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# SURF-1 (h3): 293T Lysate: sc-117415

## 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., Hoertnagel, K., Carrozzo, R., Galimberti, C., Munaro, M., Granatiero, M., Zelante, L., Gasparini, P., Marzella, R., Rocchi, M., Bayona-Bafaluy, M.P., Enriquez, J.A., Uziel, G., Bertini, E., Dionisi-Vici, C., Franco, B., Meitinger, T. and Zeviani, M. 1998. Mutations of SURF-1 in Leigh disease associated with cytochrome c oxidase deficiency. *Am. J. Hum. Genet.* 63: 1609-1621.
2. Tiranti, V., Galimberti, C., Nijtmans, L., Bovolenta, S., Perini, M.P. and Zeviani, M. 1999. Characterization of SURF-1 expression and SURF-1p function in normal and disease conditions. *Hum. Mol. Genet.* 8: 2533-2540.
3. Tiranti, V., Jaksch, M., Hofmann, S., Galimberti, C., Hoertnagel, K., Lulli, L., Freisinger, P., Bindoff, L., Gerbitz, K.D., Comi, G.P., Uziel, G., Zeviani, M. and Meitinger, T. 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., Karadimas, C., Checcarelli, N., Tanji, K., Papadopoulou, L.C., Pallotti, F., Guo, F.L., Shanske, S., Hirano, M., De Vivo, D.C., Van Coster, R., Kaplan, P., Bonilla, E. and DiMauro, S. 2000. Differential features of patients with mutations in two COX assembly genes, SURF-1 and SCO2. *Ann. Neurol.* 47: 589-595.
6. Farina, L., Chiapparini, L., Uziel, G., Bugiani, M., Zeviani, M. and Savoiodro, M. 2002. MR findings in Leigh syndrome with COX deficiency and SURF-1 mutations. *AJNR Am. J. Neuroradiol.* 23: 1095-1100.
7. Ogawa, Y., Naito, E., Ito, M., Yokota, I., Sajio, T., Shinahara, K. and Kuroda, Y. 2002. Three novel SURF-1 mutations in Japanese patients with Leigh syndrome. *Pediatr. Neurol.* 26: 196-200.
8. Agostino, A., Invernizzi, F., Tiveron, C., Fagioliari, G., Prelle, A., Lamantea, E., Giavazzi, A., Battaglia, G., Tatangelo, L., Tiranti, V. and Zeviani, M. 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.

## PRODUCT

SURF-1 (h3): 293T Lysate represents a lysate of human SURF-1 transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

## APPLICATIONS

SURF-1 (h3): 293T Lysate is suitable as a Western Blotting positive control for human reactive SURF-1 antibodies. Recommended use: 10-20 µl per lane.

Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

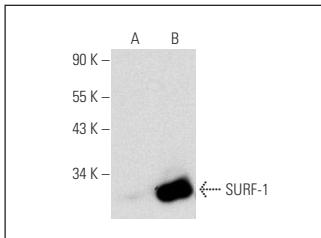
SURF-1 (H-7): sc-365159 is recommended as a positive control antibody for Western Blot analysis of enhanced human SURF-1 expression in SURF-1 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

## RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended:

1) Western Blotting: use m-IgG<sub>x</sub> BP-HRP: sc-516102 or m-IgG<sub>x</sub> 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.

## DATA



SURF-1 (H-7): sc-365159. Western blot analysis of SURF-1 expression in non-transfected: sc-117752 (**A**) and human SURF-1 transfected: sc-117415 (**B**) 293T whole cell lysates.

## STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. 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) for detailed protocols and support products.