

# EPM2AIP1 siRNA (h): sc-77976

## BACKGROUND

EPM2AIP1 (EPM2A interacting protein 1), also known as laforin-interacting protein, is the first recognized Laforin binding partner that may play a critical role in discovering the underlying pathogenesis of progressive myoclonic epilepsy type 2 (EPM2), also called Lafora disease (LD). EPM2 is an autosomal recessive disease characterized by grand mal seizures and/or myoclonus at about 15 years of age. Rapid and severe mental deterioration follows, often with psychotic features. Survival is less than 10 years after onset. Laforin is the only glycogen phosphatase in mammals that contains a carbohydrate-binding module. Mutations in the glycogen binding domain eliminate the ability of Laforin to dephosphorylate glycogen leading to EPM2 in humans. EPM2AIP1 colocalizes with Laforin to the endoplasmic reticulum. EPM2AIP1 contains two coiled-coil domains and is expressed in heart, brain, placenta, liver, pancreas, kidney and skeletal muscle.

## REFERENCES

1. Ianzano, L., et al. 2003. Identification of a novel protein interacting with laforin, the EPM2a progressive myoclonus epilepsy gene product. *Genomics* 81: 579-587.
2. Wang, W. and Roach, P.J. 2004. Glycogen and related polysaccharides inhibit the laforin dual-specificity protein phosphatase. *Biochem. Biophys. Res. Commun.* 325: 726-730.
3. Chan, E.M., et al. 2004. Laforin preferentially binds the neurotoxic starch-like polyglucosans, which form in its absence in progressive myoclonus epilepsy. *Hum. Mol. Genet.* 13: 1117-1129.
4. Hitchins, M., et al. 2005. MLH1 germline epimutations as a factor in hereditary nonpolyposis colorectal cancer. *Gastroenterology* 129: 1392-1399.
5. Worby, C.A., et al. 2006. Laforin, a dual specificity phosphatase that dephosphorylates complex carbohydrates. *J. Biol. Chem.* 281: 30412-30418.
6. Madhavan, D. and Kuzniecky, R.I. 2006. Lafora disease. *Rev. Neurol. Dis.* 3: 131-135.
7. Tagliabracci, V.S., et al. 2007. Laforin is a glycogen phosphatase, deficiency of which leads to elevated phosphorylation of glycogen *in vivo*. *Proc. Natl. Acad. Sci. USA* 104: 19262-19266.

## CHROMOSOMAL LOCATION

Genetic locus: EPM2AIP1 (human) mapping to 3p22.2.

## PRODUCT

EPM2AIP1 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 EPM2AIP1 shRNA Plasmid (h): sc-77976-SH and EPM2AIP1 shRNA (h) Lentiviral Particles: sc-77976-V as alternate gene silencing products.

For independent verification of EPM2AIP1 (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-77976A, sc-77976B and sc-77976C.

## 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

EPM2AIP1 siRNA (h) is recommended for the inhibition of EPM2AIP1 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.

## GENE EXPRESSION MONITORING

EPM2AIP1 (YS-62): sc-100651 is recommended as a control antibody for monitoring of EPM2AIP1 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).

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker<sup>™</sup> Molecular Weight Standards: sc-2035, UltraCruz<sup>®</sup> Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use m-IgG $\kappa$  BP-FITC: sc-516140 or m-IgG $\kappa$  BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz<sup>®</sup> Mounting Medium: sc-24941 or UltraCruz<sup>®</sup> Hard-set Mounting Medium: sc-359850.

## RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor EPM2AIP1 gene expression knockdown using RT-PCR Primer: EPM2AIP1 (h)-PR: sc-77976-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.