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Rabbit Anti-HEXB chain A antibody

SL10463R

Product Name:	HEXB chain A
Chinese Name:	Beta氨基己糖苷酶beta亚基蛋白A链抗体
Alias:	Beta hexosaminidase beta chain; Beta hexosaminidase subunit beta; Beta N acetylhexosaminidase; Beta-hexosaminidase subunit beta chain A; Beta-N-acetylhexosaminidase subunit beta; Cervical cancer proto oncogene 7 protein; Cervical cancer proto-oncogene 7 protein; ENC 1AS; HCC 7; HCC-7; HCC7; HEX B; Hexb; HEXB_HUMAN; Hexosaminidase B (beta polypeptide); Hexosaminidase B; Hexosaminidase subunit B; HexosaminidaseB; N acetyl beta glucosaminidase; N-acetyl-beta-glucosaminidase subunit beta; HEXB chain A.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Pig,Cow,Horse,Rabbit,Sheep,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	28/50kDa
Cellular localization:	The nucleuscytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human HEXB chain A:451-556/556
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	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). [provided by RefSeq, Jul 2008].

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.

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 a homodimer of two alpha subunits. The two beta chains are derived from the cleavage of the beta subunit.

Subcellular Location:
Lysosome.

DISEASE:
GM2-gangliosidosis 2 (GM2G2) [MIM:268800]: An autosomal recessive lysosomal storage disease marked by the accumulation of GM2 gangliosides in the neuronal cells. 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. Note=The disease is caused by mutations affecting the gene represented in this entry.

Similarity:
Belongs to the glycosyl hydrolase 20 family.

SWISS:
P07686

Gene ID:
3074

Database links:

Entrez Gene: 3074Human

GenBank: NP_000512.1Human

Omim: 606873Human

	SwissProt: P07686Human Unigene: 69293Human **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.