SANTA CRUZ BIOTECHNOLOGY, INC.

PHKA2 siRNA (m): sc-76122

BACKGROUND

PHKA2 (phosphorylase kinase, α 2), also known as PHKLA, XLG or PYK, is a 1,235 amino acid protein that is lipid-anchored to the cytoplasmic side of the cell membrane and belongs to the phosphorylase β kinase regulatory chain family. Expressed predominately in liver, but also present in other non-muscle tissues, PHKA2 exists as a component of a multi-chain polymer that functions as a phosphorylase β kinase and catalyzes the phosphorylation of target substrates, such as Troponin I. Defects in the gene encoding PHKA2 are the cause of glycogen storage disease type 9A (GSD9A), a metabolic disorder that results in glycogenosis (an abnormal accumulation of glycogen in tissue) and is characterized by hepatomegaly, growth retardation, muscle weakness, hypercholesterolemia, hypertriglyceridemia and fasting hyperketosis.

REFERENCES

1. Willems, P. 1990. Families with X-linked liver glycogenosis due to phosphorylase kinase deficiency. Clin. Genet. 38: 80.
2. Wauters, J.G., et al. 1992. Regional mapping of a liver α-subunit gene of phosphorylase kinase (PHKA) to the distal region of human chromosome Xp. Cytogenet. Cell Genet. 60: 194-196.
3. Hirono, H., et al. 1995. Isolation of cDNA encoding the human liver phosphorylase kinase α subunit (PHKA2) and identification of a missense mutation of the PHKA2 gene in a family with liver phosphorylase kinase deficiency. Biochem. Mol. Biol. Int. 36: 505-511.
4. Hendrickx, J., et al. 1995. Mutations in the phosphorylase kinase gene PHKA2 are responsible for X-linked liver glycogen storage disease. Hum. Mol. Genet. 4: 77-83.
5. Hendrickx, J., et al. 1996. X-linked liver glycogenosis type II (XLG II) is caused by mutations in PHKA2, the gene encoding the liver α subunit of phosphorylase kinase. Hum. Mol. Genet. 5: 649-652.
6. Burwinkel, B., et al. 1996. Mutation hotspots in the PHKA2 gene in X-linked liver glycogenosis due to phosphorylase kinase deficiency with atypical activity in blood cells (XLG2). Hum. Mol. Genet. 5: 653-658.

CHROMOSOMAL LOCATION

Genetic locus: Phka2 (mouse) mapping to X F4.

PRODUCT

PHKA2 siRNA (m) 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 PHKA2 shRNA Plasmid (m): sc-76122-SH and PHKA2 shRNA (m) Lentiviral Particles: sc-76122-V as alternate gene silencing products.

For independent verification of PHKA2 (m) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-76122A, sc-76122B and sc-76122C.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

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

PHKA2 siRNA (m) is recommended for the inhibition of PHKA2 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 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 PHKA2 gene expression knockdown using RT-PCR Primer: PHKA2 (m)-PR: sc-76122-PR (20 µl). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

PROTOCOLS

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