

HCCS siRNA (h): sc-91122

BACKGROUND

HCCS (holocytochrome c-type synthase), also known as CCHL (cytochrome c-type heme lyase), is a 268 amino acid mitochondrial inner membrane protein that belongs to the cytochrome c-type heme lyase family. Containing two HRM (heme regulatory motif) repeats, HCCS participates in the covalent linkage of a heme group to an apoprotein of cytochrome c. The gene encoding HCCS maps to the human X chromosome. Defects to this gene cause microphthalmia syndromic type 7 (MCOPS7), also known as MIDAS syndrome or microphthalmia with linear skin defects (MLS). MCOPS7 is an X-linked male-lethal disorder that results in eye deformation, unilateral or bilateral microphthalmia, linear skin defects in affected females, and in utero lethality for males.

REFERENCES

1. Schaefer, L., Ballabio, A. and Zoghbi, H.Y. 1996. Cloning and characterization of a putative human holocytochrome c-type synthetase gene (HCCS) isolated from the critical region for microphthalmia with linear skin defects (MLS). *Genomics* 34: 166-172.
2. Van den Veyver, I.B., Subramanian, S. and Zoghbi, H.Y. 1998. Genomic structure of a human holocytochrome c-type synthetase gene in Xp22.3 and mutation analysis in patients with Rett syndrome. *Am. J. Med. Genet.* 78: 179-181.
3. Prakash, S.K., Cormier, T.A., McCall, A.E., Garcia, J.J., Sierra, R., Haupt, B., Zoghbi, H.Y. and Van Den Veyver, I.B. 2002. Loss of holocytochrome c-type synthetase causes the male lethality of X-linked dominant microphthalmia with linear skin defects (MLS) syndrome. *Hum. Mol. Genet.* 11: 3237-3248.
4. Wimplinger, I., Morleo, M., Rosenberger, G., Iaconis, D., Orth, U., Meinecke, P., Lerer, I., Ballabio, A., Gal, A., Franco, B. and Kutsche, K. 2006. Mutations of the mitochondrial holocytochrome c-type synthase in X-linked dominant microphthalmia with linear skin defects syndrome. *Am. J. Hum. Genet.* 79: 878-889.
5. Wimplinger, I., Shaw, G.M. and Kutsche, K. 2007. HCCS loss-of-function missense mutation in a female with bilateral microphthalmia and sclerocornea: a novel gene for severe ocular malformations? *Mol. Vis.* 13: 1475-1482.

CHROMOSOMAL LOCATION

Genetic locus: HCCS (human) mapping to Xp22.2.

PRODUCT

HCCS 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 HCCS shRNA Plasmid (h): sc-91122-SH and HCCS shRNA (h) Lentiviral Particles: sc-91122-V as alternate gene silencing products.

For independent verification of HCCS (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-91122A, sc-91122B and sc-91122C.

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

HCCS siRNA (h) is recommended for the inhibition of HCCS 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 HCCS gene expression knockdown using RT-PCR Primer: HCCS (h)-PR: sc-91122-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.