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SZABO-SCANDIC HandelsgmbH

Quellenstraße 110, A-1100 Wien

T. +43(0)1 489 3961-0

F. +43(0)1 489 3961-7

mail@szabo-scandic.com

www.szabo-scandic.com

[linkedin.com/company/szaboscandic](https://www.linkedin.com/company/szaboscandic) 

PHKA1 siRNA (m): sc-152223

BACKGROUND

PHKA1 (phosphorylase kinase, α 1), also known as PHKA, is a 1,223 amino acid protein that is lipid-anchored to the cytoplasmic side of the cell membrane and belongs to the phosphorylase b kinase regulatory chain family. Expressed only in muscle, PHKA1 exists as a component of a multi-chain polymer that functions as a phosphorylase b kinase and catalyzes the phosphorylation of target substrates, such as Troponin I. PHKA1 is a key regulatory enzyme of glycogen metabolism and defects in the gene encoding PHKA1 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. Potts, M.D., et al. 1994. A BgIII polymorphism at the ovine phosphorylase kinase α subunit locus (PHKA1). *Anim. Genet.* 25: 288.
4. Gossen, M., et al. 1995. Dinucleotide repeat polymorphism within the PHKA1 gene at Xq12-q13. *Hum. Genet.* 95: 469-470.
5. 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.
6. Pallen, M.J. 2003. Glucoamylase-like domains in the α - and β -subunits of phosphorylase kinase. *Protein Sci.* 12: 1804-1807.
7. Wuyts, W., et al. 2005. Myopathy and phosphorylase kinase deficiency caused by a mutation in the PHKA1 gene. *Am. J. Med. Genet. A.* 133A: 82-84.

CHROMOSOMAL LOCATION

Genetic locus: Phka1 (mouse) mapping to X D.

PRODUCT

PHKA1 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 μ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see PHKA1 shRNA Plasmid (m): sc-152223-SH and PHKA1 shRNA (m) Lentiviral Particles: sc-152223-V as alternate gene silencing products.

For independent verification of PHKA1 (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-152223A, sc-152223B and sc-152223C.

PROTOCOLS

See our web site at 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 μ l of the RNase-free water provided. Resuspension of the siRNA duplex in 330 μ l of RNase-free water makes a 10 μ M solution in a 10 μ M Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

PHKA1 siRNA (m) is recommended for the inhibition of PHKA1 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 μ M in 66 μ 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 PHKA1 gene expression knockdown using RT-PCR Primer: PHKA1 (m)-PR: sc-152223-PR (20 μ 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.