

Recombinant Full Length Human BCKDHA Protein, GST-tagged

Cat. No. BCKDHA-1630HF **Lot. No.** (See product label)

SPECIFICATION

Product Overview	Human BCKDHA full-length ORF (AAH07878.1, 1 a.a. - 445 a.a.) recombinant protein with GST-tag at N-terminal.
Species	Human
Source	In Vitro Cell Free System
ProteinLength	445 amino acids
Description	The branched-chain alpha-keto acid (BCAA) dehydrogenase (BCKD) complex is an ininter 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.
Form	50 mM Tris-HCl, 10 mM reduced Glutathione, pH=8.0 in the elution buffer.
Molecular Mass	74.69 kDa
AA Sequence	MAVAIAAARV WRLNRGLSQA ALLLLRQPGA RGLARSHPPR QQQQFSSLDD

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KPQFPGASAE FIDKLEFIQP NVISGIPIYR VMDRQGQIIN PSEDPHLPKE
 KVLKLYKSMT LLNTMDRILY ESQRQGRISF YMTNYGEEGT HVGSAALDN
 TDLVFGQYRE AGVLMYRDYP LELFMAQCYG NISDLGKGRQ MPVHYGCKER
 HFVTISSPLA TQIPQAVGAA YAAKRANANR VVICYFGEGA ASEGDAHAGF
 NFAATLECPI IFFCRNNGYA ISTPTSEQYR GDGIAARGPG YGIMSIRVDG
 NDVFAVYNAT KEARRRAVAE NQPFLIEAMT YRIGHHSTSD DSSAYRSVDE
 VNYWDKQDHP ISRLRHYLLS QGWWDEEQEK AWRKQSRRKV MEAFEQAERK
 PKPNPNLLFS DVYQEMPAQL RKQQESLARH LQTYGEHYPL DHFDK

Storage Store at -80 centigrade. Aliquot to avoid repeated freezing and thawing.

Storage Buffer 50 mM Tris-HCl, 10 mM reduced Glutathione, pH=8.0 in the elution buffer.

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 alpha, mitochondrial; maple syrup urine disease; MSU; BCKDH E1-alpha; 2-oxoisovalerate dehydrogenase (lipoamide); branched-chain alpha-keto acid dehydrogenase E1 component alpha chain; MSUD1; OVD1A; BCKDE1A; FLJ45695

Gene ID 593

mRNA Refseq NM_000709

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Protein Refseq	NP_000700
MIM	608348
UniProt ID	P12694

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