

# GBA Antibody

Rabbit mAb

Catalog # AP91173

## Product Information

<b>Application</b>	WB, IHC
<b>Primary Accession</b>	<a href="#">P04062</a>
<b>Reactivity</b>	Rat, Human
<b>Clonality</b>	Monoclonal
<b>Other Names</b>	Alglucerase; betaGC; GBA1; GCase; GCB; GLUC; Glucosylceramidase; Imiglucerase;
<b>Isotype</b>	Rabbit IgG
<b>Host</b>	Rabbit
<b>Calculated MW</b>	59716

## Additional Information

<b>Dilution</b>	WB 1:500~1:2000 IHC 1:50~1:200
<b>Purification</b>	Affinity-chromatography
<b>Immunogen</b>	A synthesized peptide derived from human GBA
<b>Description</b>	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.
<b>Storage Condition and Buffer</b>	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.

## Protein Information

<b>Name</b>	GBA1 ( <a href="#">HGNC:4177</a> )
<b>Synonyms</b>	GBA, GC, GLUC
<b>Function</b>	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: <a href="#">15916907</a> , PubMed: <a href="#">24211208</a> , PubMed: <a href="#">32144204</a> , PubMed: <a href="#">9201993</a> ). Plays a central role in the degradation of complex lipids and the turnover of cellular membranes (PubMed: <a href="#">27378698</a> ). Through the production of ceramides, participates in the PKC-activated salvage pathway of ceramide formation (PubMed: <a href="#">19279011</a> ). Catalyzes the glucosylation of cholesterol, through a transglucosylation reaction where glucose is transferred from GlcCer to cholesterol (PubMed: <a href="#">24211208</a> , PubMed: <a href="#">26724485</a> , PubMed: <a href="#">32144204</a> ). GlcCer containing mono-unsaturated fatty acids (such as beta-D-

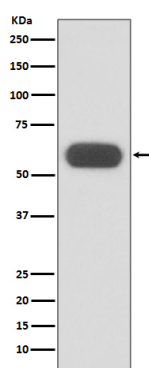
glucosyl-N-(9Z-octadecenoyl)-sphing-4-enine) are preferred as glucose donors for cholesterol glucosylation when compared with GlcCer containing same chain length of saturated fatty acids (such as beta-D-glucosyl-N-octadecanoyl-sphing-4-enine) (PubMed:[24211208](#)). Under specific conditions, may alternatively 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).

### Images

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

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