

HADHB (H-290): sc-134922

BACKGROUND

HADHB (trifunctional enzyme subunit β (mitochondrial), acetyl-CoA acyl-transferase) is a 474 amino acid protein encoded by the human gene HADHB. HADHB belongs to the thiolase family, which are ubiquitous enzymes that catalyze the reversible thiolytic cleavage of 3-ketoacyl-CoA into acyl-CoA and acetyl-CoA, a two-step reaction involving a covalent intermediate formed with a catalytic cysteine. HADHB is found in the mitochondrion as an octamer of four α (HADHA) and four β (HADHB) subunits. Defects in HADHB are a cause of trifunctional protein deficiency (TFP deficiency). The clinical manifestations are variable and include hypoglycemia, cardiomyopathy and sudden death. Phenotypes with mainly hepatic and neuromyopathic involvement can also be distinguished. Biochemically, TFP deficiency is defined by the loss of all three enzyme activities of the TFP complex.

REFERENCES

1. Ushikubo, S., et al. 1996. Molecular characterization of mitochondrial trifunctional protein deficiency: formation of the enzyme complex is important for stabilization of both α and β subunits. *Am. J. Hum. Genet.* 58: 979-988.
2. Aoyama, T., et al. 1998. Fluorescence *in situ* hybridization mapping of the α and β subunits (HADHA and HADHB) of human mitochondrial fatty acid β -oxidation multienzyme complex to 2p23 and their evolution. *Cytogenet. Cell Genet.* 79: 221-224.
3. Gevaert, K., et al. 2003. Exploring proteomes and analyzing protein processing by mass spectrometric identification of sorted N-terminal peptides. *Nat. Biotechnol.* 21: 566-569.
4. Spiekeroetter, U., et al. 2003. Molecular and phenotypic heterogeneity in mitochondrial trifunctional protein deficiency due to β subunits mutations. *Hum. Mutat.* 21: 598-607.
5. Das, A.M., et al. 2006. Isolated mitochondrial long-chain ketoacyl-CoA thiolase deficiency resulting from mutations in the HADHB gene. *Clin. Chem.* 52: 530-534.

CHROMOSOMAL LOCATION

Genetic locus: HADHB (human) mapping to 2p23.3; Hadhb (mouse) mapping to 5 B1.

SOURCE

HADHB (H-290) is a rabbit polyclonal antibody raised against amino acids 1-290 mapping at the N-terminus of HADHB 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.

STORAGE

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

APPLICATIONS

HADHB (H-290) is recommended for detection of HADHB of mouse, rat and 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

HADHB (H-290) is also recommended for detection of HADHB in additional species, including equine, canine and bovine.

Suitable for use as control antibody for HADHB siRNA (h): sc-62435, HADHB siRNA (m): sc-62436, HADHB shRNA Plasmid (h): sc-62435-SH, HADHB shRNA Plasmid (m): sc-62436-SH, HADHB shRNA (h) Lentiviral Particles: sc-62435-V and HADHB shRNA (m) Lentiviral Particles: sc-62436-V.

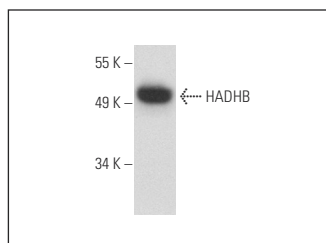
Molecular Weight of HADHB: 52 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, Jurkat whole cell lysate: sc-2204 or K-562 whole cell lysate: sc-2203.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



HADHB (H-290): sc-134922. Western blot analysis of HADHB expression in Jurkat whole cell lysate.

RESEARCH USE

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

MONOS
Satisfaction
Guaranteed

Try **HADHB (E-1): sc-271495** or **HADHB (C-6): sc-271496**, our highly recommended monoclonal alternatives to HADHB (H-290).