

Recombinant Human DBT protein, MYC/DDK-tagged

Cat. No. DBT-7061H Lot. No. (See product label)

SPECIFICATION

Product Overview	Recombinant Human DBT fused with MYC/DDK tag at C-terminal was expressed in HEK293.
Species	Human
Source	HEK293
Description	The branched-chain alpha-keto acid dehydrogenase complex (BCKD) is an inner-mitochondrial enzyme complex involved in the breakdown of the branched-chain amino acids isoleucine, leucine, and valine. The BCKD complex is thought to be composed of a core of 24 transacylase (E2) subunits, and associated decarboxylase (E1), dehydrogenase (E3), and regulatory subunits. This gene encodes the transacylase (E2) subunit. Mutations in this gene result in maple syrup urine disease, type 2. Alternatively spliced transcript variants have been described, but their biological validity has not been determined.
Form	25 mM Tris.HCl, pH 7.3, 100 mM glycine, 10% glycerol.
Molecular Mass	46.4 kDa
Purity	> 80% as determined by SDS-PAGE and Coomassie blue staining
Notes	Recombinant protein was captured through anti-DDK affinity column followed by conventional chromatography steps.

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Concentration >50 ug/mL as determined by microplate BCA method

GENE INFORMATION

Gene Name DBT dihydrolipoamide branched chain transacylase E2 [Homo sapiens]

Official Symbol DBT

Synonyms

DBT; dihydrolipoamide branched chain transacylase E2; dihydrolipoamide branched chain transacylase (E2 component of branched chain keto acid dehydrogenase complex; maple syrup urine disease); lipoamide acyltransferase component of branched-chain alpha-keto acid dehydrogenase complex, mitochondrial; BCKADE2; BCKAD-E2; BCKAD E2 subunit; dihydrolipoyl transacylase; branched chain acyltransferase, E2 component; dihydrolipoyllysine-residue (2-methylpropanoyl)transferase; branched-chain alpha-keto acid dehydrogenase complex component E2; E2 component of branched chain alpha-keto acid dehydrogenase complex; mitochondrial branched chain alpha-keto acid dehydrogenase transacylase subunit (E2b); dihydrolipoamide acetyltransferase component of branched-chain alpha-keto acid dehydrogenase complex; lipoamide acyltransferase component of mitochondrial branched-chain alpha-keto acid dehydrogenase complex; E2; E2B; BCATE2; MGC9061;

Gene ID 1629

mRNA Refseq NM_001918

Protein Refseq NP_001909

MIM 248610

UniProt ID P11182

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Chromosome Location	1p31
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 + acetyl-CoA, conserved biosystem;
Function	cofactor binding; dihydrolipoyllysine-residue (2-methylpropanoyl)transferase activity; transferase activity, transferring acyl groups;

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