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[Primary Antibody]

Rabbit Anti-HEXB chain B antibody

Catalog Number: SL15468R

FOR RESEARCH USE ONLY

[DATASHEET]

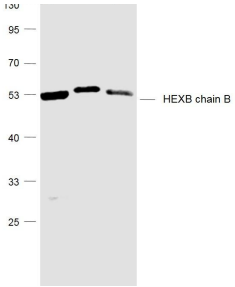
Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	HEXB chain B
Immunogen:	KLH conjugated synthetic peptide derived from human HEXB: 201-300/556
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Applications:	WB=1:500-2000 （Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Reactivity:	Human,Mouse(predicted:Rat,Dog,Cow,Sheep)
Predicted MW.:	22/50kDa
Subcellular Location:	cytoplasmic
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). [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

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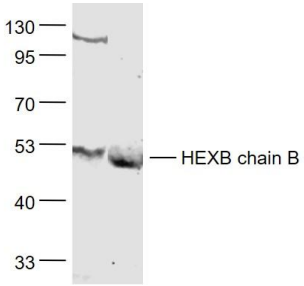
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	subunit beta chain B; hexosaminidase B is a tetramer of two subunit beta chains A and two subunit beta chains B; hexosaminidase S is a homodimer of two alpha subunits. The two beta chains are derived from the cleavage of the beta subunit. **Subcellular 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. **Database links:** **Entrez Gene:** 3074 Human GenBank: NP_000512.1 Human Omim: 606873 Human **SwissProt:** P07686 Human **Unigene:** 69293 Human

[VALIDATION IMAGES]



Sample:
Siha (Human) Cell Lysate at 30 ug
Bone (Mouse) Lysate at 40 ug
Lymph node (Mouse) Lysate at 40 ug
Primary: Anti-HEXB chain B (SL15468R) at 1/1000 dilution
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
Predicted band size: 22’50 kD
Observed band size: 22’50kD



Sample:
Hela(Human) Cell Lysate at 30 ug
Lung (Mouse) Lysate at 40 ug
Primary: Anti-HEXB chain B (SL15468R) at 1/1000 dilution
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
Predicted band size: 22/50kD
Observed band size: 22/50 kD

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