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Rabbit Anti-PMP22 antibody

SL0235R

Product Name:	PMP22
Chinese Name:	外周髓鞘蛋白-22抗体
Alias:	GAS3; CMT1A; CMT1E; DSS; GAS-3; Growth Arrest Specific 3; Growth arrest-specific protein 3; HMSNIA; HNPP; MGC20769; Peripheral Myelin Protein 22; PMP-22; PMP22; PMP22_HUMAN; Sp110; Trembler.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=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:	22kDa
Cellular localization:	The cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human PMP-22:101-160/160<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
Product Detail:	PMP22 is a 22 kDa glycoprotein expressed in the compact myelin of the peripheral nervous system. In the peripheral nervous system, PMP 22 is produced by myelinating Schwann cells and is coexpressed with the genes for myelin basic protein (MBP) during nerve development and regeneration. Alterations in the level of this protein cause several

genetic human diseases. If the protein is duplicated, patients develop Charcot Marie Tooth disease. If one copy of the gene is deleted, they suffer from the inherited tendency to pressure palsies.

Function:
Might be involved in growth regulation, and in myelinization in the peripheral nervous system.

Subcellular Location:
Cell membrane; Multi-pass membrane protein.

DISEASE:
Defects in PMP22 are the cause of Charcot-Marie-Tooth disease type 1A (CMT1A) [MIM:118220]; also known as hereditary motor and sensory neuropathy IA. CMT1A is a form of Charcot-Marie-Tooth disease, the most common inherited disorder of the peripheral nervous system. Charcot-Marie-Tooth disease is classified in two main groups on the basis of electrophysiologic properties and histopathology: primary peripheral demyelinating neuropathy or CMT1, and primary peripheral axonal neuropathy or CMT2. Neuropathies of the CMT1 group are characterized by severely reduced nerve conduction velocities (less than 38 m/sec), segmental demyelination and remyelination with onion bulb formations on nerve biopsy, slowly progressive distal muscle atrophy and weakness, absent deep tendon reflexes, and hollow feet. CMT1A inheritance is autosomal dominant.
Defects in PMP22 are a cause of Dejerine-Sottas syndrome (DSS) [MIM:145900]; also known as Dejerine-Sottas neuropathy (DSN) or hereditary motor and sensory neuropathy III (HMSN3). DSS is a severe degenerating neuropathy of the demyelinating Charcot-Marie-Tooth disease category, with onset by age 2 years. DSS is characterized by motor and sensory neuropathy with very slow nerve conduction velocities, increased cerebrospinal fluid protein concentrations, hypertrophic nerve changes, delayed age of walking as well as areflexia. There are both autosomal dominant and autosomal recessive forms of Dejerine-Sottas syndrome.
Defects in PMP22 are a cause of hereditary neuropathy with liability to pressure palsies (HNPP) [MIM:162500]; an autosomal dominant disorder characterized by transient episodes of decreased perception or peripheral nerve palsies after slight traction, compression or minor traumas.
Defects in PMP22 are the cause of Charcot-Marie-Tooth disease type 1E (CMT1E) [MIM:118300]; also known as Charcot-Marie-Tooth disease and deafness autosomal dominant. CMT1E is an autosomal dominant form of Charcot-Marie-Tooth disease characterized by the association of sensorineural hearing loss with peripheral demyelinating neuropathy.
Defects in PMP22 may be a cause of inflammatory demyelinating polyneuropathy (IDP) [MIM:139393]. IDP is a putative autoimmune disorder presenting in an acute (AIDP) or chronic form (CIDP). The acute form is also known as Guillain-Barre syndrome.

Similarity:
Belongs to the PMP-22/EMP/MP20 family.

SWISS:
Q01453

Gene ID:
5376

Database links:

