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Rabbit Anti-ARH antibody

SL6337R

Product Name:	ARH
Chinese Name:	低密度Lipoprotein受体衔接蛋白抗体
Alias:	ARH; ARH GENE; ARH_HUMAN; ARH1; ARH2; Autosomal recessive hypercholesterolemia protein; FHCB1; FHCB2; LDL receptor adaptor protein; Ldlrap1; Low density lipoprotein receptor adapter protein 1.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Pig,Cow,Horse,Rabbit,Sheep,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=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:	34kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human ARH/LDL receptor adaptor protein:41-140/308
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 protein encoded by this gene is a cytosolic protein which contains a phosphotyrosine binding (PTD) domain. The PTD domain has been found to interact with the cytoplasmic tail of the LDL receptor. Mutations in this gene lead to LDL receptor malfunction and cause the disorder autosomal recessive hypercholesterolaemia.

Function:
Adapter protein (clathrin-associated sorting protein (CLASP)) required for efficient endocytosis of the LDL receptor (LDLR) in polarized cells such as hepatocytes and lymphocytes, but not in non-polarized cells (fibroblasts). May be required for LDL binding and internalization but not for receptor clustering in coated pits. May facilitate the endocytocis of LDLR and LDLR-LDL complexes from coated pits by stabilizing the interaction between the receptor and the structural components of the pits. May also be involved in the internalization of other LDLR family members. Binds to phosphoinositides, which regulate clathrin bud assembly at the cell surface.

Subunit:
Interacts with LDLR. Binds to soluble clathrin trimers. Interacts with AP2B1; the interaction mediates the association with the AP-2 complex. Interacts with VLDLR

Subcellular Location:
Cytoplasm.

Tissue Specificity:
Expressed at high levels in the kidney, liver, and placenta, with lower levels detectable in brain, heart, muscle, colon, spleen, intestine, lung, and leukocytes.

Post-translational modifications:
Phosphorylated upon DNA damage, probably by ATM or ATR.

DISEASE:
Defects in LDLRAP1 are the cause of autosomal recessive hypercholesterolemia (ARH) [MIM:603813]. ARH is a disorder caused by defective internalization of LDL receptors (LDLR) in the liver. ARH has the clinical features of familial hypercholesterolemia (FH) [MIM:143890] homozygotes, including severely elevated plasma LDL cholesterol, tuberous and tendon xanthomata, and premature atherosclerosis. LDL receptor (LDLR) activity measured in skin fibroblasts is normal, as the LDL binding ability.

Similarity:
Contains 1 PID domain.

SWISS:
Q5SW96

Gene ID:
26119

Database links:

Entrez Gene: 26119Human

	Entrez Gene: 100017Mouse Entrez Gene: 500564Rat Omim: 605747Human SwissProt: Q5SW96Human SwissProt: Q8C142Mouse Unigene: 590911Human Unigene: 482148Mouse **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.