

HADHA (E-8): sc-374497

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 composed 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.

CHROMOSOMAL LOCATION

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

SOURCE

HADHA (E-8) is a mouse monoclonal antibody raised against amino acids 481-763 mapping at the C-terminus of HADHA 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.

HADHA (E-8) is available conjugated to agarose (sc-374497 AC), 500 μ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-374497 HRP), 200 μ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-374497 PE), fluorescein (sc-374497 FITC), Alexa Fluor[®] 488 (sc-374497 AF488), Alexa Fluor[®] 546 (sc-374497 AF546), Alexa Fluor[®] 594 (sc-374497 AF594) or Alexa Fluor[®] 647 (sc-374497 AF647), 200 μ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor[®] 680 (sc-374497 AF680) or Alexa Fluor[®] 790 (sc-374497 AF790), 200 μ g/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

HADHA (E-8) is recommended for detection of HADHA 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), 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 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.

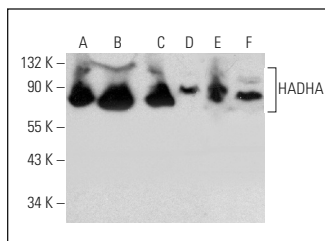
Molecular Weight of HADHA: 83 kDa.

Positive Controls: Ramos cell lysate: sc-2216, Jurkat whole cell lysate: sc-2204 or MOLT-4 cell lysate: sc-2233.

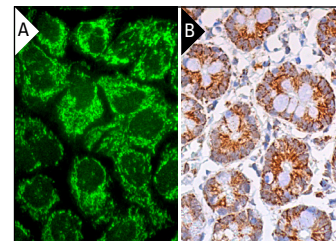
STORAGE

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

DATA



HADHA (E-8): sc-374497. Western blot analysis of HADHA expression in Ramos (A), Jurkat (B), MOLT-4 (C), 3T3-L1 (D), c4 (E) and PC-12 (F) whole cell lysates.



HADHA (E-8): sc-374497. Immunofluorescence staining of formalin-fixed A-431 cells showing mitochondrial localization (A). Immunoperoxidase staining of formalin fixed, paraffin-embedded human duodenum tissue showing cytoplasmic staining of glandular cells (B).

SELECT PRODUCT CITATIONS

- Shi, T., et al. 2013. Novel proteins associated with human dilated cardiomyopathy: selective reduction in α_1A -adrenergic receptors and increased desensitization proteins. *J. Recept. Signal Transduct. Res.* 33: 96-106.
- Kao, Y.T., et al. 2015. Japanese encephalitis virus nonstructural protein NS5 interacts with mitochondrial trifunctional protein and impairs fatty acid β -oxidation. *PLoS Pathog.* 11: e1004750.
- Han, S., et al. 2019. CPT1A/2-mediated FAO enhancement—a metabolic target in radioresistant breast cancer. *Front. Oncol.* 9: 1201.
- Santarelli, R., et al. 2020. KSHV dysregulates bulk macroautophagy, mitophagy and UPR to promote endothelial to mesenchymal transition and CCL2 release, key events in viral-driven sarcomagenesis. *Int. J. Cancer* 147: 3500-3510.
- Ow, J.R., et al. 2020. Remodeling of whole-body lipid metabolism and a diabetic-like phenotype caused by loss of CDK1 and hepatocyte division. *Elife* 9: e63835.
- Romeo, M.A., et al. 2021. p53-R273H sustains Ros, pro-inflammatory cytokine release and mTOR activation while reducing autophagy, mitophagy and UCP2 expression, effects prevented by wtp53. *Biomolecules* 11: 344.
- Amen, T. and Kaganovich, D. 2021. Stress granules inhibit fatty acid oxidation by modulating mitochondrial permeability. *Cell Rep.* 35: 109237.
- Gilardini Montani, M.S., et al. 2022. p62/SQSTM1 promotes mitophagy and activates the NRF2-mediated antioxidant and anti-inflammatory response restraining EBV-driven B lymphocyte proliferation. *Carcinogenesis* 43: 277-287.

RESEARCH USE

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

Alexa Fluor[®] is a trademark of Molecular Probes, Inc., Oregon, USA