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COX10 Rabbit pAb

Catalog # AP98387

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	Q12887
Other Accession	Q12887
Reactivity	Human, Mouse
Predicted	Rat
Host	Rabbit
Clonality	Polyclonal
Calculated MW	48910
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human COX10
Epitope Specificity	161-260/443
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	Preservative: 0.02% Proclin300, Constituents: 1% BSA, 0.01M PBS, pH7.4.
SIMILARITY	Belongs to the UbiA prenyltransferase family.
DISEASE	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. Leigh syndrome (LS) [MIM:256000]: An early-onset progressive neurodegenerative disorder characterized by the presence of focal, bilateral lesions in one or more areas of the central nervous system including the brainstem, thalamus, basal ganglia, cerebellum and spinal cord. Clinical features depend on which areas of the central nervous system are involved and include subacute onset of psychomotor retardation, hypotonia, ataxia, weakness, vision loss, eye movement abnormalities, seizures, and dysphagia. Note=The disease is caused by mutations affecting the gene represented in this entry.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Cytochrome c oxidase (COX), the terminal component of the mitochondrial respiratory chain, catalyzes the electron transfer from reduced cytochrome c to oxygen. This component is a heteromeric complex consisting of 3 catalytic subunits encoded by mitochondrial genes and multiple structural subunits encoded by nuclear genes. The mitochondrially-encoded subunits function in electron transfer, and the nuclear-encoded subunits may function in the regulation and assembly of the complex. This nuclear gene encodes heme A:farnesyltransferase, which is not a structural subunit but required for the expression of functional COX and functions in the maturation of the heme A prosthetic group of COX. This protein is predicted to contain 7-9

transmembrane domains localized in the mitochondrial inner membrane. A gene mutation, which results in the substitution of a lysine for an asparagine (N204K), is identified to be responsible for cytochrome c oxidase deficiency. In addition, this gene is disrupted in patients with CMT1A (Charcot-Marie-Tooth type 1A) duplication and with HNPP (hereditary neuropathy with liability to pressure palsies) deletion. [provided by RefSeq, Jul 2008]

Additional Information

Gene ID	1352
Other Names	MC4DN3; 2410004F01Rik; im:7145568; im:7157205; wu:fb18a03; COX10_HUMAN; COX10; Heme O synthase; 2.5.1.141; COX10_MOUSE; cytochrome c oxidase assembly factor heme A:farnesyltransferase COX10; COX10 homolog, cytochrome c oxidase assembly protein, heme A: farnesyltransferase (yeast); cytochrome c oxidase assembly homolog 10 (yeast); COX10, heme A:farnesyltransferase cytochrome c oxidase assembly factor; protoheme IX farnesyltransferase, mitochondrial
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	COX10
Function	Converts protoheme IX and farnesyl diphosphate to heme O.
Cellular Location	Mitochondrion membrane; Multi-pass membrane protein

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