

Recombinant Human DBT protein, His-tagged

Cat. No. DBT-4930H Lot. No. (See product label)

SPECIFICATION

Product Overview	Recombinant Human DBT fused with hexa-histidine purification tag was expressed in Insect cells.
Species	Human
Source	Insect Cells
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	DLST (0.91mg/ml) is supplied in 20mM HEPES buffer pH-8.0, 200mM NaCl and 20% glycerol.
Molecular Mass	47,321 Dalton
Purity	Greater than 75% as determined by SDS-PAGE.
Applications	Western blot with anti-M2-Antigen autoantibody-positive patient sera or monoclonal anti-hexa-His-tag antibody. Coating concentration; 0.4-0.8 µg/ml (depending on the

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type of ELISA plate and coating buffer). Suitable for biotinylation and iodination. Standard ELISA test (checkerboard analysis of positive/negative sera panels); immunodot test with positive/negative sera panels. 0.4-0.8 g/ml (depending on the type of ELISA plate and coating buffer). Suitable for biotinylation and iodination.

Storage

Store at 4 centigrade if entire vial will be used within 2-4 weeks. Store, frozen at -20 centigrade for longer periods of time. Avoid multiple freeze-thaw cycles.

GENE INFORMATION

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

Official Symbol DBT

Synonyms DBT; BCKADE2; BCKAD-E2; BCKAD E2 subunit; dihydrolipoyl transacylase; E2; E2B; BCATE2; MGC9061;

Gene ID 1629

mRNA Refseq NM_001918

Protein Refseq NP_001909

MIM 248610

UniProt ID P11182

Chromosome Location 1p31

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Pathway	2-oxobutanoate degradation I, organism-specific biosystem; 2-oxobutanoate degradation I, conserved biosystem; Branched-chain amino acid catabolism, organism-specific biosystem
Function	cofactor binding; dihydrolipoyllysine-residue (2-methylpropanoyl)transferase activity; transferase activity, transferring acyl groups;

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