钾离子通道蛋白家族 KCNQ2 抗体

产品货号： mlR11728

英文名称： KCNQ2

中文名称： 钾离子通道蛋白家族 KCNQ2 抗体

别　　名： BFNC; BFNS1; EBN 1; EBN; EBN1; EIEE7; ENB 1; ENB1; HNSPC; KCNA 11; KCNA11; KCNQ 2; Kcnq2; KCNQ2_HUMAN; KQT like 2; KQT-like 2; KV7.2; KVEBN 1; KVEBN1; KvLQT 2; KvLQT2; Neuroblastoma specific potassium channel alpha subunit KvLQT2; Neuroblastoma specific potassium channel protein; Neuroblastoma specific potassium channel subunit alpha; Neuroblastoma specific potassium channel subunit alpha KvLQT2; Neuroblastoma-specific potassium channel subunit alpha KvLQT2; Potassium voltage gated channel KQT like protein 2; Potassium voltage gated channel KQT like subfamily member 2; Potassium voltage gated channel subfamily KQT member 2; Potassium voltage-gated channel subfamily KQT member 2; Voltage gated potassium channel subunit Kv7.2; Voltage-gated potassium channel subunit Kv7.2.

研究领域： 神经生物学　通道蛋白　细胞膜受体

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应：　Human, Mouse, Rat, Dog, Cow, Horse, Sheep,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 Flow-Cyt=3 μg/Test ICC=1:100-500 IF=1:100-500 （石蜡切片需做抗原修复）
not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.

分 子 量： 96kDa

细胞定位： 细胞膜

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human KCNQ2:91-150/872 <Extracellular>

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： 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 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ° C.

PubMed： PubMed

产品介绍： Epilepsy affects about 0.5% of the world’s population and has a large genetic component. Epilepsy results from an electrical hyperexcitability in the central nervous system. Potassium channels are important regulators of electrical signaling, determining the firing properties and responsiveness of a variety of neurons. Benign familial neonatal convulsions (BFNC), an autosomal dominant epilepsy of infancy, has been shown to be caused by mutations in the KCNQ2 or the KCNQ3 potassium channel genes. KCNQ2 and KCNQ3 are voltage-gated potassium channel proteins with six putative transmembrane domains. Both proteins display a broad distribution within the brain, with expression patterns that largely overlap.

Function:

Probably important in the regulation of neuronal excitability. Associates with KCNQ3 to form a potassium channel with essentially identical properties to the channel underlying the native M-current, a slowly activating and deactivating potassium conductance which plays a critical role in determining the subthreshold electrical excitability of neurons as well as the responsiveness to synaptic inputs. KCNQ2/KCNQ3 current is blocked by linopirdine and XE991, and activated by the anticonvulsant retigabine. Muscarinic agonist oxotremorine-M strongly suppress KCNQ2/KCNQ3 current in cells in which cloned KCNQ2/KCNQ3 channels were coexpressed with M1 muscarinic receptors.

Subunit:

Heteromultimer with KCNQ3. May associate with KCNE2.

Subcellular Location:

Membrane; Multi-pass membrane protein.

Tissue Specificity:

In adult and fetal brain. Highly expressed in areas containing neuronal cell bodies, low in spinal chord and corpus callosum. Isoform 2 is preferentially expressed in differentiated neurons. Isoform 6 is prominent in fetal brain, undifferentiated neuroblastoma cells and brain tumors.

Post-translational modifications:

In Xenopus oocytes KCNQ2/KCNQ3 heteromeric current can be increased by intracellular cyclic AMP, an effect that depends on phosphorylation of Ser-52 in the N-terminus region.

DISEASE:

Defects in KCNQ2 are the cause of benign familial neonatal seizures type 1 (BFNS1) [MIM:121200]. A disorder characterized by clusters of seizures occurring in the first days of life. Most patients have spontaneous remission by 12 months of age and show normal psychomotor development. Some rare cases manifest an atypical severe phenotype associated with epileptic encephalopathy and psychomotor retardation. The disorder is distinguished from benign familial infantile seizures by an earlier age at onset. In some patients, neonatal convulsions are followed later in life by myokymia, a benign condition characterized by spontaneous involuntary contractions of skeletal muscles fiber groups that can be observed as vermiform movement of the overlying skin. Electromyography typically shows continuous motor unit activity with spontaneous oligo- and multiplet-discharges of high intraburst frequency (myokymic discharges). Some patients may have isolated myokymia. Defects in KCNQ2 are the cause of epileptic encephalopathy early infantile type 7 (EIEE7) [MIM:613720]. EIEE7 is an autosomal dominant seizure disorder characterized by infantile onset of refractory seizures with resultant delayed neurologic development and persistent neurologic abnormalities.

Similarity:

Belongs to the potassium channel family. KQT (TC 1.A.1.15) subfamily. Kv7.2/KCNQ2 sub-subfamily.

SWISS:

O43526

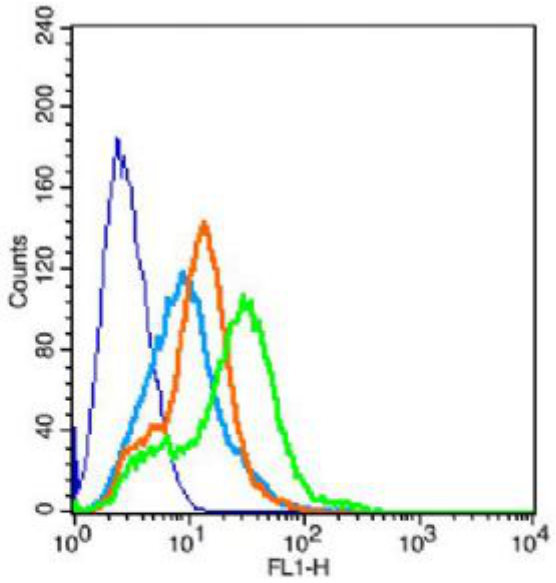
Gene ID:

3785

Important Note:

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

产品图片



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	bs-11728R-(FITC)(B)-#1FCC18.052	FL1-H	G1