

产品名称: Na⁺ CP type IV α Rabbit Polyclonal Antibody

产品货号: APRab14374

产品概述 (Summary)

描述(Description)	Rabbit polyclonal Antibody
宿主(Host)	Rabbit
应用(Application)	WB,IHC
种属反应(Reactivity)	Human,Mouse,Rat
偶联物(Conjugation)	Unconjugated
修饰(Modification)	Unmodified
同种型(Isotype)	IgG
克隆(Clonality)	Polyclonal
剂型(Form)	Liquid
保存条件(Storage)	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
储存溶液(Buffer)	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
纯化 (Purification)	Affinity purification

产品应用(Application)

稀释比(Dilution Ratio)	WB 1:500-1:2000,IHC 1:50-1:300
分子量(Molecular Weight)	200kDa

抗原信息(Antigen Information)

基因名(Gene Name)	SCN4A
别名(Alternative Names)	SCN4A; Sodium channel protein type 4 subunit alpha; SkM1; Sodium channel protein skeletal muscle subunit alpha; Sodium channel protein type IV subunit alpha; Voltage-gated sodium channel subunit alpha Nav1.4
基因 ID(Gene ID)	6329.0
SwissProt ID	P35499
免疫原(Immunogen)	The antiserum was produced against synthesized peptide derived from human SCN4A. AA range:431-480

研究背景 (Background)

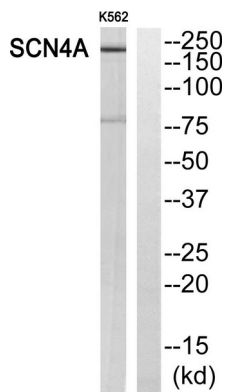
Voltage-gated sodium channels are transmembrane glycoprotein complexes composed of a large alpha subunit with 24 transmembrane domains and one or more regulatory beta subunits. They are responsible for the generation and propagation of action potentials in neurons and muscle. This gene encodes one member of the sodium channel alpha subunit gene family. It is expressed in skeletal muscle, and mutations in this gene have been linked to several myotonia and periodic paralysis disorders. [provided by RefSeq, Jul 2008],disease:Defects in SCN4A are a cause of periodic paralysis hypokalemic (HOKPP) [MIM:170400]; also designated HYPOPP. HOKPP is an autosomal dominant disorder manifested by episodic flaccid generalized muscle weakness associated with falls of serum potassium levels.,disease:Defects in SCN4A are the cause of a congenital myasthenic syndrome due to mutation in SCNA4 (CMSSCNA4) [MIM:603967]. CMSSCNA4 is a congenital myasthenic syndrome associated with fatigable generalized weakness and recurrent attacks of respiratory and bulbar paralysis since birth. The fatigable weakness involves lid-elevator, external ocular, facial, limb and truncal muscles and an decremental response of the compound muscle action potential on repetitive stimulation.,disease:Defects in SCN4A are the cause of myotonia SCN4A-related (MYOSCN4A) [MIM:608390]. Myotonia is characterized by sustained muscle tensing that prevents muscles from relaxing normally. Myotonia causes muscle stiffness that can interfere with movement. In some people the stiffness is very mild, while in other cases it may be severe enough to interfere with walking, running, and other activities of daily life. MYOSCN4A is a phenotypically highly variable myotonia aggravated by potassium loading, and often by cold. MYOSCN4A includes myotonia permanens and myotonia fluctuans. In myotonia permanens, the myotonia is generalized and there is a hypertrophy of the muscle, particularly in the neck and the shoulder. Attacks of severe muscle stiffness of the thoracic muscles may be life threatening due to impaired ventilation. In myotonia fluctuans, the muscle stiffness may fluctuate from day to day, provoked by exercise.,disease:Defects in SCN4A are the cause of paramyotonia congenita of von Eulenburg (PMC) [MIM:168300]. PMC is an autosomal dominant channelopathy characterized by myotonia, increased by exposure to cold, intermittent flaccid paresis, not necessarily dependent on cold or myotonia, lability of serum potassium, nonprogressive nature and lack of atrophy or hypertrophy of muscles. In some patients, myotonia is not increased by cold exposure (paramyotonia without cold paralysis). Patients may have a combination phenotype of PMC and HYPP.,disease:Defects in SCN4A are the cause of periodic paralysis hyperkalemic (HYPP) [MIM:170500]. HYPP is an autosomal dominant channelopathy characterized by episodic flaccid generalized muscle weakness associated with high levels of serum potassium. Concurrence of myotonia is found in HYPP patients.,disease:Defects in SCN4A are the cause of periodic paralysis normokalemic (NKPP) [MIM:170500]. NKPP is a disorder closely related to hyperkalemic periodic paralysis, but marked by a lack of alterations in potassium levels during attacks of muscle weakness.,domain:The sequence contains 4 internal repeats, each with 5 hydrophobic segments (S1,S2,S3,S5,S6) and one positively charged segment (S4). Segments S4 are probably the voltage-sensors and are characterized by a series of positively charged amino acids at every third position.,function:This protein mediates the voltage-dependent sodium ion permeability of 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. This sodium channel may be present in both denervated and innervated skeletal muscle.,online information:SCN4A entry,similarity:Belongs to the sodium channel family.,similarity:Contains 1 IQ domain.,subunit:Muscle sodium channels contain an alpha subunit and a smaller beta subunit. Interacts with the PDZ domain of the syntrophin SNTA1, SNTB1 and SNTB2.,

