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G6PC Rabbit pAb

Catalog # AP99839

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P35575
Other Accession	P35575
Reactivity	Human
Predicted	Rat, Pig, Mouse, Rabbit, Dog, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	40484
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Glucose 6 phosphatase alpha
Epitope Specificity	81-180/357
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 glucose-6-phosphatase family.
DISEASE	Defects in G6PC are the cause of glycogen storage disease type 1A (GSD1A) [MIM:232200]. A metabolic disorder characterized by impairment of terminal steps of glycogenolysis and gluconeogenesis. Patients manifest a wide range of clinical symptoms and biochemical abnormalities, including hypoglycemia, severe hepatomegaly due to excessive accumulation of glycogen, kidney enlargement, growth retardation, lactic acidemia, hyperlipidemia, and hyperuricemia.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Glucose-6-phosphatase (G6Pase) is a multi-subunit integral membrane protein of the endoplasmic reticulum that is composed of a catalytic subunit and transporters for G6P, inorganic phosphate, and glucose. This gene (G6PC) is one of the three glucose-6-phosphatase catalytic-subunit-encoding genes in human: G6PC, G6PC2 and G6PC3. Glucose-6-phosphatase catalyzes the hydrolysis of D-glucose 6-phosphate to D-glucose and orthophosphate and is a key enzyme in glucose homeostasis, functioning in gluconeogenesis and glycogenolysis. Mutations in this gene cause glycogen storage disease type I (GSD1). This disease, also known as von Gierke disease, is a metabolic disorder characterized by severe hypoglycemia associated with the accumulation of glycogen and fat in the liver and kidneys.[provided by RefSeq, Feb 2011]

Additional Information

Gene ID	2538
Other Names	G6PC; G6Pase; G6PT; GSD1; GSD1a; Glc-6-Pase; Psme3; G6PC1_CANLF; G6PC1;

	Glucose-6-phosphatase (G-6-Pase | G6Pase); 3.1.3.9; G6PC1_HUMAN; Glucose-6-phosphatase alpha (G6Pase-alpha); G6PC1_MOUSE; G6PC1_RAT;
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,Flow-Cyt=0.2 ug/test
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	G6PC1 (HGNC:4056)
Synonyms	G6PC, G6PT
Function	Hydrolyzes glucose-6-phosphate to glucose in the endoplasmic reticulum. Forms with the glucose-6-phosphate transporter (SLC37A4/G6PT) the complex responsible for glucose production in the terminal step of glycogenolysis and gluconeogenesis. Hence, it is the key enzyme in homeostatic regulation of blood glucose levels.
Cellular Location	Endoplasmic reticulum membrane; Multi-pass membrane protein

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