

NAGLU siRNA (h): sc-93564

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 IIIB (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.

CHROMOSOMAL LOCATION

Genetic locus: NAGLU (human) mapping to 17q21.2.

PRODUCT

NAGLU siRNA (h) is a pool of 3 target-specific 19-25 nt siRNAs designed to knock down gene expression. Each vial contains 3.3 nmol of lyophilized siRNA, sufficient for a 10 μ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see NAGLU shRNA Plasmid (h): sc-93564-SH and NAGLU shRNA (h) Lentiviral Particles: sc-93564-V as alternate gene silencing products.

For independent verification of NAGLU (h) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-93564A, sc-93564B and sc-93564C.

STORAGE AND RESUSPENSION

Store lyophilized siRNA duplex at -20° C with desiccant. Stable for at least one year from the date of shipment. Once resuspended, store at -20° C, avoid contact with RNases and repeated freeze thaw cycles.

Resuspend lyophilized siRNA duplex in 330 μ l of the RNase-free water provided. Resuspension of the siRNA duplex in 330 μ l of RNase-free water makes a 10 μ M solution in a 10 μ M Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

NAGLU siRNA (h) is recommended for the inhibition of NAGLU expression in human cells.

SUPPORT REAGENTS

For optimal siRNA transfection efficiency, Santa Cruz Biotechnology's siRNA Transfection Reagent: sc-29528 (0.3 ml), siRNA Transfection Medium: sc-36868 (20 ml) and siRNA Dilution Buffer: sc-29527 (1.5 ml) are recommended. Control siRNAs or Fluorescein Conjugated Control siRNAs are available as 10 μ M in 66 μ l. Each contain a scrambled sequence that will not lead to the specific degradation of any known cellular mRNA. Fluorescein Conjugated Control siRNAs include: sc-36869, sc-44239, sc-44240 and sc-44241. Control siRNAs include: sc-37007, sc-44230, sc-44231, sc-44232, sc-44233, sc-44234, sc-44235, sc-44236, sc-44237 and sc-44238.

GENE EXPRESSION MONITORING

NAGLU (54-G): sc-130383 is recommended as a control antibody for monitoring of NAGLU gene expression knockdown by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) or immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

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 MarkerTM Molecular Weight Standards: sc-2035, UltraCruz[®] Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz[®] Mounting Medium: sc-24941 or UltraCruz[®] Hard-set Mounting Medium: sc-359850.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor NAGLU gene expression knockdown using RT-PCR Primer: NAGLU (h)-PR: sc-93564-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

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.