Human Hexosaminidase A/HEXA Alexa Fluor® 532-conjugated Antibody

Monoclonal Mouse IgG_{2B} Clone # 714729

Catalog Number: FAB6237X

100 µg

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human Hexosaminidase A/HEXA in direct ELISAs and Western blots.
Source	Monoclonal Mouse IgG_{2B} Clone # 714729
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
Conjugate	Alexa Fluor 532 Excitation Wavelength: 534 nm Emission Wavelength: 553 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.*

Western Blot	Optimal dilution of this antibody should be experimentally determined.
Immunohistochemistry	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

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. Our recombinant HEXA is presumably isoform Hex S, because only α subunit was expressed. 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.

PRODUCT SPECIFIC NOTICES

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