

SURF-1 (E-7): sc-137037

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* 1492: 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.

SOURCE

SURF-1 (E-7) is a mouse monoclonal 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_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin. Also available as TransCruz reagent for Gel Supershift and ChIP applications, sc-137037 X, 200 µg/0.1 ml.

APPLICATIONS

SURF-1 (E-7) is recommended for detection of SURF-1 of human origin by Western Blotting (starting dilution 1:100, 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), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

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

SURF-1 (E-7) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

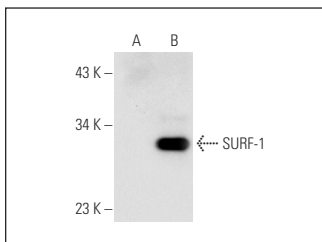
Molecular Weight of SURF-1: 31 kDa.

Positive Controls: human heart extract: sc-363763, mouse liver extract: sc-2256 or SURF-1 (h): 293T Lysate: sc-110725.

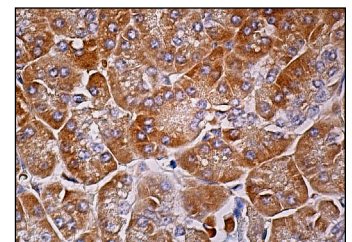
RECOMMENDED SUPPORT REAGENTS

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. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



SURF-1 (E-7): sc-137037. 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 (E-7): sc-137037. Immunoperoxidase staining of formalin fixed, paraffin-embedded human pancreas tissue showing cytoplasmic staining of glandular cells.

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.