Product Name: HEXA Rabbit Polyclonal Antibody
Catalog #: APRab11998

Summary

Production Name	HEXA Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	WB,IHC,ELISA
Reactivity	Human,Mouse,Rat

Performance

Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Purification	Affinity purification

Immunogen

Gene Name	HEXA
Alternative Names	HEXA; Beta-hexosaminidase subunit alpha; Beta-N-acetylhexosaminidase subunit alpha; Hexosaminidase subunit A; N-acetyl-beta-glucosaminidase subunit alpha
Gene ID	3073.0
SwissProt ID	P06865.Synthesized peptide derived from HEXA . at AA range: 121-170

Application

Dilution Ratio	WB 1:500 - 1:2000. IHC: 1:100-300 ELISA: 1:20000..
Molecular Weight	60kD

Background

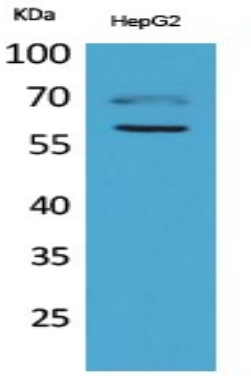
Product Name: HEXA Rabbit Polyclonal Antibody
Catalog #: APRab11998

This gene encodes a member of the glycosyl hydrolase 20 family of proteins. The encoded preproprotein is proteolytically processed to generate the alpha subunit of the lysosomal enzyme beta-hexosaminidase. This enzyme, together with the cofactor GM2 activator protein, catalyzes the degradation of the ganglioside GM2, and other molecules containing terminal N-acetyl hexosamines. Mutations in this gene lead to an accumulation of GM2 ganglioside in neurons, the underlying cause of neurodegenerative disorders termed the GM2 gangliosidoses, including Tay-Sachs disease (GM2-gangliosidosis type I). Alternative splicing results in multiple transcript variants, at least one of which encodes a preproprotein that is proteolytically processed. [provided by RefSeq, Jan 2016],catalytic activity:Hydrolysis of terminal non-reducing N-acetyl-D-hexosamine residues in N-acetyl-beta-D-hexosaminides.,disease:Defects in HEXA are the cause of GM2-gangliosidosis type 1 (GM2G1) [MIM:272800]; also known as Tay-Sachs disease. GM2-gangliosidosis is an autosomal recessive lysosomal storage disease marked by the accumulation of GM2 gangliosides in the neuronal cells. GM2G1 is characterized by GM2 gangliosides accumulation in the absence of HEXA activity, leading to neurodegeneration and, in the infantile form, death in early childhood. GM2G1 has an increased incidence among Ashkenazi Jews and French Canadians in eastern Quebec. It exists in several forms: infantile (most common and most severe), juvenile and adult (late onset).,function:Responsible for the degradation of GM2 gangliosides, and a variety of other molecules containing terminal N-acetyl hexosamines, in the brain and other tissues. The form B is active against certain oligosaccharides. The form S has no measurable activity.,online information:HEXA mutation database,online information:Tay Sachs disease website,PTM:N-linked glycan at Asn-115 consists of Man(3)-GlcNAc(2).,similarity:Belongs to the glycosyl hydrolase 20 family.,subunit:There are 3 forms of beta-hexosaminidase: hexosaminidase A is a trimer composed of one subunit alpha, one subunit beta chain A and one subunit beta chain B; hexosaminidase B is a tetramer of two subunit beta chains A and two subunit beta chains B; hexosaminidase S is an homodimer of two alpha subunits. The two beta chains are derived from the cleavage of the beta subunit.,

Research Area

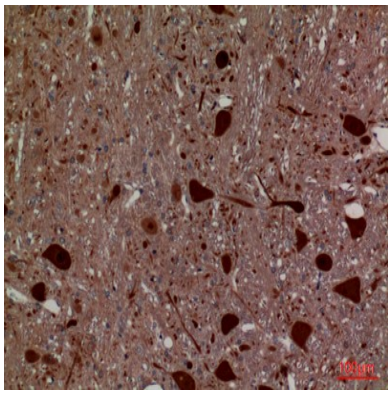
Other glycan degradation;Amino sugar and nucleotide sugar metabolism;Glycosaminoglycan degradation;Glycosphingolipid biosynthesis;Glycosphingolipid biosynthesis;Lysosome;

Image Data

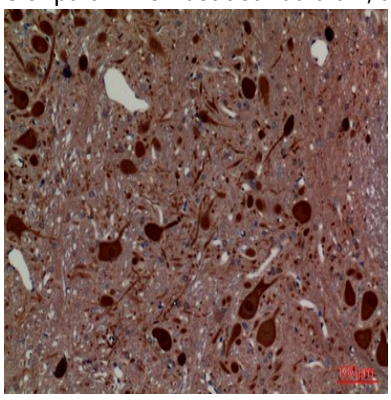


Western Blot analysis of HepG2 cells using HEXA Polyclonal Antibody. Antibody was diluted at 1:1000. Secondary antibody

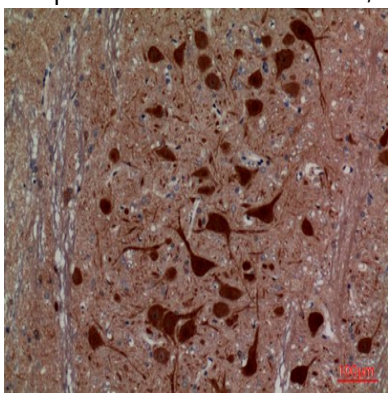
was diluted at 1:20000



Immunohistochemical analysis of paraffin-embedded rat-brain, antibody was diluted at 1:100

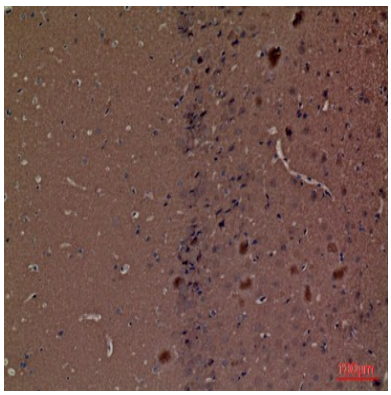


Immunohistochemical analysis of paraffin-embedded rat-brain, antibody was diluted at 1:100



Immunohistochemical analysis of paraffin-embedded rat-brain, antibody was diluted at 1:100

Product Name: HEXA Rabbit Polyclonal Antibody
Catalog #: APRab11998



Immunohistochemical analysis of paraffin-embedded mouse-brain, antibody was diluted at 1:100

Note

For research use only.