SANTA CRUZ BIOTECHNOLOGY, INC.

HIBCH siRNA (m): sc-145958

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 (mouse) mapping to 1 C1.1.

PRODUCT

HIBCH siRNA (m) is a pool of 3 target-specific 19-25 nt siRNAs designed to knock down gene expression. Each vial contains 3.3 nmol of lyophilized siRNA, sufficient for a 10 µM solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see HIBCH shRNA Plasmid (m): sc-145958-SH and HIBCH shRNA (m) Lentiviral Particles: sc-145958-V as alternate gene silencing products.

For independent verification of HIBCH (m) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-145958A, sc-145958B and sc-145958C.

RESEARCH USE

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

STORAGE AND RESUSPENSION

Store lyophilized siRNA duplex at -20° C with desiccant. Stable for at least one year from the date of shipment. Once resuspended, store at -20° C, avoid contact with RNAses and repeated freeze thaw cycles.

Resuspend lyophilized siRNA duplex in 330 µl of the RNAse-free water provided. Resuspension of the siRNA duplex in 330 µl of RNAse-free water makes a 10 µM solution in a 10 µM Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

HIBCH siRNA (m) is recommended for the inhibition of HIBCH expression in mouse cells.

SUPPORT REAGENTS

For optimal siRNA transfection efficiency, Santa Cruz Biotechnology's siRNA Transfection Reagent: sc-29528 (0.3 ml), siRNA Transfection Medium: sc-36868 (20 ml) and siRNA Dilution Buffer: sc-29527 (1.5 ml) are recommended. Control siRNAs or Fluorescein Conjugated Control siRNAs are available as 10 µM in 66 µl. Each contain a scrambled sequence that will not lead to the specific degradation of any known cellular mRNA. Fluorescein Conjugated Control siRNAs include: sc-36869, sc-44239, sc-44240 and sc-44241. Control siRNAs include: sc-37007, sc-44230, sc-44231, sc-44232, sc-44233, sc-44234, sc-44235, sc-44236, sc-44237 and sc-44238.

GENE EXPRESSION MONITORING

HIBCH (E-11): sc-515355 is recommended as a control antibody for monitoring of HIBCH gene expression knockdown by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) or immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

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) 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.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor HIBCH gene expression knockdown using RT-PCR Primer: HIBCH (m)-PR: sc-145958-PR (20 µl). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

PROTOCOLS

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