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KCNQ1 Rabbit pAb

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Catalog # AP52223

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P51787
Predicted	Rat, Dog, Human, Mouse, Chicken
Host	Rabbit
Clonality	Polyclonal
Calculated MW	74699
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human KCNQ-1
Epitope Specificity	501-600/676
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Cell membrane; Multi-pass membrane protein. Cytoplasmic vesicle membrane; Multi-pass membrane protein.
DISEASE	Defects in KCNQ1 are the cause of long QT syndrome type 1 (LQT1) ; also known as Romano-Ward syndrome (RWS). Long QT syndromes are heart disorders characterized by a prolonged QT interval on the ECG and polymorphic ventricular arrhythmias. They cause syncope and sudden death in response to exercise or emotional stress. LQT1 inheritance is an autosomal dominant. Defects in KCNQ1 are the cause of Jervell and Lange-Nielsen syndrome type 1 (JLNS1) . JLNS1 is an autosomal recessive disorder characterized by congenital deafness, prolongation of the QT interval, syncopal attacks due to ventricular arrhythmias, and a high risk of sudden death. Defects in KCNQ1 are the cause of atrial fibrillation familial type 3 (ATFB3) . Atrial fibrillation is a common disorder of cardiac rhythm that is hereditary in a small subgroup of patients. It is characterized by disorganized atrial electrical activity and ineffective atrial contraction promoting blood stasis in the atria and reduces ventricular filling. It can result in palpitations, syncope, thromboembolic stroke, and congestive heart failure.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Probably important in cardiac repolarization. Associates with KCNE1 (MinK) to form the I(Ks) cardiac potassium current. Elicits a rapidly activating, potassium-selective outward current. Muscarinic agonist oxotremorine-M strongly suppresses KCNQ1/KCNE1 current in CHO cells in which cloned KCNQ1/KCNE1 channels were coexpressed with M1 muscarinic receptors. May associate also with KCNE3 (MiRP2) to form the potassium channel that is important for cyclic AMP-stimulated intestinal secretion of chloride ions, which is reduced in cystic fibrosis and pathologically stimulated in cholera and other forms of secretory diarrhea. Involvement in disease:

Additional Information

Gene ID	3784
Other Names	Potassium voltage-gated channel subfamily KQT member 1, IKs producing slow voltage-gated potassium channel subunit alpha KvLQT1, KQT-like 1, Voltage-gated potassium channel subunit Kv7.1, KCNQ1 (HGNC:6294)
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,ELISA=1:5000-10000
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	KCNQ1 (HGNC:6294)
Function	Pore-forming subunit of the voltage-gated potassium (Kv) channel involved in the regulation of cardiomyocyte excitability and important in normal development and functions of myocardium, inner ear, stomach and colon (PubMed:10646604, PubMed:25441029). Associates with KCNE beta subunits that modulates current kinetics (PubMed:10646604, PubMed:11101505, PubMed:19687231, PubMed:8900283, PubMed:9108097, PubMed:9312006). Induces a voltage-dependent current by rapidly activating and slowly deactivating potassium-selective outward current (PubMed:10646604, PubMed:11101505, PubMed:25441029, PubMed:8900283, PubMed:9108097, PubMed:9312006). Also promotes a delayed voltage activated potassium current showing outward rectification characteristic (By similarity). During beta-adrenergic receptor stimulation, participates in cardiac repolarization by associating with KCNE1 to form the I(Ks) cardiac potassium current that increases the amplitude and slows down the activation kinetics of outward potassium current I(Ks) (By similarity) (PubMed:10646604, PubMed:11101505, PubMed:8900283, PubMed:9108097, PubMed:9312006). Muscarinic agonist oxotremorine-M strongly suppresses KCNQ1/KCNE1 current (PubMed:10713961). When associated with KCNE3, forms the potassium channel that is important for cyclic AMP-stimulated intestinal secretion of chloride ions (PubMed:10646604). This interaction with KCNE3 is reduced by 17beta-estradiol, resulting in the reduction of currents (By similarity). During conditions of increased substrate load, maintains the driving force for proximal tubular and intestinal sodium ions absorption, gastric acid secretion, and cAMP-induced jejunal chloride ions secretion (By similarity). Allows the provision of potassium ions to the luminal membrane of the secretory canaliculus in the resting state as well as during stimulated acid secretion (By similarity). When associated with KCNE2, forms a heterooligomer complex leading to currents with an apparently instantaneous activation, a rapid deactivation process and a linear current-voltage relationship and decreases the amplitude of the outward current (PubMed:11101505). When associated with KCNE4, inhibits voltage-gated potassium channel activity (PubMed:19687231). When associated with KCNE5, this complex only conducts current upon strong and continued depolarization (PubMed:12324418). Also forms a heterotetramer with KCNQ5; has a voltage-gated potassium channel activity (PubMed:24855057). Binds with phosphatidylinositol 4,5-bisphosphate (PubMed:25037568). KCNQ1-KCNE2 channel associates with Na(+)-coupled myo-inositol symporter in the apical membrane of choroid plexus epithelium and regulates the myo- inositol gradient between blood and cerebrospinal fluid with an impact on neuron

	excitability (By similarity).
Cellular Location	Cell membrane; Multi-pass membrane protein. Cytoplasmic vesicle membrane Early endosome. Membrane raft. Endoplasmic reticulum Basolateral cell membrane. Apical cell membrane {ECO:0000250|UniProtKB:P97414}; Multi-pass membrane protein. Note=Colocalized with KCNE3 at the plasma membrane (PubMed:10646604). Upon 17beta-oestradiol treatment, colocalizes with RAB5A at early endosome (PubMed:23529131). Heterotetramer with KCNQ5 is highly retained at the endoplasmic reticulum and is localized outside of lipid raft microdomains (PubMed:24855057). During the early stages of epithelial cell polarization induced by the calcium switch, it is removed from the plasma membrane to the endoplasmic reticulum, where it is retained, and redistributed to the basolateral cell surface in a PI3K-dependent manner at a later stage (PubMed:21228319). Colocalizes with SLC5A3 at the apical membrane of choroid plexus epithelium {ECO:0000250|UniProtKB:P97414, ECO:0000269|PubMed:10646604, ECO:0000269|PubMed:21228319, ECO:0000269|PubMed:23529131, ECO:0000269|PubMed:24855057}
Tissue Location	Abundantly expressed in heart, pancreas, prostate, kidney, small intestine and peripheral blood leukocytes. Less abundant in placenta, lung, spleen, colon, thymus, testis and ovaries

Background

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

References

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Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.