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Rabbit Anti-beta glucuronidase antibody

SL7980R

Product Name:	beta glucuronidase
Chinese Name:	β葡萄糖醛酸苷酶抗体
Alias:	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.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Pig,Cow,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	69kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human GUSB/beta glucuronidase:589-651/651
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	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
Product Detail:	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.

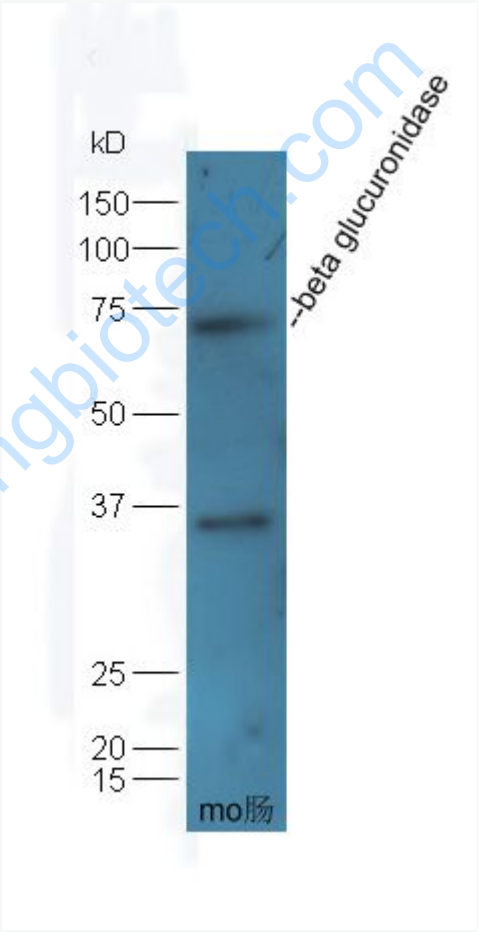
Similarity:
Belongs to the glycosyl hydrolase 2 family.

SWISS:
P08236

Gene ID:
2990

Database links:

Entrez Gene: 2990Human

	Omim: 611499Human SwissProt: P08236Human Unigene: 255230Human **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 Sample: Intestine (Mouse) Lysate at 40 ug Primary: Anti-beta glucuronidase (SL7980R) at 1/300 dilution Secondary: HRP conjugated Goat-Anti-rabbit IgG (SL7980R) at 1/5000 dilution Predicted band size: 69 kD

	Observed band size: 69 kD