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Rabbit Anti-Coproporphyrinogen III Oxidase antibody

SL8860R

Product Name:	Coproporphyrinogen III Oxidase
Chinese Name:	原卟啉氧化酶3抗体
Alias:	Coprogen oxidase; COPROPORPHYRIA; Coproporphyrinogen III oxidase; Coproporphyrinogenase; COX; CPO; CPOX; CPX; HEM 6; Hem-6; Hem6; HEM6_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Pig,Cow,Rabbit,Sheep,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	39kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human CPOX/Coproporphyrinogen Oxidase:361-454/454
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	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. **Function:** Coproporphyrinogen Oxidase (CPOX) is the sixth enzyme of the heme biosynthetic pathway. This soluble protein is localized in the intermembrane space of mitochondria and its catalytic activy mediates the coversion of Coproporphyrinogen-III + O2 + 2 H+ into Protoporphyrinogen-IX + 2 CO2 + 2 H2O. Defects in CPOX are the cause of hereditary coproporphyria **Subunit:** Homodimer. **Subcellular Location:** Mitochondrion intermembrane space. **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. **Similarity:** Belongs to the aerobic coproporphyrinogen-III oxidase family. **SWISS:** P36551 **Gene ID:** 1371 **Database links:**

	Entrez Gene: 1371Human Entrez Gene: 12892Mouse Entrez Gene: 304024Rat Omim: 612732Human SwissProt: P36551Human SwissProt: P36552Mouse SwissProt: Q3B7D0Rat Unigene: 476982Human Unigene: 291519Mouse Unigene: 19581Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.