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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 glucohydrolase, 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.

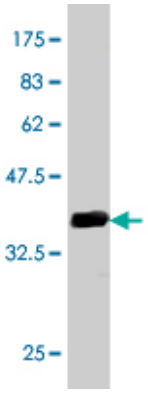
Background

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.

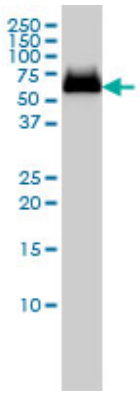
References

1.Parkin-mediated ubiquitination of mutant glucocerebrosidase leads to competition with its substrates PARIS and ARTS.Bendikov-Bar I, Rapaport D, Larisch S, Horowitz MOrphanet 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 GMBrain. 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.

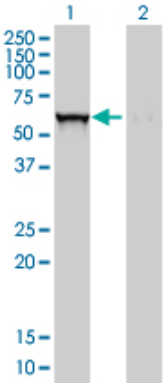
Images



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



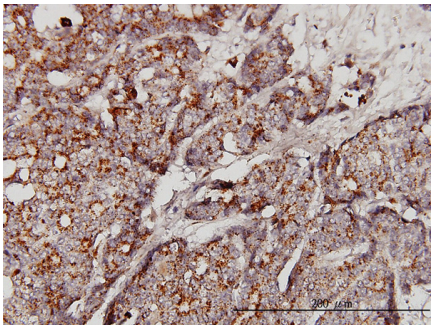
GBA monoclonal antibody (M01), clone 2E2 Western Blot analysis of GBA expression in MCF-7 ((Cat # AT2167a)

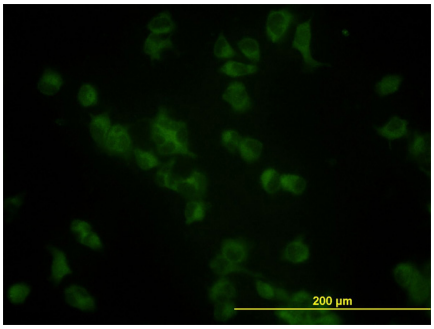


Western Blot analysis of GBA expression in transfected 293T cell line by GBA monoclonal antibody (M01), clone 2E2.

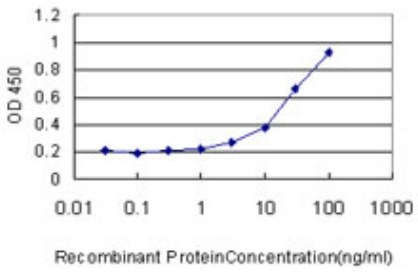
Lane 1: GBA transfected lysate(60 KDa).
Lane 2: Non-transfected lysate.

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