

NDUFV1 siRNA (h): sc-96604

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

1. 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.
2. 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.
3. 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.
4. Schuelke, M., et al. 1999. Mutant NDUFV1 subunit of mitochondrial complex I causes leukodystrophy and myoclonic epilepsy. *Nat. Genet.* 21: 260-261.

CHROMOSOMAL LOCATION

Genetic locus: NDUFV1 (human) mapping to 11q13.2.

PRODUCT

NDUFV1 siRNA (h) is a pool of 2 target-specific 19-25 nt siRNAs designed to knock down gene expression. Each vial contains 3.3 nmol of lyophilized siRNA, sufficient for a 10 μ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see NDUFV1 shRNA Plasmid (h): sc-96604-SH and NDUFV1 shRNA (h) Lentiviral Particles: sc-96604-V as alternate gene silencing products.

For independent verification of NDUFV1 (h) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-96604A and sc-96604B.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support 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 μ l of the RNase-free water provided. Resuspension of the siRNA duplex in 330 μ l of RNase-free water makes a 10 μ M solution in a 10 μ M Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

NDUFV1 siRNA (h) is recommended for the inhibition of NDUFV1 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 μ M in 66 μ 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

NDUFV1 (14.8): sc-100566 is recommended as a control antibody for monitoring of NDUFV1 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 κ BP-HRP: sc-516102 or m-IgG κ 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) Immunofluorescence: use m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz[®] Mounting Medium: sc-24941 or UltraCruz[®] Hard-set Mounting Medium: sc-359850.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor NDUFV1 gene expression knockdown using RT-PCR Primer: NDUFV1 (h)-PR: sc-96604-PR (20 μ l, 590 bp). 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.