

Recombinant Human BCKDHA cell lysate

Cat. No. BCKDHA-163HCL Lot. No. (See product label)

SPECIFICATION

Species

Human

Description

The branched-chain alpha-keto acid (BCAA) dehydrogenase (BCKD) complex is an inter 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

Size

100 ul

Storage Buffer

1X Sample Buffer (50 mM Tris-HCl, 2% SDS, 10% glycerol, 300 mM 2-mercaptoethanol, 0.01% Bromophenol blue)

Applications

Western Blot;

GENE INFORMATION

Gene Name

BCKDHA branched chain keto acid dehydrogenase E1, alpha polypeptide [Homo sapiens]

Official Symbol

BCKDHA

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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;
Gene ID	593
mRNA Refseq	NM_000709
Protein Refseq	NP_000700
MIM	608348
UniProt ID	P12694
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;

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