



NDUFA1 (G-13): sc-131623

BACKGROUND

NDUFA1 (NADH dehydrogenase (ubiquinone) 1 α subcomplex, 1), also known as ZNF183 or MWFE, is one of about 45 subunits comprising complex I of the oxidative phosphorylation electron transport chain. Consisting of 70 amino acids and expressed predominantly in skeletal muscle and heart, NDUFA1 functions as an accessory subunit of the multi-protein mitochondrial membrane respiratory chain NADH dehydrogenase complex (known as complex I). complex I plays an important role in the transfer of electrons from NADH to the respiratory chain, a process that is essential for cellular respiration. As a single-pass membrane protein, NDUFA1 localizes to the matrix side of the mitochondrial inner membrane and is a member of the complex I NDUFA1 subunit family. The gene encoding NDUFA1 maps to human chromosome Xq24 and mouse chromosome X A2. NDUFA1 defects may cause mitochondrial Complex I deficiency, a mitochondrial disorder with wide symptoms ranging from cardiomyopathy, myopathy, liver disease and neurological disorders.

REFERENCES

1. Zhuchenko, O., et al. 1996. Isolation, mapping, and genomic structure of an X-linked gene for a subunit of human mitochondrial complex I. *Genomics* 37: 281-288.
2. Au, H.C., et al. 1999. The NDUFA1 gene product (MWFE protein) is essential for activity of complex I in mammalian mitochondria. *Proc. Natl. Acad. Sci. USA* 96: 4354-4359.
3. Gaikwad, J.S., et al. 2001. Identification of tooth-specific downstream targets of Runx2. *Gene* 279: 91-97.
4. Yadava, N., et al. 2002. Species-specific and mutant MWFE proteins. Their effect on the assembly of a functional mammalian mitochondrial complex I. *J. Biol. Chem.* 277: 21221-21230.
5. Qi, X., et al. 2004. SOD2 gene transfer protects against optic neuropathy induced by deficiency of complex I. *Ann. Neurol.* 56: 182-191.
6. Fernandez-Moreira, D., et al. 2007. X-linked NDUFA1 gene mutations associated with mitochondrial encephalomyopathy. *Ann. Neurol.* 61: 73-83.
7. Online Mendelian Inheritance in Man, OMIM™. 2007. Johns Hopkins University, Baltimore, MD. MIM Number: 300078. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
8. Pagliarini, D.J., et al. 2008. A mitochondrial protein compendium elucidates complex I disease biology. *Cell* 134: 112-123.
9. Online Mendelian Inheritance in Man, OMIM™. 2010. Johns Hopkins University, Baltimore, MD. MIM Number: 252010. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>

CHROMOSOMAL LOCATION

Genetic locus: *Ndufa1* (mouse) mapping to X A2.

STORAGE

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

SOURCE

NDUFA1 (G-13) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of NDUFA1 of mouse origin.

PRODUCT

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

Blocking peptide available for competition studies, sc-131623 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

NDUFA1 (G-13) is recommended for detection of NDUFA1 of mouse and rat origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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); non cross-reactive with other NDUFA family members.

Suitable for use as control antibody for NDUFA1 siRNA (m): sc-149866, NDUFA1 shRNA Plasmid (m): sc-149866-SH and NDUFA1 shRNA (m) Lentiviral Particles: sc-149866-V.

Molecular Weight of NDUFA1: 8 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (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) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

RESEARCH USE

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

PROTOCOLS

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