Product Data Sheet:
Hexb Rabbit Polyclonal Antibody

Cat. No:	ABN11999
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human HEXB. AA range:481-530
Reactivity:	Human,Rat,Mouse
Applications:	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:200-1:1000,ELISA 1:10000-1:20000
Molecular Weight:	63kDa
Purification:	Affinity purification
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
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],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), respectively.,PTM:The beta-A and beta-B chains are produced by proteolytic processing of the precursor beta chain.,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.,
Form:	Liquid

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Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.

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