SunLong Biotech Co.,LTD
Tel: 0086-571-56623320 Fax:0086-571-56623318
E-mail:sales@sunlongbiotech.com
www.sunlongbiotech.com

[Marker Primary Antibody]

Rabbit Anti-GNS/Biotin Conjugated antibody

Catalog Number: SL13479R-Bio

FOR RESEARCH USE ONLY

[DATASHEET]

Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	Anti-GNS/Biotin
Immunogen:	KLH conjugated synthetic peptide derived from human GNS/Glucosamine 6 sulfatase
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
Applications:	IHC-P=1:100-500 IHC-F=1:100-500 ICC/IF=1:50-200 IF=1:100-500 ELISA=1:500-5000 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Reactivity:	(predicted:Human,Mouse,Rat,Cow,Horse,Rabbit)
Background:	background: GNS is a 552 amino acid lysosomal enzyme that hydrolyzes the 6-sulfate groups of the N-acetyl-D-glucosamine 6-sulfate units of keratan sulfate and heparan sulfate. A member of the sulfatase family, GNS assists in the catabolism of heparin, and binds calcium as a cofactor. GNS deficiency results in an autosomal recessive lysosomal storage disorder known as mucopolysaccharidosis type IIID (Sanfilippo D syndrome), which is characterized by mild somatic disease and severe degeneration of the central nervous system. Subject to post-translational internal peptidase cleavage, GNS is encoded by a gene mapping to human chromosome 12q14.2 and mouse chromosome 10 D2.

Important Note: This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

	Subcellular Location: Lysosome. **Post-translational modifications:** 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. 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. 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. **Similarity:** Belongs to the sulfatase family. **Database links:** **Entrez Gene:** 2799 Human Omim: 607664 Human **SwissProt:** P15586 Human **Unigene:** 334534 Human Important **Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.