

GMPPB (F-11): sc-100483

BACKGROUND

GMPPB (GDP-mannose pyrophosphorylase B) is a 360 amino acid protein that belongs to the transferase hexapeptide repeat family and is involved in protein modification pathways. Functioning as a GDP-mannose pyrophosphorylase, GMPPB enzymatically catalyzes the conversion of mannose-1-phosphate and GTP to GDP-mannose and a free phosphate, a reaction that is involved in the production of N-linked oligosaccharides. Defects in the gene encoding GMPPB that cause errors in the glycosylation pathway may lead to congenital disorders of glycosylation (CDG). CDGs are multisystemic diseases that often involve both the central and peripheral nervous systems and are often characterized by endocrine and coagulation disorders. GMPPB is expressed as two isoforms due to alternative splicing events.

REFERENCES

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3. Grubenmann, C.E., Frank, C.G., Hülsmeier, A.J., Schollen, E., Matthijs, G., Mayatepek, E., Berger, E.G., Aebi, M. and Hennet, T. 2004. Deficiency of the first mannosylation step in the N-glycosylation pathway causes congenital disorder of glycosylation type Ik. *Hum. Mol. Genet.* 13: 535-542.
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CHROMOSOMAL LOCATION

Genetic locus: GMPPB (human) mapping to 3p21.31; Gmppb (mouse) mapping to 9 F2.

SOURCE

GMPPB (F-11) is a mouse monoclonal antibody raised against recombinant GMPPB of human origin.

PRODUCT

Each vial contains 50 µg IgG_{2b} kappa light chain in 0.5 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

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.

APPLICATIONS

GMPPB (F-11) is recommended for detection of GMPPB 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), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for GMPPB siRNA (h): sc-78444, GMPPB siRNA (m): sc-145649, GMPPB shRNA Plasmid (h): sc-78444-SH, GMPPB shRNA Plasmid (m): sc-145649-SH, GMPPB shRNA (h) Lentiviral Particles: sc-78444-V and GMPPB shRNA (m) Lentiviral Particles: sc-145649-V.

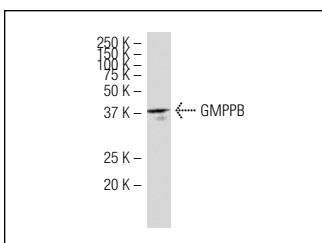
Molecular Weight of GMPPB: 40 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227.

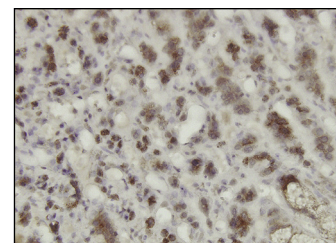
RECOMMENDED SUPPORT REAGENTS

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, 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 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



GMPPB (F-11): sc-100483. Western blot analysis of GMPPB expression in Hep G2 whole cell lysate.



GMPPB (F-11): sc-100483. Immunoperoxidase staining of formalin-fixed, paraffin-embedded human adrenal gland tissue showing cytoplasmic localization.

PROTOCOLS

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