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DMPK Rabbit pAb

Catalog # AP97847

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q09013
Other Accession	Q09013
Reactivity	Mouse
Predicted	Rat, Pig, Human, Dog, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	69385
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human DMPK
Epitope Specificity	51-120/629
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the protein kinase superfamily. AGC Ser/Thr protein kinase family. DMPK subfamily. Contains 1 AGC-kinase C-terminal domain. Contains 1 protein kinase domain.
SUBUNIT	Homodimer; homodimerization stimulates the kinaseactivity. Interacts with HSPB2; may enhance DMPK kinase activity.Interacts with PLN; phosphorylates PLN. May interact with RAC1; mayregulate DMPK kinase activity. Interacts with LMNA; may regulatenuclear envelope stability.
Post-translational modifications	Phosphorylated. Autophosphorylates. Phosphorylation by RAF1may result in activation of DMPK.Proteolytic processing of the C-terminus may remove thetransmembrane domain and release the kinase from membranesstimulating its activity.
DISEASE	Defects in DMPK are the cause of dystrophia myotonica type 1 (DM1) [MIM:160900]; also known as Steinert disease. A muscular disorder characterized by myotonia, muscle wasting in the distal extremities, cataract, hypogonadism, defective endocrine functions, male baldness and cardiac arrhythmias. Note=The causative mutation is a CTG expansion in the 3'-UTR of the DMPK gene. A length exceeding 50 CTG repeats is pathogenic, while normal individuals have 5 to 37 repeats. Intermediate alleles with 35-49 triplets are not disease-causing but show instability in intergenerational transmissions. Disease severity varies with the number of repeats: mildly affected persons have 50 to 150 repeats, patients with classic DM have 100 to 1,000 repeats, and those with congenital onset can have more than 2,000 repeats.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Myotonic dystrophy protein kinase is a multi-domain protein kinase found in muscle that is activated in response to G protein second messengers and proteolysis (1). DMPK is implicated in myotonic muscular dystrophy (DM), an autosomal dominant-inherited disorder that predominately affects skeletal and cardiac muscle and causes defects in cardiac conduction (2,3). DM arises through expansion of CTG repeats in the 3’-UTR of the DMPK gene (4). Mutant

DMPK transcripts with an extended region of CUG repeats are retained in the nucleus (5). These transcripts also influence the expression of the DM locus-associated homeodomain protein (DMAHP)/SIX5, to mediate in part the DM phenotype (6). Other substrates for DMPK include myogenin, L-type calcium channels, and Phospholemman (PLM) (1).

Additional Information

Gene ID	1760
Other Names	DM; DM1; DM1PK; DMK; MDPK; MT-PK; Dm15; DMPK_HUMAN; DMPK; DM-kinase (DMK); DM1 protein kinase; Myotonic dystrophy protein kinase; 2.7.11.1; DMPK_MOUSE; Myotonic dystrophy protein kinase (MDPK); dystrophia myotonica 1 (includes dystrophia myotonia protein kinase); dystrophia myotonica protein kinase; dystrophia myotonica 1; DM protein kinase; myotonin protein kinase A; myotonic dystrophy associated protein kinase; thymopoietin homolog; Myotonin-protein kinase
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	DMPK
Synonyms	DM1PK, MDPK
Function	Non-receptor serine/threonine protein kinase which is necessary for the maintenance of skeletal muscle structure and function. May play a role in myocyte differentiation and survival by regulating the integrity of the nuclear envelope and the expression of muscle-specific genes. May also phosphorylate PPP1R12A and inhibit the myosin phosphatase activity to regulate myosin phosphorylation. Also critical to the modulation of cardiac contractility and to the maintenance of proper cardiac conduction activity probably through the regulation of cellular calcium homeostasis. Phosphorylates PLN, a regulator of calcium pumps and may regulate sarcoplasmic reticulum calcium uptake in myocytes. May also phosphorylate FXYD1/PLM which is able to induce chloride currents. May also play a role in synaptic plasticity.
Cellular Location	Endoplasmic reticulum membrane; Single-pass type IV membrane protein; Cytoplasmic side. Nucleus outer membrane; Single-pass type IV membrane protein; Cytoplasmic side Mitochondrion outer membrane; Single-pass type IV membrane protein. Sarcoplasmic reticulum membrane. Cell membrane. Cytoplasm, cytosol. Note=Localizes to sarcoplasmic reticulum membranes of cardiomyocytes. [Isoform 3]: Mitochondrion membrane.
Tissue Location	Most isoforms are expressed in many tissues including heart, skeletal muscle, liver and brain, except for isoform 2 which is only found in the heart and skeletal muscle, and isoform 14 which is only found in the brain, with high levels in the striatum, cerebellar cortex and pons.

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