

# HADHB (C-6): sc-271496

## BACKGROUND

HADHB (trifunctional enzyme subunit  $\beta$  (mitochondrial), acetyl-CoA acyltransferase) is a 474 amino acid protein encoded by the human gene HADHB. HADHB belong 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 2-step reaction involving a covalent intermediate formed with a catalytic cysteine. HADHB is found in the mitochondrion as an octamer of four  $\alpha$  (HADHA) and four  $\beta$  (HADHB) subunits. Defects in HADHB are a cause of trifunctional protein deficiency (TFP deficiency). The clinical manifestations are very 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.

## CHROMOSOMAL LOCATION

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

## SOURCE

HADHB (C-6) is a mouse monoclonal antibody raised against amino acids 1-290 mapping at the N-terminus of HADHB of human origin.

## PRODUCT

Each vial contains 200  $\mu$ g IgG<sub>1</sub> 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.

## PROTOCOLS

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

## APPLICATIONS

HADHB (C-6) 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  $\mu$ g per 100-500  $\mu$ 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 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.

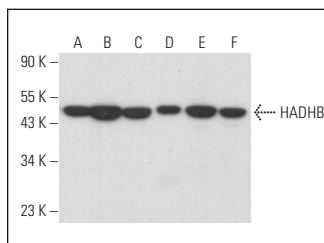
Molecular Weight of HADHB: 52 kDa.

Positive Controls: SJRH30 cell lysate: sc-2287, Jurkat whole cell lysate: sc-2204 or CCRF-CEM cell lysate: sc-2225.

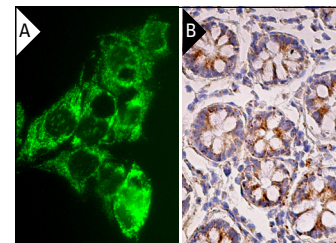
## RECOMMENDED SUPPORT REAGENTS

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. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgG $\kappa$  BP-FITC: sc-516140 or m-IgG $\kappa$  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 $\kappa$  BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

## DATA



HADHB (C-6): sc-271496. Western blot analysis of HADHB expression in Jurkat (A), SJRH30 (B), CCRF-CEM (C), 3T3-L1 (D), C2C12 (E) and BC<sub>3</sub>H1 (F) whole cell lysates.



HADHB (C-6): sc-271496. Immunofluorescence staining of formalin-fixed Hep G2 cells showing mitochondrial localization (A). Immunoperoxidase staining of formalin fixed, paraffin-embedded human colon tissue showing cytoplasmic staining of glandular cells (B).

## SELECT PRODUCT CITATIONS

- Kao, Y.T., et al. 2015. Japanese encephalitis virus nonstructural protein NS5 interacts with mitochondrial trifunctional protein and impairs fatty acid  $\beta$ -oxidation. *PLoS Pathog.* 11: e1004750.
- Li, X.X., et al. 2018. Nuclear receptor Nur77 facilitates melanoma cell survival under metabolic stress by protecting fatty acid oxidation. *Mol. Cell* 69: 480-492.e7.
- Huang, L., et al. 2018. Aberrant fatty acid metabolism in skeletal muscle contributes to Insulin resistance in zinc transporter 7 (*znt7*)-knockout mice. *J. Biol. Chem.* 293: 7549-7563.

## RESEARCH USE

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