Immunotag™ GNS Polyclonal Antibody

Antibody Specification

Catalog No.	ITN2239
Product Description	Immunotag™ GNS Polyclonal Antibody
Size	50 µg, 100 µg
Conjugation	HRP, Biotin, FITC, Alexa Fluor® 350, Alexa Fluor® 405, Alexa Fluor® 488, Alexa Fluor® 555, Alexa Fluor® 594, Alexa Fluor® 647
IMPORTANT NOTE	This product is custom manufactured with a lead time of 3-4 weeks. Once in production, this item cannot be cancelled from an order and is not eligible for return.
Target Protein	GNS
Clonality	Polyclonal
Storage/Stability	-20°C/1 year
Application	WB,ELISA
Recommended Dilution	WB 1:500-2000 ELISA 1:5000-20000
Concentration	1 mg/ml
Reactive Species	Human,Mouse
Host Species	Rabbit
Immunogen	Synthesized peptide derived from human protein . at AA range: 190-270
Specificity	GNS Polyclonal Antibody detects endogenous levels of protein.
Purification	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen
Form	Liquid in PBS containing 50% glycerol, and 0.02% sodium azide.
Gene Name	GNS
Accession No.	P15586 Q8BFR4
Description	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],

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Cell Pathway/ Category	Glycosaminoglycan degradation,Lysosome,
Protein Expression	Endothelial cell,Human uterus endothel primary cell culture,Liver,Urinary b
Subcellular Localization	lysosome,lysosomal lumen,extracellular exosome,
Protein 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 and of a cysteine residue in eukaryotes, is critical for catalytic activity.,PTM:The form A (78 kDa) is processed by internal peptidase cleavage to a 32 kDa N-terminal species (form B) and a 48 kDa C-terminal species.,similarity:Belongs to the sulfatase family.,
Usage	For Research Use Only! Not for diagnostic or therapeutic procedures.