

Product name:	Glucuronidase β Rabbit Polyclonal Antibody
Cat number:	ABN11488
Conjugate:	Unconjugated
Size:	100 μ L
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human GUSB. AA range:321-370
Reactivity:	Human,Mouse,Rat
Applications:	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:5000-1:20000
Molecular Weight:	78kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

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.,