

Recombinant Human BCKDHA, His-tagged

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

SPECIFICATION

Product Overview	Recombinant fragment, corresponding to amino acids 51-445 of Human BCKDHA with N terminal His tag; Predicted MWt 46 kDa.
Species	Human
Source	E.coli
ProteinLength	51-445 a.a.
Description	The branched-chain alpha-keto acid (BCAA) dehydrogenase (BCKD) complex is an innter 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.
Conjugation	HIS
Form	Lyophilised:Reconstitute with 69 µl aqua dest.
Storage buffer	Preservative: NoneConstituents: 0.5% Trehalose, 6M Urea, 100mM Sodium phosphate, 10mM Sodium chloride, pH 4.5

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Storage	Shipped at 4°C. Upon delivery aliquot and store at -80oC. Avoid freeze / thaw cycles.
Sequences of amino acids	KPQFPGASAEFIDKLEFIQPNVISGIPYRVMDRQGQIIN PSEDPHLPKEKVLKLYKSM TLLNTMDRILYESQRQGRI SFYMTNYGEEGTHVGSAAAALDNTDLVFGQYREAGVLM Y RDYPLELFMAQCYGNISDLGKGRQMPVHYGCKERHFVT ISSPLATQIPQAVGAAY AAKRANANRVVICYFGEGAASEG DAHAGFNFAATLECPPIFFCRNNGYAISTPTSEQ YRGD GIAARGPGYGIMSIRVDGNDVFAVYNATKEARRRAVAE NQPFLIEAMTYRIG HHSTSDSSAYRSVDEVNYWDKQD HPISRLRHYLLSQGWWDEEQEKAWRKQSR RKMVMEAFEQAE RKPKPNPNLLFSDVYQEMPAQLRKQQESLARHLQTYGE HYPLD HFDK
Sequence Similarities	Belongs to the BCKDHA family.
GENE INFORMATION	
Gene Name	BCKDHA branched chain keto acid dehydrogenase E1, alpha polypeptide [Homo sapiens]
Official Symbol	BCKDHA
Synonyms	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
Gene ID	593
mRNA Refseq	NM_000709
Protein Refseq	NP_000700

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MIM	608348
Uniprot ID	P12694
Chromosome Location	19q13.1-q13.2
Pathway	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 =>
Function	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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