

产品名称: HEXA Rabbit Polyclonal Antibody

产品货号: APRab11998

产品概述 (Summary)

描述(Description)	Rabbit polyclonal Antibody
宿主(Host)	Rabbit
应用(Application)	WB,IHC,ICC/IF,ELISA
种属反应(Reactivity)	Human,Mouse,Rat
偶联物(Conjugation)	Unconjugated
修饰(Modification)	Unmodified
同种型(Isotype)	IgG
克隆(Clonality)	Polyclonal
剂型(Form)	Liquid
保存条件(Storage)	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
储存溶液(Buffer)	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
纯化 (Purification)	Affinity purification

产品应用(Application)

稀释比(Dilution Ratio)	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:10000-1:20000
分子量(Molecular Weight)	60kDa

抗原信息(Antigen Information)

基因名(Gene Name)	HEXA
别名(Alternative Names)	HEXA; Beta-hexosaminidase subunit alpha; Beta-N-acetylhexosaminidase subunit alpha; Hexosaminidase subunit A; N-acetyl-beta-glucosaminidase subunit alpha
基因 ID(Gene ID)	3073.0
SwissProt ID	P06865
免疫原(Immunogen)	Synthesized peptide derived from HEXA . at AA range: 121-170

研究背景 (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],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 and other tissues. The form B is active against certain oligosaccharides. The form S has no measurable activity.,online information:HEXA mutation database,online information:Tay Sachs disease website,PTM:N-linked glycan at Asn-115 consists of Man(3)-GlcNAc(2),,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.,

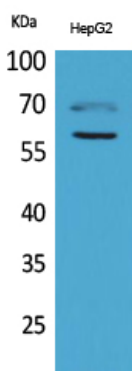
研究领域 (Research Area)

Other glycan degradation;Amino sugar and nucleotide sugar metabolism;Glycosaminoglycan degradation;Glycosphingolipid biosynthesis;Glycosphingolipid biosynthesis;Lysosome;

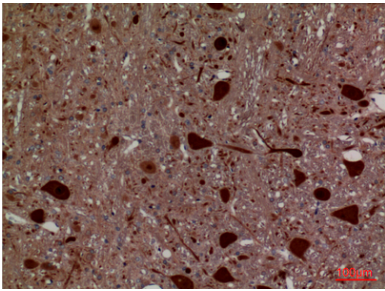
注意事项 (Note)

For research use only.

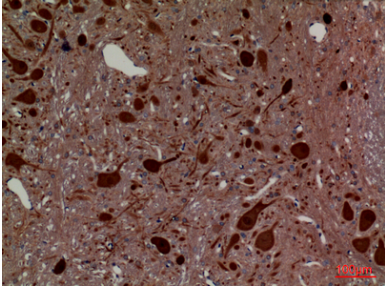
图片 (Image Data)



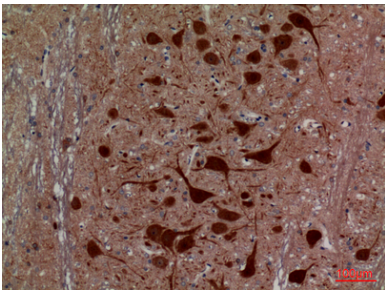
Western Blot analysis of HepG2 cells using HEXA Polyclonal Antibody. Antibody was diluted at 1:1000. Secondary antibody was diluted at 1:20000



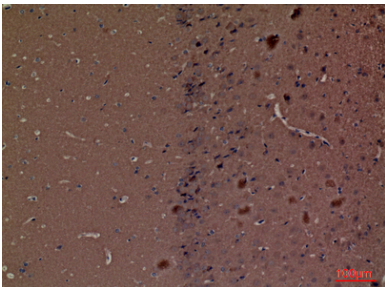
Immunohistochemical analysis of paraffin-embedded rat-brain, antibody was diluted at 1:100



Immunohistochemical analysis of paraffin-embedded rat-brain, antibody was diluted at 1:100



Immunohistochemical analysis of paraffin-embedded rat-brain, antibody was diluted at 1:100



Immunohistochemical analysis of paraffin-embedded mouse-brain, antibody was diluted at 1:100

EnkiLife 优势产品推荐

产品类别	产品货号	产品名称
WB 解决方案	RA10020	2h 极速 WB 即用型全流程试剂盒
	RA10021	4h 快速 WB 即用型全流程试剂盒
	RA10042	5h 畅享版 WB 全流程试剂盒
	RA10037	校准级彩色预染蛋白 Marker (8-180kDa)
	RA10038	校准级彩色预染蛋白 Marker (10-250kDa)
	RA10039	校准级高分子彩色预染蛋白 Marker (25-400kDa)
TSA 多重荧光染色试剂盒	RA10008	TSA 双标三色多重荧光染色试剂盒 (mIHC)
	RA10009	TSA 三标四色多重荧光染色试剂盒 (mIHC)
	RA10010	TSA 四标五色多重荧光染色试剂盒 (mIHC)
	RA10011	TSA 五标六色多重荧光染色试剂盒 (mIHC)
	RA10012	TSA 六标七色多重荧光染色试剂盒 (mIHC)
IHC 检测试剂盒	RA10006	HRP Anti-Mouse/Rabbit IHC Detection System
	RA10007	Polymer-HRP Anti-Mouse/Rabbit IHC Detection System
抗体标记试剂盒	RE80004p	辣根过氧化物酶(HRP)抗体标记试剂盒
	RE80002q	Sulfo-NHS-生物素标记试剂盒
	RE80007p	Cy3 荧光素标记试剂盒
	RE80011p	Fluor488 荧光素标记试剂盒
	RE80017p	Fluor750 荧光素标记试剂盒
	RE80005p	藻红蛋白(R-PE) 抗体快速标记试剂盒
	RE80040	PE-Cy7 串联染料抗体快速标记试剂盒
稳转细胞系构建服务 (免费赠送全膜 WB 验证)	TS-0001	过表达稳转细胞系构建
	TS-0002	敲低稳转细胞系构建
	TS-0003	敲除细胞系构建

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