# abcam

## Product datasheet

# Anti-HIBCH antibody ab153826

1 References 3 Images

Overview

Product name Anti-HIBCH antibody

**Description** Rabbit polyclonal to HIBCH

Host species Rabbit

Tested applications Suitable for: WB, IHC-P, ICC/IF

Species reactivity Reacts with: Human

Immunogen Recombinant fragment, corresponding to a region within amino acids 1-288 of Human HIBCH

(UniProt: Q6NVY1).

**Positive control** 293T, HepG2, Molt4 whole cell lysates; A431 cells; Hepatoma tissue.

**General notes**The Life Science industry has been in the grips of a reproducibility crisis for a number of years.

Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets

your needs before purchasing.

If you have any questions, special requirements or concerns, please send us an inquiry and/or

contact our Support team ahead of purchase. Recommended alternatives for this product can be

found below, along with publications, customer reviews and Q&As

**Properties** 

Form Liquid

**Storage instructions** Shipped at 4°C. Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.

**Storage buffer** pH: 7.00

Preservative: 0.01% Thimerosal (merthiolate)

Constituents: 1.21% Tris, 0.75% Glycine, 10% Glycerol (glycerin, glycerine)

Purity Immunogen affinity purified

**Clonality** Polyclonal

**Isotype** IgG

**Applications** 

The Abpromise guarantee Our Abpromise guarantee covers the use of ab153826 in the following tested applications.

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The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes	
WB		1/500 - 1/3000. Predicted molecular weight: 43 kDa.	
IHC-P		1/100 - 1/1000.	
ICC/IF		1/100 - 1/1000.	

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**Function** Hydrolyzes 3-hydroxyisobutyryl-CoA (HIBYL-CoA), a saline catabolite. Has high activity toward

isobutyryl-CoA. Could be an isobutyryl-CoA dehydrogenase that functions in valine catabolism.

Also hydrolyzes 3-hydroxypropanoyl-CoA.

**Tissue specificity** Highly expressed in liver and kidney, also detected in heart, muscle and brain (at protein level).

Not detected in lung.

Pathway Amino-acid degradation; L-valine degradation.

Involvement in disease Defects in HIBCH are the cause of HIBCH deficiency (HIBCHD) [MIM:250620]; also known as

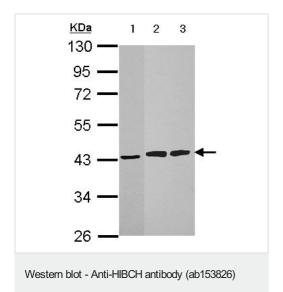
deficiency of beta-hydroxyisobutyryl CoA deacylase or methacrylic aciduria. The enzyme defect results in accumulation of methacrylyl-CoA, a highly reactive compound, which readily undergoes addition reactions with free sulfhydryl groups. Affected individuals showed delayed development of motor skills, hypotonia, initial poor feeding, and a deterioration in neurological function during

first stages of life.

**Sequence similarities** Belongs to the enoyl-CoA hydratase/isomerase family.

**Cellular localization** Mitochondrion.

#### **Images**



All lanes: Anti-HIBCH antibody (ab153826) at 1/1000 dilution

Lane 1 : 293T whole cell lysate

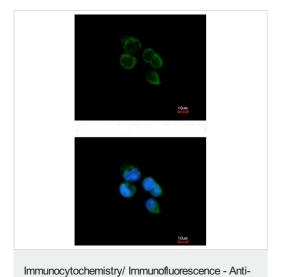
Lane 2 : HepG2 whole cell lysate

Lysates/proteins at 30 µg per lane.

Lane 3: Molt4 whole cell lysate

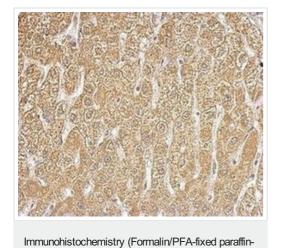
Predicted band size: 43 kDa

10% SDS PAGE



HIBCH antibody (ab153826)

Immunofluorescence analysis of methanol fixed A431 cells labeling HIBCH with ab153826 at 1/200. Lower panel shows costaining with Hoechst 33342.



embedded sections) - Anti-HIBCH antibody

(ab153826)

Immunohistochemical analysis of paraffin embedded Human hepatoma tissue labeling HIBCH with ab153826 at 1/500.

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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