

PBGS (A-7): sc-271585

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 -aminolevulinic acid 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 coproporphyrin. 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.

CHROMOSOMAL LOCATION

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

SOURCE

PBGS (A-7) is a mouse monoclonal antibody raised against amino acids 1-300 mapping at the N-terminus of PBGS of human origin.

PRODUCT

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

PBGS (A-7) is available conjugated to agarose (sc-271585 AC), 500 μ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-271585 HRP), 200 μ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-271585 PE), fluorescein (sc-271585 FITC), Alexa Fluor® 488 (sc-271585 AF488), Alexa Fluor® 546 (sc-271585 AF546), Alexa Fluor® 594 (sc-271585 AF594) or Alexa Fluor® 647 (sc-271585 AF647), 200 μ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-271585 AF680) or Alexa Fluor® 790 (sc-271585 AF790), 200 μ g/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

PBGS (A-7) 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), 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 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.

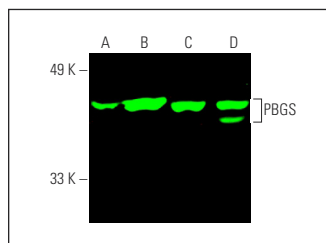
Molecular Weight of PBGS: 37-39 kDa.

Positive Controls: human liver extract: sc-363766, mouse liver extract: sc-2256 or rat liver extract: sc-2395.

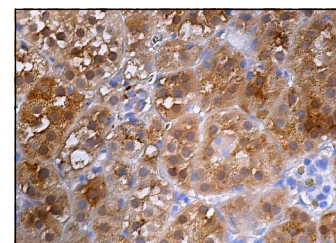
RECOMMENDED SUPPORT REAGENTS

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

DATA



PBGS (A-7): sc-271585. Near-Infrared western blot analysis of PBGS expression in human liver (A), mouse liver (B), rat liver (C) and human adrenal gland (D) tissue extracts. Blocked with UltraCruz® Blocking Reagent: sc-516214. Detection reagent used: m-IgG κ BP-CFL 680: sc-516180.



PBGS (A-7): sc-271585. Immunoperoxidase staining of formalin fixed, paraffin-embedded human adrenal gland tissue showing cytoplasmic and nuclear staining of glandular cells.

SELECT PRODUCT CITATIONS

1. Desgardin, A.D., et al. 2012. Regulation of δ -aminolevulinic acid dehydratase by Krüppel-like factor 1. PLoS ONE 7: e46482.
2. Palasuberniam, P., et al. 2019. Ferrochelatase deficiency abrogated the enhancement of aminolevulinic acid-mediated protoporphyrin IX by iron chelator deferoxamine. Photochem. Photobiol. 95: 1052-1059.

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 for detailed protocols and support products.