

Product name:	PHKB Rabbit Polyclonal Antibody
Cat number:	ABN16073
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human KPBB. AA range:661-710
Reactivity:	Human,Mouse
Applications:	WB 1:500-1:2000,IHC 1:50-1:300
Molecular Weight:	124kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

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, encoded by this gene, which is a member of the phosphorylase b kinase regulatory subunit family. The gamma subunit also includes the skeletal muscle and hepatic isoforms, 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 9B, also known as phosphorylase kinase deficiency. Defects in PHKB are the cause of glycogen storage disease type 9B (GSD9B) [MIM:261750]; also known as phosphorylase kinase deficiency of liver and muscle (PKD). GSD9B is a metabolic disorder characterized by hepatomegaly, only slightly elevated transaminases and plasma lipids, clinical improvement with increasing age, and remarkably no clinical muscle involvement. Biochemical observations suggest that this mild phenotype is caused by an incomplete holoenzyme that lacks the beta subunit, but that may possess residual activity. Enzyme regulation: By phosphorylation of various serine residues. Function: Phosphorylase b kinase catalyzes the phosphorylation of serine in certain substrates, including troponin I. The beta chain acts as a regulatory unit and modulates the activity of the holoenzyme in response to phosphorylation. Pathway: Glycan biosynthesis; glycogen metabolism. Similarity: Belongs to the phosphorylase b kinase regulatory chain family. Subunit: Polymer of 16 chains, four each of alpha, beta, gamma, and delta. Alpha and beta are regulatory chains, gamma is the catalytic chain, and delta is calmodulin.