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Rabbit Anti-PKLR antibody

SL12694R

Product Name:	PKLR
Chinese Name:	丙酮酸激酶肝型抗体
Alias:	EC 2.7.1.40; KPYR_HUMAN; L-PK; Pk-1; PK1; PKL; Pklg; Pklr; PKR; PKRL; Pyruvate kinase 1; Pyruvate kinase isozymes R/L; Pyruvate kinase liver and blood cell; Pyruvate kinase liver and RBC; Pyruvate kinase liver and red blood cell; Pyruvate kinase liver type; Pyruvate kinase type L; Pyruvate kinase, red cell type; R type/L type pyruvate kinase; R-PK; R-type/L-type pyruvate kinase; Red cell/liver pyruvate kinase; RPK.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Cow,Horse,Rabbit,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=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:	62kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human PKLR:401-500/571
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:	The protein encoded by this gene is a pyruvate kinase that catalyzes the transphosphorylation of phohsphoenolpyruvate into pyruvate and ATP, which is the

rate-limiting step of glycolysis. Defects in this enzyme, due to gene mutations or genetic variations, are the common cause of chronic hereditary nonspherocytic hemolytic anemia (CNSHA or HNSHA). Multiple transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq, Jul 2008]

Function:
Plays a key role in glycolysis.

DISEASE:
Defects in PKLR are the cause of pyruvate kinase hyperactivity (PKHYP) [MIM:102900]; also known as high red cell ATP syndrome. This autosomal dominant phenotype is characterized by increase of red blood cell ATP.
Defects in PKLR are the cause of pyruvate kinase deficiency of red cells (PKRD) [MIM:266200]. A frequent cause of hereditary non-spherocytic hemolytic anemia. Clinically, pyruvate kinase-deficient patients suffer from a highly variable degree of chronic hemolysis, ranging from severe neonatal jaundice and fatal anemia at birth, severe transfusion-dependent chronic hemolysis, moderate hemolysis with exacerbation during infection, to a fully compensated hemolysis without apparent anemia.

Similarity:
Belongs to the pyruvate kinase family.

SWISS:
P30613

Gene ID:
5313

Database links:

Entrez Gene: 5313 Human

Entrez Gene: 18770 Mouse

Omim: 609712 Human

SwissProt: P30613 Human

SwissProt: P53657 Mouse

Unigene: 95990 Human

Unigene: 383180 Mouse

Important Note:

	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.