Recombinant Human BCKDHA, His-tagged

Cat. No. BCKDHA-27442TH **Lot. No.** (See product label)

SPECIFICATION

Product Overview	Recombinant full length mature Human BCKDHA with N terminal His tag; predicted MWt 47.8kDa; 421 amino acids inclusive of tag;
Species	Human
Source	E.coli
ProteinLength	400 amino acids
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
Molecular Weight	47.800kDa inclusive of tags
Form	Liquid

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Purity	by SDS-PAGE
Storage buffer	Preservative: NoneConstituents: 30% Glycerol, 5mM DTT, 200mM Sodium chloride, 20mM Tris HCl, pH 8
Storage	Store at +4°C short term (1-2 weeks). Aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
Sequences of amino acids	MGSSHHHHHH SSGLVPRGSH MSSLDDKPQF PGASAEFIDK LEFIQPNVIS GIPIYRVMDR QGQIINPSED PHLPKEKVLK LYKSMTLLNTMDRILYESQR QGRISFYMTN YGEEGTHVGS AAALDNTDLV FGQYREAGVL MYRDYPLELF MAQCYGNISD LGKGRQMPVH YGCKERHFVTISSPLATQIP QAVGAAYAAK RANANRVVIC YFGEGAASEG DAHAGFNFAA TLECPIIFFC RNNGYAISTP TSEQYRGDGI AARGPGYGIMSIRVDGNDVF AVYNATKEAR RRAVAENQPF LIEAMTYRIG HHSTSDDSSA YRSVDEVNYW DKQDHPISRL RHYLLSQGWW DEEQEKAWRKQSRRKVMEAF EQAERKPKPN PNLLFSDVYQ EMPAQLRKQQ ESLARHLQTY GEHYPLDHFD K
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

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	dehydrogenase subunit al
Gene ID	593
mRNA Refseq	NM_000709
Protein Refseq	NP_000700
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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