Human Hexosaminidase A/HEXA Antibody

Monoclonal Mouse IgG_{2B} Clone # 714712

Catalog Number: MAB62371

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human Hexosaminidase A/HEXA in direct ELISAs.
Source	Monoclonal Mouse IgG_{2B} Clone # 714712
Purification	Protein A or G purified from hybridoma culture supernatant
Immunogen	*S. frugiperda* insect ovarian cell line *Sf* 21-derived recombinant human Hexosaminidase A/HEXA Met1-Thr529 Accession # P06865
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.

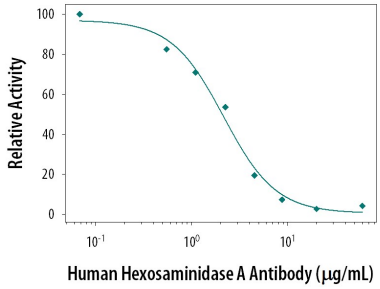
APPLICATIONS

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.*

Neutralization	Measured by its ability to neutralize Human Hexosaminidase A (2.0 μg/mL,Catalog # 6237-GH) cleavage of the fluorogenic peptide substrate 4-Methylumbelliferyl-N-Acetyl-β-D-glucosaminide (400 μM). The Neutralization Dose (ND_{50}) is typically 3.0 μg/mL.

DATA

Neutralization



Neutralization of Human Hexosaminidase A Activity by Human Hexosaminidase A Antibody. The cleavage of 4-Methylumbelliferyl-N-Acetyl-β-D-glucosaminide (400 μM) by Human Hexosaminidase A (2.0 μg/mL,Catalog # 6237-GH) is measured after it has been preincubated with increasing concentrations of Mouse Anti-Human Hexosaminidase A Monoclonal Antibody (Catalog # MAB62371). The ND50 is typically 3.0 μg/mL.

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.

BACKGROUND

β-hexosaminidases are enzymes involved in the hydrolysis of terminal N-acetyl-D-hexosamine residues in GM2 gangliosides and globo-sphingolipids in lysosomes (1-4). The enzymes are composed of two α and/or β subunits, which are coded by HEXA and HEXB genes, respectively. Different association of the α and β subunits gives rise to β-hexosaminidase isoforms A, B and S (Hex A, B and S) (5), which have the composition of αβ, ββ and αα, respectively. Hex S is suggested to releases non-reducing end N-acetylgalactosamine residues from dermatan sulfate, chondroitin sulfate and sulfated glycolipid SM2 (6). Recombinant HEXA is also highly active on 4-methylumbelliferyl-N-acetyl-β-D-glucosaminide (6). Mutations in HEXA and HEXB genes cause lysosomal lipid storage disorders. Specifically, mutations of HEXA cause Tay-Sachs disease, manifested by the harmful accumulation of ganglioside GM2 in tissues and nerve cells in the brain (7-10). Children with this disease usually die by age 4.

References:

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