SANTA CRUZ BIOTECHNOLOGY, INC.

LMBRD2 siRNA (h): sc-91692

BACKGROUND

Vitamin B12 (cobalamin) is essential in animals and humans for metabolism of methylmalonic acid, for the remethylation of homocysteine to methionine and, consequently, for all S-adenosylmethionine-dependent methylation reactions, including DNA synthesis. The lysosomal cobalamin transporter is required for the export cobalamin from lysosomes allowing its conversion to cofactors. Defects in LMBRD1 are the cause of methylmalonic aciduria and homocystinuria type cblF (MMAFHC), also known as homocystinuria-megaloblastic anemia complementation type F. MMAFHC is a disorder of cobalamin metabolism characterized by decreased levels of the coenzymes adenosylcobalamin (AdoCbl) and methylcobalamin (MeCbl) due to accumulation of cobalamin in lysosomes. Clinical features of MMAFHC include developmental delay, stomatitis, glossitis, seizures and methylmalonic aciduria in response to vitamin B12. LMBRD2 (LMBR1 domain containing 2) is a 695 amino acid multi-membrane protein that may have similar functions as LMBR1.

REFERENCES

1. Watkins, D. and Rosenblatt, D.S. 1986. Failure of lysosomal release of vitamin B12: a new complementation group causing methylmalonic aciduria (cblF). Am. J. Hum. Genet. 39: 404-408.
2. Rosenblatt, D.S. and Cooper, B.A. 1987. Inherited disorders of vitamin B12 metabolism. Blood Rev. 1: 177-182.
3. Vassiliadis, A., Rosenblatt, D.S., Cooper, B.A. and Bergeron, J.J. 1991. Lysosomal cobalamin accumulation in fibroblasts from a patient with an inborn error of cobalamin metabolism (cblF complementation group): visualization by electron microscope radioautography. Exp. Cell Res. 195: 295-302.
4. Laframboise, R., Cooper, B.A. and Rosenblatt, D.S. 1992. Malabsorption of vitamin B12 from the intestine in a child with cblF disease: evidence for lysosomal-mediated absorption. Blood 80: 291-292.
5. Waggoner, D.J., Ueda, K., Mantia, C. and Dowton, S.B. 1998. Methylmalonic aciduria (cblF): case report and response to therapy. Am. J. Med. Genet. 79: 373-375.
6. Wang, Y.H., Chang, S.C., Huang, C., Li, Y.P., Lee, C.H. and Chang, M.F. 2005. Novel nuclear export signal-interacting protein, NESI, critical for the assembly of hepatitis delta virus. J. Virol. 79: 8113-8120.
7. Rutsch, F., Gailus, S., Miousse, I.R., Suormala, T., Sagnε, C., Toliat, M.R., Nürnberg, G., Wittkampf, T., Buers, I., Sharifi, A., Stucki, M., Becker, C., Baumgartner, M., Robenek, H., Marquardt, T., Höhne, W., et al. 2009. Identification of a putative lysosomal cobalamin exporter altered in the cblF defect of vitamin B12 metabolism. Nat. Genet. 41: 234-239.

CHROMOSOMAL LOCATION

Genetic locus: LMBRD2 (human) mapping to 5p13.2.

PROTOCOLS

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

PRODUCT

LMBRD2 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 µM solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see LMBRD2 shRNA Plasmid (h): sc-91692-SH and LMBRD2 shRNA (h) Lentiviral Particles: sc-91692-V as alternate gene silencing products.

For independent verification of LMBRD2 (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-91692A, sc-91692B and sc-91692C.

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 µl of the RNAse-free water provided. Resuspension of the siRNA duplex in 330 µl of RNAse-free water makes a 10 µM solution in a 10 µM Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

LMBRD2 siRNA (h) is recommended for the inhibition of LMBRD2 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 µM in 66 µl. 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 LMBRD2 gene expression knockdown using RT-PCR Primer: LMBRD2 (h)-PR: sc-91692-PR (20 µl). 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.