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KCNQ2 Rabbit pAb

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Catalog # AP54611

Product Information

Primary Accession	O43526
Reactivity	Rat
Predicted	Dog, Human, Mouse, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	95848
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human KCNQ2
Epitope Specificity	91-150/872
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	Membrane; Multi-pass membrane protein.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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.

Additional Information

Gene ID	3785
Other Names	Potassium voltage-gated channel subfamily KQT member 2, KQT-like 2, Neuroblastoma-specific potassium channel subunit alpha KvLQT2, Voltage-gated potassium channel subunit Kv7.2, KCNQ2 (HGNC:6296)
Dilution	Flow-Cyt=3 μg/Test
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	KCNQ2 (HGNC:6296)
Function	Pore-forming subunit of the voltage-gated potassium (Kv) M- channel which is responsible for the M-current, a key controller of neuronal excitability (PubMed:24277843, PubMed:28793216, PubMed:9836639). M-channel is composed of pore-forming subunits KCNQ2 and KCNQ3 assembled as heterotetramers (PubMed:10781098, PubMed:14534157, PubMed:32884139, PubMed:37857637, PubMed:9836639). The native M-current has 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 (PubMed:14534157, PubMed:28793216, PubMed:9836639). KCNQ2-KCNQ3 M-channel is selectively permeable in vitro to other cations besides potassium, in decreasing order of affinity K(+) > Rb(+) > Cs(+) > Na(+) (PubMed:28793216). M-channel association with SLC5A3/SMIT1 alters channel ion selectivity, increasing Na(+) and Cs(+) permeation relative to K(+) (PubMed:28793216). Suppressed by activation of the muscarinic acetylcholine receptor CHRM1 (PubMed:10684873, PubMed:10713961).
Cellular Location	Cell membrane; Multi-pass membrane protein
Tissue Location	In adult and fetal brain. Highly expressed in areas containing neuronal cell bodies, low in spinal cord and corpus callosum. Isoform 2 is preferentially expressed in differentiated neurons. Isoform 6 is prominent in fetal brain, undifferentiated neuroblastoma cells and brain tumors.

Background

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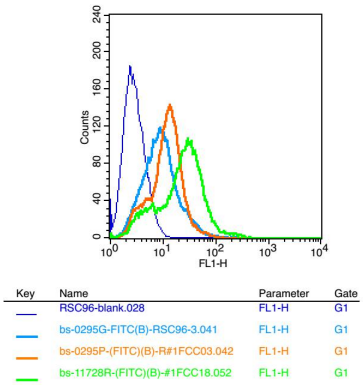
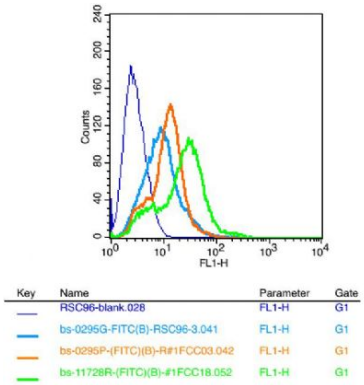
Images

Positive control: RSC96
Isotype Control Antibody: Rabbit IgG ;
Secondary Antibody: Goat anti-rabbit IgG-FITC,
Dilution: 1:100 in 1 X PBS containing 0.5% BSA ;



Primary Antibody Dilution: 3 µg in 100 µL 1X PBS containing 0.5% BSA.



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