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Diagnostik & molekulare Diagnostik



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Lieferung & Zahlungsart

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Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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PBGS (m): 293T Lysate: sc-125787

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 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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- 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 (mouse) mapping to 4 B3.

PRODUCT

PBGS (m): 293T Lysate represents a lysate of mouse PBGS transfected 293T cells and is provided as 100 μ g protein in 200 μ l SDS-PAGE buffer.

RESEARCH USE

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

APPLICATIONS

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

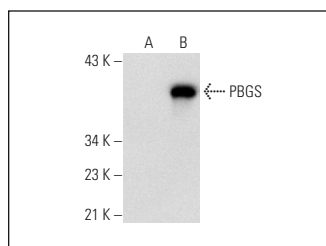
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

PBGS (G-9): sc-365095 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse PBGS expression in PBGS transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

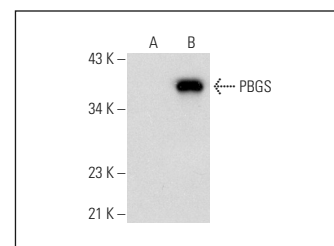
RECOMMENDED SUPPORT REAGENTS

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.

DATA



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.



PBGS (A-7): sc-271585. 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 -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

PROTOCOLS

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