



PHKA1/2 Monoclonal Antibody

Catalog No	YP-mAb-14909
Isotype	IgG
Reactivity	Human;Mouse;Rat
Applications	WB
Gene Name	PHKA1/PHKA2
Immunogen	The antiserum was produced against synthesized peptide derived from human KPB1/2. AA range:31-80
Specificity	PHKA1/2 Monoclonal Antibody detects endogenous levels of PHKA1/2 protein.
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-1:2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	533411D17;98318K24Rik;kinase PHKA1;KPB1;KPB1_HUMAN;MGC13264;Pcyt1b;PHKA;PHKA1;Phosphorylase b kinase regulatory subunit alpha;skeletal muscle isoform;Phosphorylase kinase alpha M subunit;phosphorylase kinase;alpha 1 (muscle);RP23 21E2.1;dJ499B1.2 (phosphorylase kinase;alpha 2 (liver) (PYK));GSD9A;PHK;PHKLA;Phosphorylase b kinase alpha regulatory chain;liver isoform;liver;alpha-2 subunit;Phosphorylase kinase alpha 2;phosphorylase kinase alpha L subunit;phosphorylase kinase alpha subunit;phosphorylase kinase deficiency;liver (glycogen storage disease type VIII;liver (glycogen storage disease);alpha 2 (liver);glycogen storage disease IX;PYK;PYKL;XLG;XLG2;PHKA1/2;PHKA1/PHKA2;Phosphorylase b kinase regulatory subunit alpha skeletal muscle isoform/Phosphorylase b kinase regulatory subunit alpha liver isoform;PHKA2;Phosphorylase kinase alpha L sub; liver isoform
Observed Band	137kD
Calculated Molecular Weight	137kD
Cell Pathway	Cell membrane ; Lipid-anchor ; Cytoplasmic side .
Tissue 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.



Function

disease: Defects in PHKA1 are the cause of glycogen storage disease type 9D (GSD9D) [MIM:300559]; also known as X-linked muscle glycogenosis. GSD9D is 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. enzyme regulation: By phosphorylation of various serine residues. Allosteric regulation by calcium. function: Phosphorylase b kinase catalyzes the phosphorylation of serine in certain substrates, including troponin I. The alpha chain may bind calmodulin. 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 d

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, 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 varian

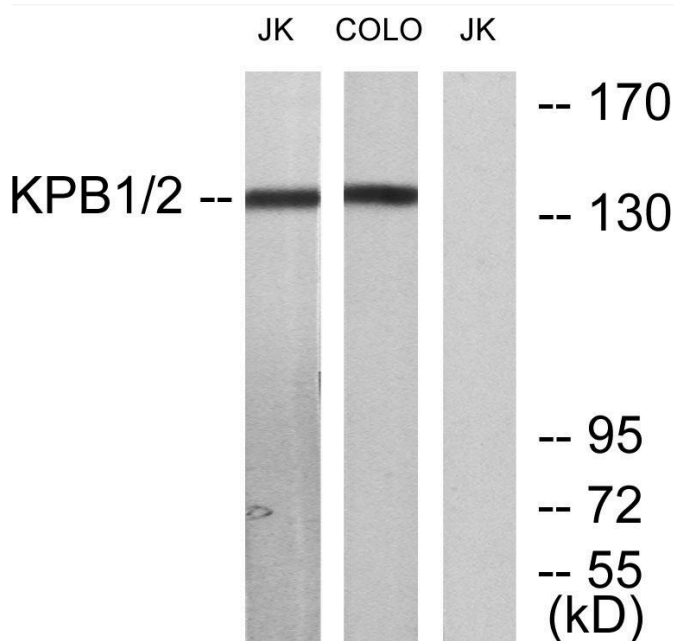
matters needing attention

Avoid repeated freezing and thawing!

Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western Blot analysis of various cells using PHKA1/2 Monoclonal Antibody