GNS Monoclonal Antibody

Catalog No	BYmab-07101
Isotype	IgG
Reactivity	Human;Mouse
Applications	WB
Gene Name	GNS
Protein Name	N-acetylglucosamine-6-sulfatase (EC 3.1.6.14) (Glucosamine-6-sulfatase) (G6S)
Immunogen	Synthesized peptide derived from human protein . at AA range: 190-270
Specificity	GNS Monoclonal Antibody detects endogenous levels of protein.
Formulation	Liquid in PBS containing 50% glycerol, 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 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	
Observed Band	60kD
Cell Pathway	Lysosome.
Tissue Specificity	Endothelial cell,Human uterus endothel primary cell culture,Liver,Urinary b
Function	catalytic activity:Hydrolysis of the 6-sulfate groups of the N-acetyl-D-glucosamine 6-sulfate units of heparan sulfate and keratan sulfate.,cofactor:Binds 1 calcium ion per subunit.,disease:Defects in GNS are the cause of mucopolysaccharidosis type 3D (MPS3D) [MIM:252940]; also known as Sanfilippo D syndrome. MPS3D is a form of mucopolysaccharidosis type 3, an autosomal recessive lysosomal storage disease due to impaired degradation of heparan sulfate. MPS3 is characterized by severe central nervous system degeneration, but only mild somatic disease. Onset of clinical features usually occurs between 2 and 6 years; severe neurologic degeneration occurs in most patients between 6 and 10 years of age, and death occurs typically during the second or third decade of life.,PTM:The conversion to 3-oxoalanine (also known as C-formylglycine, FGly), of a serine or cysteine residue in prokaryotes a

Background	glucosamine (N-acetyl)-6-sulfatase(GNS) Homo sapiens The product of this gene is a lysosomal enzyme found in all cells. It is involved in the catabolism of heparin, heparan sulphate, and keratan sulphate. Deficiency of this enzyme results in the accumulation of undegraded substrate and the lysosomal storage disorder mucopolysaccharidosis type IIID (Sanfilippo D syndrome). Mucopolysaccharidosis type IIID is the least common of the four subtypes of Sanfilippo syndrome. [provided by RefSeq, Jul 2008],
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.

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