研究领域 (Research Area)

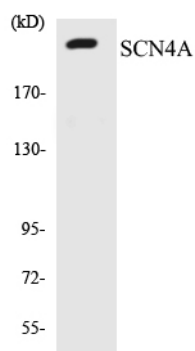
注意事项 (Note)

For research use only.

图片 (Image Data)



Western blot analysis of SCN4A Antibody. The lane on the right is blocked with the SCN4A peptide.



Western blot analysis of the lysates from COLO205 cells using SCN4A antibody.

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产品类别	产品货号	产品名称
WB 解决方案	RA10020	2h 极速 WB 即用型全流程试剂盒
	RA10021	4h 快速 WB 即用型全流程试剂盒
	RA10042	5h 畅享版 WB 全流程试剂盒
	RA10037	校准级彩色预染蛋白 Marker (8-180kDa)
	RA10038	校准级彩色预染蛋白 Marker (10-250kDa)
	RA10039	校准级高分子彩色预染蛋白 Marker (25-400kDa)
TSA 多重荧光染色试剂盒	RA10008	TSA 双标三色多重荧光染色试剂盒 (mIHC)
	RA10009	TSA 三标四色多重荧光染色试剂盒 (mIHC)
	RA10010	TSA 四标五色多重荧光染色试剂盒 (mIHC)
	RA10011	TSA 五标六色多重荧光染色试剂盒 (mIHC)
	RA10012	TSA 六标七色多重荧光染色试剂盒 (mIHC)
IHC 检测试剂盒	RA10006	HRP Anti-Mouse/Rabbit IHC Detection System
	RA10007	Polymer-HRP Anti-Mouse/Rabbit IHC Detection System
抗体标记试剂盒	RE80004p	辣根过氧化物酶(HRP)抗体标记试剂盒
	RE80002q	Sulfo-NHS-生物素标记试剂盒
	RE80007p	Cy3 荧光素标记试剂盒
	RE80011p	Fluor488 荧光素标记试剂盒
	RE80017p	Fluor750 荧光素标记试剂盒
	RE80005p	藻红蛋白(R-PE) 抗体快速标记试剂盒
	RE80040	PE-Cy7 串联染料抗体快速标记试剂盒
稳转细胞系构建服务 (免费赠送全膜 WB 验证)	TS-0001	过表达稳转细胞系构建
	TS-0002	敲低稳转细胞系构建
	TS-0003	敲除细胞系构建

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