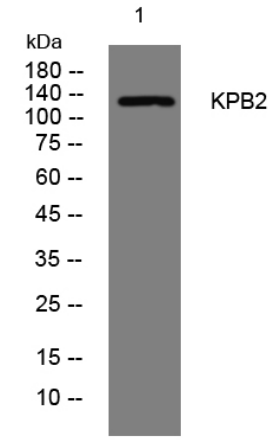
KPB2 rabbit pAb

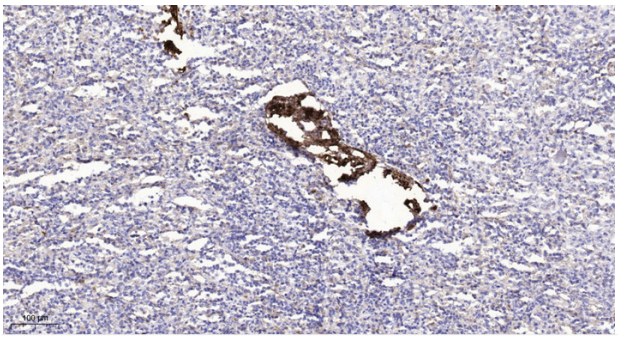
Catalog No	BYab-11540
Isotype	IgG
Reactivity	Human; Mouse
Applications	WB;IHC
Gene Name	PHKA2 PHKLA PYK
Protein Name	KPB2
Immunogen	Synthesized peptide derived from human KPB2 AA range: 911-961
Specificity	This antibody detects endogenous levels of KPB2 at Human/Mouse
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Polyclonal, Rabbit,IgG
Purification	The antibody was affinity-purified from rabbit serum by affinity-chromatography using specific immunogen.
Dilution	WB 1:500-2000;IHC-p 1:50-300
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	
Observed Band	
Cell Pathway	Cell membrane ; Lipid-anchor ; Cytoplasmic side .
Tissue Specificity	Predominantly expressed in liver and other non-muscle tissues.
Function	disease:Defects in PHKA2 are the cause of glycogen storage disease type 9A (GSD9A) [MIM:306000]; also known as X-linked liver glycogenosis (XLG). GSD9A is a metabolic disorder resulting in a mild glycogenosis with clinical symptoms that include hepatomegaly, growth retardation, muscle weakness, elevation of glutamate-pyruvate transaminase and glutamate-oxaloacetate transaminase, hypercholesterolemia, hypertriglyceridemia, and fasting hyperketosis. Two subtypes are known: type 1 or classic type, and type 2 or variant type. The variant type is characterized mainly by enlarged liver and growth retardation; patients do not show in vitro enzymatic deficiency of phosphorylase kinase. Unlike other glycogenosis diseases, GSD9A is generally a benign condition. Patients improve with age and are often asymptomatic as adults. Accurate diagnosis is therefore also of prognostic interest.,enzyme regula

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 hepatic 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 9A, also known as X-linked liver glycogenosis. Alternatively spliced transcript variants have been reported, but the full-length nature of these variants has not been determined.[provided by RefSeq, Feb 2010],
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 lysates from MCF-7 cells, primary antibody was diluted at 1:1000, 4°over night



Immunohistochemical analysis of paraffin-embedded human spleen. 1, Antibody was diluted at 1:200(4° overnight). 2, Tris-EDTA,pH9.0 was used for antigen retrieval. 3,Secondary antibody was diluted at 1:200(room temperature, 45min).