

SCO1 Antibody

Catalog # ASC10503

Product Information

Application WB, IF, E, IHC-P

Primary Accession <u>075880</u>

Other Accession 075880, 8134663
Reactivity Human, Mouse, Rat

Host Rabbit
Clonality Polyclonal
Isotype IgG
Calculated MW 33814
Concentration (mg/ml) 1 mg/mL
Conjugate Unconjugated

Application Notes SCO1 antibody can be used for detection of SCO1 by Western blot at 0.5 - 1

□g/mL. Antibody can also be used for immunohistochemistry starting at 2.5

□g/mL. For immunofluorescence start at 20 □g/mL.

Additional Information

Gene ID 6341

Other Names SCO1 Antibody: SCOD1, SCOD1, Protein SCO1 homolog, mitochondrial, SCO

cytochrome oxidase deficient homolog 1 (yeast)

Target/Specificity SCO1;

Reconstitution & Storage SCO1 antibody can be stored at 4°C for three months and -20°C, stable for up

to one year. As with all antibodies care should be taken to avoid repeated freeze thaw cycles. Antibodies should not be exposed to prolonged high

temperatures.

Precautions SCO1 Antibody is for research use only and not for use in diagnostic or

therapeutic procedures.

Protein Information

Name SCO1

Synonyms SCOD1

Function Copper metallochaperone essential for the maturation of cytochrome c

oxidase subunit II (MT-CO2/COX2). Not required for the synthesis of

MT-CO2/COX2 but plays a crucial role in stabilizing MT- CO2/COX2 during its subsequent maturation. Involved in transporting copper to the Cu(A) site on MT-CO2/COX2 (PubMed:15229189, PubMed:15659396, PubMed:16735468, PubMed:17189203, PubMed:19336478). Plays an important role in the regulation of copper homeostasis by controlling the abundance and cell

membrane localization of copper transporter CTR1 (By similarity).

Cellular Location Mitochondrion. Mitochondrion inner membrane; Single-pass membrane

protein

Tissue Location Predominantly expressed in tissues characterized by high rates of oxidative

phosphorylation (OxPhos), including muscle, heart, and brain.

Background

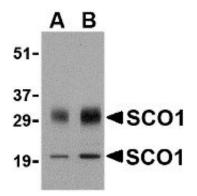
SCO1 Antibody: Synthesis of cytochrome c oxidase 1 was initially identified in yeast as one of two cytochrome c oxidase (COX) assembly proteins that enable the assembly of cytochrome c holoenzyme, a complex that catalyzes the transfer of reducing equivalents from cytochrome c to molecular oxygen and pumps protons across the inner mitochondrial membrane. Like their yeast homologs, the function of both SCO1 and SCO2 are dependent on copper ion binding. Mutations in either gene can lead to cytochrome c oxidase respiratory chain defects, with a missense mutation in human SCO1 (P174L) associated with a fatal neonatal hepatopathy when the second allele is also non-functional, suggesting the pathology is due to loss of function. It has been suggested that this mutation alters the SCO1 affinity for the copper (I) ion, thus impairing the efficiency of copper transfer to the cytochrome c oxidase. At least two isoforms of SCO1 are known to exist and both are recognized by the SCO1 antibody. This SCO1 antibody has no cross-reactivity to SCO2.

References

Glerum DM, Shtanko A, and Tzagoloff A. SCO1 and SCO2 act as high copy suppressors of a mitochondrial copper recruitment defect in Saccharomyces cerevisiae.J. Biol. Chem.1996; 271:20531-5. Horng Y-C, Leary SC, Cobine PA, et al. Human Sco1 and Sco2 function as copper-binding proteins. J. Biol. Chem.2005; 280:34113-22.

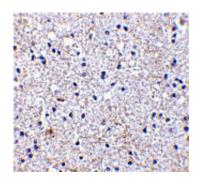
Valnot I, Osmond S, Gigarel N. Mutations of the SCO1 gene in mitochondrial cytochrome c oxidase deficiency with neonatal-onset hepatic failure and encephalopathy. Am. J. Hum. Genet.2000; 67:1104-9. Banci L, Bertini I, Ciofi-Baffoni S, et al. Human Sco1 functional studies and pathological implications of the P174L mutant. Proc. Natl. Acad. Sci. USA2007; 104:15-20.

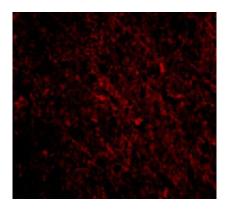
Images



Western blot analysis of SCO1 in human brain tissue lysate with SCO1 antibody at (A) 0.5 and (B) 1 µg/mL.

Immunohistochemistry of SCO1 in human brain tissue with SCO1 antibody at 2.5 µg/mL.





Immunofluorescence of SCO1 in Human Brain cells with SCO1 antibody at 20 $\mu\text{g/mL}.$

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.