

HADHSC (T-14): sc-74653

BACKGROUND

HADHSC (hydroxyacyl-coenzyme A (CoA) dehydrogenase, short chain), also known as HAD, HHF4, 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 hepatoencephalopathy, 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

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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.
7. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 601609. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
8. Molven, A., et al. 2004. Familial hyperinsulinemic hypoglycemia caused by a defect in the SCHAD enzyme of mitochondrial fatty acid oxidation. *Diabetes* 5: 221-227.
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STORAGE

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

CHROMOSOMAL LOCATION

Genetic locus: HADH (human) mapping to 4q25; Hadh (mouse) mapping to 3 G3.

SOURCE

HADHSC (T-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of HADHSC of human origin.

PRODUCT

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

Blocking peptide available for competition studies, sc-74653 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

HADHSC (T-14) is recommended for detection of HADHSC of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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).

HADHSC (T-14) is also recommended for detection of HADHSC in additional species, including canine and avian.

Suitable for use as control antibody for HADHSC siRNA (h): sc-75222, HADHSC siRNA (m): sc-75223, HADHSC shRNA Plasmid (h): sc-75222-SH, HADHSC shRNA Plasmid (m): sc-75223-SH, HADHSC shRNA (h) Lentiviral Particles: sc-75222-V and HADHSC shRNA (m) Lentiviral Particles: sc-75223-V.

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

Positive Controls: Hep G2 cell lysate: sc-2227 or HeLa whole cell lysate: sc-2200.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

RESEARCH USE

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

PROTOCOLS

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