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Rabbit Anti-PHKB antibody

SL4032R

Product Name:	PHKB
Chinese Name:	磷酸化酶β抗体
Alias:	Phosphorylase B; Phosphorylase kinase B; Phosphorylase kinase beta polypeptide; Phosphorylase kinase beta subunit; DKFZp781E15103; FLJ41698; KPBB_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,Rabbit,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	124kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human PHKB:45-160/1093
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	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 of liver and muscle. Alternatively spliced transcript variants encoding different isoforms have been identified in this gene. Two pseudogenes have been found on chromosomes 14 and 20, respectively.[provided by RefSeq, Feb 2010]. **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. **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. **Subcellular Location:** Cell membrane; Lipid-anchor; Cytoplasmic side (Potential). **Post-translational modifications:** Ser-701 is probably phosphorylated by PKA. Although the final Cys may be farnesylated, the terminal tripeptide is probably not removed, and the C-terminus is not methylated. **DISEASE:** 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 hepathomegaly, 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. **Similarity:** Belongs to the phosphorylase b kinase regulatory chain family. **SWISS:** Q93100 **Gene ID:** 5257 **Database links:**

	Entrez Gene: 5257Human Entrez Gene: 102093Mouse Entrez Gene: 361377Rat Omim: 172490Human SwissProt: Q93100Human SwissProt: Q7TSH2Mouse Unigene: 78060Human Unigene: 237296Mouse Unigene: 6488Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. 磷酸化酶A是磷酸化酶的活性状态，而磷酸化酶B是其无活性状态。6-磷酸葡萄糖结合磷酸化酶后，磷酸化酶由无活性变为活化态，的加长,之后，又回到原来无活性状态，它充当了个桥梁作用。