Product Name: GBA (1P9) Rabbit Monoclonal Antibody
Catalog #: AMRe11321

Summary

Production Name	GBA (1P9) Rabbit Monoclonal Antibody
Description	Rabbit Monoclonal Antibody
Host	Rabbit
Application	WB,ELISA
Reactivity	Human,Rat

Performance

Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Monoclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Rabbit IgG in phosphate buffered saline , pH 7.4, 150mM NaCl, 0.02% New type preservative N and 50% glycerol. Store at +4°C short term. Store at -20°C long term. Avoid freeze / thaw cycle.
Purification	Affinity purification

Immunogen

Gene Name	GBA
Alternative Names	Alglucerase; betaGC; GBA1; GCase; GCB; GLUC; Glucosylceramidase; Imiglucerase;
Gene ID	2629.0
SwissProt ID	P04062.

Application

Dilution Ratio	WB 1:500-1:2000
Molecular Weight	60kDa

Background

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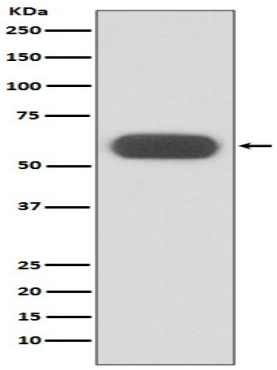
Defects in GBA are the cause of Gaucher disease (GD) [MIM:230800]; also known as glucocerebrosidase deficiency. GD is the most prevalent lysosomal storage disease, characterized by accumulation of glucosylceramide in the reticulo-endothelial system. Glucosylceramidase that catalyzes, within the lysosomal compartment, the hydrolysis of glucosylceramide/GlcCer into free ceramide and glucose (PubMed:9201993, PubMed:24211208, PubMed:15916907). Thereby, 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). Also plays a role in cholesterol metabolism (PubMed:24211208, PubMed:26724485). May either catalyze the glucosylation of cholesterol, through a transglucosylation reaction that transfers glucose from glucosylceramide to cholesterol (PubMed:24211208, PubMed:26724485). The short chain saturated C8:0-GlcCer and the mono-unsaturated C18:0-GlcCer being the most effective glucose donors for that transglucosylation reaction (PubMed:24211208). Under specific conditions, may alternatively catalyze the reverse reaction, transferring glucose from cholesteryl-beta-D-glucoside to ceramide (PubMed:26724485). Finally, may also hydrolyze cholesteryl- beta-D-glucoside to produce D-glucose and cholesterol (PubMed:24211208, PubMed:26724485).

Research Area

Image Data



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

Note

For research use only.