

NDUFS4 (1-E-4): sc-100567

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 intermembrane space to create an electrochemical proton gradient, a driving force for ATP synthesis. Complex 1 is a complicated, multi-protein, L-shaped complex composed of at least 45 different subunits and located in the mitochondrial inner membrane. NDUFS4 (NADH dehydrogenase (ubiquinone) Fe-S protein 4), also known as AQQDQ or CI-18 (complex I-18kDa protein), belongs to the complex I NDUFS4 subunit family. NDUFS4 localizes to the matrix side of the inner membrane of the mitochondrion and functions as an accessory subunit of complex I. Mutations in the gene encoding NDUFS4 can result in complex I mitochondrial respiratory chain deficiency. Patients with this deficiency may exhibit cardiomyopathy, myopathy, liver failure and neurological disorders.

REFERENCES

1. Papa, S., et al. 1996. The nuclear-encoded 18 kDa (IP) AQQDQ subunit of bovine heart complex I is phosphorylated by the mitochondrial cAMP-dependent protein kinase. *FEBS Lett.* 379: 299-301.
2. Budde, S.M., et al. 2000. Combined enzymatic complex I and III deficiency associated with mutations in the nuclear encoded NDUFS4 gene. *Biochem. Biophys. Res. Commun.* 275: 63-68.

CHROMOSOMAL LOCATION

Genetic locus: NDUFS4 (human) mapping to 5q11.2; Ndufs4 (mouse) mapping to 13 D2.2.

SOURCE

NDUFS4 (1-E-4) is a mouse monoclonal antibody raised against recombinant NDUFS4 of human origin.

PRODUCT

Each vial contains 100 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

NDUFS4 (1-E-4) is recommended for detection of NDUFS4 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), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for NDUFS4 siRNA (h): sc-75892, NDUFS4 siRNA (m): sc-75893, NDUFS4 shRNA Plasmid (h): sc-75892-SH, NDUFS4 shRNA Plasmid (m): sc-75893-SH, NDUFS4 shRNA (h) Lentiviral Particles: sc-75892-V and NDUFS4 shRNA (m) Lentiviral Particles: sc-75893-V.

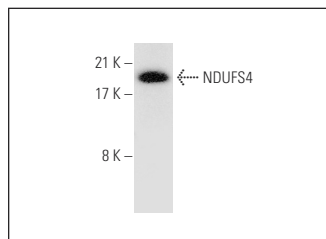
Molecular Weight of NDUFS4: 18 kDa.

Positive Controls: mouse kidney extract: sc-2255.

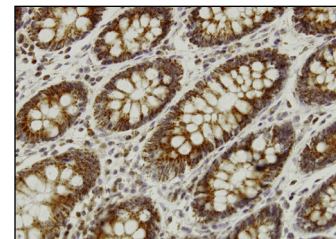
STORAGE

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

DATA



NDUFS4 (1-E-4): sc-100567. Western blot analysis of NDUFS4 expression in mouse kidney tissue extract.



NDUFS4 (1-E-4): sc-100567. Immunoperoxidase staining of formalin-fixed, paraffin-embedded human colon tissue showing cytoplasmic localization.

SELECT PRODUCT CITATIONS

1. Schlehe, J.S., et al. 2013. The mitochondrial disease associated protein Ndufaf2 is dispensable for complex-1 assembly but critical for the regulation of oxidative stress. *Neurobiol. Dis.* 58: 57-67.
2. Liu, H.F., et al. 2017. Combined LRRK2 mutation, aging and chronic low dose oral rotenone as a model of Parkinson's disease. *Sci. Rep.* 7: 40887.
3. Song, L., et al. 2017. Bipolar cell reduction precedes retinal ganglion neuron loss in a complex 1 knockout mouse model. *Brain Res.* 1657: 232-244.
4. Najjar, Y.G., et al. 2019. Tumor cell oxidative metabolism as a barrier to PD-1 blockade immunotherapy in melanoma. *JCI Insight* 4: e124989.
5. Gospe, S.M., et al. 2019. Photoreceptors in a mouse model of Leigh syndrome are capable of normal light-evoked signaling. *J. Biol. Chem.* 294: 12432-12443.
6. Wang, L., et al. 2020. Progressive optic atrophy in a retinal ganglion cell-specific mouse model of complex I deficiency. *Sci. Rep.* 10: 16326.
7. Shil, S.K., et al. 2021. NDUFS4 ablation decreases synaptophysin expression in hippocampus. *Sci. Rep.* 11: 10969.
8. Deng, Z., et al. 2021. Selective autophagy of AKAP11 activates cAMP/PKA to fuel mitochondrial metabolism and tumor cell growth. *Proc. Natl. Acad. Sci. USA* 118: e2020215118.
9. Scharping, N.E., et al. 2021. Mitochondrial stress induced by continuous stimulation under hypoxia rapidly drives T cell exhaustion. *Nat. Immunol.* 22: 205-215.

RESEARCH USE

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

PROTOCOLS

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