

SCN5A/Nav1.5 Polyclonal Antibody

Purified Rabbit Polyclonal Antibody (Pab) Catalog # AP59258

Product Information

Application Primary Accession Reactivity Host Clonality Calculated MW Physical State Immunogen Epitope Specificity Isotype Purity	IHC-P, IHC-F, IF, E Q14524 Rat, Pig, Dog, Chimpanzee Rabbit Polyclonal 226940 Liquid KLH conjugated synthetic peptide derived from human Nav1.5/SCN5A 51-150/2016 IgG affinity purified by Protein A
Buffer SUBCELLULAR LOCATION SIMILARITY	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Membrane. Belongs to the sodium channel (TC 1.A.1.10) family, Nav1.5/SCN5A subfamily.
SUBUNIT	Contains 1 IQ domain. Interacts with the PDZ domain of the syntrophin SNTA1, SNTB1 and SNTB2 (By similarity). Interacts with NEDD4, NEDD4L, WWP2 and GPD1L. Interacts with CALM. Interacts with FGF13; the interaction is direct and may regulate SNC5A density at membranes and function.
Post-translational modifications	Regulated through phosphorylation by CaMK2D (By similarity). Ubiquitinated by NEDD4L; which promotes its endocytosis. Does not seem to be
DISEASE	Defects in SCN5A are a cause of progressive familial heart block type 1A (PFHB1A) [MIM:113900]; also known as Lenegre-Lev disease or progressive cardiac conduction defect (PCCD). PFHB1A is an autosomal dominant cardiac bundle branch disorder that may progress to complete heart block. PFHB1A is characterized by progressive alteration of cardiac conduction through the His-Purkinje system with right or left bundle branch block and widening of QRS complexes, leading to complete atrioventricular block and causing syncope and sudden death. [DISEASE] Defects in SCN5A are the cause of long QT syndrome type 3 (LQT3) [MIM:603830]. 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. LQT3 inheritance is an autosomal dominant. Defects in SCN5A are the cause of Brugada syndrome type 1 (BRGDA1) [MIM:601144]. An autosomal dominant 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. Defects in SCN5A are the cause of sick sinus syndrome type 1 (SSS1) [MIM:608567]. The term 'sick sinus syndrome' encompasses a variety of conditions caused by sinus node dysfunction. The

	most common clinical manifestations are syncope, presyncope, dizziness, and fatigue. Electrocardiogram typically shows sinus bradycardia, sinus arrest, and/or sinoatrial block. Episodes of atrial tachycardias coexisting with sinus bradycardia ('tachycardia-bradycardia syndrome') are also common in this disorder. SSS occurs most often in the elderly associated with underlying heart disease or previous cardiac surgery, but can also occur in the fetus, infant, or child without heart disease or other contributing factors, in which case it is considered to be a congenital disorder. Defects in SCN5A are the cause of familial paroxysmal ventricular fibrillation type 1 (VF1) [MIM:603829]. A cardiac arrhythmia marked by fibrillary contractions of the ventricular muscle due to rapid repetitive excitation of myocardial fibers without coordinated contraction of the ventricle and by absence of atrial activity. Defects in SCN5A may be a cause of sudden infant death syndrome (SIDS) [MIM:272120]. SIDS is the sudden death of an infant younger than 1 year that remains unexplained after a thorough case investigation, including performance of a complete autopsy, examination of the death scene, and review of clinical history. Pathophysiologic mechanisms for SIDS may include respiratory dysfunction, cardiac dysrhythmias, cardiorespiratory instability, and inborn errors of metabolism, but definitive pathogenic mechanisms precipitating an infant sudden death remain elusive. Long QT syndromes-associated mutations can be responsible for some of SIDS cases. Defects in SCN5A may be a cause of familial atrial standstill (FAS) [MIM:108770]. Atrial standstill is an extremely rare arrhythmia, characterized by the absence of P waves, and a junctional narrow complex escape rhythm. Defects in SCN5A are the cause of cardiomyopathy with conduction disorder and arrhythmia or dilated cardiomyopathy with conduction defect 2. Dilated cardiomyopathy is a disorder characterized by ventricular dilatrial fibrillation is characterized by ventricular dilute
Important Note	It can result in palpitations, syncope, thromboembolic stroke, and congestive heart failure. This product as supplied is intended for research use only, not for use in
	human, therapeutic or diagnostic applications.
Background Descriptions	excitable membranes. Assuming opened or closed conformations in response to the voltage difference across the membrane, the protein forms a sodium-selective channel through which Na(+) ions may pass in accordance with their electrochemical gradient. It is a tetrodotoxin-resistant Na(+) channel isoform. This channel is responsible for the initial upstroke of the action potential.

Additional Information

Gene ID	6331
Other Names	Sodium channel protein type 5 subunit alpha, Sodium channel protein cardiac muscle subunit alpha, Sodium channel protein type V subunit alpha, Voltage-gated sodium channel subunit alpha Nav1.5, hH1, SCN5A
Target/Specificity	Found in jejunal circular smooth muscle cells (at protein level). Expressed in human atrial and ventricular cardiac muscle but not in adult skeletal muscle,

	brain, myometrium, liver, or spleen. Isoform 4 is expressed in brain.
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:50-200,ELISA=1:5000-10000
Format	0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glyce
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	SCN5A (<u>HGNC:10593</u>)
Function	Pore-forming subunit of Nav1.5, a voltage-gated sodium (Nav) channel that directly mediates the depolarizing phase of action potentials in excitable membranes. 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. The influx of Na(+) ions provokes membrane depolarization, initiating the propagation of electrical signals throughout cells and tissues (PubMed:1309946, PubMed:21447824, PubMed:23085483, PubMed:23420830, PubMed:25370050, PubMed:26279430, PubMed:26392562, PubMed:26776555). Nav1.5 is the predominant sodium channel expressed in myocardial cells and it is responsible for the initial upstroke of the action potential in cardiac myocytes, thereby initiating the heartbeat (PubMed:11234013, PubMed:11804990, PubMed:12569159, PubMed:1309946). Required for normal electrical conduction including formation of the infranodal ventricular conduction system and normal action potential configuration, as a result of its interaction with XIRP2 (By similarity).
Cellular Location	Cell membrane; Multi-pass membrane protein {ECO:0000250 UniProtKB:P15389}. Cytoplasm, perinuclear region. Cell membrane, sarcolemma, T- tubule {ECO:0000250 UniProtKB:P15389}. Cell junction {ECO:0000250 UniProtKB:P15389}. Note=RANGRF promotes trafficking to the cell membrane. Colocalizes with PKP2 at intercalated disks in the heart (By similarity). {ECO:0000250 UniProtKB:P15389, ECO:0000269 PubMed:21447824, ECO:0000269 PubMed:23420830}
Tissue Location	Found in jejunal circular smooth muscle cells (at protein level). Expressed in human atrial and ventricular cardiac muscle but not in adult skeletal muscle, brain, myometrium, liver, or spleen. Isoform 4 is expressed in brain.

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