

# GNS (T-16): sc-161670

## BACKGROUND

GNS (glucosamine (N-acetyl)-6-sulfatase), also known as G6S (glucosamine-6-sulfatase), 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 12q14.3 and mouse chromosome 10 D2.

## REFERENCES

- Tomatsu, S., Fukuda, S., Masue, M., Sukegawa, K., Fukao, T., Yamagishi, A., Hori, T., Iwata, H., Ogawa, T. and Nakashima, Y. 1991. Morquio disease: isolation, characterization and expression of full-length cDNA for human N-acetylgalactosamine-6-sulfate sulfatase. *Biochem. Biophys. Res. Commun.* 181: 677-683.
- Robertson, D.A., Freeman, C., Morris, C.P. and Hopwood, J.J. 1992. A cDNA clone for human glucosamine-6-sulphatase reveals differences between arylsulphatases and non-arylsulphatases. *Biochem. J.* 288: 539-544.
- Robertson, D.A., Freeman, C., Nelson, P.V., Morris, C.P. and Hopwood, J.J. 1988. Human glucosamine-6-sulfatase cDNA reveals homology with steroid sulfatase. *Biochem. Biophys. Res. Commun.* 157: 218-224.
- Robertson, D.A., Callen, D.F., Baker, E.G., Morris, C.P. and Hopwood, J.J. 1988. Chromosomal localization of the gene for human glucosamine-6-sulphatase to 12q14. *Hum. Genet.* 79: 175-178.
- Beesley, C.E., Burke, D., Jackson, M., Vellodi, A., Winchester, B.G. and Young, E.P. 2003. Sanfilippo syndrome type D: identification of the first mutation in the N-acetylglucosamine-6-sulphatase gene. *J. Med. Genet.* 40: 192-194.
- Jansen, A.C., Cao, H., Kaplan, P., Silver, K., Leonard, G., De Meirleir, L., Lissens, W., Liebaers, I., Veilleux, M., Andermann, F., Hegele, R.A. and Andermann, E. 2007. Sanfilippo syndrome type D: natural history and identification of 3 novel mutations in the GNS Gene. *Arch. Neurol.* 64: 1629-1634.
- Elçioğlu, N.H., Pawlik, B., Colak, B., Beck, M. and Wollnik, B. 2009. A novel loss-of-function mutation in the GNS gene causes Sanfilippo syndrome type D. *Genet. Couns.* 20: 133-139.

## CHROMOSOMAL LOCATION

Genetic locus: GNS (human) mapping to 12q14.3; Gns (mouse) mapping to 10 D2.

## SOURCE

GNS (T-16) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of GNS of human origin.

## PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-161670 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

GNS (T-16) is recommended for detection of GNS of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

GNS (T-16) is also recommended for detection of GNS in additional species, including equine, canine, bovine, porcine and avian.

Suitable for use as control antibody for GNS siRNA (h): sc-96024, GNS siRNA (m): sc-145660, GNS shRNA Plasmid (h): sc-96024-SH, GNS shRNA Plasmid (m): sc-145660-SH, GNS shRNA (h) Lentiviral Particles: sc-96024-V and GNS shRNA (m) Lentiviral Particles: sc-145660-V.

Molecular Weight of GNS: 62 kDa.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

## STORAGE

Store at 4° C, **\*\*DO NOT FREEZE\*\***. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

## RESEARCH USE

For research use only, not for use in diagnostic procedures.

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.