# HADHA (H-283): sc-292195



The Power to Question

#### **BACKGROUND**

HADHA (trifunctional enzyme subunit  $\alpha$ , mitochondrial), also known as TP- $\alpha$ , is the 763 amino acid  $\alpha$  subunit of the mitochondrial trifunctional protein, which catalyzes the last 3 steps of mitochondrial  $\beta$ -oxidation of long chain fatty acids. This mitochondrial complex is complosed of four  $\alpha$  (HADHA) and four  $\beta$  (HADHB) subunits, and the  $\alpha$  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.

## CHROMOSOMAL LOCATION

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

#### **SOURCE**

HADHA (H-283) is a rabbit polyclonal antibody raised against amino acids 481-763 mapping at the C-terminus of HADHA of human origin.

## **PRODUCT**

Each vial contains 200  $\mu g$  lgG 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.

## **RESEARCH USE**

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

### **APPLICATIONS**

HADHA (H-283) is recommended for detection of HADHA of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

HADHA (H-283) is also recommended for detection of HADHA in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for HADHA siRNA (h): sc-75220, HADHA siRNA (m): sc-75221, HADHA shRNA Plasmid (h): sc-75220-SH, HADHA shRNA Plasmid (m): sc-75221-SH, HADHA shRNA (h) Lentiviral Particles: sc-75220-V and HADHA shRNA (m) Lentiviral Particles: sc-75221-V.

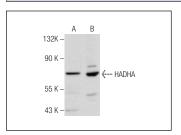
Molecular Weight of HADHA: 83 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200 or Ramos cell lysate: sc-2216.

#### **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**



HADHA (H-283): sc-292195. Western blot analysis of HADHA expression in HeLa (**A**) and Ramos (**B**) whole cell lysates.

## **PROTOCOLS**

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



Try HADHA (E-8): sc-374497 or HADHA (G-9): sc-515278, our highly recommended monoclonal alternatives to HADHA (H-283).

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