

NDUFS7 (H-57): sc-98644

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, NDUFS7 (NADH dehydrogenase [ubiquinone] iron-sulfur protein 7), also designated NADH-ubiquinone oxidoreductase 20 kDa subunit, is a 213 amino acid protein that is suggested to be required for catalytic activity. Defects in the gene encoding NDUFS7 are the cause of Leigh syndrome, a severe neurological disorder that is characterized by bilaterally symmetrical necrotic lesions in subcortical brain regions.

REFERENCES

1. Hyslop, S.J., et al. 1996. Assignment of the PSST subunit gene of human mitochondrial complex I to chromosome 19p13. *Genomics* 37: 375-380.
2. Smeitink, J., et al. 1999. Human mitochondrial complex I in health and disease. *Am. J. Hum. Genet.* 64: 1505-1510.
3. Triepels, R.H., et al. 1999. Leigh syndrome associated with a mutation in the NDUFS7 (PSST) nuclear encoded subunit of complex I. *Ann. Neurol.* 45: 787-790.
4. Bugiani, M., et al. 2004. Clinical and molecular findings in children with complex I deficiency. *Biochim. Biophys. Acta* 1659: 136-147.
5. Visch, H.J., et al. 2004. Inhibition of mitochondrial Na⁺-Ca²⁺ exchange restores agonist-induced ATP production and Ca²⁺ handling in human complex I deficiency. *J. Biol. Chem.* 279: 40328-40336.
6. Lebon, S., et al. 2007. A novel mutation in the human complex I NDUFS7 subunit associated with Leigh syndrome. *Mol. Genet. Metab.* 90: 379-382.

CHROMOSOMAL LOCATION

Genetic locus: NDUFS7 (human) mapping to 19p13.3; Ndufs7 (mouse) mapping to 10 C1.

SOURCE

NDUFS7 (H-57) is a rabbit polyclonal antibody raised against amino acids 133-189 mapping within an internal region of NDUFS7 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.

STORAGE

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

APPLICATIONS

NDUFS7 (H-57) is recommended for detection of NDUFS7 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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).

NDUFS7 (H-57) is also recommended for detection of NDUFS7 in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for NDUFS7 siRNA (h): sc-97909, NDUFS7 siRNA (m): sc-149889, NDUFS7 shRNA Plasmid (h): sc-97909-SH, NDUFS7 shRNA Plasmid (m): sc-149889-SH, NDUFS7 shRNA (h) Lentiviral Particles: sc-97909-V and NDUFS7 shRNA (m) Lentiviral Particles: sc-149889-V.

Molecular Weight of NDUFS7: 20 kDa.

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.

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.