SANTA CRUZ BIOTECHNOLOGY, INC.

PHKB siRNA (h): sc-93503

BACKGROUND

Phosphorylase kinase is a hexadecameric enzyme that is comprised of four copies of four subunits that are encoded by four separate genes: PHKA, PHKB, PHKG, and PHKD. This serine/threonine specific kinase converts glycogen phosphorylase b to glycogen phosphorylase a, resulting in the release of glucose-1-phophate from glycogen. PHKB (Phosphorylase β kinase regulatory subunit beta) is a 1,093 amino acid subunit of phosphorylase kinase that, along with PHKA, has regulatory functions controlled by phosphorylation. Defects in the gene encoding PHKB are the cause of glycogen storage disease type 9B, which is also known as phosphorylase kinase deficiency of liver and muscle. This disease is characterized by a mild phenotype of hepatomegaly with only slightly elevated transaminase and plasma lipids, no clinical muscle involvement, and generally is correlated with a gradual improvement with increasing age. There are four isoforms of PHKB that are produced as a result of alternative splicing events.

REFERENCES

1. Wüllrich-Schmoll, A. and Kilimann, M.W. 1996. Structure of the human gene encoding the phosphorylase kinase β subunit (PHKB). Eur. J. Biochem. 238: 374-380.
2. van den Berg, I.E., et al. 1997. Autosomal recessive phosphorylase kinase deficiency in liver, caused by mutations in the gene encoding the β subunit (PHKB). Am. J. Hum. Genet. 61: 539-546.
3. Burwinkel, B., et al. 1997. Phosphorylase-kinase-deficient liver glycogenosis with an unusual biochemical phenotype in blood cells associated with a missense mutation in the β subunit gene (PHKB). Hum. Genet. 101: 170-174.
4. Burwinkel, B., et al. 2003. Muscle glycogenosis with low phosphorylase kinase activity: mutations in PHKA1, PHKG1 or six other candidate genes explain only a minority of cases. Eur. J. Hum. Genet. 11: 516-526.
5. Burwinkel, B., et al. 2003. Severe phenotype of phosphorylase kinase-deficient liver glycogenosis with mutations in the PHKG2 gene. Pediatr. Res. 54: 834-839.
6. Beauchamp, N.J., et al. 2007. Glycogen storage disease type IX: high variability in clinical phenotype. Mol. Genet. Metab. 92: 88-99.
7. Daub, H., et al. 2008. Kinase-selective enrichment enables quantitative phosphoproteomics of the kinome across the cell cycle. Mol. Cell 31: 438-448.
8. Online Mendelian Inheritance in Man, OMIM™. 2009. Johns Hopkins University, Baltimore, MD. MIM Number: 172490. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/

CHROMOSOMAL LOCATION

Genetic locus: PHKB (human) mapping to 16q12.1.

PROTOCOLS

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

PRODUCT

PHKB 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 µM solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see PHKB shRNA Plasmid (h): sc-93503-SH and PHKB shRNA (h) Lentiviral Particles: sc-93503-V as alternate gene silencing products.

For independent verification of PHKB (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-93503A, sc-93503B and sc-93503C.

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 µl of the RNAse-free water provided. Resuspension of the siRNA duplex in 330 µl of RNAse-free water makes a 10 µM solution in a 10 µM Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

PHKB siRNA (h) is recommended for the inhibition of PHKB 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 µM in 66 µl. 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 PHKB gene expression knockdown using RT-PCR Primer: PHKB (h)-PR: sc-93503-PR (20 µl). 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.