

# SCO1 (B-9): sc-365380

## 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 heterogenous 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

1. Jaksch, M., et al. 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.
2. Balatri, E., et al. 2003. Solution structure of SCO1: a thioredoxin-like protein involved in cytochrome c oxidase assembly. *Structure* 11: 1431-1443.
3. Leary, S.C., et al. 2004. Human SCO1 and SCO2 have independent, cooperative functions in copper delivery to cytochrome c oxidase. *Hum. Mol. Genet.* 13: 1839-1848.
4. Horng, Y.C., et al. 2004. Specific copper transfer from the COX17 metallo-chaperone to both SCO1 and COX11 in the assembly of yeast cytochrome c oxidase. *J. Biol. Chem.* 279: 35334-35340.
5. Williams, J.C., et al. 2005. Crystal structure of human SCO1: implications for redox signaling by a mitochondrial cytochrome c oxidase assembly protein. *J. Biol. Chem.* 280: 15202-15211.
6. Horng, Y.C., et al. 2005. Human SCO1 and SCO2 function as copper-binding proteins. *J. Biol. Chem.* 280: 34113-34122.
7. Horvath, R., et al. 2005. Congenital cataract, muscular hypotonia, developmental delay and sensori-neural hearing loss associated with a defect in copper metabolism. *J. Inher. Metab. Dis.* 28: 479-492.
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## CHROMOSOMAL LOCATION

Genetic locus: SCO1 (human) mapping to 17p13.1.

## SOURCE

SCO1 (B-9) is a mouse monoclonal antibody raised against amino acids 178-243 mapping near the C-terminus of SCO1 of human origin.

## RESEARCH USE

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

## PRODUCT

Each vial contains 200 µg IgG<sub>2b</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

## APPLICATIONS

SCO1 (B-9) is recommended for detection of SCO1 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) 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.

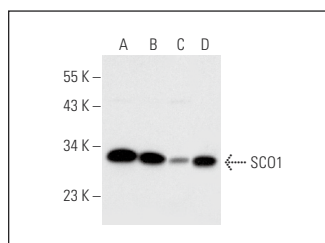
Molecular Weight of SCO1: 29 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, Hep G2 cell lysate: sc-2227 or human liver extract: sc-363766.

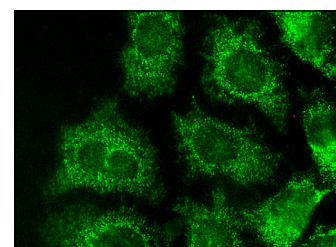
## RECOMMENDED SUPPORT REAGENTS

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.

## DATA



SCO1 (B-9): sc-365380. Western blot analysis of SCO1 expression in Hep G2 (A), HeLa (B) and IMR-32 (C) whole cell lysates and human liver tissue extract (D).



SCO1 (B-9): sc-365380. Immunofluorescence staining of methanol-fixed HeLa cells showing cytoplasmic localization.

## 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](http://www.scbt.com) for detailed protocols and support products.