

## BCKDHB Antibody (N-term)

Catalog_no :	AB0812
Applications :	WB, IHC-P
Reactivity :	H
Category :	抗原抗体
Size :	100μL/50μL
Immunogen :	HUMAN:41-70
Specificity :	This BCKDHB antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 41-70 amino acids from the N-terminal region of human BCKDHB.
Dilution :	,1:500;,1:500;IF,1:25;WB,1:1000;FC,1:10~50;
Purification :	Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.
Other_name :	2-oxoisovalerate dehydrogenase subunit beta, mitochondrial, Branched-chain alpha-keto acid dehydrogenase E1 component beta chain, BCKDE1B, BCKDH E1-beta, BCKDHB
Isotype :	Rabbit Ig
Background :	Branched-chain keto acid dehydrogenase is a multienzyme complex associated with the inner membrane of mitochondria, and functions in the catabolism of branched-chain amino acids. The complex consists of multiple copies of 3 components: branched-chain alpha-keto acid decarboxylase (E1), lipoamide acyltransferase (E2) and lipoamide dehydrogenase (E3). This gene encodes the E1 beta subunit, and mutations therein have been associated with maple syrup urine disease (MSUD), type 1B, a disease characterized by a maple syrup odor to the urine in addition to mental and physical retardation, and feeding problems.
reference :	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