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GBA Antibody
Rabbit mAb
Catalog # AP91173

Specification

GBA Antibody - Product Information

Application	**WB, IHC**
Primary Accession	P04062
Reactivity	**Rat**
Clonality	**Monoclonal**

Other Names
Alglucerase; betaGC; GBA1; GCase; GCB; GLUC; Glucosylceramidase; Imiglucerase;

Isotype	**Rabbit IgG**
Host	**Rabbit**
Calculated MW	**59716 Da**

GBA Antibody - Additional Information

Purification	**Affinity-chromatography**
Immunogen	**A synthesized peptide derived from human GBA**
Description	**Defects in GBA are the cause of Gaucher disease (GD) [MIM:230800]; also known as glucocerebrosidase deficiency. GD is the most prevalent lysosomal storage disease, characterized by accumulation of glucosylceramide in the reticulo-endothelial system.**
Storage Condition and Buffer	**Rabbit IgG in phosphate buffered saline , pH 7.4, 150mM NaCl, 0.02% sodium azide and 50% glycerol. Store at +4°C short term. Store at -20°C long term. Avoid freeze / thaw cycle.**

GBA Antibody - 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-(1<->1')-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-(1<->1')-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-(1<->1')-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).

GBA Antibody - 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

GBA Antibody - Images

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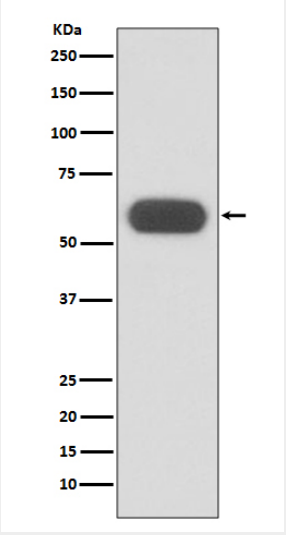

Western blot analysis of GBA expression in U87-MG cell lysate.