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GC Antibody(Center) (Ascites)

Mouse Monoclonal Antibody (Mab)
Catalog # AM2180a

Specification

GC Antibody(Center) (Ascites) - Product Information

Application	**WB,E**
Primary Accession	P04062
Other Accession	Q70KH2, Q2KHZ8, NP_000148.2
Reactivity	**Human**
Predicted	**Bovine, Pig**
Host	**Mouse**
Clonality	**Monoclonal**
Isotype	**IgM**
Calculated MW	**59716**
Antigen Region	**337-365**

GC Antibody(Center) (Ascites) - Additional Information

Gene ID 2629

Other Names
Glucosylceramidase, Acid beta-glucosidase, Alglucerase, Beta-glucocerebrosidase, Beta-GC, D-glucosyl-N-acylsphingosine glucohydrolase, Imiglucerase, GBA, GC, GLUC

Target/Specificity
This GC antibody is generated from mice immunized with a KLH conjugated synthetic peptide between 337-365 amino acids from the Central region of human GC.

Dilution
WB~~1:100~1600
E~~Use at an assay dependent concentration.

Format
Mouse monoclonal antibody supplied in crude ascites with 0.09% (W/V) sodium azide.

Storage
Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

Precautions
GC Antibody(Center) (Ascites) is for research use only and not for use in diagnostic or therapeutic procedures.

GC Antibody(Center) (Ascites) - Protein Information

Name GBA1 (HGNC:4177)

Synonyms GBA, GC, GLUC

Function Glucosylceramidase that catalyzes, within the lysosomal compartment, the hydrolysis of glucosylceramides/GlcCers (such as beta- D-glucosyl-(11')-N-acylsphing-4-enine) into free ceramides (such as N-acylsphing-4-enine) and glucose (PubMed:15916907, PubMed:24211208, PubMed:32144204, PubMed:9201993). Plays a central role in the degradation of complex lipids and the turnover of cellular membranes (PubMed:27378698). Through the production of ceramides, participates in the PKC-activated salvage pathway of ceramide formation (PubMed:19279011). Catalyzes the glucosylation of cholesterol, through a transglucosylation reaction where glucose is transferred from GlcCer to cholesterol (PubMed:24211208, PubMed:26724485, PubMed:32144204). GlcCer containing mono-unsaturated fatty acids (such as beta-D- glucosyl-N-(9Z-octadecenoyl)-sphing-4-enine) are preferred as glucose donors for cholesterol glucosylation when compared with GlcCer containing same chain length of saturated fatty acids (such as beta-D- glucosyl-N-octadecanoyl-sphing-4-enine) (PubMed:24211208). Under specific conditions, may alternatively catalyze the reverse reaction, transferring glucose from cholesteryl 3-beta-D-glucoside to ceramide (Probable) (PubMed:26724485). Can also hydrolyze cholesteryl 3-beta-D- glucoside producing glucose and cholesterol (PubMed:24211208, PubMed:26724485). Catalyzes the hydrolysis of galactosylceramides/GalCers (such as beta-D-galactosyl-(11')-N- acylsphing-4-enine), as well as the transfer of galactose between GalCers and cholesterol in vitro, but with lower activity than with GlcCers (PubMed:32144204). Contrary to GlcCer and GalCer, xylosylceramide/XylCer (such as beta-D-xyosyl-(11')-N-acylsphing-4- enine) is not a good substrate for hydrolysis, however it is a good xylose donor for transxylosylation activity to form cholesteryl 3-beta- D-xyloside (PubMed:33361282).

Cellular Location
Lysosome membrane; Peripheral membrane protein; Lumenal side. Note=Interaction with saposin-C promotes membrane association (PubMed:10781797). Targeting to lysosomes occurs through an alternative MPR-independent mechanism via SCARB2 (PubMed:18022370).

GC Antibody(Center) (Ascites) - Protocols

Provided below are standard protocols that you may find useful for product applications.

- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

GC Antibody(Center) (Ascites) - Images

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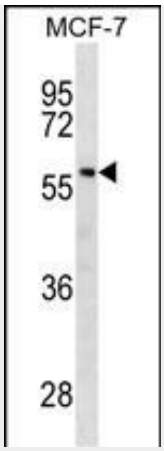

GC Antibody(Center) (Cat. #AM2180a) western blot analysis in MCF-7 cell line lysates (35µg/lane).This demonstrates the GC antibody detected the GC protein (arrow).

GC Antibody(Center) (Ascites) - Background

This gene encodes a lysosomal membrane protein that cleaves the beta-glucosidic linkage of glycosylceramide, an intermediate in glycolipid metabolism. Mutations in this gene cause Gaucher disease, a lysosomal storage disease characterized by an accumulation of glucocerebrosides. A related pseudogene is approximately 12 kb downstream of this gene on chromosome 1. Alternative splicing results in multiple transcript variants.

GC Antibody(Center) (Ascites) - References

Dos Santos, A.V., et al. Neurosci. Lett. 485(2):121-124(2010)
Bailey, S.D., et al. Diabetes Care 33(10):2250-2253(2010)
Jeong, S.Y., et al. Blood Cells Mol. Dis. (2010) In press :
Hu, F.Y., et al. Eur. J. Neurol. (2010) In press :
Velayati, A., et al. Curr Neurol Neurosci Rep 10(3):190-198(2010)