Na+ CP type IVα Rabbit pAb

CatalogNo: YT2966

Key Features

Host Species
- Rabbit

Reactivity
- Human,Mouse,Rat

Applications
- WB,IHC

MW
- 200kD (Observed)

Isotype
- IgG

Recommended Dilution Ratios

WB 1:500-2000
IHC 1:50-300

Storage

Storage*	-15°C to -25°C/1 year(Do not lower than -25°C)
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

Basic Information

Clonality	Polyclonal

Immunogen Information

Immunogen	The antiserum was produced against synthesized peptide derived from human SCN4A. AA range:431-480
Specificity	Na+ CP type IVα Polyclonal Antibody detects endogenous levels of Na+ CP type IVα protein.

Target Information

Gene name SCN4A

Protein Name Sodium channel protein type 4 subunit alpha

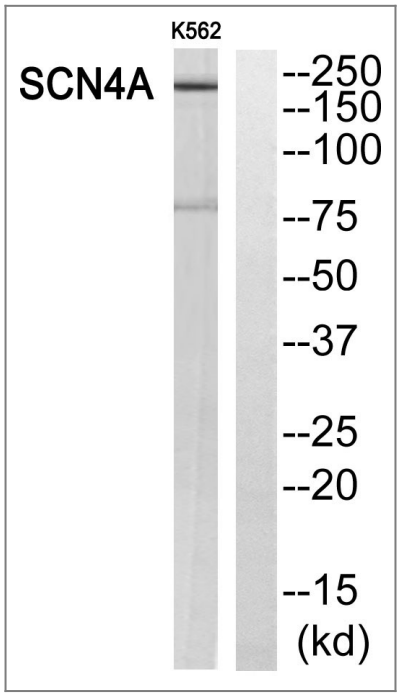
Organism	Gene ID	UniProt ID
Human	6329;	P35499;
Mouse	110880;	Q9ER60;
Rat	25722;	P15390;

Cellular Localization Cell membrane ; Multi-pass membrane protein .

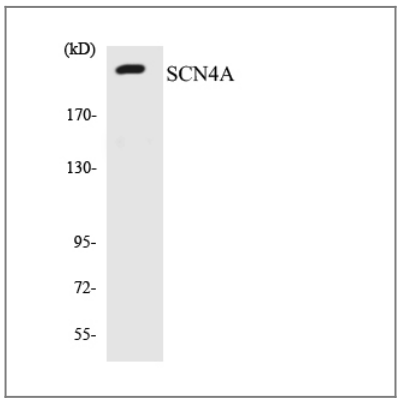
Tissue specificity Skeletal muscle,

Function 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.,

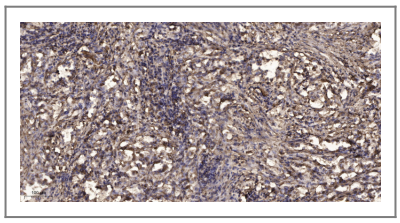
Validation Data



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.



Immunohistochemical analysis of paraffin-embedded human Squamous cell carcinoma of lung. 1, Antibody was diluted at 1:200(4° overnight). 2, Tris-EDTA,pH9.0 was used for antigen retrieval. 3,Secondary antibody was diluted at 1:200(room temperature, 45min).

Contact information

Orders: order.cn@immunoway.com
Support: support.cn@immunoway.com
Telephone: 400-8787-807(China)
Website: http://www.immunoway.com.cn
Address: 2200 Ringwood Ave San Jose, CA 95131 USA

Please scan the QR code to access additional product information:
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