

MCEE siRNA (m): sc-149318

BACKGROUND

MCEE (methylmalonyl CoA epimerase), also known as GLOD2 or DL-methylmalonyl-CoA racemase, is a 176 amino acid mitochondrial protein that belongs to the glyoxalase I family. MCEE catalyzes the interconversion of D- and L-methylmalonyl-CoA during the breakdown of branched chain amino acids. The gene encoding MCEE maps to human chromosome 2p13.3. Defects in the MCEE gene are the cause of an autosomal recessive disease known as methylmalonyl-CoA epimerase deficiency (MCEE deficiency), methylmalonyl-CoA racemase deficiency or methylmalonic aciduria type 3, which is characterized by mild to moderate methylmalonic aciduria.

REFERENCES

1. Montgomery, J.A., Mamer, O.A. and Scriver, C.R. 1983. Metabolism of methylmalonic acid in rats. Is methylmalonyl-coenzyme a racemase deficiency symptomatic in man? *J. Clin. Invest.* 72: 1937-1947.
2. Bobik, T.A. and Rasche, M.E. 2001. Identification of the human methylmalonyl-CoA racemase gene based on the analysis of prokaryotic gene arrangements. Implications for decoding the human genome. *J. Biol. Chem.* 276: 37194-37198.
3. Bikker, H., Bakker, H.D., Abeling, N.G., Poll-The, B.T., Kleijer, W.J., Rosenblatt, D.S., Waterham, H.R., Wanders, R.J. and Duran, M. 2006. A homozygous nonsense mutation in the methylmalonyl-CoA epimerase gene (MCEE) results in mild methylmalonic aciduria. *Hum. Mutat.* 27: 640-643.
4. Dobson, C.M., Gradingier, A., Longo, N., Wu, X., Leclerc, D., Lerner-Ellis, J., Lemieux, M., Belair, C., Watkins, D., Rosenblatt, D.S. and Gravel, R.A. 2006. Homozygous nonsense mutation in the MCEE gene and siRNA suppression of methylmalonyl-CoA epimerase expression: a novel cause of mild methylmalonic aciduria. *Mol. Genet. Metab.* 88: 327-333.
5. Chandler, R.J., Aswani, V., Tsai, M.S., Falk, M., Wehrli, N., Stabler, S., Allen, R., Sedensky, M., Kazazian, H.H. and Venditti, C.P. 2006. Propionyl-CoA and adenosylcobalamin metabolism in *Caenorhabditis elegans*: evidence for a role of methylmalonyl-CoA epimerase in intermediary metabolism. *Mol. Genet. Metab.* 89: 64-73.
6. Gradingier, A.B., Belair, C., Worgan, L.C., Li, C.D., Lavalley, J., Roquis, D., Watkins, D. and Rosenblatt, D.S. 2007. Atypical methylmalonic aciduria: frequency of mutations in the methylmalonyl CoA epimerase gene (MCEE). *Hum. Mutat.* 28: 1045.
7. Online Mendelian Inheritance in Man, OMIM[™]. 2009. Johns Hopkins University, Baltimore, MD. MIM Number: 608419. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>

CHROMOSOMAL LOCATION

Genetic locus: Mcee (mouse) mapping to 7 C.

PROTOCOLS

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

PRODUCT

MCEE siRNA (m) is a pool of 2 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 MCEE shRNA Plasmid (m): sc-149318-SH and MCEE shRNA (m) Lentiviral Particles: sc-149318-V as alternate gene silencing products.

For independent verification of MCEE (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-149318A and sc-149318B.

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

MCEE siRNA (m) is recommended for the inhibition of MCEE 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 MCEE gene expression knockdown using RT-PCR Primer: MCEE (m)-PR: sc-149318-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.