SZABO SCANDIC

Part of Europa Biosite

Produktinformation

- Forschungsprodukte & Biochemikalien
- Zellkultur & Verbrauchsmaterial
- Diagnostik & molekulare Diagnostik
- Laborgeräte & Service

Weitere Information auf den folgenden Seiten!
See the following pages for more information!

Lieferung & Zahlungsart

siehe unsere Liefer- und Versandbedingungen

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

SZABO-SCANDIC HandelsgmbH

Quellenstraße 110, A-1100 Wien

T. +43(0)1 489 3961-0

F. +43(0)1 489 3961-7

mail@szabo-scandic.com

www.szabo-scandic.com

linkedin.com/company/szaboscandic

SANTA CRUZ BIOTECHNOLOGY, INC.

PPOX (m): 293T Lysate: sc-122735

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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 (mouse) mapping to 1 H3.

PRODUCT

PPOX (m): 293T Lysate represents a lysate of mouse PPOX transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

PPOX (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive PPOX antibodies. Recommended use: 10-20 µl per lane.

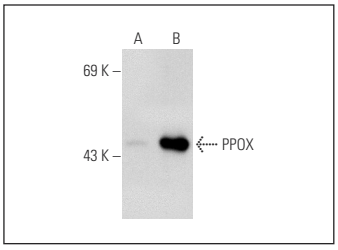
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

PPOX (42J-6): sc-100577 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse PPOX expression in PPOX transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

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.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.