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NAGA siRNA (m): sc-149799

BACKGROUND

NAGA (N-acetylgalactosaminidase, α), also known as α -galactosidase B or GALB, is a 411 lysosomal protein belonging to the glycosyl hydrolase 27 family that may exist as a homodimer and plays a critical role in glycolipid breakdown. NAGA encodes α -N-acetylgalactosaminidase, a lysosomal enzyme, which cleaves α -N-acetylgalactosaminyl groups from glycoconjugates. Mapping to human chromosome 22q13, NAGA defects are the cause of an autosomal recessive disorder with three phenotypes, known as Schindler disease (types I, II and III) or NAGA deficiency (types I, II and III). Characterized by neurologic manifestations that range in severity, Schindler disease type I is the most severe form, followed by type III, which may have mild-to-moderate effects. Schindler disease type II, also known as Kanzaki disease, is characterized by mild intellectual impairment and angiokeratoma corporis diffusum.

REFERENCES

1. de Groot, P.G., et al. 1978. Localization of a gene for human α -galactosidase B (= N-acetyl- α -D-galactosaminidase) on chromosome 22. *Hum. Genet.* 44: 305-312.
2. Geurts van Kessel, A.H., et al. 1980. Regional localization of the genes coding for human ACO2, ARSA, and NAGA on chromosome 22. *Cytogenet. Cell Genet.* 28: 169-172.
3. Wang, A.M., et al. 1990. Schindler disease: the molecular lesion in the α -N-acetylgalactosaminidase gene that causes an infantile neuroaxonal dystrophy. *J. Clin. Invest.* 86: 1752-1756.
4. de Jong, J., et al. 1994. α -N-acetylgalactosaminidase deficiency with mild clinical manifestations and difficult biochemical diagnosis. *J. Pediatr.* 125: 385-391.
5. Keulemans, J.L., et al. 1996. Human α -N-acetylgalactosaminidase (α -NAGA) deficiency: new mutations and the paradox between genotype and phenotype. *J. Med. Genet.* 33: 458-464.
6. Kodama, K., et al. 2001. A new case of α -N-acetylgalactosaminidase deficiency with angiokeratoma corporis diffusum, with Ménière's syndrome and without mental retardation. *Br. J. Dermatol.* 144: 363-368.
7. Bakker, H.D., et al. 2001. Human α -N-acetylgalactosaminidase (α -NAGA) deficiency: no association with neuroaxonal dystrophy? *Eur. J. Hum. Genet.* 9: 91-96.

CHROMOSOMAL LOCATION

Genetic locus: Naga (mouse) mapping to 15 E1.

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.

PRODUCT

NAGA siRNA (m) 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 NAGA shRNA Plasmid (m): sc-149799-SH and NAGA shRNA (m) Lentiviral Particles: sc-149799-V as alternate gene silencing products.

For independent verification of NAGA (m) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-149799A, sc-149799B and sc-149799C.

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

NAGA siRNA (m) is recommended for the inhibition of NAGA expression in mouse 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.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor NAGA gene expression knockdown using RT-PCR Primer: NAGA (m)-PR: sc-149799-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.