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PHKA1 Polyclonal Antibody

Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP58257

Product Information

Application	WB, IHC-P, IHC-F, IF, E
Primary Accession	P46020
Reactivity	Rat, Dog, Bovine
Host	Rabbit
Clonality	Polyclonal
Calculated MW	137312
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human PHKA1
Epitope Specificity	51-150/1233
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Cell membrane; Lipid-anchor; Cytoplasmic side (Potential).
SIMILARITY	Belongs to the phosphorylase b kinase regulatory chain family.
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.
Post-translational modifications	Although the final Cys may be farnesylated, the terminal tripeptide is probably not removed, and the C-terminus is not methylated (By similarity).
DISEASE	Glycogen storage disease 9D (GSD9D) [MIM:300559]: A metabolic disorder characterized by slowly progressive, predominantly distal muscle weakness and atrophy. Clinical features include exercise intolerance with early fatigability, pain, cramps and occasionally myoglobinuria. Note=The disease is caused by mutations affecting the gene represented in this entry.
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, and the skeletal muscle isoform is encoded by this gene. 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, which are encoded by two different genes. 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 9D, also known as X-linked muscle glycogenosis. Alternatively spliced transcript variants encoding different isoforms have been identified in this gene. A pseudogene has been found on chromosome 1.

Additional Information

Gene ID	5255
Other Names	Phosphorylase b kinase regulatory subunit alpha, skeletal muscle isoform, Phosphorylase kinase alpha M subunit, PHKA1, PHKA
Target/Specificity	Muscle specific. Isoform 1 is predominant in vastus lateralis muscle. Isoform 2 predominates slightly in heart, and it predominates clearly in the other tissues tested.
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Format	0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glyce
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	PHKA1
Synonyms	PHKA
Function	Phosphorylase b kinase catalyzes the phosphorylation of serine in certain substrates, including troponin I. The alpha chain may bind calmodulin.
Cellular Location	Cell membrane; Lipid-anchor; Cytoplasmic side
Tissue Location	Muscle specific. Isoform 1 is predominant in vastus lateralis muscle. Isoform 2 predominates slightly in heart, and it predominates clearly in the other tissues tested