

PTPS (D-8): sc-514628

BACKGROUND

The tetrahydrobiopterin (BH4) cofactor is essential for hepatic hydroxylase, which is involved in phenylalanine degradation and catecholamine and serotonin biosynthesis. BH4 is also an essential and limiting cofactor for all types of nitric oxide synthase. BH4 deficiency results in hyperphenylalaninemia and monoamine neurotransmitter depletion and is most commonly due to autosomal recessive mutation in 6-pyruvoyltetrahydropterin synthase (PTPS), the second enzyme for BH4 biosynthesis. The active site of PTPS consists of the pterin-anchoring Glu A107 neighbored by two catalytic motifs: a Zn(II) binding site and an intersubunit catalytic triad formed by Cys A42, Asp B88 and His B89. The active site of PTPS undergoes a Zn and Mg-dependent reaction that includes a triphosphate elimination, a stereospecific reduction and the oxidation of both side hydroxyl groups. The catalytic triad of PTPS is involved in the deprotonation of the side-chain carbons of substrates. In addition, Ser 19 of human PTPS may be a substrate for cGMP-dependent protein kinase type II phosphorylation *in vivo*, which is essential for normal activity of PTPS.

REFERENCES

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2. Thöny, B., et al. 1994. Chromosomal location of two human genes encoding tetrahydrobiopterin-metabolizing enzymes: 6-pyruvoyl-tetrahydropterin synthase maps to 11q22.3-q23.3, and pterin-4 α -carbinolamine dehydratase maps to 10q22. *Genomics* 19: 365-368.
3. Kluge, C., et al. 1996. Chromosomal localization, genomic structure and characterization of the human gene and a retropseudogene for 6-pyruvoyl-tetrahydropterin synthase. *Eur. J. Biochem.* 240: 477-484.
4. Laufs, S., et al. 1998. Retrovirus-mediated double transduction of the GTPCH and PTPS genes allows 6-pyruvoyltetrahydropterin synthase-deficient human fibroblasts to synthesize and release tetrahydrobiopterin. *J. Neurochem.* 71: 33-40.
5. Linscheid, P., et al. 1998. Regulation of 6-pyruvoyltetrahydropterin synthase activity and messenger RNA abundance in human vascular endothelial cells. *Circulation* 98: 1703-1706.
6. Ploom, T., et al. 1999. Crystallographic and kinetic investigation on the mechanism of 6-pyruvoyl tetrahydropterin synthase. *J. Mol. Biol.* 286: 851-860.

CHROMOSOMAL LOCATION

Genetic locus: PTS (human) mapping to 11q23.1.

SOURCE

PTPS (D-8) is a mouse monoclonal antibody raised against amino acids 1-145 representing full length PTPS of human origin.

PRODUCT

Each vial contains 200 μ g IgG₁ in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

PTPS (D-8) is recommended for detection of PTPS of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g 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 PTPS siRNA (h): sc-106734, PTPS shRNA Plasmid (h): sc-106734-SH and PTPS shRNA (h) Lentiviral Particles: sc-106734-V.

Molecular Weight (predicted) of PTPS: 16 kDa.

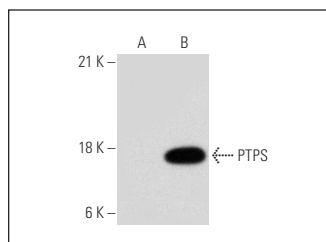
Molecular Weight (observed) of PTPS: 20 kDa.

Positive Controls: PTPS (h): 293 Lysate: sc-111356.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-mouse IgG-HRP: sc-2005 (dilution range: 1:2000-1:32,000) or Cruz Marker™ compatible goat anti-mouse IgG-HRP: sc-2031 (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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use goat anti-mouse IgG-FITC: sc-2010 (dilution range: 1:100-1:400) or goat anti-mouse IgG-TR: sc-2781 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



PTPS (D-8): sc-514628. Western blot analysis of PTPS expression in non-transfected: sc-110766 (A) and human PTPS transfected: sc-111356 (B) 293 whole cell lysates.

STORAGE

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

RESEARCH USE

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

PROTOCOLS

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