SANTA CRUZ BIOTECHNOLOGY, INC.

PBGD (H-300): sc-67037

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

PGBD (porphobilinogen deaminase), also designated Hydroxymethylbilane synthase, is a cytoplasmic enzyme found in the heme synthesis pathway. PGBD belongs to the HMBS (hydroxymethylbilane synthase) family. Deficiency of PGBD 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

1. Grandchamp, B., et al. 1987. Tissue-specific expression of porphobilinogen deaminase. Two isoenzymes from a single gene. Eur. J. Biochem. 162: 105-110.
2. Mustajoki, S., et al. 2000. Acute intermittent porphyria: expression of mutant and wildtype porphobilinogen deaminase in COS-1 cells. Mol. Med. 6: 670-679.

CHROMOSOMAL LOCATION

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

SOURCE

PBGD (H-300) is a rabbit polyclonal antibody raised against amino acids 62-361 mapping at 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.

RESEARCH USE

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

APPLICATIONS

PBGD (H-300) is recommended for detection of PBGD of mouse, rat and human 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 (H-300) is also recommended for detection of PBGD in additional species, including equine, canine, bovine and porcine.

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.

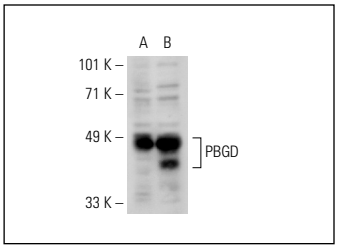
Molecular Weight of PBGD: 42-44 kDa.

Positive Controls: U-937 cell lysate: sc-2239 or Hep G2 cell lysate: sc-2227.

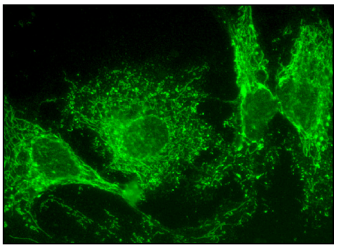
RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



PBGD (H-300): sc-67037. Western blot analysis of PBGD expression in U-937 (**A**) and Hep G2 (**B**) whole cell lysates.



PBGD (H-300): sc-67037. Immunofluorescence staining of formalin-fixed Hep G2 cells showing cytoplasmic localization.

SELECT PRODUCT CITATIONS

1. Unzu, C., et al. 2012. Transient and intensive pharmacological immunosuppression fails to improve AAV-based liver gene transfer in non-human primates. J. Transl. Med. 10: 122.

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 or our catalog for detailed protocols and support products.