

ACADVL siRNA (h): sc-72427

BACKGROUND

ACADVL (acyl-Coenzyme A dehydrogenase, very long chain), also known as VLCAD, LCACD or ACAD6, is an inner mitochondrial membrane protein that belongs to the family of acyl-CoA dehydrogenases. Involved in lipid metabolism, ACADVL has catalytic activity toward esters of long chain and very long chain fatty acids and functions in the first step of the fatty acid β -oxidation pathway. Defects in the gene encoding ACADVL are the cause of very long chain acyl-CoA dehydrogenase deficiency (VLCAD deficiency), an autosomal recessive disease that leads to impaired long-chain fatty acid oxidation. VLCAD deficiency can give rise to three different phenotypes: a severe childhood form with a high incidence of mortality and cardiomyopathy; a mild childhood form with hypoketotic hypoglycemia and low mortality rates; and an adult form characterized by myoglobinuria and rhabdomyolysis. Two isoforms of ACADVL exist due to alternative splicing events.

REFERENCES

1. Mathur, A., et al. 1999. Molecular heterogeneity in very-long-chain acyl-CoA dehydrogenase deficiency causing pediatric cardiomyopathy and sudden death. *Circulation* 99: 1337-1343.
2. Andresen, B.S., et al. 1999. Clear correlation of genotype with disease phenotype in very-long-chain acyl-CoA dehydrogenase deficiency. *Am. J. Hum. Genet.* 64: 479-494.
3. Tong, M.K., et al. 2006. Very long-chain acyl-CoA dehydrogenase deficiency presenting as acute hypercapnic respiratory failure. *Eur. Respir. J.* 28: 447-450.
4. Gobin-Limballe, S., et al. 2007. Genetic basis for correction of very-long-chain acyl-coenzyme A dehydrogenase deficiency by bezafibrate in patient fibroblasts: toward a genotype-based therapy. *Am. J. Hum. Genet.* 81: 1133-1143.
5. Spiekerkoetter, U. 2007. Effects of a fat load and exercise on asymptomatic VLCAD deficiency. *J. Inherit. Metab. Dis.* 30: 405-405.

CHROMOSOMAL LOCATION

Genetic locus: ACADVL (human) mapping to 17p13.1.

PRODUCT

ACADVL 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 ACADVL shRNA Plasmid (h): sc-72427-SH and ACADVL shRNA (h) Lentiviral Particles: sc-72427-V as alternate gene silencing products.

For independent verification of ACADVL (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-72427A, sc-72427B and sc-72427C.

PROTOCOLS

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

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

ACADVL siRNA (h) is recommended for the inhibition of ACADVL 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

ACADVL (H-7): sc-376239 is recommended as a control antibody for monitoring of ACADVL 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 ACADVL gene expression knockdown using RT-PCR Primer: ACADVL (h)-PR: sc-72427-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.