

# PPOX (E-20): sc-32364

## BACKGROUND

Protoporphyrinogen oxidase, the penultimate enzyme in the heme biosynthetic pathway, catalyzes the 6-electron oxidation of protoporphyrinogen IX to form protoporphyrin IX. The PPOX protein localizes to the inner membrane of mitochondria from various tissues, including heart, brain, placenta, lung, liver, skeletal muscle, kidney and pancreas. Genetic deficiency of PPOX results in variegate porphyria, a low penetrance, autosomal dominant disorder characterized by cutaneous photosensitivity and/or various neurological manifestations. The rare homozygous variant of VP is characterized by severe PPOX deficiency, and results in the onset of photosensitization by porphyrins in early childhood, skeletal abnormalities of the hand, and, less constantly, short stature, mental retardation and convulsions.

## REFERENCES

1. Taketani, S., et al. 1995. The human protoporphyrinogen oxidase gene (PPOX): organization and location to chromosome 1. *Genomics* 29: 698-703.
2. Nishimura, K., et al. 1995. Cloning of a human cDNA for protoporphyrinogen oxidase by complementation *in vivo* of a hemG mutant of *Escherichia coli*. *J. Biol. Chem.* 270: 8076-8080.
3. Puy, H., et al. 1996. Protoporphyrinogen oxidase: complete genomic sequence and polymorphisms in the human gene. *Biochem. Biophys. Res. Commun.* 226: 226-230.
4. Maneli, M.H., et al. 2003. Kinetic and physical characterisation of recombinant wildtype and mutant human protoporphyrinogen oxidases. *Biochim. Biophys. Acta* 1650: 10-21.
5. Wiman, A. et al. 2003. Nine novel mutations in the protoporphyrinogen oxidase gene in Swedish families with variegate porphyria. *Clin. Genet.* 64: 122-130.
6. Morgan, RR. et al. 2004. Identification of sequences required for the import of human protoporphyrinogen oxidase to mitochondria. *Biochem. J.* 377: 281-287.

## CHROMOSOMAL LOCATION

Genetic locus: PPOX (human) mapping to 1q23.3; Ppox (mouse) mapping to 1 H3.

## SOURCE

PPOX (E-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of PPOX of human origin.

## PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-32364 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## RESEARCH USE

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

## APPLICATIONS

PPOX (E-20) is recommended for detection of PPOX of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

PPOX (V-20) is also recommended for detection of PPOX in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for PPOX siRNA (h): sc-44783, PPOX siRNA (m): sc-44784, PPOX shRNA Plasmid (h): sc-44783-SH, PPOX shRNA Plasmid (m): sc-44784-SH, PPOX shRNA (h) Lentiviral Particles: sc-44783-V and PPOX shRNA (m) Lentiviral Particles: sc-44784-V.

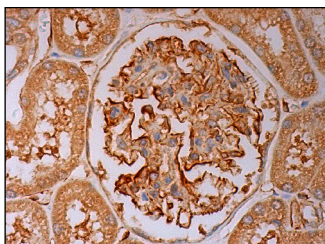
Molecular Weight of PPOX: 51 kDa.

Positive Controls: A549 cell lysate: sc-2413 or Hep G2 cell lysate: sc-2227.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941. 3) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

## DATA



PPOX (E-20): sc-32364. Immunoperoxidase staining of formalin fixed, paraffin-embedded human kidney tissue showing cytoplasmic and membrane staining of cells in glomeruli and cytoplasmic staining of cells in tubules.

## STORAGE

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