



# COX19 (C-13): sc-104167

## BACKGROUND

The cytochrome c oxidase (COX) family of proteins function as the final electron donor in the respiratory chain to drive a proton gradient across the inner mitochondrial membrane, ultimately resulting in the production of water. The mammalian COX apoenzyme is a dimer, with each monomer consisting of 13 subunits, some of which are mitochondrial and some of which are nuclear. COX19 is a 90 amino acid protein that localizes to the cytosol and contains one CHCH domain. Expressed ubiquitously with highest expression in skeletal muscle, COX19 is thought to be essential for the assembly of the COX apoenzyme, indicating an important role for COX19 in the respiratory chain. The gene encoding COX19 maps to human chromosome 7, which houses over 1,000 genes and comprises nearly 5% of the human genome. Defects in some of the genes localized to chromosome 7 have been linked to osteogenesis imperfecta, Williams-Beuren syndrome, Pendred syndrome, lissencephaly, citrullinemia and Shwachman-Diamond syndrome.

## REFERENCES

1. Taanman, J.W., Hall, R.E., Tang, C., Marusich, M.F., Kennaway, N.G. and Capaldi, R.A. 1993. Tissue distribution of cytochrome c oxidase isoforms in mammals. Characterization with monoclonal and polyclonal antibodies. *Biochim. Biophys. Acta* 1225: 95-100.
2. Lenka, N., Vijayarath, C., Mullick, J. and Avadhani, N.G. 1998. Structural organization and transcription regulation of nuclear genes encoding the mammalian cytochrome c oxidase complex. *Prog. Nucleic Acid Res. Mol. Biol.* 61: 309-344.
3. Nobrega, M.P., Bandeira, S.C., Beers, J. and Tzagoloff, A. 2002. Characterization of COX19, a widely distributed gene required for expression of mitochondrial cytochrome oxidase. *J. Biol. Chem.* 277: 40206-40211.
4. Tay, S.K., Nesti, C., Mancuso, M., Schon, E.A., Shanske, S., Bonilla, E., Davidson, M.M. and Dimauro, S. 2004. Studies of COX16, COX19, and PET191 in human cytochrome-c oxidase deficiency. *Arch. Neurol.* 61: 1935-1937.
5. Sacconi, S., Trevisson, E., Pistollato, F., Baldoim, M.C., Rezzonico, R., Bourget, I., Desnuelle, C., Tenconi, R., Basso, G., DiMauro, S. and Salvati, L. 2005. hCOX18 and hCOX19: two human genes involved in cytochrome c oxidase assembly. *Biochem. Biophys. Res. Commun.* 337: 832-839.
6. Online Mendelian Inheritance in Man, OMIM™. 2006. Johns Hopkins University, Baltimore, MD. MIM Number: 610429. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>

## CHROMOSOMAL LOCATION

Genetic locus: COX19 (human) mapping to 7p22.3.

## SOURCE

COX19 (C-13) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of COX19 of human origin.

## STORAGE

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

## 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-104167 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

COX19 (C-13) is recommended for detection of COX19 of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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 COX19 siRNA (h): sc-89654, COX19 shRNA Plasmid (h): sc-89654-SH and COX19 shRNA (h) Lentiviral Particles: sc-89654-V.

Molecular Weight of COX19: 10 kDa.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

## RESEARCH USE

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

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.