

Recombinant Human BCKDHB, GST-tagged

Cat. No. BCKDHB-10173H **Lot. No.** (See product label)

SPECIFICATION

Product Overview	Recombinant Human BCKDHB protein, fused to GST-tag, was expressed in E.coli and purified by GSH-sepharose.
Species	Human
Source	E.coli
ProteinLength	41-392a.a.
Description	Branched-chain keto acid dehydrogenase is a multienzyme complex associated with the inner membrane of mitochondria, and functions in the catabolism of branched-chain amino acids. The complex consists of multiple copies of 3 components: branched-chain alpha-keto acid decarboxylase (E1), lipoamide acyltransferase (E2) and lipoamide dehydrogenase (E3). This gene encodes the E1 beta subunit, and mutations therein have been associated with maple syrup urine disease (MSUD), type 1B, a disease characterized by a maple syrup odor to the urine in addition to mental and physical retardation, and feeding problems. Alternative splicing at this locus results in transcript variants with different 3 non-coding regions, but encoding the same isoform.
Storage	The protein is stored in PBS buffer at -20°C. Avoid repeated freezing and thawing cycles.
Storage Buffer	1M PBS (58mM Na ₂ HPO ₄ , 17mM NaH ₂ PO ₄ , 68mM NaCl, pH8.) added with 100mM GSH and 1% Triton X-100, 15% glycerol.

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GENE INFORMATION

Gene Name	BCKDHB branched chain keto acid dehydrogenase E1, beta polypeptide [Homo sapiens]
Official Symbol	BCKDHB
Synonyms	BCKDHB; branched chain keto acid dehydrogenase E1, beta polypeptide; 2-oxoisovalerate dehydrogenase subunit beta, mitochondrial; maple syrup urine disease; BCKDE1B; BCKDH E1-beta; 2-oxoisovalerate dehydrogenase beta subunit; E1b-beta subunit of the branched-chain complex; branched chain alpha-ketoacid dehydrogenase E1-beta subunit; branched-chain alpha-keto acid dehydrogenase E1 component beta chain; E1B; dJ279A18.1; FLJ17880;
Gene ID	594
mRNA Refseq	NM_000056
Protein Refseq	NP_000047
MIM	248611
UniProt ID	P21953
Chromosome Location	6q14.1
Pathway	2-oxobutanoate degradation I, organism-specific biosystem; 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 => acetoacetate +

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acetyl-CoA, conserved biosystem;

Function

3-methyl-2-oxobutanoate dehydrogenase (2-methylpropanoyl-transferring) activity;
alpha-ketoacid dehydrogenase activity; carboxy-lyase activity; protein binding; protein
complex binding;

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