GBA Antibody – Mouse Monoclonal

PRODUCT OVERVIEW

Product Name	GBA, Monoclonal Antibody
Full Product Name	GBA (Glucosidase, beta; Acid (includes glucosylceramidase), GBA1, GCB, GLUC) (MaxLight 490)
Product Gene Name	anti-GBA antibody
Product Synonym Names	Glucosidase; beta; Acid (includes glucosylceramidase); GBA1; GCB; GLUC
Clonality	Monoclonal
Isotype	IgG1,k
Clone Number	2H4
Host	Mouse
Form/Format	Supplied as a liquid in PBS, pH 7.2. No preservative added. Labeled with MaxLight490.
Purity/Purification	Purified
Specificity	Recognizes GBA.
Preparation and Storage	Store product at 4 degree C in the dark. DO NOT FREEZE! Stable at 4 degree C for 12 months after receipt as an undiluted liquid. Dilute required amount only prior to immediate use. Further dilutions can be made in assay buffer. Caution: MaxLight490 conjugates are sensitive to light. For maximum recovery of product, centrifuge the original vial prior to removing the cap.
Molecular Weight	56.4kDa (503aa) 50-70kDa (SDS-PAGE under reducing conditions)
OMIM	168600
3D Structure	ModBase 3D Structure for P04062
Immunogen	GBA (NP_000148, 146aa-235aa) partial recombinant protein with GST tag. MW of the GST tag alone is 26kD.
Immunogen Sequence	SYFSEEGIGYNIIRVPMASCDFSIRTYTYADTPDDFQLHNFSLPEEDTKLKIPLIHRALQLAQRPVSLLASPWTSPTWLKTNGAVNGKGS
Conjugate	MaxLight490
Product Categories/Family for anti-GBA antibody	Antibodies; Enzymes
Applications Tested/Suitable for anti-GBA antibody	Western Blot (WB)
Application Notes for anti-GBA antibody	Applications are based on unconjugated antibody.
Related Product Information for anti-GBA antibody	This gene encodes a lysosomal membrane protein that cleaves the beta-glucosidic linkage of glycosylceramide, an intermediate in glycolipid metabolism. Mutations in this gene cause Gaucher disease, a lysosomal storage disease characterized by an accumulation of glucocerebrosides. A related pseudogene is approximately 12 kb downstream of this gene on chromosome 1. Alternative splicing results in multiple transcript variants encoding the same protein. [provided by RefSeq]
Other Notes	Small volumes of anti-GBA antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.

Searchable Terms for GBA purchase	MBS6206369 is an antibody targeting GBA. Also known as GBA (Glucosidase, beta; Acid (includes glucosylceramidase), GBA1, GCB, GLUC) (MaxLight 490). Host: Mouse.

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for GBA. It may not necessarily be applicable to this product.

NCBI GI #	54607043
NCBI GeneID	2629
NCBI Accession #	NP_000148
UniProt Primary Accession #	P04062
UniProt Secondary Accession #	Q16545; Q4VX22; Q6I9R6; Q9UMJ8; A8K796; B7Z5G2; B7Z6S1; J3KQG4; J3KQK9
UniProt Related Accession #	P04062
GenBank Nucleotide #	NM_000157.3
NCBI Official Full Name	glucosylceramidase isoform 1
NCBI Official Synonym Full Names	glucosylceramidase beta
NCBI Official Symbol	GBA
NCBI Official Synonym Symbols	GCB; GBA1; GLUC
UniProt Protein Name	Glucosylceramidase
UniProt Synonym Protein Names	Acid beta-glucosidase; Alglucerase; Beta-glucocerebrosidase; Beta-GC; D-glucosyl-N-acylsphingosine glucohydrolase; Imiglucerase
Protein Family	Glucosylceramidase
UniProt Gene Name	GBA
UniProt Synonym Gene Names	GC; GLUC; Beta-GC
UniProt Entry Name	GLCM_HUMAN
NCBI Protein Information	glucosylceramidase
NCBI Summary for GBA	This gene encodes a lysosomal membrane protein that cleaves the beta-glucosidic linkage of glycosylceramide, an intermediate in glycolipid metabolism. Mutations in this gene cause Gaucher disease, a lysosomal storage disease characterized by an accumulation of glucocerebrosides. A related pseudogene is approximately 12 kb downstream of this gene on chromosome 1. Alternative splicing results in multiple transcript variants. [provided by RefSeq, Jan 2010]

