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Rabbit Anti-MAN2B1 antibody

SL18647R

Product Name:	MAN2B1
Chinese Name:	溶酶体α-甘露糖苷酶抗体
Alias:	EC 3.2.1.24; Laman; Lysosomal acid alpha mannosidase; Lysosomal alpha mannosidase; MANB; Mannosidase alpha class 2B member 1; Mannosidase, alpha B; Mannosidase, alpha B, lysosomal.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Rabbit,
Applications:	ELISA=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:	109kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human MAN2B1:151-250/1011
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:	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]

Function:
Necessary for the catabolism of N-linked carbohydrates released during glycoprotein turnover. Cleaves all known types of alpha-mannosidic linkages.

Subcellular Location:
Lysosome.

Post-translational modifications:
First processed into 3 peptides of 70 kDa, 42 kDa (D) and 13/15 kDa (E). The 70 kDa peptide is further processed into three peptides (A, B and C). The A, B and C peptides are disulfide-linked.

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.

Similarity:
Belongs to the glycosyl hydrolase 38 family.

SWISS:
O00754

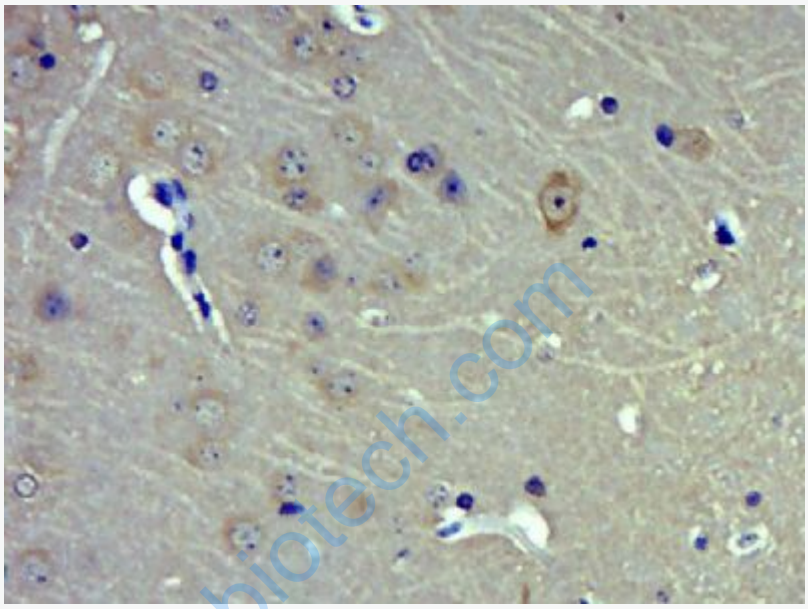
Gene ID:
4125

Database links:

Entrez Gene: 4125 Human

Omim: 609458 Human

SwissProt: O00754 Human

	Important Note: This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 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 (SL18647R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.