

Product name:	PFKM Rabbit Polyclonal Antibody
Cat number:	ABN16019
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human PFK-1. AA range:320-369
Reactivity:	Human,Mouse,Rat
Applications:	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:5000-1:20000
Molecular Weight:	85kDa
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:

Three phosphofructokinase isozymes exist in humans: muscle, liver and platelet. These isozymes function as subunits of the mammalian tetramer phosphofructokinase, which catalyzes the phosphorylation of fructose-6-phosphate to fructose-1,6-bisphosphate. Tetramer composition varies depending on tissue type. This gene encodes the muscle-type isozyme. Mutations in this gene have been associated with glycogen storage disease type VII, also known as Tarui disease. Alternatively spliced transcript variants have been described.[provided by RefSeq, Nov 2009],catalytic activity:ATP + D-fructose 6-phosphate = ADP + D-fructose 1,6-bisphosphate.,cofactor:Magnesium.,disease:Defects in PFKM are the cause of glycogen storage disease type 7 (GSD7) [MIM:232800]; also known as Tarui disease. GSD7 is an autosomal recessive disorder characterized by exercise intolerance with associated nausea and vomiting. Short bursts of intense activity are particularly difficult. Severe muscle cramps and myoglobinuria develop after vigorous exercise. Most patients obtain a "second wind" when the onset of exercise is followed by a brief rest period. In time patients adjust their activity level and are well compensated.,enzyme regulation:Allosteric enzyme activated by ADP, AMP, or fructose bisphosphate and inhibited by ATP or citrate.,miscellaneous:In human PFK exists as a system of 3 types of subunits, PFKM (muscle), PFKL (liver) and PFKP (platelet) isoenzymes.,pathway:Carbohydrate degradation; glycolysis; D-glyceraldehyde 3-phosphate and glyceraldehyde phosphate from D-glucose: step 3/4.,similarity:Belongs to the phosphofructokinase family. Two domains subfamily.,subunit:Tetramer. Muscle is M4, liver is L4, and red cell is M3L, M2L2, or ML3.,