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GNS Rabbit pAb

Catalog # AP99129

Product Information

Application	IHC-P, IHC-F, IF, E
Primary Accession	P15586
Other Accession	P15586
Predicted	Rat, Human, Mouse, Rabbit, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	62082
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human GNS/Glucosamine 6 sulfatase
Epitope Specificity	1-100/552
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the sulfatase family.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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.

Additional Information

Gene ID	2799
Other Names	G6S; GNS_HUMAN; GNS; Glucosamine-6-sulfatase (G6S); 3.1.6.14; glucosamine (N-acetyl)-6-sulfatase; Sanfilippo disease IIID; N-acetylglucosamine-6-sulfatase
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	GNS
Function	Hydrolyzes 6-sulfate groups in N-acetyl-d-glucosaminide units of heparin sulfate and keratan sulfate.
Cellular Location	Lysosome.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.