



HEXA Monoclonal Antibody

alpha 60kD Cell Pathway Lysosome. Tissue Specificity Brain,Eye,Liver,Ovary,Uterus, 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 neurona cells. GM2G1 is characterized by GM2 gangliosides accumulation in the absence of HEXA activity, leading to neurodegeneration and, in the infantile form, death in		
Reactivity Human;Mouse;Rat Applications WB Gene Name HEXA Protein Name Beta-hexosaminidase subunit alpha Immunogen Synthesized peptide derived from HEXA . at AA range: 121-170 Specificity HEXA Monoclonal Antibody detects endogenous levels of HEXA protein. Formulation Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide. Source Monoclonal, Mouse, IgG Purification The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen. Dilution WB 1:500-2000 Concentration 1 mg/ml Purity ≥90% Storage Stability -20°C/1 year Synonyms HEXA; Beta-hexosaminidase subunit alpha; Beta-N-acetylhexosaminidase subunit alpha; Hexosaminidase subunit alpha; Hexosaminidase subunit alpha Coll Pathway Lysosome. Tissue Specificity Brain, Eye, Liver, Ovary, Uterus, 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 GMZ-gangilosidosis type 1 (GMZ-gangilosidosis type 1	Catalog No	BYmab-02864
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for the degradation of GM2 gangliosides, and a variety of other molecules containing terminal N-acetyl hexosamines, in the brain

Background

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],

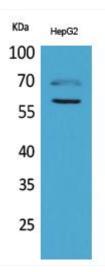
matters needing attention

Avoid repeated freezing and thawing!

Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western Blot analysis of various cells using HEXA Monoclonal Antibody

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