SANTA CRUZ BIOTECHNOLOGY, INC.

MPV17 siRNA (h): sc-94662

BACKGROUND

MPV17, also known as SYM1, is a 176 amino acid mitochondrial inner membrane protein that belongs to the peroxisomal membrane protein PXMP2/4 family. MPV17 is expressed in pancreas, kidney, muscle, liver, lung, placenta, brain and heart. MPV17 plays an important role in regulating oxidative phosphorylation and mitochondrial DNA (mtDNA) maintenance. Mutations of MVP17 have been associated with the hepatocerebral form of mitochondrial DNA depletion syndrome (MDDS). MDDS is an autosomal recessive trait characterized by a reduction in mitochondrial DNA (mtDNA) copy number. MDDS may affect single organs, typically muscle or liver. Individuals with the hepatocerebral form of MDDS have early progressive liver failure and neurologic abnormalities, hypoglycemia, and increased lactate in body fluids.

REFERENCES

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2. Meyer zum Gottesberge, A.M., et al. 1996. Inner ear defect similar to Alport's syndrome in the glomerulosclerosis mouse model MPV17. Eur. Arch. Otorhinolaryngol. 253: 470-474.
3. Karadimas, C.L., et al. 2006. Navajo neurohepatopathy is caused by a mutation in the MPV17 gene. Am. J. Hum. Genet. 79: 544-548.
4. Wong, L.J., et al. 2007. Mutations in the MPV17 gene are responsible for rapidly progressive liver failure in infancy. Hepatology 46: 1218-1227.
5. Spinazzola, A., et al. 2008. Hepatocerebral form of mitochondrial DNA depletion syndrome: novel MPV17 mutations. Arch. Neurol. 65: 1108-1113.
6. Navarro-Sastre, A., et al. 2008. Lethal hepatopathy and leukodystrophy caused by a novel mutation in MPV17 gene: description of an alternative MPV17 spliced form. Mol. Genet. Metab. 94: 234-239.
7. Spinazzola, A., et al. 2008. Lack of founder effect for an identical mtDNA depletion syndrome (MDS)-associated MPV17 mutation shared by Navajos and Italians. Neuromuscul. Disord. 18: 315-318.
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CHROMOSOMAL LOCATION

Genetic locus: MPV17 (human) mapping to 2p23.3.

PRODUCT

MPV17 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 MPV17 shRNA Plasmid (h): sc-94662-SH and MPV17 shRNA (h) Lentiviral Particles: sc-94662-V as alternate gene silencing products.

For independent verification of MPV17 (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-94662A, sc-94662B and sc-94662C.

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

MPV17 siRNA (h) is recommended for the inhibition of MPV17 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 MPV17 gene expression knockdown using RT-PCR Primer: MPV17 (h)-PR: sc-94662-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.

PROTOCOLS

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