

NAGLU (54-G): sc-130383

BACKGROUND

NAGLU (N-acetyl-alpha-glucosaminidase), also known as NAG, UFHSD1, MPS3B or MPS-IIIB, is a 743 amino acid protein that exists as both a monomer and a homodimer. Expressed in ovary, liver, testis, prostate, lung, colon, kidney, spleen, placenta and peripheral blood leukocytes, NAGLU is involved in the degradation of heparan sulfate (HS), specifically functioning to catalyze the hydrolysis of terminal N-acetyl-D-glucosamine residues in N-acetyl- α -D-glucosaminides. Defects in the gene encoding NAGLU are the cause of mucopolysaccharidosis type IIIB (MPS-IIIB), also known as Sanfilippo syndrome B. MPS-IIIB is an autosomal recessive disorder in which the body fails to degrade HS, leading to an accumulation of HS in lysosomes and urine and resulting in mental deterioration and, ultimately, death.

REFERENCES

- Weber, B., et al. 1996. Cloning and expression of the gene involved in Sanfilippo B syndrome (mucopolysaccharidosis III B). *Hum. Mol. Genet.* 5: 771-777.
- Zhao, H.G., et al. 1996. The molecular basis of Sanfilippo syndrome type B. *Proc. Natl. Acad. Sci. USA* 93: 6101-6105.
- Schmidtchen, A., et al. 1998. NAGLU mutations underlying Sanfilippo syndrome type B. *Am. J. Hum. Genet.* 62: 64-69.
- Weber, B., et al. 1999. Sanfilippo type B syndrome (mucopolysaccharidosis III B): allelic heterogeneity corresponds to the wide spectrum of clinical phenotypes. *Eur. J. Hum. Genet.* 7: 34-44.
- Bunge, S., et al. 1999. Mucopolysaccharidosis type III B (Sanfilippo B): identification of 18 novel α -N-acetylglucosaminidase gene mutations. *J. Med. Genet.* 36: 28-31.
- Yogalingam, G. and Hopwood, J.J. 2001. Molecular genetics of mucopolysaccharidosis type IIIA and IIIB: diagnostic, clinical, and biological implications. *Hum. Mutat.* 18: 264-281.
- Chinen, Y., et al. 2005. Sanfilippo type B syndrome: five patients with an R565P homozygous mutation in the α -N-acetylglucosaminidase gene from the Okinawa islands in Japan. *J. Hum. Genet.* 50: 357-359.
- Heldermon, C.D., et al. 2007. Development of sensory, motor and behavioral deficits in the murine model of Sanfilippo syndrome type B. *PLoS ONE* 2: e772.
- Ficko-Blean, E., et al. 2008. Structural and mechanistic insight into the basis of mucopolysaccharidosis III B. *Proc. Natl. Acad. Sci. USA* 105: 6560-6565.

CHROMOSOMAL LOCATION

Genetic locus: NAGLU (human) mapping to 17q21.2; Naglu (mouse) mapping to 11 D.

SOURCE

NAGLU (54-G) is a mouse monoclonal antibody raised against a partial recombinant protein mapping within amino acids 644-742 of NAGLU of human origin.

PRODUCT

Each vial contains 100 μ g IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

NAGLU (54-G) is recommended for detection of NAGLU of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) and immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)].

Suitable for use as control antibody for NAGLU siRNA (h): sc-93564, NAGLU siRNA (m): sc-149803, NAGLU shRNA Plasmid (h): sc-93564-SH, NAGLU shRNA Plasmid (m): sc-149803-SH, NAGLU shRNA (h) Lentiviral Particles: sc-93564-V and NAGLU shRNA (m) Lentiviral Particles: sc-149803-V.

Molecular Weight of NAGLU: 82 kDa

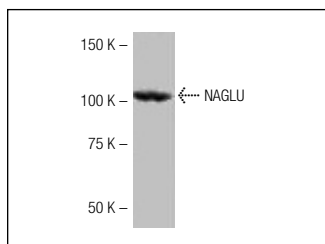
Molecular Weight of NAGLU cleavage product: 77 kDa.

Positive Controls: Jurkat whole cell lysate: sc-2204.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker[™] Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).

DATA



NAGLU (54-G): sc-130383. Western blot analysis of NAGLU expression in Jurkat whole cell lysate.

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 for detailed protocols and support products.