UniProt Comments for GBA	GBA: Defects in GBA are the cause of Gaucher disease (GD); also known as glucocerebrosidase deficiency. GD is the most prevalent lysosomal storage disease, characterized by accumulation of glucosylceramide in the reticulo-endothelial system. Different clinical forms are recognized depending on the presence (neuronopathic forms) or absence of central nervous system involvement, severity and age of onset. Defects in GBA are the cause of Gaucher disease type 1 (GD1); also known as adult non-neuronopathic Gaucher disease. GD1 is characterized by hepatosplenomegaly with consequent anemia and thrombopenia, and bone involvement. The central nervous system is not involved. Defects in GBA are the cause of Gaucher disease type 2 (GD2); also known as acute neuronopathic Gaucher disease. GD2 is the most severe form and is universally progressive and fatal. It manifests soon after birth, with death generally occurring before patients reach two years of age. Defects in GBA are the cause of Gaucher disease type 3 (GD3); also known as subacute neuronopathic Gaucher disease. GD3 has central nervous manifestations. Defects in GBA are the cause of Gaucher disease type 3C (GD3C); also known as pseudo-Gaucher disease or Gaucher-like disease. Defects in GBA are the cause of Gaucher disease perinatal lethal (GDPL). It is a distinct form of Gaucher disease type 2, characterized by fetal onset. Hydrops fetalis, in utero fetal death and neonatal distress are prominent features. When hydrops is absent, neurologic involvement begins in the first week and leads to death within 3 months. Hepatosplenomegaly is a major sign, and is associated with ichthyosis, arthrogryposis, and facial dysmorphism. Perinatal lethal Gaucher disease is associated with non-immune hydrops fetalis, a generalized edema of the fetus with fluid accumulation in the body cavities due to non-immune causes. Non-immune hydrops fetalis is not a diagnosis in itself but a symptom, a feature of many genetic disorders, and the end-stage of a wide variety of disorders. Defects in GBA contribute to susceptibility to Parkinson disease (PARK). A complex neurodegenerative disorder characterized by bradykinesia, resting tremor, muscular rigidity and postural instability. Additional features are characteristic postural abnormalities, dysautonomia, dystonic cramps, and dementia. The pathology of Parkinson disease involves the loss of dopaminergic neurons in the substantia nigra and the presence of Lewy bodies (intraneuronal accumulations of aggregated proteins), in surviving neurons in various areas of the brain. The disease is progressive and usually manifests after the age of 50 years, although early-onset cases (before 50 years) are known. The majority of the cases are sporadic suggesting a multifactorial etiology based on environmental and genetic factors. However, some patients present with a positive family history for the disease. Familial forms of the disease usually begin at earlier ages and are associated with atypical clinical features. Belongs to the glycosyl hydrolase 30 family. 3 isoforms of the human protein are produced by alternative splicing. Protein type: EC 3.2.1.45; Glycan Metabolism - other glycan degradation; Hydrolase; Lipid Metabolism - sphingolipid Chromosomal Location of Human Ortholog: 1q21 Cellular Component: lysosomal lumen; lysosomal membrane Molecular Function: glucosylceramidase activity; protein binding; receptor binding Biological Process: ceramide biosynthetic process; glucosylceramide catabolic process; glycosphingolipid metabolic process; mitochondrion organization and biogenesis; negative regulation of inflammatory response; negative regulation of interleukin-6 production; negative regulation of MAP kinase activity; negative regulation of protein homooligomerization; neurite development; positive regulation of protein amino acid dephosphorylation; positive regulation of protein complex disassembly; positive regulation of protein metabolic process; regulation of cellular protein metabolic process; regulation of macroautophagy; sphingosine biosynthetic process Disease: Dementia, Lewy Body; Gaucher Disease, Perinatal Lethal; Gaucher Disease, Type I; Gaucher Disease, Type Ii; Gaucher Disease, Type Iii; Gaucher Disease, Type Iiic; Parkinson Disease, Late-onset
Research Articles on GBA	**1.** This study shown GBA genetic variants for Parkinson's disease are associated with the risk of incident Parkinson's disease in the general population and with impairment in daily functioning in individuals without clinical parkinsonism.
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