

NDUFV1 (H-23): sc-133808

BACKGROUND

Complex 1 (also known as NADH dehydrogenase) of the electron transport chain (respiratory chain) is an enzymatic complex that catalyzes the transfer of electrons from NADH to ubiquinone. Free energy from the reaction is conserved in the transfer of protons into the inter-membrane space to create an electrochemical proton gradient, a driving force for ATP synthesis. Complex 1 is an extremely complicated, multi-protein, L-shaped complex composed of 45 different subunits located in the mitochondrial inner-membrane. NDUFV1 (NADH dehydrogenase ubiquinone flavoprotein 1), also known as UQOR1 or CI-51kD, is a 464 amino acid core subunit protein of complex 1 that is essential for the electron transport chain. The peripheral membrane protein, NDUFV1, contains a highly conserved NADH-binding site and is localized to the matrix side of the inner membrane. Defects in the gene encoding NDUFV1 can cause complex 1 deficiency in humans, which can lead to Leigh syndrome (LS), a severe neurological disorder characterized by lesions in the subcortical brain region.

REFERENCES

- Spencer, S.R., et al. 1992. The human mitochondrial NADH: ubiquinone oxidoreductase 51 kDa subunit maps adjacent to the glutathione S-transferase P1-1 gene on chromosome 11q13. *Genomics* 14: 1116-1118.
- Ali, S.T., et al. 1993. Chromosomal localization of the human gene encoding the 51 kDa subunit of mitochondrial complex I (NDUFV1) to 11q13. *Genomics* 18: 435-439.
- Schuelke, M., et al. 1998. Cloning of the human mitochondrial 51 kDa subunit (NDUFV1) reveals a 100% antisense homology of its 3'UTR with the 5'UTR of the γ -interferon inducible protein (IP-30) precursor: is this a link between mitochondrial myopathy and inflammation? *Biochem. Biophys. Res. Commun.* 245: 599-606.
- Schuelke, M., et al. 1999. Mutant NDUFV1 subunit of mitochondrial complex I causes leukodystrophy and myoclonic epilepsy. *Nat. Genet.* 21: 260-261.
- de Coo, R.F., et al. 1999. The structure of the human NDUFV1 gene encoding the 51 kDa subunit of mitochondrial complex I. *Mamm. Genome* 10: 49-53.
- Bénit, P., et al. 2001. Large-scale deletion and point mutations of the nuclear NDUFV1 and NDUF51 genes in mitochondrial complex I deficiency. *Am. J. Hum. Genet.* 68: 1344-1352.

CHROMOSOMAL LOCATION

Genetic locus: NDUFV1 (human) mapping to 11q13.2; Ndufv1 (mouse) mapping to 19 A.

SOURCE

NDUFV1 (H-23) is a Protein A purified rabbit polyclonal antibody raised against synthetic NDUFV1 peptide of human origin.

PRODUCT

Each vial contains 100 μ g IgG in 1.0 ml PBS with < 0.1% sodium azide, 0.1% gelatin and < 0.02% sucrose.

APPLICATIONS

NDUFV1 (H-23) is recommended for detection of NDUFV1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for NDUFV1 siRNA (h): sc-96604, NDUFV1 siRNA (m): sc-149891, NDUFV1 shRNA Plasmid (h): sc-96604-SH, NDUFV1 shRNA Plasmid (m): sc-149891-SH, NDUFV1 shRNA (h) Lentiviral Particles: sc-96604-V and NDUFV1 shRNA (m) Lentiviral Particles: sc-149891-V.

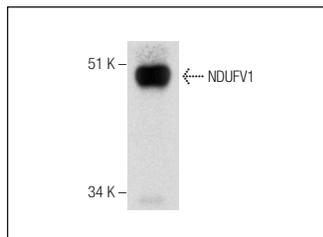
Molecular Weight of NDUFV1: 51 kDa.

Positive Controls: A-431 whole cell lysate: sc-2201, human heart extract: sc-363763 or Hep G2 cell lysate: sc-2227.

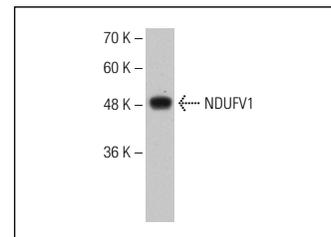
RECOMMENDED SECONDARY REAGENTS

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).

DATA



NDUFV1 (H-23): sc-133808. Western blot analysis of NDUFV1 expression in human heart tissue extract.



NDUFV1 (H-23): sc-133808. Western blot analysis of NDUFV1 expression in Hep G2 whole cell lysate.

SELECT PRODUCT CITATIONS

- García-Ruiz, I., et al. 2013. Pioglitazone leads to an inactivation and disassembly of complex I of the mitochondrial respiratory chain. *BMC Biol.* 11: 88.

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.