BOSTER BIOLOGICAL TECHNOLOGY Co.,Ltd.
3942 B Valley Ave, Pleasanton, CA, 94566
Phone: 888-466-3604 Fax: 925-215-2184 Email: boster@bosterbio.com Web: www.bosterbio.com

Polyclonal Anti-HEXA Antibody

Catalog Number: PA1787-2

Description

Gene Name	hexosaminidase A (alpha polypeptide)
Recommended Protein Name	Beta-hexosaminidase subunit alpha
Lot No.	0171312c0487111
Size	100μg/vial
Form	lyophilized
Ig type	Rabbit IgG
Specificity	No cross reactivity with other proteins.
Purification	Immunogen affinity purified.
Species	**Reacts with:** human
Immunogen	A synthetic peptide corresponding to a sequence at the C-terminus of human HEXA(513-529aa QAQPLNVGFCEQEFEQT), different from the related mouse sequence by three amino acids, and from the related rat sequences by four amino acids.
Contents	Each vial contains 5mg BSA, 0.9mg NaCl, 0.2mg Na_2HPO_4, 0.05mg Thimerosal, 0.05mg NaN_3.

Application

	Concentration	Tested Species	Predicted Species	Antigen Retrieval
Western blot	0.1-0.5μg/ml	Hu	-	-

Tested Species: In-house tested species with positive results.

Predicted Species: Species predicted to be fit for the product based on sequence similarities.

Other applications have not been tested.

Optimal dilutions should be determined by end users.

Preparation and storage

Reconstitution: 0.2ml of distilled water will yield a concentration of 500μg/ml.

Storage: At -20˚C for one year. After reconstitution, at 4˚C for one month. It can also be aliquotted and stored frozen at -20˚C for a longer time.
Avoid repeated freezing and thawing.

Relevant detection systems

Boster provides a series of assays reacted with primary antibodies. Antibody can be supported by chemiluminescence kit EK1002 in WB.

Background

HEXA (hexosaminidase A (alpha polypeptide)) is an enzyme that in humans is encoded by the HEXA gene. Hexosaminidase A and the cofactor GM2 activator protein catalyze the degradation of the GM2 gangliosides and other molecules containing terminal N-acetyl hexosamines The HEXA gene encodes the alpha subunit of hexosaminidase A , a lysosomal enzyme involved in the breakdown of gangliosides. The HEXA gene is mapped on 15q23. Even though the alpha and beta subunits of hexosaminidase A can both cleave GalNAc residues, only the alpha subunit is able to hydrolyze GM2 gangliosides. The alpha subunit contains a key residue, Arg-424, which is essential for binding the N-acetyl-neuramanic residue of GM2 gangliosides. Chimeric constructs were expressed in HeLa cells and selected constructs were produced in the baculovirus expression system to determine their ability to degrade GM2 ganglioside in the presence of GM2 activator protein. Their results allowed them to define 2 noncontiguous sequences in the alpha subunit (amino acids 1-191 and 403-529) which, when substituted into analogous positions in the beta subunit, conferred activity against the sulfated substrate.

Reference

1. Akli, S., Chomel, J.-C., Lacorte, J.-M., Bachner, L., Poenaru, A., Poenaru, L. Ten novel mutations in the HEXA gene in non-Jewish Tay-Sachs patients. Hum. Molec. Genet. 2: 61-67, 1993.
2. Beutler, E., Kuhl, W., Comings, D. Hexosaminidase isozyme in type O Gm2 gangliosidosis (Sandhoff-Jatzkewitz disease). Am. J. Hum. Genet. 27: 628-638, 1975.
3. Chern, C. J., Beutler, E., Kuhl, W., Gilbert, F., Mellman, W. J., Croce, C. M. Characterization of heteropolymeric hexosaminidase A in human x mouse hybrid cells. Proc. Nat. Acad. Sci. 73: 3637-3640, 1976.