

## REP-2 (E-4): sc-398605

### 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, 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: CHML (human) mapping to 1q43; Chml (mouse) mapping to 1 H4.

### SOURCE

REP-2 (E-4) is a mouse monoclonal antibody raised against amino acids 58-97 mapping near the N-terminus of REP-2 of mouse origin.

### PRODUCT

Each vial contains 200 µg IgG<sub>2a</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

REP-2 (E-4) is available conjugated to agarose (sc-398605 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-398605 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-398605 PE), fluorescein (sc-398605 FITC), Alexa Fluor® 488 (sc-398605 AF488), Alexa Fluor® 546 (sc-398605 AF546), Alexa Fluor® 594 (sc-398605 AF594) or Alexa Fluor® 647 (sc-398605 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-398605 AF680) or Alexa Fluor® 790 (sc-398605 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

### APPLICATIONS

REP-2 (E-4) is recommended for detection of REP-2 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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-2 siRNA (h): sc-41806, REP-2 siRNA (m): sc-41807, REP-2 shRNA Plasmid (h): sc-41806-SH, REP-2 shRNA Plasmid (m): sc-41807-SH, REP-2 shRNA (h) Lentiviral Particles: sc-41806-V and REP-2 shRNA (m) Lentiviral Particles: sc-41807-V.

Molecular Weight (predicted) of REP-2: 74 kDa.

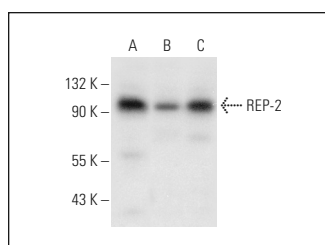
Molecular Weight (observed) of REP-2: 90 kDa.

Positive Controls: Neuro-2A whole cell lysate: sc-364185, NIH/3T3 whole cell lysate: sc-2210 or RAW 264.7 whole cell lysate: sc-2211.

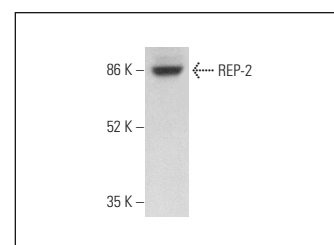
### RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 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 m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

### DATA



REP-2 (E-4): sc-398605. Western blot analysis of REP-2 expression in Neuro-2A (A), NIH/3T3 (B) and RAW 264.7 (C) whole cell lysates.



REP-2 (E-4): sc-398605. Western blot analysis of REP-2 expression in C2C12 whole cell lysate.

### 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.

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