SANTA CRUZ BIOTECHNOLOGY, INC.

HIBCH (E-11): sc-515355

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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 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 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.

REFERENCES

1. Brown, G.K., et al. 1982. β-hydroxyisobutyryl coenzyme A deacylase deficiency: a defect in valine metabolism associated with physical malformations. Pediatrics 70: 532-538.
2. Hawes, J.W., et al. 1996. Primary structure and tissue-specific expression of human β-hydroxyisobutyryl-coenzyme A hydrolase. J. Biol. Chem. 271: 26430-26434.
3. Hillier, L.W., et al. 2005. Generation and annotation of the DNA sequences of human chromosomes 2 and 4. Nature 434: 724-731.
4. Loupatty, F.J., et al. 2007. Mutations in the gene encoding 3-hydroxyisobutyryl-CoA hydrolase results in progressive infantile neurodegeneration. Am. J. Hum. Genet. 80: 195-199.
5. Choudhary, C., et al. 2009. Lysine acetylation targets protein complexes and co-regulates major cellular functions. Science 325: 834-840.
6. Meienberg, J., et al. 2010. Hemizygous deletion of COL3A1, COL5A2, and MSTN causes a complex phenotype with aortic dissection: a lesson for and from true haploinsufficiency. Eur. J. Hum. Genet. 18: 1315-1321.

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 µg 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 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

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 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (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.

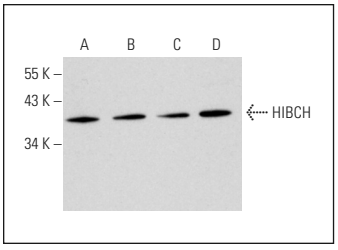
Molecular Weight of HIBCH isoforms 1/2: 43/38 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, A549 cell lysate: sc-2413 or Y79 cell lysate: sc-2240.

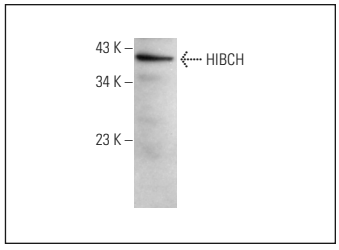
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ 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-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

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. Western blot analysis of HIBCH expression in 3T3-L1 whole cell lysate.

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.

STORAGE

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

RESEARCH USE

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