

HADHB (D-10): sc-365907

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. 1997. 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. Spiekerkoetter, U., et al. 2003. Molecular and phenotypic heterogeneity in mitochondrial trifunctional protein deficiency due to β subunits mutations. *Hum. Mutat.* 21: 598-607.
5. Adams, D.J., et al. 2003. HADHB, HuR, and CP1 bind to the distal 3'-untranslated region of human Renin mRNA and differentially modulate Renin expression. *J. Biol. Chem.* 278: 44894-44903.
6. Morris, B.J., et al. 2004. cAMP controls human Renin mRNA stability via specific RNA-binding proteins. *Acta Physiol. Scand.* 181: 369-373.

CHROMOSOMAL LOCATION

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

SOURCE

HADHB (D-10) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 63-87 near the N-terminus of HADHB of human origin.

PRODUCT

Each vial contains 200 μ g IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-365907 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

APPLICATIONS

HADHB (D-10) is recommended for detection of HADHB of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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 (D-10) is also recommended for detection of HADHB in additional species, including 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.

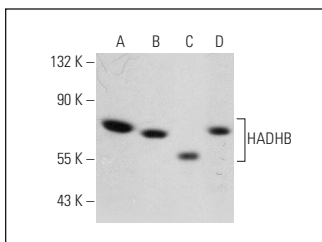
Molecular Weight of HADHB: 52 kDa.

Positive Controls: L8 cell lysate: sc-3807, H4 cell lysate: sc-2408 or K-562 whole cell lysate: sc-2203.

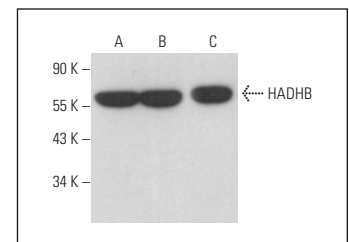
RECOMMENDED SUPPORT REAGENTS

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.

DATA



HADHB (D-10): sc-365907. Western blot analysis of HADHB expression in K-562 (A), H4 (B) and L8 (C) whole cell lysates and HeLa nuclear extract (D).



HADHB (D-10): sc-365907. Western blot analysis of HADHB expression in K-562 (A), HEL 92.1.7 (B) and SJRH30 (C) whole cell lysates.

STORAGE

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

RESEARCH USE

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