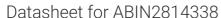
antibodies -online.com





anti-HIBADH antibody (AA 251-336) (Alexa Fluor 594)



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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 594
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

rargerbetane	
Background:	Synonyms: 3 hydroxy 2 methylpropanoate:NAD+ 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+-
	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.
Gene ID:	11112
Application Details	
Application Notes:	IF(IHC-P) 1:50-200
	IF(IHC-F) 1:50-200
	IF(ICC) 1:50-200
Restrictions:	For Research Use only
Lloudline	
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.

12 months

Expiry Date: