# SURF-1 siRNA (m): sc-63091



The Power to Question

#### **BACKGROUND**

The SURF-1 protein demonstrates a vital role in the assembly of complex IV (CIV or COX) of the mitochondrial respiratory chain. Expressed in the inner mitochondrial membrane, mutations of the SURF-1 gene generally cause cytochrome c oxidase complex IV deficiency. Shortage of complex IV leads to Leigh syndrome, a severe neurological disorder. Leigh syndrome patients are usually subject to rapidly progressive encephalopathy, characterized by necrotic lesions in subcortical brain regions. SURF-1 mutations correlate to high post-implantation embryonic lethality as well as early-onset mortality of post-natal individuals. Considerable deficit in muscle strength and motor performance is also a profound and isolated defect of SURF-1 activity in skeletal muscle and liver. Heart, brain and skeletal muscle morphological abnormalities frequently occur due to SURF-1 mutations.

# **REFERENCES**

- Tiranti, V., et al. 1998. Mutations of SURF-1 in Leigh disease associated with cytochrome c oxidase deficiency. Am. J. Hum. Genet. 63: 1609-1621.
- 2. Tiranti, V., et al. 1999. Characterization of SURF-1 expression and SURF-1p function in normal and disease conditions. Hum. Mol. Genet. 8: 2533-2540.
- Tiranti, V., et al. 1999. Loss-of-function mutations of SURF-1 are specifically associated with Leigh syndrome with cytochrome c oxidase deficiency. Ann. Neurol. 46: 161-166.
- 4. Vernon, E.G. and Gaston, K. 2000. Myc and YY1 mediate activation of the SURF-1 promoter in response to serum growth factors. Biochim. Biophys. Acta 492: 172-179.
- Sue, C.M., et al. 2000. Differential features of patients with mutations in two COX assembly genes, SURF-1 and SCO2. Ann. Neurol. 47: 589-595.
- Farina, L., et al. 2002. MR findings in Leigh syndrome with COX deficiency and SURF-1 mutations. AJNR Am. J. Neuroradiol. 23: 1095-1100.
- 7. Ogawa, Y., et al. 2002. Three novel SURF-1 mutations in Japanese patients with Leigh syndrome. Pediatr. Neurol. 26: 196-200.
- 8. Agostino, A., et al. 2006. Constitutive knockout of SURF-1 is associated with high embryonic lethality, mitochondrial disease and cytochrome c oxidase deficiency in mice. Hum. Mol. Genet. 12: 399-413.

# CHROMOSOMAL LOCATION

Genetic locus: Surf1 (mouse) mapping to 2 A3.

## **PRODUCT**

SURF-1 siRNA (m) is a pool of 3 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  $\mu$ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see SURF-1 shRNA Plasmid (m): sc-63091-SH and SURF-1 shRNA (m) Lentiviral Particles: sc-63091-V as alternate gene silencing products.

For independent verification of SURF-1 (m) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-63091A, sc-63091B and sc-63091C.

#### 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**

SURF-1 siRNA (m) is recommended for the inhibition of SURF-1 expression in mouse 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 µM in 66 µl. 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**

SURF-1 (H-7): sc-365159 is recommended as a control antibody for monitoring of SURF-1 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-lgG $\kappa$  BP-HRP: sc-516102 or m-lgG $\kappa$  BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker<sup>TM</sup> Molecular Weight Standards: sc-2035, UltraCruz<sup>®</sup> Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use m-lgG $\kappa$  BP-FITC: sc-516140 or m-lgG $\kappa$  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 SURF-1 gene expression knockdown using RT-PCR Primer: SURF-1 (m)-PR: sc-63091-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 for detailed protocols and support products.