

NDUFS6 (V-13): sc-107275

BACKGROUND

NDUFC1 (NADH-ubiquinone oxidoreductase iron-sulfur protein 6), also designated complex I-13kD-A, is one of 45 subunits comprising complex I of the oxidative phosphorylation electron transport chain. The multi-subunit NADH: ubiquinone oxidoreductase (complex I) is the first enzyme complex in the electron transport chain of the mitochondria. Complex I deficiency is the most common respiratory chain defect, resulting in various combinations of cardiac, hepatic and renal disorders. Through use of chaotropic agents, complex I can be separated into three different fractions: a flavoprotein fraction, a hydrophobic protein (HP) fraction and an iron-sulfur protein (IP) fraction. NDUFS6 is a 124 amino acid protein that belongs to the iron-sulfur protein fraction. Mutations in the NDUFS6 gene may contribute to the deleterious effects of complex I deficiency.

REFERENCES

1. Chow, W., et al. 1991. Determination of the cDNA sequence for the human mitochondrial 75-kDa Fe-S protein of NADH-coenzyme Q reductase. *Eur. J. Biochem.* 201: 547-50.
2. Duncan, A.M., et al. 1992. Localization of the human 75-kDa Fe-S protein of NADH-coenzyme Q reductase gene (NDUFS1) to 2q33-q34. *Cytogenet. Cell Genet.* 60: 212-213.
3. Kirby, D.M., et al. 2004. NDUFS6 mutations are a novel cause of lethal neonatal mitochondrial complex I deficiency. *J. Clin. Invest.* 114: 837-845.
4. Stojanovski, D., et al. 2004. Levels of human Fis1 at the mitochondrial outer membrane regulate mitochondrial morphology. *J. Cell Sci.* 117: 1201-1210.
5. Karahan, O.I., et al. 2005. Ultrasound evaluation of peritoneal catheter tunnel in catheter related infections in CAPD. *Int. Urol. Nephrol.* 37: 363-366.
6. Martin, M.A., et al. 2005. Leigh syndrome associated with mitochondrial complex I deficiency due to a novel mutation in the NDUFS1 gene. *Arch. Neurol.* 62: 659-661.
7. Smeitink, J.A., et al. 2005. Cell biological consequences of mitochondrial NADH: ubiquinone oxidoreductase deficiency. *Curr. Neurovas. Res.* 1: 29-40.
8. Sparks, L.M., et al. 2005. A high-fat diet coordinately downregulates genes required for mitochondrial oxidative phosphorylation in skeletal muscle. *Diabetes* 54: 1926-1933.
9. Flemming, D., et al. 2006. A ubiquinone oxidoreductase (complex I). *J. Mol. Microbiol. Biotechnol.* 10: 208-222.

CHROMOSOMAL LOCATION

Genetic locus: NDUFS6 (human) mapping to 5p15.33.

SOURCE

NDUFS6 (V-13) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of NDUFS6 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-107275 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

NDUFS6 (V-13) is recommended for detection of NDUFS6 of 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.

NDUFS6 (V-13) is also recommended for detection of NDUFS6 in additional species, including equine, bovine and porcine.

Suitable for use as control antibody for NDUFS6 siRNA (h): sc-91874, NDUFS6 shRNA Plasmid (h): sc-91874-SH and NDUFS6 shRNA (h) Lentiviral Particles: sc-91874-V.

Molecular Weight of NDUFS6: 13 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.

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.

PROTOCOLS

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