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PHKG2 Rabbit pAb

Catalog # AP96324

Product Information

Application	WB
Reactivity	Human
Predicted	Rat, Pig, Mouse, Rabbit, Dog, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	46 KDa
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human PHKG2
Epitope Specificity	81-180/406
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 protein kinase superfamily. CAMK Ser/Thr protein kinase family.Contains 1 protein kinase domain.
SUBUNIT	Hexadecamer of 4 heterotetramers, each composed of alpha, beta, gamma, and delta subunits. Alpha (PHKA1 or PHKA2) and beta (PHKB) are regulatory subunits, gamma (PHKG1 or PHKG2) is the catalytic subunit, and delta is calmodulin.
DISEASE	Defects in PHKG2 are a cause of glycogen storage disease type 9C (GSD9C) [MIM:613027]. A metabolic disorder manifesting in infancy with hepatomegaly, growth retardation, hypotonia, liver dysfunction, and elevated plasma aminotransferases and lipids. These symptoms improve with age in most cases; however, some patients may develop hepatic fibrosis or cirrhosis.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Phosphorylase kinase is a polymer of 16 subunits, four each of alpha, beta, gamma and delta. The alpha subunit includes the skeletal muscle and hepatic isoforms, encoded by two different genes. The beta subunit is the same in both the muscle and hepatic isoforms, and encoded by one gene. The gamma subunit also includes the skeletal muscle and hepatic isoforms, and the hepatic isoform is encoded by this gene. The delta subunit is a calmodulin and can be encoded by three different genes. The gamma subunits contain the active site of the enzyme, whereas the alpha and beta subunits have regulatory functions controlled by phosphorylation. The delta subunit mediates the dependence of the enzyme on calcium concentration. Mutations in this gene cause glycogen storage disease type 9C, also known as autosomal liver glycogenosis. Alternatively spliced transcript variants encoding different isoforms have been identified in this gene.

Additional Information

Gene ID	5261

Other Names	PHKG2_HUMAN; PHKG2; PHK-gamma-LT; PHK-gamma-T; PSK-C3; Phosphorylase kinase subunit gamma-2; 2.7.11.19; GSD9C; phosphorylase kinase catalytic subunit gamma 2; phosphorylase kinase, gamma 2 (testis)
Dilution	WB=1:500-2000
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.
Name	PHKG2 Rabbit pAb
Function	Catalytic subunit of the phosphorylase b kinase (PHK), which mediates the neural and hormonal regulation of glycogen breakdown (glycogenolysis) by phosphorylating and thereby activating glycogen phosphorylase (PYGB, PYGL or PYGM). May regulate glycogeneolysis in the testis. In vitro, phosphorylates TNNI3, TNNT2, MAPT/TAU, GAP43 and NRGN/RC3.

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