



AIPL1 siRNA (m): sc-60063

BACKGROUND

The inherited blindness associated protein, aryl hydrocarbon receptor interacting protein-like 1 (AIPL1), interacts with the cell cycle regulator protein NUB1. AIPL1 is crucial for protein folding and stabilization, as well as for protein trafficking. It localizes to the nucleus or cytoplasm and is highly expressed in the pineal gland and the retina. In the retina, AIPL1 is expressed in both developing cone and rod photoreceptors, but it is restricted to rod photoreceptors in the adult human retina. Defects in the gene encoding for AIPL1 can cause Leber congenital amaurosis type IV, an early-onset, inherited autosomal recessive disorder that results in childhood blindness.

REFERENCES

1. van der Spuy, J., et al. 2004. The Leber congenital amaurosis protein AIPL1 modulates the nuclear translocation of NUB1 and suppresses inclusion formation by NUB1 fragments. *J. Biol. Chem.* 279: 48038-48047.
2. van der Spuy, J., et al. 2004. Role of AIP and its homologue the blindness-associated protein AIPL1 in regulating client protein nuclear translocation. *Biochem. Soc. Trans.* 32: 643-645.
3. Dyer, M.A., et al. 2004. Retinal degeneration in AIPL1-deficient mice: a new genetic model of Leber congenital amaurosis. *Brain Res. Mol. Brain Res.* 132: 208-220.
4. Allikmets, R. 2004. Leber congenital amaurosis: a genetic paradigm. *Ophthalmic Genet.* 25: 67-79.
5. Silva, E., et al. 2004. A missense mutation in GUCY2D acts as a genetic modifier in RPE65-related Leber congenital amaurosis. *Ophthalmic Genet.* 25: 205-217.
6. Gallon, V.A., et al. 2004. Purification, characterisation and intracellular localisation of aryl hydrocarbon interacting protein-like 1 (AIPL1) and effects of mutations associated with inherited retinal dystrophies. *Biochim. Biophys. Acta* 1690: 141-149.
7. Liu, X., et al. 2004. AIPL1, the protein that is defective in Leber congenital amaurosis, is essential for the biosynthesis of retinal rod cGMP phosphodiesterase. *Proc. Natl. Acad. Sci. USA* 101:13903-13908.
8. Hanein, S., et al. 2005. Leber congenital amaurosis: comprehensive survey of genetic heterogeneity. A clinical definition update. *J. Fr. Ophthalmol.* 28: 98-105.
9. Galvin, J.A., et al. 2005. Clinical phenotypes in carriers of Leber congenital amaurosis mutations. *Ophthalmology* 112: 349-356.

CHROMOSOMAL LOCATION

Genetic locus: Aipl1 (mouse) mapping to 11 B4.

RESEARCH USE

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

PROTOCOLS

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

PRODUCT

AIPL1 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 AIPL1 shRNA Plasmid (m): sc-60063-SH and AIPL1 shRNA (m) Lentiviral Particles: sc-60063-V as alternate gene silencing products.

For independent verification of AIPL1 (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-60063A, sc-60063B and sc-60063C.

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

AIPL1 siRNA (m) is recommended for the inhibition of AIPL1 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 AIPL1 gene expression knockdown using RT-PCR Primer: AIPL1 (m)-PR: sc-60063-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.