

PLP1 Rabbit pAb

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Catalog # AP50859

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P60201
Predicted	Rat, Dog, Human, Mouse, Rabbit, Monkey, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	30077
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Myelin proteolipid protein
Epitope Specificity	21-120/277
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Membrane; Multi-pass membrane protein.
DISEASE	Defects in PLP1 are the cause of leukodystrophy hypomyelinating type 1 (HLD1) [MIM:312080]; also known as Pelizaeus-Merzbacher disease. HLD1 is an X-linked recessive dysmyelinating disorder of the central nervous system in which myelin is not formed properly. It is characterized clinically by nystagmus, spastic quadriplegia, ataxia, and developmental delay. Defects in PLP1 are the cause of spastic paraplegia X-linked type 2 (SPG2) [MIM:312920]. SPG2 is characterized by spastic gait and hyperreflexia. In some patients, complicating features include nystagmus, dysarthria, sensory disturbance, mental retardation, optic atrophy.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	PLP 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 Xq22. The glycoprotein zero (also designated P-zero or myelin peripheral protein) is the primary structural protein of peripheral myelin, and accounts for more than 50% of the protein present in the peripheral nerve sheath. Zero is an integral membrane glycoprotein. Expression of zero 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 primarily in compact peripheral nervous system myelin. The gene which encodes PMP22 maps to human chromosome 17p11.2.

Additional Information

Gene ID	5354
Other Names	Myelin proteolipid protein, PLP, Lipophilin, PLP1, PLP
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	PLP1
Synonyms	PLP
Function	This is the major myelin protein from the central nervous system. It plays an important role in the formation or maintenance of the multilamellar structure of myelin.
Cellular Location	Cell membrane; Multi-pass membrane protein. Myelin membrane. Note=Colocalizes with SIRT2 in internodal regions, at paranodal axoglial junction and Schmidt-Lanterman incisures of myelin sheath.

Background

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References

Diehl H.-J.,et al.Proc. Natl. Acad. Sci. U.S.A. 83:9807-9811(1986).
Simons R.,et al.Biochem. Biophys. Res. Commun. 146:666-671(1987).
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Ebert L.,et al.Submitted (JUN-2004) to the EMBL/GenBank/DDBJ databases.
Ross M.T.,et al.Nature 434:325-337(2005).

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