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COX3 Rabbit pAb

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Catalog # AP58112

Product Information

Application	WB
Primary Accession	P00414
Reactivity	Rat, Mouse
Predicted	Pig, Dog, Human, Rabbit, Chicken, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	29951
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human COX3
Epitope Specificity	25-130/261
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Mitochondrion inner membrane; Multi-pass membrane protein.
DISEASE	Leber hereditary optic neuropathy (LHON) [MIM:535000]: 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=The disease is caused by mutations affecting the gene represented in this entry. Mitochondrial complex IV deficiency (MT-C4D) [MIM:220110]: 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. Some affected individuals manifest a fatal hypertrophic cardiomyopathy resulting in neonatal death. A subset of patients manifest Leigh syndrome. Note=The disease is caused by mutations affecting the gene represented in this entry. 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. Note=The gene represented in this entry may be involved in disease pathogenesis.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	MT-CO3 (Mitochondrially Encoded Cytochrome C Oxidase III) is a Protein Coding gene. Diseases associated with MT-CO3 include Leber Hereditary Optic Neuropathy and Genetic Recurrent Myoglobinuria. Among its related pathways are Gene Expression and Respiratory electron transport, ATP synthesis by chemiosmotic coupling, and heat production by uncoupling proteins.. Gene Ontology (GO) annotations related to this gene include

cytochrome-c oxidase activity and heme-copper terminal oxidase activity.

Additional Information

Gene ID	4514
Other Names	Cytochrome c oxidase subunit 3, 7.1.1.9, Cytochrome c oxidase polypeptide III, MT-CO3, COIII, COXIII, MTCO3
Dilution	WB=1:500-2000
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.

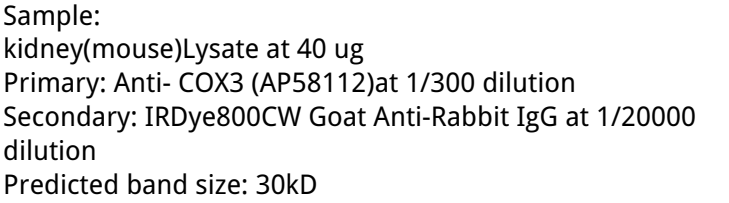
Protein Information

Name	MT-CO3
Synonyms	COIII, COXIII, MTCO3
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

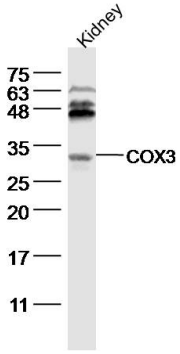
Background

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Images



Observed band size: 30 kD



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