产品名称：Glucuronidase β Rabbit Polyclonal Antibody
产品货号：APRab11488

产品概述（Summary）

产品名称（Production Name）	Glucuronidase β Rabbit Polyclonal Antibody
描述（Description）	Rabbit polyclonal Antibody
宿主（Host）	Rabbit
应用（Application）	WB,IHC,ICC/IF,ELISA
种属反应性（Reactivity）	Human,Mouse,Rat

产品性能（Performance）

偶联物（Conjugation）	Unconjugated
修饰（Modification）	Unmodified
同种型（Isotype）	IgG
克隆（Clonality）	Polyclonal
形式（Form）	Liquid
存放说明（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.
纯化方式（Purification）	Affinity purification

免疫原信息（Immunogen）

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

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 1:5000-1:20000
蛋白分子量（Molecular Weight）	78kDa

研究背景（Background）

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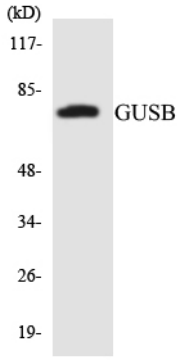
This gene encodes a hydrolase that degrades glycosaminoglycans, including heparan sulfate, dermatan sulfate, and chondroitin-4,6-sulfate. The enzyme forms a homotetramer that is localized to the lysosome. Mutations in this gene result in mucopolysaccharidosis type VII. Alternative splicing results in 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.,

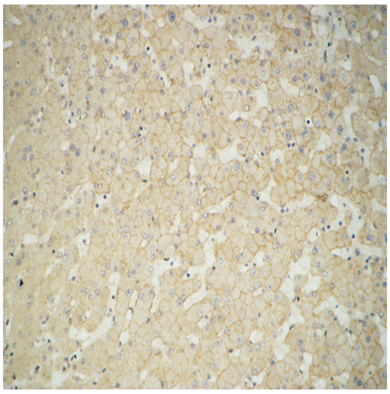
研究领域（Research Area）

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

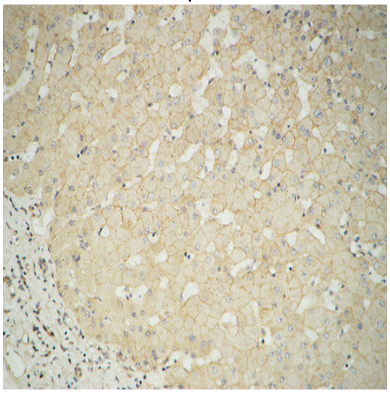
图片（Image Data）



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



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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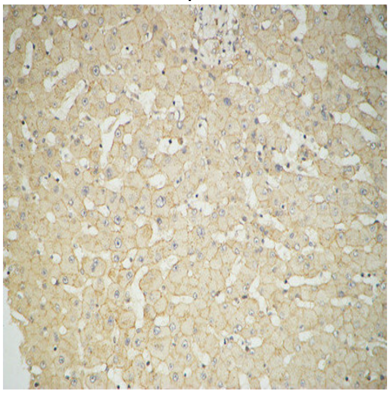

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注意事项（Note）

For research use only .