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CNGB3 Rabbit pAb

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Catalog # AP57806

Product Information

Application	WB
Primary Accession	Q9NQW8
Predicted	Rat, Pig, Dog, Human, Mouse, Chicken, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	92167
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human CNGB3
Epitope Specificity	301-400/809
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Membrane; Multi-pass membrane protein.
DISEASE	Defects in CNGB3 are the cause of Stargardt disease type 1 (STGD1) [MIM:248200]. STGD is one of the most frequent causes of macular degeneration in childhood. It is characterized by macular dystrophy with juvenile-onset, rapidly progressive course, alterations of the peripheral retina, and subretinal deposition of lipofuscin-like material. STGD1 inheritance is autosomal recessive. Defects in CNGB3 are the cause of achromatopsia type 3 (ACHM3) [MIM:262300]; also known as Pingelapese blindness. ACHM3 is a congenital complete achromatopsia and is distinct from total colorblindness mainly because of the consistent concurrence of severe myopia.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Cyclic nucleotide-gated (CNG) cation channels are heteromeric complexes made up of principal alpha and modulatory beta subunits. The alpha subunits consist of CNG1-3 and form functional cation channels by themselves. The beta subunits consist of CNG4-6 and, unlike the alpha subunits, do not form functional channels, but rather modify the properties of channels. formed by CNG1-3. CNG channels are essential components of olfactory and visual transduction. CNG proteins are present in cone and rod photoreceptors and in the pineal gland, and they contribute to modulating arterial blood pressure. CNG6, also designated cyclic-nucleotide-gated cation channel beta 3 (CNG-beta 3), is an integral membrane protein that can form a heterooligomeric complex with CNG-3. CNG-beta 3 is activated by cGMP and this activation leads to the depolarization of rod photoreceptors as a result of cation channel being opened. CNG-beta 3 is expressed in a small group of retinal photoreceptor cells and in testis. Mutations in the gene encoding for CNG-beta 3, can cause achromatopsia, an autosomal recessively inherited disease characterized by low visual acuity, photophobia, a lack of color discrimination, and nystagmus.

Additional Information

Gene ID	54714
Other Names	Cyclic nucleotide-gated channel beta-3, CNG channel beta-3, Cone photoreceptor cGMP-gated channel subunit beta, Cyclic nucleotide-gated cation channel beta-3, Cyclic nucleotide-gated cation channel modulatory subunit, CNGB3 {ECO:0000303|PubMed:37463923}
Dilution	WB=1:500-2000,ICC/IF=1:100-500,ELISA=1:5000-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	CNGB3 {ECO:0000303|PubMed:37463923}
Function	Pore-forming subunit of the cone cyclic nucleotide-gated channel. Mediates cone photoresponses at bright light converting transient changes in intracellular cGMP levels into electrical signals. In the dark, cGMP levels are high and keep the channel open enabling a steady inward current carried by Na(+) and Ca(2+) ions that leads to membrane depolarization and neurotransmitter release from synaptic terminals. Upon photon absorption cGMP levels decline leading to channel closure and membrane hyperpolarization that ultimately slows neurotransmitter release and signals the presence of light, the end point of the phototransduction cascade. Conducts cGMP- and cAMP-gated ion currents, with permeability for monovalent and divalent cations.
Cellular Location	Photoreceptor outer segment membrane; Multi-pass membrane protein. Cell membrane; Multi-pass membrane protein. Note=Localized in cone outer segment membranes
Tissue Location	Expressed specifically in the retina.

Background

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