Human Glucosamine (N-acetyl)-6-Sulfatase/GNS Antibody

Antigen Affinity-purified Polyclonal Goat IgG

Catalog Number: AF2484

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human Glucosamine (N-acetyl)-6-Sulfatase/GNS in direct ELISAs and Western blots.
Source	Polyclonal Goat IgG
Purification	Antigen Affinity-purified
Immunogen	Mouse myeloma cell line NS0-derived recombinant human Glucosamine (N-acetyl)-6-Sulfatase/GNS Val37-Leu552 Accession # P15586
Formulation	Lyophilized from a 0.2 μm filtered solution in PBS with Trehalose. See Certificate of Analysis for details. *Small pack size (-SP) is supplied either lyophilized or as a 0.2 μm filtered solution in PBS.

APPLICATIONS

Please Note: *Optimal dilutions should be determined by each laboratory for each application. General Protocols are available in the Technical Information section on our website.*

	Recommended Concentration	Sample
Western Blot	0.1 µg/mL	Recombinant Human Glucosamine (N-acetyl)-6-Sulfatase/GNS (Catalog # 2484-SUC)
Immunoprecipitation	25 µg/mL	Conditioned cell culture medium spiked with Recombinant Human Glucosamine (N-acetyl)-6-Sulfatase/GNS (Catalog # 2484-SUC), see our available Western blot detection antibodies

PREPARATION AND STORAGE

Reconstitution	Reconstitute at 0.2 mg/mL in sterile PBS.
Shipping	The product is shipped at ambient temperature. Upon receipt, store it immediately at the temperature recommended below. *Small pack size (-SP) is shipped with polar packs. Upon receipt, store it immediately at -20 to -70 °C
Stability & Storage	**Use a manual defrost freezer and avoid repeated freeze-thaw cycles.** ● 12 months from date of receipt, -20 to -70 °C as supplied. ● 1 month, 2 to 8 °C under sterile conditions after reconstitution. ● 6 months, -20 to -70 °C under sterile conditions after reconstitution.

BACKGROUND

A member of the sulfatase family, GNS is required for the lysosomal degradation of the glycosaminoglycans (GAG) heparan sulfate and keratan sulfate (1, 2). It hydrolyzes the 6-sulfate group of the N-acetyl-D-glucosamine 6-sulfate units of the GAG. GNS deficiency results in mucopolysaccharidosis type IIID (MPS IIID or Sanfilippo D Syndrome), an inborn error leading to lysosomal accumulation of heparan sulfate. MPS IIID has profound mental deterioration, hyperactivity, and relatively mild somatic manifestations. The deduced amino acid sequence of human GNS consists of a signal peptide (residues 1-36) and a mature chain (residues 37-552) that may be further processed into N-terminal and C-terminal fragments (3). Recombinant human GNS corresponds to the single chain and has sulfatase activity.

References:

1. Parenti, G. *et al.* (1997) Curr. Opin. Genet. & Dev. **7**:386.
2. Neufeld, E.F. and J. Muenzer (2001) in *The Metabolic and Molecular Basis of Inherited Disease*, Scriver, C.R. *et al.* (eds.) pp. 3421 - 3452, New York, McGraw-Hill.
3. Robertson, D.A. *et al.* (1992) Biochem. J. **288**:539.

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