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ERG/KCNH2 Rabbit pAb

Catalog # AP98068

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q12809
Other Accession	Q12809
Reactivity	Mouse
Predicted	Rat, Human, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	126655
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human ERG/KCNH2
Epitope Specificity	541-640/1159
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 potassium channel family. H (Eag) (TC1.A.1.20) subfamily. Kv11.1/KCNH2 sub-subfamily. Contains 1 cyclic nucleotide-binding domain. Contains 1 PAC (PAS-associated C-terminal) domain. Contains 1 PAS (PER-ARNT-SIM) domain.
SUBUNIT	The potassium channel is probably composed of a homo- or heterotetrameric complex of pore-forming alpha subunits that can associate with modulating beta subunits. Heteromultimer with KCNH6/ERG2 and KCNH7/ERG3. Interacts with ALG10B (By similarity). Heteromultimer with KCNE1 and KCNE2.
Post-translational modifications	Phosphorylated on serine and threonine residues. Phosphorylation by PKA inhibits ion conduction.
DISEASE	Defects in KCNH2 are the cause of long QT syndrome type 2 (LQT2) [MIM:613688]. 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. Deafness is often associated with LQT2. Defects in KCNH2 are the cause of short QT syndrome type 1 (SQT1) [MIM:609620]. Short QT syndromes are heart disorders characterized by idiopathic persistently and uniformly short QT interval on ECG in the absence of structural heart disease in affected individuals. They cause syncope and sudden death.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The potassium voltage gated channel, subfamily H (eag related), member 2 (KCNH2) gene encodes a voltage-gated potassium channel which has an important role in cardiac action potential repolarization in the mammalian heart. Mutations in KCNH2 have been shown to cause chromosome 7-linked congenital long QT syndrome, a disorder associated with delayed cardiac repolarization, prolonged electrocardiographic QT intervals, and the development of ventricular arrhythmias. KCNH2 channels are an important target for many drugs, and have emerged as a significant type of cardiac ion channel.Highly expressed in heart and brain.

Additional Information

Gene ID	3757
Other Names	ERG-1; ERG1; H-ERG; HERG; HERG1; Kv11.1; LQT2; SQT1; LQT; M-erg; Merg1; merg1a; merg1b; KCNH2_HUMAN; KCNH2; Eag homolog; Ether-a-go-go-related gene potassium channel 1 (ERG-1 | Eag-related protein 1 | Ether-a-go-go-related protein 1 | H-ERG | hERG-1 | hERG1); Potassium voltage-gated channel subfamily H member 2; Voltage-gated potassium channel subunit Kv11.1; ERG; KCNH2_MOUSE; Ether-a-go-go-related gene potassium channel 1 (ERG-1 | Eag-related protein 1 | Ether-a-go-go-related protein 1 | MERG); KCNH2_RAT; Ether-a-go-go-related gene potassium channel 1 (ERG-1 | Eag-related protein 1 | Ether-a-go-go-related protein 1 | RERG | r-ERG); potassium voltage-gated channel, subfamily H (eag-related), member 2; human ether-a-go-go-related gene; long QT syndrome type 2
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	KCNH2 (HGNC:6251)
Function	Pore-forming (alpha) subunit of voltage-gated inwardly rectifying potassium channel (PubMed:10219239, PubMed:10753933, PubMed:10790218, PubMed:10837251, PubMed:11997281, PubMed:12063277, PubMed:18559421, PubMed:22314138, PubMed:22359612, PubMed:26363003, PubMed:27916661, PubMed:9230439, PubMed:9351446, PubMed:9765245). Channel properties are modulated by cAMP and subunit assembly (PubMed:10837251). Characterized by unusual gating kinetics by producing relatively small outward currents during membrane depolarization and large inward currents during subsequent repolarization which reflect a rapid inactivation during depolarization and quick recovery from inactivation but slow deactivation (closing) during repolarization (PubMed:10219239, PubMed:10753933, PubMed:10790218, PubMed:10837251, PubMed:11997281, PubMed:12063277, PubMed:18559421, PubMed:22314138, PubMed:22359612, PubMed:26363003, PubMed:27916661, PubMed:9230439, PubMed:9351446, PubMed:9765245). Forms a stable complex with KCNE1 or KCNE2, and that this heteromultimerization regulates inward rectifier potassium channel activity (PubMed:10219239, PubMed:9230439).
Cellular Location	Cell membrane; Multi-pass membrane protein
Tissue Location	Highly expressed in heart and brain. Isoforms USO are frequently overexpressed in cancer cells

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