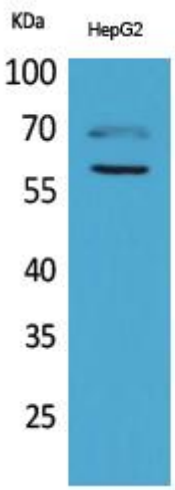
HEXA Monoclonal Antibody

Catalog No	BYmab-02864
Isotype	IgG
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 A; N-acetyl-beta-glucosaminidase subunit alpha
Observed Band	60kD
Cell Pathway	Lysosome.
Tissue Specificity	Brain,Eye,Liver,Ovary,Uterus,
Function	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
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