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BCKDHB Antibody (N-term) Blocking peptide
Synthetic peptide
Catalog # BP10672a

Specification

BCKDHB Antibody (N-term) Blocking peptide - Product Information

Primary Accession P21953

BCKDHB Antibody (N-term) Blocking peptide - Additional Information

Gene ID 594

Other Names
2-oxoisovalerate dehydrogenase subunit beta, mitochondrial, Branched-chain alpha-keto acid dehydrogenase E1 component beta chain, BCKDE1B, BCKDH E1-beta, BCKDHB

Format
Peptides are lyophilized in a solid powder format. Peptides can be reconstituted in solution using the appropriate buffer as needed.

Storage
Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C.

Precautions
This product is for research use only. Not for use in diagnostic or therapeutic procedures.

BCKDHB Antibody (N-term) Blocking peptide - Protein Information

Name BCKDHB (HGNC:987)

Function
Together with BCKDHA forms the heterotetrameric E1 subunit of the mitochondrial branched-chain alpha-ketoacid dehydrogenase (BCKD) complex. The BCKD complex catalyzes the multi-step oxidative decarboxylation of alpha-ketoacids derived from the branched-chain amino-acids valine, leucine and isoleucine producing CO2 and acyl-CoA which is subsequently utilized to produce energy. The E1 subunit catalyzes the first step with the decarboxylation of the alpha-ketoacid forming an enzyme-product intermediate. A reductive acylation mediated by the lipoylamide cofactor of E2 extracts the acyl group from the E1 active site for the next step of the reaction.

Cellular Location
Mitochondrion matrix

BCKDHB Antibody (N-term) Blocking peptide - Protocols

Provided below are standard protocols that you may find useful for product applications.

- Blocking Peptides

BCKDHB Antibody (N-term) Blocking peptide - Images

BCKDHB Antibody (N-term) Blocking peptide - Background

Branched-chain keto acid dehydrogenase is a multienzymecomplex associated with the inner membrane of mitochondria, andfunctions in the catabolism of branched-chain amino acids. Thecomplex consists of multiple copies of 3 components: branched-chainalpha-keto acid decarboxylase (E1), lipoamide acyltransferase (E2)and lipoamide dehydrogenase (E3). This gene encodes the E1 betasubunit, and mutations therein have been associated with maplesyrup urine disease (MSUD), type 1B, a disease characterized by amaple syrup odor to the urine in addition to mental and physicalretardation, and feeding problems.

BCKDHB Antibody (N-term) Blocking peptide - References

Rose, J.E., et al. Mol. Med. 16 (7-8), 247-253 (2010) :Gorzelany, K., et al. Turk. J. Pediatr. 51(2):97-102(2009)Quental, S., et al. Mol. Genet. Metab. 94(2):148-156(2008)Kang, H., et al. Fertil. Steril. 89(3):728-731(2008)Flaschker, N., et al. J. Inherit. Metab. Dis. 30(6):903-909(2007)