

COX1/MTCO1 Rabbit pAb

Catalog # AP98502

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P00395
Other Accession	P00395
Host	Rabbit
Clonality	Polyclonal
Calculated MW	57041
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human COX1
Epitope Specificity	401-500/513
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the heme-copper respiratory oxidase family.
DISEASE	Defects in MT-CO1 are a cause of Leber hereditary optic neuropathy (LHON) [MIM:535000]. LHON is a maternally inherited disease resulting in acute or subacute loss of central vision, due to optic nerve dysfunction. Cardiac conduction defects and neurological defects have also been described in some patients. LHON results from primary mitochondrial DNA mutations affecting the respiratory chain complexes. Note=MT-CO1 may play a role in the pathogenesis of acquired idiopathic sideroblastic anemia, a disease characterized by inadequate formation of heme and excessive accumulation of iron in mitochondria. Mitochondrial iron overload may be attributable to mutations of mitochondrial DNA because these can cause respiratory chain dysfunction, thereby impairing reduction of ferric iron to ferrous iron. The reduced form of iron is essential to the last step of mitochondrial heme biosynthesis. Defects in MT-CO1 are a cause of mitochondrial complex IV deficiency (MT-C4D) [MIM:220110]; also known as cytochrome c oxidase deficiency. A disorder of the mitochondrial respiratory chain with heterogeneous clinical manifestations, ranging from isolated myopathy to severe multisystem disease affecting several tissues and organs. Features include hypertrophic cardiomyopathy, hepatomegaly and liver dysfunction, hypotonia, muscle weakness, exercise intolerance, developmental delay, delayed motor development and mental retardation. A subset of patients manifest Leigh syndrome. Defects in MT-CO1 are associated with recurrent myoglobinuria mitochondrial (RM-MT) [MIM:550500]. Recurrent myoglobinuria is characterized by recurrent attacks of rhabdomyolysis (necrosis or disintegration of skeletal muscle) associated with muscle pain and weakness, and followed by excretion of myoglobin in the urine. Defects in MT-CO1 are a cause of deafness sensorineural mitochondrial (DFNM) [MIM:500008]. DFNM is a form of non-syndromic deafness with maternal inheritance. Affected individuals manifest progressive, postlingual, sensorineural hearing loss involving high frequencies. Defects in MT-CO1 are a cause of colorectal cancer (CRC) [MIM:114500].
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Background Descriptions	Contributes to cytochrome-c oxidase activity. Predicted to be involved in electron transport coupled proton transport and mitochondrial electron transport, cytochrome c to oxygen. Part of mitochondrial respiratory chain complex III and mitochondrial respiratory chain complex IV. [provided by Alliance of Genome Resources, Apr 2022]
Additional Information	
Gene ID	4512
Other Names	COI; MTCO1; CoxI; COX1_BOVIN; MT-CO1; Cytochrome c oxidase polypeptide I; 7.1.1.9; COX1_HUMAN; COX1_MOUSE; COX1_RAT; COX1_DANRE; cox1; mitochondrially encoded cytochrome c oxidase I; cytochrome c oxidase I
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	MT-CO1
Synonyms	COI, COXI, MTCO1
Function	Component of the cytochrome c oxidase, the last enzyme in the mitochondrial electron transport chain which drives oxidative phosphorylation. The respiratory chain contains 3 multisubunit complexes succinate dehydrogenase (complex II, CII), ubiquinol- cytochrome c oxidoreductase (cytochrome b-c1 complex, complex III, CIII) and cytochrome c oxidase (complex IV, CIV), that cooperate to transfer electrons derived from NADH and succinate to molecular oxygen, creating an electrochemical gradient over the inner membrane that drives transmembrane transport and the ATP synthase. Cytochrome c oxidase is the component of the respiratory chain that catalyzes the reduction of oxygen to water. Electrons originating from reduced cytochrome c in the intermembrane space (IMS) are transferred via the dinuclear copper A center (CU(A)) of subunit 2 and heme A of subunit 1 to the active site in subunit 1, a binuclear center (BNC) formed by heme A3 and copper B (CU(B)). The BNC reduces molecular oxygen to 2 water molecules using 4 electrons from cytochrome c in the IMS and 4 protons from the mitochondrial matrix.
Cellular Location	Mitochondrion inner membrane; Multi-pass membrane protein

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