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GC Antibody(Center) (Ascites)

Mouse Monoclonal Antibody (Mab)
Catalog # AM2180a

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
Clone Names	660CT8.6.6.2
Calculated MW	59716
Antigen Region	337-365

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.

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).

Background

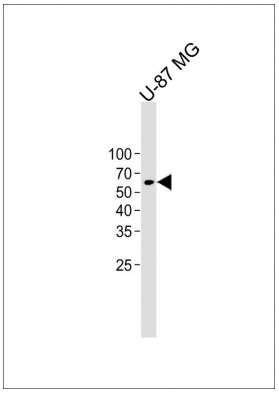
This gene encodes a lysosomal membrane protein that cleaves the beta-glucosidic linkage of glycosylceramide, an intermediate in glycolipid metabolism. Mutations in this gene cause Gaucher disease, a lysosomal storage disease characterized by an accumulation of glucocerebrosides. A related pseudogene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

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)

Images

All lanes: Anti-GC Antibody (Center) at 1:1000 dilution + U-87 MG whole cell lysate Lysates/proteins at 20 µg per lane. Secondary: Goat Anti-Mouse IgG, (H+L), Peroxidase conjugated (ASP1613) at 1/8000 dilution. Observed band



size: 60 KDa Blocking/Dilution buffer: 5% NFDM/TBST.