



Anti-BCKDHA monoclonal antibody (DCABH-10722)

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

PRODUCT INFORMATION

Antigen Description	The branched-chain alpha-keto acid (BCAA) dehydrogenase (BCKD) complex is an inner mitochondrial enzyme complex that catalyzes the second major step in the catabolism of the branched-chain amino acids leucine, isoleucine, and valine. The BCKD complex consists of three catalytic components: a heterotetrameric (alpha2-beta2) branched-chain alpha-keto acid decarboxylase (E1), a dihydrolipoyl transacylase (E2), and a dihydrolipoamide dehydrogenase (E3). This gene encodes the alpha subunit of the decarboxylase (E1) component. Mutations in this gene result in maple syrup urine disease, type IA. Multiple transcript variants encoding different isoforms have been found for this gene
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Immunogen	A synthetic peptide of human BCKDHA 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	BCKDHA branched chain keto acid dehydrogenase E1, alpha polypeptide [Homo sapiens]
Official Symbol	BCKDHA
Synonyms	BCKDHA; branched chain keto acid dehydrogenase E1, alpha polypeptide; 2 oxoisovalerate dehydrogenase (lipoamide) , branched chain keto acid dehydrogenase E1, alpha polypeptide (maple syrup urine disease) , OVD1A; 2-oxoisovalerate dehydrogenase subunit alpha, mitochondrial; maple syrup urine disease; MSU; BCKDH E1-alpha; 2-oxoisovalerate dehydrogenase (lipoamide); branched-chain alpha-keto acid dehydrogenase E1 component alpha chain; MSUD1; OVD1A; BCKDE1A; FLJ45695;
Entrez Gene ID	593
Protein Refseq	NP_000700
UniProt ID	A0A024R0K3
Chromosome Location	19q13.1-q13.2
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	3-methyl-2-oxobutanoate dehydrogenase (2-methylpropanoyl-transferring) activity; alpha-ketoacid dehydrogenase activity; carboxy-lyase activity; metal ion binding; protein binding;