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Rabbit Anti-AIPL1 antibody

SL12452R

Product Name:	AIPL1
Chinese Name:	遗传性失明相关蛋白AIPL1抗体
Alias:	A930007I01Rik; Aipl1; AIPL1_HUMAN; AIPL2; Aryl hydrocarbon interacting protein like 1; Aryl hydrocarbon receptor interacting protein like 1; Aryl-hydrocarbon-interacting protein-like 1; LCA4; MGC25485; OTTHUMP00000128207; OTTMUSP00000006382; RP23-401C17.1.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Rabbit,Sheep,
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:	44kDa
Cellular localization:	The nucleuscytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human AIPL1:165-260/384
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:	PubMed
Product Detail:	The inherited blindness associated protein, aryl hydrocarbon receptor interacting protein-like 1 (AIPL1), interacts with the cell cycle regulator protein NUB1. AIPL1 is crucial for protein folding and stabilization, as well as for protein trafficking. It localizes to the nucleus or cytoplasm and is highly expressed in the pineal gland and the retina. In

	the retina, AIPL1 is expressed in both developing cone and rod photoreceptors, but it is restricted to rod photoreceptors in the adult human retina. Defects in the gene encoding for AIPL1 can cause Leber congenital amaurosis type IV, an early-onset, inherited autosomal recessive disorder that results in childhood blindness. **Function:** May be important in protein trafficking and/or protein folding and stabilization. **Subunit:** Interacts with NUB1. **Subcellular Location:** Cytoplasm. Nucleus. **Tissue Specificity:** Highly expressed in retina. Specifically localized to the developing photoreceptor layer and within the photoreceptors of the adult retina. **DISEASE:** Defects in AIPL1 are the cause of Leber congenital amaurosis type 4 (LCA4) [MIM:604393]. 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. **Similarity:** Contains 1 PPIase FKBP-type domain. Contains 3 TPR repeats. **SWISS:** Q9NZN9 **Gene ID:** 23746 **Database links:** Entrez Gene: 23746Human Entrez Gene: 114230Mouse Entrez Gene: 59110Rat Omim: 604392Human

	SwissProt: Q9NZN9Human SwissProt: Q924K1Mouse SwissProt: Q9JLG9Rat Unigene: 279887Human Unigene: 95707Mouse Unigene: 102037Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.