SANTA CRUZ BIOTECHNOLOGY, INC.

NAGLU (C-14): sc-168707

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

NAGLU (N-acetyl-α-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

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2. Zhao, H.G., et al. 1996. The molecular basis of Sanfilippo syndrome type B. Proc. Natl. Acad. Sci. USA 93: 6101-6105.
3. Schmidtchen, A., et al. 1998. NAGLU mutations underlying Sanfilippo syndrome type B. Am. J. Hum. Genet. 62: 64-69.
4. 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.
5. 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.
6. 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.
7. 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.
8. 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.

CHROMOSOMAL LOCATION

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

SOURCE

NAGLU (C-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of NAGLU of human origin.

STORAGE

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

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-168707 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

NAGLU (C-14) is recommended for detection of NAGLU 77 kDa form, 82 kDa form and precursor of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], 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).

NAGLU (C-14) is also recommended for detection of NAGLU 77kDa form, 82kDa form and precursor in additional species, including canine.

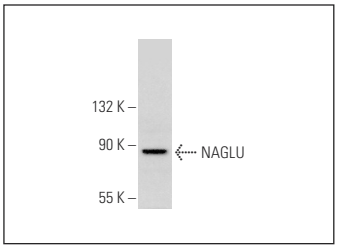
Suitable for use as control antibody for NAGLU siRNA (h): sc-93564, NAGLU siRNA (m): sc-149803, NAGLU shRNA Plasmid (h): sc-93564-SH, NAGLU shRNA Plasmid (m): sc-149803-SH, NAGLU shRNA (h) Lentiviral Particles: sc-93564-V and NAGLU shRNA (m) Lentiviral Particles: sc-149803-V.

Molecular Weight of NAGLU: 82 kDa.

Molecular Weight of NAGLU cleavage product: 77 kDa.

Positive Controls: Jurkat whole cell lysate: sc-2204 or NIH/3T3 whole cell lysate: sc-2210.

DATA



NAGLU (C-14): sc-168707. Western blot analysis of NAGLU expression in NIH/3T3 whole cell lysate.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.