



Anti-BCKDHB monoclonal antibody (DCABH-10723)

This product is for research use only and is not intended for diagnostic use.

PRODUCT INFORMATION

Antigen 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.
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Immunogen	A synthetic peptide of human BCKDHB is used for rabbit immunization.
Isotype	IgG
Source/Host	Rabbit
Species Reactivity	Human
Purification	Protein A
Conjugate	Unconjugated
Applications	Western Blot (Transfected lysate); ELISA
Buffer	In 1x PBS, pH 7.4
Preservative	None
Storage	Store at -20°C or lower. Aliquot to avoid repeated freezing and thawing.

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;
Entrez Gene ID	594
Protein Refseq	NP_000047
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; acetoacetate + 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;