

HPS-4 siRNA (h): sc-44420

BACKGROUND

Hermansky-Pudlak syndrome (HPS) is a rare, genetically heterogeneous, autosomal recessive disorder. It is characterized by oculocutaneous albinism, lysosomal storage defects and prolonged bleeding due to platelet storage pool deficiency. There are ten HPS genes encoding HPS proteins that all interact within three distinct ubiquitously expressed protein complexes or biogenesis of lysosome-related organelle complexes. Defects in these genes cause HPS. HPS-4, also designated light-ear protein homolog, is important in organelle biosynthesis. Defects in the gene encoding for the HSP-4 protein can cause Hermansky-Pudlak syndrome 4 (HPS-4).

REFERENCES

1. Hirosawa, M., et al. 2001. Identification of novel transcribed sequences on human chromosome 22 by expressed sequence tag mapping. *DNA Res.* 8: 1-9.
2. Suzuki, T., et al. 2002. Hermansky-Pudlak syndrome is caused by mutations in HPS-4, the human homolog of the mouse light-ear gene. *Nat. Genet.* 30: 321-324.
3. Huizing, M., et al. 2002. Hermansky-Pudlak syndrome: vesicle formation from yeast to man. *Pigment Cell Res.* 15: 405-419.
4. Nazarian, R., et al. 2003. Biogenesis of lysosome-related organelles complex 3 (BLOC-3): a complex containing the Hermansky-Pudlak syndrome (HPS) proteins HPS-1 and HPS-4. *Proc. Natl. Acad. Sci. USA* 100: 8770-8775.
5. Anderson, P.D., et al. 2003. Hermansky-Pudlak syndrome type 4 (HPS-4): clinical and molecular characteristics. *Hum. Genet.* 113: 10-17.
6. Bachli, E.B., et al. 2004. Hermansky-Pudlak syndrome type 4 in a patient from Sri Lanka with pulmonary fibrosis. *Am. J. Med. Genet. A* 127A: 201-207.

CHROMOSOMAL LOCATION

Genetic locus: HPS4 (human) mapping to 22q12.1.

PRODUCT

HPS-4 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 HPS-4 shRNA Plasmid (h): sc-44420-SH and HPS-4 shRNA (h) Lentiviral Particles: sc-44420-V as alternate gene silencing products.

For independent verification of HPS-4 (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-44420A, sc-44420B and sc-44420C.

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.

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

HPS-4 siRNA (h) is recommended for the inhibition of HPS-4 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

HPS-4 (A-6): sc-398070 is recommended as a control antibody for monitoring of HPS-4 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 Marker™ 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 HPS-4 gene expression knockdown using RT-PCR Primer: HPS-4 (h)-PR: sc-44420-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.