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Rabbit Anti-ERG/KCNH2/AF647 Conjugated antibody

SL23704R-AF647

Product Name	Anti-ERG/KCNH2/AF647
Chinese Name	AF647 标记的特异性钾离子 Channel protein 抗体
Alias	ERG; ERG1; H ERG; HERG 1; HERG; HERG1; LQT 2; LQT2; Potassium channel HERG; SQT1; Voltagegated potassium channel, subfamily H, member 2; KCNH2_HUMAN.
Research Area	Tumour immunology Cyclin Channel protein
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	Mouse(predicted:Human,Rat,Dog,Cow,Horse,Sheep)
Applications	IF=1:100-500 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight	127kDa
Form	Lyophilized or Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human ERG/KCNH2
Lsotype	IgG
Purification	affinity purified by Protein A
Storage Buffer	Preservative: 15mM Sodium Azide, Constituents: 1% BSA, 1M PBS, pH 7.4.
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
Product Detail	**background:** 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.

Function:
Pore-forming (alpha) subunit of voltage-gated inwardly rectifying potassium channel. Channel properties are modulated by cAMP and subunit assembly. Mediates the rapidly activating component of the delayed rectifying potassium current in heart (IKr). Isoform 3 has no channel activity by itself, but modulates channel characteristics when associated with isoform 1.

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.

Subcellular Location:
Membrane; Multi-pass membrane protein.

Tissue Specificity:
Highly expressed in heart and brain.

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.

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.

Database links:

Entrez Gene: 3757 Human

Entrez Gene: 16511 Mouse

Entrez Gene: 117018 Rat

Omim: 152427 Human

SwissProt: Q12809 Human

SwissProt: O35219 Mouse

SwissProt: O08962 Rat

Unigene: 647099 Human

Unigene: 6539 Mouse

Unigene: 10970 Rat

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