Product Name: Na+ CP type IVα Rabbit Polyclonal Antibody
Catalog #: APRab14374

Summary

Production Name	Na+ CP type IVα Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	WB,IHC-P
Reactivity	Human,Mouse,Rat

Performance

Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Purification	Affinity purification

Immunogen

Gene Name	SCN4A
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
Gene ID	6329.0
SwissProt ID	P35499.The antiserum was produced against synthesized peptide derived from human SCN4A. AA range:431-480

Application

Dilution Ratio	WB 1:500-2000, IHC-P 1:50-300
Molecular Weight	200kDa

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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

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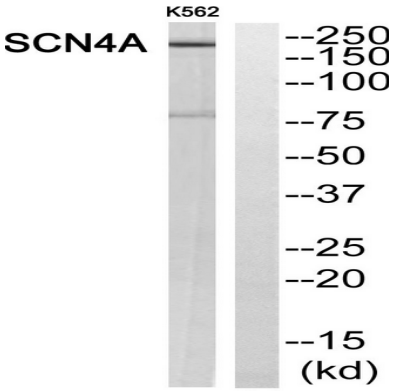
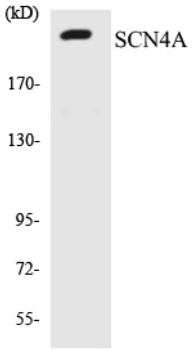
voltage-dependent sodium ion permeability of excitable membranes. Assuming opened or closed conformations in response to the voltage difference across the membrane, the protein forms a sodium-selective channel through which Na(+) ions may pass in accordance with their electrochemical gradient. This sodium 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.,

Research Area

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.

Note

For research use only.