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RPGR Antibody (C-term)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP12226b

Product Information

Application	WB, IHC-P, E
Primary Accession	Q92834
Other Accession	NP_000319.1
Reactivity	Human
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB31028
Calculated MW	113387
Antigen Region	744-772

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	WB~~1:1000 IHC-P~~1:100~500 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.

Protein Information

Name	RPGR (HGNC:10295)
Synonyms	RP3, XLRP3
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: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

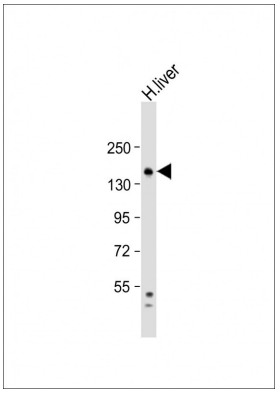
Background

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.

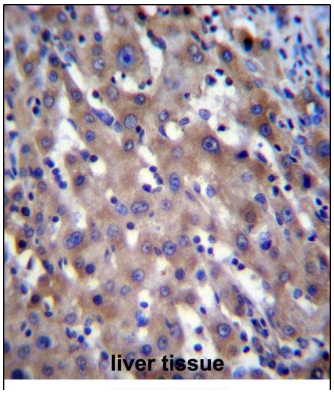
References

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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'.