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COQ2 Rabbit pAb

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Catalog # AP55367

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	Q96H96
Predicted	Rat, Pig, Human, Mouse, Rabbit, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	40475
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human COQ2
Epitope Specificity	51-150/371
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 membrane.
DISEASE	Defects in COQ2 are the cause of coenzyme Q10 deficiency, primary, type 1 (COQ10D1) [MIM:607426]. An autosomal recessive disorder with variable manifestations consistent with 5 major phenotypes. The phenotypes include an encephalomyopathic form with seizures and ataxia; a multisystem infantile form with encephalopathy, cardiomyopathy and renal failure; a predominantly cerebellar form with ataxia and cerebellar atrophy; Leigh syndrome with growth retardation; and an isolated myopathic form.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes an enzyme that functions in the final steps in the biosynthesis of CoQ (ubiquinone), a redox carrier in the mitochondrial respiratory chain and a lipid-soluble antioxidant. This enzyme, which is part of the coenzyme Q10 pathway, catalyzes the prenylation of parahydroxybenzoate with an all-trans polyprenyl group. Mutations in this gene cause coenzyme Q10 deficiency, a mitochondrial encephalomyopathy, and also COQ2 nephropathy, an inherited form of mitochondriopathy with primary renal involvement. [provided by RefSeq, Oct 2009]

Additional Information

Gene ID	27235
Other Names	4-hydroxybenzoate polyprenyltransferase, mitochondrial {ECO:0000255|HAMAP-Rule:MF_03189}, 4-HB polyprenyltransferase {ECO:0000255|HAMAP-Rule:MF_03189}, 2.5.1.39, 4-hydroxybenzoate decaprenyltransferase, COQ2 homolog, hCOQ2, Para-hydroxybenzoate--polyprenyltransferase, PHB:PPT {ECO:0000255|HAMAP-Rule:MF_03189}, PHB:polyprenyltransferase

	{ECO:0000255|HAMAP-Rule:MF_03189}, COQ2 {ECO:0000255|HAMAP-Rule:MF_03189, ECO:0000303|PubMed:15153069}
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
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	COQ2 {ECO:0000255|HAMAP-Rule:MF_03189, ECO:0000303|PubMed:15153069}
Function	Mediates the second step in the final reaction sequence of coenzyme Q (CoQ) biosynthesis (PubMed:15153069, PubMed:16400613, PubMed:17374725, PubMed:20526342). Catalyzes the prenylation of para-hydroxybenzoate (PHB) with an all-trans polyprenyl donor (such as all-trans-decaprenyl diphosphate) (PubMed:15153069, PubMed:16400613, PubMed:17374725, PubMed:20526342). The length of the polyprenyl side chain varies depending on the species, in humans, the side chain is comprised of 10 isoprenyls (decaprenyl) producing CoQ10 (also known as ubiquinone), whereas rodents predominantly generate CoQ9 (PubMed:15153069, PubMed:16400613). However, this specificity is not complete, human tissues have low amounts of CoQ9 and rodent organs contain some CoQ10 (PubMed:15153069). Plays a central role in the biosynthesis of CoQ10 (PubMed:15153069, PubMed:16400613, PubMed:17374725). CoQ10 is a vital molecule that transports electrons from mitochondrial respiratory chain complexes (PubMed:16400613, PubMed:17374725, PubMed:27493029). CoQs also function as cofactors for uncoupling protein and play a role as regulators of the extracellularly-induced ceramide-dependent apoptotic pathway (PubMed:16400613, PubMed:17374725). Regulates mitochondrial permeability transition pore (mPTP) opening and ROS production (pivotal events in cell death) in a tissue specific manner (By similarity).
Cellular Location	Mitochondrion inner membrane {ECO:0000255|HAMAP- Rule:MF_03189, ECO:0000269|PubMed:27493029}; Multi-pass membrane protein {ECO:0000255|HAMAP-Rule:MF_03189}; Matrix side {ECO:0000255|HAMAP-Rule:MF_03189}
Tissue Location	Widely expressed. Present in all of the tissues tested. Expressed at higher level in skeletal muscle, adrenal glands and the heart.

Background

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.