



## RPGR siRNA (m): sc-41896

### BACKGROUND

RPGR (retinitis pigmentosa GTPase regulator) is a retina-specific, GTP-binding protein that is involved in the maintenance of a polarized distribution of outer segment-specific proteins. RPGR contains an N-terminus that is homologous to the regulator of chromosome condensation (RCC1) and contains a conserved nucleotide binding motif as well as a putative C-terminal isoprenylation site. RPGR interacting protein (RPGRIP) is a scaffold protein that can associate with RPGR within the connecting cilium of photoreceptor cells, where both proteins influence proper polarized protein distribution across the connecting cilium. RPGR may also be involved in mediating vesicular transport-associated events in photo-receptors. Mutations in the RPGR gene can cause X-linked retinitis pigmentosa type 3 (RP3), a disease characterized by retinal dystrophy, which eventually leads to complete blindness.

### REFERENCES

1. Roepman, R., et al. 1996. Positional cloning of the gene for X-linked retinitis pigmentosa 3: homology with the guanine-nucleotide-exchange factor RCC1. *Hum. Mol. Genet.* 5: 1035-1041.
2. Meindl, A., et al. 1996. A gene (RPGR) with homology to the RCC1 guanine nucleotide exchange factor is mutated in X-linked retinitis pigmentosa (RP3). *Nat. Genet.* 13: 35-42.
3. Buraczynska, M., et al. 1997. Spectrum of mutations in the RPGR gene that are identified in 20% of families with X-linked retinitis pigmentosa. *Am. J. Hum. Genet.* 61: 1287-1292.
4. Linari, M., et al. 1999. The retinitis pigmentosa GTPase regulator, RPGR, interacts with the  $\delta$  subunit of rod cyclic GMP phosphodiesterase. *Proc. Natl. Acad. Sci. USA* 96: 1315-1320.
5. Hong, D.H., et al. 2000. A retinitis pigmentosa GTPase regulator (RPGR)-deficient mouse model for X-linked retinitis pigmentosa (RP3). *Proc. Natl. Acad. Sci. USA* 97: 3649-3654.
6. Hong, D.H., et al. 2001. Retinitis pigmentosa GTPase regulator (RPGR)-interacting protein is stably associated with the photoreceptor ciliary axoneme and anchors RPGR to the connecting cilium. *J. Biol. Chem.* 276: 12091-12099.

### CHROMOSOMAL LOCATION

Genetic locus: Rprg (mouse) mapping to X A1.1.

### PRODUCT

RPGR 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 RPGR shRNA Plasmid (h): sc-41895-SH and RPGR shRNA (h) Lentiviral Particles: sc-41895-V as alternate gene silencing products.

For independent verification of RPGR (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-41895A, sc-41895B and sc-41895C.

### 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

RPGR siRNA (m) is recommended for the inhibition of RPGR 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  $\mu$ 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.

### RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor RPGR gene expression knockdown using RT-PCR Primer: RPGR (m)-PR: sc-41896-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.