

PMP22 (PMP22H1): sc-65739

BACKGROUND

PLP (myelin proteolipid protein or lipophilin) is a major constituent of myelin. The two isoforms of the myelin proteolipid protein, PLP and DM20, are very hydrophobic integral membrane proteins that account for about half of the protein content of adult CNS Myelin. A mutation in the gene which encodes PLP is linked to Pelizaeus-Merzbacher disease (PMD), a chronic infantile type of diffuse cerebral sclerosis. The gene which encodes PLP maps to human chromosome Xq13-q22. The glycoprotein zero (also designated P-zero or myelin peripheral protein) is the major structural protein of peripheral myelin, accounting for more than 50% of the protein present in the sheath of peripheral nerves. Zero is an integral membrane glycoprotein whose expression is restricted to Schwann cells. The gene which encodes zero maps to human chromosome 1q22. PMP22 (peripheral myelin protein 22) is a growth-regulated membrane protein which is expressed by Schwann cells and is localized mainly in compact peripheral nervous system myelin. The gene which encodes PMP22 maps to human chromosome 17p12.

REFERENCES

1. Ford, F.R. 1960. Diseases of the Nervous System in Infancy, Childhood and Adolescence, 4th ed. Springfield, IL: Charles C. Thomas Publisher, 831-833.
2. Willard, H.F. and Riordan, J.R. 1985. Assignment of the gene for myelin proteolipid protein to the X chromosome: implications for X-linked myelin disorders. *Science* 230: 940-942.
3. Mattei, M.G., et al. 1986. The gene encoding for the major brain proteolipid (PLP) maps on the q-22 band of the human X chromosome. *Hum. Genet.* 72: 352-353.
4. Patel, P.I., et al. 1992. The gene for the peripheral myelin protein PMP22 is a candidate for Charcot-Marie-Tooth disease type 1A. *Nat. Genet.* 1: 159-165.
5. Suter, U., et al. 1992. A leucine-to-proline mutation in the putative first transmembrane domain of the 22 kDa peripheral myelin protein in the trembler-J mouse. *Proc. Natl. Acad. Sci. USA* 89: 4382-4386.
6. Hayasaka, K., et al. 1993. Structure and chromosomal localization of the gene encoding the human myelin protein zero (MPZ). *Genomics* 17: 755-758.

CHROMOSOMAL LOCATION

Genetic locus: PMP22 (human) mapping to 17p12.

SOURCE

PMP22 (PMP22H1) is a mouse monoclonal antibody raised against a 13-mer peptide in the second extracellular domain of PMP22 of human origin.

PRODUCT

Each vial contains 200 µg IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

APPLICATIONS

PMP22 (PMP22H1) is recommended for detection of residues 121-123 of PMP22 of human and rhesus monkey origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500); non cross-reactive with mouse, rat and bovine nerve.

Suitable for use as control antibody for PMP22 siRNA (h): sc-42036, PMP22 shRNA Plasmid (h): sc-42036-SH and PMP22 shRNA (h) Lentiviral Particles: sc-42036-V.

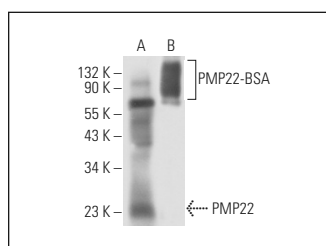
Molecular Weight of PMP22: 22 kDa.

Positive Controls: human brain extract: sc-364375.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



PMP22 (PMP22H1): sc-65739. Western blot analysis of PMP22 expression in human brain tissue extract (A) and human recombinant PMP22-BSA (B).

SELECT PRODUCT CITATIONS

1. Poitelon, Y., et al. 2018. A dual role for Integrin $\alpha 6 \beta 4$ in modulating hereditary neuropathy with liability to pressure palsies. *J. Neurochem.* 145: 245-257.
2. Di Tomaso, M.V., et al. 2022. Colocalization analysis of peripheral myelin protein-22 and lamin-B1 in the Schwann cell nuclei of Wt and TrJ mice. *Biomolecules* 12: 456.

RESEARCH USE

For research use only, not for use in diagnostic procedures.