

Product datasheet for **TA302079**

BCKDH kinase (BCKDK) Rabbit Polyclonal Antibody

Product data:

| | |
|-------------------------|--|
| Product Type: | Primary Antibodies |
| Applications: | IHC, WB |
| Recommended Dilution: | WB: 1:1000, IHC: 1:50~100 |
| Reactivity: | Human (Predicted: Mouse, Rat) |
| Host: | Rabbit |
| Isotype: | Ig |
| Clonality: | Polyclonal |
| Immunogen: | 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. |
| Formulation: | Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. |
| Concentration: | lot specific |
| Purification: | This antibody is prepared by Saturated Ammonium Sulfate (SAS) precipitation followed by dialysis against PBS. |
| Conjugation: | Unconjugated |
| Storage: | Store at -20°C as received. |
| Stability: | Stable for 12 months from date of receipt. |
| Predicted Protein Size: | 46360 Da |
| Gene Name: | branched chain ketoacid dehydrogenase kinase |
| Database Link: | NP_005872 Entrez Gene 12041 Mouse Entrez Gene 29603 Rat Entrez Gene 10295 Human O14874 |


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Background:

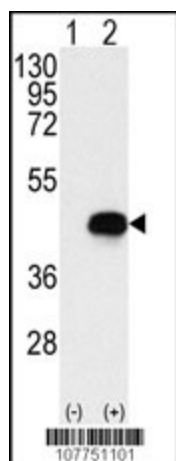
The second major step in the catabolism of the branched-chain amino acids, isoleucine, leucine, and valine, is irreversibly catalyzed by the branched-chain alpha-keto acid dehydrogenase complex (BCKD), an inner-mitochondrial enzyme complex composed of 3 catalytic components: a branched-chain alpha-keto 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:

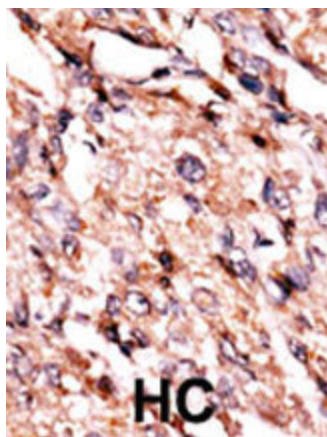
BCKDKD; BDK

Protein Families:

Druggable Genome, Protein Kinase

Product images:


Western blot analysis of BCKDK (arrow) using rabbit polyclonal BCKDK Antibody (Center) (Cat.#TA302079). 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.