



Glucuronidase β Monoclonal Antibody

Catalog No	YP-mAb-02643
Isotype	IgG
Reactivity	Human;Mouse;Rat
Applications	WB
Gene Name	GUSB
Immunogen	The antiserum was produced against synthesized peptide derived from human GUSB. AA range:321-370
Specificity	Glucuronidase β Monoclonal Antibody detects endogenous levels of Glucuronidase β protein.
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-1:2000
Concentration	1 mg/ml
Purity	$\geq 90\%$
Storage Stability	-20°C/1 year
Synonyms	Beta G1;Beta glucuronidase;glucuronidase;beta;GUSB;MPS7;Glucuronidase β ; Beta-glucuronidase;Beta-G1
Observed Band	78kD
Calculated Molecular Weight	78kD
Cell Pathway	Lysosome.
Tissue Specificity	Colon,Fibroblast,Liver,Placenta,Plasma,
Function	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

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],

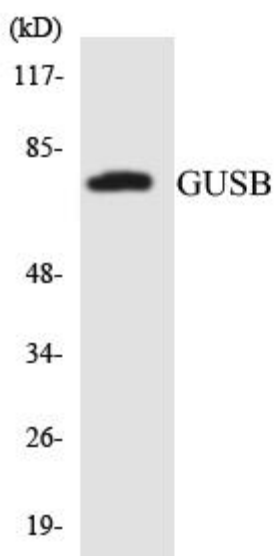
matters needing attention

Avoid repeated freezing and thawing!

Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western Blot analysis of various cells using Glucuronidase β Monoclonal Antibody