abcam

Product datasheet

Anti-GNE antibody [EPR15058] ab184963

Recombinant RabMAb

4 Images

Overview

Product name	Anti-GNE antibody [EPR15058]
Description	Rabbit monoclonal [EPR15058] to GNE
Host species	Rabbit
Tested applications	**Suitable for:** WB, Flow Cyt (Intra)
Species reactivity	**Reacts with:** Human **Predicted to work with:** Mouse, Rat
Immunogen	Synthetic peptide. This information is proprietary to Abcam and/or its suppliers.
Positive control	Human placenta, fetal liver and SW480 lysates; HeLa cells.
General notes	This product is a recombinant monoclonal antibody, which offers several advantages including: - High batch-to-batch consistency and reproducibility - Improved sensitivity and specificity - Long-term security of supply - Animal-free production For more information **see here**. Our RabMAb® technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to **RabMAb® patents**.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term. Avoid freeze / thaw cycle.
Storage buffer	pH: 7.2 Preservative: 0.01% Sodium azide Constituents: 0.05% BSA, 40% Glycerol, 59% PBS
Purity	Protein A purified
Clonality	Monoclonal
Clone number	EPR15058
Isotype	IgG

Applications

The Abpromise guarantee Our **Abpromise guarantee** covers the use of ab184963 in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
WB		1/10000 - 1/50000. Detects a band of approximately 75 kDa (predicted molecular weight: 79 kDa).
Flow Cyt (Intra)		1/100. **ab172730** - Rabbit monoclonal IgG, is suitable for use as an isotype control with this antibody.

Target

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.

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.

Pathway Amino-sugar metabolism; N-acetylneuraminate biosynthesis.

Involvement in disease Defects in GNE are a cause of sialuria (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.
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.
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.

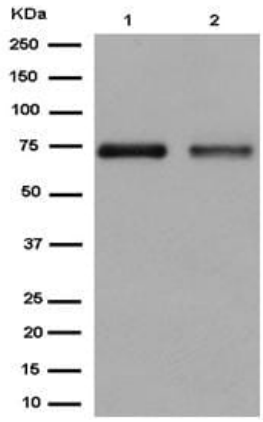
Sequence similarities In the N-terminal section; belongs to the UDP-N-acetylglucosamine 2-epimerase family.
In the C-terminal section; belongs to the ROK (NagC/XylR) family.

Post-translational Phosphorylated by PKC.

modifications

Cellular localization Cytoplasm.

Images



Western blot - Anti-GNE antibody [EPR15058] (ab184963)

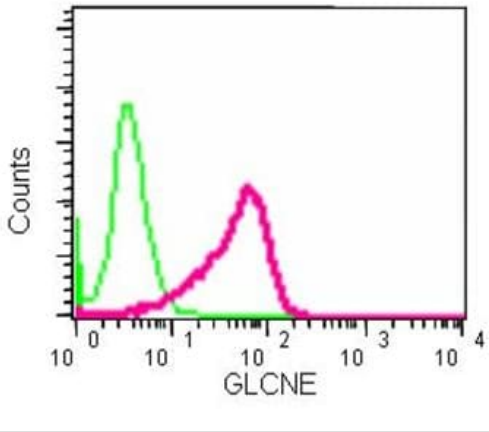
All lanes : Anti-GNE antibody [EPR15058] (ab184963) at 1/10000 dilution

Lane 1 : Human fetal liver tissue lysate
Lane 2 : Human placenta lysate

Lysates/proteins at 20 µg per lane.

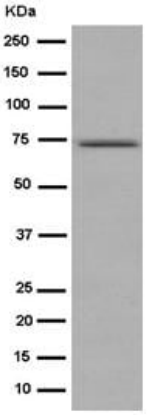
Secondary
All lanes : Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/1000 dilution

Predicted band size: 79 kDa



Flow Cytometry (Intracellular) - Anti-GNE antibody [EPR15058] (ab184963)

Intracellular flow cytometric analysis of HeLa cells (2% paraformaldehyde-fixed) labeling GNE with ab184963at 1/100 dilution (red) or a Rabbit monoclonal IgG (negative) (green)followed by Goat anti rabbit IgG (FITC) secondary at 1/150 dilution.



Western blot - Anti-GNE antibody [EPR15058] (ab184963)

Anti-GNE antibody [EPR15058] (ab184963) at 1/20000 dilution + SW480 lysate at 10 µg

Secondary

Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/1000 dilution

Predicted band size: 79 kDa

Anti-GNE antibody [EPR15058] (ab184963)

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