

# Rabbit Anti-RP1 antibody

# SL11030R

<b>Product Name:</b>	RP1
Chinese Name:	视网膜色素变性蛋白1抗体
Alias:	DCDC4A; ORP1; Oxygen-regulated protein 1; Retinitis pigmentosa 1 protein; Retinitis
	pigmentosa RP1 protein; RP1; RP1_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human, Mouse, Rat, Dog, Horse, Rabbit,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-
	500 (Paraffin sections need antigen repair)
	not yet tested in other applications.
	optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	241kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	lmg/ml
immunogen:	KLH conjugated synthetic peptide derived from human RP1/DCDC4A:451-550/2156
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized
	antibody is stable at room temperature for at least one month and for greater than a year
	when kept at -20°C. 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.
PubMed:	<u>PubMed</u>
Product Detail:	Retinitis pigmentosa 1 is a novel 2,156 amino acid oxygen-regulated photoreceptor
	specific to retina. Originally named ORP1 (for 'oxygen-regulated protein-1'), the
	expression of retinitis pigmentosa 1 has been found to be regulated by oxygen levels in
	the retina. Mutation of the retinitis pigmentosa 1 gene causes dominant retinitis
	pigmentosa which leads to degeneration of retinal photoreceptor cells and symptoms
	such as night vision blindness and deficits in the midperipheral visual field. Retinitis

pigmentosa 1 may assist in differentiation of photoreceptor cells and has been identified in the cilia of photoreceptors, possibly aiding in both cilial structure and protein transport between inner and outer segments of photoreceptors. Retinitis pigmentosa 1 contains two doublecortin domains and is encoded by a gene which maps to human chromosome 8q11-q13.

#### **Function:**

Microtubule-associated protein regulating the stability and length of the microtubule-based axoneme of photoreceptors. Required for the differentiation of photoreceptor cells, it plays a role in the organization of the outer segment of rod and cone photoreceptors ensuring the correct orientation and higher order stacking of outer segment disks along the photoreceptor axoneme.

### Subunit:

Interacts (via the doublecortin domains) with microtubules. Interacts with RP1L1 (By similarity). Interacts with MAK (By similarity).

#### **Subcellular Location:**

Cytoplasm; cytoskeleton; cilium axoneme. Cell projection; cilium; photoreceptor outer segment. Specifically localized in the connecting cilia of rod and cone photoreceptors.

## Tissue Specificity:

Expressed in retina. Not expressed in heart, brain, placenta, lung, liver, skeletal muscle, kidney, spleen and pancreas.

#### DISEASE:

Defects in RP1 are the cause of retinitis pigmentosa type 1 (RP1) [MIM:180100]. RP leads to degeneration of retinal photoreceptor cells. Patients typically have night vision blindness and loss of midperipheral visual field. As their condition progresses, they lose their far peripheral visual field and eventually central vision as well.

#### Similarity:

Contains 2 doublecortin domains.

#### SWISS:

P56715

#### Gene ID:

6101

#### Database links:

Entrez Gene: 6101 Human

Omim: 603937 Human

SwissProt: P56715 Human

Unigene: 128938 Human

# Important Note:

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

