Human Glucosylceramidase/GBA
Alexa Fluor® 488-conjugated Antibody

Monoclonal Mouse IgG_1 Clone # 812201

Catalog Number: FAB7410G

100 µg

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
Conjugate	Alexa Fluor 488 Excitation Wavelength: 488 nm Emission Wavelength: 515-545 nm
Formulation	Supplied 0.2mg/ml in 1X PBS with RDF1 and 0.09% Sodium Azide *Contains <0.1% Sodium Azide, which is not hazardous at this concentration according to GHS classifications. Refer to the Safety Data Sheet (SDS) for additional information and handling instructions.

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.*

Knockout Validated	Optimal dilution of this antibody should be experimentally determined.
Western Blot	Optimal dilution of this antibody should be experimentally determined.
Immunocytochemistry	Optimal dilution of this antibody should be experimentally determined.

PREPARATION AND STORAGE

Shipping	The product is shipped with polar packs. Upon receipt, store it immediately at the temperature recommended below.
Stability & Storage	Protect from light. Do not freeze. 12 months from date of receipt, 2 to 8 °C as supplied

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).

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