



GLCNE Monoclonal Antibody

Catalog No	BYmab-14759
Isotype	IgG
Reactivity	Human;Mouse;Rat
Applications	WB
Gene Name	GNE
Protein Name	Bifunctional UDP-N-acetylglucosamine 2-epimerase/N-acetylmannosamine kinase
Immunogen	The antiserum was produced against synthesized peptide derived from human GNE. AA range:592-641
Specificity	GLCNE Monoclonal Antibody detects endogenous levels of GLCNE protein.
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	GNE; GLCNE; Bifunctional UDP-N-acetylglucosamine 2-epimerase/N-acetylmannosamine kinase; UDP-GlcNAc-2-epimerase/ManAc kinase
Observed Band	80kD
Cell Pathway	Cytoplasm .
Tissue Specificity	Highest expression in liver and placenta. Also found in heart, brain, lung, kidney, skeletal muscle and pancreas. Isoform 1 is expressed in heart, brain, kidney, liver, placenta, lung, spleen, pancreas, skeletal muscle and colon. Isoform 2 is expressed mainly in placenta, but also in brain, kidney, liver, lung, pancreas and colon. Isoform 3 is expressed at low level in kidney, liver, placenta and colon.
Function	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

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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. 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 p

Background

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],

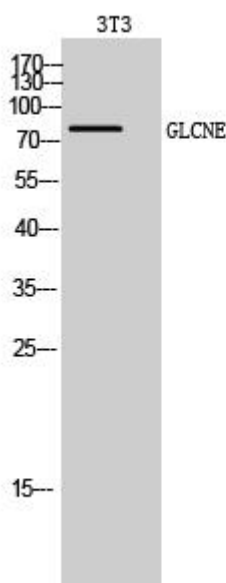
matters needing attention

Avoid repeated freezing and thawing!

Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western Blot analysis of various cells using GLCNE Monoclonal Antibody

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