SANTA CRUZ BIOTECHNOLOGY, INC.

RPGRIP1 (B-3): sc-390331

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The Power to Question

BACKGROUND

RPGRIP1 (retinitis pigmentosa GTPase regulator interacting protein 1), also known as LCA6, RGI1, RGRIP, CORD13, RPGRIP or RPGRIP1d, is a 1,286 amino acid protein that belongs to the RPGRIP1 family and localizes to the cilium. Expressed in retina, RPGRIP1 colocalizes with SEC16S in the outer segment of rod photoreceptors and cone outer segments. RPGRIP1 forms homodimers and elongated homopolymers, and exists six alternatively spliced isoforms. RPGRIP1 is required for SEC16S function and is essential for normal disk morphogenesis. Mutations in the gene encoding RPGRIP1 are the cause of Leber congenital amaurosis type 6 (LCA6) and cone-rod dystrophy type 13 (CORD13). LCA is considered the most common genetic cause of congenital visual impairment in infants and children. CORD13 is an inherited retinal dystrophy characterized by retinal pigment deposits visible on fundus examination and initial loss of cone photoreceptors followed by rod degeneration.

REFERENCES

1. Boylan, J.P., et al. 2000. Identification of a novel protein interacting with RPGR. Hum. Mol. Genet. 9: 2085-2093.
2. Roepman, R., et al. 2000. The retinitis pigmentosa GTPase regulator (RPGR) interacts with novel transport-like proteins in the outer segments of rod photoreceptors. Hum. Mol. Genet. 9: 2095-2105.
3. Gerber, S., et al. 2001. Complete exon-intron structure of the RPGR-interacting protein (RPGRIP1) gene allows the identification of mutations underlying Leber congenital amaurosis. Eur. J. Hum. Genet. 9: 561-571.
4. Mavlyutov, T.A., et al. 2002. Species-specific subcellular localization of RPGR and RPGRIP isoforms: implications for the phenotypic variability of congenital retinopathies among species. Hum. Mol. Genet. 11: 1899-1907.
5. Hameed, A., et al. 2003. Evidence of RPGRIP1 gene mutations associated with recessive cone-rod dystrophy. J. Med. Genet. 40: 616-619.
6. Pawlyk, B.S., et al. 2010. Replacement gene therapy with a human RPGRIP1 sequence slows photoreceptor degeneration in a murine model of Leber congenital amaurosis. Hum. Gene Ther. 21: 993-1004.
7. Fernández-Martínez, L., et al. 2011. Evidence for RPGRIP1 gene as risk factor for primary open angle glaucoma. Eur. J. Hum. Genet. 19: 445-451.

CHROMOSOMAL LOCATION

Genetic locus: Rpgrip1 (mouse) mapping to 14 C2.

SOURCE

RPGRIP1 (B-3) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 44-57 near the N-terminus of RPGRIP1 of mouse origin.

PRODUCT

Each vial contains 200 µg IgG_3 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-390331 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

APPLICATIONS

RPGRIP1 (B-3) is recommended for detection of RPGRIP1 isoforms 1 and 2 of mouse and rat origin by Western Blotting (starting dilution 1:100, 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 RPGRIP1 siRNA (m): sc-153101, RPGRIP1 shRNA Plasmid (m): sc-153101-SH and RPGRIP1 shRNA (m) Lentiviral Particles: sc-153101-V.

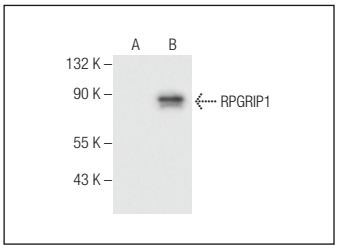
Molecular Weight of RPGRIP1: 147 kDa.

Positive Controls: RPGRIP1 (m): 293T Lysate: sc-123274.

RECOMMENDED SUPPORT REAGENTS

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

DATA



RPGRIP1 (B-3): sc-390331. Western blot analysis of RPGRIP1 expression in non-transfected: sc-117752 (**A**) and mouse RPGRIP1 transfected: sc-123274 (**B**) 293T whole cell lysates.

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 for detailed protocols and support products.