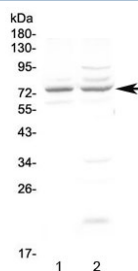


## CHM Antibody / Choroideremia protein (RQ4174)

| Catalog No. | Formulation                                           | Size   |
|-------------|-------------------------------------------------------|--------|
| RQ4174      | 0.5mg/ml if reconstituted with 0.2ml sterile DI water | 100 ug |

**Bulk quote request**

|                           |                                                                   |
|---------------------------|-------------------------------------------------------------------|
| <b>Availability</b>       | 1-3 business days                                                 |
| <b>Species Reactivity</b> | Human                                                             |
| <b>Format</b>             | Antigen affinity purified                                         |
| <b>Clonality</b>          | Polyclonal (rabbit origin)                                        |
| <b>Isotype</b>            | Rabbit IgG                                                        |
| <b>Purity</b>             | Antigen affinity purified                                         |
| <b>Buffer</b>             | Lyophilized from 1X PBS with 2% Trehalose and 0.025% sodium azide |
| <b>UniProt</b>            | P24386                                                            |
| <b>Applications</b>       | Western Blot : 0.5-1ug/ml<br>Direct ELISA : 0.1-0.5ug/ml          |
| <b>Limitations</b>        | This CHM antibody is available for research use only.             |



Western blot testing of human 1) COLO320 and 2) U-87 MG cell lysate with CHM antibody at 0.5ug/ml. Predicted molecular weight ~73 kDa.

## Description

Rab escort protein 1 (REP1) also known as rab proteins geranylgeranyltransferase component A 1 is an enzyme that in humans is encoded by the CHM gene. 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 geranylgeranylated 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. Alternatively spliced

transcript variants have been found for this gene.

## **Application Notes**

Optimal dilution of the CHM antibody should be determined by the researcher.

## **Immunogen**

A recombinant human protein corresponding to amino acids A193-Q380 was used as the immunogen for the CHM antibody.

## **Storage**

After reconstitution, the CHM antibody can be stored for up to one month at 4°C. For long-term, aliquot and store at -20°C. Avoid repeated freezing and thawing.