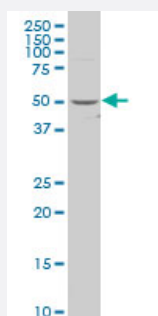


BCKDHA polyclonal antibody (A01)

Catalog # H00000593-A01

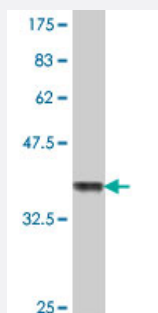
Size 50 uL

Applications



Western Blot (Cell lysate)

BCKDHA polyclonal antibody (A01), Lot # 051212JC01 Western Blot analysis of BCKDHA expression in Y-79 (Cat # L042V1).



Western Blot detection against Immunogen (37 KDa) .

Specification

Product Description	Mouse polyclonal antibody raised against a partial recombinant BCKDHA.
Immunogen	BCKDHA (NP_000700, 347 a.a. ~ 445 a.a) partial recombinant protein with GST tag.
Sequence	SVDEVNYWDKQDHPISRLRHYLLSQGWWDDEEQEKAWRKQSRRKVMFAFEQAERKPKPNPPLL FSDVYQEMPAQLRKQQESLARHLQTYGEHYPLDHFDDK
Host	Mouse
Reactivity	Human
Quality Control Testing	Antibody Reactive Against Recombinant Protein. Western Blot detection against Immunogen (37 KDa) .

Storage Buffer

50 % glycerol

Storage Instruction

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

Applications

- Western Blot (Cell lysate)

BCKDHA polyclonal antibody (A01), Lot # 051212JC01 Western Blot analysis of BCKDHA expression in Y-79 (Cat # L042V1).

[Protocol Download](#)

- Western Blot (Recombinant protein)

[Protocol Download](#)

- ELISA

Gene Info — BCKDHA

Entrez GeneID[593](#)**GeneBank Accession#**[NM_000709](#)**Protein Accession#**[NP_000700](#)**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 1A. 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

Pathway

- [Metabolic pathways](#)
- [Valine](#)

Disease

- [Maple Syrup Urine Disease](#)