SANTA CRUZ BIOTECHNOLOGY, INC.

SURF-1 (h4): 293T Lysate: sc-176677

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.
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.

PRODUCT

SURF-1 (h4): 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.

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.

APPLICATIONS

SURF-1 (h4): 293T Lysate is suitable as a Western Blotting positive control for human reactive SURF-1 antibodies. Recommended use: 10-20 µl per lane.

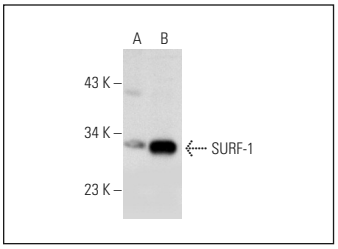
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

SURF-1 (D-9): sc-166948 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).

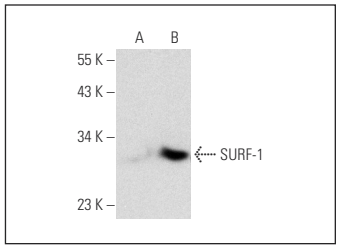
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ 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 (D-9): sc-166948. Western blot analysis of SURF-1 expression in non-transfected: sc-117752 (**A**) and human SURF-1 transfected: sc-176677 (**B**) 293T whole cell lysates.



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-176677 (**B**) 293T whole cell lysates.

PROTOCOLS

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