

## Recombinant Human BCKDHA, His-tagged

Cat. No. BCKDHA-27439TH Lot. No. (See product label)

### SPECIFICATION

<b>Product Overview</b>	Recombinant fragment, corresponding to amino acids 51-445 of Human BCKDHA with N terminal His tag; Predicted MWt 46 kDa.
<b>Species</b>	Human
<b>Source</b>	E.coli
<b>ProteinLength</b>	51-445 a.a.
<b>Description</b>	The branched-chain alpha-keto acid (BCAA) dehydrogenase (BCKD) complex is an inter mitochondrial enzyme complex that catalyzes the second major step in the catabolism of the branched-chain amino acids leucine, isoleucine, and valine. The BCKD complex consists of three catalytic components: a heterotetrameric (alpha2-beta2) branched-chain alpha-keto acid decarboxylase (E1), a dihydrolipoyl transacylase (E2), and a dihydrolipoamide dehydrogenase (E3). This gene encodes the alpha subunit of the decarboxylase (E1) component. Mutations in this gene result in maple syrup urine disease, type IA. Multiple transcript variants encoding different isoforms have been found for this gene.
<b>Conjugation</b>	HIS
<b>Form</b>	Lyophilised:Reconstitute with 69 µl aqua dest.
<b>Storage buffer</b>	Preservative: None Constituents: 0.5% Trehalose, 6M Urea, 100mM Sodium phosphate, 10mM Sodium chloride, pH 4.5

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<b>Storage</b>	Shipped at 4°C. Upon delivery aliquot and store at -80oC. Avoid freeze / thaw cycles.
<b>Sequences of amino acids</b>	KPQFPGASAEFIDKLEFIQPNVISGIPIYRVMDRQGQIIN PSEDPHLPKEKVLKLYKSM TLLNTMDRILYESQRQGRI SFYMTNYGEEGTHVGSAAAALDNTDLVFGQYREAGVLM Y RDYPLELFMAQCYGNISDLGKGRQMPVHYGCKERHFVT ISSPLATQIPQAVGAAY AAKRANANRVVICYFGEGAASEG DAHAGFNFAATLECPPIFFCRNNGYAISTPTSEQ YRGD GIAARGPGYGIMSIRVDGNDVFAVYNATKEARRRAVAE NQPFLIEAMTYRIG HHSTSDSSAYRSVDEVNYWDKQD HPISRLRHYLSSQGWWDEEQEKAWRKQSR RKVMEAFEQAE RKPKNPNLLFSDVYQEMPAQLRKQQESLARHLQTYGE HYPLD HFDK
<b>Sequence Similarities</b>	Belongs to the BCKDHA family.
<b>GENE INFORMATION</b>	
<b>Gene Name</b>	<a href="#">BCKDHA branched chain keto acid dehydrogenase E1, alpha polypeptide [ Homo sapiens ]</a>
<b>Official Symbol</b>	<a href="#">BCKDHA</a>
<b>Synonyms</b>	BCKDHA; branched chain keto acid dehydrogenase E1, alpha polypeptide; 2 oxoisovalerate dehydrogenase (lipoamide) , branched chain keto acid dehydrogenase E1, alpha polypeptide (maple syrup urine disease) , OVD1A; 2-oxoisovalerate dehydrogenase subunit al
<b>Gene ID</b>	<a href="#">593</a>
<b>mRNA Refseq</b>	<a href="#">NM_000709</a>
<b>Protein Refseq</b>	<a href="#">NP_000700</a>

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<b>MIM</b>	608348
<b>Uniprot ID</b>	P12694
<b>Chromosome Location</b>	19q13.1-q13.2
<b>Pathway</b>	2-oxobutanoate degradation I, conserved biosystem; Branched-chain amino acid catabolism, organism-specific biosystem; Leucine degradation, leucine => acetoacetate + acetyl-CoA, organism-specific biosystem; Leucine degradation, leucine =>
<b>Function</b>	3-methyl-2-oxobutanoate dehydrogenase (2-methylpropanoyl-transferring) activity; alpha-ketoacid dehydrogenase activity; carboxy-lyase activity; metal ion binding; protein binding;

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