



# D2HGDH siRNA (m): sc-142809

## BACKGROUND

D2HGDH (D-2-hydroxyglutarate dehydrogenase), also known as FLJ42195 or MGC25181, is a 521 amino acid member of the FAD-binding oxidoreductase/transferase type 4 protein family. Localized to mitochondria, D2HGDH is activated by cobalt and zinc and utilizes FAD as a cofactor. D2HGDH catalyzes the oxidation of D-2-hydroxyglutarate, resulting in  $\alpha$ -ketoglutarate. Defects in the gene that encodes D2HGDH are the cause of D-2-hydroxyglutaric aciduria (D2HGA), a rare recessive neurometabolic disorder characterized by early infantile-onset epileptic encephalopathy and cardiomyopathy. D2HGA causes developmental delay, hypotonia, epilepsy and dysmorphic features. D2HGDH contains one FAD-binding PCMH-type domain and is expressed as two isoforms produced by alternative splicing.

## REFERENCES

1. Gibson, K.M., et al. 1993. D-2-hydroxyglutaric aciduria in a newborn with neurological abnormalities: a new neurometabolic disorder? *J. Inher. Metab. Dis.* 16: 497-500.
2. Achouri, Y., et al. 2004. Identification of a dehydrogenase acting on D-2-hydroxyglutarate. *Biochem. J.* 381: 35-42.
3. Struys, E.A., et al. 2005. Mutations in the D-2-hydroxyglutarate dehydrogenase gene cause D-2-hydroxyglutaric aciduria. *Am. J. Hum. Genet.* 76: 358-360.
4. Struys, E.A., et al. 2005. Mutations in phenotypically mild D-2-hydroxyglutaric aciduria. *Ann. Neurol.* 58: 626-630.
5. Misra, V.K., et al. 2005. Phenotypic heterogeneity in the presentation of D-2-hydroxyglutaric aciduria in monozygotic twins. *Mol. Genet. Metab.* 86: 200-205.
6. Struys, E.A., et al. 2006. D-2-hydroxyglutaric aciduria in three patients with proven SSADH deficiency: genetic coincidence or a related biochemical epiphenomenon? *Mol. Genet. Metab.* 88: 53-57.
7. Wickenhagen, W.V., et al. 2009. Measurement of D-2-hydroxyglutarate dehydrogenase activity in cell homogenates derived from D-2-hydroxyglutaric aciduria patients. *J. Inher. Metab. Dis.* 32: 264-268.

## CHROMOSOMAL LOCATION

Genetic locus: D2hgdh (mouse) mapping to 1 D.

## PRODUCT

D2HGDH siRNA (m) is a target-specific 19-25 nt siRNA designed to knock down gene expression. Each vial contains 3.3 nmol of lyophilized siRNA, sufficient for a 10  $\mu$ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see D2HGDH shRNA Plasmid (m): sc-142809-SH and D2HGDH shRNA (m) Lentiviral Particles: sc-142809-V as alternate gene silencing products.

## PROTOCOLS

See our web site at [www.scbt.com](http://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  $\mu$ l of the RNase-free water provided. Resuspension of the siRNA duplex in 330  $\mu$ l of RNase-free water makes a 10  $\mu$ M solution in a 10  $\mu$ M Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

## APPLICATIONS

D2HGDH siRNA (m) is recommended for the inhibition of D2HGDH 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  $\mu$ M in 66  $\mu$ 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 D2HGDH gene expression knockdown using RT-PCR Primer: D2HGDH (m)-PR: sc-142809-PR (20  $\mu$ 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.