

NDUFS4 (A-7): sc-514002

BACKGROUND

Complex I (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 I 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 AQDQ or Cl-18 (complex I-18 kDa 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) AQDQ subunit of bovine heart complex I is phosphorylated by the mitochondrial cAMP-dependent protein kinase. *FEBS Lett.* 379: 299-301.
2. Online Mendelian Inheritance in Man, OMIM™. 1998. Johns Hopkins University, Baltimore, MD. MIM Number: 602694. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
3. 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.
4. Scacco, S., et al. 2006. Mutations in structural genes of complex I associated with neurological diseases. *Ital. J. Biochem.* 55: 254-262.
5. Piccoli, C., et al. 2006. cAMP controls oxygen metabolism in mammalian cells. *FEBS Lett.* 580: 4539-4543.

CHROMOSOMAL LOCATION

Genetic locus: NDUFS4 (human) mapping to 5q11.2.

SOURCE

NDUFS4 (A-7) is a mouse monoclonal antibody raised against amino acids 1-175 representing full length NDUFS4 of human origin.

PRODUCT

Each vial contains 200 µg IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

NDUFS4 (A-7) is available conjugated to agarose (sc-514002 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-514002 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-514002 PE), fluorescein (sc-514002 FITC), Alexa Fluor® 488 (sc-514002 AF488), Alexa Fluor® 546 (sc-514002 AF546), Alexa Fluor® 594 (sc-514002 AF594) or Alexa Fluor® 647 (sc-514002 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-514002 AF680) or Alexa Fluor® 790 (sc-514002 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

NDUFS4 (A-7) is recommended for detection of NDUFS4 of human origin by Western Blotting (starting dilution 1:100, 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).

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

Molecular Weight of NDUFS4: 18 kDa.

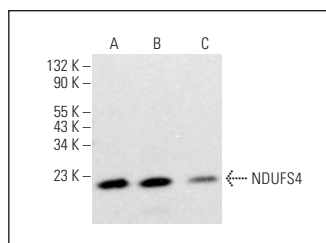
Positive Controls: A-431 whole cell lysate: sc-2201, K-562 whole cell lysate: sc-2203 or A-673 cell lysate: sc-2414.

RECOMMENDED SUPPORT REAGENTS

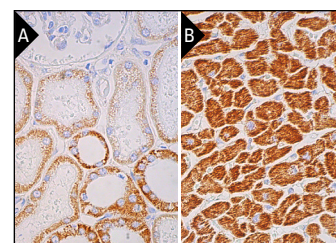
To ensure optimal results, the following support reagents are recommended:

- 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 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 m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



NDUFS4 (A-7): sc-514002. Western blot analysis of NDUFS4 expression in A-431 (A), K-562 (B) and A-673 (C) whole cell lysates.



NDUFS4 (A-7): sc-514002. Immunoperoxidase staining of formalin fixed, paraffin-embedded human kidney tissue showing cytoplasmic staining of cells in tubules (A) and human heart muscle tissue showing cytoplasmic staining of myocytes (B). Blocked with 0.25X UltraCruz® Blocking Reagent: sc-516214. Detection reagents used: m-IgGκ BP-B: sc-516142 and ImmunoCruz ABC Kit: sc-516216.

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.

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