Entrez Gene: 5376Human

Entrez Gene: 24660Rat

Omim: 601097Human

SwissProt: Q01453Human

SwissProt: P25094Rat

SwissProt: Q07066Rat

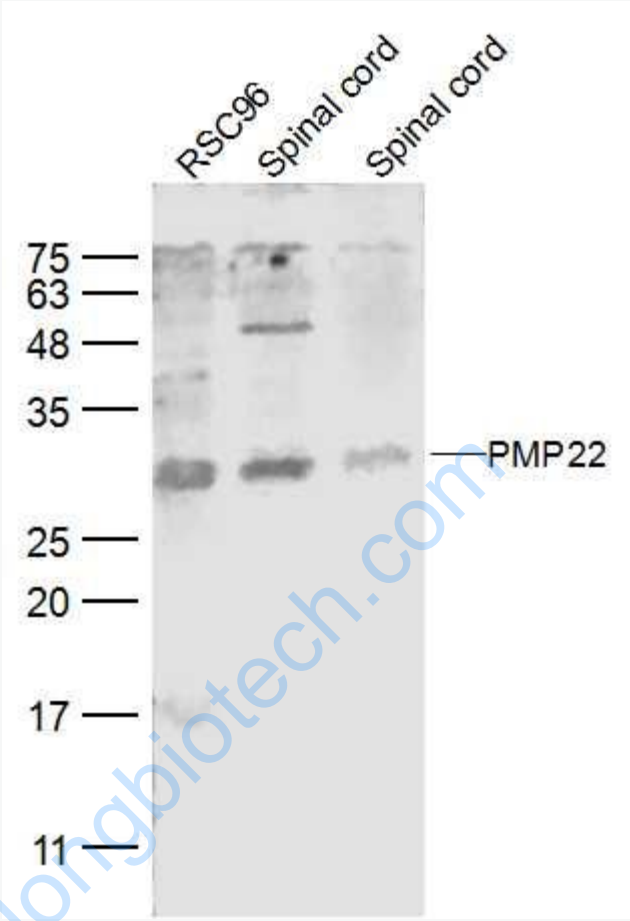
Unigene: 372031Human

Unigene: 1476Rat

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

Neurobiology相关蛋白(Neurobiology)
外周髓鞘蛋白22(PMP22)是一种glycoprotein，在外周神经系统中的致密肌纤维素中表达。它由髓鞘雪旺氏细胞产生，并在神经发育和再生过程中与MBP和Po蛋白共同表达。该蛋白表达水平变化可引起几种人类遗传性疾病，如果该蛋白增多，则病人会发生Charcot-Marie-Tooth疾病，如果缺失，则易患压力麻痹的遗传倾向。

Picture:



Sample:

RSC96 (Mouse) CellLysate at 30 ug

Spinal cord (Mouse) Lysate at 40 ug

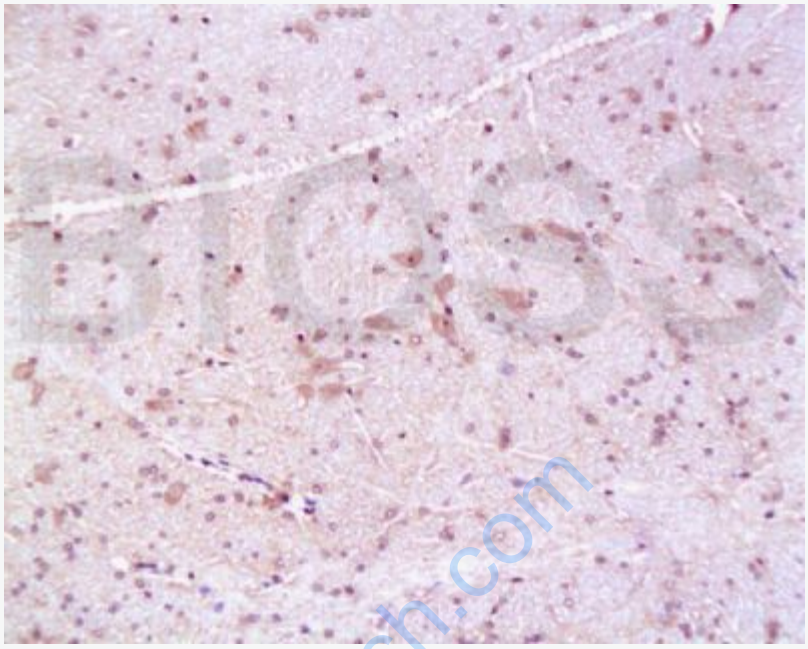
Spinal cord (Rat) Lysate at 40 ug

Primary: Anti-PMP22 (SL0235R) at 1/500 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

Predicted band size: 22 kD

Observed band size: 27 kD



Tissue/cell: rat brain tissue; 4% Paraformaldehyde-fixed and paraffin-embedded; Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min;

Incubation: Anti-PMP22 Polyclonal Antibody, Unconjugated(SL0235R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining