

# PPOX (42J-6): sc-100577

## BACKGROUND

Protoporphyrinogen oxidase, the penultimate enzyme in the heme biosynthetic pathway, catalyzes the six-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, R.R., et al. 2004. Identification of sequences required for the import of human protoporphyrinogen oxidase to mitochondria. *Biochem. J.* 377: 281-287.
7. <http://harvester.embl.de/harvester/P503/P50336.htm>

## CHROMOSOMAL LOCATION

Genetic locus: PPOX (human) mapping to 1q23.3.

## SOURCE

PPOX (42J-6) is a mouse monoclonal antibody raised against recombinant PPOX of human origin.

## PRODUCT

Each vial contains 50 µg IgG<sub>1</sub> kappa light chain in 0.5 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

## STORAGE

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

## APPLICATIONS

PPOX (42J-6) is recommended for detection of PPOX of human origin by Western Blotting (starting dilution 1:200, 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), 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).

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

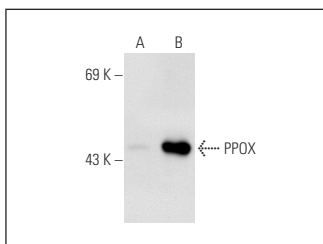
Molecular Weight of PPOX: 51 kDa.

Positive Controls: PPOX (m): 293T Lysate: sc-122735 or Hep G2 cell lysate: sc-2227.

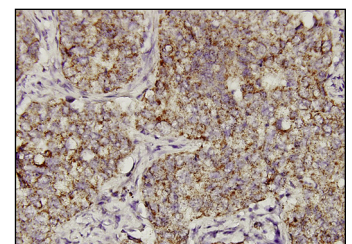
## RECOMMENDED SUPPORT REAGENTS

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. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

## DATA



PPOX (42J-6): sc-100577. Western blot analysis of PPOX expression in non-transfected: sc-117752 (A) and mouse PPOX transfected: sc-122735 (B) 293T whole cell lysates.



PPOX (42J-6): sc-100577. Immunoperoxidase staining of formalin-fixed, paraffin-embedded human lung, adenocarcinoma tissue showing cytoplasmic localization.

## RESEARCH USE

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

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) for detailed protocols and support products.