



# biotinidase siRNA (h): sc-78477

## BACKGROUND

Biotin, also known as vitamin B7, is an essential water-soluble vitamin that is a cofactor in glucogenesis and in the metabolism of fatty acids and leucine. Biotinidase is a 523 amino acid enzyme that catalyzes the hydrolysis of biocytin to biotin and lysine. Secreted into extracellular space, biotinidase is expressed in liver, heart, placenta, brain, skeletal muscle, pancreas and kidney. Biotinidase contains one carbon-nitrogen hydrolase domain, which is involved in the reduction of organic nitrogen compounds and ammonia production. Defects in the gene encoding biotinidase are the cause of biotinidase deficiency, which is characterized by skin rash, ataxia, seizures, hearing loss, hypotonia and optic atrophy. These symptoms are due to the individual's inability to reutilize biotin and can, therefore, typically be treated with the addition of free biotin.

## REFERENCES

1. Cole, H., et al. 1994. Human serum biotinidase. cDNA cloning, sequence, and characterization. *J. Biol. Chem.* 269: 6566-6570.
2. Pomponio, R.J., et al. 1997. Arg 538 to Cys mutation in a CpG dinucleotide of the human biotinidase gene is the second most common cause of profound biotinidase deficiency in symptomatic children. *Hum. Genet.* 99: 506-512.
3. Swango, K.L., et al. 1998. Partial biotinidase deficiency is usually due to the D444H mutation in the biotinidase gene. *Hum. Genet.* 102: 571-575.
4. Norrgard, K.J., et al. 1998. Double mutation (A171T and D444H) is a common cause of profound biotinidase deficiency in children ascertained by newborn screening the the United States. *Mutations in brief no. 128.* Online. *Hum. Mutat.* 11: 410.
5. Knight, H.C., et al. 1998. Structure of the human biotinidase gene. *Mamm. Genome* 9: 327-330.
6. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 609019. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
7. Hassan, Y.I. and Zemleni, J. 2006. Epigenetic regulation of chromatin structure and gene function by biotin. *J. Nutr.* 136: 1763-1765.

## CHROMOSOMAL LOCATION

Genetic locus: BTB (human) mapping to 3p25.1.

## PRODUCT

biotinidase 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  $\mu$ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see biotinidase shRNA Plasmid (h): sc-78477-SH and biotinidase shRNA (h) Lentiviral Particles: sc-78477-V as alternate gene silencing products.

For independent verification of biotinidase (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-78477A, sc-78477B and sc-78477C.

## 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

biotinidase siRNA (h) is recommended for the inhibition of biotinidase 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 biotinidase gene expression knockdown using RT-PCR Primer: biotinidase (h)-PR: sc-78477-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.