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GMPPB Rabbit pAb

Catalog # AP98240

Product Information

Application	WB, IHC-P, IHC-F, IF, E
Primary Accession	Q9Y5P6
Other Accession	Q9Y5P6
Predicted	Rat, Pig, Human, Mouse, Rabbit, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	39834
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human GMPPB
Epitope Specificity	111-210/360
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the transferase hexapeptide repeat family.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	GMPPB 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.

Additional Information

Gene ID	29925
Other Names	LGMDR19; MDDGA14; MDDGB14; MDDGC14; E430010H19; RGD1560458; GMPPB_HUMAN; GMPPB; GDP-mannose pyrophosphorylase B; GTP-mannose-1-phosphate guanylyltransferase beta; 2.7.7.13; GMPPB_MOUSE; mannose-1-phosphate guanyltransferase beta
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	GMPPB (HGNC:22932)
Function	Catalytic subunit of the GMPPA-GMPPB mannose-1-phosphate guanylyltransferase complex (PubMed:33986552). Catalyzes the formation of GDP-mannose, an essential precursor of glycan moieties of glycoproteins and glycolipids (PubMed:33986552). Can catalyze the reverse reaction in vitro (PubMed:33986552). Together with GMPPA regulates GDP-alpha-D-mannose levels (PubMed:33986552).
Cellular Location	Cytoplasm.
Tissue Location	[Isoform 1]: Ubiquitously expressed, including in brain and skeletal muscle.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.