

GBA Antibody

Rabbit mAb

Catalog # AP91173

Product Information

Application	WB, IHC
Primary Accession	P04062
Reactivity	Rat, Human
Clonality	Monoclonal
Other Names	Alglucerase; betaGC; GBA1; GCase; GCB; GLUC; Glucosylceramidase; Imiglucerase;
Isotype	Rabbit IgG
Host	Rabbit
Calculated MW	59716

Additional Information

Dilution	WB 1:500~1:2000 IHC 1:50~1:200
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.

Protein Information

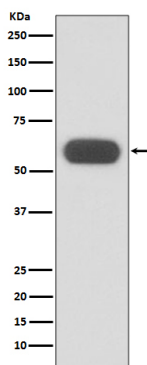
Name	GBA1 (HGNC:4177)
Synonyms	GBA, GC, GLUC
Function	Glucosylceramidase that catalyzes, within the lysosomal compartment, the hydrolysis of glucosylceramides/GlcCers (such as beta-D-glucosyl-(11')-N-acylsphing-4-ene) into free ceramides (such as N-acylsphing-4-ene) and glucose (PubMed: 15916907 , PubMed: 24211208 , PubMed: 32144204 , PubMed: 39395789 , 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-ene) 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-ene) (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](#), PubMed:[39395789](#)). Catalyzes the hydrolysis of galactosylceramides/GalCers (such as beta-D-galactosyl-(11')-N- acylsphing-4-ene), 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](#)). Can also metabolize plant glycosyl phytosterols such as glucosylstigmasterol (PubMed:[39395789](#)).

Cellular Location

Lysosome membrane; Peripheral membrane protein; Luminal 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



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

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