β 葡萄糖醛酸苷酶抗体

产品货号： mlR7980

英文名称： beta glucuronidase

中文名称： β 葡萄糖醛酸苷酶抗体

别　　名： asd; Beta G1; Beta glucuronidase; Beta-G1; Beta-glucuronidase; BG; BGLR; BGLR_HUMAN; Glucuronidase beta; Gur; Gus; Gus-r; Gus-s; Gus-t; Gus-u; GUSB; Gut; MPS7; Ac2-223.

研究领域： 细胞生物　免疫学　信号转导

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Chicken, Dog, Pig, Cow,

产品应用： WB=1:500-2000 ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 IF=1:100-500 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分 子 量： 69kDa

细胞定位： 细胞浆

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human GUSB/beta glucuronidase:589-651/651

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

PubMed： PubMed

产品介绍： Defects in GUSB are the cause of mucopolysaccharidosis type 7 (MPS7) ; 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. Mucopolysaccharidosis type 7 is associated with non-immune hydrops fetalis, a generalized edema of the fetus with fluid accumulation in the body cavities due to non-immune causes. Non-immune hydrops fetalis is not a diagnosis in itself but a symptom, a feature of many genetic disorders, and the end-stage of a wide variety of disorders.

Function:

Plays an important role in the degradation of dermatan and keratan sulfates.

Subunit:

Homotetramer.

Subcellular Location:

Lysosome.

Post-translational modifications:

N-linked glycosylated with 3 to 4 oligosaccharide chains.

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.

Note=Mucopolysaccharidosis type 7 is associated with non-immune hydrops fetalis, a generalized edema of the

fetus with fluid accumulation in the body cavities due to non-immune causes. Non-immune hydrops fetalis is not a diagnosis in itself but a symptom, a feature of many genetic disorders, and the end-stage of a wide variety of disorders.

Similarity:

Belongs to the glycosyl hydrolase 2 family.

SWISS:

P08236

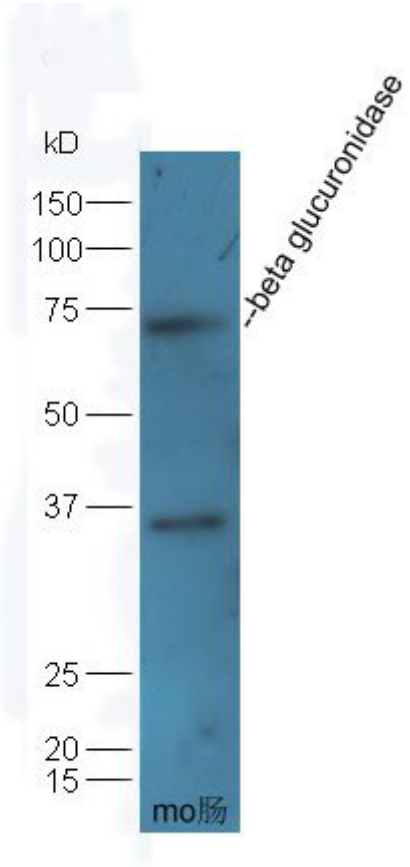
Gene ID:

2990

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

产品图片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