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HPS-1 siRNA (h): sc-44418

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 10 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. HSP-1 is a component of multiple cytoplasmic organelles and is important for their normal development and function. It plays a role in intracellular protein sorting.

REFERENCES

- Oh, J., et al. 1996. Positional cloning of a gene for Hermansky-Pudlak syndrome, a disorder of cytoplasmic organelles. *Nat. Genet.* 14: 300-306.
- Bailin, T., et al. 1997. Organization and nucleotide sequence of the human Hermansky-Pudlak syndrome (HPS) gene. *J. Invest. Dermatol.* 108: 923-927.
- Wildenberg, S.C., et al. 1998. Identification of a novel transcript produced by the gene responsible for the Hermansky-Pudlak syndrome in Puerto Rico. *J. Invest. Dermatol.* 110: 777-781.
- Oh, J., et al. 1998. Mutation analysis of patients with Hermansky-Pudlak syndrome: a frameshift hot spot in the HPS gene and apparent locus heterogeneity. *Am. J. Hum. Genet.* 62: 593-598.
- Oetting, W.S., et al. 1999. Molecular basis of albinism: mutations and polymorphisms of pigmentation genes associated with albinism. *Hum. Mutat.* 13: 99-115.
- Di Pietro, S.M., et al. 2004. Characterization of BLOC-2, a complex containing the Hermansky-Pudlak syndrome proteins HPS-3, HPS-5 and HPS-6. *Traffic* 5: 276-283.

CHROMOSOMAL LOCATION

Genetic locus: HPS1 (human) mapping to 10q24.2.

PRODUCT

HPS-1 siRNA (h) is a target-specific 19-25 nt siRNA 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-1 shRNA Plasmid (h): sc-44418-SH and HPS-1 shRNA (h) Lentiviral Particles: sc-44418-V as alternate gene silencing 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-1 siRNA (h) is recommended for the inhibition of HPS-1 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-1 (5G12G2): sc-101435 is recommended as a control antibody for monitoring of HPS-1 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-1 gene expression knockdown using RT-PCR Primer: HPS-1 (h)-PR: sc-44418-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

SELECE PRODUCT CITATIONS

- Ahuja, S., et al. 2015. MAP1LC3B overexpression protects against Hermansky-Pudlak syndrome type-1 induced defective autophagy *in vitro*. *Am. J. Physiol. Lung Cell. Mol. Physiol.* 310: L519-L531.

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.