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Rabbit Anti-GNS antibody

SL13479R

Product Name:	GNS
Chinese Name:	氨基葡萄糖6-硫酸酯酶抗体
Alias:	2610016K11Rik; AU042285; C87209; G6S; Glucosamine (N-acetyl) 6 sulfatase; Glucosamine 6 sulfatase; Glucosamine-6-sulfatase; GNS; GNS_HUMAN; MGC21274; N acetylglucosamine 6 sulfatase [Precursor]; N-acetylglucosamine-6-sulfatase; N28088.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Cow,Horse,Rabbit,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	58kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human GNS/Glucosamine 6 sulfatase:1-100/552
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
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 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	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.

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.

SWISS:
P15586

Gene ID:
2799

Database links:

Entrez Gene: 2799Human

Omim: 607664Human

SwissProt: P15586Human

Unigene: 334534Human

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