

REP-1 (N-17): sc-14759

BACKGROUND

Newly synthesized Rab proteins are bound to Rab escort proteins (REP) and presented to the Rab geranylgeranyltransferase (GGTase) type II, which mediates the prenylation of Rab proteins on two carboxy terminal cysteine residues. Rab GGTase only recognizes Rab proteins as a substrate when they are bound to REP. REP remains complexed with Rab until it is transported to the appropriate subcellular membrane, although it is still unclear whether REP participates in this targeting. Two isoforms of the REP gene have been isolated, REP-1 and REP-2. The REP-1 gene, located on chromosome Xq21.2, is prone to a wide variety of mutations, including nonsense, frameshift and splice-site mutations and deletions. In patients with choroideraemia (CHM), mutations in the REP-1 gene result in progressive dystrophy of the choroid, retinal pigment epithelium and retina. CHM is an X-linked hereditary eye disease that leads to blindness later in life. REP-2 is able to bind to several Rab proteins with the same affinity as REP-1 and may act a substitute for REP-1 to prevent widespread tissue abnormalities in patients with CHM.

REFERENCES

1. Cremers, F.P., et al. 1994. REP-2, a Rab escort protein encoded by the choroideremia-like gene. *J. Biol. Chem.* 269: 2111-2117.
2. Ohba, N. and Isashiki, Y. 1999. Clinical and genetic features of choroideremia. *Nippon Ganka Gakkai Zasshi* 103: 773-781.
3. Fujiki, K., et al. 1999. REP-1 gene mutations in Japanese patients with choroideremia. *Graefes Arch. Clin. Exp. Ophthalmol.* 237: 735-740.
4. Hayakawa, M., et al. 1999. Visual impairment and REP-1 gene mutations in Japanese choroideremia patients. *Ophthalmic Genet.* 20: 107-115.
5. Alexandrov, K., et al. 1999. Characterization of the ternary complex between Rab7, REP-1 and Rab geranylgeranyl transferase. *Eur. J. Biochem.* 265: 160-170.

CHROMOSOMAL LOCATION

Genetic locus: CHM (human) mapping to Xq21.2.

SOURCE

REP-1 (N-17) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of REP-1 of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-14759 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

APPLICATIONS

REP-1 (N-17) is recommended for detection of REP-1 of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for REP-1 siRNA (h): sc-41804, REP-1 shRNA Plasmid (h): sc-41804-SH and REP-1 shRNA (h) Lentiviral Particles: sc-41804-V.

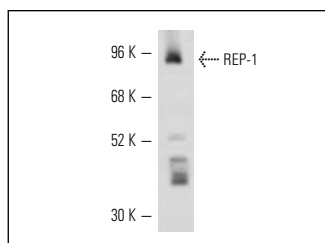
Molecular Weight of REP-1: 83 kDa.

Positive Controls: Y79 cell lysate: sc-2240 or A-431 whole cell lysate: sc-2201.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



REP-1 (N-17): sc-14759. Western blot analysis of REP-1 expression in A-431 whole cell lysate.

SELECT PRODUCT CITATIONS

1. Sergeev, Y.V., et al. 2009. The functional effect of pathogenic mutations in Rab escort protein 1. *Mutat. Res.* 665: 44-50.

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Try **REP-1 (2F1): sc-23905**, our highly recommended monoclonal alternative to REP-1 (N-17).