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| <b>Product name:</b>     | PHKA1/2 Rabbit Polyclonal Antibody                                                               |
| <b>Cat number:</b>       | ABN16072                                                                                         |
| <b>Conjugate:</b>        | Unconjugated                                                                                     |
| <b>Size:</b>             | 100µL                                                                                            |
| <b>Clone:</b>            | Polyclonal                                                                                       |
| <b>Concentration:</b>    | 1mg/ml                                                                                           |
| <b>Host:</b>             | Rabbit                                                                                           |
| <b>Isotype:</b>          | IgG                                                                                              |
| <b>Immunogen:</b>        | The antiserum was produced against synthesized peptide derived from human KPB1/2. AA range:31-80 |
| <b>Reactivity:</b>       | Human,Mouse,Rat                                                                                  |
| <b>Applications:</b>     | WB 1:500-1:2000,IHC 1:50-1:300,ELISA 1:2000-1:20000                                              |
| <b>Molecular Weight:</b> | 137kDa                                                                                           |
| <b>Purification:</b>     | Affinity purification                                                                            |
| <b>Form:</b>             | Liquid                                                                                           |
| <b>Buffer:</b>           | Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.               |
| <b>Storage:</b>          | Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.     |

**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 variant 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 delta is calmodulin. 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.