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Lieferung & Zahlungsart

siehe unsere [Liefer- und Versandbedingungen](#)

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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BCKDHA purified MaxPab mouse polyclonal antibody (B01P) MaxPab®

Catalog # : H00000593-B01P

規格 : [50 ug]

[List All](#)

Specification

Product Description: Mouse polyclonal antibody raised against a full-length human BCKDHA protein.

Immunogen: BCKDHA (NP_000700.1, 1 a.a. ~ 445 a.a) full-length human protein.

Sequence:
 MAVAIAAARVWRLNRGLSQAALLLRQPGARGLARSHPPRQQQFSSL
 DDKPQFPGASAEFIDKLEFIQPNVISGIPYRVMDRQQIINPSEDPHLPKE
 KVLKLYKSMTLLNTMDRILYESQRQGRISFYMTNYGEEGTHVGSAAALD
 NTDLVFGQYREAGVLMYRDYPLELFMAQCYGNISDLGKGRQMPVHYGC
 KERHFVTISSPLATQIPQAVGAAYAARANANRVVICYFGEAASEGDAH
 AGFNFAATLECPPIFFCRNNGYAISTPTSEQYRGDIAARGPGYGIMSIRV
 DGNDVFAVYNATKEARRRAVAENQPFLIEAMTYRIGHHSTSDSSAYRSV
 DEVNYWDKQDHPISRLRHYLLSQGWWDDEEQEKAWRKQSRRKVMFAFE
 QAERKPKPNLLFSDVYQEMPAQLRKQQESLARHLQTYGEHYPLDHF
 DK

Host: Mouse

Reactivity: Human

Quality Control Testing: Antibody reactive against mammalian transfected lysate.

Storage Buffer: In 1x PBS, pH 7.4

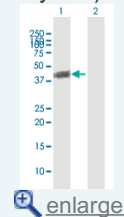
Storage Instruction: Store at -20°C or lower. Aliquot to avoid repeated freezing and thawing.

MSDS: [Download](#)

Datasheet: [Download](#)

Application Image

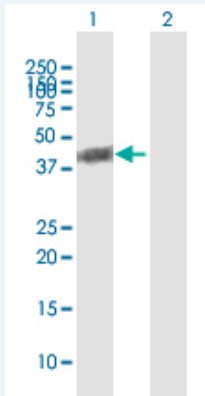
Western Blot (Transfected lysate)



[enlarge](#)

Applications

Western Blot (Transfected lysate)



Western Blot analysis of BCKDHA expression in transfected 293T cell line (H00000593-T01) by BCKDHA MaxPab polyclonal antibody.

Lane 1: BCKDHA transfected lysate(48.95 KDa).
 Lane 2: Non-transfected lysate.

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](#)