

# PBGD (H-11): sc-166842

## 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., 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 wild-type 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-11) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 340-365 at the C-terminus of PBGD of human origin.

## PRODUCT

Each vial contains 200 µg IgG<sub>2b</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-166842 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

## APPLICATIONS

PBGD (H-11) is recommended for detection of PBGD of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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-11) is also recommended for detection of PBGD in additional species, including equine.

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.

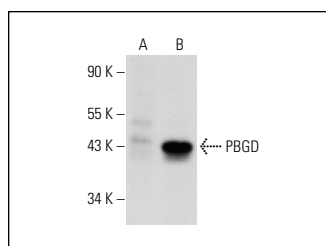
Molecular Weight of PBGD: 42-44 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, rat liver extract: sc-2395 or PBGD (h): 293 Lysate: sc-110913.

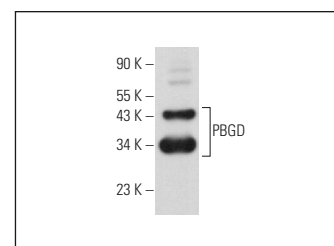
## RECOMMENDED SUPPORT REAGENTS

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. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

## DATA



PBGD (H-11): sc-166842. Western blot analysis of PBGD expression in non-transfected: sc-110760 (A) and human PBGD transfected: sc-110913 (B) 293 whole cell lysates.



PBGD (H-11): sc-166842. Western blot analysis of PBGD expression in rat liver tissue extract.

## SELECT PRODUCT CITATIONS

1. Wang, Z., et al. 2019. Neurogenic niche conversion strategy induces migration and functional neuronal differentiation of neural precursor cells following brain injury. *Stem Cells Dev.* 29: 235-248.
2. Bustad, H.J., et al. 2020. A pharmacological chaperone therapy for acute intermittent porphyria. *Mol. Ther.* 28: 677-689.

## 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](http://www.scbt.com) for detailed protocols and support products.