

SURF-1 (H-170): sc-68345

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

1. 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.
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. 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.
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.

CHROMOSOMAL LOCATION

Genetic locus: SURF1 (human) mapping to 9q34.2; Surf1 (mouse) mapping to 2 A3.

SOURCE

SURF-1 (H-170) is a rabbit polyclonal antibody raised against amino acids 81-250 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.

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

APPLICATIONS

SURF-1 (H-170) 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), 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).

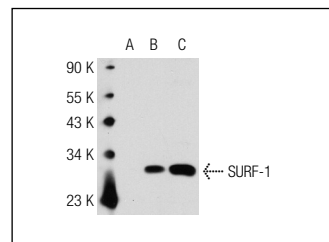
SURF-1 (H-170) is also recommended for detection of SURF-1 in additional species, including bovine, canine and equine.

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.

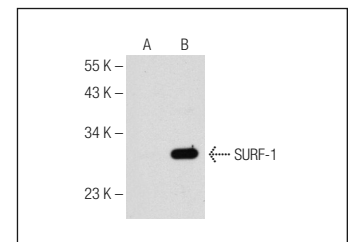
Molecular Weight of SURF-1: 31 kDa.

Positive Controls: SURF-1 (h): 293T Lysate: sc-176677, human heart extract: sc-363763 or HeLa whole cell lysate: sc-2200.

DATA



SURF-1 (H-170): sc-68345. Western blot analysis of SURF-1 expression in non-transfected 293T: sc-117752 (A), human SURF-1 transfected 293T: sc-176677 (B) and HeLa (C) whole cell lysates.



SURF-1 (H-170): sc-68345. Western blot analysis of SURF-1 expression in non-transfected: sc-117752 (A) and human SURF-1 transfected: sc-110725 (B) 293T whole cell lysates.

RESEARCH USE

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

PROTOCOLS

See our web site at 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 (H-170).