

SCO1 (L-14): sc-49106

BACKGROUND

The SCO1 and SCO2 protein homologs belong to the SCO1/2 family of proteins. SCO1 and SCO2 both localize to the mitochondrion and are inner membrane proteins crucial for copper insertion or transport to the active site of cytochrome c oxidase (COX). COX is a crucial component in energy production because it functions as the terminal enzyme in the respiratory chain. SCO1 is predominantly expressed in highly oxidative phosphorylation tissues such as brain, heart and muscle, while SCO2 is ubiquitously expressed. Defects in the gene encoding for SCO1 may cause cytochrome c oxidase deficiency, a heterogeneous disorder. Defects in the gene encoding for SCO2 may cause cardioencephalomyopathy with cytochrome c oxidase deficiency, a fatal infantile disorder characterized by hypertrophic cardiomyopathy, lactic acidosis and gliosis.

REFERENCES

- Jaksch, M., Paret, C., Stucka, R., Horn, N., Müller-Höcker, J., Horvath, R., Trepesch, N., Stecker, G., Freisinger, P., Thirion, C., Müller, J., Lunke, R., Rödel, G., Shoubridge, E.A. and Lochmüller, H. 2001. Cytochrome c oxidase deficiency due to mutations in SCO2, encoding a mitochondrial copper-binding protein, is rescued by copper in human myoblasts. *Hum. Mol. Genet.* 10: 3025-3035.
- Balatri, E., Banci, L., Bertini, I., Cantini, F. and Ciofi-Baffoni, S. 2003. Solution structure of SCO1: a thioredoxin-like protein involved in cytochrome c oxidase assembly. *Structure* 11: 1431-1443.
- Hornig, Y.C., Cobine, P.A., Maxfield, A.B., Carr, H.S. and Winge, D.R. 2004. Specific copper transfer from the COX17 metallochaperone to both SCO1 and COX11 in the assembly of yeast cytochrome c oxidase. *J. Biol. Chem.* 279: 35334-35340.
- Leary, S.C., Kaufman, B.A., Pellicchia, G., Guercin, G.H., Mattman, A., Jaksch, M. and Shoubridge, E.A. 2004. Human SCO1 and SCO2 have independent, cooperative functions in copper delivery to cytochrome c oxidase. *Hum. Mol. Genet.* 13: 1839-1848.
- Williams, J.C., Sue, C., Banting, G.S., Yang, H., Glerum, D.M., Hendrickson, W.A. and Schon, E.A. 2005. Crystal structure of human SCO1: implications for redox signaling by a mitochondrial cytochrome c oxidase assembly protein. *J. Biol. Chem.* 280: 15202-15211.
- Hornig, Y.C., Leary, S.C., Cobine, P.A., Young, F.B., George, G.N., Shoubridge, E.A. and Winge, D.R. 2005. Human SCO1 and SCO2 function as copper-binding proteins. *J. Biol. Chem.* 280: 34113-34122.

CHROMOSOMAL LOCATION

Genetic locus: SCO1 (human) mapping to 17p12-p13; Sco1 (mouse) mapping to 11 B3.

SOURCE

SCO1 (L-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of SCO1 of human origin.

RESEARCH USE

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

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-49106 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

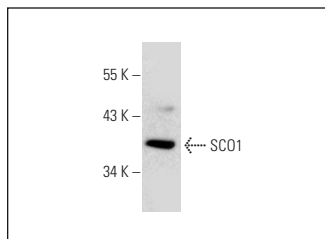
SCO1 (L-14) is recommended for detection of SCO1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for SCO1 siRNA (h): sc-61505, SCO1 shRNA Plasmid (h): sc-61505-SH and SCO1 shRNA (h) Lentiviral Particles: sc-61505-V.

Molecular Weight of SCO1: 29 kDa.

Positive Controls: mouse liver extract: sc-2256.

DATA



SCO1 (L-14): sc-49106. Western blot analysis of SCO1 expression in mouse liver tissue extracts.

STORAGE

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

PROTOCOLS

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



Try **SCO1 (D-1): sc-398001** or **SCO1 (B-9): sc-365380**, our highly recommended monoclonal alternatives to SCO1 (L-14).