

# GCSH siRNA (h): sc-93301

## BACKGROUND

GCSH (glycine cleavage system protein H (aminomethyl carrier)), also known as GCE or NKH, is a 173 amino acid mitochondrial protein that contains one lipoyl-binding domain and belongs to the gcvH family. Defects in the gene encoding GCSH are the cause of glycine encephalopathy (GCE), an autosomal recessive disease that is also referred to as non-ketotic hyperglycinemia (NKH). Characterized by severe neurological symptoms, patients with GCE have a large amount of glycine accumulated in their body fluids. The gene encoding GCSH maps to human chromosome 16, which encodes over 900 genes and comprises nearly 3% of the human genome.

## REFERENCES

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4. Koyata, H., et al. 1991. The glycine cleavage system: structure of a cDNA encoding human H-protein, and partial characterization of its gene in patients with hyperglycinemias. *Am. J. Hum. Genet.* 48: 351-361.
5. Fujiwara, K., et al. 1991. The primary structure of human H-protein of the glycine cleavage system deduced by cDNA cloning. *Biochem. Biophys. Res. Commun.* 176: 711-716.
6. Sakata, Y., et al. 2001. Structure and expression of the glycine cleavage system in rat central nervous system. *Brain Res. Mol. Brain Res.* 94: 119-130.
7. Kure, S., et al. 2001. Chromosomal localization, structure, single-nucleotide polymorphisms, and expression of the human H-protein gene of the glycine cleavage system (GCSH), a candidate gene for nonketotic hyperglycinemia. *J. Hum. Genet.* 46: 378-384.
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## CHROMOSOMAL LOCATION

Genetic locus: GCSH (human) mapping to 16q23.2.

## PRODUCT

GCSH siRNA (h) is a pool of 2 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  $\mu$ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see GCSH shRNA Plasmid (h): sc-93301-SH and GCSH shRNA (h) Lentiviral Particles: sc-93301-V as alternate gene silencing products.

For independent verification of GCSH (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-93301A and sc-93301B.

## 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

GCSH siRNA (h) is recommended for the inhibition of GCSH 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  $\mu$ 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 GCSH gene expression knockdown using RT-PCR Primer: GCSH (h)-PR: sc-93301-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.

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) for detailed protocols and support products.