



# SZABO SCANDIC

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### Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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# HADHSC (h2): 293T Lysate: sc-170845

## 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  $\beta$ -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<sup>+</sup> is simultaneously reduced to NADH. Defects in HADHSC can lead to HADH (3- $\alpha$ -hydroxyacyl-CoA dehydrogenase) deficiency and familial hyperinsulinemic hypoglycemia 4 (HHF4). HADH deficiency is characterized as a metabolic disorder with patients exhibiting hepatocerebrolopathy, 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  $\beta$ -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.

## STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) for detailed protocols and support products.

## CHROMOSOMAL LOCATION

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

## PRODUCT

HADHSC (h2): 293T Lysate represents a lysate of human HADHSC transfected 293T cells and is provided as 100  $\mu$ g protein in 200  $\mu$ l SDS-PAGE buffer.

## APPLICATIONS

HADHSC (h2): 293T Lysate is suitable as a Western Blotting positive control for human reactive HADHSC antibodies. Recommended use: 10-20  $\mu$ l per lane.

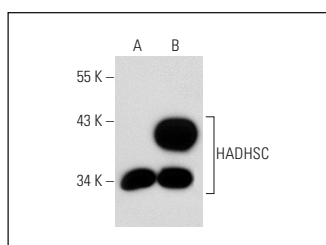
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

HADHSC (LB-7): sc-100472 is recommended as a positive control antibody for Western Blot analysis of enhanced human HADHSC expression in HADHSC transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

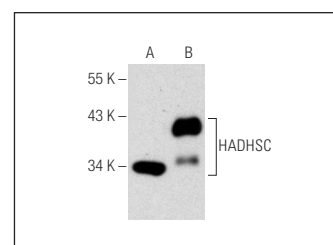
## RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  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.

## DATA



HADHSC (LB-7): sc-100472. Western blot analysis of HADHSC expression in non-transfected: sc-117752 (A) and human HADHSC transfected: sc-170845 (B) 293T whole cell lysates.



HADHSC (A-5): sc-376525. Western blot analysis of HADHSC expression in non-transfected: sc-117752 (A) and human HADHSC transfected: sc-170845 (B) 293T whole cell lysates.

## RESEARCH USE

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