

# NDUFA1 siRNA (h): sc-90892

## BACKGROUND

NDUFA1 (NADH dehydrogenase (ubiquinone) 1  $\alpha$  subcomplex, 1), also known as ZNF183 or MWFE, is one of about 45 subunits comprising Complex I of the oxidative phosphorylation electron transport chain. Consisting of 70 amino acids and expressed predominantly in skeletal muscle and heart, NDUFA1 functions as an accessory subunit of the multi-protein mitochondrial membrane respiratory chain NADH dehydrogenase complex (known as Complex I). Complex I plays an important role in the transfer of electrons from NADH to the respiratory chain, a process that is essential for cellular respiration. As a single-pass membrane protein, NDUFA1 localizes to the matrix side of the mitochondrial inner membrane and is a member of the complex I NDUFA1 subunit family. The gene encoding NDUFA1 maps to human chromosome Xq24 and mouse chromosome X A2. NDUFA1 defects may cause mitochondrial Complex I deficiency, a mitochondrial disorder with wide symptoms ranging from cardiomyopathy, myopathy, liver disease and neurological disorders.

## REFERENCES

1. Zhuchenko, O., et al. 1996. Isolation, mapping, and genomic structure of an X-linked gene for a subunit of human mitochondrial complex I. *Genomics* 37: 281-288.
2. Au, H.C., et al. 1999. The NDUFA1 gene product (MWFE protein) is essential for activity of complex I in mammalian mitochondria. *Proc. Natl. Acad. Sci. USA* 96: 4354-4359.
3. Gaikwad, J.S., et al. 2001. Identification of tooth-specific downstream targets of Runx2. *Gene* 279: 91-97.
4. Yadava, N., et al. 2002. Species-specific and mutant MWFE proteins. Their effect on the assembly of a functional mammalian mitochondrial complex I. *J. Biol. Chem.* 277: 21221-21230.
5. Qi, X., et al. 2004. SOD2 gene transfer protects against optic neuropathy induced by deficiency of complex I. *Ann. Neurol.* 56: 182-191.
6. Fernandez-Moreira, D., et al. 2007. X-linked NDUFA1 gene mutations associated with mitochondrial encephalomyopathy. *Ann. Neurol.* 61: 73-83.
7. Pagliarini, D.J., et al. 2008. A mitochondrial protein compendium elucidates complex I disease biology. *Cell* 134: 112-123.
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## CHROMOSOMAL LOCATION

Genetic locus: NDUFA1 (human) mapping to Xq24.

## PRODUCT

NDUFA1 siRNA (h) is a target-specific 19-25 nt siRNA designed to knock down gene expression. Each vial contains 3.3 nmol of lyophilized siRNA, sufficient for a 10  $\mu$ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see NDUFA1 shRNA Plasmid (h): sc-90892-SH and NDUFA1 shRNA (h) Lentiviral Particles: sc-90892-V as alternate gene silencing products.

## STORAGE AND RESUSPENSION

Store lyophilized siRNA duplex at -20° C with desiccant. Stable for at least one year from the date of shipment. Once resuspended, store at -20° C, avoid contact with RNases and repeated freeze thaw cycles.

Resuspend lyophilized siRNA duplex in 330  $\mu$ l of the RNase-free water provided. Resuspension of the siRNA duplex in 330  $\mu$ l of RNase-free water makes a 10  $\mu$ M solution in a 10  $\mu$ M Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

## APPLICATIONS

NDUFA1 siRNA (h) is recommended for the inhibition of NDUFA1 expression in human cells.

## SUPPORT REAGENTS

For optimal siRNA transfection efficiency, Santa Cruz Biotechnology's siRNA Transfection Reagent: sc-29528 (0.3 ml), siRNA Transfection Medium: sc-36868 (20 ml) and siRNA Dilution Buffer: sc-29527 (1.5 ml) are recommended. Control siRNAs or Fluorescein Conjugated Control siRNAs are available as 10  $\mu$ M in 66  $\mu$ l. Each contain a scrambled sequence that will not lead to the specific degradation of any known cellular mRNA. Fluorescein Conjugated Control siRNAs include: sc-36869, sc-44239, sc-44240 and sc-44241. Control siRNAs include: sc-37007, sc-44230, sc-44231, sc-44232, sc-44233, sc-44234, sc-44235, sc-44236, sc-44237 and sc-44238.

## GENE EXPRESSION MONITORING

NDUFA1 (3B9-1A1): sc-517006 is recommended as a control antibody for monitoring of NDUFA1 gene expression knockdown by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) or immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker<sup>™</sup> Molecular Weight Standards: sc-2035, UltraCruz<sup>®</sup> Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use m-IgG $\kappa$  BP-FITC: sc-516140 or m-IgG $\kappa$  BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz<sup>®</sup> Mounting Medium: sc-24941 or UltraCruz<sup>®</sup> Hard-set Mounting Medium: sc-359850.

## RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor NDUFA1 gene expression knockdown using RT-PCR Primer: NDUFA1 (h)-PR: sc-90892-PR (20  $\mu$ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

## RESEARCH USE

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

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) for detailed protocols and support products.