Product Name: Glucuronidase β Rabbit Polyclonal Antibody
Catalog #: APRab11488

Summary

Production Name	Glucuronidase β Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	WB,IHC-P,IF-P,IF-F,ICC/IF,ELISA
Reactivity	Human,Mouse,Rat

Performance

Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Purification	Affinity purification

Immunogen

Gene Name	GUSB
Alternative Names	GUSB; Beta-glucuronidase; Beta-G1
Gene ID	2990.0
SwissProt ID	P08236.The antiserum was produced against synthesized peptide derived from human GUSB. AA range:321-370

Application

Dilution Ratio	IHC-P 100-300.WB 1:500-1:2000, ELISA 1:10000, IF-P/IF-F/ICC/IF 1:50-200
Molecular Weight	78kDa

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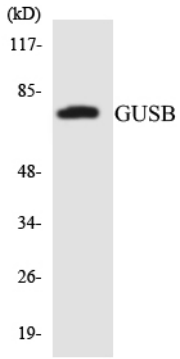
Background

This gene encodes a hydrolase that degrades glycosaminoglycans, including heparan sulfate, dermatan sulfate, and chondroitin-4,6-sulfate. The enzyme forms a homotetramer that is localized to the lysosome. Mutations in this gene result in mucopolysaccharidosis type VII. Alternative splicing results in multiple transcript variants. There are many pseudogenes of this locus in the human genome.[provided by RefSeq, May 2014],catalytic activity:A beta-D-glucuronoside + H(2)O = D-glucuronate + an alcohol.,disease:Defects in GUSB are the cause of mucopolysaccharidosis type 7 (MPS7) [MIM:253220]; also known as Sly syndrome. MPS7 is an autosomal recessive lysosomal storage disease characterized by inability to degrade glucuronic acid-containing glycosaminoglycans. The phenotype is highly variable, ranging from severe lethal hydrops fetalis to mild forms with survival into adulthood. Most patients with the intermediate phenotype show hepatomegaly, skeletal anomalies, coarse facies, and variable degrees of mental impairment.,disease:Mucopolysaccharidosis type 7 is associated with non-immune hydrops fetalis [MIM:236750]. Hydrops fetalis is a generalized edema of the fetus with fluid accumulation in the body cavities.,enzyme regulation:Inhibited by L-aspartic acid.,function:Plays an important role in the degradation of dermatan and keratan sulfates.,PTM:N-linked glycosylated with 3 to 4 oligosaccharide chains.,similarity:Belongs to the glycosyl hydrolase 2 family.,subunit:Homotetramer.,

Research Area

Pentose and glucuronate interconversions;Starch and sucrose metabolism;Glycosaminoglycan degradation;Porphyrin and chlorophyll metabolism;Drug metabolism;Lysosome;

Image Data



Western blot analysis of the lysates from COLO205 cells using GUSB antibody.

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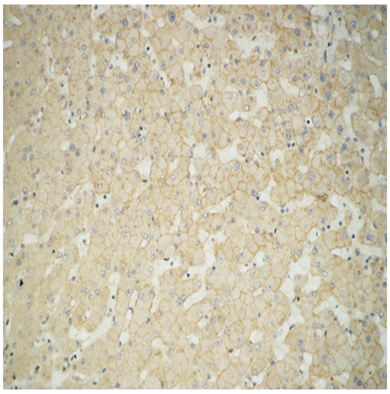
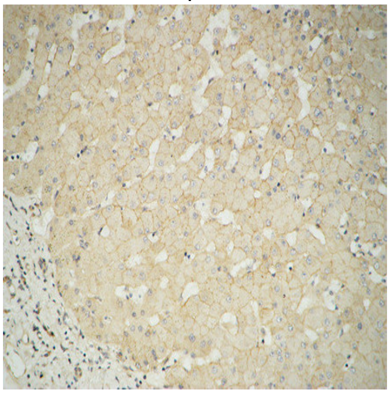


Immunohistochemical analysis of paraffin-embedded Human Liver. 1, Antibody was diluted at 1:100（4°,overnight）. 2, High-pressure and temperature EDTA, pH8.0 was used for antigen retrieval. 3,Secondary antibody was diluted at 1:200（room temperature, 30min）.



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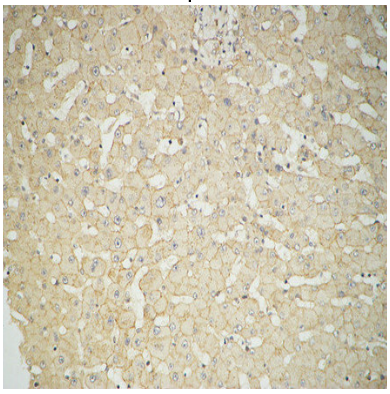

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Note

For research use only.