

BACKGROUND

Cytochrome c oxidase (EC 1.9.3.1) (COX) belongs to the superfamily of terminal oxidases present in all aerobic organisms. In eukaryotic cells, cytochrome c oxidase is embedded as a dimer in the mitochondrial inner membrane. Electron transfer by the enzyme from cytochrome c to molecular oxygen is coupled to proton pumping across the membrane, contributing to the mitochondrial membrane potential, resulting in a proton and charge gradient that is then employed by the FOF1-ATPase to synthesize ATP. The redox centers involved in electron transfer are two copper centers (CuA and CuB) and two heme A moieties (a and a₃). The eukaryotic enzyme contains three mitochondrial DNA (mtDNA)-encoded subunits: MTCO1, MTCO2, and MTCO3. These subunits are homologous to polypeptides found in bacterial cytochrome c oxidases. They form both the catalytic and structural core of the enzyme. The hydrophobic interior of enzyme subunit 1 (MTCO1) coordinates the heme a group and a binuclear center composed of heme a₃ and CuB. The intermembrane space domain of enzyme subunit 2 (MTCO2) coordinates the binuclear CuA center. In addition to the three mtDNA-encoded subunits, mammalian cytochrome c oxidase contains 10 nuclear DNA-encoded subunits: COX4, COX5A, COX5B, COX6A, COX6B, COX6C, COX7A, COX7B, COX7C, and COX8. In humans, COX6A and COX7A exist as skeletal/cardiac muscle (COX6A2 and COX7A1) and ubiquitously expressed (COX6A1 and COX7A2) isoforms. Although not confirmed at the protein level, human isoforms of COX4, COX6B, and COX8 have also been identified.¹ Cytochrome c oxidase subunit 4 isoform 1 (COX4I1) is the nuclear-encoded subunit IV isoform 1 of the human mitochondrial respiratory chain enzyme. It is located at the 3' of the NOC4 (neighbor of COX4) gene in a head-to-head orientation, and shares a promoter with it.

Cytochrome c oxidase (COX) deficiencies are one of the most common defects of the respiratory chain found in mitochondrial diseases. cytochrome c oxidase (COX) biosynthesis requires numerous assembly factors that do not form part of the final complex but participate in prosthetic group synthesis and metal delivery in addition to membrane insertion and maturation of cytochrome c oxidase (COX) subunits. Human diseases associated with cytochrome c oxidase (COX) deficiency including encephalomyopathies, Leigh syndrome, hypertrophic cardiomyopathies, and fatal lactic acidosis are caused by mutations in cytochrome c oxidase (COX) subunits or assembly factors. Owing to the importance of the enzyme, pathogenetic mutations affecting cytochrome c oxidase (COX) frequently result in severe, often fatal metabolic disorders.² In the last decade, numerous animal models have been created to understand the pathophysiology of cytochrome c oxidase (COX) deficiencies and the function of assembly factors. These animal models, ranging

from invertebrates to mammals, in most cases mimic the pathological features of the human diseases.³

References

1. Capaldi, R.A.: Annu. Rev. Biochem. 59: 569-596, 1990
2. Pecina, P. et al: Physiol. Res. 53(Suppl. 1):S213-S223, 2004
3. Diaz, F.: Biochim Biophys Acta. 1802:100-10, 2010

TECHNICAL INFORMATION

Source:

COX4I1 Antibody is a mouse monoclonal antibody raised against purified recombinant human COX4I1 fragments expressed in *E. coli*.

Specificity and Sensitivity:

This antibody detects endogenous COX4I1 proteins without cross-reactivity with other related proteins.

Storage Buffer: PBS and 30% glycerol

Storage:

Store at -20°C for at least one year. Store at 4°C for frequent use. Avoid repeated freeze-thaw cycles.

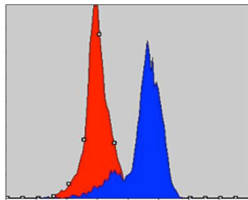
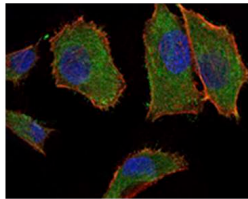
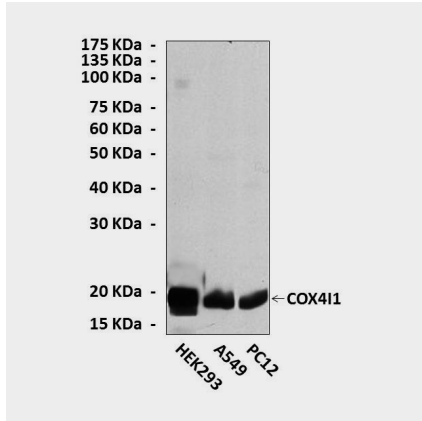
APPLICATIONS

Application:	*Dilution:
WB	1:1000
IP	1:50
IHC	n/d
ICC	1:50-200
FACS	1:50-100

**Optimal dilutions must be determined by end user.*



QUALITY CONTROL DATA



Top: Western Blot detection of COX4I1 proteins in various cell lysates using COX4I1 antibody. **Middle:** This antibody stains PANC-1 cells in confocal immunofluorescent analysis (COX4I1 antibody: Green; Actin filaments: Red; and DRAQ5: blue). **Bottom:** It also detects COX4I1 proteins specifically in K562 cells by FACS assay (COX4I1 Antibody: blue and negative control IgG: red).

