

Goat Anti-RPE65 Antibody

Peptide-affinity purified goat antibody Catalog # AF1943a

Specification

Goat Anti-RPE65 Antibody - Product Information

Application Primary Accession Other Accession Reactivity Predicted Host Clonality Concentration Isotype Calculated MW WB <u>Q16518</u> <u>NP_000320</u>, <u>6121</u> Rat Human, Dog Goat Polyclonal 100ug/200ul IgG 60948

Goat Anti-RPE65 Antibody - Additional Information

Gene ID 6121

Other Names

Retinoid isomerohydrolase, 3.1.1.64, All-trans-retinyl-palmitate hydrolase, Retinal pigment epithelium-specific 65 kDa protein, Retinol isomerase, RPE65

Format

0.5 mg lgG/ml in Tris saline (20mM Tris pH7.3, 150mM NaCl), 0.02% sodium azide, with 0.5% bovine serum albumin

Storage

Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

Precautions

Goat Anti-RPE65 Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

Goat Anti-RPE65 Antibody - Protein Information

Name RPE65 (<u>HGNC:10294</u>)

Function

Critical isomerohydrolase in the retinoid cycle involved in regeneration of 11-cis-retinal, the chromophore of rod and cone opsins. Catalyzes the cleavage and isomerization of all-trans-retinyl fatty acid esters to 11-cis-retinol which is further oxidized by 11-cis retinol dehydrogenase to 11-cis-retinal for use as visual chromophore (PubMed:16116091). Essential for



the production of 11-cis retinal for both rod and cone photoreceptors (PubMed:17848510). Also capable of catalyzing the isomerization of lutein to meso-zeaxanthin an eye- specific carotenoid (PubMed:28874556). The soluble form binds vitamin A (all-trans-retinol), making it available for LRAT processing to alltrans-retinyl ester. The membrane form, palmitoylated by LRAT, binds all-trans-retinyl esters, making them available for IMH (isomerohydrolase) processing to all-cis-retinol. The soluble form is regenerated by transferring its palmitoyl groups onto 11-cis-retinol, a reaction catalyzed by LRAT (By similarity).

Cellular Location

Cytoplasm {ECO:0000250|UniProtKB:A9C3R9}. Cell membrane; Lipid-anchor. Microsome membrane {ECO:0000250|UniProtKB:Q28175}. Note=Attached to the membrane by a lipid anchor when palmitoylated (membrane form), soluble when unpalmitoylated. Undergoes light-dependent intracellular transport to become more concentrated in the central region of the retina pigment epithelium cells.

Tissue Location

Retina (at protein level). Retinal pigment epithelium specific.

Goat Anti-RPE65 Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

- <u>Western Blot</u>
- <u>Blocking Peptides</u>
- <u>Dot Blot</u>
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- <u>Cell Culture</u>

Goat Anti-RPE65 Antibody - Images



AF1943a (0.5 μ g/ml) staining of Rat Retina lysate (35 μ g protein in RIPA buffer). Primary incubation was 1 hour. Detected by chemiluminescence.

Goat Anti-RPE65 Antibody - Background

This gene encodes a protein which is located in the retinal pigment epithelium and is involved in the production of 11-cis retinal and in visual pigment regeneration. There are two forms of this



protein, a soluble form called sRPE65, and a palmitoylated, membrane-bound form known as mRPE65. mRPE65 serves as the palmitoyl donor for lecithin retinol acyl transferase (LRAT), the enzyme that catalyzes the vitamin A to all trans retinol step of the chromophore regeneration process. Both mRPE65 and sRPE65 also serve as regulatory proteins, with the ratio and concentrations of these molecules playing a role in the inhibition of 11-cis retinal synthesis. Mutations in this gene have been associated with Leber congenital amaurosis type 2 (LCA2) and retinitis pigmentosa.

Goat Anti-RPE65 Antibody - References

Development of a Diagnostic Genetic Test for Simplex and Autosomal Recessive Retinitis Pigmentosa. Clark GR, et al. Ophthalmology, 2010 Jun 28. PMID 20591486.

FATP1 inhibits 11-cis retinol formation via interaction with the visual cycle retinoid isomerase RPE65 and lecithin:retinol acyltransferase. Guignard TJ, et al. J Biol Chem, 2010 Jun 11. PMID 20356843. Negative charge of the glutamic acid 417 residue is crucial for isomerohydrolase activity of RPE65. Nikolaeva O, et al. Biochem Biophys Res Commun, 2010 Jan 22. PMID 20043869. Differential macular morphology in patients with RPE65-, CEP290-, GUCY2D-, and AIPL1-related Leber congenital amaurosis. Pasadhika S, et al. Invest Ophthalmol Vis Sci, 2010 May. PMID 19959640.

RPE65, visual cycle retinol isomerase, is not inherently 11-cis-specific: support for a carbocation mechanism of retinol isomerization. Redmond TM, et al. J Biol Chem, 2010 Jan 15. PMID 19920137.