

Recombinant Full Length Human BCKDHB Protein, GST-tagged

Cat. No. BCKDHB-1612HF **Lot. No.** (See product label)

SPECIFICATION

Product Overview	Human BCKDHB full-length ORF (AAH40139, 1 a.a. - 392 a.a.) recombinant protein with GST-tag at N-terminal.
Species	Human
Source	In Vitro Cell Free System
ProteinLength	392 amino acids
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.
Form	50 mM Tris-HCl, 10 mM reduced Glutathione, pH=8.0 in the elution buffer.
Molecular Mass	68.86 kDa

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AA Sequence

MAVVAAAAGW LLRLRAAGAE GHWRR LPGAG LARGFLHPAA TVEDAAQRRQ
 VAHFTFQDPD EPREYGQTQK MNLFQSVTSA LDNSLAKDPT AVIFGEDVAF
 GGVFRCTVGL RDKY GKDRVF NTPLCEQGIV GFGIGIAVTG ATAIAEIQFA
 DYIFPAFDQI VNEAAKYRYR SGDLFNC GSL TIRSPWGCVG HGALYHSQSP
 EAFFAHCPGI KVVIPRSPFQ AKRLLLSCIE DKNPCIFFEP KILYRAAAEE
 VPIEPYNIPL SQA EVIQEGS DVTLVAWGTQ VHVIREVASM AKEKLGVSCE
 VIDLRTIIPW DVDTICKSVI KTGRLLISHE APLTGGF ASE ISSTVQEECF
 LNLEAPISRV CGYDTPFPHI FEPFYIPDKW KCYDALRKMI NY

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

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

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Protein Refseq	NP_000047
MIM	248611
UniProt ID	P21953

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