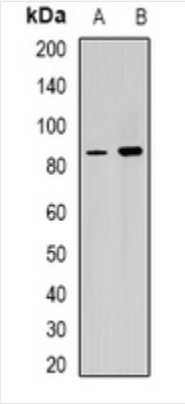
RPGR Antibody – Polyclonal

PRODUCT OVERVIEW

Product Name	RPGR, Polyclonal Antibody
Full Product Name	Anti-RPGR Antibody
Product Gene Name	anti-RPGR antibody
Product Synonym Names	RP3; XLRP3; X-linked retinitis pigmentosa GTPase regulator
Clonality	Polyclonal
Species Reactivity	Human, Mouse, Rat
Form/Format	Liquid in 0.42% Potassium phosphate, 0.87% Sodium chloride, pH 7.3, 30% glycerol, and 0.01% sodium azide.
Purity/Purification	The antibody was purified by immunogen affinity chromatography.
Specificity	Recognizes endogenous levels of RPGR protein.
Preparation and Storage	Shipped at 4 degree C. Upon delivery aliquot and store at -20 degree C for one year. Avoid freeze/thaw cycles.
Molecular Weight	127,042 Da
OMIM	300029
3D Structure	ModBase 3D Structure for Q92834
Source	Rabbit
Immunogen	Recombinant full length protein of human RPGR
Applications Tested/Suitable for anti-RPGR antibody	Western Blot (WB)
Application Notes for anti-RPGR antibody	WB: 1/500 - 1/2000
Related Product Information for anti-RPGR antibody	MBS8245501 is an antibody targeting RPGR. Reactive in Human, Mouse, Rat. Rabbit polyclonal antibody to RPGR
Other Notes	Small volumes of anti-RPGR antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.



Western Blot (WB) of anti-RPGR antibody
Western blot analysis of RPGR expression in Y79 (A), mouse heart (B) whole cell lysates.

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for RPGR. It may not necessarily be applicable to this product.

NCBI GI #	4506581
NCBI GeneID	6103
NCBI Accession #	NP_000319.1
UniProt Primary Accession #	Q92834
UniProt Secondary Accession #	O00702; O00737; Q3KN84; Q8N5T6; Q93039; Q9HD29; Q9UMR1; B1ARN3; E9PE28
UniProt Related Accession #	Q92834
GenBank Nucleotide #	NM_000328.2
NCBI Official Full Name	X-linked retinitis pigmentosa GTPase regulator isoform A
NCBI Official Synonym Full Names	retinitis pigmentosa GTPase regulator
NCBI Official Symbol	RPGR
NCBI Official Synonym Symbols	CRD; RP3; COD1; PCDX; RP15; XLRP3; orf15; CORDX1
UniProt Protein Name	X-linked retinitis pigmentosa GTPase regulator
Protein Family	X-linked retinitis pigmentosa GTPase regulator
UniProt Gene Name	RPGR
UniProt Synonym Gene Names	RP3; XLRP3
UniProt Entry Name	RPGR_HUMAN
NCBI Protein Information	X-linked retinitis pigmentosa GTPase regulator
NCBI Summary for RPGR	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. [provided by RefSeq, Dec 2008]

UniProt Comments for RPGR	RPGR: a guanine nucleotide exchange factor (GEF) that plays a role in cilia biogenesis and maintenance. Interacts with the GDP-bound form of the small GTPase RAB8A. Promotes the exchange of GDP to GTP, converting inactive GDP-bound Rab proteins into their active GTP-bound form. Probably regulates cilia formation by regulating actin stress filaments and cell contractility. Plays an important role in photoreceptor integrity. May play a critical role in spermatogenesis and in intraflagellar transport processes May be involved in microtubule organization and regulation of transport in primary cilia. Interacts with PDE6D, RPGRIP1, CEP290, and DFNB31. Defects in RPGR are the cause of retinitis pigmentosa (RP3, RP15, and RPDSI), cone-rod dystrophy, X-linked 1 (CORDX1), and macular degeneration, X-linked, atrophic (MDXLA). Six isoforms of the human protein are produced by alternative splicing. Isoform 3 is found only in the retina. Colocalizes with RPGRIP1 in the outer segment of rod photoreceptors and cone outer segments. Isoform 6 associates with microtubule organizing centers, cilium basal bodies, and cilium axonemes. Interacts with NPM1 (via C-terminus), RPGRIP1L, SMC1A and SMC3. Protein type: GEFs, Rab; GEFs Chromosomal Location of Human Ortholog: Xp21.1 Cellular Component: centrosome; Golgi apparatus; photoreceptor outer segment Molecular Function: guanyl-nucleotide exchange factor activity; protein binding Biological Process: cilium biogenesis; intracellular protein transport; intraflagellar transport; positive regulation of GTPase activity; response to stimulus; visual perception Disease: Cone-rod Dystrophy, X-linked, 1; Macular Degeneration, X-linked Atrophic; Retinitis Pigmentosa 3; Retinitis Pigmentosa, X-linked, And Sinorespiratory Infections, With Or Without Deafness
Research Articles on RPGR	**1.** Although carriers of XLRP are usually asymptomatic or have a mild disease of late onset, the proband presented here exhibited an early-onset, aggressive form of the disease.
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