

PBGS (G-9): sc-365095

BACKGROUND

PBGS (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. PBGS is inhibited by lead. A defect in the gene encoding PBGS, 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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CHROMOSOMAL LOCATION

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

SOURCE

PBGS (G-9) is a mouse monoclonal antibody raised against amino acids 1-300 mapping at the N-terminus of PBGS of human origin.

RESEARCH USE

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

PRODUCT

Each vial contains 200 μ g IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

PBGS (G-9) 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 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 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.

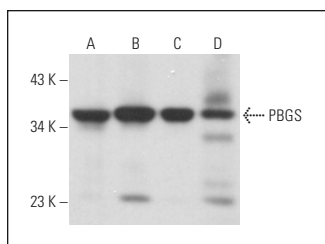
Molecular Weight of PBGS: 37-39 kDa.

Positive Controls: human liver extract: sc-363766, mouse liver extract: sc-2256 or PBGS (m): 293T Lysate: sc-125787.

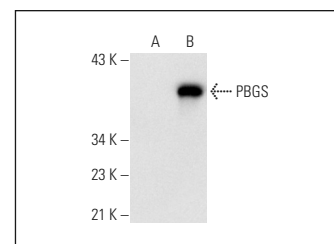
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ 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-IgG κ BP-FITC: sc-516140 or m-IgG κ 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 (G-9): sc-365095. Western blot analysis of PBGS expression in human liver (A), mouse liver (B), rat liver (C) and mouse spleen (D) tissue extracts.



PBGS (G-9): sc-365095. Western blot analysis of PBGS expression in non-transfected: sc-117752 (A) and mouse PBGS transfected: sc-125787 (B) 293T 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.

PROTOCOLS

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