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Datasheet for ABIN5004614

## anti-HIBADH antibody (AA 251-336) (Alexa Fluor 750)

### Overview

Quantity:	100 µL
Target:	HIBADH
Binding Specificity:	AA 251-336
Reactivity:	Human, Mouse
Host:	Rabbit
Clonality:	Polyclonal
Conjugate:	This HIBADH antibody is conjugated to Alexa Fluor 750
Application:	Western Blotting (WB), Immunofluorescence (Cultured Cells) (IF (cc)), Immunofluorescence (Paraffin-embedded Sections) (IF (p))

### Product Details

Immunogen:	KLH conjugated synthetic peptide derived from human HIBADH
Isotype:	IgG
Cross-Reactivity:	Human, Mouse
Predicted Reactivity:	Rat,Cow,Sheep,Horse
Purification:	Purified by Protein A.

### Target Details

Target:	HIBADH
Alternative Name:	HIBADH ( <a href="#">HIBADH Products</a> )

## Target Details

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**Background:** Synonyms: 3 hydroxy 2 methylpropanoate:NAD<sup>+</sup> oxidoreductase, 3 hydroxyisobutyrate dehydrogenase, 3 hydroxyisobutyrate dehydrogenase mitochondrial, EC 1.1.1.31, MGC40361, NS5ATP1, 3HIDH\_HUMAN.

Background: HIBADH 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.

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**Gene ID:** 11112

## Application Details

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**Application Notes:** IF(IHC-P) 1:50-200  
IF(IHC-F) 1:50-200  
IF(ICC) 1:50-200

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**Restrictions:** For Research Use only

## Handling

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**Format:** Liquid

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**Concentration:** 1 µg/µL

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**Buffer:** Aqueous buffered solution containing 0.01M TBS (pH 7.4) with 1 % BSA, 0.03 % Proclin300 and 50 % Glycerol.

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**Preservative:** ProClin

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**Precaution of Use:** This product contains ProClin: a POISONOUS AND HAZARDOUS SUBSTANCE, which should be handled by trained staff only.

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**Storage:** -20 °C

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**Storage Comment:** Store at -20°C. Aliquot into multiple vials to avoid repeated freeze-thaw cycles.

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**Expiry Date:** 12 months