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Rabbit Anti-RPE65 antibody

SL60224R

Product Name	RPE65
Chinese Name	视网膜色素 epithelial cells 特异性蛋白 65 抗体
Alias	All-trans-retinyl-palmitate hydrolase; LCA 2; LCA2; Leber congenital amaurosis; mRPE 65; mRPE65; p63; rd 12; rd12; Retinal pigment epithelium specific 61 kDa protein; Retinal pigment epithelium specific 65 kDa protein; Retinal pigment epithelium specific protein; Retinal pigment epithelium specific protein 65kDa; Retinal pigment epithelium-specific 65 kDa protein; Retinitis pigmentosa 20; Retinoid isomerohydrolase; RP 20; RP20; RPE 65; RPE65; RPE65_HUMAN; sRPE 65; sRPE65.
Research Area	Tumour Neurobiology Signal transduction
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	Mouse
Applications	WB=1:500-2000,IHC-P=1:1000-4000,IHC-F=1:400-800,IF=1:100-500 （Paraffin section need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Theoretical molecular weight	59kDa
Cellular localization	cytoplasmic The cell membrane
Form	Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from mouse RPE65
Lsotype	IgG
Purification	affinity purified by Protein A
Buffer Solution	1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol.
Storage	Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.

Attention This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

PubMed PubMed

Product Detail

The retinal pigment epithelium (RPE) is a monolayer simple epithelium in proximity to the outer surface of the retinal photoreceptor cells. Retinal pigment epithelium-specific protein (RPE65) is a 65 kDa protein belonging to the β-carotene dioxygenase family. This protein is important in 11-cis retinal production as well as in visual pigment regeneration. RPE65 is attached to the membrane by a lipid anchor when palmitoylated (membrane form) and soluble when unpalmitoylated. The soluble form of the protein binds vitamin A. Defects in RPE65 causes autosomal dominant retinitis pigmentosa and/or Leber congenital amaurosis type 2.

Function:
Plays important roles in the production of 11-cis retinal and in visual pigment regeneration. The soluble form binds vitamin A (all-trans-retinol), making it available for LRAT processing to all-trans-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. The enzymatic activity is linearly dependent of the expression levels and membrane association.

Subunit:
Interacts with MYO7A; this mediates light-dependent intracellular transport of RPE65.

Subcellular Location:
Cytoplasm. Cell membrane. Attached to the membrane by a lipid anchor when palmitoylated (membrane form), soluble when unpalmitoylated.

Tissue Specificity:
Retinal pigment epithelium specific.

Post-translational modifications:
Palmitoylation by LRAT regulates ligand binding specificity; the palmitoylated form (membrane form) specifically binds all-trans-retinyl-palmitate, while the soluble unpalmitoylated form binds all-trans-retinol (vitamin A).

DISEASE:
Defects in RPE65 are the cause of Leber congenital amaurosis type 2 (LCA2) [MIM:204100]. LCA designates a clinically and genetically heterogeneous group of childhood retinal degenerations, generally inherited in an autosomal recessive manner. Affected infants have little or no retinal photoreceptor function as tested by electroretinography. LCA represents the most common genetic cause of congenital visual impairment in infants and children.

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Similarity:
Belongs to the carotenoid oxygenase family.

SWISS:
Q91ZQ5

Gene ID:
19892

Database links:

Entrez Gene: 6121 Human

Entrez Gene: 19892 Mouse

Entrez Gene: 89826 Rat

Omim: 180069 Human

SwissProt: Q9YGX2 Chicken

SwissProt: Q28175 Cow

SwissProt: Q16518 Human

SwissProt: Q91ZQ5 Mouse

SwissProt: O70276 Rat

Unigene: 2133 Human

Unigene: 131708 Mouse

Unigene: 76724 Rat

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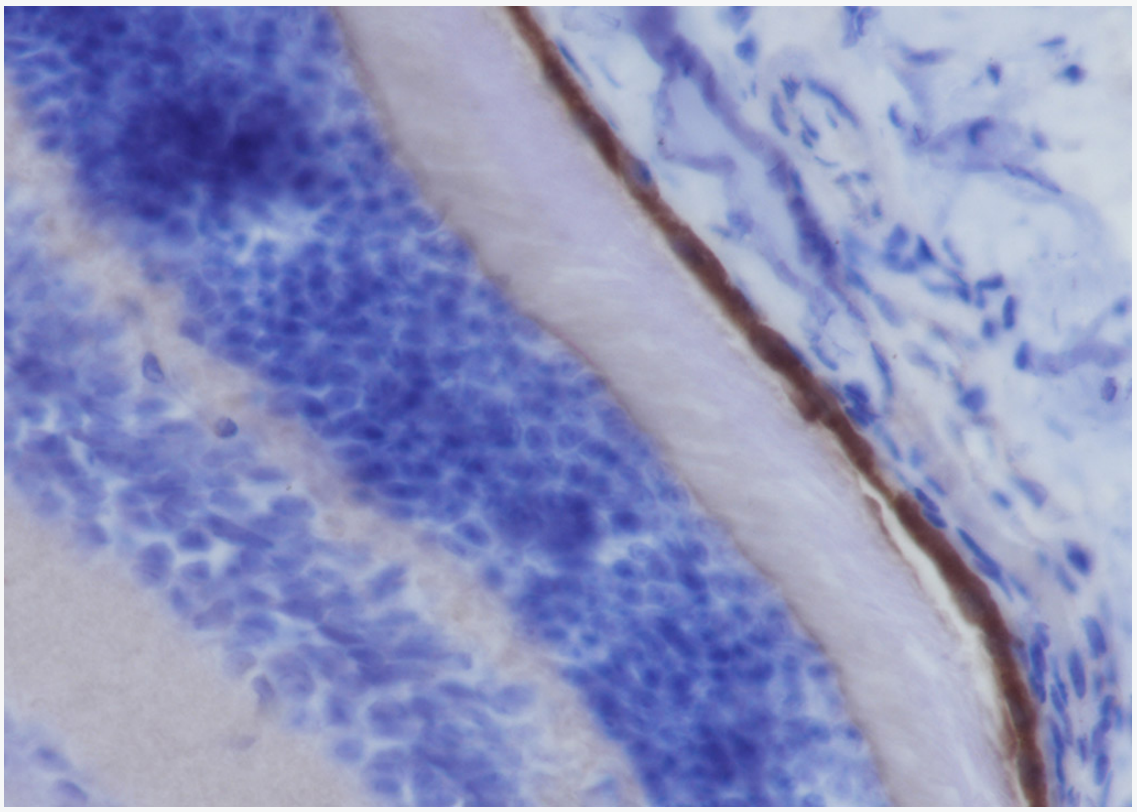
Product Picture



Tissue:Mouse eyeball

Section type: Formalin fixed & Paraffin -embedded section

Retrieval method: High temperature and high pressure

Retrieval buffer: Tris/EDTA buffer, pH 9.0 Primary ab dilution: 1:4000

Primary ab incubation condition: 1 hour at room temperature

Counter stain: Hematoxylin

Comment: Color brown is the positive signal for SLM-60224R