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MAN2B1 Rabbit pAb

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Catalog # AP57198

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	O00754
Reactivity	Mouse
Predicted	Rat, Pig, Dog, Human, Rabbit
Host	Rabbit
Clonality	Polyclonal
Calculated MW	113744
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human MAN2B1
Epitope Specificity	151-250/1011
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	Lysosome.
DISEASE	Mannosidosis, alpha B, lysosomal (MANSA) [MIM:248500]: A lysosomal storage disease characterized by accumulation of unbranched oligosaccharide chains. This accumulation is expressed histologically as cytoplasmic vacuolation predominantly in the CNS and parenchymatous organs. Depending on the clinical findings at the age of onset, a severe infantile (type I) and a mild juvenile (type II) form of alpha-mannosidosis are recognized. There is considerable variation in the clinical expression with mental retardation, recurrent infections, impaired hearing and Hurler-like skeletal changes being the most consistent abnormalities. Note: The disease is caused by mutations affecting the gene represented in this entry.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes an enzyme that hydrolyzes terminal, non-reducing alpha-D-mannose residues in alpha-D-mannosides. Its activity is necessary for the catabolism of N-linked carbohydrates released during glycoprotein turnover and it is member of family 38 of glycosyl hydrolases. The full length protein is processed in two steps. First, a 49 aa leader sequence is cleaved off and the remainder of the protein is processed into 3 peptides of 70 kDa, 42 kDa (D) and 13/15 kDa (E). Next, the 70 kDa peptide is further processed into three peptides (A, B and C). The A, B and C peptides are disulfide-linked. Defects in this gene have been associated with lysosomal alpha-mannosidosis. Alternatively spliced transcript variants encoding different isoforms have been found for this gene.[provided by RefSeq, Mar 2010]

Additional Information

Gene ID	4125
Other Names	Lysosomal alpha-mannosidase, Laman, 3.2.1.24, Lysosomal acid alpha-mannosidase, Mannosidase alpha class 2B member 1, Mannosidase alpha-B, Lysosomal alpha-mannosidase A peptide, Lysosomal alpha-mannosidase B peptide, Lysosomal alpha-mannosidase C peptide, Lysosomal alpha-mannosidase D peptide, Lysosomal alpha-mannosidase E peptide, MAN2B1 (HGNC:6826), LAMAN, MANB
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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	MAN2B1 (HGNC:6826)
Synonyms	LAMAN, MANB
Function	Can hydrolyze a variety of glycan substrates containing terminal alpha-mannosidic linkages. Cleaves alpha 1,2-, alpha 1,3-, and alpha 1,6-linked mannose residues on oligosaccharides generated by N- glycoprotein degradation pathways.
Cellular Location	Lysosome lumen. Secreted. Note=The protein may be secreted in an unprocessed but enzymatically active form (PubMed:11350179, PubMed:21505070). Secreted enzyme may enter cells via endocytosis (PubMed:11350179).

Background

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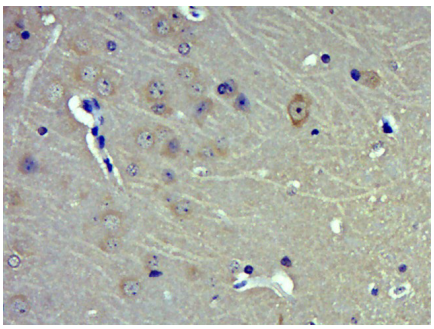
Images



Paraformaldehyde-fixed, paraffin embedded (Mouse brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (MAN2B1) Polyclonal Antibody, Unconjugated (AP57198) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.