

COA5 siRNA (h): sc-94812

BACKGROUND

COA5 (cytochrome C oxidase assembly factor 5), also known as Pet191, is a 74 amino acid protein that plays a role in the beginning stage of the mitochondrial complex IV assembly process. A member of the Pet191 family, COA5 is encoded by a gene that maps to human chromosome 2q11.2. Defects in the COA5 gene are the cause of mitochondrial complex IV deficiency (MT-C4D), a mitochondrial respiratory chain disorder characterized by exercise intolerance, hepatomegaly, developmental delay, mental retardation and hypertrophic cardiomyopathy. As the second largest human chromosome, chromosome 2 consists of 237 million bases, encodes over 1,400 genes and makes up approximately 8% of the human genome. A number of genetic diseases are linked to genes on chromosome 2 including Harlequin ichthyosis, sitosterolemia and Alström syndrome.

REFERENCES

1. Zumsteg, U., Muller, P.Y. and Miserez, A.R. 2000. Alstrom syndrome: confirmation of linkage to chromosome 2p12-13 and phenotypic heterogeneity in three affected sibs. *J. Med. Genet.* 37: E8.
2. Shulenin, S., Schriml, L.M., Remaley, A.T., Fojo, S., Brewer, B., Allikmets, R. and Dean, M. 2001. An ATP-binding cassette gene (ABCG5) from the ABCG (white) gene subfamily maps to human chromosome 2p21 in the region of the sitosterolemia locus. *Cytogenet. Cell Genet.* 92: 204-208.
3. Hearn, T., Renforth, G.L., Spalluto, C., Hanley, N.A., Piper, K., Brickwood, S., White, C., Connolly, V., Taylor, J.F., Russell-Eggitt, I., Bonneau, D., Walker, M. and Wilson, D.I. 2002. Mutation of ALMS1, a large gene with a tandem repeat encoding 47 amino acids, causes Alström syndrome. *Nat. Genet.* 31: 79-83.
4. Tay, S.K., Nesti, C., Mancuso, M., Schon, E.A., Shanske, S., Bonilla, E., Davidson, M.M. and Dimauro, S. 2004. Studies of COX16, COX19, and PET191 in human cytochrome-c oxidase deficiency. *Arch. Neurol.* 61: 1935-1937.
5. Kelsell, D.P., Norgett, E.E., Unsworth, H., Teh, M.T., Cullup, T., Mein, C.A., Dopping-Hepenstal, P.J., Dale, B.A., Tadini, G., Fleckman, P., Stephens, K.G., Sybert, V.P., Mallory, S.B., North, B.V., Witt, D.R., Sprecher, E., et al. 2005. Mutations in ABCA12 underlie the severe congenital skin disease harlequin ichthyosis. *Am. J. Hum. Genet.* 76: 794-803.
6. Khalimonchuk, O., Rigby, K., Bestwick, M., Pierrel, F., Cobine, P.A. and Winge, D.R. 2008. Pet191 is a cytochrome c oxidase assembly factor in *Saccharomyces cerevisiae*. *Eukaryotic Cell* 7: 1427-1431.
7. Huigsloot, M., Nijtmans, L.G., Szklarczyk, R., Baars, M.J., van den Brand, M.A., Hendriksfranssen, M.G., van den Heuvel, L.P., Smeitink, J.A., Huynen, M.A. and Rodenburg, R.J. 2011. A mutation in C2orf64 causes impaired cytochrome c oxidase assembly and mitochondrial cardiomyopathy. *Am. J. Hum. Genet.* 88: 488-493.

CHROMOSOMAL LOCATION

Genetic locus: COA5 (human) mapping to 2q11.2.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PRODUCT

COA5 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 COA5 shRNA Plasmid (h): sc-94812-SH and COA5 shRNA (h) Lentiviral Particles: sc-94812-V as alternate gene silencing products.

For independent verification of COA5 (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-94812A, sc-94812B and sc-94812C.

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

COA5 siRNA (h) is recommended for the inhibition of COA5 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 COA5 gene expression knockdown using RT-PCR Primer: COA5 (h)-PR: sc-94812-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

PROTOCOLS

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