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Lieferung & Zahlungsart

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Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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BCKDHA 293T Cell Transient Overexpression Lysate(Denatured)

產品編號 # : H00000593-T01

規格 : [100 uL]

List All

Specification

Transfected Cell Line: 293T

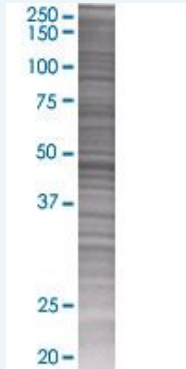
Plasmid: pCMV-BCKDHA full-length

Host: Human

Theoretical MW (kDa): 49.06

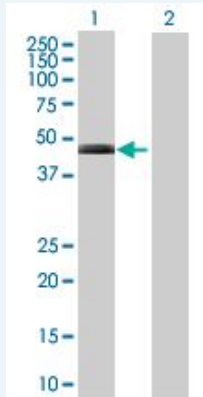
Quality Control Testing: Transient overexpression cell lysate was tested with Anti-BCKDHA antibody ([H00000593-B01](#)) by Western Blots.

SDS-PAGE Gel



BCKDHA transfected lysate.

Western Blot



Lane 1: BCKDHA transfected lysate (49.06 KDa)

Lane 2: Non-transfected lysate.

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

Storage Instruction: Store at -80°C. Aliquot to avoid repeated freezing and thawing.

MSDS:  [Download](#)

Applications

Western Blot

Gene Information

Entrez GeneID: [593](#)

GeneBank Accession#: [NM_000709.2](#)

Protein Accession#: [NP_000700.1](#)

Gene Name: BCKDHA

Gene Alias: BCKDE1A,FLJ45695,MSU,MSUD1,OVD1A

Gene Description: branched chain keto acid dehydrogenase E1, alpha polypeptide

Omim ID: [248600](#), [608348](#)

Gene Ontology: [Hyperlink](#)

Gene Summary: 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

Other Designations: 2-oxoisovalerate dehydrogenase (lipoamide), branched-chain alpha-keto acid dehydrogenase complex E1 alpha subunit, maple syrup urine disease

Gene Pathway

[Metabolic pathways Valine, leucine and isoleucine degradation](#)

Related Disease

[Maple Syrup Urine Disease](#)

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