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Rabbit Anti-CHM antibody

SL13911R

Product Name:	CHM
Chinese Name:	遗传性脉络膜缺乏症相关蛋白抗体
Alias:	CHM; Chm; Choroideraemia protein; Choroideremia; DXS540; FLJ38564; GGTA; HSD 32; MGC102710; Rab escort protein 1; Rab geranylgeranyltransferase component A; Rab proteins geranylgeranyltransferase component A 1; RAE1_HUMAN; REP 1; REP-1; REP1; TCD; TCD protein.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,
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:	73kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human CHM:301-400/653
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:	This gene encodes component A of the RAB geranylgeranyl transferase holoenzyme. In the dimeric holoenzyme, this subunit binds unprenylated Rab GTPases and then presents them to the catalytic Rab GGTase subunit for the geranylgeranyl transfer reaction. Rab GTPases need to be geranylgeranyled on either one or two cysteine

	residues in their C-terminus to localize to the correct intracellular membrane. Mutations in this gene are a cause of choroideremia; also known as tapetochoroidal dystrophy (TCD). This X-linked disease is characterized by progressive dystrophy of the choroid, retinal pigment epithelium and retina. Alternative splicing results in multiple transcript variants encoding different isoforms.[provided by RefSeq, Feb 2009] **Function:** Binds unprenylated Rab proteins, presents it to the catalytic Rab GGTase dimer, and remains bound to it after the geranylgeranyl transfer reaction. The component A is thought to be regenerated by transferring its prenylated Rab back to the donor membrane. Also a pre-formed complex consisting of CHM and the Rab GGTase dimer (RGGT or component B) can bind to and prenylate Rab proteins; this alternative pathway is proposed to be the predominant pathway for Rab protein geranylgeranylation. **Subcellular Location:** Belongs to the Rab GDI family. **DISEASE:** Defects in CHM are the cause of choroideremia (CHM) [MIM:303100]. An X-linked recessive disease characterized by a slowly progressive degeneration of the choroid, photoreceptors, and retinal pigment epithelium. Affected males develop night blindness in their teenage years followed by loss of peripheral vision and complete blindness at middle age. Carrier females are generally asymptomatic but funduscopic examination often shows patchy areas of chorioretinal atrophy. **SWISS:** P24386 **Gene ID:** 1121 **Database links:** Entrez Gene: 1121 Human Entrez Gene: 12662 Mouse Omim: 300390 Human SwissProt: P24386 Human SwissProt: Q9QXG2 Mouse Unigene: 496449 Human

	Unigene: 257316 Mouse **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.