RPGR Antibody – Rabbit Polyclonal

PRODUCT OVERVIEW

Product Name	RPGR, Polyclonal Antibody
Full Product Name	RPGR, CT (RPGR, RP3, XLRP3, X-linked retinitis pigmentosa GTPase regulator) (MaxLight 650)
Product Gene Name	anti-RPGR antibody
Product Synonym Names	RPGR; RP3; XLRP3; X-linked retinitis pigmentosa GTPase regulator
Clonality	Polyclonal
Isotype	IgG
Host	Rabbit
Species Reactivity	Human
Form/Format	Supplied as a liquid in PBS, pH 7.2. No preservative added. Labeled with MaxLight650.
Purity/Purification	Purified by Protein A and Peptide Affinity Chromatography.
Specificity	Recognizes human RPGR
Preparation and Storage	Store product at 4 degree C in the dark. DO NOT FREEZE! Stable at 4 degree C for 12 months after receipt as an undiluted liquid. Dilute required amount only prior to immediate use. Further dilutions can be made in assay buffer. Caution: MaxLight650 conjugates are sensitive to light. For maximum recovery of product, centrifuge the original vial prior to removing the cap.
Molecular Weight	113,387 Da
OMIM	phenotype 312610
3D Structure	ModBase 3D Structure for Q92834
Immunogen	KLH-conjugated synthetic peptide mapping to a fragment of residues within amino acids 744-772 in the C-terminal region of human RPGR.
Conjugate	MaxLight650
Note	Preservative Free
Product Categories/Family for anti-RPGR antibody	Antibodies; Disease Markers
Applications Tested/Suitable for anti-RPGR antibody	Immunohistochemistry (IHC) Paraffin, Western Blot (WB), FLISA
Application Notes for anti-RPGR antibody	Applications are based on unconjugated antibody.
Related Product Information for anti-RPGR antibody	MBS6343267 is an antibody targeting RPGR. Also known as RPGR, CT (RPGR, RP3, XLRP3, X-linked retinitis pigmentosa GTPase regulator) (MaxLight 650). Reactive in Human. Host: Rabbit.
Product References and Citations for anti-RPGR antibody	1. Clark, G.R., et al. Ophthalmology 117(11):2169-2177(2010). 2. Schmid, F., et al. Invest. Ophthalmol. Vis. Sci. 51(3):1628-1635(2010). 3. Ji, Y., et al. Curr. Eye Res. 35(1):73-79(2010). 4. Sheng, X., et al. Mol. Vis. 16, 1620-1628 (2010). 5. Murga-Zamalloa, C.A., et al. J. Genet. 88(4):399-407(2009).
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.

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; retinitis pigmentosa 15; retinitis pigmentosa 3 GTPase regulator
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; GEFs, Rab Chromosomal Location of Human Ortholog: Xp21.1 Cellular Component: Golgi apparatus; centrosome; photoreceptor outer segment Molecular Function: protein binding; guanyl-nucleotide exchange factor activity Biological Process: intracellular protein transport; eye photoreceptor cell development; visual perception; response to stimulus; intraflagellar transport; cilium biogenesis; positive regulation of GTPase activity Disease: Retinitis Pigmentosa, X-linked, And Sinorespiratory Infections, With Or Without Deafness; Cone-rod Dystrophy, X-linked, 1; Macular Degeneration, X-linked Atrophic; Retinitis Pigmentosa 3
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