SANTA CRUZ BIOTECHNOLOGY, INC.

PBGD (h): 293 Lysate: sc-110913

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

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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.
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CHROMOSOMAL LOCATION

Genetic locus: HMBS (human) mapping to 11q23.3.

PRODUCT

PBGD (h): 293 Lysate represents a lysate of human PBGD transfected 293 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.

APPLICATIONS

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

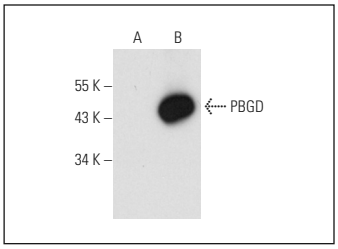
Control 293 Lysate: sc-110760 is available as a Western Blotting negative control lysate derived from non-tranfected 293 cells.

PBGD (B-6): sc-166788 is recommended as a positive control antibody for Western Blot analysis of enhanced human PBGD expression in PBGD transfected 293 cells (starting dilution 1:100, dilution range 1:100-1:1,000).

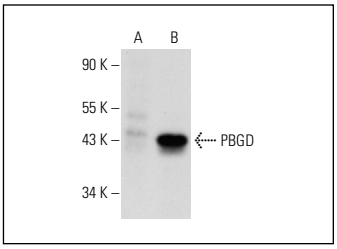
RECOMMENDED SUPPORT REAGENTS

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-6): sc-166788. 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 non-transfected: sc-110760 (**A**) and human PBGD transfected: sc-110913 (**B**) 293 whole cell lysates.

RESEARCH USE

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

PROTOCOLS

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