abcam

Product datasheet

Anti-GNS antibody [EPR8329(2)] ab154177

Recombinant RabMAb

1 Image

Overview

Product name	Anti-GNS antibody [EPR8329(2)]
Description	Rabbit monoclonal [EPR8329(2)] to GNS
Host species	Rabbit
Tested applications	**Suitable for:** WB, IP **Unsuitable for:** Flow Cyt,ICC/IF or IHC-P
Species reactivity	**Reacts with:** Human
Immunogen	Synthetic peptide, corresponding to residues in Human GNS (UniProt: P15586).
Positive control	PC3, NCI-H460 and 293T cell lysates.
General notes	Mouse, Rat: We have preliminary internal testing data to indicate this antibody may not react with these species. Please contact us for more information. Our RabMAb® technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to RabMab® patents. This product is a recombinant rabbit monoclonal antibody.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Store at -20ºC.
Storage buffer	Preservative: 0.01% Sodium azide Constituents: 9% PBS, 40% Glycerol, 0.05% BSA, 50% Tissue culture supernatant
Purity	Tissue culture supernatant
Clonality	Monoclonal
Clone number	EPR8329(2)
Isotype	IgG

Applications

Our Abpromise guarantee covers the use of **ab154177** 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. Predicted molecular weight: 62 kDa.
IP		1/10 - 1/100.

Application notes Is unsuitable for Flow Cyt,ICC/IF or IHC-P.

Target

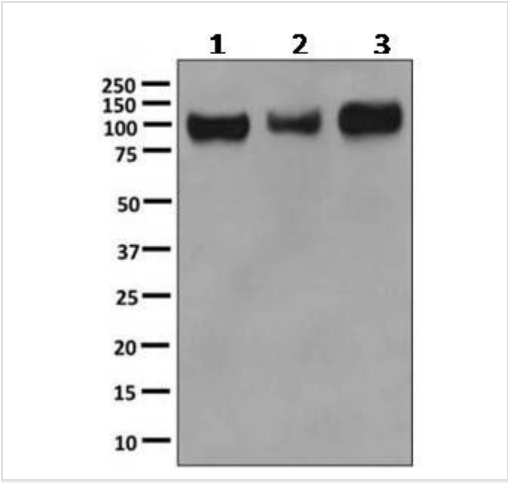
Involvement in disease Defects in GNS are the cause of mucopolysaccharidosis type 3D (MPS3D) [MIM:252940]; also known as Sanfilippo D syndrome. MPS3D is a form of mucopolysaccharidosis type 3, an autosomal recessive lysosomal storage disease due to impaired degradation of heparan sulfate. MPS3 is characterized by severe central nervous system degeneration, but only mild somatic disease. Onset of clinical features usually occurs between 2 and 6 years; severe neurologic degeneration occurs in most patients between 6 and 10 years of age, and death occurs typically during the second or third decade of life.

Sequence similarities Belongs to the sulfatase family.

Post-translational modifications The form A (78 kDa) is processed by internal peptidase cleavage to a 32 kDa N-terminal species (form B) and a 48 kDa C-terminal species.
The conversion to 3-oxoalanine (also known as C-formylglycine, FGly), of a serine or cysteine residue in prokaryotes and of a cysteine residue in eukaryotes, is critical for catalytic activity.

Cellular localization Lysosome.

Images



Western blot - Anti-GNS antibody [EPR8329(2)] (ab154177)

All lanes : Anti-GNS antibody [EPR8329(2)] (ab154177) at 1/10000 dilution

Lane 1 : PC3 cell lysate
Lane 2 : NCI-H460 cell lysate
Lane 3 : 293T cell lysate

Lysates/proteins at 10 µg per lane.

Secondary
All lanes : Goat anti-rabbit HRP at 1/2000 dilution

Predicted band size: 62 kDa

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