SANTA CRUZ BIOTECHNOLOGY, INC.

NDUFS2 (E-18): sc-109594

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The Power to Question

BACKGROUND

Located in the mitochondrial inner membrane, mitochondrial complex I is the first and largest enzyme in the electron transport chain of oxidative phosphorylation. By oxidizing NADH that is produced in the Krebs cycle, this complex utilizes the two electrons to reduce ubiquinone to ubiquinol, thereby initiating the passage of electrons to successive complexes and ultimately leading to the reduction of oxygen to water. Mitochondrial complex I consists of over 40 subunits and is of considerable clinical interest since defects in any of the subunits can lead to various myopathies and neuropathies. As a subunit of mitochondrial complex I, NDUFS2 (NADH dehydrogenase [ubiquinone] iron-sulfur protein 2), also designated NADH-ubiquinone oxidoreductase 49 kDa subunit, is a 463 amino acid protein that is suggested to be required for catalytic activity. Defects in the gene encoding NDUFS2 are the cause of complex I mitochondrial respiratory chain deficiency, which is characterized by a variety of symptoms including liver failure, cardiomyopathy and neurodegeneration.

REFERENCES

1. Procaccio, V., et al. 1998. Mapping to 1q23 of the human gene (NDUFS2) encoding the 49-kDa subunit of the mitochondrial respiratory complex I and immunodetection of the mature protein in mitochondria. Mamm. Genome 9: 482-484.
2. Smeitink, J. and van den Heuvel, L. 1999. Human mitochondrial complex I in health and disease. Am. J. Hum. Genet. 64: 1505-1510.
3. Loeffen, J., et al. 2001. Mutations in the complex I NDUFS2 gene of patients with cardiomyopathy and encephalomyopathy. Ann. Neurol. 49: 195-201.
4. Bugiani, M., et al. 2004. Clinical and molecular findings in children with complex I deficiency. Biochim. Biophys. Acta 1659: 136-147.
5. Ugalde, C., et al. 2004. Differences in assembly or stability of complex I and other mitochondrial OXPHOS complexes in inherited complex I deficiency. Hum. Mol. Genet. 13: 659-667.
6. Visch, H.J., et al. 2004. Inhibition of mitochondrial Na^+-Ca^{2+} exchange restores agonist-induced ATP production and Ca^{2+} handling in human complex I deficiency. J. Biol. Chem. 279: 40328-40336.
7. Online Mendelian Inheritance in Man, OMIM™. 2006. Johns Hopkins University, Baltimore, MD. MIM Number: 602985. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/
8. Lazarou, M., et al. 2007. Analysis of the assembly profiles for mitochondrial- and nuclear-DNA-encoded subunits into complex I. Mol. Cell. Biol. 27: 4228-4237.

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 or our catalog for detailed protocols and support products.

CHROMOSOMAL LOCATION

Genetic locus: NDUFS2 (human) mapping to 1q23.3; Ndufs2 (mouse) mapping to 1 H3.

SOURCE

NDUFS2 (E-18) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of NDUFS2 of human origin.

PRODUCT

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

Blocking peptide available for competition studies, sc-109594 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

NDUFS2 (E-18) is recommended for detection of NDUFS2 of mouse, rat and human 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 NDUFS family members.

NDUFS2 (E-18) is also recommended for detection of NDUFS2 in additional species, including equine, bovine and porcine.

Suitable for use as control antibody for NDUFS2 siRNA (h): sc-78903, NDUFS2 siRNA (m): sc-106290, NDUFS2 shRNA Plasmid (h): sc-78903-SH, NDUFS2 shRNA Plasmid (m): sc-106290-SH, NDUFS2 shRNA (h) Lentiviral Particles: sc-78903-V and NDUFS2 shRNA (m) Lentiviral Particles: sc-106290-V.

Molecular Weight of NDUFS2: 49 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.