SANTA CRUZ BIOTECHNOLOGY, INC.

SURF-1 (21H2): sc-65245

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. AJNR Am. J. Neuroradiol. 23: 1095-1100.

CHROMOSOMAL LOCATION

Genetic locus: SURF1 (human) mapping to 9q34.13; Surf1 (mouse) mapping to 2 A3.

SOURCE

SURF-1 (21H2) is a mouse monoclonal antibody raised against SURF-1 of human origin.

PRODUCT

Each vial contains 100 µg IgG_1 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.

RESEARCH USE

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

APPLICATIONS

SURF-1 (21H2) is recommended for detection of SURF-1 of mouse and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) and immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)].

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.

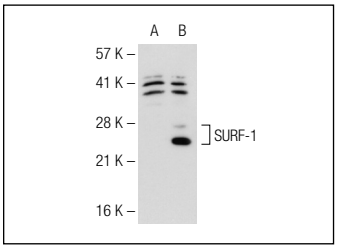
Molecular Weight of SURF-1: 31 kDa.

Positive Controls: mouse liver extract: sc-2256, SURF-1 (h): 293T Lysate: sc-110725 or HeLa whole cell lysate: sc-2200.

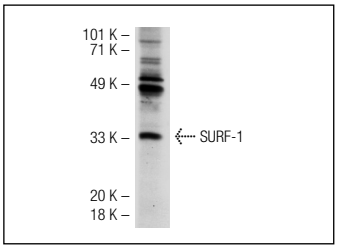
RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-mouse IgG-HRP: sc-2005 (dilution range: 1:2000-1:32,000) or Cruz Marker™ compatible goat anti-mouse IgG-HRP: sc-2031 (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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).

DATA



SURF-1 (21H2): sc-65245. 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.



SURF-1 (21H2): sc-65245. Western blot analysis of SURF-1 expression in HeLa whole cell lysate.

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.