

HIBCH (E-11): sc-515355

BACKGROUND

HIBCH (3-hydroxyisobutyryl-CoA hydrolase) 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 β -hydroxypropionyl-CoA, an intermediate in the minor pathway involved in the metabolism of propionate. 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 β -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.

CHROMOSOMAL LOCATION

Genetic locus: HIBCH (human) mapping to 2q32.2; Hibch (mouse) mapping to 1 C1.1.

SOURCE

HIBCH (E-11) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 272-296 within an internal region of HIBCH of human origin.

PRODUCT

Each vial contains 200 μ g IgM kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-515355 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

APPLICATIONS

HIBCH (E-11) is recommended for detection of HIBCH of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for HIBCH siRNA (h): sc-94322, HIBCH siRNA (m): sc-145958, HIBCH shRNA Plasmid (h): sc-94322-SH, HIBCH shRNA Plasmid (m): sc-145958-SH, HIBCH shRNA (h) Lentiviral Particles: sc-94322-V and HIBCH shRNA (m) Lentiviral Particles: sc-145958-V.

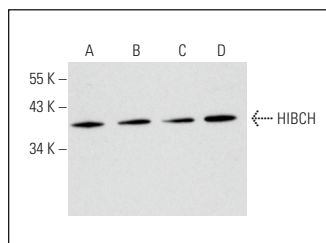
Molecular Weight of HIBCH isoforms 1/2: 43/38 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227 or A549 cell lysate: sc-2413.

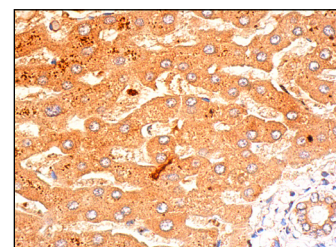
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein L-Agarose: sc-2336 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850. 4) Immunohistochemistry: use m-IgG κ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohisto-mount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



HIBCH (E-11): sc-515355. Western blot analysis of HIBCH expression in Hep G2 (A), A549 (B), Jurkat (C) and Y79 (D) whole cell lysates.



HIBCH (E-11): sc-515355. Immunoperoxidase staining of formalin fixed, paraffin-embedded human liver tissue showing cytoplasmic staining of hepatocytes and bile duct cells. Blocked with 0.25X UltraCruz® Blocking Reagent: sc-516214. Detected with m-IgG κ BP-B: sc-516142 and ImmunoCruz® ABC Kit: sc-516216.

SELECT PRODUCT CITATIONS

1. Biswas, D., et al. 2020. Adverse outcomes in obese cardiac surgery patients correlates with altered branched-chain amino acid catabolism in adipose tissue and heart. *Front. Endocrinol.* 11: 534.
2. Park, S., et al. 2022. Transcription factors TEAD2 and E2A globally repress acetyl-CoA synthesis to promote tumorigenesis. *Mol. Cell* 82: 4246-4261.e11.
3. Xu, M., et al. 2023. Molecular mechanism of valine and its metabolite in improving triglyceride synthesis of porcine intestinal epithelial cells. *Sci. Rep.* 13: 2933.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.