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Datasheet for ABIN1698221

anti-HIBADH antibody (AA 251-336) (Alexa Fluor 647)

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 647
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 (HIBADH Products)

Target Details

Background:	<p>Synonyms: 3 hydroxy 2 methylpropanoate:NAD⁺ oxidoreductase, 3 hydroxyisobutyrate dehydrogenase, 3 hydroxyisobutyrate dehydrogenase mitochondrial, EC 1.1.1.31, MGC40361, NS5ATP1, 3HIDH_HUMAN.</p> <p>Background: HIBADH is a 336 amino acid mitochondrial enzyme that catalyzes the NAD⁺-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.</p>
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Gene ID:	11112
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Application Details

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
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Handling

Format:	Liquid
Concentration:	1 µg/µL
Buffer:	Aqueous buffered solution containing 0.01M TBS (pH 7.4) with 1 % BSA, 0.03 % Proclin300 and 50 % Glycerol.
Preservative:	ProClin
Precaution of Use:	This product contains ProClin: a POISONOUS AND HAZARDOUS SUBSTANCE, which should be handled by trained staff only.
Storage:	-20 °C
Storage Comment:	Store at -20°C. Aliquot into multiple vials to avoid repeated freeze-thaw cycles.
Expiry Date:	12 months