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anti-HIBCH antibody (AA 251-350)



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Quantity:	100 μL	
Target:	HIBCH	
Binding Specificity:	AA 251-350	
Reactivity:	Human	
Host:	Rabbit	
Clonality:	Polyclonal	
Conjugate:	This HIBCH antibody is un-conjugated	
Application:	Western Blotting (WB), ELISA, Immunofluorescence (Cultured Cells) (IF (cc)), Immunofluorescence (Paraffin-embedded Sections) (IF (p)), Immunohistochemistry (Paraffin-embedded Sections) (IHC (p)), Immunocytochemistry (ICC), Immunohistochemistry (Frozen Sections) (IHC (fro))	

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)

Target Details

Background:

Synonyms: 3 hydroxyisobutyryl Coenzyme A hydrolase, 3 hydroxyisobutyryl Coenzyme A 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, 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:

WB 1:300-5000

ELISA 1:500-1000

IHC-P 1:200-400

IHC-F 1:100-500

IF(IHC-P) 1:50-200

IF(IHC-F) 1:50-200

IF(ICC) 1:50-200

ICC 1:100-500

Restrictions:

For Research Use only

Handling

Format:

Liquid

Concentration:

 $1 \mu g/\mu L$

Handling

Buffer:	0.01M TBS(pH 7.4) with 1 % BSA, 0.02 % 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:	4 °C,-20 °C	
Storage Comment:	Shipped at 4°C. Store at -20°C for one year. Avoid repeated freeze/thaw cycles.	
Expiry Date:	12 months	