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GBA Rabbit mAb

Catalog # AP77642

Product Information

Application	WB, IHC-P
Primary Accession	P04062
Reactivity	Rat, Human
Host	Rabbit
Clonality	Monoclonal Antibody
Isotype	IgG
Conjugate	Unconjugated
Purification	Affinity Chromatography
Calculated MW	59716

Additional Information

Gene ID	2629
Other Names	GBA
Dilution	WB~~1:1000 IHC-P~~N/A
Format	Liquid in 10mM PBS, pH 7.4, 150mM sodium chloride, 0.05% BSA, 0.02% sodium azide and 50% glycerol.
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.

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: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-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, PubMed:39395789). 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). Can also metabolize plant glycosyl phytosterols such as glucosylstigmasterol (PubMed:39395789).
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

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.

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