



Rabbit Anti-GJC2 antibody

SL11050R

Product Name:	GJC2
Chinese Name:	间隙连接蛋白47抗体
Alias:	Connexin 46.6; Connexin 47; Connexin-46.6; Connexin-47; Connexin46.6; Connexin47; CX 46.6; Cx 47; Cx46.6; Cx47; CXG2_HUMAN; Gap junction alpha 12 protein; Gap junction alpha-12 protein; Gap junction gamma 2 protein; Gap junction gamma-2 protein; Gap junction protein alpha 12 47kDa; Gap junction protein gamma 2 47kDa; GAP JUNCTION PROTEIN, 47-KD; gap junction protein, gamma 12, 47kDa; gap junction protein, gamma 2, 47kDa; GJA 12; GJA12; GJC 2; Gjc2; HLD 2; HLD2; PMLDAR; SPG44; CXG2_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Pig,Cow,Rabbit,Sheep,Guinea Pig,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	47kDa
Cellular localization:	The cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human GJC2/Connexin 47:21-120/439<Extracellular>
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed

This gene encodes a gap junction protein. Gap junction proteins are members of a large family of homologous connexins and comprise 4 transmembrane, 2 extracellular, and 3 cytoplasmic domains. This gene plays a key role in central myelination and is involved in peripheral myelination in humans. Defects in this gene are the cause of autosomal recessive Pelizaeus-Merzbacher-like disease-1. [provided by RefSeq, Jul 2008]

Function:

One gap junction consists of a cluster of closely packed pairs of transmembrane channels, the connexons, through which materials of low MW diffuse from one cell to a neighboring cell. May play a role in myelination in central and peripheral nervous systems.

Subunit:

A connexon is composed of a hexamer of connexins. Interacts with TJP1 (By similarity).

Subcellular Location:

Cell membrane. Cell junction; gap junction.

Tissue Specificity:

Expressed in central nervous system, in sciatic nerve and sural nerve. Also detected in skeletal muscles.

DISEASE:

Defects in GJC2 are the cause of leukodystrophy hypomyelinating type 2 (HLD2) ; also known as Pelizaeus-Merzbacher-like disease autosomal recessive type 1. HLD2 is an autosomal recessive hypomyelinating leukodystrophy characterized by nystagmus, impaired motor development, ataxia, choreoathetotic movements, dysarthria and progressive spasticity.

Defects in GJC2 are the cause of spastic paraplegia autosomal recessive type 44 (SPG44). A form of spastic paraplegia, a neurodegenerative disorder characterized by a slow, gradual, progressive weakness and spasticity of the lower limbs. Rate of progression and the severity of symptoms are quite variable. Initial symptoms may include difficulty with balance, weakness and stiffness in the legs, muscle spasms, and dragging the toes when walking. In some forms of the disorder, bladder symptoms (such as incontinence) may appear, or the weakness and stiffness may spread to other parts of the body.

Similarity:

Belongs to the connexin family. Gamma-type subfamily.

SWISS:

Q5T442

Gene ID:

57165

Product Detail:

Database links:

[Entrez Gene: 57165](#) Human

[Entrez Gene: 118454](#) Mouse

[Entrez Gene: 497913](#) Rat

[Omim: 608803](#) Human

[SwissProt: Q5T442](#) Human

[SwissProt: Q8BQU6](#) Mouse

[SwissProt: Q80XF7](#) Rat

[Unigene: 100072](#) Human

[Unigene: 40016](#) Mouse

[Unigene: 203000](#) Rat

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

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