BCKDHA rabbit monoclonal antibody

Catalog # H00000593-K Size 100 ug x up to 3

Specification

Product Description	Rabbit monoclonal antibody raised against a human BCKDHA peptide using ARM Technology.
Immunogen	A synthetic peptide of human BCKDHA is used for rabbit immunization. Customer or Abnova will decide on the preferred peptide sequence.
Host	Rabbit
Library Construction	Non-fusion antibody library from rabbit spleen (ARM Technology).
Expression	Overexpression vector and transfection into 293H cell line.
Reactivity	Human
Purification	Protein A
Isotype	IgG
Quality Control Testing	Antibody reactive against human BCKDHA peptide by ELISA and mammalian transfected lysate by Western Blot.
Storage Buffer	In 1x PBS, pH 7.4
Storage Instruction	Store at -20°C or lower. Aliquot to avoid repeated freezing and thawing.
Deliverable	Up to three rabbit IgG clones of 100 ug each will be delivered to customer.
Note	1. Customer may provide cell or tissue lysate for antibody screening. 2. Rabbit monoclonal antibody generated by ARM technology is amenable to antibody engineering in cluding $F(ab)_2$, IgG, scFv and different Fc and non-Fc conjugates per customer request.

Applications

- Western Blot (Transfected lysate)

  Protocol Download

- ELISA

Gene Info — BCKDHA

Entrez GeneID	593
GeneBank Accession#	BCKDHA
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 innter mitoch ondrial enzyme complex that catalyzes the second major step in the catabolism of the branched-c hain amino acids leucine, isoleucine, and valine. The BCKD complex consists of three catalytic c omponents: a heterotetrameric (alpha2-beta2) branched-chain alpha-keto acid decarboxylase (E 1), a dihydrolipoyl transacylase (E2), and a dihydrolipoamide dehydrogenase (E3). This gene enc odes the alpha subunit of the decarboxylase (E1) component. Mutations in this gene result in mapl e syrup urine disease, type IA. Multiple transcript variants encoding different isoforms have been f ound for this gene
Other Designations	2-oxoisovalerate dehydrogenase (lipoamide)|branched-chain alpha-keto acid dehydrogenase co mplex E1 alpha subunit|maple syrup urine disease

Pathway

- Metabolic pathways
- Valine

Disease

- Maple Syrup Urine Disease