

HGSNAT siRNA (h): sc-75251

BACKGROUND

HGSNAT (heparan- α -glucosaminide N-acetyltransferase), also known as HGNAT, MPS3C or TMEM76, is a 663 amino acid multi-pass membrane protein that localizes to the lysosome and is expressed in a wide variety of tissues, including liver, lung, placenta, heart and skeletal muscle. Functioning as a lysosomal acetyltransferase, HGSNAT acetylates the non-reducing terminal α -glucosamine residue of intralysosomal heparin or heparan sulfate, effectively producing a substrate for luminal α -N-acetyl glucosaminidase. Defects in the gene encoding HGSNAT are the cause of mucopolysaccharidosis type 3C (MPS3C), an autosomal recessive lysosomal storage disease that is caused by impaired heparan sulfate degradation and is characterized by degeneration of the central nervous system.

REFERENCES

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2. Meikle, P.J., et al. 1995. Human acetyl-coenzyme A: α -glucosaminide N-acetyltransferase. Kinetic characterization and mechanistic interpretation. *Biochem. J.* 308: 327-333.
3. Ausseil, J., et al. 2004. Localisation of a gene for mucopolysaccharidosis IIIC to the pericentromeric region of chromosome 8. *J. Med. Genet.* 41: 941-945.
4. Fan, X., et al. 2006. Identification of the gene encoding the enzyme deficient in mucopolysaccharidosis IIIC (Sanfilippo disease type C). *Am. J. Hum. Genet.* 79: 738-744.
5. Hřebíček, M., et al. 2006. Mutations in TMEM76* cause mucopolysaccharidosis IIIC (Sanfilippo C syndrome). *Am. J. Hum. Genet.* 79: 807-819.
6. Fedele, A.O., et al. 2007. Mutational analysis of the HGSNAT gene in Italian patients with mucopolysaccharidosis IIIC (Sanfilippo C syndrome). Mutation in brief #959. Online. *Hum. Mutat.* 28: 523.

CHROMOSOMAL LOCATION

Genetic locus: HGSNAT (human) mapping to 8p11.21.

PRODUCT

HGSNAT 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 HGSNAT shRNA Plasmid (h): sc-75251-SH and HGSNAT shRNA (h) Lentiviral Particles: sc-75251-V as alternate gene silencing products.

For independent verification of HGSNAT (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-75251A, sc-75251B and sc-75251C.

PROTOCOLS

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

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

HGSNAT siRNA (h) is recommended for the inhibition of HGSNAT 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.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor HGSNAT gene expression knockdown using RT-PCR Primer: HGSNAT (h)-PR: sc-75251-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.