

### **BCKDK Antibody (Center)**

Purified Rabbit Polyclonal Antibody (Pab) Catalog # AP7112a

### **Specification**

#### **BCKDK Antibody (Center) - Product Information**

Application	WB, IHC-P,E
Primary Accession	<u>014874</u>
Other Accession	<u>Q00972, O55028</u>
Reactivity	Human
Predicted	Mouse, Rat
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit Ig
Calculated MW	46360
Antigen Region	120-151

#### BCKDK Antibody (Center) - Additional Information

#### Gene ID 10295

# Other Names

[3-methyl-2-oxobutanoate dehydrogenase [lipoamide]] kinase, mitochondrial, Branched-chain alpha-ketoacid dehydrogenase kinase, BCKD-kinase, BCKDHKIN, BCKDK

#### Target/Specificity

This BCKDK antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 120-151 amino acids from the Central region of human BCKDK.

#### Dilution

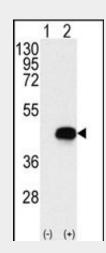
WB~~1:1000 IHC-P~~1:50~100

#### Format

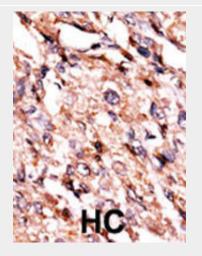
Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is prepared by Saturated Ammonium Sulfate (SAS) precipitation followed by dialysis against PBS.

#### Storage

Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.



Western blot analysis of BCKDK (arrow) using rabbit polyclonal BCKDK Antibody (Center) (Cat.#AP7112a). 293 cell lysates (2 ug/lane) either nontransfected (Lane 1) or transiently transfected with the BCKDK gene (Lane 2) (Origene Technologies).



Formalin-fixed and paraffin-embedded human cancer tissue reacted with the primary antibody, which was peroxidase-conjugated to the secondary antibody, followed by AEC staining. This data demonstrates the use of this antibody for immunohistochemistry; clinical relevance has not been evaluated. BC = breast carcinoma; HC = hepatocarcinoma.



#### Precautions

BCKDK Antibody (Center) is for research use only and not for use in diagnostic or therapeutic procedures.

#### **BCKDK Antibody (Center) - Protein Information**

#### Name BCKDK

### Function

Catalyzes the phosphorylation and inactivation of the branched-chain alpha-ketoacid dehydrogenase complex, the key regulatory enzyme of the valine, leucine and isoleucine catabolic pathways. Key enzyme that regulate the activity state of the BCKD complex.

**Cellular Location** Mitochondrion matrix. Mitochondrion

Tissue Location Ubiquitous.

# **BCKDK Antibody (Center) - Protocols**

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

- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- <u>Cell Culture</u>

# BCKDK Antibody (Center) - Background

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 ubiquitiously 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.

## **BCKDK Antibody (Center) - References**

Chang, C.F., et al., J. Biol. Chem. 277(18):15865-15873 (2002). Popov, K.M., et al., J. Biol. Chem. 267(19):13127-13130 (1992). Zneimer, S.M., et al., Genomics 10(3):740-747 (1991).