques. Branched-chain Alpha-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.

## Product Info

| Amount : | $5 \mu \mathrm{~g}$ |
| :---: | :---: |
| Purification : | Greater than $80.0 \%$ as determined by SDS-PAGE. |
| Content : | The BCKDHA solution ( $0.25 \mathrm{mg} / \mathrm{ml}$ ) contains 20 mM Tris-HCl buffer ( pH 8.0 ), 5 mM DTT, $30 \%$ glycerol and 0.2 M NaCl . |
| Storage condition : | Store at $4^{\circ} \mathrm{C}$ if entire vial will be used within $2-4$ weeks. Store, frozen at $-20^{\circ} \mathrm{C}$ 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 SSGLVPRGSH MSSLDDKPQF PGASAEFIDK LEFIQPNVIS GIPIYRVMDR QGQIINPSED |
|  | PHLPKEKVLK LYKSMTLLNT MDRILYESQR QGRISFYMTN YGEEGTHVGS AAALDNTDLV FGQYREAGVL |
|  | MYRDYPLELF MAQCYGNISD LGKGRQMPVH YGCKERHFVT ISSPLATQIP QAVGAAYAAK RANANRVVIC |
|  | YFGEGAASEG DAHAGFNFAA TLECPIIFFC RNNGYAISTP TSEQYRGDGI AARGPGYGIM SIRVDGNDVF |
|  | AVYNATKEAR RRAVAENQPF LIEAMTYRIG HHSTSDDSSA YRSVDEVNYW DKQDHPISRL |
|  | RHYLLSQGWW DEEQEKAWRK QSRRKVMEAF EQAERKPKPN PNLLFSDVYQ EMPAQLRKQQ |
|  | ESLARHLQTY GEHYPLDHFD K. |



