GNPTAB Rabbit pAb

产品详情

产品货号	产品名称	储存条件	保质期
IM72058	GNPTAB Rabbit pAb	-20℃	1 年

产品简介：

产品货号	IM72058
产品名称	GNPTAB Rabbit pAb
别名	GNPTA;ICD;EG432486;RGD1564821;GNPTA_HUMAN;GNPTAB;GlcNAc-1-phosphotransferase subunits alpha/beta;Stealth protein GNPTAB; UDP-N-acetylglucosamine-1-phosphotransferase subunits alpha/beta; 2.7.8.17;KIAA1208;GNPTA_MOUSE.
抗体来源	Rabbit
克隆类型	Polyclonal
交叉反应	predicted:Human,Mouse,Rat,Pig,Sheep,Cow,Dog.
产品应用	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,ICC/IF=1:100-500,ELISA=1:5000-10000 not yet tested in other applications.optimal dilutions/concentrations should be determined by theend user.
理论分子量	143kDa
细胞定位	细胞浆、细胞膜
性状	Liquid
浓度	1mg/ml

免疫原	KLH conjugated synthetic peptide derived from human N-acetylglucosamine-1-phosphotransferase subunit beta: 901-1000/1256.
亚型	IgG
纯化方法	Affinity purified by Protein A.
缓冲液	0.01M TBS(pH7.4)with 1% BSA,0.02% Proclin300 and 50% Glycerol.
产品介绍	This gene encodes two of three subunit types of the membrane-bound enzyme N-acetylglucosamine-1-phosphotransferase,a heterohexameric complex composed of two alpha,two beta,and two gamma subunits.The encoded protein is proteolytically cleaved at the Lys928-Asp929 bond to yield mature alpha and beta polypeptides while the gamma subunits are the product of a distinct gene(GeneID 84572).In the Golgi apparatus,the heterohexameric complex catalyzes the first step in the synthesis of mannose 6-phosphate recognition markers on certain oligosaccharides of newly synthesized lysosomal enzymes.These recognition markers are essential for appropriate trafficking of lysosomal enzymes.Mutations in this gene have been associated with both mucolipidosis II and mucolipidosis IIIA.[provided by RefSeq, May 2010].
Function	Catalyzes the formation of mannose 6-phosphate(M6P)markers on high mannose type oligosaccharides in the Golgi apparatus.M6P residues are required to bind to the M6P receptors(MPR),which mediate the vesicular transport of lysosomal enzymes to the endosomal/ prelysosomal compartment.
Subunit	Hexamer of two alpha,two beta and two gamma subunits;

Subunit	disulfide-linked. It is believed that the alpha and/or the beta subunit of the enzyme contain the catalytic portion and that the gamma subunit functions in recognition of the lysosomal enzymes.
Subcellular Location	N-acetylglucosamine-1-phosphotransferase subunit alpha:Golgi apparatus membrane;Single-pass type I membrane protein. N-acetylglucosamine-1-phosphotransferase subunit beta:Golgi apparatus membrane;Single-pass type II membrane protein.
Tissue Specificity	Expressed in the heart, whole brain, placenta, lung, liver, skeletal muscle, kidney and pancreas.
Post-translational modifications	The alpha- and beta-subunits appear to be generated by a proteolytic cleavage at the Lys-928-Asp-929 bond.
DISEASE	Defects in GNPTAB are the cause of mucolipidosis type II(MLII) [MIM:252500];also known as inclusion cell disease or I-cell disease(ICD). MLII is a fatal, autosomal recessive, lysosomal storage disorder characterized by severe clinical and radiologic features, peculiar fibroblast inclusions, and no excessive mucopolysaccharidurial. Congenital dislocation of the hip, thoracic deformities, hernia, and hyperplastic gums are evident soon after birth. Defects in GNPTAB are the cause of mucolipidosis type III complementation group A(MLIIIA)[MIM:252600];also known as variant pseudo-Hurler polydystrophy. MLIIIA is an autosomal recessive disease of lysosomal enzyme targeting. Clinically MLIII is characterized by restricted joint mobility, skeletal dysplasia, and short stature. Mildly coarsened facial features and thickening of the skin have been described. Cardiac valvular disease and corneal clouding may also occur. Half of the reported patients show learning disabilities or mental retardation.

Similarity	Belongs to the stealth family.Contains 1 EF-hand domain.Contains 2 LNR(Lin/Notch)repeats.
SWISS	Q3T906
Gene ID	79158

储存与保存：

1.保存：-20℃
2.有效期：1年

注意事项：

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