Human Glucosylceramidase/GBA Antibody

Monoclonal Mouse IgG_1 Clone # 812201

Catalog Number: MAB7410

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human Glucosylceramidase/GBA in direct ELISAs. In direct ELISAs, no cross-reactivity with recombinant human Cytosolic beta-Glucosidase/GBA3 is observed.
Source	Monoclonal Mouse IgG_1 Clone # 812201
Purification	Protein A or G purified from hybridoma culture supernatant
Immunogen	Chinese hamster ovary cell line CHO-derived recombinant human Glucosylceramidase/GBA Met1-Gln536 Accession # P04062
Formulation	Lyophilized from a 0.2 μm filtered solution in PBS with Trehalose. See Certificate of Analysis for details. *Small pack size (-SP) is supplied as a 0.2 μm filtered solution in PBS.

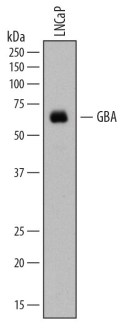
APPLICATIONS

Please Note: *Optimal dilutions should be determined by each laboratory for each application. General Protocols are available in the Technical Information section on our website.*

	Recommended Concentration	Sample
Western Blot	1 µg/mL	See Below
Immunocytochemistry	8-25 µg/mL	See Below
Simple Western	10 µg/mL	See Below

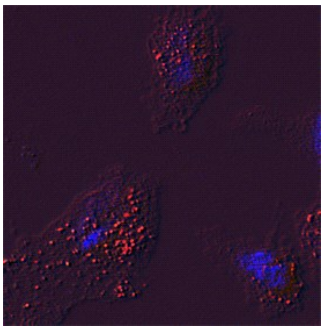
DATA

Western Blot



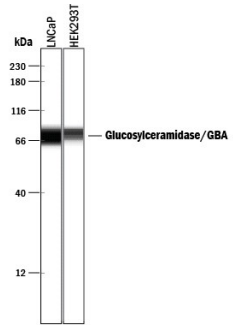
Detection of Human Glucosylceramidase/GBA by Western Blot. Western blot shows lysates of LNCaP human prostate cancer cell line. PVDF membrane was probed with 1 µg/mL of Mouse Anti-Human Glucosylceramidase/GBA Monoclonal Antibody (Catalog # MAB7410) followed by HRP-conjugated Anti-Mouse IgG Secondary Antibody (Catalog # HAF007). A specific band was detected for Glucosylceramidase/GBA at approximately 60 kDa (as indicated). This experiment was conducted under reducing conditions and using Immunoblot Buffer Group 1.

Immunocytochemistry



Glucosylceramidase/GBA in HeLa Human Cell Line. Glucosylceramidase/GBA was detected in immersion fixed HeLa human cervical epithelial carcinoma cell line using Mouse Anti-Human Glucosylceramidase/GBA Monoclonal Antibody (Catalog # MAB7410) at 15 µg/mL for 3 hours at room temperature. Cells were stained using the NorthernLights™ 557-conjugated Anti-Mouse IgG Secondary Antibody (red; Catalog # NL007) and counterstained with DAPI (blue). Specific staining was localized to lysosomes. View our protocol for Fluorescent ICC Staining of Cells on Coverslips.

Simple Western



Detection of Human Glucosylceramidase/GBA by Simple Western™. Simple Western lane view shows lysates of LNCaP human prostate cancer cell line and HEK293T human embryonic kidney cell line, loaded at 0.2 mg/mL. A specific band was detected for Glucosylceramidase/GBA at approximately 77 kDa (as indicated) using 10 µg/mL of Mouse Anti-Human Glucosylceramidase/GBA Monoclonal Antibody (Catalog # MAB7410). This experiment was conducted under reducing conditions and using the 12-230 kDa separation system.

PREPARATION AND STORAGE

Reconstitution	Sterile PBS to a final concentration of 0.5 mg/mL.
Shipping	The product is shipped at ambient temperature. Upon receipt, store it immediately at the temperature recommended below. *Small pack size (-SP) is shipped with polar packs. Upon receipt, store it immediately at -20 to -70 °C
Stability & Storage	**Use a manual defrost freezer and avoid repeated freeze-thaw cycles.** ● 12 months from date of receipt, -20 to -70 °C as supplied. ● 1 month, 2 to 8 °C under sterile conditions after reconstitution. ● 6 months, -20 to -70 °C under sterile conditions after reconstitution.

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BACKGROUND

Glucosylceramidase is a lysosomal enzyme that cleaves the beta-glucosidic linkage of glucosylceramide (1, 2), an intermediate in glycolipid metabolism. The mature enzyme has 497 amino acids with a molecular weight of 62 kDa (3). Glycosylation occurs at four of five N-glycosylation sites and is essential for the trafficking and activity of the enzyme (4). The enzyme is activated in lysosomes by saposin C, although the mechanism of activation is not well understood (5). Defects in Glucosylceramidase are the cause of Gaucher disease, also known as glucocerebrosidase deficiency (6). Gaucher disease is the most prevalent lysosomal storage disease, characterized by accumulation of glucosylceramide in the reticulo-endothelial system. Symptoms of Gaucher disease may include enlarged spleen and liver, liver malfunction, skeletal disorders and bone lesions, severe neurologic complications, swelling of lymph nodes, anemia, low blood platelets and yellow fatty deposits on the white of the eye (7). Currently, enzyme replacement therapy is used to treat patients with the disease (8, 9).

References:

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