

# Recombinant Full Length Human BCKDHA Protein, GST-tagged

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

## SPECIFICATION

<b>Product Overview</b>	Human BCKDHA full-length ORF ( AAH07878.1, 1 a.a. - 445 a.a.) recombinant protein with GST-tag at N-terminal.
<b>Species</b>	Human
<b>Source</b>	In Vitro Cell Free System
<b>ProteinLength</b>	445 amino acids
<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>Form</b>	50 mM Tris-HCl, 10 mM reduced Glutathione, pH=8.0 in the elution buffer.
<b>Molecular Mass</b>	74.69 kDa
<b>AA Sequence</b>	MAVAIAAARV WRLNRGLSQA ALLLLRQPGA RGLARSHPPR QQQQFSSLDD

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KPQFPGASAE FIDKLEFIQP NVISGIPIYR VMDRQQQIIN PSEDPHLPKE  
 KVLKLYKSMT LLNTMDRILY ESQRQGRISF YMTNYGEEGT HVGSAALDN  
 TDLVFGQYRE AGVLMYRDYP LELFMAQCYG NISDLGKGRQ MPVHYGCKER  
 HFVTISSPLA TQIPQAVGAA YAAKRANANR VVICYFGEGA ASEGDAHAGF  
 NFAATLECP IFFCRNNGYA ISTPTSEQYR GDGIAARGPG YGIMSIRVDG  
 NDVFAVYNAT KEARRRAVAE NQPFLIEAMT YRIGHHSTSD DSSAYRSVDE  
 VNYWDKQDHP ISRLRHILLS QGWWDDEEQEK AWRKQSRRKV MEAFEQAERK  
 PKPNPNULLFS 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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