SANTA CRUZ BIOTECHNOLOGY, INC.

sulfamidase siRNA (h): sc-76607

BACKGROUND

Sulfatases are enzymes that hydrolyse a diverse range of sulfate esters. Deficiency of lysosomal sulfatases leads to human diseases characterized by the accumulation of either GAGs (glycosaminoglycans) or sulfolipids. Sulfamidase, also known as HSS, SFMD, MPS3A or SGSH, is a 502 amino acid lysosome that belongs to the sulfatase family. It has been suggested that sulfamidase may be involved in the lysosomal degradation of heparan sulfate. Defects in the gene encoding sulfamidase are the cause of Sanfilippo syndrome A, an autosomal recessive lysosomal storage disease caused by impaired degradation of heparan sulfate. Sanfilippo syndrome A is characterized by severe central nervous system degeneration but relatively mild somatic manifestations.

REFERENCES

1. Scott, H.S., et al. 1995. Cloning of the sulphamidase gene and identification of mutations in Sanfilippo A syndrome. Nat. Genet. 11: 465-467.
2. Blanch, L., et al. 1997. Molecular defects in Sanfilippo syndrome type A. Hum. Mol. Genet. 6: 787-791.
3. Perkins, K.J., et al. 1999. Expression and characterization of wild type and mutant recombinant human sulfamidase. Implications for Sanfilippo (mucopolysaccharidosis IIIA) syndrome. J. Biol. Chem. 274: 37193-37199.
4. Muschol, N., et al. 2004. Transport, enzymatic activity, and stability of mutant sulfamidase (SGSH) identified in patients with mucopolysaccharidosis type III A. Hum. Mutat. 23: 559-566.
5. Mason, K.E., et al. 2006. Characterization of sulfated oligosaccharides in mucopolysaccharidosis type IIIA by electrospray ionization mass spectrometry. Anal. Chem. 78: 4534-4542.
6. Di Natale, P., et al. 2006. Gene symbol: SGSH. Disease: Sanfilippo type A syndrome, mucopolysaccharidosis IIIA. Hum. Genet. 119: 679.

CHROMOSOMAL LOCATION

Genetic locus: SGSH (human) mapping to 17q25.3.

PRODUCT

sulfamidase 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 µM solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see sulfamidase shRNA Plasmid (h): sc-76607-SH and sulfamidase shRNA (h) Lentiviral Particles: sc-76607-V as alternate gene silencing products.

For independent verification of sulfamidase (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-76607A, sc-76607B and sc-76607C.

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 µl of the RNAse-free water provided. Resuspension of the siRNA duplex in 330 µl of RNAse-free water makes a 10 µM solution in a 10 µM Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

sulfamidase siRNA (h) is recommended for the inhibition of sulfamidase 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 µM in 66 µl. 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 sulfamidase gene expression knockdown using RT-PCR Primer: sulfamidase (h)-PR: sc-76607-PR (20 µl). 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.