SANTA CRUZ BIOTECHNOLOGY, INC.

RPGR (M-20): sc-14676

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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 δ 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 (RPGRr)-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: Rpgr (mouse) mapping to X A1.1.

SOURCE

RPGR (M-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of RPGR of mouse origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-14676 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

RPGR (M-20) is recommended for detection of RPGR of mouse and rat origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for RPGR siRNA (m): sc-41896, RPGR shRNA Plasmid (m): sc-41896-SH and RPGR shRNA (m) Lentiviral Particles: sc-41896-V.

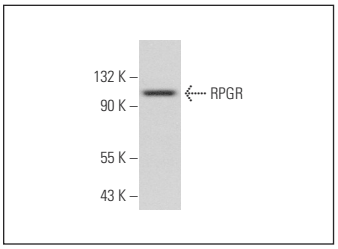
Molecular Weight of RPGR 113 kDa.

Positive Controls: AMJ2-C8 whole cell lysate: sc-364366.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



RPGR (M-20): sc-14676. Western blot analysis of RPGR expression in AMJ2-C8 whole cell lysate.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

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

PROTOCOLS

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