

## HIBADH polyclonal antibody

Catalog: BS61107

Host: Rabbit

Reactivity: Human, Mouse, Rat

### BackGround:

HIBADH (3-hydroxyisobutyrate dehydrogenase) is a 336 amino acid mitochondrial enzyme that catalyzes the NAD<sup>+</sup>-dependent, reversible oxidization of 3-Hydroxyisobutyrate to methylmalonate semialdehyde, an intermediate of valine catabolism. The enzyme functions as a homodimer between a pH of 7.0 and 10.0, with optimal activity between 8.8 and 9.0. It was previously hypothesized that defects in the gene encoding HIBADH may be the cause of 3-Hydroxyisobutyric aciduria, a rare disorder that is characterized by a variety of clinical manifestations such as neurodevelopmental problems and dysmorphic features. However, it was shown that HIBADH activity was equal in patients with 3-Hydroxyisobutyric aciduria as compared with controls.

### Product:

1 mg/ml in Phosphate buffered saline (PBS) with 0.05% sodium azide, approx. pH 7.3.

### Molecular Weight:

~ 35 kDa

### Swiss-Prot:

P31937

### Purification&Purity:

The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen and the purity is > 95% (by SDS-PAGE).

### Applications:

WB: 1:500~1:1000

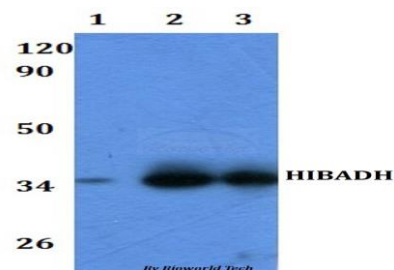
### Storage&Stability:

Store at 4 °C short term. Aliquot and store at -20 °C long term. Avoid freeze-thaw cycles.

### Specificity:

HIBADH polyclonal antibody detects endogenous levels of HIBADH protein.

### DATA:



Western blot (WB) analysis of HIBADH polyclonal antibody at 1:500 dilution Lane1:

Lane2:RAW264.7 whole cell lysate Lane3:H9C2 whole cell lysate

Lane4:HELA whole cell lysate

### Note:

For research use only, not for use in diagnostic procedure.

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