

HCS siRNA (m): sc-72199

BACKGROUND

Holocarboxylase synthetase (HCS, HLCS) catalyzes the posttranslational covalent attachment of biotin to biotin-dependent carboxylases. These carboxylases include pyruvate carboxylase, propionyl coenzyme A (CoA) carboxylase, β -methylcrotonoyl CoA carboxylase, and acetyl CoA carboxylase. HCS is an obligate participant in biotin-mediated regulation of its own expression and of biotin-dependent carboxylase mRNA levels in human cells. In addition, biotin regulates the genetic expression of HCS and mitochondrial carboxylases. Multiple carboxylase deficiency (MCD) is a rare metabolic life-threatening disease accountable for early onset biotin-responsive multiple carboxylase deficiency. MCD presents with lactic acidosis, tachypnea, temperature instability, and shock in neonates. Interestingly, MCD can be detected prenatally and, if diagnosed, prenatally administered biotin is effectively taken up by the fetus and prevents functional deficiency of the carboxylases in an affected newborn. The HCS gene maps to human chromosome 21q22.13 and encodes a cytoplasmic and mitochondrial protein.

REFERENCES

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2. Squires, L., et al. 1997. Resolution of subependymal cysts in neonatal holocarboxylase synthetase deficiency. *Dev. Med. Child Neurol.* 39: 267-269.
3. Suormala, T., et al. 1998. Late-onset holocarboxylase synthetase-deficiency: pre- and post-natal diagnosis and evaluation of effectiveness of antenatal biotin therapy. *Eur. J. Pediatr.* 157: 570-575.
4. Hwu, W.L., et al. 2000. Late-onset holocarboxylase synthetase deficiency with homologous R508W mutation. *J. Formos. Med. Assoc.* 99: 174-177.
5. Rodriguez-Melendez, R., et al. 2001. Biotin regulates the genetic expression of holocarboxylase synthetase and mitochondrial carboxylases in rats. *J. Nutr.* 131: 1909-1913.
6. Yang, X., et al. 2001. Structure of human holocarboxylase synthetase gene and mutation spectrum of holocarboxylase synthetase deficiency. *Hum. Genet.* 109: 526-534.

CHROMOSOMAL LOCATION

Genetic locus: Hlcs (mouse) mapping to 16 C4.

PRODUCT

HCS 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 HCS shRNA Plasmid (m): sc-72199-SH and HCS shRNA (m) Lentiviral Particles: sc-72199-V as alternate gene silencing products.

For independent verification of HCS (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-72199A, sc-72199B and sc-72199C.

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

HCS siRNA (m) is recommended for the inhibition of HCS 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 HCS gene expression knockdown using RT-PCR Primer: HCS (m)-PR: sc-72199-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.

PROTOCOLS

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