10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

SCN1B Rabbit pAb

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Catalog # AP58489

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q07699
Predicted	Rat, Human, Mouse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	24707
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human SCN1B
Epitope Specificity	21-120/268
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; Single-pass type I membrane protein.
DISEASE	Defects in SCN1B are the cause of generalized epilepsy with febrile seizures plus type 1 (GEFS+1) [MIM:604233]. Generalized epilepsy with febrile seizures-plus refers to a rare autosomal dominant, familial condition with incomplete penetrance and large intrafamilial variability. Patients display febrile seizures persisting sometimes beyond the age of 6 years and/or a variety of afebrile seizure types. GEFS+ is a disease combining febrile seizures, generalized seizures often precipitated by fever at age 6 years or more, and partial seizures, with a variable degree of severity.Defects in SCN1B are the cause of Brugada syndrome type 5 (BRGDA5) [MIM:612838]. A tachyarrhythmia characterized by right bundle branch block and ST segment elevation on an electrocardiogram (ECG). It can cause the ventricles to beat so fast that the blood is prevented from circulating efficiently in the body. When this situation occurs (called ventricular fibrillation), the individual will faint and may die in a few minutes if the heart is not reset.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Crucial in the assembly, expression, and functional modulation of the heterotrimeric complex of the sodium channel. The subunit beta-1 can modulate multiple alpha subunit isoforms from brain, skeletal muscle, and heart. Its association with neurofascin may target the sodium channels to the nodes of Ranvier of developing axons and retain these channels at the nodes in mature myelinated axons. Tissue specificity; Abundantly expressed in skeletal muscle, heart and brain.

Additional Information

Gene ID	6324

Other Names	Sodium channel regulatory subunit beta-1, SCN1B (HGNC:10586)
Dilution	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	SCN1B (HGNC:10586)
Function	Regulatory subunit of multiple voltage-gated sodium (Nav) channels directly mediating the depolarization of excitable membranes (PubMed:40768348). Navs, also called VGSCs (voltage-gated sodium channels) or VDSCs (voltage-dependent sodium channels), operate by switching between closed and open conformations depending on the voltage difference across the membrane. In the open conformation they allow Na(+) ions to selectively pass through the pore, along their electrochemical gradient (PubMed:40768348). The influx of Na+ ions provokes membrane depolarization, initiating the propagation of electrical signals throughout cells and tissues (PubMed:14622265, PubMed:15525788, PubMed:18464934, PubMed:19710327, PubMed:29992740, PubMed:36696443, PubMed:8125980, PubMed:8394762). The accessory beta subunits participate in localization and functional modulation of the Nav channels (PubMed:15525788, PubMed:19710327, PubMed:29992740). Modulates the activity of SCN1A/Nav1.1, SCN2A/Nav1.2, SCN3A/Nav1.3, SCN4A/Nav1.4, SCN5A/Nav1.5, SCN8A/Nav1.6, SCN9A/Nav1.7 and SCN10A/Nav1.8 (PubMed:14622265, PubMed:15525788, PubMed:18464934, PubMed:30765606, PubMed:36696443, PubMed:8125980, PubMed:8394762).
Cellular Location	[Isoform 1]: Cell membrane; Single-pass type I membrane protein. Perikaryon {ECO:0000250|UniProtKB:P97952} Cell projection {ECO:0000250|UniProtKB:P97952}. Cell projection, axon {ECO:0000250|UniProtKB:Q00954}. Note=Detected at nodes of Ranvier on the sciatic nerve. {ECO:0000250|UniProtKB:Q00954}
Tissue Location	The overall expression of isoform 1 and isoform 2 is very similar. Isoform 1 is abundantly expressed in skeletal muscle, heart and brain. Isoform 2 is highly expressed in brain and skeletal muscle and present at a very low level in heart, placenta, lung, liver, kidney and pancreas. In brain, isoform 2 is most abundant in the cerebellum, followed by the cerebral cortex and occipital lobe, while isoform 1 levels are higher in the cortex compared to the cerebellum Isoform 2 is expressed in many regions of the brain, including cerebellar Purkinje cells, cortex pyramidal neurons and many of the neuronal fibers throughout the brain (at protein level). Also detected in dorsal root ganglion, in fibers of the spinal nerve and in cortical neurons and their processes (at protein level)

Background

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