abcam

Product datasheet

Anti-GNE antibody [EPR15059] - C-terminal ab189927

Recombinant RabMAb

1 References 5 Images

Overview

Product name Anti-GNE antibody [EPR15059] - C-terminal

Description Rabbit monoclonal [EPR15059] to GNE - C-terminal

Host species Rabbit

Tested applications Suitable for: ICC/IF, IP, WB

Species reactivity Reacts with: Human

Predicted to work with: Rat

Immunogen Synthetic peptide. This information is proprietary to Abcam and/or its suppliers.

Human fetal brain, fetal kidney, fetal liver, HeLa, A549 and K562 lysates; HeLa cells. Positive control

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: 59% PBS, 40% Glycerol (glycerin, glycerine), 0.05% BSA

Purity Protein A purified

Clonality Monoclonal EPR15059 Clone number

Isotype lgG

Applications

The Abpromise guarantee

Our Abpromise guarantee covers the use of ab189927 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
ICC/IF		1/500.
IP		1/30 - 1/40.
WB		1/1000 - 1/10000. Detects a band of approximately 75 kDa (predicted molecular weight: 79 kDa).

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.

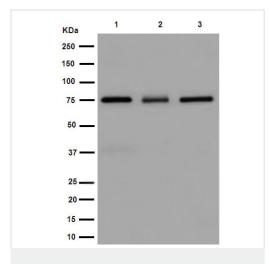
Sequence similarities

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

Post-translational

Phosphorylated by PKC.

Images



Western blot - Anti-GNE antibody [EPR15059] - Cterminal (ab189927) **All lanes :** Anti-GNE antibody [EPR15059] - C-terminal (ab189927) at 1/1000 dilution

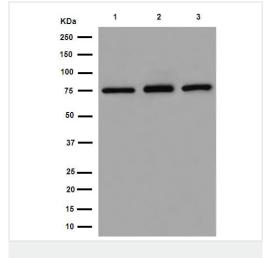
Lane 1 : Human fetal liver tissue lysate
Lane 2 : Human fetal kidney tissue lysate
Lane 3 : Human fetal brain tissue lysate

Lysates/proteins at 20 µg per lane.

Secondary

All lanes : Anti-Rabbit lgG (HRP), specific to the non-reduced form of lgG at 1/1000 dilution

Predicted band size: 79 kDa **Observed band size:** 75 kDa



Western blot - Anti-GNE antibody [EPR15059] - C-terminal (ab189927)

All lanes : Anti-GNE antibody [EPR15059] - C-terminal (ab189927) at 1/10000 dilution

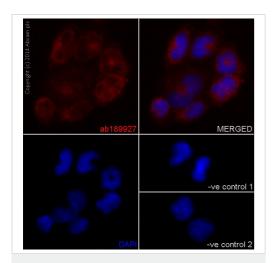
Lane 1 : HeLa cell lysate Lane 2 : A549 cell lysate Lane 3 : K562 cell lysate

Lysates/proteins at 20 µg per lane.

Secondary

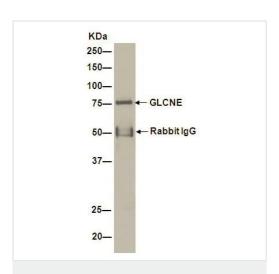
All lanes : Goat Anti-Rabbit lgG, (H+L), Peroxidase conjugate at 1/1000 dilution

Predicted band size: 79 kDa **Observed band size:** 75 kDa



Immunocytochemistry/ Immunofluorescence - Anti-GNE antibody [EPR15059] - C-terminal (ab189927)

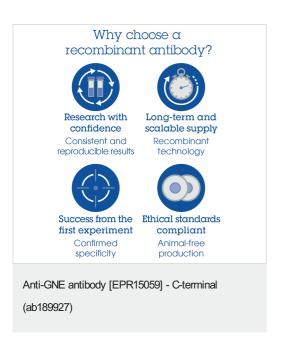
Immunofluorescent analysis of 4% paraformaldehyde-fixed HeLa cells labeling GNE with ab189927 at 1/500 dilution followed by Goat anti rabbit lgG (Alexa Fluor® 555) secondary antibody at 1/200 dilution. Counter stained with DAPI.



Immunoprecipitation - Anti-GNE antibody [EPR15059] - C-terminal (ab189927)

Western blot analysis of GNE in A549 cell lysate immunoprecipitated using ab189927 at 1/50 dilution.

Secondary antibody: Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugate at 1/1000 dilution.



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