SANTA CRUZ BIOTECHNOLOGY, INC.

HADHA (G-9): sc-515278

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

HADHA (trifunctional enzyme subunit α, mitochondrial), also known as TP-α, is the 763 amino acid α subunit of the mitochondrial trifunctional protein, which catalyzes the last three steps of mitochondrial β-oxidation of long chain fatty acids. This mitochondrial complex is complosed of four α (HADHA) and four β (HADHB) subunits, and the α subunit (HADHA) is responsible for catalyzing the 3-hydroxyacyl-CoA dehydrogenase and enoyl-CoA hydratase activities. Mutations in the HADHA gene can lead to long-chain 3-hydroxyacyl-coenzyme A dehydrogenase (LCHAD) deficiency or mitochondrial trifunctional protein deficiency. LCHAD deficiency is characterized by a deficiency of the dehydrogenase activity with normal hydratase activity and moderately decreased thiolase activity. In mitochondrial trifunctional protein deficiency, all three activities of the protein, dehydrogenase, hydratase, and thiolase, are deficient.

REFERENCES

1. 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.
2. Spiekerkoetter, U., et al. 2002. Uniparental disomy of chromosome 2 resulting in lethal trifunctional protein deficiency due to homozygous α-subunit mutations. Hum. Mutat. 20: 447-451.
3. 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.
4. Kao, H.J., et al. 2006. ENU mutagenesis identifies mice with cardiac fibrosis and hepatic steatosis caused by a mutation in the mitochondrial trifunctional protein β-subunit. Hum. Mol. Genet. 15: 3569-3577.
5. Lee, H.S., et al. 2006. Identification of a novel single nucleotide polymorphism of HADHA gene at a referred primer-binding site during prediagnostic tests for preimplantation genetic diagnosis. J. Korean Med. Sci. 21: 794-799.

CHROMOSOMAL LOCATION

Genetic locus: HADHA (human) mapping to 2p23.3.

SOURCE

HADHA (G-9) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 420-436 within an internal region of HADHA of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2b} 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-515278 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

APPLICATIONS

HADHA (G-9) is recommended for detection of HADHA of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg 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).

Suitable for use as control antibody for HADHA siRNA (h): sc-75220, HADHA shRNA Plasmid (h): sc-75220-SH and HADHA shRNA (h) Lentiviral Particles: sc-75220-V.

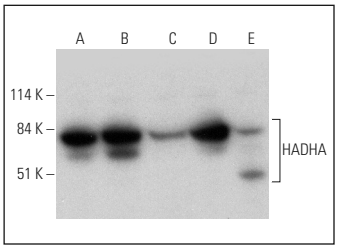
Molecular Weight of HADHA: 83 kDa.

Positive Controls: Ramos cell lysate: sc-2216, Jurkat whole cell lysate: sc-2204 or A-431 whole cell lysate: sc-2201.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), 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 m-IgGκ BP-FITC: sc-516140 or m-IgGκ 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



HADHA (G-9): sc-515278. Western blot analysis of HADHA expression in Ramos (**A**), Jurkat (**B**), MOLT-4 (**C**), Raji (**D**) and A-431 (**E**) 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.

PROTOCOLS

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