SANTA CRUZ BIOTECHNOLOGY, INC.

PBGS (E-10): sc-398308

BACKGROUND

PGBS (porphobilinogen synthase), an enzyme that belongs to the ALADH family, is composed of eight identical subunits and catalyzes the condensation of two molecules of δ-aminolevulinate to form porphobilinogen, a precursor of heme, cytochromes and other hemoproteins. It also catalyzes the second step in the porphyrin and heme biosynthetic pathway in which zinc is essential for enzymatic activity. PGBS is inhibited by lead. A defect in the gene encoding PGBS, ALAD, can cause increased sensitivity to lead poisoning and acute hepatic porphyria, a group of inherited disorders caused by partial enzyme defects in heme biosynthesis, which includes acute intermittent porphyria, variegate porphyria and hereditary coproporphyria. There are two common alleles of ALAD, ALAD*2 and ALAD*1. When exposed to environmental lead, individuals heterozygous or homozygous for ALAD*2 Asn 59 have significantly higher blood lead levels than do ALAD*1 Lys 59 homozygotes.

REFERENCES

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2. Gabriel, D., et al. 2005. Human erythrocyte δ-aminolevulinate dehydratase inhibition by monosaccharides is not mediated by oxidation of enzyme sulfhydryl groups. Cell Biol. Int. 29: 669-674.
3. Hernandez, A.F., et al. 2005. Changes in erythrocyte enzymes in humans long-term exposed to pesticides: influence of several markers of individual susceptibility. Toxicol. Lett. 159: 13-21.
4. Aisemberg, J., et al. 2005. Comparative study on two freshwater invertebrates for monitoring environmental lead exposure. Toxicology 210: 45-53.
5. Lee, M.K., et al. 2005. Du-zhong *(Eucommia ulmoides* Oliv.) cortex water extract alters heme biosynthesis and erythrocyte antioxidant defense system in lead-administered rats. J. Med. Food 8: 86-92.
6. Roza, T., et al. 2005. 2,3-dimercapto-1-propanol does not alter the porphobilinogen synthase inhibition but decreases the mercury content in liver and kidney of suckling rats exposed to $HgCl_2$. Basic Clin. Pharmacol. Toxicol. 96: 302-308.
7. Sawada, N., et al. 2005. The activation mechanism of human porphobilinogen synthase by 2-mercaptoethanol: intrasubunit transfer of a reserve zinc ion and coordination with three cysteines in the active center. J. Biol. Inorg. Chem. 10: 199-207.

CHROMOSOMAL LOCATION

Genetic locus: ALAD (human) mapping to 9q32; Alad (mouse) mapping to 4 B3.

SOURCE

PBGS (E-10) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 142-159 within an internal region of PBGS of human origin.

STORAGE

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

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-398308 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

APPLICATIONS

PBGS (E-10) is recommended for detection of PBGS 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 PBGS siRNA (h): sc-61385, PBGS siRNA (m): sc-61386, PBGS shRNA Plasmid (h): sc-61385-SH, PBGS shRNA Plasmid (m): sc-61386-SH, PBGS shRNA (h) Lentiviral Particles: sc-61385-V and PBGS shRNA (m) Lentiviral Particles: sc-61386-V.

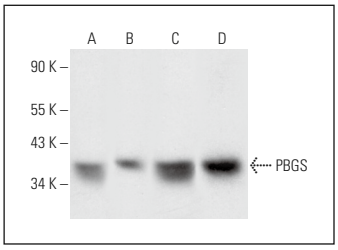
Molecular Weight of PBGS: 37-39 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, HEL 92.1.7 cell lysate: sc-2270 or human liver extract: sc-363766.

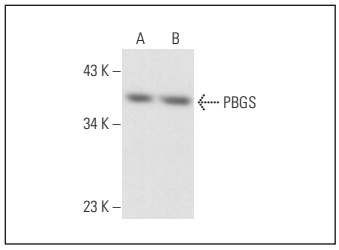
RECOMMENDED SUPPORT REAGENTS

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



PBGS (E-10): sc-398308. Western blot analysis of PBGS expression in HEL 92.1.7 (**A**), Hep G2 (**B**) and TF-1 (**C**) whole cell lysates and human liver tissue extract (**D**).



PBGS (E-10): sc-398308. Western blot analysis of PBGS expression in Hep G2 (**A**) and K-562 (**B**) whole cell lysates.

RESEARCH USE

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