abcam

Product datasheet

Anti-GNE antibody ab244403

3 Images

Overview

Product name	Anti-GNE antibody
Description	Rabbit polyclonal to GNE
Host species	Rabbit
Tested applications	**Suitable for:** ICC/IF, WB, IHC-P
Species reactivity	**Reacts with:** Human **Predicted to work with:** Mouse, Rat, Chinese hamster
Immunogen	Recombinant fragment corresponding to Human GNE aa 250-450. Database link: **Q9Y223** Run BLAST with Run BLAST with
Positive control	ICC/IF: A431 cells. WB: A549 cell lysate. IHC-P: Human liver tissue.
General notes	The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing. If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As

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: 7.20 Preservative: 0.02% Sodium azide Constituents: PBS, 40% Glycerol (glycerin, glycerine)
Purity	Immunogen affinity purified
Clonality	Polyclonal
Isotype	IgG

Applications

The Abpromise guarantee Our **Abpromise guarantee** covers the use of ab244403 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		Use a concentration of 0.25 - 2 µg/ml. Fixation/Permeabilization: PFA/Triton X-100.
WB		Use a concentration of 0.04 - 0.4 µg/ml.
IHC-P		1/50 - 1/200. Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.

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.

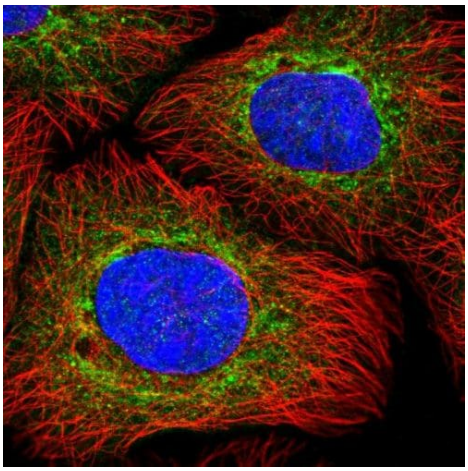
Involvement in disease Defects in GNE are a cause of sialuria (SIALURIA) [MIM:269921]; also known as sialuria French type. In sialuria, free sialic acid accumulates in the cytoplasm and gram quantities of neuraminic acid are secreted in the urine. The metabolic defect involves lack of feedback inhibition 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 modifications Phosphorylated by PKC.

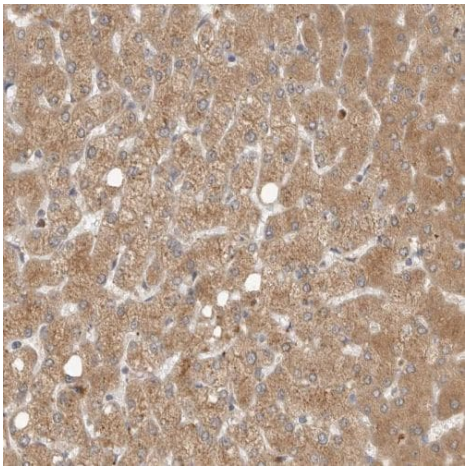
Cellular localization Cytoplasm.

Images



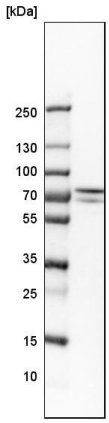
Immunocytochemistry/ Immunofluorescence - Anti-GNE antibody (ab244403)

PFA-fixed, Triton X-100 permeabilized A431 (human epidermoid carcinoma cell line) cells stained for GNE (green) using ab244403 at 4 µg/ml in ICC/IF.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-GNE antibody (ab244403)

Paraffin-embedded human liver tissue stained for GNE using ab244403 at 1/50 dilution in immunohistochemical analysis.



Western blot - Anti-GNE antibody (ab244403)

Anti-GNE antibody (ab244403) at 0.4 µg/ml + A549 (Human lung carcinoma cell line) cell lysate

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