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Coproporphyrinogen III Oxidase Rabbit pAb

Catalog # AP99708

Product Information

Application	IHC-P, IHC-F, IF, E
Primary Accession	P36551
Other Accession	P36551
Predicted	Rat, Pig, Human, Mouse, Rabbit, Chicken, Dog, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	50152
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human CPOX/Coproporphyrinogen Oxidase
Epitope Specificity	361-454/454
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 aerobic coproporphyrinogen-III oxidase family.
SUBUNIT	Homodimer.
DISEASE	Defects in CPOX are the cause of hereditary coproporphyria (HCP) [MIM:121300]. HCP is an acute hepatic porphyria and an autosomal dominant disease characterized by neuropsychiatric disturbances and skin photosensitivity. Biochemically, there is an overexcretion of coproporphyrin III in the urine and in the feces. HCP is clinically characterized by attacks of abdominal pain, neurological disturbances, and psychiatric symptoms. The symptoms are generally manifested with rapid onset, and can be precipitated by drugs, alcohol, caloric deprivation, infection, endocrine factors or stress. A severe variant form is harderoporphyria, which is characterized by earlier onset attacks, massive excretion of harderoporphyrin in the feces, and a marked decrease of coproporphyrinogen IX oxidase activity.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	CPOX is a 454 amino acid mitochondrial enzyme that is localized to the inner membrane space of erythrocytes. It participates in the sixth step of heme biosynthesis by catalyzing the formation of protoporphyrinogen IX from copropophyrinogen III. Mutations in the gene encoding CPOX are the cause of coproporphyria, an autosomal dominant disease characterized by skin photosensitivity and neurological disturbances. Symptoms are often experienced as attacks, which include severe abdominal and nerve pain. People affected by coproporphyria overexcrete copropophyrinogen III in feces and urine and the enzymatic activity of CPOX is found to be approximately half that of normal, leading to a decrease in overall heme synthesis. There is no cure for coproporphyria, but preventative treatment to relieve symptoms usually involves dietary changes and avoidance of drugs and alcohol.

Additional Information

Gene ID	1371
Other Names	COX; CPO; CPX; HARPO; HCP; M100835; Rgsc835; cac; nct; HEM6_HUMAN; CPOX; Coprogen oxidase; Coproporphyrinogenase; 1.3.3.3; HEM6_MOUSE; HEM6_RAT; coproporphyrinogen oxidase; coproporphyrinogen oxidase (coproporphyria, harderoporphyria); coproporphyria; homozygous coproporphyria
Dilution	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	CPOX (HGNC:2321)
Synonyms	CPO, CPX
Function	Catalyzes the aerobic oxidative decarboxylation of propionate groups of rings A and B of coproporphyrinogen-III to yield the vinyl groups in protoporphyrinogen-IX and participates to the sixth step in the heme biosynthetic pathway.
Cellular Location	Mitochondrion intermembrane space {ECO:0000250|UniProtKB:Q3B7D0}

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