

RPGR (L-18): sc-14672

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.

CHROMOSOMAL LOCATION

Genetic locus: RPGR (human) mapping to Xp11.4; RpgR (mouse) mapping to X A1.1.

SOURCE

RPGR (L-18) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of RPGR of human origin.

PRODUCT

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

Blocking peptide available for competition studies, sc-14672 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

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

APPLICATIONS

RPGR (L-18) is recommended for detection of RPGR of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

RPGR (L-18) is also recommended for detection of RPGR in additional species, including canine.

Suitable for use as control antibody for RPGR siRNA (h): sc-41895, RPGR siRNA (m): sc-41896, RPGR shRNA Plasmid (h): sc-41895-SH, RPGR shRNA Plasmid (m): sc-41896-SH, RPGR shRNA (h) Lentiviral Particles: sc-41895-V and RPGR shRNA (m) Lentiviral Particles: sc-41896-V.

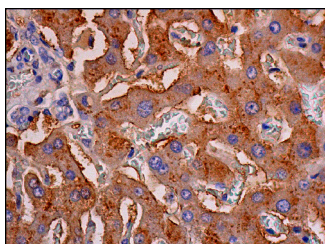
Molecular Weight of RPGR 113 kDa.

Positive Controls: Y79 cell lysate: sc-2240.

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. 4) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

DATA



RPGR (L-18): sc-14672. Immunoperoxidase staining of formalin fixed, paraffin-embedded human liver tissue showing cytoplasmic and membrane staining of hepatocytes and hepatic sinusoid cells.

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.