

GBA Antibody (monoclonal) (M01)

Mouse monoclonal antibody raised against a partial recombinant GBA.

Catalog # AT2167a

Product Information

Application	WB, IHC, IF, E
Primary Accession	P04062
Other Accession	NM_000157
Reactivity	Human
Host	Mouse
Clonality	monoclonal
Isotype	IgG2a Kappa
Clone Names	2E3
Calculated MW	59716

Additional Information

Gene ID	2629
Other Names	Glucosylceramidase, Acid beta-glucosidase, Alglucerase, Beta-glucocerebrosidase, Beta-GC, D-glucosyl-N-acylsphingosine glucosylhydrolase, Imiglucerase, GBA, GC, GLUC
Target/Specificity	GBA (NP_000148, 146 a.a. ~ 235 a.a) partial recombinant protein with GST tag. MW of the GST tag alone is 26 KDa.
Dilution	WB~~1:500~1000 IHC~~1:100~500 IF~~1:50~200 E~~N/A
Format	Clear, colorless solution in phosphate buffered saline, pH 7.2 .
Storage	Store at -20°C or lower. Aliquot to avoid repeated freezing and thawing.
Precautions	GBA Antibody (monoclonal) (M01) is for research use only and not for use in diagnostic or therapeutic procedures.

Background

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.

References

1. Parkin-mediated ubiquitination of mutant glucocerebrosidase leads to competition with its substrates PARIS and ARTS. Bendikov-Bar I, Rapaport D, Larisch S, Horowitz M, Orphanet J Rare Dis. 2014 Jun 16;9:86. doi: 10.1186/1750-1172-9-86.

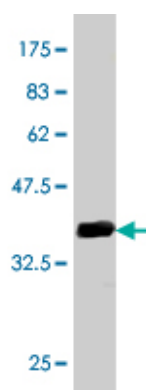
2. Reduced glucocerebrosidase is associated with increased α -synuclein in sporadic Parkinson's disease. Murphy KE, Gysbers AM, Abbott SK, Tayebi N, Kim WS, Sidransky E, Cooper A, Garner B, Halliday GM. Brain. 2014 Jan 28.

3. Gaucher disease paradigm: From ERAD to comorbidity. Bendikov-Bar I, Horowitz M. Hum Mutat. 2012 Oct;33(10):1398-407. doi: 10.1002/humu.22124. Epub 2012 Jun 11.

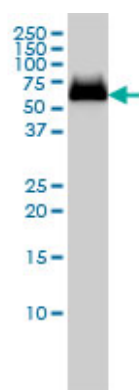
4. Characterization of the ERAD process of the L444P mutant glucocerebrosidase variant. Bendikov-Bar I, Ron I, Filocamo M, Horowitz M. Blood Cells Mol Dis. 2010 Nov 22. [Epub ahead of print]

5. Characterization of Gaucher disease bone marrow mesenchymal stromal cells reveals an altered inflammatory secretome. Campeau PM, Rafei M, Boivin MN, Sun Y, Grabowski GA, Galipeau J. Blood. 2009 Oct 8;114(15):3181-90. Epub 2009 Jul 8.

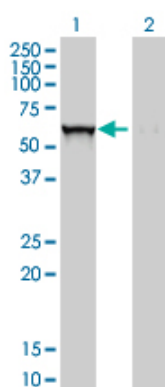
Images



Antibody Reactive Against Recombinant Protein. Western Blot detection against Immunogen (35.64 kDa) .



GBA monoclonal antibody (M01), clone 2E2 Western Blot analysis of GBA expression in MCF-7 ((Cat # AT2167a)

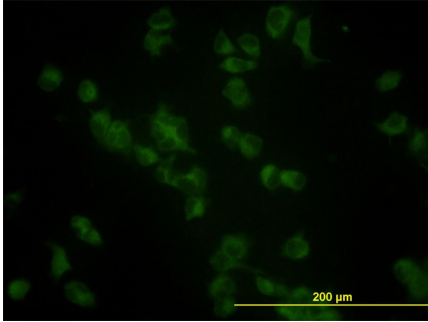
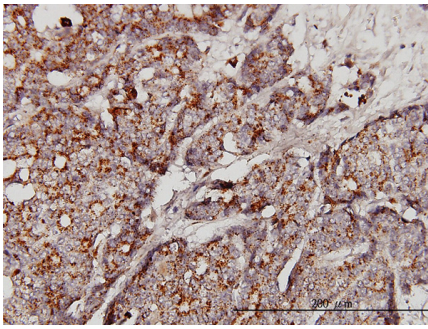


Western Blot analysis of GBA expression in transfected 293T cell line by GBA monoclonal antibody (M01), clone 2E2.

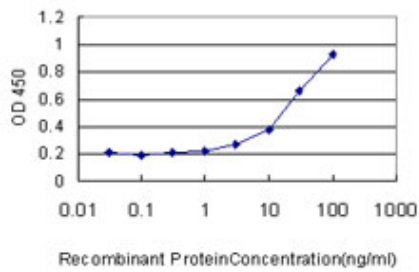
Lane 1: GBA transfected lysate (60 kDa).

Lane 2: Non-transfected lysate.

Immunoperoxidase of monoclonal antibody to GBA on formalin-fixed paraffin-embedded human breast cancer. [antibody concentration 3 μ g/ml]



Immunofluorescence of monoclonal antibody to GBA on HeLa cell. [antibody concentration 10 ug/ml]



Detection limit for recombinant GST tagged GBA is approximately 1ng/ml as a capture antibody.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.