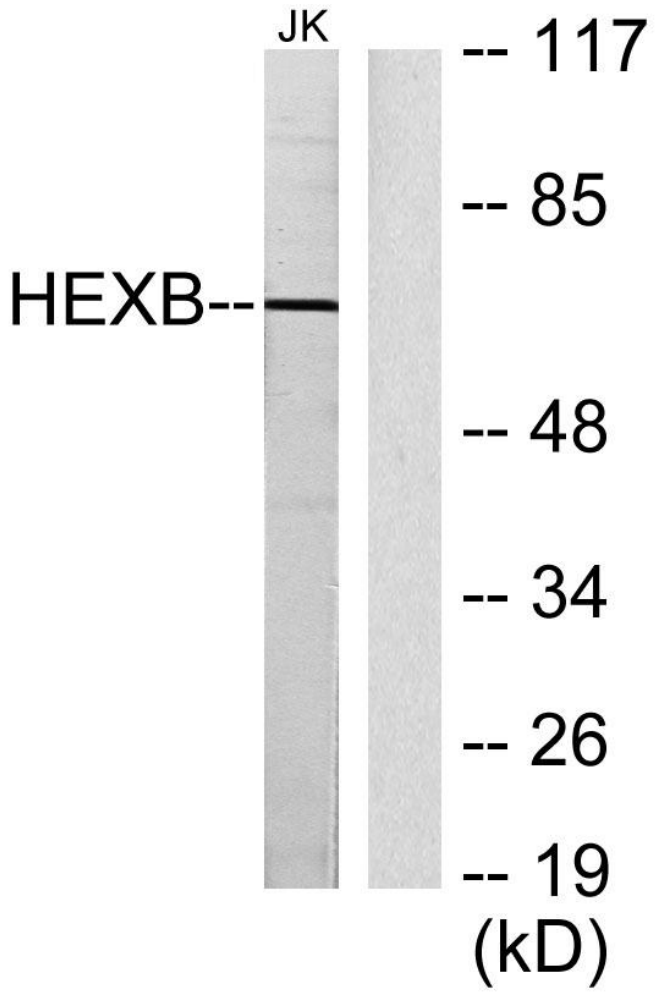
Hexb Monoclonal Antibody

Catalog No	YP-mAb-02653
Isotype	IgG
Reactivity	Human;Rat;Mouse;
Applications	WB
Gene Name	HEXB
Protein Name	Beta-hexosaminidase subunit beta
Immunogen	The antiserum was produced against synthesized peptide derived from human HEXB. AA range:481-530
Specificity	Hexb Monoclonal Antibody detects endogenous levels of Hexb 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 1:500-1:2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	HEXB; HCC7; Beta-hexosaminidase subunit beta; Beta-N-acetylhexosaminidase subunit beta; Hexosaminidase subunit B; Cervical cancer proto-oncogene 7 protein; HCC-7; N-acetyl-beta-glucosaminidase subunit beta
Observed Band	63kD
Cell Pathway	Lysosome . Cytoplasmic vesicle, secretory vesicle, Cortical granule .
Tissue Specificity	Liver,Skin,
Function	catalytic activity:Hydrolysis of terminal non-reducing N-acetyl-D-hexosamine residues in N-acetyl-beta-D-hexosaminides.,disease:Defects in HEXB are the cause of GM2-gangliosidosis type 2 (GM2G2) [MIM:268800]; also known as Sandhoff disease. GM2-gangliosidosis is an autosomal recessive lysosomal storage disease marked by the accumulation of GM2 gangliosides in the neuronal cells. GM2G2 is clinically indistinguishable from GM2-gangliosidosis type 1, presenting startle reactions, early blindness, progressive motor and mental deterioration, macrocephaly and cherry-red spots on the macula.,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.,online information:HEXB mutation database,PTM:N-linked glycans at Asn-142 and Asn-190 consist of Man(3)-GlcNAc(2) and Man(5 to 7)-GlcNAc(2)

Background	Hexosaminidase B is the beta subunit of the lysosomal enzyme beta-hexosaminidase that, together with the cofactor GM2 activator protein, catalyzes the degradation of the ganglioside GM2, and other molecules containing terminal N-acetyl hexosamines. Beta-hexosaminidase is composed of two subunits, alpha and beta, which are encoded by separate genes. Both beta-hexosaminidase alpha and beta subunits are members of family 20 of glycosyl hydrolases. Mutations in the alpha or beta subunit genes lead to an accumulation of GM2 ganglioside in neurons and neurodegenerative disorders termed the GM2 gangliosidoses. Beta subunit gene mutations lead to Sandhoff disease (GM2-gangliosidosis type II). Alternatively spliced transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq, May 2014],
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 Hexb Monoclonal Antibody