

# SURF-1 (C-14): sc-55904

## 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

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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.
3. 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., et al. 2000. Myc and YY1 mediate activation of the SURF-1 promoter in response to serum growth factors. *Biochim. Biophys. Acta* 492: 172-179.
5. 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.
6. Farina, L., et al. 2002. MR findings in Leigh syndrome with COX deficiency and SURF-1 mutations. *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 (human) mapping to 9q34.2; Surf1 (mouse) mapping to 2 A3.

## SOURCE

SURF-1 (C-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of SURF-1 of human origin.

## PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-55904 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

SURF-1 (C-14) is recommended for detection of SURF-1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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).

SURF-1 (C-14) is also recommended for detection of SURF-1 in additional species, including equine, canine, bovine and avian.

Suitable for use as control antibody for SURF-1 siRNA (h): sc-63090, SURF-1 siRNA (m): sc-63091, SURF-1 shRNA Plasmid (h): sc-63090-SH, SURF-1 shRNA Plasmid (m): sc-63091-SH, SURF-1 shRNA (h) Lentiviral Particles: sc-63090-V and SURF-1 shRNA (m) Lentiviral Particles: sc-63091-V.

Molecular Weight of SURF-1: 31 kDa.

Positive Controls: human heart extract: sc-363763, mouse liver extract: sc-2256 or HeLa whole cell lysate: sc-2200.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

## 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.

## PROTOCOLS

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



Try **SURF-1 (H-7): sc-365159** or **SURF-1 (D-9): sc-166948**, our highly recommended monoclonal alternatives to SURF-1 (C-14).