

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
Conjugate	Alexa Fluor 532 Excitation Wavelength: 534 nm Emission Wavelength: 553 nm
Formulation	Supplied 0.2mg/ml in 1X PBS with RDF1 and 0.09% Sodium Azide *Contains <0.1% Sodium Azide, which is not hazardous at this concentration according to GHS classifications. Refer to the Safety Data Sheet (SDS) for additional information and handling instructions.

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.

Western Blot	Optimal dilution of this antibody should be experimentally determined.
Immunoprecipitation	Optimal dilution of this antibody should be experimentally determined.

PREPARATION AND STORAGE

Shipping	The product is shipped with polar packs. Upon receipt, store it immediately at the temperature recommended below.
Stability & Storage	Protect from light. Do not freeze. 12 months from date of receipt, 2 to 8 °C as supplied

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.

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