

PBGD (I-17): sc-46621

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) which 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

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- 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. Inher. Metab. Dis.* 27: 625-631.
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- 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.

CHROMOSOMAL LOCATION

Genetic locus: HMBS (human) mapping to 11q23.3; Hmbs (mouse) mapping to 9 A5.2.

SOURCE

PBGD (I-17) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of PBGD of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-46621 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

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

APPLICATIONS

PBGD (I-17) is recommended for detection of PBGD of human and to a lesser extent, mouse origin by Western Blotting (starting dilution 1:200, 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).

PBGD (I-17) is also recommended for detection of PBGD in additional species, including bovine.

Suitable for use as control antibody for PBGD siRNA (h): sc-45702, PBGD siRNA (m): sc-45703, PBGD shRNA Plasmid (h): sc-45702-SH, PBGD shRNA Plasmid (m): sc-45703-SH, PBGD shRNA (h) Lentiviral Particles: sc-45702-V and PBGD shRNA (m) Lentiviral Particles: sc-45703-V.

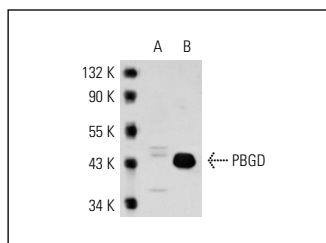
Molecular Weight of PBGD: 42-44 kDa.

Positive Controls: U-937 cell lysate: sc-2239, PBGD (h): 293 Lysate: sc-110913 or Hep G2 cell lysate: sc-2227.

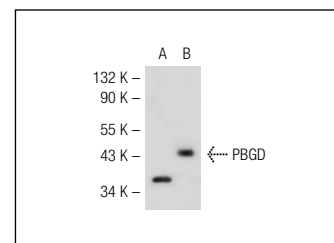
RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



PBGD (I-17): sc-46621. Western blot analysis of PBGD expression in non-transfected: sc-110760 (A) and human PBGD transfected: sc-110913 (B) 293 whole cell lysates.



PBGD (I-17): sc-46621. Western blot analysis of PBGD expression in non-transfected: sc-117752 (A) and mouse PBGD transfected: sc-122406 (B) 293T whole cell lysates.

RESEARCH USE

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

PROTOCOLS

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