

RPGR Antibody (C-term)

Affinity Purified Rabbit Polyclonal Antibody (Pab) Catalog # AP12226b

Specification

RPGR Antibody (C-term) - Product Information

Application IHC-P, WB,E **Primary Accession** 092834 Other Accession NP 000319.1 Reactivity Human Host **Rabbit** Clonality **Polyclonal** Isotype Rabbit IgG Antigen Region 744-772

RPGR Antibody (C-term) - Additional Information

Gene ID 6103

Other Names

X-linked retinitis pigmentosa GTPase regulator, RPGR, RP3, XLRP3

Target/Specificity

This RPGR antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 744-772 amino acids of human RPGR.

Dilution

IHC-P~~1:10~50 WB~~1:1000

E~~Use at an assay dependent concentration.

Format

Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.

Storage

Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

Precautions

RPGR Antibody (C-term) is for research use only and not for use in diagnostic or therapeutic procedures.

RPGR Antibody (C-term) - Protein Information

Name RPGR (HGNC:10295)

Synonyms RP3, XLRP3



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Function Acts as a guanine-nucleotide releasing factor (GEF) for RAB8A and RAB37 by promoting the conversion of inactive RAB-GDP to the active form RAB-GTP (PubMed:20631154). GEF activity towards RAB8A may facilitate ciliary trafficking by modulating ciliary intracellular localization of RAB8A (PubMed:20631154). GEF activity towards RAB37 maintains autophagic homeostasis and retinal function (By similarity). Involved in photoreceptor integrity (By similarity). May control cilia formation by regulating actin stress filaments and cell contractility. May be involved in microtubule organization and regulation of transport in primary cilia (PubMed:21933838). May play a critical role in spermatogenesis and in intraflagellar transport processes (By similarity).

Cellular Location

Cytoplasm, cytoskeleton, flagellum axoneme {ECO:0000250|UniProtKB:Q9R0X5}. Golgi apparatus. Cell projection, cilium {ECO:0000250|UniProtKB:Q9R0X5}. Note=In the retinal photoreceptor cell layer, localizes at the connecting cilium (By similarity). Colocalizes with WHRN in the photoreceptor connecting cilium (By similarity) Colocalizes with CEP290 in the photoreceptor connecting cilium (By similarity). Colocalizes with RPGRIP1 in the photoreceptor connecting cilium (By similarity). Colocalizes with RPGR at the primary cilia of epithelial cells (By similarity). {ECO:0000250|UniProtKB:Q9R0X5}

Tissue Location

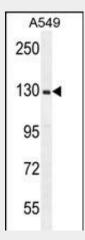
Heart, brain, placenta, lung, liver, muscle, kidney, retina, pancreas and fetal retinal pigment epithelium. Isoform 3 is found only in the retina. Colocalizes with RPGRIP1 in the outer segment of rod photoreceptors and cone outer segments

RPGR Antibody (C-term) - Protocols

Provided below are standard protocols that you may find useful for product applications.

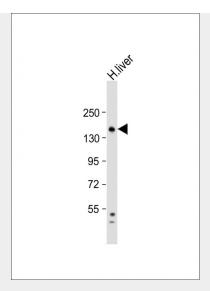
- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- <u>Immunofluorescence</u>
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

RPGR Antibody (C-term) - Images

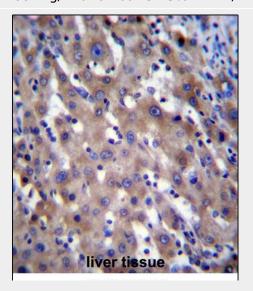


RPGR Antibody (C-term) (Cat. #AP12226b) western blot analysis in A549 cell line lysates (35ug/lane). This demonstrates the RPGR antibody detected the RPGR protein (arrow).





Anti-RPGR Antibody (C-term) at 1:1000 dilution + human liver lysate Lysates/proteins at 20 μ g per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/10000 dilution. Predicted band size : 113 kDa Blocking/Dilution buffer: 5% NFDM/TBST.



RPGR Antibody (C-term) (Cat. #AP12226b)immunohistochemistry analysis in formalin fixed and paraffin embedded human liver tissue followed by peroxidase conjugation of the secondary antibody and DAB staining. This data demonstrates the use of RPGR Antibody (C-term) for immunohistochemistry. Clinical relevance has not been evaluated.

RPGR Antibody (C-term) - Background

This gene encodes a protein with a series of six RCC1-like domains (RLDs), characteristic of the highly conserved guanine nucleotide exchange factors. The encoded protein is found in the Golgi body and interacts with RPGRIP1. This protein localizes to the outer segment of rod photoreceptors and is essential for their viability. Mutations in this gene have been associated with X-linked retinitis pigmentosa (XLRP). Multiple alternatively spliced transcript variants that encode different isoforms of this gene have been reported, but the full-length natures of only some have been determined.

RPGR Antibody (C-term) - References





Clark, G.R., et al. Ophthalmology 117(11):2169-2177(2010) Schmid, F., et al. Invest. Ophthalmol. Vis. Sci. 51(3):1628-1635(2010) Ji, Y., et al. Curr. Eye Res. 35(1):73-79(2010) Sheng, X., et al. Mol. Vis. 16, 1620-1628 (2010) : Murga-Zamalloa, C.A., et al. J. Genet. 88(4):399-407(2009)