

## RPGR Antibody (C-term)

Affinity Purified Rabbit Polyclonal Antibody (Pab)  
Catalog # AP12226b

### Product Information

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<b>Application</b>	WB, IHC-P, E
<b>Primary Accession</b>	<a href="#">Q92834</a>
<b>Other Accession</b>	<a href="#">NP_000319.1</a>
<b>Reactivity</b>	Human
<b>Host</b>	Rabbit
<b>Clonality</b>	Polyclonal
<b>Isotype</b>	Rabbit IgG
<b>Clone Names</b>	RB31028
<b>Calculated MW</b>	113387
<b>Antigen Region</b>	744-772

### Additional Information

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<b>Gene ID</b>	6103
<b>Other Names</b>	X-linked retinitis pigmentosa GTPase regulator, RPGR, RP3, XLRP3
<b>Target/Specificity</b>	This RPGR antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 744-772 amino acids of human RPGR.
<b>Dilution</b>	WB~~1:1000 IHC-P~~1:100~500 E~~Use at an assay dependent concentration.
<b>Format</b>	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.
<b>Storage</b>	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.
<b>Precautions</b>	RPGR Antibody (C-term) is for research use only and not for use in diagnostic or therapeutic procedures.

### Protein Information

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<b>Name</b>	RPGR ( <a href="#">HGNC:10295</a> )
<b>Synonyms</b>	RP3, XLRP3
<b>Function</b>	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: <a href="#">20631154</a> ). 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:Q9N1T2, 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

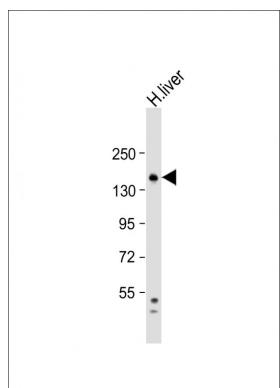
## Background

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.

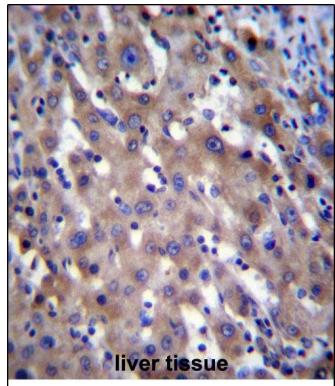
## References

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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)

## Images



Anti-RPGR Antibody (C-term) at 1:1000 dilution + human liver lysate Lysates/proteins at 20 µg 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.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.