SANTA CRUZ BIOTECHNOLOGY, INC.

REP-2 (C-20): sc-14768

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Newly synthesized Rab proteins are bound to Rab escort proteins (REPs) 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 is it 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., Armstrong, S.A., Seabra, M.C., Brown, M.S. and Goldstein, J.L. 1994. REP-2, a Rab escort protein encoded by the choroideremia-like gene. J. Biol. Chem. 269: 2111-2117.
2. Alexandrov, K., Simon, I., Yurchenko, V., Iakovenko, A., Rostkova, E., Scheidig, A.J. and Goody, R.S. 1999. Characterization of the ternary complex between Rab 7, REP-1 and Rab geranylgeranyl transferase. Eur. J. Biochem. 265: 160-170.
3. Ohba, N. and Isashiki, Y. 1999. Clinical and genetic features of choroideremia. Nippon Ganka Gakkai Zasshi 103: 773-781.
4. Fujiki, K., Hotta, Y., Hayakawa, M., Saito, A., Mashima, Y., Mori, M., Yoshii, M., Murakami, A., Matsumoto, M., Hayasaka, S., Tagami, N., Isashiki, Y., Ohba, N. and Kanai, A. 1999. REP-1 gene mutations in Japanese patients with choroideremia. Graefes Arch. Clin. Exp. Ophthalmol. 237: 735-740.
5. Hayakawa, M., Fujiki, K., Hotta, Y., Ito, R., Ohki, J., Ono, J., Saito, A., Nakayasu, K., Kanai, A., Ishidoh, K., Kominami, E., Yoshida, K., Kim, K.C. and Ohashi, H. 1999. Visual impairment and REP-1 gene mutations in Japanese choroideremia patients. Ophthalmic Genet. 20: 107-115.
6. Zhang, H., Seabra, M.C. and Deisenhofer, J. 2000. Crystal structure of Rab geranylgeranyltransferase at 2.0 A resolution. Structure 8: 241-251.
7. Overmeyer, J.H., Wilson, A.L. and Maltese, W.A. 2001. Membrane targeting of a Rab GTPase that fails to associate with Rab escort protein (REP) or guanine nucleotide dissociationinhibitor (GDI). J. Biol. Chem. 276: 20379-2086.

CHROMOSOMAL LOCATION

Genetic locus: CHML (human) mapping to 1q43.

SOURCE

REP-2 (C-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of REP-2 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-14768 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

REP-2 (C-20) is recommended for detection of REP-2 of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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-2 siRNA (h): sc-41806, REP-2 shRNA Plasmid (h): sc-41806-SH and REP-2 shRNA (h) Lentiviral Particles: sc-41806-V.

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) 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.

SELECT PRODUCT CITATIONS

1. Strunnikova, N.V., Barb, J., Sergeev, Y.V., Thiagarajasubramanian, A., Silvin, C., Munson, P.J. and Macdonald, I.M. 2009. Loss-of-function mutations in Rab escort protein 1 (REP-1) affect intracellular transport in fibroblasts and monocytes of choroideremia patients. PLoS ONE 4: e8402.

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.

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.