

Recombinant Human BCKDHA Protein, GST-tagged

Cat. No. BCKDHA-135H **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	Wheat Germ
Description	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.
Form	50 mM Tris-HCl, 10 mM reduced Glutathione, pH=8.0 in the elution buffer.
Molecular Mass	74.69 kDa
AA Sequence	MAVAIAAARVWRLNRGLSQAALLLLRQPGARGLARSHPPRQQQQFSSLDDKPQFP GASAEFIDKLEFIQPNVISGIPYRVMDRQGGQIINPSEDPHLPKEKVLKLYKSMTLLNT MDRILYESQRQGRISFYMTNYGEEGTHVGSAAALDNTDLVFGQYREAGVLMYRDYP LELFMAQCYGNISDLGKGRQMPVHYGCKERHFVTISSPLATQIPQAVGAAYAAKRA

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NANRVVICYFGEGAASEGDAHAGFNFAATLECPHFFCRNNGYAISTPTSEQYRGDGI
AARGPGYGIMSIRVDGNDVFAVYNATKEARRRAVAENQPFLIEAMTYRIGHHSTSD
SSAYRSVDEVNYWDKQDHPISRLRHYLLSQGWDEEQEKAWRKQSRRKVMEAFE
QAERKPKPNPNLLFSDVYQEMPAQLRKQQESLARHLQTYGEHYPLDHFDK

Notes	Best use within three months from the date of receipt of this protein.
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
Protein Refseq	NP_000700

MIM	608348
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UniProt ID	P12694
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