

Product datasheet for **AP13678PU-N**

BCKDH kinase (BCKDK) (C-term) Rabbit Polyclonal Antibody

Product data:

Product Type:	Primary Antibodies
Applications:	WB
Recommended Dilution:	ELISA: 1/1,000. Western blotting: 1/100 - 1/500. Flow cytometry.
Reactivity:	Human, Mouse
Host:	Rabbit
Isotype:	Ig
Clonality:	Polyclonal
Immunogen:	This antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide selected from the C-terminal region of human BCKDK.
Specificity:	This antibody reacts to BCKDK.
Formulation:	PBS with 0.09% (W/V) sodium azide State: Purified State: Liquid purified Ig
Concentration:	lot specific
Purification:	Protein G column, eluted with high and low pH buffers and neutralized immediately, followed by dialysis against PBS
Conjugation:	Unconjugated
Storage:	Store the antibody undiluted at 2-8°C for one month or (in aliquots) at -20°C for longer. Avoid repeated freezing and thawing.
Stability:	Shelf life: one year from despatch.
Gene Name:	branched chain ketoacid dehydrogenase kinase
Database Link:	Entrez Gene 10295 Human O14874



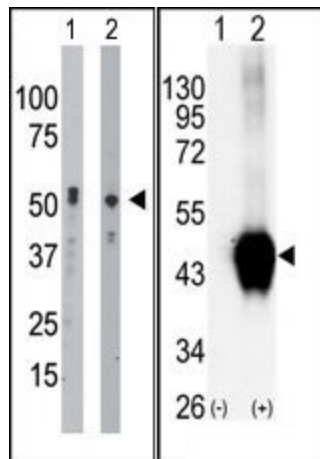
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Background:

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 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.

Synonyms:

BCKD-kinase, BCKDHKIN

Product images:


(LEFT)The anti-BCKDK Pab is used in Western blot to detect BCKDK in mouse intestine tissue lysate (Lane 1) and HeLa cell lysate (Lane 2). (RIGHT)Western blot analysis of BCKDK (arrow) using rabbit polyclonal BCKDK Antibody (C-term).293 cell lysates (2 ug/lane) either nontransfected (Lane 1) or transiently transfected with the BCKDK gene (Lane 2)