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Lieferung & Zahlungsart

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- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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SANTA CRUZ BIOTECHNOLOGY, INC.

PBGD (m): 293T Lysate: sc-122406

BACKGROUND

PBGD (porphobilinogen deaminase), also designated hydroxymethylbilane synthase, is a cytoplasmic enzyme found in the heme synthesis pathway. PBGD belongs to the HMBS (hydroxymethylbilane synthase) family. Deficiency of PBGD causes errors in pyrrole metabolism, which in turn leads to an inherited autosomal disorder called acute intermittent porphyria (AIP). AIP is characterized by acute attacks of neurological dysfunctions with hypertension, tachycardia, peripheral neurologic disturbances, abdominal pain and excessive amounts of aminolevulinic acid and porphobilinogen in the urine.

REFERENCES

1. Grandchamp, B., De Verneuil, H., Beaumont, C., Chretien, S., Walter, O. and Nordmann, Y. 1987. Tissue-specific expression of porphobilinogen deaminase. Two isoenzymes from a single gene. Eur. J. Biochem. 162: 105-110.
2. Mustajoki, S., Laine, M., Lahtela, M., Mustajoki, P., Peltonen, L. and Kauppinen, R. 2000. Acute intermittent porphyria: expression of mutant and wild-type porphobilinogen deaminase in COS-1 cells. Mol. Med. 6: 670-679.
3. Schneider-Yin, X., Hergersberg, M., Schuurmans, M.M., Gregor, A. and Minder, E.I. 2004. Mutation hotspots in the human porphobilinogen deaminase gene: recurrent mutations G111R and R173Q occurring at CpG motifs. J. Inherit. Metab. Dis. 27: 625-631.
4. Neuvians, T.P., Gashaw, I., Sauer, C.G., von Ostau, C., Kliesch, S., Bergmann, M., Hacker, A. and Grobholz, R. 2005. Standardization strategy for quantitative PCR in human seminoma and normal testis. J. Biotechnol. 117: 163-171.
5. von und zu Fraunberg, M., Pischik, E., Udd, L. and Kauppinen, R. 2005. Clinical and biochemical characteristics and genotype-phenotype correlation in 143 Finnish and Russian patients with acute intermittent porphyria. Medicine 84: 35-47.
6. Sheppard, L. and Dorman, T. 2005. Anesthesia in a child with homozygous porphobilinogen deaminase deficiency: a severe form of acute intermittent porphyria. Paediatr. Anaesth. 15: 426-428.
7. SWISS-PROT/TrEMBL (P08397). World Wide Web URL: http://www.expasy.ch/sprot/sprot-top.html
8. http://harvester.embl.de/harvester/P083/P08397.htm

CHROMOSOMAL LOCATION

Genetic locus: Hmbs (mouse) mapping to 9 A5.2.

PRODUCT

PBGD (m): 293T Lysate represents a lysate of mouse PBGD transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

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.

RESEARCH USE

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

APPLICATIONS

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

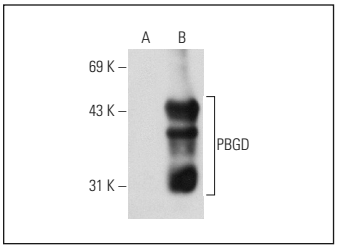
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

PBGD (B-3): sc-166742 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse PBGD expression in PBGD transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

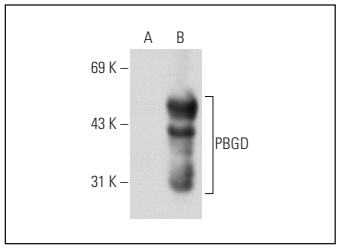
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-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.

DATA



PBGD (B-3): sc-166742. Western blot analysis of PBGD expression in non-transfected: sc-117752 (**A**) and mouse PBGD transfected: sc-122406 (**B**) 293T whole cell lysates.



PBGD (E-9): sc-166743. Western blot analysis of PBGD expression in non-transfected: sc-117752 (**A**) and mouse PBGD transfected: sc-122406 (**B**) 293T whole cell lysates.

PROTOCOLS

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