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Datasheet for ABIN1707427
anti-HIBADH antibody (AA 251-336) (Cy7)

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 Cy7
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: 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

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