Product Name: GLCNE Rabbit Polyclonal Antibody
Catalog #: APRab11459

Summary

Production Name	GLCNE Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	WB
Reactivity	Human,Mouse,Rat

Performance

Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Purification	Affinity purification

Immunogen

Gene Name	GNE
Alternative Names	GNE; GLCNE; Bifunctional UDP-N-acetylglucosamine 2-epimerase/N-acetylmannosamine kinase; UDP-GlcNAc-2-epimerase/ManAc kinase
Gene ID	10020.0
SwissProt ID	Q9Y223.The antiserum was produced against synthesized peptide derived from human GNE. AA range:592-641

Application

Dilution Ratio	WB 1:500-1:2000. ELISA: 1:5000.
Molecular Weight	80kD

Background

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The protein encoded by this gene is a bifunctional enzyme that initiates and regulates the biosynthesis of N-acetylneuraminic acid (NeuAc), a precursor of sialic acids. It is a rate-limiting enzyme in the sialic acid biosynthetic pathway. Sialic acid modification of cell surface molecules is crucial for their function in many biologic processes, including cell adhesion and signal transduction. Differential sialylation of cell surface molecules is also implicated in the tumorigenicity and metastatic behavior of malignant cells. Mutations in this gene are associated with sialuria, autosomal recessive inclusion body myopathy, and Nonaka myopathy. Alternative splicing of this gene results in transcript variants encoding different isoforms. [provided by RefSeq, Jul 2008],catalytic activity:ATP + N-acyl-D-mannosamine = ADP + N-acyl-D-mannosamine 6-phosphate.,catalytic activity:UDP-N-acetyl-D-glucosamine = UDP-N-acetyl-D-mannosamine.,disease:Defects in GNE are a cause of sialuria [MIM:269921]; also known as sialuria French type. In sialuria, free sialic acid accumulates in the cytoplasm and gram quantities of neuraminic acid are secreted in the urine. The metabolic defect involves lack of feedback inhibition of UDP-GlcNAc 2-epimerase by CMP-Neu5Ac, resulting in constitutive overproduction of free Neu5Ac. Clinical features include variable degrees of developmental delay, coarse facial features and hepatomegaly. Sialuria inheritance is autosomal dominant.,disease:Defects in GNE are the cause of inclusion body myopathy type 2 (IBM2) [MIM:600737]. Hereditary inclusion body myopathies are a group of neuromuscular disorders characterized by adult onset, slowly progressive distal and proximal weakness and a typical muscle pathology including rimmed vacuoles and filamentous inclusions. IBM2 is an autosomal recessive disorder affecting mainly leg muscles, but with an unusual distribution that spares the quadriceps as also observed in Nonaka myopathy.,disease:Defects in GNE are the cause of Nonaka myopathy (NM) [MIM:605820]; also known as distal myopathy with rimmed vacuoles (DMRV). NM is an autosomal recessive muscular disorder, allelic to inclusion body myopathy 2. It is characterized by weakness of the anterior compartment of the lower limbs with onset in early adulthood, and sparing of the quadriceps muscles. As the inclusion body myopathy, NM is histologically characterized by the presence of numerous rimmed vacuoles without inflammatory changes in muscle specimens.,enzyme regulation:Allosterically regulated (Probable); feedback inhibited by cytidine monophosphate-N-acetylneuraminic acid (CMP-Neu5Ac), the end product of neuraminic acid biosynthesis. Activity is dependent on oligomerization. The monomer is inactive, whereas the dimer catalyzes only the phosphorylation of N-acetylmannosamine; the hexamer is fully active for both enzyme activities (By similarity). Up-regulated after PKC-dependent phosphorylation.,function:Regulates and initiates biosynthesis of N-acetylneuraminic acid (NeuAc), a precursor of sialic acids. Plays an essential role in early development (By similarity). Required for normal sialylation in hematopoietic cells. Sialylation is implicated in cell adhesion, signal transduction, tumorigenicity and metastatic behavior of malignant cells.,pathway:Amino-sugar metabolism; N-acetylneuraminic acid biosynthesis.,PTM:Phosphorylated by PKC.,similarity:In the C-terminal section; belongs to the ROK (nagC/xylR) family.,similarity:In the N-terminal section; belongs to the UDP-N-acetylglucosamine 2-epimerase family.,subunit:Homodimer and homohexamer.,tissue specificity:Highest expression in liver and placenta. Also found in heart, brain, lung, kidney, skeletal muscle and pancreas.,

Research Area

Amino sugar and nucleotide sugar metabolism;

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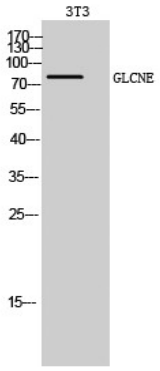
Image Data



Western Blot analysis of 3T3 cells using GLCNE Polyclonal Antibody diluted at 1：500

Note

For research use only.