

HADHSC (LB-7): sc-100472

BACKGROUND

HADHSC (hydroxyacyl-coenzyme A (CoA) dehydrogenase, short chain), also known as HAD, HHH4, HADH1, SCHAD or M/SCHAD (medium and short chain L-3-hydroxyacyl-CoA dehydrogenase), is a mitochondrial matrix protein expressed in pancreas, liver, heart, kidney and skeletal muscle. HADHSC exists as a homodimer that participates in lipid metabolism and is essential for the β -oxidation of medium and short chain fatty acids. More specifically, HADHSC catalyzes the dehydrogenation of 3-hydroxyacyl-CoAs to their corresponding 3-ketoacyl-CoAs while NAD⁺ is simultaneously reduced to NADH. Defects in HADHSC can lead to HADH (3- α -hydroxyacyl-CoA dehydrogenase) deficiency and familial hyperinsulinemic hypoglycemia 4 (HHF4). HADH deficiency is characterized as a metabolic disorder with patients exhibiting hepatocerebralopathy, hypoglycemia, myopathy or cardiomyopathy and sometimes experiencing sudden death. HHF4 is a disorder characterized by elevated Insulin secretion that, if left untreated, can cause brain damage from recurrent hypoglycemia episodes.

REFERENCES

1. He, X.Y., et al. 1989. Assay of L-3-hydroxyacyl-coenzyme A dehydrogenase with substrates of different chain lengths. *Anal. Biochem.* 180: 105-109.
2. Vredendaal, P.J., et al. 1996. Human short-chain L-3-hydroxyacyl-CoA dehydrogenase: cloning and characterization of the coding sequence. *Biochem. Biophys. Res. Commun.* 223: 718-723.
3. Bennett, M.J., et al. 1996. Mitochondrial short-chain L-3-hydroxyacyl-coenzyme A dehydrogenase deficiency: a new defect of fatty acid oxidation. *Pediatr. Res.* 39: 185-188.
4. He, X.Y., et al. 1999. Identity of heart and liver L-3-hydroxyacyl coenzyme A dehydrogenase. *Biochim. Biophys. Acta* 1437: 119-123.
5. Treacy, E.P., et al. 2000. Short-chain hydroxyacyl-coenzyme A dehydrogenase deficiency presenting as unexpected infant death: A family study. *J. Pediatr.* 137: 257-259.
6. Clayton, P.T., et al. 2001. Hyperinsulinism in short-chain L-3-hydroxyacyl-CoA dehydrogenase deficiency reveals the importance of β -oxidation in Insulin secretion. *J. Clin. Invest.* 108: 457-465.

CHROMOSOMAL LOCATION

Genetic locus: HADH (human) mapping to 4q25.

SOURCE

HADHSC (LB-7) is a mouse monoclonal antibody raised against recombinant HADHSC of human origin.

PRODUCT

Each vial contains 100 μ g IgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

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

APPLICATIONS

HADHSC (LB-7) is recommended for detection of HADHSC of human origin by Western Blotting (starting dilution 1:200, 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 HADHSC siRNA (h): sc-75222, HADHSC shRNA Plasmid (h): sc-75222-SH and HADHSC shRNA (h) Lentiviral Particles: sc-75222-V.

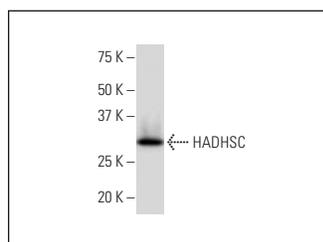
Molecular Weight of HADHSC isoforms 1/2: 34/42 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, HADHSC (h2): 293T Lysate: sc-170845 or Hep G2 cell lysate: sc-2227.

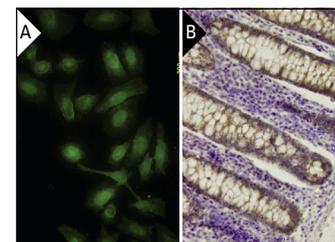
RECOMMENDED SUPPORT REAGENTS

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 A/G PLUS-Agarose: sc-2003 (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 Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



HADHSC (LB-7): sc-100472. Western blot analysis of HADHSC expression in HepG2 whole cell lysate.



HADHSC (LB-7): sc-100472. Immunofluorescence staining of paraformaldehyde-fixed HeLa cells (A) and immunoperoxidase staining of formalin-fixed, paraffin-embedded human colon tissue (B) showing nuclear and cytoplasmic localization.

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.