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CHM Antibody (Center)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP20084c

Product Information

Application	WB, E
Primary Accession	P24386
Other Accession	NP_000381.1
Reactivity	Human
Predicted	Canine
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB42411
Calculated MW	73476
Antigen Region	292-320

Additional Information

Gene ID	1121
Other Names	Rab proteins geranylgeranyltransferase component A 1, Choroideremia protein, Rab escort protein 1, REP-1, TCD protein, CHM, REP1, TCD
Target/Specificity	This CHM antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 292-320 amino acids from the Central region of human CHM.
Dilution	WB~~1:1000 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	CHM Antibody (Center) is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	CHM
Synonyms	REP1, TCD

Function	Substrate-binding subunit of the Rab geranylgeranyltransferase (GGTase) complex. Binds unprenylated Rab proteins and presents the substrate peptide to the catalytic component B composed of RABGGTA and RABGGTB, 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. Besides, 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.
Cellular Location	Cytoplasm, cytosol.

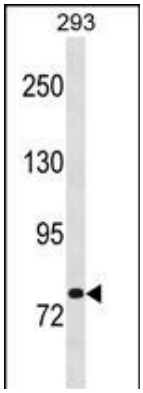
Background

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.

References

Perez-Cano, H.J., et al. Am. J. Med. Genet. A 149A (10), 2134-2140 (2009) :
Renner, A.B., et al. Arch. Ophthalmol. 127(7):907-912(2009)
Sergeev, Y.V., et al. Mutat. Res. 665 (1-2), 44-50 (2009) :
MacDonald, I.M., et al. Surv Ophthalmol 54(3):401-407(2009)
Strunnikova, N.V., et al. PLoS ONE 4 (12), E8402 (2009) :

Images



CHM Antibody (Center) (Cat. #AP20084c) western blot analysis in 293 cell line lysates (35ug/lane).This demonstrates the CHM antibody detected the CHM protein (arrow).

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.