

CPOX (B-9): sc-393388

BACKGROUND

CPOX (coproporphyrinogen III oxidase) 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 coproporphyrinogen III. Mutations in the gene encoding CPOX are the cause of coproporphyrinemia, 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 coproporphyrinemia overexcrete coproporphyrinogen 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 coproporphyrinemia, but preventative treatment to relieve symptoms usually involves dietary changes and avoidance of drugs and alcohol.

REFERENCES

1. Taketani, S., et al. 1994. Molecular cloning, sequencing and expression of cDNA encoding human coproporphyrinogen oxidase. *Biochim. Biophys. Acta* 1183: 547-549.
2. Martasek, P., et al. 1994. Homozygous hereditary coproporphyrinemia caused by an arginine to tryptophane substitution in coproporphyrinogen oxidase and common intragenic polymorphisms. *Hum. Mol. Genet.* 3: 477-480.
3. Delfau-Larue, M.H., et al. 1994. Coproporphyrinogen oxidase: gene organization and description of a mutation leading to exon 6 skipping. *Hum. Mol. Genet.* 3: 1325-1330.

CHROMOSOMAL LOCATION

Genetic locus: CPOX (human) mapping to 3q11.2; Cpx (mouse) mapping to 16 C1.2.

SOURCE

CPOX (B-9) is a mouse monoclonal antibody raised against amino acids 155-454 mapping at the C-terminus of CPOX of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

CPOX (B-9) is available conjugated to agarose (sc-393388 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-393388 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-393388 PE), fluorescein (sc-393388 FITC), Alexa Fluor® 488 (sc-393388 AF488), Alexa Fluor® 546 (sc-393388 AF546), Alexa Fluor® 594 (sc-393388 AF594) or Alexa Fluor® 647 (sc-393388 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-393388 AF680) or Alexa Fluor® 790 (sc-393388 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

APPLICATIONS

CPOX (B-9) is recommended for detection of CPOX of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for CPOX siRNA (h): sc-77907, CPOX siRNA (m): sc-142545, CPOX shRNA Plasmid (h): sc-77907-SH, CPOX shRNA Plasmid (m): sc-142545-SH, CPOX shRNA (h) Lentiviral Particles: sc-77907-V and CPOX shRNA (m) Lentiviral Particles: sc-142545-V.

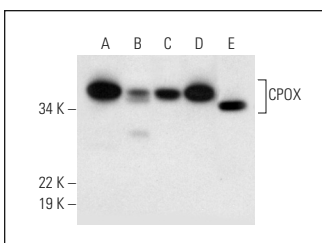
Molecular Weight of CPOX: 36 kDa.

Positive Controls: JAR cell lysate: sc-2276, Hep G2 cell lysate: sc-2227 or Jurkat whole cell lysate: sc-2204.

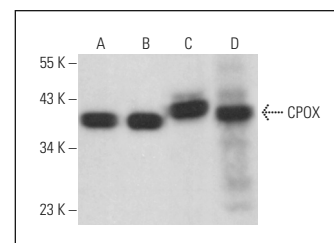
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



CPOX (B-9): sc-393388. Western blot analysis of CPOX expression in JAR (A), BJ (B), Jurkat (C) and Hep G2 (D) whole cell lysates and mouse liver tissue extract (E).



CPOX (B-9): sc-393388. Western blot analysis of CPOX expression in ALL-SIL (A), M1 (B) and C6 (C) cell lysates and rat uterus tissue extract (D).

SELECT PRODUCT CITATIONS

1. Paul, B.T., et al. 2019. Sideroflexin 4 affects Fe-S cluster biogenesis, iron metabolism, mitochondrial respiration and heme biosynthetic enzymes. *Sci. Rep.* 9: 19634.
2. Schary, N., et al. 2022. Identification and pharmacological modification of resistance mechanisms to protoporphyrin-mediated photodynamic therapy in human cutaneous squamous cell carcinoma cell lines. *Photodiagnosis Photodyn. Ther.* 39: 103004.

RESEARCH USE

For research use only, not for use in diagnostic procedures.