

# CTC1 siRNA (h): sc-93645

## BACKGROUND

Telomeres are DNA-protein structures that protect the ends of linear chromosomes and help maintain genomic stability and cell phenotype. CST complex plays an essential role in protecting telomeres from degradation. CTC1 (CTS telomere maintenance complex component 1), also known as CRMCC, AAF132, AAF-132, C17orf68 or tmp494178, is a 1,217 amino acid nuclear protein belonging to the CTC1 family. CTC1 along with OBFC1, also known as STN1, and TEN1 form the trimeric CST complex and is considered critical for telomere replication. Mutations in the gene encoding CST1 leads to an autosomal recessive pleiomorphic disorder that is characterized by intracranial calcifications, leukodystrophy, and brain cysts, which results in spasticity, ataxia, dystonia, seizures, and cognitive decline. Individuals afflicted with CTC1 gene mutation also may develop Coats disease, extraneurologic manifestations, osteopenia with poor bone healing and is at high risk of gastrointestinal bleeding and portal hypertension caused by vasculature ectasias in the stomach, small intestine, and liver. CTC1 exists as two alternatively spliced isoforms and is encoded by a gene located on human chromosome 17p13.1.

## REFERENCES

1. Surovtseva, Y.V., Churikov, D., Boltz, K.A., Song, X., Lamb, J.C., Warrington, R., Leehy, K., Heacock, M., Price, C.M. and Shippen, D.E. 2009. Conserved telomere maintenance component 1 interacts with STN1 and maintains chromosome ends in higher eukaryotes. *Mol. Cell* 36: 207-218.
2. Polvi, A., Linnankivi, T., Kivelä, T., Herva, R., Keating, J.P., Mäkitie, O., Pareyson, D., Vainionpää, L., Lahtinen, J., Hovatta, I., Pihko, H. and Lehesjoki, A.E. 2012. Mutations in CTC1, encoding the CTS telomere maintenance complex component 1, cause cerebroretinal microangiopathy with calcifications and cysts. *Am. J. Hum. Genet.* 90: 540-549.
3. Stewart, J.A., Wang, F., Chaiken, M.F., Kasbek, C., Chastain, P.D., Wright, W.E. and Price, C.M. 2012. Human CST promotes telomere duplex replication and general replication restart after fork stalling. *EMBO J.* 31: 3537-3549.
4. Anderson, B.H., Kasher, P.R., Mayer, J., Szykiewicz, M., Jenkinson, E.M., Bhaskar, S.S., Urquhart, J.E., Daly, S.B., Dickerson, J.E., O'Sullivan, J., Leibundgut, E.O., Muter, J., Abdel-Salem, G.M., et al. 2012. Mutations in CTC1, encoding conserved telomere maintenance component 1, cause Coats plus. *Nat. Genet.* 44: 338-342.
5. Chen, L.Y., Redon, S. and Lingner, J. 2012. The human CST complex is a terminator of telomerase activity. *Nature* 488: 540-544.
6. Keller, R.B., Gagne, K.E., Usmani, G.N., Asdourian, G.K., Williams, D.A., Hofmann, I. and Agarwal, S. 2012. CTC1 mutations in a patient with dyskeratosis congenita. *Pediatr. Blood Cancer* 59: 311-314.
7. Gu, P. and Chang, S. 2013. Functional characterization of human CTC1 mutations reveals novel mechanisms responsible for the pathogenesis of the telomere disease Coats plus. *Aging Cell* 12: 1100-1109.
8. Chen, L.Y., Majerská, J. and Lingner, J. 2013. Molecular basis of telomere syndrome caused by CTC1 mutations. *Genes Dev.* 27: 2099-2108.
9. Chen, L.Y. and Lingner, J. 2013. CST for the grand finale of telomere replication. *Nucleus* 4: 277-282.

## CHROMOSOMAL LOCATION

Genetic locus: CTC1 (human) mapping to 17p13.1.

## PRODUCT

CTC1 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  $\mu$ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see CTC1 shRNA Plasmid (h): sc-93645-SH and CTC1 shRNA (h) Lentiviral Particles: sc-93645-V as alternate gene silencing products.

For independent verification of CTC1 (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-93645A, sc-93645B and sc-93645C.

## 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  $\mu$ l of the RNase-free water provided. Resuspension of the siRNA duplex in 330  $\mu$ l of RNase-free water makes a 10  $\mu$ M solution in a 10  $\mu$ M Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

## APPLICATIONS

CTC1 siRNA (h) is recommended for the inhibition of CTC1 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  $\mu$ M in 66  $\mu$ 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 CTC1 gene expression knockdown using RT-PCR Primer: CTC1 (h)-PR: sc-93645-PR (20  $\mu$ 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](http://www.scbt.com) for detailed protocols and support products.