

HEXA Polyclonal Antibody

Catalog No: YT5233

Reactivity: Human; Mouse; Rat

Applications: WB;IHC;IF;ELISA

Target: HEXA

Fields: >>Other glycan degradation;>>Various types of N-glycan biosynthesis;>>Amino

sugar and nucleotide sugar metabolism;>>Glycosaminoglycan

degradation;>>Sphingolipid metabolism;>>Glycosphingolipid biosynthesis - globo

and isoglobo series;>>Glycosphingolipid biosynthesis - ganglio

series;>>Metabolic pathways;>>Lysosome

Gene Name: HEXA

Protein Name: Beta-hexosaminidase subunit alpha

P06865

P29416

Human Gene ld: 3073

Human Swiss Prot

No:

Mouse Gene ld: 15211

Mouse Swiss Prot

No:

Rat Gene Id: 300757

Rat Swiss Prot No: Q641X3

Immunogen: Synthesized peptide derived from HEXA . at AA range: 121-170

Specificity: HEXA Polyclonal Antibody detects endogenous levels of HEXA protein.

Formulation : Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

Source: Polyclonal, Rabbit, IgG

1/3



Dilution: WB 1:500 - 1:2000. IHC: 1:100-300 ELISA: 1:20000.. IF 1:50-200

Purification: The antibody was affinity-purified from rabbit antiserum by affinity-

chromatography using epitope-specific immunogen.

Concentration: 1 mg/ml

Storage Stability: -15°C to -25°C/1 year(Do not lower than -25°C)

Observed Band: 60kD

Cell Pathway: Other glycan degradation; Amino sugar and nucleotide sugar

metabolism;Glycosaminoglycan degradation;Glycosphingolipid

biosynthesis;Glycosphingolipid biosynthesis;Lysosome;

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],

Function: 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

Subcellular Lysosome.

Location:

Expression:

Brain, Eye, Liver, Ovary, Uterus,

Sort : 7329

2/3

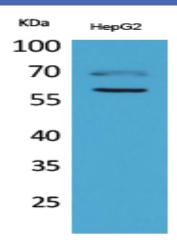


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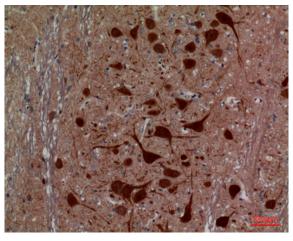
Host: Rabbit

Modifications: Unmodified

Products Images



Western Blot analysis of HepG2 cells using HEXA Polyclonal Antibody. Antibody was diluted at 1:1000. Secondary antibody(catalog#:RS0002) was diluted at 1:20000



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