



Laforin siRNA (h): sc-75405

BACKGROUND

Laforin (Lafora PTPase) is a dual specificity protein phosphatase expressed in heart, skeletal muscle, kidney, pancreas and brain. It belongs to the protein-tyrosine phosphatase family and contains one CBM20 (carbohydrate binding type-20) domain and one tyrosine-protein phosphatase domain. Laforin may be involved in the control of glycogen metabolism, particularly in monitoring for and preventing the formation of poorly branched glycogen molecules (polyglucosans). Laforin isoform 1 is primarily associated with polyribosomes at the endoplasmic reticulum, however, it is also found at the plasma membrane. Isoform 2 can be found in the nucleus. Defects in the EPM2A gene are a cause of progressive myoclonic epilepsy type 2 (EPM2), also known as Lafora disease. EPM2 is an autosomal recessive disease and a severe form of adolescent-onset progressive epilepsy.

REFERENCES

1. Ganesh, S., et al. 2000. Laforin, defective in the progressive myoclonus epilepsy of Lafora type, is a dual-specificity phosphatase associated with polyribosomes. *Hum. Mol. Genet.* 9: 2251-2261.
2. Ganesh, S., et al. 2001. Mutation screening for Japanese Lafora's disease patients: identification of novel sequence variants in the coding and upstream regulatory regions of EPM2A gene. *Mol. Cell. Probes* 15: 281-289.
3. Wang, J., et al. 2002. A unique carbohydrate binding domain targets the Lafora disease phosphatase to glycogen. *J. Biol. Chem.* 277: 2377-2380.
4. Ganesh, S., et al. 2002. Genotype-phenotype correlations for EPM2A mutations in Lafora's progressive myoclonus epilepsy: exon 1 mutations associate with an early-onset cognitive deficit subphenotype. *Hum. Mol. Genet.* 11: 1263-1271.
5. Ganesh, S., et al. 2002. Alternative splicing modulates subcellular localization of Laforin. *Biochem. Biophys. Res. Commun.* 291: 1134-1137.
6. Ki, C.S., et al. 2003. Two novel mutations in the EPM2A gene in a Korean patient with Lafora's progressive myoclonus epilepsy. *J. Hum. Genet.* 48: 51-54.

CHROMOSOMAL LOCATION

Genetic locus: EPM2A (human) mapping to 6q24.3.

PRODUCT

Laforin 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 μ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see Laforin shRNA Plasmid (h): sc-75405-SH and Laforin shRNA (h) Lentiviral Particles: sc-75405-V as alternate gene silencing products.

For independent verification of Laforin (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-75405A, sc-75405B and sc-75405C.

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 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

Laforin siRNA (h) is recommended for the inhibition of Laforin 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 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.

GENE EXPRESSION MONITORING

Laforin (k2A3): sc-135810 is recommended as a control antibody for monitoring of Laforin gene expression knockdown by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) or immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor Laforin gene expression knockdown using RT-PCR Primer: Laforin (h)-PR: sc-75405-PR (20 μ l). 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.