N-乙酰基-α葡萄糖苷酶抗体

产品货号： mlR19002

英文名称： NAGLU

中文名称： N-乙酰基-α葡萄糖苷酶抗体

别　　名: Alpha N acetylglucosaminidase; alpha N acetylglucosaminidase, lysosomal; Alpha-N-acetylglucosaminidase 77 kDa form; ANAG; ANAG_HUMAN; MPS IIIB; MPS3B; N acetyl alpha glucosaminidase; N acetylglucosaminidase, alpha; N-acetyl-alpha-glucosaminidase; NAG; NAGLU; UFHSD 1; UFHSD; UFHSD1.

研究领域： 细胞生物　神经生物学　信号转导　新陈代谢

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Dog, Pig, Horse, Rabbit, Sheep,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500 （石蜡切片需做抗原修复）
not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.

分 子 量： 80kDa

细胞定位： 细胞浆

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human NAGLU:121-220/743

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： 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

产品介绍： This gene encodes an enzyme that degrades heparan sulfate by hydrolysis of terminal N-acetyl-D-glucosamine residues in N-acetyl-alpha-D-glucosaminides. Defects in this gene are the cause of mucopolysaccharidosis type IIIB (MPS-IIIB), also known as Sanfilippo syndrome B. This disease is characterized by the lysosomal accumulation and urinary excretion of heparan sulfate. [provided by RefSeq, Jul 2008]

Function:

Involved in the degradation of heparan sulfate.

Subcellular Location:

Lysosome.

Tissue Specificity:

Liver, ovary, peripheral blood leukocytes, testis, prostate, spleen, colon, lung, placenta and kidney.

DISEASE:

Defects in NAGLU are the cause of mucopolysaccharidosis type 3B (MPS3B) [MIM:252920]; also known as Sanfilippo syndrome B. MPS3B is a form of mucopolysaccharidosis type 3, an autosomal recessive lysosomal storage disease due to impaired degradation of heparan sulfate. MPS3 is characterized by severe central nervous system degeneration, but only mild somatic disease. Onset of clinical features usually occurs between 2 and 6 years; severe neurologic degeneration occurs in most patients between 6 and 10 years of age, and death occurs typically during the second or third decade of life.

SWISS:

P54802

Gene ID:

4669

Important Note:

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