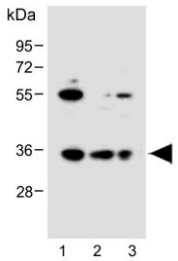
# BCKDHB Antibody (F54271)

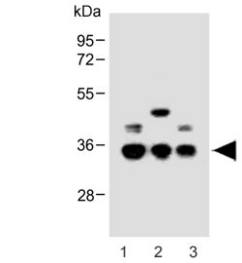
| Catalog No. | Formulation | Size |
|---|---|---|
| F54271-0.4ML | In 1X PBS, pH 7.4, with 0.09% sodium azide | 0.4 ml |
| F54271-0.08ML | In 1X PBS, pH 7.4, with 0.09% sodium azide | 0.08 ml |

**Bulk quote request**

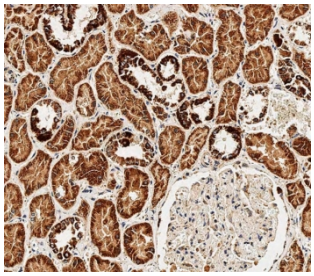
| | |
|---|---|
| **Availability** | 1-3 business days |
| **Species Reactivity** | Human |
| **Format** | Purified |
| **Host** | Rabbit |
| **Clonality** | Polyclonal (rabbit origin) |
| **Isotype** | Rabbit Ig |
| **Purity** | Antigen affinity purified |
| **UniProt** | P21953 |
| **Localization** | Cytoplasmic |
| **Applications** | Western Blot : 1:500-1:2000<br>Immunohistochemistry (FFPE) : 1:25<br>Immunofluorescence : 1:25<br>Flow Cytometry : 1:25 (1x10e6 cells) |
| **Limitations** | This BCKDHB antibody is available for research use only. |



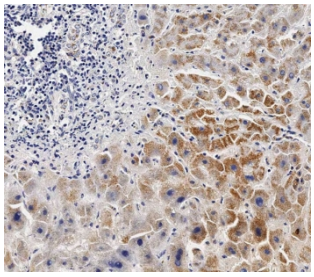

Western blot testing of human 1) HeLa, 2) 293T and 3) Jurkat cell lysate with BCKDHB antibody. Predicted molecular weight: ~43 kDa, observed molecular weight: ~35 kDa.



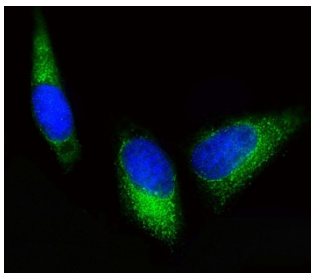

Western blot testing of human 1) HeLa, 2) Jurkat and 3) Daudi cell lysate with BCKDHB antibody. Predicted molecular weight: ~43 kDa, observed molecular weight: ~35 kDa.



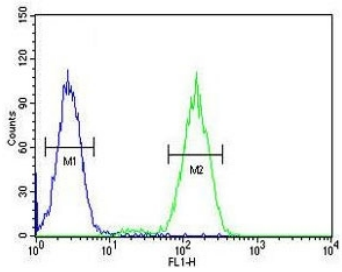
IHC testing of FFPE human kidney tissue with BCKDHB antibody. HIER: steam section in pH6 citrate buffer for 20 min and allow to cool prior to staining.



IHC testing of FFPE human hepatocarinoma with BCKDHB antibody. HIER: steam section in pH6 citrate buffer for 20 min and allow to cool prior to staining.



Immunofluorescent staining of fixed and permeabilized human HeLa cells with BCKDHB antibody (green) and DAPI nuclear stain (blue).




Flow cytometry testing of fixed and permeabilized human HeLa cells with BCKDHB antibody; Blue=isotype control, Green= BCKDHB antibody.

## Description

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.

## Application Notes

The stated application concentrations are suggested starting points. Titration of the BCKDHB antibody may be required due to differences in protocols and secondary/substrate sensitivity.

## Immunogen

A portion of amino acids 41-70 from the human protein were used as the immunogen for the BCKDHB antibody.

## Storage

Aliquot the BCKDHB antibody and store frozen at -20oC or colder. Avoid repeated freeze-thaw cycles.


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