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Rabbit Anti-GNS/Cy5 Conjugated antibody

SL13479R-Cy5

Product Name	Anti-GNS/Cy5
Chinese Name	Cy5 标记的氨基葡萄糖 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.
Research Area	Cell biology Developmental biology Neurobiology The new supersedes the old
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	(predicted:Human,Mouse,Rat,Cow,Horse,Rabbit)
Applications	ICC/IF=1:50-200,IF=1:100-500 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight	58kDa
Form	Lyophilized or Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human GNS/Glucosamine 6 sulfatase
Lsotype	IgG
Purification	affinity purified by Protein A
Storage Buffer	1M TBS(pH7.4) with 1% BSA, 3% 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 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
Product Detail	**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.

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.

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