

HMGCL siRNA (h): sc-78794

BACKGROUND

HMGCL (3-hydroxy-3-methylglutaryl-coenzyme A (CoA) lyase), also known as HMG-CoA lyase or HL, is a mitochondrial matrix protein that belongs to the HMG-CoA lyase family of proteins. Expressed in liver, lymphoblasts and fibroblasts, HMGCL exists as a homodimer and participates in leucine catabolism and ketogenesis, the hepatic synthesis of ketone bodies that, during fasting, provide a major source of energy for heart, brain and kidney. More specifically, HMGCL catalyzes the final step of these processes, the cleavage of 3-hydroxy-3-methylglutaryl CoA to acetoacetic acid and acetyl CoA. Mutations in the gene encoding HMGCL can lead to HMG-CoA lyase deficiency (also known as HL deficiency or hydroxymethylglutaricaciduria), a metabolic disease that, if left untreated, results in hypoglycemia and coma.

REFERENCES

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2. Wang, S.P., et al. 1996. 3-hydroxy-3-methylglutaryl CoA lyase (HL): mouse and human HL gene (HMGCL) cloning and detection of large gene deletions in two un-related HL-deficient patients. *Genomics* 33: 99-104.
3. Funghini, S., et al. 2001. 3-hydroxy-3-methylglutaric aciduria in an Italian patient is caused by a new nonsense mutation in the HMGCL gene. *Mol. Genet. Metab.* 73: 268-275.
4. Kim, S., et al. 2004. Hepatic gene expression profiles in a long-term high-fat diet-induced obesity mouse model. *Gene* 340: 99-109.
5. Cardoso, M.L., et al. 2004. The E37X is a common HMGCL mutation in Portuguese patients with 3-hydroxy-3-methylglutaric CoA lyase deficiency. *Mol. Genet. Metab.* 82: 334-338.
6. Al-Sayed, M., et al. 2006. Mutations underlying 3-hydroxy-3-methylglutaryl CoA lyase deficiency in the Saudi population. *BMC Med. Genet.* 7: 86.
7. Alsmadi, O., et al. 2006. LCGreen I-based real-time PCR assays for detecting common ASL and HMGCL variants. *Clin. Chem.* 52: 1439-1440.

CHROMOSOMAL LOCATION

Genetic locus: HMGCL (human) mapping to 1p36.11.

PRODUCT

HMGCL 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 HMGCL shRNA Plasmid (h): sc-78794-SH and HMGCL shRNA (h) Lentiviral Particles: sc-78794-V as alternate gene silencing products.

For independent verification of HMGCL (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-78794A, sc-78794B and sc-78794C.

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

HMGCL siRNA (h) is recommended for the inhibition of HMGCL 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.

GENE EXPRESSION MONITORING

HMGCL (63Z): sc-100548 is recommended as a control antibody for monitoring of HMGCL gene expression knockdown by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) or immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor HMGCL gene expression knockdown using RT-PCR Primer: HMGCL (h)-PR: sc-78794-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.

PROTOCOLS

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