Datasheet for ABIN1696411

anti-HIBCH antibody (AA 251-350) (Alexa Fluor 555)

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Overview

Quantity:	100 µL
Target:	HIBCH
Binding Specificity:	AA 251-350
Reactivity:	Human
Host:	Rabbit
Clonality:	Polyclonal
Conjugate:	This HIBCH antibody is conjugated to Alexa Fluor 555
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 HIBCH
Isotype:	IgG
Predicted Reactivity:	Human
Purification:	Purified by Protein A.

Target Details

Target:	HIBCH
Alternative Name:	Hibch (HIBCH Products)
Background:	Synonyms: 3 hydroxyisobutyryl Coenzyme A hydrolase, 3 hydroxyisobutyryl Coenzyme A

Target Details

	hydrolase, mitochondrial, 3-hydroxyisobutyryl-CoA hydrolase, 3-hydroxyisobutyryl-coenzyme A hydrolase, BETA HYDROXYISOBUTYRYL COENZYME A HYDROLASE, HIB CoA hydrolase, HIB-CoA hydrolase, HIBCH, HIBCH_HUMAN, HIBCoA hydrolase, HIBYL CoA H, HIBYL CoAH, HIBYL-CoA-H, HIBYLCoA H, HIBYLCoAH, mitochondrial, HIBCH_HUMAN. Background: HIBCH is a 386 amino acid protein belonging to the enoyl-CoA hydratase/isomerase family. Localizing to the mitochondria, HIBCH is highly expressed in liver and kidney, with lower levels found in heart, muscle and brain. HIBCH hydrolyzes HIBYL-CoA, a saline catabolite, and _-hydroxypropionyl-CoA, an intermediate in the minor pathway involved in the metabolism of proprionate. Existing as two alternatively spliced isoforms, the gene encoding HIBCH maps to human chromosome 2q32.2. Defects to this gene result in HIBCH deficiency (HIBCHD), known alternatively as deficiency of _-hydroxyisobutyryl CoA deacylase or methacrylic aciduria. HIBCHD is characterized by the accumulation of methacrylyl-CoA, a highly reactive compound that undergoes addition reactions with free sulfhydryl groups. Phenotypic symptoms include early deterioration of neurological function, delayed motor skill development and hypotonia.
Gene ID:	26275
Pathways:	Monocarboxylic Acid Catabolic Process

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.

Handling

Storage:	-20 °C
Storage Comment:	Store at -20°C. Aliquot into multiple vials to avoid repeated freeze-thaw cycles.
Expiry Date:	12 months

