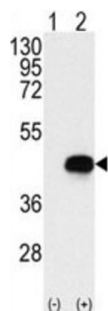
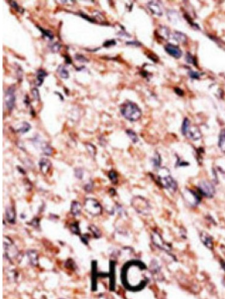


Branched Chain Alpha-Ketoacid Dehydrogenase Kinase (BCKDK) Antibody

Catalogue No.: abx033187



The second major step in the catabolism of the branched-chain amino acids, isoleucine, leucine, and valine, is irreversibly catalyzed by the branched-chain alpha-keto acid dehydrogenase complex (BCKD), an inner-mitochondrial enzyme complex composed of 3 catalytic components: a branched-chain alpha-keto acid decarboxylase (E1), a dihydrolipoyl transacylase (E2), and a dihydrolipoamide dehydrogenase (E3). The complex also contains 2 enzymes that regulated the state of activity of the BCKD complex: a kinase (BCKDK), and a phosphorylase. The ubiquitously expressed kinase contains 1 histidine kinase domain. Maple syrup urine disease (MSUD) is a pathology secondary to an enzyme defect in the catabolic pathway of leucine, isoleucine, and valine. Accumulation of these amino acids and their corresponding keto acids results in encephalopathy and progressive neurodegeneration in infants not treated for MSUD.

Target: Branched Chain Alpha-Ketoacid Dehydrogenase Kinase (BCKDK)

Clonality: Polyclonal

Reactivity: Human

Tested Applications: ELISA, WB, IHC

Host: Rabbit

Recommended dilutions: WB: 1/1000, IHC-P: 1/50 - 1/100. Not tested in IHC-F. Optimal dilutions/concentrations should be determined by the end user.

Conjugation: Unconjugated

Datasheet

Version: 1.0.0
Revision date: 03 Dec 2024



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|----------------------------|--|
| Immunogen: | KLH-conjugated synthetic peptide between 120-151 amino acids from the Central region of human BCKDK. |
| Isotype: | IgG |
| Form: | Liquid |
| Purification: | Purified Rabbit Polyclonal Antibody. |
| Storage: | Aliquot and store at -20°C. Avoid repeated freeze/thaw cycles. |
| UniProt Primary AC: | O14874 (UniProt , ExPASy) |
| Gene Symbol: | BCKDK |
| KEGG: | hsa:10295 |
| String: | 9606.ENSP00000378405 |
| Molecular Weight: | Calculated MW: 46.4 kDa |
| Buffer: | PBS containing 0.09% sodium azide. |
| Specificity: | Predicted to react with Mouse and Rat BCKDK. |
| Note: | This product is for research use only. |

For Reference Only