N 聚糖酶多肽蛋白 1 抗体

产品货号： mlR19233

英文名称： NGLY1

中文名称： N 聚糖酶多肽蛋白 1 抗体

别　　名： CDG1V; hPNGase; N glycanase 1; NGLY 1; Peptide N(4) (N acetyl beta glucosaminyl asparagine amidase; Peptide:N glycanase; PNG1; PNGase.

研究领域： 细胞生物　神经生物学　转录调节因子　泛素

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Pig, Cow, Rabbit,

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： 74kDa

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 NGLY1:231-330/654

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 catalyzes hydrolysis of an N(4)-(acetyl-beta-D-glucosaminyl) asparagine residue to N-acetyl-beta-D-glucosaminylamine and a peptide containing an aspartate residue. The encoded enzyme may play a role in the proteasome-mediated degradation of misfolded glycoproteins. Multiple transcript variants encoding different isoforms have been found for this gene.[provided by RefSeq, Feb 2009]

Function:

Specifically deglycosylates the denatured form of N-linked glycoproteins in the cytoplasm and assists their proteasome-mediated degradation. Cleaves the beta-aspartyl-glucosamine (GlcNAc) of the glycan and the amide side chain of Asn, converting Asn to Asp. Prefers proteins containing high-mannose over those bearing complex type oligosaccharides. Can recognize misfolded proteins in the endoplasmic reticulum that are exported to the cytosol to be destroyed and deglycosylate them, while it has no activity toward native proteins. Deglycosylation is a prerequisite for subsequent proteasome-mediated degradation of some, but not all, misfolded glycoproteins.

Subunit:

Component of a complex required to couple retrotranslocation, ubiquitination and deglycosylation composed of NGLY1, SAKS1, AMFR, VCP and RAD23B. Interacts with the proteasome components RAD23B and PSMC1. Interacts with directly with VCP. Interacts with DERL1, bringing it close to the endoplasmic reticulum membrane. Interacts with SAKS1

Subcellular Location:

Cytoplasm

DISEASE:

A multisystem disorder caused by a defect in glycoprotein biosynthesis and characterized by under-glycosylated serum glycoproteins. Characterized by developmental delay, hypotonia, abnormal involuntary movements and alacrima or poor tear production. Other features include microcephaly, intractable seizures, abnormal eye movements and evidence of liver dysfunction, probably due to cytoplasmic accumulation of storage material in vacuoles. The broad spectrum of features reflects the critical role of N-glycoproteins during embryonic development, differentiation, and maintenance of cell functions.

Similarity:

Belongs to the transglutaminase-like superfamily. PNGase family.

Contains 1 PAW domain.

Contains 1 PUB (PUG) domain.

SWISS:

Q96IV0

Gene ID:

55768

Important Note:

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

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